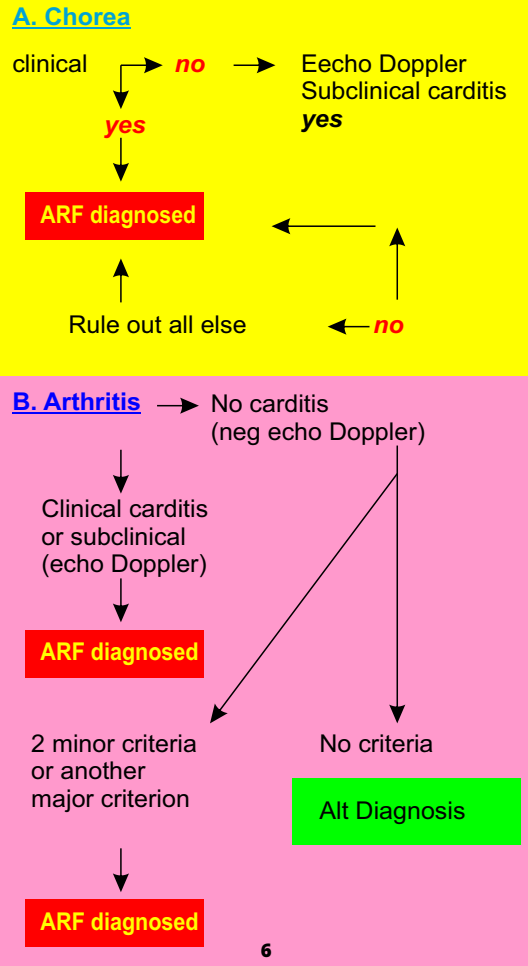
Rheumatologists working-book 3.0

Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography (May 2015)

Diagnosis strategy for acute rheumatic fever



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

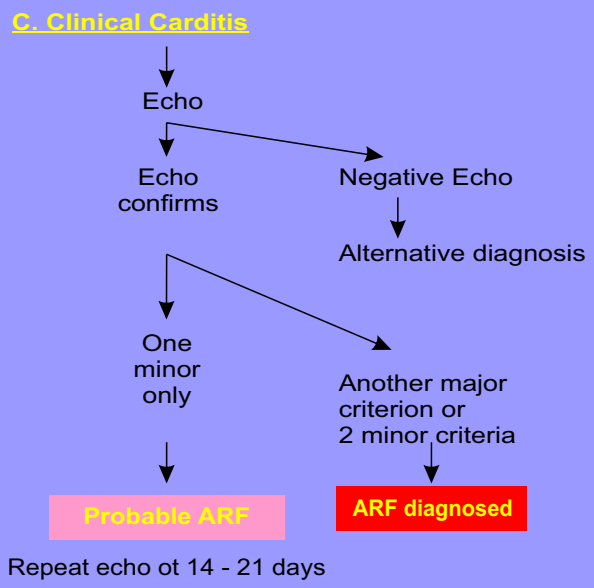
Aortic valve changes in either acute or chronic carditis

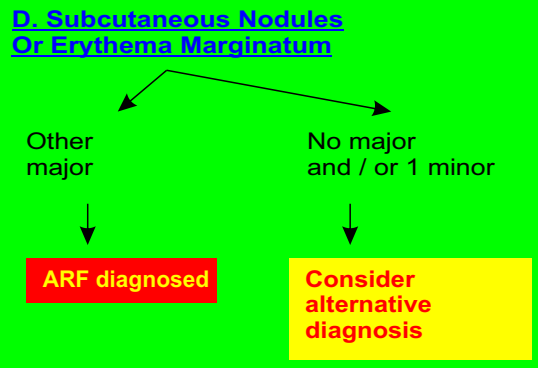
Irregular or focal leaflet thickening Coaptation defect
Restricted leaflet motion Leaflet prolapse

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.

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





Differential Diagnosis of Arthritis, Carditis, and Chorea

Arthritis	Carditis	Chorea
Septic arthritis (including gonococcal)	Physiological mitral regurgitation	Drug intixication
Connective tissue and another autoimmune diseases such as juverile idiopathic arthritis	Mitral valve prolapse	Wilson disease
Viral arthropathy	Myxomatous mitrak valve	Tic disorder
Reactive arthropathy	Fibroelastoma	Choreoathetoid cerebral palsy
Lyme diseases	Congetinal valve desease	Encephalitis
Sickle cell anemia	Congetinal aortic desease	Familial chorea (including Hurtington desease)
Infective endocarditis	Infective endocarditis	Intracranial tumor
Leukemia of limphoma	Cardiomyopathy	Lyme disease
Gout and pseudo gout	Myocarditis, viral or idiopathic	Hormonal
Poststreptococcal reactive arthritis	Kawasaki disease	Metabolic (eg. Lesch-Nyhan, hiperalaninemia, ataxia, telangiectasia)
Henoch-Schonlein pupura		Antiphospholipid anitidy syndrome
		Autoimmune: Systemic lupus erythematosus, systemic vasculitis
		Sarcoidisis
		Hyperthyroidism

Revised Jones Criteria

Arthritis	Chorea
A. For all patient populations with evidence of preceding GAS infection	
Diagnosis: initial ARF	2 Major manifestations or 1 major 2 minor manifestations
Diagnosis: recurrent ARF	2 Major or 1 major and 2 minor or 3 minor
B. Major criteria	Moderate- and high-risk populations
Low-risk populations*	Carditis
Carditis**	*Clinical and/or subclinical*
Clinical	Arthritis
Clinical and/or subclinical	*Monoarthritis or poliarthritis*
Arthritis	*Polyarthralgia***
Polyarthritis only	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)	Fever (38.5)
ESR more then 60 mm in the first hour and/or CRP more than 3.0 mg/dl	ESR more then 60 mm in the first hour and/or CRP more than 3.0 mg/dl
Prolonged PR interval, after accounting for age variability (unless carditis is major criterion)	Prolonged PR interval, after accounting for age variability (unless carditis is major criterion)

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

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

**Subclinical carditis indicates echocardiographic valvulitis as defined in table «Doppler Findings in Rheumatic Valvulitis»

***See section on polyarthralgia, with should only be considered as a major manifestation in moderate- to high-risk