Understanding Chiari Malformation

A Chiari malformation is a problem in the structure of the skull and brain. It's often present at birth. It occurs when the skull doesn't form correctly or is too small. As a result, parts of the brain are forced down into the foramen magnum. This is the hole at the base of the skull. It's where the spinal cord enters the skull to connect to the brain.

A Chiari malformation may block the flow of cerebrospinal fluid (CSF) between the brain and spinal cord. This fluid protects the brain. It also carries nutrients to the brain and takes waste out of it. A blockage in CSF can lead to a buildup of fluid and pressure in the brain. This condition is called hydrocephalus.

There are 4 types of Chiari malformation. They are based on how severe the condition is and how much of the brain is affected:

- **Type 1.** This is the most common form. In this type, the bottom part of the cerebellum dips into the foramen magnum. The cerebellum is the part of the brain that controls balance. People with this type may have no symptoms. Or they may develop symptoms as teens or adults. In some cases, the condition is found only through imaging tests, such as an MRI, for some other health reason.
- **Type 2.** In this type, parts of the cerebellum and the brain stem may be in the foramen magnum. This type is often noticed in childhood. It tends to block the flow of CSF, causing more serious symptoms. This type is often linked with a spine malformation called a myelomeningocele.
- **Type 3.** This rare form is very serious and life-threatening. It's found at birth. Parts of the brain and brain stem may stick out of a hole in the back of the skull.
- **Type 4.** This is also known as cerebellar hypoplasia. In this rare condition, the cerebellum doesn't completely form. Babies born with type 4 often die at birth. They may have other skull or spinal cord defects.

How to say it

kee-AR-ee MAL-fohr-MAY-shuhn

What causes Chiari malformation?

Experts aren't sure what causes a Chiari malformation. It's likely the result of a mutation in a gene that happens as a fetus develops.

Sometimes, a person can develop a Chiari malformation later in life. It may occur due to injury, a disease, a tumor, or an infection.

Symptoms of Chiari malformation

The symptoms of a Chiari malformation vary from person to person. Adults and children with type 1 may have no symptoms. Or they may start to have symptoms as they get older. Those with other types of the condition often have symptoms at birth or in childhood.

The main symptom of a Chiari malformation is a headache or neck pain. This pain may be worse after sudden coughing, sneezing, or straining. Other symptoms include:

- Trouble speaking or swallowing
- Trouble with balance or coordination
- Muscle weakness or numbness
- Dizziness

- Fainting
- Vomiting
- Hearing problems like tinnitus
- Sleep problems like sleep apnea
- Blurry or double vision

In babies, the condition may look like other health problems. Babies may have trouble feeding. They may cry a lot. They also may not develop or grow properly.

Treatment for Chiari malformation

Treatment is not always needed for a Chiari malformation. That's often the case for children and adults who don't have any symptoms. They may instead have regular checkups and MRIs to keep an eye on the condition.

If a Chiari malformation causes symptoms, treatment may include:

- Pain relievers. Medicines like nonsteroidal anti-inflammatory drugs (NSAIDs) and muscle relaxers can ease headaches and neck pain.
- Stress management. High levels of stress can raise pain levels in the body. Activities like meditation and yoga may help lower stress and pain levels.
- A shunt. If a Chiari malformation blocks the flow of CSF between the brain and the spinal cord, a shunt may be needed. A shunt is a tube put in the brain to drain the CSF.
- Surgery. If symptoms are severe or getting worse, surgery may be needed. The most common type of surgery is posterior fossa decompression. During this procedure, a surgeon removes a bit of bone from the bottom of the skull, called the posterior fossa. This surgery makes more room in the skull for the cerebellum. It eases pressure around the brain and spinal cord. It can also help improve the flow of CSF. Surgery is usually needed for spine malformations.

Possible complications of Chiari malformation

A Chiari malformation can lead to these problems:

- Hydrocephalus, the buildup of CSF in the brain
- Damage to nerves or muscles
- Paralysis
- Problems with the spinal cord or spine, such as scoliosis

When to call your healthcare provider

Call the healthcare provider right away if you notice any of these:

- Fever of 100.4°F (38°C) or higher, or as directed by the healthcare provider
- Symptoms that don't get better, or get worse
- New symptoms