Family Education Sheet

Long Term Care: Chiari Malformation Type I

Boston Children's Hospital
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This family education sheet gives answers to frequently asked questions about what to expect in the long-term when your child has Chiari malformation type I (CMI).

Long-term outlook

How do most children do long-term?

- CMI affects children very differently. Your child may not have any symptoms. Or they may have intense symptoms.
- Whether or not they get surgery to treat their CMI, it's important to watch for symptoms, neurological changes or complications like syringomyelia.
  - This is when a cyst filled with cerebrospinal fluid, called a syrinx, forms within the spinal cord.
- Most children with CMI don't need long-term follow-up care if they don't have symptoms and if they don't have a syrinx.

What kind of monitoring will my child need?

Neurological follow-ups

- Regular visits to a neurologist or neurosurgeon, usually every 6–12 months if your child has symptoms
- Detailed neurological exams to track changes in your child's motor skills, reflexes, sensory perception, balance and cranial nerve function

MRI tests

- Baseline MRI of the brain and cervical spine when your child is diagnosed with CMI
- Follow-up MRIs if:
  - New or worsening symptoms appear
  - Your child gets a syrinx
  - Your child needs monitoring after surgery to check on how they're healing

Monitoring for syringomyelia or scoliosis

- Syringomyelia is the development of a fluid-filled cyst within the spinal cord. If this happens, your child will probably need spinal MRIs.
- Scoliosis is a curvature of the spine. Your child may need scoliosis screenings, especially during growth spurts.

How do we manage ongoing symptoms?

Headaches and neck pain

- Medications, like NSAIDs, muscle relaxants, anticonvulsants (gabapentin) or tricyclics, can help.
- Botox injections or nerve blocks (possibly) can help.
- Watch for "cough headaches." This means that there is a disruption to cerebral spinal fluid (CSF) flows.

Neurological symptoms

- For numbness, tingling or weakness: physical therapy, occupational therapy and medication (pregabalin)
- For coordination issues: balance training and adaptive support devices

Sleep and breathing disorders

- Sleep studies monitor for:
  - Severe central sleep apnea
  - Affected breathing or lower oxygen levels
- Possible use of CPAP/BiPAP machines or an evaluation by a sleep specialist

Swallowing and voice problems

- Referral to a speech-language pathologist
- Modified diets or swallowing strategies if your child has swallowing problems

Activity changes

- Have your child:
  - Avoid high-impact activities (weightlifting, contact sports, jumping).
  - Do low-impact exercises, like walking, swimming, yoga or tai chi.
  - Avoid holding their breath and straining (Valsalva maneuvers), which can trigger symptoms

Posture and neck health

- Have your child:
  - Use a proper desk setup and practice good posture
  - Sleep with a supportive cervical pillow
  - Avoid keeping their head tilted or looking down at screens for too long

Hydration and nutrition

- Being well hydrated (drinking plenty of water) supports CSF flow.

- Healthy, balanced diets may help your child have fewer headaches.

Common cognitive challenges

- Brain fog: problems with memory, concentration and processing speed
- Fatigue, mood swings or irritability

Support strategies

- Cognitive behavioral therapy (CBT) for coping and managing symptoms
- Getting evaluations for school/work accommodations
- Focusing techniques: prioritize tasks, use reminders and take breaks

How should I keep track of symptoms and tell my child's care team?

Keep a symptom diary to help track your condition and support treatment planning. Write down details like:

- How many headaches your child gets and how intense they are
- Any sensory or motor changes
- Sleep patterns and disturbances
- Identifiable triggers (causes) or activity-related patterns

Bring this log to medical appointments to help your child's provider make informed decisions about their treatment or the need for imaging tests.

What happens after surgery, if my child gets surgery?

Even if your child has decompression surgery, they still may need long-term care:

- Follow-up imaging to check for regrowth of removed bone, scar tissue or for CSF flow issues
- Watching for ongoing or returning symptoms
- Physical therapy to restore your child's full range of motion and strength

What is important to know about school, work and how my child does socially?

School accommodations (504 plan or IEP)

- Can give extra time for assignments or exams
- Changes in how they participate in PE/gym
- Can give breaks for fatigue, pain or sensory overload

Workplace adjustments

- Supportive workstation arrangements
- Flexible schedules or remote work options
- Taking breaks now and then to manage fatigue (tiredness) or pain

Social and family life

- Talk openly and honestly about invisible symptoms.
- Encourage balance between activity and rest.
- Get mental health support if your child has stress, anxiety or depression.

