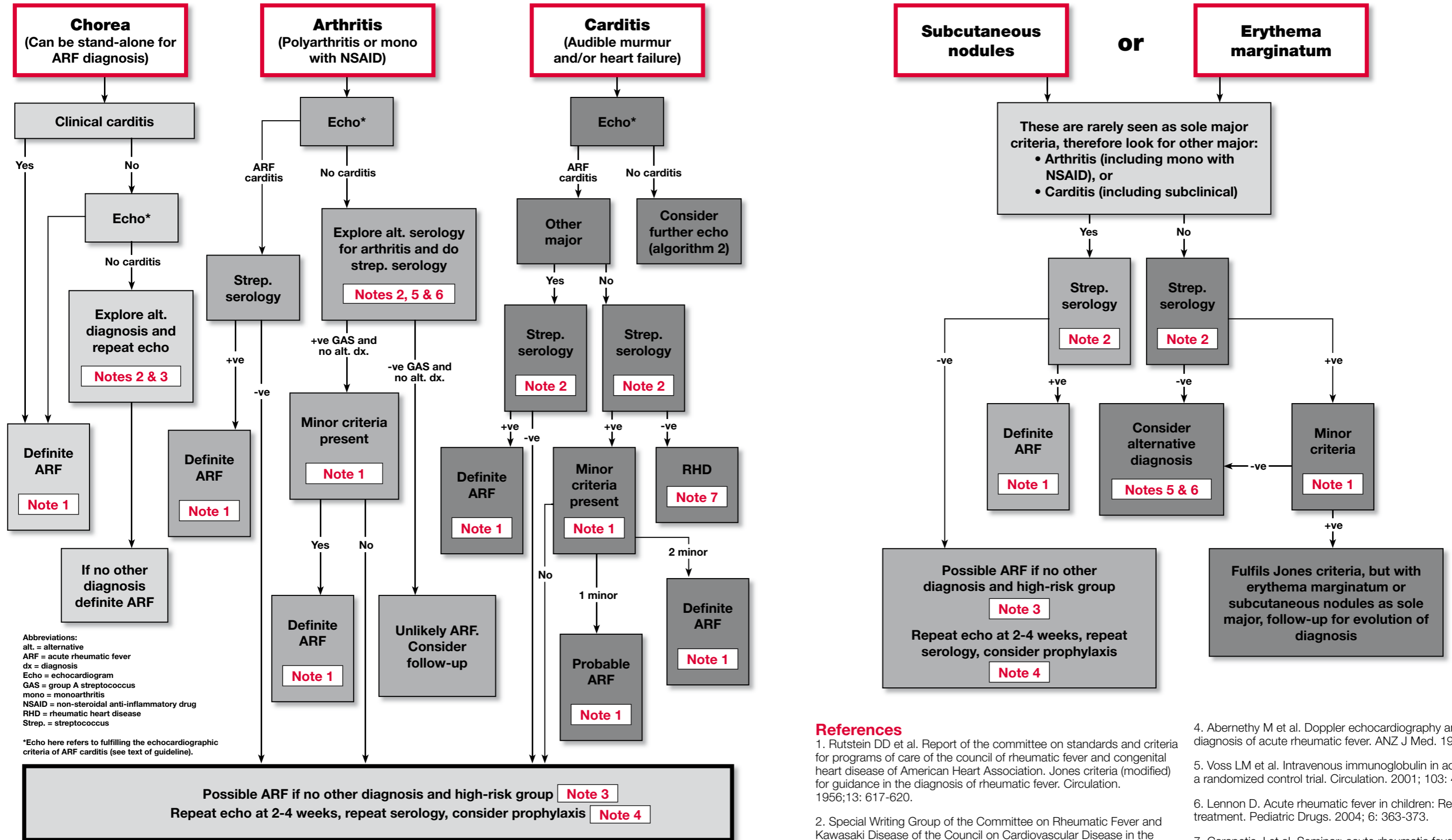


Note: This algorithm is to help in the decision-making process for the diagnosis of ARF with a clinical presentation of a major criterion. The investigations and observations are not intended to be in chronological order, and the clustering of symptoms and signs presented are commonly more complex. Note also that cases can fulfil the Jones criteria but not have ARF. Discussion with an experienced clinician, with reference to the full guideline on ARF diagnosis, is recommended.

