The Neurological Disorder of Chiari Malformation

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**Abstract**

Examination of the neurological disorder of Chiari Malformation is presented. Historical background covers the basic discovery of Chiari. Radiological evidence that includes CT scans and MRI’s, are presented and show which part of the brain is affected by the disorder. Symptoms of the disease are also examined. The causes, and treatments and outcomes are also discussed.

What is ChiariMalformation?

Chiari Malformations are structural defects in the cerebellum. The cerebellum is the part of the brain that controls coordination, balance, voluntary motor movements, sense of position of the different parts of the body, equilibrium, and muscle tone. Normally the cerebellum and parts of the brain stem sit in an indented space at the lower rear of the skull just above the funnel-like opening to the spinal canal called the foramen magnum. When part of the cerebellum is located below the foramen magnum, it is called a Chiari Malformation. The resulting pressure on the cerebellum and brain stem may affect functions controlled by these areas and possibly block the flow of the cerebrospinal fluid. Cerebrospinal fluid is the liquid that cushions or acts as a buffer for the cortex, providing basic mechanical and immunological protection to the brain inside the skull and serves as a vital function in cerebral auto-regulation of cerebral blood flow.

History of Chiari Malformation

 In 1883 a Scottish physician named John Cleland first discovered the abnormality and called it Bassilar Impression syndrome.

In 1891 Hans Chiari an Austrian scientist, classified the malformation. It was then given the name of Chiari Malformation (Donavan, Greenlee, Hasan, Menezes, 2002).

 In 1896, Hans Chiari, defined Chiari Malformation as “Peg like elongation of the tonsils and medial part of the inferior lobes of the cerebellum which go along the medulla into the cervical canal.”

In 1894, a German scientist, Julius Arnold, published a paper called Myelocyste, Transposition von Gewebskeimen und Sympodie. In this paper he described Chiari Malformation in an infant that had died right after delivery.

 In 1907, two of Julius Arnold’s students penned the term Arnold-Chiari Malformation, which is still used today to describe the disease.

Signs and Symptoms

Many people with Chiari Malformation have no signs or symptoms. This condition is often detected when other tests are performed for unrelated disorders. There are four different types of Chiari Malformations that the doctors have categorized for all ages depending on the anatomy of the brain tissue that is displaced into the spinal canal, and whether developmental abnormalities in the brain or spine are present. The more common types of CM are types 1 and 2. The difference between type 1 and 2 is that the tissue in type 2 is protruded into the spinal canal more. CM type 1 is for adults, and type 2 is for younger children, pediatric. Migraines and headaches are the classical symptoms for these types of Chiari. Other symptoms of this disorder can consist of neck pain, poor fine motor skills, balance, numbness, dizziness, vision, inflamed sinuses, pressure in the ears, and slurred speech. For Chiari types 3 and 4 they are obvious at birth or through an ultrasound. With type 4 the brain itself was never fully developed normally. Every case is very individualistic. There hasn’t been one diagnosis that is the same. All signs and symptoms differentiate between each person.

Classification (types of)

 There are four types of Chiari, and each varies on the severity of the symptoms and the parts of the brain that protrude into the spinal area.

Type I involves the cerebral tonsils extending into the foramen magnum without involving the brain stem. It is the only type of Chiari that can be acquired.

Type II involves the extension of the cerebral tonsils and the brain stem into the foramen magnum. The tissue that connects the two halves of the cerebellum might be absent too. This disorder is usually accompanied by other diseases, such as spina bifida. This is also known as Classic Chiari Malformation.

Type III is the most serious of the types. This involves the conditions of type two but also the fourth ventricle that connects with the upper parts of the brain and regulates CSF fluid can also protrude down to the cervical cavity. Severe neurological defects are caused from this type of Chiari.

Type IV is when the tonsils are in the normal position but parts of the cerebellum are missing. This is an extremely rare form on Chiari, and in some cases is not included with the other three types.

Neuropsychological Findings

Before the availability of magnetic resonance imaging (MRI), Chiari Malformation was rarely diagnosed. In one study, a rate 0.6% was reported in all age groups, and a rate of 0.9% was reported in a study of only pediatric patients. Therefore, this condition is more common in both the adult and pediatric populations than was recognized previously. A female predominance has been reported in some large case series, with a male-to-female ratio of 2:3.

Recent studies have come to show that a Chiari Malformation can be congenital. It can either be present at birth or the child can have other signs and symptoms that lead up to this diagnoses. Researchers investigated the genetic implications of the Chiari malformation, and 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. Another cause of CM would be that of developing this disease after a minor/major injury to the brain such as; a car accident, concussion, shaken baby syndrome, etc. CM is most commonly found by having an MRI.

