AD-A166 465

REVIEW OF PART 67 OF THE FEDERAL AIR REGULATIONS AND THE MEDICAL CERTIFICATION OF CIVILIAN AIRMEN

Volume II

DTIC
ELECTE
APR 0 8 1986
S D
D

Submitted to the Federal Aviation Administration

under Contract #DTFA01-83-C-20066

by the

American Medical Association

Alan L. Engelberg, MD, MPH, Project Director

and Director, Department of Public Health Policy

Theodore C. Doege, MD, MS, Senior Scientist

Special Advisor-Science, Office of the Vice President

for Science and Technology

American Medical Association

535 North Dearborn Street

Chicago, Illinois 60610

DTIC FILE COPY

86 4 8 034

Table of Contents
Volume I

Foreword ..... i-xx

Section I - Part 67 Proposed Medical Standards ..... I-1 to I-13

Section II - Recommended Changes in FAA
Form 8500-8 ..... II-1 to II-10

Section III - Recommended Changes to the
Aviation Medical Examiners Guide ..... III-1 to III-228
General Considerations 1
Questions on Medications, Visits to Health Professionals
and Smoking 2
Mental and Behavioral 5
Hematology/Oncology 25
Visual System 35
Musculoskeletal System 73
Gastrointestinal System 81
Reproductive-Urinary System 113
Ear, Nose, Throat and Related Structures 119
Dermatology 133
Respiratory System 137
Cardiovascular System 155
Nervous System 183
Endocrine System 205
Risk Factors and Qualifications for Flying 215

Volume II

Section IV - Rationale for Standards and Guidelines:
Further Recommendations to the FAA ..... IV-1 to IV-490
Endocrine System 2
Mental and Behavioral 11
Hematology/Oncology 39
Visual System 89
Reproductive-Urinary System 139
Musculoskeletal System 143
Ear, Nose, Throat and Related Structures 145
Dermatology 147
Respiratory System 149
Cardiovascular System 185
Overview 185
Arrhythmias 210
Peripheral Vascular System 231
Myocardial Disease 242
Pericardial Disease 256
Valvular Heart Disease 263
Congenital Heart Disease 282
Ischemic Heart Disease 299
Hypertension in Aviation 311
Recommended Revision of Forms 319

Nervous System 327
Epilepsy and Single Seizure 328
Head Trauma 341
Sleep and Arousal Disorders 364
Infectious Diseases of the CNS 374
Demyelinating Diseases 390
Acquired and Hereditary Ataxias 400
Movement Disorders 402
Dementia 425
Headache and Cranial Neuralgias 433
Cerebrovascular Diseases 457
Brain Tumors 488
Dysequilibrium 490

Section V - Recommendations for Research . . . . . V-1 to V-6

Accesion For
NTIS CRA&I
DTIC TAB
Unannounced
Justification
By
Distribution /
Availability Codes
Dist | Avail and/or Special
A-1

RATIONALE FOR STANDARDS AND GUIDELINES, AND FURTHER RECOMMENDATIONS TO THE FAA

In this section we present information that will be useful to the FAA in a variety of ways. First, it presents the rationale for the recommended standards, examination procedures and dispositions that were described in the first three sections of this report. Second, it provides the most recent clinical thinking on a variety of disorders that are commonly encountered by the FAA, which will guide the FAA in its review of individual cases for special issuance of medical certificates, and for follow-up of those individuals who are given special issuance certificates.

Endocrine System

Recommendations Concerning Diabetes Mellitus

The Endocrine Committee recommends that the absolute prohibition of certification of individuals requiring oral hypoglycemic agents for control of diabetes mellitus be lifted. This recommendation is based on the clinical experience of the committee members as well as the medical literature. Smith[1] states that acute or chronic malnutrition almost always accompanies the hypoglycemia caused by sulfonylureas, and that other predisposing factors for hypoglycemia include advanced age, and hepatic, renal or adrenocortical insufficiency. The action of the sulfonylureas can be potentiated by alcohol, salicylates, sulfonamides and other drugs. The American Diabetes Association states:[2] "On rare occasions hypoglycemia is associated with the use of oral hypoglycemic agents, especially in persons who have renal or hepatic disease, which makes them more susceptible to the hypoglycemic effects of sulfonylureas (particularly the long-acting and potent agent, chlorpropamide)." Steel et al,[3] in a survey of automobile drivers who had noninsulin-dependent diabetes, found an apparent absence of hypoglycemic symptoms in that group.

Persons with diabetes that is adequately controlled by diet and oral hypoglycemic agents should be well informed about their diabetes. Standards for diabetes education, which are attached, have been published by the National Diabetes Advisory Board. The committee suggests that individuals with diabetes requiring oral hypoglycemic agents for control who are permitted to fly under special issuance be required to furnish proof of having attended an educational course that conforms to these standards.

The committee recommends that absolute prohibition of certification of individuals requiring insulin for control of diabetes mellitus be continued. While eventually it may be reasonable to approve the judicious use of insulin therapy for some applicants seeking pilot certification, the committee concludes that all applicants for

Endocrine System

Recommendations Concerning Diabetes Mellitus

The Endocrine Committee recommends that the absolute prohibition of certification of individuals requiring oral hypoglycemic agents for control of diabetes mellitus be lifted. This recommendation is based on the clinical experience of the committee members as well as the medical literature. Smith[1] states that acute or chronic malnutrition almost always accompanies the hypoglycemia caused by sulfonylureas, and that other predisposing factors for hypoglycemia include advanced age, and hepatic, renal or adrenocortical insufficiency. The action of the sulfonylureas can be potentiated by alcohol, salicylates, sulfonamides and other drugs. The American Diabetes Association states:[2] "On rare occasions hypoglycemia is associated with the use of oral hypoglycemic agents, especially in persons who have renal or hepatic disease, which makes them more susceptible to the hypoglycemic effects of sulfonylureas (particularly the long-acting and potent agent, chlorpropamide)." Steel et al,[3] in a survey of automobile drivers who had noninsulin-dependent diabetes, found an apparent absence of hypoglycemic symptoms in that group.

Persons with diabetes that is adequately controlled by diet and oral hypoglycemic agents should be well informed about their diabetes. Standards for diabetes education, which are attached, have been published by the National Diabetes Advisory Board. The committee suggests that individuals with diabetes requiring oral hypoglycemic agents for control who are permitted to fly under special issuance be required to furnish proof of having attended an educational course that conforms to these standards.

The committee recommends that absolute prohibition of certification of individuals requiring insulin for control of diabetes mellitus be continued. While eventually it may be reasonable to approve the judicious use of insulin therapy for some applicants seeking pilot certification, the committee concludes that all applicants for

Classes I, II, and III certificates be denied certification permanently until better monitoring techniques are available for predicting the onset of hypoglycemic reactions.

This is strongly recommended because neuroglycopenic symptoms may develop without warning in individuals taking insulin,[4] there is an inherent unpredictability of hypoglycemic episodes that cannot be monitored readily,[5] and the effect of hypoglycemia in combination with hypoxia on cognitive functions is not known.

Therefore, the committee concludes that it is necessary first to substantiate the effectiveness and safety of the oral hypoglycemic agents before insulin therapy can be approved in the pilot population. The Federal Air Surgeon should monitor carefully all individuals who are given a special issuance certificate while taking oral hypoglycemic agents, who should report back to the FAA at 6-month intervals with a re-evaluation by a specialist in diabetes. After two years the Federal Air Surgeon, with the assistance of a panel of experts in diabetes, should review the data, and either leave these recommendations unchanged or modify them, possibly to include for special issuance some individuals who are on insulin therapy.

The Endocrine Committee also reviewed and revised the form for "Specifications for the Initial Evaluation of Abnormal Carbohydrate Metabolism" (FAA 8500-17) and "Specifications for Follow-up Evaluation of Abnormal Carbohydrate Metabolism" (FAA Form 8500-18). Also included in this section are the recommendations of the National Diabetes Data Group[6] for the interpretation of glucose tolerance test data, which should be used by the FAA in its initial and follow-up evaluations of airmen with glucose intolerance. Finally, acceptable replacement therapy for other endocrine disorders are presented.

SPECIFICATIONS FOR THE INITIAL EVALUATION OF ABNORMAL CARBOHYDRATE METABOLISM

It is important that baseline values be established for an airman who is seeking medical certification, when there is an indication of disturbance of carbohydrate metabolism from the medical history or from an abnormal screening plasma glucose value. Prior clinical information, such as hospital records, laboratory reports, and outpatient records, should also be submitted. When this information includes the data listed below, and if it is no more than 90 days old at the time of examination, the tests need not be updated. Actual ECG tracings should be forwarded with the evaluation report.

- Required data:
  1. General medical history and complaints.
  2. Family and personal history related to diabetes.
  3. Height and weight, with explanation of any recent changes in weight.
  4. Ophthalmoscopic examination.
  5. Vibration, pain, position, light touch responses in the extremities.
  6. Cardiovascular examination:
     a. History specific for cardiovascular disease.
     b. Sitting blood pressure (brachial arteries).
     c. Circulatory efficiency in extremities.
     d. Standard 12-lead resting ECG.
     e. Treadmill stress test.
     f. Blood lipid determination (total cholesterol and triglycerides).
  7. Report of chest radiograph.

8. Urinalysis for specific gravity, albumin, sugar and acetone.

9. Statement concerning present need for insulin or other hypoglycemic medication for maintenance of control. If medication has previously been required for control of carbohydrate metabolism, the AME should specify types used and the most recent date that latest medication was discontinued.*

10. Plasma glucose determination.

    a. If a prior "diagnostic" oral glucose tolerance test (OGTT) was done, the results should be submitted along with current fasting and 2-hour postprandial plasma glucose test results and glycosylated hemoglobin concentration.

    b. If no prior OGTT was done, a current OGTT should be submitted (2-hour).

Glucose determination should be performed on venous plasma in a certified laboratory. Capillary blood measurements using reagent strips with or without reflectance meters are not satisfactory for diagnostic purposes. Fasting specimens should be drawn in the morning, 10 to 14 hours following the last food intake. Postprandial

*IMPORTANT NOTE: Certification will be considered only if adequate control can be accomplished and maintained without use of insulin. If use of insulin only recently has been discontinued, control is to be demonstrated by fasting and 2-hour postprandial blood sugar tests taken at 30-day intervals during the previous 90 days. Control will be considered acceptable when fasting plasma glucose is ≤ 140 mg/dl and ≥ 60 mg/dl (a fasting plasma glucose < 100 mg/dl in a person taking an oral hypoglycemic agent may indicate excessive or inappropriate use of the sulfonyluresa, and is a cause for review and probable revision of medication), 2-hour postprandial glucose is ≤ 175 mg/dl, and glycosylated hemoglobin does not exceed the upper limits of normal in a given laboratory by more than 1% (in many laboratories the upper limit of normal is 7%). Care must be exercised to minimize the risk of hypoglycemia in persons treated with oral agents. Prolonged fasting or severe dietary restriction alone or in combination with alcohol ingestion must be avoided. Susceptibility to hypoglycemia may persist for more than 24 hours following alcohol ingestion when nutritional status compromised.

blood samples may be obtained 2 hours after a mixed meal (breakfast or lunch) containing 75 to 100 g carbohydrate.

Oral glucose tolerance tests should be conducted and interpreted as described in the reference, "Office guide to diagnosis and classification of diabetes mellitus and other categories of glucose intolerance," Diabetes Care 1981;4:335, which is reproduced below.

SPECIFICATIONS FOR FOLLOW-UP EVALUATION OF ABNORMAL CARBOHYDRATE METABOLISM

1. Significant history during interval since last examination.
2. Weight, with an explanation of any change since last examination.
3. Ophthalmoscopic examination.
4. Vibration, pain, position, light touch senses in extremities.
5. Full explanation of any problems in maintaining constant control of glucose without use of insulin.
6. Current fasting and 2-hour postprandial plasma glucose and glycosylated hemoglobin determinations. Glucose determinations should be performed on venous plasma in a certified laboratory. Capillary blood measurements using reagent strips with or without reflectance meters are not satisfactory for diagnostic purposes. Fasting specimens should be drawn in the morning, 10 to 14 hours following the last food intake. Postprandial blood samples may be obtained 2 hours after a mixed meal (breakfast or lunch) containing 75 to 100 g carbohydrate.
7. Blood pressure and any significant interim history of cardiovascular

symptoms.

8. Resting ECG. For first and second class applicants over age 40 years, a more thorough cardiovascular assessment is to be made annually, including an appraisal of circulatory efficiency. A treadmill test is required every two years unless it is medically contraindicated. The treadmill protocol should be attached to the evaluation.

9. Evidence of normal renal function initially and on a yearly basis thereafter (BUN $\leq$ 25 mg/dl or creatinine $\leq$ 1.5 mg/dl).

Control will be considered acceptable when fasting plasma glucose is $\leq$ 140 mg/dl and $\geq$ 60 mg/dl (a fasting plasma glucose $<$ 100 mg/dl in a person taking an oral hypoglycemic agent may indicate excessive or inappropriate use of the sulfonyluresa, and is a cause for review and probable revision of medication), 2-hour postprandial glucose is $<$ 175 mg/dl, and glycosylated hemoglobin does not exceed the upper limits of normal in a given laboratory by more than 1% (in many laboratories the upper limit of normal is 7%). Care must be exercised to minimize the risk of hypoglycemia in persons treated with oral agents. Prolonged fasting or severe restriction alone or in combination with alcohol ingestion must be avoided. Susceptibility to hypoglycemia may persist for more than 24 hours following alcohol ingestion when nutritional status compromised.

Recommendations Concerning Hypoglycemia

A diagnosis of hypoglycemia should not be made in the absence of neuroglycopenic results that are comfirmed by plasma glucose levels. Symptoms in the fasting state in conjunction with a fasting plasma glucose of $\leq$ 50 mg/dl suggests hypoglycemia of an organic cause, and further evaluation is indicated. A history of hypoglycemia may not be disqualifying if appropriate educational and dietary measures have alleviated the symptoms.[7]

Recommended Replacement Therapy for Individuals with Endocrine Deficiencies

Disease or Condition	Therapy
A. Addison's disease (primary adrenal insufficiency)	1. Hydrocortisone (cortisol) 13 mg/m^2/d or approx 20 mg a.m., 10 mg p.m. (may substitute prednisone 5 mg a.m., 2.5 mg p.m.; or cortisone 25 mg a.m., 12.5 mg p.m.)
	2. Florinef (9 -fluorohydrocortisone) 0.05 to 0.10 mg qd
B. Secondary adrenal insufficiency	same as for primary adrenal insufficiency but without Florinef
C. Hypothyroidism, primary or secondary	L-thyroxine 0.05-0.20 mg qd (usual replacement dose is 0.15 mg qd)
D. Hypoparathyroidism	Vitamin D_2 50,000-100,000 U.S.P. units/d (may substitute 1,25-dihydroxyvitamin D (Rocaltrol) 0.25-2.0 mcg qd (usual dose is 0.25-0.50 mcg qd)

NOTE: During the dosage titration period with Rocaltrol, serum calcium levels should be obtained at least once weekly and medication should be discontinued immediately if hypercalcemia is noted. Once a stable serum calcium is achieved, patients should be followed at least every 3 months with serum calcium determinations.

E. Diabetes insipidus

1. DDAVP (desmopressin acetate) 0.05 to 0.3 ml qd intranasally in 1-2 doses (may substitute pitressin-tannate-in-oil 1.0 ml deep intramuscular injection q 24-72 hr)

2. Diapid (lypressin) 1-2 sprays in each nostril may be used, but the effect is usually brief and must be repeated every 4-6 hr

References

1. Smith RJ: Hypoglycemia, in Marble A, Krall LP, et al (eds): Joslin's Diabetes Mellitus, ed 12. Philadelphia, Lea and Febiger, 1985, p 877.

2. The Physician's Guide to Type II Diabetes (NIDDM): Diagnosis and Treatment. New York, American Diabetes Association, p 79.

3. Steel JM, Frier BM, Young RJ, et al: Driving and insulin-dependent diabetes. Lancet 1981;2:354-356.

4. Haunz EA, Brosseau JD: Nonwarning hypoglycemia in drivers with diabetes. Amer Fam Physician 1984;30:198-197.

5. Casparie AF, Elving LD: Severe hypoglycemia in diabetic patients: Frequency, causes, prevention. Diabetes Care 1985;8:141-145.

6. Office guide to diagnosis and classification of diabetes and other categories of glucose intolerance. Diabetes Care 1981;4:335.

7. Metzer B, Freinkel N: Reactive hypoglycemia, in Conn HF (ed): Current Therapy. Philadelphia, WB Saunders Co, 1981, pp 470-472.

The Physician's Guide to Type II Diabetes (NIDDM): Diagnosis and Treatment

Copyright © 1988 by the American Diabetes Association, Inc. All rights reserved.

Office Guide to Diagnosis and Classification of Diabetes Mellitus and Other Categories of Glucose Intolerance

I. Diagnoses Associated with Glucose Intolerance
   A. Diabetes Mellitus (DM)
      1. Type I. Insulin-Dependent-Type (IDDM) Ketosis Prone. Insulin deficient due to islet cell loss. Often associated with specific HLA types, with predisposition to viral insulitis or autoimmune (islet cell antibody) phenomena. Occurs at any age, common in youth.
      2. Type II. Non-Insulin-Dependent-Type (NIDDM) Ketosis Resistant. More frequent in adults but occurs at any age. Majority are overweight. May be seen in family aggregates as an autosomal dominant genetic trait. May require insulin for hyperglycemia during stress.
      3. Diabetes Associated with Certain Conditions or Syndromes. Hyperglycemia occurring in relation to other disease states. Pancreatic diseases, drug- or chemical-induced diabetes, endocrinopathies, insulin receptor disorders, certain genetic syndromes.
   B. Impaired Glucose Tolerance (IGT). Abnormality in glucose levels intermediate between normal and overt diabetes. May "worsen to diabetes," improve toward normal, or remain unchanged on serial testing.
   C. Gestational Diabetes (GDM). Glucose intolerance with recognition of onset during pregnancy.

II. Criteria for Diagnosis
   A. Diabetes Mellitus—Adult
      1. Unequivocal elevation of plasma glucose (≥ 200 mg/dl) and classic symptoms of diabetes including polydipsia, polyuria, polyphagia, and weight loss.
      2. Fasting plasma glucose ≥ 140 mg/dl on two occasions.
      3. Fasting plasma glucose < 140 mg/dl and 2-h plasma glucose ≥ 200 mg/dl with one intervening value ≥ 200 mg/dl following a 75-g glucose load (OGTT).
   B. Impaired Glucose Tolerance (IGT)
      Fasting plasma glucose < 140 mg/dl and 2-h plasma glucose ≥ 140 mg/dl and < 200 mg/dl with one intervening value ≥ 200 mg/dl following a 75-g glucose load.
   C. Gestational Diabetes (GDM)
      Criteria of O'Sullivan. Two or more of following plasma glucose concentrations met or exceeded using a 100-g glucose load: FPG 105 mg/dl; 1-h [illegible] mg/dl; 2-h 165 mg/dl; 3-h 145 mg/dl.
   D. Diabetes Mellitus—Children
      1. Classic symptoms (see adult) with random plasma glucose ≥ 200 mg/dl.
      2. Fasting plasma glucose < 140 mg/dl and 2 OGTT and one intervening value ≥ 200 mg/dl (use 1.75 g/kg to maximum of 75-g glucose load).

III. Normal Glucose Values—Nonpregnant Adults
   FPG ≤ 115 mg/dl; 2-h PG < 140 mg/dl. OGTT values between zero time and 2 h PG < 200 mg/dl. Plasma glucose concentrations above these values but below those listed for diabetes or IGT are not diagnostic for these conditions.
   The following terms have been eliminated: latent, subclinical, and chemical diabetes mellitus; prediabetes; potential diabetes; adult-onset, maturity-onset, and juvenile-onset diabetes.

IV. Instructions for Glucose Tolerance Testing
   Ordinarily glucose tolerance testing is not recommended in the following circumstances: (1) fasting hyperglycemia; (2) persons taking medications such as thiazide diuretics, Dilantin, propranolol, birth control pills containing estrogens, birth control pills [illegible] tablets; (3) hospitalized patients or acutely ill patients or [illegible] patients. The OTT should be performed only on persons who have been on unrestricted diet and physical activity 3 days before testing. A 75-g oral glucose load should be administered in the a.m. post [illegible] The person should remain seated and not smoke during the test.

Prepared from "Classification of DM and Other Categories of Glucose Intolerance," the National Diabetes Data Group, NIH, by C. R. Shuman, M.D., and J. L. Spratt, M.D., American Diabetes Association Statistics Committee, January 1980.

Best Available Copy

No. 74

TO: Chairmen of the Board, Presidents, and Executive Directors of Affiliate Associations

FROM: Gordon T. Stulberg
Chairman of the Board

SUBJECT: AMERICAN DIABETES ASSOCIATION EMPLOYMENT POLICY STATEMENT

The American Diabetes Association policy on employment states that, "Any diabetic, whether insulin-dependent or non-insulin-dependent, should be eligible for any employment for which he or she is individually qualified." This policy is consistent with, and supports the Federal Rehabilitation Act and most state laws and important court decisions. It implies that there should be individual consideration of each candidate for employment, avoiding blanket policies with regard to people with diabetes.

The policy is also consistent with the following:

1. Diabetes as such should not exclude a person from employment.

2. Individual jobs and individual people with diabetes should be considered, weighing such factors as treatment regimen, (diet, oral hypoglycemic agents, insulin), presence of complications of diabetes, and specific job requirements or hazards.

In connection with this policy, it should be noted that:

1. Physicians should examine employees to determine whether individual factors, such as a history of severe insulin reactions (with impaired mentation), or impaired vision will contraindicate certain job situations.

2. Intensive efforts are necessary to educate physicians in evaluating the work each individual with diabetes is capable of performing.

3. Public education, especially of employers, is also important to optimize utilization of the person with diabetes in the workplace.

Best Available Copy

Approved by
American Diabetes Association, Inc.
Board of Directors
February 22, 1974

IV - 10c

National Standards for Diabetes Patient Education Programs

National Diabetes Advisory Board November 1983

National standards for diabetes patient education programs have been endorsed by the National Diabetes Advisory Board. These standards were developed under the auspices of the Board and in collaboration with the American Association of Diabetes Educators, American Diabetes Association, Centers for Disease Control, Diabetes Research and Training Centers, International Diabetes Center (Minneapolis), Juvenile Diabetes Foundation, and National Diabetes Information Clearinghouse.

This statement presents the rationale for the standards and a plan for their implementation. It includes a summary and description of the standards and a tabular presentation for easy reference.

The Need for National Standards

Major strides have been made in the treatment of diabetes during the last decade as a result of biomedical research, technological advances, and improved application of currently available knowledge and resources. Dramatic increases in our knowledge of effective approaches to prevention of some of the complications of diabetes include better methods to assess and control blood glucose. It is now possible to limit the severity of some long-term effects of the disease and thus reduce its medical, social, and economic impact.

Several barriers, however, still preclude the widespread availability of preventive approaches in self-care. These barriers include lack of patient and provider knowledge about diabetes, inadequate reimbursement policies, and lack of coordination among key components of the health care system. One major impediment has been the lack of national standards to assure that the education provided to people with diabetes is of an acceptable quality and appropriate for the individual.

The National Diabetes Advisory Board, in collaboration with experts from within and outside the diabetes community, has developed national standards for diabetes patient education programs. These standards establish specific parameters against which programs can measure themselves. The standards are rigorous enough to be acceptable to the diabetes community, yet flexible enough to be practical for the primary care community. They are applicable in any health care setting. The Board encourages adoption of these standards by all diabetes patient education programs.

7550 Wisconsin Avenue • Room 616 • Bethesda, Maryland 20205 • (301) 496-6045

National Plan for Implementation of the Standards

The National Diabetes Advisory Board (NDAB) is mandated by Congress to oversee the Long Range Plan to Combat Diabetes. In addition to its advisory role, the Board has come to serve as a forum through which the diabetes community can focus on common needs and problems and share in their solutions. Through a series of workgroups, the Board and cooperating organizations determined that the availability of the standards would be enhanced by a process to ensure their widespread application. They are therefore developing a national system of recognition for diabetes patient education programs that meet the standards. Recognition is a voluntary process through which programs meeting the standards are formally identified for a level of performance, integrity, and quality entitling them to the confidence of the community they serve. The process is flexible enough to apply to programs that conform to other standards, provided the other standards adhere to the national consensus standards or are modified to do so.

The standards and the recognition process will be pilot tested during 1984 and 1985. Pilot testing will be conducted under the auspices of a Board-appointed steering committee consisting of representatives of the diabetes-related organizations involved with the Board in the development of both the standards and the recognition process. These organizations include the American Association of Diabetes Educators, American Diabetes Association, Centers for Disease Control, Diabetes Research and Training Centers, International Diabetes Center (Minneapolis), Juvenile Diabetes Foundation, and National Diabetes Information Clearinghouse. The results of the pilot testing will form the basis for modifications in the standards and for any required adjustments in the recognition process prior to nationwide implementation. Support materials will be available to provide diabetes patient education programs with additional information on (1) how to meet the standards, (2) how to initiate or upgrade a program to meet them, and (3) how to apply for recognition.

Summary and Description of the Standards

Diabetes is a serious and common disease that is treated directly or indirectly in practically every health care facility in the nation, regardless of size or location.

In chronic diseases, especially diabetes, patients are required to assume a major share of responsibility for their own care. Only an informed and well-motivated person who has the support of the primary health care provider can carry out this responsibility effectively. Evidence is growing that inadequate patient knowledge results in multiple hospital admissions, excessive use of emergency rooms, unnecessary medication, and a high incidence of long-term complications of diabetes, all of which increase human suffering and escalate the costs of care. Studies testing patient education as the variable component of the treatment regimen have shown consistent reductions in these measures. Education for self-care is therefore recognized to be a fundamental component of quality treatment for the individual with diabetes.

At the present time, both the quantity and quality of education offered to people with diabetes vary considerably in the United States. Experience in other fields has demonstrated the ability of uniform standards to improve the quality, effectiveness, and availability of programs. It is hoped that the implementation of national standards will result in increased access to this fundamental component of treatment by stimulating adequate reimbursement for diabetes patient education.

The diabetes patient education standards consist of 10 components that will enable an institution to establish a new program or modify an existing one. Each standard offers the flexibility required to tailor a patient education program to the type of diabetes, its duration, and the life-stage of the diabetic person. Many of the standards are overlapping, reflecting the interdependence among all components of an effective diabetes education program.

1. Needs Assessment. A successful program is the product of a flexible policy based upon the needs of the community it is intended to serve. Since the diabetes caseload varies from one institution to another, each institution should assess its own needs and match its resources to the needs of its caseload. The needs assessment should be performed initially to guide the management of the program and to form the basis for program planning. It should be a continuing process that will allow the program to adapt to changing service requirements. In addition to the needs of the program, the needs of the individual patient should be assessed to provide the basis for the instructional program offered to each patient. The person with diabetes is recognized to be an equal partner in all aspects of the educational process.

2. Planning. Planning is an essential component of a diabetes patient education program. The planning process should describe the program's goals and objectives, target audience, setting (inpatient, outpatient), patient-referral mechanisms, procedures, and evaluation methods. The planning process should be a cooperative effort involving people with diabetes as well as health professionals.

3. Program Management. Effective management is required to implement a patient education program. A variety of health care professionals is involved in the total care of people with diabetes. Clear lines of authority and efficient systems for communication should be established among everyone involved in the program. The ultimate responsibility for all aspects of program management should rest with one person designated as the program coordinator. In addition, an advisory committee should be established to assist the coordinator and other members of the program staff in setting policy and managing the program.

4. Communication and Coordination. Several levels of communication are essential to the effective coordination of the program. Physician leadership and participation are necessary to ensure the integration of patient education into the treatment regimen. A physician should be identified to serve as the liaison between the education program coordinator and the medical staff. In addition, the institution should maintain regular channels of communication with its staff and the community it serves to inform diabetes patients and their families about the availability of the program. All information on the patient's educational experience should be incorporated into the medical record.

At the present time, both the quantity and quality of education offered to people with diabetes vary considerably in the United States. Experience in other fields has demonstrated the ability of uniform standards to improve the quality, effectiveness, and availability of programs. It is hoped that the implementation of national standards will result in increased access to this fundamental component of treatment by stimulating adequate reimbursement for diabetes patient education.

The diabetes patient education standards consist of 10 components that will enable an institution to establish a new program or modify an existing one. Each standard offers the flexibility required to tailor a patient education program to the type of diabetes, its duration, and the life-stage of the diabetic person. Many of the standards are overlapping, reflecting the interdependence among all components of an effective diabetes education program.

1. Needs Assessment. A successful program is the product of a flexible policy based upon the needs of the community it is intended to serve. Since the diabetes caseload varies from one institution to another, each institution should assess its own needs and match its resources to the needs of its caseload. The needs assessment should be performed initially to guide the management of the program and to form the basis for program planning. It should be a continuing process that will allow the program to adapt to changing service requirements. In addition to the needs of the program, the needs of the individual patient should be assessed to provide the basis for the instructional program offered to each patient. The person with diabetes is recognized to be an equal partner in all aspects of the educational process.

2. Planning. Planning is an essential component of a diabetes patient education program. The planning process should describe the program's goals and objectives, target audience, setting (inpatient, outpatient), patient-referral mechanisms, procedures, and evaluation methods. The planning process should be a cooperative effort involving people with diabetes as well as health professionals.

3. Program Management. Effective management is required to implement a patient education program. A variety of health care professionals is involved in the total care of people with diabetes. Clear lines of authority and efficient systems for communication should be established among everyone involved in the program. The ultimate responsibility for all aspects of program management should rest with one person designated as the program coordinator. In addition, an advisory committee should be established to assist the coordinator and other members of the program staff in setting policy and managing the program.

4. Communication and Coordination. Several levels of communication are essential to the effective coordination of the program. Physician leadership and participation are necessary to ensure the integration of patient education into the treatment regimen. A physician should be identified to serve as the liaison between the education program coordinator and the medical staff. In addition, the institution should maintain regular channels of communication with its staff and the community it serves to inform diabetes patients and their families about the availability of the program. All information on the patient's educational experience should be incorporated into the medical record.

5. Patient Access to Teaching. It should be the policy of the institution to facilitate access to patient education for the target audience specified in the plan. This is promoted by a commitment to routinely inform both patients and staff about the availability and benefits of patient self-care programs. Diabetes patient education should be regularly and conveniently accessible, and the instructional program should be able to respond to patient-initiated requests for information. The program permits referral by health professionals, health care agencies, or individual patients. The instructional design encourages active patient participation.

6. Content/Curriculum. The individual patient's needs assessment provides the basis for the instructional program offered to each patient. The assessment should be documented and should include all relevant information regarding the patient's treatment, education, and support systems. Responsibility for various facets of the assessment can be divided among the instructional team members. Curriculum and instructional materials should be appropriate for the specified target audience, taking into consideration the type and duration of diabetes and the age and learning ability of the individual. Both curriculum and available community resources should be reviewed and updated periodically. The institution should provide the program with adequate space, personnel, budget, and materials.

7. Instructor. Qualified personnel are essential to the success of a diabetes patient education program. Each institution should be responsible for identifying and evaluating its instructors. Instructors should be skilled professionals with recent experience and training in both diabetes and educational principles. The number of instructors should be proportional to the caseload requirements. Instructors should be allotted sufficient time to complete the instructional program.

8. Followup. Followup services are important because diabetes requires a lifetime of proper care. The institution should provide followup services that include periodic reassessment of the patient's knowledge and skills and should offer supplementary educational services when warranted. Written communication between the program staff and the primary care physician is essential for ongoing identification of the patient's needs. This is especially appropriate in regard to referral for early diagnosis and treatment of the complications of diabetes. Referral to community resources may also provide ongoing support for long-term psychosocial needs and behavioral modification skills. If a patient changes care settings, the institution should request the patient's permission to send his/her records to the new health care setting.

9. Evaluation. The institution should review the educational program periodically to ascertain that it continues to meet the national standards. This review should be conducted by the advisory committee. The results of this review should be utilized in subsequent program planning and modification. An assessment of each patient's needs and progress should also be conducted at regular intervals.

10. Documentation. Program planning and evaluation should be documented to provide the basis for future program development and modification. All information about the patient's educational experience should be documented in the patient's medical records, as should communication among treatment and education professionals.

National Standards for Diabetes Patient Education Programs

This table presents the standards in a form for easy reference. Standards applicable to the facility offering the program are designated "institution standards" and are separated from those applicable to the education program itself, which are designated "program standards."

COMPONENTS	STANDARDS	
	Institution	Program
1. Needs Assessment	• The institution shall assess its diabetic caseload to determine the allocation of personnel and resources to serve the instructional needs of the caseload. • There shall be a reasonable match between caseload requirements and resources allocated.	• An individualized and documented ongoing assessment of needs shall be developed with the patient's participation. This shall include medical history, present health status, previous diabetes education, health services utilization, associated medical conditions or risk factors, diabetes knowledge, skills, attitudes, self-assessment, identification of support system, barriers to learning, and financial status. • The needs assessment shall be the basis for the education program delivered to each patient.
2 Planning	• The institution shall have a written policy that affirms patient education as an integral component of quality diabetes care.	• The participants in planning shall include health professionals involved in the care and education of persons with diabetes and persons with diabetes and their families. • The planning process shall define (in order): a) Program goals and objectives b) Target audience c) Program setting d) Patient access mechanisms e) Instructional methods f) Resource requirements g) Patient followup mechanisms h) Evaluation
3 Program Management	• A coordinator shall be designated and responsible for all aspects of the program. • The organizational relationships, lines of authority, staffing, and operational policies shall be defined. • A standing advisory committee with both medical and community/consumer representation shall be established.	• Not applicable.
4 Communication/ Coordination	• A physician shall be identified to serve as liaison between the program coordinator and the medical staff. • The institution shall regularly inform its staff and the patients (and potential patients) it serves of the availability of its diabetes patient education program.	• All information about the patient's educational experience shall be permanently incorporated into the patient's (medical) records maintained by the institution. • The role of each education team member shall be clearly defined, and the intercommunication between each shall be documented in the patient's record. • There shall be written evidence of coordination between different care settings.
5 Patient Access to Monitoring	• The applicant institution shall have a policy to inform patients routinely about the benefits and availability of patient education.	• The program shall be regularly and conveniently available. • The program shall be responsive to patient-initiated requests for information and/or participation in the program's activities.

COMPONENTS	STANDARDS	
	Institution	Program
6 Content/Curriculum	• The institution shall provide space, personnel, budget, and instructional materials adequate for the program. • Assessment of available community resources shall be performed periodically.	• The program shall be capable of offering information on each of the following content items as needed: a) General facts b) Psychological adjustment c) Family involvement d) Nutrition e) Exercise f) Medications g) Relationship between nutrition/exercise/medication h) Monitoring i) Hyperglycemia and hypoglycemia j) Illness k) Complications (prevent, treat, rehabilitate) l) Hygiene m) Benefits and responsibilities of care n) Use of health care systems o) Community resources • The institution shall specify the mechanism by which the curriculum shall be reviewed, approved, and updated.
7 Instructor	• The institution shall identify appropriate instructional personnel and ascertain their competence. • The numbers of personnel identified shall be suitable for the diabetic caseload within the institution. • Designation of time for identified instructors shall be appropriate to accomplish the necessary educational objectives.	• Instructors (health professionals and others) shall be part of a comprehensive care and education program. • Instructors shall have recent experience and training in diabetes and knowledge and skills in educational principles and their application.
8 Follow-up	• The institution shall transmit the educational record to other appropriate health care settings when a patient transfers his or her care responsibilities.	• The program shall provide followup services for those patients who wish to maintain continuity of education within the institution. These services shall include: a) Periodic reassessment of knowledge and skills b) Timely reeducation based on reassessment c) Communication with the primary care provider about the need for professional and nonprofessional services.
9 Evaluation	• The institution shall review periodically the performance of the instructional program and ascertain that it continues to meet national standards.	• The program shall conduct and record an individualized assessment of each patient's original needs and progress at regular intervals. • The program shall be reviewed in ongoing fashion for both process and outcome, and the results of this review shall be used in subsequent planning and program modification.
10 Documentation	• All aspects of the evaluation shall be recorded by the sponsoring institution and reviewed periodically to ascertain that national standards are being maintained.	• All aspects of the educational program offered to each patient shall be recorded in that patient's medical record as maintained by the institution.

PREVIOUS PAGE IS BLANK

Mental and Behavioral

I. DSM III

The Diagnostic and Statistical Manual of the American Psychiatric Association, Third Edition (DSM III) presents several problems to the FAA for regulatory purposes. Foremost among these is the listing of explicit criteria that are required for each diagnosis. While this increases the reliability of psychiatric diagnosis, it makes it more difficult to establish a diagnosis that can be defended in administrative proceedings.

For example, in a psychiatric evaluation, which is somewhat adversary in nature, it is often difficult to elicit a history of delinquent behavior before the age of 15, which is required for the diagnosis of antisocial personality disorder. As a result, this diagnosis may be rejected in administrative proceedings, even in the face of ample evidence of adult antisocial acts. Another problem exists with the term, "alcoholism." DSM III has eliminated this term and substituted the diagnoses of "substance abuse" and "substance dependence." The term, "alcoholism," has a strong traditional connotation as well as a specific definition in the Federal Air Regulations (FARs), and was clinically useful and consistent with the identification of individuals who are a threat to aviation safety. This change in terminology necessitates new guidelines concerning the approval or denial of certification of pilots with either of these diagnoses.

A third problem concerns the psychotic disorders, which are no longer listed as a category in DSM III. Whereas these conditions as a group could previously be listed as disqualifying, DSM III has developed new categories, such as borderline and schizotypal personality disorders, which include diagnostic criteria that would previously have been considered evidence of psychosis. The result was to remove certain psychosis-like conditions from a category that always was disqualifying and place them in the group of personality disorders.

For these and other reasons, some consideration was given to retaining DSM II for

the purpose of the FARs. However, because DSM III is now so widely accepted, despite its arguable deficiencies this would not be an acceptable solution. Rather, the FARs should state the limitations of DSM III for regulatory purposes and support diagnoses based upon good clinical judgment, even though it may not be possible to document all of the specific criteria. As noted in DSM III, the criteria "are offered as useful guides," and "for most of the categories the criteria are based on clinical judgment, and have not yet been fully validated." This wording, if included in the regulations, would legitimize this approach, and make it possible to sustain in administrative hearings diagnoses that are based upon sound clinical judgment.

Disqualifying Conditions

The standards for psychiatric conditions should be the same for all classes of medical certification. Mental disorders adversely affect judgment and behavior in ways that create a potential hazard to others in the complex airway system. For this reason the private pilot with a mental disorder represents a risk as potentially significant to others as a commercial pilot.

- Substance use disorders

  The following conditions are disqualifying:

  1. Alcohol abuse
  2. Alcohol dependence
  3. Any other substance abuse, substance dependence or related substance use disorders including, but not limited to, those associated with barbiturates; other sedative/hypnotics; muscle relaxants; the anxiolytics; opioids; central nervous system stimulants such as cocaine and amphetamines; and hallucinogens such as phencyclidine, cannabis, and volatile solvents and gases.

The DSM III makes a distinction between substance abuse, which is the use of a substance without increased tolerance, and substance dependence, which is the use of a substance with increased tolerance. One should not view the former as a minor disorder and the latter as a major disorder. In fact, many substance abuse cases are more serious, more refractory to treatment, and have a poorer prognosis than many substance dependence cases. The term, "alcoholism," has been replaced with the terms, "alcohol abuse" and "alcohol dependence." While it is not yet clear that these new terms have any greater utility for prescribing treatment or establishing prognosis, they are nonetheless more specific and do represent current diagnostic practice.

The two-year period of sustained abstinence that is recommended for the new standard may be too severe in some cases. Early treatment of relatively minor cases is increasingly common, and some of these pilots could return safely to flight status at an earlier time if they are carefully assessed and closely monitored. Another problem with using the criterion of a two-year period of sustained abstinence is that it is very difficult to prove or disprove. Other criteria for reconsideration of certification might include continued involvement with a treatment program, episodic examination of body fluids for drugs or for organic damage from the drugs, with tests such as the GGT or mean corpuscular volume, or documentation of a stable life style.

Any and all use of illicit substances should be proscribed among pilots. From an aeronautical standpoint there are two critical

hazards with such substances. First, no quality control measures exist. Thus, a substance sold as "marijuana" may be dandylions laced with phencyclidine; or "amphetamines" may be caffeine laced with strychnine. Second, unlike alcohol, the dose-effect relationships are either unknown or extremely variable. Some substances such as alcohol and morphine produce fairly predictable central nervous system effects in most individuals at low to moderate doses. This is not true of substances such as cocaine, cannabis, amphetamines, LSD, and phencyclidine.

Random urine tests should be considered as a regulation reinforcement technique in airline pilots and controllers; the FAA might consider doing a small study of urine tests during medical examinations of randomly selected private pilots. Urine tests have an advantage over blood tests in that they reflect the status of the blood levels over a period of time. Many drugs will be detected in the urine as long as 24 hours after use, even when they are no longer detectable in the blood. Amphetamines can be detected as long as 48 hours after use. Tetrahydrocannabinol may occasionally be detected two to three weeks after the last use in a chronic abuser.

The history and examination Form 8500 should be expanded to include items relevant to drug abuse. Signs that indicate the presence of substance abuse include trauma to the head, perforation of the nasal septum, nystagmus, reddened conjunctivae, coarse rales or ronchi, enlarged or tender liver, unexplained epilepsy beginning in an adult, tremor and ataxia. Mental status items include dirty or disheveled

appearance; uncooperative, bizarre or inexplicable behavior; labile mood; slurred speech; impaired memory; and difficulty with abstract thinking. A formal mental status examination should be routinely performed. We provide for the AME a short, four-item mini-mental status examination that has appropriate complexity for a pilot population. Should the applicant not answer all four items correctly, we suggest the AME follow up with the mini-mental status examination of Folstein, Folstein and McHugh (see appendix in the recommended AME Guide mental and behavioral section).

The report by a professional of a substance-abuse problem should be prepared by a psychiatrist who is skilled in the evaluation and treatment of substance abuse and familiar with its manifestations. The psychiatrist's credentials may be assessed by considering fellowship training in substance abuse, staff affiliation with a substance abuse treatment program, association with related professional groups such as the American Society for Alcoholism, and clinical, teaching or research activities in substance abuse. The FAA should specify that the report contain a review of treatment records, detailed history of pattern of substance use; psychosocial consequences of use; a complete, formal mental status examination; a description of associated psychiatric syndromes; and a complete physical examination listing evidence of neurological abnormalities, summarizing a psychological evaluation, and listing laboratory studies.

Recertification of individuals with a history of substance abuse, substance dependence and related substance-use disorders should require sustained total abstinence from alcohol for not less than the preceding two years, and from other substances of abuse for not less than the preceding five years; also there should be stable social and occupational functioning and the absence of mental disorder or psychopathology as demonstrated by psychiatric evaluation and psychological testing. In selected individual cases judged to have an excellent prognosis and if the means for careful monitoring exists, a shorter period of abstinence may be acceptable.

- Psychotic disorders

The following conditions are disqualifying:

1. Schizophrenic disorders

   Disorganized

   Catatonic

   Paranoid

   Undifferentiated

   Residual

2. Paranoid disorders

   Paranoia

   Shared paranoid disorder

   Acute paranoid disorder

   Atypical paranoid disorder

3. Psychotic disorders, not otherwise classified

   Schizophreniform disorder

   Brief reactive psychosis

Schizoaffective disorder

Atypical psychosis

Any active psychotic disorder should be grounds for disqualification. The criteria for the schizophrenic disorders are emotional blunting, social withdrawal, eccentric behavior, illogical thinking, and loosening of associations. In addition, the paranoid disorders and the pervasive developmental disorders should be disqualifying. Also, an individual with a history of schizophrenia according to DMS III criteria should not normally be considered for medical certification. It is recognized that an occasional individual may have a single psychotic episode, make a full recovery, have no recurrences and require no medication. Such a pilot, who has been symptom-free, off medication and making an excellent social and occupational adjustment for a significant period of time, as reflected by superior or very good functioning on DSM III Axis V, may be recertified on a special issuance basis by the Federal Air Surgeon.

A pilot with a brief reactive psychosis may be recertified after two years, and one with schizophreniform psychosis recertified after five years if the individual remains symptom free, functions well in all spheres, does not require medication, and has no residual symptoms evident on psychiatric evaluation and psychological testing. Psychiatric evaluation should confirm freedom from all signs and symptoms of the illness, based upon a psychiatric history and a formal mental status examination, including testing for attention and concentration, abstraction, and judgment. Psychological testing

should confirm the freedom from any significant underlying psychopathology and vulnerability to decompensation. Such individuals should be re-evaluated periodically for several years after recertification. Long term follow-up of this group would be useful in establishing future policy.

- Affective disorders

  1. Major affective disorders

     Bipolar disorder

     Major depression

  2. Other specific affective disorders

     Cyclothymic disorder

  3. Atypical affective disorders

     Atypical bipolar disorder

     Atypical depression

Bipolar disorders should be disqualifying. As with the schizophrenic disorders, the regulations should contain no explicit criteria for recertification, but cases could be considered on an individual basis by the Federal Air Surgeon.

Major affective disorders tend to recur, and follow-up studies reveal a significant relapse rate. Comparative studies of persons with bipolar disorders, using lithium and placebo show relapse rates after one year on placebo of 45-100% (mean about 75%) and 0-58% on lithium (mean 35%). Although lithium works well in many individuals, the risk of relapse in the first year is still high. In depression, follow-up studies

of one to three years duration point to an annual relapse rate of about 29% and a cumulative rate of about 50%. The corresponding rates with placebo are 40% and 77%. The literature may indicate different results from those in clinical practice, because the use of psychotherapy in clinical practice may make a major difference. Also, persons who become study subjects may be sicker and not typical of most persons with bipolar disorders.

Dysthymic disorders need not be listed specifically as a disqualifying condition. They come under the category of disorders that should be disqualifying if they are manifested by symptoms that might adversely affect flying safety.

It is not possible to recommend that individuals taking heterocyclic antidepressants or monoamine oxidase inhibitors be allowed to pilot aircraft. One of the significant risks of heterocyclic antidepressants is cardiac arrhythmia. Monoamine oxidase inhibitors also have a significant risk of causing sudden incapacitation. Lithium has potential central nervous system effects that create significant risks. Certain individuals might be reconsidered for certification while on these medications, if they have a long history of freedom from symptoms, good functioning, absence of significant side effects, and evidence of strict compliance with treatment. In the event pilots are certified while taking lithium, periodic neurologic examinations, including evaluation of cognitive functioning, should be required.

o Personality disorders

The following types of personality disorders are disqualifying:

Paranoid

Schizoid

Schizotypal

Histrionic

Narcissistic

Antisocial

Borderline

Persons with personality disorders other than those listed above might be certified if during the preceding five years they were free from any behavior reflecting impaired judgment or creating legal or disciplinary problems, had stable social and occupational functioning, and had no other mental disorder or psychopathology as demonstrated by psychiatric evaluation and psychological testing. Also, such individuals should have no evidence of significant impairment on DSM III Axis V having implications for flying safety.

The personality disorders represent a critical problem because of the significant implications for flying safety and the difficulties of assessment. The basic hallmark of personality disorders is "disordered behavior." One does not observe the pathology in terms of formal mechanisms of cognition, perception, and regulation of affect. In fact, such persons are likely to demonstrate normal functioning on a mental status examination, on routine psychometric tests and even on projective tests. It is only when the individual is seen in action in real life that one observes problems and

disordered and dysfunctional behavior. Thus, diagnostic assessment and prognostic evaluation are based on the history of the person's behavior in ongoing life events. The problematic behavior may include aberrant emotional reactions, processing of data or motor actions. The dysfunctional behavior is the final common pathway of deficits in the appropriate processing of internal and external stimuli. Stimuli are not appropriately identified, held in abeyance for scrutiny, and evaluated and assessed in terms of context; therefore, a reasonable set of alternatives for action are not considered. Rather, one of two major responses occur: an immediate stimulus/context bound response, or a stereotyped programmed response to all stimuli, which ignores the uniqueness of the situation. The consequence of the first response is "impulsive behavior," and the response to the second is rigid, nonconforming behavior that is totally predictable.

Rountine psychiatric and psychological testing does not identify these disorders. Observations of everyday life reactions by peers and supervisors are the most important data in identifying personality disorders that disqualify. Observations of pilot behavior during training and flight simulation or in actual flight operations are critically important. Unusual incidents, deviant flight actions, and questionable judgments often reflect personality disorders. In seeking to assess the presence and significance of a personality disorder, items such as the following would merit further investigation:

1. Unusual cognitive and affective attitudes and values: poor moral judgment, poor problem-solving, high anxiety responses, untoward emotional lability, and biased and prejudiced attitudes.

2. Disturbances in social behavior: traffic violation tickets, crashes, bad checks, overdrawn bank accounts, poor credit ratings, neighborhood disturbances, unusual or conflictual behavior at social events, marital or family conflict.

3. Pathological gambling, kleptomania, high risk stunts, thrill-seeking behavior, isolated explosive behavior.

4. Disturbances in vocational behavior: poor interpersonal relations with coworkers, peers, supervisors or subordinates conflicts with administrative authorities; untoward inattention to or challenge of policies and procedures.

Each of these pieces of behavioral evidence by itself does not necessarily indicate a personality disorder. Rather it is the observation of chronic, repetitive, multiple, diverse actions indicative of either impulsive or stereotyped behavior patterns that establishes a data base for the diagnosis of personality disorder.

It is obvious that no arbitrary line can be drawn between the normal variations of personality organization or style and a psychopathological personality disorder. Also, a person might have a personality disorder that might not impair function as a pilot. For example, an obsessive-compulsive man might have typical problems in maintaining an intimate relationship with a woman, but be able to function effectively as a pilot. On the other hand, an individual might have a nondiagnosable character style, yet exhibit

flight stress responses that are pathological. For example, a passive-aggressive pilot might shy away from a critical in-flight decision because of fear of taking action.

In summary, personality disorders should be considered as a class of potentially disqualifying disorders. Evaluation should stress ongoing evidence of either stereotyped or impulsive response behavior. Evaluation should focus on the functional significance of behavioral patterns for pilot performance at an acceptable safety level, including stress response behavior. Clearly it is very difficult to make a diagnosis of personality disorder during a periodic physical examination, in the absence of any outside information about the individual's behavior. Any such information from outside sources that suggests impulsive or stereotyped maladaptive behavior should be investigated. Any available incident reports, records of arrest or legal proceedings, or medical records should be obtained prior to evaluation of the applicant. This is important because an interview without these data is likely to be worthless.

Routine psychological and psychometric testing is often unrevealing. The Millon Clinical Multiaxial Inventory, which is an objective instrument for diagnosing personality disorder, may be useful in the pilot population, and it should be considered for use on an experimental basis.

o Anxiety, somatiform and dissociative disorders

The following are disqualifying:

1. Anxiety disorders

Agoraphobia

Panic disorder

2. Dissociative disorders

Psychogenic amnesia

Psychogenic fugue

Multiple personality

Depersonalization disorder

Atypical depersonalization disorder

Most individuals with panic disorders and agoraphobia will not attempt to fly. Yet, because of the risk of sudden incapacitation from a spontaneous panic attack, these disorders must be considered disqualifying. Individuals with a history of panic disorder or agoraphobia who have sustained freedom for at least two years from any symptoms that would be a hazard to flying safety and who are taking no psychotropic medication may be certified. While tricyclic antidepressants, monoamine oxidase inhibitors or benzodiazapines are often successful in treating these disorders, the medications constitute a risk for aviation due to psychomotor impairment, the possibility of incapacitating symptoms if the drugs are not taken properly or in the case of the monamine oxidase inhibitors, the possibility of sudden severe side effects. Persons requiring continuing treatment with one of these drugs should be disqualified. The rare individual who is symptom-free for a period of two or more years while on medication, who has had no significant side effects, and who has demonstrated excellent compliance with treatment might be considered for recertification on an individual basis. Follow-up on a regular basis would be required.

Dissociative disorders are rare, but they cause such pervasive impairment of judgment and performance that they must be considered disqualifying. Routine recertification of individuals with a diagnosis of dissociative disorder cannot be recommended because of the gross incapacitation caused by the disorder and its common association with underlying psychopathology. An exceptional case might be considered for recertification.

In most cases, simple phobias of objects or situations that are easily avoided and that are not encountered in the aviation environment would be unlikely to create any specific risk for flight safety. Phobias that carry some risk of incapacitation can be handled by the regulatory statement covering other mental disorders that make the applicant unable to perform aviation duties safely.

Generalized anxiety disorders are characterized by persistent anxiety without recurrent spontaneous panic attacks. Impairment in occupational functioning is rarely more than mild, and most persons in this category are unlikely to constitute a risk to aviation safety. Individuals with severe anxiety that might interfere with judgment and performance and that is apparent to examiners because of a high degree of motor tension, autonomic hyperactivity, and a history of impairment in functioning can be disqualified under the provision of other conditions that make the applicant unable to perform aviation duties safely. It is unlikely that individuals with such symptoms would seek certification to fly. Individuals who require treatment with anti-anxiety drugs, including benzodiazapines, must be disqualified. An individual who requires treatment with beta adrenergic blocking drugs to

control automonic anxiety symptoms should be disqualified, on the basis of the severity of the underlying disorder and on the potential of the drug to cause central nervous system side effects.

Individuals with somatiform disorders are usually eager to prove they are sick, not that they are healthy. Most of these disorders have an early age of onset and a chronic course. It is unlikely that persons with these disorders would attempt to become qualified to fly, and the exceptions can be handled under the provision for other disorders that make the individual unable to perform flying duties safely.

Persons with mild obsessive-compulsive symptoms constitute no threat to aviation. A degree of compulsiveness is necessary in aviation. Persons with severe obsessive-compulsive disorders are unlikely to seek certification and can be disqualified under the general provision for other conditions that make the individual unable to perform flying duties safely.

Specific phobias of flying usually constitute an administrative problem and arise when professional flying personnel complain of developing phobias and use them as a basis for seeking medical retirement or financial compensation. The natural history of simple phobias is that they usually arise in childhood or adolescence and rarely have onset after early adulthood. Consequently, there is a high likelihood that either conscious malingering or unconscious secondary gain plays an important role when a pilot seeks economic compensation on the basis of flying "phobia."

Since simple phobias usually can be treated successfully with behavior

therapy, the thrust of policy in this area should be towards vigorous treatment and early return to duty and away from compensation for disability. While a pilot who claims to develop a fear of flying should not be forced to fly, this subjective symptom alone should not usually constitute a basis for medical retirement.

- Disorders of impulse control

The following are disqualifying:

Pathological gambling

Kleptomania

Pyromania

Intermittent explosive disorder

Isolated explosive disorder

The explosive disorders are particularly significant because of the occurrence of serious loss of control of aggressive behavior that is grossly out of proportion to the precipitating circumstances. The other impulse control disorders should also be considered disqualifying because they are often associated with signficiant underlying personality disorder and psychopathology. No established criteria for recertification can be stated. In rare instances, an individual with an established diagnosis of one of these disorders might be considered for certification if there has been freedom from symptoms and a high level of social and occupational functioning for a long duration, and there is evidence of freedom from other psychiatric symptoms or underlying psychopathology based upon psychiatric and psychological assessment.

- Disorders usually first evident in infancy, childhood or adolesence

  These disorders are included, since several of them will continue into adulthood or, as in the case of eating disorders, may first manifest themselves in adulthood. A history of any of the following disorders is grounds for deferral pending further evaluation, which may include psychiatric and psychological consultation:

  Mental retardation (IQ below 70) or borderline intellectual functioning (IQ 71-84)

  Attention deficit disorders

  Conduct disorders

  Eating disorders

  Stereotyped movement disorders

  Pervasive developmental disorders

- Organic brain syndrome

  This category includes a heterogeneous group of disorders due to transient or permanent dysfunction of the brain, associated with a wide variety of emotional, motivational and behavioral abnormalities. A history of or current evidence of these disorders is cause for deferral pending psychiatric and psychological testing, to include appropriate tests of intellectual functions.

Psychiatric and Psychological Evaluation

The Federal Air Surgeon, in making determinations about medical qualifications for flying, relies upon psychiatric and psychological evaluations. Since these evaluations are conducted outside the more customary confidential atmosphere of a therapeutic relationship and may not be entirely familiar to many psychiatrists and psychologists, the

professionals who are called upon to conduct them should receive written instructions about the procedure, its purpose, and the type of information that should be obtained. The report from a psychiatric evaluation should document specifically all past psychiatric history and any significant past behavior. The history should be supplemented by a review of all available medical records that have been obtained by the Federal Aviation Administration. Apparent inconsistencies between the known history and the applicant's statements, or overt contradictions within the interview itself, should be probed in depth. Statements about the applicant's apparent reliability, rather than a blanket acceptance of the applicant's unverified statements, are helpful. The report should include the following:

1. Current or recent psychiatric symptoms, aberrent behavior, or psychiatric or other medical findings;

2. A review of the applicant's past and current adjustment in the occupational, marital and social aspects of life;

3. The need for or the use or abuse of any pharmacologically active agent, either for therapeutic or recreation purposes;

4. Any personality traits or other unrecognized factors that affect the risk of future occurrence of a problem or other adverse events;

5. The current psychiatric and psychological functional status and stability of the applicant.

In the evaluation of applicants with a history of alcohol or other substance abuse or dependence, the specific subjects of inquiry to be considered by the examining psychiatrists should be listed.

A battery of tests should be selected by a psychologist that is designed to answer specific questions being asked by the examining psychiatrist; these might include the Rorschach test, the complete Wechsler Adult Intelligence Scale (WAIS) and any other additional tests that are considered appropriate by the psychologist.

The evaluations must be conducted by qualified psychiatrists and psychologists. A qualified psychiatrist generally is one who has been certified by the American Board of Psychiatry and Neurology or who has a background equivalent to board certification. A qualified clinical psychologist generally is one who has earned a PhD from a clinical training program approved by the American Psychological Association (APA), who is licensed in the state in which he or she practices, and who is a diplomate of the American Board of Professional Psychology. A qualified neuropsychologist should have earned a PhD and should be trained in the specialty areas of neurospychology, neurobehavior and clinical neuropsychology. He or she also should be a graduate from an APA-approved clinical training program, be licensed in the state in which he or she is practicing, and be a diplomate of the American Board of Clinical Neuropsychology.

Diagnostic Tests

- Clinical laboratory tests

  Although diagnostic clinical tests, such as the dexamethasone suppression test, are coming into use in psychiatry, they must be considered experimental at this time and cannot be recommended for routine use as part of the aviation medicine evaluation.

  Laboratory tests are increasingly valuable in identifying substance abuse and

assessing recovery. Urine testing for drugs is widely available and relatively inexpensive, and reflects the individuals drug use for the last 12 to 48 hours.

A second group of tests reflects endocrine, nutritional and target organ dysfunction caused by excessive drug or alcohol use. These tests remain abnormal for some days, weeks or even months following excessive use. They include the following:

1. Nutritional: low serum carotene, low folic acid, increased mean corpuscular volume of red blood cells.

2. Endocrine: hypoglycemia or hyperglycemia, positive dexamethasone suppression test, electrolyte abnormalities, high uric acid and low blood urea nitrogen, and abnormal thyroid tests in clinically euthyroid patients.

3. Hepatic: increased GGT and SGOT.

4. Bone marrow suppression: tests for decreased white blood cell count and decreased hemoglobin.

o Mental status testing

Although psychiatric disorders are a major cause for denying certification, and pilot error is a major cause of aircraft crashes, to date no routine psychiatric examination or mental status examination has been required. A simple, routine screening test of higher mental functions should be required during physical examinations. In the AME Guide is a four-item examination

of a complexity that is appropriate for the pilot population.

The items come from psychological tests whose reliability and validity are known. For example, the task of drawing figures that have been presented and removed from view is a part of the Benton Visual Retention Test.[1] The task of repeating 6 numbers is part of the digit-span test Wechsler Adult Intelligence Scale-revised.[2] These tests are used in their full version by psychologists in a clinical setting as part of a thorough mental evaluation of an individual, and each test is scored according to scales that have been validated by field tests. We are proposing that parts of these psychological tests be used as a screening, rather than diagnositic, test for diminished cognitive functions, and that only a perfect score be considered "passing" for screening purposes. The validity of the use of mental status examination questions in this manner is not known and the FAA may consider testing this procedure on a small scale to determine its validity.

References

1. Benton AL: The Revised Visual Retention Test (ed 4). New York, The Psychological Corporation, 1974.
2. Wechsler Adult Intelligence Scale-Revised. New York, The Psychological Corporation, 1981.

Appendix

Lists of three items

Boat	Pencil	Table	Shoe
Horse	Fish	House	Phone
Whistle	Banana	Penny	Picture

Three Serial Subtractions

13 from:

100	200	250
87	187	237
74	174	224
61	161	211

17 from:

100	200	250
83	183	233
66	166	216
49	149	199

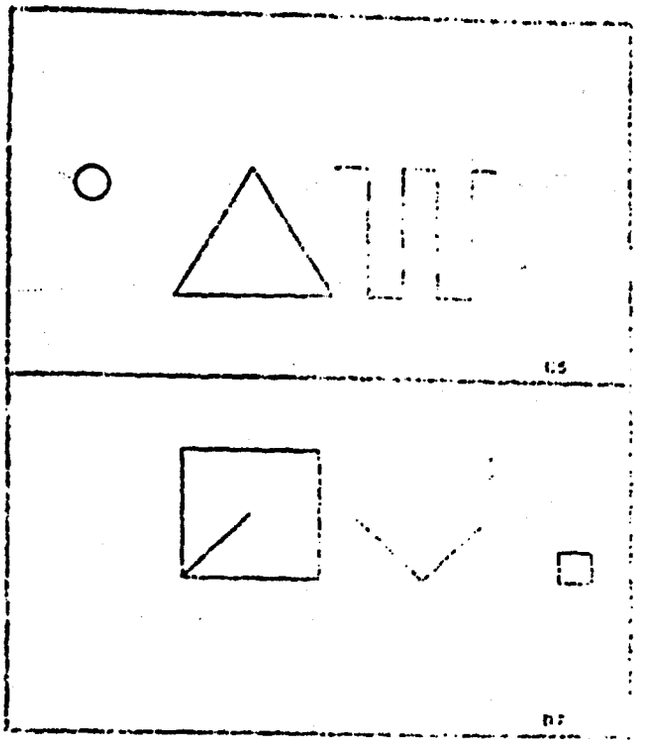
19 from:

100	200	250
81	181	231
62	162	212
43	143	193

Series of 6-digit

6-1-9-4-7-3
3-9-2-4-8-7
5-3-9-4-1-8
7-2-4-8-5-6

FIGURE 1



Two representative items of the Benton Visual Retention Test

Source: Benton AL: The Revised Visual Retention Test (ed 4). New York, The Psychological Corporation, 1974

Best Available Copy

DEPARTMENT OF TRANSPORTATION

FEDERAL AVIATION ADMINISTRATION

SPECIFICATIONS FOR PSYCHIATRIC AND PSYCHOLOGICAL EVALUATION

The Office of Aviation Medicine of the Federal Aviation Administration develops and enforces medical regulations establishing the criteria for medical certification of more than 800,000 pilots and air traffic controllers. Portions of the medical regulations relate to mental disorders that may represent a significant threat to flight safety. Certain conditions, such as psychotic disorders, panic disorders and some personality disorders, are specifically mentioned in the regulations as causes for denial of certification. In addition, the Federal Air Surgeon is required to deny certification to a person with any other mental condition that makes the applicant unable to perform the duties safely or exercise the privileges of the airman certificate that he or she holds or for which he or she is applying. The determination is to be based upon the case history and appropriately qualified medical judgment relating to the condition involved.

The regulations also provide for the special issuance of a medical certificate to an individual who has a mental disorder or a history of a mental disorder that would normally be disqualifying, if he or she can show to the satisfaction of the Federal Air Surgeon that he or she can perform safely the duties authorized by the class of medical certificate for which he or she is applying. Psychiatric and psychological evaluations are among the information considered by the Federal Air Surgeon.

It is obvious that evaluation for FAA medical certification is conducted outside the more customary confidential atmosphere of a therapeutic relationship. The examination is conducted in an adversary manner, and the applicant is required to participate in the evaluation process. Therefore, many applicants might think it to be in their best interest to present biased pictures of their mental health status, if not to

attempt to withhold information and distort previous histories. If the psychiatrist and psychologist feel uncomfortable with the necessarily probing nature of the examination and potentially adverse effect of their reports on these applicants, they are free to refuse to engage in such evaluations. However, those who accept the responsibility of performing the evaluations may feel they are undertaking a task that serves the public's interest and safety, even if their services are being sought by the applicants.

The report should deal specifically with all past psychiatric history and any significant past behavior. This history should be supplemented by a review of available medical records, which have been obtained by the Federal Aviation Administration and may be requested by the evaluator. Apparent inconsistencies between the known history and the applicant's statements, or overt contraindictions within the interview itself, should be probed in depth. Statements about the applicant's apparent reliability, rather than acceptance of the applicant's unverified statements, are helpful.

The evaluator's report should include consideration of the following: 1) any current or recent psychiatric symptoms, aberrant behavior, or psychiatric or other medical findings; 2) a review of the applicant's past and current adjustment in occupational, marital and social and other aspects of his life; 3) the use or abuse of any pharmacologically active agent, either for therapeutic or recreational purposes; 4) any personality traits or other recognized factors that might affect the risk of future recurrence of the problem, or the risk of other adverse events; 5) the current psychiatric and psychological status and stability of the applicant.

In the evaluation of an applicant with a history of alcohol or other substance abuse or dependence, the factors to be considered would include: 1) the period of the applicant's abstention from alcohol or other substances of abuse; 2) severity of the problem and its duration; 3) number of times treatment was sought and that relapses occurred; 4) an outline of the most recent treatment; 5) presence of residual medical complications; 6) progress in marital, social, vocational, educational and other spheres,

since rehabilitation began; 7) commitment to rehabilitation by virtue of continuing contacts with social or professional agencies; 8) any other personality difficulties that would be disqualifying or that would adversely affect sustained abstinence.

While the examiner's diagnostic statements and opinions are desirable, the decisions about medical certification are made by the Federal Air Surgeon with the assistance of FAA psychiatrists and professionals who are not only aware of the particular disorders but also familiar with the demands of the aviation environment and the levels of flying responsibilities. Therefore the examiner's recommendations about certification are neither binding nor necessary.

The evaluation must be conducted by psychiatrists and psychologists who generally have the following qualifications: A qualified psychiatrist is one who has been certified by the American Board of Psychiatry and Neurology, or one who has the background equivalent to board certification. A qualified clinical psychologist is one who has earned a PhD from a clinical training program approved by the American Psychological Association (APA), is licensed in the state in which he or she practices, and holds diplomate status from the American Board of Professional Psychology. A qualified neuropsychologist should have earned a PhD and have trained in a specialty area of neuropsychology, neurobehavior or clinical neuropsychology. He or she also should be a graduate from an APA-approved clinical training program, be licensed in the state in which he or she is practicing and be a diplomate of the American Board of Clinical Neuropsychology.

The evaluator's report, including a copy of the test protocol, should contain a detailed psychological evaluation, based on an appropriate battery of tests that are designed to answer specific questions; these might include the Rorschach, the complete Wechsler Adult Intelligence Scale (WAIS), or the revised Wechsler Adult Intelligence Scale (WAIS-r). In addition, any of the following tests could be administered, depending upon the nature of the referral:

a. Thematic Apperception Test (TAT)

b. Sentence Completion Test

c. Minnesota Multiphasic Personality Inventory (MMPI)

d. Bender Gestalt Visual Motor Test

e. Graham-Kendall Memory for Designs Test

f. Wechsler Memory Scale, form 1

g. Halstead Neuropsychological Battery

h. Halstead-Reitan Neuropsychological Battery

Hematology/Oncology

Introduction

General Hematologic Conditions

The ability of blood to transport oxygen effectively to tissues is dependent upon an adequate hemoglobin concentration and on the ability of blood to perfuse organs. Abnormalities of this function are important in aviation safety for two reasons: (1) they affect the transport of oxygen from the lung to tissues, and (2) they affect blood rheology such that debilitating or incapacitating organ infarcts can occur.

Hemoglobin is only one link in the oxygen transport chain. The presence of anemia, polycythemia, or abnormal hemoglobin must be placed in the context of the other links. These include pulmonary function, cardiac output and its regulation, and end-organ oxygen utilization. For example, anemic states can be compensated by increased cardiac output in normal persons, but the limit of the compensation will vary, depending on the presence or absence of heart disease and lung disease. Thus, it is impossible to establish rigid criteria for acceptable hemoglobin concentrations in the context of such interacting conditions. As a result, it is difficult to state exact criteria for denials and exemptions for medical certification of individuals with abnormalities of the hematologic system. Instead, general guidelines are provided about potentially dangerous conditions that need further evaluation:

1. The hematocrit must not fall outside the range stated in the standard if a certificate is to be issued without further evaluation. The etiology of any abnormality that leads to hematocrit values outside of this range must be established and well documented.
2. Some hematocrits that fall within the acceptable range, but that are still low, should be evaluated further before a certificate is issued.

3. When abnormalities or diseases of other organ systems lead to hematological abnormalities, the standards and guidelines established for those conditions must be met, as well as those established for the hematological abnormalities.

4. When an applicant's medical history suggests a previous or present abnormality of red blood cells, white blood cells or blood platelets, the AME must defer certification to the FAA for further evaluation.

Specific Hematologic Conditions

o Anemia

1. General considerations

Decreased hemoglobin concentration decreases the amount of oxygen that can be carried in the blood. Various classification schemes for understanding the physiology of disorders of anemia have been proposed. The simplest and easiest to understand is based upon the principle that anemia results from decreased red blood cell production, accelerated red blood cell destruction, or red blood cell loss. These processes may occur together or separately.

The body can compensate for anemia as long as the hemoglobin concentration is at least 10 g/dl or the hematocrit is at least 30%. However, in the presence of heart disease, lung disease or peripheral vascular disease, the ability to compensate is somewhat less. Furthermore, the decreased oxygen tension that occurs at altitudes thousands of feet above sea level that are commonly reached in general aviation also affects the body's ability to compensate. Thus,

we recommend a hematocrit of 32% as the absolute minimum for certification, and that hematocrits of less than 38% for males and 36% for females should be evaluated for etiology.

2. Iron deficiency

It is imperative that the cause of iron deficiency anemia be determined, particularly in males, because of its association with intestinal malignancies and peptic ulcer disease. If the cause is identified and is not disqualifying, and treatment is undertaken, a certificate may be issued when the hematocrit is greater than 32%.

3. Vitamin B_{12} and folic acid deficiency

The etiology must be established, and the candidate must be examined by a neurologist to exclude impairment that might interfere with airman's duties. After restoration of body stores of vitamin B_{12} or folic acid, a certificate may be issued. Class III certificate holders should be re-examined annually. For individuals found to have a lifelong requirement for vitamin B_{12} replacement, reports from their physicians showing strict compliance with treatment programs should be required. Such treatment must include the parenteral administration of 1000 micrograms of vitamin B_{12} by injection at least once each month. Persons with a lifelong requirement for vitamin B_{12} often are noncompliant and signs and symptoms of pernicious anemia may recur.

4. Anemia of chronic disease

The underlying disease must be identified and evaluated because it is

a potential reason for a denial of certification. If the hematocrit standard is satisfied, a certificate may be issued.

5. Sideroblastic anemia

Only carriers of the familial type should be certified, and then only if the hematocrit guidelines are satisfied. Sideroblastic anemias that result from alcoholism, malignancies, or drugs should be evaluated under the recommendations for those specific problems. The entire spectrum of "acquired sideroblastic anemias," "myelodysplastic syndromes" and "chronic refractory anemias," is automatically and permanently disqualifying due to the known propensity for acute leukemic transformation that often occurs suddenly and without warning. Frequently, an elevated mean corpuscular volume (MCV) may be noted in low grade anemias, and such individuals will have normal folic acid and vitamin B_{12} blood levels.

6. Alcohol dependence or abuse

Anemia due to alcohol dependence or abuse should be considered under the hematocrit standard, and the alcohol dependence or abuse should be evaluated under the recommendations for those disorders. An MCV above 100 cubic microns may be the earliest sign of alcohol dependence or abuse.

7. Hemolysis

The etiology of hemolytic anemia must be established. If the hemolytic anemia is acquired, the underlying conditions must be evaluated and treated. Congenital hemolytic anemia that is not due

to a hemoglobinopathy, and that has not caused the hematocrit to fall below 32%, is not a cause for denial.

A person with hereditary spherocytic anemia should not be denied certification if a splenectomy has been performed. Glucose-6-phosphate dehydrogenase (G6PD) deficiency is not a cause for denial, unless there is a history of severe hemolysis in the presence of oxidant drugs that results in a hematocrit of less than 32%. Microangiopathic hemolytic anemia should be evaluated according to the guidelines established for the conditions causing it, such as the malignancies. In individuals with chronic autoimmune hemolytic anemia, denial of certification should be automatic and permanent because of the unpredictable nature of decompensation, often with devastating effect.

8. Other conditions

Other conditions that are either rare or of obscure etiology should be evaluated on an individual basis. These include paroxysmal nocturnal hemoglobinuria, porphyria, and other rare causes of reduced hemoglobin concentration, red cell synthesis, or red cell destruction.

o Polycythemia

1. General considerations

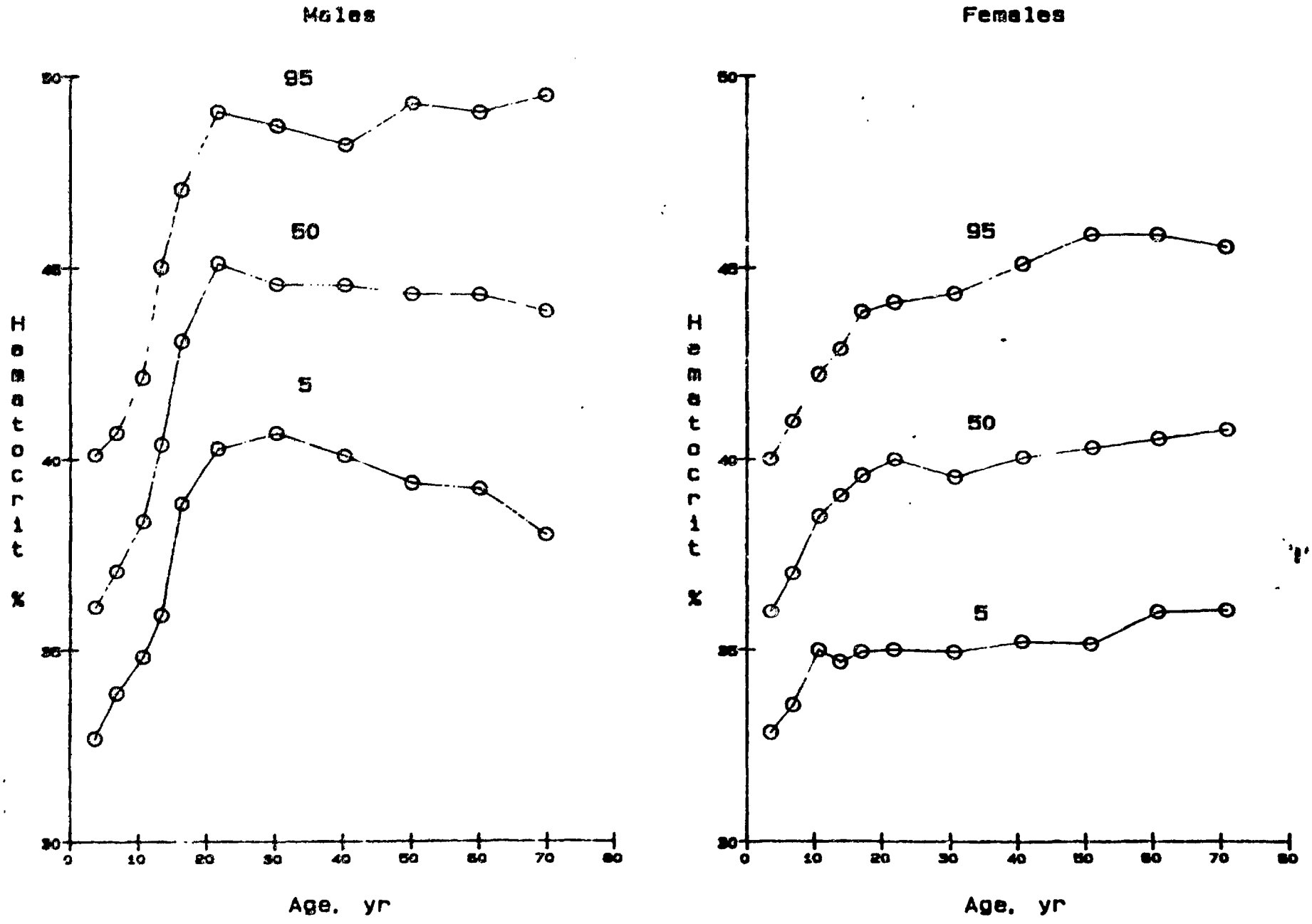
Increased hemoglobin concentration increases the viscosity of the blood and, therefore, the resistance to flow of the blood. When the hematocrit is abnormally high, decreased cerebral blood flow is particularly dangerous, and the risk of cerebrovascular accident is

elevated. Regardless of the cause of the polycythemia, there is good evidence that cerebral blood flow and some mental functions decrease continuously and in linear fashion as the hematocrit increases. There is no absolute elevated hematocrit value below which these findings do not occur. Conversely, the evidence is strong that treatment to reduce the hematocrit is associated with a linear increase in cerebral blood flow and improvement in intellectual function. These data refer to people at sea level under normal ambient oxygen tension and having no associated cerebrovascular disease.[1-4]

There is a known tendency for the hematocrit to increase in normal individuals who reside at altitudes above sea level. However, sufficient data are not available to establish criteria for normality. According to the National Center for Health Statistics, the 95th percentile of hematocrit for nonsmoking males over 20 years of age at sea level does not exceed 50% at any age (See Figure). For females the comparable hematocrit does not exceed 47%. Although normal hematocrits will be somewhat higher at elevations above sea level, very few normal persons will have hematocrits above 55%. Thus, any male or female who has a hematocrit above 55% should be denied certification pending further investigation to establish the etiology of the polycythemia. If treatment has been undertaken, the underlying disorder is itself not disqualifying, and if the hematocrit is less than 55%, a certificate may be issued. For holders of all classes of certificates, yearly re-evaluation and certification of compliance with the treatment and follow-up regimen should be required.

FIGURE

Fifth, Fiftieth and Ninety-fifth Percentiles of Hematocrit in Non-smoking



Source: National Health Survey, DHHS Publication No. (PHS) 83-1682, National Center for Health Statistics, 198

2. "Spurious" (stress) polycythemia

Applicants in whom an elevated hematocrit is due to contracted plasma volume may be issued certificates if the absence of true polycythemia has been demonstrated by a red cell volume determination, and if the hematocrit is no higher than 55%. A radioisotope red blood cell volume determination (red cell mass) should be used in the evaluation of persons with an elevated venous hematocrit. Some hematologists also suggest the simultaneous use of red blood cell mass and plasma volume to determine whether there is a true increase in red blood cell mass or only an artifact of increased hematocrit caused by alterations in plasma volume or maldistribution of plasma.

3. Cigarette smoking

Polycythemia associated with cigarette smoking should be documented by measurement of carboxyhemoglobin concentration. The hematocrit should be no more than 55% for issuance of a medical certificate.

4. Polycythemia due to hypoxic lung disease

For polycythemia due to hypoxic lung disease, the guidelines for the underlying lung abnormality must be followed for issuance of a certificate; also, the hematocrit should be no greater than 55%.

Note: Persons whose polycythemia is due to stress, smoking or lung disease and who are being treated to lower the hematocrit to

acceptable levels, may be issued a certificate under special issuance procedures. At each recertification a letter from the responsible treating physician certifying compliance with the treatment program and follow-up regimen should be required. Follow-up should be at 6-month intervals for all classes of certification.

5. Polycythemia vera

Persons in whom a diagnosis of polycythemia vera has been made should be disqualified permanently. Treatment may lower the hematocrit to acceptable levels, but the risk of thromboembolic complications or of rapid unpredictable progression makes certification not acceptable.

6. Erythropoietin-secreting tumors

Renal, hepatic, pulmonary, and cerebellar tumors are known to secrete erythropoietin in some instances, and lead to polycythemia. In these rare cases, the guidelines for the underlying malignancy should be followed.

7. High-affinity hemoglobins - See the following section on hemoglobinopathies.

o Hemoglobinopathies and thalassemias

1. General considerations

The hemoglobinopathies and thalassemias are heterogeneous sets of disorders of hemoglobin structure and synthesis. Over 300 mutant forms of hemoglobin are known, but only a few have clinically

important consequences. The most severe and controversial are the sickling disorders. These are particularly important in aviation medicine, because of the unique property of sickle hemoglobin to form aggregates at reduced oxygen tension. Applicants who may be at increased risk should be questioned about sickle cell anemia and related disorders in himself or herself and in the immediate family.

If laboratory testing is carried out, it is imperative that procedures be used that can differentiate among the various sickling syndromes, such as SS, AS, and SA, S/B+ and S/Bo thalassemias, and double heterozygous conditions in which hemoglobin S interacts with other abnormal hemoglobins. Procedures include electrophoresis in two systems and either a solubility test or a direct visualization of sickled cells in conditions of oxygen deprivation ("the sickle prep"). The FAA might seek the assistance of other governmental agencies, such as the Centers for Disease Control or the National Institutes of Health, in determining which tests would be sufficient for accurate differentiation and which reference laboratories could be certified to perform such tests for the FAA.

2. Sickling disorders

All subjects with documented sickle cell disease (hemoglobin SS) should be denied medical certification.

3. Hemoglobin SC disease

These persons should also be denied certification because of the high incidence of retinal hemorrhages and splenic infarction.

4. S/B thalassemia

Persons with either S/B+ or S/Bo thalassemia should be denied medical certification.

5. Hemoglobin AS (sickle cell trait)

These individuals should be certified unless other conditions disqualify them. Splenic infarction can occur at altitudes in the 8,000 ft to 10,000 ft range, and persons known to have sickle cell trait should be advised to pay strict attention to the use of supplementary oxygen. Also, they should be informed about the risks and symptoms of splenic infarctions.

6. Other double heterozygous conditions involving hemoglobin S

No generalizations can be made. If the presence of hemoglobin S and another hemoglobin is known, the subject should be carefully evaluated to determine the exact diagnosis. In general, if the candidate has no symptoms or history of vaso-occlusive disease, and if the hematocrit is within the acceptable range, then a medical certificate may be issued.

7. Polycythemia due to high affinity hemoglobins

These disorders are very rare, and little is known about their natural history. Although stroke and other vaso-occlusive problems have not been demonstrated convincingly, hematocrits over 55% carry the same risks as discussed for other polycythemias. Certification of these persons should be denied permanently.

8. Unstable hemoglobins

Unstable hemoglobins that do not result in anemia and whose carriers do not have splenomegaly should not lead to certification restrictions. In those cases in which splenomegaly exists or existed before splenectomy, certification should be denied.

9. Thalassemias

Simple, uncomplicated beta thalassemia trait should not be a basis for denial of certification. Likewise, a thalassemia trait in which the only manifestation is microcytosis, in the absence of hemoglobin H, should not be denied. If a candidate with hemoglobin H meets the criterion for hematocrit, he or she should also be certified. Persons with homozygous beta thalassemia and thalassemia intermedia, in whom there is biochemical evidence of beta thalassemia trait, splenomegaly or mild hemolysis, should not be certified.

10. Other hemoglobinopathies

Other rare hemoglobinopathies cause cyanosis or methemoglobinemia. Carriers of these abnormalities are almost always aware of their conditions and will not be likely to seek certification. If they do, it should be denied.

o Coagulation and thrombotic disorders

1. General considerations

Applicants with inherited disorders of coagulation should be disqualified if there is any history of factor replacement or serious bleeding episodes.

2. Hemophilia

Applicants with Factor VIII deficiency should be denied certification. Applicants with von Willebrand's disease should be denied certification if there is history of factor replacement or serious bleeding episodes. Applicants with other specific factor deficiencies should follow the same criteria.

3. Iatrogenic thrombosis

These cases are local thrombotic episodes associated with intravenous needles or catheters. After anticoagulant therapy has been discontinued, the applicant need not be disqualified.

4. Deep vein thrombosis

A history of deep venous thrombosis should be disqualifying for a period up to one year from the episode, and for 6 months after all anticoagulant therapy has been discontinued. Any underlying contributing factors, such as malignancies, must be evaluated according to the criteria for those conditions.

5. Pulmonary embolism

A history of a single episode of pulmonary embolization, not associated with chronic deep venous thrombosis, should be considered disqualifying from the date of the embolization and for at least 6 months after all anticoagulant therapy has been discontinued.

6. Multiple pulmonary emboli

If an applicant has had more than one episode of pulmonary

embolization documented by radio-isotopic or angiographic methods, no certificate should be issued. Even if the candidate is currently not taking anticoagulant therapy and meets the criteria in paragraphs 4 and 5 above, the occurrence of more than one discrete episode of pulmonary embolization should be permanently disqualifying.

7. Recurrent arterial emboli - Disqualifying under any circumstances.

8. Anticoagulant medication

Regardless of dose, route, or reason for administration, the use of anticoagulant drugs of the heparin class or coumadin/warfarin class should be disqualifying while they are in use and for 6 months after they are discontinued. The use of aspirin alone for its prophylactic, antiplatelet effect should not be considered disqualifying unless the underlying condition itself is disqualifying. Antiplatelet agents of the dypyridamole or sulphinpyrazone classes, or yet-to-be marketed antiplatelet agents, also should be considered nondisqualifying while being used either by themselves or in combination with aspirin. The AME and FAA should inquire carefully about the conditions for which medications are being taken, and letters from the responsible treating physicians concerning diagnosis and treatment should be obtained.

9. Hemorrhagic platelet abnormalities

A decreased circulating platelet count due to any cause may result in debilitating hemorrhagic episodes. Hemorrhage can also occur when platelet counts are normal but platelet function is abnormal. If platelet abnormalities are suggested by history, the AME should defer

the application, pending a more thorough investigation by the FAA.

Quantitative platelet disorders in which the platelet count is below 75,000/mm^3 should be disqualifying permanently. These disorders include the hypoproliferative disorders, disorders of ineffective thrombopoiesis, disorders of abnormal platelet distribution and disorders of abnormal platelet destruction. An applicant with idiopathic thrombocytopenic purpura (ITP), who has previously been treated by splenectomy and who has had stable platelet counts for 6 months after therapy has been discontinued, may be considered for special issuance. Platelet counts should be repeated at 6-month intervals. Applicants who have had thrombocytopenia due to abnormal destruction or consumption, as with disseminated intravascular coagulation (DIC), vasculitis, or thrombotic thrombocytopenic pupurpa (TTP), should be denied certification permanently.

Persons with thrombocytosis greater than 750,000/mm^3 should be disqualified. Some temporary episodes of thrombocytosis can occur in persons with underlying iron deficiency anemia or other temporary disorders such as recovery from alcoholic bone marrow suppression.

If there is temporary, secondary thrombocytosis that has resolved and platelet counts have been consistently normal, special issuance may be considered. Patients with "essential" thrombocytosis without apparent explanation, who continue to have platelet counts above 750,000/mm^3, should be permanently and automatically disqualified.

Hematologic Neoplasia

Since hematologic neoplasias are serious systemic disorders and often are associated with chronic and sudden incapacitation, certification of an applicant with a hematologic neoplasm should be denied by the AME, and decisions regarding fitness for flying should be made by the FAA with appropriate consultation from specialists. Individuals with histories of hematologic neoplasia not requiring continuous therapy may be certified to fly; however, adequate follow-up and reassessment is necessary because of the risk of relapse and/or progression.

Individuals receiving cytotoxic chemotherapy should be declared unfit to perform as pilots, because the use of cytotoxic chemotherapy implies a serious, progressive disorder. Also, cancer chemotherapy itself can cause anemia, thrombocytopenia, and granulocytopenia, all of which could precipitate an acute incapacitating event. Thus, the risk of sudden incapacitation in an individual taking cancer chemotherapy is too great to permit certification.

Similarly, the use of glucocorticosteroids as therapeutic agents in hematologic disorders should preclude certification. Pharmacologic doses of corticosteroids often lead to incapacitation, such as toxic effects on the cerebrum, which can cause sudden and unexpected alterations in mood and mentation. Also, when corticosteroids are used to suppress lymphoid malignancies and to control autoimmune thrombocytopenia and anemia, there can be sudden loss of control and acute anemia or thrombocytopenia with bleeding can occur.

- Leukemias

  1. Acute lymphocytic leukemia

     A person with the diagnosis of acute lymphocytic leukemia (ALL) as an adult should not be certified. Although treatment is effective, the

relapse rate is high and long-term survival is presently uncommon. A history of ALL during childhood should not preclude certification, but the applicant should be deferred to the FAA for a thorough evaluation by a hematologist or oncologist. Approximately 50% of children with ALL are cured of their disease. To be certified an individual must have had ALL treated only as a child and be in complete remission without any treatment for at least 10 years.

Prior to initial certification, a careful and detailed neurological evaluation by a qualified neurologist should be done, as well as a psychiatric or psychological evaluation, especially in applicants who have had cranial irradiation. The individual should be re-examined every 6 months because of the risk of a second malignancy and relapse. If the individual has had cranial irradiation, particular attention should be paid to re-examination of the neurologic system and mental status, because the late effects of cranial irradiation can affect mentation and performance.

2. Acute myelogenous leukemia

Acute myelogenous leukemia (AML) or acute nonlymphocytic leukemia is a very serious disorder; long-term survival is uncommon. Treatment is effective, yet the relapse rate is high and remissions last only about 15 months on the average. Thus, a diagnosis of AML should result in permanent denial of certification.

3. Preleukemia or myelodysplastic syndromes

The preleukemic or myelodysplastic syndromes are a group of

hematopoietic disorders that frequently evolve to acute myelogenous leukemia. They are characterized by hypercellular bone marrow and various degrees of peripheral blood cytopenias. Persons with these conditions are prone to infection and bleeding. Because of the relatively poor prognosis and high risk of sudden incapacitation, individuals with these disorders should not be certified (see also the discussion of sideroblastic anemia).

4. Chronic myelogenous leukemia and myeloproliferative syndromes

An applicant with a confirmed diagnosis of either Ph chromosome-positive or -negative chronic myelogenous leukemia (CML) should be denied certification permanently. While an individual may function normally during the chronic phase, the development of a "blast crisis" is unpredictable and may be sudden. Also, most individuals with chronic myelogenous leukemia in the chronic phase require cytotoxic chemotherapy. Persons with related myeloproliferative syndromes, such as essential thrombocythemia and polycythemia rubra vera, also should be disqualified. In these latter two conditions, the risk of thrombosis, bleeding, or sudden neurologic deficit is prohibitively high.

5. Chronic lymphocytic leukemia

A common staging system for chronic lymphoctyic leukemia (CLL) is as follows:

Stage 0 - bone marrow and blood lymphocytosis only

Stage I - lymphocytosis with enlarged nodes

Stage II - lymphocytosis with enlarged spleen or liver, or both

Stage III - lymphocytosis with anemia

Stage IV - lymphocytosis with thrombocytopenia

Individuals with disease in Stages II through IV should not be certified. In these stages of the disease cytotoxic therapy is often necessary and the cytopenias present a serious risk of sudden incapacitation. Persons with Stage 0 and Stage I disease may be certified, providing there is no hemolytic anemia and no requirement for chemotherapy or corticosteroids. Re-examination at intervals of three months should be required, with documentation by the treating physician of physical findings, laboratory status, and compliance with the follow-up program. T- or B-cell CLL should be treated in the same way. Because the individuals may progress from one stage to another unpredictably, frequent reassessment will be necessary to determine the stability of the disease.

7. Hairy cell leukemia

Hairy cell leukemia may be clinically benign, and persons with this disorder may live for decades with no impairment. Other forms of the disease, however, are more aggressive. Individuals who are stable after splenectomy, or without treatment,[5] or with chronic low-dose interferon treatment, and who have normal hemoglobin and platelet counts and more than 750 granulocytes/mm^3 may be fit to fly. They should be re-examined every three months and recertified at 6-month intervals.

Airmen requiring treatment for progressive disease should not be certified. These individuals are at risk of developing sudden cytopenias and opportunistic infections that may lead to acute incapacitation. Thus, the need for therapy other than splenectomy or interferon should preclude certification. Such therapy includes, but is not limited to lithium, androgenic steroids, leukapheresis, and cytotoxic drugs. Because an individual's condition may deteriorate unpredictably, frequent reassessment will be necessary; and at each recertification a statement from the responsible physician should be required that the patient is compliant with treatment and the follow-up program.

- Lymphomas

  1. Hodgkin's disease

     Hodgkin's disease is a serious but often curable lymphoid neoplasm. Because of the risk of sudden incapacitation, individuals with active Hodgkin's disease or individuals undergoing therapy for Hodgkin's disease should not be certified. Since cure rates in stages I and II-A are as high as 95%, persons with these stages who have had no evidence of disease for two years after completion of treatment are certifiable. Persons with disease in Stages II-B through IV-B are also curable, although their prognosis may be poor. About 60% to 75% of persons with disease in Stages III-B, IV-A and IV-B that is treated with standard chemotherapy will achieve complete remission. The median duration of remission is in excess of three and one-half years. Also, at least half of those persons who respond completely to treatment will be cured. Persons with bone marrow involvement and have a poor

prognosis and are rarely cured.