When should we get medical attention right away?

Contact a doctor or go to the nearest Emergency Department if your child:

- Gets sudden worsening headaches or neurological symptoms
- Feels new weakness or numbness in their arms or legs
- Pees/poops without meaning to
- Has a hard time swallowing or breathing
- Faints or collapses without a reason

Helpful resources

- Conquer Chiari: conquerchiari.org
- Online patient groups and local support networks

Long-term success tips

- Go to all of your child's follow-up visits and imaging tests.
- Balance your child's activities and have them take breaks to help prevent symptoms from getting worse.
- Build a support network. You're not alone!
- Advocate for your child at school, work and medical settings.

FAQ: Chiari I

What is Chiari malformation type I?

Chiari malformation type I CMI) is a condition where a part of the brain (the cerebellar tonsils) pushes down into the opening at the bottom of the skull. This can press on the spinal cord and block the normal flow of fluid around the brain and spine.

Is Chiari type I serious?

Sometimes it is, but not always. Some people have mild symptoms, or no symptoms at all. Others may have more serious symptoms, like fluid building up in the spinal cord (called a syrinx), trouble breathing during sleep, problems swallowing or trouble with balance and hand coordination.

What causes Chiari malformation type I?

CMI is usually congenital (a child is born with it) and may be caused by:

- A structurally small or misshapen skull
- Genetic conditions like Ehlers-Danlos syndrome
- Connective tissue disorders
- In rare cases, it may develop later in life because of a trauma, CSF leak or spinal problems.

What are common symptoms?

Symptoms often depend on a child's age but may include:

- Occipital headaches (at the back of the head), that get worse with coughing, sneezing, laughing or straining
- Neck pain or stiffness
- Dizziness, vertigo or poor balance
- Numbness or tingling in the hands, arms or feet
- Muscle weakness, especially in upper limbs
- Difficulty swallowing (dysphagia)
- Central sleep apnea

How is Chiari malformation type I diagnosed?

CMI is usually diagnosed with MRI imaging, which can show:

- The extent of tonsillar herniation (how far the cerebellum is displaced)
    - These "tonsils" aren't the ones in your throat that many people think of! They are two very different structures.
- Whether CSF flow is blocked
- If there is a syrinx (syringomyelia)
- If your child has other conditions like scoliosis

A neurological exam may also check your child's reflexes, balance and coordination.

Does everyone with this condition need treatment?

No, most children don't need treatment. Treatment depends on:

- If your child has symptoms and how intense they are
- Whether there's a syrinx or CSF blockage
- How the condition affects your child's daily functioning and quality of life
- If your child has mild or no symptoms, they may only need regular monitoring. If your child has symptoms, they may benefit from surgery or other treatments.

What is the surgery and when is it recommended?

The most common surgery is called posterior fossa decompression. This involves:

- Removing a small part of skull at the base (and sometimes part of the top cervical vertebra)
- Possibly opening the dura (membrane covering the brain) to allow more room
- Restoring normal CSF flow

Surgery is recommended when:

- Symptoms are moderate to severe
- Neurological symptoms are getting worse
- Imaging shows CSF blockage or a syringomyelia

What is recovery like after surgery?

- Hospital stay: 2–5 days
- Recovery time: 4–12 weeks
- Your child may need to stop activities like driving, lifting or working for a little while.
- Some symptoms get better right away. Others slowly get better over time.

Can Chiari malformation type I cause long-term problems?

Yes, if your child has symptoms and it isn't treated, it can lead to:

- Chronic (ongoing) pain
- Permanent nerve damage
- Scoliosis
- In severe cases, a hard time breathing or walking

Can young children have Chiari malformation type I?

Yes. CMI can affect infants, children or adolescents. In children, it may be associated with:

- Developmental delays
- Swallowing or breathing problems
- Spinal deformities (like scoliosis)

Can my child live a normal life with Chiari malformation type I?

Yes, many people do. With regular monitoring, symptom management and appropriate treatment, most people can lead a full and active life. Your child may need to:

- Avoid heavy lifting or contact sports
- Stay hydrated (drink plenty of fluids) and minimize strain
- Get regular follow-up MRIs

Which doctors should my child see?

- **Neurologist:** to get a diagnosis and manage symptoms
- **Neurosurgeon:** to go over surgical options
- Look for specialists who have experience treating children with Chiari malformation type I and CSF disorders.

This Family Education Sheet is available in Spanish.