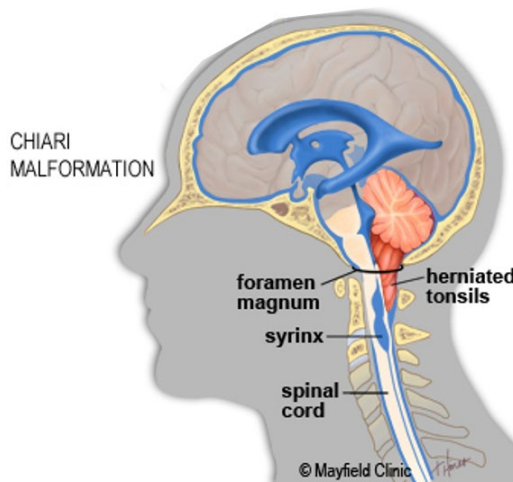


CT Children's CLASP Guideline

Chiari Malformation

INTRODUCTION

Chiari Malformation is 'crowding' of the junction between the brain and spinal cord and can lead to Valsalva-induced occipital headaches and/or signs/symptoms related to dysfunction of the brainstem, cerebellum, or spinal cord. It is characterized on neuroimaging (typically MRI) by descent of the cerebellar tonsils 5 or more millimeters below the base of the skull at foramen magnum. Chiari malformation is found incidentally on neuroimaging in 1-3% of pediatric patients. The objective of this CLASP tool is to aid primary care providers in establishing appropriate neurosurgery follow up and educate caregivers on this diagnosis.



INITIAL EVALUATION AND MANAGEMENT

History/Neurologic Exam:

- **Asymptomatic:** Patients are commonly found to have Chiari malformation incidentally with no symptoms
- **Headaches:** Headache pattern is characteristically occipital in location, precipitated by Valsalva maneuver, and brief in duration. Other patterns of headache are unlikely related to Chiari
- **Brainstem-localized symptoms:** Include oropharyngeal dysfunction such as choking/gagging (more commonly with liquids), sleep disordered breathing, and diplopia
- **Cerebellar or spinal cord dysfunction:** Including ataxic/spastic gait and myelopathy with weakness, sensory loss and/or hyperreflexia
- **Musculoskeletal asymmetry:** Most commonly scoliosis which may be present in patients with Chiari-mediated syringomyelia

Initial diagnostic evaluation

- **Diagnosis:** Typically diagnosed incidentally with cross sectional imaging studies such as MRI or CT of the brain. Due to a lack of standardization in the measurement of the position of the cerebellar tonsils relative to foramen magnum, radiology reports may be either equivocal or inaccurate in assigning a diagnosis of Chiari malformation.

Please provide Chiari Malformation FAQ sheet to caregivers ([See Appendix A: FAQ](#)).

WHEN TO REFER	<p>ROUTINE REFERRAL: Patients with findings on imaging consistent with Chiari malformation should be seen by neurosurgery for evaluation. Most patients (to include all asymptomatic patients) can be seen in a routine fashion.</p> <p>URGENT REFERRAL: Can be considered for patients with acute onset or progressive neurologic findings. Direct physician to physician communication is requested in these cases. Please call the one-call access center (see below).</p>
HOW TO REFER	<p>Referral to Neurosurgery via CT Children's One Call Access Center Phone: 833.733.7669 Fax: 833.226.2329 For more information on how to place referrals to Connecticut Children's, click here.</p> <p>Appointments available in, South Hadley and Westport</p> <p>Information to be included with the referral:</p> <ul style="list-style-type: none"> ▪ Demographic information ▪ Clinical findings, if any ▪ Radiology report for outside imaging
WHAT TO EXPECT	<p>What to expect from CT Children's Visit:</p> <ul style="list-style-type: none"> ▪ Initial evaluation <ul style="list-style-type: none"> ○ Initial visits will include targeted history, physical/neurologic examination and review of imaging ○ The majority of patients seen for imaging-proven Chiari malformation do not require surgical intervention. Several published series discuss rate of surgical recommendation for these referrals is in the range of 15%. ○ Additional referrals and diagnostic testing may be required in some patients, including: complete spinal MRI, scoliosis radiographs, and referrals to neurology/pain management, sleep medicine, aerodigestive services, and ophthalmology. ▪ Recommendations for surgical management <ul style="list-style-type: none"> ○ May be indicated based on history, exam or imaging findings. Connecticut Children's neurosurgeons belong to the Park Reeves Syringomyelia Research Consortium and offer surgical options tailored to each patient. ▪ Follow-up <ul style="list-style-type: none"> ○ Asymptomatic children with a diagnosis of Chiari malformation <i>without syringomyelia</i> are typically followed for two years OR until the age of 5 based on literature examining longitudinal follow-up of patients with a diagnosis of Chiari in neurosurgery clinic. The rate of de novo Chiari-related symptoms in this population is exceedingly low (less than 3%) with no patient developing new symptoms more than 2 years after initial presentation. Our team has created a reference document (Appendix 1) covering FAQs (frequently asked questions) for primary care providers and families.

Appendix A: Frequently asked questions in the patients with Chiari 1 malformation

Q: What is a Chiari malformation?

A: A Chiari malformation is something a child is usually born with. It means part of the brain is lower than normal and gets crowded at the base of the skull, where the brain meets the spine.

Q: What problems can Chiari malformation cause for my child?

A: Many kids with Chiari malformation have no symptoms at all! It's often found when they have an MRI for another reason. If there are symptoms, they may include:

- Quick, sharp headaches in the back of the head, especially after coughing or sneezing
- Trouble swallowing or breathing (like snoring)
- Weakness, numbness, or trouble walking
- A curved spine, called scoliosis

Q: Is Chiari malformation common?

A: Yes. About 1 to 3 out of every 100 kids who get a brain or spine MRI are found to have Chiari 1 malformation. Sometimes MRI reports may not be clear, and a neurosurgeon might say everything looks normal.

Q: Does every Chiari malformation need treatment?

A: No. Most kids who see a neurosurgeon for this don't need surgery. In a large study, fewer than 15 out of 100 kids with Chiari needed an operation. The doctors at Connecticut Children's will talk with you if surgery is ever needed.

Q: Will Chiari malformation get worse over time?

A: Usually not. Most kids stay the same or even feel better over time.

Q: Does my child need more MRIs after the first one?

A: Most of the time, no. But if your child also has something called syringomyelia (a fluid-filled space in the spinal cord), they might need follow-up MRIs.

Q: Can my child play sports or do normal activities?

A: It depends. Some activities like gymnastics or contact sports might need to be limited. Your doctor will look at your child's MRI and talk with you to decide what's safe.

Q: The neurosurgeon said to come back only if new symptoms appear. What should I and my pediatrician watch for?

A: It's rare for new symptoms to show up. In one study, fewer than 3 out of 100 kids had new problems after seeing a neurosurgeon. None had problems after two years. Doctors usually stop follow-up visits after that. Kids under 5 are followed longer.

You should come back to the neurosurgeon if your child has:

- New headaches in the back of the head that happen with coughing or sneezing
- New trouble swallowing, especially liquids
- New weakness, numbness, or trouble walking

If your child has a curved spine (scoliosis), see an orthopedic doctor first. They can check if it might be related to Chiari.