New trends in diagnosis and treatments of Arnold Chiari Malformations

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List of Abbreviations

CSF	:	*Cerebrospinal fluid*
ICP	:	*Intracranial pressure*
ISF	:	*Interstitial fluid*
MRI	:	*Magnetic resonant imaging*
CMI	:	*Chiari malformation type I*
CM II	:	*Chiari malformation type II*
CM III	:	*Chiari malformation type III*
CM VI	:	*Chiari malformation type VI*
CT	:	*Computed tomography*
CAT	:	*Computerized axial tomography*
ACM	:	*Arnold Chiari malformation*
ICP	:	*Intra cranial pressure*
NTD	:	*Neural tube defects*
AFP	:	*alpha fetoprotein*
DSA	:	*Digital subtraction angiography*
VP	:	*Ventriculoperitoneal*

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Introduction

Arnold-Chiari malformation is a genetic disorder in which parts of the brain are formed abnormally. Malformations may occur in the lower portion of the brain cerebellum or in the brain stem **(Spillane, et al, 2007).**

An Austrian pathologist named Arnold-Chiari was the first to describe Arnold-Chiari malformation in 1891. Normally, the brain stem and cerebellum are located in the posterior fossa, an area at the base of the skull attached to the spinal cord. In Arnold-Chiari malformation, the posterior fossa does not form properly. Because the posterior fossa is small, the brain stem, cerebellum, or cerebellar brain tissues called the cerebellar tonsils are squeezed downward through an opening at the bottom of the skull. The cerebellum and/or the brain stem may extend beyond the skull or protrude into the spinal column. The displaced tissues may obstruct the flow of cerebrospinal fluid (CSF), the substance that flows around the brain and spinal cord. **(Pillay, et al, 2007).**

Although this malformation is present at birth, there may not be any symptoms of a problem until adulthood. For this reason, Arnold-Chiari malformation is often not diagnosed until adulthood. Women have a higher incidence of this disorder than men **(Rengachary, et al, 2006).**

Other names for Arnold-Chiari malformation are Arnold-Chiari syndrome, herniation of the cerebellar tonsils, and cerebellomedullary malformation syndrome. When doctors diagnose Arnold-Chiari malformation, they classify the malformation by its severity. An Arnold-Chiari I malformation is the least severe. In an Arnold-Chiari I malformation, the brain extends into the spinal canal. Doctors measure the length of brain stem located in the spinal canal to further define the malformation **(Rhoton, 2004).**

A type II malformation is more severe than a type I. It is almost always linked with a type of spina bifida. A sac protrudes through an abnormal opening in the spinal column. The sac is called a myelomeningocele. It may be filled with part of the spinal cord, spinal membranes, or spinal fluid. Unlike many cases of Arnold-Chiari I malformation, Arnold-Chiari II malformation is diagnosed in childhood. Doctors have identified Arnold-Chiari III and IV malformations, but they are very rare **(Saez, et al, 2008).**

Arnold-Chiari malformations may occur with other conditions. There may be excessive fluid in the brain (hydrocephalus), opening in the spine(spina bifida) or excessive fluid in the spinal cord (syringomyelia), but many people with Arnold-Chiari malformations do not have other medical problems **(Tobin and Sencer , 2003).**

Scientists do not know what causes Arnold-Chiari malformations. One hypothesis is that the base of the skull is too small, forcing the cerebellum downward. Another theory focuses on overgrowth in the cerebellar region. The overgrowth pushes the cerebellum downward into the spinal canal **(Tokime, et al, 2008).**

Some people with Arnold-Chiari I malformations have no symptoms. Typically, with an Arnold-Chiari I malformation symptoms appear as the person reaches the third or fourth decade of life. Symptoms of this disorder vary. Most symptoms arise from the pressure on the cranial nerves or brain stem. The symptoms may be vague or they may resemble symptoms of other medical problems, so diagnosis may be delayed **(Walsh, et al, 2006).**

One of the most common symptoms of Arnold-Chiari malformations is a headache. The headache generally begins in the neck or base of the skull and may radiate through the back of the head. Coughing, sneezing, or bending

forward may bring on these headaches. The headaches can last minutes or hours and may be linked with nausea **(Swanson and Fincher , 2008).**

There may be pain in the neck or upper arm with Arnold-Chiari malformations. Patients often report more pain on one side, rather than equal pain on both sides. There may also be weakness in the arm or hand. Patients may also report tingling, burning, numbness, or pins and needles. Balance can be affected as well. A person may be unsteady on their feet or lean to one side (**Walsh, et al, 2006).**

Some people with Arnold-Chiari malformation may have difficulty in swallowing. They may say that food 'catches' in their throat when they swallow. Another common complaint linked with Arnold-Chiari malformations is hoarseness **(Stovner, et al, 2006).**

People with Arnold-Chiari malformations may have visual problems, including blurred vision, double vision, or blind spots. There may be bobbing of the eyes **(Wolpert, et al, 2005).**

