CONDITION

Arnold-chiari malformation

Learn about Chiari malformation, including symptoms, causes, MRI diagnosis, treatment options, possible complications, and when to seek urgent care.

WHAT IT IS
Structural malformation of the lower brain

BODY SYSTEM
Nervous system

MAIN SYMPTOMS
Back-of-head pain, balance problems, weakness, swallowing difficulty

WHAT CAUSES IT
Usually developmental, sometimes genetic or acquired

HOW IT IS DIAGNOSED
Neurological examination and MRI

MAIN TREATMENT
Observation, pain management, or surgery

CAN IT BE CURED?
Sometimes - surgery helps but symptoms remain

IS IT AN EMERGENCY?
Sometimes - severe breathing difficulty or sudden paralysis

LONG-TERM OUTLOOK
Symptoms may improve, but nerve damage may persist

Overview

Arnold-Chiari malformation, more often called Chiari malformation , is a structural condition involving the lower part of the brain and the opening at the base of the skull.

The cerebellum is the part of the brain that helps control balance and coordination. In Chiari malformation, tissue from the lower cerebellum extends through the opening where the spinal cord leaves the skull. This can crowd the area and interfere with the normal flow of cerebrospinal fluid, the clear fluid surrounding the brain and spinal cord.

The name "Arnold-Chiari malformation" was historically used mainly for Chiari type II. Today, people often use it for several types of Chiari malformation.

The main types include:

Some people with Chiari type I never develop symptoms. Others have headaches, balance problems, weakness, swallowing difficulties, or other neurological symptoms. The condition matters because significant crowding can affect the brainstem, spinal cord, or cerebrospinal fluid flow. It can also contribute to a fluid-filled cavity inside the spinal cord called a syrinx .

- Chiari type I: The lower cerebellar tissue, called the cerebellar tonsils, extends into the upper spinal canal. It may be found in childhood or adulthood.
- Chiari type II: More brain tissue extends downward. It is usually associated with myelomeningocele, a severe form of spina bifida in which the spinal cord and its coverings develop outside the spine.
- Chiari type III: Brain tissue extends through an opening in the back of the skull. This type is rare and generally more serious.
- Chiari type IV: An older term used for severe underdevelopment of the cerebellum. It is now considered distinct from the usual forms of Chiari malformation.

Symptoms

Symptoms vary widely. The amount of cerebellar tissue extending below the skull does not always match how a person feels. A person with a larger descent may have few symptoms, while someone with less descent may have important symptoms.

A typical symptom is pain at the back of the head or upper neck. This pain may become worse with coughing, sneezing, laughing, bending, lifting, or straining. These actions briefly increase pressure inside the head and spinal canal.

Other possible symptoms include:

Headaches are common in the general population and may have causes unrelated to Chiari malformation. A clinician will consider the headache's location, triggers, duration, and associated symptoms rather than assuming every headache comes from the MRI finding.

Infants and young children with Chiari type II may have feeding problems, weak crying, choking, noisy breathing, pauses in breathing, arm weakness, or abnormal eye movements. Children may not be able to describe pain clearly, so changes in feeding, sleep, movement, school performance, or behavior may be important.

Arrange prompt medical assessment for new or worsening weakness, repeated choking, increasing difficulty walking, loss of hand function, or new bladder or bowel control problems.

Call emergency services for severe trouble breathing, inability to swallow with ongoing choking, sudden paralysis, loss of consciousness, or a sudden severe headache accompanied

by new confusion, weakness, vision loss, or difficulty speaking. These symptoms can have causes other than Chiari malformation and need immediate assessment.

- Neck pain, dizziness, or problems with balance and coordination
- Numbness, tingling, weakness, or reduced temperature sensation in the arms or hands
- Trouble with fine hand movements, such as writing or fastening buttons
- Ringing in the ears, hearing changes, blurred vision, or abnormal eye movements
- Difficulty swallowing, choking, hoarseness, or changes in speech
- Sleep-related breathing problems, including sleep apnea
- Curvature of the spine, called scoliosis, especially when a syrinx is present
- Bladder or bowel problems when the spinal cord is affected

Causes and risk factors

Most Chiari malformations are thought to begin during fetal development. The space in the lower back part of the skull may be smaller or shaped differently than usual. This leaves less room for the cerebellum and may push its lower portion toward the spinal canal.

Chiari type II develops in connection with myelomeningocele. Changes in the developing brain, skull, and spinal cord are linked rather than being separate problems.

Some cases of Chiari type I appear to run in families, suggesting that genetic factors may contribute. However, there is no single known inheritance pattern for most people. Having Chiari malformation does not mean that a person's children will necessarily develop it.

Less commonly, downward movement of the cerebellar tonsils can develop later because pressure around the brain has changed. Possible causes include a cerebrospinal fluid leak, a mass, hydrocephalus, or another condition affecting pressure inside the skull. Hydrocephalus means that too much cerebrospinal fluid collects in the brain's fluid spaces.

Connective tissue or bone conditions may occur alongside Chiari malformation in some people. A medical evaluation is needed to decide whether an associated condition is relevant. Everyday activity, posture, or something a person did does not cause a congenital Chiari malformation.

