AHA Scientific Statement

Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography

A Scientific Statement From the American Heart Association

Endorsed by the World Heart Federation

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Background—Acute rheumatic fever remains a serious healthcare concern for the majority of the world's population despite its decline in incidence in Europe and North America. The goal of this statement was to review the historic Jones criteria used to diagnose acute rheumatic fever in the context of the current epidemiology of the disease and to update those criteria to also take into account recent evidence supporting the use of Doppler echocardiography in the diagnosis of carditis as a major manifestation of acute rheumatic fever.

Methods and Results—To achieve this goal, the American Heart Association's Council on Cardiovascular Disease in the Young and its Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee organized a writing group to comprehensively review and evaluate the impact of population-specific differences in acute rheumatic fever presentation and changes in presentation that can result from the now worldwide availability of nonsteroidal anti-inflammatory drugs. In addition, a methodological assessment of the numerous published studies that support the use of Doppler echocardiography as a means to diagnose cardiac involvement in acute rheumatic fever, even when overt clinical findings are not apparent, was undertaken to determine the evidence basis for defining subclinical carditis and including it as a major criterion of the Jones criteria. This effort has resulted in the first substantial revision to the Jones criteria by the American Heart Association since 1992 and the first application of the Classification of Recommendations and Levels of Evidence categories developed by the American College of Cardiology/American Heart Association to the Jones criteria.

Conclusions—This revision of the Jones criteria now brings them into closer alignment with other international guidelines for the diagnosis of acute rheumatic fever by defining high-risk populations, recognizing variability in clinical presentation in these high-risk populations, and including Doppler echocardiography as a tool to diagnose cardiac involvement. **(*Circulation*. 2015;131:1806-1818. DOI: 10.1161/CIR.0000000000000205.)**

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Although acute rheumatic fever (ARF) has declined in Europe and North America in incidence over the past 4 to 6 decades, the disease remains one of the most important causes of cardiovascular morbidity and mortality among socially and economically disadvantaged populations all over the world, especially in the developing countries that are home to the majority of the world's population. Incidence rates in these countries still reach epidemic levels.[1] The Jones criteria, used for guidance in the diagnosis of ARF since 1944, were last modified by the American Heart Association (AHA) in 1992.[2] They were reconfirmed in principle at an AHA-sponsored workshop in 2000[3] and historically have represented the clinical standard to establish the diagnosis of ARF. However, in the past few years, developments in several areas have prompted reexamination of the traditional Jones criteria. For example, the limited diagnostic role for echocardiography in the diagnosis of carditis as expressed in the Jones criteria revision of 1992[2] is a major area of focus. This position may no longer be appropriate, because echocardiographic techniques and applications, including quantitative Doppler and color flow mapping, have evolved worldwide during the past 2 decades. Other national and regional guidelines for the diagnosis of ARF have recently included the use of echocardiography/Doppler methodologies.[4,5] Numerous studies from

Table 1. Applying Classification of Recommendations and Level of Evidence.

ESTIMATE OF CERTAINTY (PRECISION) OF TREATMENT EFFECT	SIZE OF TREATMENT EFFECT: CLASS I *Benefit >>> Risk* Procedure/Treatment **SHOULD** be performed/administered	CLASS IIa *Benefit >> Risk* *Additional studies with focused objectives needed* **IT IS REASONABLE** to perform procedure/administer treatment	CLASS IIb *Benefit ≥ Risk* *Additional studies with broad objectives needed; additional registry data would be helpful* Procedure/Treatment **MAY BE CONSIDERED**	CLASS III *No Benefit* or CLASS III *Harm* Procedure/Test / Treatment COR III: No benefit — Not Helpful / No Proven Benefit COR III: Harm — Excess Cost w/o Benefit or Harmful / Harmful to Patients
LEVEL A Multiple populations evaluated* Data derived from multiple randomized clinical trials or meta-analyses	■ Recommendation that procedure or treatment is useful/effective ■ Sufficient evidence from multiple randomized trials or meta-analyses	■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from multiple randomized trials or meta-analyses	■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from multiple randomized trials or meta-analyses	■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Sufficient evidence from multiple randomized trials or meta-analyses
LEVEL B Limited populations evaluated* Data derived from a single randomized trial or nonrandomized studies	■ Recommendation that procedure or treatment is useful/effective ■ Evidence from single randomized trial or nonrandomized studies	■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from single randomized trial or nonrandomized studies	■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from single randomized trial or nonrandomized studies	■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Evidence from single randomized trial or nonrandomized studies
LEVEL C Very limited populations evaluated* Only consensus opinion of experts, case studies, or standard of care	■ Recommendation that procedure or treatment is useful/effective ■ Only expert opinion, case studies, or standard of care	■ Recommendation in favor of treatment or procedure being useful/effective ■ Only diverging expert opinion, case studies, or standard of care	■ Recommendation's usefulness/efficacy less well established ■ Only diverging expert opinion, case studies, or standard of care	■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Only expert opinion, case studies, or standard of care
Suggested phrases for writing recommendations	should is recommended is indicated is useful/effective/beneficial	is reasonable can be useful/effective/beneficial is probably recommended or indicated	may/might be considered may/might be reasonable usefulness/effectiveness is unknown/unclear/uncertain or not well established	COR III: No Benefit is not recommended is not indicated should not be performed/administered/other is not useful/beneficial/effective COR III: Harm potentially harmful causes harm associated with excess morbidity/mortality should not be performed/administered/other
Comparative effectiveness phrases†	treatment/strategy A is recommended/indicated in preference to treatment B treatment A should be chosen over treatment B	treatment/strategy A is probably recommended/indicated in preference to treatment B it is reasonable to choose treatment A over treatment B		

A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Although randomized trials are unavailable, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as sex, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use.