Thus, persons with disease in Stages II-B through IV-B should be free of disease after completion of therapy for at least five years before consideration for certification, and they should be re-evaluated every 6 months for 10 years. There are numerous long-term complications of treatment for Hodgkin's disease, including the development of acute leukemia and second malignancies of other types, radiation-related heart disease, pulmonary fibrosis, and hypothyroidism. Frequent re-evaluations, even in applicants with long intervals of disease-free survival, are required. After 10 years there should be annual appraisals.

2. Non-Hodgkin's lymphoma

Persons with well-differentiated and poorly-differentiated lymphocytic lymphoma, mixed lymphocytic lymphoma, and histiocytic lymphoma of either the nodular or diffuse type, are usually not curable, and these individuals should be disqualified permanently from certification. Persons with B-cell, diffuse histiocytic lymphoma, particularly in the early stages, may be cured by radiation therapy and/or chemotherapy and, if they are free of disease without therapy for at least three years, they may be certified with re-evaluation to occur every three months for three years and then every 6 months. Persons with T-cell, diffuse histiocytic lymphoma, including immunoblastic lymphoma and T-cell lymphoblastic sarcoma, should not be certified because of the high degree of malignancy of these disorders and their unpredictability. Similarly, persons with Burkitt's

lymphoma should not be certified.

3. Mycosis fungoides and Sezary syndrome

Mycosis fungoides and Sezary syndrome are chronic neoplastic disorders involving mature helper T-cells. The disorders are not curable, although in early stages the disease may cause little impairment. A TNM staging system has been proposed for mycosis fungoides (Bunn PA Jr, Lamberg SI: Report of the committee on staging and classification of cutaneous T-cell lymphomas. Cancer Treatment Report 1979;63:725-728). Individuals with visceral disease of N_3 or T_3 stages or greater should not be certified. Persons at lower stages requiring only local therapy for skin disease, in whom there is no systemic impairment, may be certified but should have careful follow-up. Persons with a compromised hematopoietic system or requiring cytotoxic therapy are not certifiable.

o Plasma-cell dyscrasias

Persons with multiple myeloma, Waldenstrom's macroglobulinemia or multiple plasmacytomas should not be certified. These disorders are not curable, require frequent therapy that is toxic, and are associated with side effects such as neurologic impairment that may lead to sudden incapacitation. Persons with a single plasmacytoma may be cured and, if they are free of disease more than three years after therapy has been discontinued, they may be considered for certification with frequent follow-up. Persons with benign monoclonal gammopathy with a monoclonal spike comprising less than 2 g/dl of protein, with fewer than 5% plasma cells in the bone marrow, and with no hematopoietic compromise or osteolytic

lesions, may be certified if they have no evidence of progression of the disease for three years; they should be recertified every 6 months. The major risks of monoclonal gammopathy are progression to multiple myeloma and an increase in serum viscosity leading to neurologic impairment.

Persons with amyloidosis associated with plasma cell dyscrasia should not be certified because of the high incidence of organ infiltration and the risk of sudden impairment. Persons with gamma or alpha heavy chain disease should not be certified; the median survival is approximately 12 months for gamma heavy chain disease, and the alpha chain disease is often associated with abdominal lymphoma, which is a progressive and fatal disorder. Mu heavy chain disease is rare and may be associated with chronic lymphocytic leukemia. Those individuals whose disorder resembles lymphoma or multiple myeloma with amyloidosis should not be certified. Persons with cold agglutinin disease should not be certified because of the risk of sudden hemolysis. Persons with cryoglobulinemia associated with myeloma and persons with the mixed cryoglobulinemia syndrome should not be certified because of the risk of sudden vascular incidents and neurologic dysfunction.

- Immunodeficiency syndromes

The criteria for the diagnosis of acquired immunodeficiency syndrome (AIDS) include:

1. immune deficiency with no specific known cause;
2. a history of opportunistic infections;
3. Kaposi's sarcoma;
4. age under 60 years.

Criteria for the diagnosis of lymphadenopathy syndrome include:

1. enlarged lymph nodes (excluding inguinal) with no apparent cause;
2. being in a high risk group for AIDS;
3. adenopathy present for more than 6 months.

Since 1982, when these criteria were established, the human T-cell lymphotropic virus type III/lymphadenopathy-associated virus (HTLV III/LAV) has been recognized as the cause of AIDS. Thus, in June 1985 the Centers for Disease Control expanded the definition of AIDS to include a positive serologic or virologic test for HTLV III/LAV in conjunction with other serious diseases such as disseminated histoplasmosis, isosporiasis, bronchial or pulmonary candidiasis, non-Hodgkin's lymphoma of high-grade pathologic type, and confirmed Kaposi's sarcoma in person's who are 60 years of age or older. The CDC also recognizes that there are other, milder forms of disease caused by HTLV III/LAV that cannot be classified as AIDS such as AIDs Related Complex (ARC). Individuals may also exhibit serologic evidence of exposure to HTLV III/LAV and have no symptoms; these persons may be infective for years. (CDC: Revision of the case definition of acquired immunodeficiency syndrome for national reporting-United States. MMWR 1982;34:373-375).

Clearly our understanding of AIDS, lymphadenopathy syndrome and other manifestations of HTLV III/LAV infection grows and changes quite rapidly. At present we recommend that persons with the AIDS should not be certified because of the high risk of opportunistic infections, which can appear suddenly and cause acute incapacitation. Persons with the lymphadenopathy syndrome without evidence of previous opportunistic infection may be

certified with follow-up every 6 months. Persons who have undergone bone marrow transplantation should not be certified because of the risk of sudden opportunistic infections or late effects of radiation therapy. The FAA should follow closely the developments in AIDS research and review these recommendations accordingly.

Persons with common variable immunodeficiency who do not have bronchiectasis and who are controlled with regular gamma globulin therapy may be certified, but they should be re-evaluated every 6 months.

- Generalized lymphadenopathy and splenomegaly

  There are a variety of disorders characterized by generalized lymphadenopathy and/or splenomegaly, some of which are not associated with abnormalities of leukocytes or immunoglobulins. Various non-neoplastic disorders, infections, inherited immune deficiency diseases, acquired immune deficiency diseases and other generalized reactive processes may be associated with splenomegaly or lymphadenopathy. An individual with these physical findings warrants deferral for careful evaluation to rule out disqualifying conditions.

Nonhematologic Malignancies

In the interest of aviation safety malignant disorders must be considered individually according to the predictability (or lack thereof) of incapacitating events associated with the disease or its treatment. Basic to the understanding of the natural history of malignant disorders is proper classification and staging. The Manual for Staging of Cancer, Second Edition, American Joint Commission on Cancer (AJCC),[6] states: "A classification scheme for cancer must encompass all attributes of the tumor

that define its life history. The AJCC classification is based on the premise that cancers of similar histology or site of origin share similar patterns of growth and extension." For the majority of nonhematologic malignancies, the concept of a cancer's origination in a particular location, with extension locally by direct extension, regionally via the lymphatics, and distantly by blood and lymphatics provides a workable framework upon which a simple classification scheme may be constructed. This scheme can provide a framework for gathering and assembling data about an applicant with a history of malignancy. The following is a summary of the AJCC staging scheme.

With most tumors the untreated primary cancer or tumor is first classified according to its size, which is designated by the capital letter "T" with various subscripts.

The presence or absence of regional lymph node involvement is indicated by the use of a capital "N" with subscripts. Distant metastasis is indicated by a capital letter "M" with subscript and description of involved organs. For example, a small 1.5 cm breast cancer nodule would be classified as T_1. If it is fixed to underlying pectoral fascia or muscle, it is classified as T_1B. If no regional lymph nodes can be palpated and no nodes seem to be suspicious for tumor, the "N" classification would be N_0. If, however, on chest radiograph pulmonary nodules indicative of metastatic disease were seen, the "M" classification would be M_1 (PUL), for pulmonary. If the metastatic evaluation were negative, the "M" classification would be M_0. The clinical stage of the tumor for the $T_1B\ N_0\ M_0$ breast tumor would be Stage I. For the $T_1B\ N_0\ M_1$ tumor with the presence of metastatic disease in the lung, the clinical stage would be Stage IV.

For many tumors the histologic grades of well-differentiated, poorly- and very-poorly differentiated provide some idea as to virulence and potential for relaspe, and have a bearing on expected clinical course. Cancer staging schemes may be based upon clinical measurements, in which case the TNM classification is preceded by the small letter "c." The breast cancer example above would be Stage I clinically, or $cT_1B\ N_0\ M_0$.

After surgical treatment a pathological staging system can be used. In addition, there is a surgical-evaluative staging system, a re-treatment staging system, and an autopsy staging system.

The natural biological history of cancer is based upon the theory that at some point in the person's life an event occurred that created a malignancy and the cancer process began. Generally, there is a silent or latent period during which tumor growth occurs and tumor burden and tumor size increase. Often, prior to the period of clinical manifestation, primary lymph node involvement occurs, invasion of blood vessels and lymphatics begins, and microscopic dissemination occurs. As the period of clinical manifestation proceeds, tumor burden can increase, and distant metastases manifest themselves.

With early cancer detection the malignant process can be ascertained by physical examination or diagnostic study early in the period of clinical manifestation. When a malignant process appears to be localized according to the TNM clinical staging process, the majority of cancer treatments are directed toward "local control," which, depending upon tumor size, location, and histologic type, may consist of surgical treatment, radiation treatment, or a combination of the two. With certain localized tumor types or more advanced size, chemotherapeutic agents may be used preoperatively or postoperatively in combination with radiation.

In addition to "local control," some persons with malignant disease that has a high likelihood of local or distant relapse may be given adjunctive radiation or chemotherapy. This is commonly done for persons with Stage II or higher carcinomas of the breast.

Therefore, the assessment of the impact that a malignant process may have on a pilot applicant requires not only consideration of the organ of origin and the clinical or surgical stage, but also the treatments that are being or have been used to accomplish the "local control."

Most surgical procedures for cancer should disqualify an applicant for aviation flight duties for a variable time, depending upon the extent of surgery and the site of operation. Individuals should not be considered qualified for piloting of any type for at least one to two months following any craniotomy, thoracotomy, or opening of the abdominal cavity, even for nonmalignant lesions. In the reduced atmospheric pressure at flight altitudes, small, usually insignificant collections of air, gas, or hematomas could cause pain or incapacitation. Otherwise, after complete healing, assuming that major organ dysfunction does not exist, a surgical procedure itself should not be considered disqualifying for flight. The condition for which the surgery was performed, however, might be disqualifying.

The use of cytotoxic chemotherapeutic agents should be considered incompatible with flying. Such chemotherapy indicates a serious underlying disorder. The therapy itself can lead to anemia, thrombocytopenia, and granulocytopenia, which could precipitate an incapacitating event while flying. In addition, nausea and vomiting are well-recognized, widely-accepted and predictable side effects of cancer hemotherapy that may create difficulty in performing flight duties.

As with hematologic neoplasms, the use of corticosteroids for solid tumors, regardless of dose, should be disqualifying. Also, unpredictable toxic effects can occur with pharmacologically high doses of female or male hormones.

Radiation therapy is generally delivered to a localized area over a finite period of time. The immediate side effects of nausea, blood count depression, and other dose-related toxicities usually disappear within a few weeks after completion of radiation treatment. Thus, during the course of radiation therapy, and for a reasonable time thereafter, an individual should be considered unfit to fly.

In general, we recommend that the staging system for cancers be used to evaluate the fitness of individuals with a solid tumor to pilot an aircraft. However, the consultants on this medical standards review project could not agree completely on the

disposition of individuals with certain stages of certain neoplasms. Therefore, in the following discussion of specific tumors we will present the conflicting views.

- Neurological tumors

  1. Brain tumors

     The diagnosis of primary or metastatic malignancy to the brain should be permanently disqualifying. One-third of all persons with malignant brain tumor experience seizures, which are unexpected and totally incapacitating. Survival rates for malignant gliomas approach 20% after one year. Disability due to therapy is high.

  2. Spinal cord tumors

     Diagnosis of either primary or secondary spinal cord malignant tumors should be disqualifying for certification. Small masses may produce very rapid and extensive neural dysfunction, and there is a risk of rapid recurrence after therapy.

  3. Malignant tumors of the peripheral nervous system

     Certification may be granted if no appreciable impairment results. These tumors are not usually confined like the brain and spinal cord tumors. The applicant should be re-examined at 6-month intervals for the evaluation of progressive neurological dysfunction that might preclude the safe performance of the pilot's duties.

- Tumors of the head and neck

  1. General considerations

     Cancers of the lip, tongue, salivary gland, floor of mouth, oropharynx,

unspecified areas of the mouth, and generally of the head and neck are seen with increased incidence in cigarette smokers. The earliest invasive tumor, T_1, is generally defined as a primary tumor less than 2 cm in diameter. Early TIS (carcinoma in situ) tumors and superficial tumors of the lip and some of the tongue may be totally curable by local means. In true invasive cancer even in the earliest stages, the five-year survival is about 75%. Some percentage of the survivors will have detectable disease at five years. Predicting who will relapse is impossible. Persons who have no clinical evidence of disease after careful study by a competent head and neck oncologic surgeon may be certified for flight duties. Re-examinations should be considered for continued certification as discussed below.

2. Oral cavity cancer: Stage I (T_1 N_0 M_0) and Stage II (T_2 N_0 M_0) carry fairly good five-year survival rates (76% and 67%, respectively), and recurrences are primarily local. Adequate treatment should be followed by a careful examination four months after the termination of appropriate therapy. The applicant may be certified when there is no evidence of recurrence or dysfunction of speech or airway. **Oncologists' recommendations:** Re-examination should be required at three to 6 month intervals for all classes of airmen. Persons with Stages III and IV disease should not be certified. **ENT specialists' recommendations:** Medical recertification should require an examination by a competent head and neck oncologic surgeon at each regularly-scheduled certification examination for Classes I and II and annually for 5 years for Class III applicants. Persons with Stages III and IV cancers may also be certified with similar follow-up.

3. Pharyngeal cancer: These lesions are usually diagnosed late and individuals with them have only a 33% rate of survival after 5 years. **Oncologists' recommendations:** Because of the involvement of the upper airway, applicants with pharyngeal cancer should not be certified. **ENT specialists' recommendations:** Certification should depend upon the results of therapy and should be handled on an individual basis. Class I and II pilots should be re-examined by a head and neck oncologic surgeon at the time of their regularly-scheduled certification examination. Class III pilots should be re-examined annually.

3. Laryngeal cancer: **Oncologists' recommendations:** Stage I ($T_1 N_0 M_0$) disease carries an excellent prognosis with radiation alone. A greater than 76% five year survival rate is expected and recurrence is local. Certification for all classes of airmen is recommended if phonation is not affected appreciably. Re-certification examinations at 6 month intervals for five years, then at yearly intervals, are indicated. Persons with Stage II, III, and IV lesions should not be certified because of compromise to the airway and phonation as a result of adequate treatment of these lesions. **ENT specialists' recommendations:** Early laryngeal carcinomas have an excellent prognosis when treated with radiation or surgery. Certification for all classes of airmen is recommended if phonation is not affected appreciably. Class I and II pilots should be examined at their regularly-scheduled recertification examination, and Class III pilots should be examined annually for five years.

4. Miscellaneous carcinomas of head a' n- k: **Oncologists' recommendations:** Carcinoma of the he- and neck requiring extensive surgery, such as radical neck dissection or mandibulectomy, should be evaluated on an individual basis due to the possibility of airway obstruction, difficulty in phonation, or difficulty in using an oxygen mask. Persons with only the earliest clinical stages of miscellaneous head and neck tumors should be considered qualified to fly and have frequent reassessments. **ENT specialists' recommendations:** Carcinomas of the head and neck requiring extensive surgery must be evaluated on an individual basis due to potential airway obstruction, difficulty with phonation or difficulty using an oxygen mask. Radical neck surgery generally does not interfere with respiratory function or phonation.

o Lung cancer

Lung cancer, with all of its various cell types and stages taken together, has only a 9% five-year survival rate. Persons who have had curative surgery for localized cancer of the lung and in whom all disease is confined to the lung without spread to regional, hilar, or mediastinal nodes have a five-year survival rate of only 42%. All applicants with the diagnosis of lung cancer should be denied certification because of the risk of brain metastasis that might lead to seizures. The site of first relapse of lung cancer is frequently within the brain and central nervous system. In one study of 247 patients, at autopsy, the following incidences of brain metastasis for the various types of lung tumors were: epidermoid, 14%; small cell, 30%; adenocarcinoma, 25%; large cell, 29%.[7] Thus, precluding all of the other potential complications of

these tumors, which would include diminished pulmonary function, bone metastases, hemorrhage, and secondary complications of surgery, the risk of brain metastasis alone should be a cause of denial of certification for at least five years for all classes of airmen.

An individual with early stage carcinoma of the lung who after initial local therapy and who after a five-year period has no evidence of disease may be considered for special issuance. The applicant should be examined carefully by a qualified specialist with training in the diagnosis and therapy of pulmonary neoplastic disease. A minimum examination would include a chest radiograph, a computed axial tomogram (CT Scan) or magnetic resonance image (MRI) of the brain, chemical tests of liver function, and complete medical history and physical examination. Endoscopic and special surgical evaluations and biopsies may be necessary to assure a disease-free state.

o Gastrointestinal tract cancers

Cancers of the esophagus, stomach, small intestines, large intestines, rectum, liver, biliary passages and pancreas constitute a major group of malignant disorders with poor prognoses. Carcinomas of the digestive organs are staged according to several common but different classifications. The TNM classification system is applicable to most tumors of the alimentary tract.

1. Esophageal carcinoma

Carcinoma of the esophagus has a very poor prognosis. The five year survival rate for treated and untreated disease is approximately 3%.

Applicants with the diagnosis of esophageal cancer should not be certified because of the risk of inanition and of sudden hemorrhage, and aspiration. An applicant who has survived five years and has no evidence of disease following local therapy might be considered for special issuance. The applicant should be examined by a specialist in gastroenterology or other specialist with interest and expertise in the diagnosis and treatment of malignant disease, and a minimum examination should consist of a complete history and physical examination, a chest radiograph, barium swallow, biochemical tests of liver function, a test for occult blood in the stool, and a CT Scan of the mediastinum and the abdomen. Special endoscopic or surgical evaluations and biopsies may be necessary to establish a disease-free state.

2. Gastric carcinoma

Because of the risk of sudden hemorrhage and the side effects of palliative therapy and local treatment, persons with the diagnosis of carcinoma of the stomach should not be certified. The five-year survival rate with cancer of the stomach of any stage is approximately 12%. Disqualification should last for a minimum of five years. An applicant who has had five years of survival with no evidence of disease following local therapy might be considered for special issuance. The applicant should be examined by a specialist with particular interest in the diagnosis and evaluation and treatment of persons with gastrointestinal malignancy. The examination should contain at a minimum a complete history and physical examination, a test for occult blood in stool, a chest radiograph, biochemical tests of

liver function, and evaluation of the upper gastrointestinal tract either by endoscopy or barium contrast studies.

3. Primary hepatic carcinoma

Primary hepatocellular carcinomas are usually unresectable and have a mean five-year survival rate of near zero percent. Because of the complications of pain, hemorrhage into the tumor, and hemorrhage in general due to liver disease, applicants should be denied certification. An applicant who has demonstrated five years of survival and has no evidence of disease following initial treatment might be considered for licensing. The examination should include a complete history and physical examination, chest radiograph, biochemical tests of liver function, and an assessment of the liver by either CT Scan or isotope liver scan.

4. Carcinoid syndrome

This discussion about carcinoid syndrome concerns only metastatic carcinoid disease and not carcinoid tumors that were removed incidental to other surgery in portions of the gastrointestinal tract. Carcinoid disease is a common disease of the intestinal tract, and it also can involve the lung and other organs. The carcinoid syndrome implies the presence of metastatic disease with the production of vaso-active and other hormones. Because the syndrome may produce sudden dyspnea and altered mental function, the diagnosis of the carcinoid syndrome should result in denial of certification. Persons with carcinoid tumors that are totally resected without metastases and who do not have evidence of the syndrome may be considered for

certification.

5. Gallbladder and biliary cancer

Gallbladder and biliary cancers carry a 2.5% five-year survival rate, making applicants with these types of malignancy uncertifiable. For applicants who have had five years of survival with no evidence of disease following local therapy, licensing may be considered. The minimum testing described above for primary hepatic carcinoma should be followed.

6. Pancreatic cancer

Only 10% to 25% of persons with carcinoma of the pancreas have tumors that are resectable, and only 8% of that proportion survive five years; thus, this disease has a poor prognosis. Unresected tumors are characterized by three disorders that would affect a pilot's proficiency: 10% to 20% develop diabetes mellitus, a significant percent develop thrombophlebitis and other thrombotic phenomena, and there is also an overall incidence of serious psychiatric disorder approaching 76%. Thus, the diagnosis of carcinoma of the pancreas should preclude certification. Applicants who have demonstrated five years of survival with no evidence of disease following local therapy might be considered for special certification. Such certification would require evaluation for diabetes mellitus, and the criteria for a certificate for diabetes should apply to the evaluation. Testing should also be done for pancreatic exocrine insufficiency. Recommendations regarding malabsorption and pancreatic insufficiency should apply. The re-evaluation for carcinoma of the pancreas should include at a

minimum a history and physical examination, chest radiograph, biochemical tests of liver function, a stool examination for occult blood, and a CT scan of the entire upper abdomen, pancreatic region, and retroperitoneum with special attention to the liver and pancreatic bed. Special endoscopic, surgical, or contrast studies may be needed to establish existence of a disease-free state.

7. Small bowel cancer

Small bowel carcinoma is an unusual tumor. Cases should be considered on an individual basis depending on cell type, location, stage, and therapy given.

8. Carcinoma of the colon and rectum

Carcinoma of the colon and rectum is staged according to several classifications. The TNM classification for colon-rectal carcinoma recognizes an early TIS (carcinoma in situ) where the primary tumor involves no invasion of the lamina propria. The T_1 tumor is confined to the mucosa or the submucosa, and the T_2 tumor is limited to the wall of the colon and rectum. The T_3 tumor involves all layers of bowel wall including serosa, with or without extension to adjacent or contiguous tissues.

Oncologists' recommendations

a. Carcinoma of the colon Stage I (T_1 N_0 M_0)

Persons with carcinomas of the colon in early stages, which are treated with or without colostomy, should be certified for all classes of airmen. A measurable proportion of patients can be

expected to develop either local or distal tumors, but it is difficult to predict who will have relapses. Most relapses occur within the first few years after surgical treatment. Re-examination with liver function studies, carcinoma embryonic antigen (CEA) and chest radiograph should be required at three-month intervals for at least three years. At that time, the interval could be lengthened to 6 months. Colonoscopy or barium studies of the colon should be required yearly.

b. All colonic malignancies beyond Stage I should be denied certification. An applicant who has had five years of survival with no evidence of disease following local therapy may be considered for special issuance. The examination should contain at a minimum a complete history and physical examination; a chest radiograph; a stool examination for occult blood; biochemical tests of liver function; a CT scan of the entire abdomen with special attention to the liver, retroperitoneum and colonic region; and an examination of the entire colon, either by colonoscopy or barium enema with air contrast technique.

The recommended schedule for re-examination of applicants with early stage colon carcinoma may be difficult or impossible to follow. If the follow-up program is impractical, the prudent course would be to deny certification, even to early stage patients, for at least the first three years following initial local treatment.

Gastrointestinal specialists' recommendations: Persons with a palliative resection of an adenocarcinoma of the colon, or recurrent adenocarcinoma should be disqualified. Persons with a curative resection of an adenocarcinoma may be qualified three to 6 months after the operation, depending upon the procedure.

An applicant who has a successful resection of carcinoma of the colon, with the tumor limited to the bowel and regional lymph nodes, and who has been issued a certificate, will be required to have follow-up examinations with a carcinoma embryonic antigen (CEA) done at least every 6 months following issuance. The result must be forwarded to the FAA along with results of any other studies relating to the status of the airman's tumor. The airman should understand that any suspicion of recurrence of tumor is disqualifying until tests are obtained indicating he or she is free of disease.

9. Miscellaneous gastrointestinal tract tumors

Other malignancies of the gastrointestinal tract occur infrequently. Applicants with anal or other carcinomas should be considered for certification on an individual basis. The overriding consideration should be the predictability or lack of predictability of relapse, the common sites and timing of relapse, and the adverse impact that such relapse might have on aviation duty.

o Urinary tract malignancy

1. Kidney

Carcinomas of the kidney are infrequently diagnosed in a truly localized curable stage. With localized disease, the five-year survival rate is reported at 72%. The smallest T_1 N_0 M_0 tumors that show minimal renal and caliceal distortion or deformity and that are surrounded by normal renal parenchyma have good five year survival rates after curative surgery. However, these individuals do have a significant risk of relapse and should be disqualified by the AME for consideration for special issuance. One-third of patients already have hematogenous metastases at the time of diagnosis, involving the lung in 50% of cases, bone in 30% of cases, liver in 30% of cases and the brain in 25% of cases. The risk of rapid development of abnormal pulmonary function or neurological deficits with seizures is significant. Therefore, applicants with a diagnosis of carcinoma of the kidney with extension outside the kidney should be permanently disqualified.

Persons with T_1 intrarenal lesions that are completely removed at surgery may be recertified if they have no evidence of disease five years after surgery. Patients with T_1 intrarenal neoplasms identified incidentally at surgery, and those with primary nonrenal cell malignancies, such as sarcoma, may be recertified if they have no evidence of disease two years after surgery. An examination should include at a minimum a complete history and physical examination, a chest radiograph, biochemical tests of liver function, and a CT scan of the entire abdominal contents and retroperitoneum. Other special

studies, surgical procedures, or biopsies may be required to establish the presence of a disease-free state.

2. Urinary bladder

Overall, malignancies of the urinary bladder are associated with a five-year survival rate of 67%. True carcinoma in situ or papillary noninvasive carcinoma are associated with a high probability of cure. Therefore, Stage I and II malignancies that have been treated with intent to cure should be certified. Recurrence is primarily local, and no sudden symptoms except hematuria occur. Re-examination for certification should be required at three month intervals for five years. After five years examinations could be extended to intervals of 6 months. Persons with Stage III and IV disease should be disqualified until they have been shown to be free of disease five years after local curative therapy, at which point they should have an examination that includes at a minimum a history and physical examination, chest radiograph, biochemical tests of liver function, endoscopy and contrast studies of the entire urinary tract if appropriate, and computer scanning of the entire abdominal contents and pelvis.

3. Prostate

Cancer of the prostate frequently occurs in older males. The degree of histologic differentiation has a high bearing on relapse potential and mortality. For the earliest localized stage of truly invasive carcinoma of the prostate, the five-year survival rate is reported to be 77%. When all stages are considered, the five-year survival rate is

63%. Young individuals with early stage disease at presentation generally have more poorly-differentiated tumors and do less well than older individuals. Clinical staging takes into account the suspicion for malignancy on digital rectal examination.

Stage A disease occurs when carcinoma is not suspected, but the individual has a transurethral prostate resection for symptoms of urinary obstruction thought to be due to benign hypertrophy, and on microscopic analysis malignancy is noted. The special issuance procedure should allow persons with Stage A to be certified after careful study for local or systemic disease. Such persons should be re-examined frequently, although many will be permanently cured of malignant disease. Persons with Stage I, II, and III carcinoma of the prostate, who have no distant metastases and who are treated with curative intent, should be certified when the effects of treatment have subsided. However, treatment with estrogen compounds should result in denial due to the recognized propensity for thrombosis.

Persons with localized disease should be re-examined at 6 month intervals with determinations of alkaline phosphatase, radioactive immunoassay for the prostatic fraction of acid phosphatase, serum calcium, urinalysis, and clinical examination of the prostate. The examination should be done by a physician with expertise in the examination and treatment of individuals with prostatic malignancy. Renal function testing, including blood urea nitrogen, creatinine, and intravenous pyelography, should be done annually. Individuals with Stage IV disease should not be certified.

In summary, the predictability of the recurrence of prostate cancer is difficult; determinants include not only tumor size at presentation but also histologic differentiation and age of the individual. Hard and fast rules about special issuance certification for some tumor types are difficult to develop, and the FAA needs to rely on a panel of experts with special knowledge in prostatic malignancy to recommend action on a case-by-case basis.

4. Carcinoma of the testicle

With present chemotherapeutic regimens persons with advanced testicular cancer or with recurrent cancer after local therapy have a good chance of cure. The treatment of early-stage carcinoma of the testicle may consist simply of surgery with or without radiation. However, both seminomatous and nonseminomatous testicular cancers may recur soon after the initial therapy. Persons who have two years of survival after treatment with curative intent and have no evidence of disease have a statistically high likelihood of cure, and they may be certified if a thorough examination discloses no evidence of recurrence.

It would be prudent to wait three years after completion of the initial treatment or treatment after the first relapse before a major re-evaluation for certification is undertaken. The examination should be done by a physician with expertise in evaluating and testing persons with testicular malignancy. A major examination for metastases using the appropriate serum markers should be done. The long-term

complications of radiation and/or use of multiple chemotherapeutic agents are similar to those of Hodgkin's disease. Therefore, frequent reassessment is prudent.

o Gynecological cancer

1. Carcinoma of the ovary

Since less than 28% of ovarian cancers are localized at the time of inital diagnosis, prognosis is usually guarded, and continuous therapy with cytotoxic chemotherapeutic agents precludes certification of applicants. An applicant who has demonstrated five years of survival with no evidence of disease following local therapy for carcinoma of the ovary might be considered for special issuance. The applicant should be examined by a specialist with expertise in diagnosis and therapy of patients with malignant ovarian disease. The minimum tests should include a history and physical examination, including a thorough pelvic exam; a chest radiograph; biochemical tests of liver function; a CT scan of the entire abdomen, retroperitoneum, and pelvis; an intravenous pyelogram; and a urinalysis. Other special tests may be indicated in selected cases.

Unusual types of ovarian tumors should be considered on a case by case basis.

2. Cancer of the corpus uteri

The most common carcinoma of the corpus uteri is adenocarcinoma of the endometrium. Staging depends upon the size of the primary tumor, the presence of affected regional nodes, and the presence or

absence of distant metastases. Spread of adenocarcinoma of the endometrium may be slow and tumor recurrence is usually local for long periods of time. If these tumors do not cause obstruction of the ureter or bowel, a pilot should not be adversely affected. On the other hand, effects of radiation may create a significant problem. The earliest truly invasive carcinoma of the endometurium, Stage I, has a 90% cure rate. Unfortunately, recurrence is unpredictable for tumors of all grades of differentiation. Therefore, it is prudent to consider all persons with a history of truly invasive carcinoma of the corpus uteri as not fit for certification.

Certification of persons with Stage I and II disease might be considered two years after adequate local therapy, if they have been free of disease. The minimum examination would include a history and physical examination, chest radiograph, urinalysis, biochemical tests of liver and renal function, and a careful pelvic examination by an individual with particular training and experience in gynecological malignancies. Specialized local studies such as intravenous pyelography, abdominal pelvic ultrasound, or CT scan should be done as well. Those persons with Stage III and IV disease at the time of initial diagnosis should not be certified until they have been free of disease for a period of five years after local therapy. All persons should be re-examined at intervals of 6 months.

4. Carcinoma of the cervix

As a site of frequent cytologic screening, the uterine cervix lends itself to diagnosis of carcinoma in early stages. The five-year

survival rate of Stage 0 carcinoma of the cervix (carcinoma in situ) is virtually 100%. The five-year survival rate of localized carcinoma of the cervix is approximately 82%. The five-year survival of all stages taken as a group is 59%.

Applicants with Stage 0 or I carcinoma of the cervix may be certified immediately after curative treatment has been completed, whether with radiation or surgery. Follow-up re-examinations at 6 month intervals for three years, and then at yearly intervals, are recommended. Persons with Stage II, III and IV disease should not be certified due to local complications of treatment and of unpredictable likelihood for recurrent disease. Individuals who have been demonstrated to be free of disease for a period of two years may be certified by special issuance. Minimum testing to assure a disease-free state is the same as with examination of the corpus uteri and includes examining cytology of the vaginal cuff.

o Carcinoma of the breast

Breast cancer is the most common malignancy in females, and one percent of breast cancers occurs in males. Various features are important when assessing the prognosis of a person with a history of breast cancer. The most common histologic group represents infiltrating carcinoma of ductal structures. Prognostic features include the amount of differentiation of the tumor and the presence or absence of lymphatic and vascular invasion. However, there have not been good studies correlating these features with relapse potential and survival.

In even the earliest Stage I cancer (T_1 N_0 M_0) with a small initial tumor and no involvement of axillary lymph nodes, the relapse rate at five years can approach 20%. With standard therapy Stage I carries approximately an 80% five-year survival rate with no evidence of disease. This is a low enough risk to justify certification with frequent re-evaluation. Re-evaluation should be done at a minimum interval of 6 months and should consist of a chest radiograph, liver function tests, serum calcium and alkaline phosphatase and a careful clinical examination by an individual with particular training and expertise in dealing with breast malignancies. Mammography of the opposite breast should be required annually. Special studies may be required based upon clinical or laboratory abnormalities.

Persons with Stage II, III and IV disease should be permanently disqualified because of the unpredictable and high incidence of metastatic disease, especially cerebral metastases with potential for causing seizures. In early Stage I disease certain histologic features have been associated with an increased incidence of recurrence, but predicting in advance who may or may not relapse is impossible.

o Malignant melanoma

Malignant melanoma is a potentially serious disorder with increasing incidence. Staging depends upon depth of penetration of the local tumor, the presence or absence of nodal disease, and the presence or absence of distal metastatic disease. The primary tumor is classified as T_0 when atypical melanocytic hyperplasia (Clark's Level I) is present. This is not considered a malignant lesion. T_1 requires invasion of the papillary dermis (Level II) or penetration of 0.75 mm thickness or less. T_2 implies invasion of the

papillary-reticular-dermal interface (Level III) of penetration of 0.76 to 1.5 mm in thickness. T_3 implies invasion of the reticular dermis (Level IV) or invasion from 1.51 to 4.0 mm in thickness. T_4 implies invasion of the subcutaneous tissues (Level V) or penetration greater than 4.1 mm, or satellite nodules within 2 cm of the primary melanoma. T_1 or T_2 lesions with clinically negative lymph nodes and no evidence of metastatic disease are called Stage I, and they are the only tumors that do not preclude certification. Stage I tumors should be diagnosed only by a pathologist with particular interest and expertise in melanoma analysis.

Oncologists' recommendations: A person with all stages above Stage I should be denied certification because of metastatic propensity: 87% of persons with melanoma have metastases to the lung and 54% have metastases to the brain. A significant number of these persons will have seizure activity as the first manifestation of metatasis. Individuals with Stage II-A or II-B (T_3 or T_4 with N_0M_0 clinical staging) who have demonstrated a survival of five years with no evidence of disease might be considered for certification. An examination for recurrence of melanoma should consist of a complete history and physical examination, chest radiograph, biochemical tests of liver function, computer assisted tomography of the brain, and liver scanning. These examinations should be repeated at yearly intervals for a minimum of 10 years.

Dermatologists' recommendations: Those persons with a malignancy of the skin with evidence of regional or systemic metastases should be disqualified from flying, until such time that the tumor and its metastases are considered cured.

References

1. Golde DW, Hocking WG, Koeffler HP, Adamson JW: Polycythemia: mechanisms and management. Ann Int Med 1981;95:71-87.
2. Kannel WB, Gordon T, Wolf PA, McNamara P: Hemoglobin and the risk of cerebral infarction: The Framingham study. Stroke 1972;3:409-420.
3. Willison JR, duBoulay GH, Paul EA, Ross-Russell RW, et al: Effect of high haemoglobin on alertness. Lancet 1980;1:846-848.
4. Winslow R: Circulation in Winslow R, Monge CC (eds): Hypoxia, Polycythemia and Chronic Mountain Sickness, Baltimore, Johns Hopkins University Press, in press.
5. Jacobs AD, Champlin RE, Golde DW: Recombinant alpha-2-interferon for hairy-cell leukemia. Blood 1985 (in press).
6. American Joint Committee on Cancer: Manual for Staging of Cancer, ed 2. Philadelphia, JB Lippincott Co, 1983.
7. Newman SJ, Hansen HH: Proceedings: Frequency, diagnosis and treatment of brain metastases in 247 consecutive patients with bronchogenic carcinomas. Cancer 1974;33:492-496.

PREVIOUS PAGE IS BLANK

Visual System

Introduction

This report consists of two broad subjects: 1) discussion of and rationale for the recommended standards, and; 2) description of and recommended disposition of specific ophthalmologic diseases. In addition, the special report forms for ophthalmologic disorders were reviewed and some minor revisions recommended.

Discussion and Rationale for Recommended Standards

- Distant visual acuity for Classes I and II

  We do not recommend a refractive error limit. Rather, an applicant should be able to correct from 20/200 to 20/20 or better in each eye with either conventional spectacle lenses or contact lenses.

- Distant visual acuity for Class III

  In most states, a visual acuity of 20/40 is required to obtain a driver's license. Thus, it is justifiable to change the visual acuity limit from 20/50 to 20/40 in order to obtain a pilot's license. We do not recommend a refractive error limit or uncorrected acuity limit that would prohibit an applicant from obtaining a Class III medical certificate.

- Near visual acuity for Class I

  Sixteen inches is an appropriate reading distance. The V = 1.00 vision terminology is antiquated and is not familiar to most AMEs and ophthalmologists. Furthermore, the near vision acuity chart which is on Form 8500-1 contains Sloane letters that are to be used at 16 in.

After age 50 years, we recommend a near vision standard of 20/40 or better at both 16 in and 32 in with or without corrective lenses. This additional requirement would correct for one of the major deficiencies in the visual examination for flying: it is important while piloting to be able to see clearly at close distances, as when looking at maps, and at intermediate distances, as when viewing the instrument panel. This is especially important in night flying. Diminished intermediate visual acuity due to presbyopia in an individual 50 years of age or older may be further compromised by bifocal correction. Trifocals or progressive power lenses may be necessary for clear vision at distance, intermediate, and near.

- Near visual acuity for Class II

  The comments above concerning the need for testing for intermediate distance visual acuity apply to Class II as well.

Testing for visual acuity using the Snellen notation of 20/40 is based on the fact that on official aeronautical maps the characters are of 20/40 linear Snellen scale-size or larger. Thus, the recommended standard provides a uniform and standard method to determine whether a Class II pilot can read aeronautical charts.

- Near visual acuity for Class III

  Near vision requirements for the Class III medical certificate should be established because at least 20/40 near vision is necessary to read charts, emergency procedure guidelines, and aeronautical maps accurately. Since not as much instrument flying is done by private pilots, an intermediate vision requirement after age 50 for Class III pilots is not necessary.

o Color vision for Classes I and II

Genetic color deficiencies occur in approximately 8% of the male population and in less than 1% of the female population. These individuals experience varying degrees of color confusion, including shifts in chromaticity or color temperature, especially when colors are desaturated by environmental conditions.[1]

A normal observer requires the ability to perceive the three standard colors, red, green and blue, in order to match an unknown wavelength to these three known standards. This person is classified as being trichromatic. A small percentage of persons with abnormal color vision can perceive only two of the three standard colors. These individuals are classified as dichromatic. Three types of dichromatic individuals exist: those lacking red-sensitive, those lacking green-sensitive, and those lacking blue-sensitive cones. An individual is a protanope if he or she has red-green color deficiency related to red-sensitive cone loss. A deuteranope also has red-green deficiency related to green-sensitive cone loss. A tritanope has blue-yellow deficiency related to blue-sensitive cone loss.

The largest group of color deficient persons are trichromatic, and they have anomalous color identification. These individuals use all three standard colors to match unknown hues, but in "anomalous" amounts compared to normal trichromats. They are actually "color-weak" rather than "color-blind." Each of the three types of anomalous trichromats has a defect analogous to that of the dichromats. For example, the protanomalous trichromat is "red-weak," requiring more red in a red-green mixture to

match an unknown hue.

There are two forms of severe color deficiency, or achromatopsia. These individuals are monochromatic and have complete loss of color discrimination. Rod monochromatism is a condition in which the individual has no functioning cones from birth. An individual with this condition has very low visual acuity, totally absent color vision, aversion to bright lights, and nystagmus. Cone monochromatism is extremely rare, and an individual with this condition has no hue discrimination, but has normal visual acuity. The following table shows the prevalence of these color deficiencies in the general population:

Prevalence of Color Deficiency

Classification	Percent
Protanopia	1.0
Deuteranopia	1.4
Protanomalous Trichromat	0.78
Deuteranomalous Trichromat	4.6
Total	8.0
Monochromat	1 per 100,000

Source: Reference 2

An early report of the significance of color vision in aviation was written by Colonel W. H. Wilmer and Major Conrad Berens, in 1919: "The proper recognition of color plays an important part in the success of all types of flyers."[3] Wilmer and Berens indicated that color vision was useful in map-

reading, recognizing sky rockets and flares, and identification of red, green, or white lights at airports. They noted also that it was useful to have reliable color vision to select a place for forced landing. Of course, aviation technology has increased multifold since that time, yet many of the initial concepts described in this early report about color vision still apply to aviation.

The use of color vision in aviation can be briefly outlined as follows:

1. Aircrew color discrimination
    a. Identification of color light signals and navigation lights
    b. Airport beacons
    c. Approach lights
    d. Runway lights (end lights, center lights, stopping distance markers)
    e. Taxi strips
    f. VASI (Visual Approach Slope Indicator)
    g. Instrument panel (navigation and warning lights)
    h. Instrument panel color TV-type instrument displays, and radar

2. Identification of colors on reflecting surfaces
    a. Instrument zone markers on panels
    b. Smoke
    c. Flares

3. Map and chart reading

4. Color differences in terrain

The use of VASI requires recognition of minute changes in a red-white color relationship. Both deuteranopes and protanopes may have difficulty with VASI, although they may not be aware of any difficulty. The deuteranopes will probably experience less difficulty, although there are some who will not be able to utilize the VASI system safely. Unfortunately, it is not possible to predetermine which deuteranopes will not be able to utilize VASI.

A change from the present use of edge taxi lights to the increasingly more popular center taxi lights may make night taxing easier for the pilot with normal color vision. However, the concomitant change from blue to green may lead the deuteranopes to straddle the runway lights, perceiving them to be the green taxiway lights, with obvious dangerous consequence.[4]

End markers denoting the amount of remaining runway are colored lights. Colored lights also mark a high speed taxiway exit.[5] Where red lights are used protanopes will have particular difficulty interpreting the red light cues.

Aircraft navigation lights may be particularly confusing to color-deficient individuals. Since navigation lights are small point sources at usual ranges, in normal individuals color perception might be diminished by the physiological phenomenon of small field tritanopia, which may lead to confusion of blue and green, and white and yellow. Dichromats might confuse a green signal with either a red or white signal, or even both, depending upon the precise chromaticities of the colors used. As a result dichromats, who may already have reduced color vision, may be further

reduced to monochromaticity, or total color-blindness, when the physiological phenomenon of small field tritanopia is added.

Another problem is that protanopes may fail to see the red navigation lights at a useful range because of their lowered sensitivity to red light.[6] Protanopic pilots are denied reliable information that is provided by the navigation light code and need to rely entirely on a fix-of-bearing for a determination of a potential collision course at night, which is not always as reliable.

Any pilot with a protanopic color vision defect will be further disabled by the inability to detect a red anticollision light at a range that allows sufficient time to make a fix-of-bearing judgment prior to a possible collision. White strobe anticollision lights may assist this protanopic observer, but the white color is permitted only in the United States. Bowman and Cole reported that the navigation light system at night serves as a crude screening method to place another aircraft into a potential threat vs a no-threat category (collision vs no collision) based on the observation of the aircraft's navigation lights.[7]

It is difficult to establish whether or not the present color vision requirements and screening for them have resulted in enhanced flight safety in the past. It has not been possible to find any reported aircraft crash or collision that has been attributed primarily to a color vision defect. The US Air Force reported on a group of twelve aviators with considerable flying experience who failed the Dvorine color plate test. Eight were pilots with an average of 3,274 hours each, for a total of 26,196 flying hours. The other

four were a navigator, a flight engineer, a boom operator, and a flight surgeon. They denied ever having any difficulty due to defective color vision in performing their jobs. None of these experienced men was grounded after the discovery of the defect. Eleven of the 12 had a deuteranopic defect and one, the flight surgeon, had a protanopic defect.[8]

This study led the Air Force to study which types of color deficiencies, if any, were acceptable for flight. This study, combined with the examination results of 2,591 additional pilots and navigators, led to the conclusion that persons with protanopia, deuteranopia, any degree of protanomaly, and all but the mildest degree of deuteranomaly, are almost never successful as pilots or navigators. The Air Force could not establish with certainty how mild a deuteranomaly would be acceptable, but those individuals who scored above 50 on the SAM Color Threshold Tester were considered very mildly color deficient, and probably safe and acceptable for flight.[9]

Dille and Booze reviewed data from 1974 through 1976 on accident experiences of civilian pilots with selected static visual defects, including deficient color vision. They noted that the group with deficient color vision and no operational restrictions had a significantly higher accident rate than expected. Individual accident records were reviewed to determine any possible relationship between visual defects of the pilot and the accident cause, such as phase of light, type of flying, time of day and weather. No unusual associations were determined.[10]

The visual system committee for this review project sent a questionnaire to airline medical directors seeking their opinions on the importance of color

vision in aviation. Two responses were received from these medical directors, and they were on opposite sides. One director stated there is a considerable need for normal color vision in commercial pilots and airline captains, since many aircraft systems, charts, and manuals are color coded. In addition, new instrument systems include features such as visual display terminals which are color-dependent.[11]

The other medical director stated that prospective studies of the importance of color vision in aviation show no apparent relationship between aircraft accidents and color-deficient individuals. This medical director pointed out the less critical role of color vision in night flying, since anticollision lights are strobe lights and airports are located by rotating beacons. Furthermore, most operations now use radar. This director also pointed out that although in the modern-day cockpit color may aid in the recognition of instrument readings, the classical method of glancing at a pointer location, rather than color indication, is still probably more important. He concluded that color vision may play a small role, if any, in modern aviation.[12]

In summary, the hazard to aviation safety of anomalous color vision is not clear. No studies have shown that color deficiency has been a direct cause of accidents.[13] On the other hand, color is an important constituent of aircraft devices such as instrument panel gauges and warning lights, and of airport landmarks, such as beacons and runway lights. Thus, we recommend that testing for color vision remain in the standards and in the routine AME examination, and that the standard for Class II pilots, who fly frequently and who are responsible for transporting humans, be as strict as the standard for Class I.

Screening for color vision has its problems, too. The US Army, which has greater control over testing procedures than is possible in civil aviation, reported that testing is 23-25% effective in screening out anomalous color vision. Persons with dichromatic deficiency were most easily detected.[14]

Another problem is the great variety of color vision tests and the inconsistencies among their scores for normal color vision. This problem is compounded by the different recommended standards for color vision in civilian aviation, that is, "normal" color vision for Class I and II and "ability to distinguish aviation signals red, green and white" for Class III. A test in the physician's office that would predict a pilot's ability to distinguish the aviation signals (ie, a test that would correlate with the signal light gun test at air traffic control towers) is most desirable, but presently unavailable.

The current acceptable equipment for testing color vision includes the Eldridge-Green lantern, Farnsworth lantern, Keystone Orthoscope, Keystone Telebinocular, School of Aerospace Medicine Color Threshhold Tester (SAM-CTT), Titmus vision tester, and various isochromatic plates (Dvorine, HRR, Isihara). We have reviewed these color vision tests to assess their value toward detecting both normal color vision and the ability to distinguish aviation signals. This review has led to the recommendations for testing and disposition that are found in the proposed AME Guide.

Eldridge-Green Lantern

The Eldridge-Green lantern was first described in 1891 and used by the British Navy, and later by the US Navy and the British Railway Authority.

However, there are no reports in the literature since that time that demonstrate the validity of this device. Eldridge-Green claimed that all persons with color vision deficiency are readily detected, and that his device enables the examiner to pass those with milder forms of color vision defects. However, he also stated that about one-third of the population will have some difficulty with the lantern, which obviously demonstrates some difficulty with the interpretation of test results. The test is complex and lacks standardization, and therefore it is unsuitable as a practical test for color screening.[15]

Farnsworth Lantern

During World War II the Eldridge-Green lantern became unavailable and the US Navy explored possible replacements. The task of developing a new lantern went to Commander Dean Farnsworth in New London, Connecticut. His lantern uses three colors, a slightly desaturated red, a yellowish white, and a yellow green. He theorized that these three colors serve as an index of confusion from which the other confusions that protanopes and deuteranopes possess can be predicted.

The lantern does not simulate actual signal recognition, but it is a test of color vision. The Farnsworth lantern is unlikely to fail subjects with normal color vision.

Unpublished data at the Naval Submarine Medical Research Laboratory indicate that those persons with color deficiencies who fail isochromatic plates but pass the Farnsworth lantern are as accurate as those with normal vision in judging typical colors used by the services for coding, signaling and

communication.

The Farnsworth lantern differentiates the degree of defect very effectively and can detect persons with mild deficiencies. Since the Farnsworth lantern became the final validating test for color vision in the US Navy in 1954, 30% of the 10 men in 100 who are deemed to have a color deficiency by some other test have been salvaged for training as line officers, submariners, electronic technicians and pilots. It has the additional advantages of a brief testing time and of not requiring the use of a darkroom.

The test cannot be learned, so persons with color deficiency are unlikely to be able to achieve a passing result from prior practice and learning. It has also been shown to be a good predictor of performance on the daytime signal light gun test.[15,16]

School of Aviation Medicine Color Threshold Tester (SAM-CTT)

Also at the onset of World War II, other studies were in progress to develop a new lantern test at the School of Aviation Medicine. Louise Sloan developed the SAM-CTT. Her objective was to devise a simple instrument to provide a quantitative test of the ability to recognize aviation signals so that persons with color deficiencies who were "color safe" could be identified reliably. Sloan argued that aviation signal colors are saturated and differ in hue from each other, so that many persons with color vision deficiencies are able to recognize the colors easily under optimal conditions. However, at threshold visibility, their difficulty in recognizing color becomes more apparent. Thus her lantern presents colors at a range of intensities. Normals would be able to recognize the colors reliably even at the lowest intensities; those with

color deficiencies would be more likely to make errors as the intensity was reduced. As was not the case with other tests, considerable effort was made to validate the test scores of the SAM-CTT test by comparison with field tests. From the data of Sloan, a score of 60 is necessary if a color deficient individual is not likely to make errors at any of the practical field tests, but a score of 50 is sufficient for some tasks and also seems to be the level at which pilots do not report difficulty identifying aviation signals.[16,17]

There is a precedent in the Air Force to pass approximately 3% of the 8% who fail standard vision testing for aviators. These apparently are deuteranopic individuals who, judged by Air Force experience, do not pose an increased risk to aviation safety.[7,18] Detecting these individuals is, however, quite difficult with common testing procedures. If these mildly color deficient aviators desired first- or second-class medical certificates, testing by the SAM-CTT would be most appropriate if they first failed the screening isochromatic plates. A score of 50 or better on the SAM-CTT would provide justification for passing these individuals.

The SAM-CTT lantern is particularly useful for testing in the aviation setting, since it has been designed to simulate aviation signals. However, the cost of the lantern and training that is required to administer the test are prohibitive for practical use. As an alternative, an applicant could travel to an institution where the instrument is available. Despite these disadvantages, the test is a proven standard and should be retained.

Titmus Vision Tester

The Titmus vision tester was designed as a quick, accurate and reliable

method for identifying persons in need of more extensive ophthalmic evaluation. Five special test slides are appropriate to aeromedical testing. The color vision plate consists of 6 "accurately and authentically reproduced Isihara pseudoisochromatic plates." The test detects the presence of a color deficiency, but not the type.[19]

Keystone Orthoscope/Telebinocular

The Keystone Orthoscope/Telebinocular was designed in cooperation with the Special Products Division, Medical Department, of the Civil Aeronautics Administration. It incorporates tests for distant and near visual acuity, vertical phoria, lateral phoria, and color vision.

Color vision is tested by two test slides, one for "severe" color deficiency (red, green), and one for "mild" (blue, violet). The subject must correctly identify numbers in two out of three test circles on each slide.[20]

Pseudoisochromatic Plates

Generally, humans have the ability to distinguish hues, or wavelengths of colors, under various intensities and saturation against a series of confusing colors. This is the basis for the pseudoisochromatic plates. If an individual can identify the appropriate pattern in a pseudoisochromatic plate, then he or she should also be able to distinguish colors under usual circumstances found in flying activities. There is some objection to this approach, in that not every instance of color discrimination in aviation involves distinguishing pigments under confusing circumstances, as with these plates. Very often individuals are required only to distinguish colored lights, as with a signal.

In the pseudoisochromatic plates certain signs, in the form of figures, letters, or geometric shapes on the test plate are undecipherable by individuals with color deficiency, because they cannot distinguish colors of the signs and those of the background. If the differences between the colors are slight, the test is very sensitive and serves as a preliminary screening device. The type of deficiency, however, can be established only if the colors are more dissimilar.

Isihara Pseudoisochromatic Plates

The Isihara Atlas was designed to classify congenital protanopes and deuteranopes. Among all of the atlases presently available, it is best for this purpose. It does not, however, allow for sufficient distinction between the types of protanopes and deuteranopes, nor does it show the severity of the deficiency. It also does not distinguish the tritanopic deficiency, although it does reveal the acquired red/green dyschromatopsias. However, the acquired blue/yellow deficiency is frequently missed.[21]

The Hardy, Rand, Rittler Atlas (HRR) Plates

For congenital dyschromatopsias this test has mediocre specificity and sensitivity. Some normal subjects cannot decipher the proof plates in this atlas, and these may sometimes be interpreted properly by abnormal subjects. However, in the acquired dyschromatopsias, the HRR is one of the best tests presently available. In these disorders, it is able to distinguish both the green/red and the blue/yellow deficiencies, and lead to assessment of their severity. The instruction booklet notes that this test was designed as a qualitative as well as quantitative diagnostic test.[22,23] There are no data to correlate the test's qualitative results to the signal light gun test

performance.[14]

Dvorine Pseudoisochromatic Plates

Dvorine has reported that misclassification of normal subjects as color deficient is more likely to occur with the HRR test than with either the Isihara or Dvorine plates. Others have found that the Dvorine test is the most discriminating and reliable plate test. The results from this test are very closely correlated to results obtained on the anomaloscope, which is an instrument designed to detect anomalous trichromatics. The anomaloscope is also very useful in detecting dichromatics, since it works by mixing red and green colors.[24] It has also been determined that a quantitative diagnosis can be made with the Dvorine plates based on the number of plates that are interpreted incorrectly.[25]

The term, "normal," is applied to an individual who identifies 12 of the 14 plates of the first section. The terms, "color-blind" and "color-deficient," are applied to individuals who fail to identify three or more plates of the first section.[26]

Recent data have shown that if this test is used for predicting passing performance on the signal light gun test, these criteria may be too strict. For this purpose, nine or more errors would be classed as failing on the series of 14 plates.[27]

American Optical Pseudoisochromatic Plates (Richmond Plates)

The AO Plates, now produced by Richmond Optical, are a screening test for red/green, but not for blue/yellow, deficiencies. Therefore, they are neither

qualitative nor totally quantitative. However, they are easy to administer. The series consists of 14 plates, and a person who makes five or more errors is considered color deficient.[25] This level may be too strict in predicting performance on the signal light gun test. A score of 6 or more errors would be more appropriate for this test.[25]

In summary, using pseudoisochromatic plates is one of the simplest methods of distinguishing between normal and red/green color deficient perception. The pseudoisochromatic plates are an extremely effective tool in the hands of either expert or relatively untrained examiners, are inexpensive, and are the least time-consuming.[29]

- Visual fields for Classes I and II

The Class I and II criteria for field of vision should fall within the generally accepted norms for each eye. With average normal bony configuration of the face, position of the eyes within the orbits, and usual prominence of the nose, the acceptable minimal normal extent of the visual field from the point of fixation is as follows:[30]

Temporally in a horizontal line	95°
Down and temporally	90°
Directly downward in a vertical line	55°
Down and nasally	50°
Nasally in a horizontal line	60°
Up and nasally	55°
Directly upward in a vertical line	50°
Up and temporally	65°

These figures have a tolerance of $\pm 5^{\circ}$.

Reductions in visual fields may be expected in older airmen.[30] However, even slight reductions in visual fields may indicate active disease within the eye or the rest of the visual system. Maintaining a standard for normal visual fields is a method of detecting early ocular pathology.

In the present standard for Class III certification there is no visual field requirement. The committee does not recommend a specific visual field requirement for a Class III medical certificate, since eye disease serious enough to cause significant visual field loss will cause a denial under requirement that no serious pathology of the eyes be present.

- Ocular pathology for Classes I, II and III

It is not necessary to list specific disqualifying ocular abnormalities in the standards. If an ocular problem interferes with visual acuity, visual fields, motility, or color vision, or increases intraocular pressure, then it would be covered under one of the other present or proposed standards.

The ocular pathology standards for Class II and Class III medical certificates should be identical to the standard for the Class I medical certificate. If there is an acute or chronic pathological condition of the eye that might interfere with its proper function, might progress to that degree, or might be aggravated by flying, then this condition could be dangerous while performing the duties of all classes of airmen.

An applicant who gives a history of any acute or chronic pathological condition of the eye should be examined more carefully and completely by an ophthalmologist, at which time a decision can be made as to whether the

ocular abnormality is one that might interfere with flying.

o Ocular motility

We recommend no changes in the present standards for ocular motility.

The present ocular motility standards consist of limits for horizontal and vertical heterophorias. A phoria (or heterophoria) is a tendency for the eyes to deviate from each other, and is measured in prism diopters once the eyes have been dissociated. To dissociate the eyes, binocular viewing, or fusion, must be disrupted, which can be accomplished in several ways: an alternate-cover test; presenting the eyes with dissimilar objects, as in the Mattox rod test; and presenting the same object to both eyes, but in different colors, such as the red/white or red/green test.[31]

Some degree of lateral phoria is present in almost 100% of the population. By contrast, a tropia, which is a manifest deviation of the eyes, is present in an estimated 2% of the population. Orthophoria is the very rare situation where both visual axes are perfectly aligned even when fusion is disrupted. Orthophoria may be considered as the physiological ideal, which rarely, if ever, exists. The practical reality is that small degrees of phoria are normal.[31]

The upper limits of normal for phoria values are vague. The following are values given by several authoritative sources:

1. In the Binocular Vision and Ocular Motility Section of the Ophthalmology Basic and Clinic Science Course 1982-1983, prepared

by the American Academy of Ophthalmology, normal limits for phorias are:[32]

Esophoria: 1-2 prism diopters

Exophoria: 1-4 prism diopters

Hyperphoria: 0.5 prism diopter

Testing distance is not specified.

2. In a 1950 edition of the same manual, Saul Sugar gave the following as norms for muscle balance:[33]

   a. At 6 meters:

      Esophoria not greater than 3 prism diopters

      Exophoria not greater than 2 prism diopters

      Hyperphoria not greater than 2 prism diopters

   b. At 33 cm

      Esophoria not greater than 1 prism diopter

      Exophoria not greater than 7 prism diopters

      Hyperphoria not greater than 1 prism diopter

3. In a 1955 discussion of Air Force Vision Standards, Brigadier General Victor A. Byrnes stated that, in the general population, "96% will have less than 4-5 diopters of esophoria or exophoria."[34]

4. David D. Michaels concurs with the opinion that heterophoria of some degree is probably universal. He states that the prevalence of lateral phorias (eso-or exophoria) is probably 86-98%, and approximately 20% for vertical phorias. He states that "it is not possible to attach 'clinical significance' to a particular phoria value, although this is

frequently specified on drivers' and fliers' examinations. Even when the distance is stipulated, it is not the absolute phoria but its relation to the compensating fusional vergence that determines visual efficiency."[31]

Dr. Michaels' comments allude to another problem with phoria measurements: their variability under various testing conditions.

5. The testing method, fixation distance, control or lack of control of accommodation, general well-being or fatigue, as well as hypoxia, can influence the amount of heterophoria.[31]

Fusional Amplitudes

As stated in the recommended Part 67 Medical Standards, motility standards are a part of the certification examination in order to ensure "bifoveal fixation and vergence-phoria relationships sufficient to prevent a break in fusion under conditions that may reasonably occur in performing aviation duties." In other words, the certification procedure is attempting to exclude any individuals who might develop diplopia while flying an airplane. In order to assess adequately an individual's chances of developing diplopia, an examiner needs information on horizontal and vertical fusional amplitudes, as well as phoria measurements. Only by comparing phoria values to fusional amplitudes can an assessment be made of one's ability to compensate for a phoria. Vergence eye movements are used continuously to maintain binocular fixation, and are necessary to prevent horizontal and vertical phorias from becoming tropias. They also assist in changes in position of gaze, as in fixation from distance to near.

The average normal fusional amplitudes, in prism diopters, according to von Noorden, are as follows:[35]

Testing Distance	Convergence Amplitudes	Divergence Amplitudes	Vertical Fusion
6 m	14	6	2.5
25 cm	38	16	2.6

Fusional convergence controls an exophoria, fusional divergence controls an esophoria, and vertical fusional vergence controls a vertical phoria. Fusional vergence amplitudes can be measured by a rotary prism or prism bar. Accommodation must be controlled with either testing method.

Fusional vergences are not absolute values, and they can vary under adverse conditions such as fatigue, illness, or hypoxia. It is well known that exercising the fusional vergence mechanisms with orthoptics increases their magnitude.[35]

Sheard states that fusional vergence amplitudes should be twice the phoria value in order to maintain comfortable single binocular vision.[36] This is certainly not a universally agreed upon absolute. In fact, most authorities emphasize the significance of the relationship between fusional amplitude and heterophoria, but do not state what the optimum relationship between these two values should be.

Thus, by measuring only phoria values, we may or may not be evaluating adequately an individual's capability for maintaining single binocular vision while flying an airplane. However, there are no known instances of flight crew incapacitation in international civil aviation due to ocular motility disorders. In a joint study by the International Civil Aviation Orgnization,

the International Federation of Airline Pilots Associations and the International Air Transport Association of "pilot malfunctions" during the years 1960-1968 and 1969-1973, the only ocular causes for nonfatal incapacitation were one case of eye injury, one case of conjunctivitis, and one case of corneal ulceration. These occurred out of a total of 66 cases of nonfatal incapacitation due to all causes from 1960-1968, and 80 cases from 1969-1973.[37]

The present FAA standards are even more stringent than the US Air Force standards, and compare favorably with other standards as well, as seen in the following table:[38]

Selected Motility Standards

Standards	Allowable Phorias (in diopters)		
	Esophoria	Exophoria	Hyperphoria
FAA	6	6	1
US Air Force	10	6	1.5
US Navy	10	10	1.0*
			1.5**
Belgium	8	6	1
France	6	6	1
United Kingdom	6	8	1
Federal Republic of Germany	5	10	1.5
Canada	6	6	1

* For pilot-candidates ** For pilots

In summary, the present standards have been adequate so far, in that no known problems of diplopia in pilots have been reported. The present standards also seem reasonable when compared with US military and international standards.

Therefore, we recommend that present standards be maintained. Because of the variability of the factors that enter into binocular vision and fusion, it is difficult to recommend any better method of evaluating ocular motility than the present ones. One might recommend performing fusional amplitudes on every candidate and relating these to phoria values. However, only a very small percentage of aviation medical examiners are ophthalmologists who have the equipment to do this. Since approximately 96% of the population should pass the present standards, we feel it is reasonable to examine the remaining 4% more closely by requiring comprehensive ocular motility examinations.

o Intraocular pressure for Classes I, II and III

The measurement of intraocular pressure is not presently required in the standards for any class of medical certificate.

Increased intraocular pressure and glaucoma may appear in an acute, painful, and suddenly impairing form known as angle-closure glaucoma, or in a subtle, painless and progressive form known as chronic open-angle glaucoma. Because either form of glaucoma can be destructive to night vision, contrast sensitivity, and central or peripheral visual fields, they are of major concern to pilots' safety.[39] The prevalence of glaucoma increases with advancing age, particularly after age 40.[40] Infrequently, acquired glaucoma may

appear in early adult life, usually in individuals with a strong family history of glaucoma.[41] Intraocular pressure of greater than 24 mm Hg mercury occurs in less than 0.15% of the US population.[42]

The course of glaucoma is an insidious one, and in most cases it remains asymptomatic until large changes in the visual fields have taken place. Glaucoma is an ideal disease to be approached in a preventive manner, and tonometry has been shown to be an effective screening method for chronic open-angle glaucoma. Because of the impact glaucoma may have on a pilot's career, it is particularly advisable for these individuals to be screened, and to be aware of any elevation of intraocular pressure in the early stage, when visual field damage can be avoided.

Therefore, we recommend that evaluation of intraocular pressure by instrumental tonometry be required for all applicants age 40 years or over. We recommend instrumental tonometry for an applicant at any age if there is a positive family history for glaucoma in a first-degree relative.[43] Aviation medical examiners also examine air traffic controllers, who are required to have tonometry as part of their examination. Therefore, AMEs already have tonometers and are familiar with their use.

References

1. American Academy of Ophthalmology: Retina and Vitreous, Ophthalmology Basic and Clinical Science Course. San Francisco, American Academy of Ophthalmology, 1982, Section 4, pp 92-100.
2. US Air Force School of Aerospace Medicine, Brooks AFB, Texas. Ophthalmology Handout #OPH 09, page 111.

3. Wilmer WH, Berens C: Aviation Medicine in the American Expeditionary Forces. Office of Director of Air Services, Washington Printing Office, Feb 1920, pp 166-167.

4. Dept of Transportation: Taxiway Centerline Lighting System-AC 150/5340-19.

5. Dept of Transportation: Evaluation of Centerline Lighting - Indications for Runway Distance Remaining and Taxiway Exits - AD 656594.

6. Gibbons HL, Lewis MF: A Discussion of Color Signals and Their Flight Safety Relationships to Radio Communications in Civil Aviation. Dept of Transportation, FAA Civil Aeromedical Institute.

7. Bowman KJ, Cole BL: Recognition of the aircraft navigation light color code. Aviat Space Environ Med 1981;52:658-665.

8. Ibid.

9. Tredici TJ, Mims JL, Culver JF: History, rationale, and verification of color vision standards and testing in the US Air Force. Ophthalmology Branch, USAF School of Aviation Medicine, Brooks Air Force Base, Texas 78235.

10. Dille JR, Booze CF: The 1976 accident experience of civilian pilots withs tatic physical defect. Aviat Space Environ Med 1980;51:182-184.

11. McCann JP, Pan American World Airways, personal communication.

12. Wick RL, American Airlines, personal communication.

13. Gibbons and Lewis, op cit.

14. Bailey RW: Color vision deficiencies in Army fliers. USAARU Report #65-2, April 1965.

15. Pauson HM: Comparison of color vision tests used by the military services. Naval Submarine Medical Research laboratory, Naval Submarine Medical Center, Groton, CT 16340.

16. Cole BL, Vingyus AJ: A survey and evaluation of lantern tests of color vision. Am J Opt and Phy Optics 1982;59:346-374.

17. Sloan LL: Selection of color vision tests for the Army Air Forces. Arch Ophth 1946;36:263-83.

18. Tredici Col TJ: Visual standards for flying in the USAF. USAF, MC, Ophthalmology Branch, USAF School of Aerospace Medicine, Brooks AFB, Texas.

19. Titmus Vision Tester Manual, Titmus Optical Company, Petersburg, Virginia.

20. Keystone Orthoscope/Telebinocular Manual 73-5402, Keystone Company, 2212 East 12th Street, Davenport, Iowa 52803.

21. Ishihara S: A series of plates designed as a test for color blindness. Konehara Shuppon Co, Ltd, Tokyo Japan.

22. Fronceschetti A, Francois BJ: Chorioretinal Degenerations. Springfield IL, Charles C Thomas Co, pp 49-55.

23. Hardy LH, Rand G, Rittler MC: AO-HRR pseudoisochromatic plates, 1957. American Optical Co, Cat #13398.

24. Cruz-Coke, MD: Color Blindness: An Evolutionary Approach. Springfield IL, Charles C Thomas Co, 1970, p 69.

25. Dvorine I: Quantitative Classification of the color-blind. J Gen Psy 1963;58:255-265.

26. Dvorine I: Preliminary report on the diagnostic efficiency of the second edition of the Dvorine pseudoisochromatic plates. Am J Opt and Arch Amer Acad Opt 1955;32:259-261.

27. Jones KN, Steen JA, Collins WE: Predictive validities of several clinical color vision tests for aviation signal light gun performance. Aviat Space Environ Med 1975;46:660-67.

28. Personal communication, Mr. Powel, Richmond Products, 4089 South Rogers Circle, Suite 6, Boca Raton, FL 33431.

29. Sloan LL, Habel AB: Tests for color deficiency based on the pseudoisochromatic principle. Arch Opth 1956;55:229-239.

30. Harrington DO: The Visual Fields, St. Louis, CV Mosby Co, 1976, pp 97-104.

31. Michaels DD: Visual Optics and Refraction, St. Louis, CV Mosby Co, 1975, pp 304-309.

32. American Academy of Ophthalmology: Binocular Vision and Ocular Motility, Ophthalmology Basic and Clinical Science Course. San Francisco, American Academy of Ophthalmology, 1982-83, Section 6, pp 59-60.

33. Sugar H, Saul MD: The Extrinsic Eye Muscles, Home study Course. American Academy of Ophthalmology and Otolaryngology, 1950, pp 28-29.

34. Byrnes, Brig Gen VA: Discussion of papers by Major JJ Claro and Dr HW Rose Agarda. USAF School of Medicine, 1955.

35. von Noorden GK: Burian-von Noorden Binocular Vision and Ocular Motility, St. Louis, CV Mosby Co, 1980, p 205.

36. Michaels DD, op cit, pp321-338.

37. Manual of Civil Aviation Medicine, Part III, ch 2, Flight Crew Incapacitation.

38. Tredici Col TJ, op cit.

39. Kolker AE, Hetherington JH: Becker-Shaffer's Diagnosis and Therapy of the Glaucomas, St. Louis, CV Mosby Co, 1976, pp 59-68.

40. Ibid, p 220.

41. Ibid, p 223.

42. Ibid, p 59.

43. Ibid, p 61-68.

Ophthalmologic Evaluation Forms

- "Near Vision Acuity" Form 8500-1

  The FAA should keep Form 8500-1 as the standard test chart for near visual acuity. It is calibrated with Snellen letters that are held at 16 in to correlate with a linear Snellen 20/20 scale. It is useful for testing near

vision at both 16 in and at 32 in.

- "Report of Eye Evaluation" Form 8500-7

  This form is acceptable, except that there should be a space for family history, especially family history of glaucoma.

- "Ophthalmological Evaluation of Glaucoma" Form 8500-14

  This form is comprehensive and requires no change.

- "Defective Color Vision" Form 8500-9

  Under the recommended standards this form would be appropriate only for Class III pilots.

- "Defective Near Vision" Form 8500-0

  This form should be changed to: "Holder shall wear correcting glasses for near vision while exercising the privileges of his or her airman certification."

Discussion of Specific Diseases or Ocular Conditions

Based on our discussion with personnel from the Aeromedical Certification Branch in Oklahoma City, certain problem areas with regard to certification and special issuance for applicants with ocular abnormalities were delineated:

1. One-eyed pilots. It is estimated that there are 500 to 800 one-eyed pilots, and in some of these individuals ocular pathology exists in the "good eye."

2. Glaucoma or cataract with decreased visual acuity. The FAA requested guidance in the disposition of individuals with cataract, particularly with reference to the different types of cataract and their potential impact on

visual acuity.

FAA also requested guidance for determining when an abnormal visual field due to glaucoma was severe enough to preclude certification, as well as advice on which glaucoma medications may interfere with vision and those which are considered safe to be used by a pilot.

3. Identified retinal pathology. FAA requested guidance on the disposition of these individuals and recommendations for the frequency of ophthalmologic follow-up examinations.

4. Radial keratotomy. Approximately 300 applicants have had this procedure, and the FAA requested guidelines for disposition.

o Cataract

A cataract is a defect in the transparency of the crystalline lens, which may be stationary but is usually progressive at a variable rate. The effect on visual acuity depends on location and severity of the opacity. It is the most common and one of the most easily treated causes of visual impairment.

The location of the cataract is either nuclear or cortical. There is a form of cortical cataract that is posterior and subcapsular in location. Advanced cataracts frequently have a combination of nuclear, cortical and posterior subcapsular locations.

The severity of the opacity is usually graded on a 1+ (mild) to 4+ (advanced) scale. The color change of nuclear cataracts is also described. In general,

when the nuclear opacity is 1+, the color change is white or yellow, and it advances to a yellow-brown, or brunescent color, as the opacity progresses.

The normal lens gradually hardens and acquires more pigment with age. The transition from a normally aging nucleus to a nuclear cataract can be subtle, and this is manifested primarily by change in visual function rather than by any observable microscopic change.

The first change is usually a myopic shift in refractive error. Later, vision becomes blurred, and there is a gradual decline in best correction of visual acuity. Usually distance vision is impaired earlier than near vision. The usual course of nuclear cataract formation is a gradual decline in visual acuity, which may cause poor color discrimination and, occasionally, monocular diplopia. Visual acuity may be slightly worse in dimly lit situations. Persons with nuclear cataracts usually do not complain of problems with glare, or with bright lights.[1]

We recommend that Class I or II applicants with nuclear cataracts have their near and distant visual acuity and color vision checked yearly, if the best corrected acuity is 20/20. If the best corrected acuity is not 20/20, then follow-up should be required every 6 months. Class III applicants who meet the vision standards should be checked yearly.

The most significant and common type of cortical lens opacity is the posterior subcapsular cataract. These can occur at any age, and they are frequently associated with the nuclear lens changes of older individuals. As isolated lens opacities they occur in middle-aged persons more frequently

than any other form of cataract. When the opacity involves the pupillary zone, visual acuity can be drastically diminished, especially in bright sunlight and conditions with glare.[1] Because of the extreme variability of vision under different lighting situations with this type of lens opacity, it is very important to measure both near and distant visual acuity when the person's pupils are not dilated. The person's vision should also be measured in bright lights, as well as in the usually dimly lit examination room. This can be done by checking acuity while shining a penlight at the eye being tested, in order to induce pupillary constriction. These individuals should be re-tested every 6 months.

The following case from Rubin's Optics for Clinicians underscores the variability of vision with posterior subcapsular cataracts: "A healthy 45-year-old prison guard complains of a gradual decrease in his visual acuity over the past year, but interestingly, only when working in bright sunlight...A check of the guard's acuity in your refraction lane reveals 20/20 OU with his present corrective lens...A more careful check finds substantiating acuity of 20/400 OU in bright light levels, and your slit lamp examination confirms your suspicions of small posterior subcapsular cataracts OU."[3]

Cortical lens opacities in locations other than the posterior subcapsular region are not usually significant, unless they are associated with changes in the nucleus of the lens. However, when these opacities are irregular and whitish, they may scatter and absorb light, especially under bright illumination, resulting in decreased acuity. The most common type of cortical change is the separation of lens fibers by clear water vacuoles, which later evolve into cloudy liquid. The configuration of the opacification

is usually spike-like, aligned radially among the lens fibers. The opacification is usually concentrated peripherally rather than centrally.[1] Class I or II applicants with cortical cataracts should have near and distant acuity examined annually in both dim and bright lights. If best corrected acuity is not 20/20 at distance and near, follow-up examination should be performed every 6 months. Class III applicants who meet the vision standards should be tested yearly.

Modern microsurgical techniques of extracapsular cataract extraction with intraocular lens implantation yield one-year follow-up results of 20/40 or better best-corrected visual acuity in over 90% of cases who do not have other peroperative eye pathology or posteroperative macular degeneration.[4] With such a high surgical success rate for the treatment of cataracts, it does not seem justifiable to allow pilots with visually significant cataracts, whose vision may be extremely poor under the certain lighting and glare situations encountered in aviation, to continue as pilots.

1. Duke-Elder S: System of Ophthalmology. London, Henry Kimpton, 1969, Vol XI, p 63.
2. Duone TD: Clinical Ophthalmology. New York, Harper and Row, 1976, Vol 1, Chapter 71-73.
3. Rubin ML: Optics for Clinicians. Gainesville, FL, Triad Scientific Publishers, 1974, p 349.
4. Start WJ, et al: The FDA report on intraocular lenses. Ophthalmology 1983;311-317.

o Retinal Abnormalities

Central retinal vein obstruction (CRVO) produces symptoms from mild peripheral visual blurring to severe visual field loss. Varying signs on examination are present from marked venous engorgement to a "blood-and-thunder" hemorrhagic retinopathy. These variations are most likely attributable to differing degrees of occlusion in the central retinal vein and central retinal artery. The etiology is most likely atherosclerotic vascular disease and diabetes, especially when associated with aging; however, CRVO has also been related to congestive failure, hypotension, blood dyscrasias, emphysema, polycythemia, multiple myeloma, and dysgammaglobulinemias. Prognosis for vision is usually quite poor in the extensive venous obstruction pattern, and it is variable in the less-obstructed patterns. Persons with CRVO must be observed closely for development of neovascular glaucoma during the first year of follow-up. Once stable, after one to two years, the pilot with CRVO should not require further FAA specialist examinations. However, he or she may have varying degrees of peripheral field loss in the affected eye.

Branch retinal vein occlusion (BRVO) occurs three times more frequently than central retinal vein obstruction. It is frequently seen in persons with longstanding hypertension, but the exact pathophysiology remains controversial. The prognosis for visual recovery in BRVO is somewhat better than for CRVO occlusion. Visual acuity usually improves in approximately 60% of persons with BRVO, and three studies demonstrated visual acuity of 20/40 or better in approximately 50% one year after the occlusion. This reduced acuity is usually a result of macular edema and usually remains stable on long-term follow-up. Once stable, the patient should not need

further examinations by eye specialists for recertification unless new symptoms develop.

Central serous retinopathy is spontaneous detachment of the macula occurring in the 25 to 50 year old age group. This produces variable reduction of acuity, from mild distortion of central vision to 20/60 or 20/100. Recovery is usually spontaneous, but recurrences are frequent and the brief attacks usually cause no lasting damage to vision. However, prolonged or recurrent disease may cause significant long-term central visual loss. Forty percent of individuals with central serous retinopathy recover completely within one month. Approximately 20% of the patients may have the condition for over 6 months. The duration of the disease and incidence of further attacks may be shortened considerably with focal laser photocoagulation. Repeated checks of an asymptomatic individual are not necessary.

Disciform macular degeneration and dominant macular drusen, a form of hereditary drusen, has an autosomal dominant form of transmission and may be seen in both eyes between the ages of 20 and 30. However, vision is minimally affected, if at all, and there is an excellent prognosis for vision. No special follow-up is needed.

Serous pigment epithelial detachment (PED) is a localized detachment of the pigment epithelium of varying size with distinct edges. This produces a varying degree of visual loss, which may completely subside and revert to essentially normal central vision. The duration of symptoms and remission is variable. PED occurs in conjunction with, or is a percursor to, neovascular

disciform macular degeneration. A person whose PED is subsiding and whose vision is stable should not need further specialist examinations for recertification unless new symptoms develop.

Hemorrhagic detachment of the pigment epithelium is associated with new subretinal vessels of disciform macular degeneration. Visual prognosis is poor if the detachment affects the central macular region.

Disciform macular degeneration (DMG) is an aging process characterized by varying combinations and degrees of serous and hemorrhagic detachments of the pigment epithelium and sensory retina, often leading to loss of central vision due to the formation of a central hypertrophic scar. When the condition is off center, it may be remedied by laser photocoagulation. This is a temporary measure, as it only treats the disease process at that particular time, and recurrences are common. The person's other eye has a 12% to 15% per year risk of developing DMG if the preliminary changes of drusen and pigment epithelial disruption are present. Continued ophthalmologic follow-up is recommended. Testing visual acuity is a good method of identifying active disease.

Presumed histoplasmic choroiditis is a macular disease affecting individuals from 15 to 50 years of age that is characterized by stages of remissions and exacerbations. It is symptomatic only when a neovascular process is underway adjacent to the fovea. The outcome may be improved by laser photocoagulation if the associated neovascular process is not central. Once a scar has developed with no evidence of persistent neovascularization, the vision should be stable unless another acute episode develops centrally.

Once stable, an individual should not need further specialist examinations for recertification unless new symptoms develop.

The characteristic feature of surface wrinkling retinopathy, also known as cellophane maculopathy or macular pucker, is wrinkling of the internal limiting membrane of the retina on or near the macula. It is usually associated with aging and is more common in the 50 to 60 year-old age group. Patients usually complain of minor loss of vision with distortion, and have visual acuity in the 20/30 to 20/100 range. More severe cases of macular pucker are occasionally seen with worse than 20/200 acuity. Progression in this disease is usually quite slow, and these persons will be stable for many years. Close follow-up is not necessary. For the more severe cases surgical therapy is probably indicated.

Macular holes, lamellar holes and macular cysts are losses of a circular area of retinal tissue in the macular region that vary in thickness from a portion of the retina to full thickness macular holes. They are associated with a variable loss of central vision. These are usually considered to be related to aging, and once they occur the visual acuity is usually very stable. Treatment is not indicated. Once stable, an individual should not need further specialist examinations for recertification unless new symptoms develop.

Retinoschisis can be divided into senile degenerative retinoschisis and juvenile sex-linked retinoschisis.

Typical senile degenerative retinoschisis is present in approximately 1% of adults; 33% of these people have bilateral disease. It is usually seen far to the periphery and remains quite stable throughout the aging process, rarely resulting in significant visual field change and even less frequently in full thickness retinal detachment. Typically, it does not extend posteriorly to threaten the macula, and it rarely requires treatment. A person without secondary retinal detachment or significant visual field loss should not require repeated follow-up by a specialist.

Juvenile sex-linked retinoschisis is a vitreoretinal dystrophy that produces extensive splitting of the peripheral retinal layers, causing peripheral visual field defects. Almost 100% of these cases have associated macular foveal schisis; and many patients have normal central vision. However, the overall prognosis for central vision remains guarded, with the final visual acuity often diminishing to 20/200.

Pavingstone degeneration of the retina is characterized by one or more discrete, round foci of depigmentation and retinal thinning located in the far retinal periphery. It is present in approximately 17% of the population. The prevalence increases markedly with advancing age. Pavingstone does not predispose to retinal breaks or retinal degeneration and is basically a benign condition.

Lattice degeneration is typically a sharply demarcated, linear degenerative process in the retinal periphery characterized by retinal thinning and accompanied by abnormalities in the overlying vitreous. The extent of involvement varies according to the size and number of lesions, from small

islands to a lattice that completely encompasses the periphery of the retina. Lattice degeneration is present in 11% of individuals at autopsy and is bilateral in 48% of those. Lattice degeneration may be responsible for retinal detachment when round holes within the degenerated areas permit a detachment to begin and progress. The great majority of persons with lattice degeneration do not develop retinal detachment and, therefore, continued ophthalmic evaluation of this condition in asymptomatic patients is unnecessary.

Retinal holes of both round and horseshoe types are most likely to remain stable if they have been present for a long time, there is no evidence of detachment and the individual is asymptomatic. There is considerable debate regarding the necessity of treating these holes. Most likely, if the retinal holes are stable, they have very little chance of progression. If the holes are treated with either laser photocoagulation or cryopexy, the chance of extension or retinal detachment is almost nil. Round asymptomatic retinal holes are present in approximately 0.4% of adults and retinal tears are present in 3.3% of persons at autopsy. A pilot with stable retinal holes should not need further specialist examination for recertification unless new symptoms develop.

The vitreous gel normally is attached to the retinal surface in broad, loose attachments at the optic nerve, macula, retinal vessels and ora serrate. Frequently during the aging process the posterior attachments separate and cause symptoms of flashing lights and "floaters." This process, known as posterior vitreous detachment, usually evolves over a one-to two-month period. An examination must be done to evaluate the possibility of

associated retinal tears. If symptomatic retinal tears occur with this condition, prophylactic laser or cryopexy treatment is necessary. If a retinal tear does not occur over this time period, the individual has very little chance of having such a tear in the future and can be deemed stable unless new symptoms develop. Once stable, a pilot with posterior vitreous detachment should not need further specialist examinations for recertification unless new symptoms develop.

Rhegmatogenous retinal detachment stems from the development of a retinal tear with fluid passing through the tear to elevate the retina. The overall incidence of retinal detachment varies from 0.005% to 0.01% per year. Detachment is present in 1-3% of persons with aphakia. Once a retinal detachment is treated with a successful scleral buckling procedure with adequate retinopexy scarring, that is, scarring of the retina to the outer pigment epithelium, the condition is most likely to be stable. Chances for redetachment 6 to 12 months later are small. In cases of uncomplicated retinal detachment with excellent recovery of visual acuity, continued examinations for recertification are not necessary as long as the condition is deemed stable by an ophthalmologist through a period of one year. Peripheral visual fields may be affected if the retina was detached for an extended period or if extensive retinopexy scarring was present. A careful examination of the visual fields is advisable.

Spontaneous vitreous hemorrhage in the nondiabetic patient most likely is due either to posterior vitreous detachment with avulsion of a small retinal vessel, which is possibly associated with a retinal tear, or to associated occlusive vascular disease. Vitreous hemorrhage is usually worked up by

ocular echography to identify signs of retinal detachment. If no signs of detachment are present, the hemorrhage is periodically observed. If no clearing occurs, pars plana vitrectomy surgery to remove the hemorrhage may be indicated. When the hemorrhage clears and the retina appears intact without evidence of vascular disease, the condition is most likely to remain stable. The periphery of the retina must be examined for evidence of retinal tears or holes, and these should be treated appropriately. If occlusive vascular disease, such as central retinal vein occlusion or branch vein occlusion, were the etiology for the vitreous hemorrhage, these conditions would have to be treated appropriately.

Diabetic retinopathy can be divided into the classifications of background and proliferative. Background retinopathy can be regarded as a stable, or as a very slowly progressing condition in which sudden changes in visual acuity are highly unlikely. The principal impairing visual problem of background retinopathy is macular edema resulting from vascular leakage. This macular edema is a slowly progressive disorder, and results of treatment by laser are poor. Proliferative retinopathy may be regarded as a more active, advanced condition that is likely to lead to sudden visual change as a result of vitreous hemorrhage. It is associated with the development of new pre-retinal blood vessels on the optic nerve and in the retinal periphery. This is a dangerous ocular condition, because the vessels may bleed spontaneously; this may lead to preretinal or vitreous hemorrhages, and further vitreous contraction and retinal detachment are possible. This condition may be helped by laser photocoagulation. Yearly ophthalmic examinations are indicated.

o Refractive surgery

There have been a variety of approaches to refractive eye surgery over the past 200 years, such as lens modification, eye shortening, corneal stroma modification, corneal releasing incision, anterior corneal grinding, corneal thermal procedures, corneal intralamellar lenses, and epikeratophakia.[1] Presently, there are several refractive surgical techniques, including radial keratotomy, epikeratophakia or keratophakia, and keratomileusis.

Radial keratotomy consists of making deep radial cuts in the cornea, sparing a 3 to 4 mm central optical zone. The number of cuts may vary, but usually it is 8 or 16. As the cuts heal and scar the central cornea flattens, decreasing the axial length of the eye and the degree of myopia.

Studies presently available are not adequate to assess fully the risks and long-term effects of radial keratotomy. The study with the longest follow-up (five years) is not adequate in design, because the surgeon is the same individual who performed the follow-up examinations. The National Eye Institute has organized a study, the Prospective Evaluation of Radial Keratotomy (PERK Study), which is well-designed and allows for independent follow-up evaluation of patients by an observer other than the surgeon.[2] Results of this study are pending.

Surgical results with radial keratotomy are variable. Stability of postoperative refraction is also variable, especially in the first few months, when night glare and fluctuating vision are significant complaints. Glare results from diffraction of light by the corneal scarring along the incision lines. Fluctuations in vision are probably related to diurnal changes in the

intraocular pressure and their effects on the shape of the healing cornea. In the early postoperative phase, significant glare has been reported in 50% to 79% of cases and significant visual fluctuation in 33% to 60% of cases.[2] These complications decrease with longer follow-up, but they are not transient in many cases. In a recent study of 290 patients, 26% reported mild to moderate problems with glare, and 23% reported mild to moderately fluctuating vision one year later. At one-year follow-up, 40% of these patients had uncorrected distance acuity of 20/20 or better, and 83% had uncorrected acuity of 20/40 or better. Ninety-one percent of these patients had a best-corrected visual acuity of 20/15 to 20/20.[3]

Ninety one percent of individuals who have undergone radial keratotomy would meet the visual acuity standards for a Class I certificate. However, approximately one-fourth of these persons have some mild to moderate difficulty with glare, visual fluctuation, and night driving. These are subjective phenomena that cannot be detected or quantified with present-day examination techniques. For this reason, persons who have undergone radial keratotomy should be excluded from obtaining Class I or II certificates. Those individuals applying for Class III certificates should not be certified until at least one year after radial keratotomy. Visual acuity and refraction should be stable when tested repeatedly over an eight-hour time period. The applicant must be queried about difficulties with glare and night driving. If these symptoms are present, the applicant should be denied certification.

These recommendations are in accordance with the recommendations of the National Advisory Eye Council of the National Eye Institute, which has urged

restraint by both patients and ophthalmic surgeons in using radial keratotomy until the long-term results of controlled clinical trails are known.[2]

In keratophakia and epikeratophakia a lathe-cut donor corneal button is sutured either within the recipient's corneal stroma (keratophakia), or on the de-epithelialized surface of the recipient's cornea (epikeratophakia). Both procedures are highly experimental and are utilized primarily in unilaterally aphakic children who are not tolerant of contact lenses.[1]

Preliminary data suggest the epikeratophakia may have some application in myopia and keratoconus, but this research is in its infancy and not clinically significant at this time.[4]

Keratomileusis was developed over 25 years ago by Jose Barraquer in Bogota, Colombia. The technique requires very sophisticated instruments and extensive training and experience by the surgeon to be performed safely. Recently, there has been increased interest in keratomileusis in the United States, and there are now a few American ophthalmologists who are performing this procedure.

The surgical technique consists of using a specially-designed microkeratome to shave off the central outer layers of the patient's cornea. This corneal tissue is frozen, placed on a cryolathe, and a lathe-cut to the predetermined shape for correction of the patient's refractive error. The tissue is then thawed, and resutured to the patient's cornea. Keratomileusis can be used for hyperopia, but it is primarily used for correction of myopia between 6

and 18 diopters.[5]

Most complications occur during the operation and involve irregular cutting of the lenticule by the keratome, or damage to the tissue by the cryolathe. In a few cases, there has been opacification at the tissue interface and a resultant decrease in visual acuity.

Although keratomileusis is effective in reducing refractive errors, the results are rather unpredictable. At a mean postoperative follow-up of 5.5 years, Swinger and Barraquer reported improvement of the preoperative myopic refractive error in 51% of 85 patients.[6] One advantage of keratomileusis is the absence postoperatively of glare and visual fluctuation.

If an individual does get a good result, it seems to be stable. Therefore, we recommend certification of applicants at least one year after keratomileusis, with best-corrected visual acuity that meets the standards, and with no other ocular pathology.

1. Binder PS: International Ophthalmology Clinics. Boston, Little, Brown and Company, 1983, Vol 23, No 3, pp 1-9.
2. Bores LE, Stark WJ, et al: Radial keratotomy. Survey of Ophthalmology, 1983; 28(2):101-111.
3. Dietz MR, et al: Radial keratotomy: An overview of the Kansas City study. Ophthalmology 1984;91:467-478.
4. Binder PS: op cit, pp 45-58.
5. Ibid, pp 75-92.
6. Ibid, pp 167-192.

o Glaucoma

Although both surgery of the iris and laser iridotomy are highly effective in the control of acute congestive or angle-closure glaucoma, there is a continuing dispute about the medical and surgical management of open-angle glaucoma. Traditional drugs used in the control of glaucoma such as pilocarpine, eserine, carbachol, phospholine iodide and others, have had an undesirable side effect of reducing pupillary diameter and inducing myopia. These drugs, instilled topically in the eye, interfere with needed pupillary enlargement under reduced illumination, which may lead to fibrosis of the iris and reduced mobility of the pupil. Pilots would be at a visual disadvantage at night or in dark, cloudly weather, and in processing remote visual cues when their pupils are constricted by the miotic drugs. Fortunately, there are other effective drugs for the reduction of seriously elevated intraocular pressure: the levo-epinephrine group of drugs (Epifrin, Epinal, Epitrate, Glaucon, Eppy-N, Propine) and the beta adrenergic blocking drugs such as timolol (Timoptic).

Normal intraocular pressure is generally between 15 and 25 mm Hg, though occasional persons tolerate pressures slightly over 25 mm Hg. When intraocular pressure is elevated above 25 mm Hg or when the pressure gradient in the two eyes is larger than 5 mm Hg, the person has either pre-glaucoma or open-angle glaucoma. Normal visual function may be maintained when there is no pathologic enlargement of the optic cups or visual field defect, including vertical enlargement of the blind spot, scotomata in the arcuate distribution of optic nerve fibers above or below the macula, or small nasal step defects.

Pilots who have a diagnosis of primary glaucoma, but whose pressure is completely controlled by surgical or laser therapy without optic nervehead or visual field damage, may receive clearance for flying, but they must be re-examined annually. If open-angle glaucoma has been well controlled by the topical use of sympathomimetic drugs and/or beta adrenergic receptor blocking agents, airmen may be certified for flying if there is no optic cup or visual field damage. The use of miotic medications or systemic carbonic anhydrase inhibitors such as Diamox, Daranide or Neptazane, is grounds for denial of medical certification.

o Anisometropia

Anisometropia is the term applied to the condition wherein the refractions of the two eyes are unequal. Mild anisometropia is a common condition, especially if astigmatic errors are present. More severe anisometropia may affect the vision in one of three ways: 1) vision may remain binocular; 2) vision may alternate from one eye to the other or; 3) vision may be exclusively uniocular with complete suppression and amblyopia in one eye.

