

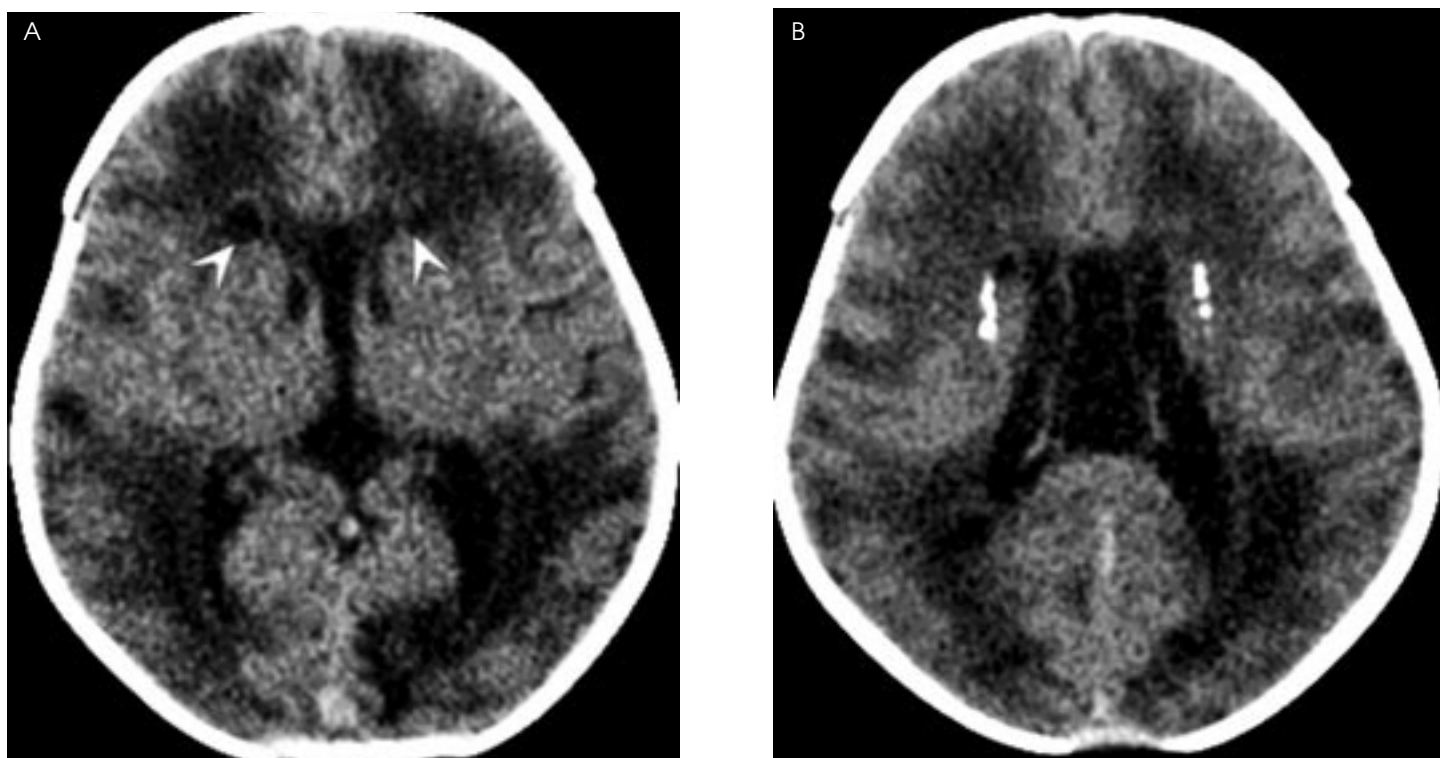
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# Case W5

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## Clinical Presentation

A newborn infant with hypotonia, microcephaly, microphthalmia, and a patent ductus arteriosus presents for CT scanning.