Recent studies from the Wisconsin Chiari Center have shown us that the neurological symptoms of Chiari malformation may also occur because of other disorders such as cervical spinal stenosis, tethered cord syndrome and syringomyelia. Spinal Stenosis is a progressive narrowing of the spinal canal that causes compression of the spinal nerve cord. Tethered Cord Syndrome is also known as Occult Spinal Dysraphism sequence. A congenital condition that causes the spinal cord, before or after birth, to become attached to the spinal column at some point along its length, most often in the lower (lumbar) portion. TSCS is related to spina bifida, a major birth defect and a type of neural tube defect that involves an opening in the vertebral column caused by the failure of the neural tube to close properly during embryonic development. Lastly, syringomyelia is the development of a fluid-filled cyst within your spinal cord. These different disorders may occur independently of Chiari malformation but frequently occur in conjunction with Chiari malformation in the same patient. The true incidences of Chiari malformations are not very well known, but continuous research is being done to find out more information about these diagnoses.

Causes

 Chiari Malformation can be caused my structural defect in the brain and spinal cord during fetal development (Amato, Hunt, 2007). Some cases of Chiari are familial, but no known gene has been found in connection with the disease. This would be called congenital Chiari Malformation.

 Injury, exposure to harmful substances, or infection can also cause Chiari Malformation. This would be considered acquired Chiari malformation because a person was not born with it.

Epidemiology

Chiari Malformation was once considered very rare, but with MRI technologies improving, the instances have risen. Estimates are anywhere from 200,000-500,000 cases per year nationally. Most reports put the instances at around 300,000 per year. This equals to about 1 in 1000. Women are more likely than men to be diagnosed with Chiari Malformation, especially with type two in which women are twice as likely to be diagnosed (Hadley, D. 2002). No significant information was found as to why women are more likely than men to have Chiari.

Treatment

Treatment for Chiari Malformation differs for each person that suffers from it. It can depend on the length of the herniation, and also the symptoms the person exhibits.

 If a person is asymptomatic, all that might be necessary is routine MRI’s and examination by a patient’s doctor. A patient may well be able to live with this malformation and not ever produce any symptoms. Routine MRI’s and exams can keep a patient informed in case anything with the disease changes.

 If there are headaches, backaches, or any other type of pain, anti-inflammatory medication, or pain relievers might be an option for the patient. Medication can treat and control the severity of the pain. There is no specific medication for treating Chiari Malformation; it is different for each person.

 There are a surgery options available to people also. The most common type of surgery is

Posterior Fossa Decompression (Achawal, Goel, 1995). The pressure can be relieved by a suboccipital craniectomy (Amato, Hunt, 2007). This is where a small portion of the back of the skull is removed to allow more room. Many times, part of the C1 vertebrae and occasionally the C2 vertebrae are partially removed. This helps to relieve the pressure of the herniated tonsils against the medulla. With this Cerebral Spinal Fluid flow around the foramen magnum improves (Chen, Chen, Li, Wang, Wu, Zhang, 2008). The dura mater can also be opened and the herniated tonsils can be shrunk by an electrical current. They can also be removed if necessary. Then a patch harvested from the patient’s body, or made of synthetic material is then sewn into place (Briani, Cagnin, Luchetta, 2009). This will also help relieve the pressure around the foramen magnum and allow CSF to flow more easily.

 A second procedure that is done is placing a shunt into the patient. This channels the flow of CSF fluid and relives pressure from the area. The shunt is placed in the brain and drains fluid to another part of the body. This procedure can be done if there is a shrinx involved with the Chiari abnormality. This is usually not the preferred method for treating Chiari, and is usually done if Posterior Fossa Decompression did not fix the shrinx.

 Other alternative treatments exist, but these are the most common and widely used.

Outcomes

 The outcomes of the treatment vary from patient to patient. In most studies that have been done, about 50% of those that have Posterior Fossa Decompression become symptom free. Another 10%

30% report some relief of symptoms, and another 10%-20% report no relief, and additional surgeries might be necessary. (Chen, Chen, Li, Wang, Wu, Zhang, 2008).

There have been a number of studies done where the surgery of CM has helped certain problems with the body, but created pain in other areas. Having the surgery is a high risk factor so at a young age while the spinal cord and brain are continually growing neurologists have suggested to wait on having surgery until the brain is fully developed, if it ever gets fully developed.

Some of the complications that can from surgery are leakage of CSF fluid, bleeding, vomiting. This happens in more than 5% of the patients after surgery. Between 1-5% of patients might experience stroke like symptoms (such as weakness of face muscles, that may be temporary or permanent) or a heart attack due to strain on the heart.

Chiari has been a rare illness and not very common, but as researchers look more into the brain disorders more and more people every day are getting diagnosed with CM.

Again, the outcomes vary from patient to patient, and more extensive research needs to be done on the outcomes of this surgery.

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