



Australian Paediatric Surveillance Unit
STUDY INFORMATION SHEET
Congenital Varicella Syndrome

Updated
2021

BACKGROUND

Congenital varicella may cause spontaneous abortion or premature delivery and may also cause embryopathy. Typical features of the Congenital Varicella Syndrome (CVS) include hypoplasia of one limb with cicatricial skin lesions in a dermatomal distribution affecting that limb, intrauterine growth retardation, neurological abnormalities (including: microcephaly, hydrocephalus and cerebellar hypoplasia, motor and sensory deficits and sphincter dysfunction), eye lesions (including microphthalmia, cataracts, Horner's Syndrome, chorioretinitis, retinal scars, nystagmus and optic atrophy) and occasional gastrointestinal (hepatic failure) and genito-urinary abnormalities^(1,2). Although most CVS results from maternal infection early in the pregnancy, CVS has been reported following maternal infection at 26 weeks of pregnancy⁽¹⁾.

There has been no systematic surveillance of congenital varicella in Australia since the end of the APSU study in 1997. Given that varicella vaccination is now recommended in the latest NHMRC immunisation schedule (2, this study gives a unique opportunity to compare current rates of congenital and neonatal infection and the sources of infection in Australia to rates reported in 1995-1997.

STUDY OBJECTIVES

1. To estimate the incidence of congenital varicella syndrome seen by Australian paediatricians and its associated morbidity and mortality.
2. To compare results with those from a previous APSU study of congenital varicella that concluded in 1997, prior to the availability of varicella vaccination.
3. To document the source of maternal infection and the management and short-term outcome of congenital varicella syndrome.
4. To estimate the need for screening to identify non-immune women antenatally
5. To identify CVS in women who had been exposed to varicella during pregnancy and had not received Zoster immunoglobulin (ZIG)

CASE DEFINITION AND REPORTING INSTRUCTIONS

*Report 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 and meets **at least one of the following criteria:***

1. Cicatricial skin lesions in a dermatomal distribution and/or pox-like skin scars and/ or limb hypoplasia
2. Development of Herpes Zoster in the first year of life
3. Spontaneous abortion, termination, stillbirth or early death following varicella infection during pregnancy

Confirm varicella by one or more of the following:

- Detection of varicella-specific IgM antibodies in cord blood or in serum specimen taken in the first 3 months of life (only 25% of cases are positive)

- Persistence of varicella specific IgG antibody in a child aged beyond 6 months of age^(3,4)
- Identification of varicella virus in skin lesions or autopsy tissue
- History of maternal varicella during pregnancy or maternal contact with varicella in pregnancy in the mother of an infant with congenital abnormalities

The following clinical signs may also be present in cases of congenital varicella syndrome:

- microcephaly, hydrocephalus, cerebellar hypoplasia, motor or sensory deficits, sphincter dysfunction and peripheral nervous system defects
- microphthalmia, cataracts, Horner's Syndrome, chorioretinitis, nystagmus, retinal scars, optic atrophy
- gastrointestinal abnormalities including colonic atresia, hepatitis, liver failure
- genito-urinary abnormalities
- cardiovascular abnormalities
- intrauterine growth retardation

PRINCIPAL INVESTIGATOR

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FURTHER INFORMATION

For further information related to this study or assistance completing the Case Report Form, please contact the APSU by either:

- email: SCHN-APSU@health.nsw.gov.au or
- phone: (02) 9845 3005

REFERENCES

1. Forrest JM, Mego S, Burgess MA. Congenital and neonatal varicella in Australia. *J Paediatr. Child Health* 2000; 36:108-113.
2. Australian Government, Department of Health and Ageing. Immunisation Provider Guidelines, National Varicella (Chickenpox) vaccination program 10th October 2005. Accessed on www.immunise.health.gov.au
3. Birthistle K, Carrington D. Fetal Varicella Syndrome – a Reappraisal of the Literature. *Journal of Infection* 1998; 36(Suppl1): 25-29.
4. Leinweber B, Grote V, Schaad UB, Heininger U. Transplacentally acquired immunoglobulin G antibodies against measles, mumps, rubella and varicella-zoster virus in preterm and full term newborns. *Pediatr Inf Dis J* 2004; 23:361-363.