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The rare birth defect that's triggering panic over the Zika virus, explained

By Julia Belluz | @juliaoftoronto | julia.belluz@voxmedia.com | Jan 28, 2016, 2:40pm EST



A baby in Brazil has his head measured to test for microcephaly. | Mario Tama/Getty Images

Up until recently, **Zika** was considered a benign virus. It was discovered in Uganda in 1947 and for decades caused only small and sporadic outbreaks in far-off places like Micronesia. At worst, people would suffer from a rash and fever with some aches and pains. Most people never got any symptoms at all.

But when the mosquito-borne virus showed up in Brazil in 2015, everything changed.

Unlike in Africa, where **Zika** has been circulating for decades, the population in Brazil had never been exposed to the mosquito-borne virus and therefore had no immunity. Brazil is also home to many hot and densely packed cities, where the mosquito that carries that virus thrives. These conditions helped Zika **"spread explosively,"** as the World Health Organization put it. The virus is now expected to reach almost every country in the Americas (except for Canada and Chile).

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But here's the really worrisome part: Over the past year, pediatric neurologists in Brazil began to notice that some **pregnant women** infected with Zika have given birth to babies with a terrible birth defect called microcephaly, which is characterized by a shrunken head and incomplete brain development. (The virus has also been linked with Guillain-Barré, a rare neurological condition that can lead to paralysis.)

Researchers still don't fully understand the link between Zika and microcephaly, but this serious birth complication is a big part of why Zika is suddenly dominating headlines — and shaping up to be a **global public health emergency**.

1) Microcephaly means "small head" — and it's difficult to diagnose

Microcephaly, which comes from the Greek words for **"small head,"** is a broad term used to describe a smaller-than-average head circumference in babies (at least two standard deviations below the mean for his or her sex and age). It can be caused by a wide range of things, from diseases to malnutrition, as noted below.

According to the Centers for Disease and Control and Prevention, microcephaly is rare in the United States, ranging from two to 12 babies per 10,000 live births.





An X-ray comparing a normal baby skull to "Patient 1," who was born to a mother infected with Zika virus during her pregnancy. (**Latin American Science**)

It's also difficult to diagnose: The fact that a fetus has microcephaly is often not apparent until late in the pregnancy (ultrasounds often can't detect the condition until well into the second trimester) or after birth (when doctors measure the circumference of the baby's head). Sometimes babies born with normal heads can acquire microcephaly later, through a subsequent brain injury or infection.

The looseness of this condition, and the fact that some babies simply have small heads, means doctors look for potential underlying causes of microcephaly.

2) The condition can be caused by everything from viruses to malnutrition



A microcephaly baby born to a mother who had Zika in pregnancy. (AP Photo/Felipe Dana)

Microcephaly can be **caused by a wide range** of things — from chromosomal disorders such as Down syndrome to viruses like rubella or toxoplasmosis or even maternal alcoholism, drug abuse, diabetes, and malnutrition.

Now doctors think Zika may be another possible trigger. Since the virus arrived in Brazil in the spring of 2015, more than 4,000 cases of microcephaly have been reported — a twentyfold increase from previous years. (These cases still need to be confirmed by the health ministry.)

The fact that Zika has the potential to cause brain injury in fetuses is what has the public health community very concerned.

There are still many questions around exactly how Zika affects fetuses, explained Dr. **Jeanne Sheffield**, the director of maternal-fetal medicine at Johns Hopkins. "Right now we still don't know the true causal link, just that there's an association. And the science behind that association is still being worked out."

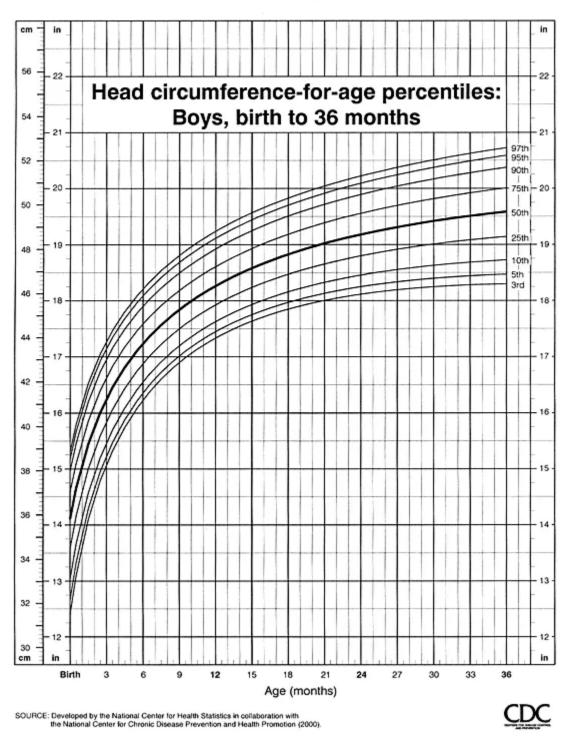
That leaves plenty of unknowns: It's not clear how many women who contract Zika give birth to babies with microcephaly. It's not clear *when* women are most at risk for passing on an infection (first trimester? second trimester?). It's also not clear whether women who get the virus but don't show symptoms pass the virus to their baby.

Right now, officials at the Centers for Disease Control and Prevention (CDC) say that birth defects seem to be more common in women who are infected earlier in their pregnancy and show symptoms of the virus. But that may not be a complete picture of the risks involved, and they've emphasized that there's still a lot we don't know.