†For comparative effectiveness recommendations (Class I and IIa; Level of Evidence A and B only), studies that support the use of comparator verbs should involve direct comparisons of the treatments or strategies being evaluated.

a broad range of clinical circumstances have suggested that there be more widespread use of echocardiography as a way to diagnose carditis even in the absence of overt clinical findings ("subclinical carditis").[6–30] Furthermore, echocardiography has become a cornerstone in worldwide screening programs to evaluate the prevalence of rheumatic heart disease (RHD).[31–35]

In addition to consideration of the proper role of echocardiography in ARF, issues have been raised regarding other clinical areas. For example, whereas in the 1992 version of the Jones criteria,[2] monoarticular arthritis was offered for consideration when a patient had been treated with nonsteroidal anti-inflammatory drugs before diagnosis, evidence has been published since then that indicates that in selective high-risk populations, monoarticular arthritis may be an indicator of the major manifestation of arthritis.[36] Furthermore, previous AHA ARF guidelines did not categorize recommendations using the currently favored Classification of Recommendations and Levels of Evidence categories. The writing group was charged with the task of performing an assessment of the evidence and assigning a Classification of Recommendation according to the American College of Cardiology/AHA classification system.[37] The Classification of Recommendations is an estimate of the size of the treatment effect that considers risks versus benefits in addition to evidence and/or agreement that a given treatment or procedure is or is not useful/effective or, alternatively, may cause harm. The Level of Evidence is an estimate of the certainty or precision of the treatment effect. The writing group reviewed and ranked evidence supporting each recommendation, with the weight of evidence ranked as Level of Evidence A, B, or C according to specific definitions that are included in Table 1. For conditions for which inadequate data are available, recommendations are based on expert consensus and clinical experience and are ranked as Level of Evidence C. This system also provides suggested phrases for writing recommendations within each Classification of Recommendations.

Finally, recent perspectives regarding the diagnosis of acute streptococcal pharyngitis itself, as reviewed in the AHA scientific statement of 2009,[38] need to be referenced as part of the discussion regarding in whom the diagnosis of ARF can be established.

As with past AHA statements concerning the Jones criteria, this revision focuses on the diagnosis of ARF and not on issues concerning the surveillance for and diagnosis of chronic RHD or its consequences.

Epidemiological Background

Insight into how to best define the appropriate application of diagnostic criteria for ARF within a given population requires a brief review of the current epidemiology of ARF.

It is well established that during the 20th century, the incidence of ARF and the prevalence of RHD declined substantially in Europe, North America, and developed nations in other geographic locations.[39,40] This decline has been attributed to improved hygiene, improved access to antibiotic drugs and medical care, reduced household crowding, and other social and economic changes.[39,41] Changes in the epidemiology of specific group A streptococcal strains that cause infections may also have played a role.[42] Although sporadic cases of ARF continue to be seen in affluent nations, the major burden is currently found in low- and middle-income countries and in selected indigenous populations elsewhere. The pattern of disease in the high-prevalence regions is often hyperendemic, with cases occurring throughout the year and a virtual absence of outbreaks. This is in contrast to high-income settings, which experience a low background incidence of ARF with periodic outbreaks.[28,43]

There is also evidence of differences in incidence even in populations within the same country, which further demonstrates the disproportional disease burden. For example, although the overall mean incidence of ARF in New Zealand rose by 55% over the past 2 decades, the incidence of ARF among the non-Maori/Pacific New Zealand populations declined by 70% over the same period.[44] Similar discrepancies in disease burden exist in Australia, where the indigenous population experiences one of the world's highest reported incidences of ARF at 153 to 380 cases per 100 000 people per year in the 5- to 14-year-old age group,[45] whereas in other Australian populations, the incidence approximates European and North American levels.

In summary, the global distribution of ARF/RHD is clearly disproportionate. Certain geographic regions and specific ethnic and socioeconomic groups experience very high rates of ARF incidence, whereas in other regions, the disease has virtually disappeared. This has led to concern regarding the uniform sensitivity of the Jones criteria, even as revised over the years, when applied to geographic areas or to populations within those areas, or elsewhere, where ARF is hyperendemic.

Implications of Epidemiological Considerations

Because the clinical utility of a diagnostic test is determined by a number of factors, including its pretest probability and background disease prevalence, and in view of the heterogeneity in global disease burden noted above, a single set of diagnostic criteria may no longer be sufficient for all population groups and in all geographic regions. To avoid overdiagnosis in low-incidence populations and to avoid underdiagnosis in high-risk populations, variability in applying diagnostic criteria in low-risk compared with high-risk populations is reasonable, as has been promulgated by the Australian rheumatic fever guidelines.[4] The epidemiological data appear to indicate the following:

1. **It is reasonable to consider individuals to be at low risk for ARF if they come from a setting or population known to experience low rates of ARF or RHD *(Class IIa; Level of Evidence C)*.**
2. **It is reasonable that where reliable epidemiological data are available, low risk should be defined as having an ARF incidence <2 per 100 000 school-aged children (usually 5–14 years old) per year or an all-age prevalence of RHD of ≤1 per 1000 population per year *(Class IIa; Level of Evidence C)*.**
3. **Children not clearly from a low-risk population are at moderate to high risk depending on their reference population *(Class I; Level of Evidence C)*.**

Clinical Manifestations of ARF

Generally, the clinical profile of ARF in low- and middle-income countries closely resembles that of high-income countries.[46–48] Universally, the most common major manifestations during the first episode of ARF (the "major criteria" for diagnosis) remain carditis (50%–70%) and arthritis (35%–66%).[1,9,28,46–48] These

are followed in frequency by chorea (10%–30%), which has been demonstrated to have a female predominance, and then subcutaneous nodules (0%–10%) and erythema marginatum (<6%), which remain much less common but highly specific manifestations of ARF.[9,46–48] Despite this general consistency for each of the classic major manifestations, recent data have suggested the possibility of substantial variability of manifestations in specific circumstances and populations.

