Chiari Malformation in Pregnancy

SHERYL E. PARFITT
CHERYL K. ROTH

You've just arrived in the labor and birth unit to receive report on a 20-year-old woman, gravida 1 para 0, who presented to her obstetrician's office at approximately 15 weeks gestation for her first prenatal visit. At that time she stated that she had experienced headaches over the past year, but they were now increasing in frequency and severity. She also mentioned that her neck was beginning to hurt. The physician instructed her to take acetaminophen or ibuprofen for pain and to try to rest more in the evening after work. Within 2 weeks, she again presented to the physician stating that she was now experiencing muscle weakness in her extremities. Upon exam, the woman was noted to have motor weakness in the upper limbs and a slightly irregular gait. She was referred to a neurologist for consultation.

Magnetic resonance imaging (MRI) of her brain showed a 6 mm downward displacement of the cerebellar tonsils through the foramen magnum into the upper cervical canal. The presence of a syrinx, a fluid-filled cyst in

Abstract The diagnosis of Chiari malformation is on the rise owing to the increased frequency of brain imaging for concussion and trauma. This abnormal brain physiology can have a significant impact on the care management of a pregnant woman during the gestational period. Here we present a case example of a pregnant woman presenting in labor with a history of Chiari malformation with surgical treatment during her pregnancy. Antepartum, intrapartum and postpartum considerations are reviewed. This brief article is meant to be used as an easy-reference tool in the clinical setting. DOI: 10.1111/1751-486X.12189

Keywords anesthesia | Chiari malformation | intrapartum | pregnancy

the spinal cord, was also noted. A diagnosis of Chiari malformation, Type 1, was made. The woman was advised to undergo decompression surgery at that time, but declined due to her pregnancy.

Chiari malformations involve herniation or hypoplasia of the cerebellum. Thus, symptoms that pregnant women present with may involve some degree of difficulty with balance, coordination, muscle weakness and speech

Eight weeks later, she presented to the neurologist for a follow-up visit. It was noted that motor weakness in both her upper and lower extremities was progressively worsening. The woman agreed to surgical intervention at that time and decompression of the posterior fossa was performed under general anesthesia without any complications. While her symptoms didn't completely disappear, they subsided greatly following the surgical intervention.

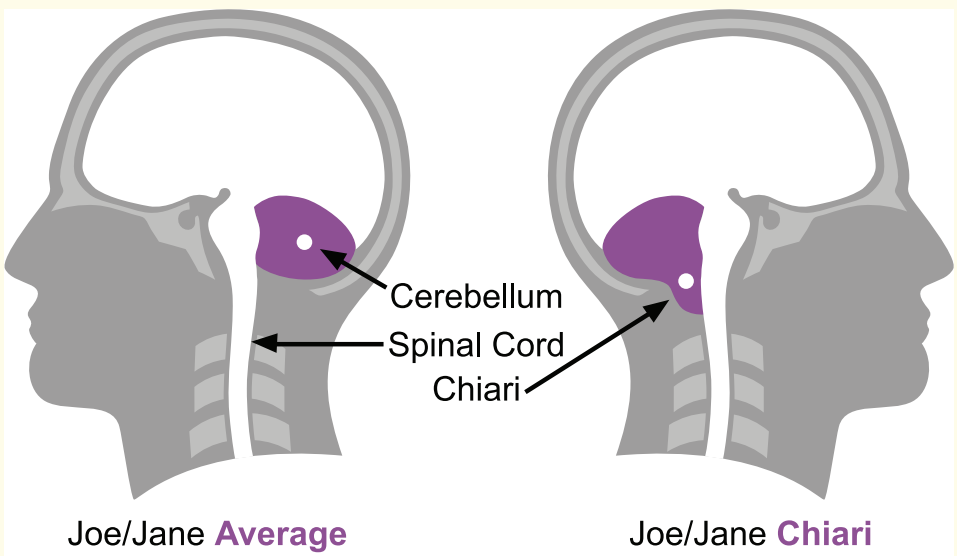
She is now 39.5 weeks gestation in active labor and is requesting an epidural.

What Is Chiari Malformation?

Chiari malformations, also known as Arnold-Chiari malformations, occur when a fetus is developing and are defined by anomalies of the craniocervical junction. In particular, the posterior fossa is abnormally underdeveloped causing downward displacement of brain tissue (see Figure 1). The pathogenesis is the subject of much debate with many proposed theories. A molecular genetic theory asserts that genetic programing of the hindbrain is defective, leading to growth abnormalities. The crowding theory suggests that restricted growth of the posterior fossa compresses neural tissue, which in turn gets pushed through the foramen magnum. Other suggested theories include downward pressure on the brainstem and cerebellum from progressive fetal hydrocephalus, and defective closure of the neural tube causing cerebrospinal fluid leak, which may result in a small posterior fossa (Khoury, 2013).

The malformation causes areas surrounding the cerebellum to be underdeveloped or deformed, displacing the cerebellar tonsils downward by at least 3 mm beneath the foramen magnum and into the cervical spinal canal. This defect can potentially block normal flow of cerebral spinal fluid between the intracranial space

Figure 1.
Chiari Malformation



Reprinted with permission of Conquer Chiari.

Sheryl E. Parfitt, MSN, RNC-OB, is a clinical educator; Cheryl K. Roth, PhD, WHNP-BC, RNC-OB, RNFA, is a nurse practitioner; both authors are at Scottsdale Healthcare in Scottsdale, AZ. The authors report no conflicts of interest or relevant financial relationships. Address correspondence to: croth@shc.org.