COMMITTEE REPORT

Jones Criteria (Revised) for Guidance in the Diagnosis of Rheumatic Fever

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Supporting Evidence of Preceding Streptococcal Infection (Increased ASO or other streptococcal antibodies; positive throat culture for Group A streptococcus; recent scarlet fever)

The presence of two major criteria, or of one major and two minor criteria, indicates a high probability of the presence of rheumatic fever if supported by evidence of a preceding streptococcal infection. The absence of the latter should make the diagnosis doubtful, except in situations in which rheumatic fever is first discovered after a long latent period from the antecedent infection (e.g., Sydenham's chorea or low-grade carditis).

RHEUMATIC FEVER is a sequel to a Group A streptococcal infection, but its pathogenesis is unknown. There is no single laboratory test, symptom, or sign which is pathognomonic of the disease, although several combinations of them are diagnostic. Various clinical manifestations of rheumatic fever are considered to be part of the same disease because they occur together with a frequency that far exceeds chance. They may occur singly, however, or in various combinations in any individual patient.

The diagnostic criteria originally proposed by Jones have proved valuable in preventing overdiagnosis and have been retained. However, some patients present a clinical syndrome which fulfills the original Jones Criteria, but which may not

*By the same token, other streptococcal sequels, typically acute glomerulonephritis, are not considered part of rheumatic fever because they seldom coexist with polyarthritis, carditis, etc.

†Jones, T. D.: Diagnosis of Rheumatic Fever, JAMA 126: 481-484 (Oct. 21) 1944. These criteria were subsequently modified by a Committee of the American Heart Association in 1955.

Circulation, Volume XXXII, October 1963
be due to rheumatic fever. Acute polyarthritis presents, the most common problem. In this revision, therefore, the importance of establishing antecedent streptococcal infection has been emphasized.

The criteria are designed to establish the diagnosis in patients during the acute stage of rheumatic fever. The categories into which clinical and laboratory criteria have been divided are based on the diagnostic importance of a particular finding. They do not relate to prognosis or severity of the disease. They are not a means of measuring rheumatic activity nor of establishing the diagnosis of inactive rheumatic heart disease.

The presence of two major criteria, or of one major and two minor criteria, indicates a high probability of the presence of rheumatic fever if supported by evidence of a preceding streptococcal infection. The absence of the latter should always make the diagnosis doubtful, except in specific situations described in the section on "Supporting Evidence of Streptococcal Infection." Because the prognosis may differ according to the major manifestations, the diagnosis of rheumatic fever should be followed by a list of the major manifestations present, e.g., rheumatic fever, manifested by polyarthritis and carditis. Also, an indication of the severity of carditis in terms of presence or absence of congestive heart failure and cardiomegaly is advisable.

In addition to the criteria to be used in the recommended formula, other manifestations have been listed which may support the diagnosis. These criteria are not meant to substitute for the judgment of the clinician. They are designed to guide him in the diagnosis of the disease, with the suggestion that he follow questionable cases carefully and restrict diagnosis to illnesses which meet acceptable criteria (see "Over-diagnosis of Rheumatic Fever"). In addition to the anxiety it creates, the diagnosis of rheumatic fever implies the indication for prolonged chemophylaxis.*

Major Manifestations

Carditis

Rheumatic carditis is almost always associated with a significant murmur. Consequently, the other manifestations listed below, when not associated with a significant murmur, should be labeled rheumatic carditis with caution.

Murmurs

1. In an individual without previous rheumatic fever or rheumatic heart disease, a significant apical systolic murmur, apical mid-diastolic murmur, or basal diastolic murmur.

2. In an individual with previous rheumatic fever or rheumatic heart disease a definite change in the character of any of these murmurs or the appearance of a new significant murmur.

Cardiomegaly

Unequivocal cardiac enlargement in an individual without a history of previous rheumatic fever, or an obvious increase in cardiac size in a patient with a past history of rheumatic heart disease.

Pericarditis

Manifested by a friction rub, pericardial effusion, or definite electrocardiographic evidence.

