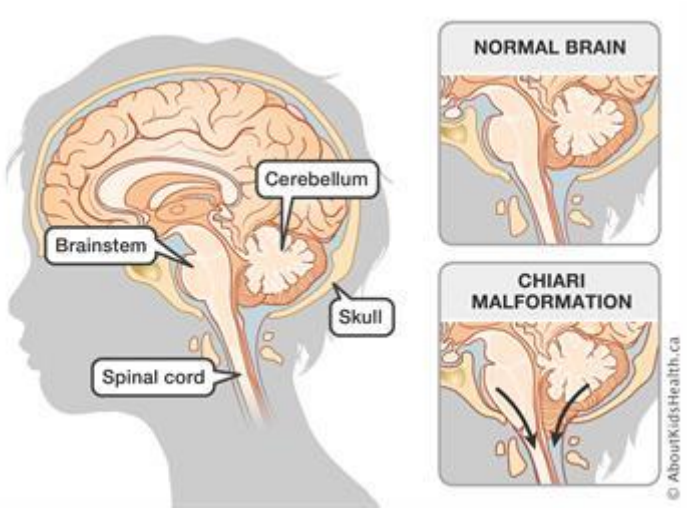
Chiari Malformations

What are Chiari Malformations?

A Chiari malformation (kee-AH-ree mal-for-MAY-shun), is a condition in which brain tissue extends into the spinal canal. It occurs when part of the skull is abnormally small, pressing on the brain and forcing it downward. The malformation is named after Austrian pathologist Hans Chiari.



Types of Chiari Malformations?

There are four types of Chiari malformations:

Type I This is by far the most observed type in children. In this type, the lower part of the cerebellum (the part of the brain at the back of the skull in vertebrates, which coordinates and regulates muscular activity.) -- but not the brain stem -- extends into an opening at the base of the skull. The opening is called the foramen magnum (Latin: great hole - is a large oval opening in the occipital bone of the skull). Normally, only the spinal cord passes through this opening.

Type I is the only type of Chiari malformation that can be acquired.

Type II This is usually only seen in children born with spina bifida. Spina bifida is the incomplete development of the spinal cord and/or its protective covering.

Type II is also known as ‘classic’ Chiari malformation or Arnold-Chiari malformation. In Type II Chiari malformation, both the cerebellum and the brain stem extend into the foramen magnum.

Arnold–Chiari malformation was named in honour of Hans Chiari and German pathologist Julius Arnold.

Type III This is the most serious form of Chiari malformation. It involves the protrusion or herniation of the cerebellum and brain stem through the foramen magnum and into the spinal cord. This usually causes severe neurological defects. Type III is a rare type.

Type IV This involves an incomplete or undeveloped cerebellum. It sometimes is associated with exposed parts of the skull and spinal cord. Type IV is a rare type.

What is the cerebellum?

The cerebellum is at the back of the brain, below the cerebrum. It is a lot smaller than the cerebrum. But it is a very important part of the brain. It controls balance, movement, and coordination (how our muscles work together).

Other conditions

In addition to spina bifida, other conditions sometimes associated with Chiari malformations include:

Hydrocephalus: An excessive build-up of cerebrospinal fluid in the brain.

Syringomyelia: A disorder in which a cyst develops in the central canal of the spinal cord.

Tethered cord syndrome: A progressive disorder in which the spinal cord attaches itself to the bony spine.

How is Chiari Malformation diagnosed?

Chiari malformations can be difficult to diagnose since the symptoms can be vague, or there may be no symptoms at all. A definitive diagnosis is generally made after an MRI scan, where the abnormal protrusion of the cerebellum toward the spinal cord can be seen.

In some cases, the diagnosis may be incidental, meaning that a person who undergoes an MRI scan for another reason may be diagnosed with Chiari when the scan reveals the abnormality – in these cases the person may not have experienced any symptoms at all.

In addition to an MRI scan of the brain, an MRI of the entire spinal column is useful.

In some people with Chiari, a cyst/syrinx (a fluid-filled cavity within the spinal cord) forms within the spinal column. Symptoms of leg numbness or scoliosis are more likely when a syrinx exists.

What are the symptoms of Chiari Malformations?

Chiari malformation is associated with a wide range of symptoms which vary by type.

Type I Chiari malformation usually causes no symptoms. Most people with the condition do not even know that they have it unless it is incidentally discovered during a diagnostic imaging test.

But if the malformation is severe, Type I may cause symptoms such as:

- Pain in the lower back of the head into the neck; it usually develops quickly and intensifies with any activity that increases pressure in the brain, such as coughing and sneezing
- Dizziness and problems with balance and coordination
- Swallowing difficulties
- Sleep apnea

Type II Most children born with Type II Chiari malformation have hydrocephalus. Older children with Type II Chiari malformation may develop head pain associated with:

- coughing or sneezing
- bending over
- strenuous physical activities
- straining to have a bowel movement

Some of the most common symptoms are linked to problems with the function of nerves in the brain stem. These include:

- weakness of vocal cords
- swallowing difficulties
- breathing irregularities
- serious changes in the function of nerves in the throat and tongue

What are the treatments for Chiari Malformations?