Generally, a difference in the refraction between the two eyes of greater than 2.50 diopters is probably the limit of tolerance for binocularity, although in rare instances binocular vision has existed with a difference of 5.00 or 6.00 diopters. Each 0.25 diopter difference between the refraction of the two eyes causes a 0.5% difference in size between the retinal images, and it is believed that a difference of 5%, which would occur at 2.50 diopters, is probably the limit that can be tolerated. This is not an absolute rule, because one must know how much of the difference is due to the axial

component and how much is due to the refractive component before one can arrive at a definite percentage difference in size of the images on the retina. A more simple and direct test for binocularity, such as a test for stereopsis, would be more significant from a performance standpoint than consideration of the factors mentioned above.

If the difference in acuity between the two eyes is above where binocular vision can be maintained, then the individual may alternate vision with each eye being used one at a time. This is apt to occur when both eyes have good and/or nearly equal visual acuity. In fact, this type of anisometropia, in which one eye is myopic and can be used for near vision and the other eye is emmetropic or hypermetropic and can be used for distance, may even be considered advantageous. This individual would not have true binocularity or stereopsis. If the acuity in one eye is poor, then that eye may be excluded completely from vision at an early age in life; that is, this eye becomes amblyopic and the individual relies on the opposite, better eye to see. Anisometropia is an important factor in the development of strabismus in children.

The present FAA examination does not include tests for stereopsis, and the proposed revisions eliminate refractive error limits and use visual acuity as the limiting standard. Thus the topic of anisometropia becomes academic; the AME gathers no information to diagnose it, and there is not a performance test to detect this condition.

This leads to a discussion of the frequency of this condition. Anisometropia is the rule rather than the exception. However, until one has about a 1.50

diopter difference, the anisometropia is not clinically significant. A difference in the refractive error of 1.00 to 1.25 diopters occurs in about 7% to 8% of a randomly selected population; of 1.50 to 2.00 diopters in about 3%; and of 2.00 diopters and over in about 1.5%. Therefore, a 2.50 diopter difference occurs in less than 1% of the population. Present US Air Force regulations limit the amount of anisometropia for Class I entrance into pilot training to a 2.00 diopter difference. However, the refractive standards for entrance are quite strict, ranging from plano to +2.00 diopters of hypermetropia. It is possible to waive an individual into training with greater than that amount of difference if he or she passes a performance test, such as a test of stereopsis either by the orthorater or other vision screeners, or the Verhoeff depth perception test.

Individuals with high anisometropia resulting in amblyopia can be considered monocular. These individuals, however, still use the peripheral field of the poorer eye, which is obviously useful in aviation. Therefore they should be able to perform adequately. Individuals who are monocular with loss of central vision due to causes such as trauma, retinal problems such as central serous retinopathy, and macular degeneration, still have the benefit of peripheral vision, and in these cases their loss is simply one of stereopsis. Stereopsis is good to have and to use in flying. Yet it has been shown that one can adequately ascertain position and depth in a three-dimensional space with clues other than stereoscopic vision.

The monocular pilot who has lost an eye does have the disadvantage of also having lost the peripheral field on that side. However, the FAA has long allowed one-eyed pilots to demonstrate their ability to compensate for the

loss and perform airman duties.

The present standards for monocularity are acceptable and should be modified only slightly: uncorrected distant visual acuity should not be worse than 20/200, and the 3.50 diopter refractive error limit should be deleted. Individuals whose refractive error is greater than 4.00 diopters will not be able to see 20/200 and therefore would fail this part of the standards. Certification should be granted only on special issuance after a thorough ophthalmologic examination.

References on Refraction

Borish IM: Clinical Refraction, ed 34. Chicago, Professional Press Inc, 1970.

Duke Elder S: Practice of Refraction, ed 9. New York, Churchill Livingston, 1978.

Milder B, Rubin ML: The Fine Art of Prescribing Glasses. Gainesville, FL, Scientific Publications, 1978.

Michaels DD: Visual Optics and Refraction: A Clinical Approach, ed 2. St. Louis, CV Mosby Co, 1978.

Reproductive-Urinary System

Renal Calculi

An applicant with a renal calculus or history of a renal calculus should have complete studies to determine the etiology and prognosis prior to issuance of a medical certificate. These studies should be left to the discretion of a qualified specialist and should include an excretory urogram and metabolic studies to include serum calcium, uric acid, phosphorus and creatinine, and 24-hour urine measurements of calcium, uric acid, oxalate, phosphorus and urine pH.

The factors in the history that should be documented are the number of times stones were passed during the past 6 months, history of surgery for stone disease, history of medical treatment for stone disease, history of gastrointestinal bypass surgery, and composition of the previously passed urinary calculi.

The applicant should be disqualified only under the following circumstances:

1. If there is a urinary calculus that causes pain, urinary obstruction, or urinary infection;
2. If there is an uncorrected metabolic abnormality that is related to stone formation;
3. When there are two or more episodes per year of passage or removal of urinary calculi.

The applicant should not be disqualified for stones that have not resulted in pain, obstruction or fever during the preceding 6 months, or stones that are incarcerated within the renal parenchyma or caliceal diverticula.

Most series have estimated the prevalence of renal stones in the general population to be about 6%. The greatest prevalence is found in young males, who also are the majority of the population of pilots. At the present time, it is estimated that

there are approximately 300,000 licensed civilian pilots. If they are representative of the population as a whole, it follows that 6%, or 48,000, would have stones. In addition, there are about 24,000 Air Force pilots and similar numbers in the Navy and Army. This would seem to provide an ample population for problems to have surfaced if they actually existed.

We contacted several sources in an attempt to find such problems, including the Air Force Inspection and Safety Center at Norton Air Force Base in California, the chief medical officers of three major airlines, and the National Transportation Safety Board (NTSB). The NTSB data bank from 1964 to the present had no record of aircraft incidents caused by kidney stones. That is, to date no one has been able to document a single case in which renal colic was found to be the cause of a crash or collision.

Although there is one case in which an autopsy revealed the presence of a small stone in the ureter of a pilot who was killed along with his entire family on a cross-country flight, the pilot had mild bilateral hydronephrosis and was reported to have back pain for several hours prior to the flight. If this truly represents a crash secondary to renal colic, it is the only one of which our sources are aware, and it would serve to substantiate the fact that unlike heart attack, stroke, and syncope, symptoms of renal colic rarely strike suddenly. There is usually a gradual onset of back, flank, or abdominal pain over an hour or more before acute colic. This is usually plenty of time for the pilot to decide not to take off, or if already in the air, to land. Obviously, the pilot must be aware of the cause of the back pain. Thus, the pilots at greatest risk are those who do not know they have stones, and who give a negative history. It is the lack of knowledge of the condition, rather than the condition itself, that puts the pilot at risk.

Unlike strokes, heart attacks, peptic ulcer disease, and certain forms of asthma and psychiatric disorders, stone passage is not related to stress. Thus, the flight environment does not increase the likelihood of incapacitation from the passage of a stone.

In spite of the risk of sudden incapacitation, over 1300 pilots have been returned to the cockpit after already having had one myocardial infarction. Similarly, over 400 pilots who have had coronary artery bypass surgery have returned to flying status, although their risks of sudden incapacitation and death have not diminished greatly as a result of the surgery.

Unfortunately, urologists often have been put in the position of having to make a statement regarding the likelihood of stone passage. This is very inexact in all but a minority of cases. The physician's true responsibility should be the identification of the stone and education of the pilot about his or her condition. A pilot with a history of renal stones should be taught what the symptoms of renal colic are, so that he or she would know not to take off, or if in the air, to land the aircraft as soon as possible.

Renal Dialysis

Persons on hemodialysis should be disqualified to fly. Whereas most persons on dialysis are stable metabolically and are able to carry out normal activities, they should not be allowed to operate an aircraft for the following reasons:

1. All of these individuals are significantly uremic, having serum creatinine levels of greater than 2.5 mg/dl. Such levels are associated with significantly high risks of electrolyte abnormalities, such as hypo- and hyperkalemia, hypo- and hypercalcemia and hypo- and hypernatremia, all of which can cause changes in awareness and neuromuscular coordination, and increase the likelihood of sudden, incapacitating cardiac or neurologic events.

2. Dialysis is usually done at the point at which aberrations in blood chemistry begin to pose a threat to the patient in his or her normal activities. This is far beyond the threshold of aviation safety. This is consistent with the

prohibition on flying after drinking or after having ingested any medications that produce central nervous system effects. It is quite conceivable that a pilot would operate his or her aircraft enroute to dialysis treatment, at which time his or her metabolic status would be at its worst.

Genitourinary Malignancy

Genitourinary malignancy, even if metastatic, that is adequately treated and documented to be stable, should not be disqualifying. Current treatments for most genitourinary malignancies result in a sufficiently high cure rate and/or stabilization that most persons can live many years without disability. The medical certification branch should request a report every 6 months documenting the stability of the malignancy.

Musculoskeletal System

- The medical flight test for musculoskeletal disorders.

Provided all other aspects of the medical examination would qualify the applicant for issuance of a medical certificate, an applicant with a physical limitation due to a musculoskeletal disorder may be authorized to undergo a medical flight test (MFT). The MFT is usually done in conjunction with the flight test for a pilot's certificate. Should the pilot not satisfy all nonmedical requirements for a flight or if he or she already holds a pilot's certificate but requires a medical flight test to evaluate a deterioration in physical condition, only the medical flight test will be administered.

The AME should defer issuance of a certificate to a person with a musculoskeletal disorder unless the otherwise qualified applicant is applying for student pilot status. In this situation the AME may issue a certificate bearing the limitation: "Valid for Student Pilot Purposes Only," which will permit the applicant to proceed with flight training until such time as a private pilot flight test is appropriate. The applicant should then request through the FAA an MFT in conjunction with the regular flight test. This affords the student an opportunity to demonstrate ability to control the aircraft despite physical limitations.

The MFT consists of procedures specifically designed to ensure safe operation of the aircraft despite the physical limitations of the applicant. The applicant will be tested under marginal or simulated marginal conditions to ensure satisfactory operations in emergencies, in adverse weather, at twilight, and at night.

The applicant may use special external devices or prostheses to assist in the control of the aircraft. The controls of the aircraft may be modified to compensate for physical limitations of the applicant. Should external devices be employed, it is important that the device does not interfere with safe operation of the aircraft, as by emitting electronic signals that could interfere with aircraft navigation instruments. If such devices are used satisfactorily during the flight test, the use of the devices becomes a condition of issuance of the medical certificate. Should an aircraft have its controls modified to compensate for physical limitations, the applicant's medical certificate may be limited to operations involving only that specific aircraft.

The results of all MFTs will be reported by the FAA flight examiner to the medical office authorizing the test. A pilot's certificate issued after an MFT must bear any limitations the instructor conducting the test finds necessary for safety. Operating limitations required by physical limitations may restrict holders to certain aircraft types, special equipment or control arrangements, or special operating conditions. These limitations should be as general as possible to reduce the necessity of additional special medical tests when the pilot desires to fly aircraft types other than that for which he or she is physically competent.

Although he or she is unable to authorize or conduct an MFT, the AME should be familiar with the policies and procedures involved. The routine AME evaluation provides an excellent opportunity to discuss with the examinee the aspects of the MFT. This will assist the examinee in developing realistic expectations and reducing anxiety.

Ear, Nose, Throat and Related Structures

An applicant who is seeking certification for the first time after mouth, tongue, nose, pharynx, or larynx surgery, or who uses an artifical voice-producing device, should be assessed carefully to ensure intelligibility of voice communication. If there is a question that intelligibility is such that the applicant may not be understood over the radio, he or she might still be considered for a Statement of Demonstrated Ability (SODA) or a special issuance certificate.

A history of, or surgery for, benign tumors is not disqualifying. A history of treatment for chronic infections, tumors, or recurrent acute infections is not disqualifying, unless the disease or its treatment has affected speech to a degree that would compromise radio communications.

The AME should be allowed to make the decision about certification in most instances. In situations where there is a doubt, the AME should order a voice recording and send it with a request for a SODA to the FAA.

Conditions that should be considered as absolutely disqualifying include acute, paroxysmal, unpredictable attacks of vertigo, which may arise as a result of endolymphatic hydrops, or Meniere's disease, perilymph fistula, benign and malignant temporal bone tumors, and multiple sclerosis. Malignant tumors of the head and neck with metastases outside the region should also be considered as absolutely disqualifying.

A history of malignant tumor of the oral cavity, sinuses, throat or larynx without distance metastasis is not necessarily disqualifying. The decision for certification should be made on a case-by-case basis. The following factors should be considered:

1. The extent of the tumor when first treated, specifically whether there was spread to the glands of the neck or other sites.

2. The time elapsed since treatment. A period of 24 months without recurrence is satisfactory in determining success of treatment. Applicants with a history of early cancers from sites with a limited potential for metastasis, such as the lip, the vocal cord or skin, need not be permanently disqualified.

3. The effect of treatment on the voice.

Individuals who have been treated two or more years prior to examination, and who have not experienced recurrence or required more treatment, and whose voice quality is satisfactory to the AME for the tasks that are required by the class of airman for which he or she is applying, may be certified by the AME. Others should be referred to the FAA with a voice recording, a pathology report, dates and description of treatment, and comments about health status and prognosis from the treating physician.

A certificate may be given to an individual 6 weeks after middle ear and/or mastoid surgery, including stapedectomy, unless the person experiences vertigo with a Valsalva maneuver or has a positive fistula test. The person must also meet minimum hearing standards.

The hearing standards should be the same for all three classes of airman. Applicants who fail to meet the minimum hearing standards should have complete audiometric testing, including tests for both speech discrimination and speech reception threshold. A speech discrimination score of 70% or better should be considered acceptable, even though the airman's hearing does not meet the pure tone air conduction audiometric standards. A waiver should be considered for those airmen who do not meet the hearing standards even after thorough evaluation. If applicable, they might be restricted to flying outside controlled air space, or with another certified pilot who can perform the radio communication.

Dermatology

The addition of syphilis as an item in medical history requires the examiner to obtain a history about the occurrence of syphilis and its treatment. Records from the treating clinic or physician may have to be obtained, as well as laboratory data to support the diagnosis and the treatment protocol. Minimal workup would require a VDRL, with titers if positive, and FTA-ABS. No-titer or low-titer VDRL with documentation of reversal in VDRL titer with treatment would suffice as evidence of adequacy of treatment. If there is any question about the adequacy of previous treatment or if the current titer is elevated, retreatment using current Centers for Disease Control guidelines might be necessary. Any evidence of tertiary syphilis requires referral for neurological and cardiovascular evaluation.

Acute and chronic urticarial eruptions are common and are usually benign unless complicated by angioedema. An airman would disqualify himself or herself from flying while experiencing the dermatosis or its treatment. However, hereditary angioedema and acquired angioedema are disorders with catastrophic symptoms and signs, which would endanger the safety of a pilot and passengers. Therefore anyone with a history of acquired angioedema for which no etiology has been documented, or who has not been free of angioedema for a period of two years without drug therapy, would be disqualified. A family history of angioedema, or a history of urticaria associated with abdominal pain or explosive diarrhea, merits further diagnostic testing by consultants familiar with the disease.

Familial or acquired cold urticaria is associated with localized or generalized edema that could interfere with the proper use of the aircraft control systems. In extreme cases it is accompanied by profound syncope and hypotension. Cold urticaria can usually be diagnosed by the application of ice to the skin for as little as 15 seconds,

but the skin may require rewarming before symptoms and signs become clinically evident. History is important in the diagnosis, and only an applicant with a positive family history or a personal history of cold-induced eruptions should be tested.

An individual with skin lesions or a history suggestive of underlying systemic disease should be referred to an internal medicine or collagen vascular disease consultant for testing before a certificate is issued.

Most persons with a history of skin cancer that has been completely removed may be certified to fly. The exception is a person with malignant melanoma. The reader is referred to the oncology section for recommendations about follow-up of persons with malignant melanoma.

Respiratory System

Introduction

The evaluation of the respiratory system of the applicant for an airman medical certificate is critically important to aviation safety. Since altitude affects respiratory function, careful assessment of pulmonary status is required to prevent incapacitation during flight. Two general considerations are useful in approaching this evaluation:

(1) Maintenance of adequate oxygenation throughout the period of flight. A variety of disease processes can have major impact on oxygenation, either acutely or chronically. The AME must recognize those individuals who are at risk for chronic hypoxia and assess the likelihood of dangerous deterioration of oxygenation during flight. Furthermore, acute lung diseases such as asthma that cause hypoxia must be evaluated with respect to the likelihood that hypoxia will occur without warning and affect the ability of the airman to perform his or her duties.

(2) Carbon dioxide is a substance that diminishes the airman's ability to fly safely, and its retention is a warning sign of serious abnormality of respiratory function. Since direct measurement of arterial oxygen and carbon dioxide levels is costly and generally unavailable in the AME's office, clinical assessment of respiratory function, including the medical history, physical examination and spirometry, will ordinarily be used to separate those applicants requiring further evaluation of their arterial oxygen and carbon dioxide levels from the majority who do not. Furthermore, a history of prior severe respiratory tract disease of any type, dyspnea on mild exertion, persistent cough, hemoptysis, and prior diagnosis of pneumonia within the past month, or a neoplasm, should result in deferral of issuance of a certificate until a chest radiograph has been obtained and interpreted.

The most common threat to normal pulmonary function is cigarette smoking. A

major responsibility of the AME is to actively discourage applicants for medical certification from smoking cigarettes and encourage them to seek assistance in quitting smoking through community or employer-sponsored smoking cessation programs. A history of cigarette smoking totalling 20 pack-years or more should result in deferral of certificate issuance until spirometry has been obtained and evaluated. Spirometry should also be done when there is a history of chronic obstructive pulmonary disease (COPD), asthma, dyspnea on exertion or findings at physical examination of thoracic deformity, wheezing, rales, diminished breath sounds or hyperresonance. Pulmonary dysfunction presumed to be moderate or severe requires that the applicant be examined by a qualified pulmonary specialist, whose report must include an opinion of the severity of pulmonary impairment (mild, moderate or severe). Severe COPD, pulmonary fibrosis, previous pulmonary resection, recent pulmonary emboli, or cyanosis should be evaluated with spirometry and arterial blood gas measurement prior to issuance of a medical certificate, to assure that there is adequate oxygenation, which is defined as an arterial oxygen tension (PaO_2) greater than or equal to 65 mm Hg.

Spirometric analysis of lung function is an important tool for the evaluation of applicants for medical certificates. Simple spirometers should be available to the AME for routine use in these examinations. In order to assure accurate data collection, a water sealed spirometer or a rolling seal spirometer is recommended for use. Use of other types of spirometers is specifically discouraged. Peak flow meters are not capable of providing the type of information required for accurate assessment of lung function, and their use is unacceptable. The measurements required for analysis are the forced vital capacity (FVC) and the forced expiratory volume in one second (FEV_1).

The danger to public safety of certifying individuals with serious pulmonary disease is great: both judgment and the ability to perform complex tasks are affected adversely by poor respiratory function. The AME Guide contains recommended dispositions for each of the various types of lung diseases. This section presents the

rationale for these recommendations.

Infectious Diseases of the Lung

- Definition

  The lung and pleura are susceptible to multiple types of infections, whose causative agents, precipitating events, courses, pathology and prognoses vary widely. Only general principles can be addressed in this section. Pneumonia represents the prototypic lung infection, since it is common and may be associated with complications or sequelae.

- Aeromedical significance

  Pneumonias are a major cause of morbidity, disability and death in the United States, despite the availability and widespread use of many effective antimicrobial agents. It is likely that many applicants for certification will have a history of pneumonia in the recent or distant past. The acute or chronic symptoms of pneumonias and the possible involvement of the disease process in other organ systems could adversely affect flying performance and endanger the individual's well-being if he or she is permitted to fly. For example, mucus plugging of bronchi, when it occurs in the course of any pulmonary infection, may disrupt equilibration of airways pressure and result in pneumothorax during changes in altitude. Also, applicants with untreated tuberculosis or those in the early stage of treatment for this disease may present a contagious risk to other individuals.

- Clinical manifestations and diagnosis

  Symptoms of infectious pneumonia are usually obvious, although their onset

may be insidious in persons with viral infections. Fever, cough and shortness of breath are common and may be associated with sputum production, particularly in bacterial processes. Purulent sputum may suggest a bacterial infection, but it is diagnostically nonspecific. However, foul-smelling sputum suggests a mixed anaerobic infection and possibly a lung abscess. With bacterial pneumonia, physical examination usually shows tachypnea, rales or signs of pulmonary consolidation. There is often a paucity of physical findings in nonbacterial pneumonias. The chest radiograph may show areas of alveolar filling or interstitial infiltrates, or both. Large pleural effusions more commonly accompany bacterial pneumonias than viral infections. Leukocytosis with a shift toward immature leukocyte forms may occur with either bacterial or nonbacterial pneumonias, although significantly elevated white cell counts are more common with the former. Viral pneumonias may depress the leukocyte count and cause a relative lymphocytosis. Arterial blood gas analysis invariably shows some hypoxemia, which is caused by pulmonary arterial blood shunting through relatively unventilated, microatelectatic areas of the lung caused by inflammatory edema or mucous obstruction of the airways.

Although the history, physical examination, complete blood count, chest radiograph, appearance of the sputum and skin tests may lead to a diagnosis of pneumonia, these data themselves do not yield an exact diagnosis. A specific microbiological diagnosis is important since antimicrobial therapy is most effective when it is directed toward the specific infecting pathogen. Sputum smears and cultures, blood cultures and appropriate serologic tests should be obtained early. Although there is controversy about the usefulness of sputum analysis, the Gram stain showing white cells and bacteria may be

acceptable and reliable if the respiratory secretions contain alveolar macrophages rather than squamous epithelial cells. On the other hand, the absence of white cells or organisms in the sputum smear may suggest a nonbacterial infection.

Depending on the severity of the illness and other factors, more invasive diagnostic methods may be necessary. These procedures include transtracheal aspiration, thoracentesis with or without pleural biopsy, flexible fiberoptic bronchoscopy, percutaneous needle aspiration, and open-lung or -pleural biopsy. Extrapulmonary involvement associated with pneumonia should also be assessed and properly cultured. The institution of empirical antimicrobial therapy may be warranted and even essential prior to determining the microbiological etiology in persons with a rapidly progressive course of disease.

o Prognosis

In the average healthy person the prognosis for most pneumonias is generally excellent with appropriate antimicrobial therapy. Most courses of antibiotics are limited in duration, because the acute pneumonia usually resolves without residual effects. Viral pneumonias usually resolve spontaneously with treatment of symptoms. Repeated or chronic antimicrobial therapy is usually necessary in persons with significant tracheobronchitis, slowly responding lung abscess, bronchiectasis or fungal pneumonias, and in persons having chemoprophylaxis or treatment of tuberculosis.

References

1. American Throacic Society. Treatment of tuberculosis and other mycobacterial disease. Am Rev Respir Dis 1983;**127**:706-9.
2. Bartlett JG: Anaerobic bacterial pneumonitis. Am Rev Respir Dis 1979;**119**:19-23.
3. Bartlett JC, Gorbach SL, Tally FP, Finegold SM: Bacteriology and treatment of primary lung abscess. Am Rev Respir Dis 1974;**109**:510-18.
4. Bayer AS: Fungal pneumonias. Pulmonary coccidioidal syndromes. Chest 1901;**79**:575-84; 686-91.
5. Davidson M, Tempest B, Palomar DL: Bacteriologic diagnosis of acute pneumonia: comparison of sputum, transtracheal aspirate, and lung aspirates. JAMA 1976;**235**:158-63.
6. Finland M: Pneumonia and pneumococcal infections, with special reference to pneumococcal pneumonia. Am Rev Respir Dis 1979;**120**:481-502.
7. Glassroth J, Robins AG, Snider DE: Tuberculosis in the 1980s. N Engl J Med 1980;**302**:1441-50.
8. Goodwin RA, Jr, Shapiro JL, Thurman GH, et al: Disseminated histoplasmosis: clinical and pathologic correlations. Medicine 1980;**59**:1-33.
9. Murray HW, Masur H, Senterfit LB, Roberts RB: The protean manifestations of Mycoplasma pneumoniae infection in adults. Am J Med 1975;**58**:229-42.
10. Pierce AK, Sanford JP: Aerobic gram-negative pneumonias. Am Rev Respir Dis 1974;**110**:647-58.
11. Wimberly N, Faling LJ, Bartlett JG: A fiberoptic bronchoscopy technique to obtain uncontaminated lower airway secretions for bacterial culture. Am Rev Respir Dis 1979;**119**:337-43.

Asthma

o Definition

The term, asthma, represents a large, heterogeneous group of disorders characterized by hyperreactivity of the tracheobronchial tree to various stimuli, and manifested by widespread narrowing of the airways, or bronchoconstriction, which changes in severity either spontaneously or as a result of therapy. Constriction of the smooth muscle of the bronchioles is the most common functional abnormality in asthma, but other changes, such as mucosal edema, mucous hypersecretion and hypertrophy of bronchial smooth muscle cells also contribute to diffuse airways obstruction.

o Aeromedical significance

Asthma is a common disorder, affecting approximately 5% of the US population. Therefore, some candidates for medical certificates will have active asthma or a history of asthma. In addition, a transient period of hyperreactivity of the airways may occur in susceptible, nonasthmatic individuals following viral upper respiratory tract infections. For unknown reasons these infections occasionally lead to persistent asthma in some individuals.

The occurrence of an asthmatic attack, regardless of severity, or the presence of chronic, symptomatic asthma is obviously incapacitating and affects flying performance and safety. Recurrence or worsening of asthma is frequently unpredictable due to the variable nature of the disorder.

- Clinical manifestations and diagnosis

  Asthma is characterized by episodic wheezing, dyspnea and cough of varying severity and duration, depending on exposure to stimuli and the nature, intensity and duration of such stimuli. Symptoms range from mild and infrequent, that is, occurring only with upper respiratory tract infections, to severe and intractable, which is known as "status asthmaticus." An asymptomatic asthmatic individual may have subtle or minimal physiologic abnormalities on pulmonary function testing. During an asthmatic attack the person commonly has some respiratory distress; inspiratory and expiratory airflow obstruction, which is characterized by audible, generalized wheezes and reduced spirometric flow rates; hyperinflation of the lungs, characterized by hyperresonance on percussion and elevated total lung capacity and residual volume; and arterial hypoxemia with hypocapnia. The severity of the asthma may be characterized objectively by evaluating expired flow rates:

Degree of Obstructive Impairment	FEV_1 % predicted	FEV_1/FVC %
Normal	80 or greater	69 or greater
Mild	70 - 79	61 - 68
Moderate	60 - 69	50 - 60
Severe	59 or less	50 or less

Other laboratory findings may include tenacious, mucoid sputum with mucous plugs and eospinophils, blood eosinophilia, and transient arrhythmias or electrocardiographic changes of right-heart strain. Chest radiographs are

usually normal or show reversible hyperinflation of the lungs. Transient pulmonary infiltrates or pneumothorax rarely are present.

The development of pulsus paradoxus, disappearance of wheezes (not due to therapy), or normocapnia during an asthma attack is a sign of advanced respiratory fatigue and failure, which calls for immediate and vigorous hospital management.

The diagnosis of asthma is suggested by a history of episodic wheezing and shortness of breath. Confirmation is provided by spirometry showing an obstructive ventilatory pattern that is partially or completely reversed by the administration of an inhaled beta adrenergic agonist such as isoproterenol, isoetharine or metaproterenol. A history of family members with asthma or atopy is often present. A subpopulation of asthmatic individuals may present with unexplained breathlessness or cough that is often associated with postnasal drip or atopy. These individuals may have a normal physical examination and spirometry. A bronchial provocation test with inhaled methacholine, histamine, cold air, a selected antigen, or other potential bronchoconstrictor agent frequently establishes the presence of airway hyperreactivity and the diagnosis of bronchial asthma.

It is important to distinguish asthma from other disorders that may cause similar symptoms, such as chronic bronchitis, emphysema, aspirated foreign body, upper airway obstruction and congestive heart failure. This differentiation can be made on the basis of reversibility of airway obstruction in response to inhaled or systemic bronchodilator therapy. Many persons have chronic sputum production and wheezing and are classified as

"asthmatic bronchitics."

- Prognosis

  The prognosis is determined by the severity of the asthma and its repsonse to therapy. Persons with infrequent or mild asthma generally do very well with or without maintenance bronchodilator therapy. Persons with moderate or severe asthma usually require continuous therapy throughout life, particularly when the onset of the asthma is during mid- or late-adulthood. Inadequate control aggravated by infection or unmodified environmental factors contributes to persistent asthma or to incapacitating or life-threatening complications.

References

1. American Thoracic Society: Definitions and classification of chronic bronchitis, asthma, and pulmonary emphysema. Am Rev Respir Dis 1962;85:762-8.
2. Boushey HA, Holtzman MJ, Sheller JR, Nadel JA: Bronchial hyperreactivity. Am Rev Respir Dis 1980;121:389-413.
3. Corrao WM, Braman SS, Irwin RS: Chronic cough as the sole presenting manifestation of bronchial asthma. N Engl J Med 1979;300:633.
4. Feldman NC, McFadden ER: Asthma: therapy old and new. Med Clin North Am 1977;61:1239-50.
5. McFadden ER: Aerosolized bronchodilators and steroids in the treatment of airway obstruction in adults. Am Rev Respir Dis 1980;122 (Part 2):89-96.
6. Lawlor GJ, Tashkin DP: Asthma. in: Lawlor GJ, Fischer TJ (eds), Manual of Allergy and Immunology. Boston, Little Brown and Company, 1981, pp 113-56.

7. McFadden ER, Feldman NT: Asthma: pathophysiology and clinical correlates. Med Clin North Am 1977;61:1229-38.

8. Spector SL, Farr RS: Bronchial inhalation procedures in asthmatics. Med Clin North Am 1974;58:71-84.

9. Webb-Johnson DC, Andrews JL: Bronchodilator therapy. N Engl J Med 1977;297:476-82, 758-64.

10. Weiss EB, Segal MS (eds): Bronchial Asthma: Mechanisms and Therapeutics. Boston, Little Brown and Company, 1976.

Chronic Obstructive Pulmonary Disease

o Definition

The term, chronic obstructive pulmonary disease (COPD), does not refer to one specific entity but rather to a group of illnesses characterized by chronicity of obstruction to airflow that is mostly irreversible and usually most severe during expiration. Pulmonary emphysema, chronic bronchitis and chronic asthma comprise the group of diseases called COPD. Many other lung diseases, such as tuberculosis, bronchiectasis and sarcoidosis, also produce irreversible airflow limitation but differ from COPD in etiology, prognosis and treatment. These diseases should not be categorized as COPD merely because of superficial similarities of symptoms such as cough, dyspnea, wheezing and/or the identification of expiratory airflow obstruction.

Many persons with COPD have both emphysema and chronic bronchitis, which share cigarette smoking as the most important etiologic factor. In nonsmokers the clinical picture and pathology of chronic bronchitis may be indistinguishable from that of chronic asthma. Finally, some persons may

have varying degrees of all three obstructive lung diseases. It is often virtually impossible and usually clincally unnecessary to separate the contribution of each of these specific diseases. Therefore, from a practical point of view, COPD is a convenient grouping or syndrome of diseases.

o Aeromedical significance

Hypoxemia, resulting in tissue hypoxia, is the main pathophysiologic effect of COPD that limits or hinders an airman from functioning safely. Cerebral hypoxia can adversely affect psychomotor skills, memory, judgment and cognition. These effects are modified by many variables including acuteness, severity and duration of hypoxemia. In general, when the PaO_2 approximates 65 mm Hg or less at sea level, the candidate for a medical certificate should be considered severely impaired, having potentially suboptimal central nervous system functioning at altitude. Predictable decreases occur in PaO_2 when ascending to high altitudes, and this should be considered carefully before the AME issues a medical certificate:

Minimal acceptable PaO_2 when applicant is breathing ambient air at sea level and at higher altitudes

Altitude (in Feet)	Lower limit of normal PaO_2 (mm Hg)*	PaO_2 (mm Hg) must be equal to or greater than
Sea level	67	65
1000	64	61
2000	61	58
3000	58	55
4000	55	52
5000	52	50

6000	50	48
7000	47	46
8000	—	45

Source: Reference 4

*The normal PaO_2 is lowest in elderly individuals at all altitudes; these lower limits of normal are for persons at age 80 years.

o Clinical manifestations and diagnosis

The main symptoms of COPD are exertional dyspnea, chronic persistent cough, expectoration of sputum and wheezing. The abnormal physical findings of decreased breath sounds, rhonchi and wheezes may be absent or minimal until moderate or severe disease has developed. Even then, the only abnormal physical sign may be decreased breath sounds detectable only by an experienced observer during a maximal forced exhalation.

Persons with a history of symptoms that have lasted 6 months or longer or who show signs of COPD should be suspected of having significant disease. Under these circumstances pulmonary function testing must be performed in order to quantify the severity of the disease. The best screening tests are the FVC and FEV_1, and the ratio of these may be used to classify the severity of the COPD as follows:

Degree of Obstructive Impairment	FEV_1 % Predicted		FEV_1/FVC%
Normal	80 or greater		69 or greater
Mild	70 - 79	61 - 68	
Moderate	60 - 69	50 - 60	
Severe	59 or less		50 or less

Since persons with COPD usually have a moderate or greater degree of obstruction before significant hypoxemia develops, arterial blood gas analysis is usually not indicated unless the spirometry reveals moderate or severe obstruction. Spirometric results characterized by diminished FEV_1 percent predicted or diminished FEV_1/FVC ratio should be evaluated further with repeat spirometry following the administration of a bronchodilator.

o Prognosis

The prognosis of COPD is quite variable, but in general the disease tends to progress slowly over a period of months to years; progression occurs more rapidly in those who continue to smoke cigarettes. Various prediction formulas for prognosis have been published that are helpful from a general point of view; however, the most practical approach is to monitor airmen with documented mild, moderate or severe obstruction by serial examinations and spirograms at one-year intervals. The applicant with COPD should be advised to have yearly immunization against influenza.

References

1. Pardee NE, Martin CJ, Morgan EH: A test of practical value of estimating breath sound intensity; breath sounds related to measured ventilatory function. Chest 1976;70:341-344.
2. Proceedings of American College of Chest Physicians - National Heart, Lung, and Blood Institute Conference on Oxygen Therapy. Chest 1984;86:August.
3. Krop HD, Block AJ, Cohen E, et al: Neuropsychologic effects of continuous oxygen therapy in the aged. Chest 1977;72:737-743.

4. Morris AH, Kanner RE, Crapo RO, Gardner RM (eds): Clinical Pulmonary Function Testing - A Manual of Uniform Laboratory Procedures for the Intermountain Area, ed 2. Salt Lake City, Intermountain Thoracic Society, 1984.

5. Neff TA: The diagnoses and management of chronic obstructive lung disease. Cleveland Clinic Quarterly 1980;47:17-24.

6. Burrows B, Earle RH: Course and prognosis of chronic obstructive lung disease. New Engl J Med 1969;280:397-404.

7. Petty TL, Pierson DJ, Dick NP, et al: Follow-up evaluation of a prevalence study for chronic bronchitis and chronic airway obstruction. Am Rev Resp Dis 1976;114:881-890.

8. Petty TL, Hudson LD, Neff TA: Methods of ambulatory care (maintenance management for patients with emphysema and chronic bronchitis). Med Clin No America 1973;57:751-762.

9. Nocturnal Oxygen Therapy Group: Continuous or nocturnal oxygen therapy in hypoxemia chronic obstructive lung disease. Ann Int Med 1980;93:391-398.

10. Wasserman K: Cardiovascular manifestations of respiratory insufficiency. Clinical Notes on Respiratory Disease, American Thoracic Society Fall 1973 12(2).

11. Schwartz JS, Bencowitz HZ, Moser KM: Air travel hypoxemia with chronic obstructive pulmonary disease. Ann Int Med 1984;100:473-476.

12. Gong H, Jr: Air travel and patients with chronic obstructive pulmonary disease (editorial). Ann Int Med 1984;100:595-597.

Disorders of the Control of Respiration

Hypoventilation Syndromes

- Definition

  Alveolar hypoventilation is comprised of hypercapnia, defined as a PCO_2

greater than 45 mm Hg, and hypoxia, defined as a PaO_2 less than 65 mm Hg. When this condition is due to a malfunction of the central nervous system mechanisms for regulation of respiration, the arterial blood gas abnormalities are present chronically and are not necessarily associated with significant malfunction of the lung, the bony thorax or the respiratory muscles. These syndromes are often associated with pulmonary hypertension and right ventricular heart failure. The diagnosis is made by analysis of arterial blood and by ruling out abnormal function of lungs, thorax and respiratory muscles by appropriate tests, which are ordinarily performed in pulmonary function laboratories.

Hypoventilation syndromes are divided into at least three general categories:

1. Alveolar hypoventilation due to a pre-existing nervous system disorder, such as encephalitis or poliomyelitis;
2. Primary (idiopathic) hypoventilation;
3. Obesity hypoventilation syndrome

o Aeromedical significance

Hypersomnolence, which is defined as excessive daytime sleepiness or irresistible sleepiness at inappropriate times, may be a prominent feature of the syndrome and may adversely affect the airman's attention span and ability to remain awake during flight. In addition, the presence of chronic hypoxia renders the airman more vulnerable to the deleterious effect of further diminution of arterial oxygenation that may be encountered in flight. Both the hypersomnolence and the hypoxia encountered in these syndromes makes operation of an aircraft prohibitively hazardous.

- Clinical manifestations and diagnosis

  The diagnosis is suspected because of a history of easy fatiguability, hypersomnolence, morning headache, peripheral edema and cyanosis. Physical findings are those of cor pulmonale, including an accentuated pulmonic component of the second heart sound, peripheral venous distension and peripheral edema. In addition, there may be peripheral cyanosis. Associated laboratory findings are those characteristic of cor pulmonale on ECG examination and right-sided cardiac enlargement on chest radiograph. The primary (idiopathic) syndrome is distinguished from the others by absence of history of neurologic disease, such as encephalitis or poliomyelitis, and absence of obesity.

- Prognosis

  Progressive deterioration of the ability to maintain adequate blood gas levels is to be expected unless the condition is treated. The secondary effects of cor pulmonale become more important and eventually lead to cardiac decompensation and death.

Hypersomnolence Sleep Apnea Syndrome

- Definition

  The syndrome is defined as hypersomnolence in the absence of primary narcolepsy, alveolar hypoventilation and other recognized causes of sleep disorders. It is a relatively common syndrome that occurs primarily in obese males. It may also occur in postmenopausal females and as a result of a variety of conditions associated with narrowing of the upper airway, such as hypertrophied tonsils and adenoids, small trachea and malformations of the

mandible.

The syndrome is caused by a disturbance of breathing during sleep in which there are frequent cessations of breathing, called apneas. These apneas are usually obstructive, that is, due to a closure of the airway at the pharynx in the presence of active respiratory efforts. Less common are the central apneas that are due to cessation of respiratory efforts. Individuals with a mixed pattern of central and obstructive apnea have been observed rarely. The apneas are ended by an arousal or near arousal from sleep and are repeated frequently throughout the night. Daytime hypersomnolence is the result of sleep deprivation caused by frequent arousals during nocturnal sleep.

o Aeromedical significance

The irresistible urge to sleep at inappropriate times is an obvious threat to aviation safety, as are the visual hallucinations and personality disorders that are part of this syndrome.

o Clinical manifestations and diagnosis

The syndrome should be suspected when there is a history of excessive sleepiness. A history of loud snoring is virtually always present. A restless sleep pattern is commonly described either by the examinee or by his or her bed partner. Severe sleep deprivation may cause symptoms such as change of personality, visual hallucinations and impotence. These symptoms in an obese male should raise the examiner's suspicion of the diagnosis.

The diagnosis is made by appropriate studies in a diagnostic sleep disorders laboratory. Arterial blood gases are measured to exclude the other hypoventilation syndromes that also may be associated with this disorder.

o Prognosis

Hypersomnolence is the hallmark of the disorder and typically worsens in the untreated individual.

References

1. Mellins RB, Balfour HH, Turino GM, Winter RW: Failure of automatic central ventilation. Medicine 1970;49:487.
2. Rodman T, Resnick ME, Berkowitz, RD, et al: Alveolar hypoventilation due to involvement of the respiratory center by obscure disease of the central nervous system. Am J Med 1962;32:398.
3. Senior RM, Fishman AP: Disturbances of alveolar ventilation. Med Clin North Amer 1967;51:403-425.
4. Guilleminault C, Dement WC: Sleep Apnea Syndromes. New York, Alan R. Liss, 1978.

Pulmonary Thromboembolism

o Definition

The term, pulmonary thromboembolism, denotes a dynamic event during which a fibrin-based venous clot dislodges from its anchor point, usually in a vessel of the lower extremity or pelvis, flows through the inferior vena cava and impacts in the pulmonary vascular bed. This is an extremely common event, even among healthy individuals. It is estimated that only 20% to 30%

of pulmonary thromboemboli are accompanied by symptoms; the remainder go unrecognized.

Pulmonary infarction is an uncommon event that implies tissue necrosis resulting from the presence of thromboemboli that occlude the vascular supply to a portion of the lung. Infarction occurs only when the vasculature is substantially compromised by another pathologic process, since the dual blood supply to the lung through the pulmonary and the bronchial circulation usually protects against tissue death. Whether or not infarction occurs, significant pulmonary thromboembolism causes physiologic abnormalities including transient pulmonary hypertension, hypoxia and occasionally acute cor pulmonale.

Following a major pulmonary thromboembolic event there is significant danger that another thromboembolism may occur and lead to more significant physiologic abnormalities and death.

o Aeromedical significance

Acute incapacitation may occur as a result of chest pain, shortness of breath, hypoxia or cardiac arrhythmias. Hypoxia may be present for extended periods of time following pulmonary thromboembolism and it represents a potential threat to safe operation of an aircraft. Furthermore the danger of recurrent thromboembolism must be considered in the evaluation of candidates for medical certification. Another significant factor that must be kept in mind is the potential effect of the anticoagulant medications that are used to treat and prevent recurrent pulmonary thromboembolism.

o Clinical manifestations and diagnosis

Dyspnea is the most common symptom of pulmonary thromboembolism, occurring in approximately 90% of individuals. Tachypnea and tachycardia often accompany the dyspnea. Syncope occasionally occurs at the time of embolization. Pleuritic chest pain may occur even in the absence of pulmonary infarction. Cough and hemoptysis are also common whether or not infarction has occurred. Physical examination is frequently unhelpful but it may disclose tachycardia and tachypnea, the presence of rales and wheezes in the area of embolization, and possibly a pleural rub. Evidence of deep vein thrombophiebitis in the lower extremities substantially increases the likelihood of pulmonary thromboembolism. Although the chest radiograph and electrocardiographic findings are sometimes helpful, they are far from definitive. Perfusion and ventilation lung scans are the mainstays of diagnostic testing, and pulmonary angiography remains the definitive procedure in difficult diagnostic cases.

o Prognosis

Once the acute pulmonary embolism has been recognized and treated with anticoagulant medications, the prognosis for recovery is good. Pulmonary hemodynamics ordinarily revert to pre-embolization levels in approximately two to eight weeks. Surgical patients who experience pulmonary thromboembolization during a period of enforced bedrest are likely to recover completely after its treatment. However, those patients with underlying medical conditions such as congestive heart failure are likely to have recurrent episodes following treatment. Chronic cor pulmonale is a feature of unrecognized and untreated showers of small pulmonary emboli.

Pulmonary Arterial Hypertension

o Definition

Pulmonary arterial hypertension is defined as an increase in the pressure within the pulmonary vessels to a level more than 5 to 10 mm Hg above the accepted normal values. Specifically, while normal values range from 10 to 25 mm Hg, with a mean of 18 mm Hg, the person with pulmonary arterial hypertension has values that range from 15 to 35 mm Hg or greater, with a mean of 25 mm Hg or greater. These changes may occur as a result of a decrease in the cross-sectional area of the pulmonary vessels or as a result of vasoconstriction of the pulmonary vessels.

Factors that may contribute to the presence of pulmonary artery hypertension include left ventricular cardiac failure, large increases in pulmonary blood flow and increased viscosity of the blood. The normal lung can compensate for major changes in blood flow through the pulmonary vessels without an increase in pulmonary artery pressure. This is probably due to the physiologic ability to recruit previously closed vessels as the blood flow increases. The loss of this ability to compensate for changes in blood flow denotes a serious alteration in pulmonary physiology that may lead to cor pulmonale and finally to right-sided cardiac failure. It is generally recognized that there are two major forms of pulmonary arterial hypertension, an idiopathic, primary pulmonary arterial hypertension (PPAH), and a secondary pulmonary hypertension due to a variety of recognized disease processes in the lung and in the heart.

o Aeromedical significance

The presence of either primary or secondary pulmonary artery hypertension indicates the presence of severe cardiopulmonary disease that may be characterized by hypoxia, inability to respond to physical stress, syncope and sudden death. Hypoxia such as that encountered in routine aviation activities is an extremely powerful vasoconstrictor of the pulmonary vessels. While the effects of mild hypoxia on a normal lung are not significant with respect to cardiopulmonary performance, individuals with pathologic pulmonary artery hypertension may decompensate acutely when presented with this added stress.

o Clinical manifestations and diagnosis

Primary pulmonary arterial hypertension is a rare disorder of unknown etiology. It is a diagnosis of exclusion, after having ruled out secondary pulmonary arterial hypertension. PPAH is most common in young adult females. Early in the course of the disease there are no characteristic symptoms or physical findings. As the hypertension progresses the individual may notice easy fatiguability, chest discomfort and exertional dyspnea.

On physical examination no evidence of primary pulmonary or cardiac disease is noted. A prominent a-wave may be noted in the jugular venous pulse. The pulmonic component of the second heart sound is accentuated. The chest radiograph is normal in the early stages of the disease, but later there is enlargement of the pulmonary trunk and an attenuation of the peripheral pulmonary arterial branches. The electrocardiogram is characterized by right ventricular enlargement and usually right artrial

enlargement. The final diagnosis rests on the results of cardiac catheterization and the demonstration of pulmonary arterial hypertension associated with a normal pulmonary capillary wedge pressure and a normal cardiac output. Other cardiopulmonary diseases must be excluded before the diagnosis is considered proven.

Secondary pulmonary arterial hypertension may result from a variety of cardiopulmonary diseases, including thromboembolic disease, talc granulomatosis, pulmonary arteritis, pulmonary fibrosis, kyphoscoliosis, chronic high altitude sickness, left ventricular failure, mitral valve disease and idiopathic veno-occlusive disease. In addition to the manifestations of the causative disease process, secondary pulmonary arterial hypertension superimposes the same findings noted in the section on primary pulmonary arterial hypertension.

o Prognosis

Primary pulmonary arterial hypertension is nearly always fatal. The usual course of the disease includes progressive pulmonary and cardiac decomposition over a period of several years. To date, no therapy has been useful in halting or slowing the progress of the disease. Secondary pulmonary arterial hypertension also is usually a progressive and fatal complication of the underlying disease process. Chronic oxygen therapy is helpful in slowing the progress of the disease in those patients found to be hypoxic.

Neoplastic Lung Disease

- Definition

Neoplastic abnormalities in the lung are either benign or malignant. Benign neoplasms of the lung are important only insofar as they may be difficult to differentiate from malignant masses and because they occasionally produce bronchial obstruction. Bronchial adenomas, including carcinoid tumors, are low grade malignant lesions that may declare their presence by hemoptysis or bronchial obstruction. A finding of a solitary pulmonary nodule on chest radiograph should never be ignored or presumed to be benign. Age more than 35 years or a history of cigarette smoking increases the likelihood of malignancy.

Malignant neoplasms may be classified as primary tumors of the lung or metastatic tumors to the lung from another site in the body. Primary lung malignancies are most commonly of the bronchogenic type, with three subcategories being recognized according to the cellular pattern noted on biopsy: squamous cell carcinoma, anaplastic carcinoma of small cell and large cell varieties, and adenocarcinoma. It is unusual to find these malignancies in individuals who have not smoked cigarettes, except possibly in the instance of adenocarcinoma. A fourth type of primary lung malignancy originates in the peripheral airways and alveoli and is designated bronchiolo-alveolar carcinoma. Other malignancies with a primary site of origin in the chest include malignant mesotheliomas of the pleura and primary maligant mediastinal neoplasms. Malignancies arising from sites outside the chest frequently metastasize to the lungs, mediastinum and pleura and can have major effects on respiratory function.

o Aeromedical significance

Malignancy involving the chest can affect an airman's performance through several mechanisms. Hypoxia is common with lung malignancies and can adversely affect performance during flight. Chest pain and shortness of breath also can diminish the airman's performance. Cerebral metastases from lung tumors are relatively common and may suddenly cause seizures.

o Clinical manifestations and diagnosis

Cough and sputum production are the most common early clinical manifestations of pulmonary malignancies. Since bronchogenic carcinomas occur most commonly in cigarette smokers, the significance of these symptoms is often missed. Hemoptysis, soaking night sweats, dyspnea, chest pain and unexplained weight loss are common clinical complaints. On physical examination metastatic foci may be palpated in the supraclavicular foci. Occasionally, localized, unilateral wheezing can be heard in the area of the primary tumor. Signs of pulmonary consolidation can accompany a postobstructive pneumonia and egophany may be heard in the area of a malignant pleural effusion. The diagnosis is often suggested by chest radiographic evidence of a mass or pulmonary infiltrate. However, confirmation of the diagnosis can be made only by the finding of malignant cells on biopsy of the lesion or on cytologic examination of sputum, pleural fluid or needle aspirate.

o Prognosis

Surgical resection of primary lung tumors may produce a cure. Other modes of therapy are rarely curative. The five-year survival rates of all

bronchogenic malignancies are approximately 5 to 10%. Ninety percent of localized bronchiolo-alveolar carcinomas are curable once they have been resected. Malignant mesotheliomas, however, are uniformly fatal, usually within 18 months of diagnosis. The prognosis of mediastinal malignancies depends on the specific cell type.

References

1. MacMahon H, et al: Diagnostic methods in lung cancer. Semin Oncol 1983;10(1):20-33.
2. Bone RC, Balk R: Staging of bronchogenic carcinoma. Chest 1982;82(4):473-480.
3. Hoffman PC, Albain KS, Bitran JD, Golomb HM: Current concepts in small cell carcinoma of the lung. Cancer 1984;34(5):269-281.

Chronic Interstitial Lung Disease

o Definition

Chronic interstitial lung disease is a heterogeneous group of disorders characterized by disruption of the normal lower respiratory tract architecture. Although the designation "interstitial" lung disease implies abnormality of interstitial tissues alone, all portions of the gas exchanging apparatus of the pulmonary parenchyma may be involved. The cellular elements that make up the alveolar structures are changed dramatically, both in absolute numbers and in proportion to one another. The result of these changes is thickening and distortion of the alveolar septae. This type of disease process can best be understood as an inflammatory response to lower respirtory tract injury in its early stages and a scar tissue repair of damage in its late stages.

More than 150 different inciting events have been identified as producing the type of injury recognized as interstitial lung disease, but these account for only about one-third of the cases seen in the United States. The other two-thirds of cases are idiopathic. The known causes include inhalation of inorganic particles such as asbestos, silica and beryllium; organic particles such as fungi and bird droppings; gases such as oxygen and oxides of nitrogen; various fumes, vapors and aerosols; infectious agents; administration of drugs; ingestion of poisons such as paraquat; and exposure to ionizing radiation. Disorders of unknown etiology that cause this disease pattern include idiopathic pulmonary fibrosis, collagen vascular disorders, sarcoidosis and a variety of rare lung diseases.

o Aeromedical significance

Chronic interstitial lung disease decreases the ability of the lung to oxygenate the blood. The severity of this process determines a pilot's ability to operate an aircraft safely. When the disease has progressed to the point of causing an arterial PaO_2 of 65 mm Hg or less while breathing room air at sea level, relatively small changes in the inspired oxygen pressure can cause major changes in the arterial oxygen content. Such changes in inspired oxygen level are usually encountered during routine flight.

o Clinical manifestation and diagnosis

The disease is usually suspected on the basis of the history and physical findings described above. A chest radiograph showing diffuse bilateral linear or reticulonodular pulmonary parenchymal markings is supportive of the diagnosis. Pulmonary function tests show a decrease in the FVC to below 80% of the predicted value and a decrease in the static lung volume

measurements, namely, the total lung capacity, functional residual capacity and residual volume. Diffusing capacity of the lung may also be diminished. Arterial blood gas analysis is sometimes normal at rest but usually shows some degree of hypoxia and hypocapnea. In people with chronic interstitial lung disease, exercise leads to a fall in the arterial oxygen level, while the normal response is a rise in oxygenation. Diagnosis may be aided by an open lung biopsy.

The usual clinical manifestation of chronic interstitial lung disease is shortness of breath. At first the affected individual will notice decreased exercise tolerance, and as the disease progresses, the person will note dyspnea at rest. Nonproductive cough is common and may occur in uncontrollable paroxysms. Soaking night sweats are frequent. On physical examination the most consistent finding is crackling rales heard at the posterior lung bases. The rales do not clear on deep breathing or coughing. Late in the disease process the physical findings of cor pulmonale are seen, including a right parasternal cardiac impulse, accentuation of the pulmonic component of the second heart sound, hepatomegaly and peripheral edema. Clubbing of the fingers and toes may occur when the disease is well established. Cyanosis is present, which denotes severe abnormality of gas exchange.

o Prognosis

This is such a diverse group of diseases that no single statem[illegible] prognosis can be considered accurate. Some of the diseas[illegible] sarcoidosis or hypersensitivity pneumonitis due to organic [illegible] are potentially reversible. Others such as asbestosis are slowly but inexorably

progressive over many decades. Idiopathic pulmonary fibrosis is ordinarily fatal within 6 years of diagnosis.

References

1. Crystal RG, Gadek JE, Fer[illegible]ans VJ, et al: Interstitial lung disease: Current concepts of pathogenesis, staging and therapy. Am J Med 1981;70:542-568.
2. Keogh BA, Crystal RG: Clinical significances of pulmonary function studies: Pulmonary function testing in interstitial pulmonary disease. What does it tell us? Chest 1980;78:856-865.
3. Fulmer JD: An introduction to the interstitial lung diseases. Clinics in Chest Med 1982;3:457-473.

Pneumothorax

- Definition

Pneumothorax can be defined as air in the pleural space. Air may gain access to the pleural space via a defect or a rent in the visceral pleura lining the lung or the parietal pleura lining the chest wall, diaphragm and mediastinum.

Primary Spontaneous Pneumothorax (PSP) is one that "occurs in a patient who exhibits no clinical findings (other than the pneumothorax per se) referable to the underlying etiology of the spontaneous pneumothorax."[1] Secondary Spontaneous Pneumothorax (SSP) is one that "occurs as a consequence of a manifest disease process."[1] Even in PSP there is undoubtedly an underlyling pathological process, such as a visceral pleural bleb, but a pneumothorax is classified as a SSP only if the underlying process is "manifest," that is, has its own symptoms and signs, as with chronic

obstructive lung disease, tuberculosis or eosinophilic granuloma.

- Aeromedical significance

  PSP or SSP may produce acute onset of chest pain and shortness of breath, which during flight are likely to be distracting and possibly incapacitating. Hypoxia can result from a change in ventilation-perfusion ratios, and this clearly poses a serious threat to safe operation of the aircraft.

- Clinical manifestations and diagnosis

  PSP is a disease mainly of young men. The most common symptoms are pain which occurs in more than 90% of cases, and dyspnea, which occurs in 80%. Cough occurs less frequently, in 10% of individuals. Major serious pathophysiologic processes and/or complications of any pneumothorax that may acutely incapacitate a person include the degree of hypoxemia; the underlying condition of the lung; the development of a tension pneumothorax, which occurs in about 5% of persons with PSP; the development of hemopneumothorax, which occurs in about 2.5% of persons with PSP; and pain.

  Pneumothorax should be suspected when there is a history of sudden onset of shortness of breath associated with chest or shoulder pain. On physical examination the affected hemithorax moves less with each respiratory excursion than the opposite side. Breath sounds are diminished on the affected side. If a tension pneumothorax has occurred, the trachea may be shifted away from the side of the pneumothorax. The diagnosis is confirmed by observing air in the pleural space on the chest radiograph; accentuation of this radiographic abnormality can often be observed more easily when the

film is exposed during complete expiration.

o Prognosis

The overall incidence of mortality due to PSP is difficult to establish accurately because the majority of episodes of spontaneous pneumothorax that were reported before 1950 included both primary and secondary types, and they were treated without surgery and with bedrest, observation and symptomatic measures. However, more recent series of cases have reported no deaths. Of major concern and importance for the aiman after an initial episode of PSP is the likelihood of recurrence. The incidence of recurrence varies widely, from 5% to 60%, because of such variables in reporting as duration of follow-up, inclusion of suspected rather than proven episodes, the description of contralateral vs ipsilateral recurrences and of initial vs subsequent episodes, and the form of treatment of previous episodes. Ipsilateral recurrence rates of about 50% have been reported in persons followed for 6 months to 10 years when no specific therapy was provided. Most of these recurrences (62%) occurred within the first two years after the initial episode.

References

1. Killen DA, Gobbel WG: Spontaneous Pneumothorax. London, J & A Churchill Ltd, 1968.
2. Gobbel WG, Jr, Rhea WG, Nelson LA, Daniel RA: Spontaneous pneumothorax. J Thorac Cardio Surg 1963;46:331.
3. Leach JE: Pneumothorax in young adult males. Descriptive characteristics in 126 cases. Arch Int Med 1945;76:264.

4. Sahn SA (ed): Pneumothorax, in Pulmonary Emergencies. New York, Churchill Livingstone,Inc, 1982.

Drugs Affecting the Respiratory System

o Definition

Numerous types of medications are available for over-the-counter or prescribed treatment of upper and lower respiratory tract disorders. Some medications are used for a brief period and others may be required frequently, intermittently, on a long-term basis, or depending on the underlying disorder. The AME should inquire about all medications being used currently by the applicant and should assess the drugs' clinical efficacies, side effects and possible hypersensitivity and idiosyncratic reactions. The applicant may not consider nonprescription medications to be potential or serious contraindications for flying.

o Aeromedical significance

A number of respiratory system related drugs may have an adverse effect on flying performance and safety by affecting the state of consciousness, depressing ventilation, increasing cardiovascular demands or participating in drug-drug interactions. For example, antihistamine medications are used widely to treat the symptoms of upper respiratory tract infections and allergies; these drugs should not be used within 24 hours of taking control of an aircraft because of their sleep-producing effects. Finally, the AME should determine why the drugs in question are being used, because the underlying conditions themselves may be disqualifying.

References

1. American Medical Association: AMA Drug Evaluations, ed 5. Chicago, American Medical Association, 1983.
2. Harding RM, Mills FJ: Is the crew fit to fly? II: Specific medical considerations. Brit Med J 1983;287:192-5.
3. International Air Transport Association: Medical Manual. Montreal, International Air Transport Association, 1979, p 13.
4. Ziment I: Respiratory Pharmacology and Therapeutics. Philadelphia, WB Saunders, 1978.

Disorders of the Respiratory Muscles and Bony Thorax

- Definition

  Congenital and acquired hernias of the diaphragm are important disorders in which the bowel enters the thoracic cavity through a defect in the membranous or muscular portion of the diaphragm. Important disorders involving all respiratory muscles include amyotrophic lateral sclerosis, the various muscular dystrophies (especially those associated with myotonia) multiple sclerosis, myasthenia gravis and myositis. Important disorders of the bony thorax are kyphoscoliosis, fibrothorax and surgical complications of the procedures such as thorocoplasty.

- Aeromedical significance

  Disorders of the respiratory muscles and bony thorax are important in two ways. Mild and moderately severe disorders decrease the airman's respiratory reserve and make it more difficult to respond to emergency situations. Severe disorders cause abnormalities of arterial blood gases, and

hypoxia is a frequent finding. Very severe disorders are often accompanied by hypercapnia. The arterial blood gas abnormalities significantly increase the risk of incapacitation during flight.

- Clinical manifestations and diagnosis

All of these disorders may exist in mild forms that do not produce respiratory embarrassment. In the generalized muscle disorders, symptoms of general weakness usually occur prior to onset of respiratory symptoms, but this is not always true. The first symptom of respiratory problems is usually dyspnea on effort. As the processes progress to a severe stage, alveolar hypoventilation may be present with the characteristics discussed above.

Disorders of the diaphragm are diagnosed by appropriate radiologic study. Muscle disorders are diagnosed by appropriate anatomic and neurophysiologic studies usually involving muscle biopsy, electromyography and studies of nerve conduction. Disorders of the bony thorax are usually apparent on physical examination or ordinary chest radiograph.

The degree of respiratory impairment usually can be determined adequately with simple pulmonary function tests. A FVC less than 70% of predicted indicates the possibility of significant respiratory impairment and requires additional evaluation by appropriate specialists.

References

1. Bergofsky EH: Respiratory insufficiency in neuromuscular and skeletal disorders of the thorax. In, Rusk HA (ed): Rehabilitation Medicine. St. Louis, Mosby, 1971, pp 542-568.
2. Caro CC, Bubois AB: Pulmonary function in kyphoscoliosis. Thorax 1961;16:282-290.
3. Newsom DS, Goldman M, Loh L, Casson M: Diaphragm function and alveolar hypoventilation. QJ Med 1976;45(177):87-100.

Postoperative State

All surgical procedures of the thorax and many of those involving the upper peritoneal cavity compromise respiratory function postoperatively for various periods of time. The abnormalities produced are similar to those of disorders of the respiratory muscle and bony thorax. Criteria for evaluation of the postoperative state, therefore, should follow those described in the preceding section. Specifically, all persons who have had pulmonary resections should be tested by spirometry. An FVC less than 70% of predicted requires additional testing. Abnormal arterial blood gases should result in disqualification.

An additional problem in the recent postoperative state is pain. All persons whose pain requires analgesics that have effects of depressing of the central nervous system should be disqualified.

Reference

1. Mossberg B, Bjork VO, Holmgren A: Working capacity nd cardiopulmonary function after extensive lung resections. Scand J Thorac Cardiovasc Surg 1976;10:247-256.

Cardiovascular System

Overview

Cardiovascular disease poses the most important medical threat to flight safety,[1] and is the leading cause of pilots' loss of certification.[2,3] The current standards for medical certification were written in 1959. The FAA amended these standards in 1984,[4] and by keeping abreast of advances in cardiology, it has granted special issuances of medical certificates to selected pilots over the years. However, the Cardiovascular Committee has readdressed the entire issue of cardiovascular disease and aviation safety, especially to review the enormous advances that have been made in diagnosis and treatment over the last twenty years.

Our assigned task was to: 1) modify, if necessary, the Part 67 Regulations, and the forms and procedures used by the FAA and its Airman Medical Examiners (AMEs); 2) advise the FAA regarding special issuances of certificates to pilots with heart disease, so that aviation safety is maintained; 3) advise the FAA regarding potential research efforts that would clarify the status and maintain the integrity of aviation safety as it relates to cardiovascular disease.

These tasks were accomplished by subcommittees that addressed specific topics in cardiology, including ischemic heart disease, hypertension, valvular heart disease, congenital heart disease, myocardial-pericardial disease, peripheral vascular disease and arrhythmias. Each member of the entire committee had the opportunity to provide input to and to deliberate the positions of each subcommittee. We also carefully considered the opinions expressed by our colleagues who had previously addressed the question of cardiovascular disease as it relates to aviation safety.[5,6]

There was not always unanimity of opinion. The disagreements that most often arose were related to our primary objective of maintaining the public safety while not

unnecessarily constraining the medical certification process.

Certain general decisions were made that guided our thinking throughout our deliberations:

1. Current medical certification practices of the FAA have resulted in aviation safety, and we drew heavily on FAA experience in certifying pilots with cardiovascular problems, as the system has worked.[7]

2. Commercial pilots have an even greater responsibility for public safety than do private pilots and, hence, the requirements for Class I and II medical certificates should be more rigid than for Class III.[4]

3. Crew redundancy enhances aviation safety and, hence, may allow the more liberal granting of special issuances for Class I an II pilots who fly in a multicrew environment. During the special issuance procedure, we believe consideration should be given to the fact that a pilot is functioning in such an environment. To make this a mandatory requirement may be unnecessarily restrictive. For example, pilots with very mild forms of certain cardiovascular diseases that are entirely uncomplicated and who are followed every 6 months for the purpose of detecting disease progression, in many cases function at a standard risk. While our assignment was to make purely medical judgments, it is also true that, in response to a legal decision, Federal Aviation Regulations do not allow Class I special issuances to have functional limitations.[4,8]

4. All special issuances of medical certificates to pilots with cardiovascular disease should require medical follow-up.

5. In principle, a system of risk factor screening for pilots is a desirable goal. However, the value of such procedures is limited by Bayes' theorem of conditional probability when the prevalence of the disease in the population

treated is low.[9,10] For example, the random use of exercise electrocardiography for the detection of asymptomatic coronary artery disease usually results in a large number of false positive results.[11-14] Therefore, we made an effort to suggest selective screening procedures that would have a reasonable probability of enhancing aviation safety, while at the same time not unduly burden a generally effective system.

The current system already screens for such major risk factors as hypertension and electrocardiograhic (ECG) abnormalities (the latter for Class I certification). We advise ECG screening for all classes, and have suggested that the serum cholesterol be determined in a very selected population of pilots seeking Class I and II certification. In addition, we suggest that additional historical information be included in the applications of pilots, including a history of coronary disease in family members less than 50 years old, and cigarette smoking habits. We make an appeal to all pilots to utilize this information and to undergo complete screening for cardiovascular risk factors including serum lipids, blood sugar and the selective use of voluntary stress testing early in their career, in an effort to avoid subsequent cardiovascular disease and continue to fly aircraft for as long as possible.

The topic of risk factors and their role in an aviation safety examination was the most difficult one we faced and the area into which we invested a good deal of our time trying to balance the goal of aviation safety with reason.

What follows is an outline of our most important decisions, addressing those areas in which we have suggested a change in current practice.

Regulations, Forms and Procedures

- Currently, ECGs are required for Class I certification only, and are performed at ages 35 and 40 years, and annually thereafter.

It is now well established that up to 20% of heart attacks fail to produce symptoms that bring a person to a physician.[15] The resting ECG often shows a prior myocardial infarction, and patterns of left ventricular hypertrophy and nonspecific ST and T-wave abnormalities that are also associated with an increased risk of coronary artery disease.[9,11,16,17] Furthermore, many of the most common alterations of intracardiac conduction, such as right and left bundle branch blocks and Wolff-Parkinson-White, are not associated with symptoms or with easily discerned physical findings. However, each of these electrocardiographic findings triggers special concerns regarding certification and special recommendations for assessment and followup.

The ECG requirement should be modified by the recognition of these facts, in an effort to increase the assurance that frequent and potentially significant cardiac diseases in pilots will be detected. Therefore, in addition to the current requirement for Class I pilots, we suggest than an ECG be done on all pilots at the time they enter the system. These tracings will serve as a valuable baseline for future comparison. In addition, we recommend that an ECG be done on Class II applicants at ages 35 and 40 years and every two years thereafter, and an ECG should be done on Class III applicants at age 40 and every 6 years* thereafter.

---

*During our discussions we were informed that the Department of Transportation was considering extending the interval between routine Class III examinations to five years, and therefore we recommend that the ECG be repeated every five years. However, the status of this change is unclear. Apparently there is new thought of increasing the interval from two years to three years. Should this occur, we would recommend that the ECG be repeated every three years. However, until the time interval is clarified, we recommend that the ECG be repeated every 6 years, which is a multiple of both the present two-year and proposed three-year intervals.

- We have produced a list of significant cardiovascular disease states that should result in an initial denial of a certificate. This list replaces the less-well-defined regulation that "a degree of circulatory efficiency that is compatible with the safe operation of aircraft" be demonstrated.

- A single sitting blood pressure standard of 150/95 mm Hg is recommended for all pilots, with the further suggestion that the systolic level never exceed 160 mm Hg, regardless of the diastolic blood pressure. This is a less rigid requirement for younger pilots and a more rigid requirement for the older pilot than now stands. The decision regarding younger pilots is supported by our assignment to address aviation safety more than preventive medicine. However, younger pilots with blood pressures near the level of 150/95 mm Hg should modify their dietary and exercise habits, and may require treatment with medication. The more rigid requirement for older pilots relates to data supporting the significance of such a level of blood pressure in this age group.[18] The blood pressure should be taken in both arms to assess differences that could detect a subclavian steal syndrome, and to provide additional readings of value in determining the average pressure. It should be taken sitting and standing to detect significant postural changes, especially in persons on antihypertensive therapy.

- Various changes were suggested regarding forms and procedures, as well as equipment and testing methods to be used by AMEs, internists and cardiologists who evaluate pilots with cardiovascular problems. The goal of these suggestions is to update these practices to the current state of the art, including: 1) a more complete medical history; 2) deletion of routine post-exercise pulse rate readings; 3) the preference for a 12-lead 3-channel ECG

done within 30 days of the routine aeromedical examination; 4) standards for treadmill stress testing, when indicated; and 5) the description of the role of various currently-used, non-invasive testing procedures and angiography.

For example, when indicated, stress testing should be limited by symptoms or by 90% of predicted maximum heart rate (MHR) for age (the previous standard was 85% of MHR) and, when feasible, it should be carried out when the person is taking no medication. Asymptomatic hypotension is deleted as a cause for denial of certification, and a more complete listing is made of electrocardiographic abnormalities that should cause a denial of a medical certificate pending further investigation.

o All members of our committee agree with the importance of the serum cholesterol as a risk factor for coronary artery disease[19,20] and urge all pilots to determine their serum cholesterol as part of their own health maintenance program at any age.

Whether a mandatory requirement to perform a serum cholesterol on all pilots would favorably impact aviation safety is less clear to us. Some of us do not favor such a requirement, as the proposed list of abnormalities that trigger a comprehensive cardiovascular evaluation already includes ECG changes, hypertension and a history of any cardiovascular disease. In addition, laboratory standards for the performance of a serum cholesterol on a vast scale may be difficult to enforce.

Age is the most powerful predictor of coronary artery disease, and the risk increases rapidly above the age of 50 years.[21] Therefore, in an effort to

select out the population of pilots at highest risk, and where that risk especially impacts the public safety, our committee recommends that a serum cholesterol be determined at age 50 years for Class I and II pilots who are performing in a single-crew commercial operation. If the serum cholesterol exceeds 300 mg/dl, a treadmill stress test is advised.

Ischemic Heart Disease

- The scintigraphic assessment of myocardial perfusion and function by thallium 201 imaging and radionuclide ventriculography plays an important role in the evaluation of abnormal stress tests, and in the follow-up of patients after infarction, angioplasty and aortocoronary bypass surgery. Along with continuous electrocardiographic monitoring, scintigraphy assists in the stratification of such individuals into risk categories for future disease.[9,22-25] When properly performed, in some cases these procedures obviate the need for invasive angiography.

Therefore angiography is not felt to be a requirement for Class III special issuance after infarction or after aortocoronary bypass surgery, if scintigraphic studies demonstrate adequate myocardial perfusion and function. However, angiography is advised for special issuance to Class III pilots after angioplasty, because of the relative newness of the procedure.

- The current waiting period for a special issuance after aortocornary bypass surgery, angioplasty and myocardial infarction is one year. We suggest that this be changed to 6 months for aortocoronary bypass surgery and angioplasty, since during this interval the great majority of bypass graft occlusions[26] and re-stenoses of dilated vessels[27] should occur.

- In addition to the angiography that is currently required at the time of the initial examination for special issuance for all Class I and II pilots who have had coronary bypass surgery, we suggest that angiograms should be performed at five, 8 and 10 years after surgery. This judgment is based on the palliative nature of the procedure and the progressive atherosclerotic changes in the grafts and native vessels that tend to occur five to 10 years after surgery.[28,29] For angioplasty, in addition to the initial follow-up angiography, angiography is recommended after five years, as long-term follow-up data on this procedure are not yet available.

  The recommendation to perform postoperative angiography is, in part, based on the conclusion that scintigraphic studies have not yet reached a standard level of excellence at all medical institutions. However, the scintigrapic measurement of perfusion and function could replace the requirement for angiography in individual cases, when the highest quality standards of technical excellence are available and used.

- For aortocoronary bypass surgery, the number of grafts appears to be a less important determinant of special issuance than graft patency and adequate myocardial perfusion and function.[28,29]

- Follow-up of Class I and II pilots who have had an infarction, aortocoronary bypass surgery or angioplasty requires stress testing and monitoring of arrhythmias every 6 months, and scintigraphy annually. Follow-up of Class III pilots requires, at a minimum, annual stress testing and monitoring of arrhythmias.

Hypertension

- A single blood pressure standard of 150/95 mm Hg is recommended with the systolic level not to exceed 160 mm Hg, regardless of the diastolic pressure. The diagnosis of high blood pressure is confirmed when the average of two or more blood pressure determinations, ideally determined on separate visits, exceeds these standards.

- Once hypertension is established, a cardiovascular evaluation is indicated, and the initial evaluation should attempt to answer the questions posed in the 1984 Report of the Joint National Committee on the Detection, Evaluation and Treatment of High Blood Pressure:[18]

  1. Is target organ involvement present?
  2. Are cardiovascular risk factors other than hypertension present?
  3. Does the patient have a primary or secondary form of hypertension?

  We do not favor routine, extensive evaluation for secondary causes of hypertension. Rather, the extent of the assessment depends on the results of the history, physical examination, routine laboratory screening work and a treadmill stress test.

- If there is no significant target organ damage and the blood pressure is controlled either by nonpharmacologic methods or by certain acceptable drugs without significant side effects, the pilot can be certified. Annual re-evaluation and treadmill stress testing at least every three years are advised.

- Maintenance of ideal weight, restriction of salt intake, avoidance of alcohol, cessation of smoking and an appropriate program of aerobic exercise are among the important components of the nonpharmacologic treatment of hypertension.[18]

- Hydralazine and calcium channel blockers are added to the currently acceptable diuretics and beta blockers for the treatment of hypertension.

- When a pilot is taking a diuretic, the serum potassium should be checked four weeks after initiation of therapy and every 6 months thereafter. Whenever hypokalemia, defined as a serum postassium less than 3.5 mg/dl, is detected, it should be treated with potassium supplementation or with the addition of a potassium-sparing diuretic. Serum potassium should then be checked again within four weeks to be sure that the correction was successful. Such close monitoring of the serum potassium is required, because thiazide diuretics cause vetricular ectopic beats in direct proportion to the hypokalemia they induce.[30]

Valvular Heart Disease

- Only pilots with functional pulmonary or aortic outflow tract murmurs may be certified without special issuance. Initial and follow-up cardiovascular evaluation for pilots with valvular heart disease should include at a minimum echocardiography, continuous ECG monitoring and treadmill stress testing. Echo Doppler studies have replaced the need for cardiac catheterization in many patients with valvular heart disease, and the value of the procedure is likely to become even greater in the future.[31] Crew redundancy is a

important consideration for special issuance for Class I and II pilots.

- Because of the risk of atrial tachyarrhythmia and thromboembolism, only pilots with very mild and uncomplicated mitral stenosis may be considered for special issuance.[32,33] Because of its more favorable prognosis, pilots with mild and occasionally moderate, uncomplicated mitral regurgitation, including that due to mitral valve prolapse, might qualify for special issuance. In the latter group, there are rare persons with a family history of sudden death or a history of thromboembolism. This history, or the presence of a significant arrhythmia, is disqualifying. Scintigraphic studies may be necessary in some patients with mitral valve prolapse to clarify ST segment and T wave abnormalities that occur at rest and with stress testing.[34-36]

- Persons with aortic stenosis present special concerns of risk of syncope and sudden death.[37] Therefore, only pilots with mild forms of the disease, defined as a resting left ventricular aortic gradient of less than 40 mm Hg with a normal cardiac index, may be considered for special issuance with appropriate follow-up. Pilots with mild to moderate aortic regurgitation may be considered, unless the lesion is due to diseases of the aortic root. Due to the poor prognosis, those with the latter disorders should not be certified.[37]

- Selected pilots who have had mitral valvuloplasty, aortic valvotomy or tissue valve replacement may be considered for special issuance. Mechanical valves have an unacceptable risk of thromboembolism and most often require anticoagulation. Neither is consistent with aviation safety.[38]

Congenital Heart Disease

- The recommendations for discrete aortic outflow tract obstruction below or above the aortic valve follow the same general guidelines outlined for aortic stenosis. The type and extent of the cardiovascular evaluation of pilots with congenital heart disease also follows the same general guidelines described for pilots with valvular heart disease.

- Pilots with coarctation of the aorta may qualify if the lesion is minimal, with less than 10 mm Hg systolic pressure difference between the arms and legs, and if they have no hypertension, a normal ECG and no significant bicuspid aortic valve disease. Similar criteria apply after surgery. There should be extra caution with those pilots in whom repair was carried out after the age of 12 years, for whom there appears to be a greater risk of sudden death and cerebrovascular accidents.[39,40]

- Due to excellent prognoses, pilots with small (pulmonary-to-systemic flow ration less than 1.5:1) uncomplicated ostium secundum and sinus venosus defects may qualify for special issuance, as may those persons one year after having a successful surgical repair. Pilots with ostium primum defects may also be certified using similar criteria, with special attention to the degree and progression of mitral regurgitation and conduction defects.[41]

- Pilots with small ventricular septal defects may be certified for all classes. Those with moderate defects may qualify for Class III certification. After surgical closure of the defect a pilot may be certified for any class, but the criteria are more strict for Classes I and II than for Class III. Monitoring for

conduction delay and dysrhythmia should be carried out after surgery.[42]

- Pilots with mild forms of patent ductus arteriosus and those in whom the ductus has been successfully corrected surgically may qualify for special issuance.[41]

- Pilots with discrete pulmonary outflow obstruction, whether treated surgically or not, may qualify for special issuance if the obstruction is mild.[43]

- In very rare instances, applicants who have had ideal results following surgery for selected types of cyanotic congenital heart disease may qualify for certification, especially for Class III certificates. The presence of pulmonary hypertension is disqualifying for all classes.

Myocardial-Pericardial Disease

- Pilots with acute myocarditis may be granted a special issuance 6 months after the illness, if it has been transient. They must also be asymptomatic, have a normal physical examination and normal laboratory evaluation, including a noninvasive assessment of ventricular function at rest and exercise and electrocardiographic monitoring.

- Certification for all classes should be denied to applicants who have a chronic dilated or restrictive cardiomyopathy, because it is usually progressive with a significant risk of sudden incapacitation from arrhythmia, heart failure or embolus.[44] Special issuance may be granted in selected cases in which left ventricular function is reduced only mildly and is

maintained during exercise, as shown by radionuclide ventriculographic study. Echocardiography should demonstrate the absence of a significant arrhythmia. In some persons, often because of excessive alcohol intake, mildly reduced ventricular function is associated with electrocardiographic abnormalities of conduction, repolarization or rhythm. These persons should be assessed in accordance with the guidelines established for these specific electrocardiographic abnormalities.

- Applicants with hypertrophic myopathy with or without obstruction and with or without medical or surgical treatment should be denied certification, as they have a shortened life expectancy, often dying suddenly and unexpectedly from ventricular arrhythmia. Heart failure may be precipitated suddenly by the onset of artrial fibrillation.[45,46] There is a subset of individuals in whom the diagnosis is uncertain, whose disorder may be discovered on routine ECG due to changes that suggest left ventricular hypertrophy. Two-dimensional echocardiography may also demonstrate a minor degree of hypertrophy. Such persons may be considered for special issuance with close follow-up, if radionuclide ventriculography at rest and during exercise demonstrates normal or near normal function and no significant arrhythmia is observed during 48 to 72 hours of continuous ECG monitoring.

- Special issuance may be granted to pilots who have had acute pericarditis when all clinical symptoms and findings have cleared and when there is no echocardiographic evidence of pericardial effusion or continuous electrocardiographic evidence of significant arrhythmia. Since the disease frequently recurs,[47] at a minimum follow-up should include an assessment

one year after the inital episode.

- Applicants with constrictive pericarditis should be considered for special issuance only after successful surgical resection of the pericardium, which is documented by near normal postoperative intracardiac pressures, the absence of a significant arrhythmia on continuouos electrocardiographic monitoring, and a well-maintained ejection fraction at rest and during exercise.[48]

Peripheral Vascular Disease

- Any evidence of significant peripheral atherosclerotic disease, including an aneurysm, atheroembolic or chronic occlusive disease, or a history of surgery for these conditions, is initially disqualifying because of the associated incidence of cerebrovascular and coronary artery disease.[49] Appropriate diagnostic procedures should be carried out to evaluate the carotid and coronary circulation before a certificate is granted. At a minimum this should include a treadmill stress test and digital subtraction carotid angiography. Individuals who cannot perform an adequate stress test due to their peripheral vascular disease may require coronary angiography.

- Significant unoperated aneurysms of the large vessels, such as abdominal aneurysms greater than 4 cm as detected by ultrasonography, are disqualifying due to the risk of rupture that may lead to sudden incapacitation or death.[50,51]

- Pilots with aortic dissection should not be granted a medical certificate even after surgery, because the risk of recurrent dissection or other

cardiovascular complications is significant.[52]

- Special issuances can be considered four months after aneurysmectomy and four months after surgery for occlusive peripheral vascular disease, providing there are no complications or coexisting disqualifying conditions.

- Special issuances may be considered 6 months after an episode of deep vein phlebitis. They may also be considered for persons who have had one episode of pulmonary emboli, as long as these individuals have not been on anticoagulant therapy for at least 6 months. See the respiratory system section.

Arrhythmias

- Some pilots with intermittent supraventricular tachyarrhythmias may be certified by special issuance, if they are asymptomatic and rigorous noninvasive testing demonstrates no evidence of underlying cardiac disease. Atrial fibrillation that is idiopathic and paroxysmal appears to cause no excess mortality.[53] Persons with this disorder should have no recurrence for 6 months, documented at three months and 6 months by 24-hour ECG monitoring; they may take acceptable medication or no medication during this time. While the risk of systemic embolus is low in this population,[54] any such history would disqualify the applicant, as would echocardiographic evidence of a left atrial thrombus. Re-evaluation should be carried out every 6 months and should include at a minimum annual stress testing and electrocardiographic monitoring.

- For pilots with chronic atrial fibrillation with a controlled ventricular response, the same guidelines apply, including a normal exercise capacity and heart rate response to exercise. These persons may take acceptable medication.

- Pilots with pre-excitation associated with a history of arrhythmia or underlying heart disease should not be certified, because such persons may develop supraventricular tachycardia with a catastrophically rapid rate.[55] If pre-excitation occurs as an isolated observation, special issuance may be considered for all classes, with crew redundancy preferable for Class I and II applicants. Certification may also be considered after successful surgery for pre-excitation.

- Pilots with simple ventricular ectopic activity (based on the Lown Classification for 24-hour continuous ECG monitoring[56]) that occurs in the absence of underlying heart disease may be certified. Such simple ventricular ectopic activity, evidenced by frequent, isolated, nonrepetitive and unifocal premature ventricular contractions with no R-on-T phenomenon, has no known adverse prognostic significance.[57,58]

- For pilots in whom complex ventricular ecotopy occurs in the absence of underlying heart disease, special issuance in highly selected cases with rigorous follow-up may be considered.[57,59] Seventy-two hours of continuous ECG recording and stress testing may be required to exclude complex ectopy with a reasonable certainty.[60]

- Pilots who are asymptomatic with first degree atrial-ventricular (AV) block, or Type I second degree AV block that is reversible with exercise and occurs in the absence of heart disease or QRS prolongation, may be certified. Those with other types of AV block should be disqualified, due to the risk of bradyarrhythmia and/or syncope.[61]

- Right bundle branch block (RBBB) present for many years in an asymptomatic person with a normal cardiovascular examination carries no increased risks of sudden incapacitation, and such pilots should be certified.[62] Pilots may also be certified when RBBB is recently acquired or of unknown duration, if there is no underlying heart disease by perfusion scintigraphy and echocardiography, and no significant additional ECG abnormalities on continuous monitoring.[63] Pilots with left anterior and posterior fascicular block should be evaluated and certified by the same criteria.

Pilots with left bundle brach block (LBBB) have a greater risk of sudden incapacitation, and a significant percentage of those in whom LBBB is recently acquired will have underlying coronary artery disease.[64,65] The evaluation and criteria for special issuance should generally follow the guidelines for acquired RBBB. However, in addition to the significant incidence of underlying coronary artery disease, persons with LBBB may have false positive thallium perfusion studies.[66] Hence, coronary angiography may be necessary to evaluate these individuals. Annual follow-up is required.

- Pilots with pacemakers should not be certified.[5] Special issuance may be considered only if it can be demonstrated that the pacemaker was implanted in the absence of currently accepted indications and provides no therapeutic benefit, and the applicant has no other disqualifying cardiovascular disorder.

- In general, drug therapy for arrhythmias should not immediately disqualify a pilot. The following factors should be considered during the special issuance process: 1) the type of arrhythmia; 2) the presence or absence of associated cardiac disease; 3) the type and dose of the therapeutic agent; 4) the degree of control that has been achieved and the period of time that adequate control has lasted; 5) the presence or absence of side effects; 6) the blood level of the drug; 7) the documentation of the pilot's medical status at the time of consideration for special issuance, and; 8) the program of medical monitoring.

References

1. Bennett G: Aviation accident risk and aircrew licensing. Eur Heart J 1984;5(suppl A):9-13.
2. Characteristics of Medically Disqualified Airline Pilots. Civil Aeromedical Institute, Federal Aviation Administration, 1983;5.
3. Characteristics of Medically Disqualified Airmen Applicants in Calendar Years 1977 and 1978. Civil Aeromedical Institute, Federal Aviation Administration, AM 1980;19.
4. Special Issuance of Airman Medical Certificates and Revision of Cardiovascular and Alcoholism Standards. Federal Aviation Regulations Amendment 67-11 (47-FR-16298) April 15, 1982.

5. Cardiovascular problems associated with aviation safety: Eighth Bethesda Conference of the American College of Cardiology. Am J Cardio 1975;36:573-628.

6. The First United Kingdom Workshop in Aviation Cardiology. Eur Heart J 1984;5(suppl A):1-164.

7. Federal Aviation Administration Response to the Comptroller General of the United States (Report No CED-76-154, November 3, 1976) September 22, 1978.

8. Special Issuance of Medical Certificates. Federal Aviation Regulations 67.19 effective May 17, 1982.

9. Uhl GS, Froelicher V: Screening for asymptomatic coronary artery disease. J Am Coll Cardio 1983;1:946-955.

10. Vecchio JT: Predictive value of a single diagnostic test in unselected populations. N Engl J Med 1966;274:1171-1173.

11. Zoltic JM, McCallister HA, Bedynek JL: The United States Army cardiovascular screening program. J Cardiac Rehab 1984;4:530-535.

12. Borer JS, Brenski JF, Redwood DR: Limitations of the electrocardiographic response to exercise in predicting coronary artery disease. N Engl J Med 1975;293:367-371.

13. Bruce RA, DeRoven TA, Hossack HF: Value of maximal exercise tests in the risk assessment of primary coronary heart disease in healthy men: Five years experience of the Seattle Heart Watch Study. Am J Cardiol 1980;46:371-378

14. Piepgrass SR, Uhl GS, Hickman JR: The limitations of the exercise stress test in the detection of coronary artery heart disease in apparently healthy men. Aviat Space Environ Med 1982;53:379-382.

15. Margolis JR, Kannel WB, Feinleib M: Clinical features of unrecognized myocardial infarction, silent and symptomatic: Eighteen year follow-up: The Framingham Study. Am J Cardiol 1973;32:1-7.

16. Kannel WB, Doyle JT, McNamara PM, et al: Precursors of sudden cardiac death: Factors related to the incidence of sudden death. Circulation 1975;51:606-614.

17. Hampton JR: The importance of minor abnormalities in the resting electrocardiogram. Eur Heart J 1984;5(suppl A):57-63.

18. The 1984 Report of the Joint National Committee on Detection, Evaluation, and Treatment of High Blood Pressure. Arch Int Med 1984;144:1045-1057.

19. Stamler J: Diet, serum lipids, and coronary heart disease: The epidemiologic evidence, in Levy RI, Rifkind BM, Dennis BH, et al (eds): Nutrition, Lipids and Coronary Heart Disease - A Global View. New York, Raven Press, 1979.

20. Kannel WB, Castelli WP, Gordon T: Cholesterol in the prediction of atherosclerotic disease. New perspectives based on the Framingham Study. Ann Intern Med 1979;90:85.

21. Tunstall-Pedoe H: Risk of coronary heart attack in the normal population and how it might be modified in flyers. Eur Heart J 1984;5(suppl A):43-49.

22. Hung J, Goris ML, Nash E, et al: Comparative value of maximum treadmill testing, exercise thallium myocardial perfusion scintigraphy and exercise radionuclide ventriculography for distinguishing high- and low-risk patients soon after acute myocardial infarction. Am J Cardiol 1984;53:1221-1227.

23. Gibson RS, Watson DD, Taylor GJ, et al: Prospective assessment of regional myocardial perfusion before and after coronary revascularization surgery by quantitative thallium-201 scintigraphy. J Am Coll Cardiol 1983;1:804-815.

24. Lindsay J, Nolan NG, Kotlyarov EV, et al: Radionuclide ventriculography following coronary bypass surgery: Correlation with arteriographic findings. Ann Thorac Surg 1982;33:238-243.

25. The Multicenter Postinfarction Group: Risk stratification after myocardial infarction. New Engl J Med 1983;309:331-336.

26. Lawrie GM, Morris GC: The role of surgery in the treatment of coronary artery disease: A surgical perspective. Cardiology Series Baylor College of Medicine 1983;6(4).

27. Libow M, Gruentzig AR, Greene L: Percutaneous transluminal coronary angioplasty. Current Prob in Cardiol 1985;X(6).

28. Loop FD: Progress in surgical treatment of coronary arteriosclerosis. Chest 1983;84:611-624, 740-755.

29. Hall RJ, Elayda MA, Gray A: Coronary artery bypass: Long term follow-up of 22,284 consecutive patients. Circulation 1984;68(suppl II):208-212.

30. Holland OB, Nixon JV, Kuhnert L: Diuretic-induced ventricular ectopic activity. Am J Med 1981;70:762-768.

31. Richards KL: Doppler echocardiographic diagnosis and quantification of valvular heart disease. Current Probl in Cardiol 1985;X(2).

32. Probst P, Goldschlager N, Selzer A: Left atrial size and atrial fibrillation in mitral stenosis: Factors influencing their relationship. Circulation 1973;48:1282.

33. Coulshed N, Epstein EJ, McKendrick CS, et al: Systemic embolism in mitral valve disease. British Heart J 1970;32:26.

34. Rappaport E: Natural history of aortic and mitral valve disease. Am J Cardiol 1975;35:221.

35. Cheitlin MD: Mitral valve prolapse. Circulation 1979;59:610-612.

36. Jeresaty RM: Mitral Valve Prolapse. New York, Raven Press, 1979.

37. Hancock EW: Diseases of the aortic valve: Diagnosis, natural history and surgical treatment. Cardiology Series, Baylor College of Medicine 1983;6:5-24.

38. McClung JA, Stein JH, Ambrose JA, et al: Prosthetic heart valves: A review. Prog Cardiovasc Dis 1983;26:237-270.

39. Maron BJ: Coarctation of the aorta in the adult, in Roberts WC (ed): Congential Heart Disease in Adults. Philadelphia, FA Davis, 1979, pp 311-319.

40. Maron BJ, Humphries JON, Rowe RD, et al: Prognosis of surgically corrected coarctation of the aorta. A 20-year postoperative appraisal. Circulation 1973;47:119-126.

41. McNamara DG, Latson LA: Long-term follow-up of patients with malformations for which definitive surgical repair has been available for 25 years or more, in Engle MA and Perloff JK (eds): Congenital Heart Disease After Surgery: Benefits, Residua, Sequelae. New York, Yorke Medical Books, 1983, pp 77-99.

42. Engle MA, Kline SA: Ventricular septal defect in the adult, in Roberts WC (ed): Congenital Heart Disease in Adults. Philadelphia, FA Davis, 1979, pp 279-309.

43. Nugent EW, Freedmon RM, Nora JJ, et al: Clinical course in pulmonary stenosis. Circulation 1977;56:38-47.

44. Goodwin JF: The frontiers of cardiomyopathy. Br Heart J 1983;48:1-18.

45. McKenna W, Deanfield J, Faruqui A, et al: Prognosis in hypertrophic cardiomyopathy: Role of age and clinical electrocardiographic and hemodynamic features. Am J Cardiol 1981;47:1532-1538.

46. Shah PM, Adelman AG, Wigle ED, et al: The natural (and unnatural) course of hypertrophic cardiomyopathy - A multicenter study. Circulation 1975;34/35(suppl II):179-195.

47. Shabetai R: The Pericardium. New York, Grune and Stratton Inc, 1981.

48. Boltwood CM, Shah PM: The pericardium in health and disease. Current Problems in Cardiology 1984;9(5).

49. Hertzer NR, Beven EG, Young JR, et al: Coronary artery disease in peripheral vascular patients. A classification of 1000 coronary angiograms and results of surgical management. Ann Surg 1984;199:223-233.

50. Joyce JW, Fairbairn JF II, Kincaid OW, et al: Aneurysms of the thoracic aorta. A clinical study with specific reference to prognosis. Circulation 1964;29:176-181.

51. Schatz IJ, Fairbairn JF II, Juergens JL: Abdominal aorta aneurysms: A reappraisal. Circulation 1962;26:200-205.

52. Appelbaum A, Karp RB, Kirklin JW: Ascending vs descending aortic dissections. Ann Surg 1976;183:296-300.

53. Gajewski J, Singer RB: Mortality in an insured population with atrial fibrillation. JAMA 1981;245:1540-1544.

54. Wolf PA, Dawber TR, Thomas E, et al: Epidemiological assessment of chronic atrial fibrillation and risk of stroke: The Framingham Study. Neurol 1978;28:973-977.

