

# Chiari FAQ's

*These FAQ's are for informational purposes only and in no way represent an attempt to provide medical advice. This information may or may not apply to your case and anyone with a question or concern about their health is strongly encouraged to consult with a medical professional.*

## **1. What is a Chiari Malformation?**

Chiari Malformation Type I (CM) is a neurological disorder where part of the brain, the cerebellum (or more specifically the cerebellar tonsils), descends out of the skull into the spinal area. This results in compression of parts of the brain and spinal cord, and disrupts the normal flow of cerebrospinal fluid (a clear fluid which bathes the brain and spinal cord).

## **2. Is Arnold-Chiari different from Chiari?**

Not everyone uses the same terminology when describing Chiari. Some people use Arnold-Chiari (ACM) interchangeably with Chiari. Others only use Arnold-Chiari to refer to Chiari Type II which involves more of the brain descending out of the skull, is predominantly diagnosed in children, and is commonly associated with Spina Bifida. Other terms for Chiari include tonsillar ectopia and hindbrain herniation, meaning the cerebellar tonsils are out of position.

## **3. What are the symptoms?**

Because Chiari involves the nervous system, symptoms can be numerous and varied. In fact, one large study showed that the vast majority of Chiari patients reported 5 or more symptoms, and 49 distinct symptoms were reported by 2 or more patients. Despite this variety, the most common Chiari symptom, and the hallmark of the disease, is a headache. Usually, the Chiari headache is described as an intense pressure in the back of the head and is brought on, or aggravated, by exercise, straining, coughing, sneezing, laughing, bending over, or similar activities. Other common symptoms include balance problems and fullness in the ears. In very young children, trouble swallowing is one of the most frequent symptoms. When thinking about symptoms, it is also important to keep in mind that once a person's health is compromised in one way (with Chiari for example), secondary problems are more likely to develop, especially if a person is in chronic pain. Research has shown that people with chronic pain are much more likely to develop other chronic conditions, so not every symptom may be a direct result of a Chiari Malformation.

## **4. Does the size of the malformation matter?**

Traditionally, Chiari Malformation has been defined as the cerebellar tonsils descending more than 3-5mm out of the skull. However, research has shown there is no real correlation between the amount of descent (or herniation) and clinical symptoms. Some people with herniations of less than 3mm are extremely symptomatic and some people with quite large herniations are symptom free. Because of this, doctors are now focusing on whether the cerebellar tonsils block the normal flow of cerebrospinal fluid (CSF). The current theory is that disruption of CSF flow is a more important measure than the size of the herniation.

## **5. How is a Chiari Malformation diagnosed?**

An MRI (Magnetic Resonance Imaging - a non-invasive test which uses a large magnet to create a picture of internal organs) can clearly show if the cerebellar tonsils are out of position. However, since the definition of Chiari is changing, most doctors will use a combination of reported symptoms, a neurological exam, MRI results, and their experience and judgment to determine if a person has Chiari. Unfortunately, there is no single, objective test which can clearly say that someone has a Chiari malformation which is causing problems.

## **6. What is a cine MRI?**

Cine MRI is a type of MRI where the machine is programmed to measure the flow of cerebrospinal fluid (CSF). Doctors use this to see if the cerebellar tonsils are blocking the normal flow of CSF from the brain to the spinal area and back. While many doctors now consider cine MRI to be a routine test in diagnosing Chiari, some experts question it's usefulness and are reserving judgment.

## **7. What is a borderline Chiari?**

Since Chiari was traditionally defined based on the size of tonsillar herniation, a borderline Chiari can refer to when the cerebellar tonsils are descended only a couple of millimeters out of the skull. Alternatively, borderline Chiari can refer to someone with mild symptoms which may not be directly attributable to Chiari.

## **8. How did I get this condition?**

Chiari was originally thought to be a congenital condition - meaning you are born with it. While this may be true for many people, published case studies have also demonstrated that Chiari can be acquired and even reverse itself if the source of the problem is removed. It is not known how many cases are congenital and how many are acquired. Complicating the situation is that for reasons that aren't clear, some people develop symptoms as children, and some people develop symptoms as adults. What triggers symptoms is not fully understood.

