



Acute rheumatic fever

Principal investigator

Christina G. Templeton, MD, FRCPC

Janeway Children's Health & Rehabilitation Centre, 300 Prince Philip Dr.,
St. John's NL A1B 3V6; tel.: 709-777-4462; fax: 709-777-4747; e-mail:
christina.templeton@hccsj.nl.ca

Co-investigators

Austin Rick Cooper, MD, FRCPC, Chair, Department of Paediatrics, Janeway
Children's Health & Rehabilitation Centre

Derek G. Human, BM, MRCP (UK), FRCPC, Head, Division of Cardiology,
Department of Paediatrics, University of British Columbia

Proton Rahman, MD, FRCPC, Associate Professor of Medicine, Memorial
University of Newfoundland

Background

Acute rheumatic fever is a post-infectious collagen vascular disease affecting the heart, joints and central nervous system. It follows untreated group A streptococcal (GAS) pharyngitis after a latent period of approximately three weeks, but it does not occur after other GAS infections, such as skin infection (impetigo). Worldwide, acute rheumatic fever remains the commonest cause of acquired heart disease in children, yet the incidence varies widely from region to region with the vast majority of cases now occurring in developing countries.

While the incidence of acute rheumatic fever in developed countries has decreased dramatically since its last peak in the 1970s, it has not disappeared, and in fact still remains an important public health issue, as outbreaks have occurred in school-aged children as recently as in the 1990s. Nor is the reason for its decrease fully understood, as the decline in incidence in the early 20th century had already begun prior to the introduction of effective antimicrobial agents. It may be that the common use of penicillin to treat symptomatic sore throat contributed somewhat to the decline.¹ Socioeconomic factors, such as overcrowding and low income, are known to be significant risk factors.² The majority of cases of rheumatic fever follow cases of pharyngitis due to specific M serotypes of GAS, most commonly 1, 3, 5, 6, 18, 19 and 24, and spontaneous fluctuation of the prevalence of these serotypes is known to occur.^{3,4}



Rheumatic fever is not a reportable condition in Canada, and so in the current era of evidence-based, judicious use of antibiotics, ongoing surveillance of this now rare, but serious, condition is crucial. Rheumatic heart disease is a lifelong complication of the condition, which can lead to ongoing medical and surgical needs and can interfere with employment, causing a significant socioeconomic impact. However, the risk of developing rheumatic fever must be balanced against the risk of encouraging microbial antibiotic resistance, which is a growing problem in all developed nations and carries its own impact.

No current Canadian incidence data is available, but recent American reports would suggest an expected number of cases of about 240 per year in Canada.⁵ This is a sufficiently rare condition that only a national reporting system could gather statistically significant numbers.

Methods

Paediatricians and paediatric subspecialists, including rheumatology, neurology and cardiology, will be asked to report cases of new onset initial attack of rheumatic fever through the CPSP monthly survey. Reporting physicians will then be asked to complete a straightforward, concise follow-up questionnaire with specific non-nominal details of the diagnosis, treatment and outcome of each case.

Case definition

Report any child up to and including 18 years of age that meets the most recent modifications of the Jones Criteria for diagnosis of an initial attack of rheumatic fever as follows:^{6,7}

Major manifestations	Minor manifestations
Carditis Polyarthrititis Chorea Erythema marginatum Subcutaneous nodules	Clinical Arthralgia Fever Laboratory findings Increased acute phase reactants: Increased erythrocyte sedimentation rate Increased C-reactive protein Prolonged P-R interval
<p>All cases, except Sydenham's chorea, will require documentation of antecedent group A streptococcal infection, either by positive throat culture, rapid antigen test, or an elevated or rising antibody titre. Antistreptolysin O titre measurement is the preferred test to distinguish recent streptococcal infection from chronic pharyngeal carriage.</p> <p>If there is evidence of recent streptococcal infection, the presence of two major manifestations or one major and two minor manifestations will be considered diagnostic.</p>	



Acute rheumatic fever (continued)

The definition of carditis will require clinical evidence of cardiac involvement in the form of a pathological murmur, pericarditis, or congestive heart failure. Current literature is divided as to whether silent echocardiographic findings should be included;⁸ the questionnaire will include this information but the case definition will remain faithful to current international consensus requiring clinical manifestations.

Objectives

1. To determine the incidence of rheumatic fever among Canadian children.
2. To determine the relationship between modern rheumatic fever and demographic features, such as overcrowding and low household income.
3. To describe current Canadian treatment practices.
4. To determine the morbidity and mortality of first episode rheumatic fever in Canada.

Duration of study

April 2004 to March 2007

Expected number of cases

Based on recent American reports and the current Canadian paediatric population, the expected number of cases would be up to 240 per year.

Ethical approval

Human Investigation Committee, Memorial University of Newfoundland

Data for analysis and publication

The investigators will analyze data and report any important findings promptly to the CPSP. Quarterly progress reports and annual summaries will be submitted for distribution. Data will be published in a peer-reviewed journal on completion of the study.

References

1. Massell BF, Chute CG, Walker AM, et al. Penicillin and the marked decrease in morbidity and mortality from rheumatic fever in the United States. *N Engl J Med* 1988; 318: 280.
2. World Health Organization. Rheumatic fever and rheumatic heart disease. WHO Technical Report Series 764. Geneva, World Health Organization, 1998.
3. Schwartz B, Facklam RR, Breiman RF. Changing epidemiology of group A streptococcal infection in the USA. *Lancet* 1990; 336: 1167.



4. Colman G, Tanna A, Efstatiou A, et al: The serotypes of *Streptococcus pyogenes* present in Britain during 1980-1990 and their association with disease. *J Med Microbiol* 1993; 39: 165.
5. Veasy GL, Tani LY, Hill HR. Persistence of acute rheumatic fever in the intermountain area of the United States. *J Pediatr* 1994; 124: 9-16.
6. Dajani AS, Ayoub E, Bierman FZ, et al. Guidelines for the diagnosis of rheumatic fever: Jones criteria, updated 1992. *Circulation* 1993;87:302-7.
7. Ferrieri P, for the Jones criteria working group. Proceedings of the Jones criteria workshop. *Circulation* 2002; 106: 2521-3.
8. Ozkutlu S. Can subclinical valvitis detected by echocardiography be accepted as evidence of carditis in the diagnosis of acute rheumatic fever? *Cardiol Young* 2001; 11(3): 255-60.