



Department of Health and Social Services
William J Streur, Commissioner

Division of Public Health
Ward B Hurlburt, MD, MPH, CMO/Director

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

3601 C Street, Suite 540
Anchorage, AK 99503

<http://www.epi.Alaska.gov>

Local (907) 269-8000
24 Hour Emergency 1-800-478-0084

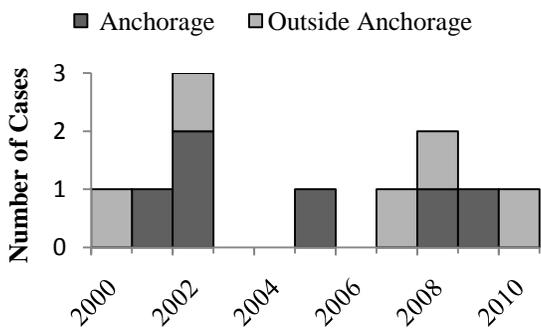
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Two Cases of Rheumatic Fever in Alaska, 2009–2010

Introduction

Since the 1950s, the incidence of rheumatic fever (RF), a sequela of group A streptococcal pharyngitis, has decreased sharply.¹ Nevertheless, RF remains a leading cause of acquired heart disease in the developing world.¹ Several environmental and social determinants of RF, including limited health care resources, are prevalent in Alaska. During 2000–2010, RF cases were reported in Alaska during all but 3 years (Figure). In November 2010, the Section of Epidemiology (SOE) was notified of two cases of RF. The purpose of this *Bulletin* is to describe these cases, to provide current guidelines for diagnosis and treatment, and to remind clinicians to *report all suspected and confirmed RF cases* to SOE to facilitate prompt outbreak detection and response.

Figure. Reported Rheumatic Fever Cases in Alaska, 2000–2010



Case Reports

Patient A. On June 27, 2009, a previously healthy adolescent female Pacific Islander was seen in the Providence Alaska Medical Center (PAMC) emergency department (ED) complaining of “twitching” in her left arm and leg. Patient A was discharged from the ED and referred for follow-up with a neurologist. On July 10, Patient A was admitted to PAMC following a second ED visit with a diagnosis of “possible chorea.” On admission, the attending neurologist noted numerous “choreoathetoid movements” that were especially prominent distally. Despite the absence of a murmur by auscultation, an echocardiogram revealed mild tricuspid and mitral valve insufficiency. Patient A had an elevated ASO titer and anti-DNase B titers. Patient A was diagnosed with Sydenham’s chorea and rheumatic heart disease and started on intravenous penicillin and high-dose aspirin; she was later transitioned to oral penicillin. Following the course of oral penicillin, she was prescribed monthly intramuscular bicillin “at least until age 25.” Patient A is currently stable from a cardiac standpoint, but has considerable cardiac dysfunction. She is on carbamazepine for her movement disorder. She is being followed by neurology and cardiology.

Patient B. On October 12, 2010, a previously healthy adolescent Alaska Native female was referred to the Alaska Native Medical Center (ANMC) for neurologic and cardiac evaluation after choreiform movements were observed by her primary care provider. Patient B reported experiencing abnormal movements in her extremities for approximately two months, and had a history of recurrent streptococcal infections. On exam, she had choreiform movements. She had an elevated erythrocyte sedimentation rate (ESR). An echocardiogram on October 18 showed an abnormal mitral valve with thickened distal leaflets and mild-to-moderate mitral insufficiency. The cardiologist concluded that Patient B’s history was consistent with RF and prescribed intramuscular bicillin and aspirin.

Discussion

Rheumatic fever is an uncommon and serious sequela of group A *Streptococcus pyogenes* (GAS) infection that can result in considerable morbidity and mortality. Appropriate treatment of streptococcal pharyngitis reduces the risk of RF.²

A diagnosis of a primary episode of RF requires that a patient have either two major criteria, or one major and two minor criteria, plus evidence of previous GAS infection (Table).¹ Evidence of previous infection can include a positive culture or rapid test, recent scarlet fever, a prolonged P-R interval on electrocardiogram, or an elevated/increasing streptococcal antibody test.¹ A follow-up antibody test performed 3–4 weeks after RF symptom onset is recommended to confirm the previous streptococcal infection.¹

Table. Jones Criteria for the Diagnosis of Rheumatic Fever

Major Criteria	Minor Criteria
Carditis	Arthralgia
Polyarthritits	Fever
Subcutaneous nodules	Laboratory results suggesting acute inflammation (e.g., elevated WBC count or ESR)
Chorea	
Erythema marginatum	

WBC=white blood cell; ESR=erythrocyte sedimentation rate

Recommendations

1. If a patient is suspected of having RF, evaluate evidence of previous GAS infection (see Discussion above). Laboratory tests for markers of inflammation are also useful (Table).
2. Consider ordering an echocardiogram to evaluate for subclinical vasculitis.
3. A patient with irregular movement should be evaluated by someone aware of the clinical appearance of Sydenham’s chorea to avoid confusion with other conditions where positive GAS serology is an unrelated finding.
4. Because cardiac sequelae can develop after a movement disorder, re-evaluation of cardiac health 3–6 months after the development of irregular movements may be prudent.
5. The recommended treatment for RF includes anti-inflammatory therapy (e.g., an analgesic, aspirin, or prednisone, depending on clinical severity) and intramuscular benzathine penicillin G every 4 weeks to prevent additional attacks. The penicillin dosing should be 1.2 million units for persons weighing >67 kg, or 600,000 units for persons weighing ≤67 kg. The duration of therapy depends on the degree of cardiac involvement during the most recent attack.³ A macrolide or an azalide can be substituted for patients allergic to penicillin, depending on bacterial antibiotic sensitivity patterns.³
6. Rheumatic fever is reportable to SOE by health care providers (7 AAC 27.005). To report, please call 907-269-8000, 561-4234, or 1-800-478-1700 if outside Anchorage, or fax information to 907-561-4239.

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