55. Gallagher JJ, Pritchett ELC, Sealy WC, et al: The pre-excitation syndromes. Prog Cardiovasc Dis 1978;20:285-327.

56. Lown B: Clinical management of ventricular arrhythmias. Hospital Practice April, 1982:73-85.

57. Crow R, Blackburn H: The prognostic significance of ventricular ectopic beats among the apparently healthy. Amer Heart J 1982;105:244-248.

58. Rabkin SW, Mathewson FAL, Tate RB: Relationship of ventricular ectopy in men without apparent heart disease to occurrence of ischemic heart disease and sudden death. Amer Heart J 1981;104:135-142.

59. Rodstein M, Wolloch L, Gubner RS: A mortality study of the significance of extrasystoles in an insured population. Trans Assoc Life Insurance Med Dir Amer 1971;54:91-111.

60. Moss AJ: Detection, significance and management of VPB's in ambulatory patients. Cardiovasc Clinics 1980;11:143-158.

61. Strasberg B, Amat-y-Leon F, Dhingra RC, et al: Natural history of chronic second-degree atrioventricular nodal block. Circulation 1981;63:1043-1049.

62. Barrett PA, Peter CT, Swan HJC, et al: The frequency and prognostic significance of electrocardiographic abnormalities in clinically normal individuals. Prog Cardiovasc Dis 1981;23:299-319.

63. Lancaster MC, Schechter G, Massing GK: Acquired complete right bundle branch block without overt cardiac disease. Am J Cardiol 1972;30:32-36.

64. Singer RB: Mortality in 966 life insurance applicants with bundle branch block or wide QRS. Trans Assoc Life Ins Med Dir Amer 1970;52:94-114.

65. Rabkin SW, Mathewson FAL, Tate RB: Natural history of left bundle branch block. Br Heart J 1980;43:164-169.

66. Iskandrian AS, Hamid Hakki A: Thallium-201 myocardial scintigraphy. Am Heart J 1985;109(1):113-129.

Arrhythmias

Introduction

The most frequent symptoms described by individuals who are experiencing alterations of rate, rhythm or conduction include palpitations, episodes of light-headedness or syncope, and either chest discomfort or shortness of breath. Palpitations are often described as skipped beats, jumps, thumps, fluttering or racing of the heart, and they may be frequent or infrequent, episodic or sustained, or regular or irregular. Light-headedness is often described as dizziness or "grey out" and is only rarely accompanied by any focal neurological signs. Syncope represents a complete loss of consciousness. When due to alterations of cardiac rhythm, syncope is usually not accompanied by seizure activity or by loss of continence, except in the case of asystole or ventricular fibrillation. Occasionally, disturbances of rhythm present with substernal oppressive chest discomfort mimicking coronary insufficiency, or as shortness of breath mimicking heart failure, or simply as fatigue and a decrease in exercise tolerance. Often, disturbances of rate, rhythm or conduction are detected on physical examination or on a routine ECG without any symptoms that can be attributed to the arrhythmia or conduction abnormality.

Other problems in evaluating the history are that the incidence of certain alterations of rhythm increases with age in the absence of heart disease, and that episodes of arrhythmias are separated by days, weeks or years. Symptoms attributable to arrhythmia often mimic cerebrovascular disease, "functional illness" or anxiety. Occasionally, the only clue to arrhythmia is that the subject is taking a medication that was prescribed for the arrhythmia in the absence of any symptoms attributable to the disturbance rhythm.

On physical examination the AME is expected to evaluate the rate and regularity of the pulse. When present, alterations of rate or rhythm are characterized by

tachycardia or bradycardia, or by irregularity of the arterial pulse or heart sounds. Simultaneous auscultation of the heart sounds and visual observation of the neck veins can provide important clues to the diagnosis of alterations of rhythm, although an ECG should be obtained in any applicant suspected of having an arrhythmia. In modern cardiological practice the electrocardiogram provides the definitive evidence regarding the nature of any suspected alteration of rhythm and it provides the only evidence for most alterations of conduction that should concern the AME.

It has been noted above that a history or physical finding indicating an alteration of rhythm may be the only clue to underlying heart disease. Of particular significance on physical examination is a conscious effort by the AME to evaluate the candidate for evidence of asymptomatic congenital heart disease, mitral or aortic valve disease, pericardial disease, mitral valve prolapse, and ventricular hypertrophy with or without cardiomyopathy. Alterations of rhythm may be significant in an otherwise normal individual; however, they take on added significance when they are a manifestation of underlying heart disease.

The history, the physical examination and the ECG should be regarded as minimal components of a comprehensive cardiovascular flight assessment. In the absence of any one of these components significant or potentially significant heart disease cannot be diagnosed or ruled out with reasonable certainty. In addition to these a chest radiography is required to assess and evaluate the potential significance of any disturbance of rhythm or conduction.

Among applicants for airman certification the most frequently detected disturbances of rhythm or conduction in descending order of prevalence are ventricular or atrial extrasystoles, atrial fibrillation, and episodic supraventricular tachyarrhythmias. Less prevalent are intraventricular conduction defects, pre-excitation and bradycardia that is due to either sinus node disease or advanced levels of atrioventricular block.

We recommend no restriction on certification for an applicant with either a sinus arrhythmia or infrequent simple ventricular extrasystoles, as described below, as long as all other components of the routine examination provide no evidence of underlying heart disease. Any other disturbance of rate, rhythm or conduction mandates either deferral or denial of certification with issuance contingent upon additional data, and an interpretation of these data that is compatible with aviation safety.

Sinus Arrhythmia

Normal sinus rhythm is not typically absolutely regular. The cycle length may vary up to 120 milliseconds, while the average heart rate varies between 60 and 100 beats per minute. When the average heart rate remains within these limits but individual cycle lengths vary by more than 120 milliseconds, sinus arrhythmia is present. This variation of cycle length may be either in phase or out of phase with respiration. Both forms are typically observed in situations of increased vagal tone. Sinus arrhythmia is more frequent in the young and most common in well-conditioned individuals, both young and old. It is not pathological and carries no prognostic implications. Sinus arrhythmia should not constitute a basis for denial.

Simple Ventricular Ectopic Activity

Lown has proposed a classification scheme for venticular ectopic activity based upon a 24-hour period of continuous ECG monitoring.[1] This scheme is noted below:

Grade	Characteristics
0	no ventricular premature beats (VPBs)
1A	occasional isolated VPBs less than 30/hr and less than 1/min
1B	occasional isolated VPBs less than 30/hr but greater than 1/min
2	frequent isolated VPBs greater than 30/hr

3	multiform VPBs
4A	two VPBs in a row
4B	three or more VPBs in a row
5	R-on-T, where R-VPB/QT is less than 1.0 sec

Grades 1 and 2 are regarded as simple. Grades 3, 4, and 5 are regarded as complex. Shortcomings to this classification scheme have been identified, but it remains the best currently available approach to a classification with documented predictive power.[2]

Experience over many years confirms that ectopic beats are common in otherwise healthy adults.[3-5] The following generalizations characterize this experience. First, on a routine 12-lead ECG approximately 1% to 5% of healthy individuals demonstrate some ectopic activity. This increases to 20% to 30% during a symptom-limited maximal exercise test, and to 40% to 60% during a 24-hour continuous Holter ECG.[6] The prevalence of ventricular ectopy far exceeds that of supraventricular ectopy. The incidence of ventricular ectopy and its frequency (beat per hour) increase exponentially with age.[7] Among those with any venticular ectopy 5% to 10% will show complex ventricular ectopy; the corollary is that 90% to 95% of ventricular ectopy is of the simple type. Simple ventricular ectopy in the absence of other evidence of heart disease has no known prognostic significance.[8,9] On the basis of these data it is recommended that simple ectopy should not constitute a basis for denial in the absence of other clinical evidence of heart disease.

- Supraventricular tachycardia

  Supraventicular tachycardia is defined as a heart rate at rest in excess of 100 beats per minute with the impulse originating above the bundle of His; thus, it is capable of producing a normal QRS complex. Included in this class

of disturbances of rhythm are sinus tachycardia, nonparoxysmal atrial tachycardia, nonparoxysmal junctional tachycardia, paroxysmal junctional tachycardia and multifocal or chaotic atrial tachycardia.

Sinus tachycardia is almost never caused by sinus node disease and is nearly always a manifestation of neurogenic or humoral stimuli acting on a normal sinus node. Thus, the evaluation of the subject with sinus tachycardia is focused on conditions such as anxiety, anemia, or hyperthyroidism, and treatment of these underlying conditions corrects the disturbances of rhythm and eliminates the reason for deferral or denial.

Nonparoxysmal atrial tachycardia is usually a manifestation of excess digitalis with or without associated potassium depletion. It will usually resolve with discontinuation of digitalis and supplemental potassium. Once resolved, the issue of certification will be determined not by the arrhythmia, but by the nature of the underlying condition for which digitalis was prescribed.

Nonparoxysmal junctional tachycardia generally has the same etiology and significance as nonparoxysmal atrial tachycardia; hence, the same recommendations apply.

Paroxysmal junctional tachycardia usually occurs in persons without heart disease and has been associated with anxiety, fatigue or excessive use of caffeine. Perhaps as many as 20% to 30% of these people have an atrial-ventricular (AV) nodal bypass tract producing a condition that is either associated with the Wolff-Parkinson-White (WPW) Syndrome or is

mechanistically identical to WPW but has no ventricular pre-excitation during sinus beats. Treatment is directed toward removing obvious stimuli that trigger attacks, and toward using drugs such as digitalis, beta blockers, or calcium antagonists that are designed to inhibit mechanisms responsible for this arrhythmia. In the absence of other evidence of heart disease certification might be considered if the subject is free of arrhythmia for 6 to 12 months while on treatment. Certification should be re-evaluated every 6 months and contingent upon the pilot's being free of arrhythmia. Any recurrence while on treatment should be a basis for denial.

Multifocal atrial tachycardia is usually a manifestation of clinically significant heart disease or its treatment. It should be a basis for denial unless underlying heart diseae can be excluded and the arrhythmia is controlled by treatment for 6 to 12 months.

Among applicants for certification in whom there is either a history or documentation of supraventricular tachycardia other than sinus tachycardia and for whom special issuance is being considered, it is recommended that a chest radiography and echocardiogram be included in the initial evaluation, to exclude underlying heart disease, and that an exercise test and 24-hour Holter ECG be included initially and annually thereafter to exclude recurrent arrhythmias. For persons taking mediation to prevent tachycardia, special issuance should be considered only if the candidate is free of significant side effects, and if drug blood levels are determined, these do not exceed the accepted therapeutic level.

o Atrial fibrillation and flutter

Atrial fibrillation and flutter are best considered as two ends of a spectrum resulting from re-entry of conduction within atrial muscle. When the atrial rate is 200 to 250 beats per minute with a regular or regularly irregular ventricular response, the rhythm is referred to as flutter. When the atrial rate is 300 to 360 beats per minute and the ventricular response is regular or regularly irregular, the rhythm is referred to as flutter-fibrillation. When the atrial rate is in excess of 400 beats per minute and the ventricular response is irregularly irregular, the rhythm is called atrial fibrillation. Among applicants for certification the prevalence of atrial fibrillation is about 2 per 1000.[10,11] Its prevalence increases with age.

Flutter and fibrillation may be paroxysmal or persistent. In a general population atrial fibrillation is most often a manifestation of underlying disease and usually is associated with atrial dilatation. Left ventricular failure, mitral valve disease, pericardial disease and chronic obstructive lung disease are common causes. Occasionally, flutter or fibrillation is due to sinus node disease or hyperthyroidism, or it presents as the first arrhythmic manifestation of the WPW syndrome. Atrial fibrillation is sometimes seen in the absence of any of the above conditions and is then referred to as "lone or idiopathic" atrial fibrillation. Among pilots, at least 50% of atrial fibrillation is idiopathic.

When atrial fibrillation or flutter is detected by history, physical examination or ECG, the first responsibility of the AME should be to evaluate the pilot for evidence of underlying heart disease. The

echocardiogram is particularly valuable, as is the chest radiograph, to assess left atrial size, the status of the mitral valve and left ventricular dimensions. Echocardiography or radionuclide ventriculography may be used as noninvasive tests to assess left ventricular performance, but either test is performed optimally on persons who have sinus rhythm and is less precise in persons with sustained atrial fibrillation. In individuals under the age of 45 years who have no history of chest pain or significant coronary risk factors, a negative exercise ECG or thallium scan is probably sufficient to exclude coronary artery disease. In some persons, especially in males over the age of 45 years, cardiac catheterization and coronary angiography may be required to exclude or quantify significant heart disease, most notably coronary heart disease.

Persons who have atrial fibrillation that is idiopathic and paroxysmal have no excess motality compared to normals.[12] For those with idiopathic persistent atrial fibrillation, the literature yields conflicting reports on prognosis for sruvival. Systemic emboli occasionally complicate the course of idiopathic atrial fibrillation, but the risk is low and seems to increase with age and with the chronicity of atrial fibrillation.[13] By contrast, the risk of embolization is very significant among those with atrial fibrillation and any concomitant underlying heart disease.

Therefore, evidence of any underlying heart disease in a person with atrial fibrillation should be a basis for denial of certification. For individuals with paroxysmal fibrillation or flutter and no evidence of heart disease, certification may be considered if they have had no recurrence for 6 to 12 months without medication, or have had no recurrence with medication. In

addition, they may be considered if they have had recurrences despite medication that have not caused symptoms, and it can be demonstrated that their heart rates at rest and during exercise testing, and their exercise capacity are appropriate and not significantly different from those that are expected or observed during sinus rhythm.

For those persons with chronic idiopathic atrial fibrillation the same guidelines should apply; that is, the fibrillation should be slow, the heart rate response to exercise should be controlled at a level comparable to that expected or observed in sinus rhythm, and exercise capacity should be reasonable and adequate. The blood level of drugs used to treat atrial fibrillation and flutter should be measured at the time of exercise testing and be within the "therapeutic range." Any history of a documented or probable systemic embolus should be a basis for denial. Pilots certified with idiopathic atrial fibrillation should be re-evaluated every 6 months.

o Complex ventricular ectopic activity

Ventricular ectopic beats and ventricular tachycardia arise below the bundle of His either from re-entry, enhanced automaticity or abnormal automaticity. According to the Lown criteria, complex ectopic activity includes instances of multiform VPBs, VPBs with an R-on-T pattern and two or more VPBs in a row, including ventricular tachycardia. Complex ectopic activity is infrequent, but it is occasionally seen in normal individuals. Among the most frequently detected underlying heart diseases are coronary artery disease, mitral valve prolapse, ventricular hypertrophy, cardiomyopathy, and naturally occurring or drug-induced prolongation of the QT interval. Complex ectopy may be detected on a routine ECG, but the

most sensitive test is a continuous ECG.[14] A single 24-hour tape is probably adequate for screening purposes, but three separate recordings plus an exercise test are sometimes required to exclude complex ectopy with reasonable certainty in persons with a suggestive history.

Complex ectopy documented by ECG, Holter or exercise test carries an increased risk of heart disease[15,16] or sudden death. However, complex ectopy that is observed in young healthy runners, in asymptomatic subjects during near-maximal exercise, or in persons without clinical evidence of heart disease, has never been demonstrated to be of prognostic significance.[3,8] Isolated VPBs that have been classified as complex because of more than one form or because of R-on-T are of no known prognostic significance in otherwise healthy subjects. In view of these considerations the following recommendations seem prudent: in asymptomatic subjects with isolated VPBs that are detected and classified as complex either because they are multiform or because they satisfy the R-on-T criterion, the ECG should be used to exclude the long Q-T interval syndrome; echocardiographic studies or radionuclide studies should be used to exclude left ventricular dysfunction; and either an exercise test or thallium image should be obtained to reasonably exclude significant coronary artery disease.[17] Evidence of heart disease by any of these tests should be a basis for denial, but in the absence of heart disease, isolated VPBs of multiform or R-on-T should not disqualify.

In contrast, when ventricular ectopic activity is detected and classified as complex because of two or more VPBs in a row, including ventricular tachycardia, certification should be denied regardless of the presence or

absence of underlying heart disease. The literature fails to provide evidence that any form of therapy for ventricular tachycardia alters prognosis or reduces risk of sudden death.[18] Further testing including echocardiography, radionuclide scanning, cardiac catheterization and invasive electrophysiologic testing may be indicated for diagnosis and as guides to therapy. However, none of these tests is presently known to provide data that will guide therapy in a way that reduces risk to a "normal" level. Because there is no test result that justifies certification in an individual disqualified because of venticular tachycardia, the above tests have no practical role in the certification process.[19]

o Atrioventricular conduction disturbances

Atrioventricular conduction disturbances include conditions resulting from either the delay or interruption of conduction from the atrium to the ventricle. The mechanisms responsible may be located in the atrioventricular node, the bundle of His, the bundle branch system, or a combination of these. First degree atrioventricular (AV) block refers to a delay of AV conduction with PR intervals greater than 0.22 seconds. Second degree AV block describes a condition in which the ratio of atrial to ventricular beats is 2:1, 3:2, 4:3, etc, that is, some atrial beats are conducted in the ventricle and some are not. Second degree AV block is further classified into Type I or Wenckeback pattern, and Type II, in which there is an abrupt interruption of the AV conduction without prior progressive lengthening of the PR interval. Third degree and advanced AV block describe a condition in which two or more consecutive atrial impulses fail to propagate to the ventricle.

Occasionally it may be difficult to determine the "degree" of AV block, and an atrial impulse may not be conducted even though AV conduction is normal. Such a phenomenon occurs in individuals who develop sinus node slowing, or an "escape" AV junctional pacemaker that competes with the normal SA node impulse. Dissociation of atrial and ventricular electrical events need not indicate any AV conduction disturbance, and careful assessment is required to distinguish between AV dissociation and AV block.

AV conduction disturbances may be observed in individuals without heart disease. Most notably, first degree AV block or Type I second degree AV block can result from drug effects or from increased vagal tone. These situations are typically reversible either by removing the drug or by reducing vagal tone by exercise or atropine. They are typically associated with a normal QRS duration and morphology. The presence of first degree AV block or Type I second degree block that does not respond to exercise or atropine or is associated with prolongation of the QRS complex, or Type II second degree AV block or third degree AV block, suggests significant underlying heart disease. These conditions progress to either bradycardia or syncope with sufficient frequency to be of concern. Reversible first degree AV block or Type I second degree AV block in the absence of heart disease or QRS prolongation are conditions for which the pilot may be certified. Persons with first or second degree AV block that is not reversible and that is associated with widening of the QRS complex, and persons with Type II second degree or third degree AV block, should be disqualified.

The degree and type of AV block are usually evident from a routine ECG, but individuals suspected of having AV block and those with any degree of AV

block on the resting ECG should have a Holter recording and an exercise test to assess adequately the highest degree of AV block. An echocardiogram or radionuclide ventriculogram should be obtained to exclude significant myocardial dysfunction. Invasive electrophysiologic studies are required to define the anatomic site of block, and these studies may aid in subsequent treatment. However, no known test result in a person with Type II AV block or third degree AV block provides reasonable assurance of negligible risk for bradycardia or syncope, and no test result assures a normal prognosis. Hence, invasive electrophysiologic studies have no practical value in the certification process.

o Intraventricular conduction disturbances

Abnormalities in the sequence of ventricular activation may be due to either delayed excitation or pre-excitation. The former results from a spectrum of anatomical-pathological conditions causing conduction delay or block, including septal fibrosis, prior myocardial infarction, ventricular hypertrophy and infiltrative diseases of the myocardium. Conduction defects, most notably right bundle branch block (RBBB), may be congenital and not associated with any progressive pathological process. Pre-excitation, such as the WPW syndrome or its variants, is the consequence of a congenitally persistent accessory pathway(s) for atrioventricular conduction.

If an ECG reveals an intraventricular conduction disturbance, the disturbance should be classified according to standard electrocardiographic criteria. The most frequently encountered abnormality is right bundle branch block (RBBB). Its prevalence is 1 to 2 per 1000. It frequently is present at birth or develops only at high heart rates, such as during exercise,

but it then disappears at lower rates. A special effort should be made to review ECGs that were obtained on the pilot at a younger age. Wher RBBB is known to have been present for years, is not associated with any cardiac symptoms, and is accompanied by an otherwise normal cardiac examination, it carries no known adverse risk or prognostic significance.[20,21] Therefore, it should not be a basis for denial. When RBBB is of unknown duration or is known to be recently acquired, a more comprehensive evaluation is indicated to exclude progressive cardiac pathology. A minimum evaluation should include exercise stress testing, a thallium scan to screen for potentially significant coronary artery diseae, an echocardiogram or radionuclide ventriculogram to exclude a cardiomyopathy, and a 24-hour Holter recording to exclude a more advanced block. In the absence of evidence of heart disease, acquired RBBB carries no known increased risk for death or syncope.[22,23]

Disturbed conduction in the left bundle may present as left anterior fascicular block, left posterior fascicular block or complete left bundle branch block (LBBB). Accepted criteria are available to classify most electrocardiograms into one of these groups. The most common of these abnormalities is left anterior fascicular block, which has a prevalence of 1 per 100. There is no increased risk of death and no known increased risk of syncope in persons with isolated left anterior fascicular block.[24]

Isolated left posterior fascicular block has a prevalence of approximately 1 per 1000 and there is no follow-up experience of sufficient size to justify a prognostic conclusion.

Isolated acquired left anterior or posterior fascicular block should be evaluated in the same manner as acquired or presumably acquired RBBB, using exercise tests, echocardiograms and nuclear imaging to complement a routine cardiac evaluation. Evidence of progressive cardiac pathology should be excluded with reasonable certainty. When these tests are normal the applicant may be considered for certification.

In the fourth decade of life complete LBBB is detected in about 1 to 2 per 10,000 ECGs, which is a prevalence of about one-tenth that of RBBB. Isolated LBBB is associated with only a very slight increase in mortality risk;[25] however, persons with known, recently acquired LBBB have a 10-fold increase in risk of mortality.[26] From 10% to 20% of persons with asymptomatic LBBB have coronary artery disease demonstrated by catheterization.[27] Persons with LBBB should be evaluated with echocardiography or nuclear studies to exclude a cardiomyopathy, and with thallium scans to reasonably exclude significant coronary disease. For LBBB acquiried after age 45 years catheterization and coronary arteriography may be indicated. When heart disease has been satisfactorily excluded, the applicant with LBBB may be certified. Recertification should be contingent upon an annual follow-up examination.

o Pre-excitation

The prevalence of pre-excitation, or Wolff-Parkinson-White (WPW) and its variants, is estimated at 1 to 3 per 1000 ECGs,[28] although this prevalence varies depending on the population selected for study.[29] Furthermore, the incidence of tachyarrhythmias varies with the type of population studied. The lowest incidence of arrhythmias is among young adults without a past

history. The best estimate is that 10% to 20% of such individuals will experience tachycardia at some time in later life. Although pre-excitation identifies a group with an increased incidence of troublesome tachyarrhythmias, actuarial studies have so far failed to provide evidence that pre-excitation among those without other evidence of heart disease and without prior arrhythmias is associated with an increased risk of premature death.[29,30]

The prevalence of pre-excitation is higher in certain forms of heart disease, most notably Ebstein's malformation and hypertrophic cardiomyopathy, than in the general population.[28] For this reason, the first obligation of the examiner who is evaluating an applicant with pre-excitation is to exclude these types of underlying heart disease. If these abnormalities are not obvious on physical examination and chest radiograph, the most sensitive test would be an echocardiogram.

Individuals with a past history of arrhythmia are prone to have repeated attacks. Therefore, a second obligation of the medical examiner is to obtain a detailed history of either documented or probable past episodes of tachycardia. In the absence of a history of tachycardia there is no evidence that exercise testing is a useful provocative test.[31] A 24-hour continuous ECG should be obtained; however, one that fails to show arrhythmia provides no assurance that arrhythmias will not occur at some other time.[32]

A majority of subjects with pre-excitation have inducible arrhythmias during invasive electrophysiological studies.[28] While such studies may be a useful guide to therapy, they do not help in defining those who are prone to

arrhythmia among a group with pre-excitation and no known history of arrhythmia.

Supraventricular reciprocating tachycardia is the most common arrhythmia in subjects with pre-excitation. Frequently, the heart rate during such attacks and the hemodynamic consequences are greater than in paroxysmal supraventricular tachycardia that is not associated with pre-excitation. The second most frequent arrhythmia in subjects with pre-excitation is atrial fibrillation. Although atrial fibrillation is infrequent, it is difficult to predict who is prone to this arrhythmia and which of those who develop atrial fibrillation will have a catastrophically rapid ventricular response. Invasive electrophysiologic techniques have been used to induce atrial fibrillation in subjects with pre-excitation and a very rapid ventricular response has been used as one indication for surgical therapy. Unfortunately, a relatively slow ventricular response to induced atrial fibrillation does not assure that a more rapid response will not occur under circumstances of stress other than those that occur in a catetherization laboratory.

Given these considerations it is not surprising that in the United States, Canada and Great Britain, pre-excitation has generally been regarded as a basis for disqualification. Certainly any applicant with pre-excitation and either a history of arrhythmia or underlying heart disease should be disqualified. Special issuance might be considered for those with pre-excitation and no history of arrhythmia or evidence of underlying heart disease. Under such circumstances the special issuance should be limited to situations where the applicant would not be the sole pilot, that is, situations

in which there is crew redundancy, and it should be contingent upon annual follow-up. Fortunately, pre-excitation and the risk of arrhythmia can be eliminated in 90% to 95% of persons who undergo surgery. If symptoms warrant such intervention and if postoperative electrophysiological studies confirm that pre-excitation and susceptibility to arrhythmia have been eliminated, certification could be considered.

- Pacemakers

  It is recommended that persons with an implanted cardiac pacemaker be denied certification or recertification regardless of age.[33] Any applicant who seeks special issuance should be referred to a cardiologist for a complete cardiovascular evaluation. Special issuance may be considered if it can be demonstrated that the pacemaker was inserted in the absence of currently accepted indications, and that the pacemaker currently provides no therapeutically desired or beneficial advantage, and that there is no significant underlying coronary or myocardial disease or disturbance of rhythm or conduction.

References

1. Lown, B: Clinical management of ventricular arrhythmias. Hospital Practice April 1982;73-85.
2. Bigger JT, Weld FM, Rolnitzky LM: Problems with the Lown grading system for observational and experimental studies in ischemic heart disease, in Harrison DC (ed): Cardiac Arrhythmias: A Decade of Progress, Boston, GT Hall, 1980;48:653-670.
3. Pantano JA, Oriel RJ: Prevalence and nature of cardiac arrhythmias in apparently well trained runners. Amer Heart J 1982;104:762-768.

4. Moss AJ: Clinical significance of ventricular arrhythmia in patients with and without coronary artery disease. Prog Cardiovasc Dis 1980;23:33-36.

5. Kastor J: Ventricular premature beats. Adv Int Med 1983;28:63-91.

6. Pilcher G: Twenty-four hour continuous ECG during exercise and free activity in eighty apparently healthy runners. Amer J Cardio 1983;[illegible]:859-861.

7. Sherman H, Sandberg S, Fineberg HV: Exponential increase in age specific prevalence of ventricular dysrhythmias among males. J Chronic Dis 1982;35:743-750.

8. Crow R, Blackburn H: The prognostic significance of ventricular ectopic beats among the apparently healthy. Amer Heart J 1983;105:244-248.

9. Rabkin Sw, Mathewson FAL, Tate RB: Relationship of ventricular ectopy in men without apparent heart disease to occurrence of ischemic heart disease and sudden death. Amer Heart J 1981;104:135-142.

10. Bennett D: Atrial fibrillation. Eur Heart J 1984:5(suppl A):89-93.

11. Busby DE, Davis AW: Paroxysmal and chronic atrial fibrillation in airman certification. Aviat Space Environ Med 1976;47:185-186.

12. Gajewski J, Singer RB: Mortality in an insured population with atrial fibrillation. JAMA 1981;245:1540-1544.

13. Wolf PA, Dawber TR, Thomas E, et al: Epidemiological assessment of chronic atrial fibrillation and risk of stroke: The Framingham Study. Neurol 1978;28:973-977.

14. Moss AJ: Detection, significance and management of VPBs in ambulatory patients. Cardiovasc Clinics 1980;11:143-158.

15. Kennedy HL, Pescarmona JE, Bouchard RJ, et al: Objective evidence of occult myocardial dysfunction in patients with frequent ventricular ectopy without clinically apparent heart disease. Amer Heart J 1982;104:57-65.

16. Rodstein M, Wolloch L, Gubner RS: A mortality study of the significance of extrasystoles in an insured population. Trans Assoc Life Ins Med Dir Amer 1971;54:91-111.

17. Kennedy HL, Pescarmona JE, Bouchard RJ, et al: Coronary artery status of apparently healthy subjects with frequent ventricular ectopics. Ann Int Med 1980;92:179-185.

18. Vlay S, Reid P: Ventricular ectopy: Etiology, evaluation and therapy. Amer J Med 1983;73:899-913.

19. Campbell RWF: Ventricular ectopic activity and its relevance to aircrew licensing. Eur Heart J 1984;5(suppl A):95-98.

20. Rowlands DJ: Left and right bundle branch block. Eur Heart J 1984;5(suppl A):99-105.

21. Barrett PA, Peter CT, Swan HJC, et al: The frequency and prognostic significance of electrocardiographic abnormalities in clinically normal individuals. Prog Cardiovasc Dis 1981;23:299-319.

22. Massing GK, Lancaster MC: Clinical significance of acquired complete right bundle branch block in 59 patients without overt heart disease. Aerospace Med 1969;40:967-976.

23. Lancaster MC, Schechter G, Massing GK: Acquired complete right bundle branch block without overt cardiac disease. Am J Cardiol 1972;30:32-36.

24. Rabkin SW, Mathewson FAL, Tate RB: Natural history of marked left axis deviation—left anterior hemiblock. Amer J Cardiol 1979;43:605-611.

25. Singer RB: Mortality in 966 life insurance applicants with bundle branch block or wide QRS. Trans Assoc Life Ins Med Dir Amer 1970;52:94-114.

26. Rabkin SW, Mathewson FAL, Tate RB: Natural history of left bundle branch block. Brit Heart J 1980;43:164-169.

27. Froehlicker VF, Thompson AJ, Wolthins R: Angiographic findings in asymptomatic airmen with electrocardiographic abnormalities. Amer J Cardiol 1977;39:32-398.

28. Gallagher JJ, Pritchett ELC, Sealy WC, et al: The pre-excitation syndromes. Prog in Cardiovasc Dis 1978;20:285-327.

29. Hiss RG, Averill KH, Lamb L: Electrocardiographic findings in 67,375 asymptomatic subjects. Amer J Cardiol 1960;6:96-107.

30. Sherf L, Neufeld HN: The Pre-excitation Syndrome: Facts and Theories. New York, Yorke Medical Books, 1978.

31. Strasberg B, Ashley WW, Wyndham CC: Treadmill exercise testing in the Wolff-Parkinson-White syndrome. Amer J Cardiol 1980;45:742-748.

32. Force T, Grayboys TB: Exercise testing and ambulatory monitoring in patients with pre-excitation syndrome. Arch Int Med 1981;141:88-90.

33. Surawicz B, Donoso E, Escher D, et al: Task force on arrhythmias, Eighth Bethesda Conference: Cardiovascular problems associated with aviation safety. Amer J Cardiol 1975;36:24-628.

Diseases of the Peripheral Vascular System

Arterial Disease

o History

The medical examiner should inquire about intermittent claudication, which is the most common presenting manifestation of chronic occlusive arterial disease in the lower extremities. Pain in the extremities at rest occurs only late in the course of occlusive arterial disease when there are other obvious manifestations of peripheral ischemia.

Aneurysms of the aorta and its branches usually cause no symptoms until they are large enough to exert pressure on surrounding structures and cause pain. When aneurysms are large enough to cause pain, rupture is usually imminent. A few persons become aware of their aneurysms because of the abnormal pulsations they feel in the abdominal, femoral or popliteal areas. A history of penetrating injury, such as from bullets, shrapnel, birdshot, a knife, or glass, followed by the development of a pulsating mass suggests an arteriovenous (AV) fistula. Aortic, iliac, femoral and popliteal aneurysms sometimes give rise to microemboli distally, which is usually manifested by purple discoloration and pain in the toes. Microemboli can also lodge in the skin of the lower extremities, giving rise to petechial hemorrhages. The examiner should also inquire about the presence of Raynaud's phenomenon in the extremities, usually the fingers.

o Physical examination

During examination of the abdomen, the medical examiner should attempt to palpate the abdominal aorta and to delineate its size. The femoral,

popliteal, posterior tibial and dorsalis pedis arteries should be palpated and the amplitude of pulsation should be graded from zero (absent) to four (normal). Evidence of aneurysmal dilatation of the peripheral arteries should be noted, especially in the aortoliliac, popliteal and femoral areas where they are most common. Arteries in the upper extremities should also be examined for patency. Systolic bruits heard in the abdominal aorta or in the femoral areas are much less significant than those heard in the neck. They do not necessarily indicate that an aneurysm or occlusive disease is present. A continuous bruit in the abdomen suggests either severe stenosis of a visceral artery, usually the renal artery, or an AV fistula. The AME should look for postural color changes (pallor on elevation and rubor on dependency) in persons who have diminished or absent peripheral pulses, as an estimation of the degree of ischemia of the skin.

o Recommendations

When the diagnosis of peripheral atherosclerotic disease is made, whether it be an aneurysm, atheroemboli or chronic occlusive disease, appropriate diagnostic procedures should be carried out to evaluate the coronary and carotid circulation before a certificate is granted.[1]

A maximal treadmill test should be performed. If intermittent claudication, chest pain, fatigue or any other condition contraindicates the maximal stress test, or if the results are positive or inconclusive, coronary angiography is indicated. An intravenous digital subtaction angiogram of the carotids or conventional carotid arteriography is warranted.

Thoracic or abdominal (or thoracoabdominal) aneurysms are predisposed to rupture leading to sudden death or incapacitation.[2-5] A person with an abdominal aneurysm that is more than four cm in diameter as measured by ultrasonography should be denied a medical certificate for any class.[4,5] Aortic aneurysms smaller than this should not preclude granting a certificate providing the pilot agrees to submit to ultrasonography every 6 months in order to monitor the progress of the aneurysm.[6] Following aneurysmectomy the pilot should wait for a period of four months after which the certificate can be issued, provided there have been no disabling complications from the operation, ultrasonography shows no evidence of residual aneurysmal dilatation, and no other disqualifying conditions are present.

Aneurysms of the iliac artery are also predisposed to rupture[7] and therefore to producing sudden disability or incapacitation. Applicants with aneurysms in this site may be granted a medical certificate four months after successful aneurysmectomy providing no other disqualifying conditions exist.

Femoral and popliteal aneurysms are much less likely to rupture but are likely to cause thrombosis, embolization, and venous congestion.[8-10] Consequently, they are not a common source of sudden incapacitation. Sudden arterial occlusion of a lower extremity is not a desirable event for the pilot, but it would not likely result in incapacitation to the point that the pilot and passengers would be in danger. A medical certificate should not be issued to a pilot who has an aneurysm that is more than three cm in diameter until it has been successfully removed, because aneurysms of this size are likely to cause complications.

Splenic, hepatic, renal and other visceral artery aneurysms are rare, and rupture is infrequent except during pregnancy; therefore they are not a major source of concern with regard to sudden death or incapacitation. They are usually discovered incidentally during angiography for other conditions or, when the walls are calcified, on plain radiographs of the abdomen. Without complications they should not be a reason for denying a medical certificate. Following surgical removal of a visceral artery aneurysm, a medical certificate may be issued within four months, if there have been no complications.

Aortic dissection is a catastrophic event that can disable or kill its victim.[11,12] Except for Marfan's syndrome, hypertension and congenitally bicuspid or unicommissural aortic valves,[13] there are no identifiable risk factors for this condition, which usually appears suddenly. For this reason applicants with Marfan's syndrome should not be granted a medical certificate. Hypertension and congenital abnormalities of the aortic valve are addressed elsewhere.

"Healed" dissection of the aorta results when acute dissection stops spontaneously with or without the benefit of antihypertensive therapy. The risk of further dissection or other cardiovascular complications is great enough to disqualify an applicant with this history, as well as applicants who have had surgical treatment of dissecting aneurysm.[14]

With or without intermittent claudication, occlusive peripheral arterial disease (aortoiliac, femoropopliteal or distal to the popliteal) should not be a reason to deny a medical certificate unless there are ischemic lesions that

cause so much pain that the pilot would be distracted. Chronic occlusive arterial disease itself rarely causes complications that would be suddenly incapacitating. Sudden arterial occlusion of an important collateral artery, such as the profunda femorus, might lead to abrupt worsening of ischemia, but this would not likely incapacitate the pilot to the point that he or she could not manage an aircraft.

In the absence of other disqualifying conditions, a medical certificate may be issued four months after successful revascularization of the lower extremities, even if peripheral pulses have not been fully restored, provided that there are no painful ischemic lesions.

Showers of minute atheromatous emboli, consisting of debris from aneurysms or mural thrombi within the thoracic or abdominal aorta, may lodge in multiple viscera and in the lower extremities. Sometimes this is spontaneous but most of the time it is induced by intra-arterial catheters during angiography or sometimes by anticoagulant therapy. These episodes may be isolated and single but more often are recurrent. They can lead to renal failure and perforation of a viscus, and to minute cutaneous infarcts in the skin of the lower extremities, especially the toes.

Applicants who have a history of findings in the lower extremities suggesting the diagnosis of atheromatous emboli should not be issued a certificate until further investigation excludes the diagnosis or confirms that it is an .lated episode that has not caused a significant impairment.

Raynaud's phenomenon and livedo reticularis may be primary or secondary to a multitude of underlying conditions.[15] Primary Raynaud's disease and livedo reticularis do not subject the applicant to danger of sudden disability and they are not reasons for denying a medical certificate, unless there are painful ischemic lesions on the extremities that would distract the pilot or require his or her taking opiates for relief. Medication that is sometimes prescribed for these conditions, such as alpha receptor blocking agents, can cause orthostatic hypotension, and therefore applicants who require medication to control their symptoms should not be granted a certificate.

When Raynaud's phenomenon or livedo retricularis is secondary to another disease, the underlying disease should be evaluated and the applicant should meet the criteria for that disease.

Acrocyanosis is another vasospastic disease and, although it is cosmetically unattractive, it is a benign condition that rarely produces any type of disability; it is not a reason for denying a certificate.

Venous Disease

o History

The medical examiner should question the applicant about previous episodes of thrombophlebitis and should inquire about recurrences. A remote episode following an operation, prolonged bed rest or trauma, which leaves residua, is of no concern. Recent episodes and recurrent episodes might well be a reason for disqualification. The examining physician should also inquire about late sequelae of thrombophlebitis, including chronic swelling, varicose veins, stasis dermatitis and stasis ulceration of the lower extremities. A

history of a pulmonary embolus associated with an episode of thrombophlebitis is important, especially if it has been recent or if there have been recurrent episodes.

It is important to try to establish by history, when possible, whether the thrombophlebitis was confined to the superficial veins or whether it involved the deep veins. Diffuse swelling and cyanosis of the extremity would suggest involvement of the deep veins. When thrombophlebitis is confined to the superficial veins it is readily apparent as a "red streak" along the course of the vein, which is easily palpable as a tender cord. Superficial thrombophlebitis usually occurs in varicose veins.

- Physical examination

The lower extremities should be examined for evidence of incompetent varicose veins, which can be either primary or secondary to previous deep venous thrombosis. The combination of incompetent superficial veins, chronic edema of the involved extremity and brownish discoloration around the ankle suggests chronic venous insufficiency from previous deep venous thrombosis. Stasis ulcers typically occur just above the internal malleolus, usually in an area of indurated cellulitis. Frequently, a single episode of deep vein thrombosis will leave no residual whatsoever.

- Recommendations

An applicant requesting a certificate to fly is not likely to be examined at the time when he or she has acute venous thrombosis. Therefore, the recommendations below apply more to pilots who have already been certified and who develop acute venous thrombosis.

Superficial thrombophlebitis usually occurs in varices and sometimes as a result of minor trauma, but it can occur in normal and competent superficial veins as a result of direct trauma or systemic illness, such as cancer, blood dyscrasias, or thromboangiitis obliterans. Since superficial thrombophlebitis seldom gives rise to pulmonary emboli, this disease is not likely to lead to sudden death or incapacitation and should not disqualify the applicant once the acute episode has subsided, provided serious underlying disease has been ruled out.

Deep venous thrombosis can led to pulmonary emboli, sudden incapacitation or even death, and therefore this condition should disqualify the applicant until the acute event has subsided and it is determined that this is a recurrent condition or an isolated episode. Since most physicians now prescribe coumarin anticoagulants following an episode of deep venous thrombosis, the certificate should be withheld until the applicant can safely discontinue long-term anticoagulant therapy. In no case should a certificate be issued sooner than 6 months after an acute episode of deep venous thrombosis.

If deep venous thrombosis is complicated by pulmonary embolism, coumarin-type anticoagulants are almost always prescribed for periods up to one year. This therapy itself should disqualify the applicant until it can safely be withdrawn. If there was an identifiable event responsible for the deep venous thrombosis and pulmonary embolus, such as trauma or surgery, and pulmonary function has not been compromised by the embolus, a certificate may be issued when long-term anticoagulant therapy is discontinued; in no

case should this be sooner than 6 months after the embolic event.

Chronic venous insufficiency is the late result of deep venous thrombosis. It is not a reason to disqualify an applicant for a medical certificate, since it will not lead to sudden disability or incapacitation, provided that at least 6 months have elapsed since the acute event and the applicant is no longer on anticoagulant therapy and has no history of recurrent thrombophlebitis.

Incompetent superficial veins with or without varicosities are not a reason to disqualify an applicant for a medical certificate, since this condition will not lead to sudden incapacitation. Varicosities sometimes are complicated by superficial thrombophlebitis but since this rarely gives rise to emboli, this also should not be a reason for disqualifying an applicant.

Arteriovenous (AV) fistulas are either congenital or caused by trauma. They are characterized by loud continuous bruits, and when they are close to the surface, by an accompanying thrill. Large fistulas can lead to heart failure, which would disqualify the applicant. In the absence of cardiac complications, an AV fistula is not likely to lead to sudden incapacitation and therefore is not a reason to disqualify an applicant. Since it is often desirable to resect AV fistulas to prevent complications, a medical certificate may be granted after four months if there have been no complications from the surgery, there are no other disqualifying conditions, and the applicant has recovered completely.

References

1. Hertzer NR, Beaven EG, Young JR, et al: Coronary artery disease in peripheral vascular patients: A classification of 1000 coronary angiograms and results of surgical management. Ann Surg 1984;199:223-233.
2. Joyce JW, Fairbairn JF II, Kincaid OW, et al: Aneurysms of the thoracic aorta: A clinical study with specific reference to prognosis. Circulation 1964;29:176-181.
3. Estes JE Jr: Abdominal aortic aneurysm: A study of 102 cases. Circulation 1950;2:258-264.
4. Sommerville RL, Allen EV, Edwards JE: Bland and infected atherosclerotic abdominal aortic aneurysms: A clinical pathologic study. Medicine 1950;38:207-221.
5. Schatz IJ, Fairbairn JF II, Juergens JL: Abdominal aortic aneurysms: A reappraisal. Circulation 1962;26:200-205.
6. Hertzer NR, Beven EG: Ultrasound aortic measurement and elective aneurysmectomy. JAMA 1978;240:1966-1968.
7. Markowitz AM, Normal JC: Aneurysms of the iliac artery. Ann Surg 1961:154:777-787.
8. Pappas G, Janes JM, Bernatz PE, Schirger A: Femoral aneurysms. JAMA 1964;190:489-493.
9. Gifford RW Jr, Hines EA Jr, Janes JM: An analysis and follow-up study of 100 popliteal aneurysms. Surgery 1953;33:284-293.
10. Wychulis AR, Spittell JR Jr, Wallace RB: Popliteal aneurysms. Surgery 1970;68:942-952.
11. Hirst AE Jr, Johns VJ Jr, Kime WS Jr: Dissecting aneurysm of the aorta: A review of 505 cases. Medicine 1958;37:217-279.

12. McCloy RM, Spittell JA Jr, McGoon DC: The prognosis in aortic dissection (dissecting aortic hematoma or aneurysm). Circulation 1965;31:665-669.

13. Larson EW, Edwards WD: Risk factors for aortic dissection: A necropsy study of 162 cases. Am J Cardiol 1984;53:849-855.

14. Appelbaum A, Karp RB, Kirklin JW: Ascending vs descending aortic dissections. Ann Surg 1976;183:296-300.

15. Gifford RW Jr, Hines EA Jr: Raynaud's disease among women and girls. Circulation 1957;16:1012-1021.

Myocardial Disease - Cardiomyopathy

Introduction

In some areas of the world cardiomyopathy is a common type of heart disease. In the United States, it is an important but not frequent cause of death from heart disease. It may present either as an acute and transient insult to the myocardium or as a long-standing chronic disease. Because of the risks of incapacitation from heart failure and arrhythmia, it is of great concern to aviation personnel.[1,2]

Some confusion has resulted from terminology and various classifications of myocardial disease. "Cardiomyopathy," "myocardiopathy," and "primary myocardial disease" are the names that have been most commonly used and, for all practical purposes, they are synonymous. They mean that the disease primarily affects the myocardium and that it is not vascular, valvular, hypertensive, or pericardial in origin. While it is known that anatomic changes in the myocardium and myocardial failure may occur as the result of vascular diseases, such as coronary artery disease; valvular disease, such as aortic valve disease or mitral regurgitation; hypertensive disease; and pericardial disease, such as constrictive pericarditis with myocardial involvement, those forms of secondary myocardial failure are not considered to be true cardiomyopathies. There are many known causes of cardiomyopathy, but in most cases the etiology is not known, that is, it is "idiopathic."

Classification

A classification based on the hemodynamic presentation of the disease state divides cardiomyopathy into dilated (which was formerly called congestive), restrictive, and hypertrophic (which was formerly called obstructive) forms.[3] There are no data available to compare the frequencies of occurrence of these forms of cardiomyopathy. Their incidence undoubtedly varies in different regions of the world. However, in the

United States the vast majority are of the dilated type, a few are of the hypertrophic type, and only rare cases are of the restrictive type. Those persons with the dilated type of cardiomyopathy have dilatation of the left, right or both ventricles. The dilatation often becomes quite severe. In some cases, mild degrees of hypertrophy may accompany it. Systolic ventricular function becomes impaired. Congestive heart failure may or may not be present. Both atrial and ventricular cardiac arrhythmias are very common.

In contrast, the cardiomyopathies of the restrictive type cause stiffening of the ventricules and prevent ventricular diastolic filling, which is very similar to the pattern seen in constrictive pericarditis. Examples are endomyocardial fibroelastosis; Loeffler's cardiomyopathy, or eosinophilic endomyocardial disease; amyloidosis; sarcoidosis; and hemochromatosis.

In the hypertrophic form of cardiomyopathy, the diseased muscle is hypertrophied and impairs ventricular filling and may obstruct ventricular ejection. The hypertrophy usually involves the left ventricle, but it may also occasionally affect the right ventricle. The septum is usually more hypertrophied than the free wall, but in some cases the hypertrophy may be concentric. Left ventricular volume is normal or reduced. Systolic pressure gradients are common.

This classification is purely descriptive, not etiologic, and is based solely on the hemodynamic and anatomic characteristics of the ventricles. There are some persons with cardiomyopathy who do not fit neatly into anyone of these categories and theirs has been called "unclassified cardiomyopathy." This term applied particularly to the individual with ECG abnormalities such as repolarization changes, conduction defects, large voltage of ventricular hypertrophy, or cardiac arrhythmias, but in whom ventricular function is normal. Progression to overt cardiomyopathy of the dilated, restrictive, or hypertrophic types may or may not occur.

Acute and Chronic Cardiomyopathy

Cardiomyopathy can be seen in both acute and chronic stages.[4] Usually, acute myocarditis runs a brief course, clears, and leaves no clinical evidence of residual damage. Infrequently, it may pass from an acute phase into a chronic form, either shortly after the acute phase or after a latent period.[5] However, in most cases of chronic cardiomyopathy there is no history of a previous acute illness that might have been identified as acute myocarditis.

Acute Cardiomyopathy or Myocarditis

- Etiology

  Acute myocarditis may be due to numerous causes, such as viral infections, especially Coxsackie B, and less commonly Coxsackie A, echo, polio, and influenza; peripartum; toxins; idiopathic; rheumatic fever; Chagas' disease or American trypanosomiasis; toxoplasmosis; diphtheria; and typhoid fever. Acute pericarditis may accompany the myocarditis of infectious diseases.

  Heart failure and arrhythmias occurring toward the end of pregnancy and in the postpartum period sometimes have been considered a specific form of cardiomyopathy. It is controversial whether this cardiomyopathy is directly due to the pregnancy or merely the coincidental occurrence of an idiopathic or viral form of myocarditis in the peripartum period.

  Toxic myocarditis has occurred in cobalt poisoning when cobalt was used in excessive amounts as an additive to beer. Doxorubicin (Adriamycin^{R}) is a cancer chemotherapy drug of the anthracycline group that may lead to irreversible cardiotoxicity.[6] It is particularly hazardous in persons who have

received or who are concurrently receiving radiation therapy to the chest, cyclophosphamide or mithramycin.

Rheumatic fever can produce a myocarditis that leads to heart failure. Fortunately, severe myocarditis occurs only in a small percentage of persons with acute rheumatic fever. After the acute attack heals, many of these individuals have residual valvular abnormalities due to the rheumatic endocarditis that accompanied the myocarditis.

- Natural history

  In severe acute myocarditis there may be a relentless progression of heart failure and even death, but in the more typical case there is either a total or partial healing of the acute process. When there is significant residual injury to the myocardium, varying degrees of cardiomegaly, heart failure or arrhythmia may result.

- Recommendations for certification

  Complete evaluation should include an ECG, chest radiograph, treadmill exercise test, echocardiogram, gated heart pool scan, and 24-hour ambulatory ECG. Certificates for all classes of airmen should be denied to individuals who have recently had myocarditis because of the potential risk of arrhythmia and heart failure.

  Special issuance may be granted 6 months after the illness if it has been transient and if the person has become asymptomatic, has a normal physical examination, and has an entirely normal laboratory evaluation.

Chronic Dilated and Restrictive Cardiomyopathy

Acute myocarditis may progress to a chronic cardiomyopathy. However, in most chronic forms of the disease, an acute phase either did not occur or was unrecognized. Chronic cardiomyopathy may affect myocardial function, and when it does, the classification into the dilated, restrictive, and hypertrophic types is helpful.

- Etiology

  Some causes of the chronic form of cardiomyopathy are: idiopathic; viruses; alcohol; collagen vascular diseases, including systemic lupus erythematosus, scleroderma, polyarteritis nodosa, and rheumatoid arthritis; neuromuscular diseases, such as progressive muscular dystrophy, Friedreich's ataxia and myotonia atrophica; familial cardiomyopathy; Chagas' disease; hemochromatosis; sarcoidosis; amyloidosis; and eosinophilic or Loeffler's disease.

  Those cases that are idiopathic or due to alcohol or previous viral infections are the most common and present as dilated cardiomyopathies. Of these, the idiopathic cases are most frequently seen.

  Hemochromatosis, sarcoidosis, amyloidosis, and eosinophilic (Loeffler's) myocarditis are uncommon types of cardiomyopathies; they produce a restrictive hemodynamic pattern.

- Natural history

  The chronic dilated and restrictive cardiomyopathies usually become progressively worse over a period of months or years, even with appropriate

medical management. However, their course is quite unpredictable. Heart failure and arrhythmias are their usual manifestations, but pulmonary or systemic emboli may occur, particularly in the presence of atrial fibrillation. Sudden death is not uncommon.

The term "ischemic cardiomyopathy" has been used by some to describe coronary artery disease with previous myocardial infarction and extensive myocardial scarring that presents predominantly with severe heart failure and little or no angina pectoris. The clinical presentation of this problem may be identical to any of the forms of dilated cardiomyopathy listed above. Strictly speaking, this is an incorrect use of the term "cardiomyopathy" because the true cardiomyopathy disorders are not of coronary artery origin.

o Recommendations for certification

Complete evaluation should include an ECG, chest radiograph, treadmill exercise test, echocardiogram, and a gated heart pool scan. Because of the high risk of cardiac arrhythmia, a 48 to 72 hour ambulatory ECG should be obtained. When heart failure is present and the diagnosis of coronary artery disease cannot be excluded, a coronary arteriogram may be indicated.

Certification for all classes of airmen should be denied to individuals who have a chronic dilated or restrictive cardiomyopathy because of the risk of sudden incapacitation from an arrhythmia, heart failure, or embolus.

Special issuance may be granted to those persons whose left ventricular function is only mildly reduced, as evidenced by a near-normal ejection

fraction and by a failure to reduce the ejection fraction with exercise during the gated heart pool scan.

In some cases, cardiomyopathy is suspected solely because of ECG abnormalities such as repolarization abnormalities, conduction disturbances, or cardiac arrhythmia. If there is no clinical or laboratory evidence to categorize the disorder as a dilated, restrictive or hypertrophic cardiomyopathy, recommendations for certification should be made according to the guidelines for those specific ECG abnormalities.

Any applicant with a history of any form of cardiomyopathy should be denied certification even though he or she may be asymptomatic and have no apparent residual effects of a previous cardiomyopathy. A complete evaluation including cardiovascular examination, chest radiograph, ECG, treadmill exercise test, echocardiogram, 24-hour ambulatory ECG, and gated heart pool scan at rest and with exercise should be performed. Special issuance may be granted when the only significant abnormality is a mild decrease in the left ventricular function, as evidence by a near-normal ejection fraction, and by a failure to reduce the ejection fraction with exercise during the gated heart pool scan.

Chronic Hypertrophic Cardiomyopathy

Hypertrophic cardiomyopathy is a myocardial disorder characterized by disproportionate hypertrophy of portions of the left ventricle and occasionally of the right ventricle, usually affecting the upper intraventricular septum more than the free wall. Occasionally it is concentric.[3] There is usually a bizarre form of myocardial fiber hypertrophy, which results in myocardial fiber and myofibrillar disarray.[7]

o Background

When this disease was first recognized, it was thought that its major consequences were due to obstruction of the left ventricular outflow tract.[8-11] It was called by many names, including idiopathic hypertrophic subaortic stenosis (IHSS), hypertrophic obstructive cardiomyopathy (HOCM), and muscular subaortic stenosis (MSS). Systolic gradients were found in the left ventricular outflow tract in many cases and it was therefore considered to be similar to valvular aortic stenosis. Subsequently, it has been appreciated that the myocardial disorder itself is the major problem and that true left ventricular outflow tract obstruction is not necessarily present.[12,13] Symmetrical left ventricular hypertrophy, and even apical hypertrophy, have been found and have therefore greatly expanded the specturm of this disorder.[14,15]

o Outflow tract obstruction

The parameter that previously had been used to measure outflow tract obstruction, the intraventricular pressure gradient, may not be an accurate indicator of obstruction, since in some cases the gradient has been found to be due to cavity obliteration without true obstruction.[16,17] There are certain features about the intraventricular pressure gradient of hypertrophic cardiomyopathy that make it different from the transvalvular pressure gradient of aortic stenosis. Unlike persons with valvular aortic stenosis, persons with hypertrophic cardiomyopathy have very rapid rates of ejection, even more rapid than subjects with normal hearts.[18] This rapid ejection is found in persons with hypertrophic cardiomyopathy regardless of whether or not a gradient is present. In some individuals, a gradient may be extreme-

variable and labile, being absent at rest but provocable with intropic drugs, the Valsalva maneuver, or amyl nitrite inhalation.[19] When an intraventricular pressure gradient is found, it is usually in individuals with asymmetric septal hypertrophy, but some individuals with symmetric left ventricular hypertrophy also have cavity obliteration and pressure gradients.[20] Intraventricular pressure gradients have been produced in normal animals by infusing subhypertensive levels of norepinephrine.[21] In some studies, a smaller gradient has actually been associated with a worse prognosis.[22] For these reasons, the term "obstructive" has been dropped from the original names of this disorder and it is now usually known as hypertrophic cardiomyopathy.

- Diastolic function

  A major hemodynamic defect in this cardiomyopathy is found during ventricular diastole[23-25] and is associated with prolongation of the isovolumetric relaxation time, delay in left ventricular filling, and a slowing of the rate of mitral valve opening. The diminished ventricular filling may reduce cardiac output, and the signs and symptoms of heart failure, which are infrequently present, are due to reduced left ventricular filling.

- Cardiac arrhythmias

  Ventricular arrhythmias are a major problem and the main cause of death in this disorder. Sudden death is frequent.[5] Atrial fibrillation often leads to congestive heart failure because the effect of the loss of atrial contractions is magnified due to the already existing impairment of ventricular filling from the hypertrophied ventricle.

o Establishing diagnosis

There is a broad clinical spectrum to this disease, ranging from mild to very severe. Those persons with the most severe disease may clearly demonstrate the clinical, echocardiographic, and angiographic features, but those with only mild or moderate disease have diagnostic features that are much less obvious. Furthermore, the difficulty in establishing a diagnosis is increased because there is no single criterion that is specific for the disease. The clinical, echocardiographic, and angiographic findings must all be considered and the weight of the evidence should then determine the suitablity of the diagnosis. Although there has been a great emphasis on the echocardiographic diagnosis, it should be stressed that all of the individual echocardiographic findings have also been seen in other conditions, and none of them alone, including the finding of disproportionate septal thickening or asymmetric septal hypertrophy, is diagnostic of hypertrophic cardiomyopathy. Angiographic evidence may be helpful in establishing the diagnosis when it is clearly present, but the effects of minor degrees of hypertrophy may not be apparent on the ventriculogram.

The clinical signs are a systolic ejection murmur at the left sternal border and apex; an arterial pulse of bifid character, known as "spike and dome;" and a palpable presystolic ventricular impulse. The echocardiographic findings are a disproportionate thickness of the interventricular septum, or asymmetric septal hypertrophy; a systolic anterior motion of the mitral valve appartus; a reduced diastolic closure rate of the mitral valve; a small left ventricular cavity; a displacement of the mitral valve towards the septum; and a midsystolic closure of the aortic valve. The angiographic findings are left ventricular cavity elimination in systole, a massive

hypertrophy of the free wall and septum, large papillary muscles, and an angulated banana-shaped ventricle in diastole.

Because of the wide range of abnormalities within this disease category and because methods of diagnosis are sometimes limited, we recognize varying levels of certainty in the diagnoses, ranging from "definite" to "probable" to "uncertain." The more positive the clinical, echocardiographic, and angiographic data, the greater the certainty of diagnosis. When all the findings are present, we can make a "definite" diagnosis, but if the evidence in all these categories is no more than equivocal, then the diagnosis must remain "uncertain." In the mid-ground lies a category of "probable" in which some, but not all, of the features in each category are present.

o Prognosis

People with hypertrophic cardiomyopathy have a shortened life expectancy, often dying suddenly and unexpectedly from a ventricular arrhythmia. Heart failure is relatively uncommon, but it frequently occurs after the onset of atrial fibrillation. McKenna et al followed 254 patients for up to 23 years (mean of 6 years): 48 died and 32 of those died suddenly.[26] Maron et al reported the clinical profiles on 78 patients who had either died suddenly or who had experienced a cardiac arrest and survived.[27] Seventy-one percent of those patients were less than 30 years old, and 54% had been on adequate doses of propranolol. The echocardiogram and ECG were no different in those patients who died than in age-and-sex matched controls with hypertrophic cardiomyopathy who had not died, so that there was no clinical sign or hemodynamic variable that could identify the high risk patient. However, McKenna et al have shown that individals with ventricular

arrhythmias and those with a family history of sudden death are in the high risk category.[28] A study of natural history of hypertrophic cardiomyopathy showed an overall mortality rate of 16% (31/190) and an annual mortality rate of 3.4% in those subjects who were not treated with surgery. Twenty-six of the 31 deaths were sudden.

Dividing subjects with hypertrophic cardiomyopathy into those "with obstruction" and those "without obstruction" is an unrealistic classification because the gradient is often quite variable, the methods of determining the extent of obstruction are not always reliable, and because there is a high risk of sudden unexpected death whether or not obstruction is present.

Beta adrenergic receptor blockade has improved symptoms of dyspnea and chest pain but has not provided an improvement in the treatment of arrhythmia.[30] Likewise, verapamil has not been effective in treating cardiac arrhythmias.[31] The only drug that has consistently helped arrhythmia is amiodarone,[31] and that drug is not yet available in the US.

Surgery has been helpful in improving the outflow tract gradient in many individuals and has improved their symptoms at a variable range of operative risk.[29,32-34] However, even in the surgical "successes" there has been no definite improvement in the long-term prognosis.[33,34]

- Recommendations for certification

  Complete evaluation should include a chest radiograph, ECG, treadmill exercise test, gated heart pool scan, echocardiogram, 24-hour ambulatory ECG, and left heart catheterization with left ventriculogram.

All applicants with definite hypertrophic cardiomyopathy should be denied certification.

All applicants with probable hypertrophic cardiomyopathy should also be denied certification. This recommendation is made because of the high risk of sudden death in hypertrophic cardiomyopathy and because individuals in this category have many of the clinical, echocardiographic and angiographic findings that are highly suggestive of the disease.

Special issuance may be made only if findings become less convincing and the person then qualified for the category of "uncertain" diagnosis of hypertrophic cardiomyopathy.

An individual in the "uncertain" category should be denied initially until a complete cardiac evaluation can be made. It should include a chest radiograph, ECG, treadmill exercise test, gated heart pool scan, echocardiogram, 48-hour ambulatory ECG, and left heart catheterization and left ventriculogram. If the diagnosis of definite or probable hypertrophic cardiomyopathy cannot be established, a certificate may be issued but the applicant should be re-evaluated every two years with a chest radiograph, ECG, treadmill exercise test, echocardiogram, and 48-hour ambulatory ECG.

Subjects with definite or probable hypertrophic cardiomyopathy who have had any of the forms of medical or surgical therapy should not be issued certificates on the basis of any expected improvement in their cardiac

status. None of the forms of therapy currently available have had any effect on improving survival rates and those subjects are still subject to sudden and unexpected death.

Pericardial Diseases

Introduction

Acute and chronic diseases of the pericardium are being recognized more frequently than before, mainly because echocardiography now provides a more accurate diagnosis. Persons with pericardial disease are at risk for sudden incapacitation because of pericardial tamponade, chest pain, arrhythmias, and reduced cardiac output. It is for these reasons that diseases of the pericardium are important to aviation safety.

Acute Pericarditis

Acute pericarditis is an inflammation of the visceral and/or parietal pericardium. It has numerous causes, and the natural history of the pericarditis depends to a great extent on the specific etiologic factor.

- Etiology

  The etiologies include idiopathic; infectious, either viral, bacterial, tuberculous, or fungal; acute rheumatic fever; acute myocardial infarction; uremia; collagen diseases, including rheumatoid arthritis, systemic lupus erythematosus, or polyarteritis; metastatic tumor; postmyocardial infarction syndrome; postpericardiotomy syndrome; myxedema; radiation, and trauma.

  The idiopathic and viral forms of pericarditis are quite similar. They are independent of any other underlying process and these types of pericarditis are the most common in aviation personnel. Chest pain, cardiac arrhythmia, and pericardial tamponade are all serious effects that occur frequently with sudden onset and are potentially disabling. It is for that reason that pericarditis is a hazard to aviation safety.

The diagnosis should be established with a complete evaluation including a chest radiograph, ECG and echocardiogram.

- Recommendations for certification

  Applicants for all classes who have acute pericarditis should be denied certification. Follow-up evaluation is required, including a chest radiograph, ECG, echocardiogram, and 24-hour ambulatory ECG for arrhythmia. Special issuance may be granted when all symptoms and signs have cleared and when there is no echocardiographic evidence of pericardial effusion or ECG evidence of serious arrhythmia.

  Acute pericarditis frequently recurs[35] and for that reason it has been called chronic relapsing pericarditis. Therefore, follow-up evaluation is required at one year after the initial episode and should include a chest radiograph, ECG, echocardiogram and 24-hour ambulatory ECG for arrhythmia.

Chronic Constrictive Pericarditis

Constrictive pericarditis may be idiopathic or due to viral pericarditis, hemopericardium, tuberculosis, cardiac surgery, and, rarely, uremic pericarditis.

Cardiac restriction is caused either by marked thickening of the pericardium, dense scarring of the pericardium with pericardial sac obliteration, or by calcification of the pericardium. The pathologic process commonly extends into the myocardium, occasionally causing a decrease in myocardial contractility,[36] but the major fault in constrictive pericarditis is impairment of diastolic filling of the ventricles.[37] Right and left ventricular diastolic pressures are elevated, leading to elevation of mean atrial pressures and congestion in both the systemic and pulmonary veins. However, the major

physical findings are related to systemic venous congestion; namely, cervical venous distention, hepatosplenomegaly, and leg edema.[38] Cardiac output is limited and cardiac arrhythmias, particularly atrial arrhythmias, are frequent, thereby placing pilots at risk for sudden incapacitation.

- Recommendations for certification

  Applicants with constrictive pericarditis in all classes should be denied certification. Special issuances should not be considered unless there has been successful surgical resection of the pericardium. In such cases, follow-up evaluation is required with chest radiography, ECG, treadmill exercise test, gated heart pool scan at rest and with exercise, echocardiogram, 24-hour ambulatory ECG, and right heart catheterization. Certification should be allowed if the following criteria are met: 1) intracardiac pressures are normal or only mildly elevated; 2) the ejection fraction is normal or near normal; 3) the ejection fraction does not diminish with exercise and; 4) no significant arrhythmias occur on treadmill exercise test and 24-hour ambulatory ECG.

  Follow-up re-evaluations should occur annually. Although right heart catheterization need not be repeated annually, all of the other tests should be done.

References

1. Harvey WP, Segal JP, Gurel T: The clinical spectrum of primary myocardial disease. Prog Cardiovasc Dis 1964;7:17-42.
2. Mattingly TW: Diseases of the myocardium (cardiomyopathies): The viewpoint of a clinical cardiologist. Am J Cardiol 1970;25:70-80.

3. Report of the WHO/ISFC Task Force on the definition and classification of cardiomyopathies. Br Heart J 1980;44:672-673.

4. Goodwin JF: Cardiac function in primary myocardial disorders. Br Med J 1964;1:1527-1533, 1595-1597.

5. Goodwin FJ: The frontiers of cardiomopathy. Br Heart J 1982;48:1-18.

6. Proceedings of the symposium on anthracycline toxicity. Cancer Treatment Reports 1978;62:855.

7. Maron J, Ferrans VJ, Henry WL, et al: Differences in distribution of myocardial abnormalities in patients with obstructive and nonobstructive asymmetric septal hypertrophy (ASH): Light and electron microscopic findings. Circulation 1974;50:436-446.

8. Teare RD: Asymmetrical hypertrophy of the heart in young adults. Br Heart J 1958;20:1-8.

9. Brock RC: Functional obstruction of the left ventricle (acquired aortic subvalvular stenosis). Guys Hosp Rep 1957;126:221-238.

10. Morrow AG, Braunwald E: Functional aortic stenosis: A malfunction characterized by resistance to left ventricular outflow without anatomic obstruction. Circulation 1959;20:181-189.

11. Goodwin JF, Hollman A, Cleland WP, et al: Obstructive cardiomyopathy simulating aortic stenosis. Br Heart J 1960;22:403-414.

12. Goodwin JF: An appreciation of hypertrophic cardiomyopathy. Am J Med 1980;68:797-800.

13. Criley JM, Lennon PA, Abbasi AS, Blaufus AH: Hypertrophic cardiomyopathy, in Levine HJ (ed): Clinical Cardiovascular Physiology, New York, Grune and Stratton, 1976.

14. Maron BJ, Bonow RO, Seshagiri TNR, et al: Hypertrophic cardiomyopathy with ventricular septal hypertrophy localized to the apical region of the left ventricle (apical hypertrophic cardiomyopathies). Am J Cardiol 1982;49:1838-1849.

15. Yamaguchi H, Ishimura T, Nishiyama S, et al: Hypertrophic nonobstructive cardiomyopathy with giant negative T-waves (apical hypertrophy): Ventriculographic and echocardiography features in 30 patients. Am J Cardio 1979;44:401-412.

16. Criley JM, Lewis KB, White RI Jr, et al: Pressure gradients without obstruction. A new concept of "hypertrophic subaortic stenosis." Circulation 1965;32:881-887.

17. Krasnow N, Rolett E, Hood WB Jr, et al: Reversible obstruction of the ventricular outflow tract. Am J Cardiol 1963;11:1-7.

18. Murgo JP, Alter BR, Dorethy JF, et al: Dynamics of left ventricular ejection in obstructive and nonobstructive hypetrophic cardiomyopathy. J Clin Invest 1980;66:1369-1381.

19. Braunwald E, Lambrew Cj, Morrow AG, et al: Idiopathic hypertrophic subaortic stenosis. American Heart Association, Monography No. 10, 1964.

20. Raizner AE, Chahine RA, Ishimori T, et al: Clinical correlates of left ventricular cavity obliteration. Am J Cardiol 1977;40:303-309.

21. Blaufus AH, Laks MM, et al: Production of ventricular hypetrophy simulating idiopathic hypertrophic subaortic stenosis (IHSS) by subhypertensive infusion of norepinephidrene (NE) in the consious dog. Clin Res 1979;23:77A.

22. Swan DA, Bell B, Oakley CM, et al: Analysis of symptomatic course and prognosis and treatment of hypetrophic obstructive cardiomyopathy. Br Heart J 1971;33:671-685.

23. Sanderson JE, Gibson DG, Brown DJ, et al: Left ventricular filling in hypertrophic cardiomyopathy: An angiographic study. Br Heart J 1977;39:661-670.

24. Sanderson JE, Traill TA, Sutton MG, et al: Left ventricular relaxation and filling in hypertrophic cardiomyopathy. An echocardiographic study. Br Heart J 1978;40:596-601.

25. Hanrath P, Mathey DG, Siegert R, et al: Left ventricular relaxation and filling pattern in different forms of left ventricular hypertrophy: an echocardiographic study. AM J Cardiol 1980;45:15-23.

26. McKenna W, Deanfield J, Faruqui A, et al: Prognosis in hypertrophic cardiomyopathy: Role of age and clinical electrocardiographic and hemodynamic features. Am J Cardiol 1981;47:1532-1538.

27. Maron BJ, Roberts WC, Epstein SE: Sudden deaths in hypertrophic cardiomyopathy: A profile of 78 patients. Circulation 1982;65:1388-1394.

28. McKenna WJ, England D, Doi YL, et al: Arrhythmia in hypertrophic cardiomyopathy: I. Influence on prognosis. Br Heart J 1981;46:168-172.

29. Shah PM, Adelman AG, Wigle ED, et al: The natural (and unnatural) course of hypertrophic obstructive cardiomyopathy - A multicenter study. Circulation Res 1975;34/35(suppl II):179-195.

30. McKenna WJ, Chetty S, Oakley CM, et al: Arrhythmia in nypertrophic cardiomyopathy: Exercise and 48-hour ambulatory electrocardiographic assessment with or without beta adrenergic blocking therapy. Am J Cardiol 1980;45:1-5.

31. McKenna WJ, Harris L, Perez G, et al: Arrhythmia in hypertrophic cardiomyopathy: II. Comparison of amiodarone and verapamil in treatment. Br Heart J 1981:46:173-178.

32. Morrow AG, Roberts WC, Ross J Jr, et al: Obstruction to the left ventricular outflow. Current concepts of management and operative tretment. Ann Int Med 1968;69:1255-1286.

33. Maron BJ, Merrill WH, Freier PA, et al: Long-term clinical course and symptomatic status of patients after operation for hypertrophic subaortic stenosis. Circulation 1978;57:1205-1213.

34. Trajik AJ, Giuliani ER, Weidman WH, et al: Idiopathic hypertrophic subaortic stenosis: Long-term surgical follow-up. Am J Cardiol 1974;34:815-822.

35. Shabetai R: The Pericardium, New York, Grune and Stratton, 1981.

36. Dines DE, Edwards JE, Burchell HB: Myocardial atrophy in constrictive pericarditis. Proc Staff Meeting Mayo Clinic, 1958;39:93-99.

37. Shabetai R, Fowler NO, Guntheroth WG: The hemodynamics of cardiac tamponade and constrictive pericarditis. Am J Cardiol 1970;26:480-489.

38. Boltwood CM, Shah PM: The pericardium in health and disease. Current Problems in Cardiology 1984;9(5).

Valvular Heart Disease

General Considera[illegible]

Any applicant who has a heart murmur other [illegible]n innocent pulmonary ejection murmur or functional left heart ejection (outflow) murmur as described below should have a two dimensional and M-mode echocardiogram and an evaluation by a board-certified cardiologist or internist.[1-4]

Murmurs occurring in the absence of cardiac disease, that is, innocent or functional murmur, must be distinguished from those due to organic disease. The final determination that a murmur is innocent depends upon the absolute exclusion of any cardiac abnormality, and this may not always be possible. Absence of radiographic, electrocardiographic and echocardiographic abnormalities, while not absolute proof of the functional nature of the murmur, are important corraborative findings.

The innocent pulmonary ejection murmur is usually short and is due to an increased flow or enhanced audibility of turbulent flow in the pulmonary outflow tract. Such a murmur is usually found in young individuals or in persons with conditions like pectus excavatum or the straightback syndrome. Since the murmur is similar to that of mild pulmonary valve stenosis and atrial septal defect, normal splitting of the second heart sound is an important feature of the innocent murmur. If the applicant has no symptoms and if the chest radiograph and electrocardiogram are normal, this murmur should not disqualify an applicant. If there is any question about the etiology of the murmur, a two dimensional and M-mode echocardiogram should be performed, and if it is normal, a certificate should be issued.

The functional left heart ejection murmur is also characterized by its early timing in systole, by the absence of an ejection click, and by a normal electrocardiogram and chest radiograph. The murmur may also be heard at the cardiac apex.

Diastolic murmurs never occur in the normal heart. Two dimensional and M-mode

echocardiography should be performed to exclude aortic leaflet eccentricity or thickening and organic causes of the murmur, such as hypertrophic cardiomyopathy or aortic valvular stenosis. If the two-dimensional and M-mode echocardiogram (and Doppler evaluation, if available) are normal, in addition to a normal chest radiograph and electrocardiogram, this murmur would not disqualify an asymptomatic applicant.

Detection of valvular heart disease is dependent upon the competence of the physician conducting the physical examination. Persons with suspected valvular abnormalities should be evaluated more completely by a consultant in cardiology. The clinical evaluation should include a careful history for both rheumatic fever and other conditions known to cause valvular heart disease (Table 1). The history should seek a past knowledge of cardiac murmurs, palpitation, cardiac rhythm disturbances, syncope, episodes of transient cerebral ischemia and symptoms of angina, dyspnea, effort fatigue, paroxysmal dyspnea, and hemoptysis. The physical examination should include measurement of the blood pressure in both arms, evaluation of the jugular vein and peripheral arterial pulsations, and careful inspection, palpation, and auscultation of the heart. The cardiac examination should be made with the patient in the supine, sitting and left semi-decubitus positions. The examining clinician should be prepared to evaluate the effects of exercise, position changes, and the Valsalva maneuver if systolic murmurs suggesting organic origin are present at the base or apex of the heart.

Screening noninvasive studies should include a postero-anterior and left lateral chest radiograph and a 12-lead electrocardiogram. If palpitation or cardiac irregularities are suggested by the history or examination, a 24-hour Holter electrocardiographic monitoring procedure should be performed. Standard graded treadmill exercise testing may be advisable in selected patients with valvular heart disease. This is particularly true if there is a history suggesting angina pectoris or concern about the functional capacity of the individual. The study may also be useful in persons with a history of cardiac arrhythmias.

Echocardiography has become the single most important noninvasive study. It is particularly valuable in supporting the diagnosis of mitral valve stenosis, mitral valve prolapse, hypertrophic cardiomyopathy, aortic valve disease, some types of mitral valve incompetence, and tumors of the heart. In addition, it gives an accurate estimate of chamber size, wall thickness, and ventricular function. The echocardiographic study should include both M-mode and two-dimensional procedures. Doppler evaluation, in addition, gives valuable information regarding gradients and regurgitant fractions. Phonocardiography is commonly combined with echocardiography and is useful in accurately showing the relationship of abnormal sounds to the heart sounds, as well as documenting the audible murmurs. Radionuclide studies are valuable in assessing left ventricular function in persons with aortic and mitral valve regurgitation.

Invasive examinations such as cardiac catheterization and angiography, including coronary angiography, may occasionally be indicated. The addition of two-dimensional echocardiography with Doppler studies has significantly reduced the indications and need for cardiac catheterization in the evaluation of valvular heart disease. The need to evaluate the coronary arteries in the presence of valvular heart disease is now one of the most important and common reasons for doing cardiac catheterization with ventriculography and coronary arteriography.

Complications of Valvular Heart Disease

The risk of these disorders constitutes the basis for disqualification. Infective endocarditis can complicate any valvular lesion with the possible exception of mitral stenosis. The most serious complications of valvular disease are cardiac decompensation secondary to destruction of valvular tissue, valvular insufficiency, and systemic emboli.

Cardiac decompensation and acute pulmonary edema may be precipitated in individuals with valvular disease by unusual physical or emotional stresses or the onset of supraventricular tachycardia.

Systemic embolization occurs most often in the presence of mitral stenosis, having been noted in 10% to 20% of persons with this lesion. It most often occurs in the presence of a significantly enlarged left atrium and chronic atrial fibrillation, but occasionally it is noted with only minor degrees of obstruction and no recognized or sustained atrial fibrillation.

Atrial arrhythmias are most likely to occur with mitral valve disease because of its effect on left atrial anatomy and function. However, they are not uncommon in persons with aortic valve disease. The sudden onset of supraventricular tachycardia could lead to syncope or an inability to perform all airman tasks properly. Ventricular arrhythmias are associated with myocardial ischemia, and may occur with severe aortic stenosis. They also may be noted in association with the mitral valve prolapse syndrome. When ventricular ectopy is advanced, it may lead to near-syncope or syncope.

Myxomatous valvular degeneration, which occurs in Marfan's syndrome and in some cases of the mitral valve prolapse syndrome, may be associated with valve or chordal rupture leading to acute cardiac decompensation.

Acute aortic dissection with associated acute aortic valve incompetence, acute coronary insufficiency, cerebral ischemia, and aortic rupture, is a significant complication of certain hereditary connective tissue disorders, such as Marfan's syndrome. Careful follow-up is indicated in persons with these disorders despite the apparent absence of any cardiovascular abnormalities.

Mitral Stenosis

The classic history of mitral stenosis is for ausculatory evidence of the disease first to be noted about one decade after an acute attack of rheumatic fever. Another decade passes before the onset of symptoms, and another decade before the onset of disabling symptoms.[5-7] The disease has become less frequent, and this classic pattern is seen less often; a milder form is now seen in older persons. The valve area must be

reduced by one-half before symptoms normally develop. Asymptomatic individuals may develop acute pulmonary edema with the onset of atrial fibrillation, however. Atrial fibrillation becomes chronic in over 50% of patients with mitral stenosis.[8] Occasionally a systemic embolus is the first sign of the disease. Ten percent to 20% of persons with mitral stenosis, including those with mild disease, will develop systemic emboli.[9] Since the rheumatic process tends to be progressive and since the valve area is usually reduced by 50% or more before the diagnosis is made clinically, persons with this diagnosis, even though they are asymptomatic and have no history of surgery, should usually be disqualified. The rare individual with congenital mitral stenosis must meet the same standards as the individual with rheumatic mitral stenosis.

A person who may be considered for special issuance is an asymptomatic individual who has mild disease, as evidenced by an A_2-opening snap interval of 10 seconds or more, and a presystolic and short mid-diastolic murmur on physical examination; who has mild orifice reduction, pliable valve, and slight left atrial enlargement on echocardiography; and who performs a good stress test, and has normal sinus rhythm and no significant arrhythmia. Rarely, such a person may need an exercise study with the determination of wedge and pulmonary artery pressure. Another person who may be considered for special issuance is an individual who has had a successful mitral valvotomy, who is asymptomatic and has a normal sinus rhythm, no significant mitral regurgitation, and no postoperative atrial thrombi. The echocardiogram should show only mild mitral valve narrowing and thickening and no more than mild left atrial enlargement. The stress test must show normal exercise tolerance and the Holter study no significant arrhythmia. Follow-up should be yearly with cardiovascular evaluation, echocardiography, stress test, and Holter study.

Class I and II airmen may be considered for special issuance especially if they perform airman duties in situations in which there is crew redundancy.

Mitral Regurgitation

While mitral stenosis is almost invariably due to rheumatic fever, mitral regurgitation has a host of etiologies, which are usually grouped under four primary headings.[10] The first is disorders of the leaflets. These may be due to disease processes, such as rheumatic fever, infective endocarditis, trauma, and spontaneous rupture; abnormalities in development, such as clefts, redundant valves, or anomalous attachments of the leaflet; and defects in the connective tissue, such as occurs in Hurler's, Marfan's and Ehlers-Danlos syndromes. The second main heading is that of disorders of the annulus, which may be due to dilatation, calcification, destruction from infection, and disruption of the ring from prosthetic valves. The third major heading is disorders of the chordae tendineae, including rupture, which is most often idiopathic, but which may be due to infective endocarditis, trauma, Marfan's syndrome, and myocardial infarction. Thickened chordae occur in some congenital mitral valve problems of the atrioventricular cushion type, and also with a hypoplastic left ventricle. Occasionally the chordae become elongated, leading to mitral regurgitation. Chordae may also arise from unusual locations. The fourth major heading is disorders of the papillary muscle. This includes dysfunction or rupture from myocardial infarction, abscess, trauma, malalignment, and congenital abnormalities.

The degree of regurgitation depends on the size of the regurgitant orifice and the gradient of pressure between the left ventricle and left atrium. Both the size of the orifice and the gradient are labile.

In persons with rheumatic mitral regurgitation, the interval between rheumatic fever and symptoms of regurgitation is longer than with mitral stenosis, often exceeding two decades. Other differences are that the incidence of pulmonary edema is lower and hemoptysis and systemic emboli are much less common than with mitral stenosis.[11] Persons with mild mitral regurgitation may remain asymptomatic forever.[12] The

majority with the rheumatic type will have only mild impairment unless there is a complication of recurrent acute rheumatic fever, infective endocarditis or ruptured chordae.[12] The natural history will vary considerably depending on the underlying etiology, the volume of regurgitation, and the state of the myocardium. While there is no progression in many individuals, severe regurgitation may result from infective endocarditis or ruptured chordae tendineae. Progression is also more rapid when the disorder is due to connective tissue disease, such as Marfan's syndrome. In one unselected group of patients with mitral regurgitation treated medically from the time of diagnosis, 80% were alive after five years and 60% after 10 years. With severe regurgitation, however, the five-year survival rate is less than 50%.

Therefore, we recommend that all individuals with severe mitral regurgitation and most individuals with moderately severe mitral regurgitation should be disqualified.

Since mild and occasionally moderate mitral regurgitation, usually of rheumatic origin, has an excellent prognosis, a person with this lesion may be qualified for flying. Such a person should be asymptomatic and have only a grade 3/6 apical systolic murmur. A third heart sound may be present, but there must be no diastolic murmur. The radiograph should show no more than slight cardiac enlargement and no more than slight enlargement of the left atrium by echocardiography. Echocardiography or a left ventricular angiogram should show normal left ventricular function and no evidence of mitral stenosis. The electrocardiogram should show no more than minor atrial abnormalities. A maximal graded treadmill test should produce no significant arrhythmia and no decrease in exercise tolerance. In rare instances hemodynamic studies may be necessary to establish the modest degree of mitral regurgitation. If so, these should show normal pressures and the ventriculogram should disclose no more than 2+ mitral regurgitation. Follow-up examinations should be yearly, with a cardiovascular evaluation, echocardiogram, stress test, and Holter study.

Class I and II airmen may be considered for special issuance especially if they

perform airman duties in situations in which there is crew redundancy.

Mitral Valve Prolapse

This represents a spectrum of disorders from simple, mild redundancy of the mitral leaflet to a severe degree of valve prolapse that is often associated with myxomatous changes in the valve tissue.[14,15] In the majority of instances, mitral valve prolapse is characterized by a mid-systolic crescendo murmur, which may have to be elicited by causing variations in ventricular volume. In the majority of persons with this syndrome who are asymptomatic and have a normal cardiac silhouette and echocardiogram (except for the redundancy of the mitral valve leaflets), no treatment except prophylaxis for infective endocarditis is indicated and there is no contraindication to flying.[16] In those persons with more serious mitral valve prolapse, and consequently more significant mitral valve regurgitation, the same criteria apply as with mitral regurgitation.[17]

Therefore, we recommend that persons who are asymptomatic, who on physical examination are found to have only a systolic click or a click and a late apical systolic murmur, who on echocardiogram have mild to moderate prolapse of the leaflets with no associated abnormalities, and who have no significant arrhythmias on Holter monitoring, should be certified. Follow-up examinations should occur yearly with a cardiovascular evaluation, stress test and Holter monitoring; the echocardiogram should be repeated every three years.

Class I and II airmen may be considered for special issuance especially if they perform airman duties in situations in which there is crew redundancy.

Those persons with significant and complex atrial and ventricular arrhythmias should be evaluated as other individuals with arrhythmias. A very small number of persons, especially those with a family history of significant mitral disease, have the potential for sudden death. These persons should be disqualified. Those individuals with

mitral valve prolapse syndrome who are asymptomatic but who have ST segment depression and T wave inversion on resting ECG, and who have a normal thallium perfusion study or radionuclide stress test (MUGA), should not be disqualified, but should have an annual re-evaluation.

Mitral Valvuloplasty

Occasional pilots may be considered for special issuance. They should be asymptomatic, have a normal sinus rhythm, have no more than slight cardiomegaly and left atrial enlargement, normal left ventricular function by echocardiogram, a normal stress test, and no significant arrhythmia by Holter monitoring.

Follow-up evaluation should be yearly, with a cardiovascular evaluation, normal stress test and no significant arrhythmia by Holter monitoring; echocardiography should be done every three years.

Class I and II airmen may be considered for special issuance especially if they peform airman duties in situations in which there is crew redundancy.

Tissue Valves

The development of tissue valve replacements in the past decade has resulted in a decrease in the incidence of thromboembolic complications compared with mechanical valve prostheses, but the durability of the tissue valve seems to be less than that of mechanical prostheses.[18] The majority of thromboembolic episodes with tissue valves in the mitral position occur in the first 6 to 12 weeks following surgery, and consequently, anticoagulant treatment is recommended during this period.[19] The anticoagulant is discontinued unless there are persisting thrombogenic factors not related to the prosthesis, such as atrial fibrillation, a markedly dilated left atrium, a calcified left atrial wall, a postoperative thromboebolic event, or a clot present in the atrium at the time of operation.[20] If any of these additional thrombogenic factors are present, the

person should be on chronic anticoagulant treatment. Despite anticoagulant treatment, the incidence of postoperative systemic emboli with the porcine valve in the mitral position is three times as high in persons with concomitant atrial fibrillation as in those with normal sinus rhythm.

We recommend that, in view of these problems, most persons with tissue valves should be excluded from piloting.

A possible exception is an individual who is asymptomatic, has normal sinus rhythm, and is free of evidence of significant mitral regurgitation or valve obstruction. In addition, the cardiac silhouette should not be significantly enlarged on the chest radiograph. There must be no anticoagulant treatment and no history of thromembolic complications. Consideration for approval would be deferred until one year after the operation. Evaluation would include a cardiovascular evaluation, Holter monitoring, treadmill stress test, and echocardiography. Follow-up should be yearly with all the above studies.

Aortic Stenosis

Aortic stenosis may be congenital or acquired, and the latter is usually due to rheumatic fever.

Minimal Aortic Valvular Stenosis or Sclerosis

Airmen discovered to have minimal aortic valve stenosis or sclerosis can be considered for certification if they are asymptomatic and have no evidence of significant outflow tract obstruction on physical examination. That is, the applicant has a short systolic murmur of grade 3/6 of less, a normal carotid upstroke, no palpable fourth heart sound, and no left ventricular hypertrophy on physical examination, electrocardiography, or chest radiograph. A graded submaximal treadmill test should be normal. Occasionally persons with a high pressure gradient due to a small valve area can present with a normal

electrocardiogram and physical findings suggesting mild aortic valvular stenosis. Two-dimensional, M-mode and Doppler echocardiography are invaluable in the assessment of the severity of aortic valvular stenosis. Cardiac catheterization would ordinarily not be necessary in the asymptomatic pilot when all other studies indicate that the left ventricular-aortic gradient would be mild, that is, less than 20 mm Hg. If the echocardiographic study is equivocal, cardiac catheterization studies should be performed. The left ventricular-aortic systolic gradient should be less than 20 mm Hg at rest with a normal cardiac index and a normal left ventricular end-diastolic pressure of 12 mm Hg. If chest pain is a symptom, coronary arteriography should be done and should reveal no more than minor luminal irregularities.

Therefore, we recommend that persons with minimal aortic valve sclerosis or stenosis may be certified. Yearly re-evaluation is mandatory, since the rate of progression of this disease is not well documented.[21]

Mild Aortic Valvular Stenosis

For Class III applicants with mild aortic valve stenosis, certification should be given if: 1) based on cardiac catheterization or Doppler echocardiographic studies the left ventricular-aortic gradient is less than 40 mm Hg at rest with a normal cardiac index; 2) the chest radiograph and resting electrocardiogram show no significant left ventricular hypertrophy or ST segment or T wave changes; 3) left ventricular function is normal on two dimensional and M-mode echocardiography; 4) a near normal maximal graded treadmill test shows no significant arrhythmias or significant ST segment changes, and the predicted heart rate response and level of exercise are within normal limits for the person's age; and 5) 24-hour Holter monitoring also shows no significant arrhythmias. An annual cardiovascular evaluation would be necessary for renewal of the certificate. There should be no evidence of clinical change on re[illegible]yearly examination. If clinical change is noted, cardiac catheterization [illegible]pler

echocardiographic studies would be indicated.

We recommend that Class I or Class II certificates generally should not be issued to applicants with mild aortic valvular stenosis, but could be granted by a special issuance with regular follow-up, especially if these pilots perform their duties as airmen in situations in which there is crew redundancy.

Moderate or Severe Aortic Valve Stenosis

This is defined by a resting left ventricular-aortic gradient of 40 mm Hg or greater, with a normal cardiac index. There is abundant evidence of a poor prognosis for symptomatic individuals with severe or moderately severe aortic valve stenosis.[3,22,23] These individuals are usually candidates for valve replacement, and sudden death is highest in the group that has symptoms.[22] The time from appearance of symptoms to death is usually short. The asymptomatic person with severe aortic stenosis also has a poor prognosis and is also subject to sudden death.

We recommend that all pilot applicants with moderate or severe aortic stenosis should be disqualified.

Aortic Valvular Regurgitation

Aortic valvular regurgitation can be either congenital or acquired, and the etiology should be identified because of differences in prognosis. Congenital causes may be associated with other lesions, such as coarctation of the aorta, a patent ductus arteriosus or a ventricular septal defect. Acquired causes include rheumatic fever, syphilis, dissecting aneurysm, bacterial endocarditis, hypertension, cystic medial necrosis, connective tissue disorders such as Marfan's or Ehlers-Danlos syndrome, and rheumatoid spondylitis.

Mild Aortic Valvular Regurgitation

Persons with mild aortic valvular regurgitation may be considered qualified when they are asymptomatic and have had a normal cardiovascular evaluation, which includes: 1) a normal blood pressure, that is, a pulse pressure equal to or less than 55 mm Hg and a diastolic pressure equal to or greater than 65 mm Hg; 2) a normal chest radiograph and electrocardiogram with no evidence of cardiac enlargement; 3) a normal graded submaximal treadmill test to 90% of the predicted heart rate, or which is symptom limited; 4) a two-dimensional echocardiographic study that excludes disproportionate aortic root dilatation. (If disease of the aortic root is suspected, an aortogram may be necessary. Diseases of the aortic root due to connective tissue disorders such as Marfan's or Ehlers-Danlos syndromes, thoracic dissection or cystic medial necrosis have a poor and unpredictable prognosis and should be disqualified); 5) hemodynamic studies that may be required to confirm the mild nature of the aortic valve regurgitation in some instances. These studies should demonstrate normal intracardiac pressures, defined as a mean pulmonary artery pressure of 20 mm Hg, a mean pulmonary arterial wedge pressure of 15 mm Hg or less, and a left ventricular end-disatolic pressure of 15 mm Hg or less; and an absence of a left ventricular-aortic valve gradient both at rest and on supine exercise. An aortogram should reveal 2+ or less regurgitation or a regurgitant fraction of less than 25% of the left ventricular stroke volume.

Therefore, we recommend that approval for certification may be granted to applicants with mild aortic valvular regurgitation if there are no other disqualifying factors; regular follow-up examination should be required.

Moderate Aortic Valvular Regurgitation

Applicants with moderate aortic valve regurgitation, that is individuals who have slight cardiomegaly and no symptoms, may be considered for Class III certificates if

electrical stability is confirmed on an electrocardiogram and if a radionuclide angiogram or two-dimensional sector echocardiogram shows a normal ejection fraction and no significant decrease in the ejection fraction with exercise. A near maximal graded treadmill test should demonstrate a normal exercise tolerance with a normal heart rate response for the applicant's age. If the certificate is granted, an annual examination is required to document a stable clinical course. Class I or II airmen may be considered for special issuance especially if they perform airman duties in situations in which there is crew redundancy.

Severe Aortic Valvular Regurgitation

All persons should be permanently disqualified for all classes of certification.[24]

Aortic Stenosis with Aortic Regurgitation

Persons with both aortic stenosis and aortic regurgitation should be disqualified unless they qualify under the separate criteria for aortic stenosis and aortic regurgitation.

