

Congenital Varicella Syndrome and Neonatal Chicken Pox (Varicella)

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ABSTRACT

Varicella is an infection perceived to be a benign disease. But if adults get the disease, especially if pregnant mother gets chicken pox, it could be fatal or could cause disease in the unborn baby varying from fetal varicella syndrome to neonatal varicella—both causing significant morbidity and mortality. Understanding of the disease is very important, especially at what age it can cause problems. Unlike many other viral infections, varicella during pregnancy can cause different problems at different gestations.

Keywords: Congenital varicella, Fetal varicella syndrome, Neonatal varicella, Varicella zoster, Varicella zoster immunoglobulin.

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Chicken pox also called varicella is the THIRD disease that was described and is a highly infectious disease caused by varicella zoster virus (VZV). It was not differentiated from smallpox until the late 19th century. The first documented record of the word “Chicken pox” was in 1658, although various explanations have been given for the word “Chicken,” one of them was the relative mildness of the disease compared to smallpox and the other one was that the rash looks like the peck marks caused by a chicken!

There are references in ancient Babylonia more than 2,000 years ago of a disease similar to chicken pox. In the late 800 AD, Muhammad Zakariya Razi recorded the first known information on chicken pox and also noted the differences between measles and small pox. In 1500s, Giovanni Filippo gave a more detailed description of chicken pox. Although chicken pox is one of the “oldest diseases”, it was in 1767 that William Heberden demonstrated smallpox being different from chicken pox. In 1875, Rudolf Steiner showed that chicken pox was caused by an infectious agent. In 1909, Von Bokay was the first one to observe the clinical difference between herpes zoster and varicella.

Dr Michiaki Takahashi had spent several years studying measles and polio viruses in Japan. He was on a research fellowship in 1964 at Baylor Medical College in Houston, when his son, Teruyuki, developed severe chicken pox while playing with a friend who had the virus. In 1965, he returned to Japan and within 5 years he developed first version of the vaccine. By 1972, he had developed the vaccine for clinical trials and officially it was licensed in 1984 in Japan. Since then, the vaccine use is gradually spreading in rest of the world.

Chicken pox is very important for obstetricians, neonatologists, and pediatricians—as it is one of the “diseases thought to be benign”—but can cause devastating problems, especially in the perinatal period. Children under 1 year of age or over 15 years tend to have more severe disease and more complications.¹ Although it is a common childhood infection, the disease can have its incidence in adults, especially susceptible population. Also, it is one of the few virus diseases that can result in the following two forms of clinical disease: primary infection such as chicken pox (varicella) or secondary reactivation from latent disease in the dorsal root of sensory ganglia such as herpes zoster (shingles). With increasing survival of people with cancer, contact of a person undergoing

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chemotherapy with a person suffering from chicken pox will be devastating.

India poses a peculiar problem, with chicken pox being quite common in some states—most children are immune to chicken pox. But in some areas such as Kerala and coastal Karnataka, for example, “adult chicken pox” is relatively common, suggesting that many adults are susceptible to chicken pox. Hence, the morbidity is reasonably high in India, considering that the adult chicken pox is so common. This means we can potentially have lot of pregnant women susceptible to be contacting this disease during their pregnancy.²

If a pregnant woman gets chicken pox during pregnancy, it can have deleterious effects on the unborn baby depending upon which period of gestation she is in.³ Primary infection in pregnancy can cause maternal mortality too or serious morbidity. Even the mother can get severe pneumonia or secondary respiratory compromise. Pneumonia occurs in up to 10% of pregnant women with chicken pox and the severity increases as the gestation increases. Chicken pox during the periconceptional period until 20 weeks can cause embryopathy and possible teratogenic effects including miscarriage. Congenital varicella syndrome or fetal varicella syndrome is a relatively rare disease in which the affected babies have distinctive abnormalities at birth due to maternal varicella early during pregnancy (i.e., up to 20 weeks gestation).^{4,5} The affected newborns will generally have a low birth weight and characteristic abnormalities of the skin—scarring in a dermatome distribution; the arms, legs, hands, and/or feet (extremities); the brain; the eyes (cataracts, chorioretinitis, or microphthalmia); and/or,

in rare cases, other areas of the body. Some babies may also exhibit hypoplasia of fingers and/or toes or rudimentary digits. Some babies may have cortical atrophy, ventriculomegaly, microcephaly, to name a few, among other changes. The range and severity of associated symptoms and physical findings may vary greatly from case to case depending upon when maternal varicella infection occurred during fetal development, with the earlier the infection, the more marked the damages.⁶

