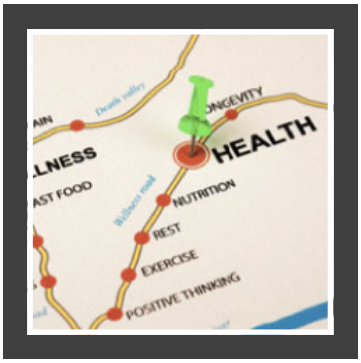
I HAVE Chiari
(kee-AR-ee)



Taking Chiari to School: A Primer for School Nurses

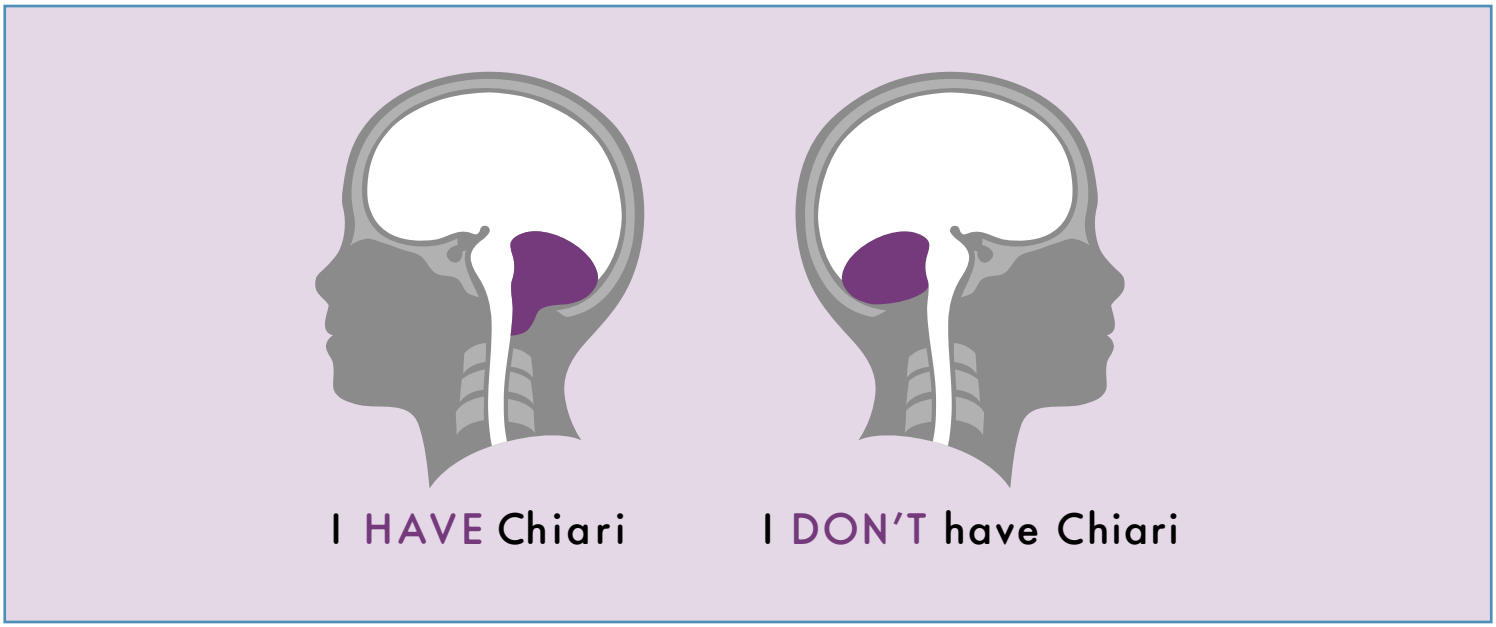
Contents

CONQUER CHIARI®
From Knowledge, Strength. Through Research, Hope.

This primer is for school nurses who may be responsible for the care of a child with the Chiari malformation type one (CM1). CM1, although not a common condition, may present challenges for the child in the school setting. Even though most children with CM1 are generally able to attend school with their peers, some require nursing care and accommodations in the school setting to promote their safety and success. Your familiarity with the CM1 diagnosis, symptoms, treatments, and available resources will reinforce parent efforts and provide continuity of care for the child with CM1 in school.

Chiari Malformation Type 1

The Chiari Malformations (CM), a classification of defects that affect the hindbrain, involves the bony structure of the skull, the brain itself, or both.



Definition

The Chiari Malformation Type One (CM1) is the dropping of the cerebellar tonsils of the brain into the opening from the brain to the spinal canal. Normally, only the spinal cord passes through this opening, the foramen magnum. CM1, usually accidentally diagnosed in adolescence or adulthood by physicians searching for other conditions, is identified as incidental Chiari.

