



Department of Health and Social Services
Valerie J. Davidson, Commissioner

Division of Public Health
Jay Butler, MD, Chief Medical Officer
and Director

Editors:
Joe McLaughlin, MD, MPH
Louisa Castrodale, DVM, MPH

3601 C Street, Suite 540
Anchorage, Alaska 99503 <http://dhss.alaska.gov/dph/Epi>

Local (907) 269-8000
24 Hour Emergency (800) 478-0084

Bulletin No. 18 August 9, 2016

Acute Rheumatic Fever — Two Case Reports and New Jones Criteria

Introduction

Acute rheumatic fever (ARF) is an immune-mediated sequelae of untreated group A streptococcal (GAS) infection, e.g., pharyngitis, that can result in serious long-term health consequences. The incidence of ARF has decreased in the U.S., but remains high in low-income countries with crowding and inadequate hygiene,¹ and among certain ethnic groups (e.g., Pacific Islanders and Samoans).² The purpose of this *Bulletin* is to raise awareness of ARF, to underscore the variable presentation of ARF by relaying two recent case reports, and to update clinicians on the new Jones criteria for establishing an ARF diagnosis.³

Case Reports

Patient A On August 1, 2015, a 16-year-old female Pacific Islander was hospitalized after experiencing collapse without loss of consciousness; a new erythematous, raised rash on shin and thighs; and swelling with paresthesias of her hands and feet. Directly preceding this, she had a 2-week history of headache, fever, sore throat, back pain, and emesis. Extensive diagnostic workup was only remarkable for first degree AV block on electrocardiogram. Her discharge diagnosis was “viral syndrome”. Nine days later she was readmitted with acute onset of right knee pain, generalized malaise, persistent rash, and paresthesias. A joint aspirate was culture-negative. Her C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were elevated. Target lesions on her hands were diagnosed as possible erythema marginatum. A systolic ejection murmur was heard and echocardiography revealed polyvalvular disease. Anti-streptolysin O (ASO) and anti-DNase B titers were elevated. The diagnosis of ARF was made and she was treated with high-dose aspirin and intramuscular (IM) LA benzathine penicillin G (Bicillin).

Patient B On March 4, 2016, a 6-year-old Alaska Native female presented to her village clinic with cough, fever, and wheezing, and was prescribed acetaminophen and nebulizer treatments. Her symptoms progressed to include labored breathing, tachypnea, anorexia, and fatigue. She was seen on March 23 and continued on supportive care. A rapid streptococcal screen was reportedly negative at that visit. On March 27, with new onset of facial swelling, she was sent to a hub hospital and found to have a prominent cardiac murmur and cardiomegaly on chest x-ray. An echocardiogram showed diffuse chamber enlargement, severe mitral insufficiency, moderate aortic regurgitation, and pulmonary hypertension. An ASO titer was elevated. She was transferred to Seattle Children’s Hospital and treated for congestive heart failure (CHF); she then underwent mitral valve replacement on April 15 for refractory CHF. After a complicated postoperative course, she was discharged on June 17 to the Alaska Native Medical Center to optimize her anticoagulation regimen before discharge home with Bicillin therapy every 3 weeks.

New Jones Criteria

The 2015 Jones criteria reflect the new role of echocardiography and differences in ARF presentation between low risk and moderate-high risk populations (Table).³ Echocardiography can detect valvar dysfunction in patients without physical signs, leading to inclusion of subclinical carditis as a major diagnostic criteria. In moderate-high risk populations, aseptic monoarthritis and polyarthralgia can be included as criteria.³ Chorea occurs in 10%–30% of cases. Erythema marginatum (painless, non-pruritic, red, blanching, annular rash) and subcutaneous nodules (0.5–2 cm painless lesions over extensor surfaces) occur in <10% of cases.³

Table. Revised Jones Criteria for Diagnosing ARF*

	Population Risk	Low [†]	Mod-High
Major Criteria			
Carditis (clinical/subclinical)		x	x
Arthritis (polyarthritits)		x	x
Arthritis (mono- or polyarthritits/arthralgia)			x
Chorea		x	x
Erythema marginatum		x	x
Subcutaneous nodules		x	x
Minor Criteria			
Polyarthralgia		x	x
Fever ≥ 38.5 °C		x	x
ESR ≥ 60 mm/hr, CRP ≥ 3.0		x	x
Monoarthralgia			x
Fever ≥ 38 °C			x
ESR ≥ 30 mm/hr, CRP ≥ 3.0			x
Prolonged PR interval for age		x	x

*2 major, or 1 major and 2 minor Jones criteria are diagnostic for ARF in persons with evidence of preceding GAS infection (e.g., ASO/anti-DNase B, positive strep test/culture, or scarlet fever). Recurrent ARF in a person with a reliable past history of ARF or established rheumatic heart disease (RHD) can be diagnosed by 2 major + 1 minor criteria or 3 minor criteria.

[†]Low risk populations are those with ARF incidence ≤ 2 per 100,000 school-aged children or all-age RHD prevalence of ≤ 1 per 1000 population per year.

Discussion

While the number of reported cases of ARF in Alaska has been low in recent years (6 reported cases during 2006–2015; <1 per 100,000 school-aged children), many cases may go undiagnosed/unreported. Moreover, some Alaska populations, including rural Alaska Native people in overcrowded conditions with lack of running water and Alaskans of Pacific Islander/Polynesian ethnicity, may be at higher risk for ARF. Lastly, as the case reports illustrate, ARF should be considered in patients with a clinically compatible syndrome even in the absence of a clear history of preceding GAS infection.

Recommendations

1. Obtain ASO and anti-DNase B titers in suspected ARF cases to facilitate detection of GAS infection.⁴
2. Obtain an echocardiogram on all patients with ARF, even if they lack clinical evidence of carditis (murmur).³
3. ARF patients should be treated with anti-inflammatory medications (e.g., aspirin, NSAIDs, or prednisone) and supportive cardiac care, and be placed on long-term antibiotic prophylaxis. The antibiotic of choice is IM Bicillin (other regimens are also acceptable under special circumstances).⁴ The duration of secondary prophylaxis depends on the degree of cardiac involvement.⁴
4. All household contacts of ARF patients should have throat cultures for GAS (and be treated if positive).⁴
5. Report all suspect or confirmed cases of ARF to the Section of Epidemiology (7 AAC 27.005). Call 907-269-8000 or fax information to 907-561-4239.

References

1. Carapetis JR, et al. Acute rheumatic fever and rheumatic heart disease. *Nat Rev Dis Primers* 2016;2:1-24.
2. Coffey S, et al. The modern epidemiology of heart valve disease. *Heart* 2016;102(1):75-85.
3. Gewitz MH, et al. Revision of the Jones Criteria for the diagnosis of acute rheumatic fever in the era of Doppler Echocardiography. *Circulation* 2015;131(20):1806-18.
4. Gerber MA, et al. Prevention of rheumatic fever and diagnosis and treatment of acute Streptococcal pharyngitis. *Circulation* 2009;119(11):1541-51.