

Table 1. AHA 2015 statement for revision of Jones criteria. 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

Table 2. AHA 2015 statement for revision of Jones criteria. 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†	Carditis
• Clinical and/or subclinical	• Clinical and/or subclinical
Arthritis	Arthritis
• Polyarthritis only	• 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 ($\geq 38.5^{\circ}\text{C}$)	Fever ($\geq 38^{\circ}\text{C}$)
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.

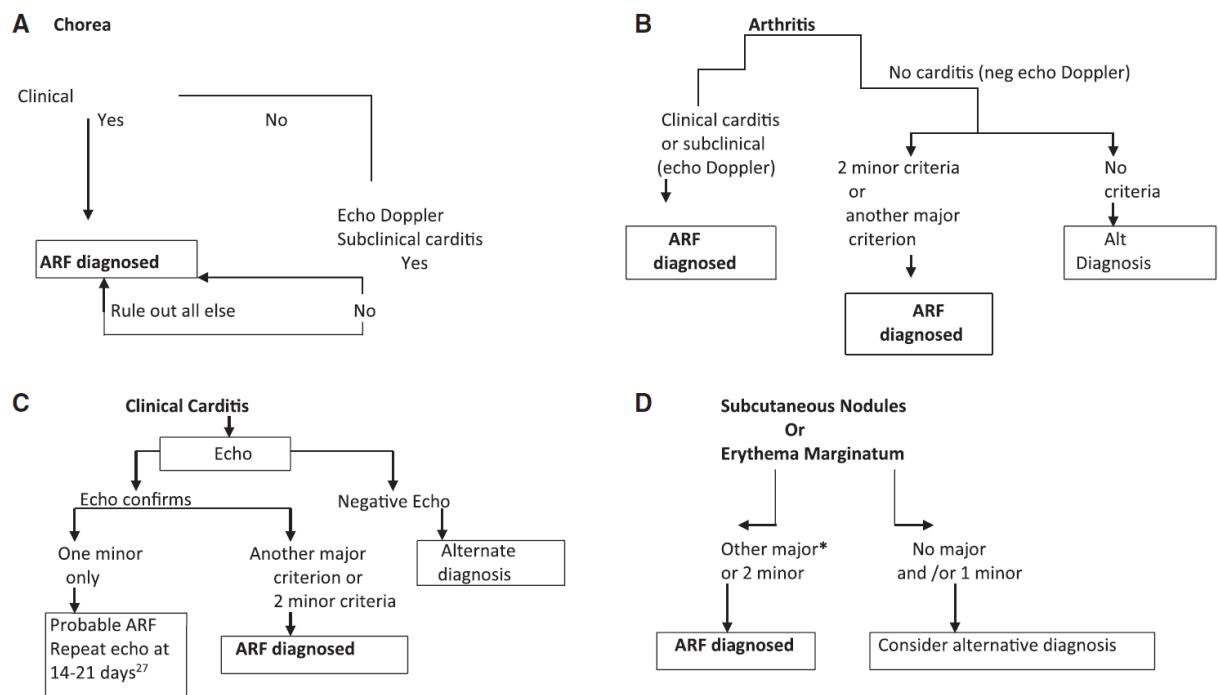
*Low-risk populations are those with ARF incidence ≤ 2 per 100 000 school-aged children or all-age rheumatic heart disease prevalence of ≤ 1 per 1000 population per year.

†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.

Gewitz MH, et al. 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. *Circulation*. 2015;131:1806-18.



B, C, and D require evidence of GAS infection.

Figure 1. AHA 2015 statement for revision of Jones criteria. 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.

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Table 3. AHA 2015 statement for revision of Jones criteria. Echocardiography in ARF

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. These findings can also be seen in chronic rheumatic heart disease.

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.

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Table 4. AHA 2015 statement for revision of Jones criteria. Evidence of preceding streptococcal infection

Increased or rising anti-streptolysin O titer or other streptococcal antibodies (anti-DNASE B) I-B
A rise in titer is better evidence than a single titer result.

