Chiari Academy Video Transcription
Chiari Bootcamp- Treatment & Outcomes

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[Music]

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In this lesson we will cover deciding whether to have surgery, what decompression surgery entails,

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Surgical and non-surgical outcomes, the surgical experience and residual symptoms. Unfortunately

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when it comes to Chiari there are not a lot of treatment options the only real treatment is

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surgery performed by a neurosurgeon to relieve the pressure caused by the herniated tonsils.

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Because of this deciding on treatment boils down to deciding on whether to have surgery or not

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by default then absent surgery the only other option is to wait and see and see what happens

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and manage individual symptoms as best you can. While it seems like a simple situation with only

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one decision to make the reality is that in some cases the decision of whether to have surgery or

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not can become quite complicated. This is partly because the surgical decision involves both the

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patient or the parents in the case of a child and the doctor both of whom must agree agree

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to proceed with surgery. In making this decision each party will look at it from their own point of

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view and consider both objective and subjective factors the patient must decide how much their

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symptoms are impacting their quality of life, how they feel about the risk of the procedure,

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and how much faith they have in their doctor. Some people are eager to have surgery while for others

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the thought of it is too overwhelming to consider even in the face of severe symptoms. Meanwhile the

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doctor will decide based on the patient's reported symptoms what the Imaging shows whether there are

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clear signs the nervous system is being affected and their own experience in treating Chiari. It

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may be surprising but when evaluating Chiari neurosurgeons will often recommend surgery less

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than half of the time. Unfortunately for patients research has shown that surgeons often disagree

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on when surgery should be recommended so while some cases are straightforward many are not.

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From a practical point of view this means that if a patient or family seeks additional opinions they

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need to be prepared to hear different things from different doctors. In those situations the burden

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shifts to the patient in deciding which opinion they feel the most comfortable with. Next let's

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look at what the surgery entails and then we will talk about the surgical experience for

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patients. Chiari surgery is often referred to as decompression surgery decompression is shorthand

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for posterior fossa decompression which is the technical name for Chiari surgery. The posterior

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fossa is the region of the skull where the cerebellum is situated and decompression

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refers to making more space around the herniated cerebellar tonsils. Decompression surgery has two

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major goals the first is to create more space around the herniated tonsils so that neural

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tissue is not being squeezed and the second is to restore the natural flow of cerebrospinal fluid.

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While in general this is accomplished in several steps it is important to note that there are

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many variations some minor and some major we will highlight some of the more common major variations

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but before undergoing surgery patients and parents should make sure they have a clear understanding

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from their surgeon of what specifically will be done and why. The first part of the surgery

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involves removing bone from the skull and spine. After the patient is prepped for surgery they are

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placed on the operating table and their head is secured in a clamp. Next an incision is made down

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the center of the back of the skull from near the top down to the top area of the neck. Once the

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skull or Cranium is exposed a piece of it near the bottom is removed this is called a craniectomy.

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The amount of bone removed may vary but for an adult it is usually between three to four

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centimeters in length in addition the surgeon will likely also remove a piece of the top vertebra in

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what's called a laminectomy. If the herniation is large part of the second vertebra may be

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removed as well in some cases especially among children this is all that will be done this type

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of surgery is called a bone-only decompression. The advantages and disadvantages of bone-only

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decompressions will be reviewed a little later but for now let's assume the surgery will continue.

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With the bone removed the dura or covering of the brain and spinal cord is exposed the dura is cut

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and pulled back in an upside down triangle shape exposing where the spinal fluid flows and also the

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cerebellar tonsils themselves. This allows the surgeon to look for and remove any adhesions or

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scars that are interfering with the flow of the spinal fluid. In addition some surgeons

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will choose to reduce the size of the tonsils themselves by cauterizing them. Next in what is

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called a duroplasty a patch is sewn over the Dural opening essentially expanding the space underneath

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and providing more room for the tonsils and spinal fluid. The specific material that is used for the

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patch again varies from surgeon to surgeon some surgeons like to use tissue taken from the patient

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while others will use one of several commercial Dural substitute products. Then before closing

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everything up some surgeons will attach a special metal plate to replace the cranial bone that was

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removed and give the neck muscles something to attach to other than the dura. Finally the

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surgical side is closed in layers and stapled shut down the initial incision this gives the

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appearance of a zipper on the patient's skull and has led to the term zipperhead being used

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among some in the Chiari Community to refer to patients who have undergone surgery. While

