Congenital rubella syndrome

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Background
Since rubella vaccine became available in 1969, the number of reported cases of rubella and congenital rubella syndrome (CRS) has declined dramatically in Canada. During the mumps and rubella consensus conference in 1994, a national goal was set for the elimination of indigenous rubella infection during pregnancy by the year 2000 and thus preventing fetal damage, congenital rubella syndrome and other negative outcomes of infection. A key element in achieving this goal is to have in place sensitive surveillance systems for congenital rubella syndrome to help monitor the effectiveness of immunization programs and to identify any need for change in elimination strategies.

Before the Canadian Paediatric Surveillance Program (CPSP), there were two surveillance systems monitoring CRS in Canada — the Notifiable Diseases Reporting System (NDRS) and the Immunization Monitoring Program, ACTive (IMPACT). Passive reporting of CRS cases to the NDRS began in 1979. From 1987 to 1996, an average of three cases of CRS per year (with very limited epidemiologic information) were reported to this system. However, under-reporting of CRS occurs, as was shown in a study carried out in Quebec in which only five of nine CRS cases identified through an active search of laboratory and hospital discharge databases were reported to public health authorities. Since 1992, active surveillance for CRS has been carried out by the IMPACT network of 11 paediatric hospitals (representing approximately 85% of tertiary care paediatric beds in Canada). IMPACT monitors at each hospital review laboratory reports, admission and discharge records to identify cases, and complete case report forms. The IMPACT network provides valuable information on the CRS cases diagnosed in these specialized settings, but such cases will likely represent those with more severe clinical presentations.

CRS surveillance under the CPSP was initiated in January 1996 to expand active surveillance into the paediatric community setting. Since April 1996, all CRS cases reported to IMPACT have been forwarded to the CPSP, thus integrating paediatric hospital-based and community-based surveillance. The CPSP offers the opportunity to overcome the under-reporting of CRS to the passive surveillance system. The information gathered from the detailed case report forms would allow an assessment...
of the risk factors for CRS in the Canadian population and contribute to the development of improved prevention strategies.

Paediatricians participating in the CPSP are also asked to report newborns with congenital rubella infection (CRI), defined as cases with no clinically compatible manifestations present, but with laboratory confirmation of infection. Children infected with rubella during the intrauterine period may not have congenital defects (especially if infection occurred after the first 16 weeks of gestation) but may develop late-onset manifestations of congenital rubella, including sensorineural hearing loss, peripheral pulmonary artery stenosis, mental retardation, diabetes mellitus, thyroid disorders, growth hormone deficiency and progressive panencephalitis. Therefore, it is important that all children born to mothers suspected or proven to have rubella infection during pregnancy receive appropriate laboratory investigations at the time of birth even if they have normal clinical examinations.

Methods

Physicians or IMPACT investigators who report CRS and CRI cases through the CPSP monthly survey are asked to complete detailed case report forms. Epidemiologic data collected include demographic, clinical and laboratory information on the case, in addition to maternal demographic, immunization and pregnancy histories. If more than one physician reports on a particular case, the information supplied on all case report forms is reviewed and collated. Provincial/territorial public health authorities are consulted to determine if cases have been previously reported.

Objectives

• To improve the reporting of congenital rubella syndrome in Canada.
• To estimate the incidence of congenital rubella syndrome and congenital rubella infection in Canada.
• To obtain detailed epidemiologic data, including maternal histories, on the reported cases of congenital rubella syndrome and congenital rubella infection.

Case definitions

Confirmed case

Live birth

Two clinically compatible manifestations (any combination from Table 1, Columns A and B) with laboratory confirmation of infection:
• Isolation of rubella virus from an appropriate clinical specimen;
or
• Detection of rubella-specific IgM in the absence of recent immunization with rubella-containing vaccine;
or
• Rubella-specific IgG persisting at elevated levels for longer than would be expected from passive transfer of maternal antibody, or in the absence of recent immunization.
Congenital rubella syndrome (continued)

Stillbirth
Two clinically compatible manifestations with isolation of rubella virus from an appropriate clinical specimen.

N.B.: The following cannot be classified as a CRS case:
- Rubella antibody titre absent in the infant.
  or
- Rubella antibody titre absent in the mother.
  or
- Rubella antibody titre declining in the infant consistent with the normal decline after birth of passively transferred maternal antibody.

<table>
<thead>
<tr>
<th>Column A</th>
<th>Column B</th>
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</thead>
<tbody>
<tr>
<td>1. Cataracts or congenital glaucoma (either one or both count as one)</td>
<td>1. Purpura</td>
</tr>
<tr>
<td>2. Congenital heart defect</td>
<td>2. Hepatosplenomegaly</td>
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<td></td>
<td>5. Mental retardation</td>
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<td></td>
<td>6. Meningoencephalitis</td>
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<td>7. Radiolucent bone disease</td>
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<td></td>
<td>8. Developmental or late onset conditions such as diabetes and progressive panencephalitis and any other conditions possibly caused by rubella virus.</td>
</tr>
</tbody>
</table>

Congenital rubella infection

Confirmed case
A case with laboratory confirmation of infection but with no clinically compatible manifestations:
- Isolation of rubella virus from an appropriate clinical specimen.
  or
- Detection of rubella-specific IgM in the absence of recent immunization with rubella-containing vaccine.
  or
- Persistence of rubella-specific IgG at elevated levels for longer than would be expected from passive transfer of maternal antibody, or in the absence of recent immunization.

Duration
January 1996 to December 2004

Expected number of cases
Approximately 10 cases per year
**Ethical approval**
Bioethics Committee, Canadian Paediatric Society

**Funding**
Centre for Infectious Disease Prevention and Control, Health Canada

**Date for analysis and publication**
Data will be analyzed by the principal investigator, and annual reports (along with quarterly updates) will be distributed to CPSP participants, provincial and territorial epidemiologists and Chief Medical Officers of Health.

Publication of data will be at the discretion of the Division of Immunization, Bureau of Infectious Diseases, Centre for Infectious Disease Prevention and Control, Population and Public Health Branch, Health Canada.

**References**
Available from the investigator or the CPSP office.