For example, in very high-risk populations, such as the indigenous Australian population, variability in typical Jones criteria manifestations has been described.[9,36,45] As discussed below, these include presentations with aseptic monoarthritis, polyarthralgia, and low-grade (as opposed to traditionally considered high-grade) fevers. These variable manifestations were reinforced in the 2012 Australian criteria[4] to increase the sensitivity of diagnosis in patients from those specific high-risk populations. To date, however, the applicability of these variable clinical manifestations in low-risk populations has not been tested and is not recommended.

In general, it remains standard practice to maintain continuing vigilance in the application of the clinical manifestations for the diagnosis of ARF. Ongoing reassessment of evolving clinical information is important in any specific patient, because there always has been the potential for "diagnosis overlap" in application of the Jones criteria. In addition to the above, much attention has also been focused on the appropriate role of noninvasive cardiac imaging, namely, echocardiography combined with Doppler flow assessment, in the diagnosis of carditis in ARF.

Carditis: Diagnosis in the Era of Widely Available Echocardiography

Classically, as discussed in the 1992 AHA revised Jones criteria statement, carditis as a major manifestation of ARF has been a clinical diagnosis based on the auscultation of typical murmurs that indicate mitral or aortic valve regurgitation, at either valve or both valves. Thus, although the carditis of ARF has been considered to be a pancarditis and can involve the endocardium, myocardium, and pericardium, valvulitis is by far the most consistent feature of ARF, and isolated pericarditis or myocarditis should rarely, if ever, be considered rheumatic in origin. Clinical carditis remains universally accepted as a major manifestation in all populations; however, on the basis of emerging evidence, several issues have come to prominence that require at least some modification of the classic view. In addition, in an era when clinical auscultatory skills may be declining at the same time that widespread availability of reliable cardiac ultrasound is increasing, echocardiography is being used increasingly to diagnose carditis. Thus, the concept of subclinical carditis has become incorporated into other guidelines and consensus statements as a valid rheumatic fever major manifestation,[4,5] as shown in Table 2.

Subclinical carditis refers exclusively to the circumstance in which classic auscultatory findings of valvar dysfunction either are not present or are not recognized by the diagnosing clinician but echocardiography/Doppler studies reveal mitral or aortic valvulitis. The development of these echocardiographic findings and the rationale for their application to help identify changes in valvar status associated with ARF are discussed below. These changes are listed in Tables 3 and 4 and are analogous to valvular abnormalities also described in RHD in the recent World Heart Federation statement on that condition.[51]

Clinical Studies Assessing the Role of Echocardiography

Numerous studies over the past 20 years have addressed the role of echocardiography (compared with purely clinical assessment) in the diagnosis of ARF. Specific reports (with a minimum of 20 cases of ARF) are reviewed in Table 5. In general, >25 studies have reported echocardiography/Doppler evidence of mitral or aortic valve regurgitation in patients with ARF despite the absence of classic auscultatory findings. These studies have included various geographic locations and population characteristics. The reports of the ARF outbreak in Utah were among the first in a developed world population to indicate the validity of Doppler echocardiography in diagnosing carditis in ARF.[23] In contrast to all of these reports, during the same time period, only 1 study found that echocardiography had no incremental diagnostic utility in patients without traditional, clinically evident carditis.[25] In support of the findings of these multiple single studies is a meta-analysis of subclinical carditis in ARF.[52] The prevalence of subclinical carditis ranged from 0% (1 study) to 53% in this review of 23 articles. The weighted pooled prevalence of subclinical carditis was 16.8% (95% confidence interval 11.9%–21.6%). This increased slightly to 18.1% when the analysis was limited to the 10 studies that used the full World Health Organization[49] criteria. The weighted pooled persistence or worsening of carditis in patients with subclinical carditis was 44.7% (95% confidence interval 19.3%–70.2%).[52] The

Table 2. Evolving Role of Echocardiography in the Diagnosis of ARF

Year	Guidelines	Perform Echo in All Confirmed Cases of ARF Without Clinical Carditis?	Perform Echo in All Suspected Cases of ARF?	Use Echo to Confirm Carditis as Major Criterion in Absence of Murmur?
1992	Jones criteria 1992[2]	No	No	No
2000	Jones Criteria Workshop[3]	No	No	No
2001	WHO guidelines[49]	Yes	No	No
2008	Indian Working Group[50]	Yes*	No	No
2008	New Zealand guidelines[5]	Yes†	Yes‡	Yes§
2012	Australian guidelines[4]	Yes‖	Yes¶	Yes#

ARF indicates acute rheumatic fever; Echo, echocardiography; and WHO, World Health Organization.

*Importance suggested, but not required.

†Repeat in 2 to 4 weeks if negative in all cases of chorea.

‡Repeat in 2 to 4 weeks as necessary.

§All groups.

‖Repeat serially in cases with chorea.

¶Repeat in 1 month if negative in all cases.

#High-risk populations (see the section Epidemiologic Considerations).

Table 3. Doppler Findings in Rheumatic Valvulitis

Pathological mitral regurgitation (all 4 criteria met)
Seen in at least 2 views
Jet length ≥2 cm in at least 1 view
Peak velocity >3 m/s
Pansystolic jet in at least 1 envelope
Pathological aortic regurgitation (all 4 criteria met)
Seen in at least 2 views
Jet length ≥1 cm in at least 1 view
Peak velocity >3 m/s
Pan diastolic jet in at least 1 envelope

Loading conditions should be accounted for at time of echocardiography/Doppler assessment (see the section Differential Diagnosis of ARF for a full discussion). This table reflects an amalgam of the findings from the references listed in Table 5 and other guideline statements[4,5] and also resembles findings described in rheumatic heart disease.[51]

authors noted, however, that the quality of follow-up data in most studies was poor, with inconsistent follow-up intervals and lack of ongoing follow-up in patients who showed signs of improvement.

