

# Varicella Syndrome

BIRTH DEFECT RESEARCH FOR CHILDREN



## What is Varicella?

Varicella, also known as Chickenpox, is a highly contagious viral infection caused by first exposure to the varicella zoster virus. The virus is transmitted by respiratory droplets and close personal contact. The most common symptom of Chickenpox is an itchy rash which first appears as small reddish spots. The appearance of the rash is sufficient to establish a diagnosis. New spots appear for up to 3-5 days, usually in small groupings first on the trunk and then spreading to the extremities, face, and scalp. The spots blister and eventually scab over. Chickenpox is contagious from 2 days prior to the appearance of the rash until all the spots are scabbed over (approximately 6 days after the appearance of the first spots). Fever and body aches may occur before the rash. The incubation period (time between exposure and the appearance of symptoms) ranges from 10 to 21 days. After infection, the virus remains dormant in the body and lifelong immunity is usually present.



# Varicella



What is a woman's risk of infection if exposed to Chickenpox during pregnancy?

A woman who previously had Chickenpox is immune and most likely will not develop Chickenpox if exposed again, and there is no risk to the unborn baby. If a woman is unsure whether she ever had Chickenpox, she should see a doctor for a blood test to see if she has any antibodies to the virus. If antibodies are present, she is immune to the infection. 80-95% of women who don't know or don't think they've had Chickenpox will show antibodies when tested. If the test does not show antibodies, she is not immune and is at risk of being infected.

What are the effects on a pregnant woman who develops Chickenpox?

Infection of a pregnant woman can result in Chickenpox pneumonia (10% chance) which can be very severe and sometimes life-threatening.

What are the effects on the fetus when a pregnant woman develops chickenpox?

The effects on the fetus depend on the timing of the illness. If the infection is within the first 20 weeks of pregnancy, the growth of various fetal organs may be affected. The greatest risk for birth defects is when a woman develops Chickenpox between 8 and 20 weeks of pregnancy. Up until 12 weeks the risk is about 0.4%; between 13 and 20 weeks it is 2%. The infection can cause a pattern of birth defects called Congenital Varicella Syndrome (also called Varicella Embryopathy and Fetal Varicella Syndrome). There is also a risk of pregnancy loss.

If a woman develops Chickenpox within 5 days before delivery or 2 days after delivery, there is a 20% chance the baby will develop Chickenpox. The baby receives the virus but not the ready-made maternal antibodies. Infection at this time is called Congenital Varicella and the baby will be present with symptoms between 5-10 days of age. Congenital Varicella can be very severe and the baby is more likely to develop serious complications such as pneumonia and an increased risk of

death. The mortality rate is 20-30%.

Maternal Chickenpox between 6 and 21 days before delivery could result in Congenital Varicella but it would probably be mild because the baby would receive some of the mother's newly-made Chickenpox antibodies.

Between 20 weeks and approximately 3 weeks prior to delivery, there is little risk to the baby from a maternal case of Chickenpox. However, the baby could develop herpes zoster (shingles) during infancy.

How many pregnancies are affected?

The incidence of Varicella in pregnancy is estimated to be about 5-10 cases per 10,000 pregnancies in the United States. It is estimated to be about 3 cases per 1,000 pregnancies in the United Kingdom.

How many babies are affected?

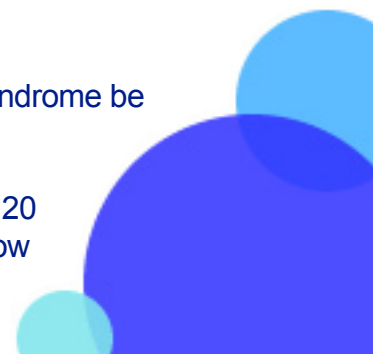
Most babies born to women who had Chickenpox in pregnancy are normal. 1-2% of babies whose mothers had Chickenpox in pregnancy have birth defects due to the mother's infection.

What is Congenital Varicella Syndrome?

Birth defects can be manifested in the baby's growth, skin, eyes, musculoskeletal system, and central nervous system. Abnormalities can include intrauterine growth retardation, low birth weight, cataracts, abnormally small sized eye(s), skin scarring, small head size, defective development of legs or arms, underdevelopment of fingers or toes, paralysis, psychomotor retardation, problems with the brain, seizures, and mental retardation.

Can Congenital Varicella Syndrome be diagnosed prenatally?

Ultrasound between 18 and 20 weeks of pregnancy can show limb defects and brain problems that may have been



# Varicella



caused by Chickenpox infection. Sometimes ultrasound can show liver problems, extra amniotic fluid, and severe swelling of the fetus that may have been caused by Chickenpox infection. Ultrasounds over a period of time may show growth problems related to the infection. However, ultrasounds cannot pick up all birth defects caused by Chickenpox in pregnancy.

What is the prognosis for a baby with Congenital Varicella Syndrome?

The prognosis depends on the type and extent of the abnormalities. Limited scarring abnormalities usually have good prognosis. Brain abnormalities usually have a less favorable prognosis. Severely affected infants usually do not survive.

What is the treatment for a pregnant woman exposed to Chickenpox?

If an “at-risk” pregnant woman (one with no antibodies) is exposed to Chickenpox, she should contact her doctor immediately. Man-made antibodies called Varicella-Zoster Immune Globulin (VZIG) can be given to the woman no later than 4 days after exposure to significantly reduce the severity of the disease. It is not known whether VZIG will prevent Chickenpox infection in the fetus. Some doctors use intravenous acyclovir therapy for pregnant women; however, acyclovir is classified as Category C in the Food and Drug Administration’s use-in-pregnancy rating. This classification indicates that risk cannot be ruled out, but potential benefits may justify the possible risk. Aspirin should not be used because its use during Chickenpox illness has been associated with a risk of developing Reye’s Syndrome. Depending on the woman’s condition, hospitalization may be necessary.

How can you help a newborn who has been exposed to Chickenpox prenatally?

Exposed infants are usually given VZIG to reduce the disease severity. If administered early enough, VZIG may modify the illness. Infants with moderate to severe cases of Chickenpox are sometimes treated with intravenous acyclovir.

How can you prevent Congenital Varicella Syndrome?

The primary method of prevention is through the establishment of immunity to the virus prior to conception. Vaccination against Chickenpox offers long-term protection against the disease. Because there is not enough information to determine the vaccine’s effect on a fetus, it should not be given to a woman during her pregnancy. The Varicella Vaccine in Pregnancy Registry has been established to help study the vaccine’s effect on the fetus. The vaccine maker recommends that women wait 3 months after vaccination before trying to conceive. A woman can be vaccinated immediately after delivering a baby.

Chickenpox is a highly contagious virus so susceptible pregnant women should avoid those infected. Individuals are most infectious before the rash develops when there is no reason to suspect that they have Chickenpox. 90% of susceptible individuals who come in close contact with a case can develop the infection. A woman can be given VZIG if she comes into contact with a case but it has to be given within 3-4 days of exposure, is costly, and offers protection for only a few weeks. Susceptible children living in a household with a susceptible pregnant woman can be vaccinated to protect her from the disease. Although there is a small risk that a vaccinated person with a weakened immune system could get a vaccine rash and spread the vaccine strain virus, this risk is much smaller than the risk that the pregnant woman could contract “natural” varicella.

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