Acute Rheumatic Fever

Case Definition

Based on the updated 2012 Australian Guidelines

Definite initial episode of ARF	2 major OR 1 major AND 2 minor manifestations (Jones criteria) PLUS evidence of a preceding GAS infection (ie. elevated antiDNAase or ASOT* [repeat 10-14 days later if first test is not confirmatory] OR group A streptococcus grown from throat swab)	
Definite recurrent attack of ARF in a patient with known past ARF or RHD	2 major OR 1 major AND 1 minor OR 3 minor manifestations PLUS evidence of a preceding GAS infection	
Probable ARF (first episode or recurrence)	A clinical presentation that falls short by either one major or one minor manifestation, or in the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such cases should be further categorised according to the level of confidence with which the diagnosis is made: highly-suspected ARF uncertain ARF (see national guidelines for more detail)	
Major manifestations	High-risk groups	All other groups
	• Carditis (clinical or echocardiographic evidence of pericarditis, pericardial effusion, myocarditis or heart failure, including evidence of subclinical carditis on echocardiogram) • Polyarthritis OR aseptic mono-arthritis OR polyarthralgia • Chorea1 • Erythema marginatum • Subcutaneous nodules	• Carditis (as per high risk group but excluding evidence of subclinical carditis on echocardiogram) • Polyarthritis • Chorea1 • Erythema marginatum • Subcutaneous nodules
Minor manifestations	High-risk groups	All other groups
	• Mono-arthralgia • Fever >38˚C • ESR >30 mm/h or CRP >30 mg/L •Prolonged P-R interval** on ECG (if no carditis present)	• Fever >38˚C • Polyarthralgia or aseptic mono-arthritis • ESR >30 mm/h or CRP >30 mg/L • Prolonged P-R interval** on ECG (if no carditis present)

[1]Rheumatic (Sydenham's) chorea does not require other manifestations or evidence of preceding GAS infection, provided other causes of chorea are excluded. Sydenham's chorea is characterised by jerky, uncoordinated movements, particularly affecting the hands, feet, face and tongue. The movements should disappear during sleep, The movements may only affect one side of the body (hemichorea).

Helpful clinical tests for chorea are:

Patient unable to maintain tongue protrusion

Milkmaid's grip sign- rhythmic squeezing on grasping examiner's fingers

Pronator sign- arms/palms held above the head will turn outwardsSpooning sign- wrists flex and fingers extend when the hands are extended

ASSESSMENT AND MANAGEMENT

Discuss all cases of suspected ARF with the regional paediatrician / physician - likely to require admission

Investigations:

- Microbiology – swab throat and any infected skin sores; blood cultures if temperature ≥ 38˚c.
- Bloods – FBE, ESR, CRP, ASOT, anti-DNAase.
- ECG
- Joint effusions – if significant, aspirate for MC&S, cell count and differential and crystals.
- Order echo (URGENT for patients with clinical suspicion of carditis), otherwise within 3 months of diagnosis

Management:

1. Give Benzathine penicillin IMI stat

   Dose:

- Adults and children > 20kg: LA Bicillin 900mg stat.
- Children < 20kg: LA Bicillin 450mg stat.

  If possible hypersensitivity to penicillin refer/discuss with paediatrician or physician]

2. Analgesia for arthritis/arthralgia:

First line:

- Oral Paracetamol (+/- codeine- if > 12 years old)
- Adults and children >45kg: 500mg-1000mg QID
- Children <45kg:15mg/kg every 4-6 hours. (max 60mg/kg (up to 4G) per day) QID

Once diagnosis has been confirmed, if arthritis/arthralgia is severe and not responding to paracetamol use:

- Aspirin -
  Adults: 4-8g/day in 4-5 divided doses
  Children: starting dose 50-60mg/kg/day in 4-5 divided doses. Increase to 80-100mg/kg/day only if required
- Reduce to 60–70mg/kg/day when symptoms improve
- OR alternative Ibuprofen if > 6 months old – 10mg/kg (max 400mg) 6-8 hourly with food.

Acute Rheumatic Fever

- Avoid aspirin use prior to confirmation of diagnosis as it may mask the presentation of polyarthritis
- *Discuss treatment for chorea with the regional Paediatrician / Physician.*

NOTIFICATION

ARF/RHD is a notifiable disease. Under WA Health (*Rheumatic Heart Disease Register of Western Australia*) Regulations 2015 you must report this diagnosis to the WA RHD Register - see forms in MMEx or online http://www.public.health.wa.gov.au/cproot/2848/2/acute_rheumatic_fever_notification_form_june2014.pdf

Patient consent is no longer required for notification and failure to notify can result in fines.

Completed notifications can be sent to the register by either fax or email

- fax to 9193 5260 or
- email RHDRegister@health.wa.gov.au

You must also provide a copy of each diagnostic test (including an echocardiogram) within 15 days for a new diagnosis and 30 days for a recurrence.

