

Arnold-Chiari Malformation
By Myrtle Ross

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Arnold-Chiari Malformation was discovered by an Austrian pathologist named Hans Chiari (Answers.com). Hans Chiari was born in 1851 and studied medicine in Vienna from 1874 to 1879. During a period of forty years (1876 to 1916), Chiari published over 177 writings of medical literature (Who Named It). *“In 1891 in Prague Chiari published a series of observations called Über Veränderungen des Kleinhirns, der Pons und der Medulla oblongata infolge von Hydrocephalie des Grosshirns. Chiari had thus accepted the theory that the cause of the deformities in the posterior skull cave was the child's hydrocephalus. Five years later he published a richly illustrated monograph, in which he expanded his theories on the basis of a number of cases. In this he made the distinction between the deformities of the brainstem and the cerebellum which is still in use”* (Who Named It).

A medical college of Chiari's named Julius Arnold also published major writings on the same subject. Arnold's work was published three years after Chiari's. The article was about an infant that died after birth. *“The child had a large herniation of the spinal cord thoracolumbally. He also described the brainstem as underdeveloped with a downward draw of some of the lower parts of the cerebellum and the 4th ventricle in the spinal canal. Arnold reasoned that this was a case of monogerminal terathomatous deformity. He was, though, not able to explain the deformity of the brainstem”* (Who Named It). Both of these medical professionals inspired Gredig and Schwalbe, who published Arnoldsche und Chiarische Missbildung in 1907. This was when the name Arnold-Chiari Malformation was applied to the disorder (Who Named It).

Arnold-Chiari Malformation is basically a genetic disorder that causes parts of the cerebellum to form abnormally (Answers.com). Most of the medical profession believes

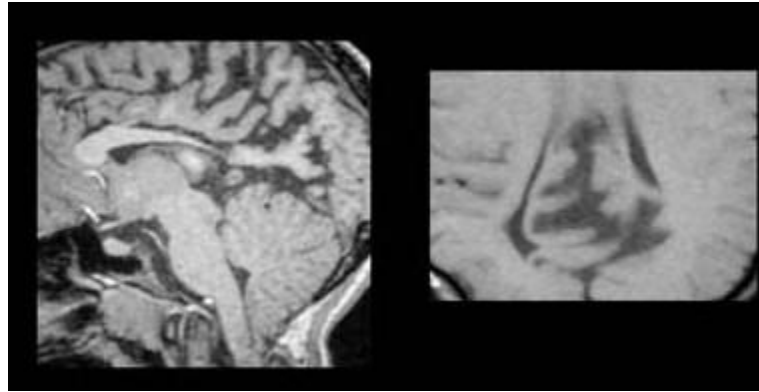
Arnold-Chiari Malformation to be genetic, but some think otherwise. Some of the other suspected causes of Arnold-Chiari Malformation are: the mother was exposed to toxic/hazardous chemicals, unborn infant did not receive the proper nutrients, or the mother used illegal substances or drank during pregnancy. The children of someone who has Arnold-Chiari Malformation have a greater likelihood of having it, but someone with no family history can have it as well. People with the malformation are born with it, but it can take years for symptoms to manifest (Columbia University). It is estimated that about 1 in every 1000 people has some form of Arnold-Chiari. Most of these people are thought to never develop symptoms (University of Southern California). There are four types of Arnold-Chiari Malformation.



Images of Arnold-Chiari Malformation Type I

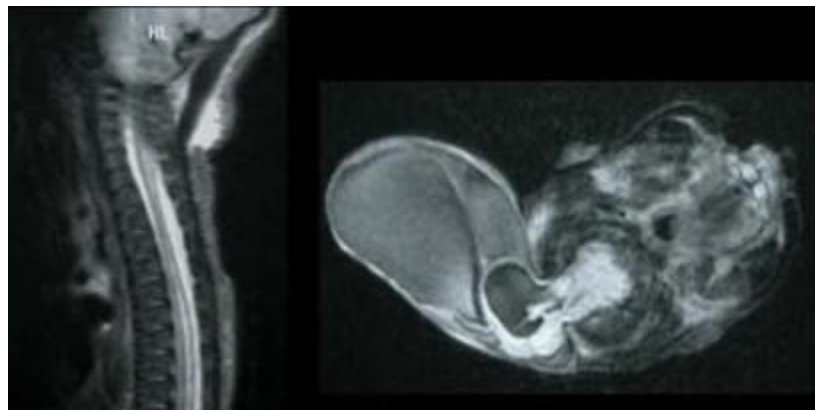
The least severe and most common type is **Arnold-Chiari Malformation Type I** (Pau). With this type of Chiari, symptoms may not surface until the person is in their teens or even into adulthood. The abnormality with this type is usually the cerebellum and cervical vertebrae are not formed correctly (Columbia University). The most common symptoms of this type of Chiari are: headache at the back of and base of the skull, inability to feel pain or temperature change in the hands/arms, or upper body muscle weakness, vision problems (light sensitivity, double vision), and a loss of coordination

and balance (University of Southern California).



Images of Arnold-Chiari Malformation Type II

Arnold-Chiari Type II is where in the ventricles of the brain there is too much cerebral spinal fluid. This is caused by the creation of too much cerebral spinal fluid or there is a problem with the ability to absorb it. The excess fluid causes intracranial pressure (Columbia University). The symptoms of this type are: intense headaches, breathing problems (such as Apnea), uncontrollable eye movements, muscle weakness, and weak gag reflex (University of Southern California).



Images of Arnold-Chiari Malformation Type II

Arnold-Chiari Type III is when the cerebellum grows down through the foramen magnum. The symptoms are: headaches, neck pain and/or stiffness, muscle weakness in the extremities, loss of feeling in the extremities, and breathing problems (Columbia University).

Arnold-Chiari Type IV is when the occipital lobe and the cerebellum develop abnormally. This is the rarest and most severe type. The symptoms are similar to the other types, but are far more intense (Columbia University).

A person can suffer from just one or more of the symptoms. The most common symptom of the types is headaches. These are chronic headaches that occur almost on a daily basis. They can be so painful that the person is unable to function or even get out of bed. The over the counter medications (such as Tylenol or Excedrin) will do little to nothing for the headaches.

There are several ways to diagnose Arnold-Chiari Malformation. The most common is an *MRI (Magnetic Resonance Imaging)*. This is a test that creates a three-dimensional image of the brain, skull, and spinal column. Type I can be seen on a *CAT scan (Computed Tomography Scan)*. It uses x-rays to create images of the brain. There are other less used tests that can diagnose Arnold-Chiari, such as *Brainstem Auditory Evoked Potential* or a *Myelogram*. These tests take longer to do and the MRI is just as (or more) effective (University of Southern California).

Arnold-Chiari is often treated surgically. The surgeries try to relieve or eliminate the symptoms entirely. The patient is put under a local anesthesia and then kept at the hospital for observation after the surgery. In most cases, a portion of the back of the skull is removed. If the symptoms persist, then the entire back of the skull may be removed. In

the case of Arnold-Chiari Type III and Type IV, a section of the foramen magnum is removed and in some cases, part of the back of the skull is also removed (University of Southern California).