Chiari malformations (CM) are a group of complex neurological disorders where the cerebellum descends beyond the base of the skull through an opening called the foramen magnum and into the spinal canal. It occurs when part of the skull is abnormally small or misshapen, pressing on the brain and forcing it downward. The structural defect can cause complex brain abnormalities and disrupt the flow of the cerebrospinal fluid (CSF).

The exact cause of CM is unknown although in children it is believed to be abnormal brain formation during fetal development. It has been known to run in families so there is belief that there could be a genetic cause. CM is usually present at birth but in rare cases CM can be acquired later in life due to traumatic injury, disease, or infection. Any condition that takes up space within the skull can cause a CM including tumors, arachnoid cysts and hematomas. Children with CM may also suffer from comorbid conditions including spina bifida, hydrocephalus, and tethered cord syndrome.

Chiari malformations are classified by their severity and the part of the brain protruding into the spinal canal. The 4 types of Chiari malformations include:

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>The most common form of CM. The base of the skull and upper spinal area are not properly formed, causing the lower part of the cerebellum to extend into the foramen magnum. Type 1 may not cause symptoms and can go undetected until adolescence or adulthood.</td>
</tr>
<tr>
<td>Type II</td>
<td>Both the cerebellum and brain stem tissue protrude into the foramen magnum. Symptoms are generally more severe and appear during childhood. Type II can cause life-threatening complications during infancy or early childhood and treating it requires surgery. 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.</td>
</tr>
<tr>
<td>Type III</td>
<td>Vary rare but most serious form causing life-threatening complications. Some of the cerebellum and the brain stem herniate through an abnormal opening in the back of the skull.</td>
</tr>
<tr>
<td>Type IV</td>
<td>Incomplete or underdeveloped cerebellum. Type IV is usually fatal during infancy.</td>
</tr>
</tbody>
</table>

Similar to most disorders, symptoms will vary from child to child. Symptoms may begin during infancy but can also be delayed and not present until adolescence or adulthood. Some of the most common symptoms include:

- Headache, most common symptom
- Hearing problems, loss, ringing in the ears
- Vision problems including blurred or double
- Difficulty swallowing or speaking
- Insomnia, depression
- Spinal curvature
- Neck and shoulder pain
- Muscle weakness or numbness
- Difficulty with balance and coordination
- Problems with fine motor skills

For infants and children who cannot verbalize their symptoms, crying and irritability is common. In children under age 3, symptoms can include aspiration, choking, regurgitation, dysphagia, chronic cough, sleep apnea, reflux, gagging, trouble swallowing and poor weight gain.
What is the treatment?

Treatment for CM is directed towards the child’s specific symptoms. Treatment teams usually consist of pediatricians, neurosurgeons, neurologists, eye specialists and other healthcare professionals. Some children with CM will need surgery. A decompression surgery is usually performed during which more space is created around the cerebellar tonsils and restoring normal flow of CSF is the goal. Sometimes a shunt (insert link to shunt devices page) is placed to relieve the pressure of cerebral spinal fluid. A team of medical and developmental specialists are key to the best possible outcome.

Suggested school accommodations

Supporting students with CM in the school setting requires the educators and parents/guardians to work as a team. A student with CM may experience difficulties with attention, motor coordination, and time management. A student with CM may require additional cognitive, behavioral, and emotional support. A well coordinated plan promotes success for the student in the educational setting. Some accommodations to consider for an 504/IEP might include:

- Early intervention assessment for young children
- PT/OT/SLP/Vision specialist consults
- Emotional support
- Staff education and training as appropriate
- Offer clear, concise direction
- Break down complex tasks
- Allowing extended time for a response
- Consider assistive technology
- Extended time for testing
- Accommodations if student is having surgery
- Extra time for assignments as needed
- Offer emotional support
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including all affected systems and symptoms and any other associated conditions
- Communicate with school staff, parents, and provider any changes or concerns about the disease
- Current medication list for home and school
- Orders from provider for any physical activity or positioning restrictions
- Child specific signs and symptoms of increased intracranial pressure
- Orders for emergency medications, when to administer, dose, route
- PT/OT/SLP/Vision services and hearing specialist assessment
- Orders for bowel and bladder program, if applicable
- Plan for monitoring neurological changes, headaches, anxiety
- Plan/protocol for chronic pain
- Provide rest area if needed
- Medical device information (see SHNIC’s “Medical Device Information Guide”)
- Emergency Care Plan (ECP) related to medical needs in the school setting and staff training as appropriate for each

Resources & Manuals

American Association of Neurological Surgeons– Chiari Malformation

Children’s National– Pediatric Chiari Malformation
https://childrensnational.org/visit/conditions-and-treatments/brain--nervous-system/chiari-malformations

Conquer Chiari
https://conquerchiari.org/resources/education-sheets.asp

National Organization for Rare Disorders (NORD)–Chiari Malformations
https://rarediseases.org/rare-diseases/chiari-malformations/

NIH National Institute of Neurological Disorders and Stroke-Chiari Malformation Factsheet
https://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Chiari-Malformation-Fact-Sheet