Combined Valvular Lesions

Persons with mitral stenosis combined with aortic stenosis or aortic insufficiency, or mitral regurgitation combined with aortic stenosis or aortic regurgitation, should be disqualified unless they qualify under the separate criteria for each valvular disorder.

Other Acquired Valvular Disease

Tricuspid stenosis and tricuspid regurgitation of rheumatic origin are almost always associated with mitral or aortic valve disease, or both, and are disqualifying. Occasionally, tricuspid regurgitation of nonrheumatic origin that is secondary to trauma or infectious endocarditis is mild and the person has a normal-sized heart and a normal

systemic venous pressure. Persons with this condition have an excellent prognosis and are qualified for flight certification after the findings are confirmed by a cardiovascular consultant.

Pulmonic valve insufficiency and tricuspid valve insufficiency that are secondary to pulmonary hypertension are disqualifying.

Mechanical Heart Valves

Valve replacement has proven successful in improving well being and exercise tolerance, as well as in prolonging life.[25-27] However, the complications and problems of valve replacement continue to be a major concern. Long-term studies have shown a 31% mortality in the first three years following aortic valve replacement and a 15% mortality in the first year following mitral valve replacement.[26] The most common modes of death are sudden death after aortic valve replacement and cardiac failure following mitral valve replacement.[26] In a 15-year follow-up 44% of patients with aortic valve replacement and 48% of patients with a mechanical mitral valve replacement had experienced a thromboembolic event;[26] thus, thromboembolism remains a persistent problem in patients with mechanical heart valves, occurring at a rate of 1.5-2.0% per year.[25] In addition, there are reports of strut fractures and other late structural complications.

Therefore, we recommend that all applicants with a mechanical artificial valve of any type are disqualified to pilot aircraft.

Summary

We are recommending four changes from the Bethesda Conference of 1975:[28] 1) the majority of persons with tissue artificial valves should be disqualified, but certain exceptions with tissue valves could be made by special issuance. The First United Kingdom Workshop in Aviation Cardiology recommended the certification of certain

applicants with a homograft valve replacement.[2] 2) Most applicants with mitral valve stenosis should be disqualified. Unusual exceptions may be made, such as mild mitral valve stenosis and after valvotomy. 3) Applicants who have had successful valvuloplasty for mitral regurgitation may be certified if there are no other disqualifying factors. 4) Follow-up periodic re-evaluations for recertification are mandatory for all pilots with demonstrated valvular disorders.

Four other recommended changes deviate from both the Bethesda Conference and the United Kingdom Workshop: 1) Class I and II applicants with mild aortic valve stenosis (defined as 20 to 40 mm Hg left ventricular-aortic systolic gradient) may be qualified by special issuance if there is crew redundancy; 2) Class I and II applicants with moderate aortic valve insufficiency may be certified if there is crew redundancy; 3) applicants with moderate mitral regurgitation may be qualified by special issuance; and 4) pilots with mitral valve prolapse may be certified in most instances, providing they have no significant arrhythmias and they have no other contraindications related to their mitral regurgitation.

There is one recommended change from the United Kingdom Workshop which agrees with the Bethesda recommendations: applicants with mild mitral regurgitation may be qualified by special issuance. The United Kingdom Workshop has stricter qualifications if the etiology is related to rheumatic heart disease or coronary artery disease. There have been no recommended changes from the previous Bethesda Conference in certain categories: 1) persons with any murmur other than an innocent or functional murmur should have a cardiovascular consultation and periodic follow-up examinations. 2) Persons with minimal aortic valve sclerosis or stenosis may be certified. 3) Moderate or severe aortic stenosis is a disqualifying condition. 4) Severe aortic and mitral valve regurgitation are disqualifying disorders. 5) Diseases of the aortic root are a disqualifying disorder. 6) Applicants with mechanical heart valves should be disqualified.

Table 1

Classification of Causes of Valvular Heart Disease

Congenital

Rheumatic

Infectious

- Syphilitic
- Infective endocarditis

Traumatic

Heritable disorders, such as Marfan's syndrome

Tumors (myxoma)

Rheumatoid arthritis and ankylosing spondylitis

Carcinoid heart disease, right and left valvular lesions

Degenerative

- Calcified aortic stenosis
- Degenerative and inflammatory lesions of the root with secondary aortic regurgitation

Mitral valve prolapse

- Ruptured chordae tendineae (multiple causes)
- Hypertrophic cardiomyopathy

References

1. Dreifus LS: Introduction to Eighth Bethesda Conference on cardiovascular problems associated with aviation safety. Am J Cardiol 1975;36:617-620.
2. Joy J: The First United Kingdom Workshop in Aviation Cardiology. Eur Heart J 1984;5:5-6, 135-147.

3. Hancock EW: Diseases of the aortic valve: Diagnosis, natural history and surgical treatment. Cardiology Series, Baylor College of Medicine 1983;6:5-24.

4. Waller BF: Morphologic aspects of valvular heart disease. Current Problems in Cardiology 1984;9:6-66.

5. Wood P: An appreciation of mitral stenosis. Brit Med J 1954;1:1051.

6. Rowe JC, Bland F, Sprague HB, White PD: Course of mitral stenosis without surgery: Ten and twenty year perspectives. Annals Int Med 1960;52:741.

7. Selzer A, Cohn KE: Natural history of mitral stenosis: A review. Circulation 1972;45:878-890.

8. Probst P, Goldschlager N, Selzer A: Left atrial size and atrial fibrillation in mital stenosis: Factors influencing their relationship. Circulation 1973;48:1282-1287.

9. Coulshed N, Epstein EJ, McKedrick CS, et al: Systemic embolism in mitral valve disease. Brit Heart J 1970;32:26-34.

10. Pierpont GL, Talley RC: Pathophysiology of valvular heart disease. Arch Int Med 1982;142:998-1001.

11. Reichek N, Shelburne JC, Perloff JR: Clinical aspects of rheumatic valvular disease. Prog Cardiovasc Dis 1973;14:491-537.

12. Allen H, Harris A, Leatham A: Significance and prognosis of an isolated late systolic murmur. Brit Heart J 1974;36:525-532.

13. Rappaport E: Natural history of aortic and mitral valve disease. Am J Cardiol 1975;35:221-227.

14. Jeresaty RM: Mitral valve prolapse-click syndrome. Prog Cardiovasc Dis 1973;15:623-652.

15. Cheitlin MD, Byrd RC: Prolapsed mitral valve: The commonest valve disease? Curr Prob Cardiol 1984;8:1-54.

16. Devereux RB, Perloff JK, Reichek N, Josephson MD: Mitral valve prolapse. Circulation 1976;54:3-11.

17. Jeresaty RM: Sudden death in the mitral valve prolapse-click syndrome. Am J Cardiol 1976;37:317-318.

18. Cohn LH, Mudge GH, Pratter F, Collins JJ: Five to eight year follow-up of patients undergoing porcine heart valve replacement. N Engl J Med 1981;304:258-262.

19. Edmunds LH Jr: Thromboembolic complications of current cardiac valvular prostheses. Ann Thorac Surg 1982;34:96-106

20. Janusz MT, Jamieson WRE, Burr LH, et al: Thromboembolic risks and role of anticoagulants in patients in chronic atrial fibrillation following mitral valve replacement with porcine bioprosthesis. J Am Coll Cardiol 1983;1:587.

21. Harries AD: Assessment of chronic aortic valve disease in adults. Post Grad Med J 1982;58:1-3.

22. Frank S, Johnson A, Ross J Jr: Natural history of valvular aortic stenosis. Br Heart J 1973;35:41-46.

23. Shah PM, Roberts DL: Diagnosis and treatment of aortic valve stenosis. Curr Probl Cardiol 1977;2:1-49.

24. Bonow RO, Rosing DR, McIntosh CL, et al: The natural history of asymptomatic patients with aortic regurgitation and normal left ventricular function. Circulation 1983;68:509-517.

25. McClung JA, Stein JH, Ambrose JA: Prosthetic heart valves: A review. Prog Cardiovasc Dis 1983;26:237-270.

26. McGoon MD, Fuster V, McGoon DC, et al: Aortic and mitral valve incompetence: Long-term follow-up (10 to 19 years) of patients treated with a Starr-Edwards prosthesis. J Am Coll Cardiol 1984;3:930-938.

27. Schwarz F: The effect of aortic valve replacement on survival. Circulation 1982;66:1105-1110.

28. Eighth Bethesda Conference on Cardiovascular Problems Associated with Aviation Safety Task Force IV: Valvular heart disease. Am J Cardiol 1975;36:617-620.

Congenital Heart Disease

Introduction

The following recommendations for applicants for medical certification as pilots are based on what is known about the natural history of congenital heart disease and about the long-term follow-up of post-operative patients. Regrettably, there are few specific data available about the risk of sudden death or incapacitation. This is true whether a defect has been corrected surgically or not. Since 1954 both early and late mortality results of surgery for congenital heart disease have steadily improved.[1] In addition, there are data to indicate that the older a person is at the time of surgical correction, the greater the chance of a late complication. This has been demonstrated for coarctation of the aorta,[2] tetralogy of Fallot,[3] and atrial septal defect.[4] Therefore it is difficult to make categorical judgments regarding the suitablity of individuals for pilot certification who have had heart disease or who have had surgical correction of a heart defect.

Success in the operative treatment of congential heart disease, with resulting increased longevity, will undoubtedly result in increasing numbers of persons with congenital heart disease who apply for flight training. It is hoped that applicants suspected of having congenital cardiovascular disease will be identified and evaluated prior to entry into pilot training. Therefore, the question of recertification of airmen with congenital heart disease will depend upon serial evaluation of existing residua. For this reason, it is particularly important that initial screening be completed by a physician who is knowledgeable in the field of cardiovascular disease.

Diagnostic Evaluation

The presence of congenital heart disease should not automatically disqualify applicants as medically unfit for pilot certification. This decision should be based on the

specific anatomic diagnosis and its severity and whether the condition has or has not been treated surgically. The decision for certification should consider not only the applicant's present status but also the possibility of late onset of functional derangements or impairment. These judgments may be modified as more data become available on postoperative patients.[5] When initial screening by an AME suggests the presence of a congenital cardiac anomaly, the applicant should be referred for further cardiac evaluation. The definitive diagnosis of the condition and its severity should be established by a cardiologist expert in the field of congenital heart disease. This evaluation should utilize appropriate noninvasive as well as invasive techniques.

The following noninvasive studies should be carried out in every applicant suspected of having congenital heart disease: 1) a complete history and physical examination; 2) a 12-lead electrocardiogram; and 3) a routine chest radiograph. If these procedures suggest the presence of a congenital heart defect, two-dimensional echocardiography should also be done. In those persons considered at risk for the development of arrhythmia, continuous electrocardiographic monitoring at rest and during exercise should be carried out. Stress testing is useful in most cases of significant congenital heart disease. Individuals who have had open heart surgery should have stress testing in order to evaluate cardiac function and in order to elicit cardiac rhythm disturbances. Intracardiac electrophysiologic studies may be necessary to evaluate certain conditions properly. In selected cases, hemodynamic studies with contrast visualization should be performed.

Recommendations

Applicants for certification are likely to be those with mild forms of congenital heart disease for whom surgery is not indicated or in whom the condition has spontaneously resolved, such as spontaneous closure of a ventricular septal defect. Also, those persons who previously have had surgical repair of a malformation may apply for

pilot training. Persons with more significant congenital heart disease are not likely to apply for a Class I or Class II certificate but might, nevertheless, apply for a Class III certificate. Persons with uncorrected cyanotic congenital heart disease should not be qualified for any class because of their already compromised oxygen concentration. Altitude would further decrease arterial oxygen tension, placing the individual at risk to the effects of hypoxemia. Similarly, individuals with pulmonary hypertension should not be certified. By decreasing arterial oxygen tension, altitude will cause increased pulmonary resistance and further raise pulmonary arterial pressure. Such individuals would be at risk to right heart failure and even sudden death.[6]

The presence of an isolated, small, left-to-right shunt, defined as a pulmonary-to-systemic flow ratio of less than 1.5:1, should not disqualify an individual for certification, whether this shunt is at the atrial or ventricular level. Such individuals will have normal electrocardiograms, normal heart size on chest radiograph, and no evidence of increased pulmonary artery pressure. Risk of a sudden incapacitating event due to the presence of such a heart defect is almost nonexistent.[7]

In general, the presence of prosthetic devices, such as right ventricle-to-pulmonary artery conduits, or artificial valves, are considered disqualifying because of the substantial risk of complications.[8] Prosthetic materi. per se is not disqualifying. For example, many persons with coarctation of the aorta undergo repair with Dacron patch grafts. Surgical closure of a ventricular septal defect usually includes a material patch. In both instances, the results of surgery, not the presence of prosthetic material, should determine eligibility for flying status.

Criteria for evaluating applicants for all classes of airman with the following common congenital cardiovascular anomalies should serve as guidelines for assessing all forms of congenital heart disease. For postoperative applicants, a minimal waiting period of one year following surgery is recommended to permit clinical and hemodynamic evaluation of results.

Minimal Aortic Valvular Stenosis

Personnel with a nonstenotic to minimally stenotic aortic valve, including a bicuspid aortic valve, may be qualified for Class I, II, or III if they are asymptomatic, have a systolic murmur of grade 3/6 or less, and have no left ventricular hypertrophy on physical examination, on ECG or on chest radiograph. Two-dimensional and Doppler echocardiography should show no increase in left ventricular dimensions and no increase in left ventricular posterior wall or ventricular septal thickness. A graded submaximal treadmill exercise test should be normal (to 90% of maximal heart rate, or symptom limited, reached with a normal heart rate response and normal exercise tolerance for the individual's age). If these studies are equivocal, cardiac catheterization should be performed. The pressure difference between the left ventricle and aorta should be less than 20 mm Hg under conditions of normal cardiac output, and left ventricular end-diastolic pressure should be less than 12 mm Hg. Persons who meet these criteria have no increased risk of sudden death or incapacitation.[7] However, progression with age is common and it has been estimated that 20% of individuals with minimal aortic stenosis will develop calcification by age 45 years.[9] Therefore, annual cardiovascular examinations by a specialist would be necessary for renewal of the certificate.

Mild to Moderate Aortic Valvular Stenosis

This category is defined by the presence of a pressure difference across the aortic valve not exceeding 40 mm Hg under conditions of normal cardiac output. Although the risk of sudden death or incapacity is not great, a small percentage of such persons do die suddenly, without previous symptoms.[10] Class I and II certificates should ordinarily not be issued for this category, although special issuance may be considered especially for pilots who perform their duties in situations in which there is crew redundancy.

Personnel may qualify for Class I, II, and III certification if they meet the following criteria: 1) auscultation should demonstrate a typical systolic murmur and an aortic ejection click; 2) the resting ECG should show neither left ventricular hypertrophy nor ST segment or T wave changes; 3) the chest radiograph should demonstrate a normal heart size, but may show poststenotic dilatation of the ascending aorta as well as mild left ventricular contour to the heart shape; 4) echocardiography should show normal left ventricular function; 5) the near maximal graded treadmill exercise test should be normal, demonstrating neither significant cardiac rhythm disturbances nor ST segment changes, and the predicted heart rate response and level of exercise should be within normal limits for the individual's age; 6) cardiac catheterization should demonstrate a pressure difference of 40 mm Hg or less across the aortic valve. Doppler echocardiography may be acceptable to quantify the pressure difference provided the examination is done by a physician who is expert in echo-Doppler diagnosis. If inconsistencies exist between the Doppler findings and other clinical data, catheterization should be done. An annual cardiovascular examination by a specialist would be necessary for renewal of the certificate. If clinical change is noted, repeat cardiac catheterization or echo-Doppler study would be indicated.

Moderate to Severe Aortic Valvular Stenosis

Moderate to severe aortic stenosis is defined as a pressure difference across the aortic valve exceeding 40 mm Hg with normal cardiac output. Persons with this disorder would not qualify for certification because of the recognized risk of sudden death or incapacity, and because of the recognized risk of progression of the lesion.[10]

Postoperative Aortic Valve Stenosis

Although the results of aortic valvotomy are good, with an operative mortality of approximately 2%, long-term prognosis remains guarded. Postoperative sequelae, such as

recurrent obstruction, aortic regurgitation, endocarditis, and late sudden death may affect as many as 50% of such individuals.[11] Therefore, individuals who have been operated upon for aortic valve stenosis ordinarily would not qualify for Class I or II certificates, although they may qualify for Class I, II and III certificates one year after aortic valvotomy if they meet the criteria for a minimally stenotic valve as defined above. In addition, they must not have more than trivial aortic regurgitation.

Subvalvular Aortic Stenosis

This is defined as discrete, membranous subvalvular aortic stenosis, without associated valvular abnormalities or secondary hypertrophic cardiomyopathy. Such individuals may receive Class I, II, and III certificates if the obstruction is trivial as defined by lack of symptoms, a normal chest radiograph and ECG, no more than trivial aortic regurgitation, and a pressure difference across the left ventricular outflow tract of less than 20 mm Hg with a normal cardiac output. Following surgical correction, individuals ordinarily would not qualify for Class I or II certificates, again because of significant late complications.[11] Personnel may qualify for Class I, II and III certificates if one year after surgery they are asymptomatic, have a normal chest radiograph and ECG, normal exercise ECG, no more than trivial aortic regurgitation, and a pressure difference across the left ventricular outflow tract of less than 10 mm Hg, with a normal cardiac output.

Discrete Supravalvular Aortic Stenosis

Few data exist for this defect because it is rare. The decision for certification should be in keeping with recommendations for valvular and subvalvular aortic stenosis. Persons with uncorrected supravalvular aortic stenosis would not qualify. Following surgical correction individuals may qualify for Class I, II and III certificates provided they meet the criteria described for persons following correction of subvalvular aortic

stenosis.

Atrial Septal Defect

Persons with either an ostium secundum or sinus venosus atrial septal defect may be qualified for Class I, II or III certification if they are asymptomatic and have cardiac catheterization findings of normal pulmonary artery pressure, a pulmonary-to-systemic flow ratio of less than 1.5:1, and no right-to-left shunt. Such individuals have no increased risk of sudden death or incapacitation.[7] Persons who have had surgical correction of one of these two types of atrial septal defects may be qualified for Class I, II, or III certification if they are asymptomatic, have normal physical findings, no cardiac rhythm disturbance other than respiratory sinus dysrhythmia, or occasional atrial or ventricular ectopic beats, minimal residual chest radiograph or ECG findings, and no evidence of residual defect by echocardiography. Such individuals have an excellent prognosis with little evidence to suggest subsequent cardiac deterioration.[12] However, there are some data indicating that individuals who have had surgical closure after age 11 years have persistent abnormalities of right ventricular function. The significance of these findings is at present unknown.[12]

Persons with an ostium primum atrial septal defect may be qualified for Class I, II, or III certification if they are asymptomatic and have cardiac catheterization findings of normal pulmonary artery pressure, pulmonary-to-systemic flow ratio of less than 1.5:1, and no more than trivial mitral regurgitation. Some caution should be exercised with this group, however, as the degree of mitral regurgitation can increase with age, and conduction system disorders may occur.[7] Annual cardiovascular examinations by a specialist would be necessary for renewal of the certificate. Following surgical correction, such individuals may be qualified for Class I, II, or III certification if they are asymptomatic, have no cardiac rhythm disturbance, other than respiratory sinus dysrhythmia or occasional atrial or ventricular ectopic beats, minimal residual

radiographic or ECG findings, such as the expected left anterior hemiblock pattern, and have post-operative cardiac catheterization demonstration of normal pulmonary artery pressure, no significant residual left-to-right shunt (pulmonary-to-systemic flow ratio less than 1.2:1), and no more than trivial mitral regurgitation by angiography or quantitative noninvasive evaluation.[12] Individuals with a greater degree of mitral regurgitation following surgical correction can qualify for Class III certification but not Class I or II, if they meet the criteria just described. For all such persons, annual cardiovascular examinations by a specialist would be necessary for renewal of the certificate.

Coarctation of the Aorta

Persons with uncorrected coarctation of the aorta have a significantly shortened life expectancy, with an average age at death of 35 years.[13] Because of the presence of systemic hypertension, these individuals are at risk to develop its complications, including intracranial hemorrhage, aortic rupture, and congestive heart failure. They may qualify for Class I, II and III certification if they are asymptomatic, have a normal ECG, have normal blood pressure at rest without medication, and have less than a 10 mm Hg resting systolic pressure difference between the arms and legs. They should also have normal treadmill exercise tests, including a normal blood pressure response. Persons with a bicuspid aortic valve must meet the criteria described for this lesion as well. Because each of these lesions can be progressive, an annual cardiovascular examination by specialist is necessary for renewal of the certificate.

Individuals whose coarctation has been corrected surgically may qualify for Class I, II or III certification if they are asymptomatic, have normal blood pressure and a less than 10 mm Hg systolic pressure difference across the coarctation repair, a normal heart size on chest radiograph, normal resting and exercise ECGs, and a normal blood pressure response to exercise. Persons with residual systemic hypertension would not qualify

because of the significant risk of complications, including sudden incapacitation.[13] If an abnormal aortic valve is present, criteria must be met for this lesion also. Some data exists that suggest that surgical repair of coarctation of the aorta after the age of 12 years is associated with a subsequent risk of sudden death or cerebral vascular accident; these individuals should be evaluated carefully and certified cautiously.

Patient Ductus Arteriosus

Persons with an uncorrected patent ductus arteriosus can qualify for Class I, II or III certification if they are asymptomatic, have a typical continuous murmur at the left base, a normal chest radiograph and ECG, and normal left ventricular size and function on echocardiogram. These individuals have no increased risk of sudden incapacitation.[7]

Persons whose ductus has been closed by surgery may qualify for Class I, II or III certification if the results of the physical examination, ECG and chest radiograph are normal.[12]

Minimal Pulmonic Valvular Stenosis

These individuals may qualify for Class I, II and III certification if the obstruction is trivial, as defined by the absence of symptoms, the presence of a grade 3/6 or less systolic ejection murmur at the left base, a systolic ejection click that varies with respiration, minimal widening of the second sound with normal intensity of the pulmonic component, a normal ECG, a normal heart size on chest radiograph with prominence of the pulmonic trunk, and echocardiographic criteria consistent with less than 25 mm Hg pressure difference across the pulmonic valve. If clinical assessment of a Class I or II applicant is unclear, cardiac catheterization is necessary to document a pressure difference of less than 25 mm Hg across the pulmonic valve in the presence of normal cardiac output. Minimal pulmonic stenosis does not progress, and such individuals are not at risk of sudden incapacitation.[14]

Mild Pulmonic Valvular Stenosis

Class III certification may be considered for individuals with mild pulmonic valve stenosis, as defined by physical examination findings of minimal pulmonic valve stenosis just described, in addition to normal arterial oxygen tension, mild right ventricular hypertrophy on ECG and echocardiogram, and cardiac catheterization findings that demonstrate a pressure difference of less than 45 mm Hg across the pulmonic valve in the presence of normal cardiac output. Class I and II certification may be considered especially if the pilot performs duties in situations in which there is crew redundancy.

Moderate to Severe Pulmonic Valvular Stenosis

Moderate to severe pulmonic valve stenosis is defined as a pressure difference across the pulmonic valve exceeding 45 mm Hg, with normal cardiac output. Individuals with this disorder would not qualify for certification, because of the likely presence of right ventricular dysfunction, and the likelihood of progression of the obstruction.

Postoperative Pulmonic Valvular Stenosis

Persons who have had surgery or nonsurgical valvotomy may qualify for Class I, II, or III certification if they are asymptomatic, have a chest radiograph that shows only a prominence of the pulmonic trunk, an ECG that shows no more than mild right ventricular hypertrophy, an echocardiogram that shows no evidence of right ventricular dysfunction and cardiac catheterization that demonstrates a pressure difference of less than 25 mm Hg across the pulmonic valve in the presence of normal cardiac output. The presence of mild pulmonic valve regurgitation should not disqualify.[12] Following valvotomy, individuals may qualify for Class III certification if they meet these criteria but have a pressure difference across the pulmonic valve of greater than 25 mm Hg but less than 45 mm Hg in the presence of normal cardiac output. Such individuals may be

considered for special issuance of Class I and II certificates, especially if they are permitted to perform the duties of an airman in a situation in which there is crew redundancy.

Discrete Right Ventricular Infundibular Stenosis

Long-term follow-up data for this defect are not clear, but, in general, the recommendations for valvular pulmonic stenosis should be followed. These persons may qualify for Class I, II or III certification if the obstruction is mild, as defined by the absence of symptoms, the presence of only minimal right ventricular hypertrophy on ECG, and a pressure difference of less than 25 mm Hg across the outflow tract in the presence of normal cardiac output. A small ventricular septal defect may be present with a pulmonic-to-systemic flow ratio of less than 1.5:1 as long as the other criteria are met.

Individuals who have had surgical correction of discrete right ventricular outflow tract obstruction may qualify for Class I, II or III certification if they are asymptomatic, have a normal heart size on chest radiograph, no more than minimal right ventricular hypertrophy and/or right ventricular conduction delay on ECG, a normal exercise ECG, and less than 15 mm Hg pressure difference across the right ventricular outflow tract in the presence of normal cardiac output. A residual ventricular septal defect is allowable provided the pulmonic-to-systemic flow ratio is less than 1.2:1. Individuals exceeding these criteria may qualify for Class III certification if there is no more than a 25 mm Hg pressure difference across the right ventricular outflow tract in the presence of normal cardiac output, and if a ventricular septal defect is present, the pulmonic-to-systemic ratio must not exceed 1.5:1. Pilots may be given Class I or II certificates, especially if they perform their duties as airmen in the situation in which there is crew redundancy.

Supravalvular Pulmonic Stenosis, including Coarctation of the Pulmonary Arteries

Persons may qualify for Class I, II or III certification if the obstruction is trivial, as defined above for trivial pulmonic valve stenosis. Class III certification may be given if the obstruction is mild as defined above for mild pulmonic valve stenosis, and Class I and II certification may be considered, especially if the airmen perform their duties in situations in which there is cockpit redundancy. Following correction, certification may be given in accordance with criteria described for surgical correction of pulmonic valve stenosis.

Pulmonary Hypertension

Pulmonary hypertension from any cause is disqualifying for all classes. Persons with primary pulmonary hypertension are continually at risk of sudden death. Exposure to low environmental oxygen tension, as at altitude, increases this risk. Similarly, individuals with secondary pulmonary hypertension, such as those with Eisenmenger's syndrome, are also at risk of incapacitation and sudden death.[6] Exposure to altitude is also deleterious for this group.

Tetralogy of Fallot

Very few individuals with tetralogy of Fallot survive beyond the second decade of life without surgical intervention.[15] Persons with uncorrected tetralogy of Fallot do not qualify. Following surgical correction, few individuals will be eligible for certification because of the significant incidence of postoperative complications.[15] However, an individual may qualify for Class I, II or III certification if the following criteria are met: 1) the surgical repair must be done without the use of a prosthetic device, homograft, or transannular patch; 2) transient third degree heartblock must not have occurred in the postoperative period; 3) there must be a normal heart size and normal pulmonary vascularity on chest radiograph, as well as absence of ECG evidence of bifascicular

block, atrioventricular conduction delay, or dysrhythmia at rest or with exercise.

Continuous ambulatory 24 hour ECG monitoring is required prior to certification and annually thereafter; treadmill exercise testing is also required annually. Echocardiographic documentation of normal right and left ventricular dimensions and function is required prior to certification and annually thereafter. It is also necessary to have documentation by cardiac catheterization of normal right ventricular function and dimensions, absence of more than trivial tricuspid valve regurgitation, right ventricular systolic pressure less than 40 mm Hg in the presence of normal cardiac output, no right to left shunt, and no residual left to right shunt exceeding a pulmonary to systemic flow ratio of 1.2:1.

Individuals who have had surgical repair may qualify for Class III certificates if they meet the criteria just described but exceed cardiac catheterization requirements. However, the catheterization must document no more than minimal right ventricular dilatation, no more than mild tricuspid regurgitation, right ventricular systolic pressure less than 40 mm Hg in the presence of normal cardiac output, no right-to-left shunt, and if a residual left-to-right shunt is present, it must not exceed a pulmonary to systemic flow ratio of 1.5:1. Because of the continuing risk of postoperative complications, annual cardiovascular examinations by a specialist are required for renewal of certification.

Simple Transposition of the Great Arteries, without Associated Ventricular Septal Defect or Pulmonic Stenosis

Because of the presence of substantial cyanosis, individuals with uncorrected transposition of the great arteries do not qualify. Persons who have undergone surgical correction of transposition of the great arteries do not qualify for Class I or II certification because of the risk of postoperative complications.[5]

These individuals may be considered for Class III certification if they are asymptomatic and are receiving no cardiac medication, have no more than mild

cardiomegaly on chest radiograph, demonstrate sinus rhythm on routine ECG, an absence of significant cardiac dysrhythmia or significant conduction disturbance on 24-hour ambulatory ECG monitoring and an absence of significant dysrhythmia on exercise testing. These individuals must undergo postoperative cardiac catheterization with intracardiac electrophysiological studies that demonstrate no more than trivial tricuspid regurgitation, no right-to-left or left-to-right shunting, normal right ventricular dimensions with right ventricular end-diastolic pressure less than 12 mm Hg, left ventricular systolic pressure less than 40 mm Hg, normal sinus node function, and normal atrioventricular conduction.

Since the long term effects of the right ventricle's functioning as the systemic ventricle are not known, it is recommended that right ventricular performance be assessed periodically by appropriate techniques.

Small Ventricular Septal Defect

Persons with a small ventricular septal defect may qualify for Class I, II or III certification.[7] Such individuals must be asymptomatic, have typical ausculatory findings of a ventricular septal defect with normal splitting and intensity of the second sound, and no diastolic murmur of aortic regurgitation or increased mitral valve flow. They must have a normal heart size on chest radiograph, a normal ECG and a normal left ventricular size and function by two-dimensional echocardiography. Cardiac catheterization is not required, but if it is performed, it must document a normal pulmonary artery pressure, with a pulmonary-to-systemic flow ratio of less than 1.5:1 and no right-to-left shunt. Persons with previous cardiac catheterization data that exceed these criteria, who undergo a spontaneous closure or clinically significant reduction in size of a ventricular septal defect, may qualify if they currently meet all of the other criteria described.

Moderate Ventricular Septal Defect

Individuals with a moderate size ventricular septal defect, that is, who exceed the criteria for a small ventricular septal defect, will not qualify for Class I or II certification. Although the risks of sudden incapacitation are not great, such events do occur.[16] They may qualify for Class III certification if they meet the following criteria: 1) they must be asymptomatic and be taking no cardiac medication; 2) they must have no more than mild cardiac enlargement on chest radiograph, and no more than mild left atrial enlargement and/or left ventricular hypertrophy on ECG, with no right ventricular hypertrophy; 3) there should be cardiac catheterization documentation of normal pulmonary arteriolar resistance, pulmonary artery systolic pressure less than 40 mm Hg and left to-right shunt with a pulmonary-to-systemic flow ratio of less than 2:1.

Postoperative Surgical Closure of a Ventricular Septal Defect

Persons who have had surgical closure of a ventricular septal defect may qualify for Class I, II or III certification if they meet the following criteria: 1) they must be asymptomatic and taking no cardiac medication; 2) there must be no residual auscultatory evidence of a ventricular septal defect, and there must be normal splitting and intensity of the second heart sound; 3) there must be a normal heart size on chest radiograph, and a normal ECG at rest and with exercise. If a mild right ventricular conduction delay (QRS complex duration less than 0.12 seconds) exists on ECG, an individual may still qualify if no dysrhythmia occurs on 24-hour ambulatory ECG monitoring; this monitoring should be repeated annually. If a more significant right ventricular conduction delay is present, the individual may qualify if invasive His bundle studies show no prolongation of the H-V interval. Postoperative catheterization is not required provided preoperative catheterization shows normal pulmonary arteriolar resistance.[16]

Persons who exceed the postoperative criteria just described may qualify for Class III certification, but not Class I or II certification, if they meet criteria described for individuals with a moderate ventricular septal defect and if they have no significant dysrhythmia or conduction disturbance on exercise electrocardiogram.[16]

References

1. Macartney FJ, Taylor JFN, Graham GR, et al: The fate of survivors of cardiac surgery in infancy. Circulation, 1980;62:80-91.
2. Maron BJ, Humphries JON, Rowe RD, et al: Prognosis of surgically corrected coarctation of the aorta. A 20-year post-operative appraisal. Circulation 1973;47:119-126.
3. Deanfield JE, McKenna WJ, England D, et al: Factors affecting ventricular arrhythmia after correction of tetralogy of Fallot. Circulation 1981;64:IV-227.
4. Esscher E, Michaelsson M: Long-term results following closure of isolated ostium secundum atrial septal defect in children and adults. Eur J Cardiol 1977;6:109-116.
5. Engle MA, Perloff JK (eds): Congenital Heart Disease After Surgery: Benefits, Residua, Sequelae. New York, Yorke Medical Books, 1983.
6. Graham TP: The Eisenmenger reaction and its management, in Roberts WC (ed): Congenital Heart Disease in Adults. Philadelphia, FA Davis, 1979, pp 531-542.
7. Macartney FJ: Flying and congenital heart disease. Eur Heart J 1984;5:147-154.
8. McGoon DC: Long-term effects of prosthetic materials, in Roberts WC (ed): Congenital Heart Disease in Adults. Philadelphia, FA Davis, 1979, pp 177-201.
9. Keith JD: Bicuspid aortic valve, in Keith JD, Rowe RD, Vlad P (eds): Heart Disease in Infancy and Childhood. New York, Macmillan, 1978, pp 729-731.
10. Friedman WF, Novak V, Johnson AD: Congenital aortic stenosis in adults, in Roberts WC (ed): Congenital Heart Disease in Adults. New York, FA Davis, 1979, pp 235-251.

11. Jones M, Barnhart GR, Morrow AG: Late results after operations for left ventricular outflow obstruction, in Engle MA, Perloff JK (eds): Congenital Heart Disease After Surgery: Benefits, Residua, Sequelae. New York, Yorke Medical Books, 1983, pp 105-132.

12. McNamara DG, Latson LA: Long-term follow-up of patients with malformations for which definitive surgical repair has been available for 25 years or more, in Engle MA, Perloff JK (eds): Congenital Heart Disease After Surgery: Benefits, Residua, Sequelae. New York, Yorke Medical Books, 1983, pp 77-99.

13. Maron BJ: Coarctation of the aorta in the adult, in Roberts WC (ed): Congenital Heart Disease in Adults. Philadelphia, FA Davis, 1979, 311-319.

14. Nugent EW, Freedmon RM, Nora JJ, et al: Clinical course in pulmonary stenosis. Circulation 1979;56:38-47.

15. Garson A, McNamara DG, Cooley DA: Tetralogy of Fallot in adults, in Roberts WC (ed): Congenital Heart Disease in Adults. Phildelphia, FA Davis, 1979, pp 341-364.

16. Engle MA, Kline SA: Ventricular septal defect in the adult, in Roberts WC (ed): Congential Heart Disease in Adults. Philadelphia, FA Davis, 1979, pp 279-309.

Ischemic Heart Disease

Nearly ten years have elapsed since participants of the Eighth Bethesda Conference of the American College of Cardiology addressed the issues of cardiovascular problems associated with aviation safety.[1] In this document we re-examine pilot certification practices in light of more current knowledge about coronary artery disease and the results of medical and surgical treatment. These issues also have been considered extensively by a recent workshop in the United Kingdom on Aviation Cardiology.[2]

Coronary artery disease in individuals less than age 65 years remains predominantly a disease of men, and its incidence is strongly determined by age. From age 40 to 60 years, men experience a risk of myocardial infarction from seven to three times greater than that experienced by women of similar ages.[3] Additionally, among men the risk of death from ischemic heart disease increases exponentially with age, and is 8, 36 and nearly 100 times greater at ages 40, 50 and 60 years, respectively, than it is at the age of 30 years.[3] There is a well-known latent period, during which persons with coronary heart disease are asymptomatic, before the first onset of clinical manifestations. The frequency with which the onset of coronary artery disease occurs with incapacitating rapidity, or as sudden death, is less readily identified, although more than 50% of those who die from an acute myocardial infarction will do so in the first one to two hours after the onset of the attack.[4] Similarly, in male population groups who were studied prospectively, nearly one-half of the deaths caused by coronary heart disease occurred suddenly within one hour of the symptoms.[5] The risk of sudden death in these populations was positively correlated with high blood pressure, the electrocardiographic patterns of left ventricular enlargement, obesity and heavy cigarette usage.[5] It is only natural periodically to re-examine the question whether better methods are available, or need be utilized, to screen pilot populations in an

attempt to enhance the detection of coronary artery disease in the latent, asymptomatic phase.

The diagnosis of coronary artery disease usually is not difficult when manifested by typical angina pectoris or by classic myocardial infarction. Even the presence of prior silent or clinically unrecognized myocardial infarction is usually evident on the subsequent resting electrocardiogram. However, during the latent asymptomatic phase of coronary artery disease or when the symptoms of coronary artery disease are atypical, recognition continues to be the challenge.

It is well known and generally accepted that the presence of certain identifiable risk factors individually and collectively increase the probability of coronary artery occlusive disease,[6] these being age; male sex; a family history of coronary artery disease, especially of early or premature coronary disease; elevated blood pressure; cigarette smoking; diabetes mellitus; and an elevated level of serum cholesterol. The awareness of the role of these risk factors has played an important part in the health education of the general population in this country and has probably had an influence in the decreasing mortality from coronary artery disease.[7-10] This message should not be overlooked in the aviation community.

Detection of Latent Coronary Artery Disease

Detection of individuals with asymptomatic coronary artery disease is not only a national challenge because of the devastating consequences of myocardial infarction and sudden death, but is an especially challenging issue in the air transportation industry because of the potential consequences of incapacitation of an air crew member due to a a sudden coronary event. The occurrence of air transportation incidents resulting from sudden incapacitation of an air crew member because of a cardiovascular event is infrequent and has all but been eliminated completely in multi-crew operations. Overstringent standards of fitness could increase public safety risks due to the

replacement of senior, more experienced pilots by younger pilots who may be more fit medically but less experienced.[11,12]

The Resting 12-Lead Electrocardiogram (ECG)

The resting ECG consistently demonstrates changes of prior myocardial infarction when one has occurred. Additionally, the patterns of left ventricular enlargement on the resting ECG[5] and even of nonspecific ST segment and T wave abnormalities are associated with an increased risk of coronary artery disease.[6,13,14] In the United States Army cardiovascular screening program of 42,752 males over the age of 40 years, 4,747 (11%) failed the primary screening because of an anormal resting electrocardiogram.[6] Presently, FAA regulations require an ECG only on examinations for Class I certificates at 35 years of age and annually beginning at 40 years of age. Broader use of the resting ECG as a primary screening mechanism is strongly recommended for applicants of all classes upon entry. Class II pilots should have an ECG performed at age 35 and 40 years, and every two years thereafter. Class III pilots should have an ECG performed at age 40 years and every 5 years thereafter.

Exercise Electrocardiography

The use of routine exercise electrocardiographic testing after some predetermined age, as a screening technique to detect latent coronary artery disease in asymptomatic persons, received considerable deliberation by this committee, and a wide range of opinions was considered. Unfortunately, the value of any screening procedure is limited by Bayes' theorem of conditional probability.[14-16] That is, the random use of exercise electrocardiographic testing for the detection of asymptomatic coronary artery disease usually gives rise to a large number of false positive results when the prevalence of disease in the study population is relatively low.[6,17-19] The committee recommends a thorough cardiovascular evaluation, including exercise electrocardiographic testing, of

those individuals with symptoms that are either typical or atypical for coronary artery disease, and of those individuals who are found to have hypertension or resting electrocardiographic abnormalities. Such testing would currently include one of the standardized graded exercise treadmill test protocols that has displaced prior techniques of testing such as the Masters Two-step test. The low prevalence of coronary artery disease in younger pilots and the resultant poor predictive value of routine exercise testing would make the use of this test in the group unwise.

Since the risk of coronary artery disease increases rapidly in men above the age of 50 years, most of the members of this committee, as well as some participants in the United Kingdom Workshop in Aviation Cardiology,[3,20,21] were of the opinion that risk factor assessment and exercise electrocardiographic testing are appropriate screening measures for male pilots age 50 years and older, who hold Class I and II certificates and who are performing in a single-crew commercial operation. We recommend that the serum cholesterol be determined at age 50 years and that exercise electrocardiographic testing be done on those persons whose serum cholesterol is 300 mg/dl or greater. An abnormal response to electrocardiographic exercise testing would require radionuclide stress testing and, if this is abnormal, should lead to performance of coronary angiography.[14,15] Extension of risk factor assessment and exercise electrocardiographic testing to pilots holding Class I and II certificates operating in multi-crew operations does not presently appear justified. There exists, however, a need for research into methods of collecting risk factor information in pilots and the predictive value of such information. Further research into the frequency of coronary events among pilots, while on and off duty, and the prediction of these events by risk factor assessment, is strongly needed. Such investigations would also emphasize to the pilot population the importance of correctable risk factors, such as cigarette smoking.[8]

Radionuclide Stress Testing

Radionuclide ventriculography, a noninvasive procedure, provides excellent information about regional and global ventricular function at rest and following exercise. Thallium 201 myocardial perfusion imaging provides information about both prior myocardial infarction and stress induced myocardial perfusion defects.[14,22] In the cardiovascular evaluation of the asymptomatic subject, thallium 201 exercise imaging is indicated if the electrocardiographic treadmill response is abnormal. An ischemic response to stress would strongly favor the subsequent use of coronary angiography.[14] Both tests have also been used to risk-stratify patients following myocardial infarction[23] and to evaluate patients after coronary artery bypass surgery.[24,25] Neither test has a role in the routine screening of the asymptomatic subject;[14] however, both tests have enjoyed a role in the secondary screening of coronary artery disease with some evidence favoring a higher sensitivity and specificity with thallium 201 exercise imaging.[14,15]

Individuals with Coronary Artery Disease

The presence of known coronary artery disease is the single most powerful predictor of subsequent coronary events. Therefore, the reissuance of a certificate that has once been denied because of coronary artery disease dictates that the evaluation for the potential of recurrent events be stringent, that reissuance of Class I and II certificates be considered preferentially for pilots in multi-crew operations, and, since coronary disease is not static, that a mechanism for effective, periodic re-evaluation be initiated and followed. Fortunately, our knowledge about the course of coronary artery disease, both after infarction and after coronary artery bypass surgery continues to grow, and presently permits sound risk stratification, based not only upon coronary and ventricular anatomy, but also upon arrhythmia monitoring and functional testing.[23-31] Such evaluation permits the selection of individuals with known coronary artery disease, who are at minimal risk from their disease, and allows their return to flying status.

Asymptomatic and otherwise healthy individuals with recognized coronary artery disease who desire reissuance generally fall into one of three categories: 1) individuals who have successfully recovered from an acute myocardial infarction, or who have been identified as having had a clinically "silent" infarction, or who have angiographic evidence of coronary artery disease discovered in some unrelated fashion; 2) individuals who have successfully recovered from coronary artery bypass surgery; and 3) individuals who have been successfully treated by coronary balloon angioplasty. Criteria for evaluation and return to flying status as well as for periodic evaluation follow.

- Individuals who have recovered from previous myocardial infarction

  For Classes I, II and III, individuals must be asymptomatic one year after the event. At a minimum, results of a thorough cardiovascular evaluation should demonstrate a normal cardiovascular system at rest that responds normally to an exercise test to at least Stage III[18] and a heart rate of 90% of the predicted maximum heart rate for age[32,33] without evidence of stress-induced ischemia. Ambulatory electrocardiographic monitoring[27] should reveal no significant rhythm disturbance. Exercise thallium scintigraphy and/or exercise radionuclide ventriculography[23,26] should demonstrate no significant evidence of a stress-induced myocardial perfusion defect and/or significant stress-induced regional or global systolic ventricular dysfunction.

  For Classes I and II, coronary angiography should demonstrate no evidence of significant coronary occlusive disease, defined as greater than or equal to 50% narrowing of lumen diameter, other than that producing the index infarction; and left ventriculography should demonstrate near-normal global systolic ventricular function with no more than a single area of regional hypokinesia or akinesia. At 6 month intervals follow-up cardiovascular

assessment and treadmill electrocardiographic stress testing should remain normal and at 12 month intervals electrocardiographic monitoring should show no significant rhythm disturbance, and exercise thallium scintigraphy and/or exercise gated blood pool radionuclide ventriculography should demonstrate no evidence of a stress-induced myocardial perfusion defect or of stress-induced regional or global ventricular dysfunction. Repeat coronary angiography should be performed after five years or earlier if there are coexisting risk factors known to accelerate atherogenesis.

For Class III follow-up cardiovascular assessment should include normal stress electrocardiographic testing at yearly intervals.

- Individuals who have undergone coronary artery bypass (CAB)

  For Classes I, II and III individuals must be asymptomatic 6 months after CAB.[34,35] At a minimum, results of a thorough cardiovascular evaluation should demonstrate a normal cardiovascular system at rest that responds normally to an exercise test to at least Stage III[18] and a heart rate of 90% of the predicted maximum heart rate for age[32,33] without evidence of stress-induced ischemia. Ambulatory electrocardiographic monitoring should reveal no significant rhythm disturbance. Exercise thallium scintigraphy and/or exercise radionuclide ventriculography[24,25] should demonstrate no significant evidence of a stress-induced myocardial perfusion defect and/or a significant stress-induced regional or global systolic ventricular dysfunction.

  For Classes I and II coronary angiography should demonstrate wide patency of all bypasses and no significant unbypassed disease. The number of bypasses is a less important factor than adequate graft patency. Left

ventriculography should demonstrate near normal global systolic ventricular function with no more than a single area of regional hypokinesia or akinesia. Follow-up examinations should be identical to those for previous myocardial infarction with an additional requirement of repeat coronary artery graft angiography and left ventriculography five, 8, and 10 years after CAB. A somewhat less stringent schedule for repeat angiography might be permitted if the internal mammary artery(ies) was used as the bypass conduit(s).[36] Occurrence of significant graft occlusion or of native vessel disease distal to the graft or of significant progress of native vessel disease in unbypassed vessels would preclude special issuance. These stringent follow-up requirements are necessary because of the palliative nature of CAB and the likelihood of progressive atherosclerotic changes in either the graft or the native vessels, especially in the second half decade after surgery.[37,38]

Class III requirements for follow-up cardiovascular assessment are the same as for myocardial infarction.

- Individuals who have been treated successfully by intracoronary balloon angioplasty

  For Classes I and II requirements are as outlined above for those following CAB.[39] Coronary angiography should demonstrate wide patency of the dilated vessel and no other major occlusive disease, defined as greater than or equal to 50% luminal diameter narrowing. Left ventriculographic criteria should be identical to those for myocardial infarction and CAB. Follow-up should be identical to the requirements for CAB. The long-term status of individuals who have undergone balloon angioplasty is still unknown.

Accordingly, follow-up angioplasty at the fifth year presently appears advisable.[40]

Class III requirements are the same as those following myocardial infarction, although special issuance may be considered at 6 months after surgery. Because of the newness of this procedure, coronary angiography should be performed at 6 months and the results should meet the criteria set fourth for Classes I and II.

References

1. Cardiovascular Problems Associated with Aviation Safety: Eighth Bethesda Conference of the American College of Cardiology. Am J Cardiol 1975;36:573-628.
2. The First United Kingdom Workshop in Aviation Cardiology. Eur Heart J 1984;5(suppl A):1-164.
3. Tunstall-Pedoe H: Risk of coronary heart attack in the normal population and how it might be modified in flyers. Eur Heart J 1984;5(suppl A):43-49.
4. Armstrong A, Duncan B, Oliver MF, et al: Natural history of acute heart attacks: A community study. Brit Heart J 1972;34:67-80.
5. Kannel WB, Doyle JT, McNamara PM, et al: Precursors of sudden cardiac death: Factors related to the incidence of sudden death. Circulation 1975;51:606-614.
6. Zoltick JM, McAllister HA, Bedynck JL: The United States Army Cardiovascular Screening Program. J Cardiac Rehab 1984;4:530-535.
7. Kannel WB, Thom TJ: Implications of the recent decline in cardiovascular mortality. Cardiovasc Med 1979;4:983-997.
8. Ebert RV, McNabb McE: Cessation of smoking in prevention and treatment of cardiac and pulmonary disease. Arch Intern Med 1984;144:1558-59.

9. Chobanian AV: The influence of hypertension and other hemodynamic factors in atherogenesis. Prog Cardiovasc Dis 1983;26:177-196.

10. Brown WV, Goldberg JJ, Ginsberg HN: Treatment of common lipoprotein disorders. Prog Cardiovasc Dis 1984;27:1-20.

11. FAA Reponse to the Comptroller General of the United States. Report No CED-76-154, November 3, 1976.

12. Bennett G: Aviation accident risk and aircrew licensing. Eur Heart J 1984;5(suppl A):9-13.

13. Hampton JR: The importance of minor abnormalities in the resting electrocardiogram. Eur Heart J 1984;5(suppl A):61-63.

14. Uhl GS, Froelicher V: Screening for asymptomatic coronary artery disease. J Am Coll Cardiol 1983;1:946-955.

15. Epstein SE: Implications of probability analysis on the strategy used for noninvasive detection of coronary artery disease (combined use of exercise electrocardiographic testing, radionuclide cineangiography and myocardial perfusion imaging). Am J Cardiol 1980;46:491-499.

16. Vecchio JT: Predictive value of a single diagnostic test in unselected populations. N Engl J Med 1966;274:1171-1173.

17. Borer JS, Brenski JF, Redwood DR: Limitations of the electrocardiographic response to exercise in predicting coronary artery disease. N Engl J Med 1975;293:367-371.

18. Bruce RA, DeRoven TA, Hossack HF: Value of maximal exercise tests in the risk assessment of primary coronary heart disease in healthy men: Five years experience of the Seattle Heart Watch Study. Am J Cardiol 1980;46:371-378.

19. Piepgrass SR, Uhl GS, Hickman JR: The limitations of the exercise stress test in the detection of coronary artery heart disease in apparently healthy men. Aviat Space Environ Med 1982;53:379-382.

20. Poole-Wilgon PA: Natural history of minor coronary events. Eur Heart J 1984;5(suppl A):69-72.

21. Chamberlain DA: Routine and exercise electrocardiography in aircrew: Techniques, interpretation, and recommendations. Eur Heart J 1984;5(suppl A):55-60.

22. Robinson P: Radionuclide imaging. Eur Heart J 1984;5(suppl A):65-68.

23. Hung J, Goris ML, Nash E, et al: Comparative value of maximum treadmill testing, exercise thallium myocardial perfusion scintigraphy and exercise radionuclide ventriculography for distinguishing high- and low-risk patients soon after acute myocardial infarction. Am J Cardiol 1984;53:1221-1227.

24. Gibson RS, Watson DD, Taylor GJ, et al: Prospective assessment of regional myocardial perfusion before and after coronary revascularization surgery by quantitative thallium 201 scintigraphy. J Am Coll Cardiol 1983;1:804-815.

25. Lindsay J, Nolan NG, Kotlyarov EV, et al: Radionuclide ventriculography following coronary bypass surgery: Correlation with arteriographic findings. Ann Thorac Surg 1982;33:238-243.

26. Gibson RS, Watson DD, Graddock GB: Prediction of cardiac events after uncomplicated myocardial infarction: A prospective study comparing predischarge thallum 201 scintigraphy and coronary angiography. Circulation 1983;68:321-336.

27. The multicenter postinfarction group: Risk stratification after myocardial infarction. N Engl J Med 1983;309:331-336.

28. DeFeyten PJ, van Eenige Mj, Dighton DH, et al: Prognostic value of exercise testing, coronary angiography and left ventriculography 6-8 weeks after myoardial infarction. Circulation 1982;66:527-536.

29. Balcon R: Prognostic significance of coronary angiography. Eur Heart J 1984;5:(suppl A):73-75.

30. Parker DJ: The airline pilot after coronary artery bypass grafting. Eur Heart J 1984;5(suppl A):77-79.

31. DePace NL, Iskandrain AS, Hakki AH: One-vessel coronary artery disease: Anatomic, functional, and prognostic considerations. Arch Intern Med 1984;144:1233-1238.

32. Ellestad MH, Allen W, Wan MCK: Maximum treadmill stress testing for cardiovascular evaluation. Circulation 1969;39:517.

33. Ellestad MH, Wan MCK: Predictive implications of stress testing; Follow-up of 2700 subjects after maximum treadmill stress testing. Circulation 1975;5:363-369.

34. Sheldon WC, Rincon G, Pichard AD, et al: Surgical treatment of coronary artery disease: Pure graft operations, with a study of 741 patients followed 3-7 years. Prog in Cardiovasc Dis 1975;18:237-53.

35. Sheldon WD: Factors influencing patency of coronary bypass grafts. Cleveland Clinic Quarterly 1978;45:109-111.

36. Grondin CM, Campeau L: Comparison of late changes in internal mammary artery and saphenous vein grafts in two consecutive series of patients 10 years after operation. Circulation 1984;70(suppl I):208-212.

37. Loop FD: Progress in surgical treatment of coronary arteriosclerosis. Chest 1983;84:611-624 and 740-755.

38. Hall RJ, Elayda MA, Gray A: Coronary artery bypass: Long-term follow-up of 22,284 consecutive patients. Circulation 1984;68(suppl II):208-212.

39. Libow M, Gruntzig AR, Greene L: Percutaneous transluminal coronary angioplasty. Curr Prob in Cardiol 1985;10:1-55.

40. Meier B, King SB III, Gruentzig AR: Repeart coronary angioplasty. J Am Coll Cardiol 1984;4:463-466.

Hypertension in Aviation

Since hypertension affects approximately 25% of the adult population in the United States,[1] this important risk factor for cardiovascular disease is a major consideration in the medical qualifications for airmen.[2]

As of January 1983, increased blood pressure was the cardiovascular condition with highest prevalence (6206 cases) among airmen seeking medical certification in all three classes. Therefore, it is not surprising that, among all cardiovascular conditions, hypertension caused the highest number (169) of denials for medical certification.[3]

As reported by the 1975 Bethesda Conference,[4] in the absence of clinical or laboratory evidence of cardiovascular disease, blood pressure may be the best predictor of subsequent cardiovascular events that might cause unexpected disability or death in a pilot or other flight crew members during the performance of duty. The risk of adverse cardiovascular events is directly related to the level of the blood pressure and is heightened by factors such as age, sex, race, family history, smoking, blood lipid levels, glucose intolerance, target organ damage and alcohol intake. The interactions of the major risk factors are well known through the results of the Framingham study.[5] The importance of initiating procedures leading to the detection and prevention of the controllable risk factors, such as smoking and lipid levels, is obvious and should be considered by regulatory agencies responsible for the health of civilian airmen and the safety of civilian aviation.

The diagnosis of hypertension in adults is confirmed when the average of two or more diastolic blood pressures measured in the sitting position on at least two separate visits or on separate measurements is equal to or greater than 95 mm Hg, and when the average of multiple systolic blood pressures on two or more separate visits is consistently greater than 150 mm Hg.[6,7] Systolic pressure should not exceed 160 mm Hg regardless of the level of diastolic blood pressure.[7] Casual or isolated blood pressure measurements

should not be used to establish a diagnosis of hypertension. Identical blood pressure criteria for all classes of certification are justified based on the clear increased risk for the development of cardiovascular disease in persons with hypertension.

Blood pressure should be measured with the subject seated comfortably with the arm bared at heart level and well supported. Blood pressure should be measured in both arms. The disappearance of sound (phase V) should be reported as the diastolic blood pressure. Sphygmomanometer standards and appropriate cuff sizes should be used according to criteria set forth by the American Heart Association.

Intermittent or occasional blood pressure measurements above acceptable levels are frequent, and persons with intermittently variable or labile blood pressure present a potential problem because of the well-recognized fact that these persons might develop sustained hypertension and thereby later be disqualified from medical certification. The purpose of confirming elevated blood pressure and of repeating measurements is to determine whether initial elevations remain high and require close observation, evaluation and/or therapy, or whether the blood pressure has returned to normal levels and thereafter requires only periodic monitoring.

Once the presence of a sustained and significant elevation in blood pressure has been documented, then an evaluation should attempt to answer at least the following three questions:[6]

1. Is target organ involvement present?
2. Are cardiovascular risk factors other than hypertension present?
3. Does the patient have a primary or secondary (possibly a reversible or surgically correctable) form of hypertension?

Prior to pursuing an expensive and extensive laboratory investigation to answer these questions, a careful history is mandatory. It is most important to review the use of all prescribed and "over the counter" medications for any effect they may have on

current blood pressure measurements or their possible interference with the detection of angina, transient ischemic attacks, dyspnea and edema, and with the subsequent effectiveness of antihypertensive drug regimens. The medical history should elicit a family history of hypertension and associated cardiovascular disease, renal disease or diabetes mellitus. The duration and levels of increased blood pressure, the response to or side effects of previous antihypertensive therapy, any weight changes, dietary habits, especially sodium and potassium intake, and use of alcohol, are important historical items. In addition, the presence or absence of other cardiovascular risk factors such as obesity, smoking, hyperlipidemia and carbohydrate intolerance are important.

The physical examination should include blood pressure measurements with the patient sitting. Blood pressure should be measured in both arms. There should also be a recording of height and weight; a careful funduscopic examination for the presence of arteriolar narrowing, arteriovenous compression, hemorrhages, exudates and papilledema; palpation and auscultation of the neck for the amplitude of carotid pulsations as well as for bruits and the presence of cervical venous distention; examination of the lungs for the presence of pulmonary congestion; assessment of the heart for incresed heart rate, size, arrhythmia, murmurs and gallop sounds; examination of the abdomen for bruits and palpable masses such as enlarged kidneys; careful examination of the extremities for diminished or absent peripheral arterial pulses and the presence of edema; and a brief neurological exam.

The extent of the initial and subsequent laboratory investigations will depend upon the level of blood pressure, the details of the medical history and the findings on physical examination. However, the tests will generally be grouped in categories to assist in determining the severity of any associated vascular disease and/or the possible cause(s) for hypertension. Laboratory tests to assess cardiovascular risk factors may provide appropriate and necessary baseline values for judging initial and future adverse biochemical effects of therapy. The laboratory investigation should likewise lead to an

assessment of the presence or absence of significant target organ involvement in the hypertensive process. The usual initial laboratory studies should include a hemogram; urinalysis; a biochemical profile, including at a minimum a fasting plasma glucose, creatinine, serum electrolytes and cholesterol; chest radiograph, ECG and exercise treadmill test. Annually thereafter the medical re-evaluation should include a history, physical examination, ECG and biochemical profile. At intervals of three to five years chest radiographs and exercise treadmill tests are recommended. Because evidence of target organ damage increases the risk of complications, the following findings might be considered disqualifying for medical certification regardless of the levels of blood pressure or its ability to be controlled by appropriate medication:[4]

1. Left ventricular hypertrophy, as manifested in the ECG by voltage changes and ST segment T wave abnormalities (voltage changes alone may not be disqualifying).
2. Radiographic evidence of cardiomegaly greater than 20% by the Underleider criteria or a cardiothoracic ratio of greater than 50%.
3. Azotemia as reflected by a serum creatinine greater than 2.5 mg/dl.
4. Hypertensive retinopathy of group 3 or 4 (Keith-Wagner-Barker-classification).

If the person is determined to have a form of surgically correctable hypertension, then following such treatment he or she should be re-evaluated for certification.

Such surgically correctable conditions include pheochromocytoma, primary aldosteronism (unless the surgery was a bilateral adrenalectomy), renovascular disease and renal parenchymal disease. Three months or more after the operation, if the blood pressure is normal and there is an improvement or clearing of the target organ damage, the applicant should be certified. If residual hypertension is present following these surgical procedures and the blood pressure can be controlled with acceptable drugs and

there is no target organ disease, then certification should be granted on the same basis as for persons with essential hypertension. Because of the likelihood of a recurrence of hypertension following transluminal balloon angioplasty of the renal artery for atherosclerotic renovascular hypertension, the blood pressure should be monitored every six months following this procedure, and more often for those persons who are receiving antihypertensive drug therapy as well. Blood pressure surveillance may need to be extended to three years.

Persons who have undergone either surgical treatment for Cushing's syndrome or sympathectomy for hypertension should not be certified even if they become normotensive as a result of the operation.

Antihypertensive Treatment and Medications

"The goal of treating patients with hypertension is to prevent the morbidity and mortality attributable to high blood pressure. This means the reduction of elevated blood pressure to the extent that excess cardiovascular risk is eliminated. Although the benefits of therapy have been demonstrated in numerous clinical trials, the decision to initiate therapy in any patient requires the physician to consider at least two factors: the severity of the blood pressure elevation and the presence of other complications or additional risk factors. The effectiveness of antihypertensive drugs in reducing elevated blood pressure is well established."[6] This statement from the 1984 Report of the Joint National Committee in Detection, Evaluation and Treatment of High Blood Pressure clearly outlines the generally accepted principles and concepts of antihypertensive therapy. The ultimate goal of antihypertensive therapy is to achieve and maintain, if feasible, a diastolic blood pressure at or lower than 90 mm Hg.[6,7] It is strongly recommended that effective treatment of hypertension should include nonpharmacologic modalities in conjunction with currently available antihypertensive drugs.

Considerable interest has developed in the nonpharmacologic treatments for hypertension.[8] Such factors as weight reduction, dietary sodium restriction combined with increased dietary potassium, reduction in alcohol intake, reduction in dietary saturated fats, cessation of cigarette smoking, appropriate exercise programs and other modifications of behavior should be incorporated into antihypertensive regimens when appropriate.

Presently the Federal Aviation Administration (FAA) considers diuretics and low doses of approved beta adrenergic blocking agents to be acceptable treatment of hypertension, if special cardiovascular evaluation reveals no target organ disease. Allowable doses, recommended periods of surveillance and appropriate laboratory monitoring are the subjects of continuing discussion and will be determined by levels of blood pressure, co-existing conditions and patient compliance. Considerable interest and information have developed regarding the importance of potassium depletion in the risk of actual or potential cardiac arrhythmias, especially ventricular ectopy, following the administration of thiazide drugs as a part of hypertensive therapy. The maintenance of a normal serum potassium concentration is mandatory in flight personnel treated with diuertics for hypertension. Supplemental oral potassium preparations should be given, if necessary, to accomplish this goal. Following the initiation of therapy with any diuretic, the serum potassium should be monitored in four weeks and thereafter at 6 month intervals, and these values should be reported to the FAA. Consideration should also be given toward the potential negative biochemical effect of thiazides on lipid levels, carbohydrate metabolism, magnesium and serum uric acid concentrations.

Beta adrenergic receptor blocking agents may cause sedation and also have other side effects. The hydrophilic beta blockers may cause fewer side effects, particularly as related to the central nervous system, since they do not readily penetrate the blood-brain barrier. There are probably theoretical reasons why beta blocking agents should be preferred over diuretics as so-called "first-line" drugs for airline pilots requiring

antihypertensive therapy. Should such a firm recommendation be made, the demonstration of unimpaired critical task performances while using these drugs would seem to be a reasonable requirement.

Hydralazine and calcium channel blockers should be accepted as antihypertensive therapy for airmen. The potential usefulness of other classes of compounds, such as low-dose angiotensin-converting enzyme inhibitors, may also be considered. Based on previous experience, guanethidine, guanadryl, ganglionic blocking agents, rauwolfia derivatives, prazosin, methyldopa, clonidine and guanabenz are considered unsuitable.

If pilots are to be granted special issuance certificates contigent upon their taking acceptable antihypertensive medications for control of their blood pressure, there must be some method to ensure that the medication is taken regularly and the blood pressure remains under satisfactory control. The recommendations from the Bethesda conference are suitable and desirable in this regard.[4]

References

1. Narins RG: Mild hypertension: A therapeutic dilemma. Kidney International 1984;26:881-894.
2. Robertson JIS: Hypertension and biochemical risk factors in airline pilots. Eur Heart J 1984;5(suppl A):15-16.
3. FAA Statistics, 1983.
4. Cardiovascular problems associated with aviation safety: Eighth Bethesda Conference of the American College of Cardiology: Task Force V: Hypertension. Am J Cardiol 1975;36:613-616.
5. Kannel WB: Importance of hypertension as a major risk factor in cardiovascular disease, in Genest J, Koiw E, Kuchel O (eds) Hypertension: Physiopathology and Treatment. New York, McGraw-Hill, 1977, pp 838-909.

6. The 1984 Report of the Joint National Committee on Detection, Evaluation, and Treatment of High Blood Pressure. Arch Int Med 1984;144:1045-1057.

7. Bulpitt CJ: Risk indicators for death in patients treated for hypertension: Emphasis on consequences for airline pilots. Eur Heart J 1984;5(suppl A):33-35.

8. Block HR: Nonpharmacologic therapy for hypertension. Am J Med 1979;66:837-842.

Recommended Revision of Form 8065-1

Instructions for Preparation and Submittal of Electrocardiogram

1. Submit only original ECG tracings. Photocopies are not acceptable.
2. ECG must be taken within 30 days prior to FAA physical examination.
3. Chest electrode placement as follows:

   V-1 At the 4th right interspace at the sternal border

   V-2 At the 4th left interspace at the sternal border

   V-3 Halfway between leads V-2 and V-4

   V-4 At the 5th left interspace on the midclavicular line

   V-5 Halfway between V-4 and V-6

   V-6 On a line dropped perpendicularly from V-4 to the midaxillary line

4. Show standardization on leads I and V-1.
5. Supply 12-lead 3-channel ECG, or if one channel recording:

   a. Cut leads I, II and III six inches long; leads AVR, AVL, AVF, and all V leads to two inches long. (Guide provided above for measurements.)

   b. Arrange leads in the order shown in line 3 above; mark lead number in upper left hand corner on the front of each segment.

6. Print applicant's name on the FRONT of the lead I portion of tracing.
7. Staple all tracings to identification card below at point indicated; tear off identification card along perforation; attach to Form FAA-8500-8, and mail to:

   Federal Aviation Administration

   Aeromedical Certification Branch

   PO Box 25082

   Oklahoma City, Oklahoma 73125

Recommended Revision of Form 8500-19

Cardiovascular Evaluation Specifications

These specifications have been developed by the Federal Aviation Administration (FAA) to determine an applicant's eligibility for airman medical certification. Standardization of examination methods and reporting is essential to provide sufficient basis for making this determination and the prompt processing of applicants. This cardiovascular evaluation, therefore, must be reported in sufficient detail to permit a clear and objective evaluation of the cardiovascular disorder(s) with emphasis on the functional capacity, prognosis, and risk for future serious incapacitating events. It must be performed by a specialist in internal medicine or cardiovascular diseases and should be forwarded to the FAA immediately upon completion. Inadequate evaluation or reporting or failure to submit the report to the FAA promptly may delay the certification decision. At a minimum the evaluation must include the following:

1. Medical History - Particular reference should be given to cardiovascular abnormalities, including cerebral or peripheral vascular diseases, a history of hypertension, stroke, myocardial infarction, syncope, edema, heart murmur, chest pain, dyspnea and arrhythmias. The results of special diagnostic procedures, such as exercise treadmill tests, angiograms, echocardiograms and nuclear studies, should be documented. A review of all prior hospitalizations should be recorded. All medications currently or recently used should be identified including the name and purpose of the medication. Side effects of permitted medication should be identified, such as drowsiness or decreased mental acuity, particularly if the patient is on a beta-receptor blocking drug.

2. Family, Personal and Social History - A statement of the ages and health status of parents and sibilings is necessary; if deceased, age at death and cause should be included. Also, an indication of whether any blood relative has had heart or vascular disease, hypertension, diabetes or known disorders of lipid metabolism should be provided. Smoking, drinking and recreational habits of the applicant are pertinent, as well as whether a program of physical fitness is being maintained. Comments on the level of physical activities, functional limitations, occupation and avocational pursuits are essential.

3. Records of Previous Medical Care - If not previously furnished to the FAA, a copy of pertinent hospital records as well as outpatient treatment records, with clinical data, radiographic and laboratory observations and copies of serial ECG tracings and exercise electrocardiograms should be provided. Detailed reports or surgical procedures as well as cerebral and coronary arteriography and other major diagnostic studies are of prime importance.

4. General Physical Examination - A brief description of any personal characteristics worthy of comment: height, weight, blood pressure readings in both arms while sitting and standing; funduscopic examination of retinal arteries; condition of peripheral arteries; carotid artery auscultation; heart size; rate; rhythm and description of murmurs (location, intensity, timing and opinion as to significance) and other findings of consequence must be provided.

5. Laboratory Data - A minimum must include actual test values of:

   A. Routine urinalysis and complete blood count.

B. Blood chemistries (values and normal ranges of the laboratory).

1. Serum cholesterol and triglycerides after 12 to 16 hour fast.

2. Fasting plasma glucose (If the fasting plasma glucose is elevated, include a two hour oral glucose tolerance test following a 75 to 100 gram carbohydrate load).

3. Serum creatinine or BUN.

4. T3 and T4, if indicated.

5. Serum electrolytes if patient has been on diuretics or has a history of hypertension.

C. Recent PA and lateral chest radiographs (provide films if abnormal).

D. Electrocardiograms

1. Resting tracing.

2. Near maximal treadmill exercise stress test, unless contraindicated, with achievement of at least 90% of maximal heart rate or symptom limited. (If applicant is taking a beta receptor blocking drug, it is suggested that if possible the applicant be tested after the beta blocker drug has been stopped if 90% of the maximal heart rate cannot be achieved during exercise testing on the drug, especially if there has not been a prior treadmill test recorded off medication).

a. State methodology used, including protocol.

b. Describe symptoms that occurred during the test.

c. Provide blood pressure and pulse determinations at rest and at each stage of the exercise stress test and during each minute of the recovery period.

d. Submit copies of the representative ECG tracings for the control, immediately at peak exercise, and for each

one minute interval for six minutes following cessation of exercise.**

E. Two dimensional and M-mode echocardiogram should be obtained in all applicants who have a significant heart murmur or cardiomegaly on the PA chest radiograph. Echocardiography may be indicated also at the cardiovascular specialist's recommendation in the evaluation of certain other suspected problems (such as VPB's or other arrhythmias).

F. Thallium exercise and resting myocardial perfusion scans may be useful in the evaluation of certain conditions (left bundle branch block, resting ECG repolarization abnormalities, atypical chest pain problems) and should be at the option of the cardiovascular specialist.

G. Exercise and resting MUGA scans or first pass ventriculogram: At the option of the cardiovascular specialist one of these tests may be also used in addition to or in place of the thallium scan.

H. Ambulatory electrocardiogram: A 24-hour ambulatory electrocardiogram should be obtained in all applicants with a history of an arrhythmia, as well as in applicants with certain other conditions such as those who have had coronary artery bypass surgery

---

**NOTE: The information obtained through a determination of current cardiovascular capacity and an evaluation of exercise end points under the stress of rhythmic exercise is considered essential to the determination of fitness of any applicant with suspected or known cardiovascular disease. Current practice indicates that a stress test on a treadmill, using either a Bruce or Blake protocol, is best for providing the desired performance data. Alternatively, an ergometer test that results in a degree of work of approximately 90% of the age-predicted maximum capacity using heart rate end points is acceptable. All usual medical precautions should be followed in pre-screening, election to test, testing and follow-up on applicants who undergo exercise stress testing. The resting tracing should be reviewed by the examining physician for evidence of acute coronary insufficiency, recent myocardial infaction, or repolarization abnormalities. ECG evidence of recent, unsuspected myocardial ischemia or infarction would contraindicate exercise testing. Please state reasons if the exercise stress test is medically contraindicated.

or a myocardial infarction, and in certain applicants with valvular heart disease. The analysis of certain arrhythmias may require 48 to 72 hour recordings.

L. Cardiac catheterization and coronary arteriograms may be required for certain applicants who have had a myocardial infarction, coronary artery bypass surgery or coronary angioplasty. Cardiac catheterization may be necessary in certain other conditions such as aortic or pulmonary stenosis if noninvasive clinical data are not conclusive.

Recommendations for Cardiovascular Drugs

On the following page is a table of commonly used cardiovascular drugs. The recommendations of the cardiovascular committee regarding the acceptability of these drugs for special issuance represent their best judgments at this time. New drugs for cardiovascular conditions are always being formulated and experiences with older drugs may call for alterations in treatment regimens and expected side effects. Therefore, the table should not be interpreted strictly, and the FAA should frequently consult cardiovascular specialists for updated recommendations.

RECOMMENDATIONS FOR CARDIOVASCULAR DRUGS

Drug	Allowable(+) Not Allowable(-)	Recommended Maximum Allowable Dose (mg/day)*	Period of Surveillance prior to certification	Surveillance Tests: BP, sitting and standing	Surveillance Tests: Serum Potassium	Evidence of Drowsiness and Diminished Mental Capacity
Metalazone	+			+	+	
Amiloride/ Dyrenium	+			+	+	
Chlorthalidone	+			+	+	
Thiazide	+			+	+	
Furosemide	+			+	+	
Atenolol	+	50	90	+		+
Propranolol	+	160	90	+		+
Nadolol	+	80	90	+		+
Metoprolol	+	100	90	+		+
Pindolol	+	20	90	+		+
Timolol	+	20	90	+		+
Norpace	+		6 months**	+		
Quinidine	+		6 months**	+		
Pronestyl	+		6 months**	+		
Heparin	-					
Coumadin	-					
Nitrates	-					
Prazosin	-					
Apresoline	+	200	6 months	+		
Captopril	+		6 months	+		
Methyldopa	-					
Clonidine	-					
Ganglionic Blockers	-					
Reserpine	-					
Minoxidil	-					
Verapamil	+	320	6 months	+		
Nifedipine	+	60	6 months	+		
Diltiazem	+	240	6 months	+		
Digitalis	+		6 months			
Aspirin	+					
Dypridamole	+					
Anturane	+					

*If no dose is given, the drug may be used within its usual therapeutic range

**Approved for use for supraventricular tachycardia. There should be proof of electrical stability throughout observation period before special issuance is granted

The Nervous System

The purpose of this report is to provide the FAA with current information on neurological conditions most likely to be encountered in civilian airmen, and to update the AMA report of 1979 on "Neurological and Neurosurgical Conditions Associated with Aviation Safety."[1] The emphasis in this report is placed on providing information that the AME and FAA must have to make appropriate dispositions of airmen with neurological conditions. This information includes personal and family histories, the results of neurological and laboratory investigations, consultations, follow-up examinations, and most important, prognosis, particularly as they apply to flying performance and/or the risk of sudden incapacitation in flight. Studies on the natural history of certain neurological conditions were not designed specifically to answer questions about flying safety. Therefore, one has to extrapolate from the available clinical literature.

This report of neurological conditions did not go through the thorough internal review process that was mentioned in the foreword (p i). The information contained within on specific neurological conditions is scientifically and clinically sound, and the recommendations for aeromedical disposition are appropriate given that information, but the FAA should be aware that the recommendations and the rationale for them did not benefit from peer review.

The most common neurological disorders by average annual incidence and point prevalence rates per 100,000 population and at all ages, are listed in Tables I and II. Only those conditions that are prevalent in the pilot age group and that may affect flight safety are addressed below in subsequent sections.

TABLE I

MOST COMMON NEUROLOGICAL DISORDERS: APPROXIMATE AVERAGE ANNUAL INCIDENCE RATES PER 100,000 POPULATION. ALL AGES

Disorder	Rate
Herpes zoster	400
Migraine	250
Brain trauma	200
Other severe headache[a]	200
Acute cerebrovascular disease	150
Other head injury[a]	150
Transient postconcussive syndrome	150
Lumbosacral herniated nucleus pulposus	150
Lumbosacral pain syndrome[a]	150
Neurological symptoms without defined disease	75
Epilepsy	50
Febrile fits	50
Dementia	50
Meniere's disease	50
Mononeuropathies	40
Polyneuropathy	40
Transient ischemic attacks	30
Bell's palsy	25
Single seizures	20
Parkinsonism	20
Cervical pain syndrome[a]	20
Persistent postconcussive syndrome	20
Alcoholism[a]	20
Meningitides	15
Encephalitides	15
Sleep disorders[b]	15
Subarachnoid hemorrhage	15
Cervical herniated nucleus pulposus	15
Metastatic brain tumor	15
Peripheral nerve trauma	15
Blindness	15
Benign brain tumor	10
Deafness[a]	10

[a]Rates are for those a neurologically competent physician should see (10% of total)

[b]Narcolepsies and hypersomnias

source: REF 1

TABLE II

MOST COMMON NEUROLOGICAL DISORDERS: APPROXIMATE POINT PREVALENCE RATES PER 100,000 POPULATION. ALL AGES

Disorder	Rate
Migraine[a]	2000
Other severe headache[a]	1500
Brain injury	800
Epilepsy	650
Acute cerebrovascular disease	600
Lumbosacral pain syndrome[a]	500
Alcoholism[a]	500
Sleep disorders[b]	300
Meniere's disease	300
Lumbosacral herniated nucleus pulposus	300
Cerebral palsy	250
Dementia	250
Parkinsonism	200
Transient ischemic attacks	150
Febrile fits	100
Persistent postconcussive syndrome	80
Herpes zoster	80
Congenital CNS malformations	70
Single seizures	60
Multiple sclerosis[c]	60
Benign brain tumor	60
Cervical pain syndrome[a]	60
Down's syndrome	50
Subarachnoid hemorrhage	50
Cervical herniated nucleus pulposus	50
Transient postconcussive syndrome	50
Spinal cord injury	50

[a]Rates are for those a neurologically competent physician should follow (20% of migraine, 10% of all others)

[b]Narcolepsies and hypersomnias

[c]Rate is for high-risk areas

source: REF 1

Epilepsy and Single Seizure

Epilepsy is derived from a Greek word meaning "a condition of being overcome, or seized, or attacked." Although there is no universally accepted definition, it can be defined as "a group of conditions characterized by recurring convulsions for a group of disorders in which the common factor is a paroxysmal excessive, neuronal discharge within the brain."[2] Another definition is "convulsive disorders characterized by sudden, brief, repetitive and stereotyped alterations of behavior which are presumed to be due to a paroxysmal discharge of cortical or subcortical neurones."[3] Therefore, epilepsy means more than one convulsion. Up to 5% of a general population will at some time suffer a well-defined, non-febrile epileptic convulsion. The prevalence rate for chronic epilepsy ranges between 4/1000-10/1000.

An International Classification of Epileptic Seizures was proposed in 1981, and is shown in Table I. More recently, the Commission on Classification and Terminology of the International League Against Epilepsy has proposed a new classification of epilepsies and epileptic syndromes as shown in Table II. Since both classifications are used, the ME and FAA should become familiar with both.

Considerations for airman certification

Most persons with epilepsy and epileptic syndromes as shown in Tables I or II will never apply for certification because of persistence of seizures, the nature of the disease causing the fits, or other associated neurological abnormalities. This is particularly true for persons with symptomatic localization-related seizures; with some of the idiopathic, generalized epilepsies and symptomatic, generalized epilepsies; and with some of the special syndromes. The AME and FAA from time to time will have to make decisions concerning applicants with a past history or family history of seizures, or who have experienced their first seizures after initial medical licensure. Since pilots taking anti-

TABLE I

INTERNATIONAL CLASSIFICATION OF EPILEPTIC SEIZURES

I. Partial seizures (Seizures beginning locally)

- A. Simple partial seizures (consciousness not impaired)
  - 1. With motor symptoms
  - 2. With somatosensory or special sensory symptoms
  - 3. With autonomic symptoms
  - 4. With psychic symptoms
- B. Complex partial seizures (with impairment of consciousness)
  - 1. Beginning as simple partial seizures and progressing to impairment of consciousness
  - 2. With impairment of consciousness at onset
    - a. With impairment of consciousness only
    - b. With automatisms
- C. Partial seizures secondanly generalized

II. Generalized seizures (bilaterally symmetrical and without local onset)

- A. 1. Absence seizures
  2. Atypical absence
- B. Myoclonic seizures
- C. Clonic seizures
- D. Tonic seizures
- E. Tonic-clonic seizures
- F. Atonic seizures

III. Unclassified epileptic seizures (incomplete data)

source: REF 4 with modifications

TABLE II

INTERNATIONAL CLASSIFICATION OF EPILEPSIES AND EPILEPTIC SYNDROMES

1. Localization-related (focal, local, partial) epilepsies and syndromes

1.1 Idiopathic with age-related onset

At present, two syndromes are established, but more may be identified in the future:

- o Benign childhood epilepsy with centrotemporal spike
- o Childhood epilepsy with occipital paroxysms

1.2 Symptomatic

This category comprises syndromes of great individual variability, which will mainly be based on anatomical localization, clinical features, seizure types, and etiological factors (if known). Major examples and descriptions of varieties subsumed under this heading are given in Appendix I.

2. Generalized epilepsies and syndromes

2.1 Idiopathic, with age-related onset, listed in order of age

- o Benign neonatal familial convulsions
- o Benign neonatal convulsions
- o Benign myoclinic epilepsy in infancy
- o Childhood absence epilepsy (pyknolepsy)
- o Juvenile absence epilepsy
- o Juvenile myoclonic epilepsy (impulsive petit mal)
- o Epilepsy with grand mal seizures (GTCS) on awakening

Other generalized idiopathic epilepsies, if they do not belong to one of the above syndromes, can still be classified as generalized idiopathic epilepsies.

2.2 Idiopathic and/or symptomatic, in order of age of appearance

- o West syndrome (infantile spasms, Blitz-Nick-Salaam Krampfe)
- o Lennox-Gastaut syndrome
- o Epilepsy with myoclonic-astatic seizures
- o Epilepsy with myoclonic absences

2.3 Symptomatic

2.3.1 Nonspecific etiology

- o Early myoclonic encephalopathy

2.3.2 Specific syndromes

- o Epileptic seizures may complicate many disease states

Included under this heading are those diseases in which seizures are a presenting or predominant feature. These are detailed in Appendix II.

TABLE II CONT'D

3. Epilepsies and syndromes undetermined as to whether they are focal or generali
   - 3.1 With both generalized and focal seizures
     - o Neonatal seizures
     - o Severe myoclonic epilepsy in infancy
     - o Epilepsy with continuous spike-waves during slow wave sleep
     - o Acquired epileptic aphasia (Landau-Kleffner syndrome)
   - 3.2 Without unequivocal generalized or focal features
     This heading covers all cases with GTCS whare clinical and EEG findings do not permit classification as clearly generalized or localization-related, such as in many cases of sleep grand mal.

4. Special syndromes
   - 4.1 Situation-related seizures (Gelegenheitsanfalle)
     - o Febrile convulsions
     - o Seizures related to other identifiable situations such as stress, hormonal changes, drugs, alcohol, or sleep deprivation
   - 4.2 Isolated, apparently unprovoked epileptic events
   - 4.3 Epilepsies characterized by specific modes of seizure precipitation
   - 4.4 Chronic progressive epilepsia partialis continua of childhood

source: REF 5

convulsant medications are not allowed to fly, the major question to be dealt with is, whether the epilepsies ever go into permanent remission and, if so, which types?" Another way of asking this question is, "How long does a person not presently taking medication need to be seizure-free before his or her epilepsy is considered permanently cured, or his or her risk of having a convulsion is no greater than the risk found in the population at large?" We will try to answer this question for specific epileptic syndromes.

Childhood and adolescent epilepsy

The studies to date indicate a favorable prognosis for some forms of epilepsy, particularly those beginning in childhood or adolescence. However, they do not provide a definite answer to our question. In Table III the relapse rates of seizures following discontinuance of anti-convulsant therapy are shown. These data show that the relapse rate ranges from 24% to 36%. However, the duration of follow-up in each study is limited with the exception of the study by Holowach Thurston, et al, who followed their patients from 15 to 23 years. The largest percentage of relapses occur in the first year after withdrawal. In the study by Holowach Thurston et al, only two relapses occurred after 8 years after discontinuation of anti-convulsant medication, one in the 16th and one in the 18th year. Because of the long remission in these patients, they questioned whether these late relapses were related to the original epileptic process, or whether they represented new convulsive disorders. Annegers et al state that all relapses occur in the first 20 years.[6] There is some disagreement in the literature on the factors predictive of a relapse. In Table IV, the relapse rates according to seizure type from three studies are shown. It is clear that there is a wide variation. However, most authorties feel that the chances for relapse are greater for seizures that begin focally, with the exception of benign Rolandic epilepsy excluded, and for mixed seizures. Other important factors are duration of illness before control, the age of onset, the presence of

TABLE III

RELAPSE RATE AFTER WITHDRAWAL OF ANTICONVULSANTS IN CHILDREN AND ADOLESCENTS WITH EPILEPSY

Authors	Duration of Follow-up	% Relapse	Time of Relapse
Emerson et al[6]	6 months to 6 years	26%	78% in first yr
Holowach et al[7]	5 to 12 years	24%	56% in first yr 68% in first 2 yr 85% within 5 yrs
Holowach-Thurston et al[8]	15 to 23 years	28%	Only 2 relapses after 8 years (16th and 18th yr
Todt[9]	5 to 6 years average	36.3%	86% in first yr 98% within 3 yrs 1.3% in 4th yr
Skinner et al[10]	6 months to 5 years	25%	82% in first yr
Annegers et al[11]	-----	-----	Most relapses in first 10 years and all in the first 20 years

TABLE IV

RELAPSE RATES BY SEIZURE TYPE

Seizure Type	AUTHORS * Holowach Thurston[8] et al	Emerson[6] et al	Skinner[10] et al	Total
Grand Mal	5/36 (14%)	11/25 (44%)	15/47 (32%)	31/108 (
Jacksonian or Focal Motor	11/19 (58%)	0/10 (0%)	8/30 (27%)	19/59 (3
Psychomotor	5/16 (31%)	4/4 (25%)	3/27 (11%)	9/47 (19
Simple Febrile	4/32 (12%)	-----	-----	4/32 (12
Petit Mal	1/8 (12%)	0/1 (0%)	1/6 (17%)	2/15 (13
Myoclonic	0/2 (0%)	0/1 (0%)	-----	0/3 (0%)
Mixed Seizure Types	15/35 (43%)	5/19 (26%)	-----	20/54 (3
Atypical Absence	-----	1/4 (25%)	5/18 (28%)	6/22 (27
Complex or Atypical Febrile	-----	0/4 (0%)	6/13 (46%)	6/17 (35

$$\text{Relapse Rate} = \frac{\text{\# Patients with Relapse}}{\text{\# Patients with Seizure Type}}$$

*In addition, Todt (9) showed an increased incidence of relapse in patients with partial epilepsies, in patients with secondary generalized partial seizures, and in patients with combined seizures.

other neurological dysfunctions, and EEG abnormalilties.

The data presented suggests that permanent remissions of epilepsy of childhood and adolescence may occur, although longer follow-up studies are still needed. The data also suggest that if a patient with onset of seizures in childhood or adolescence remains seizure free while off medication for approximately 20 years, his or her chance of having a subsequent seizure is probably no greater than the chance in the population at large.

Therefore, for an applicant with a past history of epilepsy, the following criteria should be met: 1) the applicant must be seizure-free for at least 20 years off all anticonvulsant medication; 2) the applicant must have no abnormalities on a neurologic examination, performed by a neurologist; 3) the applicant must have a normal awake EEG with hyperventilation and photic stimulation, as well as with sleep; 4) the applicant must have a negative blood screen for anti-convulsants. In some applicants a CT scan with or without enhancement may be indicative, if so, then it must be normal. All of the applicants past medical records including consultations, EEGs and radiographs must be obtained before the final decision is made.

Single unprovoked seizure in childhood

The risk of a second convulsion after a first unprovoked seizure in childhood is approximately 52%.[12] Of those persons who had a recurrence, 79% had additional seizures. The lowest rate of recurrence followed a generalized tonic-clonic seizure with a normal EEG and neurological examination. Therefore, in general, an applicant with a history of a single unprovoked seizure in childhood should meet the same criteria that the applicant with childhood epilepsy must meet before a certificate may be issued.

Other epileptic syndromes of neonatology, children and adolescents

Benign neonatal familial convulsions: This is a dominant, inherited disorder with clonic or apneic seizures beginning on the second or third day of life. The seizures are

self-limited. Fourteen percent of these infants develop epilepsy late. If an applicant with a history of this seizure disorder has had no further seizures over a 20-year period, while off medication and has a normal neurological examination and EEG, he or she may be considered for certification.

Benign neonatal convulsions: These are repeated clonic or apneic seizures that first occur around the fifth day of life, without a known etiology. The seizures are self-limited. The disposition should be the same as for benign neonatal familiar convulsions.

Febrile seizures: A febrile seizure is defined as "a seizure in infancy or childhood, usually occurring between three months and five years of age, associated with fever, but without evidence of intracranial infection or recognized acute neurological illness."[13] Approximately 2 to 5% of all children will experience convulsions with febrile illnesses before age 5,[14] and approxmately 30% of these children experience more than one febrtile seizure.[13] Approxmately 75% of febrile seizures are brief and self-limited, and are generalized tonic-clonic convulsions; these are known as simple febrile seizures.[13] Complex febrile convulsions last longer than 15 minutes, occur more than once in a 24-hour period or have focal features.[13] Approximately 25% of febrile convulsions are complex.[13] An EEG performed soon after a febrile seizure is likely to show either bilateral or unilateral diffuse, slow waves. In EEG follow-ups of persons with febrile seizures, a surprisingly large proportion of persons with febrile seizures (40% or more) will show spike waves on subsequent EEGs.[14] However, it should be pointed out that the finding of definite epileptiform abnormalities or the EEG cannot be used solely as a predictor of the subsequent development of either recurrent febrile seizures or epilepsy.

Genetic factors are important. Febrile convulsions occur in parents or siblings at a rate 2 to 3 times higher than the general population.[14] Data are inconclusive as to whether non-febrile seizures are more common in families of individuals with febrile seizures.

In cohort studies of persons with febrile convulsions, the incidence of the subsequent development of non-febrile seizures ranges between 2% and 6%.[13] Thus, the risk for non-febrile convulsions is increased several fold over that expected for the general population. The increased risk is most marked in the first three years after the initial febrile seizure, during which time 75% of these individuals will experience their first non-febrile convulsions.[13] However, a slight increase in risk persists at least until the third decade of life.[15] Neurological abnormalities prior to the febrile seizure, prolonged febrile seizures, a family history of epilepsy, and the occurrence of complex features during the febrile seizure increase the risk for the subsequent development of epilepsy.[13,14] There is no evidence to date that more than one febrile seizure in the absence of the risk factors mentioned above increases the risk of subsequent epilepsy.[13]

Therefore, an applicant who has a history of one or more febrile seizures may be granted a certificate if: a) the febrile seizure(s) occurred between the ages of three months and five years; b) the seizure did not last longer than 15 minutes; c) not more than one seizure occurred in a 24-hour period; d) there are no atypical features, such as focal seizures with or without post-ictal transient paralysis; e) there is no first-degree relative with a history of epilepsy and; f) the present neurological examination is normal and the EEG, which should include an awake and sleep recording as well as hyperventilation and photic stimulation, shows no paroxysmal abnormalities.

Family history of epilepsy: Annegers et al, reported that the risk of epilepsy through age 20 among the siblings and children of persons with childhood epilespy is three times the rate in the general population.[16] Furthermore, they report that the risk of any type of seizure, which includes isolated seizures, febrile seizures and acute cerebral insult seizures, is 11% in the siblings and 13.1% in the children. Therefore, applicants with a history of childhood-onset epilespy in either their parents or siblings, should be examined by a neurologist and undergo an EEG examination. If EEG shows a specific epileptiform abnormality, no certificate should be issued.

Benign childhood epilepsy with centro-temporal spikes (benign rolandic epilepsy of childhood): This is a seizure disorder with onset between three and 13 years of age; usually there is recovery before ages 15 or 16.[17] It is genetically determined with male predominance. The seizures are brief, simple, partial, hemi-facial motor seizures, which may become generalized. They often occur during sleep. Although it is an apparently self-limiting type of epilepsy, there is one report[18] of relapse in adulthood in a patient who had been seizure free for 8 years, the last 6 years without medication.

Before an applicant with this type of epilepsy may be granted a certificate, he or she must be seizure-free for 10 years off medication, have a normal awake and sleep EEG, and have a normal neurologic examination, performed by a neurologist.

Childhood epilepsy with occipital paroxysms: The seizures in this condition start with visual symptoms, such as amaurosis, phosphenes, illusions, or hallucinations. They are often followed by a hemi-clonic seizure or automatisms. A migraine-like headache follows the seizure in 25% of cases. The EEG shows rhythmical, paroxysmal, high amplitude spike-and-wave, or sharp waves over the occipital and posterior temporal areas. Because this is a newly recognized type of epilepsy,[19] no statement on prognosis can be made. Therefore, an applicant with this type of seizure disorder should not be issued a certificate regardless of how long he or she has been free of seizures.

Adult epilepsy

Approximately 25% of persons with epilepsy will have their first seizure after the age of 25 years. The major etiologies for these unprovoked seizures are unknown (38%), alcohol non-withdrawal seizures (25%), brain tumor (16%), cerebrovascular infarctions (14%), head trauma (4%), and miscellaneous (5%).[20]

When evaluating an applicant with adult-onset multiple seizures, the physician must first make sure that a treatable cause has not been overlooked, particularly for provoked convulsions (see below). Therefore, if the applicant has not seen a neurologist,

a consultation should be obtained. In those candidates in which a provoked cause is not responsible for the convulsions, or if the neurological assessment does not reveal any abnormalities which by themselves would prohibit flying, then the same recommendations as for childhood and adolescent epilepsy apply.

Provoked seizures

Provoked seizures are those that occur in association with an acute neurological insult, such as head trauma, stroke or infection, and that occur with an acute systemic metabolic disturbance, such as hypoglycemia, uremia, or drug or alcohol withdrawal. Stress, particularly sleep deprivation, may also precipitate a seizure.

