Chiari Malformation

Background

Chiari malformations (CM) are a group of complex neurological disorders affecting the structure of the base of the skull and the cerebellum. A CM occurs when part of the cerebellum, the part of the brain controlling balance, extends below the opening in the skull and into the spinal canal. Structural defects can occur, causing complex brain abnormalities and disrupting the flow of the cerebrospinal fluid (CSF).

The exact cause of CM is unknown although research does support abnormal brain formation during fetal development. Genetics has also been considered as malformations are known to have familial links. CM is usually present at birth, but a diagnosis can be made later in life due to an acquired injury, disease, or infection.

There are several types of Chiari malformations classified by their severity and the part of the brain protruding into the spinal canal.

- Type 1 is the most common form that develops as the brain and skull are growing. Type 1 is usually asymptomatic and can go undetected until adolescence or adulthood.
- Type II and Type III are pediatric forms of CM present at birth. In Type II, tissue from both the cerebellum and brainstem protrudes into the foramen magnum, leading to potentially life-threatening complications in infancy. Symptoms are generally more severe, requiring early surgical intervention. Type II is usually accompanied by a myelomeningocele, a form of spina bifida that occurs when the spinal canal and backbone do not close before birth. The term Arnold-Chiari malformation is specific to Type II malformations. Type III is the most severe with life-threatening complications and neurological impairment.

Symptoms will vary from child to child depending on the degree of structural defect, compression of nerves, and disruption of the flow of CSF. The most common symptom is headaches near the base of the skull. Severe pain may radiate to the neck and shoulders and has been reported to worsen with coughing, sneezing, or straining.

Sensory symptoms affecting hearing and vision include hearing loss, ringing in ear, and blurred or double vision. Other physical symptoms include muscle weakness, numbness, pain, and difficulties with both gross and fine motor skills affecting balance and coordination.

In young children, symptoms more commonly include reflux, chronic cough, difficulty swallowing, poor weight gain and sleep apnea. For infants and younger children unable to verbalize their symptoms, crying and irritability is common.

Children with CM may also suffer from comorbid conditions including spina bifida, hydrocephalus, tethered cord syndrome and scoliosis. Surgical decompression or placement of a shunt may be considered to alleviate progression of nerve damage and improve symptoms.

Top Takeaways for School

Chiari malformations (CM) are abnormal brain structures affecting the posterior skull where the brain and spinal cord connect.

The most common presenting symptom in children is pain. It is often reported as a headache or neck and shoulder pain.

CM can have both a physical and cognitive impact.

Children can experience difficulties with auditory processing, sensory integration, and visual perception that create learning challenges.

Children who have Type II-Arnold-Chiari malformation most often experience learning difficulties in the classroom related to language, concentration, and organization.

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnoses: Risk for disturbed sensory perception, risk for injury, impaired physical mobility, pain (acute/chronic), imbalanced nutrition: less than body requirements and impaired thought process
- Assessment of implanted medical device (consider location, date of surgical placement, and device-specific information)
- Use of specialized equipment, adaptive equipment, and orthotics
- Activity and positioning precautions and/or restrictions
- Equipment troubleshooting (consider equipment/device user manual, battery, charger)
- Consider emergency action plans (EAPs) and emergency evacuation plans (EEPs) related to special health care needs, including staff education/training

Discussion Starters for the Educational Team

1. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
2. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
3. Can strategies be implemented to assist the student with executive function (e.g., plan, prompts, organizers, agendas)?
4. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?
5. Will staff receive education/training to implement the student-specific emergency plan?

Resources

Kennedy Krieger Institute: Center for Spina Bifida and Related Disorders
kennedykrieger.org/patient-care/centers-and-programs/center-for-spina-bifida-and-related-conditions

Conquer Chiari
conquerchiari.org/index.asp

For more information, please scan the QR code or visit: KennedyKrieger.org/SHNIC

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