

Explainer: what is microcephaly and what is its relationship to Zika virus?

Written by The Conversation

This week the [World Health Organisation](#) declared Zika virus a public health emergency of international concern.

Despite high rates of infection, the outbreak would not have been particularly alarming – since the infection is usually asymptomatic (80% of cases) or mild and self-limiting – had it not been for the sudden and (apparently associated) [increase](#) in numbers of infants born with microcephaly.

What is microcephaly?

Microcephaly is a condition in which the infant's head is smaller than “normal” for the infant's age and gender, because of delayed or arrested brain growth. There is no universally agreed definition. Most authorities suggest it should be defined by a head circumference of two – but some say three – standard deviations or more below the average.

It is often first diagnosed by ultrasound examination during pregnancy. The incidence of microcephaly – in the absence of Zika virus infection – is difficult to determine.

Apart from the lack of an agreed definition or definitive diagnostic test, there is probably significant [under-reporting](#) of the condition. State-based surveillance in the United States – where Zika virus is not endemic – [suggests it occurs](#) in between two and 12 infants per 10,000 live births. Rates vary from 0.5 to 19 in 10,000 live births in different states.

If the incidence were similar in Brazil, where about three million infants are born each year, this would represent 600-3,600 cases a year. This is more than estimates based on [recent review of birth certificates](#) – approximately 0.5 per 10,000 live births.

Some of the [approximately 4,000 cases](#) reported in Brazil during 2015 may have been due to increased awareness and reporting – although there appears to have been a real increase also.

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Microcephaly [is often associated with](#) other developmental abnormalities and with varying degrees of intellectual and developmental delay, seizures, and visual and hearing loss. In severe cases it can be life-threatening.

Causes

There are many recognised causes of microcephaly including a number of [other infections](#) in pregnant women. These include rubella, cytomegalovirus (a common virus that causes asymptomatic infection or a mild glandular fever-like illness in otherwise healthy people and severe disease in people with severe immune suppression such as AIDS), herpes simplex virus infections, syphilis and toxoplasmosis (a parasitic disease).

Chikungunya, a virus spread by the same mosquito responsible for spreading Zika (the *Aedes aegypti*, or yellow fever mosquito), [has also been shown](#) to cause brain damage in infants of women infected during pregnancy in a naïve population (one without previous exposure to the virus).

Noninfective causes of microcephaly [include](#) a variety of genetic disorders, maternal exposure to drugs, alcohol, chemical toxins and radiation and severe malnutrition.

Is Zika to blame?

Although Zika virus has not yet been definitively proven to be the cause of the increased numbers of infants with microcephaly in Brazil, there is strong circumstantial and epidemiological evidence that it is, at least partly, responsible.

Many of the mothers of affected babies in Brazil reported an illness consistent with Zika virus infection in early pregnancy. However, this was often mistaken for dengue and not confirmed by laboratory tests.

The [peak incidence](#) of microcephaly occurred in the same geographic region (northeastern Brazil) about a year after an outbreak of dengue-like illness, with fever and rash, started. Six months later Zika virus was identified as the cause.

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There have been [several reports of detection](#) of Zika virus genetic material (nucleic acid) in amniotic fluid, placentas, tissues of infants who have died with microcephaly and in live-born infants, with or without microcephaly, of mothers who have had Zika virus infection during pregnancy. It is highly likely that maternal Zika virus infection can damage the developing foetal brain. But the level of risk is unknown.

The other major uncertainty about Zika virus infection and microcephaly is the level of risk at different stages of pregnancy. Because the infection is so frequently asymptomatic or easily mistaken for other viral infections, the number of pregnant women infected and the stage of pregnancy at which infections occur are unknown.

For most intrauterine (within the uterus) infections that cause foetal damage (such as rubella or cytomegalovirus, for which these risks are well-known), the risk of the foetus being infected from the mother is relatively low in early pregnancy and increases with increasing gestation.

However, if foetal infection does, in fact, occur early in pregnancy, the foetus is more likely to be severely affected than if it occurs in the later stages of pregnancy. This is yet to be determined for Zika virus infection.

Hopefully, studies and enhanced surveillance of Zika virus infection and birth defects will provide answers to these questions. These are underway in Brazil.

In the meantime, while overall rates of Zika virus remain high, pregnant women are being advised to defer travel to Zika-affected countries if possible. Those who live there are advised to defer pregnancy or take extra precautions to avoid mosquito bites.

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