Acute neurology insults: Generalized tonic-clonic seizures and/or partial seizures may occur during the course of acute neurological insults. They also may have their onset following recovery from the acute injury, which is called remote systematic epilepsy. Unfortunately, with the exception of head trauma, there is little information in the literature on the late development of post-injury epilepsy for the other conditions. In a study of 65 cases of remote symptomatic epilepsy (24 with head trauma, 19 with cerebrovascular disease, 3 with central nervous system infection, and 19 with other causes), 31% of the cases suffered a recurrence of seizures, all of which occurred within 20 months of the initial seizure.[21]

Acute systemic metabolic and toxic encephalopathy: Seizures that occur during metabolic disturbances usually are self-limited, and if the underlying cause is permanently corrected, will not occur again. They also may occur as an idiosyncratic reaction to a drug(s) or during withdrawal from alcohol and certain drugs, particularly hypnotics and sedatives. In a study of 53 patients with drug induced seizures, 45% had single seizures, 40% had multiple convulsions and 15% had status epilpeticus.[22] The authors noted that generalized seizures with focal features were common. The most common drugs that caused seizures were isoniazid, insulin lidocaine, and psychotropic

medications. In Table V drugs reported to cause convulsions are listed.

Before issuing a certificate the FAA must have irrefutable evidence that the convulsions were secondary to acute metabolic or toxic causes. In addition, the underlying abnormality must be evaluated and corrected, and the applicant must satisfy the certification criteria for that abnormality. Finally, the applicant should not have suffered any significant neurological deficit as a result of the acute illness or seizures. Psychometric testing, EEGs and/or CT scan may be required if there are any doubts about the cause or residual neurological deficits.

Stress-precipitated seizures: Seizures may occur after various stresses, for example, sleep deprivation,[23] fatigue, overexertion or emotional stress. Friis and Lund estimate the incidence of "stress convulsions" to be about 1 per 100,000 patients. Of 37 patients they followed for 1-12 years, 24 remained seizure free, 10 of whom were not taking anti-convulsant medications. The remainder of the patients had one or more subsequent stress-provoked attacks. Two of the 36 patients developed spontaneous convulsions.[24]

In a study of sleep deprivation convulsions in 40 soldiers, Gunderson et al, found a history suggestive of previous sleep deprived seizures in six and possibly seven of the soliders.[25]

Although somewhat limited, the data on stress-precipitated seizures indicate a propensity for recurrence and for the development of unprovoked seizures. Therefore, any applicant with a history of a stress-induced seizures should not be certified, unless a period of 20 years has elapsed since the last seizure and the applicant has been off anti-convulsant medication for this period of time.

Other seizures

Convulsive syncope: Syncope is caused by an acute reduction in brain oxygenation, most often as a result of a drop in systemic blood pressure. In

TABLE V

DRUGS REPORTED TO CAUSE CONVULSIONS

Aqueous iodinated contrast agents

Anticholinesterase agents (organophosphates, physostigmine)

Antihistamines

Antidepressants

Antipsychotics

Baclofen

Beta blockers (propranolol, oxpreolol)

Camphor

Chlorambucil

Cocaine

Cycloserine

Cyclosporin A

Ergonovine

Folic acid

General anesthetics (ketamine, halothane, Althesin, enflurane, propanidid)

Hyperbaric oxygen

Hypoglycemic agents

Hyposmolar parenteral solutions

Isoniazid

Local anesthetics (bupivacaine, lidocaine, procaine, etidocaine)

Mefenamic acid

Methylxanthines

Metronidazole

Misonidazole

Nalidixic acid

Narcotic analgesics (fentanyl, meperidine, pentazocine, propoxyphene)

Oxytocin (secondary to water intoxication)

Penicillins

Phencyclidine

Phenobarbital

Phenytoin

Prednisone (with hypocalcemia)

Sympathomimetics (amphetamines, ephedrine, phenylpropanolamine, terbutaline)

Vitamine K oxide

source: REF 22

approximately 12% of patients with syncope, some type of convulsive movements may occur.[26] In the study by Lin et al, tonic spasms occurred in 23 patients, during which the patients become pale and diaphoretic. The motor activity may resemble that seen in epileptic seizures. Knowledge of the precipitating event, and the presence of facial pallor and the rapid recovery of consciousness without amnesia help distinguish convulsive syncope from epilepsy. Persons who faint may fall and suffer a secondary head injury, which can produce a concussion causing delayed recovery and amnesia. If the concussion is associated with convulsive movements, the physician may mistakenly make a diagnosis of epilepsy. More serious complications such as intracranial hematoma have also been reported.

An applicant with a history of convulsive syncope may be granted a certificate if: 1) the cause of the syncope is not disqualifying; 2) the applicant presently has a normal neurological examination; and 3) an awake EEG with hyperventilation and photic stimulation, and a sleep EEG do not show any epileptiform discharges. If the applicant suffered a head injury during the syncopal episode, criteria for certification for head injuries should be used (see below).

Pseudoseizures: In neurological practice, a not uncommon problem is the differentiation of true seizures from hysterical events. Several recent publications have addressed this problem.[27-29] Although a hysterical cause can be suspected from observation of the seizure, the absolute test is the recording of a normal EEG during the episode. A normal ictal EEG with true seizures is exceedingly rare. Motor activity occurring during the pseudo-ictal event varies considerably. Tonic posturing and tremulousness followed by jerking, grimacing, thrashing, bicycling-like movements, pelvic thrusts and partial opisthotonic postures may be seen. The movements may be bilateral or unilateral. They may be sterotyped from seizure to seizure. Many of these persons will have their eyes tightly closed during the event, a finding not often seen in true seizures. Urinary and fecal incontinence is very rare. Hysterical seizures may

occur in persons with known epilepsy. Suggestion may precipitate the seizures. Although the pseudo-seizure itself might not disqualify an applicant, the underlying psychopathology would preclude the issuance of a license.

Single Unprovoked Seizure in Adults

The data on the recurrence of seizures after an attack of a single, unprovoked seizure is found in Table VI.

It is apparent that the wide variation in percent recurrence, ranging from 27%-82% reflects different study designs and duration of follow-up. Most of the patients in these studies were placed on anti-convulsant medication after their initial seizure. However, in the investigation by Hauser et al, of the 64 patients with idiopathic epilepsy who were not started on anti-epileptic medication, 15% suffered relapses, as compared to 20% of those patients who were treated.[31] In the study by Johnson et al, of 77 enlisted Navy men whose first seizures were considered idiopathic and who were not placed on anti-convulsant medications, there was a 64% recurrence of seizure within the subsequent three years, with 77% of the seizures occurring within the first year.[32]

Thus, the literature shows quite clearly that, regardless of whether a person is taking anti-convulsant medication, the likelihood of suffering convulsions subsequent to a single, unprovoked seizure is high. Thus, an applicant with a history of a single seizure must remain seizure-free and off all anti-convulsant medications for at least 20 years before he or she should be certified.

TABLE VI

STUDIES OF THE RECURRENCE OF SEIZURES AFTER A FIRST ATTACK

Author	"	Patient Selection	Follow up*	% Recurrence+
HOSPITAL BASED STUDIES				
Thomas (1959)	48	Referred to EEG dept	ns	27
Johnson et al (1972)	77	Young adult males,	6 mns	34
		naval recruits (excluding	12 mns	49
		organic causes, alcohol,	24 mns	57
		drug abuse)	36 mns	58
Saunders and Marshall (1975)	33	Referred to EEG dept	10-48 mns	33
Cleland et al (1981)	70	Adult referrals to a neurology clinic, with untreated "major" seizures, (excluding head injury, drug overdose)	3-120 mns	39
Hauser et al (1982)	244	"Unprovoked seizures"	12 mns	16
		(incl multiple seizures	24 mns	21
		on a single day, excl	36 mns	27
		acute symptomatic fits)	> 36 mns	27
COMMUNITY BASED STUDIES				
Hauser & Kurland (1975)	769	Survey of Rochester MN including recurrent provoked seizures as single seizures)	> 24 mns	67
Goodridge & Shorvon (1983)	114	Survey of Tonbridge, Kent (all seizures regardless of actiology)	> 36 mns	82

*Period of follow up after first seizure

+Percentage of patients in whom a second seizure occurs during the specified follow up period

source: REF 30

References

1. Kurtzke JF: Neuroepidemiology. Ann Neurol 1984;16:265-277.
2. Robb JP: Epilepsy: A Review of Basic and Clinical Research. Bethesda MD, National Institute of Neurological Diseases and Blindness, 1965.
3. Kurland LT, Kurtzke JF, Goldberg ID (eds): Epidemiology of Neurologic and Sense Organ Disorders. Cambridge, Havard University Press, 1973.
4. Dreifuss FE, Bancaud J, Henriksen D, et al: Proposal for revised clinical and electroencephalographic classification of epileptic seizures. Epilepsia 1981;22:489-501.
5. Commission on Classification and Terminology of the International League Against Epilepsy. proposal for classification of epilepsies and epileptic syndromes. Epilepsia 1985;26:268-278.
6. Emerson R, D'Souza BJ, Vining EP, et al: Stoping medication in cildren with epilepsy. New Engl J Med 1981;304:1125-1129.
7. Holowach J, Thurston DL, O'Leary J: Prognosis in childhood epilepsy: Follow-up study of 148 cases in which therapy had been suspended after prolonged anti-convulsant control. New Engl J Med 1972;286:169-174.
8. Holowach-Thurston J, Thurston DL, Hixon BB, Keller AJ: Additional follow-up of 184 children 15-23 years after withdrawal of anti-convulsant therapy. New Engl J Med 1982;306:831-836.
9. Todt H: The late prognosis of epilepsy in childhood: Results of a prospective follow-up study. Epilepsia 1984;25:137-144.
10. Skinner S, Vining EPG, Mellits ED, et al: Discontinuing anti-epileptic medication in children with epilepsy after two years without seizures: A prospective study. New Engl J Med 1985;313:976-980.

11. Annegers JF, Hauser WA, Elveback LR: Remission of seziures and relapse in patients with epilepsy. Epilepsia 1979;20:729-737.

12. Camfield PR, Camfield CS, Dooley JM, et al: Epilepsy after a first unprovoked seizure in childhood. Neurology 1985;35:1657-1660.

13. Nelson KB, Ellenberg JH: Febrile seizures, in: Dreifuss FE (ed): Pediatric Epileptology. Boston, John Wright PSG Inc, 1983.

14. Hauser WA: The natural history of febrile seizures, in: Nelson KB, Ellenberg JH (eds): Febrile Seizures. New York, Raven Press, 1981.

15. Annegers JF, Hauser A, Elveback LR, et al: The risk of epilepsy following febrile convulsions. Neurology 1979;29:297-308.

16. Annegers JF, Hauser WA, Anderson VE, Kurland LJ: The risks of seizure disorders among relatives of patients with childhood onset epilepsy. Neurology 1984;32:174-179.

17. Loiseau P, Pestre M, Dartigues JF, et al: Long-term prognosis in two forms of childhood epilepsy: Typical absence seizures and epilepsy with Rolandic (centrotemp oral) EEG foci. Ann Neurol 1983;13:642-648.

18. Ambrosetto G, Tiniyser P, Baruzzi, A: Relapse of benign partial epilepsy of children in adulthood: Report of a case (letter). J Neurol Neurosurg and Psychiatry 1985;48:90.

19. Gastaut H: A new type of epilepsy: Benign partial epilepsy of childhood with occipital spike waves. Clin Electroencephalography 1982;13:13-22.

20. Dam AM, Fuglsany-Frederiksen A, Svarre-Olsen V, Dam M: Late-onset epilepsy: Etiologies, types of seizures, and value of clinical investigation, EEG, and computerized tomographic scan. Epilepsia 1985;26:227-231.

21. Hauser WA, Anderson VE, Loweenson RB, McRoberts SM: Seizure recurrence after a first unprovoked seizure. New Engl J Med 1982;307:522-528.

22. Messing RO, Closson RG, Simon RP: Drug induced seizures: A 10 year experience. Neurology 1984;34:1582-1586.

23. Bennett DR: Sleep deprivation and major motor seizures. Neurology 1963;13:953-958.

24. Friis ML, Lund M: Stress convulsions. Arch Neurol 1974;31:155-159.

25. Gunderson CH, Dunne P, Feyer TL: Sleep deprivation seizures. Neurology 1973;23:678-686.

26. Lin JT-Y, Ziegler DK, Lai CW, Bayer W: Convulsive syncope in blood donors. Ann Neurol 1982;11:525-528.

27. Lesser RP, Lueders H, Dinner D: Evidence for epilepsy is rare in patients with psychogenic seizures. Neurology 1983;33:502-504.

28. Golic TA, Spinks IP, King DW: Pseudoseizures: Ictal phenomena. Neurology 1982;32:24-30.

29. Cohen RJ, Suter C: Hysterical seizures: Suggestion as a provocative EEG test. Ann Nueorl 1982;11:391-395.

30. Shorvon SD: The temporal aspects of prognosis in epilepsy. J Neurol; Neurosurg and Psychiatry 1984;47:1157-1165.

31. Hauser WA, Anderson VE, Loewenson RB, et al: Seizure recurrence after a first unprovoked seizure. N Engl J Med 1982;307:522-528.

32. Johnson LC, DeBolt WL, Long MT, et al: Diagnostic factors in adult males following initial seizures: A 3-year follow-up. Arch Neurol 1972;27:193-197.

Head Trauma

Introduction

Head injuries occur very frequently in our modern, mechanistic society. Because of the great diversity of acute neurological abnormalities, complications and long range sequelae that may result, the AME and FAA often are confronted with difficult decisions in making an appropriate decision about certification of an airman with a history of head injury. For an excellent review of the acute and late effects of head trauma, the reader is referred to the monograph by Jennett and Teasdale.[1]

Epidemiology

Traumatic head injuries from all causes exceed 1,000,000 new cases annually. Approximately 200-300 per 100,000 population are admitted to hospitals each year in the United States and Great Britain. Approximately 20% of head injury cases seen the the emergency room are admitted to the hospital. The most frequent injured are males between the ages of 15 and 24 years.

Minimal criteria for brain injury

When evaluating a candidate with a history of head injury, the AME and FAA must first determine that a brain injury has occurred. The minimal criteria for brain injury following head trauma are: 1) loss of consciousness or amnesia (antegrade or post-traumatic) even if lasting only several seconds (concussion); 2) evidence of a neurological deficit resulting from the trauma, such as a hemiparesis or aphasia; 3) evidence of focal brain damage demonstrated by special diagnostic studies such as CT scan; 4) impairment of brain function demonstrated by appropriate psychological studies.[2]

Severity of brain injury

Since the majority of persons with a history of head trauma will be evaluated by the FAA at some time after the traumatic event, the assessment of the severity of the inital brain injury, as well as intermediate and late sequelae, will have to be determined by a critical review of the applicant's records. These should include emergency room or hospital in-patient records, reports of consultants, a review of the radiographs, EEGs, psychometric tests and other studies. The details surrounding the head injury; the cause, such as missile vs nonmissile; and the type, that is, closed or open, should be determined. Almost all closed head injuries are associated with a disturbance of consciousness due to diffuse cerebral hemisphere or brain stem injury. The single most important index of severity is the degree of non-responsiveness. This can be determined retrospectively by determining the duration of post-traumatic amnesia or knowng the score on the Glasgow Coma Scale.

Post-traumatic amnesia (PTA)

The duration of posttraumatic amnesia is the time interval after the head injury during which the patient is unable to recall on-going events; that is, new memories can not be formed. This state includes persons in varying levels of coma as well as those who, although appearing awake and alert, cannot remember recent events. Based on the duration of PTA, the severity of head injury is classified as follows:

Duration	Severity
Less than 5 minutes	Very mild
5 - 60 minutes	Mild
1 - 24 hours	Moderate
1 - 7 days	Severe

1 - 4 weeks	Very severe
More than 4 weeks	Extremely severe

The longer the duration of PTA, the greater the chance of serious sequelae.

Retrograde amnesia, which is the inability to recall events prior to the injury, is a less accurate indicator of severity of brain injury. The duration of this type of amnesia may vary from a few seconds or minutes to several weeks. Over a course of several weeks this amnesia tends to shrink, so that the only permanent memory loss is for the events several seconds to a few minutes prior to the injury.

Concussion

Since the term, concussion, is still frequently used in head trauma parlance and may appear on an applicant's records, a brief discussion is in order. As defined by the Committee to Study Head Injury Nomenclature of the Congress of Neurological Surgeons, cerebral concussion is "a clinical syndrome characterized by immediate and transient impairment of neural function, such as alteration of consciousness, disturbance of vision, equilibrium, etc, due to mechanical forces."[3] One of the hallmarks of cerebral concussion is the presence of variable degrees of retrograde and post-traumatic amnesia with or without loss of consciousness. With this type of injury there is no evidence of structural damage on CT scan. Yarnell and Lynch describe two different amnestic events with mild head injuries. In one, the patients cannot recall events surrounding the incident moments later. They have a sense of bewilderment with cognitive difficulty and only patchy recollection of the experience on recovery one hour later. In the second group, the patients can recall the events surrounding the accident when questioned immediately, but lose this information permanently when questioned later.[4] In addition to deficits in memory recall, Gronwall and Wrightson reported that patients with

cerebral concussion (defined by them as an injury not associated with a neurological deficit and without post-traumatic amnesia lasting 24 hours or less) have difficulty in processing sequential information that may persist for several weeks.[5] This problem was more pronounced in patients with multiple concussions.

The term, concussion, therefore, usually means a mild or moderate brain injury depending on the duration of PTA. Although the neurological examination and CT scan are usually normal, because of the problems a person who suffered a concussion has with processing new information, the FAA should wait three months before certifying the applicant as fit to fly. The high propensity for seizures in the first 12 weeks after "trivial" head trauma also supports the duration of this waiting period. If there is any question of residual deficits, then a neurological evaluation, including a CT scan and psychometric tests, should be obtained. An applicant who has a history of multiple concussions should also have complete neurological evaluation before a disposition is made.

Glasgow coma scale

The scale is used in many centers to assess the severity of injury as well as to establish a baseline for follow-up and is graded as follows:

E	Eye Opening	Grade
	Spontaneous	4
	To sound	3
	To pain	2
	None	1

M	Best Motor Response	
	Follows commands	6
	Localized stimulus	5
	Withdraws	4
	Flexion posturing	3
	Extension posturing	2
	No movement	1
V	Best Verbal Response	
	Oriented	5
	Confused	4
	Words	3
	Sounds	2
	None	1

Coma Score (E+M+V) = 3 to 15

The least severe injury would receive a total score of 15, the most serious 3. In addition to the above observations, vital signs, pupil size and reactivity and eye movements are documented. The coma score within the first 24 hours after injury, for the most part, correlates closely with long-term outcome.[1]

Radiographs

Plain skull radiographs are used to detect linear or depressed skull fractures. The major value of the CT scan during the acute stage is to identify surgically remedial lesions such as epidural, subdural, and intracerebral hematomas. In addition, other structural changes may be seen that may have important implications not only in the assessment of the severity of the injury, but in the determination of flying status after

recovery. These include cerebral or brain stem contusions, cerebral infarctions, cerebral edema, subarachnoid hemorrhage, and intracranial air. The finding of bilateral dense intracerebral lesions on the early CT scans usually indicates that a permanent impairment is likely. For example, bifrontal lesions may herald a persistent personality disorder and bitemporal lesions may herald permanent intellectual and memory deficits.[2]

Severity of injury does not always correlate with CT findings. French and Dublin reported that almost 50% of patients who were in deep coma with abnormal motor activity had a normal scan.[6] In another study of 60 severely injured patients without an intracranial hematoma, the CT scan was normal in 35% of cases.[7] This may be explained by the limited value of the CT scan in detecting white matter lesions. Diffuse white matter axonal injury is a major pathological finding in acute, severe, diffuse closed head trauma, with or without intracranial hemorrhage or hematomas.

Radiographic and pathological studies on persons who survived the acute injury have shown cerebral atrophy with lateral ventricular enlargement, that is, hydrocephalus ex vacuo, attributed to periventricular white matter injury, which can be seen as early as one month after injury.

Magnetic resonance imaging (MRI) is a more sensitive radiographic technique for detecting cerebral white matter disease, as well as small subdural hematomas and subacute, hemorrhagic (isodense) lesions, than is the CT scan. Since MRI has been in use for only a few years, little substantive information correlating the MRI findings in acute and recovery stages with severity of head injury, neurological signs, and neuropyschological deficits is available.[8-10]

The vast majority of persons with head trauma who have CT or MRI abnormalities will have other neurological complications or behavioral problems that may preclude certification. However, from time to time a person with a remote history or recent recovery from a closed head injury, which according to information available would be classified as a moderate or severe, and who may have a radiographic abnormality, but

who otherwise appears normal, may apply for a certificate. In this case, an extensive neurological evaluation should be performed, which should include a neurological consultation, EEG, CT or MRI with comparison with previous studies; neuropsychological studies and a flight simulation test. If no abnormalities are detected or if the CT or MRI scans remain unchanged or are now normal, the candidate may be granted a special issuance certificate.

Complications or sequelae

The presence or absence of the following additional findings are important for estimating prognosis of a head injury:

A. Early signs

1. Generalized and/or partial seizures.
2. Focal neurological deficits such as hemiparesis or aphasia.
3. Cranial nerve dysfunction.
4. Skull fracture, either linear or depressed.
5. Intracranial hemorrhage, either epidural, subdural, or intracerebral.
6. CSF oto- or rhinorrhea.
7. Abnormalities on CT scan other than hemorrhage.
8. Other injuries.

B. Intermediate and late sequelae

1. Post-concussion syndrome.
2. Post-traumatic headaches.
3. Post-traumatic epilepsy
4. Persistent focal cerebral hemisphere or cranial nerve abnormalities.
5. Mental sequelae.
6. Chronic subdural hematoma.

7. Obstructive or communicating hydrocephalus.

8. Meningitis.

9. Traumatic aneurysms or arteriovenous malformations.

Classifications

With the above information, a physician can retrospectively classify the injury into one of the following categories, and also list the complications or sequelae. A decision on flying status may then be made.

1. Mild brain injury

There is transient loss or alteration of consciousness, but no focal neurologic deficit, and fast return of alertness or orientation. Persons who have some evidence of traumatic brain injury and brain dysfunction, but who do not lose consciousness, would have mild injury. Post-traumatic amnesia (PTA) lasts less than one hour.

2. Moderate brain injury

One hour after injury there is still impaired consciousness or disorientation, but the individual can follow some commands. Alternatively, the individual may be alert, but have a focal neurologic deficit. Post-traumatic amnesia lasting one to 24 hours usually occurs.

3. Severe brain injury

After injury these comatose or stuporous individuals are unable to follow any commands; they may use words, but inappropriately. Their motor responses vary from localizing stimuli to posturing or nothing. If they recover, PTA usually lasts one to seven days, but it may last longer.

4. Very severe brain injury

After injury these unresponsive individuals keep their eyes closed even to intense stimuli. They utter no words or sounds and follow no commands. On stimulation, they exhibit either no motor movement or a posturing response. A person in this state often dies, unless a clot is responsible for the clinical presentation and is removed. If the person lives, he or she is frequently in a vegetative state. PTA lasts longer than seven days and often much longer.

The above classification focuses on degrees of unresponsiveness as an index of severity. Perforating or penetrating injuries to the skull and brain may not result in a disturbance of consciousness. However, they may cause neurological deficits, which themselves may be severely impairing.

Sequelae

Impact seizures

Convulsive activity occurring within a few seconds of impact are referred to as impact or instantaneous seizures. Approximately 50% of persons with impact seizures will have repeated attacks during the first hours or days after injury.[11] These persons usually have evidence of structural damage, such as an intracranial hematoma, cerebral contusion or a depressed skull fracture. For those with no structural damage the injury may be considered trivial.

The mechanism of an impact seizure with a mild head injury is unknown; it may be vasovagal. Although studies are limited, there does not appear to be an increased risk for subsequent convulsion.[12] Therefore, an applicant with mild brain trauma, and who otherwise satisfies the criteria for head injury may, be granted a certificate provided a

neurological evaluation and an EEG are normal. A CT scan may be needed, depending on the recommendations of the neurological consultant.

Early seizures

Approximately five percent of persons who sustain a nonmissile head injury will have a generalized and/or focal seizures(s) within the first seven days after a nonmissile head injury; 60% occur within the first 24 hours. Early epilepsy occurs more frequently after serious head injuries, especially those associated with skull fractures, intracerebral hematoma or post-traumatic amnesia that lasts longer than 24 hours. Mild brain injuries, particularly in children under 5 years of age, may be associated with seizures.[13] Oka et al[14] reported that 28 of 37 patients, the majority of whom were under 14 years of age, developed convulsive attacks after trivial brain injuries. Most of these occurred within two hours of the head injury. After early seizures, there is a 17% chance of developing late epilespy in persons under 16 years old and 33% in persons over 16 years.[15]

Although early seizures are usually associated with severe head injuries, and each seizure increases the risk for the development of late epilepsy, in some persons, especially children and adolescents, early seizures may be benign. Eighty percent of individuals who develop late epilespy will do so within 2 years after injury.[16] The new cases after this time occur at decreasing frequencies, so that by 10 years, the risk of having a seizure is probably the same as in the non-injured population. Therefore, provided the applicant with a head injury meets all other criteria, a period of 10 years with no seizures and no anti-convulsant medication must elapse before the applicant should be certified. Before a decision is made, a complete neurological consultation should be done, as well as an EEG, CT or MRI scan, and a blood screen for anti-convulsant medications.

Late epilepsy (Post-traumatic epilepsy - PTE)

The incidence of PTE, defined as seizure activity occurring later than 7 days after the traumatic event, is 5% for a closed head injury not caused by missles. For missile injuries, if the dura is penetrated, the incidence is 42%, and 23% if the dura remains intact. The risk of PTE is also increased with depressed fractures, intracranial hematoma, early epilepsy and with focal cerebral hemisphere damage. Severity of injury as estimated from the duration of post-traumatic amnesia itself does not necessarily increase the risk of PTE. However, in general, the more severe the injury, the greater the likelihood of PTE. If there is a history of a febrile convulsion in childhood or a history of epilepsy in parents, siblings or offspring, the risk for PTE is further increased. The EEG is of little value in predicting PTE.

Approximately 40% to 50% of persons develop PTE within 6 months of the injury, 70% by one year, and 80% by two years. Thereafter, there is a continuing decreasing incidence. In a recent unpublished study on war missile injuries, a group was identified who experienced their first seizure on the average of 7.7 years after the insult.[17] Thus, the onset of PTE can be delayed appreciably. The interval of time that must elapse before the incidence of epilepsy is the same as the general population has yet to be determined. Post-traumatic epilepsy may remit temporarily, and approximately 50% of persons with PTE will stop having seizures altogether. Data on permanent remissions are lacking, although a seizure-free interval of 20 years off all anti-convulsant medications would most likely constitute a permanent remission.

The AME and FAA are confronted with two major problems when evaluating candidates with head injuries: 1) identification of those persons at risk for developing PTE and, 2) disposition of those candidates with PTE who are in apparent remission.

With regard to the first problem:

1. Those applicants with early epilepsy, a depressed skull fracture, intracranial hematoma, or focal cerebral hemisphere deficits as detected from neurological examination or CT or MRI scans should be denied certification. They should be considered for certification only after a 20-year interval has elapsed since the injury, provided they satisfy the other criteria for head injury.

2. Since incidence of PTE after moderate to severe, diffuse closed head injuries without complications is not definitely known, but seem to be higher than the incidence of nontraumatic seizures, it is advisable to restrict applicants with moderate or severe injury, who otherwise satisfy the head injury criteria, for an extended period of time, to be determined by the neurological consultant.

With regard to the second problem:

1. Applicants with a history of PTE may be considered for certification only after 20 years have elapsed since the last seizure, during which time the applicant has been off all anti-convulsant medication.

To assist in resolving both problems, a neurological consultation, EEG, CT or MRI scan must be done before a disposition is made.

Transient neurological signs and symptoms associated with trivial head trauma

The AME and FAA may encounter applicants with a history of transient neurological signs and symptoms following minor head trauma that is usually not associated with amnesia or unconsciousness. This presentation is more common in

children and young adults. Transient confusion, hemiparesis, hemisensory symptoms, as well as visual scotomata, hemianopsias, and total blindness have been reported. These symptoms last usually less than 1/2 hour and are followed by a headache. This syndrome resembles migraine, although spontaneous migraine has been reported in less than 50% of the cases. This syndrome may run in families.[18] The FAA should request a neurological consultation before making a disposition. The consultant may request further studies such as an EEG or CT scan. If the results of the evaluation are normal, provided the applicant does not have disqualifying migraine, he or she may be certified.

Prolonged retrograde amnesia

Retrograde amnesia is a feature of concussion. Usually the amnestic period (the period of time that is not remembered) is of short duration (several minutes to hours), but it may extend for days. However, over a course of days or months the amnestic period gradually shrinks, so that only few seconds or minutes remain lost. In persons in whom a prolonged period of retrograde amnesia remains unchanged, a psychogenic cause should be suspected. This can be determined by a sodium amytal or hypnotic interview. Amnesia resulting from an organic disease cannot be recalled, whereas with amnesia from functional causes, the person can remember events using either one of these two techniques. Therefore, in applicants with a prolonged, fixed period of retrograde amnesia, a psychiatric consultation should be obtained.

Post-traumatic headaches (PTH)

Post-traumatic headaches are a frequent sequelae of head injuries. Brenner et al[19] reported that 69% of 200 consecutive hospitalized persons with head injuries complained of PTH. Thirty percent of the headaches did not persist beyond the hospital stay. Thirty-two percent of the headaches continued beyond 2 months, and 6% of the headaches began after discharge. Headaches are far less frequent after major than after

trivial head trauma. Four distinct types have been defined:[20] 1) steady pressure with cap-like distribution; 2) circumscribed superficial tenderness around impact site; 3) episodic unilateral aching or throbbing pain (migraine-like); and 4) episodic unilateral with ipsilateral mydriasis and facial hyperhydrosis. With this last type of headache, when the pain subsides, partial ptosis and miosis remain. All persons who have this type of headache had anterior neck injuries.[21] Chronic migraine after minor head trauma also has been reported.[20] Dispostion of applicants with the various types of PTH should follow the guidelines listed in the headache section (below).

Post-traumatic or postconcussion syndrome

The post-traumatic syndrome can be a very disabling sequela of minor head injuries. Symptoms of this syndrome include headache, dizziness, fatigue, reduced concentration, memory deficits, irritability, anxiety, insomnia, hyperacusis, photophobia, depression and slowed information processing. In some persons positional nystagmus may be the only abnormal neurological findings. The symptoms may begin within 24 hours after injury or may be delayed for several weeks. Often the symptoms last only a few days to several weeks. However, persistence of the symptoms for months is not uncommon.

In assessing a person with post-traumatic syndrome, the FAA must make sure that a structural abnormality, such as a clot or obstructive hydrocephalus, has not been overlooked. If there is no structural cause for the syndrome, then the decision to certify an applicant depends on the complete resolution of symptoms followed by an appropriate follow-up period of two to three months. Since cognitive deficits and difficulties in processing information may persist, neuropsychological (see section on dementia) studies as well as flight simulator tests may be necessary before a final disposition can be made. For applicants with positional nystagmus, a thorough ENT evaluation is required before a disposition can be made.

Cranial nerve abnormalities

Cranial nerve abnormalities are frequently overlooked as complications of head injury. Before issuing a certificate to applicants with a history of head injury, the FAA must determine the functional integrity of the cranial nerves. A careful history is a prerequisite, since asymptomatic cranial nerve dysfunction in an otherwise neurologically normal individual is exceedingly rare; a routine neurologic examination of the cranial nerves should uncover any deficit. The degree of abnormality can then be determined by quantitiative tests. Whether the cranial nerve abnormality is sufficient to compromise flying performance will have to be determined according to FAA standards and guidelines for the cranial nerve's paticular function.

Cranial Nerve I: Anosmia occurs in about 7% of persons admitted to hospitals with head injury. Most persons with anosmia have frontal fracture and have been unconscious. Approximately 50% of individuals with CSF rhinorrhea from anterior fossa fractures suffer from lack of smell. With surgical repair, the incidence of anosmia increases.

With trivial head trauma the first cranial nerve is the one most frequently damaged, particularly when the impact is to the occipital region. Countercoup damage to the olfactory filaments in the cribiform plate without an associated fracture is postulated as the mechanism.

Prognosis for recovery is particularly poor if post-traumtic amnesia exceeds 24 hours. Recovery usually occurs within the first three months, although some persons will complain of distortion of smell.

Visual pathways - cranial nerve II: The optic nerve can be damaged by both blunt and penetrating head trauma; the usual site of damage in the optic canal. There may be an associated orbital or anterior fossa fracture. Various degrees of visual loss as well as field deficits have been reported. Most often the blindness is complete. Delayed onset

of visual loss may result from increased intracranial pressure or from adhesive arachnoiditis, which results from subarachnoid blood.

Oculomotor nerves - cranial nerves III, IV, VI: Transient symptoms of dysfunction of the motor nerves to the eye are not uncommon immediately after head trauma. However, they usually dissipate quickly. Persistence of diplopia means injury to one or more of these nerves has occurred. The third nerve may be directly injured by stretching or contusion or secondarily by tentorial herniation. Isolated fourth nerve paresis is a rare complication of closed head injuries. The 6th nerve, because it is the longest intracranial nerve, is the most frequently injured oculomotor nerve. The injury may be associated with fractures of the petrous or sphenoid bones or present as a false localizing sign with increased intracranial pressure.

Cranial nerve VII: Injury to the facial nerve is most commonly caused by fractures of the petrous bone. This is often associated to the 8th cranial nerve. Two types of injury have been identified, one with immediate onset of complete paralysis, and the other with delayed onset occurring within several days of the accident. With the latter type, total or partial recovery occurs, usually over a period of 6 to 8 weeks.

Cranial nerve VIII: The 8th cranial nerve is frequently injured in closed head injuries, with or without evidence of the temporal bone fracture. Symptoms and signs of vestibular dysfunction have been reported in approximately 50% of individuals with closed head injuries. Often the symptoms and signs are mild, although many of these persons have other symptoms of the postconcussion syndrome. The auditory component of the 8th cranial nerve is also involved, manifesting as sensorineural deafness. A concussion of the organ of Corti is postulated as the mechanism. Transverse petrous fractures are commonly associated with this type of injury. Conductive type deafness may be seen with longitudinal temporal fractures that cause bleeding in the middle ear.

Cranial nerves IX, X, XI, XII: These cranial nerves are infrequently damaged in closed head injuries. Gunshot wounds of the extracranial structures are the most

frequent causes.

Persons presenting with persistent cranial nerve deficits must be evaluated to assess their ability to function in a cockpit. This is especially true for vision, oculomotor function, hearing, and equilibrium.

CSF rhinorrhea and otorrhea

Approximately 25% of persons with an anterior basal fracture will develop CSF rhinorrhea. This occurs usually within several days after injury and in most cases only persists for several days. Rarely, delayed rhinorrhea can occur several weeks or months after the injury.

CSF otorrhea occurs in 7% of basal skull fractures and usually resolves spontaneously in 5 to 10 days.

The major complication of CSF fistulas is menigitis. It is generally accepted that if the CSF leak persists for more than one to two weeks, then it should be repaired surgically in order to prevent meningeal infection. After an appropriate post-operative recovery period, and provided there are no other complications that would negate return to flying status, a pilot may be certified medically. However, he or she should be re-evaluted every 6 months for two years.

Vascular complications

The vascular complications of head trauma are arterial dissections, aneurysms and fistulas. Arterial dissections frequently involve the extracranial arteries and are caused by blunt or penetrating injuries to the neck. Rarely, dissections of intracranial arteries, particularly the basilar artery, may occur with closed head injuries. The dissections occur at the time of or soon after the injury. The signs and symptoms may be difficult to distinguish from other complications. The effects are usually devastating and if the patient survives, the residual deficits are so severe that there is no question about

certification.

Traumatic aneurysms are rare. They can develop on any artery; however, the cervical and the intracavernous portion of the internal carotid artery are most frequently involved. Most intracranial traumatic aneurysms rupture within three weeks of injury. Therefore, the FAA should not be concerned about this complication after the recovery period.

Seventy-five percent of carotid artery-cavernous sinus fistulas are due to trauma. Of concern to aviation safety is the finding that 20% of persons with this condition present with symptoms later than 2 months after injury, and that the symptoms may occur after even minor trauma. Proptosis, chemosis, visual impairment, ocular palsies and orbital pain are the major clinical findings. Carotid artery-cavernous sinus fistulas may resolve spontaneously. More often they must be repaired surgically. Before being certified with this condition, the applicant must document correction by angiographic studies, and there must be no other complications that would prevent certification. The pilot should be evaluated every 6 months for two year.

Hydrocephalus

Ventricular enlargement develops in 40% to 80% of adults with head injury.[23] Fifty percent of persons with ventricular enlargement were moderately to severely impaired. In contrast, 68% of persons without ventricular enlargement had good recovery.[24] The ventricular enlargement in the greatest majority of these patients was a result of loss of brain substance, particularly white matter (hydrocephalus ex vacuo).

Before certifying applicants in whom ventricular enlargement has developed, as demonstrated by serial CT scans, the FAA must wait at least two years to assure that the ventricular enlargement is not progressive. Certification of these individuals depends on the results of a thorough neurological evaluation including EEG, psychometric tests and perhaps a flight simulator test.

Obstructive hydrocephalus is a rare complication. The progressive ventricular dilatation is caused by adhesions in the subarachnoid space. The symptoms are delayed and usually are manifestations of increased intracranial pressure, such as decreased mentation, lethargy and headaches. These persons must be shunted, and therefore are at great risk for shunt malfunction with sudden increase in intracranial pressure, and therefore are disqualified permanently.

Normal pressure hydrocephalus, or communicating hydrocephalus, develops in approximately one to two percent of persons with head injuries. This condition occurs late, usually after the person has, for the most part, recovered from his injuries. The clinical triad of mental changes, gait apraxia and urinary incontinence is characteristic of this condition.[23] Because shunting procedures are the only treatment, the candidate is disqualified permenantly.

Intracranial hematomas

The incidence of intracranial hematomas in persons with head injuries who are admitted to the hospital is approximately 1.2%, with an annual incidence rate of 4.5 per 100,000 population.[25] Approximately 25% are extradural only, 31% subdural only, 9% both extra and subdural, 23% subdural and intracerebral, and 13% intracerebral only.[26] Ninety percent of the hematomas are supratentorial in location and the remainder are in the posterior fossa. Excluding the posterior fossa hematomas, the one with the best prognosis for moderate or good recovery is the epidural clot followed by the extradural and subdural hematomas. Subdurals have the worst prognosis because of other associated brain injuries. Intracerebral hematomas also carry a poor prognosis for full recovery. Unfortunately, follow-up studies on persons with hematomas have been carried out only for short periods of time, for example, two years. Long-term follow-up would be helpful for aeromedical disposition.

Based on the literature, persons with the following hematomas may be considered

for special issuance certification:

1. Uncomplicated epidural hematoma without evidence of other complications as determined by neurological examination, EEG, serial CT scans or MRI, and neuropsychological examinations. The applicant may be considered two years after injury. The incidence of post-traumatic convulsions with uncomplicated epidurals is low.

2. Because acute subdural hematomas and intracerebral clots have high risk of late seizures, persons with these conditions should not be considered for certification until 20 years has elapsed from the time of injury and all neurological investigations are normal. (See section on epilespy).

3. Persons with posterior fossa sub- or epidural hematomas may be considered for special issuance two years after the injury, provided a complete neurological investigation including a CT scan or MRI, shows no disqualifying abnormalities.

Chronic subdural effusions, hygromas and hematomas

Up to one half of persons with chronic subdural hematomas have no history of trauma. These persons usually are elderly or chronic alcoholics, epileptics, and individuals on anti-coagulation therapy. Chronic subdural hematomas are rare after severe impact injury probably because the acute subdural hematomas are diagnosed early. If an applicant with a history of chronic subdural hematoma presents for certification, he or she may be considered for special issuance if a period of at least five years has elapsed since either medical or surgical treatment, he or she has had no seizures, he or she has no other disqualifying conditions as determined from the results of a neurological and/or neurosurgical consultation, EEG, CT scan and neuropsychological

studies. This recommendation pertains also to subdural hygromas whether they have been treated by surgery or not. Some subdural hygromas may resolve spontaneously.

Applicants with a history of subdural effusions in infancy and childhood, who otherwise are neurologically normal as determined by the aforementioned tests, may be considered for certification. Many of these persons have significant brain damage as well as seizures. However, it has been reported that approximately one half of these persons may develop normally.

References

1. Jennett B, Teasdale G: Management of Head Injuries. Contemporary Neurology series Number 20. Philadelphia, FA Davis, 1981.
2. Walker AE, personal communication.
3. Gurdjian EJ, Voris HC: Report of ad hoc committee to study head injury nomenclature. Clinic Neurosurg 1966;12:386-387.
4. Yarnell PR, Lynch S: The "ding": Amnestic state in football trauma. Neurology 1973;23:196-197.
5. Gronwall D, Wrightson P: Delayed recovery of intellectual function after minor head injury. Lancet 1974;2:605-609.
6. French BN, Dublin AB: The value of computerized tomography in 1000 consecutive head injuries. Surg Neurol 1977;7:171-183.
7. Jennett B, Teasdale G: Management of Head Injuries. Contemporary Neurology Series. Philadelphia, FA Davis, 1981, No 20, pp 111-151.
8. Levin HS, Kalisky Z, Handell SF, et al: Magnetic resonance imaging in relation to the sequelae and rehabilitation of diffuse closed head injury: Preliminary findings, in Goldblatt D (ed): Seminars of Neurology, Head Injury. New York, Thieme-Stratton, 1985, vol 5, pp 211-232.

9. Gandy SE, Snow RB, Zimmerman RT, Deck MDF: Cranial nuclear magnetic resonance imaging in head trauma. Ann Neurol 1984;16:234-257.

10. Hans JS, Kaufman B, Alfidi RJ, et al: Head trauma evaluation by magnetic resonance and computed tomography: A comparison. Radiology 1984;150:70-71.

11. Kollevold T: Immediate and early seizures after head injuries. I. J Oslo City Hospitals 1976;12:99-113.

12. Jennett B: Epilepsy After Non-missle Head Injuries (ed 2). London, Heinemann, 1975.

13. Jennett BW: Early traumatic epilepsy: Incidence and significance after nonmissile head injuries. Arch Neurol 1974;30:394-398.

14. Oka H, Kato M, Matsushimo M, Ando K: Traumatic spreading depression syndrome: Review of a particular type of head injury in 37 patients. Brain 1977;100:287-298.

15. Jennett B, Teasdale G, op cit, 1981, pp 271-288.

16. Jennett B, op cit, 1975.

17. Weiss GH: Predicting post-traumatic epilepsy. Arch Neurol, in press.

18. Bennett DR, Fuenning SI, Sullivan G, Weber J: Migraine precipitated by head trauma in athletes. Am J Sport Medicine 1980;8:202-205.

19. Brenner C, Fedlan RG, Friedman P, et al: Post-traumatic headache. J Neurosurg 1944,1:379.

20. Scherokamn B, Massey W: Post-traumatic headaches, in: Packard RC (ed): Neurologic Clinics, Philadelphia, WB Saunders 1983, vol 1, pp 457-463.

21. Vijayan N: A new post-traumatic headache syndrome: Clinical and therapeutic observations. Headache 1977;17:19-22.

22. Weiss HD, Stern BJ, Goldberg J: Chronic migraine after minor head trauma. Neurology 1985;35(suppl 1):113.

23. Levin HS, Meyer C, Grossman RG, Sarwar M: Ventricular enlargement after closed head injury. Arch Neurol 1981;38:623-629.

24. Gudeman SK, Kishore RS, Becker DP, et al: Computed tomography in the evaluation of incidence and significance of post-traumatic hydrocephalus. Radiology 1981;141:397-402.

25. Jennett B, Murray H, MacMillan R, et al: Head injuries in Scottish hospitals. Lancet 1977;2:696-698.

26. Teasdale G, Galbraith S: Acute traumatic intracranial hematomas, in: Progress in Neurological Surgery 10. Basel Switzerland, Karger, 1980.

Sleep and Arousal Disorders

Introduction

Of all conditions that compromise flight safety, the disorders of sleep and arousal may be the least recognized. Whereas sudden incapacitation may occur with narcolepsy and cataplexy, the insidious and subtle mental and physical changes associated with other sleep and arousal disorders are just as devastating. The diagnsotic problems are very complex, since these disorders bridge the disciplines of psychiatry, neurology, and internal medicine. The causes are multiple, there are no anatomical or biochemical markers, and diagnoses are based on historical information and confirmed only in some cases by physiological tests such as polysomnography. However, regardless of the cause, the ultimate result of these conditions is an impairment of psychomotor performance.

The neurological deficits accompanying structural diseases of the central nervous system that may cause alterations in the sleep-wake cycles, such as trauma, cerebral vascular accidents, infections, and tumors, are by themselves sufficiently impairing to disqualify the candidate. Therefore, only those sleep disorders due to nonstructural diseases and that are most frequently referred to the neurologist will be considered. Sleep disorders associated with psychiatric and medical diseases are discussed in other chapters in this report. There are a number of excellent review articles on sleep and arousal disorders.[1-4]

Criteria for diagnosis

Diagnostic criteria are often difficult to establish. Complaints of insomnia or excessive sleepiness would suffice in some instances. However, in some individuals the problem is transient and situational and requires no further evaluation. In others, the complaint is chronic and more serious. In addition, other symptoms such as personality changes, diminished mental performance and motivation, slowness of thought, sexual

difficulties, or headaches may be a result of sleep disorders; it is only after a careful history of sleep habits and other medical conditions that sleep abnormalities may be implicated as the cause. The FAA should become familiar with the various sleep questionnaires used in sleep centers for evaluation of persons with suspected sleep disorders.

Classification

The diagnostic classification of sleep and arousal disorders presently in use[5] is in the process of revision; no publication date has been set. The following is a list of those disorders that may present as neurological diagnostic problems.

A. Disorders of Initiating and Maintaining Sleep (Insomnias)
   1. Associated with sleep induced respiratory impairment
   2. Associated with sleep related myoclonus and "restless legs"

B. Disorders of Excessive Somnolence
   1. Associated with sleep induced respiratory impairment
   2. Associated with sleep related myoclonus and "restless legs"
   3. Narcolepsy
   4. Idiopathic CNS hypersomnolence

C. Dysfunctions Associated with Sleep, Sleep Stages or Partial Arousals (Parasomnias)
   1. Sleepwalking (somnambulism)
   2. Sleep terrors (pavor nocturnus)
   3. Sleep related enuresis
   4. Other dysfunctions
      a. Sleep related epileptic seizures
      b. Sleep related headaches

Fifty-one percent of persons referred to sleep disorder clinics suffer from hypersomnia, 31% from insomnia and 15 % from parasomnia. Of those with hypersomnia 43% have sleep apnea, 25% narcolepsy, and 9% idiopathic CNS hypersomnia.[6]

Sleep apnea

Persons with sleep apnea may present with insomnia and/or excessive daytime sleepiness. Because of the latter they are frequently misdiagnosed as having narcolepsy. The sleep apnea syndromes have been divided into central, obstructive and mixed.

Insomnia is the major complaint of persons with the central type. They awaken many times during the night with a choking sensation, anxious feelings and gasping for air. They usually do not nap during the day. Persons with obstructive sleep apnea suffer from excessive daytime sleepiness. During sleep, long pauses in respirations are interrupted by loud snoring. On awakening these persons may be disoriented for several minutes. Early morning headches are not uncommon and hallucinations or misperceptions may occur during the day when the person is drowsy. Thus, they may suffer from mental disorders. Also, alcohol ingestion may increase the severity of obstructive sleep apnea.

Unlike persons with central apnea, who have normal or below normal weight, persons with obstructive apneas may be overweight, and have short, fat necks. The sleep apnea syndromes, particularly the obstructive type, may be associated with hypothyroidism, acromegaly, micrognathia, basilar invagination, platybasia, neck tumors, enlarged tonsils and adenoids, and chest deformities. Major complications of sleep apnea are cardiac arrhythmias, which can be life threatening, and hypertension.[7-8]

Applicants with a diagnosis of sleep apnea should be disqualified permanently from flying. A possible exception is when the apnea is secondary to a treatable disease, such as enlarged tonsils or adenoids or hypothyroidism. In these cases, a decision to return to flight status must depend on the recommendations from an accredited sleep disorder

laboratory following a thorough evaluation, including polysomnography.

Sleep related myoclonus and "restless legs"

As defined by Coleman et al, periodic movements in sleep (PMS), also known as nocturnal myoclonus, are stereotyped, repetitive, nonepileptiform movements of one or both lower extremities occurring primarily in non-rapid-eye-movement (NREM) sleep.[9] These should not be confused with sleep jerks (hypnic jerks), which are single and sudden myoclonic jerks that occur on falling asleep and which are normal. Nocturnal myoclonic jerks occur approximately ever 30 seconds and last 1.5 to 2.5 seconds. They may persist for a period of several hours. The contraction consists of extension of the big toe and partial flexion of the ankle, knee and sometimes the hip. Nocturnal myoclonus is associated with insomnia and hypersomnolence. Therefore, it is not a distinct disorder, but only one part of a number of sleep-wake disorders. It may be confused with sleep epilepsy; however, the history and electrophysiologic studies distinguish between these two conditions.

The restless legs syndrome (RLS) may or may not be associated with PMS.[10] Persons with RLS complain of an uncomfortable dysasthetic, creeping sensation in their legs that appears only at rest. For example, at bedtime, in order to obtain relief, they frequently have to move their legs continually or pace around the room. Onset of sleep is delayed and the person suffers from insomnia. RLS has been reported in a number of conditions, such as iron deficiency anemia, folate deficiency, peripheral neuropathies, uremia, and diabetes mellitus. It has been reported as occurring in families.[11] Candidates with a history of RLS are disqualified from flying unless the cause for their problem has been defined and permanently cured and the sleep disorder secondary to the syndrome has resolved. Clonazepam (Clonapen^{R}) has been used to control "the restless legs." Airmen taking on this medicine should not be allowd to fly because of its sedative effects.

Narcolepsy

Narcolepsy is a disorder of excessive daytime somnolence characterized by irresistable sleep attacks, which may occur either as the only symptom, or in combination with the other features, such as cataplexy, sleep paralysis and hypnagogic hallucinations (the "narcoleptic tetrad"). Automatic behavior and lapses of memory have been reported.[12] Some persons complain of blurred or double vision and droopy eye lids. The prevalence rate of narcolepsy is 4:10,000 persons. There is evidence of a genetic transmission via an autosomal recessive trait since relatives of index cases having a 60-fold greater risk of having the condition than the general population.

Narcolepsy usually becomes manifest toward the end of puberty. Eighty percent of all persons with narcolepsy develop their first symptoms by age 35 years. Cataplexy will develop at some point in approximately 85% of cases. Sleep paralysis occurs in 70% and hypnagogic hallucinations in 50% of persons with narcolepsy and cataplexy. The characteristic physiological abnormality is REM-onset sleep, which is found in over 90% of persons with narcolepsy and cataplexy. Without the symptom of cataplexy, only 10% of narcoleptics have REM onset sleep. It is these persons who represent the most difficult diagnostic problem, although, over a period of time, at least one of the other symptoms develops.

Narcolepsy is a chronic, unremitting disease. Applicants with a diagnosis of narcolepsy, even if treated with medications, should be permanently disqualified from flying. A family history of narcolepsy is not by itself disqualifying; however, the airman with a family history should be evaluated on a regular basis for symptoms of this disorder.[13]

Idiopathic CNS hypersomnolence

This disorder is characterized by excessive daytime sleepiness and is often confused with narcolepsy. However, the sleep periods are deeper (stages 3 and 4) and much longer. In addition, the other symptoms of the narcoleptic tetrad are lacking. During the waking state, polygraphic monitoring has shown brief periods of "micro-sleep." When sleep is resisted, automatic behavior may occur. This disorder may be associated with migraine, syncope, and Raynaud's syndrome. There may be a familial predisposition. In contrast to narcolepsy, this conditon does not respond readily to CNS stimulants. It is also chronic and applicants with this condition should be disqualified permanently.

Parasomnias

Somnambulism is usually a disorder of childhood and adolescence. It is reported that 15% of all children experience one or more sleepwalking episodes, and frequent attacks occur in 1% to 6% of children. Although usually limited only to childhood, somnambulism may recur in adulthood, in which case it is associated with psychiatric disorders.[14] Somnambulism is also frequently associated with sleep related enuresis, sleep terrors and sleep drunkeness. In children with this disorder, there is an increased incidence of convulsive disorders, CNS infections and head trauma.[5]

An applicant with a history of these disorders in childhood may be certified provided there are no other associated neurological conditions. If the episodes recur in adulthood, then a psychiatric consult must be obtained before granting certification. Sleep walking may be confused with sleep epilespy, particularly complex partial seizures, because of persevering motor acts, automatisms and amnesia. If there are any questions about other causes for sleep walking episodes, then the applicant should be referred to either a neurologist or an accredited sleep diagnotic laboratory.

Sleep terrors (pavor nocturnus) usually begin in childhood and disappear in early

adolescence. When they begin in adulthood, they usually occur between 20 years to 40 years of age. The terrors occur in the first third of the night when the individual is aroused from deep NREM sleep. Because of a terrifying scream that signals the onset, the panic and confusion that follows as well as autonomic signs such as tachycardia and mydriasis, the episodes may be confused with epileptic seizures. An applicant with confusing symptoms should be referred to a neurologist or an accredited sleep laboratory for evaluation to rule out epilepsy. Persons with sleep terrors persisting or beginning in adulthood should receive psychiatric clearance before being certified.[15]

Sleep related enuresis occurs in approxmately 10% to 15% of children. In most cases, it is idiopathic and resolves spontaneously by puberty. Enuresis may also be secondary to structural disease of the genitourinary tract or nervous system. The onset of sleep enuresis in adulthood should alert the FAA to structural causes, psychiatric disorders, or drugs.

A history of enuresis in childhood is not diqualifying unless it is caused by a neurological disease or is associated with nocturnal epilepsy. With the former, other neurological deficits such as mental retardation or paralysis are often present which, by themselves, would be disqualifying. With the latter, other historical information will suggest the cause.

Sleep enuresis in the adult is a cause for concern. If diseases of the genitourinary system have been ruled out, then the candidate should have a thorough psychiatric and neurological evaluation. Incontinence and/or sleep enuresis may be the initial manifestation of potentially serious neurological diseases.

Other conditions

Approximately 6% of persons with epilepsy will have seizures only in sleep. The seizures usually are infrequent, of the generalized type, and are responsive to anti-convulsant therapy. Approximately 11% to 30% of persons whose initial seizures occur

only during sleep will develop daytime convulsions within the first six years after the initial nighttime seizure.[17,18]

Therefore, persons with a history of seizures occurring only during sleep are disqualified unless the they have been free of seizures and off medication for 20 years and the results of the neurological exam and EEG are normal.

Migraine, and especially cluster headaches, may awaken a person from sleep. This usually occurs during the REM and post-REM sleep periods; therefore, sleep disturbance is most pronounced toward the end of the sleep period. Cluster headaches may awaken a person from sleep on a nightly basis for several weeks. Therefore, cluster headaches may cause excessive daytime somnolence. Certification is dependent on the guidelines established for headaches (see below).

Sleep laboratories

Within the last 10 to 15 years there has been a proliferation of laboratories for the polysomnographic evaluation of individuals with sleep disorders. A list of the accredited centers is available from the Association of Sleep Disorders Centers. There are now 23 accredited centers with multidisciplinary expertise, and 25 more centers hold probationary status.[19] In evaluating the results of sleep laboratory investigations, it is extremely important that the FAA be knowledgable about the competency of the various centers.

References

1. William SRL, Karlan I (eds): Sleep Disorders: Diagnosis and Management. New York, John Wiley and Sons, 1978.

2. Guilleminault C (ed): Sleeping and Waking Disorders. Menlo Park CA, Addison-Wesley, 1982.

3. Guilleminault C, Lugaresi E (eds): Sleep/Wake Disorders: Natural History, Epidemiology and Long Term Evaluation. New York, Raven Press, 1983.

4. Guides to the Evaluation of Permanent Impairment, ed 2. Chicago, American Medical Association, 1984, pp 64, 229-239.

5. Association of Sleep Disorders Centers Sleep Disorders Classification Committee: Diagnostic Classification of Sleep and Arousal Disorders, ed 1, in Sleep 1979;2:1-137.

6. Miller MM: Sleep disorders medicine: How neurological? Arch Neurol 1985;42:601-602.

7. Guillemenault C, Cumminsky J, Dement WC: Sleep apnea syndrome: Recent advances. Adv Intern Med 1980;26:346-372.

8. Guillemenault C: Sleep and breathing, in Guillemenault C (ed): op cit.

9. Coleman RM, Pollak CP, Weitzman ED: Periodic movements in sleep (nocturnal myoclonus): Relation to sleep disorders. Ann Neurol 1980;8:416-421.

10. Coleman RM: Periodic movements in sleep (nocturnal myoclonus and restless legs syndrome, in Guillemenault C (ed): op cit.

11. Montplaisir J, Gadbout MA, Boghen D, et al: Familial restless legs with periodic movements in sleep: Electrophysiologic, biochemical and pharmacologic study. Neurology 1985;35:130-134.

12. Kales A, Cadieux RJ, Soldatos CR, et al: Narcolepsy-cataplexy I: Clinical and electrophysiologic characteristics. Arch Neurol 1982;39:164-168.

13. Kales A, Soldatos CR, Bixler EO, et al: Narcolepsy-cataplexy II: Psychosocial consequences and associated psychopathology. Arch Neurol 1982;39:169-171.

14. Kales A, Soldatos CR, Caldwell AB: Somnambulism: Clinical characteristics and personality patterns. Arch Gen Psychiat 1980;37:1406-1410.

15. Kales JD, Kales A, Soldatas CR, et al: Night terrors: Clinical characteristics and personality patterns. Arch Gen Psychiat 1980;37:1413-1417.

16. Young GB, Blume GT, Wells GA, et al: Differential aspects of sleep epilepsy. Journal Canadian des Sciences Neurologiques 1985;12:317-320.

17. Gibberd FB, Bateson MC: Sleep epilepsy: Its patterns and prognosis. Br Med J 1974;2:403-405.

18. D'Allesandro R, Sintini M, Pazzaglia P, Lugaresi E: Sleep epilepsies: Prognostic features, in Nistice G, DiPerri R, Meinard H (eds): Epilepsy: An Update on Research and Therapy. Progress in Clinical and Biological Research Vol 124. New York, Alan R. Liss, 1983, pp 235-239.

19. Ferris G: Sleep disorders. Neurologic Clinics 1984;2:51-69.

Infectious Diseases of the Central Nervous System

Introduction

Infectious diseases of the central nervous system are caused by such organisms as bacteria, viruses, fungi, rickettsia and parasites. The signs and symptoms may be a result of tissue invasion with destruction of brain tissue, or to an abnormal host immune response, such as post-infectious and postvaccinal encephalomyelitis. The infectious agent may diffusely affect the meninges, the parenchyma of the brain and spinal cord, or both. Focal lesions, such as brain abscesses and subdural empyema, may also occur. The majority of CNS infections are blood-borne; however, spread from contiguous infected structures such as the mastoid, may also occur.

Since most of the infectious agents are capable of producing the same clinical syndrome, from the aeromedical point of view it is not as important to know the etiologic organism as it is the part(s) and extent of CNS involvement and the complications and neurological residuals. Before granting a certificate to applicants with a past history of meningitis, encephalitis or brain abscess, the FAA should obtain all past records, including the history and reports of neurological examinations, the CSF results, EEG, radiographs, and treatments, including any surgical procedures. In some instances it may become clear that the applicant did not have a CNS infection; in others that infection was mild and self-limited, such as aseptic meningitis; while in others the disease was more serious, such as encephalitis, bacterial meningitis or brain abscess. For the serious diseases, a complete neurological re-evaluation is indicated before a decision on flying status can be made.

In this section, the most commonly encountered clinical syndromes, their minimal criteria for diagnosis, their complications, and the specific aeromedical problems they present will be discussed. A thorough review of CNS infectious diseases may be found in the book by Molavi and LeFrock.[1] Infectious diseases of the peripheral nervous system,

and muscles will be discussed in another section.

Minimal criteria for diagnosis

Signs and symptoms of diffuse meningeal irritation include headache, nuchal rigidity, photophobia, and fever. There may also be evidence of diffuse or focal parenchymal disease, including disturbances of consciousness, convulsions, dysphasia, or hemiparesis. These signs should be corroborated by appropriate CSF abnormalities with or without positive cultures, or diagnostic biopsies with or without cultures. For suspected brain abscesses, subdural effusions, or subdural or epidural abscesses, a compatible head CT or MRI scan with or without positive cultures or biopsy is sufficient to confirm the diagnosis.

Acute bacterial meningitis

Acute bacterial meningitis may result during the course of septicemia by hematogenous spread of bacteria to the meninges from distant infected organs, such as the heart or lung, or by extension from infected contiguous skull or spinal structures, such as the orbit, sinuses, mastoid or a dermal sinus tract. Bacteria can also enter the subarachnoid space in penetrating head injuries. Although any pathogenic bacteria can produce meningitis, Neisseria meningitidis, Hemophilus influenzae, and Streptococcus pneumoniae are the most common causative organisms. Bacterial meningitis occurs more frequently in children than in adults. Irrespective of the pathogen, the clinical signs and symptoms are, for the most part, similar. These consist of fever and signs of meningeal irritation. As the disease progresses, signs of cerebral edema and cortical inflammation develop, including irritability, lethargy, confusion ar ' convulsions (which are most common in children). Focal signs, such as hemiparesis and aphasia, are secondary to infarcts caused by thrombosis of inflamed arteries or veins. Basilar inflammations may cause various cranial nerve palsies.

The diagnosis of bacterial meningitis is established by examination and culture of the CSF. Countercurrent immunoelectrophoresis may also be of help. Approximately 20% of individuals with purulent meningitis will have a sterile CSF. These persons usually have received antibiotics for several days prior to the lumbar puncture.[2]

Bacterial meningitis may occur in normal as well as compromised hosts. The causative pathogen may alert the physician to the possibility of an underlying systemic disease, particularly in the adult. For example, meningitis caused by Listeria monocytogenes or H influenzae in the adult may be associated with chronic alcoholism, diabetes, connective tissue diseases, chronic renal diseases, or acquired immunological deficiency states. Anaerobic meningitis may be secondary to chronic otitis media with mastoiditis, chronic sinusitis, head trauma, head and neck neoplasms, and immunosupressed conditions. Staphylococcus aureus meningitis may be secondary to endocarditis.

Approximately 5% to 30% of children who recover from acute bacterial meningitis have neurological complications or persistent residua,[3] which can be categorized as follows:

1. Complications
    a. subdural effusions
    b. hydrocephalus
    c. brain abscess
2. Persistent residua
    a. mental retardation
    b. personality disorders or other psychiatric disorders
    c. seizure disorders
    d. visual, language and motor impairment
    e. cranial nerve palsies

Complications usually occur in infants and children and are discovered when the child fails to respond to appropriate antibiotic therapy. Of possible concern to aviation is hydrocephalus, which may be slowly progressive and which may eventually evolve into the normal pressure hydrocephalus syndrome of dementia, gait apraxia and urinary incontinence. Unfortunately, serial head CT scan studies during both the acute phase and long term follow-up periods are limited; these studies show that progressive ventricular enlargement can occur.[4-6]

Of the peristent residuals, seizure disorders and cranial nerve palsies deserve special attention; the others are usually obvious and do not present serious problems concerning disposition.

Seizures during the acute phase, particularly in children, are not uncommon. In some individuals they may result from cortical damage caused by brain infarction, necrosis, abscess formation, or subdural effusions or empyema. In others, they may result from metabolic abnormalities, such as hyponatremia. Usually it is impossible to implicate one definite cause. Data are not available on the percentage of persons with seizures during the acute phase who persist in having attacks after recovery. There is also no information on the percentage of persons without seizures during the acute illness who later develop post-meningitic epilepsy. Persons who subsequently develop epilepsy are most often the most seriously ill, who have prolonged periods of altered states of consciousness and signs of focal cerebral hemisphere damage, such as hemiparesis. Thus it is appropriate to apply the guidelines established for seizures and head trauma to those persons most seriously ill with an intracranial infection.

Although any cranial nerve may be permanently damaged in acute bacterial meningitis, the 8th cranial nerve is the most vulnerable. Six percent to 30% of children will suffer permanent hearing loss.[7] Therefore, in evaluating applicants with a history of bacterial meningitis, a careful audiological examination must be performed.

Because the optic (cranial nerve 2) as well as the oculomotor nerves (cranial

nerves 3, 4 and 6) might be damaged, a thorough ophthalmologic evaluation is indicated. Other cranial nerve abnormalities, such as peripheral 7th nerve paresis, should be evaluated for the potential to compromise flight safety.

The articles by Feigin et al,[8] Sangster et al,[9] Davey et al,[10] Cherubin et al,[11] and Geisler et al[12] are excellent reviews of acute bacterial meningitis.

Aeromedical Disposition

An applicant with a history of definite or presumed bacterial meningitis, who has no history of seizures, may be considered for licensure two years after recovery, at which time the medical and neurological examinations should be normal and the audiologic and ophthalmologic examination should meet minimum standards. For some persons neuropsychological tests and a head CT scan or MRI may be indicated. If there is a history of prolonged unresponsiveness during the acute illness, or signs of focal neurological deficit, the applicant should wait 10 years from the acute event. (See section on post-traumatic epilepsy.)

For persons with a history of definite or presumed bacterial meningitis who also had seizures during the acute episode, the guidelines for early post-traumatic epilepsy should be followed.

Persons with post-meningitic seizures only, must be seizure free and off anti-convulsants for 20 years. The guidelines in the section on epilepsy should be followed.

Subacute or chronic infectious meningitis

This form of meningitis is usually caused by the tubercle bacilli or mycotic organisms. Other associated diseases are leptospirosis, sarcoidosis, syphilis, brucellosis, nocardia, and toxoplasmosis. Immunosuppressed persons and those with chronic diseases such as diabetes are at risk for this form of meningitis. The neurological complications and residua are similar to those of acute bacterial meningitis; however, they are more

common, since subacute or chronic meningitis also frequently involves the brain parenchyma. Communicating hydrocephalus may be more frequent.[13]

The guidelines for certification are the same as for acute bacterial meningitis. Because of the association of this type of meningitis with chronic diseases, a thorough medical evaluation is also required.

Recurrent meningitis

Recurrent episodes of infectious meningitis are most frequently the result of seeding of the meninges from parameningeal inflammatory foci. Congenital dermal defects, osteomyelitis of the skull or spine, cerebrospinal fluid fistulas, and chronic sinusitis and otitis media are among the most common causes. Immunodeficiency conditions and previously inadequate treatment for acute, subacute and chronic meningitis may also cause recurrence. Mollaret's meningitis, a rare, recurrent and poorly understood cause of benign aseptic meningitis may also be responsible.

Befor applicants with a history of recurrent episodes of infectious meningitis may be certified, the cause of the condition must be determined and corrected. In addition, the other medical and neurological criteria for infectious meningitis must be satisifed.

Aseptic meningeal reactions

Signs and symptoms of meningeal inflammation with sterile CSF cultures may be secondary to parameningeal infectious foci, seeding of the subarachnoid space from primary or metastatic tumors, connective tissue disorders, or from the introduction of foreign substances, such as dyes, drugs or blood, into the CSF. With parameningeal foci and primary tumors the attacks may be recurrent. Aeromedical certification depends on first establishing the cause and then applying the guidelines for that particular entity.

Aseptic meningitis

Aseptic meningitis is a nonpyogenic inflammation of the meninges, usually a result of enteroviruses, although it has been reported with mumps, lymphocytic choriomeningitis, Epstein-Barr virus and arboviruses. Of the 3000 to 4000 cases reported annually, approximately only 25% are etiologically defined. The major symptoms and signs are fever, headache, malaise, nuchal rigidity, and photophobia. The disease is self-limited and the person with the disease usually recovers within several weeks. A pilot may return to flight status two to three months after the symptoms have subsided as long as the physical examination is normal. A repeat lumbar puncture is not indicated at that time. If symptoms persist after one to two weeks, and/or if there are signs or symptoms of parenchymal involvement, other causes should be considered. The examining physician must be aware of involvement of other organs, such as the heart or liver, in the disease process.

Acute viral encephalitis

Of the approximately 2000 cases of viral encephalitis reported each year, a specific agent is identified in only 30% to 60%. Herpes simplex encephalitis is the most common nonseasonal, sporadic form reported in the United States. Epidemics are usually caused by the arthropod-borne togaviruses. A list of the major encephalitic viruses is found in Table I.

The viral encephalitides may occur in otherwise normal individuals as well as in persons with immunodeficiency conditions (see section on AIDS). There are no distinguishing clinical or cerebrospinal fluid abnormalities that permit an etiological diagnosis; this depends on positive serological tests or cultures. Alterations of consciousness, convulsions, myoclonus, focal neurological deficits, aphasia, fever, nausea and vomiting, and nuchal rigidity characterize most viral encephalitides. In herpes simplex encephalitis the signs and symptoms can mimick an acute temporal lobe mass

TABLE I MAJOR ENCEPHALITIS VIRUSES

Virus	Virus Group	Distribution	Approx Number Cases Yearly
Herpes simplex type 1	Herpes	Worldwide	100-1000
Epstein-Barr	Herpes	Worldwide	10-100
Cytomegalovirus	Herpes	Worldwide	?
Varicella-zoster	Herpes	Worldwide	50-200
Eastern equine encephalitis	Toga(Alpha)	N & S America	0-12
Western equine encephalitis	Toga(Alpha)	N & S America	3-50
St. Louis encephalitis	Toga(Flavi)	N & S America	20-2500
California (LaCrosse) encephalitis	Bunya	United States	50-200
Venezuelan equine encephalitis	Toga(Alpha)	N & S America	0-5
Japanese encephalitis	Toga(Flavi)	Asia	0
Murray Valley encephalitis	Toga(Flavi)	Australia, New Guinea	0
Rabies	Rhabdo	Worldwide*	0-1
Mumps	Paramyxo	Worldwide	5-50
Measles	Paramyxo	Worldwide	0-20
Polio	Picorna	Worldwide	0-20
Coxsackie	Picorna	Worldwide	0-20
Echo	Picorna	Worldwide	0-20

*Excluding Great Britain

source: REF 14

because of the predilection of the herpes type 1 virus to invade rhinencephalic structures. Herpes encephalitis is one of the few encephalitides in which red blood cells are found in the CSF.

Although the data vary somewhat from virus to virus, permanent neurological sequelae occur in approximately 25% to 50% of persons with viral encephalitis. These abnormalities include seizures, behavioral changes, mental retardation, hemiparesis and coordination defects. In addition, MRI may show temporal lobe shrinkage. Complete neurological recovery after herpes type 1 encephalitis is so unusal that this does not present a problem for medical certification at this time.

The major aeromedical problem in granting a medical certificate to an applicant with a definite or presumed history of an acute viral encephalitis is the question of postencephalitic seizure disorder. Seizures occur in approximately 25% of persons during the acute phase. However, data are not available on what is recurrence or occurrence of late seizure(s) in persons who did not experience them during the acute disease. It is known that in 27% of persons with seizures between the ages of one month and 21 years, infection is the etiology. Therefore, the guidelines established for acute bacterial meningitis are also applicable to acute viral encephalitis.

Chronic viral encephalitis

The chronic viral encephalitides, or slow virus infections, such as subacute sclerosing panencephalitis, progressive rubella panencephalitis, and progressive multifocal leukoencephalopathy are so neurologically devastating that they do not present a problem for aeromedical certification.

Parainfectious encephalitis (vasculomyelinopathy)

This entity is thought to be caused by a hypersensitivity reaction to an acute viral illness or vaccination. It is estimated that as many as 20% of encephalitis may be of this

type. The symptoms begin four to 14 days after an infectious illness or vaccination, are similar to those fo acute viral encephalits and are related to the portion of the nervous system most severely involved.

Parainfectious encephalitis has been subdivided into disseminated encephalomyelitis, acute hemorrhagic leukoencephalitis, transverse myelitis, and acute cerebellar ataxia. These syndromes are most frequently associated with the common viral exanthems, mycoplasma, bacterial infections, infectious mononucleosis, mumps, influenza, leptospirosis, upper respiratory and other obscure febrile illnesses, and vaccinations.

With the exception of measles, the neurologic sequelae of these syndromes are less frequent and severe than those of the acute viral encephalitidies. Applicants with a history of a parainfectious syndrome require a complete neurological evaluation before being granted a certificate. Those applicants with acute cerebellar ataxia or transverse myelitis may be certified after recovery, provided they satisfy all other neurological criteria. Because of the high incidence of seizures with disseminated encephalomyelitis (37%) and acute hemorrhagic leukoencephalitis in the acute phase, the guidelines listed for the acute viral encephalitides should apply to applicants with history of these disorders. Those applicants whose disease was related to immunizations should not receive further immunizations.

Postinfectious encephalopathy (Reye's syndrome)

This is a disease of children characterized by encephalopathy and fatty infiltration of the viscera, particularly the liver. It most frequently follows influenza and varicella infections, although it can follow a host of other viral diseases. The majority of the survivors are neurologically intact. Although convulsions may occur in the acute episode, there are no reports of recurrence. An applicant with a history of Reye's syndrome may be issued a medical certificate if found to have no neurologic deficits. Reviews of viral

encephalitides may be found in publications by Johnson, Weiner and Fleming, Kennard and Swash, and Evans.[15-18]

Brain abscess

Brain abscess may result from a hematogenous spread of infection in otherwise normal individuals, or in individuals with cyanotic congenital heart disease, or in persons with pulmonary diseases, such as lung abscesses or bronchiectasis. Other sources for hematogenous spread are septic abortions, tooth and facial infections, and chronic foci such as osteomyelitis. Subacute bacterial endocarditis is a rare cause. Abscesses may also result from spread from infected contiguous structures, such as the mastoid, and from penetrating brain injuries or following neurosurgical procedures. Ten percent of abscesses are multiple, and 80% to to 90% are supratentorial. Most of the supratentorial abscesses resulting from hematogenous spread are located within the distribution of the middle cerebral arteries. Posterior fossa abscesses are found most frequently in the cerebellum, usually as a direct extension from the mastoids. Rarely are they found in the brain stem.

The signs and symptoms are related to location. In some cases they may also be related to increased intracranial pressure due to a mass, cerebral edema or hydrocephalus. Generalized or focal seizures are not uncommon with supratentorial lesions. The diagnosis of abscess is suggested by a head CT scan and confirmed by neurosurgical intervention. The most common organisms isolated are streptococcus; S aureus; gram-negative anaerobic bacilli, particularly Bacteriodes fragilis; and gram-negative aerobic bacilli. Multiple organisms are not uncommon. Approximately 30% of cultures are sterile. Abscesses may also be caused by Nocardia and various fungi, as well as by parasitic infection. Opportunistic infections such as toxoplasmosis, cryptococcus, Candida albicans and bacteria of low virulence should alert the physician to an immunodeficiency state. Therapy is usually drainage or excision with appropriate

antibiotic treatment. There have been recent reports of success with only medical treatments.[19,20]

The neurological residual in persons treated either medically or surgically are focal deficits from cerebral, cerebellar or brain stem damage; seizures; and mental and personality changes.

Additional information may be found in articles by Yang,[21] Harrison[22] and Kaplan.[23]

Guidelines for aeromedical disposition are the same as for meningitis and encephalitis when the abscess is located in the cerebral hemispheres. A history of multiple abscesses is permanently disqualifying. For persons with cerebellar abscesses, return to flight status may be considered only if the cause for the abscess, such as mastoiditis, has been cured, there are no disqualifying neurological residuals that would compromise flight safety, and hydrocephalus is not present. Return to flight status after a two year wait seems appropriate. A complete medical and neurological evaluation is necessary not only to detect residuals, but also to find the source of infection. There is an increasing incidence of brain abscesses in IV drug abusers.

A collection of pus in the subdural space (subdural empyema) is a rare condition usually resulting from trauma, septicemia, or extension of an infection from the sinuses. An aerobic streptococcus is the most frequent pathogen. The course is usually acute and fulminating. Even with early recognition and treatment, neurological residuals are common. The guidelines for menigitis are applicable to this condition.

Spinal epidural abscesses result from direct spread from vertebral osteomyelitis or other contiguous structures or from hematogenous spread, particularly from furuncles or other septic foci such as the teeth and tonsils. In many cases, the cause cannot be determined. Depending on the organism, the course may be acute or subacute. Fever, pain and localized spinal tenderness, and signs of spinal cord compression are typical features. Persons with chronic debilitating illnesses and heroin addicts are predisposed

to this condition. Treatment consists of drainage and appropriate antibiotic therapy. S aureus is the most frequent pathogen.

Aeromedical disposition depends on identification and successful treatment of the source and the abscess, and the absence of spinal cord dysfunction that would compromise flight safety. If all of the above are satisfied, an applicant may be certified following a complete neurological evaluation, performed one year after recovery.

Venous sinus thromboses are exceedingly rare, and have such devastating neurological residuals that they should not present any disposition problems.

Acquired immunodeficiency syndrome (AIDS)

Approximatey 40% of individuals with AIDS have neurological complications. One third of these persons present because of the neurological abnormality. The most frequent CNS complications are infectious in etiology. Neoplasms, vascular diseases and peripheral nervous system disease are not as common. Of the viral syndromes, the most frequent is a subacute encephalitis, followed by a typical aseptic menigitis and herpes simplex encephalopathy. Of the non-viral infections, Toxoplasma gondii and Cryptococcus neoformans are the major offenders.

The finding of opportunistic organisms in persons with CNS infections should alert the physician to the possibility of AIDS. To date, this is a fatal disease. The candidate with AIDS is permanently disqualified from flying (see also hematology section).[23,24]

Neurosyphilis

Neurosyphilis today is a rare disease; it is estimated that two new symptomatic cases per 100,000 persons will occur each year. This disease should always be considered in the differenital diagnosis of meningitis, meningovasculitis, dementia, and spinal cord diseases. An applicant with a diagnosis of vascular syphilis, general paresis or tabes dorsalis is disqualified from flying, irrespective of arrest of the disease with appropriate

antibiotic therapy. The neurological deficits at the time of diagnosis and the initiation of treatment are such that they would jeopardize flight safety. The only possible exception is acute meningitis that may occur with the rash of secondary syphilis. If the disease is arrested by appropriate therapy, and there are no other disqualifying neurological sequelae or complications, then the applicant may be considered for certification two years after recovery. This decision should be based on a complete neurological evaluation including CSF analysis. The airman should then have yearly evaluations.

Perhaps the most difficult diagnostic problem is the evaluation of a positive non-treponema serologic test, such as the VDRL, in an otherwise asymptomatic individual. Transient false positive tests may occur with acute infections or pregnancy. Persistent false positive tests occur in persons with autoimmune diseases such as lupus erythematosus and other collagen vascular disorders. Approximately 55% "false" positive VDRLs are of unknown cause.

The FTA-ABS test is a specific test for detecting specific antibodies to Treponema pallidum. The test is more specific and sensitive than the VDRL, although false positive reactions have been reported. It has also been documented that if the blood FTA-ABS reaction is negative, the CSF reaction will be the same, thereby excluding central nervous system syphilis.

Therefore, if the blood VDRL is positive and the FTA-ABS is negative, a CSF examination is not required. If both are positive and a cause of the false positive reactions is not determined, a CSF analysis must be performed. If the CSF FTA-ABS is positive, irrespective of other changes in the CSF, such as pleocytosis or elevation of the protein, it must be assumed that the CNS has been invaded by T pallidum and the person must be treated accordingly. Provided that the neurological evaluation, including neuropsychological studies, is normal, an applicant with these laboratory findings and treatment history may be certified. A CT scan may be indicated, particularly if other

abnormalities are present. The applicant should be evaluated annually thereafter until a neurology consultant determines that the disease is inactive.[25]

Progressive multifocal leukoencephalopathy (PML)

Progressive multifocal leukoencephalopathy is a subacute demyelinating disease caused by the ubiquitous papovirus. Symptoms are insidius at onset, with progressive paralysis, mental deterioration, visual disturbances, and sensory abnormalities. In some persons, brain stem and cerebellar signs are present. CT scan of the head may show multiple radiolucent defects in the white matter of the brain. Approximately 50% of the persons have lymphproliferative diseases. PML has also been associated with chronic disease states such as sarcoidosis, tuberculosis, acquired or drug-induced immunosuppression, and malignancies. Brain biopsy is the only definitive diagnostic test. There is no treatment and the patients usually die within three to 6 months after onset, although remissions have been reported. Therefore, diagnosis of PML is disqualifying.

References

1. Molavi A, LeFrock JL (eds): Infections of the central nervous system. Medical Clinics of North America. Philadelphia, WB Saunders, 1985, Vol 69, No. 2.
2. Geisler PJ, Nelson KE, Levin S: Community acquired purulent meningitis of unknown etiology. Arch Neurol 1981;28:749-753.
3. Shohet I, Davidson S, Shahar ML, et al: Bacterial meningitis: Follow-up study of 115 children. Isr J Med Sci 1982;18:779-784.
4. Cockrill HH, Dreisbach J, Lowe B, Tamauchi T: Computed tomography in leptomeningeal infections. AJR 1978;130:511-515.
5. Snyder RD, Stouring J: The follow-up CT scan in childhood meningitis. Neuroradiology 1978;16:22-23.

6. Ahmadi J, Segall HD, Zea CS, Haa JS: Computed tomography in childhood intracranial infections, in Bell WE (ed): Seminars of Neurology: Central Nervous System Infections in Childhood. New York Thieme Stratton Inc, Vol 2, No 4, 1982.

7. Borkowski WJ, Goldgar D, Gorga M, et al: Cerebrospinal fluid parameters and auditory brain stem responses following meningitis. Pediatric Neurology 1985;1:134-139.

8. Feigin RD, Stechenberg BW, Chang MJ: Prospective evaluation of treatment of hemophilus influenzae meningitis. J Pediat 1976;88:542-548.

9. Sangster G, Murdoch J-MC, Gray JA. Bacterial meningitis 1949-1979. J Inf Diseases 1982;5:245-255.

10. Davey PG, Cruikshank JK, McManus IC, et al: Acute meningitis: Ten years experience. J Hyg (London) 1982;88:383-401.

11. Cherubin CE, Marr JS, Sierra MF, Becker S: Listeria and gram-negative bacillary meningitis in New York City 1972-1979: Frequent causes of meningitis in adults. Am J Med 1981;71:199-209.

12. Geisler PJ, Nelson KE, Levin S: Community-acquired purulent meningitis: A review of 1316 cases during the antibiotic era 1954-1976. Rev Inf Diseases 1980;2:725-745.

13. Ellner JJ, Bennett JE: Chronic meningitis. Medicine 1976;55:341-369.

14. Hirsch MS, Cutler RWP: Acute viral central nervous system diseases, in Rubenstein E, Federman DD (eds): Scientific American Medicine. New York, Scientific America, 19, Vol XXVII.

15. Johnson RT: Viral Infections of the Nervous System. New York, Raven Press, 1982.

16. Weiner LP, Fleming JO: Viral infections of the nervous system. Neurosurgery 1984;61:207-224.

17. Kennard C, Swash M: Acute viral encephalitis: Its diagnosis and outcome. Brain 1981;104:129-148.

18. Evans OB: Parainfectious neurological diseases, in Seminars in Neurology: Neurological Manifestations of Systemic Diseases. New York, Thieme-Stratton, Inc, Vol 5, No 4, 1985.

19. Berg B, Franklin G, Cuneo R, et al: Nonsurgical cure of brain abscess: Early diagnosis and follow-up with computerized tomography. Ann Neurol 1978;3:474-481.

20. Rosenblum ML, Hoff JR, Norman D, et al: Nonoperative treatment of brain abscesses in selected high risk patients. J Neurosurgery 1980;52:217-225.

21. Yang S-Y: Brain abscess: A review of 400 cases. J Neurosurgery 1981;55:794-799.

22. Harrison JJG: The clinical presentation of intracranial abscesses. Quarterly J Med 1982;204:461-468.

23. Kaplan K: Brain abscess. Med Clinic of North America 1985;69:345-360.

24. Levy RM, Bredesen DE, Rosenblum ML: Neurological manifestations of the acquired immunodeficiency syndrome (AIDS): Experience at UCSF and review of the literature. J Neurosurgery 1985;62:475-495.

25. Snider WD, Simpson DM, Nielsen S, et al: Neurological complications of acquired immunodeficiency syndrome: Analysis of 50 patients. Ann Neurol 1983;14:403-418.

26. Simon RP: Neurosyphilis. Arch Neurol 1985;42:606-613.

Demyelinating Diseases

Multiple Sclerosis

Introduction

Multiple sclerosis (MS), a primary demyelinating disease of the central nervous system, is one of the more frequent causes of neurological impairment in the adult. The overall prevalence rate in the United States is approximately 58/100,000 with the highest rates reported in the northern tier states. Higher rates are reported in first degree relatives of persons with MS; however there is no conclusive evidence that MS is a genetically determined disease. Females are slightly more affected than males. The cause is unknown, although epidemiologic data suggest that it may result from an altered cell-mediated immune response to an environmental cause, perhaps a virus, acquired before the age of 15 years. The average age of onset of MS is 33 years, and symptoms only rarely first appear before the age of 15 years or after 50 years. A mean delay in diagnosis from 1 year for acute-type MS to 10 to 12 years for the other types has been reported.[1]

Multiple sclerosis is the third most common cause (after trauma and rheumatologic disorders) of severe impairment between the ages of 15 years and 60 years. With improved methods of treating complications, the average survival has increased from 25 years to 35 years in the last several decades. Life expectancy of persons with MS remains only slightly less than for the normal poplation.[2,3]

Clinical presentation

The separation of signs and symptoms in time and location is the hallmark of the clinical presentation of MS. In Table I the initial symptoms, course and clinical category in 461 patients are tabulated.[4]

TABLE I

INITIAL SYMPTOMS, CLINICAL COURSE, AND PREDOMINANT CLINICAL CATEGORY IN 461 MS PATIENTS*

	Frequency					
	Women (n = 279)		Men (n=182)		TOTAL (n=461)	
	No.	%	No.	%	No.	%
Symptom						
Visual loss in one eye	54	18	24	13	78	17
Double vision	27	10	35	19	62	13
Disturbance of balance and gait	38	14	45	25	83	18
Sensory disturbance in limbs	72	26	79	43	151	33
Sensory disturbance in face	10	4	6	3	16	3
Acute myelitis syndrome	20	7	6	3	26	6
Lhermitte's symptom	7	3	6	3	13	3
Pain	5	2	3	2	8	2
Progressive weakness	27	9	18	8	45	10
Clinical course						
Relapsing and remitting	164	59	93	51	257	56
Chronic progressive	67	24	60	33	127	28
Combined	66	24	29	16	95	21
Benign	39	14	16	9	55	12
Predominant clinical category						
Spinal	128	46	134	74	262	57
Cerebellar	23	8	35	19	58	13
Cerebral	11	4	7	4	18	4

*These patients satisfied the criteria for clinically definite MS of Schumacher and colleagues and were seen at the MS clinic, University of Western Ontario, London between 1972 &

source: REF 4

Persons with MS usually complain initially of impairment of vision or other ocular disturbances, difficulties with gait and coordination, loss of sphincter control, fatigue, dysarthria, and vague sensory symptoms. Specific signs or symptoms that strongly suggest the diagnosis of MS in the young adult are acute optic neuritis, internuclear ophthalomoplegia, Lhermitte's sign, trigeminal neuralgia, and acute transverse myelitis. Single or multple symptoms may be associated with an attack. The symptoms and signs associated with acute attacks usually progress over a period of 24 hours to several days and resolve within 4 to 8 weeks, generally without residua after the first episode. It is rare for the impairments to reach their maximums during the first acute episode.

Paroxysmal events of short duration, usually lasting less than 5 minutes, may also occur in MS patients. These include brief periods of amaurosis, vertigo, dysarthria, ataxia, diplopia, and sensory disturbances. Two to five percent of persons with MS have generalized, usually tonic seizures. Symptoms can be exacerbated with an increase of body or ambient temperature, infectious diseases, fatigue and stress.

Diagnosis and classification

The clinical diagnosis is based primarily on signs and symptoms, and is categorized into definite, probably and possible multiple sclerosis.[5] Recent technological advancements, including evoked potentials, CSF immunological assays, and radiographic techniques such as CT scans and MRI scans of the head improve, diagnostic accuracy, and a new classification has been proposed. Although it has been developed as a guideline for research protocols, this diagnostic classification (Table II) will probably be used in clinical practice in the future.

TABLE II
CLASSIFICATION OF MULTIPLE SCLEROSIS

A. Clinically definite MS (CDMS)
   1. Two attacks and clinical evidence of two separate lesions.
   2. Two attacks; clinical evidence of one lesion and paraclinical evidence of another, separate lesion.

B. Laboratory-supported definite MS (LSDMS)
   1. Two attacks; either clinical or paraclinical evidence of one lesion; and CSF OB/IgG (Oligoclonal Bands/Immune globulin G).
   2. One attack; clinical evidence of two separate lesions; and CSF OB/IgG.
   3. One attack; clinical evidence of one lesion and paraclinical evidence of another, separate lesion; and CSF OB/IgG.

C. Clincially probable MS (CPMS)
   1. Two attacks and clinical evidence of one lesion.
   2. One attack and clinical evidence of two separate lesions.
   3. One attack; clinical evidence of one lesion and paraclinical evidence of another, separate lesion.

D. Laboratory-supported probable MS (LSPMS)
   1. Two attacks and CSF OB/IgG

source: Ref 6, as modified from Ref 7

In this classification "paraclinical evidence" refers to the results of tests such as CSF abnormalities, evoked potentials and neuroimaging, which demonstate a lesion(s) in the CNS that may not have produced signs of recent dysfunction, but that may have been responsible for symptoms in the past. There is no one laboratory marker that is specific for MS. In addition to lacking high specificity, the paraclinical tests also have varying degrees of sensitivity, particularly in the early stages of the disease. The hot bath test has been used in the past to bring out clinical signs; however, with more sophisiticated tests now available, it is seldom used today.

In Table III, the probability of finding CSF abnormalities and evoked potentials are shown.

TABLE III
CLINICAL CLASSIFICATION OF MS

CSF Abnormality	Definite	Probable-Possible
Globulin Fraction	60-75%	45-55%
IgG Index	80-90%	56-60%
Oligoclonal Proteins	85-90%	55-70%
Evoked Potentials		
Visual	80-90%	35-48%
Somatosensory	70-85%	50-60%
Brain Stem Auditory	50-65%	15-20%

source: Ref 8

The oligoclonal bands are the most sensitive CSF test, while the visual is the most sensitive evoked potential test. Evoked potentials may be abnormal in persons with MS who have had no clinical signs or symptoms related to the function being evaluated. For example, approximately 30% to 50% of persons with suspected MS who have no history of optic neuritis and who have normal ophthalmologic examination will have abnormal visual evoked potentials.

Neuroimaging techniques are helpful ancillary diagnostic techniques. However, cost has limited their routine use. Periventricular white matter hypodense lesions, often multiple, have been reported in 40% to 60% of CT scans of persons with definite multiple sclerosis.[8] These lesions are often clinically silent, and are usually seen in patients with severely impairing MS. Contrast-enhanced lesions of the white matter may occasionally be seen in patients with acute MS. The CT scan abnormalities are not specific for MS, and the sensitivity is not as great as with the other studies. In early, mildly impairing MS, the CT scan of the head is usually normal.

Recent studies have shown that the MRI scan of the head is much more sensitive than the CT scan for detecting MS lesions, especially in the early stages of the disease. Some investigators feel that MRI may prove to be the single most sensitive test, more so than CSF analysis and evoked potentials; data to support this are too early to be

conclusive.[9-10] For example, Tramo et al recently reported that evoked potentials are more sensitive than MRI in detecting subclinical MS lesions.[11]

The studies to date, although not in total agreement, have shown that the lesions seen by MRI are most often multiple in the white matter of the cerebral hemispheres, and can also be seen in the brain stem and/or cerebellum. The MRI demonstrates more lesions than the CT scan, and often the observed lesions do not correlate with the person's signs and symptoms, or with the severity of the disease. To date, the MRI has not be of particular value in detecting abnormalities in the optic nerves or spinal cord. In addition, it is not possible to determine if the lesions represent active demyelination or old sclerotic plaques.

In summary, paraclinical tests have contributed significantly to the capabilities in of diagnosing MS. However, because they lack high specificity and sensitivity, clinical assessment will continue to be the most important diagnostic technique.

Clinical course

Because of the marked variability in the manner in which MS expresses itself, it is impossible to predict the time or frequency of attacks, and when the disease has been arrested. In Table IV, the disease has been subdivided into four categories according to course.[12]

In the early stage of the disease, the attack rate is approximately 0.5 episodes/year. This gradually drops to 0.25/year in the intermediate years. In 5% of persons with MS there is latent interval of several years duration between the first and second attack. Acute attacks eventually become quite rare or cease; however, the disease enters a slowly progressive phase. With each successive attack, the degree of neurological impairment increases. Generally, the course of MS is established during the first several years of the disease. A person with frequent exacerbations in this period is likely to incur severely impairing neurological deficits, while the person with infrequent

TABLE IV

DISEASE COURSE SEEN IN MULTIPLE SCLEROSIS

Type of Course	Characteristics	Approximate Frequency (%)
Benign	Sudden onset. One or two mild exacerbations with complete or nearly complete remission. No permanent functional disability.	20
Exacerbating-Remitting	Sudden Onset. Symptoms remit partially or totally after exacerbations. Long periods of stability; months, even years.	20-30
Remitting-Progressive	Same as exacerbating/remitting in the beginning. At some time during the course, symptoms no longer remit and disability slowly increases.	40
Progressive	Insidious onset. Steady progression of symptoms.	10-20

source: REF 12

attacks may have only minimal or no impairment. An examining physician must determine if an attack is a new episode or represents a pseudoexacerbation, that is, a recurrence of previous symptoms caused by metabolic or physiological influences, such as increase in body or ambient temperature or stress, on a previously demyelinated area.