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decompression surgery is the standard treatment for Chiari some patients may face other procedures

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as well according to the Chiari 1000 13 percent of patients both children and adults also had a

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shunt placed in the brain to divert spinal fluid a similar percentage had stabilization of the

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cervical spine and nearly 5 percent underwent a tethered cord release. In all more than one

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in four surgical patients reported having more than one Chiari related procedure and 15 percent

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reported having more than two. As with any procedure Chiari decompression surgery involves

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risks and complications can and do occur therefore it is very important that patients and parents

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understand the risks involved before deciding to have surgery. How often complications occur with

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decompression surgery isn't entirely clear and depends somewhat on the source of the data when

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surgeons publish papers based on their own case series, they often report complication rates of

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five percent or less and a database maintained by the American College of Surgeons showed a similar

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complication rate for 1500 pediatric surgical cases. However studies based on Hospital records

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tend to show higher complication rates. One such study found that of 900 pediatric Chiari

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surgeries across three states there was a 90-day complication rate of nearly 13 percent similarly a

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10-year study of over 13 000 pediatric cases found a complication rate of over 8 percent for adults.

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Studies have found the complication rate can be as high as 18 percent and that nearly 1 in 10 adults

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are readmitted to the hospital within 30 days the most common complications are infections and CSF

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leaks infections are a problem for any Hospital procedure but CSF leaks arise because it can be

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difficult to get a watertight seal when placing the Dural patch. One of the more puzzling and

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troubling complications is hydrocephalus or excess fluid in the brain for unknown reasons this occurs

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after about five percent of Chiari surgeries in both children and adults. Unfortunately in these

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cases four out of five patients will require a permanent shunt to be placed in the brain

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to divert the extra fluid. Now that we have a picture of some of the potential complications

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let's Circle back to the question of opening the dura during surgery. As mentioned in recent

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years some surgeons have explored performing decompressions where the dura isn't opened or

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at least not fully opened. While there has been extensive research comparing the two approaches

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a definitive answer has not emerged. Rather it appears that there are pros and cons to each

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specifically not opening the dura dramatically reduces the chance of surgical complications and

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results in shorter hospital stays and less pain. However there is a an increased risk of needing a

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second operation where the Dura will likely be open to achieve good results on the other hand

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opening the dura increases the risk of surgical complications and results in longer hospital stays

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but is also more likely to be successful the first time. Therefore while bone only decompressions

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have become more popular in pediatric cases they are not as commonly used for adults. Now that we

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have covered the two treatment options surgery or no surgery let's look at how patients do.

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With each for those In the no surgery or wait and see group one way to assess outcomes is to

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see how often surgery is required in the future. For Pediatric cases Studies have shown that in the

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medium term meaning several years less than 10 percent of conservatively treated children end

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up requiring surgery. In addition for children who do not have symptoms when the herniation is

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identified only five percent develop any symptoms or neurological deficits. Over time there are two

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important caveats to this: First no study has has followed incidentally diagnosed or conservatively

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treated children into adulthood so it is possible that some of these cases become symptomatic as

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adults. The second is that parents should be aware that when children are monitored with regular MRIs

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it is very common for the amount of herniation to change from year to year. While this is valuable

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data for Pediatric cases less data is available on conservatively treated adults. However one study

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that involved both children and adults treated conservatively found that the children did much

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better than the adults with 95 percent of the kids remaining stable compared to 64 percent

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of the adults. For surgical outcomes there are several options for measuring outcomes the most

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common although perhaps not the most useful for patients is the general clinical impression of

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the surgeon this is often reported as improved no change or Worse. There are many published

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studies like this and in general surgeons report Improvement 80 to 85 percent of the time for both

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children and adults but what does improved mean a couple of Studies have gone further and added the

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category of resolved to their reports based on very limited numbers close to half of children

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and 20 percent of adults experienced complete resolution of their symptoms after surgery.

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A second way to measure surgical outcomes is by using any number of health-related scales

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including the Chicago Chiari outcome scale which as the name implies was designed specifically for

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Chiari results from these types of studies are generally in line with the clinical impression

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type findings in that about 80 percent of patients will score as improved or having a good outcome.

