

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 (part of the brain controlling balance). A CM occurs when part of the cerebellum extends below the opening in the skull called the foramen magnum and into the spinal canal. Structural defects can occur when part of the skull is smaller or misshapen, 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 malformation 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, both the cerebellum and brain stem tissue protrude into the foramen magnum causing cause life-threatening complications during 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 CSF flow. Some symptoms may begin during infancy while others present as the child grows and develops. The most common symptom is headaches near the base of the skull. Severe pain may radiate to the neck and shoulders and have 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 can be considered to alleviate progression of nerve damage and improve symptoms.

Top Takeaways for School Considerations

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

CM can have both a physical and cognitive impact.

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

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.

Kennedy Krieger Institute's Specialized Health Needs Interagency Collaboration

The Specialized Health Needs Interagency Collaboration (SHNIC) program is a collaborative partnership between Kennedy Krieger Institute and the Maryland State Department of Education.



Kennedy Krieger Institute

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of risk for disturbed sensory perception, risk for injury, impaired physical mobility, pain (acute/chronic), imbalanced nutrition: less than body requirements and impaired thought process
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- 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 care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Has the school staff been trained to implement the student-specific emergency plan?
2. 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?
3. 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)?
4. Can strategies be implemented to assist the student with executive function (e.g., plan, prompts, organizers, agendas)?
5. 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)?

Resources

Kennedy Krieger Institute: Center for Spina Bifida and Related Conditions
kennedykrieger.org

Conquer Chiari
conquerchiari.org/index.asp

Chiari Medicine: Chiari to School
chiarimedicine.com/workflow



Scan QR code or visit **KennedyKrieger.org/Redirect** for more information.