Several features suggest a favorable prognosis, although it is impossible to predict with any great degree of accuracy the ultimate course in any one person. These are: 1) onset of disease before age 35 years, 2) acute onset, particularly with only one symptom; 3) symptoms that are predominantly sensory, including optic neuritis; and 4) low score on the "disability status scale," which is based on functional deficits determined by physical examination as they relate to the activities of daily living.[13]

Features that point to an unfavorable prognosis are: 1) onset after age 35 years; 2) more than one symptom with each attack; 3) early onset of motor signs, including weakness and cerebellar abnormalities, especially impaired gait and spasticity; 4) pyramidal or cerebellar signs present within 5 years of onset of disease; 5) and male sex.

Optic neuritis is the first symptom in 16% to 30% of persons with MS. It is usually unilateral. The risk of developing definite MS after bilateral optic neuritis in childhood is low; in adulthood, the risk in uncertain. Optic neuritis has been reported to occur at some time in the course of the disease in approximately 30% to 70% of persons with MS; approximately 25% of these persons will experience a second attack. In 90% of persons with optic neuritis, recovery is complete. The interval between the first episode of optic neuritis and the onset of other signs and symptoms of MS may be as long as 35 years. However, for 60% of persons the interval is 8 years or less. Approximately 15% of persons who experience acute, idiopathic optic neuritis have been found to have other causes, such as Leber optic atrophy and optic nerve ischemia.

Studies evaluating paraclinical tests of persons who have experienced a single episode of optic neuritis are few. Oligoclonal bands have been reported in approximately 25% of these individuals and indicate an increased tendency for dissemination of the

disease. Ten percent will show abnormalities in somatosensory evoked potentials. MRI studies on this condition are in progress.

Therefore, individuals who experience acute episodes of optic neuritis whose cause is not determined, should be considered as having MS.

Additional information on optic neuritis and MS may be found in articles by McDonald,[14] Compston et al,[15] Ebers,[16] and Kurtzke.[17]

Aeromedical disposition

The FAA should obtain all medical records and laboratory studies of applicants with a history of MS. The FAA should classify the applicant's condition according to the scheme of Smith et al.[5] Also, the course of the disease should be determined, such as benign, exacerbating-remitting,[12] etc. The applicant should undergo a complete neurological evalution in order to verify the diagnosis and to determine the presence of activity or neurological residuals that would compromise flight safety. Neuropsychological tests, evoked potentials, CSF analysis for oligoclonal bands and IgG synthesis and/or a MRI may be necessary before these questions can be answered.

The following guidelines are suggested for applicants with a clinically or laboratory-supported definite diagnosis: a) applicants with progressive MS should not be certified; b) applicants with remitting-progressive MS should not be certified; c) applicants with either exacerbating-remitting or benign MS may be certified provided a period of one year has elapsed since the last attack and the candidate otherwise satisfies the neurological criteria. However, persons with either benign or exacerbating-remitting MS who have a history of "paroxysmal attacks" such as seizures, visual disturbances, vertigo, trigeminal neuralgia, ataxia, or dysarthria, should be permanent disqualified. Persons with a diagnosis of definite MS who are certified should be examined yearly by a neurological consultant.

Applicants with a clinically or laboratory-supported diagnosis of probable MS

cannot be certified until a definite diagnosis is made. When it is established, the crtieria listed previously should be followed. Persons with multiple sclerosis under teatment with immunosuppressive drugs, or medications to treat symptoms such as trigeminal neuralgia, bladder dysfunction, or spasticity, are disqualified. An applicant with a history of optic neuritis may be certified if: 1) the diagnosis is confirmed and there is a full return of visual function that satisfies the visual standards and guidelines and 2) a neurological evaluation uncovers no other disqualifying deficits. Since persons with optic neuritis may be MS suspects, in all likelihood paraclinical tests will be performed. Depending on which test is positive, applicants may be classified as laboratory supported definite MS or clinically probable MS. If so, the recommendations for those forms of MS should be followed. If these studies are normal, the applicant should be examined at yearly intervals.

Other Primary Demyelinating Diseases

The demyelinating diseases of infectious origin are covered in the section on infectious diseases. Other causes are the genetic metabolic diseases of myelin listed in Table V. Because of the severe neurological impairment associated with all of these diseases, all of them are disqualifying.

TABLE V

CLASSIFICATION OF GENETIC METABOLIC DISORDERS OF MYELIN

1. Krabbe's globoid leukodystrophy

   Infantile
   Late Onset

2. Metachromatic leukodystrophy

   Congenital?
   Late Infantile
   Juvenile
   Adult
   Multiple sulfatase deficiency
   Cerebroside sulfate sulfatase activator deficiency

3. Adrenoleukodystrophy

   Sex linked

      Adrenoleukodystrophy
      Adrenomyeloneuropathy
      Familial Addison's disease
      Symptomatic female carriers

   Autosomal recessive

      Neonatal adrenoleukodystrophy

4. Refsum's disease

5. Pelizaeus-Merzbacher disease

   Classic
   Neonatal Seitelberger type

6. Alexander's disease

7. Spongy degeneration of the nervous system

8. Nonspecific hypomyelination

source: REF 18

References

1. Izuierdo G, Hauw, JJ, Lyon-Caen O, et al: Value of multiple sclerosis diagnostic criteria: 70 autopsy-confirmed cases. Arch Neurol 1985;42:848-850.
2. Poser CM (ed): The diagnosis of multiple sclerosis. New York, Thieme-Stratton, Inc, 1984.
3. Joynt RJ (ed): Disorders of Myelin. Seminars of Neurology. New York, Thieme-Stratton, Vol 5, 1985.
4. Paty DW, Ebers, GC, Wonnacott TH: Prognostic markers in multiple sclerosis. 12th World Congress of Neurology. Excerpta Medica, International Congress Series, 1981;548:56.
5. Rose AS, Ellison GW, Myers LW, et al: Criteria for the clinical diagnosis of multiple sclerosis. Neurology 1976;26:20-22.
6. Smith CR, Scheinberg LC: Clinical features of multiple sclerosis, in: Joynt RJ (ed): Disorders of Myelin, op cit, pp 85-93.
7. Poser CM, Paty DW, Scheinberg L, et al: New diagnostic criteria for multiple sclerosis: Guidelines for research protocols. Ann Neurol 1983;13:227-231.
8. Hart RG, Scherman DG: The diagnosis of multiple sclerosis. JAMA 1982;247:498-503.
9. Gebarski SS, Gabrielsen TO, Gilman S, et al: The initial diagnosis of multiple sclerosis: Clinical impact of magnetic resonance imaging. Ann Neurol 1985;17:469-474.
10. Kirshner HS, Tsai Si, Rung VM, Price AC: Magnetic resonance imaging and other techniques in the diagnosis of multiple sclerosis. Arch Neurol 1985;42:859-863.
11. Tramo MJ, Schneck MJ, Lee BCP, et al: Evoked potentials and MRI in the diagnosis of multiple sclerosis. Neurology 1985;35 (Suppl 1):105.

12. Kraft GH, Freal JE, Coryell JK, et al: Multiple sclerosis: Early prognostic guidelines. J Chronic Diseases 1977;30:819-830.

13. Kurtzke JF: Rating neurological impairment in multiple sclerosis: An expanded disability status scale (EDSS). Neurology 1983;33:1422-1427.

14. McDonald WI: The significance of optic neuritis. Tran Ophthal Soc of United Kingdom 1983;103:230-246.

15. Compston DAS, Batchelor JR, Earl CJ, McDonald WI: Factors influencing the risk of MS developing in patients with optic neuritis. Brain 1978;101:495-511.

16. Ebers GC: Optic neuritis and multiple sclerosis. Arch Neurol 1985;42:702-704.

17. Kurtzke JF: Optic neuritis or multiple sclerosis. Arch Neurol 1985;42:704-710.

18. Hollander J. Genetic metabolic disease of myelin, in Joynt RJ (ed): Disorders of Myelin, op cit, pp 155-167.

Acquired and Hereditary Ataxias

Ataxia is defined as a decomposition of movement associated with lack of coordination, unsteadiness and irregularity of muscular action. Acquired or hereditary diseases of the cerebellum or its connections are primarily responsible for this neurological abnormality. The acquired diseases span most categories of neurological diseases, including infection, demyelination, cerebral vascular disease, and tumors.

The dominantly or recessively inherited ataxic disorders are characterized by spinocerebellar atrophy. Although classifications based on clinical, pathological and, in some cases, metabolic defects are not entirely satisfactory because of genetic heterogeneity, the broad classification in Table I contains the diseases most commonly encountered.

In addition to the major disorder, cerebellar ataxia, other associated signs and symptoms are caused by spinal cord, brain stem, basal ganglia, and cerebral cortex involvement. Impairment of vision and hearing may occur. Skeletal abnormalities, particularly scoliosis, pes cavus, and hammer toes are common. Cardiomyopathy is frequently found in Friedreich's ataxia.

Dominant or recessive ataxias account for approximately one third of cases, sporadic ataxias also about one third, and the rest are due to other causes. In the dominant forms, the onset of symptoms occurs typically between adolescence and the fifth decade. The recessive syndromes, the most frequent being Friedreich's disease, usually begin in childhood. The sporadic may occur at any age. In all cases, the hereditary ataxias are relentlessly progressive.[2]

Persons with hereditary ataxic diseases should be permanently disqualified from flying. The aeromedical disposition of persons whose primary, persistent neurological deficit is acquired ataxia of the limbs or trunk is based on the underl ing disease and the degree of neurological impairment.

TABLE I

THE CLASSIFICATION OF HEREDITARY ATAXIAS

I. Friedreich's ataxia and variants

II. Cerebellcolivary atrophy
   1. Dominant
   2. Recessive (Holmes)
   3. Sporadic (Marie-Alajouanine)

III. Olivopontocerebellar atrophy
   1. Dominant
      a. Menzel's type
      b. With retinal degeneration
      c. With slow saccades
      d. Spinopontine type
   2. Recessive
   3. Sporadic (Dejerine-Thomas)
   4. Sporadic with progressive autonomic failure (Shy-Drager syndrome)

IV. Familial spastic paraplegia (Strumpell-Lorrain and variants)

V. Familial spastic ataxia (Sanger Brown)

source: REF 1

Acquired and Hereditary Ataxias

Ataxia is defined as a decomposition of movement associated with lack of coordination, unsteadiness and irregularity of muscular action. Acquired or hereditary diseases of the cerebellum or its connections are primarily responsible for this neurological abnormality. The acquired diseases span most categories of neurological diseases, including infection, demyelination, cerebral vascular disease, and tumors.

The dominantly or recessively inherited ataxic disorders are characterized by spinocerebellar atrophy. Although classifications based on clinical, pathological and, in some cases, metabolic defects are not entirely satisfactory because of genetic heterogeneity, the broad classification in Table I contains the diseases most commonly encountered.

In addition to the major disorder, cerebellar ataxia, other associated signs and symptoms are caused by spinal cord, brain stem, basal ganglia, and cerebral cortex involvement. Impairment of vision and hearing may occur. Skeletal abnormalities, particularly scoliosis, pes cavus, and hammer toes are common. Cardiomyopathy is frequently found in Friedreich's ataxia.

Dominant or recessive ataxias account for approximately one third of cases, sporadic ataxias also about one third, and the rest are due to other causes. In the dominant forms, the onset of symptoms occurs typically between adolescence and the fifth decade. The recessive syndromes, the most frequent being Friedreich's disease, usually begin in childhood. The sporadic may occur at any age. In all cases, the hereditary ataxias are relentlessly progressive.[2]

Persons with hereditary ataxic diseases should be permanently disqualified from flying. The aeromedical disposition of persons whose primary, persistent neurological deficit is acquired ataxia of the limbs or trunk is based on the underlying disease and the degree of neurological impairment.

References

1. Plaitakis A, Gudesblatt M: The hereditary ataxias, in: Appel SH (ed): Current Neurology. Philadelphia, Wiley and Sons, 1984, Vol 5, No 4, pp 471-509.

2. Stumpf DA: The inherited ataxias. Pediat Neurol 1985;1:129-133.

Movement Disorders

Introduction

Movement disorders are classified according to their physical characteristics, as shown in Table I. They are important because of their high prevalence in the general population, their interference with motor and coordination functions, and their intensification with stress and fatigue. In some cases, their causes are acquired and in others genetically determined. The acquired movement disorders may result from an acute event, from which the person either recovers completely or is left with a residual deficit. Chronic acquired and the hereditary movement disorders are slowly progressive. The type of movement is not specific to a particular disease; several types may be seen in the same person as well as in the same disease.

With the acquired conditions, the decision to certify is dependent on the cause as well as the persistence of neurological deficits. The hereditary movement disorders present two problems: 1) determining the degree of impairment and rate of progression, and the frequency of follow-up examinations and; 2) the frequency at which pilot applicants with a positive family history should be followed.

In this section only the more frequent disorders will be discussed. The reader is referred to standard neurological textbooks or review articles for further information.[2-4]

The incidence of various types of disorders evaluated at the Baylor Movement Disorder Clinic, for example, is listed in Table II.[5]

Minimal criteria for diagnosis and classification

The physical findings of a generalized or focal, hypo- or hyperkinetic disturbance of motor function should suggest the possibility of a movement disorder. A classification of movement disorders is shown in Table III. The FAA should become familiar with terms and eponyms that may appear in pilot-candidate's application or in a physician's report.

TABLE I

DIFFERENTIATING INVOLUNTARY MOVEMENTS

Movement	Speed	Quality	Predominant Topography	Contraction Pattern
Tremor	3-8/sec	Rhythmic oscillation	Distal	Alternating agonist and antagonist
Chorea	Fast	Forceful jerking	Face distal	Intermittent agonist
Athetosis	Slow	Continuous endulation	Face distal	Irregular sustained agonist and antagonist
Ballismus	Fast	Flinging	Proximal (unilateral)	Repetitive agonist
Dystonia	Slow	Sustained contraction	Axial and proximal	Sustained agonist and antagonist
Myoclonus	Fast	Jerking	Diffuse	Irregular arrythmic angonist

source: REF 1

TABLE II

INCIDENCE OF MOVEMENT DISORDERS*

	% of Total
Parkinsonism	53.2
Parkinson's disease	47.7
Progressive supranuclear palsy	2.0
Shy-Drager syndrome	0.8
Neuroleptic-induced	0.6
Other	2.1
Tremor	15.8
Essential	11.0
Other	4.8
Dystonia	14.2
Dyskinesia	5.4
Tics	5.1
Chorea	4.4
Myoclonus	4.1
Hemiballism	0.6
Athetosis	0.3
Other	4.6

*First 1000 patients evaluated at the Movement Disorder Clinic, Baylor College of Medicine

source: REF 5

Table III
CLASSIFICATION OF MOVEMENT DISORDERS

Hypokinetic-akinetic (bradykinetic) or parkinsonian disorders

I. Dopamine deficiency rates (presynaptic)
   A. Primary (idiopathic) Parkinson's disease

   B. Secondary (symptomatic)
      1. Postencephalitic
      2. Syringomesencephalalia
      3. Drugs - reserpine, tetrabenazine, a-methyl-dopa, lithium, MPTP

II. Dopamine receptor disorders (postsynaptic)
   A. System degenerations (parkinsonism plus)
      1. Shy-Drager or multiple system atrophy (autonomic failure)
      2. Progressive supranuclear palsy (ophthalmoplegia and pseudobulbar palsy)
      3. Olivopontocerebellar atrophy (ataxia)
      4. Striatonigral degeneration
      5. Parkinsonism-dementia-ALS complex
      6. Juvenile parkinsonism (rare)
      7. Hemiatrophy - hemiparkinsonism
      8. Hereditary disorders - Huntington; Wilson; Hallervorder-Spatz; spinocerebellar degenerations; familial basal ganglia calcification; familial parkinsonism with peripheral neuropathy; neuronal ceroid lipofuscinosis; glutamate dehydrogenase deficiency

   B. Secondary (symptomatic)
      1. Drugs and toxins - antipsychotics, antiemetics, tetrabenazine, CO MN, CS_2, methanol, ethanol
      2. Other - parathyroid abnormalities, hypothyroidism, hepatocerebral degeneration, brain tumor, Creutzfeldt-Jakob disease, normal pressure hydrocephalus, akinetic mutism

Hyperkinetic
I. Paroxysmal
   A. Choreoathetosis - dystonia
      1. Familial - kinesigenic or nonkinesigenic
      2. Acquired - multiple sclerosis, static encephalopathy, hypoparathyroidism, head trauma, seizures, sporadic paroxysmal choccoathetrosis-dystonia, oculogyric crises

   B. Myoclonus
      1. Physiologic
      2. Essential
      3. Epileptic
      4. Symptomatic progressive or static encephalopathy
      5. Segmental (bulbar, spinal)

   C. Tics -Tourette's syndrome

   D. Tremor

E. Hyperreflexia

F. Other - Habit spasms, mannerisms, hemifacial spasm

II. Nonparoxysmal
- A. Tremor
  - 1. At rest - parkinsonian
  - 2. Action - physiologic, essential, cerebellar, parkinsonian
- B. Choreoathetosis
  - 1. Sterotyped tardive dyskinesia
  - 2. Nonstereotyped
    - a. Hereditary - Huntington's disease
    - b. Symptomatic - Sydenham, SLE, chorea gravidarum, metabolic, toxic and static encephalopathy
    - c. Sporadic
- C. Dystonia
  - 1. Primary
    - a. Hereditary (dystonia musculorum deformans, Hallervorden-Spatz, etc)
    - b. Sporadic (idiopathic torsion dystonia) - generalized, segmental, focal (Meige)
  - 2. Secondary
- D. Ballism - hemiballism

III Other
- A. Akathisia
- B. Restless legs
- C. Disorders of gait and posture
- D. Disorders of muscle tone - rigidity, gegenhalten (paratonia) spasticity, hypotonia, myotonia, carpopedal spasms, stiff person syndrome, rigid spine syndrome

---

Source: Ref 5

The number of conditions that may cause hyper- and hypokinetic movement disorders is large. The rest of this section describes those movement disorders most likely to be encountered in the pilot age group that present difficult disposition problems, or that have recently received much publicity in the lay press, despite their low prevalence. These conditions are: 1) acute drug-induced movement disorders; 2) chronic drug-induced movement disorders; 3) Sydenham's chorea; 4) Parkinson's disease; 5)

essential tremor; 6) hemifacial spasm; 7) tics and Tourette's syndrome; 8) focal dystonias; 9) Huntington's disease; and 10) Wilson's disease.

For aeromedical purposes, the decision to certify is based on the following considerations; 1) is the cause reversible, that is, can it be permanently cured? 2) are the neurological residuals, if present, static and do they compromise flying safety? 3) is the disease inherited and therefore progressive? 4) is the applicant's movement disorder being treated with drugs, even if the disease is static, which would disqualify him as long as he is under treatment? 5) does the candidate have a family history of a movement disorder, in which case he should be followed at frequent intervals? 6) has the candidate had surgical treatment, in which case, with some specific exceptions, he should be denied certification?

Acute drug induced movement disorders

Medications should always be suspected as a cause for the sudden onset of dyskinesias, particularly in the younger age group. In those persons with an idiosyncratic reaction, the abnormal movements, which are usually dystonic or choreiform, begin within several hours after a single dose and are not dose-dependent. Unless treated, they resolve spontaneously over a period of several hours or days. Persistent dyskinesias are rare. With neuroleptic drugs the overall incidence of movement disorders is approximately 2.5% to 5%. However, with the more potent piperzine phenothiazines and butyrophenones, it may approach 50%. The dyskinesias may recur with re-introduction of the same drug or other medications of equal potency.

Rarely, the neuroleptic malignant syndrome may occur. This is associated with the onset of fever, movement disorder and altered mentation. Most cases occur following a single or combined administration of haloperiodol (Haldol^{R}), fluphenazine (Prolixin^{R}) or chlorpromazine (Thorazine^{R}). Serious medical and neurological sequelae may occur, although patients may recover completely.

In the dose-related (toxic) group, the dyskinesias including tremors may or may not be associated with other signs and symptoms of drug toxicity. The most frequent drugs associated with acute, idiosyncratic or toxic dyskinesias, are phenothiazines, butyrophenones, levodopa, bromocriptine, amphetamines, methylphenidate, metoclopramide, anticholinergic containing drugs, lithium, and anticonvulsants.

The following guidelines are suggested for evaluation of candidates with a history of acute drug induced dyskinesias: 1) determine the drug(s) that is responsible for the dyskinesias; 2) ascertain if the drug(s) was taken as prescribed or was taken as an intentional overdose; 3) identify the underlying cause for which the drug(s) was prescribed; and 4) do a complete neurological examination.

If the reaction was idiosyncratic or secondary to an unintentional overdose, if the underlying disease is not disqualifying, and if the neurological examination is normal, the applicant may be certified. However, he should be told that he is at risk for a recurrence of dyskinesias if the same or related drugs are taken again.

Chronic drug induced movement disorders

The chronic neurological syndromes associated with drugs, particularly the neuroleptics, should not present problems for aeromedical disposition, since the underlying conditions for which the medication was given, such as psychotic disorders, are, for the most part, disqualifying. However, occasionally applicants may have a history of a transient, self-limiting psychiatric or medical disorders, and may have developed drug-induced dyskinesias while under treatment, although at the time of evaluation they may be neurologically normal. Examples of this might be drug-induced parkinsonism resulting from the treatment of hypertension with alpha methyldopa (Aldomet^{R}) or gastrointestinal disorders with metoclopramide (Reglan^{R}).

Tardive dyskinesia (TD) is characterized by involuntary, choreiform movements, affecting primarily the facial muscles, and also the extremities. This extrapyramidal

syndrome is caused by chronic use of medications that block dopamine receptor sites. Most of these drugs are neuroleptic and are prescribed for psychiatric conditions. However, prochlorperazine (Compazine^{R}) and metoclopramide (Reglan^{R}) are frequently prescribed to nonpsychiatric patients with acute or chronic gastrointestinal diseases. TD usually begins after three months of continuous drug use. It is estimated that 10% to 20% of persons taking neuroleptic drugs develop this condition. The incidence increases with age. Remission may occur in 50% to 90% of persons under 40 years of age following drug withdrawal; in older individuals, the figure is 33%. In the vast majority of cases, if improvement occurs, it will do so within the first three months after the drug(s) is discontinued. A minimum of 6 months should elapse before the condition is considered irreversible. There is no effective medical treatment for this disorder.

Dyskinesias may follow the abrupt discontinuance of neuroleptics (withdrawal dyskinesias) and usually resolve spontaneously within several weeks to three months.

Akathisia is a condition characterized by motor restlessness such as continual shuffling of the feet and a feeling of inner tension. This usually occurs soon after the initial administration of or an increase in dosage of neuroleptic drugs, most commonly reserpine, phenothiazines (particularly fluphenazine (Prolixin^{R})), and butyrophenones. Its incidence is about 20%. Ninety percent of persons who develop this disorder will do so after two to three months. Akathisia usually improves slowly after medication withdrawal, although persistent cases have been reported.

All the major features of parkinsonism can be produced by neuroleptic drugs, particularly reserpine, phenothiazines, thioxanthenes, and the butyrophenones. Metoclopramide (Reglan^{R}) and alphamethyldopa (Aldomet^{R}) can also cause this syndrome. Of recent interest is 1 methyl 4 phenyl 1,2,3,6-tetrahydropyridine (MPTP)-induced parkinsonism. Symptoms may begin within several days after treatment is initiated, reaching their peak by three months, at which time 90% of cases will have developed. The patients are usually free of their extrapyramidal signs within several

weeks after the drug is discontinued, although slower rates of recovery occur in the older population. Persistent cases have been reported. Patients with this condition may be treated with antiparkinson drugs, primarily the centrally active anticholinergic drugs such as trihexyphenidyl (Artane^{R}). However, the only possible complete cure is the discontinuance of the responsible drug.

Applicants with a history of tardive dyskinesias, withdrawl dyskinesias, akathisias, and drug-induced parkinsonism are permanently disqualified from flying, unless 1) the cause of the disorder can be attributed directly to a drug(s); 2) the medical or psychiatric condition for which the drug(s) was given is not disqualifying; 3) the neurological evaluation performed by a consultant is normal; 4) the candidate is not taking antidyskinesia or antiparkinsonian medications.

Sydenham's chorea

Although rheumatic fever is rare today, chorea, carditis and migratory polyarthritis are still the major clinical signs of this disease. The age group between 7 and 14 years are usually affected. It is rare after puberty. The disease usually runs its course within one to two months, although minor movements may persist for six months. In one third of the cases recurrences after several months or years have been reported. Recovery is usually complete.

A candidate with a history of Sydenham's chorea may be certified if 1) the initial or the last recurrent episode occurred prior to one year of the date of application; 2) there are no neurological residuals as determined by a neurological consultant; and 3) the applicant satisfied all other medical criteria, primarily those for the heart.

Parkinson's disease (paralysis agitans)

Parkinson's disease is a common, progressive, degenerative disease of the central nervous system. Prevalence rates range between 66 and 200/100,000 population, and

with annual incidence is approximately 5 to 24/100,000. The mean age of onset is 55 years. Males are affected more than females. A positive family history is found in 5% to 15% of probands; however, there is no conclusive evidence that the disease is genetically determined.

The clinical hallmarks of this disease are bradykinesia, rigidity, and resting tremor. Other common features are disturbances of posture, equilibrium, autonomic functions and ocular motility. In 10% of persons with this condition, the onset of tremor and/or rigidity is unilateral. As the disease progresses, despite therapeutic intervention, deficits in cognition, perception and memory may appear in approximately 30% of individuals. Dementia is common in individuals with an older age at time of onset. It has been suggested that the dementia associated with Parkinson's disease is co-existent with Alzheimer's disease (Parkinsons-Alzheimer complex). Signs of dementia are more frequent in persons with bradykinesia and postural and gait abnormalities than in those in which tremor predominates.

The initial symptoms in Parkinson's disease are listed below in Table IV, according to one study.[6]

TABLE IV
INITIAL SYMPTOMS IN PARKINSON'S DISEASE

	No. of Cases (183)	(%)
Tremor	129	70.5
Stiffness or slowness of movement	36	19.7
Loss of dexterity and/or handwriting disturbance	23	12.6
Gait disturbance	21	11.5
Muscle pain, cramps, aching	15	8.2
Depression, nervousness, or other psychiatric disturbance	8	4.4
Speech disturbance	7	3.8
General fatigue, muscle weakness	5	2.7
Drooling	3	1.6
Loss of arm swing	3	1.6
Facial masking	3	1.6
Dysphagia	1	0.5
Paresthesia	1	0.5

Average number of initial symptoms per patient	1.4

source: Ref 6

When the disease has been present for several years, the diagnosis is relatively easy. However, other causes for the clinical signs and symptoms, as shown in Table V, must be ruled out.[7]

TABLE V
CONDITIONS THAT MAY PRESENT WITH PARKINSONIAN FEATURES

Multiple System Degenerations
- Striatonigral degeneration
- Olivopontocerebellar atrophy
- Shy-Drager syndrome
- Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy)
- Joseph disease (Machado disease, nigral-spinal-dentatal degeneration with nuclear ophthalmoplegia, Azorean disease)
- Corticodentatonigral dengeneration
- Parkinsonism - dementia of Guam

Other Diseases
- Drug or toxin-induced parkinsonism
  - Neuroleptic-induced parkinsonism
  - Carbon monoxide poisoning
  - Chronic manganese poisoning
  - MPTP-induced parkinsonism
- Normal pressure hydrocephalus
- Jakob-Creutzfeldt disease
- Hypoparathyroidism with basal ganglia calcification
- Chronic hepatocerebral degeneration
- Brain neoplasms or arteriovenous malformations
- Rigid form of Huntington's chorea
- Wilson's disease

source: Ref 7

Of aeromedical importance is the early recognition of the disease before the classic signs are apparent. In most cases, the symptoms that precipitate the intial visit to a physician are tremor and disturbances in equilibrium and gait. At this time, the

disease may be mildly to moderately advanced. The early signs of the disease, such as lack of facial expression, decreased frequency of eye blinks, voice changes, paucity of associated movements, slowness in initiating and maintaining motor actions, and sialorrhea, may not be appreciated by the patient. In addition, non-specific symptoms, such as fatigue, muscle aches, cramps and weakness, sleep disturbances, and mental changes, may not be recognized by the physician as early signs of the disease. Early recognition is important because the neurological abnormalities associated with Parkinson's disease, as well as the pseudoparkinson diseases, adversely affect reaction and movement times, coordination and postural reflexes, voice, ocular motility, mentation and behavior. These changes may be accentuated by alterations in environment, sleep and mood, as well as with fatigue and degree of motor activity.

The disease is slowly progressive. Prior to the treatment with levodopa, the course from onset to death was approximately 11 years. Now, with this and other drugs, life expectancy has increased, approaching normal values. However, these medications do not alter the ultimate progression of the disease, but only temporarily ameliorate the signs and symptoms. For the most part, they have been very effective in reducing some of the most disabling signs, such as bradykinesia, during the first several years of treatment. Unfortunately, with prolonged use, troublesome side effects like dyskinesias or psychiatric disturbances occur as the disease progresses.

The drugs that ar used to treat this as well as related conditions are, by themselves, disqualifying because of their side effects. The following list should alert the FAA to the possibility that applicants in this age group who are taking or who have taken these medications in the past may have Parkinsons' disease or related disorders.

1. Anticholinergics
    a. trihexyphenidyl
    b. benztropine mesylate
    c. biperiden

d. procyclidine

e. cycrimine

f. diphenhydramine

g. chlorphenoxamine

h. orphenadine

2. Levodopa drugs

a. levodopa

b. levodopa/carbidopa (Sinemet^{R})

3. Bromocriptine

4. Amantadine

Since the advent of levodopa for the treatment of this disease, the indications for surgical intervention are very limited. Applicants with a history of surgical treatment should be disqualified permanently because the disease is usually too far advanced at the time the procedure is performed.

An applicant with a history of idiopathic parkinsonism (Parkinson's disease) should be permanently disqualified from flying. The only possible exception is the applicant who is in the early stages of the disease with mild neurological deficits that have been judged, after flight simulation tests, not to interfere with reaction time, voice control, and visual tracking. If certified, these candidates must be followed at six month intervals by a neurological consultant.

Applicants under chronic treatment with any of the aforementioned drugs, or who have been treated surgically, should not be certified.

The other conditions that may present with parkinsonian features (Table V) are disqualifying with the exception of those induced by drugs, toxins or endocrine

abnormalities in some cases.

Essential tremor

Essential tremor is an action or postural-induced tremor. It most frequently involves the upper extremities (in 95% to 100% of cases), head (34%), voice (20%), and lower limbs (25%). The prevalence rate in the US is 3/1,000, and it is slightly more common in women. In 50% of cases it is known to be inherited as an autosomal dominant condition. The mean range of age of onset is 35 to 46 years, with earlier onset in the hereditary cases. A bimodal onset has been suggested, with the first peak at age 15 years and the second at 60 years.

With the exception of the tremor, the neurological examination is usually normal. The course is variable but in most cases the tremor progresses slowly with an increase in amplitude and spread to other body parts. It has been reported that essential tremor beginning at age 50 years will be of moderate severity by age 65 years and severe by 70 years. A moderate-to-severe tremor can be functionally incapacitating, affecting one's ability to write, eat or coordinate fine hand movements. The tremor also is embarassing to the individual, particularly when the neck and voice are involved. The tremor increases under stress, exercise and fatigue and with the use of stimulants such as caffeine-containing beverages or drugs, or with amphetamines. It is transiently improved by the use of small to moderate amounts of alcohol. The tremor then intensifies as the blood alcohol level falls. Because of the inital "benefit" chronic alcohol us may become a problem, and 67% of persons with essential tremor may abuse alcohol.

The drugs that are currently used to treat this condition are beta-adrenergic blockers, particularly propranolol, sodium valproic acid, clonazepam, primidone, tranquilizers, and sedatives and hypnotics. Pilot applicants taking these drugs should be disqualified from flying not only because of their side effects, but because their use suggests that the disease is already impairing.

Essential tremor shas been reported in other neurological conditions such as Charcot-Marie-Tooth, and may be coincidentally seen in persons with parkinsonism and with dystonia muscular deformans. It should not be confused with tremors associated with cerebellar disease such as in multiple sclerosis. An accentuated physiologic tremor secondary to drugs or other conditions may be confused with essential tremor. These are listed in Tables VI and VII.

TABLE VI
SUBSTANCES PRODUCING ENHANCED PHYSIOLOGIC TREMOR

Dextroamphetamine
Epinephrine and beta-2 agonists
- Isoetharine
- Isoproterenol
- Metaproterenol
- Terbutaline

Levodopa
Lithium
Neuroleptics
- Butyrophenones
- Phenothiazines

Prednisone
Thyroid hormones
Tricyclic antidepressants
Xanthines in coffee or tea

source: Ref 8

TABLE VII
CIRCUMSTANCES ASSOCIATED WITH ENHANCED PHYSIOLOGIC TREMOR

Increased activity around the segmental stretch reflex arc
Cold shivering
Exercise
Fatigue
Forceful contraction (including Jendrassik's maneuver)
Hypoglycemia
- Insulin
- Nondiabetic

Pheochromocytoma
Thyrotoxicosis
Vibration of muscle
Withdrawal of opiate or sedative compounds

source: Ref 8

Additional information may be obtained from Larson and Caine[9] and Koller.[10]

Candidates with a history of sporadic or hereditary essential tremor may be

certified if they have no other neurological cause and the examination is otherwise normal.

Hemifacial spasm

Hemifacial spasm is characterized by hyperactive unilateral brief contractions of facial muscles. It usually starts in the obicularis oculi muscle, but over a period of time it spreads to other muscles innervated by the 5th cranial nerve. With recurrent sustained contractions the eyes will close and the mouth will be pulled upwards. Emotion, stress, fatigue and stimulants will increase with spasm. The onset is usually in the sixth decade of life, and the condition is more common in females. It is generally a chronic progressive disease, which is annoying and embarrassing to the patient. The current accepted cause is pulsatile compression of the facial nerve by elongated arterial loops, most commonly of the posterior inferior cerebellar artery. Drug management has, for the most part, been ineffective. Medications such as carbamazapine (Tegretol^{R}), phenytoin (Dilantin^{R}), and phenobarbital are contraindicated for flying. The currently accepted treatment is microvascular decompression, or the Jannetta procedure, which has afforded immediate relief for approximately 90% of persons with this disease. However, recurrence as early as one to two years after the operation is not uncommon. The major surgical complication is deafness.[11-12]

Candidates with a diagnosis of hemifacial spasm are disqualified from flying unless the spasms have been relieved by surgical intervention, the audiological examination meets the standards, there is no significant facial weakness, and a symptom-free period of six to 12 months has elapsed since the surgical procedure. Thereafter, the applicant should be followed on a yearly basis.

Tics

Tics are involuntary, brief, rapid contractions of skeletal muscles in one or more

parts of the body. In many respects they resemble myoclonus. It is estimated that "tics" may be present in 28% of the total population, with the majority of tic disorders occurring in children and adolescents. The various tic disorders and diagnostic criteria are listed in Table VIII.

The tic disorder that has gained the most attention is Tourette's syndrome. This is a chronic disorder in which there are both involuntary motor and vocal tics. Its onset is between five and 10 years. Males are significantly more affected than females. The initial tic is usually one of repetitive eyelid blinking or movement of the head and neck. As the disease progresses, tics of the extremities may occur. Vocal tics are present in all cases. Coprolalia and copropraxia are found in approximately 30% of cases. Persons with Tourette's syndrome also exhibit compulsive motor activities such as skipping and touching objects. The symptoms usually reach a peak in late adolescence and thereafter regress or remain stable. The symptoms usually are chronic and lifelong, although spontaneous remissions have been reported in 7% to 19% of patients. These individuals are usually neurologically and intellectually normal. The drug of choice for treatment is haloperidol (Haldol^{R}), which is contraindicated for flying.

An additional reference on Tourette's syndrome is Butler.[14]

Candidates with a diagnosis of Tourette's syndrome should not be certified unless they are free of symptoms and off all medications for five years and the results of neurological as well as neuropsychological studies are normal.

The other tic disorders listed in Table VIII should be evaluated on an individual basis. Most of these are self-limiting.

Dystonias

Generalized as well as focal dystonias can be found in a number of acquired and genetically determined diseases (Table IX).

The hereditary or idiopathic dystonias usually begin focally, for example in the

TABLE VIII

DIAGNOSTIC CRITERIA FOR TIC DISORDERS

Transient Tic Disorder

- Onset during childhood or early adolescence
- Presence of recurrent, involuntary, repetitive, rapid, purposeless motor movements (tics)
- Ability to suppress the movements voluntarily for minutes to hours
- Variation in the intensity and type of symptoms over weeks or months
- Duration of at least 1 month but not more than 1 year

Tourette Syndrome

- Age at onset between 2 and 15 years
- Presence of recurrent, involuntary, repetitive, rapid, purposeless motor movements affecting multiple muscle groups
- Multiple vocal tics
- Ability to suppress movements voluntarily for minutes to hours
- Variations in the intensity and type of symptoms over weeks or months
- Duration of more than 1 year

Confirmatory but not essential for the diagnosis

- Coprolalia (involuntary swearing)
- Copropraxia (involuntary obscene gesturing)
- Echolalia (involuntary repetition of words or sounds heard by the patient)
- Echopraxia (involuntary imitation of the movements of others)
- Palilalia (involuntary repetition of patient's own words or sounds)

Multiple Tic Disorder

- Age of onset between 2 and 15 years
- Presence of recurrent, involuntary, repetitive, rapid, purposeless motor movements affecting multiple muscle groups. Rarely do patients have only vocal tics
- Ability to suppress movements voluntarily for minutes to hours
- Variations in the intensity and type of symptoms over weeks or months
- Duration of more than 1 year

Chronic Motor Tic Disorder

- Presence of recurrent, involuntary, repetitive, rapid, purposeless movements (tics) involving no more than three muscle groups at any one time
- Unvarying intensity and type of tics over weeks or months
- Ability to suppress the movements voluntarily for minutes to hours
- Duration of at least 1 year

source: REF 13

TABLE IX

CLASSIFICATION OF THE DYSTONIC STATES

I. Primary Dystonia
   A. Hereditary
      1. Autosomal dominant
      2. Autosomal recessive
      3. X-linked recessive
      4. Paroxysmal dystonia
   B. Idiopathic
      1. Distribution pattern: generalized, segmental, focal
      2. Onset: childhood, adult

II. Secondary Dystonia
   A. Associated with other hereditary neurologic disorders
      1. Wilson's disease
      2. Huntington's disease (Westphal variant)
      3. Hallervorden-Spatz disease
      4. GM-1 gangliosidosis
      5. Hexosamidase A and B deficiency
      6. Juvenile dystonic lipidosis
      7. Glutaric acidemia
      8. Joseph disease
   B. Environmental dystonia
      1. Perinatal cerebral injury
      2. Infection
      3. Postinfectious
      4. Reye's syndrome (Personal observation)
      5. Head trauma
      6. Focal cerebral vascular injury
      7. Brain tumor
      8. Toxins: manganese, carbon monoxide
      9. Drugs: levodopa, antipsychotics, metoclopramide, anticonvulsants
   C. Psychologic dystonia (personal observations)

source: REF 15

foot; however, over a period of time, they become generalized. This secondary generalization also pertains to other primary, hereditary, progressive neurological diseases caused by either metabolic or enzymatic defects. In addition to focal or generalized dystonia, these patients have other neurological abnormalities. An example of this is torsion dystonia, a genetically determined disease most prevalent in Ashkenazic Jews.

There is no effective treatment for these disorders with the exception of Wilson's disease (see below). Candidates with primary or secondary dystonia should not be certified.

Persistent dystonias, particularly focal ones, rarely may follow an acute event such as a cerebral vascular accident, head trauma, or infection. Dispositions in these cases is related to determining the primary cause as well as the degree of neurological impairment. As with all movement disorders, the movement is increased with stress, fatigue, and exercise. Applicants on drugs to control the disorder are disqualified. It should be noted that the control of these movements by medications is not very satisfactory.

Perhaps the major aeromedical disposition problem is the evaluation of persons with idiopathic focal dystonias. These usually occur in adults, are not secondary to structural disease, involve primarily facial or neck muscles, and are difficult to treat. The focal idiopathic dystonias have been classified by Marsden,[16] and are found in Table X.

TABLE X

Cranial dystonia
- Blepharospasm
- Oromandibular dystonia
- Laryngeal dystonia ("Spastic dysphonia")
- Pharyngeal dystonia

Spasmodic torticollis
Truncal dystonia

Writer's cramp and other craft palsies

source: Ref 16

All persons with dystonias should be disqualified permanently.

Essential blepharospasm

Essential blepharospasm begins at an average age of 52 years in men and 60 years in women. Women tend to be more adversely affected. Its onset is insidious, usually with brief, infrequent painless closures of one or both eyes. If the onset is unilateral, the condition will eventually involve both eyes. With time, the episodes become more frequent and prolonged, and vary from day to day, but tend to become more severe in the afternoon, in bright light and during periods of physical and emotional stress and fatigue. The condition may be so severe as to affect visual function.

The drug treatment of this disorder is not particularly effective. Several surgical procedures are currently used, including open division of the facial nerve branches to the eyelide or avulsion or thermolytic fractional destruction. Although the studies are limited, it is estimated that approximately 70% of persons with essential blepharospasm improve for an average of 20 months; however, recurrences may occur in as many as 50% of these individuals within three years. There are other ophthalmologic complications, such as ectropion of the lower eyelids, or exposure keratonathy. Most recently, the subcutaneous injection of botulinum Type A toxin into the eyelids has been reported to be an effective treatment. However, the reports are preliminary and there are no long-term follow-up studies.

It should be emphasized that blepharospasm may be the initial manifestation in an approximatly 50% of persons who later develop orofacial-cervical dystonia, or Meige's syndrome, which is a more incapacitating movement disorder. The differential diagnosis of blepharospasm is contained in Table XI.

TABLE XI
DIFFERENTIAL DIAGNOSIS OF BLEPHAROSPASM

Reflex blepharospasm
Essential blepharospasm
Meige's syndrome
Tardive dyskinesia and dystonia
Parkinson's disease
Huntington's disease
Wilson's disease
Encephalitis
Midbrain infaction or demyelination
Drugs: antipsychotics, antiemetics, anorectics, nasal decongestants, levodopa
Habit spasms and tics (Gilles de la Tourette's syndrome)
Hemifacial spasm
Facial nerve misdirection
Myokymia
Myotonia
Tetany
Tetanus
Schwartz-Jampel syndrome
Ocular disease
Seizures (absence, complex, partial)
Functional (hysterical)

Source: Ref 17

A candidate with a history of blepharospasm may be certified if 1) the cause has been determined to be essential; 2) the candidate is not on medication; 3) the candidate has not had a recurrence within one year after a corrective operation (botulinum injection procedures excluded); and 4) there are no ophthalmological complications from the procedure. Once certified the candidate should be evaluated annually for recurrence, and for the development of other movement disorders.

Orofacial-cervical dystonia (Meige's syndrome)

Meige's syndrome is dystonic movement disorder characterized by blepharospasm and involuntary spasms involving facial muscles, including the jaw, neck and, in some

cases, the larynx. These movements consist of blinking, facial grimacing, trismus, jaw opening, closing and protrusion, head turning and speech arrest. The onset of this condition is between the ages of 40 years and 70 years and is more common in women. Drug treatment is, for the most part, ineffective. Spontaneous remissions are rare.

Candidates with this disorder should be disqualified permanently.

Spasmodic torticollis

Spasmadic torticollis is a chronic form of focal dystonia with either repetitive or continous contractions of neck muscles that force the head laterally. Ten percent of persons with this disorder will develop dystonia in other body parts. The age of onset is similar to the other focal dystonias of adulthood.

Remissions are infrequent. Drug treatment as well as various surgical techniques have, for the most part, been ineffective. The results of biofeedback treatment are inconclusive.

Candidates with a history of this disorder should be disqualified permanently from flying.

Occupational dystonias

Writer's cramp is the most common of these disorders. This is characterized by spasms of the hand and forearm when attempting to write. Although initially confined to this activity, it may eventually involve other tasks, such as using tools. The disease usually does not spread to involve other parts. Spontaneous remissions are rare. Drugs, as well as biofeedback therapy, are not very effective.

Other occupational dystonias arise in typists, pianists, and percussionists.

Candidates with occupational dystonias may be certified provided 1) there is no other neurological abnormality or disease that would preclude certification; 2) the candidate is not on medication; and 3) the particular dystonia will not compromise flight

safety. Once certified, the candidate should be re-evaluated at periodic intervals.

Huntington's disease

Huntington's disease is an autosomal dominant disease with complete penetrance, whose hallmarks are mental and personality disturbance, choreic movements and positive family history. The prevalence rate in the US is 4 to 8/100,000. The onset is usually between 35 years and 40 years of age. The disease resembles parkinsonism. In approximately two thirds of cases, the first signs are chorea, and in the remaining third, personality changes or intellectual deterioration. The neurological signs initially are very subtle, consisting of minor choreic movements in the hands and face, personality changes and memory impairment. After five years, work performance decreases and the chorea is more pronounced. By five to 12 years individuals with Huntington's disease are unable to work. Death occurs usually 15 years after once. Suicide during the early stages, while the person still has insight about the disease, is not uncommon.

A review of Huntington's disease may be found in the chapter by Fahn.[18] Recently a test using recombinant DNA has been developed to detect the genetic marker for Huntington's disease. The clinical use of this test is still being debated.[19]

A candidate with a history of Huntington's disease should be disqualified permanently. Candidates with a family history of Huntington's disease should be evaluated periodically, preferably every year after the age of 35 years.

Cerebral palsy

A variety of static movement disorders, particularly choreoathetosis, may result from a number of pre- and perinatal diseases. In some individuals the movement disorder may be the only problem and these individuals are otherwise intellectually normal.[20] Aeromedical disposition rests with the determination whether the movement disorder is of such magnitude that it compromises flight safety.

Wilson's disease (hepatolenticular degeneration)

Although rare, this condition is important because if it is diagnosed early, the various movement disorders can be reversed by drug treatment. This disease is caused by an autosomal recessive defect in copper metabolism, causing abnormal deposition of this mineral in tissues, particularly the liver and central nervous system. In the vast majority of cases, the onset of symptoms is between the ages of 10 years and 25 years although persons whose onset is in the 4th and 5th decasdes have been reported.

The neurological signs and symptoms are variable, but movement disorders are the hallmark of this disorder. Intellectual and psychiatric disturbances are not uncommon. The Kayser-Fleischer ring in the cornea is pathognomonic of this disease. Treatment with penicillamine can effect a dramatic reversal of the neurological and hepatic abnormalities, if they are not far advanced. There are persons who have been treated with continuous penicillamine for at least 10 years, and who are virtually free of neurological and liver abnormlaities. The treatment is lifelong. Liver transplantation has been reported curative. A review of Wilson's disease can be found in the text by Scheinberg and Sternlieb.[21]

Candidates with Wilson's disease being treated with penicillamine may be certified provided 1) the neurological and psychological evaluations reveal no disqualifying defects; 2) there is no evidence of disqualifing liver abnormalities; and 3) there are no side effects from the treatment (penicillamine can produce a lupus-like syndrome, nephrosis, pemphigus and a mysathenic-like-condition). Once certified the candidate should be followed annually.

In investigations of families with Wilson's disease asymptomatic individuals are encountered. These persons are treated with penicillamine and remain asymptomatic as long as they take the medication. Candidates with asymptomatic Wilson's disease who are treated with penicillamine, may be certified, provided there are no side effects from

the treatment and they are followed at periodic intervals.

References

1. Stewart RM: Treatment of movement disorders, in Rosenberg RN (ed): Treatment of Neurological Diseases. New York, SP Medical and Scientific Books, 1979, p 406.
2. Marsden C, David T, Fahn S (eds): Movement Disorders. London, Butterworths International Medical Reviews, 1982.
3. Jankovic J (ed): Movement disorders, in Neurologic Clinics. Philadelphia, WB Saunders, Vol 2, No 3, 1984.
4. Joynt RJ (ed): Movement disorders, in Seminars in Neurology. New York, Thieme-Stratton, Vol 3, No 4, 1982.
5. Jankovic J: Parkinsonian disorders, in Appel SH (ed): Current Neurology. Philadelphia, John Wiley and Sons, 1984, pp 1-49.
6. Yahr MD: Parkinsonism, in Rowland LP (ed): Merritt's Textbook of Neurology, ed 7. Philadelphia, Lea and Febiger, 1984, Vol 113, pp 526-537.
7. Burton K, Calne DB: Pharmacology of Parkinson's disease, in Jankovic J (ed): Movement Disorders, in Neurologic Clinics. Philadelphia, WB Saunders. 1984, pp 461-472.
8. Young RR: Essential-familial tremor and other action tremors. Seminars in Neurology, New York, Thieme-Stratton, 1983. Vol 2, No 4, pp 386-391.
9. Larson TA, Calne DB: Essential tremor. Clinical Neuropharmacology 1983;6:185-206.
10. Kollier WC: Diagnosis and treatment of tremor, in Symposium on Movement Disorders. Neurologic Clinics. Philadelphia, WB Saunders, 1984, pp 499-514.
11. Auger R: Hemifacial spasm: Clinical and electrophysiologic observations. Neurology 1979;29:1261-1272.

12. Moller MB, Miller AR: Loss of auditory function in microvascular decompression for hemifacial spasm: Results in 143 consecutive cases. J Neurosurgery 1985;63:17-20.

13. Shapiro AK, Shapiro E: Tourette's syndrome: Clinical aspects, treatment and etiology in Yahr MD (ed): Movement Disorders in Seminars in Neurology. New York, Thieme-Stratton, 1983, Vol 2, No 4, pp 373-385.

14. Butler IJ: Tourette's syndrome in, Jankovic J (ed): Neurologic Clinics. Philadelphia, WB Saunders, 1984, Vol 2, No 3, pp 571-580.

15. Fahn S: Torsion dystonia: Clinical spectrum and treatment, in Yahr MD (ed): Movement Disorders in Seminars in Neurology. New York, Thieme-Stratton, 1982, Vol 2, No 4, pp 316-323.

16. Marsden CD: The focal dystonias, in Yahr MD (ed): Movement Disorders in Seminars of Neurology. New York, Thieme-Stratton, 1982, Vol 2, No 4, pp 324-333.

17. Jankovic J, Havins WE, Wilkens RB: Blinking and blepharospasm: Mechanisms, diagnosis and managment. JAMA 1982;248:3160-3164.

18. Fahn S. Huntington's disease and other forms of chorea, in Roland LP (ed), op cit, pp 517-521.

19. Merz B: Markers for disease genes open new era in diagnostic screening. Medical News JAMA 1985;254:3153-3159.

20. Carter S. Low NL: Cerebral palsy and mental retardation, in Rowland LP (ed), op cit, pp 355-362.

21. Scheinberg IH, Sternlieb I: Wilson's Disease. Philadelphia, WB Saunders, 1984.

Dementia

Introduction and Definition

Dementia is a "syndrome" manifested by deficits in cognition, memory and visual-spatial skills, as well as changes in language functions and personality. It has been proposed that at least three of these five deficits must be documented before a diagnosis of dementia can be made. However, these criteria have not been universally accepted. The Diagnostic and Statistical Manual (DSM III) of the American Psychiatric Association defines dementia as: 1) a loss of intellectual abilities of sufficient severity to interfere with social or occupational functioning; 2) memory impairment; 3) at least one of the following: (a) impairment of abstract thinking; (b) inability to find similarities and differences between related words, and difficulty in defining words in concepts and other similar tasks; (c) impaired judgment and other disturbances of high cortical function, such as aphasia, apraxia, agnosia, and constructional difficulty; (d) personality change; and (e) unclouded state of consciousness, that is, the person is alert and aware until the late stages of the syndrome (in contrast to delirium or intoxicated states, in which there is a fluctuating level of consciousness).

Recently, it has been customary to divide the dementias into subcortical and cortical types. The former is associated with impairment of memory and learning that is associated with slowness of intellectual function, inertia and apathy. In addition to these, with cortical dementias there are praxias and difficulties with language perception. Some diseases associated with subcortical dementia are progressive supranuclear palsy, Huntington's disease, and parkinsonism. The major disease causing cortical dementia is Alzheimer's. Recently this anatomical classification of the dementias has been challenged.[1]

Dementia can occur with many acquired and hereditary neurological diseases, and with systemic disorders, either as a presenting sign or during the course of the disease.

No age is spared and over 50 disorders may cause dementia. In diseases like Alzheimer's, dementia may be the only neurological abnormality, while in others it is associated with other neurological signs, for example, chorea in Huntington's disease, and gait and bladder disturbances in normal pressure hydrocephalus. Dementia may be a residual of an acute neurological insult such as head injury, CNS infection, or cerebrovascular accident. In the sections dealing with these diseases, the importance of obtaining thorough neuropsychological studies before a decision on flying status is made is discussed. In other diseases the onset can be insidious and slowly progressive. Consequently, the diagnosis will often be delayed. It is the slowly progressive dementias that pose the greatest medical problem.

Minimal criteria for diagnosis

It has been suggested by Mesulam[2] that, "when the performance of an individual falls at least one standard deviation below the average for his peer group, the presence of pathology is strongly suspected, especially if there are reasons that infer prior performance had been normal."

Since it is rare that a person with early dementia will seek medical attention, the FAA should consider this diagnosis when information provided by supervisors, other aircrew, or family members, or contained in medical or work records indicates diminished work performance, difficulties in adapting to changes, mood alterations, increased absenteeism, judgment errors, forgetfulness, preoccupation, increased use of alcohol, sleep disturbances, or general lack of interest. An applicant for certification should be referred to appropriate specialists, preferably a neurologist, psychiatrist, or neurospsychologist, not only to confirm the diagnosis and to determine the degree of impairment, but also to find a cause.

Evaluation

Neurological evaluation should include a complete personal and family history. At the time of the examination, a member of the applicant's immediate family should be present. The neurological evaluation should include the mini-mental status examination.[3] If this examination confirms or suggests the diagnosis, then an EEG and MRI, or at a minimum CT scan of the head should be performed. In addition, a complete blood count, sedimentation rate, serology for syphilis, serum electrolytes, thyroid profile, blood urea nitrogen, plasma glucose, vitamin B_{12} level and chest radiograph should be obtained. Depending on the results from the history, physical and neurological evaluation, other studies such as a cerebrospinal fluid examination may be required.

Neuropsychological studies

In Table I are listed the various neuropsychological studies commonly used to evaluate the cognitive deficits associated with Alzheimer, which may be used with other types of dementia. There is some disagreement about which tests are most appropriate for measuring a particular cognitive function. There is also a lack of random sample, population-based standards for many of these tests, so that abnormalities can be determined only by comparison with a control group, with adjustment for age, sex and educational background. According to the work group of the National Institute of Neurological and Communicative Disorders and Stroke/Alzheimer's Disease and Related Disorders Association (NINCD/ADRDA)[4] an individual's score that falls in the lowest 5th percentile of the range for the appropriate control group may be designated as "abnormal." Therefore, the results of neuropsychological studies cannot stand by themselves for making a diagnosis; they must be used in conjunction with the other clinical and laboratory findings. They provide confirmatory evidence and can be used as a baseline information for follow-up. They are also helpful in separating out depressed

persons whose symptoms suggest dementia.

TABLE I NEUROPSYCHOLOGICAL EVALUATION

The major cognitive processes that are impaired in the dementias, with examples of the kinds of tests used to assess these functions include:

- o Orientation to place and time, graded by a test such as the mini-mental status examination of Folstein and Folstein.
- o Memory evaluated by tests such as the free-recall test of concrete nouns, a 3-4 paired-associate learning test (verbal and nonverbal) by use of a recognition paradigm, the Recognition Span Test, and the Brown-Peterson Distractor Text (stopping the task when the patient fails or begins to produce the distractor instead of the stimulus trigrams).
- o Language skills tested by examination of verbal fluency of the semantic or category types, with the examiner writing repsonses, and by other tests such as the Boston Naming Test (preferably one of the abbreviated forms), the Boston Diagnostic Aphasia Examination, the Western Aphasia Test, and the Token Test with Reporter's Test.
- o Praxis evaluated by tests such as those in which the patient copies a drawing (cube, daisy, clock, or house) or performs the block designs subtest of the Wechsler Adult Intelligence Scale.
- o Attention monitored by tests as a reaction-time task or by the Continuous Performance Test.
- o Visual perception studied by use of a variety of tasks, such as the Gollin Incomplete-Pictures Test and the Hooper Test.
- o Problem-solving skills determined by tests such as the Wisconsin Card Sorting Test or the Poisoned Food Problem Task of Arenberg.
- o Social function, activities of daily living, assessed by methods similar to those described in the Philadelphia Geriatrics Center forms

source: Ref 4

Aeromedical disposition

Once a diagnosis of dementia is made, the first step in aeromedical disposition is to determine the cause, if it is progressive or non-progressive and if progressive, whether it is potentially reversible. The major conditions that can cause dementia are intrinsic brain disease, toxic-metabolic encephalopathies and depression.

Nonprogressive dementias

These usually are the result of acute neurological insults such as head trauma, CNS infections, and cerebrovascular accidents. They also may result from severe systemic illnesses such as cardiopulmonary arrest. Candidates with a history of nonprogressive dementia should be disqualified from flying unless: 1) the degree of dementia is minimal as determined by thorough neuropsychological studies, and judged not to compromise flight safety (a flight simulation test may be required); and 2) the disease causing the dementia is not disqualifying.

Progressive dementias

Approximately 80% of the progressive dementias are irreversible. In Table II are listed the remaining, potentially reversible dementias.

Roughly 66% of persons with these potential reversible dementias will improve with appropriate therapy. However, despite potential reversibility, most of the diseases listed in Table II, particularly the brain disorders, are still disqualifying. Dementia associated with endocrine abnormalities, particularly hypothyroidism, or deficiency states such as pernicious anemia, may be completely reversible if the diagnosis is made early in the course of the disease, before the dementia becomes moderate to severe.

TABLE II - REVERSIBLE CAUSES OF DEMENTIA

Depression (pseudodementia)

Intoxication

- Therapeutic drugs
- Alcohol
- Other substances (heavy metals, carbon monoxide)

Metabolic-endocrine derangements

- Renal failure
- Hyponatremia
- Volume depletion
- Hypoglycemia
- Hepatic failure
- Hypothyroidism
- Hyperthyroidism
- Hypercalcemia
- Cushing's syndrome
- Hypopituitarism

Brain disorders

- Stroke
- Subdural hematoma
- Infection (meningitis, neurosyphilis, abscess)
- Tumors (primary or metastatic)
- Normal pressure hydrocephalus

Cardiopulmonary disorders (congestive heart failure, arrhythmias, chronic obstructive pulmonary disease)

Generalized infections (tuberculosis, endocarditis)

Deficiency states (vitamin B_{12}, folate, niacin)

Miscellaneous causes

- Sensory deprivation (blindness, deafness)
- Hospitalization (isolation or anesthesia)
- Fecal impaction
- Anemia
- Remote effects of cancer

source: REF 5

Depression may also masquerade as dementia (pseudodementia). It is estimated that approximately 20% of individuals diagnosed as having dementia actually suffer depression. With appropriate psychiatric treatment, it may be reversible. The decision for certification of these persons is based on the criteria for depression found in the mental and behavioral section.

Candidates with a history of progressive dementia should be disqualified from flying unless: 1) the cause is correctable and intellectual functions have returned to pre-illness levels, as determined by thorough neurological and neuropsychological evaluations (a flight simulator test may be required); and 2) the causative disease or medications required to treat the condition are not disqualifying.

Alzheimer's disease

Alzheimer's disease, the most common cause of primary neurodegenerative dementia, is age-related, with the highest incidence occurring in the population over 65 years of age. It is estimated that approximately 1.3 to 1.8 million Americans over the age of 65 suffer from this disorder. Of aeromedical importance is that at least 80,000 people in their forties and fifties suffer from this malady. In fact, the first reported case by Alzheimer was a man only 51 years of age. Post-mortem investigations in both presenile (under 65 years of age) and senile (over 65 years of age) persons with dementia have shown the neurofibrillary tangles and plaques of Alzheimer's disease in half the cases. Thus, this disease is probably the most common cause of progressive dementia in the age group involved in recreational or commercial flying as well as support activities.

The disease effects both sexes about equally. The rate of progression of the disease is variable with marked incapacitation occurring within one year to 10 years after onset. The subject of heredity is somewhat controversial. A first degree relative of a proband with this disease has a several fold chance of developing the disease, particularly if the onset in the proband occurred at an early age. In addition, there are a

small number of families in which the disease presents as an autosomal dominant disorder.

Since there are no physiological or biochemical markers for this disease, the clinical diagnosis is mainly one of excluding the other causes of dementia. The definitive diagnosis ultimately rests with histopathologic identification either from brain biopsies or at autopsy. Biopsy is rarely used for diagnosis, the exception being unexplained progressive dementia in the younger age groups.

The NINCDS/ADRDA workgroup has recently suggested the criteria listed in Table III for the clinical diagnosis of Alzheimer's disease.[4] Additional references are articles by Terry and Katzman[6] and Khachaturian.[7]

The aeromedical disposition of persons with progressive dementia, which includes Alzheimer's, has already been discussed. Because of the hereditary aspects, applicants with a stong family history of dementia should be followed at frequent intervals.

Pick's disease

Pick's disease is a rare cause of presenial dementia, particularly in the United States, with the onset of symptoms occurring most frequently in the 6th decade of life. Pathologically, the disease is characterized by lobar atrophy, particularly of the frontal and temporal lobes. The signs and symptoms are similar to those with other dementing diseases. The disease is slowly progressive and there is no cure. It has been reported in families.[8-9]

TABLE III

Criteria for clinical diagnosis of Alzheimer's disease

I. The criteria for the clinical diagnosis of PROBABLE Alzheimer's disease include:

dementia established by clinical examination and documented by the Mini-Mental Test,[1] Blessed Dementia Scale,[2] or some similar examination, and confirmed by neuropsychological tests;

deficits in two or more areas of cognition;

progressive worsening of memory and other cognitive functions;

no disturbance of consciousness;

onset between ages 40 and 90, most often after age 65; and

absence of systemic disorders or other brain diseases that in and of themselves could account for the progressive deficits in memory and cognition.

II. The diagnosis of PROBABLE Alzheimer's disease is supported by:

progressive deterioration of specific cognitive functions such as language (aphasia), motor skills (apraxia), and perception (agnosia);

impaired activities of daily living and altered patterns of behavior;

family history of similar disorders, particularly if confirmed neuropathologically; and

laboratory results of:

normal lumbar puncture as evaluated by standard techniques,

normal pattern or nonspecific changes in EEG, such as increased slow-wave activity, and

evidence of cerebral atrophy on CT with progression documented by serial observation.

III. Other clinical features consistent with the diagnosis of PROBABLE Alzheimer's disease, after exclusion of causes of dementia other than Alzheimer's disease, include:

plateaus in the course of progression of the illness;

associated symptoms of depression, insomnia, incontinence, delusions, illusions, hallucinations, catastrophic verbal, emotional, or physical outbursts, sexual disorders, and weight loss;

other neurologic abnormalities in some patients, especially with more advanced disease and including motor signs such as increased muscle tone, myoclonus, or gait disorder;

seizures in advanced disease; and

CT normal for age.

IV. Features that make the diagnosis of PROBABLE Alzheimer's disease uncertain or unlikely include:

sudden, apoplectic onset;

focal neurologic findings such as hemiparesis, sensory loss, visual field deficits, and incoordination early in the course of the illness; and

seizures or gait disturbances at the onset or very early in the course of the illness.

V. Clinical diagnosis of POSSIBLE Alzheimer's disease:

may be made on the basis of the dementia syndrome, in the absence of other neurologic, psychiatric, or systemic disorders sufficient to cause dementia, and in the presence of variations in the onset, in the presentation, or in the clinical course;

may be made in the presence of a second systemic or brain disorder sufficient to produce dementia, which is not considered to be *the* cause of the dementia; and

should be used in research studies when a single, gradually progressive severe cognitive deficit is identified in the absence of other identifiable cause.

VI. Criteria for diagnosis of DEFINITE Alzheimer's disease are:

the clinical criteria for probable Alzheimer's disease and

histopathologic evidence obtained from a biopsy or autopsy.

VII. Classification of Alzheimer's disease for research purposes should specify features that may differentiate subtypes of the disorder, such as:

familial occurrence;

onset before age of 65;

presence of trisomy-21; and

coexistence of other relevant conditions such as Parkinson's disease.

source: REF 4

References

1. Whitehouse PJ: The concept of subcortical and cortical dementia: Another look. Ann Neurology 1986;19:1-6.
2. Mesulam MM: Dementia: Its definition, differential diagnosis, and subtypes. JAMA 1985;253(17):2559-2561.
3. Folstein M, Folstein S, McHugh PR: Mini-mental status. A practical method for grading the cognitive state of patients for the clinician. J Psychiatric Res 1975;12:189-198.
4. McKhann G, Drachman D, Folstein M, et al: Clinical diagnosis of Alzheimer's disease. Report of the NINCDS-ADRDA workgroup. Neurology 1984;34:939-944.
5. Beck JC, Benson DF, Scheibel AB, et al: Dementia in the elderly: The silent epidemic. Ann Intern Med 1982;97:231-241.
6. Terry RD, Katzman RG: Senile dementia of the Alzheimer type. Ann Neurol 1983;14:497-506.
7. Khachaturian ZS: Diagnosis of Alzheimer's disease. Arch Neurol 1985;42:1097-1105.
8. Wechsler AF: Pick's disease: A clinical, computed tomographic and histologic study with Golgi impregnation observations. Arch Neurol 1982;39:287-290.
9. Groin JJ: Hereditary Pick's disease. Brain 1982;105:443-459.

Other references

Wells CE (ed): Dementia, ed 2. Philadelphia, FA Davis Co, 1977.

Katzman R, Terry RD (eds): Neurology of Aging. Philadelphia, FA Davis Co, 1983.

Mesulam M: Principles of Behavioral Neurology. Philadelphia, FA Davis Co, 1985.

Mayeux R, Rosen WG (eds): The dementias in, Advances in Neurology, New York Raven Press. Vol 38, 1983.

Headache and Cranial Neuralgias

Introduction

Headache is one of the most frequently reported medical symptoms. It is rare to find a person who has never experienced a headache. As noted by Goldstein anbd Chen, approximately 75% of the general and patient populations experience headaches; of these, around 18% will have severe and 9% very severe, recurrent headaches.[1] Headaches can be associated with both acute and chronic structural neurological diseases, such as acute cerebral vascular disease, hemorrhage, infections, and brain tumors. More often the neurological evaluation of persons who present with a history of recurrent headaches is normal and the diagnosis is based primarily on personal and historical information.

Approximately 1% of the general population may have the onset of disabling headaches, including migraine, each year. Females are more predisposed to develop headaches, and suffer more severe pain than males. The highest frequency of recurrent headaches is reported between the ages of 15 and 54 years.[2]

In a study of women with migraine, only 23% had visited a physician because of their headaches during the previous year, and 46% had not sought medical advice about their headaches at any time in their lives. These percentages are most likely much higher for males.[3]

Therefore, headaches present a most difficult and challenging aeromedical problem. Not only can the pain interfere with proficiency of performing airman duties, but the transient neurological deficits, particularly with migraine, can jeopardize safety. In addition, the large number of prescription and nonprescription medications that are available for treatment can interfere with performance.

Appenzeller et al presented the FAA with an in-depth review of migraine headache and related conditons.[4] In this section will be additional information on those

conditions that cause the major problems for aeromedical disposition, such as benign headaches that suggest structural neurological diseases, and the pr ... with drug treatment. Only those recurrent headaches not secondary to structura ... disease will be covered. Reviews of headache can be found in the publications of Packard,[5] Critchley, et al,[6] Ziegler,[7] Diamond and Dalessio,[8] and Selby.[9]

After reviewing an applicant's records the FAA should determine the type of headache and its frequency, severity and treatment (prescribed or nonprescribed), and determine that structural CNS disease has been ruled out as a cause of the headache. If the information on these points is lacking or unclear, then a complete neurological evalution is required before a disposition can be recommended.

Classification

The classification and definition of headaches proposed by the National Institute of Neurological Disease and Blindness (NINDB) in 1962 is still, for the most part, used today (see attachment). Since there are no biological markers for headaches, this classification provides physicians with a set of useful guidelines. However, errors in diagnosis are not uncommon since the diagnoses are based primarily on history. They are dependent on the person's ability to describe symptoms, as well as the bias and judgment of the physician. This problem is documented by the study of Hirayama and Ito, who found prevalence rates of migraine ranging from 5% to 34% in different Japanese districts.[10] In all likelihood, this disparity is not due to different population characteristics in the districts, but to varying physicians' interpretations of histories.

Recently, Ziegler addressed two other problems related to migraine in particular and to headache in general: 1) the accuracy of diagnosing an individual attack and; 2) the accuracy with which a eprson can be identified as having a disease entity.[11] He mentions that the neurological symptoms and temporal profile of a classic migraine attack are relatively easy to determine from the history. However, criteria for the

common migraine episode are less specific since there is a great deal of variation in the location of the headache and associated symptoms during individual attacks in the same person. He also raises the as yet unanswered question as to the occurrence of several types of headache in the same individual. That is, how many people with common migraine may also have one, two, three or more attacks of classic migraine or muscle contraction headaches? If they do have different forms of headache is there a common mechanism or different mechanisms? These questions can be answered only by future physiological and biochemical investigations.

Because of these problems with the headache classification guidelines, the FAA should place more emphasis on determining the frequency and severity of headaches and the presence of neurological deficits, rather than on definitive diagnoses, which is a more holistic approach to the problem.

Minimum criteria for diagnosis

Because headache is such a common complaint, it is difficult to establish minimal criteria for diagnosis. Since many people with recurrent headaches never consult a physician, the emphasis should be placed on ancillary information that may be helpful in identifying the headache victim or potential victim: 1) history of recurrent heaches in childhood; 2) strong family history of headaches, particularly "migraine;" 3) history of transient visual or neurological symptoms; 4) consultation(s) with a physician because of headaches; 5) a history of frequent use of over-the-counter drugs; 6) absenteeism from work; and 7) change in personality or work performance.

Childhood headaches

Recurrent headaches in children are not uncommon. Sillanpaa reported a 6% incidence of headaches occurring at least once or more per month in children 7 years of age.[12] The greatest percentage of headaches in children are either muscle contraction

or migraine. The prevalence of the latter is estimated to be approximately 4% in children between 7 and 15 years of age.[13]

It nas been reported that common migraine is more frequent in children than is classic migraine. However, Hockaday reported a 59% incidence of the latter in his 122 childhood migraneurs.[14] Basilar artery and acute confusional migraine are more common in children and adolescents that in the adult. In addition, the migraine syndrome precipitated by minor head injuries is more common in chidlren (see section on head trauma). A history of migraine is found in 73% of mothers and 21% of fathers of children with this disorder.

The incidence of abnormal EEGs is higher in children than adults with migraine. Although most of the abnormalties are non-specific, consisting of diffuse slow-wave patterns, 17 of the 64 children with migraine studied by Prensky and Sommer showed spike or spike and wave paroxysms. In a follow-up period ranging from three to nine years, seven of these children had no personal or family history of epilepsy.

The relationship between migraine and epilepsy remains controversial. Some studies indicate an increased incidence of seizures in persons with migraine as well as family members, while others do not.[16] The frequency of reported seizures in migraine has varied between one percent and 25%. Recently, Seshia et al reported seizures occurring in 54 of 240 children with migraine. Nineteen of the 54 experienced partial, compex seizures not related to a migraine attack.[17] Therefore, it appears that children with migraine are predisposed to seizures. These may occur independent of a migraine attack or during one. However, the relationship between migraine and seizures is still controversial.

The prognosis for childhood migraine is generally favorable. Approximately one third become symptom-free by puberty, and one third remain unchanged. However, not uncommon is the person with childhood migraine who undergoes a remission during the teenage years, and experiences a relapse in early adulthood.

Studies on the prognosis of muscle contraction headaches in children that are severe or frequent enough to warrant medical attention are lacking. However, one would suspect that these would likely continue into adulthood because of the probability of persistence of the psychogenic determinants of these headaches.

Aeromedical disposition

An applicant with a childhood history of migraine is qualified for certification unless: 1) the migraine was associated with epilepsy; 2) the candidate has been left with a residual neurological deficit (complicated migraine), such as a persistent visual or motor deficit, which by itself is disqualifying; 3) the candidate is taking antimigrainous prophlactic medication; or 4) the last attack of classic or complicated migraine occurred within 5 years prior to application. Candidates with a past history of childhood migraine who are certified should be evaluated on a periodic basis in order to check for recurrence.

An applicant with a childhood history of other types of headaches, particularly muscle contraction headaches, may be certified if: 1) there is no psychiatric contraindication; 2) the candidate is not taking prophylactic drugs; and 3) if the headaches persist, they are infrequent and not severe.

The migraine complex in adults

Introduction

The prevalence of migraine is approximately 9% for males and 16% for females. Because of problems with definition, these percentages may be somewhat high. Migraine is generally considered a hereditary disorder, although the mode of inheritance has not been clarified. Forty-six percent of migraineurs have a positive family history of the disorder in either parents or siblings. When grandparents are included, the percentage raises to 55%. The familial association is higher for children with migraine than for

adults. Ninety percent of persons with migraine suffer the first attack before the age of 40 years, and 20% have thir first attack in the first decade of life. A number of precipitants to attacks have been identified, including foods, stress, hormonal changes, oral contraceptives, and bright lights. Drugs such as reserpine and vasodilators may also precipitate attacks in an already migrainous person. However, most of the time attacks occur without identificable predipitants.

The definitions of the various forms of migraine (common, classic, cluster) are contained in the AMA's 1979 report to the FAA. The term, complicated migraine, has crept into the literature and has caused some confusion. Selby uses it to describe a rare migraine variant in which visual or focal neuological deficits outlast the headache and may even become permanent.[9] Others use it to refer to migraine with dramatic neurological deficts, and as such, the term overlaps with classic migraine. Persons with persistent defict of vision, ophthalmoplegia, hemiplegia, and acute confusional (dysphrenic) migraine are included in Selby's complicated migraine syndrome. Persistent neurological deficits have been reported in migraine, usually in the older age group.[18]

Because of the high prevalence rate of migraine in the general population, and the varying presentation of attacks in the same individual, and among other migraneurs, it is difficult to establish guidelines that would allow migraineurs to pilot aircraft, yet not unduly compromise flight safety. For aeromedical purposes, it is best to consider all types of migraine as one entity, and base certification on the frequency and severity of attacks, and associated neurological deficitis, rather than on specific diagnoses.

Frequency and severity

The frequency of attacks is extremely variable from one migraineur to another. There are no recent data that address this issue. Selby[9] states that 50% of individuals with common migraine will experience one to four headaches per month, 10% to 15% of these people will experience less than 12 attacks per year, and 30% to 50% will

experience episodes as frequently as three times per week. With classic migraine the attack rate is much lower, usually less than 10 episodes per year. Some persons may experience only one to three episodes per year and is not uncommon for symptom-free periods longer than one year to be reported. Data on the frequency of attacks in persons with complicated migraine are not available.

The severity of an attack also varies within the same person. Over sixty percent of individuals with common migraine will be pain free in less than 24 hours, 16% within 48 hours, and in the remaining the headache can persist for longer than 48 hours. With classic migraine the headache rarely exceed 8 to 12 hours. Assessment of severity can be difficult since interpretation and reaction to pain are extremely variable.

In assessing the severity of migraine attacks, the physician should be mindful of and obtain information about the following: 1) migraines are more severe in women; 2) migraines are more severe at onset of sleep; 3) the intensity and frequency may be inferred from the number and amount of drugs used to obtain relief; 4) the intensity and frequency may be inferred from sick leave and absenteeism; 5) there may be associated, nonfocal, neurological symptoms such as nausea, vomiting, and lightheadness; 6) there may be increased sensitivity to light, noises and odors; 7) there may be problems with concentration, slowness of thought processes, and other problems with cognition; and 8) there may be prolonged and disabling postheadache malaise.

Associated neurological deficits

The following major focal neurological deficits are associated with migraine (classic or complicated): 1) visual; 2) motor, sensory, language disorders; 3) vertigo; 4) syncope; and 5) acute confusional states.

Visual

Visual disturbances are the most frequent neurological problems that occur with

classic migraine. Also during the course of common migraine, a person may experience occasional episodes of visual disturbances. This fact has raised the question that all migraine may be one entity.[2]

The visual symptoms may also occur independent of a headache, which is called migraine equivalent. The visual symptoms with migraine usually include positive or negative scotomata, visual field defects, and amaurosis from retinal migraine. Visual hallucinations, illusions and disorders or cortical visual function have been occasionally described.

The attacks are generally not stereotyped. The visual aura prodrome (aura) usually lasts ten to 30 mintues and is followed by the migraine headache. On occasion visual symptoms persist into the headache phase. The visual deficits may be permanent.[19]

A rare visual accompaniment of migraine is involvement of the extraocular nerves. Most common is involvement of the 3rd nerve, then the 6th, and finally the 4th nerve. The headache usually develops first and the extraocular movement deficit usually lasts for 1 to 4 weeks thereafter. Therefore, this type of migraine is classified as complicated.

Sensory, motor and language symptoms

Focal sensory, motor and language symptoms can occur either during the prodromal period, during the headache, or after the headache has dissipated. Sensory symptoms occur in approximately 50% of people with migraine. They usually begin unilaterally in a hand and progress to involve the entire arm and face (cheiro-oral migraine). When severe, function of the extremity can be impaired.

Focal motor deficits during the prodrome are less frequent and severe, although if the prodromal period is prolonged, profound weakness may occur. Dysphasic and dysarthric symptoms may also occur during the prodrome.

Focal sensory, motor or language deficits that persist after the headache has abated are classified in the category of complicated migraine. Sporadic or familial hemiplegic migraine has received the most attention. Although rare, the impairment it causes can be devastating. The paresis can shift from side to side in different attacks. Although it may be a single event, it usually recurs. Persons with this disorder have a history of common migraine. It is more frequent in children and adolescents.

Vertigo

True vertigo occurs in approximately 10% of migrainous persons during the prodromal phase. It usually lasts less than 15 minutes. It may be the only aura, or it may be associated with the other neurological symptoms, particularly those caused by the brain stem ischemia, of basilar artery migraine. This form of migraine is more frequent in children and adolescents. Unlike hemiplegic migraine, the symptoms clear before the headache begins. Drop attacks in migraine have been attributed to this form of migraine.

Syncope

Although loss of consciousness can occur in basilar artery migraine, it is usally independent of this disorder. Females are more predisposed than males. It occurs most often in the headache phase. The exact incidence is not known.

Migraine equivalents

Auras not followed by headache are referred to as migraine equivalent. These occur primarily in classic and less frequently in common migraines. The symptoms are most often visual or vertiginous. Transient disturbances of motor, sensory or speech functions are less common and should alert the physician to other causes.

As people age, the auras of migraine, particularly visual auras, may persist

without headaches. Middle aged women with recurrent, short, vertiginous episodes may have a history of past migraine attacks. Migraine equivalent s are infrequent but they may be confused with cerebral vascular disease.[20]

Mitral valve prolapse

Mitral valve prolapse has been reported in approximately 21% of persons with vascular headaches, which is significantly higher than its occurrence in the general population.[21]

MRI or CT scans

Selby[9] summarizes the results of investigations on the CT evaluations of migraineurs during an acute attack as well as during asymptomatic periods. Most of the studies were on selected groups of individuals, usually those suffering from severe migraine. Therefore, the true incidence of CT abnormalities in the general migraine population is unknown. The CT findings that have been reported are mild cerebral edema during acute attacks, cerebral infarction, and focal or generalized cerebral atrophy. In some cases the radiographic signs of infarction did not correlate with the symptoms, most likely indicating a previous event.

CT scans are not routinely performed in a migraineur unless the physician suspects another cause for the person's headaches. In the event that one of the aforementioned abnormalities is noted and the subject otherwise satisfied the criteria for certification, a complete neurological evalution should be performed.

Drug treatment of migraine

In Table I and II are listed the drugs most often used in the treatment of acute attacks, as well as prophylaxis, of migraine. Recent reports indicate that calcium-channel blockers such as verapamil may be beneficial.

TABLE 1 *Drugs for effective treatment of acute attack of migraine headache*[a]

Drug	Form	Dosage	Route	Action	Side effects	Contraindications
Gynergen (Sandoz) Ergotamine tartrate, 0.05 mg/cc	Ampul	1–2	Parenteral	Vasoconstrictive	Nausea, vomiting, weakness in legs, muscle pains in extremities, numbness and tingling in fingers and toes, angina-like precordial distress and pain, transient tachycardia or bradycardia, localized edema and itching	Septic infections, vascular diseases (e.g., marked arteriosclerosis, coronary artery disease, thrombophlebitis, Raynaud or Buerger syndrome, pregnancy)
Ergomar (Cooper) Ergotamine tartrate, 2 mg	Tablet	1–3	Sublingual	Same		
Medihaler-ergotamine (Riker) Ergotamine tartrate, 0.35 mg per dose	Aerosol	2–6	Oral inhalation	Same		
D.H.E. 45 (Sandoz) Dihydroergotamine Methanesulfonate, 1 mg/cc	Ampul	1–2	IM or IV	Same		
Cafergot P-B (Sandoz) Ergotamine tartrate, 1 mg Caffeine, 100 mg l-Belladonna alkaloids (belladonine), 0.125 mg Phenobarbital sodium, 30 mg	Tablet	2–6	Oral	Vasoconstrictive Antispasmodic Sedative	Nausea, vomiting, numbness and tingling of hands and feet, muscle pain in thighs and neck, abdominal pain, prostration, dryness of mucous membranes and skin, drowsiness	Septic infections, vascular diseases, coronary sclerosis, history of angina pectoris, pregnancy, hypertension, impaired renal or hepatic function, glaucoma
Ergotamine tartrate, 2 mg Caffeine, 100 mg l-Belladonna alkaloids, 0.25 mg Pentobarbital, 60 mg	Suppository	1–2	Rectal			
Wigraine (Organon) Ergotamine tartrate, 1 mg Caffeine, 100 mg l-Belladonna alkaloids, 0.1 mg Phenacetin, 130 mg	Tablet or suppository	1–2	Oral or rectal	Vasoconstrictive	Same as for ergotamine tartrate + dryness of mucous membranes and skin	Same as for ergotamine tartrate + glaucoma
Migral (Burroughs Wellcome) Ergotamine tartrate, 1 mg Caffeine, 50 mg Cyclizine hydrochloride, 25 mg	Tablet	2–4	Oral	Vasoconstrictive Antiemetic	Same as for Wigraine	Same as for ergotamine tartrate
Midrin (Carnick) Isomethheptene mucate, 65 mg Dichloralphenazone, 100 mg Acetaminophen, 325 mg	Capsule	2–5	Oral	Vasoconstrictive Analgesic Sedative	Drowsiness, dizziness, palpitations, weakness	Glaucoma, severe renal disease, hypertension, organic heart disease, hepatic disease, patients on MAO inhibitor therapy

TABLE 2. *Drugs for prophylactic treatment of migraine headache*

Drug	Dosage
Those acting as competitive serotonin inhibitors by simulating action of serotonin on receptor sites	
Methysergide maleate (Sansert)	2 mg t.i.d.
Cyproheptadine (Periactin)	4–16 mg daily as tolerated
Those producing vasoconstriction, sedation; antispasmodic	
Ergotamine, phenobarbital, and belladonna	
Bellergal	1 tablet q.i.d.
Bellergal-S	1 tablet b.i.d.
Those preventing vasodilatation by blocking beta-adrenergic receptors on blood vessels	
Propranolol hydrochloride (Inderal)	40–120 mg daily in divided doses
Those inhibiting uptake of norepinephrine and serotonin, including tricyclic depressants	
Imipramine hydrochloride (Tofranil)	(50–150 mg in divided doses)
Amitriptyline hydrochloride (Elavil)	(or once daily at bedtime)
Those having a stimulating effect on arterial alpha receptors	
Clonidine (Catapres)	0.1 mg b.i.d. or t.i.d. (dosage may be increased gradually by increments of 0.1 mg)
Miscellaneous group	
Tranquilizers	
Sedatives	
Muscle relaxants	
Heparin	
Lithium carbonate	
Levodopa	
Bromocriptine	
Indomethacin	
Prednisone	
Estrogens	
Serotonin precursors	
Papaverine hydrochloride	
Anticonvulsants	

source: REF 22

With the exception of mild, non-narcotic analegesics, the drugs for the treatment and prevention of migraine are disqualifying for flying.

Aeromedical disposition

It is extremely difficult to establish strict medical certification criteria for a neurological condition so common and varied as migraine. The data presented in this section have come from investigations that have relied on historical information. There are no recent studies on the natural history of this disease that can be used for aeromedical disposition. Until such time when biological or physiological markers are discovered that will allow us to predict with greater accuracy the natural history of this malady, we will have to rely on these data, imperfect as they may be. With this in mind, we propose the following guidelines:

Acute migraine: An airman with an acute migraine attack should not be allowed to engage in airman's duties until he or she has recovered completely and has been granted clearance by a physician.

Recurrent headaches without focal neurologcial deficits (common migraine): Applicants with a history of more than four to six severe headaches per year should be disqualified. This is an artibtrary figure. There may be persons with more frequent but less disabling headaches who, after neurological evalution, may be granted a certificate (an example of this exception is a woman with menstrual migraine). Candidates using any of the drugs listed in Table II prophylactically should be permanently disqualified.

Focal neurological deficits: Candidates with a history of any focal signs or symptoms should be disqualified permanently from flying. This includes symptoms that occur prior to or during the headache, or persist afterward. A possible exception to this would be applicants who only rarely experience (perhaps once every 2 years) classic visual prodromata. These candidates must receive neurological clearance before certification. This also applies to applicants who suffer from nondisqualifying common

migraine who, on rare occasions, have focal symptoms.

Candidates with a history of vertigo or syncope with attacks of migraine should be disqualified permanently, unless these sympotms are extremely infrequent. Candidates with a history of acute confusional or vertebral-basilar artery migraine should be permanently disqualified. Candidates with a history of seizure activity associated with a migraine episode or occurring at any other time should be permanently disqualified (see section on epilepsy).

Persons with migraine who otherwise satisfy the neurological criteria and have mitral valve prolapse should satisfy the cardiovascular criteria for this entity. Candidates with migraines and abnormal CT scans who otherwise satisfy the neurological criteria should receive clearance from a neurologist before certification. Finally, candidates with a diagnosis of migraine equivalent should be disqualified permanently unless their episodes are extremely rare.

Cluster headaches

Cluster headaches occur predominantly in men. Mean age of onset is about 30 years. The prevalence rate has been estimated to be 0.45% for males and .08% for females. The signs and symptoms are shown in Table III.

Despite their short duration, cluster headaches are perhaps the most severe and disabling. Several attacks may occur in a 24 hour period and not infrequently they may awaken the individual from sound sleep. In remission periods, which may last one year or longer, the individual is entirely normal. Some perosns will not experience remissions and suffer from cluster headaches almost daily (chronic migrainous neuralgia). This condition has been treated with retrogasserian injection of alcohol.

Persons with clusters may also experience typical migraine attacks. An increased incidence of peptic ulcer disease has been reported and it has been suggested that coronary artery disease may also be more prevalent in persons with this disease.

TABLE III

PROFILE OF CLUSTER HEADACHE ATTACK

Cluster periods	6–12 weeks*
Remission periods	12 months*
Attacks	
Frequency	1–3 days*
Duration	45 minutes*
Location	Unilateral
	Oculotemporal
Severity	Excruciating
Character	Boring, nonthrobbing
Associated symptoms	Unilateral
	Lacrimation
	Rhinorrhea
	Partial Horner's
Induction	Vasodilators, alcohol, REM†
Behavior in attack	Walking, sitting, rocking

*Average values
†REM = Rapid eye movement

source: REF 23

The treatment modalities both for the acute phase as well as for prophylaxis are similar to those for migraine. A short course of prednisone is today the most popular treatment regimen. Recently, an association between cluster headache and herpes simplex has been postulated.[24] In all likelihood, acyclovir will be used in the future to manage these attacks.

A more chronic variant of cluster headache is chronic migrainous neuralgia, which is severely debilitating. The surgical treatment of this condition has recently been described.[25]

Aeromedical disposition

For persons with attacks of cluster headaches that occur less frequently than one per year: Candidates with a history of cluster headaches should not engage in aviation activities during the headache period. They may return to flying during the cluster-free period provided they are not taking prophylactic medication.

Candidates with more frequent cluster periods should be disqualified permanently. Applicants with chronic migrainous neuralgia should be disqualified permanently from flying. If they are treated surgically, they must be symptom-free for a period of one year before they may be considered for certification. A neurological consultation is required.

Muscle contraction headaches

This is the most common cause of recurrent and chronic headaches. Most people experience infrequent muscle contraction headaches, which are self-limited and not severe. The most frequent cause of recurrent or chronic muscle contraction headaches is mental problems, particularly depression, which is thought to be the cause of headaches in 84% of cases.[26] The headache is located at points of insertion around the head, particularly at the mastoid line, superior insertion of the temporalis, and along the

eyebrows.[27]

The most reliable historical features of this type of headache are: 1) a sensation of pressure or tightness with pain, which is worse in the posterior head and neck region; 2) pain that increases as the day progresses; and 3) a preheadache history of anxiety or tension. Nausea and vomiting may accompany the headache. Muscle contraction headaches can occur in persons with migraine. In Kudrow's study,[28] (Table IV) persons with subacute or chronic muscle contraction headaches often had a positive history of migraine. Persons with chronic muscle contraction headaches present many problems in pain management and their rehabilitation is not very satisfactory.

On occasion, headaches resembling the muscle contraction type may be caused by disease of the cervical spine and contiguous structures, as well as intracranial neoplastic, developmental and inflammatory diseases. These can be ruled out by appropriate neurological studies, including radiographs and CSF analysis.

Aeromedical disposition

The major aeromedical problems are determining the frequency and severity of the headaches, ruling out organic causes, identifying predisposing psychiatric conditions, and determining if the candidate is abusing medication. Airmen with infrequent, acute, severe muscle contraction headaches should not engage in aviation activities during the acute attack. Airmen with recurrent muscle contraction headches should not be certified if they occur more than once every one to two months (this is an arbitrary figure) and are severe. Certification of these airman is dependent on a normal neurological examination, no evidence of a disqualifying psychiatric condition and no evidence of chronic drug abuse. Airmen with chronic muscle contraction headaches should be disqualified permanently.

TABLE IV

CHARACTERISTICS OF SCALP MUSCLE CONTRACTION HEADACHE

Features	Scalp muscle contraction headache		
	Acute[a]	Subacute[b]	Chronic
Frequency	Occasional	2–4/Wk	Daily
Duration	Hours	1 Day	Constant
Intensity	Dull	Dull	Dull
Location	General	Front-occ	Front-occ
Laterality	Bilateral	Bilateral	Bilateral
Associated symptoms	None	None	None
Migraine status	Negative	Positive	Positive
Analgesic use	Occasional	Often	Excessive
Response to			
Analgesics	Excellent	Fair	Poor
Amitriptyline	—	Excellent	Good

[a] Data obtained from general medical population.
[b] Data obtained from headache clinic population.

source: REF 28

Acute headaches simulating structural CNS disease

From time to time the FAA will encounter a candidate with a history of acute headaches related to a specific event, particularly exertion. These headaches are severe, but brief. Because of these features, they suggest the presence of neurological disease, such as a subarachnoid hemorrhage, or disease causing increased intracranial pressure, like brain tumors. They are more frequent in males and in persons over 40 years of age. Exertional headaches have been subdivided by Diamond and Medina into three categories based upon activity: 1) those activities that increase intrathoracic pressure, such as coughing, defacating, and lifting; 2) those activities that raise blood pressure, such as orgasm and excessive exercise, and; 3) those activities that cause traction of intracranial structures, such as jumping and head rotation.[29]

In addition to the short duration headaches, a prolonged benign exertional headache has been recently described by Diamond and Medina. The mean duration of this type of headache is approximately four hours. Exertional headaches frequently occur in persons with migraine and cluster headaches.

The prevalence of associated neurological disease varies according to the type of exertional headache. With cough headache, 6 of 27 persons had organic disease. Of the 21 persons with neurological disease, more than one half experienced improvement while in the remaining cases, the symptoms persisted.[30]

In Rooke's study of 103 patients with exertional headaches, 10 subsequently developed anomalies of the posterior fossa, such as an Arnold-Chiari malformation. This study was performed in the pre-CT scan era. Therefore, the prevalence of structural neurological disease associated with exertional headaches may be higher. The natural history of exertional headaches is not known.[31]

Migraine may occur with acute effort, particularly at high altitudes. Vascular headaches in untrained athletes are not uncommon. Persons with exertional headaches may also suffer from underlying endocrinologic disease, such as a pheochromocytoma.

The initial coital-induced headache simulates a spontaneous subarachnoid hemorrhage because of its explosive nature. However, it is of short duration and not associated with vomiting, and the individual recovers without residua. It is extremely variable in frequency and runs a benign course.

Aeromedical disposition

Candidates with exertional headaches may be certified provided that: 1) CNS disease has been ruled out (a neurological consutation and CT scan or MRI scan of the head must be obtained); 2) there is no associated disqualifying medical condition, such as hypertension or pheochromocytoma; 3) if the candidate also has migraine, he or she should satisfy the criteria for that entity; and 4) the attacks of acute headaches are infrequent and would not jeopardiaze flight safety.