Ongoing reports from cardiologists/physicians/ paediatricians and surveillance echoes also need to be sent through to the register as they occur.

Patients who have had more than one episode require a new notification to be made for EACH new episode.

Pregnancy should be notified to the register in the third trimester.

For further information please call the WA RHD Register on 1300 622 745 or email RHDRegister@health.wa.gov.au or view http://ww2.health.wa.gov.au/Articles/U_Z/WA-rheumatic-heart-disease-register

Follow Up

ALL PATIENTS WITH SUSPECTED OR CONFIRMED ARF REQUIRE FOLLOW UP WITH THEIR LOCAL HEALTH CARE PROVIDER

For those managed as an outpatient, this review should take place one week after initial presentation.

For those admitted to hospital, this review should take place one week after discharge.

At the follow up appointment:

- Confirm and document initial presentation, detailing major and minor diagnostic features clearly in the medical record.
- Assign a care plan –[see RHD PROTOCOL for information on priority allocation]
- Ensure the WA RHD Register has been notified and results of any investigations sent through [see NOTIFICATION]
- Repeat ECG
- Repeat CRP/ESR
- Organise repeat ASOT and antiDNAase titres 10-14 days after initial titres (if initial titres were inconclusive)
- Note any response to aspirin
- Clinical assessment to look for any features of developing carditis
- Consider joint aspiration
- Organise echo (ASAP after diagnosis, but no longer than 3 months after diagnosis)
    - If initial echo is normal, repeat in 1-3 months
    - If this second echo is normal - repeat at 1 year
    - If this second echo is abnormal - discuss with paediatric cardiology/adult physician
- Organise review by physician/cardiologist/paediatrician within 3 months of presentation (preferably AFTER echo has been done)
- Education about rationale for long term prophylaxis and the risks of no prophylaxis

SECONDARY PROPHYLAXIS (to prevent further ARF):

- All patients who have had ARF need intramuscular LA Bicillin every 3-4 weeks. Monthly injections are less effective so every attempt must be made to adhere to injections every 3-4 weeks
- If patient attends from days 21-28 give dose LAB

Dose:

Adults and children > 20kg: 900mg

For children < 20kg: 450mg

If possible hypersensitivity to penicillin refer/discuss with paediatrician or physician

Benzathine penicillin is superior to any oral prophylaxis and should be used except when there is severe documented allergy to penicillin when oral erythromycin 250mg twice a day is indicated (all ages).

NB: Patients who have confirmed breakthrough ARF, despite full adherence to 4-weekly Benzathine Penicillin should be considered for 3 weekly Benzathine Penicillin. Discuss with the physician or paediatrician.

6.DURATION OF SECONDARY PROPHYLAXIS

All persons with probable ARF	**Highly-suspected ARF:** Minimum of 10 years after most recent suspected episode of ARF or until alternative diagnosis confirmed **Uncertain ARF:** 12 months after diagnosis, and then reassess (including echocardiography and specialist review) at that time. If evidence of RHD at that time, manage as for

Acute Rheumatic Fever

	highly-suspected ARF. If no evidence of RHD, consider ceasing prophylaxis
RHD Priority 3	Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer)
RHD Priority 2	Continue until 35 years of age or 10 years after last episode of ARF (whichever is longer)
RHD Priority 1	Continue until 40 years old or 10 years after last episode of ARF (whichever is longer).

NB -All patients who are due to cease secondary prophylaxis (including those who are well) require an echo and specialist review prior to ceasing.
NB: Patients >25 years of age who are diagnosed with RHD, and without any documented history of prior ARF, should receive prophylaxis until the age of 35. At this time, they should be reassessed to determine whether prophylaxis should be continued. This decision is based upon the severity of the RHD at age 35.
NB - ALL patients who have ever had ARF should have their RHD classified as priority 1-4 as per the Rheumatic Heart Disease protocol. This includes patients who have had ARF but have no RHD (priority 3), and those patients with no evidence of RHD for whom secondary prophylaxis has been ceased (priority 4)

For schedule of ongoing follow up after acute rheumatic fever (including for those patients with no evidence of rheumatic heart disease) see "Rheumatic Heart Disease" protocol

REFER DISCUSS

To physician / paediatrician :

All people with confirmed or suspected ARF.

