



Overview

- Definition
 - An acute, contagious, exanthematous disease caused by Rubrivirus
 - Spread only by humans, via respiratory route or transplacentally
 - Essentially extinct in the United States after the introduction of the measles, mumps, and rubella (MMR) vaccines in 1969; about 20,000 were born with congenital disease in mid-1960s
 - In the United States, acquired cases are typically contracted from foreign visitors or travel
- Symptoms
 - Redness
 - Purulent discharge
 - Photophobia
 - Sudden vision loss
- Laterality
 - Unilateral or bilateral
- Course
 - Acute and self-limited
- Age of onset
 - All age groups affected
- Gender/race
 - No gender or racial predilection
- Systemic association
 - Congenital rubella syndrome (CRS)
 - Cardiac, hearing, and ocular defects
 - Severity depends on time of maternal disease contraction: most severe during first trimester (miscarriage may result); defects unlikely after 20 weeks gestation

- Acquired rubella (German measles)
 - Spread by respiratory route; common in spring and winter
 - Incubation period of 2–3 weeks; hosts are contagious 1 week prior to rash onset and throughout active symptoms

Exam: Ocular

Congenital Rubella

Ocular findings can be present at birth, shortly after birth, or later in life

- Salt-and-pepper retinopathy (inactive chorioretinitis, usually confined to RPE)
 - Unilateral or bilateral
 - Typically stable but may progress later in life
 - Vision ranges from normal to 20/200
 - Rarely complicated by CNV. May have focal necrosis of ciliary epithelium, pars plicata, or pars plana
- Nuclear cataract
 - Significant AC inflammation after cataract extraction due to liberation of live virus from lens. Some cataracts have been known to resorb, leaving KP's behind
- Glaucoma
 - Corneal clouding and buphthalmos
- Microphthalmia

Acquired Rubella

- Conjunctivitis
 - Most common ocular finding (70%)
- Epithelial keratitis
 - Infrequent and resolves without sequelae within 1 week
- Retinitis
 - Rare
 - May present as multifocal exudative RD and RPE detachment
 - No retinal hemorrhage
- Chronic rubella has been implicated as a cause of Fuchs' heterochromic iridocyclitis (Chap. 11)

Exam: Systemic

Congenital Rubella

- Unilateral or bilateral deafness (>80%)
- Cardiac malformations: patent ductus arteriosus, peripheral pulmonary artery stenosis, and interventricular septal defects

Acquired Rubella

- Rubella exanthem
 - Erythematous and maculopapular
 - Spreads from face down to hands and feet over 24 hours
 - Resolves by day 3 (thus second nickname “3-day measles”)
 - Not always present
- Fever
 - Variable
 - After rash in children; before rash in adolescents and adults
- Postauricular and suboccipital lymphadenopathy
- Other complications: arthritis, encephalitis, thrombocytopenic purpura

Imaging

- FA
 - Abnormal hyperfluorescence and hypofluorescence in salt-and-pepper retinopathy
 - Early hyperfluorescence with late leakage in CNV
 - Hyperfluorescence associated with area of retinitis without vascular leakage in acquired rubella
- ERG
 - Normal in salt-and-pepper retinopathy

Laboratory and Radiographic Testing

- Maternal serum rubella titers
- Viral culture and/or reverse-transcriptase PCR of amniotic fluid, nose, throat, urine, blood, or cerebrospinal fluid (CSF)
- Serum IgM titer is useful in children with anomalies from uneventful pregnancies
- Hearing test
- Echocardiogram

Differential Diagnosis

Congenital Anomalies

- TORCH infections
 - *Toxoplasmosis*
 - Other infections (syphilis, parvovirus, varicella zoster, Zika)
 - *Rubella*
 - *Cytomegalovirus*
 - *Herpes simplex*

Salt-and-pepper Retinopathy

- Congenital syphilis
- Congenital measles
- Leber's congenital amaurosis
- Retinitis pigmentosa carrier
- Choroideremia carrier
- Albinism carrier
- Cystinosis
- Drug toxicity (e.g., phenothiazine)
- Kearns–Sayre syndrome

Salt-and-pepper Retinopathy and Hearing Loss

- Congenital syphilis
- Usher's syndrome

Treatment

Congenital Rubella

- Mothers infected during first trimester should be counseled about possible birth defects
- Immune globulin within 72 hours of exposure during pregnancy
- Rubella vaccination is contraindicated during pregnancy
- CRS infants are contagious at birth and need isolation

Acquired Rubella

- Treatment is supportive, as there is no known antiviral therapy for rubella
- Topical NSAIDs and artificial tears to reduce conjunctival hyperemia
- Retinitis can benefit from corticosteroids

Referral/Co-management

- Audiology
- Cardiology
- Infectious Disease