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Not surprisingly they have also shown that in general kids have better outcomes than

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adults a third way to measure outcomes is to ask patients directly how the surgery impacted their

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symptoms. Patient reported studies including the Chiari 1000 show that 40 to 45 percent of

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adult patients report a significant Improvement in symptoms and another 20 to 30 percent report

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as slight Improvement. While these results are not quite as good as when doctors report outcomes the

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same studies indicate that knowing the outcome in advance 80 to 90 percent of patients would

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choose to have the surgery again. Finally we can look at symptom-specific Improvement while Chiari

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patients often suffer from many symptoms research has clearly demonstrated that headaches especially

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cough or valsalva headaches are the most likely symptom to improve with surgery decompression

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surgery is also effective at reducing the size of or eliminating syrinxes where fluid collects in

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the tissue of the spine itself this is important because syrinx can lead to nerve damage and even

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paralysis if not treated. Decompression surgery has also proven to be effective in improving sleep

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apnea however for many Chiari related symptoms such as visual disturbances and balance issues

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the results are mixed with improvements only seen about half the time or less. One common question

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that people have is whether it is possible to predict who will do well with surgery and who

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won't. While many research researchers have tried to find an objective way to accomplish

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this unfortunately at this point in time it is not possible however certain factors have been

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shown to negatively affect outcomes. As previously mentioned children tend to do better than adults

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in terms of symptom improvement in addition people with certain related conditions such as

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syringomyelia or ehlers-danlos syndrome tend to have poorer outcomes than patients with simpler

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cases. Time before diagnosis appears to be a very important factor as well with adults who

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experience symptoms for more than two years before surgery having significantly poorer outcomes.

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Finally surgical skill is also important as studies have shown that outcomes even within the

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same practice vary from surgeon to surgeon. These are just some of the factors that have been shown

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to be related to surgical outcome but there are likely many more as is clear. In looking at the

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outcomes data sometimes surgery fails to improve symptoms at all. This can happen for a number of

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reasons one possibility is that the surgery itself did not provide enough room meaning not enough

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bone was removed or the dura wasn't opened failure can also occur due to surgical complications such

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as issues with the Dural patch or the development of hydrocephalus another possibility is that the

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the patient's primary symptoms were actually due to something other than Chiari. Regardless

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of the reason for the initial failure up to 15 percent of Chiari patients will undergo a

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second decompression. Now that we've reviewed the technical aspects of Chiari surgery let's look at

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what the experience is like for patients. While the experience will vary from person to person

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based on what specifically is done, whether there are surgical complications and what the

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patient's overall health is going into surgery, it is important to realize that Chiari decompression

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is major surgery performed under general anesthesia. Before the surgery there will likely

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be pre-operative testing and of course the hair on the back of the head is shaved where the surgeon

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will cut. In general the procedure itself can take a few hours and patients will likely spend at

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least one night in the Intensive Care Unit where it can be quite shocking for family members to

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see their loved one. After patients are moved to a regular room they will likely stay in the hospital

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for another couple of days. For example one study found that 75 percent of of pediatric patients

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were discharged by day four. However another study found that one in four adults visit the

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ER at least once within 30 days of discharge most commonly for headaches or CSF related issues. So

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it is important not to leave too soon. A detailed discussion of recovery is beyond the scope of this

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lesson but while most people can recover from the surgery itself in a matter of weeks or months it

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can take years for the nervous system to recover from the effects of Chiari itself and while a lot

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of children may bounce back fully research has shown that most adults will not. Specifically,

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pain, depression, anxiety and neck related disability are common for adult patients

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even after what may be considered a successful surgery. For example Conquer Chiari research has

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shown that 80 percent of adult women with Chiari experience at least moderate levels of neck

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related disability whether they have had surgery or not. This means that the vast majority of adult

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patients struggle with one or more aspects of daily living. Something else patients should be

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aware of is that symptoms can come back over time. Good data on this subject doesn't exist for kids

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but among adults a large German study found that 10 years after surgery 15 percent of the patients

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had experienced neurological deterioration. To summarize the primary treatment for Chiari is

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posterior fossa decompression surgery. Many surgeons disagree on when surgery should be

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recommended. The most common complications after surgery are infections and CSF leaks. While

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bone-only decompressions are popular in pediatric cases they are not as commonly used for adults. In

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general symptoms will improve with surgery about 80 percent of the time. Valsalva headaches are the

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most likely symptom to improve with surgery. At this time it is not possible to predict who will

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and who will not do well with surgery. While most people can recover from surgery in a matter of

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weeks or months it can take years for the nervous system to recover. Pain, depression, anxiety,

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and neck related disability are common for adult patients even after a successful surgery.