Will you spend five minutes to change a patient’s life?

That’s all it takes to learn what you
need to know about the disorder
that claimed our daughter

SHANNON’S HOPE

CHIARI MALFORMATION AWARENESS

SHANNON

CHIARI MALFORMA

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Austin, Te
(512)90

Shannon's Hope Foundatio

Dear Physician,

Like many people, we had never heard of Chiari malformation – until its complications claimed the life of our daughter, Shannon, in the summer of 2011. In this serious neurological disorder, the cerebellum descends out of the skull and crowds the spinal cord, causing severe headaches and other debilitating symptoms.

As a physician, you can play a critical part in fighting this terrible disease. Chiari malformation is notoriously difficult to diagnose, but awareness of the symptoms and other facts about it across the physician community will hopefully help more people get a proper diagnosis on a timely basis.

There is a wide range of common Chiari malformation symptoms, but specific symptoms and their severity vary, and an MRI is necessary to verify a diagnosis. Consequently, people often go for years without a proper diagnosis, delaying the opportunity to seek treatment. We experienced firsthand the frustration of the lack of information and awareness during the long period that the family struggled to find the correct diagnosis for Shannon. Please visit www.Shannons-Hope.org to learn more about her story.

Our goal is to raise awareness of Chiari malformation among physicians like you. To that end, Dr. K. Michael Webb, a neurosurgeon with Neuro Texas PLLC, Austin, Texas, has agreed to generously share his extensive knowledge about the disease. On the following pages, you'll find:

- Valuable facts from Dr. Webb about Chiari malformation
- Additional detailed information about symptoms, diagnosis, and treatment
- A personal appeal from another of your physician colleagues, Dr. Richard Parker

Please take five minutes to review this information. If we can help just one person get the proper diagnosis, all efforts will have been rewarded. Thank you for your time, consideration, and help in the effort to conquer Chiari.

Sincerely,

Prudence E. Hudson

Prudence Hudson

David Hudson
DHudson@Shannons-Hope.org

What you need to know about Chiari malformation

By K. Michael Webb, M.D.

Dr. Webb is a neurosurgeon with extensive experience in Chiari malformation diagnosis and treatment, including decompression surgery. Learn more about his practice at www.neurotexas.net.

What is Chiari malformation?

A Chiari Type I malformation occurs when the cerebellum protrudes through the opening in the skull where the spinal cord exits, which can cause increased pressure on the brain stem, spinal cord, or cerebellum. It occurs in roughly one out of 1,000 births, more commonly in females. Even though Chiari malformation is present at birth, symptoms generally do not develop until adolescence or adulthood, and patients can go undiagnosed for years.

What are the symptoms?

The symptoms of Chiari malformation vary from person to person, and can range from mild to severe or debilitating. Some people have no symptoms at all. The most common symptom is a severe headache, which is usually in the back of the head and can be brought on by straining, coughing, sneezing, and laughing. Another common symptom is neck pain which radiates across the shoulders and down the spine. Other symptoms include difficulty swallowing, trouble speaking or hoarseness, respiratory problems and sleep apnea, frequent urination and/or loss of bladder control, irritable bowel syndrome and/or lack of bowel control, weakness and stiffness in the arms and/or legs, numbness in the hands and/or feet, and vertigo and/or trouble balancing.

Chiari malformation can also occasionally lead to syringomyelia, a disorder in which a cyst forms within the spinal cord, which can compress and damage the spinal cord, resulting in weakness or stiffness in the arms, legs, and cause chronic, severe pain.

How is the condition diagnosed?

Anyone who consistently experiences any of the symptoms described here should receive a neurological examination, including a complete medical history and physical exam. The best diagnostic tool to date for detecting Chiari malformation is magnetic resonance imaging (MRI). Recent advances in MRI techniques, most notably the ability to measure the flow of cerebrospinal fluid (CSF), allow doctors to identify many cases that would otherwise go undiagnosed.

What is the treatment?

Treatment options vary according to the severity of the disease. Many people who have Chiari malformation experience no symptoms at all and therefore require no treatment. Patients who complain of mild symptoms can sometimes be effectively treated with medication. However, medication can only relieve the symptoms and does not correct the problem.