Between 20 weeks and 38 weeks, generally chicken pox is reasonably benign or thought to be benign from the fetus point of view. But if a pregnant mother has severe disease, the possibility of intrauterine death is higher.^{7,8} But if a woman gets chicken pox between 2 weeks before delivery and 1 week after delivery, the neonate is at the risk of getting “vertical transmission” and possibly “congenital or neonatal chicken pox”, which can have disastrous effect including death of the neonate. Hence, most perinatologists would agree to treat the baby with intravenous varicella zoster immunoglobulin along with acyclovir and strict isolation for both the mother and baby, if the mother gets chicken pox during this period. Delivering a pregnant woman during the viremic period is hazardous and increases maternal risks to include bleeding, thrombocytopenia, disseminated intravascular coagulation, hepatitis, and so on. Also, the risk of varicella in the newborn is very high with significant mortality and morbidity. Hence, it is better to deliver the baby after 5 days of the onset of maternal illness to allow for passive transfer of antibodies. But if the delivery happens within 5 days of maternal varicella, then the baby should be given varicella zoster immunoglobulin and treated with acyclovir. Maternal shingles or varicella zoster infection is not a risk to the newborn because it is protected by the transplacental transfer of maternal antibodies.

Neonatal or congenital chicken pox is basically infection with varicella zoster (chicken pox) virus soon after birth. In most cases, the affected babies exhibit varicella infection due to the active infection in the mother with chicken pox during the last weeks or days of pregnancy. Many a times, neonatal chicken pox may result due to baby’s exposure to the virus soon after birth. Because of the time span that typically occurs between exposure to the virus and initial symptoms (i.e., approximately 10–14-day incubation period), if the mother initially contracted chicken pox more than a week before giving birth, the affected newborn may have received antibodies from the mother that help fight the virus (transplacental anti-VZV antibody). However, if the interval from maternal VZV infection to birth was less than a week, the newborn may not have received protective antibodies from the mother. In such cases, neonatal chicken pox may result in severe symptoms. Chicken pox tends to be a severe illness in the first year of life, purely attributed to the lack of antibodies from the mother. Symptoms of neonatal chicken pox include a low-grade fever, listlessness, lack of appetite (anorexia), and the appearance of an itchy (pruritic)

rash. The rash consists of small, red spots that become fluid-filled blisters, eventually dry, scab, and drop-off, potentially leaving small pits in the skin. Babies with neonatal chicken pox are at increased risk for more serious complications such as hepatitis, pneumonia, and encephalitis.

Chicken pox vaccine became available in the mid-1980s in Japan, but it was not until 1989 that USA started using the vaccine. European countries were slow in adopting this vaccine—in fact, countries such as the UK were known and encouraging children to have “chicken pox” parties, so that children get exposed to chicken pox and get life long immunity after contracting the disease! The initial data of the chicken pox vaccination which became available after 10 years of regular use in USA were so encouraging that many other countries started recommending the vaccine to their population based on this experience. But soon, the studies revealed that the immunity from the vaccine is not lifelong and breakthrough infections were more common than it was previously thought. Hence, the current recommendation is for two doses, with the second dose being given or recommended after 3 months, preferably in the preschool age after the first dose having been given in the end of the first year. The most skeptic of all, the UK is finally convinced and will be introducing the vaccine soon in their national program. India is at crossroads and sooner we take action on this, we will save lot of morbidity and mortality at this preventable disease. Currently, the chicken pox vaccine is not part of Universal Immunisation Programme (UIP) and is an optional vaccine for the parents in India.

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