

Rubella (German Measles)

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Definition

Rubella (German Measles) is a common communicable viral disease of childhood. It tends to be mild in children causing a rash and swollen, tender lymph glands. In adults the infection may be more severe and may cause arthritis especially in women. Rubella in early pregnancy is a much more serious situation for the fetus because the virus may infect the placenta and then the unborn baby. It may cause miscarriage, stillbirth, intrauterine growth retardation, prematurity, low birth weight, and/or a multisystem spectrum of abnormalities that together are called Congenital Rubella Syndrome (CRS).

Introduction

Thanks to the development of a vaccine for Rubella, 80-90% of the population of the United States is immune. However, outbreaks continue to occur, usually among adolescents and young adults attending college or in workplaces with close person-to-person contact.

The risk of congenital malformations with CRS depends on the time during the pregnancy that the unborn baby (fetus) is infected. For example, congenital heart disease is common when the infection occurs during the first trimester (first 3 months of pregnancy.) When the infection occurs in the last trimester (last 3 months of pregnancy) the risk of congenital malformations is very small, but the baby may develop a chronic infection with dysfunction of the eyes, ears, brain, and liver. The baby may excrete virus for months to years after birth. Some manifestations of CRS are limited to the newborn period; others remain or develop over the lifespan of the individual with CRS.

Transient abnormalities that may be found in the newborn with CRS include:

1. Intrauterine growth retardation
2. Jaundice
3. Purpura and/or a blueberry muffin rash
4. Swollen lymph glands
5. Low platelet count
6. Abnormalities at the ends of the femur and humerus on X-ray
7. Enlarged liver and spleen (hepatosplenomegaly)
8. Meningoencephalitis (infection of brain)
9. Pneumonia
10. Myocarditis (inflammation of the heart muscle)
11. Anemia
12. Cloudy cornea of the eye

Permanent abnormalities that may be present at birth or manifest over a lifetime include:

1. Growth retardation
2. Hearing loss – sensorineural hearing loss and deafness
3. Visual loss – including cataracts, microphthalmia (small eye size), retinopathy, corneal opacity, glaucoma, nystagmus (abnormal eye movement), retinal detachment, sub retinal neovascularization, keratoconus, lens absorption, diabetic retinopathy and blindness

4. Congenital heart defects – including patent ductus arteriosus (PDA), peripheral pulmonary stenosis (narrowing), pulmonary valve stenosis (narrowing), ventricular septal defect (VSD), and atrial septal defect (ASD)
5. Abnormalities of blood vessels – including renal artery stenosis (narrowing), high blood pressure and arteriosclerosis (hardening of the arteries)
6. Genitourinary abnormalities – including undescended testicles, inguinal hernia, hypospadias and polycystic ovaries
7. Gastrointestinal problems – including GERD (Gastroesophageal reflux), esophageal stricture, gagging, cyclic vomiting, swallowing difficulties, and cirrhosis of the liver
8. Endocrine abnormalities – including hyperthyroidism, hypothyroidism thyroiditis, Addison's Disease (adrenal insufficiency), excessive hairiness in females (hirsutism), polyglandular autoimmune disease, growth hormone deficiency, diabetes, and precocious (early onset) puberty
9. Abnormal teeth
10. CNS abnormalities – including microcephaly (small head size), mental retardation (mild, moderate, severe, profound), autism, progressive rubella panencephalitis (PRP), cerebral palsy, behavior problems (self stimulatory, self-abusive, self-injurious, impulsivity, tantrums), intracranial calcifications and seizures

With so many possible manifestations of this syndrome that can show up at different times over the lifespan doctors may have their hands full caring for individuals with CRS! Some of these abnormalities are much more common than others. It can be helpful to plan regular assessments so that common conditions do not slip unnoticed between the cracks. We recommend the following:

1. Hearing screening in newborns, frequent repeat audiology evaluations throughout lifespan.
2. Ophthalmology consultation in newborns, frequent follow-up examinations throughout lifespan. Eye pain from glaucoma may present as increased behavior problems, SIB and head banging.
3. Regular developmental assessment in children.
4. Pediatric cardiology consult as newborn, corrective cardiac surgery may be needed, and SBE (sub acute bacterial endocarditis) prophylaxis may be needed before dental procedures, etc. Check with cardiologist.
5. Check BP (blood pressure) every visit.
6. GERD is very common among individuals with CRS and other developmental disabilities. It may present with classic symptoms of burping and heartburn, but it may present with weight gain or weight loss, increased crying or behavior problems. A high index of suspicion is key to diagnosis.
7. Annual urine test or fasting blood sugar to rule out diabetes.
8. Annual TSH, T4 to rule out thyroid dysfunction.

Diagnosis

Diagnosis of CRS is usually made in the newborn period when virus can be isolated. There are also blood tests such as rubella-specific IgM antibody or a persistent rubella hemagglutination inhibition (HI) titer.

Prevention and Treatment

Congenital Rubella Syndrome can be prevented by immunization with MMR or Rubella vaccine.

There is no specific treatment for CRS. Individual manifestations of the condition will be treated as they occur. For example:

1. Hearing loss may require SLP therapy, hearing aids, and special education
2. Visual problems will require frequent ophthalmologic follow up, possible surgery, glasses, early intervention for the visually impaired, special education, and mobility training

3. Cardiac problems may require cardiology follow up, surgery and SBE (Subacute Bacterial Endocarditis) prophylaxis
4. Hypertension may require anti-hypertensive medication
5. Genitourinary tract abnormalities may require urology consultation and surgery
6. GERD will require medication, swallowing disorders may require SLP or OT therapy, a specific mealtime program or consultation by GI specialist or SAFE feeding clinic
7. Thyroid dysfunction, diabetes, growth hormone deficiency, Addison's disease and polycystic ovarian disease will require appropriate endocrine evaluation and treatment
8. CNS problems will require treatment such as anticonvulsant medication for seizures. EI (early intervention), special education and OT, PT, SLP therapy for individuals with mental retardation. OT, PT and SLP services for individuals with cerebral palsy. Behavior therapy and/or psychiatric consultation and medication for individuals with behavior problems.

Emergency Situations – What can go wrong?

Seizures can lead to:

- **Status epilepticus** – prolonged seizure activity such as a seizure that lasts for more than 10 minutes or several seizures that occur one after another for 20-30 minutes.
- **Injury** – including bruising, concussion, fractures or even drowning if the seizure occurs during a bath.
- **Trouble breathing** – individual's lips may turn blue.

What to Do:

1. Clear the area around the individual, stay with him/her to prevent injury. DO NOT put anything in his/her mouth.
2. Try to write down what happened before, during and after the seizure and how long the seizure lasts.
3. Notify agency nurse/supervisor as soon as possible.
4. Call 911 if the seizure lasts longer than 5 minutes, if individual is injured or if he/she stops breathing.
5. Begin rescue breathing if you are certified to do so.

Acute Glaucoma:

Notify individual's ophthalmologist as soon as you suspect this complication.

Conclusion

Congenital Rubella Syndrome is a constellation of abnormalities found in individuals who were infected with the rubella virus before birth. Manifestations may be transient and limited to the newborn period or permanent and be present at birth or develop over the lifespan of an involved individual. Almost every body system can be involved – brain, eyes, ears, heart, lungs, liver, endocrine glands, gastrointestinal and genitourinary tract. Repeated developmental, auditory, visual and endocrinologic assessment is required. No cure is available but therapy is aimed at improved quality of life and may include corrective surgery, hearing aids, OT, PT and SLP therapy, special education, behavioral therapy and others. CRS can be prevented by immunization with MMR or Rubella vaccine.

References

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O'Donnell, Nancy, "History of congenital rubella syndrome", Journal of Vocational Rehabilitation 6 (1996) 149-157.