If the symptoms are severe and debilitating, affecting the patient's overall quality of life or neurologic function, surgery may be considered. Surgery generally involves removal of the bone over the Chiari malformation to create more room for the cerebellum and brain stem. Additionally, the lining of the spinal cord is usually expanded to further improve the flow of spinal fluid.

What are the patient's long-term prospects?

Though not everyone experiences the same severity of symptoms, living with Chiari malformation is a life-long struggle that can place a tremendous strain on patients and their families. With proper knowledge of the symptoms, diagnosis, and multidisciplinary treatment, Chiari malformation can be managed, allowing patients to achieve their best quality of life.

Most Common Symptoms

Symptom	Percentage
Headache	98%
Dizziness	84%
Difficulty Sleeping	72%
Weakness in Arms/Hands	69%
Neck Pain	67%
Numbness/ Tingling in Arm, Hands	62%
Fatigue	59%
Nausea	58%
Shortness of Breath	57%
Blurred Vision	57%
Tinnitus	56%
Difficulty Swallowing	54%
Leg Weakness	52%

Source: www.conquerchiari.org
C&S Patient Education Foundation

More about Chiari malformation
symptoms, diagnosis, and treatment

Definition and types of Chiari malformation

Chiari malformation (also known as Arnold-Chiari malformation, tonsillar herniation, tonsillar ectopia, or hindbrain herniation) is a congenital condition in which the posterior fossa, or lower part of the skull, is too small to accommodate the lower part of the brain, causing part of the cerebellum to protrude into and crowd the spinal column. The resulting compression of the brain stem and spinal cord is the main reason for most of the symptoms that are associated with Chiari malformation. Even a minimal amount of protrusion – as little as one millimeter – can result in enough compression to cause symptoms.

- Chiari Type I malformation: The lower part of the cerebellum extends into the foramen magnum. This type of Chiari malformation is present at birth but may not be recognized at the time. It is the only type of Chiari malformation that presents in adulthood.

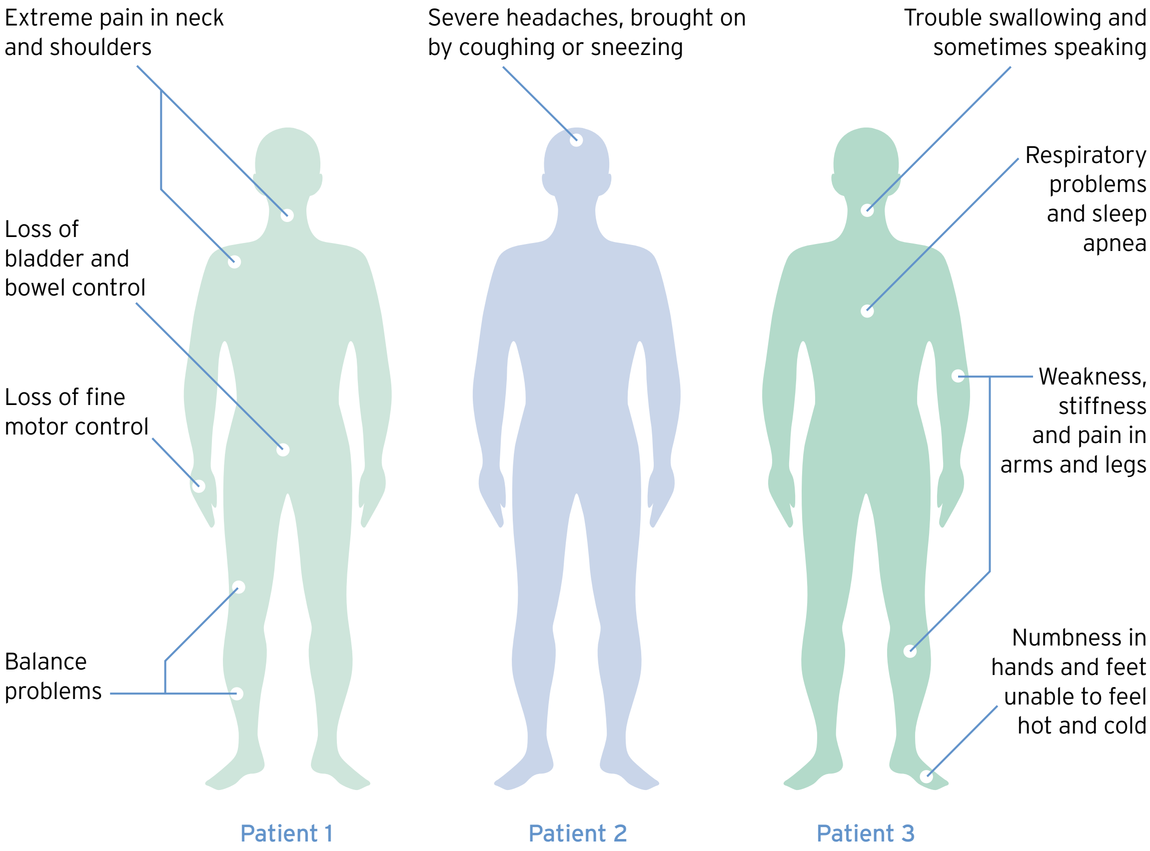
- Chiari Type II malformation: Both the cerebellum and brain stem tissue extend into the foramen magnum in this type of Chiari malformation. Type II is invariably associated with myelomeningocele, a severe form of spina bifida.