If a Chiari malformation is suspected, a doctor will perform a physical exam. The doctor will also check functions controlled by the cerebellum and spinal cord. These functions include:

- balance
- touch
- reflexes
- sensation
- motor skills

The doctor may order diagnostic tests, such as:

- X-ray
- CT scan
- MRI

An MRI is the test most often used to diagnose Chiari malformations.

If Chiari malformations cause no symptoms and do not interfere with activities of daily living, no treatment is necessary. In other cases, medications can be used to manage symptoms such as pain. Surgery is the only treatment that can correct functional defects or stop progression of damage to the central nervous system.

In both Type I and Type II Chiari malformations, the goals of surgery are to:

- Relieve pressure on the brain and spinal cord
- Re-establish normal fluid circulation through and around the area

In adults and children with Chiari malformations, several types of surgery can be performed. These include:

Posterior fossa decompression surgery: This involves the removal of a small portion of the bottom of the skull and sometimes part of the spinal column to correct the irregular bony structure. The surgeon also may open and widen the dura. That is the firm covering of the brain and spinal cord tissues. This creates additional space for the cerebrospinal fluid to circulate.

Spinal fusion: This may be performed in addition to posterior fossa decompression surgery in certain people with spine instability. The neck area of the spine may be unstable due to scoliosis, Ehler-Danlos syndrome (EDS) is a disease that weakens the connective tissues of your body. These are things like tendons and ligaments that hold parts of your body together), or another bone abnormality. Rods and screws are inserted to structurally reinforce the skull and neck vertebrae.

Shunting: This is used to reroute CSF. The shunt includes a flexible tube with a 1-way valve that directs the fluid out in the desired direction. For a syrinx in the spinal cord, one end of the tubing is placed in the syrinx. The other end is placed outside the spinal cord. A shunt remains inside the body after surgery. However, shunts pose risks and often become clogged or dislodge. Repeated surgeries may be necessary.

Treatment options vary depending on the severity of the symptoms and the presence of other conditions such as syringomyelia (a disorder in which a cyst or cavity forms within the spinal cord), hydrocephalus, and disorders of the skull and spine.

Observations

If a person has mild or no symptoms, monitoring by regular check ups and MRI scans may be recommended. Some people experience headache relief with taking anti-inflammatory or pain-relieving drugs. Her are some self-care tips to minimise neck strain:

- Ice packs for 20 minutes can help relieve neck and shoulder pain
- Get at least 8 hours sleep and use a good pillow

- Have a sleep study and evaluation for sleep apnea. A CPAP (Continuous Positive Airway Pressure) machine can improve your sleep and reduce fatigue.
- If you are overweight, shed some pounds to reduce the strain on your arms and legs – this will help with numbness/tingling sensations.
- Eat a healthy diet, include fibre, and drink plenty of water.
- Stay as active as possible with low impact activities such as walking, cycling or water aerobics.
- Play cards, crosswords, or Sudoku puzzles to sharpen your thinking.
- Tai Chi or Yoga can help stretch and tone muscles, improve balance, and reduce stress. Avoid poses that aggravate your symptoms.

Activities to avoid if you have a Chiari, with or without syringomyelia:

- High- velocity chiropractic manipulation - a chiropractic manual adjustment meant to restore normal range of motion in the joint
- Cervical traction - is a treatment often used in physical therapy to help treat neck pain and cervical radiculopathy (pinched nerves). It involves gently stretching your neck and separating the disc and joint surfaces in your cervical spine (neck).
- Trampolines, roller coasters, scuba diving and other activities that apply G forces to the neck
- Contact sports such as football, diving, running, weightlifting etc.
- Constipation and straining during bowel movements. Straining can cause worsening of the syrinx. A high fire diet, plenty of water and bowel management can help
- Lumbar punctures (spinal taps) and epidurals can be dangerous for a person with Chiari. Discuss these procedures with a neurosurgeon.
- Childbirth (bearing down and pushing) can also increase cerebellar herniation and formation of a syrinx. Make sure your obstetrician is aware of your Chiari and inform your neurosurgeon if you become pregnant.

It is important for people with Chiari to closely monitor their symptoms. Some people find it helpful to keep a symptom diary. By keeping track of how you feel and what you do, you may be able to find patterns, identify triggers and notice subtle changes over time. Bring your symptom diary to each appointment this will help you communicate more clearly with your doctor. Knowing what symptoms, you experience most and to what degree may help your diagnosis and treatment.

If symptoms worsen or if new ones develop call your neurosurgeon to inform them.