Congestive Heart Failure

In a child or young adult in the absence of other discernible causes.

Polyarthritis

Polyarthritis is almost always migratory and is manifested by swelling, heat, redness and tenderness, or by pain and limitation of motion, of two or more joints. (Arthralgia alone, without other evidence of joint involvement, may occur in rheumatic fever, but is not considered a major manifestation.)

Chorea

Purposeless, involuntary, rapid movements often associated with muscle weakness are characteristic of chorea. These must be differentiated from tics, athetosis, and restlessness. Chorea is a delayed manifestation of rheumatic fever, and other rheumatic manifestations may or may not be present.

Erythema Marginatum

This evanescent, pink rash is characteristic of rheumatic fever. The erythematous areas often have pale centers and round or serpiginous margins. They vary greatly in size and occur mainly on the trunk and proximal part of the extremities, never on the face. The erythema is transient, migrates from place to place, and may be brought out by the application of heat. It is non-pruritic, not indurated, and blanches on pressure.

Subcutaneous Nodules

These firm, painless nodules are seen or felt over the extensor surface of certain joints, particularly elbows, knees, and wrists, in the occipital region, or over the spinous processes of the thoracic and lumbar vertebrae. The skin overlying them moves freely and is not inflamed.

Minor Manifestations

Clinical

These are clinical features which occur fre-
quent in rheumatic fever. Because they also occur in many other diseases, their diagnostic value is minor. Their usefulness consists in supporting the diagnosis of rheumatic fever when this diagnosis rests mainly on a single major manifestation.

History of previous rheumatic fever or evidence of pre-existing rheumatic heart disease increases the index of suspicion in evaluating any rheumatic complaint. The history must be well-documented, or the evidence of pre-existing rheumatic heart disease clear-cut.

Arthralgia constitutes pain in one or more joints (not in the muscles and other periarticular tissues) without evidence of inflammation, tenderness to touch, or limitation of motion. The presence of arthralgia, in addition to polyarthritis, does not make the latter any more indicative of rheumatic fever, but in the presence of monoarticular arthritis, arthralgia in other joints strengthens the diagnosis of rheumatic fever.

Fever—temperature in excess of 100.4° F. (38° C.) rectally—is usually present early in the course of untreated rheumatic fever.

Laboratory

The acute phase reactants offer objective but nonspecific confirmation of the presence of an inflammatory process. The erythrocyte sedimentation rate and C-reactive protein test are most commonly employed. Unless the patient has received corticosteroids or salicylates, these tests are almost always abnormal in patients who present with polyarthritis or acute carditis, whereas they are often normal in patients presenting with chorea.

The erythrocyte sedimentation rate (ESR) may be markedly increased by anemia and may be decreased in congestive heart failure. The C-reactive protein (CRP) test is a sensitive indicator of inflammation and is negative in uncomplicated anemia. Heart failure, due to any cause, is often accompanied by a positive CRP test. Sera from normal individuals do not contain this protein, but relatively minor inflammatory stimuli may result in a positive reaction. Leukocytosis, anemia, or other nonspecific responses to inflammation may also occur in acute rheumatic fever.

Electrocardiographic changes, mainly P-R interval prolongation, are frequent, but may occur in other inflammatory processes. Furthermore, ECG changes that are not associated with clinical evidence of carditis (see “Major Manifestations”) have a benign prognosis with regard to the ultimate development of rheumatic heart disease. Such changes by themselves, therefore, do not constitute adequate criteria for carditis.

Supporting Evidence of Streptococcal Infection

The diagnosis of acute rheumatic fever should never be made solely on the basis of laboratory findings plus minor clinical manifestations. On the other hand, since laboratory indications of recent streptococcal infection and current inflammation occur so regularly with this disease, their unexplained absence should make the physician question the diagnosis of rheumatic fever.

Laboratory Evidence of Preceding Streptococcal Infection—by specific antibody tests or by identification of the offending organism—greatly strengthens the possibility of acute rheumatic fever.