Health researchers in Brazil and at the CDC are doing a careful review of information on the babies recognized with birth defects to make sure the diagnoses and links with Zika were accurate. They're also following pregnant women currently infected with the disease to see what happens to their babies and exploring other environmental factors that may have contributed to the birth defects.

3) Microcephaly is a spectrum — and Zika is associated with severe cases

CDC Growth Charts: United States



Head circumference-for-age percentiles, boys birth to 36 months. (CDC)

Microcephaly is a spectrum and can range from mild to severe, mostly depending on the cause. In borderline cases, the head is small but the baby has few long-term problems and normal intelligence. (In about 15 percent of cases, a child with microcephaly will have normal intelligence.)

In severe microcephaly, the head is extremely small because the brain has essentially been destroyed by the virus or underlying cause, and the baby's long-term health outcomes are very poor.

"A lot of the Zika virus cases reported so far are truly severe microcephaly," says Sheffield. "We are concerned that these cases are going to be more severe than the mild microcephaly cases we've seen before."

4) There's no cure for microcephaly

According to **Boston Children's Hospital**, some children with microcephaly have normal intelligence "and experience no particular difficulty with schoolwork, physical activity, relationships or any other aspect of their lives."

But other children with the condition — particularly severe cases — can face learning disabilities, impaired motor functions, difficulty moving their bodies, and even speech delays. Severe cases are also associated with facial deformities, seizures, and very short stature and dwarfism.

There's no treatment or cure for microcephaly. But depending on the severity of the condition, physical therapy (to improve strength and motor function) and speech and occupational therapy can help.

5) If you're pregnant or thinking of getting pregnant, avoid traveling to places where the virus is circulating



Countries with active transmission. (CDC)

Because of the **potential for birth defects**, the **CDC issued travel guidance** for pregnant American women and women of childbearing age who may become pregnant, warning them to avoid visiting places where the virus is currently circulating.

Right now the list includes Barbados, Bolivia, Brazil, Colombia, Dominican Republic, Ecuador, El Salvador, French Guiana, Guadeloupe, Guatemala, Guyana, Haiti, Honduras, Martinique, Mexico, Panama, Paraguay, Puerto Rico, Saint Martin, Suriname, and Venezuela — and officials warn that more countries will be added soon. (You can find **updates here**.)

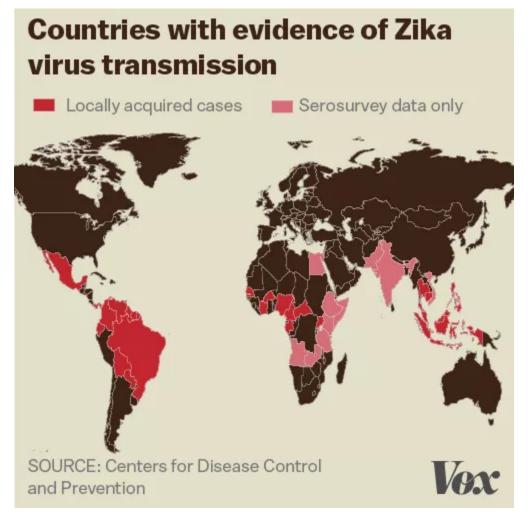
There's no vaccine or medicine to prevent Zika, so simply avoiding mosquitoes in countries where the virus has been circulating is the best defense. The **CDC also issued guidance** on how to care for pregnant women during a Zika outbreak. (Read **more on that here**.)

In addition, health officials recommend *everyone* traveling to or living in areas with Zika virus take precautions to protect themselves — wearing mosquito repellent, using screens to keep mosquitoes outside, wearing long pants and long-sleeved shirts, and making sure there's no standing water inside or outside the home.

6) If you're pregnant and traveled in a Zika-infected country, talk to your doctor

In general, the CDC advises pregnant women who have visited countries where the virus is circulating consult a doctor. Those who had symptoms of Zika should be tested for the virus (a controversial suggestion, since many people never experience any symptoms), and all pregnant women who traveled in Zika-infected regions should also seek out ultrasound scans to make sure their fetuses are developing normally. Unfortunately, however, microcephaly does now show up in scans until later in the pregnancy (just before the third trimester).

7) The link with brain injury has been found in reexaminations of past Zika outbreaks



Javior Zarraoina/Matt Moore

Since microcephaly started showing up in large numbers in Brazil, researchers have been looking at data from past Zika outbreaks to see if this had previously been an issue that no one had noticed.

Signs point to yes. During a Zika outbreak in French Polynesia in 2013 to 2014, there were **12 identified cases of microcephaly**. Previous Zika outbreaks have been much smaller than Brazil's, and have happened in places that don't always have great disease surveillance systems, which may explain why this wasn't noticed until now.

8) Expect this information to evolve in the coming months

Information about Zika and its link with birth defects and other health conditions is going to evolve and change in the coming weeks and months as researchers learn more.

We might learn that Brazil seriously overestimated the burden of microcephaly, and that it's an extremely rare complication. We might learn microcephaly was actually triggered by

an interaction with another virus or some unaccounted-for environmental factor. We might learn that Zika causes other severe birth problems and side effects. (The virus has been found in babies who died within 24 hours of birth.)

Health authorities have already noted an increase in Guillain-Barré associated with Zika. This is a neurological syndrome where a person's own immune system damages his nerve cells, leading to muscle weakness or paralysis. The symptoms can last weeks or months, but most people recover fully — though it can take years to do so.

Again, though, researchers are studying this link, and a direct causal relationship has not yet been established. So expect the information about Zika to evolve as scientists scramble to understand this new virus.

Correction: A previous version of this article misstated the number of microcephaly cases in French Polynesia.