[GUIDE]

for diagnosis of acute rheumatic fever (ARF)



Abbreviations:
 alt. = alternative
 ARF = acute rheumatic fever
 dx = diagnosis
 Echo = echocardiogram
 GAS = group A streptococcus
 mono = monoarthritis
 NSAID = non-steroidal anti-inflammatory drug
 RHD = rheumatic heart disease
 Strep. = streptococcus

*Echo here refers to fulfilling the echocardiographic criteria of ARF carditis (see text of guideline).

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Algorithm based on the evidence-based, best practice *New Zealand Guideline for Acute Rheumatic Fever: Diagnosis, Management and Secondary Prevention* (2006), produced by The National Heart Foundation of New Zealand and The Cardiac Society of Australia and New Zealand.



Note 1

New Zealand Guidelines For The Diagnosis of Acute Rheumatic Fever (ARF)

	DIAGNOSTIC REQUIREMENTS	CATEGORY
Initial episode of ARF	2 major or 1 major and 2 minor criteria plus evidence of a preceding GAS infection	Definite ARF
Initial episode of ARF	1 major and 2 minor with the inclusion of evidence of a preceding GAS infection as a minor criteria (Jones, 1956) ¹	Probable ARF
Initial episode of ARF	Strong clinical suspicion of ARF, but insufficient signs and symptoms to fulfil diagnosis of definite or probable ARF	Possible ARF
Recurrent attack of ARF in a patient with known past ARF or established RHD	2 major or 1 major and 2 minor or several* minor plus evidence of a preceding GAS infection (Jones, 1992) ²	
Major criteria modified** from Jones, 1992. (See guideline for further information on major criteria).	Carditis (including evidence of subclinical rheumatic valve disease on echo) ^{***} Polyarthritis# (or aseptic monoarthritis with history of NSAID use) Chorea (can be stand-alone for ARF diagnosis) Erythema marginatum Subcutaneous nodules	
Minor criteria (See guideline for further information on minor criteria).	Fever Raised ESR or CRP Polyarthralgia# Prolonged P-R interval on ECG	

All categories assume that other more likely diagnoses have been excluded.

CRP = C-reactive protein; ECG = electrocardiogram; ESR = erythrocyte sedimentation rate; GAS = group A streptococcus; RHD = rheumatic heart disease

- * WHO (2004) recommendations state that where there is established RHD, a recurrent attack can be diagnosed by the presence of two minor manifestations plus evidence of a preceding group A streptococcal infection³
- ** Acceptance of echocardiographic evidence of carditis as a major criterion is a New Zealand modification to the Jones (1992) update
- *** When carditis is present as a major manifestation (clinical and/or echocardiographic), prolonged P-R interval cannot be considered an additional minor manifestation in the same person
- # If polyarthritis is present as a major manifestation, polyarthralgia cannot be considered an additional minor manifestation in the same person.

Note 2

Upper Limits of Normal For Serum Streptococcal Antibody Titres Used in New Zealand

ANTIBODY TEST	TITRE (IU/ML)
ASO (anti-streptolysin O)	≥ 480
Anti-DNase B	≥ 680

Established from residual sera from children (under 15 years) hospitalised in Auckland in 1982. Lower levels may be acceptable in the very young or those over the age of 15 years. A two-tube (two-fold) rise or fall in antibody titres after 10-14 days would also be diagnostic. Note that evidence of a preceding GAS infection is not necessary for the diagnosis of chorea as ARF.

Note 3

Special consideration should be given to high-risk population groups such as Māori and Pacific people, and those residing in poor socio-economic circumstances who may not be readily available for review. In these cases, it may be important to err on the side of diagnosis and prophylaxis.

Note 4

Patients who do not fulfil these criteria, but in whom the clinician remains suspicious that the diagnosis may be ARF, should be maintained on oral penicillin and reviewed in 2-4 weeks with a repeat echocardiogram to detect the appearance of new lesions.^{4,5} If there is evidence of rheumatic valve disease clinically or on echocardiogram, the diagnosis is confirmed, and long-term secondary prophylaxis can be commenced. If there is no evidence of carditis and no alternative diagnosis has been found then ARF is possible. Those with epidemiological risk factors (Maori, Pacific, low socio-economic status) should be commenced on secondary prophylaxis with due consideration of an alternative diagnosis (such as rheumatological), and the need for ongoing review.