FOR SUPPORT OR FURTHER INFORMATION CONTACT:

WA RHD Register:

- pH: 1300 622 745
- email: RHDRegister@health.wa.gov.au

RESOURCES

RHD Australia Website – www.rhdaustralia.org.au

Download the app – "ARF / RHD Guideline" from Google Play or the App Store or follow the link from the RHD Australia Website

- Includes diagnosis calculator

Appendix

*Age adjusted reference range for ASOT and anti-DNAase

Age (in years)	Upper limit of normal	
	ASOT	Anti-DNAase
1-4	170	366
5-14	276	499
15-24	238	473
25-34	177	390
≥35	127	265

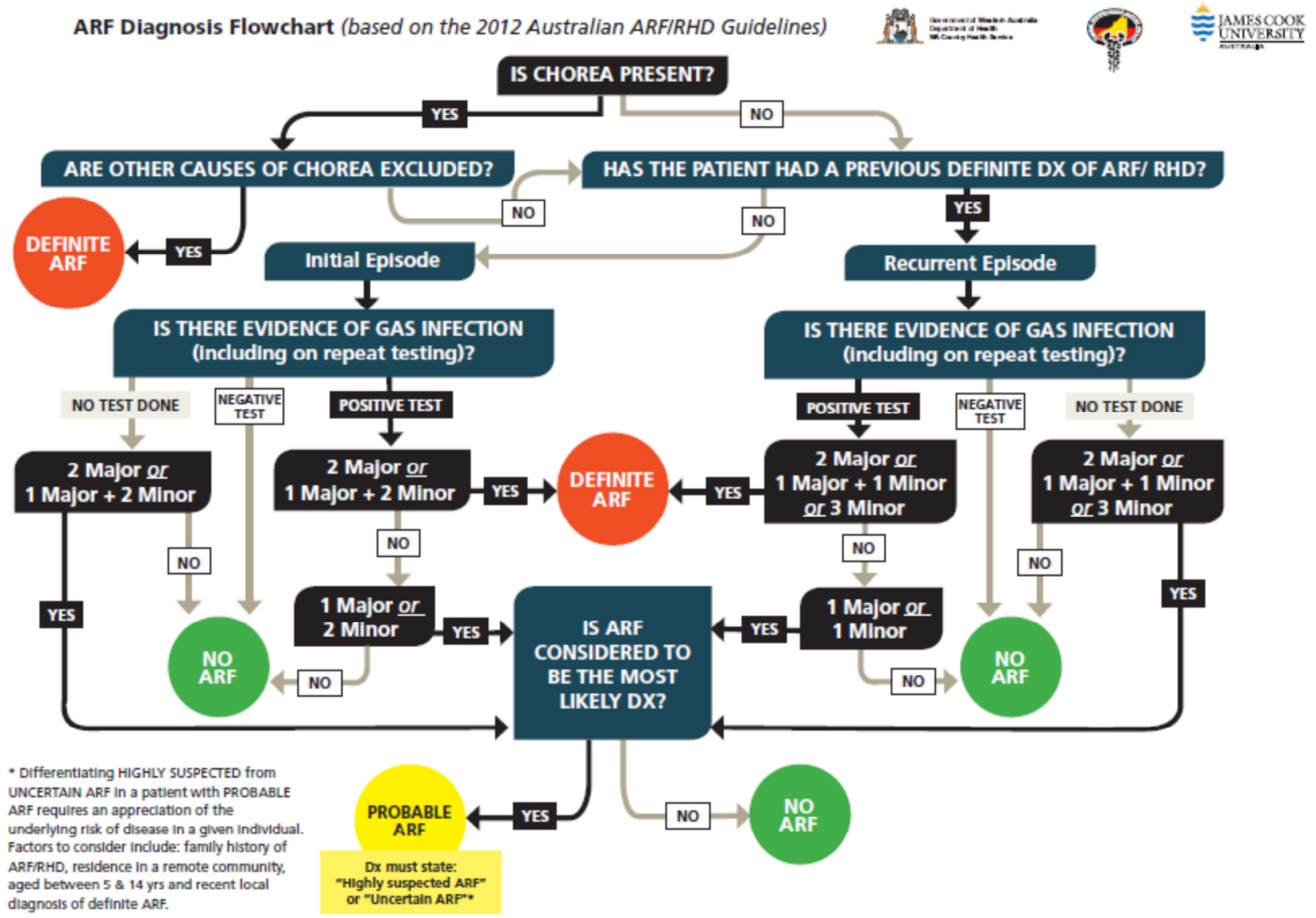
**Upper limit of normal of PR interval

Age (in years)	ULN for PR interval
3-12	0.16 secs
12-16	0.18 secs
17+	0.20 secs

*Table 3.6, page 38. RHDAustralia (ARF/RHD Writing group), Nation Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (second edition).

**Table 3.4, page37. RHDAustralia (ARF/RHD Writing group), Nation Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (second edition).

Acute Rheumatic Fever



This flow chart was developed with the assistance of Marc Rémond (James Cook University), Rhona Dawson (KAMSC), Michael Dawson (KAMSC) & Graeme Maguire (Baker IDI). For suggestions or feedback, please email marc.remond@my.jcu.edu.au
Flowchart & tables based on: RHDAustralia (ARF/RHD writing group), National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). 2012