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Chiari Malformations

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Underlying Pathophysiology

“Chiari malformations are caused by insufficient development of the posterior fossa of the skull” (Elam & Vaughn, 2011, p. 758). They may be a congenital or acquired anomaly. The malformations are imperfections in the structural makeup of the posterior fossa which then causes a portion of the cerebellum to extrude inferiorly through the foramen magnum (Elam & Vaughn, 2011, p. 758). There is speculation that (congenital) Chiari malformations are possibly due to a mesodermal defect or that the imperfection sparks from the neural crest (Labuda, Loth, & Slavin, 2011, p. 227). An acquired Chiari I malformation is seen when cerebellar tonsillar herniation is secondary to downward displacement by some force from above (Graham, Davis, Gouvernayre, & Thomas, 2012, p. 1002). An acquired Chiari malformation can occur from things such as: minor trauma, hydrocephalus, shunt failure, or neoplasms- supratentorial or infratentorial tumors (Graham et al., 2012, p. 1002).



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Classification

They are classified by degree of severity and the specific anatomy involved as follows:

CM-I is a result of the cerebellar tonsils extending through the foramen magnum (Klein, Hopewell, & Oien, 2014).

CM-II is “herniation of both cerebellum and lower brainstem” (Ramón, Gonzáles-Mandly, & Pascual, 2011, p. 157).

CM-III is a “rare type of brainstem herniation in association with a cervical or occipital encephalocele” (Ramón et al., 2011, p. 158).

CM-IV “involves extreme cerebellar hypoplasia and caudal displacement of the posterior fossa contents” (Ramón et al., 2011, p. 158).

“**Chiari type 0** is defined as syringohydromyelia with distortion of contents in posterior fossa without cerebellar tonsillar herniation” (Ramón et al., 2011, p. 158).

Chiari type 1.5 includes caudal migration of the brainstem and cerebellar tonsils often associated with syringomyelia but without spinabifida” (Ramón et al., 2011, p. 158).

Significance of Pathophysiology

“Chiari malformations may result in downward pressure on the cerebellum or brainstem, with subsequent blockade of cerebrospinal fluid flow” (Graham et al., 2012, p. 1000). Cerebellar and brainstem dysfunction. Development of syringomyelia in 15–65% of the cases, which is characterized by a fluid filled cyst, or syrinx, in the spinal cord (Labuda et al., 2011, p. 228). Development of hydrocephalus and/or central nervous system lesions (Graham et al., 2012, p. 1003). CM that shows positive progression may severely impair central nervous system functioning (NIH, 2013, para. 28).

Signs & Symptoms

Adults/Children

neck pain
hearing or balance problems
muscle weakness or numbness
dizziness
difficulty swallowing or speaking
vomiting
ringing or buzzing in the ears (tinnitus)
curvature of the spine (scoliosis)
insomnia
depression
problems with hand coordination and fine motor skills” (NIH, 2013, para. 3)

Additional Signs and symptoms:

severely limited extension of the cervical spine (Robertson & Stanley, 2012, p. 483)
debilitating headaches (Fischbein et al., 2015, p. 1617)

Signs and Symptoms- Infants

difficulty swallowing
irritability when being fed
excessive drooling
a weak cry
gagging or vomiting
arm weakness
a stiff neck
breathing problems
developmental delays
an inability to gain weight (NIH, 2013, para. 4)

Introduction

The well prepared advanced practice nurse will have a solid understanding of anatomy and physiology, as well as pathophysiological principles. The APN that practices in the emergency department or in the acute care setting should be familiar with a variety of common neurological disorders. The National Institute of Neurological Disorders and Stroke (NIH) estimate that greater than 1 out of every 1000 persons will be identified with a Chiari malformation (CM) and the greater majority diagnosed will be female (National Institute of Neurological Disorders and Stroke [NIH], 2013, para. 6). Chiari malformations can be congenital or acquired and are staged as I, II, III, IV, 0 or 1.5. Stage I is the most common and it is the only type that can be acquired. Not all CMs require treatment. There are CMs that only require monitoring as they are asymptomatic; however, other CMs can cause severe impairment and require surgical intervention. Effective management of chronic conditions, such as CM, can have profound effects on a variety of factors such as patient morbidity, mortality rates and health care utilization. For a nurse with a strong background in neurological intensive care, the study of neurological conditions is an especially significant area of interest.



FIGURE 1. Sagittal T1-weighted magnetic resonance image of the cervical spine demonstrating protrusion of the cerebellar tonsils (orange arrow) below the foramen magnum (black arrow), consistent with a type I Chiari malformation.

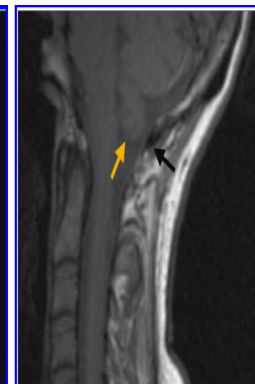


FIGURE 2. Sagittal T1-weighted magnetic resonance image of the cervical spine demonstrating normal positioning of the cerebellum (orange arrow) in relation to the foramen magnum (black arrow) in an unrelated patient. Reproduced with permission from Walk.

From: <http://dx.doi.org/10.2519/jospt.2008.00410>

Implications for Nursing Care

The greater the severity of the Chiari malformation the greater the impact will be on a patient’s activities of daily living and instrumental activities of daily living. One study found that as many as 96.1% of study participants reported “at least one physical activity they can no longer engage in or enjoy due to CM symptoms” (Meeker, Amerine, Kropp, Chyatte, & Fischbein, 2015, p. 524).

The following considerations for efficacious advanced practice nursing (APN) care will be imperative for early diagnosis and impactful treatment and care of the patient with CM:

- Detailed assessment to assist with early detection and cause analysis (if applicable)
- Ordering an MRI for suspected CM
- Pain management
- Provide education to patient and family
- Initiate an interdisciplinary care team to ensure holistic approach to meeting a patient’s needs (neurologist, neurosurgeon, physical/occupational therapy, speech pathologist, home health, community services, etc.)

It is also the responsibility of the APN to consider alternative therapies that may be in the best interest of the patient. Headaches and dizziness stemming from a CM-I malformation were completely resolved within three chiropractic visits in one case (Sergent & Cofano, 2014).

Conclusion

In order to effectively diagnosis and treat Chiari malformations it is imperative for advanced licensed professionals to understand exactly what a Chiari malformation is. A thorough understanding of the pathophysiological process as well as the signs and symptoms to look for will be invaluable. This knowledge will guide the provider in making the correct diagnostic and treatment decisions for the patient. For the APN, it is equally important to follow the medical diagnosis with application of the nursing process to address all patient’s needs. This will ensure better patient outcomes and higher levels of satisfaction.

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