## **9. I don't have any symptoms, but an MRI shows a malformation. What does this mean?**

This is sometimes referred to as an incidental finding. Someone has an MRI for an unrelated reason, but it shows a Chiari malformation - meaning the cerebellar tonsils are descended - yet the person has no Chiari type symptoms. As MRI's become more common, this is happening more frequently and is one reason that diagnosing Chiari can be difficult and can not be based on an MRI alone.

## **10. Does Chiari run in families?**

An ongoing study at Duke University has identified more than 100 families where two or more members are affected by Chiari. This implies that for some cases there is a genetic basis for Chiari. It is not known, however, what percent of cases may have a genetic component. In other words, this does not mean that the family of everyone with Chiari is carrying a Chiari gene.

## **11. Will my children get it? Is there a genetic test to see if someone has Chiari?**

A Chiari gene has not yet been identified, so there is currently no genetic test.

## **12. How is Chiari treated?**

If the symptoms aren't severe, doctors may recommend just monitoring the situation with regular MRI's and treating the symptoms individually. However, if symptoms are interfering with quality of life, are getting worse, or if the nervous system is being impaired, doctors may recommend surgery. The most common surgical treatment, performed by a neurosurgeon, is known as decompression surgery (see details in Question 14). An alternative surgery involves placing a shunt (a tube like device) to channel the flow of CSF and relieve pressure

## **13. How do I know whether to have surgery?**

The decision whether to have surgery is up to each individual and their doctor. Some of the factors that are considered are the severity of symptoms, whether the symptoms are getting worse, whether the nervous system is being compromised, whether there are any complicating issues, and the surgeon's own experience and judgment. Unfortunately, there is no single, objective measure to say whether someone should have surgery and many patients will find that different doctors may have different opinions. Some doctors are more aggressive in their treatment approach and some are more conservative. A recent survey about when to recommend surgery showed that there was general agreement among surgeons in the extreme cases - no or mild symptoms, don't operate; severe, progressive symptoms or syringomyelia, operate - but there was little agreement in the middle. In one of the survey's hypothetical cases, the surgeons were split almost evenly down the middle on whether to operate or not.

## **14. What is the surgery like?**

Decompression surgery is a general term used to refer to any of a number of variations on the same basic procedure. The goal of the surgery is to create more space around the cerebellar tonsils and restore the normal flow of CSF. The procedure involves removing a piece of the skull in the back of the head near the bottom (craniectomy). Often part of the top one or two vertebra are also removed (laminectomy). At this point, depending on the individual case and doctor, some doctors will also open the covering of the brain, the dura, and sew a patch in to make it larger (duraplasty). There are many variations in how the surgery is performed, including (but not limited to) how much bone to remove, whether to open the dura, what type of material to use for a dural patch, whether to shrink or remove the cerebellar tonsils, and whether to replace the missing piece of skull with anything. Unfortunately, there is no consensus, and no strong evidence, on which technique(s) is the best. Because of this, it is important for patients to understand specifically what their surgeon will be doing and why. The procedure itself lasts several hours and most people will spend a night in the ICU and an additional couple of days in the hospital.

## **15. Is the surgery always successful?**

As with any surgery, the chance of success depends on the individual case, so each person should ask their doctor what their chance of having a successful surgery is. It should be noted that success can mean different things to different people, so it is best to ask specific questions such as what are the odds I will be symptom free; what are the odds I will be mostly better; and what are the odds I will get worse.

Unfortunately, there is not a lot of strong surgical outcome research, but there are enough reports to get a general idea of the overall success rates. For patients with just Chiari (no syringomyelia), up to 50% become symptom free after surgery, with another 10%-30% improving significantly. On the flip side, for 10%-20%, the surgery will be a failure and they will likely require additional surgeries. Keep in mind these are not scientific numbers and each patient should discuss their own chance of success with their doctor.

## **16. What are the possible complications of surgery?**

This is another question that is important for every patient to ask their doctor so that they fully understand the risks and potential outcomes of surgery. Many of the complications of decompression surgery have to do with opening the dura and research has shown that opening the dura does increase the complication rate. There is a risk of infection and sometimes the patch that is sewn in leaks or becomes scarred. A more serious complication - not necessarily related to opening the dura - occurs when the brain slumps further into the spinal area after the surgery.