Note 5

Differential Diagnoses of Common Major Manifestations of ARF^{6,7}

	POLYARTHRITIS AND FEVER	CARDITIS	PRESENTATION	CHOREA
Differential diagnoses	<ul style="list-style-type: none"> • Other infections* • Connective tissue and other auto-immune disease** • Reactive arthropathy • Sickle cell anaemia • Infective endocarditis • Leukaemia or lymphoma • Gout and pseudogout • Henoch-Schonlein purpura • Post-streptococcal reactive arthritis*** • Other, e.g. HIV/AIDS, leukaemia 	<ul style="list-style-type: none"> • Innocent murmur • Mitral valve prolapse • Congenital heart disease • Infective endocarditis • Hypertrophic cardiomyopathy • Myocarditis — viral or idiopathic • Pericarditis — viral or idiopathic 		<ul style="list-style-type: none"> • Systemic lupus erythematosus • Drug ingestion (extrapyramidal syndrome)# • Wilson's disease (usually adult onset) • Tic disorder (see guideline) • Congenital, e.g. hyperbilirubinaemia • Choreoathetoid cerebral palsy • Encephalitis • Familial chorea (including Huntington's) • Intracranial tumour • Hormonal§ • Metabolic, e.g. Lesch-Nyhan, hyperalanaemia, ataxia, telangiectasia • Antiphospholipid antibody

- * Includes bacterial arthritis, influenza B, cytomegalovirus, Epstein-Barr Virus, mycoplasma, rubella (also post-vaccination), hepatitis B, parvovirus, Yersinia spp and other gastrointestinal pathogens
- ** Includes rheumatoid arthritis, juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis and sarcoidosis, among others
- *** In these cases the arthritis may affect joints that are not commonly affected in ARF, such as the small joints of the hand, and is less responsive to anti-inflammatory treatment. It is recommended that the diagnosis of post-streptococcal reactive arthritis should rarely, if ever, be made in high-risk populations (**Grade C** evidence - see guideline)
- # Drugs and toxins include anticonvulsants, antidepressants, lithium, scopolamine, calcium channel blockers, methylphenidate, theophylline and antihistamines
- § Includes oral contraceptives, pregnancy (chorea gravidarum), hyperthyroidism and hypoparathyroidism.

Note 6

Investigations in Suspected ARF

RECOMMENDED FOR ALL CASES
<ul style="list-style-type: none"> • White blood cell count • Erythrocyte sedimentation rate (repeat weekly once diagnosis confirmed) • C-reactive protein • Blood cultures if febrile • Electrocardiogram (repeat as necessary if conduction abnormality more than first degree) • Chest x-ray if clinical or echocardiographic evidence of carditis • Echocardiogram (repeat as necessary in 2-4 weeks if equivocal, or if serious carditis) - see Algorithm 2 • Throat swab (preferably before giving antibiotics) - culture for group A streptococcus • Anti-streptococcal serology: both anti-streptolysin O and anti-DNase B titres, if available (repeat 10-14 days later if first test not confirmatory)
TESTS FOR ALTERNATIVE DIAGNOSES, DEPENDING ON CLINICAL FEATURES
<ul style="list-style-type: none"> • Repeated blood cultures if possible endocarditis or septic arthritis • Joint aspirate (microscopy and culture) for possible septic arthritis* • Joint X-ray • Copper, caeruloplasmin, anti-nuclear antibody, drug screen, and consider CT/MRI head for choreiform movements** • Serology and auto-immune markers for auto-immune or reactive arthritis (including ANA - Anti Nuclear Antibody).

- * Typically, the synovial fluid in joints affected by ARF contains 10,000 to 100,000 white blood cells/mm³ (predominantly neutrophils). The protein concentration is approximately 4g/dL, glucose levels are normal, gram stain negative and a good mucin clot is present⁸
- ** The chorea of ARF can be readily diagnosed on the basis of history, physical examination and laboratory evaluation. Neuroimaging is seldom necessary and should be reserved for patients who have an atypical presentation, such as hemichorea.⁹

Note 7

It may be difficult to distinguish between indolent carditis and established rheumatic heart disease. The presence of any minor criterion makes indolent carditis more likely.