- Chiari Type III malformation: In this type of Chiari malformation, which is both rare and serious, the hind part of the brain protrudes through a defect in the skull. Like Type II, it is apparent at birth.

- Chiari Type 0 malformation: This is the same as a Chiari Type I malformation, except that the cerebellum does not extend into the foramen magnum by the standard five millimeters.

Patients with Chiari Type 0 malformation are often misdiagnosed because current diagnostic criteria do not accurately reflect the neurological condition underlying the malformation.

Every case is different



Extreme pain in neck and shoulders

Loss of bladder and bowel control

Loss of fine motor control

Balance problems

Severe headaches, brought on by coughing or sneezing

Trouble swallowing and sometimes speaking

Respiratory problems and sleep apnea

Weakness, stiffness and pain in arms and legs

Numbness in hands and feet unable to feel hot and cold

Patient 1

Patient 2

Patient 3

Source: www.conquerchiari.org, C&S Patient Education Foundation

Symptoms of Chiari malformation

Most people with Chiari malformation do not experience symptoms until adulthood, usually (although not exclusively) in their 20s or 30s. The specific symptoms experienced by a patient will vary, as will the severity of the symptoms. One patient may only have severe headaches brought on by coughing or sneezing, while another may present with trouble swallowing, respiratory problems, weakness in the arms and legs, and numbness in the hands and feet. The following describes the various symptoms.

Headache

This is by far the most common symptom. In one study (Mueller, Oro, 2004, as cited in www.conquerchiari.org), 98% of 265 patients reported suffering headaches. Headaches associated with Chiari malformation are typically described as causing a feeling of intense pressure. They often start in the back of the head and may also radiate to the area behind the eyes. They can be aggravated by activities such as straining, coughing, sneezing, or laughing, or by bending forward or looking up. These headaches may be mistaken for migraines. In fact, some experts believe there is some type of connection between Chiari malformation and migraine headaches.

Dizziness or vertigo

In the study cited above (Mueller, Oro), 84% of patients reported dizziness. Extending the neck may make the symptom worse.

Neck pain

Some Chiari patients report pressure-like neck pain that radiates down the spine and across the shoulders. Neck pain occurred in 67% of the patients in the Mueller, Oro study.

Other symptoms

More than half of the patients in the Mueller, Oro study reported the following additional symptoms:

- Difficulty sleeping
- Arm/hand weakness
- Arm/hand numbness/tingling
- Fatigue
- Nausea
- Shortness of breath
- Blurred vision
- Tinnitus
- Difficulty swallowing
- Leg weakness

Still other symptoms include auditory problems beyond tinnitus, such as a lowered ability to hear or an increased sensitivity to sound; visual symptoms including double vision, sensitivity to light, and blind spots; and vocal issues such as hoarseness or an inability to regulate the voice when shouting or singing. 95% of patients in the Mueller, Oro study reported experiencing at least five symptoms.

In the Mueller, Oro study, 20%-50% of patients specifically reported experiencing depression, body weakness, balance problems, memory problems, leg/foot numbness, hoarseness, chest pain, facial numbness, anxiety, slurred speech, arm pain, abdominal pain, and photophobia.