Additionally, none of these studies questioned the utility of echocardiography/Doppler for the evaluation of cardiovascular status in patients with ARF confirmed by usual clinical criteria or for its use in long-term management. In sum, aside from the singular 1996 report cited above, all the studies reviewed overwhelmingly support the use of echocardiography/Doppler results as part of the diagnostic criteria for confirmation of the presence of carditis in patients with suspected ARF. Accordingly, this writing group concludes the following:

1. **Echocardiography with Doppler should be performed in all cases of confirmed and suspected ARF *(Class I; Level of Evidence B)*.**
2. **It is reasonable to consider performing serial echocardiography/Doppler studies in any patient with diagnosed or suspected ARF even if documented carditis is not present on diagnosis *(Class IIa; Level of Evidence C)*.**
3. **Echocardiography/Doppler testing should be performed (strictly fulfilling the findings noted in Tables 2 and 3) to assess whether carditis is present in the absence of auscultatory findings, particularly in moderate- to high-risk populations and when ARF is considered likely *(Class I; Level of Evidence B)*.**
4. **Echocardiography/Doppler findings not consistent with carditis should exclude that diagnosis in patients with a heart murmur otherwise thought to indicate rheumatic carditis *(Class I; Level of Evidence B)*.**

Arthritis

Typically, as described in the Jones criteria revision of 1992,[2] the arthritis of ARF is a migratory polyarthritis, and the joints most frequently involved are larger ones, including knees, ankles, elbows, and wrists. A history of rapid improvement with salicylates or nonsteroidal anti-inflammatory drugs is also characteristic. Generally, the arthritis in ARF runs a self-limited course, even without therapy, lasting ≈4 weeks.[53]

Table 4. Morphological Findings on Echocardiogram in Rheumatic Valvulitis

Acute mitral valve changes
Annular dilation
Chordal elongation
Chordal rupture resulting in flail leaflet with severe mitral regurgitation
Anterior (or less commonly posterior) leaflet tip prolapse
Beading/nodularity of leaflet tips
Chronic mitral valve changes: not seen in acute carditis
Leaflet thickening
Chordal thickening and fusion
Restricted leaflet motion
Calcification
Aortic valve changes in either acute or chronic carditis
Irregular or focal leaflet thickening
Coaptation defect
Restricted leaflet motion
Leaflet prolapse

On occasion, particularly early in the course of acute rheumatic fever, mitral or aortic valve morphology may be normal on echocardiogram while Doppler shows regurgitation, as defined in Table 3. These findings can also be seen in chronic rheumatic heart disease.[51]

There is absence of long-term joint deformity. Involvement of small joints of the hands and feet and the spine is much less common in ARF than in other arthritic illnesses.

Reactive Arthritis

In the 1944 original Jones criteria[54] arthralgia was considered to be a major manifestation of ARF, but since the 1956 modification,[55,56] only migratory polyarthritis has been considered to be a major manifestation to fulfill the Jones criteria, and arthralgia has been classified as a minor manifestation. Patients with group A β-hemolytic streptococcal infection and articular disease that does not fulfill the classic Jones criteria for the diagnosis of ARF are sometimes classified as having poststreptococcal reactive arthritis/arthralgia, and currently, there is controversy about secondary prophylaxis for these patients.[57] Some pediatric patients with poststreptococcal reactive arthritis have later developed episodes of ARF or RHD,[58,59] which indicates that the initial diagnosis should probably have been ARF. In contrast, a prospective study in low-risk white adults in the Netherlands demonstrated that poststreptococcal reactive arthritis was not associated with long-term cardiac sequelae.[60]

Aseptic Monoarthritis

Studies from India, Australia, and Fiji have indicated that aseptic monoarthritis may be important as a clinical manifestation of ARF in selected high-risk populations.[9,36,61–64] In the high-risk indigenous Australian population, aseptic monoarthritis has been found to be present in 16% to 18% of confirmed cases of ARF. In this population, according to 1 study,[36] 55% of cases (15/27) who would have satisfied the Jones criteria if monoarthritis had been considered to be a major criterion subsequently developed either ARF or RHD. There has only been 1 North American report of a small case series of aseptic monoarthritis.[65]