Although CM1 may be asymptomatic, it is the most common form of CM (Vannemreddy, Nourbakhsh, Willis, & Guthikonda, 2010). The National Institute for Neurological Disorders and Stroke reports the incidence of CM1 at 1:1000 live births (NINDS, 2012).

Causes

CM1 causes are either congenital or acquired. Congenital defects occur during fetal development (Vannemreddy et al, 2010). CM1 is the only type of CM acquired during a lumbar puncture (NINDS, 2012).

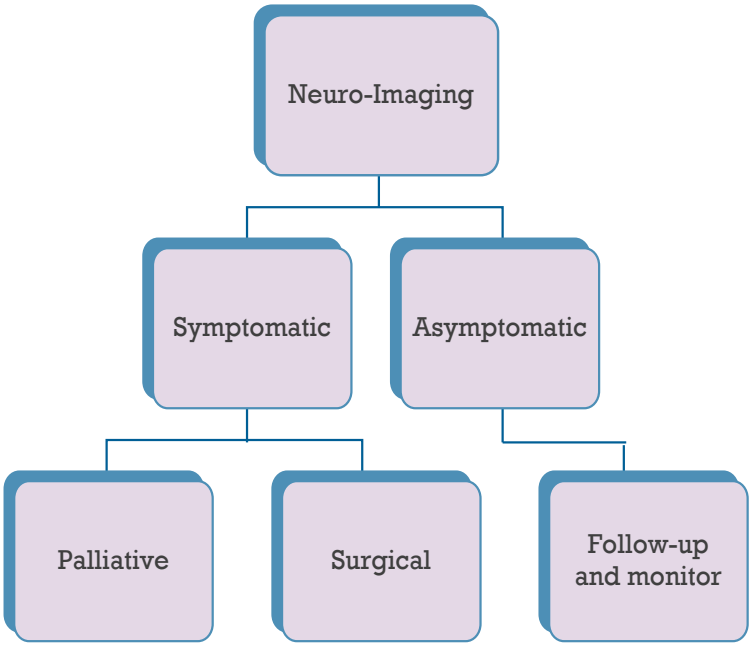
Comorbidities

1. ***Syringomyelia*** *(sear-IN-go-my-EEL-ya) occurs when a cyst forms within the spinal cord (NINDS, Syringomyelia, 2012)*

2. ***Hydrocephalus*** *is the build-up of CSF in the ventricles of the brain causing them to dialate and increase intracranial pressure.*

3. ***Tethered cord syndrome*** *is the limited mobility or attachment of the spinal cord within the spinal canal (NINDS, Tethered Cord, 2012).*

4. ***Spinal curvatures*** *include scoliosis and kyphosis (NINDS, 2012).*

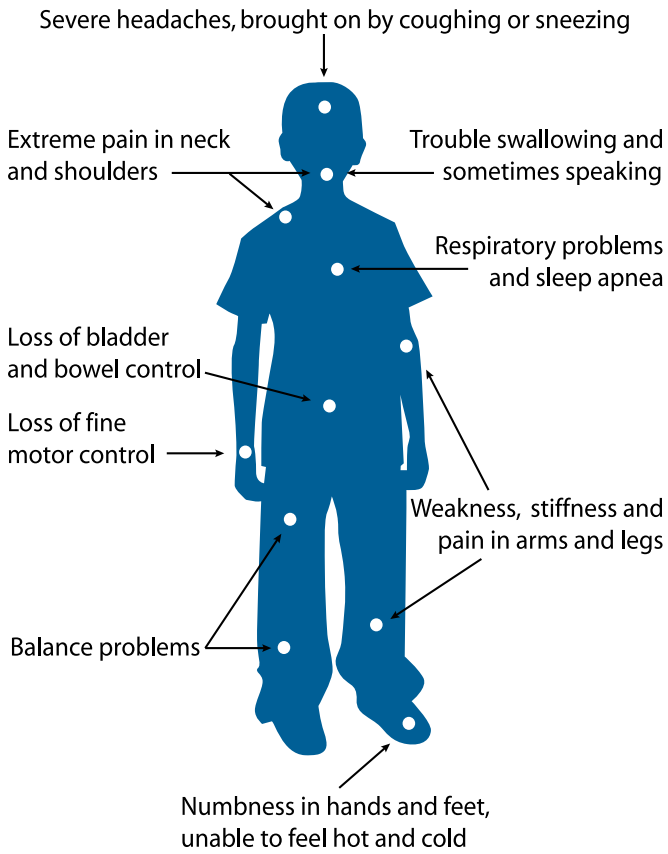


Diagnosis and Treatment

Suspicion of CM1 may be based upon presenting symptoms. However, at times, CM1 is discovered accidentally with magnetic resonance imaging (MRI) for other health concerns. Definitive diagnosis comes from neuro-imaging, including X-ray, computed tomography (CT), or MRI (Fernández et al, 2009). Asymptomatic children are conservatively monitored with regular neurological exams and neuro-imaging (Vannreddy et al, 2010). Children exhibiting symptoms may undergo a decompression surgery with or without duraplasty. Current best research evidence indicates that this surgery generally produces good outcomes and relief of symptoms (McGirt et al. 2008).

