Chiari Decompression Surgery

Overview
Chiari decompression surgery removes bone at the back of the skull to widen the foramen magnum and create space for the brain. The dura overlying the herniated tonsils is opened and a patch is sewn to expand the space, similar to letting out the waistband on a pair of pants. The goals of surgery are to control the progression of symptoms, relieve compression, and restore the normal flow of cerebrospinal fluid (CSF). The surgery takes about 2 to 3 hours and recovery in the hospital usually lasts 2 to 4 days.

What is Chiari decompression?
Posterior fossa decompression is a surgical procedure that removes bone at the back of the skull and spine to widen the space for the tonsils and brainstem (Fig. 1 and 2).

Many patients ask about minimally invasive or endoscopic surgery. Minimally invasive can mean different things: shorter skin and muscle incision, no dura opening, no shrinkage of the tonsils, or use of ultrasound and endoscopes. Despite what the words “minimally invasive” suggest, the amount of bone removal needed to effectively restore normal CSF flow depends on the individual patient’s anatomy and size of Chiari. The amount of bone removal should be the same in any procedure, endoscopic or standard “open” technique. It’s also important to understand that some minimally invasive techniques used for children (whose skulls are still growing) may or may not be appropriate for adults.

Spinal fusion may be performed in addition to posterior fossa decompression surgery in certain patients with spine instability due to scoliosis, Ehler-Danlos syndrome, or other bone abnormality. Rods and screws are inserted to structurally reinforce the skull and neck vertebrae.

Who is a candidate?
You may be a candidate for surgery if you have:

- An abnormal collection of CSF in the spinal cord called a syrinx.
- A Chiari malformation obstructing CSF flow (confirmed by cine MRI) and is causing severe or worsening symptoms.
What happens before surgery?
During the office visit, the neurosurgeon will explain the procedure, its risks and benefits, and answer any questions. Next, you will sign consent forms and complete paperwork to inform the surgeon about your medical history (i.e., allergies, medicines, vitamins, bleeding history, anesthesia reactions, prior surgeries). Discuss all medications (prescription, over-the-counter, and herbal supplements) that you are taking with your healthcare provider. Some medications will need to be continued or stopped the day of surgery. You will be scheduled for presurgical tests (e.g., a blood test, electrocardiogram, chest X-ray, and CT scan) several days before surgery.

Stop taking all non-steroidal anti-inflammatory medicines (Naprosyn, Advil, Motrin, Nuprin, Aleve) and blood thinners (coumadin, Plavix, aspirin) 1 week before surgery. Stop smoking and chewing tobacco 1 week before and 2 weeks after surgery because these activities can cause bleeding problems. Wash your hair with Hibiclens® (chlorhexidine) antiseptic soap for 3 consecutive days before surgery. No food or drink is permitted past midnight the night before surgery.

What happens during surgery?
Step 1: prepare the patient
You will lie on the operating table and be given anesthesia. Once you are asleep, your head will be placed in a 3-pin skull-fixation device that attaches to the table and holds your head in position during surgery. An inch wide strip of hair is shaved along the planned incision. The scalp is prepped with an antiseptic.

Step 2: make a skin incision
A skin incision is made down the middle through the neck muscles so that the surgeon can see the skull and top of the spine. The skin incision is about 3 inches long (Fig. 3). The skin and muscles are lifted off the bone and folded back.

Step 3: remove bone
The surgeon removes a small section of skull at the back of your head (suboccipital craniectomy). In some cases the bony arch of the C1 vertebra may be removed (laminectomy). These steps expose the protective covering of the brain and spinal cord called the dura (Fig. 4). Bone removal relieves compression of the tonsils.

Morning of surgery
• Shower using antibacterial soap. Dress in freshly washed, loose-fitting clothing.
• Wear flat-heeled shoes with closed backs.
• If you have instructions to take regular medication the morning of surgery, do so with small sips of water.
• Remove make-up, hairpins, contacts, body piercings, nail polish, etc.
• Leave all valuables and jewelry at home (including wedding bands).
• Bring a list of medications with dosages and the times of day usually taken.
• Bring a list of allergies to medication or foods.
• Bring your CPAP machine to the hospital if you have one.

Arrive at the hospital 2 hours before your scheduled surgery time to complete the necessary paperwork and work-ups. The nurse will explain the preoperative process and discuss any questions you may have. An anesthesiologist will talk with you to explain the effects of anesthesia and its risks. An intravenous (IV) line will be placed in your arm before transport to the operating room.
Step 4: open the dura
Next, the surgeon opens the dura to view the tonsils and cisterna magna (Fig. 5). Some surgeons perform a Doppler ultrasound study during surgery to determine if opening the dura is necessary. Sometimes bone removal alone may restore normal CSF flow.