Streptococcal Antibody Tests

The most reliable evidence of a specific infection capable of producing acute rheumatic fever is an increased or, even better, a rising streptococcal antibody titer. These titers differentiate preceding streptococcal from other acute respiratory infections and are increased following asymptomatic as well as symptomatic streptococcal infections.

These antibody levels are generally increased in the early stages of acute rheumatic fever, but may be declining, or low, if the interval from the acute streptococcal infection to the detection of rheumatic fever has been longer than two months. This occurs most often in patients whose presenting rheumatic manifestation is chorea. Also, patients whose only manifestation is rheumatic carditis may have low antibody titers when first seen. Their rheumatic attack may have been in progress several months before becoming symptomatic and thus recognized. Except in the latter two instances, one should be reluctant to make the diagnosis of acute rheumatic fever in the absence of serological evidence of a recent streptococcal infection.

The antistreptolysin O test (ASO) is the most widely used and best standardized streptococcal antibody test. In general, single titers of at least 250 Todd units in adults and at least 333 units in children over five years of age are considered to be increased. Depending on the general prevalence of streptococcal infections, a varying per cent of the normal population may show titers of this magnitude.

About 20 per cent of patients in the early stages of acute rheumatic fever, and most patients who present with chorea, have a low or borderline ASO titer. In these instances, it is advisable to obtain another streptococcal anti-
body test.* When two or more different streptococcal antibody tests are performed, it is possible to show an increased titer in almost all cases of acute rheumatic fever within the first two months of onset, and in about half the cases presenting with chorea. Antibody determinations on serum samples obtained at two-week intervals, preferably performed at the same time, are very useful in documenting a streptococcal infection, especially in patients with very low preinfection titers. A rise in titer of two dilution tubes or more can be demonstrated for at least one of the streptococcal antibodies in almost all recurrent, as well as primary attacks of rheumatic fever.

Isolation of Group A Streptococci

Many patients continue to harbor Group A streptococci at the onset of acute rheumatic fever, but these organisms are usually present in small numbers and may be difficult to isolate by a single throat culture. Their demonstration may require special techniques. The administration of penicillin or other antibiotics may also result in failure to isolate the infecting organism. In addition, a significant number of normal individuals, particularly children, may harbor Group A streptococci in the upper respiratory tract. For these reasons, throat cultures† are less satisfactory than antibody tests as supporting evidence of recent streptococcal infection.

Clinical Evidence of Preceding Streptococcal Infection by a history of a recent attack of scarlet fever is the best clinical indication of antecedent streptococcal infection.

Other Clinical Features

These include abdominal pain, rapid sleeping pulse rate (tachycardia out of proportion to fever), malaise, anemia, epistaxis, and precordial pain. They may be even more common in other diseases than they are in rheumatic fever so their usefulness is less than that of the Minor Criteria. Although they are not to be considered diagnostic, they provide additional evidence of the presence of rheumatic fever, as does a family history of rheumatic fever.

Combinations of major and minor manifestations and features may be caused by other diseases which may have to be ruled out before a definitive diagnosis of rheumatic fever is made. One combination in particular—polymyositis, fever, and elevated sedimentation rate—is common in a variety of other disorders. Diseases to be ruled out include rheumatoid arthritis, systemic lupus erythematosus, subacute bacterial endocarditis, serum sickness (including manifestations of penicillin hypersensitivity), gonococcal arthritis, sickle cell anemia, viral pericarditis or myocarditis, leukemia, tuberculosis, undulant fever, and septicemia, particularly meningococceemia. Most of these diseases can be diagnosed with assurance by appropriate tests. Streptococcal antibody determinations are often useful in these differential diagnoses, especially in stimulating the search for other causes when they show no increase.

Murmurs Indicating Carditis

Significant Apical Systolic Murmur (Mitral Regurgitation)

This is a long murmur, filling most of systole. Its blowing quality and high pitch are its most important characteristics. It is heard best in the apical region and is transmitted toward the axilla. The intensity of the murmur is variable, particularly in the early stages of illness, but is at least of grade two on a scale of six. It does not change substantially with position or respiration.