Symptoms

Children with CM1 have their own unique symptoms that may require specific accommodations in the school setting



CM1 symptoms vary. The amount of brain pressing down into the spinal canal does not determine symptom severity. For example, a child with a cerebellar tonsillar herniation of 5 mm may show more severe symptoms than one with 10 mm herniation. Children with CM1 may require individualized accommodations in the school setting.

The most common symptom in children is headache or neck and shoulder pain (Tubbs et al, 2007). Infants and children unable to communicate verbally may cry excessively and be irritable. In patients under three-years, symptoms are related to the mouth and throat (oropharyngeal) including aspiration, choking, regurgitation, dysphagia, abnormal vocal chord function, and chronic cough (Greenlee, Donovan, Hasan, & Menezes, 2002). Other symptoms include sleep apnea and feeding problems such as reflux, gagging, trouble swallowing, and poor weight gain.

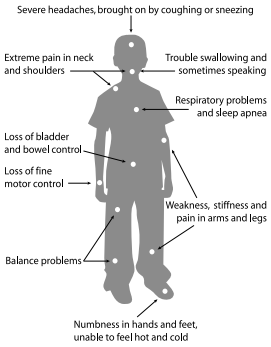
General symptoms include loss of fine motor control (handwriting), balance (clumsy), and sensation, as well as changes in bowel and bladder control, vision, and hearing. Older children may present with headache and/or spinal curvature. The Valsalva maneuver increases symptom severity. Sneezing, laughing, crying, and straining increase intracranial pressure, causing more pain and symptoms (Tubbs et al, 2007).

What is an Individual Health Plan (IHP)?

Chiari malformation type 1 (CM1) Individual Health Plan

I HAVE Chiari

The Chiari Malformation Type One (CM1) is the dropping of the cerebellar tonsils of the brain (not the tonsils in your throat) into the opening from the brain to the spinal canal.
Normally, only the spinal cord passes through this opening called the foramen magnum.
The most common presenting symptom in children is pain reported by the child as a headache or neck and shoulder pain.
Circle symptoms that apply to this student.



An individual health plan (IHP) is a document prepared for students that outlines how health conditions are managed in the school setting. The development of the individual health plan is a collaborative effort that includes families, school nurses, school staff, and health care providers and is reviewed annually by this collaborative team. An IHP is not an educational plan like an IEP or 504-plan; however, it may be included in the IEP and 504 as a supporting document for the provision of accommodations (NASN, 2008).

The child's need for an IHP is based on required nursing care. The IHP will include doctor's orders that are implemented in the school setting, medication administration, communication with the parent, and staff direction on proper response to the child's presenting symptoms in both chronic and emergency situations. Also noted on the IHP is the special training of school personnel to assist the child with timely and appropriate interventions. A school nurse may not always be present in the school to respond to the child's needs. Therefore a health aide or secretary will undergo training, known as delegation, to safely implement interventions for the child (NASN, 2008). Pictured above is a draft of a standardized IHP developed for the CM1. It includes a definition of CM1 and symptoms. Data fields allow for the customization of the plan for the child. Children with CM1 will have different symptoms, different needs, and their IHP will require customization to reflect this.

Chiari malformation type 1 (CM1) Individual Health Plan

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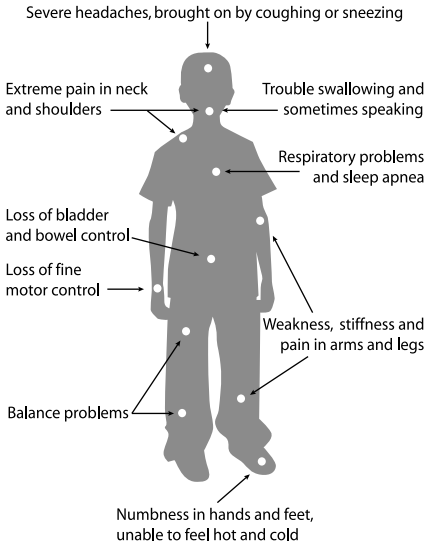

Health Concern:

Student name:

Parent or Guardian:

Grade/ Student ID #:

Contact information:

Academic year:

Emergency Contact:

Teacher:

Doctor:

Allergies:

Other health conditions:

Nursing Assessment:

Expected Outcome:

Intervention:

Staff Persons Involved: nurse, delegated staff, staff working w/ child

Implementation:

Evaluation Plan:

Items needed:

Persons Responsible:

Deadline:

Nursing Assessment:

Expected Outcome:

Intervention:

Staff Persons Involved: nurse, delegated staff, staff working w/ child

Implementation:

Conclusions:

Items:

Persons Responsible:

Deadline:

Nursing Assessment:

Expected Outcome:

Intervention:

Staff Persons Involved: nurse, delegated staff, staff working w/ child

Implementation:

Conclusions:

Items:

Persons Responsible:

Deadline:

I give permission for the information on this care plan to be shared with the listed health care provider of my child and adult staff at the school that will be working with my child. This plan will remain in force for 1 year from approval date. It is the responsibility of the parent to notify the school nurse whenever there is a change in the health status of the child. Signed parent permission allows the school nurse or delegated staff to contact the health care provider as necessary and authorizes staff members to seek emergency medical assistance for my child

Signature of Parent/Guardian Date

Signature of School Nurse Date

CHIARI
frequently used terms

Parents of children with Chiari are Chiari experts. They speak its medical language. The school nurse should become familiar with the common CM1 terms.

The following non-medical definitions copied with permission from the Conquer Chiari website.

apnea - temporary stop in breathing, sleep apnea.

brainstem - the lowest part of the brain which connects with the spinal cord and controls automatic functions such as breathing and swallowing.

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape.

cerebellum - part of the brain at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance.

cerebrospinal fluid (CSF)- clear liquid in the brain and spinal cord, acts as a shock absorber.

Chiari malformation (CM) - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow.

decompression surgery- common term for any of several variations of a surgical procedure to alleviate a Chiari malformation.

duraplasty - surgical technique where a patch is sewn into the dura

foramen magnum - large opening at the skull base where the spinal cord passes and joins with the brain.

syringomyelia (SM) - neurological condition where a fluid filled cyst forms in the spinal cord **syrinx** - fluid filled cyst in the spinal cord.

tonsillar herniation - displacement of the cerebellar tonsils out of the skull; usually measured in mm below the skull base.

Valsalva – straining against a closed airway increasing intra-abdominal and intra-cranial pressure.

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www.conquerchiari.org

For more information:

Governmental and not-for-profit organizations provide information and services for CM1 patients and their family members.

Four primary organizations are:

- **National Institute of Neurological Disorders & Stroke, Brain Resources and Information Network**
  ninds.nih.gov
- **C&S Patient Education Foundation /Conquer Chiari**
  conquerchiari.org
- **American Syringomyelia and Chiari Alliance Project**
  ASAP.org
- **Chiari and Syringomyelia Foundation**
  csfinfo.org/

References

C& S Patient Education Education Foundation. (2003-2012). *Glossary of Medical Terms*. Retrieved from http://conquerchiari.org/education/glossary.html

Fernández, A., Guerrero, A., Martínez, M., Vázquez, M., Fernández, J., Chesa i Octavio, E., De la Cruz Labrado, J., Escribano Silva, M., Araoz, M., García-Ramos, R., Valdivia, J., Valbuena, R., &Ramón, J. (2009). *Malformations of the craniocervical junction (Chiari type I and syringomyelia: classification, diagnosis and treatment)*. BMC Musculoskeletal Disorders, 10Suppl 1S1.

Greenlee, J. W., Donovan, K. A., Hasan, D. M., & Menezes, A. H. (2002). *Chiari I Malformation in the very young child: The spectrum of presentations and experience in 31 children under age 6 years.* Pediatrics, 110(6), 1212.

McGirt, M., Attenello, F., Atiba, A., Garces-Ambrossi, G., Datoo, G., Weingart, J., Carson, B., & Jallo,G. (2008).*Symptom recurrence after suboccipital decompression for pediatric Chiari I malformation: analysis of 256 consecutive cases.* Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery, 24(11), 1333-1339.

National Association of School Nurses. (2008). Position Statement- Individualized Health Plans. Retrieved from: https://www.nasn.org/PolicyAdvocacy/PositionPapersandReports/NASNPositionStatementsFullView/tabid/462/ArticleId/32/Individualized-Healthcare-Plans-IHP-Revised-2008

National Institute of Neurologic Disorders and Stroke. (2012). NINDS Chiari Malformation Fact Sheet. Retrieved from http://www.ninds.nih.gov/disorders/chiari/detail_chiari.htm

Tubbs, R., Doyle, S., Conklin, M., & Oakes, W. (2006). *Scoliosis in a child with Chiari I malformation and the absence of syringomyelia: case report and a review of the literature*. Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery, 22(10), 1351-1354.

Vannemreddy, P., Nourbakhsh, A., Willis, B., & Guthikonda, B. (2010). Congenital Chiari malformations. Neurology India, 58(1), 6-14. doi:10.4103/0028-3886.60387

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