

# Rubella, Congenital Syndrome

## 2010 Case Definition

### Case classification

**Suspected:** An infant that does not meet the criteria for a probable or confirmed case but who has one or more of the following clinical findings:

- cataracts or congenital glaucoma,
- congenital heart disease (most commonly patent ductus arteriosus or peripheral pulmonary artery stenosis),
- hearing impairment,
- pigmentary retinopathy
- purpura,
- hepatosplenomegaly,
- jaundice,
- microcephaly,
- developmental delay,
- meningoencephalitis, or
- radiolucent bone disease.

**Probable\*:** An infant without an alternative etiology that does not have laboratory confirmation of rubella infection but has at least 2 of the following:

- cataracts or congenital glaucoma,\*
- congenital heart disease (most commonly patent ductus arteriosus or peripheral pulmonary artery stenosis),
- hearing impairment, or
- pigmentary retinopathy;

### OR

An infant without an alternative etiology that does not have laboratory confirmation of rubella infection but has at least one or more of the following:

- cataracts or congenital glaucoma,\*
- congenital heart disease (most commonly patent ductus arteriosus or peripheral pulmonary artery stenosis),
- hearing impairment, or
- pigmentary retinopathy

**AND** one or more of the following:

- purpura,
- hepatosplenomegaly,
- jaundice,
- microcephaly,
- developmental delay,
- meningoencephalitis, or
- radiolucent bone disease.

**Confirmed:** An infant with at least one symptom (listed above) that is clinically consistent with congenital rubella syndrome; and laboratory evidence of congenital rubella infection as demonstrated by:

- isolation of rubella virus, or
- detection of rubella-specific immunoglobulin M (IgM) antibody, or
- infant rubella antibody level that persists at a higher level and for a longer period than expected from passive transfer of maternal antibody (i.e., rubella titer that does not drop at the expected rate of a twofold dilution per month), or
- a specimen that is PCR positive for rubella virus.

**Infection only:** An infant without any clinical symptoms or signs but with laboratory evidence of infection as demonstrated by

- isolation of rubella virus, or
- detection of rubella-specific immunoglobulin M (IgM) antibody, or
- infant rubella antibody level that persists at a higher level and for a longer period than expected from passive transfer of maternal antibody (i.e., rubella titer that does not drop at the expected rate of a twofold dilution per month), or
- a specimen that is PCR positive for rubella virus.

\*In probable cases, either or both of the eye-related findings (cataracts and congenital glaucoma) count as a single complication. In cases classified as infection only, if any compatible signs or symptoms (e.g., hearing loss) are identified later, the case is reclassified as confirmed.

### **Epidemiologic Classification of Internationally-Imported and U.S.-Acquired**

Congenital Rubella Syndrome (CRS) cases will be classified epidemiologically as internationally imported or U.S.-acquired, according to the source of infection in the mother, using the definitions below, which parallel the classifications for rubella cases.

*Internationally imported case:* To be classified as an internationally imported CRS case, the mother must have acquired rubella infection outside the U.S. or in the absence of documented rubella infection, the mother was outside the United States during the period when she may have had exposure to rubella that affected her pregnancy (from 21 days before conception and through the first 24 weeks of pregnancy).

*U.S.-acquired case:* A US-acquired case is one in which the mother acquired rubella from an exposure in the United States. U.S.-acquired cases are subclassified into four mutually exclusive groups:

**Import-linked case:** Any case in a chain of transmission that is epidemiologically linked to an internationally imported case.

**Import-virus case:** a case for which an epidemiologic link to an internationally imported case was not identified but for which viral genetic evidence indicates an imported rubella genotype, i.e., a genotype that is not occurring within the United States in a pattern indicative of endemic transmission. An endemic genotype is the genotype of any rubella virus that occurs in an endemic chain of transmission (i.e., lasting  $\geq 12$  months). Any genotype that is found repeatedly in U.S.-acquired cases should be thoroughly investigated as a potential endemic genotype, especially if the cases are closely related in time or location.

**Endemic case:** a case for which epidemiological or virological evidence indicates an endemic chain of transmission. Endemic transmission is defined as a chain of rubella virus transmission continuous for  $\geq 12$  months within the United States.

**Unknown source case:** a case for which an epidemiological or virological link to importation or to endemic transmission within the U.S. cannot be established after a thorough investigation. These cases must be carefully assessed epidemiologically to assure that they do not represent a sustained U.S.-acquired chain of transmission or an endemic chain of transmission within the U.S.

**Note:** Internationally imported, import-linked, and imported-virus cases are considered collectively to be import-associated cases.

States may also choose to classify cases as “out-of-state-imported” when imported from another state in the United States. For national reporting, however, cases will be classified as either internationally imported or U.S.-acquired.