

Surveillance Highlights

Small bite, big problem: Understanding severe microcephaly in Canada

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CLINICAL VIGNETTE

A 6-week-old male infant is brought to clinic for a checkup. He has severe microcephaly. On physical exam, the infant is afebrile and has normal vital signs. Head circumference is below the first percentile. Weight and height are at the 50th percentile. He is alert though irritable and difficult to console. Examination of the eyes demonstrated that the pupils react normally, red reflexes are present, there are no opacities, and extraocular movements are normal. He transiently fixes on faces. Facial movements are normal. There is no drooling. Hearing is grossly intact. Motor examination reveals diminished axial tone with increased tone in the four extremities and a paucity of spontaneous movement in all four limbs. Muscle stretch reflexes are increased throughout the four extremities with spread and sustained clonus at the ankles. He withdraws the four limbs briskly to light touch. The atonic neck reflex is not elicited. Seizures were not observed during the examination.

Both parents went on a hiking trip in Northeastern Brazil for 2 weeks. The child's mother recalls having a rash and some muscle pain during her trip. The rash was not bothersome, and went away after a few days. Approximately 2 weeks after returning to Canada, she discovered that she was pregnant. Ultrasound findings showed intracranial calcifications at the 20-week scan. The pregnancy was otherwise uneventful. Routine antenatal serology was normal. There were no chemical or toxic exposures in utero, and his mother did not consume alcohol, smoke or use drugs around time of conception or during pregnancy. There is no positive family history of microcephaly.

As a result of the clinical presentation and travel history of the mother, serum samples were collected from both mother and infant for testing. Plaque Reduction Neutralization Test serology confirmed the presence of Zika virus-specific antibodies in both the mother and infant's serum.

LESSONS LEARNED

- The epidemiology of severe microcephaly in Canada is not well known. In response to the emerging Zika virus public health threat, the Canadian Paediatric Surveillance Program (CPSP) launched a multi-year study in June 2016 to gain a better understanding of the epidemiology and etiology of severe microcephaly in Canada (1).
- In the first 6 months of the CPSP severe microcephaly study, 24 possible cases were identified. Of those, 17 cases were confirmed with severe microcephaly and questionnaires were completed by the reporting physicians. To date, no Zika-associated severe microcephaly cases have been identified by the study.
- Severe microcephaly is often detected prenatally; however, it may also be detected at birth or in early infancy. Severe microcephaly is a more extreme form of microcephaly, where a baby's head is much smaller than expected (less than three standard deviations below the expected mean for gestational age and sex). Children born with severe microcephaly can experience a range of complications including: seizures, developmental delay, intellectual disability, hearing loss and/or vision problems (2).

- There is no single cause of microcephaly and causality is often not readily discernible.
- Infants and children can acquire Zika virus infection congenitally or postnatally (3). The impact of postnatal infection on the developing central nervous system remains unclear. Zika virus testing is recommended for infants born to women with laboratory evidence of confirmed or probable Zika virus infection regardless of the presence or absence of phenotypic abnormalities (4).
- Infants should be tested for Zika virus infection when born to women with confirmed or suspected Zika virus infection in pregnancy, or if they have unexplained microcephaly, intracranial calcifications, ventriculomegaly or major structural central nervous system abnormalities or other symptoms of congenital Zika virus infection in whom the mother had potential exposure to the virus. This testing should include serology (IgM and PRNT) and polymerase chain reaction (PCR) of serum (umbilical cord or infant sample) and placental tissue. If cerebrospinal fluid is sampled, it can also be sent for PCR and serology (5).
- If an infant has microcephaly, arrange for MRI of the head, as well as audiologic and ophthalmologic assessments.
- All physicians should inquire about possible etiology, including details of travel history that include countries visited, and dates of departure and return.
- If a child is seen in clinic with severe microcephaly, a CPSP severe microcephaly study questionnaire should be completed.
- In January 2017, the CPSP launched a congenital Zika syndrome study (6). Physicians who see a case of severe microcephaly suspected to be associated with Zika virus should complete a questionnaire for the severe microcephaly study AND the congenital Zika syndrome study.
- For more information, please reference the Canadian Paediatric Society Practice Point entitled "Zika Virus: What does a physician caring for children in Canada need to know?" (7).

References

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