Acute rheumatic fever

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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. Socioeconomic factors, such as
overcrowding and low income, are known to be significant risk factors.
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.
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. 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:

<table>
<thead>
<tr>
<th>Major manifestations</th>
<th>Minor manifestations</th>
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<tbody>
<tr>
<td>Carditis</td>
<td>Clinical</td>
</tr>
<tr>
<td>Polyarthritis</td>
<td>Arthralgia</td>
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<tr>
<td>Chorea</td>
<td>Fever</td>
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<tr>
<td>Erythema marginatum</td>
<td>Laboratory findings</td>
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<tr>
<td>Subcutaneous nodules</td>
<td>Increased acute phase reactants:</td>
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<tr>
<td></td>
<td>Increased erythrocyte sedimentation rate</td>
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<td></td>
<td>Increased C-reactive protein</td>
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<td>Prolonged P-R interval</td>
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</table>

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.

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.
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; 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**