Drug abuse and headaches

One of the more challenging areromedical problems is the identification of the airmen who resorts to the daily or frequent use of over-the-counter or prescribed analgesics to control headaches. Detecting these individuals is not easy, particularly in the early stages of abuse. Usually they first seek medical help when their pain has become refractory to drugs and side effects have occurred. At this point treatment and management become exceedingly difficult. It is estimated that approximately 50% of persons who are referred to a headache clinic have chronically overindulged in various types of pain relieving drugs.[32]

Obtaining an accurate history of abuse is very difficut. This is probably more of a problem in aviation, since detection usually means suspension from flying activities. That a pilot is abusing drugs may only be discovered when a complication has occurred, for example, a gastrointestinal hemorrhage from overuse of aspirin, nephropathy with phenacetin, or symptoms of withdrawal from barbiturate preparations. A decrease in

work performance, change in personality and increased absenteeism may signify a drug abuse problem. In some cases, concerned family members or fellow aircrew members may report the problem to a physician. If the FAA suspects a drug problem in the management of headaches, appropriate blood and urine screens should be obtained.

In Table V the analgesics used by 200 patients with chronic scalp muscle contraction headache are listed. Thirty-eight percent of these patients used analgesics combined with sedatives, tranquilizers or muscle relaxants; 31.5% used analgesics-narcotic combinations; 28% used mixed or single antipyretic analgesic compounds, and 24% used propoxyphene or pentazocine chronically. Approximately six pills were consumed on the average each day.

In Table VI are listed the most common non-prescribed analgesics that persons may take for chronic headaches. Table VII contains a selected list of the most commonly prescribed analgesics.

In addition to acute CNS toxic side effects that are dose related, central nervous system symptoms such as mental changes, depression, impairment of psychomotor performance, decreased reaction times, and sleep disturbances can occur with chronic use. Withdrawal symptoms usually occur with both barbiturate and nonbarbiturate sedatives. One of the more commonly prescribed analgesics for headaches is Fiorinal^{R}. Most physicians do not realize that it contains a barbiturate derivative. Withdrawal symptoms have been reported in persons who have used this drug for 6 months to 2 years. Saper mentions that in his experience, withdrawal symptoms may follow rapid discontinuance of chronically administered nonsedative over-the counter medicines.

Two other medications are worthy of mention, caffeine and ergot, since they are frequently used in the management of headaches, particularly migraine. Most analgesic preparations contain caffeine mainly because it is reported to enhance analgesia, although this is debatable. The caffeine content of common drugs is listed in Table VIII. In Table IX the content of some foods is listed.

TABLE V *Analgesics used by 200 patients with chronic scalp muscle contraction headache*[a]

Analgesics		Patient distribution	
Class	Generic	*N*	%
Antipyretics solely (single or combination)	Aspirin, acetaminophen, or phenacetin	56	28.0
Plus sedative, tranquilizer, or muscle relaxant	Barbiturate, meprobamate, orphenadrine, etc.	76	38.0
Plus narcotic	Codeine oxycodone	63	31.5
Plus narcotic-antagonist	Propoxyphene, pentazocine	48	24.0
Others		37	18.5

[a] Based on 1-month prestudy record.

source: REF 28

TABLE VI *Composition of Selected Nonprescription Analgesics*

	ASPIRIN MG	ACETAMINOPHEN MG	CAFFEINE MG	OTHER
Plain aspirin	300–325	—	—	—
Tylenol	—	325	—	—
Datril	—	325	—	—
Excedrin P.M.	—	500	—	Pyrilamine maleate
Excedrin E.S.	250	250	65	—
Bufferin	324	—	—	Buffers
Empirin	325	—	—	—
Anacin	400	—	32	—
Percogesic	—	325	—	Phenyltoloxamine citrate, 30
Cope	421	—	32	Buffers
Vanquish	227	194	37	Buffers
Midol	454	—	32.4	Cinnamedrine, 14.9
Sinus drugs	Simple analgesics	—	—	Decongestants & antihistamines

source: REF 32

TABLE VII *Composition of Selected Prescription Analgesics*

DRUG	ASPIRIN MG	ACETAMINOPHEN MG	PHENACETIN MG	CAFFEINE MG	AVAILABLE WITH CODEINE	OTHER
Fiorinal	200	—	130	40	Yes	Butalbital, 50 mg
Empirin	325	—	—	—	Yes	
Darvon Compound	227	—	167	32	No	Propoxyphene, 65 mg
Percodan	224	—	160	32	No	Oxycodone
Percocet	—	325	—	—	No	Oxycodone hydrochloride
Esgic	—	325	—	40	No	Butalbital, 50 mg
Dialog	—	300	—	—	No	Alobarbital, 15 mg
Synalgos	356	—	—	30	No*	Promethazine hydrochloride
Micrainin	325	—	—	—	No	Meprobamate, 200 mg

*Available with semisynthetic narcotic analgesic.

source: REF 32

TABLE VIII *Caffeine Content of Common Drugs*

DRUG	Mg	DRUG	Mg
A.P.C.	32	Anacin	32
Cafergot	100	Bromoseltzer	32
Darvon Compound	32	Cope	32
Fiorinal	40	Midol	32
Migral	50	Vanquish	60
Wigraine	100	Excedrin	66
Pre-Mens	30		

source: REF 32

TABLE IX *Caffeine Content of Food*

SOURCE	ESTIMATED CAFFEINE MG
Brewed coffee (cup)	100–150
Instant coffee (cup)	85–100
Tea (cup)	60–75
Decaffeinated coffee (cup)	2–4
Cola (6 oz)	40–60
Cocoa (cup)	45–35
Chocolate bar	25

source: REF 32

Caffeine, although a stimulant, has been reported to decrease the pain of migraine by causing constriction of the extracranial arteries, although this theory has not been verified. Some persons with migraine can abort a headache with several cups of coffee and over-the-counter medications, which, unknown to them, also contain this compound. If they use caffeine on a frequent basis, they can develop symptoms of caffeinism, which include mood disturbances like anxiety, irritability, agitation, disorders of the sleep cycle, lighteadness, palpitations, and gastrointestinal complaints. Withdrawal causes headaches, malaise, and lethargy. Caffeine withdraw headaches can also occur in subjects who are heavy coffee drinkers when they rather abruptly stop their intake, or in persons who abuse ergot compounds containing caffeine (Cafergot^{R}) for the treatment of migraine. A vicious cycle is thus established; the physician is confronted with not only treating the migraine headache, but also the headache resulting from withdrawal of medication.

Ergotamine is one of the more useful drugs for the management of the acute migraine attack. The signs and symptoms of ergotism are well documented and include mental changes, seizures, muscle aches, ophthalmological complications, brain ischemia and periperal vasoconstriction leading to ischemic changes in the heart and extremities, and peripheral neuropathy. A not well-recognized chronic effect is the ergot headache, a withdrawal headache that develops insidiously in individuals who not only use large amounts daily, but also in those who use relatively small and infrequent doses, such as two to three times per week. Over time, they become more dependent on the medication and require increasing amounts to control the headaches. Finally, the other complications of ergotism develop.

There are no ready solutions to the problem of drug abuse in the treatment of chronic headaches. The problem of detection have been discussed. Increased emphasis on physician and lay person education is needed.

Aeromedical disposition

Candidates with a documented history of drug abuse for the treatment of headaches are permanently disqualifed from flying unless: a) the cause of the headache is not disqualifying; b) the drug abuse problem has been cured; c) no permanent medical or neurological residuals from the drugs, as determined by a thorough examination including neuropsychological studies, are found.

Cranial neuralgias

Trigeminal and glossopharyngeal neuralgias are the two most frequently encountered neuralgias in clinical practice. Although greater occipital neuralgia has been considered as a definite entity, its existence has been refuted.

Trigeminal neuralgia (tic douloureux)

Trigeminal neuralgia occurs most frequently in the 6th and 7th decades of life. Females are twice more likely to develop this than males. Familial cases have been reported, and in 20% of these, the pain is bilateral. The pain is far more frequently unilateral in nonfamilial cases. The cause is usually idiopathic in the older age groups. Recently, Janetta stated that the most frequent cause of this disease is vascular pulsatile compression of the trigeminal nerve, particularly by the superior cerebellar artery. the onset of the classic symptoms of trigeminal neuralgia in the younger age group should alert the physician to other causes, such as multiple sclerosis, arteriovenous malformations and brain tumors. The branches of the 5th cranial nerve that are most frequently involved in the neuralgia are mandibular and maxillary combined (36%), mandibular alone (20%), all three divisions (16%), maxillary alone (14%), maxillary and ophthalmic (11.5%), and ophthalmic alone (3%).[34] The primary pain site is listed in Table X.

The pain with trigeminal neuralgia is of short duration, but extremely severe. It

TABLE X

PRIMARY PAIN SITE IN TRIGEMINAL NEURALGIA*

Site	No. of Cases	Percent
Supraorbital	22	5
Infraorbital	200	46
Inferior dental	57	13
V_1 other	8	2
V_2 other	53	12
V_3 other	35	8
Multiple	63	14
Total	438	100

*V_1 indicates fifth nerve, first division; V_2, fifth nerve, second divisio
and V_3, fifth nerve, third division.

source: Ref 35

usually occurs in runs of repetitive attacks over a period of minutes. Most often it is triggered by touching the involved area or by talking and eating. In the beginning the attacks are infrequent with long periods of remission. By the time the patient consults a physician, he or she is usually functionally disabled by the pain. Rarely will the disease run a benign course. Medical treatment, particularly the prophylactic use of carbamazapine (Tegretol^{R}) is very helpful in the early course of the disease. Other drugs that have been used are phenytoin (Dilantin^{R}) and baclofen. Over time, the favorable response to medication decreases in 25% to 50% of patients, and surgical relief is required. A number of surgical techniques have been employed over the years. At this time percutaneous radio-frequency gangliolysis of the gasserian ganglion and microvascular decompression of the trigeminal root are the tones most often used. With the latter, approximately 80% of patients remain pain free for at least 24 months. With the former procedure, 69% were asymptomatic after one year; however after 5 years this percentage was reduced to 35%.[36] Postoperative complications with the above procedures occur in about 10% of patients.

Aeromedical disposition

Candidates with a history of trigeminal neuralgia should be permanently disqualified from flying unless: 1) the disease has been determined to be benign, that is, only rare paroxyms occur that are not incapacitating and that do not require prophylactic drugs for management (a neurological consultation is recommended). Certified candidates should be followed at 6-month intervals; 2) if surgical procedures are performed, candidates may be considered for certification if they have remained pain free off prophylactic medicines for a minimum of six months after recovery from the operation. In addition, they must not have suffered postoperative complications, which themselves, are disqualifying.

Glossopharyngeal neuralgia

This conditon, much less frequent than trigeminal neuralgia, begins after 40 years of age. The pain characteristics and course are similar to those with trigeminal neuralgia. Howver, the pain is located in the tonsillar area and base of the tongue with radiation to the ear. Swallowing is the main triggering event. This form of neuralgia may be associated with cardiac arrest and syncope.[37] The cause is unkown. Carbanazapine (Tesretol^{R}) or phenytoin (Dilatin^{R}) may be effective in preventing the painful paroxyms. Several surgical procedures have been tried; however, the number of cases treated is limited. Therefore, there are insufficient data on long term followup.

Aeromedical disposition

Candidates with a history of glossopharyngeal neuralgia should be disqualified permanently from flying.

Other causes for headaches and face pain

There are other causes for headaches and face pain, namely intra-and extracranial diseases that are not covered in this section. The reader is referred to the references listed in the introductory section of this chapter for further information. Dispostion is dependent on establishing the cause, the frequency and severity of pain, the use of medications and reversibility of the conditon after appropriate medical and/or surgical treatment.

References

1. Goldstein M, Chen TC: The epidemiology of disabling headache, in Critchley M, Friedman AP, Gorini S, et al (eds): Headache: Physiopathological and Clinical Concepts. Advances in Neurology, New York, Raven Press, 1982, Vol 22, pp 377-390.

2. Ziegler DK, Hassanein RS, Couch JR: Characteristics of life headache histories in a non-clinic population. Neurology 1977;27:265-269.

3. Waters WE, O'Connor PJ: Epidemiology of headache and migraine in women. J Neurol Neurosurg and Psychiat 1971;34:148-153.

4. Appenzeller O, Feldman RG, Friedman AP: Migraine, headache and related conditions. Arch Neurol 1979;36:784-805.

5. Packard RC (ed): Symposium on Headache. Neurological Clinics. Philadelphia, WB Saunders, 1983, Vol 1, No 2.

6. Critchley M, Friedman AP, Gorini S, et al: op cit.

7. Ziegler DK (ed): Headache Seminars in Neurology. New York, Theime-Stratton, 1982, Vol 1 No 1.

8. Diamond S, Dalessio DJ: The Practicing Physician's Approach to Headache (ed 3). Philadelphia, William and Wilkens, 1982.

9. Selby G: Migraine and Its Variants. Sidney Australia, ADIS Health Science Press, 1983.

10. Hirayama K, Ito N: Clinical aspects of migraine in Japan in, Rose FC (ed): Adavnces in Migraine Research and Therapy. New York, Raven Press, 1982.

11. Zielger DK: The headache symptom: How many entities? Arch Neurol 1985;42:273-274.

12. Sillanpaa M: Prevalence of migraine and other headache in Finnish children starting school. Headache 1975;15:877-880.

13. Billie B: Headaches in, children in: Viken PJ, Bruyn GW (eds): Handbook of Clinical Neurology. Amsterdam, North Holland Publishing Company, 1968.

14. Hockaday JM: Basilar migraine in children. Develop Med and Child Neurol 1979;21:455-463.

15. Prensky AL, Sommer D: Diagnosis and treatment of migraine in children. Neurology 1979;29:506-510.

16. Selby, op cit, pp 16-17.

17. Seshia SS, Reggin JD, Stanwick RS: Migraine and complex seizures in children. Epilepsia 1985;26:232-236.

18. Bartleson JD: Transient persistent neurological manifestations of migraine. Curr Concepts Cerebrovascular Disease and Stroke 1983;XVIII (5):21-25.

19. Corbett JJ: Neuro-opthalmic complications of migraine and cluster headaches, in: Smith CH, Beck RW (eds): Neuro-opthalmology. Neurologic Clinics. Philadelphia, WB Saunders, 1983, Vol 1 No 4, pp 973-995.

20. Fisher CMG. Late life accompaniments as a cause of unexplained tranisent ischemic attacks. Les Journal Canadian des Sciences Neurologies 1980;7:9-17.

21. Amat G, Jean Louis P, Loisy C, et al: Migraine and the mitral valuve prolapse syndrome, in: Critchley M, Friedmen AP, Gorini S, et al: op cit, pp 27-29.

22. Friedman AP: Overview of migraine, in: Critchley M, Friedman AP, Gorini S, et al: op cit, pp 1-17.

23. Kudrow L: Cluster headache: New concepts, in: Packard RC (ed): op cit, pp 369-383.

24. Joseph R, Rose FC: Cluster headache and herpes simplex: An association? Brit Med J 1985;290:1625-1626.

25. Ekbom K, Lindgren L, Nilsson BY, et al: Chronic migrainous neuralgia treated by retrogasserian injection of glycerol. Second International Migraine Symposium, London, 1984.

26. Diamond S, Dalessio DJ: op cit, pp 99-108.

27. Riley TL: Muscle-contraction headaches, in Packard RC (ed): op cit, pp 489-500.

28. Kudrow L: Paradoxical effects of frequent analgesic use, in: Critchley M, Friedman AP, Gorini S, et al (eds): op cit, pp 335-341.

29. Diamond S, Medina JL: Prolonged benign exertional headache: Clinical characteristics and response to indomethacin, in Critchley M, Freidman AP, Gorini S: op cit, pp 145-149.

30. Symonds C: Cough headache. Brain 1956;79:557-568.

31. Rooke ED: Benign exertional headache. Med Clin N America 1968;52:801-808.

32. Saper JR: Drug overuse among patients with headache, in: Packard RC (ed): op cit, pp 465-477.

33. Janetta PJ: Microvascular management of trigeminal neuralgia. Arch Neruol 1985;42:800.

34. Frank RA: Cranial neuralgias, in Packard RC (ed): op cit, pp 501-509.

35. Morley TP: Case against microvascular decompression in the treatment of trigeminal neuralgia. Arch Neurol 1985;42:801-802.

36. Burchiel KJ, Steege TD, Howe JF, et al: Comparison of percutaneous radio frequency gangliolysis and microvascular decompression for the surgical management of tic douloureux. Neurosurgery 1981;9:111-119.

37. St John JN: Glossopharyngeal neuralgia associated with syncope and seizures. Neurosurgery 1982;10:380-383.

Special Communication

Classification of Headache

Ad Hoc Committee on Classification of Headache

THE TERM "HEADACHE" commonly denotes head pain from brow level up. This outline discusses headaches somewhat more broadly; it covers both painful and nonpainful discomforts of the entire head, including the face and upper nucha. Since so much that a man describes as headache may be any abnormal head sensation, it is essential, for proper treatment, to determine whether the complaint is actually one of pain.

A useful scheme for the classification of the varieties of headache is one based on pain mechanisms. The divisions rest on experimental and clinical data together with reasonable inference. The story is far from complete; yet the arrangement can serve as a framework for diagnostic criteria for the major clinical types of headache and, by emphasis on basic mechanisms, it offers a logical approach to the planning of therapeutic trials. For convenience, short and simple names are suggested for certain major entities and are indicated in boldface type.

Essential in the study of headache, in most instances, is an appraisal of its close link to the patient's situation, activities, and attitudes. Sometimes in obvious ways, more often in subtle ones, headache may be the principal manifestation of temporary or sustained difficulties in life adjustment. These relationships are notably evident in Groups 1 through 3.

Table of Contents

1. Vascular Headache of Migraine Type
   A. "Classic" Migraine
   B. "Common" Migraine
   C. "Cluster" Headache
   D. "Hemiplegic" and "Ophthalmoplegic" Migraine
   E. "Lower-Half" Headache
2. Muscle-Contraction Headache
3. Combined Headache: Vascular and Muscle-Contraction.
4. Headache of Nasal Vasomotor Reaction
5. Headache of Delusional, Conversion, or Hypochondriacal States.
6. Nonmigrainous Vascular Headaches
7. Traction Headache
8. Headache Due to Overt Cranial Inflammation
9-13. Headache Due to Disease of Ocular, Aural, Nasal and Sinusal, Dental, or Other Cranial or Neck Structures.
14. Cranial Neuritides
15. Cranial Neuralgias

The committee was composed of Drs. Arnold P. Friedman (chairman), Knox H. Finley, John R. Graham, E. Charles Kunkle, Adrian M. Ostfeld, and Harold G. Wolff.

From the National Institute of Neurological Diseases and Blindness, Bethesda, Md.

Classification

1. Vascular Headaches of Migraine Type.—Recurrent attacks of headache, widely varied in intensity, frequency, and duration. The attacks are commonly unilateral in onset; are usually associated with anorexia and, sometimes, with nausea and vomiting; in some are preceded by, or associated with, conspicuous sensory, motor, and mood disturbances; and are often familial.

Evidence supports the view that cranial arterial distention and dilatation are importantly implicated in the painful phase but cause no permanent changes in the involved vessel. Listed below are particular varieties of headache, each sharing some, but not necessarily all, of the above-mentioned features:

(A) "Classic" Migraine.—Vascular headache with sharply defined, transient visual, and other sensory or motor prodromes or both.

(B) "Common" Migraine.—Vascular headache without striking prodromes and less often unilateral than *A* and *C*. Synonyms are: "atypical migraine" or "sick" headache. Calling attention to certain relationships of this type of headache to environmental, occupational, menstrual, or other variables are such terms as: "summer," "Monday," "weekend," "relaxation," "premenstrual," and "menstrual" headache.

(C) "Cluster" Headache.—Vascular headache, predominantly unilateral on the same side, usually associated with flushing, sweating, rhinorrhea, and increased lacrimation; brief in duration and usually occurring in closely packed groups separated by long remissions. Identical or closely allied are: erythroprosopalgia (Bing); ciliary or migrainous neuralgia (Harris); erythromelalgia of the head or histaminic cephalgia (Horton); and petrosal neuralgia (Gardner et al.).

(D) "Hemiplegic" Migraine and "Ophthalmoplegic" Migraine.—Vascular headache featured by sensory and motor phenomena which persist during and after the headache.

(E) "Lower-Half" Headache.—Headache of possibly vascular mechanism, centered primarily in the lower face. In this group there may be some instances of "atypical facial" neuralgia, sphenopalatine ganglion neuralgia (Sluder), and vidian neuralgia (Vail).

Best Available Co

2. Muscle-Contraction Headache.—Ache or sensations of tightness, pressure, or constriction, widely varied in intensity, frequency, and duration, sometimes long-lasting, and commonly suboccipital. It is associated with sustained contraction of skeletal muscles in the absence of permanent structural change, usually as part of the individual's reaction during life stress. The ambiguous and unsatisfactory terms "tension," "psychogenic," and "nervous" headache refer largely to this group.

3. Combined Headache: Vascular and Muscle-Contraction.—Combinations of vascular headache of the migraine type and muscle-contraction headache prominently coexisting in an attack.

4. Headache of Nasal Vasomotor Reaction.—Headaches and nasal discomfort (nasal obstruction, rhinorrhea, tightness, or burning), recurrent and resulting from congestion and edema of nasal and paranasal mucous membranes, and not proven to be due to allergens, infectious agents, or local gross anatomic defects. The headache is predominantly anterior in location, and mild or moderate in intensity. The illness is usually part of the individual's reaction during stress. This is often called "vasomotor rhinitis."

5. Headache of Delusional, Conversion, or Hypochondriacal States.—Headaches of illnesses in which the prevailing clinical disorder is a delusional or a conversion reaction and a peripheral pain mechanism is nonexistent. Closely allied are the hypochondriacal reactions in which the peripheral disturbances relevant to headache are minimal. These also have been called "psychogenic" headaches.

N.B.: *The foregoing represent the major clinical disorders dominated by headache—those which are particularly common, and in which headache is frequently recurrent and disabling.*

6. Nonmigrainous Vascular Headaches associated with generally nonrecurrent dilatation of cranial arteries:

(A) Systemic infections, usually with fever.

(B) Miscellaneous disorders, including hypoxic states, carbon monoxide poisoning, effects of nitrites, nitrates, and other chemical agents with vasodilator properties, caffeine-withdrawal reactions, circulatory insufficiency in the brain (in certain circumstances), postconcussion reactions, postconvulsive states, "hangover" reactions, foreign-protein reactions, hypoglycemia, hypercapnia, acute pressor reactions (abrupt elevation of blood pressure, as with paraplegia or pheochromocytoma), and certain instances of essential arterial hypertension (e.g., those with early morning headache).

7. Traction Headache.—Headaches resulting from traction on intracranial structures, mainly vascular, by masses:

(A) Primary or metastatic tumors of meninges, vessels, or brain.

(B) Hematomas (epidural, subdural, or parenchymal).

(C) Abscesses (epidural, subdural, or parenchymal).

(D) Post-lumbar-puncture headache ("leakage" headache).

(E) Pseudotumor cerebri and various causes of brain swelling.

8. Headache Due to Overt Cranial Inflammation.—Headaches due to readily recognized inflammation of cranial structures, resulting from usually nonrecurrent inflammation, sterile or infectious.

A. Intracranial disorders: infectious, chemical, or allergic meningitis, subarachnoid hemorrhage, postpneumoencephalographic reaction, arteritis, and phlebitis.

B. Extracranial disorders: arteritis and cellulitis.

9-13. Headache Due to Disease of Ocular, Aural, Nasal and Sinusal, Dental, or Other Cranial or Neck Structures:

9. Headache due to spread of effects of noxious stimulation of *ocular* structures (as by increased intraocular pressure, excessive contraction of ocular muscles, trauma, new growth, or inflammation).

10. Headache due to spread of effects of noxious stimulation of *aural* structure (as by trauma, new growth, or inflammation).

11. Headache due to spread of effects of noxious stimulation of *nasal* and *sinusal* structures (as by trauma, new growth, inflammation, or allergens).

12. Headache due to spread of effects of noxious stimulation of *dental* structures (as by trauma, new growth, or inflammation).

13. Headache due to spread of pain from noxious stimulation of *other* structures of the cranium and neck (periosteum, joint, ligaments, muscles, or cervical roots).

14. Cranial Neuritides.—Caused by trauma, new growth, or inflammation.

15. Cranial Neuralgias.—Trigeminal (tic douloureux) and glossopharyngeal. The pains are lancinating ("jabbing"), usually in rapid succession for several minutes or longer; are limited to a portion or all of the domain of the affected nerve; and are often triggered by end-organ stimulation. Trigeminal neuralgia must be distinguished, in particular, from cluster headache (1C), with which it is often confused.

NB.: So-called chronic post-traumatic headache *may arise from any one of several mechanisms. Such headache may represent sustained muscle contraction (2), recurrent vascular dilatation (1B), or, rarely, local scalp or nuchal injury (13). In some patients, the post-traumatic pain is part of a clinical disorder characterized by delusional, conversion, or hypochondriacal reactions (5).*

This study was supported by a grant from the National Institute of Neurological Diseases and Blindness.

Best Available Copy

Cerebrovascular Diseases

Introduction

Despite a declining incidence, cerebrovascular disease is the third most common cause of death in North American and most European countries. In the United States, the prevalence is 1.7 million persons, and the incidence of cerebrovascular "accidents" (CVAs or strokes) is 414,000 new cases per year. The incidence markedly increases after age 55 years. Approximately 50% of strokes are thrombotic in origin, with 2/3 involving large arteries resulting in infarction and 1/3 involving small, penetrating arteries producing smaller infarcts known as lacunes. Eighteen percent of thrombotic CVAs result from atherosclerotic disease of the carotid artery, 16% from disease of the vertebral and basilar arteries, and 19% from disease of the small, penetrating arteries. Thirty-one percent of all strokes are embolic in origin, either from the heart (15% to 25% of cases) or from an artery; neck arteries, especially the carotid, are most frequently the site of origin. Sixteen percent of CVAs are caused by brain hemorrhage, with 10% resulting from hypertension and 6% resulting from ruptured aneurysms or arteriovenous (AV) malformations. Therefore, the major risk factors for stroke are atherosclerosis, hypertension and heart disease. Other factors are diabetes, hyperlipidemia and cigarette smoking.

The classification and definitions of the types of cerebrovascular disease are continuing to be revised as new scientific information becomes available. For the present time, it is recommended that the FAA use the classification suggested by the ad hoc committee on stroke of the Advisory Council for the National Institute of Neurological and Communicative Disorders and Stroke.[1]

In the section on cerebrovascular disease that is found in the AMA's 1979 report to the FAA, the minimal criteria for the diagnosis of the various types of stroke as well as recommendations for flying are discussed.[2] For the most part, these criteria and

recommendations are applicable today. Therefore, in this section are discussed only those categories of cerebrovascular disease that present the most difficult decisions about aeromedical disposition. These are: 1) transient ischemic attacks (TIAs); 2) intracranial aneurysms; 3) arteriovenous (AV) malformations; 4) subarachnoid hemorrhage; 5) asymptomatic neck murmurs; and 6) stroke in young people.

Ischemic infarctions and parenchymal hemorrhages will not be covered, since in the great majority of persons with these conditions will be disqualified either because of their underlying disease or neurologic sequelae. However, candidates with a remote history of an infarction or hemorrhage due to the self-limited cause, such as drugs like as amphetamines or oral contraceptives, or a nonatherosclerotic embolism, such as fat, may be considered for certification provided they satisfy the neurological, neuropsychological, and CT or MRI criteria as determined from appropriate consultations.

Transient ischemic attacks (TIAs)

TIAs are reversible, focal, neurological deficits, lasting minutes to 24 hours, followed by a complete recovery of function. They may be secondary to embolization or hemodynamic factors such as low flow in a diseased vessel. They can be subdivided symptomatically into retinal (amaurosis fugax), cerebral and vertebral-basilar artery events. The major cause, particularly after age 45 years, is atherosclerotic disease, particularly of the neck arteries. There is no question that individuals with TIAs are at risk for the subsequent development of stroke. Mohr, for example, estimated that 10% of strokes are preceded by TIAs.[3] Approximately 30% to 33% of the strokes occur within 5 years and of these, 20% will occur within one month, and 50% will occur within one year.[4-5]

In the Framingham study the incidence of TIAs in the general population was reported to increase from 4/100,000/year for persons under 40 years, to 8/100,000/year

for those age 40 to 49 years, to 14/100,000/year for those age 50 to 62 years.[6] In this prospective study, stroke developed in 40% of persons with TIAs, 1/2 of which occurred within 3 months of the onset of TIAs and 2/3 within 6 months. Hass estimates the risk of a stroke after a well-defined TIA at 7% per year.[7]

In addition to the increased risk for stroke with TIAs, approximately 30% of persons with TIAs will experience a myocardial infarction or sudden death within seven years.[8]

Retinal TIA (amaurosis fugax)

Transient monocular blindness in middle age and older age strongly suggests atheromatous disease at the origin of the internal carotid arteries, although significant disease may not alway bedeomonstrated angiographically.[9] A follow-up study of individuals with only this type of transient ischemic event showed a seven-year cumulative rate of cerebral infarction of 14%. Four of the 52 (8%) people who were treated surgically and four of 35 (11%) who were treated medically subsequently developed cerebral infarction. The five year cumulative rate of recurrence of retinal TIAs was 37%, and some of these occurred in persons who were treated with endarterectomy. There was also an increased incidence of myocardial infarction and sudden death.[10]

In a retrospective follow-up study (median 8 years) of 110 patients with amaurosis fugax who were treated medically, Poole and Russell report an increased incidence of stroke, decreased life expectancy and increased ischemic heart disease.[11]

Aeromedical disposition: Candidates with a history of amaurosis fugax due to atherosclerotic disease should be disqualified permanently even if they have been treated surgically by endarterectomy.

Cerebral transient ischemic attack

Transient disturbances of motor, sensory or language functions characterize cerebral ischemic attacks. Most of the ischemic events, whether embolic or hemodynamic, occur within the distribution of the middle cerebral artery. In the same person, the attacks tend to be stereotyped. In addition, they may be associated with attacks of amaurosis fugax.

With cerebral TIAs, the seven-year cumulative risk of cerebral infarction is 27%.[10] Of 123 persons treated medically, 23 (19%) developed cerebral infarction, compared to nine of the 79 (11%) treated surgically. All these individuals were at risk for ischemic heart disease, and TIAs also occurred after surgery.

Although the studies mentioned in the beginning of this section did not specifically separate amaurosis fugax from cerebral TIAs, the evidence is overwhelming that TIAs are a significant risk factor for the development of cerebral infarction and myocardial ischemia.

The increased risk also applies to persons treated with surgery. Muuronen followed 100 persons who had vascular surgery (mostly endarterectomies) for periods of time with a mean of 1.7 years. Fifteen (14%) suffered TIAs, 6 (5%) cerebral infarcts, and 9 (8%) acute myocardial infarcts. The annual rate of brain infarction was 3.3% and of myocardial infarction, 4.4%.[12] In a study by Whisnant et al[13] of persons who had clearly defined TIAs in one carotid territory and who underwent carotid endarterectomy on the appropriate side, 15% continued to have TIAs. The ischemic stroke rate was 2% per year.

Aeromedical disposition: Candidates with a history of cerebral TIAs due to atherosclerotic disease should be disqualified permanently even if the condition was treated surgically.

Vertebrobasilar artery insufficiency

TIAs due to vertebrobasilar artery insufficiency are 1.6 time less frequent than carotid TIAs. Infarction is 6 times less frequent. The major symptoms are intermittent attacks of vertigo, diplopia, ataxia, visual disturbances, dysarthria, alternating hemiplegia and/or sensory disturbances. A small percent of individuals will experience a disturbance of consciousness. The natural history of this disease has not been as extensively investigated as carotid artery TIAs.[14]

In the best available study, Whisnant et al reported on a 15-year follow-up of persons with vertebrobasilar artery attacks. In persons not treated with anticoagulants the incidence of infarction was similar to that in persons with carotid artery TIAs. With treatment, the probability of infarction was significantly lower from three months through 5 years of observation. However, beginning at 6 months of observation of stroke-free patients, no significant difference was noted.[15] Surgical treatment of this disorder, including bypass operations, have not proved to be effective.[16] From the limited information available, transient ischemic attacks preceed brain stem infarcts in approximately 50% of patients.[17]

Aeromedical disposition: Candidates with a history of vertebrobasilar artery ischemic attacks should be disqualified permanently.

Intracranial aneurysms

Intracranial aneurysms are subdivided into the following types: a) berry or saccular; b) mycotic; c) fusiform; d) atherosclerotic; e) traumtic and; f) neoplastic. Of these, the berry aneurysm is the most common, with an age-corrected prevalence rate for the North American population reported as high as 2,000/100,000. From autopsy studies there is a uniform prevalence at all ages over 20 years.[18] Berry aneurysms can be found in 0.2% to 7.9% of autopsies; only 50% will have been ruptured.[18-19] Aneurysms have also been detected as incidental findings in 0.65% of arteriograms. This

reported prevalence may increase because of the increased utilization of contrast-enhanced CT scans and other neuroimaging techniques.

Most berry aneurysms become clinically symptomatic between the ages of 40 and 60 years. They are slightly more common in females, by a ratio of 3:2. Ten to twenty percent of berry aneurysms are multiple. This type of aneurysm is considered to be congenital, although acquired factors like hypertension may play a role in their enlargement and subsequent rupture.

An increased incidence of saccular aneurysms has been reported in persons with polycystic kidney disease, coarctation of the aorta, Ehlers-Danlos syndrome, arteriovenous malformations of the brain, fibromuscular hyperplasia, and Moya-Moya disease. A familial occurrence has also been reported. Saccular aneurysms occur at arterial bifurcations with 95% of single aneurysms occurring on the anterior portion of the Circle of Willis. Internal carotid aneurysms, primarily at the junction with the poster or communicating artery, is the most frequent site. Next are anterior communicating artery aneurysms, followed by middle cerebral artery defects. The aneurysms may solely rupture into the subarachnoid space; however, rupture into the parenchyma of the brain or ventricular system can occur.

The major clinical signs and symptoms of intracranial aneurysms are related to subarachnoid hemorrhage. Ten to fifteen percent of persons will present with cranial nerve palsies or signs of a space occupying lesion, especially when a large aneurysm remains intact. Approximately 50% of persons with a major subarachnoid hemorrhage will experience symptoms of a "warning leak" that precedes the major event by several hours or days, consisting of a sudden onset of headache associated with nausea and neck stiffness, which clears in several days. Most often these are mistaken as being caused by the "flu" or tension headaches.

The diagnosis of intracranial aneurysms is suggested by the clinical presentation, and confirmed by four-vessel angiography. However, angiograms may be normal in

persons with subarachnoid hemorrhage.

The natural history of ruptured and unruptured aneurysms has been extensively studied in the last several decades.[21] The data from this as well as other studies to be mentioned later are of utmost importance in determining whether an applicant with an intracranial aneurysm(s), whether ruptured or not, may be certified.

Ruptured aneurysms treated conservatively

In a follow-up study of 568 persons with ruptured intracranial aneurysms who were managed conservatively, 378 (68%) died. Forty percent of the deaths occurred within six months of hemorrhage. In the next two decades, the patients' survival curve was significantly worse than that of a matched US population. The probable recurrent bleeding rate after 6 months was 2.2% per year for the first decade, and 0.86% per year for the second decade. Seventy-eight percent of the recurrent bleeds were fatal.[22] These data are in general agreement with the results of other investigations.[23,24]

Symptomatic unruptured aneurysm

Fifteen percent of symptomatic, intact aneurysms rupture within 6 months of onset of symptoms.[25]

Incidental unruptured aneurysms

Incidental unruptured aneurysms and those found in association with a ruptured aneurysm (multiple aneurysms) bleed at a rate of 1% per year.[26] Winn et al estimate the rate of bleeding to be 2% to 3% per year for intact multiple or incidental aneurysms.[27]

Wiebers et al, in a study of 65 persons with 81 unruptured aneurysms (a mean follow-up period of 8.2 years) reported subsequent rupture in 8 (12%). None of the hemorrhages occurred in persons with an aneurysm smaller than 10 millimeters in diameter.[25] However, in the Cooperative Aneurysm Study, the average diameter of

ruptured aneurysms was 8.5mm, with 69% having a diameter of less than 10 mm and 13% less than 5 mm.[28]

Probabilities for risk of hemorrhage have been developed by Dell. Based on his data, for a person 20 years old the lifetime risk of rupture is 16.6%; 30 years old, 16.1%; 40 years old, 14.4%; 50 years old, 10.3%; and 60 years old, 4.7%.[29]

Ruptured and non-ruptured aneurysms treated with surgery

Aneurysms that are surgically isolated from the arterial tree are considered cured and the chance of recurrent hemorrhage is negligible. However, with nonisolation techniques, such as carotid artery ligation for posterior communicating artery aneurysms and anterior cerebral artery clipping for anterior communicating aneurysms, the recurrence of bleeding is approximately 3% per year[26] (see Table I).

The data on long-term follow-up of patients treated with surgery in the Collaborative Aneurysm Study have not yet been published. Although at this time it seems that aneurysms that are isolated from the arterial circulation are considered "cured," the FAA should re-evaluate this statement when these results are in.

In addition to the concern for rebleeding, aeromedical disposition also depends on the determination of neurological and neuropsychological residua in those individuals who have been treated surgically. Assessment of cranial nerve, motor and cognitive functions, psychosocial impairment and the presence of hydrocephalus by neurological examination, neuropsychological studies and CT scans or MRI, is mandatory.

It is generally assumed that persons with benign or uncomplicated subarachnoid hemorrhage (grades I and II according to the Hunt-Hess[30] classification) who undergo successful surgery will, by one year, be at a functional state comparable to their pre-illness level. However, in a recent study, Saveland et al reported that one in five such patients, despite favorable neurologic outcome, at one year suffered from cognitive or psychosocial deficits.[31] Therefore, it is imperative that before certification, potential

TABLE I

RATE OF REBLEEDING FOR ANEURYSMS AND AVM's

Cerebrovascular Disease*	Rate of Rebleeding
Ruptured aneurysms	
early ($\leq$ 6 months)	50% will rebleed
late ($>$ 6 months)	3%/year
Unruptured aneurysms	
incidental findings	1%/year
multiple aneurysms	1%/year
Treated aneurysms+	
early ($\leq$ 6 months)	3%/year
late ($>$ 6 months)	3% /year
SAH of unknown etiology	1%/year
Ruptured AVM	3%/year

*SAH = subarachnoid hemorrhage: AVM = arteriovenous malformation

+Rebleeding rate for posterior communicating artery aneurysms treated by carotid ligation and anterior communicating artery aneurysms treated by anterior cerebral artery clipping.

source: REF 26

psychological complications be evaluated thoroughly.

Aeromedical disposition: 1) Applicants with a history of ruptured aneurysm treated conservatively should be disqualified permanently; 2) applicants with a history of ruptured aneurysm treated surgically should be disqualifed unless: a) the aneurysm(s) has been successfully isolated from the arterial circulation, as determined by arteriography one year after recovery from surgery, and; b) there is no evidence of neurological, cognitive or psychosocial deficits that would interfere with flying performance as determined from the results of neurological and neuropsychological consultations, as well as CT and/or MRI investigations; 3) candidates with symptomatic, unruptured aneurysms should be disqualified, unless the aneurysm is isolated from the arterial circulation and at one year after the operation the applicant satisfies the criteria listed in 2b; 4) candidates with multiple aneurysms should be disqualified permanently unless all of the aneurysms are isolated surgically from the arterial circulation, whatever the diameters of the unruptured aneurysms, and the candidate satisfies all the criteria listed in 2b; 5) applicants with a single asymptomatic aneurysm should be disqualified permanently unless the diameter of the aneurysm is less than 5 mm in diameter. If an aneurysm larger than 5 mm in diameter is treated surgically, the criteria in 2b must be satisfied. For asymptomatic aneurysms small than 5 mm in diameter, the candidates may be certified, but they should be evaluated by angiogrpahy every one to two years. (However, because of the risk of repeated angiography, it would seem appropriate to disqualify all applicants with asymptomatic aneurysms regardless of size, unless they are corrected surgically); 6) candidates with a history of mycotic, fusiform, atherosclerotic, traumatic or neoplastic aneurysms should be disqualified permanently because of the underlying disease; 7) candidates with a history of aneursym associated with another disease, such as polycystic disease of the kidneys should be disqualified permanently; 8) candidates with a strong family history of familial aneurysms should be evaluated on an individual basis by a neurologist or neurosurgeon.[32]

Arteriovenous (AV) malformations

Non-aneurysmal vascular malformations have been classified pathologically into capillary telangiectasias, cavernous and venous angiomas, and arteriovenous (AV) malformations. The last is the most common and important of these embryonic malformations. Morphologically, they consist of abnormal arteriovenous connections without intervening capillaries. They are located within the brain or spinal cord parenchyma, as well as the dura. Their size varies from very small anomalies that are difficult to identify angiographically and pathologically (cryptic malformations), to massive lesions fed by multiple vessels.

The prevalence of AV malformations in the general population is estimated to be 0.14%. In most cases they are an isolated vascular anomaly; however, approximately 10% of persons with AV malformations also have saccular aneurysms. There is a familial predisposition, and an association with neurofibromatosis. Seventy percent of AV malformations are located above the tentorium and primarily within the distribution of the middle cerebral artery.

The major initial symptoms of hemorrhage from an AV malformation are seizures, headache and focal neurological deficits. Seven percent of persons with these anomalies bleed again within on year of the intital hemorrhage. Thereafter, the risk of rebleeding is 3% per year.

The natural history of AV malformations in unoperated individuals was recently reviewed by Crawford, et al.[33] These authors followed 217 patients with nonoperable AV malformations for varying periods of time (mean 10.4 years). They found a 42% risk of hemorrhage, 29% risk of death, 18% risk of epilepsy and a 27% risk of having neurological impairment by 20 years after diagnosis.

For operated cerebral and spinal cord malformations the natural history is less clear. Location, size and variation of intervention techniques (the employment of

arterial embolization with or without subsequent surgery as well as occasional treatment with radiation) make it extremely difficult to assess prognosis.

In a study by Parkinson and Bachers of 86 individuals with operated supratentorial AV malformations who were followed from four to 26 years, seizures were present in 35 (28 had preoperative seizures), and neurological deficits in 18. These authors do not mention the incidence of recurrent hemorrhage in the operated cases.[34]

Guidetti and Delitala followed 95 patients with operated intracranial AV malformations for two to 25 years, they reported no episodes of recurrent bleeding.[35]

The studies of Graf et al and Fults and Kelly, although they include the follow-up of persons treated both surgically and non-surgically, do not specifically address the long-term postoperative complications.[36,37] The information on spinal cord vascular malformations is even less complete.

Aeromedical disposition: Based on the available scientific information, candidates with a history of arteriovenous or other non-aneurysmal malformations of the brain and spinal cord should be disqualified permanently.

Vascular fistulas, particularly carotid artery-cavernous sinus fistulas, are not considered in this section, because they are exceedingly rare. Disposition will depend on the results of a complete neurosurgical evaluation one year after surgical treatment or after the fistula has closed spontaneously. Angiograms are required.

Subarachnoid hemorrhage

The incidence of subarachnoid hemorrhage is approximately 15/100,000/year. Most episodes occur between the ages of 20 and 60 years. The greatest majority of nontraumatic subarachnoid hemorrhages are a result of ruptured aneurysms or AV malformations.

The diagnosis of a subarachnoid hemorrhage is based on the history, physical findings, CSF findings and, in some cases, CT scan of the head. The following

classification proposed by the National Survey of Stroke is helpful:[38]

Definite: A saccular aneurysm or AV malformation confirmed by angiography, surgery or autopsy, and with a blood clot in the fissure of Sylvius, between the frontal lobes, in basal cisterns, or within a ventricle observed by CT scan.

Highly probable: A bloody or xanthochromic CSF plus two of the following signs or symptoms occurring at or shortly after onset: severe headache, depressed consciousness, signs of meningeal irritation, subhyaloid hemorrhages, weakness on one side or third nerve palsy.

Probable: One or more of the symptoms or signs mentioned for "highly probable" above. However, a lumbar puncture is not performed, or is traumatic, or a CSF examination is done more than two weeks after onset of symptoms and is negative.

Undocumented: Insufficient clinical evidence to be included in the other categories.

In the National Stroke Survey, 55% of the cases were definite, 33% highly probable, 10% probable and 2% undocumented.

Once it has been determined that a subarachnoid hemorrhage has occurred, then the next step is to determine the cause. These can be subdivided into: 1) ruptured aneurysms and AV malformations; 2) subarachnoid hemorrhage due to other known causes; 3) and subarachnoid hemorrhage of undetermined etiology. Ruptured aneurysms and AV malformations have already been discussed.

Subarachnoid hemorrhage due to other known causes

The other causes of subarachnoid hemorrhage are listed in Table II.[2] Most of these diseases are themselves disqualifying, regardless of whether they produce subarachnoid hemorrhage. Therefore, those candidates with subarachnoid hemorrhage resulting from the diseases listed in Table II should be disqualified permanently because of the nature of the disease itself. Exceptions to this are: 1) hemorrhage due to

TABLE II

CAUSES OF SUBARACHNOID HEMORRHAGE

Diseases of the blood-coagulopathies
- Hemophilia
- Thrombocytopenia
- Leukemia
- Waldenstrom's macroglobinemia
- Lymphoma
- Myeloma
- Aplastic anemia
- Hereditary spherocytosis
- Sickle cell anemia
- Pernicious anemia
- Polycythemia
- Afibrinogenemia
- Hypofibrinogenemia-hepatic disease or malignancy
- Consumption coagulopathies/disseminated intravascular coagulopathy
- Anticoagulant therapy

Angiopathies
- Lupus angiitis, lupus erythematosus
- Giant cell cerebral arteritis
- Dysproteinemias-amyloidosis
- Hypertension-all cases
- Atherosclerosis

Infectious diseases
- Bacterial meningoencephalitis, as with tuberculosis, leptospirosis, listeriosis
- Mycotic infection of brain, eg, aspergillosis
- Rarely, brucellosis, typhoid fever, yellow fever, dengue
- Viral encephalitis due to influenza, pertussis, herpes simplex; cytomegalic inclusion disease
- Bacterial endocarditis
- Anthrax
- Malaria

Intoxications
- Hypertensive crises pharmacologically induced, as with adenaline, monamine oxidase inhibitors, and amphetamine
- Alcohol
- Ether
- Carbon monoxide
- Morphine, nicotine poisoning, hydrocyanic acid, lead, quinine, phosphorus, pentamethylenetetrazol, amphetamine
- Snake bites (Bothrops sp)
- Uremia

Subarachnoid hemorrhage associated with pregnancy

Tumors
- Gliomas
- Pituitary tumros
- Meningiomas
- Pinealomas
- Choroid plexus papillomas
- Angioblastomas
- Melanomas
- Metastatic tumors

TABLE II cont'd

Craniocerebral trauma
- Direct or indirect blows to head
- Electrical injury
- Caisson disease
- High altitude anoxia
- Radiation
- Ultrasound
- Strangulation
- Thermal injury

Intracranial venous thrombosis
- Oral contraceptives
- Pregnancy
- Trauma
- Infection
- Coagulopathies
- Marasmus

Other conditions
- Physical stress-Valsalva's maneuver, as with coitus, defecation, parturition, etc
- Vitamin deficiency: scurvy, vitamin K deficiency

source: REF 2

transient coagulation disorders, most of which are drug induced; 2) transient, pharmacologically-induced hypertensive crises and; 3) trauma (see section on head injuries). In the first two categories, four-vessel angiography must be done before considering certification because of the possibility of a pre-existing aneurysm or AV malformation. In addition, a neurological consultation, neuropsychological studies and a CT scan or MRI are required. If the results of these investigations are normal, the candidate may return to flight status six to 12 months after complete recovery.

Subarachnoid hemorrhage of undetermined etiology

It has been reported that in 20% to 30% of individuals with subarachnoid hemorrhage no cause can be demonstrated, even after pan-angiography.[39]

In the Cooperative study of Intracranial Aneurysms and Subarachnoid Hemorrhage, 477 of 6,638 cases (7%) followed up to 24 years had unexplained subarachnoid hemorrhage.[39] The authors followed 464 patients up to 24 years after hemorrhage in whom pan-angiography was normal. One and one half percent of these patients had a subsequent hemorrhage. Repeat angiograms in 61 of these patients demonstrated lesions that were not detected initially. Based on the results of this comprehensive study, the investigators concluded that the first six-month interval is the period of greatest risk for recurrent hemorrhage. Thereafter, the life expectancy for normotensive individuals equals that of a comparable age-sex matched US population. If the person survives the first six months, the rate of subsequent hemorrhage is at a maximum 0.86% per year, which is lower than for persons with ruptured aneurysms who are managed conservatively. However, the cumulative probability of recurrent hemorrhage after a definite subarachnoid hemorrhage increases from 3.1% in the first six months to 7.3% by 20 years after the initial event. Persons with hypertension are at a definite increased risk.

However, studies on the long-term prognosis of complications other than risk of

recurrent bleeding and death are limited. Brismar and Sundbarg followed 127 persons with unexplained subarachnoid hemorrhages for an average of 5.4 years.[39] After the first week, only three persons experienced a recurrence. Eighty-eight percent of the persons, who at the end of the second week were fully awake and without symptoms of delayed cerebral ischemia, returned to full activity, and 98% returned to at least part-time work. These authors also found the important relationship of arterial hypertension to prognosis.

The question of the necessity of repeating pan-angiography if the intitial study is normal is controversial. Certainly, the results of Nishioka's study would support this.[40] However, many of these individuals were studied at a time when angiographic techniques were not as sophisticated as they are today. Forster et al[41] reported a false negative rate of 1.8% in the angiograms of 56 individuals whose initial four-vessel angiograms were normal. However, Ishii et al[42] found no false negative results in 24 individuals who had repeated angiograms. At times, it may be impossible to determine if an aneurysm that is seen on a repeated angiogram was responsible for the initial subarachnoid hemorrhage or had developed subsequently.

Therefore, based on the available data, we recommend that candidates with unexplained subarachnoid hemorrhage should be disqualified permanently even if the initial four-vessel angiograms are normal. Exceptions to this are candidates who have fully recovered without neurological and neuropsychological abnormalities as determined by appropriate consultations as well as CT scans and/or MRI, and in whom four-vessel angiography performed one year after recovery is normal, and who are normotensive.

Asymptomatic neck murmurs

Auscultation of the neck has become a routine physical examination procedure, particularly for persons over 40 years of age, and for those with symptoms of cerebral vascular disease. The prevalence of cervical murmurs in persons over 45 years has been

reported to be 12.6%. There are a number of causes of neck murmurs, which are listed in Table III.

In the Mayo Clinic study of 509 patients,[43] cervical murmurs were heard in 64 asymptomatic individuals. In three persons, the murmurs were classified as venous hums, in 39 as transmitted supraclavicular murmurs, and in 22 as cerebrovascular disease midcervical bruits. Seven of these 22 persons had previous symptoms of cerebrovascular disease.

It is the midcervical bruit that has created the most medical interest because of its reported association with stroke and heart disease.[44,45] The prevalence rate of 2.9% of midcervical artery bruits in the Mayo Clinic Study is slightly lower than the 4% and 4.4% reported in the Framingham and Evans County studies, respectively.[44,45] Midcarotid bruits increase from a prevalence rate of 0.9% in persons 45 to 54 years of age, to 2.1% in persons between 55 and 64 years of age, to 3.8% in persons between 65 and 74 years of age. The prevalence is higher in females than males — 4.4% versus 1.6%.[43]

It is thought that before a bruit can be detected by auscultation, the lumen of the artery must be reduced by 40%. Neither loudness nor pitch is a reliable indicator of the degree of stenosis. In fact, persons with high grade stenosis (90% or greater) may not even have a bruit. It should also be emphasized that not all carotid lesions will produce a bruit even if the stenosis is greater than 50%. In a study of the relationship between ausculation and Doppler examination of the neck, Hennerici et al[46] found concordance in only 9% of persons with abnormal and 52.1% with normal Doppler results. The false positive rate (negative Doppler with positive bruit) was 23%; the false negative rate was 16%.

However, in the study by Lo et al[47] only two persons with asymptomatic bruits had normal real-time ultrasound and continuous-wave Doppler studies. Of interest is that 60% of these individuals had significant atheromas ipsilateral to the bruit (ie, they

TABLE III

CLASSIFICATION OF CERVICAL BRUITS

I. Venous

Base of neck and above; abolished by jugular with compression

II. Arterial

A. Supraclavicular bruits

1. Transmitted from the heart: aortic stenosis and other aortic outflow murmurs (interstitial hypertrophic subaortic stenosis, supravalvular aortic stenosis); mitral insufficiei and related problems (mitral valve prolapse, rupture of chorda tendineae of papillary muscle); coarctation of the aorta; patent ductus arteriosus

2. Base of the neck: physiologic, atherosclerosis, thoracic outlet syndrome, arteritis

3. Hyperdynamic states: hemodialysis, fever, anemia, hyperthyroidism

B. Vertebral bruits (posterior to the sternocleidomastoid muscle):

1. Diffuse--upward continuation of supraclavicular murmurs

2. Localized--structural anomaly, atherosclerosis

C. Carotid bruits (anterior to the sternocleidomastoid muscle):

1. Diffuse--upward continuation of supraclavicular murmurs; obstruction of the opposite carotid (augmentation bruit)

2. Localized (midupper carotid; below angle of the jaw)--atherosclerosis (internal, external carotid); tortuosity, kinking, fibromuscular dysplasia, cranial arteritis, trauma, other conditions

source: REF 43

had plaques causing greater than a 50% reduction in flow, or ulcerated plaques, or total occlusion).

Therefore, the first step in the evaluation of an applicant with an asymptomatic neck bruit is to determine its cause. Since auscultation is not very reliable, noninvasive studies are indicated.

Noninvasive studies

Within the past decade, noninvasive techniques for evaluating the extracranial circulation have become an important part of the diagnostic armamentarium. Technological advancements have increased the sensitivity and specificity of these tests. The two techniques most frequently used today are real-time B-mode ultrasound, which displays actual images of all tissues in the neck and visualizes the carotid artery pulsations, and the Doppler study, which gives information on flow velocity. Most laboratories use both techniques or variations thereof. In addition, indirect tests for evaluating superficial and deep orbital flow characteristics are frequently employed. The noninvasive tests are listed in Table IV.

The studies relating noninvasive tests to arterial angiograms show a high degree of correlation. This is illustrated in Table V, which compares duplex scanning (B-mode imager and a pulsed Doppler unit with real-time spectral analysis) and angiography, from a study by Roederer et al.[49] These investigators found a concordance of 82%. The specificity of the non-invasive duplex was 84% and the sensitivity was 99%.

A comparison of continuous wave form Doppler (CWD) and real-time ultrasound (RTU) with angiography, from the study by Lo et al,[47] is shown in Tables VI and VII.

These studies also showed excellent correlation of noninvasive techniques with arteriographic findings. The tests are most sensitive when the degree of stenosis is greater than 40%. Ulcerated plaques are usually not detected unless they are associated with severe stenosis.

TABLE IV
APPLICABILITY OF DIFFERENT CLASSES OF NONINVASIVE TESTS

Test	Residual Lumen (mm)
Indirect	
Periorbital directional Doppler ultrasonography (PDU)	0-2.0
Oculoplethysmography (OPG)	0-2.5
Direct	
Bruit analysis	0.5-3.0
Direct Doppler exam	0.0-3.5
Imaging systems	0.0-7.0

source: REF 48

TABLE V
CONCORDANCE DUPLEX SCANNING VS. ANGIOGRAPHY

	Duplex results (% diameter reduction)						
Angiography	Normal	1-15%	16-49%	50-79%	80-99%	100%	
Normal	47	9					56
1-15%	4	49					61
16-49%		14	62	4			80
50-79%		1	7	56	8		72
80-99%				5	22	1	28
100%					1	38	39
	51	73	77	65	31	39	336

source: REF 49

TABLE VI
COMPARISON OF CWD WITH ANGIOGRAPHY IN 102 CAROTID ARTERIES

	Degree of stenosis by angiography		
CWD	<50%	>50%	Occlusion
<50%	62 (97%)	2	0
>50%	2	31 (94%)	0
Occlusion	0	0	5

TABLE VII
COMPARISON OF RTU WITH ANGIOGRAPHY IN 102 CAROTID ARTERIES

RTU	Normal	Plaque	Ulcerated Plaque	Occlusion
Normal	9 (100%)	0	0	0
Plaque	7 (9.6%)	62 (85%)	4 (5.4%)	0
Ulcerated Plaque	0	3 (20%)	12 (80%)	0
Occlusion	0	0	0	5 (100%)

source: REF 47

Recently, digital subtraction angiography has been used to visualize the extracranial vessels. Studies comparing noninvasive studies with this technique are in their infancy, although a recent study suggests that the degree of concordance is less than with conventional angiography.[50]

Fischer et al compared digital subtraction angiogrpahy and ultrasound and found a concordance of 89% in categorizing stenosis at less than or equal to 50% and greater than 50%. Correlation iwth fatal occlusion and ulceration was poor.[51] Further studies are required.

The FAA should recognize that these studies of concordance have come from laboratories with excellent equipment and expertise in this field. In the last decade there has been a proliferation of laboratories using noninvasive techniques in most medium- and large-sized hospitals, and even in physicians' offices. It is extremely important that the FAA determine the quality of the laboratory before referring a candidate. Unfortunately, these laboratories are not certified by any authority or association.

The prognosis for persons with asymptomatic bruits without ultrasound confirmation

The Framingham[44] and Evans County[45] studies of persons with asymptomatic cervical bruits revealed a stroke rate more than twice as great as expected for an age-matched, general population. Both studies also showed that the correlation between the location of the bruit and type of subsequent stroke was poor. In the Framingham study, 50% of the cardiovascular accidents were not ischemic in origin, not located or related to the side of the bruit, or were secondary to cardiogenic emboli. Cervical bruits in men were an added risk factor for death resulting from ischemic heart disease. The conclusion from the two studies is that asymptomatic carotid bruits are markers for the presence of diffuse atherosclerotic vascular disease.

Prognosis for persons with asymptomatic bruits secondary to carotid artery disease diagnosed by noninvasive studies

Roederer et al[49] followed 162 persons with asymptomatic bruits with serial ultrasonic duplex scans. By 36 months 10 of them became symptomatic (see Table VIII); all symptoms correlated with the side of the bruit.

Table VIII

RELATIONSHIP BETWEEN DISEASE PROGRESSION BEYOND 80% DIAMETER REDUCTION AND SYMPTOMS/OCCLUSION

Clinical status	Disease status on follow-up	
	< 80%	≥ 80%
No complication	258	13
TIA only	1	4
TIA with occlusion	0	1
TIA followed by stroke and occlusion	0	1
Stroke with occlusion	0	3
Asymptomatic occlusion	3	3
Total complications	4	12

source: REF 49

The rate of complications in this study is higher than in the studies of asymptomatic bruits not followed by noninvasive studies. Most likely this is due to the sensitivity of the scanner in detecting the source of the murmur.

Also of interest in this study is the rate of progression of the atherosclerotic process, particularly as it relates to the development of symptoms. Thirty-one percent

of the subjects showed some disease progression on one side and 7% on both sides. When a lesion is initially classified as causing less than 50% stenosis, 33% will progress to causing greater than 50% stenosis after 3 years. The progression of a lesion to greater than 80% stenosis is an important warning sign because the risk of developing ischemic symptoms or ipsilateral total occlusion is 35% at six months and 46% at 12 months. Of the lesions that did not progress to 80% stenosis, only 1.5% caused complications. The major risk factors for disease progression were diabetes mellitus, age and cigarette smoking. Persons under 65 years of age were most likely to show progression.

In the Toronto Asymptomatic Bruit Study,[52] the risk of a cerebral vascular event was approximately 15% per year in individuals who had a stenosis larger than 75%, compared to 3% for those who had stenosis less than 75%.

The results of these serial noninvasive studies correlate with the only angiographic study on the progression of carotid arteryarteriosclerosis.[53] In this study, 51 of 86 subjects who were followed for up to 9 years had increased severity of the stenosis; in 32 of these persons the stenosis prognosed at a rate of greater than 25% per year.

Aeromedical disposition: Based on the available data, the following sequence of evaluation should be followed with a candidate with an asymptomatic bruit: 1) the asymptomatic bruit should be determined to be of arterial origin, particularly midcervical in location; 2) if the bruit is midcervical and not transmitted, then noninvasive studies should be performed at a reputable laboratory; 3) if these studies show a degree of stenosis that is greater than 75% to 80% of the diameter of the lumen of the artery, either in one carotid or in both, the applicant should be denied certification (see the discussion on carotid endarterectomy); 4) if the studies show a degree of stenosis less than 75% to 80% of the diameter of the lumen, the candidate may be certified, provided there are no other disqualifying conditions. A thorough cardiovascular examination and evaluation for diabetes mellitus is mandatory, as is a thorough neurological evaluation; and 5) if the candidate meets all standards and

guidelines for cardiovascular disease and diabetes mellitus, he or she may be certified. However, the candidate should be re-evaluated neurologically and medically and with noninvasive studies performed at the same laboratory every six months thereafter. If the degree of stenosis progresses to greater than 75% to 80%, or if neurologically or medically disqualifying conditions develop, the airman should be denied certification permanently.

CT scan studies of persons with asymptomatic bruits have not been published. Since it is possible that persons with asymptomatic bruits may have experienced previous cerebral vascular events that either were not reported or were not recognized, a CT scan or an MRI study of the head may be considered by the FAA as part of the evaluation. If changes consistent with infarctions are detected, then the applicant should not be certified.

Strokes in the young adult

From time to time, the FAA will encounter young adult airmen with a history of transient ischemic attacks or completed strokes. The relative incidence of all TIAs or strokes between the ages of 16 and 45 years is approximately 3.7%,[38] which is similar to the figures found in the study in Toronto.[54] The causes of stroke or TIA in the Toronto study were as follows: 28% were due to cardiac disease, 23% were due to unknown cause (with normal angiograms in 2/3 of these individuals), 15% due to migraine, and 11% assumed to be a result of premature athreosclerosis. Miscellaneous causes comprised the remainder. Causes of premature TIAs and strokes in the younger age group are found in Table IX.

TABLE IX - CAUSES OF STROKES AND TIAs IN YOUNG ADULTS

1. Migraine
2. Hemorrhages
   a. Aneurysms
   b. Arteriovenous malformations
   c. Hypertensive hemorrhage

3. Hypertensive encephalopathy
4. Premature atherosclerosis
   a. Hypertension
   b. Diabetes
   c. Hyperlipidemia
   d. Hypothyroidism
   e. Nephrotic syndrome
5. Fibromuscular
6. Takayasu disease
7. Arrythmias
   a. Atrial fibrillation
8. Myocardial infarction
   a. Mural thrombus
   b. Akinetic segments
9. Cardiomyopathies
10. Blood disorders and dyscrasias
   a. Hemoglobinopathies such as sickle cells
   b. coagulopathies
   c. Thrombocytic thrombocytopenia purpura
   d. Leukemia
   e. C_2 deficiency
   f. Homocystinuria
11. Arteritis
   a. Infection such as syphilis, TB, virus
   b. Collagen vascular disease
   c. Giant cell arteritis
   d. Drug abuse
   e. Moyamoya disease
12. Venous infarctions
13. Arterial dissections
   a. Trauma
   b. Spontaneous
   c. Atherosclerotic
14. Trauma
   a. Accidents
   b. Chiropractic manipulation
15. Pulmonary venous disease
   a. Pulmonary venous thrombosis
   b. Paradoxical embolism
16. Valvular heart disease
   a. Mitral valve stenosis
   b. Mitral valve prolapse
   c. Bicuspid aortic valve
   d. Infective or non-infective endocarditis
   e. Valvular prostheses.
17. Cardiac tumors
   a. Atrial myxoma

source: REF 54

Many of the diseases in Table IX are themselves disqualifying, or the neurological residuals are so devastating that there is no question about certification. Ischemic

events associated with migraine have already been discussed. In cases in which there is a single event, such as arterial dissection due to trauma, or thrombosis due to chiropractic manipulation, the decision to certifiy is dependent on complete neurological and/or neurosurgical evaluation. CT scan or MRI, angiograms and neuropsychological studies may be indicated. Candidates with an unexplained transient ischemic episode(s) should be disqualified permanently unless a treatable, nondisqualifying deficit is discovered on subsequent evaluations, or a sufficient symptom-free observation period has elapsed (an arbitrary figure would be five years), from the time of the initial event(s).

Several entities deserve special discussion. These are: a) mitral valve prolapse; b) drugs and cerebrovascular disease; c) fibromuscular dysplasia; and d) transient global amnesia.

Mitral valve prolapse (MVP)

MVP is a common disorder, reported in 5% of the general population and 17% of young women and girls. It is reported to be associated with a number of conditions, including migraine, transient global amnesia, transient ischemic attacks, autonomic dysfunction, muscular dystrophies and intracranial aneurysms. However, for most of these conditions, the numbers reported are insufficient and a relationship has not been firmly established. A number of investigators have reported an increased incidence of cerebral ischemic attacks, particularly in younger persons with this entity, as shown in Table X. Thromboembolism has been implicated as the cause, based on clinical angiographic and postmortem examinations. The risk of stroke in young persons is estimated to be 1/6,000 per year.

Hart and Easton found a relationship of MVP with cerebral infarction,[56] and Jackson et al report recurrent episodes of TIA in 44% of subjects with MVP.[57]

The studies listed in Table X were on individuals who already had experienced an ischemic episode. Recently, Nishimura et al reported on long-term follow-up of 237

TABLE X

Mitral Valve Prolapse in Cerebral Ischemia

Investigators	No. of Patients	Age (yrs.)	% of Patients with MVP	% of Controls with MVP
Barnett et al[15]	60	6-45	40	6.8 ($p<.001$)
Barnett et al[15]	143	49-87	5.7	7.1 ($p>.05$)
Scharf et al[16]	47	≤45	28	8.5 ($p<.01$)
Egeblad and Sorensen[17]	30	24-39	10	0 ($p>.05$)
De Bono and Warlow[18]	117	25-70	11.1	3.8 ($p>.05$)
Bernard et al[19]	20	28-40	20	No Controls
Bonsal et al[20]	116	40-70	5.2	No Controls
[illegible] et al[20]	14	<45	21.5	No Controls
Fiesch et al[20]	106	>45	2.9	No Controls
Kouvaras and Baroulas[21]	66	<50	34.8	No Controls
Thareken et al[22]	38	<40	13	No Controls
Greenland et al[23]	100	Mean 70	1	No Controls
Smith and McKnight[24]	96	Unknown	5.2	No Controls
Coghurn et al[25]	88	14-68	23.9	No Controls

source: REF 54

persons with MVP demonstrated by echocardiography.[58] These subjects were either asymptomatic, or minimally symptomatic. The follow-up periods had a mean of 6.2 years. Only 10 persons (4.3%) developed an ischemic event. Six of these were in atrial fibrillation at the time of the event, one had a ventricular aneurysm with an apical thrombus, one had infective endocarditis, and the remaining two were not in atrial fibrillation and had no other cause. However, in both of these individuals, multiple embolic occlusions were seen in carotid angiograms. For the neurological as well as the other complications, those who had redundant valve leaflets were at the greatest risk.

Aeromedical disposition: Based on the information available, it is recommended that candidates with documented MVP who have TIAs should be disqualified permanently even if they are taking otherwise acceptable medications, such as aspirin and dipyrimidole (Presantine^{R}). The candidate should also satisfy the criteria discussed in the section on cardiovascular diseases.

Drugs and cerebrovascular disease

Elicit drugs: It is not often appreciated that abuse of elicit drugs can cause cerebrovascular complications. The relative incidence of CVAs with these various drugs is listed in Table XI.[59] Amphetamines are associated with increased risk of intracranial, and especially intracerebral hemorrhage. Other complications of these drugs are cerebral embolization and ischemic strokes. Candidates with a history of CVAs resulting from drug abuse should be disqualified permanently for obvious reasons.

Oral contraceptives: Although not commonly thought of today as a drug, oral contraceptives can cause CVAs. Case-control studies of oral contraceptives and stroke are listed in Table XII. The estimated relative risk of stroke ranges from zero to 16, depending on the type of study and length of follow-up. Both ischemic strokes and subarachnoid hemorrhage have been reported. Women who are older and who have other risk factors for stroke, such as hypertension and cigarette smoking habits, are the most

TABLE XI

RELATIVE INCIDENCE OF DOCUMENTED CEREBROVASCULAR COMPLICATIONS OF ABUSED DRUGS*

Drug	Intracerebral hemorrhage	Subarachnoid hemorrhage	Emboli	Ischemic stroke	Hypertension
Heroin	—	—	—	+	—
Amphetamines	+ + + +	+ + +	—	—	+ +
T's & Blues	+ +	—	+ + +	+	—
Cocaine	+	—	—	—	+ +
Methylphenidate	—	—	+ + +	—	+
LSD	—	—	—	+	+
PCP	+	—	—	—	+ +

*Not including complications of endocarditis.

source: REF 59

TABLE XII

CASE-CONTROL STUDIES OF ORAL CONTRACEPTIVES AND STROKE

Studies	Types of strokes (N)	Estimated relative risk
Vessey and Doll 1969[4]	Ischemic (19)	6
Sartwell et al 1969[5]	Ischemic (13)	19
Collaborative 1973[6]	Ischemic (140)	9
	Hemorrhagic (195)	2
Collaborative 1975[7]	Hemorrhagic	
	Never smoked (41)	1.2
	Heavy smoking (78)	7
Jick et al 1978[8]	Ischemic (13)	26
	Hemorrhagic (1)	
Inman 1979[9]	Subarachnoid hemorrhage (134)	1.5
Thorogood et al 1981[10]	Subarachnoid hemorrhage (168)	1.4

source: REF 60

susceptible. Oral contraceptive-induced migraine was discussed in the headache section. The exact mechanism is not known, although a hypercoaguable state has been postulated.

Candidates with a history of stroke assumed to be a result of oral contraceptives should be disqualified permanently unless: a) the resulting neurological deficits are not disqualifying as determined from neurological and neuropsychological evaluations as well as from radiological procecdures such as a CT scan or MRI; b) four vessel angiograms are normal; c) the candidate is no longer taking oral contraceptives; and d) the applicant satisfies all other medical criteria, particularly the cardiovascular criteria.

Fibromuscular dysplasia (FMD)

Fibromuscular dysplasia is a non-atheromatous and non-inflammatory angiopathy of unknown etiology. It affects the primary and distant branches of the aorta, including the internal carotid, vertebral and intracranial arteries. It is a rare disorder with reported frequencies of 0.25% to 0.77%. In two thirds of cases, the internal carotid artery is involved.[61] Bilateral involvement has been reported in 86% of persons with this condition. Approximately 90% of the cases occur in women, with the average age of onset around 58 years. The natural history of this disease is not known. Focal cerebral symptoms, spontaneous arterial dissections, and mural and saccular aneurysms have been reported. In a clinical and angiographic study of 32 persons with this disease, So et al report a 56% incidence of focal neurological symptoms and 22% incidence of saccular aneurysms with rupture in 5 of the 7 persons. Follow-up angiograms in 6 individuals showed progression of the disease in two.[62]

In other studies reviewed by Sandok[60] angiographic progression of the lesion was noted in five of 15 persons and the development of new lesions in two. In 79 persons with carotid artery FMD followed for an average period of five years, only three experienced subsequent ischemic events. The association of atherosclerotic disease with this

entity has been noted by Sandok; thus it may be difficult to determine whether these symptoms were due to FMD or atherosclerosis.

In some persons surgery has been performed, particularly bypass procedures; however, numbers are too few to evaluate the results. Thus, based on the available information, candidates with angiographic evidence of FMD, whether it is related to their symptoms or is an incidental finding, should be disqualified permanently.

Transient global amnesia (TGA)

Transient global amnesia is included in the section on cerebrovascular disease because many authorities feel that it is caused by ischemia within the posterior circulation. It is characterized by: 1) an acute onset of disturbed memory; 2) a variable period of retrograde amnesia; 3) preservation of immediate recall; 4) preservation of remote memory; 5) resolution of all deficits, generally within 24 hours; 6) a residual period of amnesia after the attack; and 7) absence of exclusionary criteria.[63] TGA usually occurs in themiddle-to-late age groups. It has been reported in association with a number of conditions: migraine; mitral valve prolapse; cerebral and cardiac angiography; polycythemia; digitalis, diazepam and iodochlorhydroxyquin drug intoxications; and TIAs within the posterior circulation. It has been reported in 3.5% of persons with TIAs.[64] Recurrent attacks occur in approximately 10% of persons with TGA. Kushner and Hauser[63] conclude from the clinical, electroencephalographic and radiological investigations of 18 patients that TGA is closely linked to cerebrovascular disease. The recurrence rate was 7.0% for both TGA and subsequent cerebral ischemia. In 5 of 13 patients, CT scans showed focal thalamic and temporal lobe abnormalities.

Although the pathogenesis of this disease remains controversial, there is sufficient evidence to support the recommendation that candidates with this condition should be disqualified permanently. Possible exceptions to this are candidates with drug or arteriography-induced episodes, provided that the drug-provoked attacks were accidental

(eg, the drug was not abused) and diseases confirmed by the angiograms are themselves not disqualifying.

References

1. Committee on Cerebrovascular Disease: A classification and outline of cerebrovascular disease. Stroke 1975;6:563-616.
2. Whisnant JP, Crowel RM, Patterson RH, et al: Cerebrovascular disease. Arch Neurol 1979;36:734-738.
3. Mohr JP: Transient ischemic attacks and the prevention of strokes (editorial). New Engl J Med 1978;299:93-95.
4. Thompson JE, Talkington CM: Carotid endarterectomy. Ann Surg 1976;184:1-15.
5. Sandok BA, Furlan AJ, Whisnant JP, et al: Guidelines for the management of transient ischemic attacks. Mayo Clinic Proc 1978;53:665-674.
6. Wolf PA, Kannel WB, Dawber TR: Epidemiology of stroke: The Framingham Study, in Schoenberg B (ed): , Neurological Epidemiology: Principles and Clinical Applications. Advances in Neurology. New York, Raven Press, 1978, Vol 19, 107-120.
7. Hass WK: Aspirin for the limping brain (editorial). Stroke 1977;8:299.
8. Heyman A, Wilkinson WE, Hurtwitz BJ, et al: The risk of ischemic heart disease in patients with TIA. Neurology 1984;34:626-630.
9. Adams HP, Putnam SF, Corbett JJ, et al: Amaurosis fugax: The results of arteriography in 59 patients. Stroke 1983;14:742-744.
10. Hurwitz BJ, Heyman A, Wilkinson WE, et al: Comparison of amaurosis fugax and transient cerebral ischemia: A prospective clinical and arteriographic study. Ann Neurol 1985;18:698-704.
11. Poole CJM, Ross Russell RW: Mortality and stroke after amaurosis fugax. J Neurol Neurosurg and Psychiat 1985;48:902-905.

12. Muuronen A: Outcome of surgical treatment of 100 patients with transient ischemic attacks. Stroke 1984;15:959-964.

13. Whisnant JP, Sandok BA, Surodt TM: Carotid endarterectomy for unilateral carotid system transient cerebral ischemia. Mayo Clinic Proc 1983;58:171-175.

14. Ausman JI, Shrontz CE, Pearce JE, et al: Vertebrobasilar insufficiency: A review. Arch Neurol 1985;42:803-808.

15. Whisnant JP, Cartlidge NEF, Elvebach LR: Carotid and vertebralbasilar transient ischemic attacks: Effects of anticoagulants, hypertension and cardiac disorders on survival and stroke occurrence: A population study. Ann Neurol 1978;3:107-115.

16. Failure of extracranial-intracranial arterial bypass to reduce the risk of ischemic stroke: Results of an international randomized trial: The EC/IC bypass study group. New Engl J Med 1985;313:1191-1200.

17. Thompson JR, Simmons CR, Hass AN, et al: Occlusion of the intradural vertebrobasilar artery. Neuroradiol 1978;14:219-229.

18. McCormick FW: Pathology and pathogenesis of intracranial saccular aneurysms, in Saks AL, Adams HP (eds): Seminars in Neurology. New York, Thieme-Stratton, 1984, Vol 4, pp 291-303.

19. Fox JL: Intracranial Aneurysms. New York, Springer-Verlag 1983, Vol 1, pp 15-18.

20. Winn HR, Taylor J, Kaiser DL: Prevalence of asymptomatic incidental aneurysms: Review of 4568 arteriograms (abstract). Stroke 1983;14:21.

21. Saks AL, Nishioka H, Torner JC, et al: Cooperative study of intracranial aneurysms and subarachnoid hemorrhage: A long-term prognostic study. I: Introduction. Arch Neurol 1984;41:1140-1141.

22. Nishioka H, Torner JC, Graf CJ: Cooperative study of intracranial aneurysms and subarachnoid hemorrhage: A long-term prognostic study. II: Ruptured intracranial aneurysms managed conservatively. Arch Neurol 1984;41:1142-1146.

23. Winn HR, Richardson AE, Jane JA: The long-term prognosis in untreated cerebral aneurysms. I: The incidence of late hemorrhage in cerebral aneurysms: A ten-year evaluation of 364 patients. Ann Neurol 1977;1:358-370.

24. Winn HR, Richardson AE, O'Brien ', et al: The long-term prognosis in untreated cerebral aneurysms. II: Late morbidity and mortality. Ann Neurol 1978;4:418-426.

25. Wiebers DO, Whisnant JP, O'Fallon WM: The natural history of unruptured intracranial aneurysms. N Engl J Med 1981;304:696-698.

26. Jane JA, Kassell NF, Torner JC, et al: The natural history of aneurysms and arteriovenous malformations. J Neurosurg 1985;62:321-323.

27. Winn HR, Berga SL, Richardson AE, et al: Long term evaluation of patients with cerebral aneurysms. Ann Neurol 1981;10:106.

28. Kassell NP, Torner JC: Size of intracranial aneurysms. Paper presented at Conress of Neurological Surgeons, Los Angeles, October 1981.

29. Dell S: Asymptomatic cerebral aneurysm: Assessment of its risk of rupture. Neurosurgery 1982;10:162-166.

30. Hunt WE, Hess RM: Surgical risk as related to time of intervention in the repair of intracranial aneurysms. J Neurosurg 1968;28:14-20.

31. Saveland H, Sonesson B, Ljunggren B, et al: Outcome evaluation following subarachnoid hemorrhage. J Neurosurg 1986;64:191-196.

32. Acosta-Rua G: Familial incidence of ruptured intracranial aneurysms. Arch Neurol 1978;35:656-677.

33. Crawford PM, West CR, Chadwick DW, et al: Arteriovenous malformations of the brain: Natural history in unoperated patients. J Neurol Neurosurg and Psychiat 1986;49:1-10.

34. Parkinson D, Bachers G: Arteriovenous malformations: Summary of 100 consecutive supratentorial cases. J Neurosurg 1980;53:285-299.

35. Guidetti B, Delitala A: Intracranial arteriovenous malformations: Conservative and surgical treatment. J Neurosurg 1980;53:149-152.

36. Graf CJ, Perrett GE, Torner JC: Bleeding from the cerebral arteriovenous malformations as part of their natural history. J Neurosurg 1983;58:331-337.

37. Fults D, Kelly DL: Natural history of arteriovenous malformations of the brain: A clinical study. Neurosurg 1984;15:658-662.

38. Walker AE, Robins M, Weinfeld FD: Clinical findings: National Survey of Stroke. Stroke 1981;12(suppl):13-44.

39. Brismar J, Sundbarg G: Subarachnoid hemorrhage of unknown origin: Prognosis and prognostic factors. J Neurosurg 1985;63:349-354.

40. Nishioka H, Torner JC, Graf CJ, et al: Cooperative study of intracranial aneurysms and subarachnoid hemorrhage: A long term prognostic study. III: Subarachnoid hemorrhage of undertermined etiology. Arch Neurol 1984;41:1147-1151.

41. Forster DML, Steiner L, Hakenson S, et al: The value of repeat pan-angiography in cases of unexplained subarachnoid hemorrhage. J Neurosurg 1978;48:712-716.

42. Ishii R, Kuroki M, Tanaka R, et al: Subarachnoid hemorrhage of unknown cause. Neurol Med Chir 1983;23:262-266.

43. Sandok B, Whisnant JP, Furlan AJ, et al: Carotid artery bruits: Prevalence survey and differential diagnosis. Mayo Clinic Proc 1982;57:227-230.

44. Wolf PA, Kannel WB, Sorlie P, et al: Asymptomatic carotid bruit and risk of stroke: The Framingham study. JAMA 1981;245:1442-1445.

45. Heyman A, Wilkinson WE, Heyden S, et al: Risk of stroke in asymptomatic persons with cervical arterial bruits: A population study in Evans County, Georgia. N Engl J Med 1980;302:838-841.

46. Hennerici M, Aulich A, Sandermann W, et al: Incidence of asymptomatic extracranial disease. Stroke 1981;12:750-758.

47. Lo LY, Ford S, McKinney WM, et al: Asymptomatic bruit, carotid, and vertebral-basilar transient ischemic attacks: A clinical and ultrasonic correlation. Stroke 1986;17:65-68.

48. Ackerman RH: Non-invasive diagnosis of carotid disease in the era of digital subtraction angiography, in Barnett HJM (ed): Symposium on Cerebrovascular Disease. Neurologic Clinics. Philadephia, WB Saunders, 1983, Vol 1, pp 263-278.

49. Roederer GO, Langlois YE, Jager KA, et al: The natural history of carotid arterial disease in asymptomatic patients with cervical bruits. Stroke 1984;15:605-613.

50. Guidotti M, Landi G, Scotti G, et al: Digital subtraction angiography in patients with cerebral ischemic attacks and normal continuous wave Doppler studies. J Neurol Neurosurg and Psychiat 1985;480:39-43.

51. Fischer CG, Anderson DC, Farber R, et al: Prediction of carotid disease by ultrasound and digital subtraction angiography. Arch Neurol 1985;42:224-227.

52. Kuller LH, Sutton KC: Cartoid artery bruit: Is it safe and effective to auscultate the neck? Stroke 1984;15:944-947.

53. Javid H, Ostermiller WE, Hengesh JW, et al: Natural history of carotid artery bifurcation atherosclerosis. Surgery 1970;67:80-86.

54. Hachinski V, Norris JW: The Acute Stroke. Philadelphia, FA Davis, 1985, Chapter 9, pp 141-163.

55. Jackson AC: Neurological disorders associated with mitral valve prolapse. Can J Neurol Sci 1986;13:15-20.

56. Hart RG, Easton JD: Mitral valve prolapse and cerebral infarction. Stroke 1982;13:429-430.

57. Jackson AC, Boughner DR, Barnett HJM: Mitral valve prolapse and cerebral ischemic events in young patients. Neurology 1984;34:784-787.

58. Hishamura RA, McGoon MD, Shub C, et al: Echocardiographically documented mitral valve prolapse: Long-term follow of 237 patients. N Engl J Med 1985;313:1305-1309.

59. Caplan LR, Hier DB, Banks G: Current concepts of cerebrovascular disease - stroke: Stroke and drug abuse. Stroke 1982;13:869-872.

60. Longstreth WT Jr, Swanson PD: Oral contraceptives and stroke. Stroke 1984;15:747-750.

61. Sandok BA: Fibromuscular dysplasia of the internal carotid artery, in Barnett HJM (ed): Symposium on Cerebrovascular Disease. Neurologic Clinics. Philadelphia, WB Saunders, 1983, Vol 1, pp 17-26.

62. So EL, Toole JF, Dalal P, et al: Cephalic fibromuscular dysplasia in 32 patients. Arch Neurol 1981;38:619-622.

63. Kushner MJ, Hauser WA: Transient global amnesia: A case-control study. Ann Neurol 1985;18:684-691.

64. Crowell GF, Stump DA, Biller J, et al: The transient global amnesia-migraine connection. Arch Neurol 1984;41:75-79.

Brain Tumors

The AMA reviewed the subject of brain tumors for the FAA in 1979.[1] The neurology consultants for this present project conclude than, in general, the clinical descriptions and recommendations for medical disposition of persons with brain tumors are applicable today as well. The consultants would like to update the 1979 review with the following:

Computed Tomography (CT) Scans and Magnetic Resonance Imaging (MRI)

Scant mention was made of the benefit of CT scans, and no mention was made of MRI, in the diagnosis of brain tumors. These imaging techniques are non-invasive, and are now indispensible in the evaluation of primary and metastatic intracranial tumors. Although these studies may need to be supplemented by cranial angiography and, rarely, pneumoencephalography, CT scanning and MRI essentially have supplanted these two invasive techniques. The FAA should obtain CT scans or MRI on all persons with presumed primary or metastatic intracranial tumors.

Aeromedical disposition

In general, all applicants with intracranial tumors should be disqualified initially, and most likely, permanently. This applies especially to persons with malignant primary brain tumors and metastatic tumors such as from cancer of the lung (see hematology/oncology section, pulmonary section, and discussions of solid tumors in the other organ-system-based sections of this report).

Persons with "benign" tumors should be disqualifed. After removal of the tumor, these persons should be followed for at least 5 years before certification should be considered.[2] A person may be certified if there is no history of pre-operative or post-operative seizures, and the neurological examination, EEG, CT scan and/or MRI, and

neuropsychological studies at the time of application are normal. The applicant shall have taken no medications for the five year observation period.

Persons with "benign" posterior fossa tumors, pituitary adenomas and acoustic neurinomas should be disqualified and observed for at least one year, at which time certification may be considered. The neurologic examination should be normal at that time. If replacement therapy of endocrine hormones is required, the applicant should be evaluated by an endocrinologist (see endocrine system section).

Pseudotumor cerebri

Persons with pseudotumor cerebri should be disqualified for at least six months following the termination of corrective therapy. The neurological evaluation at the time of application should show no residual neurologic and visual deficits. Since some pseudotumors tend to recur, these applicants, once certified, should be evaluated every 6 months for 5 years. Those persons whose pseudotumor is associated with morbid obesity should also be certified according to the guidelines for obesity that are recommended by the risk factor committee (see section on risk factors).

References

1. Neurologic and neurosurgical conditions associated with aviation safety. Arch Neurol 1979;**36**:739-749.
2. Adegbite AB, Khan MI, Paine KWE, et al: The recurrence of intracranial meningiomas after surgical treatment. J Neurosurg 1983;**58**:51-56.

Dysequilibrium

The subject of dizziness and dysequilibrium was discussed by the AMA in its 1979 report to the FAA,[1] as well as by the ENT consultants for this review. The neurological consultants reviewed both reports and concur with their conclusions, especially the recommendation of the ENT committee that a person who has any condition that may cause sudden paroxysms of dizziness or dysequilibrium be disqualified permanently. A list of disorders that cause these symptoms is found in the 1979 report.

The 1979 report discusses in more detail dysequilibrium caused by transient ischemic attacks (TIAs), multiple sclerosis (MS), brainstem migraine and vertigenous epilepsy. These four entities are discussed in other sections of this present review (sections on cerebrovascular disease, demyelinating disease, headaches, and epilepsy, respectively). The 1979 report also discusses cerebellar-pontine angle tumors. The neurology consultants agree with its recommendations, adding only that an applicant with such a tumor undergo a thorough neurological evaluation, including EEG, at least one year after the removal of the tumor, or after the tests of vestibular function have returned to normal. The one-year period of observation should be sufficient to detect delayed onset symptoms, especially if the tumor or surgery affected the brainstem or brainstem blood supply.

Reference

1. Neurological and neurosurgical conditions associated with aviation safety. Arch Neurol 1979;36:806-810.

RECOMMENDATIONS FOR RESEARCH

Endocrine System

- The FAA should devise a surveillance program to monitor diabetic individuals taking oral hypoglycemic agents, who may be granted medical certificates if the recommendations set forth in this report are adopted. This surveillance should include special medical monitoring as well as interviews with those pilots about incidents of possible hypoglycemia while in flight status.

- Both hypoxia and hypoglycemia affect mentation and psychomotor function. The FAA should devise a study in a controlled setting of the combined affects of hypoglycemia and hypoxia on the ability of persons to perform the tasks of airmen.

Mental and Behavioral

- The five-question mini-mental state examination that is proposed as a screening tool for cognitive function is comprised of techniques from psychological test batteries that are standardized and validated. However, the five-question examination itself is new and untested.

- The FAA should monitor the use of this screening mini-mental state examination, to determine its acceptance by pilots and AMEs, and assess its validity and reliability as a screening tool in detecting diminished cognitive functions in the pilot population.

- The FAA should also consider developing a computerized test of cognitive function, which could be administered and scored in the AME's office. This appears feasible, since computers are becoming more commonplace in physicians' offices. The test items could be changed randomly to prevent the examinees from knowing what item will be presented. As with the five-item test described in this report, the computerized test would be a screening test that would detect significant cognitive impairments that may otherwise go unrecognized during a routine physical examination.

- The FAA should study the usefulness of routine GGT determination and urine screening for alcohol abuse or dependence. This could be attempted on a sample of pilot applicants, to assess acceptance, logistics, quality control, and validity.

Visual System

- Progressive power lenses, or Varilux lenses, are considered conventional lenses, and as such are allowed by the FAA to provide best-corrected visual acuity for a pilot. However, these lenses also induce a built-in trapezoidal distortion. We recommend further research into the induced visual disturbances by such lenses to assess their potential hazard to aviation safety.

- It is well known that kinetic disturbances are induced with contact lenses. The FAA should study these disturbances in pilots who perform acrobatics.

- There is a pressing need for an office method for testing aviation signal red, green, and white in Class III pilots. A simple lantern test incorporated into

the popular Titmus vision screening tester, or a simple hand-held tester would seem most appropriate. For the Titmus, all that would be required is an additional slide that would permit recognition of the aviation signal green, red and white.

- The Titmus slide or the hand-held device should incorporate the same chromaticity and luminosity as the signal light gun. It would have to be field-tested first on a group of color-deficient and color-normal individuals, and correlated to the signal light gun test. The office plate tests, such as Dvorine and American Optical, are very strict and fail a number of individuals who pass the signal light gun test. Therefore, a simple office signal light gun-type test would be ideal.

- The FAA may have to consider the implications of new four-color weather radar and color cathode-ray tube visual displays in the cockpits of modern aircraft, and may have to amend color vision standards in the future, depending on standardization of the colors in these instruments.

- The FAA should consider changing the tower signal light controls to red and white only. The present system has been shown to be very confusing and is seldom in use. This is well-documented in the article by HL Gibbons and MF Lewis in Aerospace Medicine, 1969;40(6):662-669. Similarly, a change in the standard aviation lighting systems has been suggested as a means of better signal light recognition for color-deficient pilots.

- Some persons, mainly those with deuteranopia, or green-blindness, have no trouble seeing red, provided that the red is sufficiently intense. Unless they

are also blue-yellow blind (which is very rare), these persons have no trouble seeing blue lights. The elimination of green signals will enable them to function well when an intense red light is seen and when objects or signals that were formerly green are changed to blue. The added purification of red with elimination of yellow, keeping the red close to the 650 mu wavelength, should also help those individuals to see red. With these changes, only a very small number of color-deficient persons will be uncertified, and these will be persons with protanopia, or red-blindness.

The FAA should recognize that no aviation crash has ever been attributed directly to a color deficiency. Therefore, a change in some color signal lights may cause an increase in crashes, not because the lights would not be seen, but because the pilot may not interpret the new lights correctly. Clearly, a re-education effort would have to accompany a signal light change.

- Dille and Booze state in the October 1984 issue of Aviation, Space and Environmental Medicine: "The relationships between heterophoria, fatigue, dim light, a break in fusion, decreased depth perception, diplopia and accidents are unknown." The FAA should consider studying these factors and their significance in flight safety.

- It can be argued that heterophoria testing is costly, time-consuming and unnecessary, since no aircraft crash has ever been attributed to motility abnormalities. However, since we cannot show that another method of assessing motility is more meaningful to the visual tasks in flying, present methods of motility testing should be continued.

- There are many newly emerging visual acuity testing procedures, including contrast testing, dynamic visual acuity testing, and glare testing. The FAA should evaluate these new testing procedures for their possible applicability to aviation. If the use of the new technology were found to be applicable, then perhaps persons who have undergone radial keratotomy, in whom we cannot presently distinguish those who have problems with glare and those who do not, might be given different recommended dispositions regarding certification. Such testing devices would also have application in the evaluation of persons with cataracts.

Ear, Nose and Throat

- The FAA should devise and test the use of an audio cassette tape for speech discrimination, which reflects more accurately the type of speech a pilot hears and the environment of speech discrimination in which the pilot functions. That is, the tape should be a series of instructions from an air traffic control tower as they would be heard on a cockpit radio. The pilot-applicant would be expected to understand and repeat exactly the series of instructions on the tape.

- The FAA should study the environmentally-induced hearing loss of pilots by reviewing serial audiograms.

Cardiovascular System

- The FAA should develop a surveillance program that would test the reliability of risk factor monitoring (blood pressures, electrocardiogram findings, serum lipids, etc) to predict sudden, incapacitating cardiovascular

events. This information would guide future judgments regarding the need for and extent of risk factor screening.

- The FAA should carefully monitor the progress of pilots with special issuances, especially for myocardial infarction, coronary artery bypass surgery, coronary angioplasty and arrhythmias. With this information the validity of the special issuance recommendaions and practices could be assessed.

- With the assistance of the airline industry and pilots' organizations, the FAA should develop a program to educate airmen about the risk factors for and the prevention of cardiovascular and other preventable diseases, which may adversely affect public safety, pilot safety and the ability of a pilot to enjoy operating aircraft to the fullest.

- Because of the ever-increasing knowledge of the prevention, diagnosis, treatment, rehabilitation and monitoring of cardiovascular diseases, the FAA should review and upgrade its cardiovascular standards, guidelines and practices every five years.