Table 5. Studies Reporting Subclinical Carditis

Country (Reference)	No. of Patients With Clinical Carditis/No. With Rheumatic Fever	No. of Patients With Subclinical Carditis/No. Without Clinical Carditis	Criteria Used for Mitral Regurgitation	Criteria Used for Aortic Regurgitation
Turkey[8]	39/80	25/41	2 Planes, jet >1 cm, holosystolic, peak velocity >2.5 m/s	2 Planes, jet >1 cm, holodiastolic, peak velocity >2.5 m/s
Australia[9]	46/98	27/52	2 Planes, jet >1 cm, holosystolic, mosaic jet by color, peak velocity >2.5 m/s	2 Planes, jet >1 cm, holodiastolic, mosaic jet, peak velocity >2.5 m/s
India[7]	220/333	52/113	2 Planes, jet >1 cm, holosystolic, mosaic jet	NS
Brazil[11]	27/56	11/29	Systolic jet into LA	Diastolic jet into LVOT
Pakistan[10]	0/30	21/30	2 Planes, jet >1 cm, holosystolic, mosaic jet, peak velocity >2.5 m/s	2 Planes, jet >1 cm, holodiastolic, mosaic jet, peak velocity >2.5 m/s
Brazil[30]	22/31	9/9	>2 Of the following: 2 planes, jet >1 cm, jet area >1 cm^2, holosystolic, peak velocity >3.2 m/s, flow convergence	Jet wider than 0.1 cm in LVOT, holodiastolic
Nepal[12]	38/51	9/13	2 Planes, jet >1 cm	2 Planes, jet >0.5 cm
India[6]	237/452	116/215	2 Planes, well beyond valve leaflets, holosystolic	2 Planes, well beyond valve leaflets, holodiastolic
Turkey[13]	84/129	19/45	2 Planes, jet >1 cm, holosystolic, mosaic jet	2 Planes, well beyond valve leaflets, holodiastolic
Thailand[14]	17/44	3/27	2 Planes, holosystolic, mosaic jet, high velocity	2 Planes, high velocity, mosaic jet, diastolic
Turkey[15]	NS/189	40/NS	2 Planes, jet >1 cm, holosystolic, peak velocity >2.5 m/s, mosaic jet	2 Planes, holodiastolic, peak velocity >2.5 m/s
Turkey[16]	51/104	23/53	2 Planes, jet >1 cm, holosystolic, peak velocity >2.5 m/s, mosaic posterolaterally directed jet	Holodiastolic, peak velocity >2.5 m/s
Jordan[17]	24/50	4/26	2 Planes, jet >1 cm, mosaic jet	2 Planes, jet >1 cm, mosaic jet
Brazil[18]	28/40	2/12	2 Planes, jet >1 cm, duration >200 ms, peak velocity >2.5 m/s	Jet >1 cm, duration >200 ms, peak velocity >2.5 m/s
Chile[19]	15/35	10/20	2 Planes, holosystolic, mosaic jet	NS
Brazil[20]	8/22	5/14	Mosaic systolic jet in LA (jet area/LA area >20%)	Diastolic jet into LVOT
Turkey[21]	5/22	9/17	Mosaic, 2 planes, holosystolic, high velocity	NS
Brazil[22]	396/786	144/390	NS	NS
United States[23]	68/113	25/37	2 Planes, jet >1 cm, holosystolic, mosaic jet	NS
United States[24]	24/30	2/6	Flow back to LA wall, holosystolic, high velocity, turbulent	NS
India[25]	80/108	0/28	Jet >1 cm, high velocity, turbulent jet	NS
France[26]	50/100	>30/50	At least mild	At least mild
New Zealand[27]	15/47	4/32	Flow well into LA, >80% systole, high velocity	High-velocity diastolic jet
United States[28]	189/274	45/85	Flow back to LA wall, holosystolic, high velocity, turbulent	NS
New Zealand[29]	36/66	20/30	2 Planes, jet well into LA, holosystolic, high velocity	2 Planes, well beyond valve leaflets, high velocity, holodiastolic

LA indicates left atrium; LVOT, left ventricular outflow tract; and NS, not stated.

1. At present, consideration that monoarthritis may be part of the ARF spectrum should be limited to patients from moderate- to high-risk populations *(Class I; Level of Evidence C)*.

Polyarthralgia

Polyarthralgia is a very common, highly nonspecific manifestation of a number of rheumatologic disorders. Until 1956, it was considered to be a major criterion for the diagnosis of

ARF, but as the Jones criteria were modified over the decades to fulfill Dr Jones' original intention not to overdiagnose ARF, polyarthralgia was reclassified as a minor manifestation. The present writing group has not found compelling evidence to amend this conclusion in low-risk populations.

As noted previously, arthritis caused by ARF is highly responsive to salicylates and nonsteroidal anti-inflammatory agents, which are now readily available worldwide over the counter and therefore have often been used before clinical evaluation. Use of such drugs before diagnosis may mask the development of the classic migratory nature of polyarthritis and underlines the need for a careful history to be taken in all patients with suspected ARF. Additionally, patients susceptible to develop ARF are often at elevated risk for other infectious and inflammatory diseases that may be associated with arthralgia or arthritis. Therefore, clinicians should be aware of the extensive differential diagnosis for joint problems and should be particularly careful to exclude other causes of arthritis, especially septic arthritis (Table 6).

As noted in other sections of this statement, the positive predictive value of any sign or symptom increases as the incidence of disease increases in the population. Thus, children with polyarthralgia are more likely to have ARF if they come from a population with a high incidence of ARF than if they come from a low-incidence population. In the latter case, the writing group affirmed that polyarthralgia is almost always a symptom of an illness other than ARF and favored retaining polyarthralgia as a minor manifestation for low-risk populations, as per the historic Jones criteria.

1. **The inclusion of polyarthralgia as a major manifestation is applicable only for moderate- or high-incidence populations and only after careful consideration and exclusion of other causes of arthralgia such as autoimmune, viral, or reactive arthropathies (Table 6) *(Class IIb; Level of Evidence C).***

Chorea (Sydenham Chorea)

Chorea in ARF is characterized by purposeless, involuntary, nonstereotypical movements of the trunk or extremities.[66] It often is associated with muscle weakness and emotional lability. Table 6 reviews the differential diagnosis of chorea. In some patients, chorea can be predominantly unilateral and may require careful neurological examination to confirm that other neurological disorders are not present. Huntington chorea, systemic lupus erythematous, Wilson disease, and drug reactions are to be excluded, and the movements should be differentiated from tics, athetosis, conversion reaction, and hyperkinesis. Evidence of a recent group A streptococcal infection may be difficult or impossible to document because of the long latent period between the inciting streptococcal infection and the onset of chorea. Worsening of choreiform movements in a child with previous low-grade residual chorea may be hard to distinguish from a new attack of chorea.

Skin Findings

Erythema marginatum is the unique, evanescent, pink rash seen with pale centers and rounded or serpiginous margins. The rash usually is present on the trunk and proximal extremities and is not facial. Heat can induce its appearance, and it blanches with pressure. As with other rashes, erythema marginatum may be harder to detect in dark-skinned individuals. Subcutaneous nodules are firm, painless protuberances found on extensor surfaces at specific joints, including the knees, elbows, and wrists, and also are seen in the occiput and along the spinous processes of the thoracic and lumbar vertebrae. They have not been found to have racial or population variability. Nodules are more often observed in patients who also have carditis, and as with erythema marginatum, subcutaneous nodules almost never occur as the sole major manifestation of ARF.