The murmur of mitral regurgitation must be differentiated from functional (innocent) murmurs which frequently occur in normal individuals, especially children. Functional murmurs usually occupy only a portion of systole. They may be quite loud, particularly in anxious or febrile patients, and are rather widely transmitted in thin-chested individuals. These murmurs are heard at times only intermittently and tend to vary with position and respiration. They are usually of two types: an ejection type murmur heard best over the pulmonic area, and a low-pitched, vibratory, groaning or musical murmur heard best along the lower left sternal border. The former is frequently transmitted to the neck and may be mistaken for aortic stenosis. The latter is frequently transmitted to the apex and is most likely to be confused with mitral regurgitation by those unfamiliar with its characteristic quality.

Apical Mid-Diastolic Murmur

Mitrail regurgitation and cardiac dilatation accentuate the third heart sound as a result of

*Available secondary tests include the antihyaluronidase (ASH) and antistreptokinase (ASK) tests which have been in use for some time, but are difficult to standardize from laboratory to laboratory, and the new antidesoxyribonuclease B (anti-DNase B) and anti-diphosphopyridine nucleotidase (anti-DPNase) or anti-nicotinamide adenine dinucleotidase (anti-NADase) tests which may be easier to standardize.

†A separate brochure, entitled "A Method of Culturing Beta Hemolytic Streptococci from the Throat," is available from the American Heart Association.
rapid flow of blood from atrium to ventricle in diastole. During tachycardia, this may produce a protodiastolic gallop rhythm. Frequently, however, in acute rheumatic fever with marked mitral regurgitation, the third heart sound is followed, or replaced, by a low-pitched diastolic rumble. This can be heard best with the patient in the left lateral recumbent position with the breath held in expiration. The same murmur may occur in other forms of acute carditis or in conditions causing rapid blood flow into the left ventricle, such as left-to-right shunts, hyperthyroidism, sickle cell and other forms of severe anemia, etc. It must be differentiated from the low-pitched, crescendo apical presystolic murmur followed by an accentuated mitral first heart sound, which is indicative of an established mitral stenosis rather than of acute carditis.

**Basal Diastolic Murmur**

This murmur of aortic regurgitation begins early in diastole. It is high-pitched, blowing, decrescendo, and is heard best along the left sternal border after deep expiration with the patient leaning forward. It is of great diagnostic importance, but may be difficult to hear and may be present only intermittently.

**Over-Diagnosis of Rheumatic Fever**

Following a well-documented streptococcal infection, the conscientious physician may note suggestive evidence of rheumatic fever, such as, vague pains in the extremities, borderline temperature elevations, increased intensity of a functional murmur, tachycardia during the physical examination of an anxious or hyperactive patient, an increased erythrocyte sedimentation rate, and prolonged P-R interval in the electrocardiogram. Follow-up of such patients has not revealed the delayed appearance of rheumatic heart disease. In the vast majority of cases, significant murmurs of rheumatic carditis appear within the first few weeks of the disease; very rarely do they appear later than three months after the onset of the rheumatic attack, and almost never after six months. Patients without significant cardiac murmurs during acute rheumatic fever have a good chance to escape rheumatic valvular disease.

The diagnosis of acute rheumatic fever should be made, therefore, with conservatism and with insistence upon clearly expressed major clinical manifestations. A common error is the premature, vigorous administration of corticosteroids or salicylates before the signs and symptoms of rheumatic fever are unmistakable. This often leaves an ill-defined syndrome, only presumptively rheumatic fever, and the subsequent management of the patient, particularly the indications for long-term chemoprophylaxis, is in doubt. In the absence of a curative agent, one should not suppress the signs and symptoms of rheumatic fever until they are clearly expressed.

*Ad hoc Committee to revise the Jones Criteria (Modified) of the Council on Rheumatic Fever and Congenital Heart Disease of the American Heart Association.*

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Circulation. 1965;32:664-668
doi: 10.1161/01.CIR.32.4.664
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1965 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/32/4/664.citation

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