## **17. What will happen to me if I don't have surgery?**

The natural progression of Chiari - as doctors call it - varies from person to person and is not well understood. For example, why do some people develop symptoms in their 30's while others have symptoms their whole life? For many people with no or mild symptoms, the symptoms will not get worse and surgery will not be necessary. However, there are also anecdotal reports of symptoms becoming rapidly worse, sometimes after a sneeze or a fall. If a patient does not have surgery, many doctors will recommend monitoring the situation with routine MRI's and neurological exams.

## **18. How long will it take to recover from surgery?**

As to be expected, recovery will vary from person to person and will depend in part on a person's overall health and fitness before the surgery. Barring any complications, some people recover from a successful surgery in a few weeks, others take a few months, and others may take more than a year. Your doctor may suggest a physical rehabilitation program to regain strength and flexibility in your neck and may refer you to a physiatrist - a doctor of physical medicine and rehabilitation. One factor that people sometimes overlook during recovery is that if they were inactive due to severe symptoms for a long period of time prior to surgery, they will need time to regain a general level of strength and conditioning.

## **19. How can I find a doctor with a lot of Chiari experience?**

The American Association of Neurological Surgeons (AANS) does not recognize Chiari as a sub-specialty. This, combined with liability issues and the difficulty in establishing expert-level criteria (what does it take to qualify as an expert?) make it difficult to put together a list of Chiari experts. Each person must find a doctor they are comfortable with. Some people like to see university based researchers, some would prefer a regular neurosurgeon; some are willing to travel for surgery, others aren't; some want a surgeon they can relate to, others think surgical skill is more important. When trying to find a doctor, some things to consider are how many Chiari surgeries they do a year, how many total surgeries they do a year, are they up to speed on the latest thinking on Chiari, how they relate to patients, and what type of reputation they have among patients and the medical community (this is by no means comprehensive). There is no right answer to these questions; they are just intended as a way for a patient to feel comfortable with their doctor. One way to find a doctor is to ask

around. Ask people in your community, ask any medical professionals you know, or go on the internet to find what you are looking for.

## **20. I had surgery, but I'm still in a lot of pain. What can I do?**

One possibility is to see a pain specialist. A certified pain doctor will perform a thorough examination to determine the exact cause of your pain and may recommend therapies such as acupuncture, trigger point injections, over the counter medications, or prescription medications. Unfortunately, neuropathic pain - pain caused by damage to a nerve - can be very difficult to treat. Anti-seizure drugs, like Neurontin, work for some people but can have strong side effects. Many Chiari patients have found that they must try different things and see what works best for them.

## **21. Am I eligible for disability?**

Many people with Chiari have qualified for government disability. Some people have been able to get disability easily, while others have had to fight for it.

## **22. How many people have Chiari?**

There is no exact answer to this, because a rigorous study to determine this has not been performed. Once thought to be rare, the increased use of MRI's has shown that Chiari is much more common than originally believed. Confusing the issue is the question of how you define Chiari. Many people may have cerebellar tonsils that descend out of the skull, but they have no symptoms and probably never will. Studies have shown the incidence of this tonsillar ectopia may be as high as .5%-.7% of the general population. However, this does not mean that all these people have Chiari. Estimates for the number of people with true Chiari range as high as 500,000 in the United States. A more conservative estimate of 300,000 would mean that 1 in 1,000 people have Chiari, or 0.1% of the population.

## **23. I'd like to talk with other people who have this. Is there a support group?**

Conquer Chiari has built an on-line meeting place for people in the Chiari community to confidentially connect. It is not a message board, but rather a way to meet people like yourself, exchange contact information, and develop supporting friendships. The meeting place can be found at the On-line Meeting Place.

You can visit our Support Group page and look for a group in your area or let others know you are interested in forming a local support group.

Facebook is a great place to meet others with Chiari, check out the Conquer Chiari Facebook page to converse with other chiari patients, and for the latest information in the Chiari Community.

Additionally, the Pediatric Chiari Facebook page is a great place for parents to connect with other parents and talk about whats going on with their child.



**For More Information Visit: [www.conquerchiari.org](http://www.conquerchiari.org)**