Other Clinical Features: Minor Manifestations

In the 1965[56] revision of the Jones criteria, the authors commented that during an episode of ARF, temperature usually exceeds 38°C, and in the 1992 revision,[2] that was revised to 39°C. However, in the aforementioned Aboriginal Australian population, a high-risk population, the definition of fever as a temperature >38°C has resulted in improved sensitivity, with 75% of individuals with ARF meeting this criterion compared with only 25% when a cutoff value of >39°C was used. A cutoff value of >37.5°C would have allowed the diagnosis of fever in 90% of suspected cases of ARF. This is of potential importance, because 41% of individuals in this particular population who were not diagnosed as having ARF because of the absence of fever when defined as 38°C or 39°C subsequently developed ARF or RHD.[36] However, in most settings, including all low-risk populations, fever associated with ARF usually exceeds 38.5°C orally. As with arthritis, the widespread availability of antipyretic agents requires that a detailed history be taken to put the presentation of fever in the proper context.

Generally, there appear to be no differences in other minor clinical manifestations (raised C-reactive protein, erythrocyte sedimentation rate, prolonged PR interval on ECG, a past history of rheumatic fever or RHD) between that of low- and higher-risk populations and geographies.[2,4,45,61] For most populations, an erythrocyte sedimentation rate >60 mm in the first hour and C-reactive protein >3.0 mg/dL are considered typical of ARF.

In ARF, C-reactive protein values should always be higher than the upper limit of normal for any specific laboratory and are commonly >7.0 mg/dL or even higher, depending on the laboratory method used. Some experts, however, consider an erythrocyte sedimentation rate >30 mm/h as consistent with the diagnosis of ARF.[4] Normal erythrocyte sedimentation rate and C-reactive protein levels prompt serious reconsideration of the diagnosis of ARF, because except for patients with isolated chorea, these values are almost never normal in ARF.

Abdominal pain, rapid sleeping pulse rate, tachycardia out of proportion to fever, malaise, anemia, leukocytosis, epistaxis, and precordial pain also may be noted in patients with ARF. Although these clinical and laboratory features are not diagnostic, they are certainly compatible with the presence of ARF. Because these signs and symptoms frequently are noted in many diseases, their usefulness is less

Table 6. Differential Diagnosis of Arthritis, Carditis, and Chorea

Arthritis	Carditis	Chorea
Septic arthritis (including gonococcal)	Physiological mitral regurgitation	Drug intoxication
Connective tissue and other autoimmune diseases such as juvenile idiopathic arthritis	Mitral valve prolapse	Wilson disease
Viral arthropathy	Myxomatous mitral valve	Tic disorder
Reactive arthropathy	Fibroelastoma	Choreoathetoid cerebral palsy
Lyme disease	Congenital mitral valve disease	Encephalitis
Sickle cell anemia	Congenital aortic valve disease	Familial chorea (including Huntington disease)
Infective endocarditis	Infective endocarditis	Intracranial tumor
Leukemia or lymphoma	Cardiomyopathy	Lyme disease
Gout and pseudo gout	Myocarditis, viral or idiopathic	Hormonal
Poststreptococcal reactive arthritis	Kawasaki disease	Metabolic (eg, Lesch-Nyhan, hyperalaninemia, ataxia telangiectasia)
Henoch-Schonlein purpura		Antiphospholipid antibody syndrome
		Autoimmune: Systemic lupus erythematosus, systemic vasculitis
		Sarcoidosis
		Hyperthyroidism

than that of the principal minor manifestations. A family history of rheumatic fever also may heighten the suspicion of this disease.

Evidence of Preceding Streptococcal Infection

Because other illnesses may closely resemble ARF, laboratory evidence of antecedent group A streptococcal infection is needed whenever possible, and the diagnosis is in doubt when such evidence is not available. Exceptions to this include chorea, which may be the only manifestation of rheumatic fever at the time of its presentation, and rarely, individuals with chronic, indolent rheumatic carditis with insidious onset and slow progression. This latter problem refers to patients without an identifiable history of ARF who have had subclinical carditis that was not detected previously, and it may be the only manifestation of prior ARF in a patient who presents with cardiovascular sequelae of an ARF attack at a time remote from the initial episode.[34] Interpretation of streptococcal serology results can be difficult in populations with endemic skin or upper respiratory group A streptococcal infections. In these settings, a negative streptococcal antibody test helps to exclude a recent infection, but a positive test does not necessarily indicate an infection in the past few months.

Any 1 of the following can serve as evidence of preceding infection, per a recent AHA statement[38]:

1. **Increased or rising anti-streptolysin O titer or other streptococcal antibodies (anti-DNASE B) *(Class I; Level of Evidence B)*.[38] A rise in titer is better evidence than a single titer result.**
2. **A positive throat culture for group A β-hemolytic streptococci *(Class I; Level of Evidence B)*.[38]**
3. **A positive rapid group A streptococcal carbohydrate antigen test in a child whose clinical presentation suggests a high pretest probability of streptococcal pharyngitis *(Class I; Level of Evidence B)*.[38]**