A small number of patients in the Mueller, Oro study (less than 20%) specifically reported experiencing tachycardia, trouble hearing, vomiting, double vision, vision loss, blackouts, apnea, vertigo, loss of peripheral vision, nystagmus, earache, snoring, thoracic pain, hypotension, waking up choking, leg pain, palpitations, hypertension, gag reflex, and face pain/tingling.

Causes of Chiari malformation symptoms
Symptoms associated with Chiari malformation are the result of compression of the cranial nerves, brain steam and cerebellum, as well as of disruption of the flow of CSF or elevation of CSF pressure in the skull. Nerve damage in the spine can also cause symptoms.

No one knows exactly why some people who are born with Chiari malformation experience symptoms while others never do. It's possible that head and neck trauma, or some medical conditions, may trigger or aggravate symptoms.

Research indicates that the amount of protrusion of the cerebellum into the spinal column has no relation to the occurrence or severity of symptoms. A patient with as little as one millimeter of protrusion may have symptoms, yet someone with a much larger degree of herniation may have no symptoms at all.

Diagnosing Chiari malformation

A complete medical history and physical examination, followed by a complete neurological evaluation to assess symptoms and neurological function, are required to begin the diagnostic process. Some patients may have problems with coordination, balance, eye movement, sensation, and strength. The reflexes may be overactive or underactive. Reflexes that are present only when the spinal cord is not functioning properly may also be detected.
It is also entirely possible that none of the preceding problems will be present and that the patient will have an entirely normal examination. This can make it extremely difficult to determine whether Chiari malformation is the problem. In that case, an experienced neurosurgeon's judgment is vital. If the neurological examination is abnormal, a neurosurgeon may detect a pattern of abnormal findings that make it possible to pinpoint the problem in the cervical spinal cord and brain stem.

MRI is the most valuable resource for diagnosing Chiari malformation. It provides detailed images about the anatomy of the areas at the base of the brain. The longtime standard for diagnosing Chiari malformation is a measurement of five millimeters of protrusion of the cerebellum into the spinal column; if this is detected, the doctor may be able to confidently make the diagnosis. However, in some cases, the measurement is less – and this can cause patients to go undiagnosed and untreated.

Advances in detecting Chiari malformation using MRI and other diagnostic tools have been reported by various sources. The Wisconsin Chiari Center (www.wichiaricenter.org), for example, reports having developed MRI imaging strategies designed to demonstrate brain stem compression more clearly. These strategies involve using three-dimensional imaging and cine imaging, which essentially makes a movie of the brain, to determine if the brain stem is being compressed. Others have reported on the use of spinal taps as a diagnostic tool (www.chiarione.org).

What causes Chiari symptoms?

Direct compression of the cranial nerves

Direct compression of the brainstem

Direct compression of the cerebellum

Disruption of the natural flow of CSF

Elevated CSF pressure in the skull/brain

Damage to
nerves in the spine

Source: www.conquerchiari.org, C&S Patient Education Foundation

Treating Chiari malformation

Some symptoms of Chiari malformation, such as headaches and dizziness, may be treated with medication. However, medication alone will not address the brain stem compression that is the underlying cause of Chiari malformation symptoms. When a patient's symptoms are serious enough to have a significant impact on the quality of life, it is best to consider decompression surgery.

The goal of surgery is to make more room for the cerebellum, in order to relieve compression of the brain stem and spinal cord and facilitate better CSF circulation. The surgery involves removing a small part of the bottom skull to correct the existing structure. The neurosurgeon may also use electrocautery to shrink the parts of the cerebellum that protrude into the spinal column. Another procedure involves removing part of the lamina of the spinal canal to relieve pressure on the spinal cord.

An appeal from our friend and your colleague, Dr. Richard Parker

Dear Colleague,

Too often, we as physicians are personally touched by the death of a loved one. Consequently, this tragic loss becomes a positive force in educating and enlightening people and physicians about an often rarely diagnosed and misunderstood illness.

Recently, my dearest friends, David and Prudence Hudson, lost a daughter to the complications of Chiari malformation. They have begun to honor their daughter, Shannon Hudson Terry, by starting a campaign of love and caring to conquer Chiari. The start has been a web site, www.shannons-hope.org, dedicated to Shannon and Chiari.