## Radiologic Findings

A non-contrast CT scan (Fig. A) demonstrates accentuation of low density in the white matter as compared with a normal neonate. In addition, bifrontal subependymal cysts (*arrowheads*) are present. The superior sagittal sinus is slightly dense, probably related to mild dehydration and hemoconcentration. An image at a slightly higher level (Fig. B) demonstrates calcification along the lateral aspect of the caudate nuclei, as well as mild ventricular prominence. Incidental note is made of a cavum septum pellucidum.

## Diagnosis

Congenital rubella infection

## Differential Diagnosis

- Congenital cytomegalovirus (periventricular calcifications, cerebral cortical malformations)

## Pearls

- CT best demonstrates calcifications and inner ear malformations, while MR best demonstrates delayed myelination and white matter hyperintensities.
- Cranial ultrasound may demonstrate linear increased echoes in the basal ganglia. These are associated with congenital rubella but are nonspecific. Their detection, however, should prompt a workup for congenital infection.

## Pitfall

- Postnatally acquired rubella may rarely cause a postinfectious encephalomyelitis or a progressive panencephalitis similar to subacute sclerosing panencephalitis (SSPE).

- Congenital toxoplasmosis (randomly scattered calcification, often extensive parenchymal destruction)

## Discussion

### Background

Congenital rubella is now rare in Western nations because of childhood vaccination and mass screening of pregnant women, with an incidence of ~1 per 100,000 live births in the United States. Placental transmission of the rubella virus occurs at the time of primary maternal infection, with the risk of fetal infection being greatest when the maternal rash appears between 3 and 6 weeks from the last menstrual period. The gestational age of the fetus at the time of infection has a significant influence on the outcome. Early infection may kill the fetus or result in severe abnormalities, while third trimester infection may be without apparent effect. Since most maternal rubella infection is subclinical and most of the infants with congenital rubella syndrome are minimally symptomatic at birth, timely diagnosis may be difficult.

### Etiology

The rubella virus is a togavirus that contains enveloped RNA.

### Clinical Findings

The infant with congenital rubella is usually lethargic and hypotonic at or soon after birth. Ten to 20% of affected infants will manifest symptoms of an acute meningoencephalitis. Other features include microphthalmia, microcephaly, cataracts, glaucoma, cardiac anomalies, motor and mental retardation, and seizures. Deafness is the most common feature, but is often not detected until the infant is older.

### Pathology

#### Gross

- Microcephaly, thought to be secondary to a direct inhibitory effect of the rubella virus on mitosis, is often striking

#### Microscopic

- Generalized vasculitis with relatively little cellular necrosis results in ischemic brain damage. The small penetrating arteries supplying the deep white matter and basal ganglia are most commonly affected. Perivascular calcification and subependymal cysts may result.
- Leptomeningitis with infiltration of chronic inflammatory cells

### Imaging Findings

Both CT and MR show microcephaly and ventriculomegaly.

#### CT

- Punctate calcification in basal ganglia, periventricular white matter, cortex
- Malformations of the inner ear

## MR

- Delayed myelination, decreased volume of cortical gray matter
- Multifocal white matter hyperintensities
- Subependymal cysts not related to prior hemorrhage have been described

## Treatment

- None
- Prevention via vaccination is the best strategy. Unfortunately, not all immunized patients develop antibodies to rubella, and the presence of antibodies does not always prevent rubella infection.

## Prognosis

Varies with the severity of brain involvement

## Suggested Readings

Chang Y-C, Huang C-C, Liu C-C. Frequency of linear hyperechogenicity over the basal ganglia in young infants with congenital rubella syndrome. *Clin Infect Dis* 22:569–571, 1996.

Lane B, Sullivan EV, Lim KO, et al. White matter MR hyperintensities in adult patients with congenital rubella. *AJNR* 17:99–103, 1996.

Shaw DWW, Cohen WA. Viral infections of the CNS in children: imaging features. *AJR* 160:125–133, 1993.