Appendix 1: Case Definitions and Disease-Specific Information

Disease: Rubella, congenital syndrome

Effective: May 2022
Rubella, congenital syndrome

☒ Communicable
☐ Virulent

Health Protection and Promotion Act (HPPA)  
Ontario Regulation (O. Reg.) 135/18 (Designation of Diseases)

Provincial Reporting Requirements

☒ Confirmed case
☒ Probable case

Ontario is currently documenting the elimination of rubella and CRS and is involved in enhanced surveillance for this disease. Any confirmed or probable case of CRS identified by the board of health should be reported immediately via telephone to Public Health Ontario (PHO).

As part of elimination documentation, it is essential to document maternal rubella and travel history to assess the potential source of infection for every CRS case.

As per Requirement #3 of the “Reporting of Infectious Diseases” section of the Infectious Diseases Protocol, 2018 (or as current), the minimum data elements to be reported for each case are specified in the following:

- O. Reg. 569 (Reports) under the HPPA;
- The iPHIS User Guides published by PHO; and
- Bulletins and directives issued by PHO.

Type of Surveillance

Case-by-case
Case Definition

Confirmed Case

Live birth: Two clinically compatible manifestations (any combination from Table 1: Congenital Rubella Syndrome: Clinically Compatible Manifestations (Table 1, Columns A and B) with laboratory confirmation of infection:

- Isolation of rubella virus from an appropriate clinical specimen (e.g., throat swab, urine, nasopharyngeal aspirate/wash/swab);
  OR
- Detection of rubella virus ribonucleic acid (RNA) by nucleic acid amplification test (NAAT) from an appropriate clinical specimen;
  OR
- Positive serologic test for rubella Immunoglobulin M (IgM) antibody in the absence of recent immunization with rubella-containing vaccine;
  OR
- Rubella Immunoglobulin G (IgG) persisting for longer than would be expected (approximately 6 months following birth) from passive transfer of maternal antibody, or in the absence of recent immunization.

Still birth: Two clinically compatible manifestations with isolation and/or detection of rubella virus RNA from an appropriate clinical specimen (e.g., placenta and autopsy material)

Probable Case

In the absence of appropriate laboratory tests, a case that lacks evidence of any other etiology and has at least:

- Two clinically compatible manifestations listed in Table 1, column A (See Clinical Evidence section);
  OR
- One manifestation listed in Table 1, column A, plus one listed in Table 1, column B (See Clinical Evidence section).
Outbreak Case Definition

Rubella is not an endemic disease in Canada; therefore one confirmed case of Congenital Rubella Syndrome (CRS) is considered an outbreak.

The outbreak case definition varies with the outbreak under investigation. Please refer to the *Infectious Diseases Protocol, 2018* (or as current) for guidance in developing an outbreak case definition as needed.

The outbreak case definitions are established to reflect the disease and circumstances of the outbreak under investigation. The outbreak case definitions should be developed for each individual outbreak based on its characteristics, reviewed during the course of the outbreak, and modified if necessary, to ensure that the majority of cases are captured by the definition. The case definitions should be created in consideration of the outbreak definitions.

Outbreak cases may be classified by levels of probability (*i.e.*, confirmed and/or probable).

Clinical Information

Clinical Evidence

Table 1: Congenital Rubella Syndrome: Clinically Compatible Manifestations

<table>
<thead>
<tr>
<th>Congenital Rubella Syndrome</th>
<th>Clinically Compatible Manifestations</th>
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<tbody>
<tr>
<td>• Cataracts or congenital glaucoma (either one or both count as one)</td>
<td>• Purpura</td>
</tr>
<tr>
<td>• Congenital heart defect</td>
<td>• Hepatosplenomegaly</td>
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<td>• Sensorineural hearing loss</td>
<td>• Microcephaly</td>
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<td>• Pigmentary retinopathy</td>
<td>• Micro ophthalmia</td>
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<td></td>
<td>• Developmental delay</td>
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<tr>
<td></td>
<td>• Meningoencephalitis</td>
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<td>• Radiolucent bone disease</td>
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Clinical Presentation

CRS can result in miscarriage, stillbirth and fetal malformations, including congenital heart disease, cataracts, deafness and intellectual disabilities. Fetal infection can occur at any stage of pregnancy. The greatest risk of fetal damage following maternal infection is highest in the first trimester (90%) which is reduced as the pregnancy progresses and is very uncommon after the 20th week.\(^1\) Infected infants who appear normal at birth may later show eye, ear or brain damage. Congenital infection may give rise to such problems as diabetes mellitus and panencephalitis later in life. Congenitally infected infants may shed the virus in the urine and in nasopharyngeal secretions for one year or more.\(^2\)

Laboratory Evidence

Laboratory Confirmation

Any of the following will constitute laboratory confirmation:

- Positive for rubella IgM in the absence of recent (i.e., 7 - 42 days) immunization with rubella-containing vaccine;
- Rubella IgG persisting longer than would be expected (approximately 6 months) from passive transfer of maternal antibody, or in the absence of recent immunization;
- Isolation of rubella virus from an appropriate clinical specimen;
- Detection of rubella virus RNA.

