

Single ventricle physiology in a pediatric patient with maternal Zika virus exposure: A case report and literature review regarding the anesthetic considerations of newborns and infants with congenital Zika syndrome

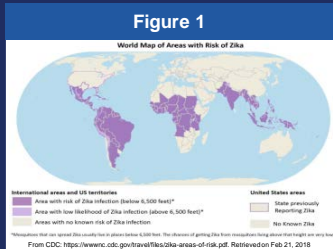
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Background

- Zika virus was first reported in 1947
- Named after the Zika forest in Uganda
- Arbovirus that belongs to the *Flaviviridae* family
- 1952 - First human cases of Zika virus, remained endemic in tropical areas of Africa, Southeast Asia, and the Pacific Islands
- 2015 - Global pandemic since its identification in Brazil leading the World Health Organization (WHO) to declare a Public Health Emergency of International Concern (PHEIC) on February 1, 2016
- Spread via vector borne (*Aedes* species mosquito), congenital, and sexual transmission^{1,2,3,4}
- All pregnant women who have traveled to or live in endemic areas are recommended to undergo Zika serology testing and serial fetal ultrasound screenings.⁵



Case Presentation

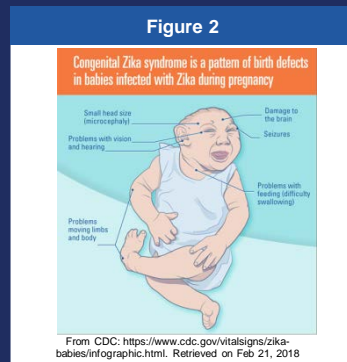
- 10 month old male with single ventricle physiology from a large ventricular septal defect, subaortic stenosis, hypoplastic ascending aorta and transverse aortic arch, bidirectional patent ductus arteriosus, head sparing failure-to-thrive, and heart failure requiring diuretics.
- His mother had serologically confirmed Zika virus infection at 20+ weeks gestation. Exact time of viral inoculation is unknown.
- Our patient did not exhibit the usual constellation of birth defects seen with congenital Zika syndrome (CZS) though he was never tested for the virus.
- Due to maternal Zika virus exposure, the patient underwent extra fetal scans and an echocardiogram after birth which demonstrated his congenital heart disease (CHD). No family history of CHD was reported.
- He underwent diagnostic cardiac catheterization and initial repair of his CHD with a **Norwood procedure and use of a modified Blalock-Taussig (BT) shunt.**
- The Norwood procedure was ultimately completed successfully. However, when initially coming off of bypass in the operating suite, the patient had ST segment elevation with multiple episodes of ventricular fibrillation that required internal defibrillation prior to the return of normal sinus rhythm. He was then noted to be hypoxemic and went back on bypass for revision of the BT shunt.
- His postoperative course was complicated by refractory hypoxemia and persistent need for inhaled nitric oxide, but extubation was eventually achieved.

Case Discussion

- Though the full range of congenital defects related to Zika virus exposure is unknown, the currently accepted phenotype of CZS includes microcephaly, CNS and eye abnormalities, neural tube defects, joint contractures, and sensorineural deafness.⁶
- Maternal Zika exposure does not always result in CZS, but CHD may be seen.⁷
- In a study of 103 infants in Brazil with presumed congenital Zika syndrome (microcephaly and head CT findings consistent with congenital Zika syndrome, serologic diagnosis not available for all infants), 14 transthoracic echocardiograms performed by an experienced pediatric cardiologist demonstrated congenital heart disease including atrial septal defects (ASDs) and ventricular septal defects (VSDs). Infants with persistent foramen ovale (PFO) or minimum patent ductus arteriosus (PDA) were considered normal. They concluded that "infants with presumed congenital Zika syndrome were almost three times the expected rate of CHD in a general population less than a year of age, at 135 per 1000 live births".⁷ However, this study is the first to assess for congenital heart defects in infants with presumed congenital Zika syndrome and further studies are warranted to corroborate the results.
- Many questions remain regarding pregnancy and Zika virus exposure. These questions require further study to fully understand the complete spectrum of birth defects that may be directly linked to virus exposure and the risk to mother and fetus with virus exposure during different stages of pregnancy and fetal development.²

Perianesthetic Considerations when treating CZS patients⁸

Concern	Notes	What to do about it?
Blood-borne pathogen	Viremia high in newborns Virus transmission can occur through blood transfusions	Use of Personal Protective Equipment (PPE) including splash guards and eye shields Safe needle handling practices Proper hand hygiene
Micrognathia and craniocervical biomechanical limitations	Could make mask ventilation, direct laryngoscopy, and even placement of a laryngeal mask airway challenging	Availability of video laryngoscopy and other advanced airway techniques are recommended during induction of anesthesia or during neonatal resuscitation
Arthrogryposis	Congenital joint contractures in two or more areas of the body may present challenges regarding positioning and IV placement	Alternative positioning methods or vein finding devices may be needed
Craniosynostosis	Skull suture premature fusion	Medications that have the potential to increase intracranial pressure should be avoided or used with caution
Epileptic activity	Cautious use of medications that lower the seizure threshold is recommended	Cognizant use of medications that have the potential to lower the seizure threshold including some antibiotics and opioid pain medications



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