

OBJECTIVES

To determine the incidence of congenital and neonatal varicella in Australia; evaluate the morbidity and mortality rate of congenital and neonatal varicella and; assess the need for varicella vaccination of non immune women prior to pregnancy.

INVESTIGATOR CONTACT DETAILS (*Principal Investigator)

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SUMMARY PROTOCOL

It is estimated that varicella occurs in about 1 in 2,000 pregnancies in the United States and about 3 in 1,000 in the United Kingdom, however the incidence in Australia is unknown. In Australia 90-95% of adult women are immune to varicella. Varicella may cause spontaneous abortion or premature delivery. If the infection occurs in the first 20 weeks of gestation, the fetus may be damaged. The risk of embryopathy is about 2% when maternal infection occurs between 13-20 weeks gestation and 0.4% when it occurs before 13 weeks. There are only two reports of embryopathy following maternal infection later than 20 weeks. Typical features of the *fetal varicella syndrome* include hypoplasia of one limb with cicatricial skin lesions in a dermatomal distribution affecting that limb, often forming a zig-zag pattern, pox-like skin scars, intrauterine growth retardation, neurological abnormalities (microcephaly, hydrocephalus and cerebellar hypoplasia, motor or sensory deficits and sphincter dysfunction) eye lesions (microphthalmia, cataracts, Horner's Syndrome, chorioretinitis, retinal scars, nystagmus, and optic atrophy) and occasional gastrointestinal (hepatic failure) and genito-urinary abnormalities.

Varicella embryopathy cannot always be confirmed by serology. Varicella zoster specific IgM antibody is found in about 25% of infants with clinical manifestations of intrauterine infection and specific IgG antibody in about 70%. Infants who appear normal after maternal infection may also have detectable levels of specific IgM antibody.

Neonatal varicella (varicella developing in the first month of life) may be due to either intrauterine or post natal infection. When maternal varicella occurs 1-4 weeks before delivery the fetal infection rate is about 50%; one third of these infants develop varicella. Their illness may be severe, with fever, haemorrhagic rash and generalized visceral involvement. Neonatal varicella has a high morbidity and mortality rate even when treated with acyclovir. A live attenuated varicella is licensed in some European countries. It is expected that it will also become available in Australia.

This study aims to document the incidence of congenital and neonatal varicella infection and its associated morbidity and mortality rate and to assess the need for varicella vaccination of non-immune women prior to pregnancy.

CASE DEFINITION

Any stillbirth, newborn infant, or child up to the age of 2 years who, in the opinion of the notifying paediatrician, has definite or suspected congenital varicella syndrome, with or without defects or neonatal varicella, based on history, clinical and laboratory findings.

Factors which may lead to a suspicion of congenital or neonatal varicella infection are:

- Single or multiple congenital abnormalities, particular hypoplasia or one limb with skin scarring affecting that limb, poxlike skin scars, intrauterine growth retardation, microcephaly, hydrocephalus, cerebellar hypoplasia, microphthalmia, cataracts, Horner's syndrome, chorioretinitis, nystagmus, optic atrophy, gastrointestinal and genito-urinary abnormalities.
- Stillbirth following varicella in pregnancy
- Development of herpes zoster in the first year of life
- Varicella with onset in the first two weeks of life

Diagnosis may be confirmed by:

- Detection of varicella specific IgM antibodies in cord blood or in a serum specimen in the first months of life (only 25% of cases are positive)
- Persistence of varicella specific IgG antibody in a child aged 6-12 months (only 70% of cases)
- Identification of varicella virus in skin lesions (rarely) or autopsy tissue
- A history of varicella during pregnancy or of maternal contact with varicella in the first 20 weeks of pregnancy in an infant with congenital anomalies

REPORTING INSTRUCTIONS

Initial 3 mailings

Please report any **new** patient with definite or suspected congenital or neonatal varicella infection you have seen since 1 January 1995.

Subsequent mailings

Please report any patient seen this month with definite or suspected congenital or neonatal varicella infection, whom you have not already reported.