Differential Diagnosis of ARF

It is important to have a working differential diagnosis when considering each of the major criteria in the diagnosis of ARF. Table 6, modified from the Australian and New Zealand guidelines,[4,5] provides a list of alternative diagnoses to consider in the evaluation of patients with arthritis, carditis, or chorea. Acceptance of echocardiography-based criteria to diagnose carditis in the absence of clinical findings requires knowledge of other findings that could resemble rheumatic carditis, especially in low-risk populations. The echocardiographic diagnosis of carditis is best made in strict accordance with Tables 3 and 4 referenced above. In this respect, accounting for circulatory loading conditions is considered part of the echocardiographic assessment. Three of the 4 criteria used to diagnose pathological mitral or aortic regurgitation (jet length, velocity, and completeness of the Doppler envelope) are influenced by the systemic blood pressure.[67] Because blood pressure may change rapidly in a febrile or agitated patient, it is reasonable whenever circumstances allow to measure blood pressure at the time of the echocardiogram to recognize the presence of an abnormal circulatory load (high or low) and to include blood pressure data when serial echocardiograms are performed to assist in the appropriate comparison. Other nonrheumatic mitral valve findings to be considered include physiological mitral regurgitation, mitral valve prolapse, myxomatous mitral valve, Barlow syndrome, and congenital mitral valve disease. Endocarditis and annular dilation from conditions associated with left-sided heart dilation, including myocarditis and cardiomyopathy, are also in the differential diagnosis. Continuous-wave Doppler of the mitral regurgitant jet can help discriminate physiological from pathological regurgitation. Signals that are not

Table 7. Revised Jones Criteria

A. For all patient populations with evidence of preceding GAS infection	
Diagnosis: initial ARF	2 Major manifestations or 1 major plus 2 minor manifestations
Diagnosis: recurrent ARF	2 Major or 1 major and 2 minor or 3 minor
B. Major criteria	
Low-risk populations*	Moderate- and high-risk populations
Carditis† • Clinical and/or subclinical	Carditis • Clinical and/or subclinical
Arthritis • Polyarthritis only	Arthritis • Monoarthritis or polyarthritis • Polyarthralgia‡
Chorea	Chorea
Erythema marginatum	Erythema marginatum
Subcutaneous nodules	Subcutaneous nodules
C. Minor criteria	
Low-risk populations*	Moderate- and high-risk populations
Polyarthralgia	Monoarthralgia
Fever (≥38.5°C)	Fever (≥38°C)
ESR ≥60 mm in the first hour and/or CRP ≥3.0 mg/dL§	ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL§
Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)	Prolonged PR interval, after accounting for age variability (unless carditis is a major criterion)

ARF indicates acute rheumatic fever; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; and GAS, group A streptococcal infection.

*Low-risk populations are those with ARF incidence ≤2 per 100 000 school-aged children or all-age rheumatic heart disease prevalence of ≤1 per 1000 population per year.

†Subclinical carditis indicates echocardiographic valvulitis as defined in Table 3.

‡See section on polyarthralgia, which should only be considered as a major manifestation in moderate- to high-risk populations after exclusion of other causes. As in past versions of the criteria, erythema marginatum and subcutaneous nodules are rarely "stand-alone" major criteria. Additionally, joint manifestations can only be considered in either the major or minor categories but not both in the same patient.

§CRP value must be greater than upper limit of normal for laboratory. Also, because ESR may evolve during the course of ARF, peak ESR values should be used.

holosystolic and peak velocity <3.0 m/s are more likely to be physiological than pathological. The mitral valve prolapse seen in ARF patients differs from the redundant, myxomatous mitral valve and prolapse seen with Barlow syndrome.[68] In valvulitis from ARF, only the coapting portion of the anterior mitral valve leaflet tip prolapses, and there is no billowing of the medial portion or body of the leaflet. This leaflet tip prolapse results in abnormal leaflet coaptation, a regurgitant orifice, and a jet of mitral regurgitation that is typically directed posterolaterally.

Isolated congenital mitral valve abnormalities are relatively uncommon but are in the differential diagnosis of newly identified mitral regurgitation. These include cleft mitral valve, double-orifice mitral valve, parachute mitral valve variants, and fibroelastomas. Congenital aortic valve anomalies should be in the differential diagnosis of newly identified aortic regurgitation; however, isolated aortic regurgitation is rarely the sole valvular finding in rheumatic carditis. Congenital diagnoses to consider include bicuspid aortic valve, spontaneously closed ventricular septal defect with aortic valve prolapse, subaortic membrane, and syndromic-related aortic root dilation. Infective endocarditis can be mistaken for rheumatic carditis if there is no obvious vegetation and valve damage has already occurred.

Rheumatic Fever Recurrences

As stated in the 1992 guidelines,[2] patients who have a history of ARF or RHD are at high risk for "recurrent" attacks if reinfected with group A streptococci. Such an attack is considered a new episode of ARF, but one in which the complete set of Jones criteria, even as revised, may not be completely fulfilled.

1. **With a reliable past history of ARF or established RHD, and in the face of documented group A streptococcal infection, 2 major or 1 major and 2 minor or 3 minor manifestations may be sufficient for a presumptive diagnosis *(Class IIb; Level of Evidence C)*.**
2. **When minor manifestations alone are present, the exclusion of other more likely causes of the clinical presentation is recommended before a diagnosis of an ARF recurrence is made *(Class I; Level of Evidence C)*.**

"Possible" Rheumatic Fever

In some circumstances, a given clinical presentation may not fulfill these updated Jones criteria, but the clinician may still have good reason to suspect that ARF is the diagnosis. This may occur in high-incidence settings where, for