A positive throat culture for group A β -hemolytic streptococci I-B

A positive rapid group A streptococcal carbohydrate antigen test in a child whose clinical presentation suggests a high pretest probability of streptococcal pharyngitis I-B

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Table 5. AHA 2009 Statement on rheumatic fever prevention. Primary prevention of rheumatic fever (treatment of streptococcal tonsillopharyngitis)

Agent	Dose	Mode	Duration	Rating
Penicillins				
Penicillin V (phenoxymethylpenicillin)	Children: 250 mg 2–3 times daily for ≤ 27 kg (60 lb); children >27 kg (60 lb), adolescents, and adults: 500 mg 2–3 times daily	Oral	10 days	I-B
	or			
Amoxicillin	50 mg/kg once daily (maximum 1 g)	Oral	10 days	I-B
	or			
Benzathine penicillin G	600 000 U for patients ≤ 27 kg (60 lb); 200 000 U for patients >27 kg (60 lb)	Intramuscular	Once	I-B
For individuals allergic to penicillin				
Narrow-spectrum cephalosporin† (cephalexin, cefadroxil)	Variable	Oral	10 days	I-B
	or			
Clindamycin	20 mg/kg per day divided in three doses (maximum 1.8 g/d)	Oral	10 days	IIa-B
	or			
Azithromycin	12 mg/kg once daily (maximum 500 mg)	Oral	5 days	IIa-B
	or			
Clarithromycin	15 mg/kg per day divided bid (maximum 250 mg bid)	Oral	10 days	IIa-B

† To be avoided in those with immediate (type I) hypersensitivity to a penicillin. sulfonamides, trimethoprim, tetracyclines, and fluoroquinolones are not acceptable alternatives.
* Similar recommendations have been provided by ACC/AHA GL on valve disease.

Table 6. AHA 2015 Statement. Possible Rheumatic Fever

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 IIa-C

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 IIa-C

Table 7. AHA 2015 Statement. Rheumatic Fever Recurrences

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 IIb-C

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. I-C

Table 8. ACC/AH 2020 guideline on valve disease. Secondary Prevention of Rheumatic Fever

Antibiotics for Prevention	Dosage*
Penicillin G benzathine	1.2 million U intramuscularly every 4 wk†
Penicillin V potassium	200 mg orally twice daily
Sulfadiazine	1 g orally once daily
Macrolide or azalide antibiotic (for patients allergic to penicillin and sulfadiazine)‡	Varies

~~Adapted from Gerber et al. (1)~~

*In patients with documented valvular heart disease, the duration of rheumatic fever prophylaxis should be ≥ 10 y or until the patient is 40 y of age (whichever is longer). Lifelong prophylaxis may be recommended if the patient is at high risk of group A streptococcus exposure. Secondary rheumatic heart disease prophylaxis is required even after valve replacement.

†Administration every 3 wk is recommended in certain high-risk situations.

‡Macrolide antibiotics should not be used in persons taking other medications that inhibit cytochrome P450 3A, such as azole antifungal agents, HIV protease inhibitors, and some selective serotonin reuptake inhibitors.

Table 9. ACC/AH 2020 guideline on valve disease. Duration of Secondary Prophylaxis for Rheumatic Fever

Type	Duration After Last Attack*
Rheumatic fever with carditis and residual heart disease (persistent VHD †)	10 y or until patient is 40 y of age (whichever is longer)
Rheumatic fever with carditis but no residual heart disease (no valvular disease †)	10 y or until patient is 21 y of age (whichever is longer)
Rheumatic fever without carditis	5 y or until patient is 21 y of age (whichever is longer)

~~Adapted from Gerber et al. (1)~~

*Lifelong prophylaxis may be recommended if the patient is at high risk of group A streptococcus exposure. Secondary rheumatic heart disease prophylaxis is required even after valve replacement.

†Clinical or echocardiographic evidence.

VHD indicates valvular heart disease.