An Arnold-Chiari malformation is diagnosed with magnetic resonance imaging (MRI). An MRI uses magnetism and radio waves to produce a picture of the brain and show the crowding of the space between the brain and spinal cord that occurs with Arnold-Chiari malformations. In addition to an MRI, patients will also have a thorough neurologic examination **(Tokime, et al, 2008).**

Individuals who begin to experience symptoms from an Arnold-Chiari malformation are usually first seen by their primary care physician, who may send them on to a neurologist for further evaluation. If the patient is deemed to require surgery, a neurosurgeon will be consulted **(Walsh, et al, 2006).**

The recommended treatment for an Arnold-Chiari I malformation is surgery to relieve the pressure on the cerebellar area. During the surgery, the surgeon removes a small part of the bone at the base of skull. This enlarges and decompresses the posterior fossa. This opening is patched with a piece of natural tissue. In some people with Arnold-Chiari malformation, displaced brain tissue affects the flow of cerebrospinal fluid. Doctors may evaluate the flow of cerebrospinal fluid during surgery for Arnold-Chiari malformation. If they find that brain tissue is blocking the flow of cerebrospinal fluid, they will shrink the brain tissue during surgery **(Stovner, et al, 2006).**

Individuals who are recovering from surgery to repair an Arnold-Chiari malformation may require physical and/or occupational therapy as they try to regain strength and fine motor control in their arms and hands. A speech therapist may be helpful in improving both speech and swallowing **(Wolpert, et al, 2005).**

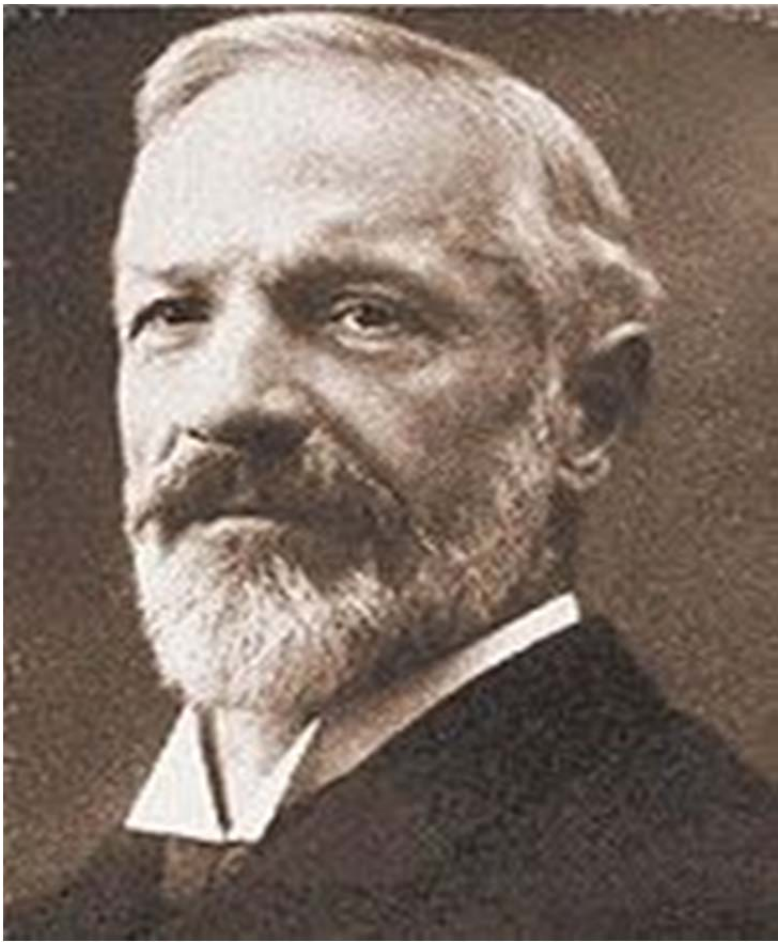
The National Institutes of Health are undertaking several research studies exploring aspects of Arnold-Chiari malformations. Efforts are being made to delineate a possible genetic defect leading to such malformations; studies are further exploring the anatomy and physiology of the malformations; and comparisons of the efficacy of various surgical treatments are being made **(Swanson and Fincher, 2008).**

Long-term prognosis for persons with Arnold-Chiari I malformations is excellent. Full recovery from surgery may take several months. During that time, patients may continue to experience some of the symptoms associated with Arnold-Chiari malformations **(Stovner, et al, 2006).**

Prognosis for Arnold-Chiari II a malformation depends on the severity of the myelomeningocele and will be equivalent to that of spina bifida **(Wolpert, et al, 2005).**

Historical Overview

Hans Chiari (1851 _1916) was born in Vienna, Austria (fig. 1) His father was the famous gynecologist J. B. V. L. Chiari and his brother was the rhino laryngologist Ottokar Chiari. He graduated from medical school in 1875 and became assistant to the Austrian pathologist Karl Rokitansky at the Institute of Pathology in Vienna **(Appleby et al, 2005)**.



(Fig. 1) Hans Chiari (1851 – 1916) ***(Georg,et al,2002)***