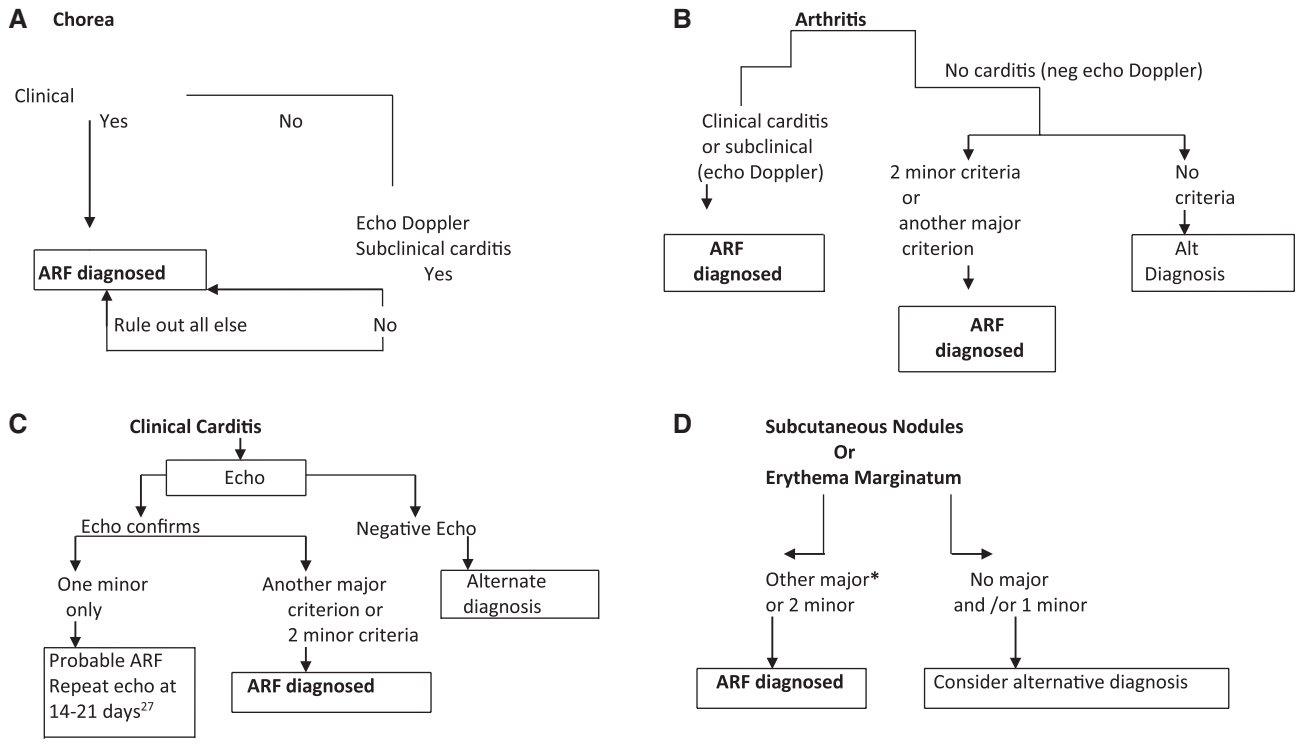

Figure. Diagnosis strategy for acute rheumatic fever. *Subclinical carditis can be considered. Alt indicates alternative; ARF, acute rheumatic fever; echo, echocardiography; GAS, group A streptococcal; and neg, negative.

example, laboratory tests for acute phase reactants or for confirmation of recent streptococcal infection are not available, documentation of clinical features is not clear, or the history is not considered to be reliable. In such situations, clinicians should use their discretion and clinical acumen to make the diagnosis that they consider most likely and manage the patient accordingly.

1. **Where there is genuine uncertainty, it is reasonable to consider offering 12 months of secondary prophylaxis followed by reevaluation to include a careful history and physical examination in addition to a repeat echocardiogram *(Class IIa; Level of Evidence C)*.**
2. **In a patient with recurrent symptoms (particularly involving the joints) who has been adherent to prophylaxis recommendations but lacks serological evidence of group A streptococcal infection and lacks echocardiographic evidence of valvulitis, it is reasonable to conclude that the recurrent symptoms are not likely related to ARF, and discontinuation of antibiotic prophylaxis may be appropriate *(Class IIa; Level of Evidence C)*.**

Impact of Modifications of Jones Criteria in High-Risk Populations

A retrospective study in North Queensland, Australia, investigated the impact of the addition of subclinical carditis, monoarthritis, and low-grade fever (>37.5°C) to the 1992 revised Jones criteria.[36] Of the 98 cases with a clinical diagnosis of ARF, only 71.4% met the revised Jones criteria. Modification of the criteria, as discussed above, increased the proportion of the cases that satisfied diagnostic criteria to 91.8%. Of the 28 people who did not meet the traditional Jones criteria, 12 (42%) developed evidence of chronic RHD. This study, if confirmed, may suggest that the addition of monoarthritis and subclinical carditis as major manifestations and low-grade fever as a minor manifestation to the Jones criteria could increase sensitivity when applied specifically to high-risk populations. Additionally, study of the impact of the application of the New Zealand guidelines resulted in a 16% increase in the diagnosis of ARF compared with the 1992 revision of the Jones criteria.[29] There are no additional data that corroborate these results in populations with a lower incidence of ARF.

In summary, in the context of the previous discussion, revision of the Jones criteria to meet current technological advances and clinical needs is warranted. Thus, strict application of echocardiography/Doppler findings (Tables 3 and 4) may be used to fulfill the major criterion of carditis, even in the absence of classic auscultatory findings, providing that ambient loading conditions are taken into consideration. In addition, monoarthritis or polyarthralgia could be accepted as fulfilling the major criterion of arthritis, but only in moderate- to high-risk populations. For low-risk populations, monoarthritis is not included, and polyarthralgia remains a minor criterion. Similarly, the requirement for the presence of fever can be fulfilled with oral, tympanic, or rectal temperature documented at 38°C in moderate- to high-risk populations, but only at ≥38.5°C in others. The

writing group confirms the appropriateness of retaining the time-honored approach initially advocated by Dr Jones that favors low sensitivity and high specificity in assessing the criteria for the diagnosis of ARF in low-risk populations. Table 7 and the Figure summarize diagnostic strategies using these revised criteria.

Future Considerations

In addition to the broad epidemiological issues and the widespread careful application of echocardiography that have led to the suggested revisions in the Jones criteria described in this statement, recent findings suggesting genetic susceptibility factors in ARF[69–71] may one day point to a totally new set of diagnostic tools. Future revisions should continue to honor Dr Jones' initial goal, particularly in low-risk populations, to avoid overdiagnosis and its consequences.[54]

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Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography: A Scientific Statement From the American Heart Association

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