ANMC Acute Rheumatic Fever Evaluation and Management

Background Information & Definitions

- Acute Rheumatic Fever (ARF): Immune-mediated inflammatory complication of group A streptococcal (GAS) infection. Usually follows strep pharyngitis, may follow other GAS infections
- Rheumatic Heart Disease (RHD): Long-term cardiac complications of ARF – most commonly mitral and aortic valves
- Increased risk of ARF/RHD is likely due to socioeconomic factors including high number of household members, and/or hauled or collected water. Current data suggest that the AI/AN population is at increased risk. → ***Consider ARF and/or RHD for any child ≥3 yo with arthritis/arthralgia, systemic inflammation, new murmur, or symptoms of HF.***

Initial Evaluation

- Throat swab for GAS PCR testing "Strep A "Molecular" (regardless of pharyngitis symptoms) – if not available collect Rapid Strep *and* culture
- Serologies: ASO and anti-DNase B
- Inflammatory Markers: ESR and CRP
- Electrocardiogram

Diagnosis – Jones Criteria

Initial diagnosis: 2 major *or* 1 major plus 2 minor || Recurrent diagnosis: 2 major *or* 1 major plus 2 minor criteria *or* 3 minor

Current Recommendation: Use Moderate/High-Risk Jones Criteria for individuals from northern and western Alaska and individuals identifying as AI/AN & NH/PI. Consider using Moderate/High-Risk Jones Criteria for others with risk factors.

Moderate/High Risk Major Criteria	Moderate/High Risk Minor Criteria
• Carditis (clinical or subclinical) ♥ • Arthritis (polyarthritis or monoarthritis; polyarthralgia) * • Chorea ¥ • Erythema marginatum • Subcutaneous nodules	• Monoarthralgia (unless arthritis is a major criteria) * • Fever ≥38°C/100.4°F • ESR ≥30 mm/h and/or CRP ≥3 mg/dL • Prolonged PR interval (in absence of carditis)

♥Urgent pediatric cardiology referral: if Jones Criteria are met *OR* if high clinical suspicion for ARF (even if Jones Criteria are not fully met and no cardiac symptoms/murmur)

*Consider pediatric cardiology referral for arthritis/arthralgia with systemic inflammation as ARF may present similarly to autoimmune conditions.
-For painful arthritis, if >2 yo, treat with Naproxen 10 mg/kg/dose PO BID (MAX 500mg/dose) OR if <2 yo or naproxen is unavailable, treat with ibuprofen 13mg/kg/dose PO TID (MAX 800mg/dose). Scheduled dosing x2 months then PRN.

¥Urgent neurology referral for Sydenham's chorea, consider starting levetiracetam 10mg/kg/dose PO BID until seen by neurology

Consider consult to infectious diseases if additional input would aid in clarifying the diagnosis.

Household Contacts: At the time of diagnosis, all household members should be screened for GAS carriage with a throat swab. Positive family members should be treated.

Treatment

Initial Treatment

GAS treatment should be provided at the time of initial diagnosis regardless of symptoms. See **ANMC GAS Pharyngitis Treatment Guidelines**

Long Term GAS Prophylaxis

Preferred Treatment	**IM penicillin G benzathine** • ≤27 kg: **600,000 units** IM q28 days • >27 kg: **1.2 million units** IM q28 days *Decrease interval to q21 days if recurrent ARF*
Unable to Tolerate IM¶ *or* IM unavailable (shortage, delay, etc)	**Penicillin VK 250mg** PO BID
Penicillin Allergic∞	**Azithromycin 6mg/kg/dose** PO Daily (MAX 250mg)

Duration of Prophylaxis

Category	Duration
ARF without carditis	Until 21yo or 5 years after last ARF (whichever is longer)
ARF with carditis but *no* residual heart disease	Until 21yo or 10 years after last ARF (whichever is longer)
ARF with carditis *and* residual heart disease	Until 40yo or 10 years after last ARF (whichever is longer); lifetime prophylaxis may be needed

Pain Reduction Techniques for IM Injections

- Bring IM penicillin G benzathine to room temperature before administration
- Locally applied vibratory device (ex. Buzzy Bee)
- Topical numbing with EMLA cream
- Comfort positioning for younger children (examples available www.megfoundationforpain.org)
- Distraction
- Routine (ex. same day of the week, same time of day)

Considerations

¶ Including severe symptomatic valvular disease, NYHA class III or IV heart failure symptoms, and/or ventricular dysfunction (ie, EF <50%). Consultation by cardiology with documentation on a case-by-case basis is recommended prior to administering medications IM.

∞ Refer to allergist for evaluation and consideration of penicillin allergy testing

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1. Alaska Epi bulletin: https://epi.alaska.gov/bulletins/docs/rr2026_01.pdf; 2. Centers for Disease Control and Prevention. Clinical Guidance for Acute Rheumatic Fever. Accessed September 09, 2025. https://www.cdc.gov/group-a-strep/hcp/clinicalguidance/acute-rheumatic-fever.html; 3. Gewitz MH, Baltimore RS, Tani LY, 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. May 19 2015;131(20):1806–18. doi:10.1161/cir.0000000000000205; 4. Alaska Section of Epidemiology. Streptococcal Control Program - Winning the Fight Against Rheumatic Fever. Epidemiology Bulletin. 1985;3; 5. Christiansen J, Singleton R. Acute Rheumatic Fever — Two Case Reports and New Jones Criteria. State of Alaska Epidemiology Bulletin. 2016;(18); 6. Rwebembera J, Marangou J, Mwita JC, et al. 2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease. Nat Rev Cardiol. Apr 2024;21(4):250–263. doi:10.1038/s41569-023-00940-9; 7. Baker MG, Gurney J, Moreland NJ, et al. Risk factors for acute rheumatic fever: A case control study. Lancet Reg Health West Pac. Sep 2022;26:100508. doi:10.1016/j.lanwpc.2022.100508