I am asking you to become involved in any capacity you wish. This would include advice as to how we should proceed, information to your patients, or a wider role with investment of your time and expertise.

There is a saying, "You can easily judge the character of a man by how he treats those who can do nothing for him." We cannot do anything for you, but you can do a great deal for these people who suffer from the effects of Chiari malformation.

Thank you,

Rich Parker

E. Richard Parker, M.D.
Austin Plastic Surgery Institute

The Mission of Shannon's Hope Foundation

Shannon's Hope Foundation is dedicated to improving awareness of Chiari malformation to better facilitate early diagnosis of this condition. Understanding the complexity of the disease is extremely critical to diagnosis and treatment. To that end, our objective is to provide the medical community with ongoing informational documentation about Chiari malformation. Shannon's diagnosis took five years before her decompression surgery; unfortunately, it was not successful. What might have been with an early diagnosis?

Shannon's Hope Foundation was created to help others avoid the debilitating and excruciating pain that can result from undiagnosed Chiari malformation. Awareness is the key to early diagnosis and treatment. Shannon's goal was lifestyle improvement for patients; achieving that goal depends largely upon the physician's willingness to recognize and treat this disease. Our long-term objective in this cause is to provide or assist with low-cost MRIs, so vital to accurate diagnosis.

Chiari malformation is not a rare disease, just rarely diagnosed on a timely basis. Please help us in fulfilling our mission to provide the information and understanding that are essential in the quest to conquer Chiari.

Shannon's Hope Foundation is a 501 (c) (3) non-profit organization.

Stephanie's Story of Chiari Survival

My name is Stephanie Barzellone. I am the daughter of Texas State Representative Jerry Madden. I am a wife, mother of four children, and I am also a Chiarian. My journey with Chiari malformation started on June 2, 2008, when I was injured in a work accident. The saga includes multiple visits and repeated examinations by a variety of physicians over a two year period.

- Three general practitioners with exams and X-Rays
- Three orthopedic surgeons with exams, X-Ray's and MRI's
- Two neurosurgeons with exams, X-Rays and MRI's
- Received brain decompression surgery for Type 1 Chiari malformation exactly 2 years and 5 days from the start of this frustrating and painful ordeal

The initial examination and X-Rays were taken at the Family Medical Center, revealing no fractures. I was given a neck brace and ibuprofen, and released. Still suffering with severe neck and head pain, the next day I went to two of my personal doctors. One prescribed an anti-inflammatory and the other recommended an MRI. Due to the fact I was injured at work, Workers' Compensation took over and sent me to two of their orthopedic surgeons. Thirteen days after the accident I saw the two specialists. One doctor recommended physical therapy and an MRI. The second doctor took over treatment and, two months after the accident, finally ordered the MRI. Chiari malformation was not a part of the diagnosis.

Because of ongoing headaches, nausea, vomiting, and numbness in my right arm, I asked to see a neurosurgeon. Instead, I was sent to a third orthopedic surgeon, who on October 7, 2008, re-read the MRI and found the Chiari malformation. Due to Workers' Compensation, it took a while to get additional treatment. I did not receive the second MRI and EMG testing until May 2009. Based on the results, it was recommended that I see a neurosurgeon for a CSF flow study.

On October 7, 2009, I was allowed to consult a neurosurgeon. He ordered the 3TESLA MRI with flow study and new X-rays. On November 18, 2009, the flow study results were evaluated and decompression surgery was prescribed. Insurance required a second opinion, but I didn't see the second neurosurgeon until April 2010. He confirmed the diagnosis and recommendation for immediate surgery. On June 7, 2010, I received brain decompression surgery for Chiari malformation Type 1.

Today is December 15, 2011. I am one year, six months, and eight days from my surgery. I have spent the last year and a half in recovery. It has been the toughest battle of my life but I am here and I am alive. Looking back on this journey, I can only dream of what might have been with early detection. There are so many lessons to be learned from my story and changes that should be made in the system. My goal is to improve Chiari malformation awareness and education to better facilitate early diagnosis for the thousands of people affected by this disease. One day, with your help, we will conquer Chiari.

SHANNON’S HOPE

CHIARI MALFORMATION AWARENESS

www.shannons-hope.org