Approved/Validated Tests

- Standard culture for rubella virus.
- Commercial tests for anti-rubella IgM and IgG antibodies.
- NAAT for rubella virus RNA.
- Consult with laboratory about appropriate specimens for each testing methodology.
Indications and Limitations

- Rubella IgM may not always be detectable at birth following congenital infection. Virus isolation and/or detection of rubella RNA and monitoring of IgG response may be necessary.

- Do not use cord blood. Many of the commercial kits used are not necessarily approved for testing cord blood and validation studies have not been done at the Public Health Ontario Laboratories.

For further information about human diagnostic testing, contact the Public Health Ontario Laboratories.

Case Management

Confirm the diagnosis and ensure that appropriate specimens have been collected for diagnosis according to case definition.

Investigate the maternal history according to the Appendix for Rubella.

In addition to the requirements set out in the Requirement #2 of the “Management of Infectious Diseases – Sporadic Cases” and “Investigation and Management of Infectious Diseases Outbreaks” sections of the Infectious Diseases Protocol, 2018 (or as current), the board of health shall investigate cases to determine the source of infection. Refer to Provincial Reporting Requirements above for relevant data to be collected during case investigation. The following disease-specific information should also be obtained during case management:

- Determine whether the mother received rubella containing vaccine 4 weeks prior to conception;

- Antenatal serological test results; and

- Travel history or exposure to a person who travelled 30 days prior to conception or during pregnancy.

Infants with congenital rubella infection should be isolated from non-immune pregnant women, infants and children, and should be considered infectious until there are 2 sets of negative tests. Urine and nasopharyngeal (NP) specimens in addition to serology should be collected shortly after birth and again in 1-2 months.
If the test results are not negative the infant is considered infectious and should continue to be isolated from non-immune persons. Regular testing should be done until tests are negative.

There is no specific treatment for congenital rubella except for symptomatic and supportive care.³

**Contact Management**

Refer to the Appendix for Rubella.

**Outbreak Management**

Not applicable

**Prevention and Control Measures**

In the event that publicly funded vaccine doses are needed for case and contact management, the board of health should contact the Ministry of Health’s (ministry) immunization program at vaccine.program@ontario.ca as soon as possible.

**Personal Prevention Measures**

Refer to the Appendix for Rubella for prevention of maternal infection during pregnancy.

Prevention strategies:

- Women should avoid pregnancy for at least 4 weeks following immunization.²
- Susceptible women should be discouraged from traveling to rubella-endemic countries the month prior to conception and during pregnancy.

**Infection Prevention and Control Strategies**

Hospitals should obtain documented proof of immunity to rubella as a condition of employment for reasons of patient safety as per the Rubella Surveillance Protocol for Ontario Hospitals.⁵

Routine practices and respiratory isolation precautions are recommended for hospitalized CRS cases; only persons with documented immunity to rubella should have contact with these infants.
**Disease Characteristics**

**Aetiologic Agent** - Rubella virus (family Togaviridae; genus Rubivirus).\(^1\)

**Modes of Transmission** - Transplacental passage of rubella virus from maternal blood.\(^1\)

**Incubation Period** – Not applicable

**Period of Communicability** - Birth to 9-12 months of age, rarely longer. A small number of infants with CRS continue to shed virus in nasopharyngeal secretions and urine for one year or more and can transmit infection to susceptible contacts.\(^1\)

**Reservoir** - Humans; source is maternal viremia.\(^3\)

**Host Susceptibility and Resistance** - Fetuses of rubella-susceptible pregnant women who have not received at least one dose of rubella-containing vaccine. Immunity is usually permanent after immunization and natural infection.\(^1\)

Please refer to [PHO's Reportable Disease Trends in Ontario reporting tool](https://www.phac-aspc.gc.ca/rdre-rdqa/cmnts-cmns/cmrss-syss/ribell-crs-eng.php) for the most up-to-date information on infectious disease trends in Ontario.

Canada, as well as the Americas, have successfully eliminated transmission of rubella virus and CRS. Endemic transmission of rubella and CRS has been interrupted by high vaccine coverage as a part of routine infant and childhood immunization programs. Rubella and CRS continue to be endemic in other areas of the world and therefore importation of rubella is an ongoing concern.

For additional national and international epidemiological information, please refer to the Public Health Agency of Canada and the World Health Organization.

**Comments**

Provincial and territorial ministries of health provide active, weekly case-by-case notification (including zero-notification) to the Canadian Measles/ Rubella Surveillance System (CMRSS).

Weekly reporting is completed from CMRSS to the Pan-American Health Organization, in accordance with the elimination of rubella and congenital rubella syndrome in the Western Hemisphere.
References


Case Definition Sources


Document History

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<tr>
<td>April 2022</td>
<td>Entire Document</td>
<td>New template. Appendix A and B merged. No material content changes.</td>
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<td>Epidemiology: Occurrence section</td>
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