

What is Chiari Malformation?



Also known as Arnold Chiari Malformation, Chiari malformation affects the part of the brain called the cerebellum (see 'What is Hydrocephalus' for further information). There are several different forms of Chiari Malformation but most cases are congenital, meaning they are present from birth.

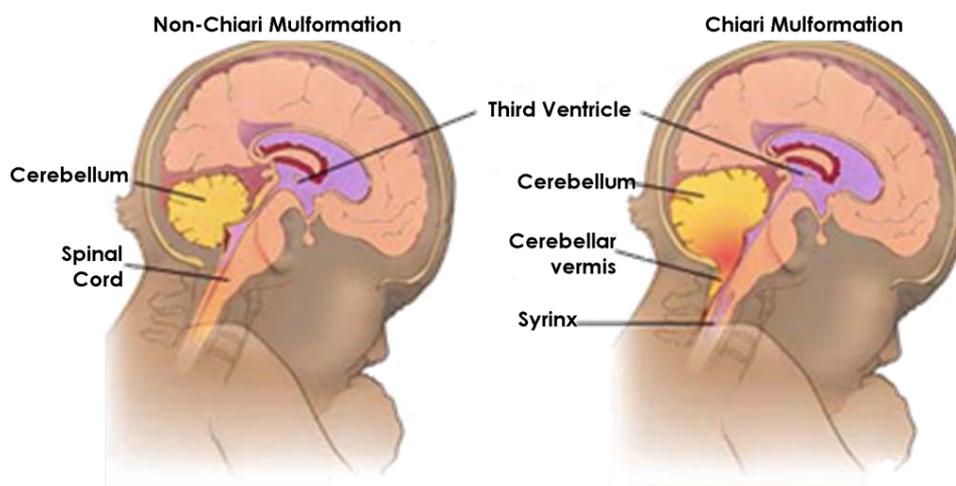
Although Chiari malformation can be present at birth, there may not be any symptoms until adulthood. For this reason, Chiari malformation is often not diagnosed until adulthood. There is a higher incidence of diagnosis in women than in men, the reasons for which are unknown.

What are the symptoms?

The most common symptom of Chiari malformation is a headache, which begins at the back of the head (neck) and radiates upward. The pain is often made worse or can be brought on by coughing, sneezing or straining. These activities are known as valsalva maneuvers.

Visual problems such as nystagmus (involuntary eye movements), double or blurred vision

may occur. Balance difficulties, vertigo and dizziness also may be present. Some people may have cranial nerve compression. This can result in apnea (cessation of breathing), gagging, swallowing difficulties, facial numbness or syncope (temporary loss of consciousness).



Symptoms may present as muscle weakness, particularly in the upper extremities, coordination problems, and gait abnormalities. Imaging of the spine may reveal a fluid collection inside of the spinal cord, known as a syrinx. Some individuals may have Hydrocephalus, a build-up of fluid in the ventricles of the brain.

What is Chiari Malformation?



Is there a Treatment?

The first step after diagnosis is to consult with a neurosurgeon who has experience treating and managing this disorder. Be aware that you may need to travel and you may wish to consult with more than one specialist.

If symptoms are mild and not progressing, your doctor may recommend conservative management. Supportive care such as headache and pain management, physical therapy or a reduction in activities can help manage symptoms.

An operation may be recommended. This is referred to as a posterior fossa decompression. The surgeon makes more room in the back of the head by removing small pieces of the skull bones. This reduces compression of the brain stem and allows the tonsils to move back into their natural position. The specific surgical techniques will vary among surgeons; no consensus yet exists on the best variation on this surgical procedure.

Is this condition hereditary?

Researchers investigated the genetic implications of the Chiari malformation with or without syringomyelia. A genetic prevalence has been identified in some families. Researchers continue to search for the gene(s) that are responsible for producing the Chiari malformation.

MRI scanning is recommended for family members who have signs or symptoms of the disorder.