Diagnosis

Chiari type I is sometimes found unexpectedly when imaging is performed for another reason.

An MRI finding alone does not show whether Chiari malformation is causing a person's symptoms.

Diagnosis usually begins with a medical history and neurological examination. The clinician may assess eye movements, swallowing, speech, strength, sensation, reflexes, coordination, balance, and walking. They will also ask whether headaches are triggered by coughing or straining and whether symptoms have changed over time.

Magnetic resonance imaging, or MRI, is the main imaging test. It provides detailed pictures of the brain, the junction between the skull and spine, and the spinal cord. MRI can show the position of the cerebellar tonsils, crowding around the brainstem, and related problems such as a syrinx or hydrocephalus.

The distance that the tonsils extend below the skull is one part of the assessment, but no measurement should be considered alone. Symptoms, neurological findings, cerebrospinal fluid spaces, age, and other anatomical features also matter.

A clinician may recommend MRI of more of the spine to look for a syrinx or other spinal cord changes. In selected cases, a specialized MRI called a cine flow study may be used to examine cerebrospinal fluid movement. Its value depends on the clinical situation and it does not replace a full evaluation.

Other tests may include a sleep study for suspected sleep apnea, a swallowing assessment, hearing or eye testing, or X-rays to monitor scoliosis. A neurosurgeon or neurologist may review the images when symptoms are significant, the diagnosis is uncertain, or a syrinx is present.

Treatment and management

Treatment depends on the type of Chiari malformation, symptoms, examination findings, associated conditions, and changes seen on follow-up imaging.

People who have no symptoms and no concerning neurological or spinal cord findings may not need surgery. Their clinician may recommend observation, education about symptoms, and periodic follow-up. Repeat MRI is not required on the same schedule for everyone.

Headache or neck pain may be treated with general pain-management approaches when appropriate. Medication may reduce pain, but it does not change the underlying crowding or restore cerebrospinal fluid flow. Because headaches can have several causes, treatment should be based on the headache pattern and medical evaluation.

Activity advice should be individualized. Many people can remain active. A clinician may suggest avoiding an activity if it repeatedly causes marked symptoms or if there is another

concern, such as spinal instability. People should not assume that all exercise, lifting, air travel, or routine daily activity is unsafe.

Surgery may be considered when symptoms are significant or worsening, there is neurological impairment, cerebrospinal fluid flow is blocked, or a syrinx is present or progressing. The most common operation is posterior fossa decompression . The surgeon removes a small amount of bone from the back of the skull to create more space.

Depending on the anatomy and the surgeon's plan, the operation may also include:

The goal is to relieve crowding and restore cerebrospinal fluid movement, not simply to change a measurement on MRI. Surgery often helps pressure-related cough headaches and may prevent further neurological damage, but it cannot guarantee that every symptom will resolve. Long-standing nerve or spinal cord damage may not fully recover.

Associated problems may need separate treatment. These include hydrocephalus, myelomeningocele, spinal instability, tethering of the spinal cord, or a persistent syrinx. Direct drainage of a syrinx is usually reserved for selected situations because treating the cause of blocked fluid flow is often the first approach.

- Removal of part of the first neck vertebra, called a C1 laminectomy
- Opening the dura, the protective covering around the brain and spinal cord
- Sewing in a patch to enlarge that covering, called duraplasty
- Additional procedures in selected cases to improve fluid flow

Complications

Untreated, symptomatic Chiari malformation can sometimes lead to progressive neurological problems. A syrinx may enlarge and damage the spinal cord, causing weakness, muscle wasting, altered sensation, stiffness, pain, scoliosis, or bladder and bowel dysfunction.

Crowding near the brainstem may contribute to swallowing problems or sleep-related breathing disorders. Repeated choking can allow food or liquid to enter the airway. This is called aspiration.

Surgery also has risks. Possible complications include infection, bleeding, cerebrospinal fluid leakage, a fluid collection under the skin, meningitis, injury to nearby nervous system structures, or continued symptoms. Scar tissue or renewed obstruction can occur, and some people need another operation. A neurosurgeon can explain how the expected benefits and risks apply to the specific findings.

Follow up

Follow-up is based on symptoms, age, examination findings, and whether a syrinx or another associated condition is present. Care may involve primary care, neurology, neurosurgery, rehabilitation, sleep medicine, or other specialists.

At routine visits, discuss changes in headaches, balance, swallowing, sleep, strength, sensation, hand function, and bladder or bowel control. Children may also need monitoring for scoliosis, growth, school difficulties, and changes in motor development.

After surgery, follow the care team's instructions about wound care, activity, return to work or school, and imaging. Recovery time varies. Improvement may be gradual, and some symptoms may remain even when cerebrospinal fluid flow has improved.

Contact the treating team promptly for worsening weakness, repeated falls, increasing numbness, new swallowing problems, drainage or swelling at a surgical incision, fever after surgery, or a major change in bladder or bowel control.

Seek emergency care for severe breathing difficulty, ongoing choking, sudden major weakness, loss of consciousness, or sudden neurological symptoms such as facial drooping or difficulty speaking. Routine headaches or stable long-term symptoms can usually be discussed at a scheduled visit unless their pattern changes significantly.

Sources

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