Step 5: reduce the tonsils (optional)
Depending on the size of herniation, the stretched and damaged tonsils may be shrunk with electrocautery. This shrinkage ensures that there is no blockage of CSF flow out of the 4th ventricle.

Step 6: attach dura patch
A patch of synthetic material or the patient’s own pericranium (a piece of deep scalp tissue just outside the skull) is sutured into place (Fig. 6). This patch enlarges the dura and the space around the tonsils. The dural patch is sutured in a watertight fashion. The suture line is covered with a dural sealant to prevent CSF leak (Fig. 7).

Step 7: close the incision
The strong neck muscles and skin are sutured together. A dressing is placed over the incision.

What happens after surgery?
You will wake up in the recovery area. Your throat may feel sore from the tube inserted to assist your breathing during surgery. Once awake, you’ll be moved to your room. Your blood pressure, heart rate, and breathing will be monitored. If you feel nausea or headache after surgery, medication can be given. When your condition stabilizes, you will be discharged in the care of family or a caregiver, usually 1 or 2 days after surgery. Discharge instructions for home:

Discomfort
- After surgery, pain is managed with narcotic medication. Narcotics can be addictive and are used for a limited period of time. Thereafter, pain is managed with acetaminophen (e.g., Tylenol) and nonsteroidal anti-inflammatory drugs (NSAIDs) (e.g., aspirin; ibuprofen, Advil, Motrin, Nuprin; naproxen sodium, Aleve.
- Narcotics can also cause constipation. Drink lots of water and eat high-fiber foods. Stool softeners and laxatives such as Dulcolax, Senokot, Colace, and Milk of Magnesia are available without a prescription.
- Ice packs for 20 minutes 3 times a day can help relieve neck and shoulder pain and muscle spasms. Muscle relaxants may be prescribed.

Restrictions
- Avoid activities that increase pressure in the head:
  - Bending over, with head low
  - Straining, bearing down when having a bowel movement; avoid constipation
  - Prolonged coughing; hold your face to sneeze
  - Nodding in the “yes” position

Figure 5. The dura is opened.
Figure 6. A patch is sewn to the dura to enlarge the CSF space.
Figure 7. A dural sealant applied to the suture line prevents CSF leak.
• Do not drive until after your follow-up appointment.
• Do not lift anything heavier than 5 pounds for 2 weeks after surgery.
• No strenuous activity for the next 2 weeks including yard work, housework and sex.
• Do not drink alcohol for 2 weeks after surgery or while you are taking narcotic medication.

Activity
• Begin balance exercises and neck stretches as instructed.
• Get up and walk 5-10 minutes every 3-4 hours. Gradually increase your walking time, as you are able. Avoid getting over heated.

Bathing/Incision Care
• Shower and wash hair with mild shampoo after surgery unless otherwise directed.
• Do not submerge or soak the incision in water (bath, pool or tub).
• Sutures, steri-strips or staples, if used, will be removed at your follow-up appointment.

When to Call Your Doctor
• Fluid may accumulate under the skin around the incision. A visible swelling that is soft and squishy may be a sign of cerebrospinal fluid (CSF) leakage. A clear sticky fluid may leak from the incision. Call the surgeon should any drainage occur.
• If you experience any of the following:
  • A temperature that exceeds 101º F
  • An incision that shows signs of infection, such as redness, swelling, pain, or drainage.
  • Decreased alertness, increased drowsiness, weakness of arms or legs, increased headaches, vomiting, or severe neck pain that prevents lowering your chin toward the chest.

Recovery
Before you leave the hospital, appointments with the neurosurgeon will be scheduled 10 to 14 days after surgery to remove your sutures and check your recovery. Recovery from the actual surgery varies from 4 to 6 weeks, depending on your general health.

After surgery, you can expect headache and neck pain from the incision that may last several weeks. You will be given neck exercises to do at home. These will help with neck mobility and healing.

Patients typically return to work in 4 to 6 weeks, but be sure to check with your surgeon. A follow-up cine MRI is planned for 6 months to 1 year.

Recovery from the Chiari syndrome and its symptoms may take months or longer. Returning to "normal" is gradual – time is your best ally. Slowly increase activity, avoid strenuous lifting, adhere to instructions and maintain a positive attitude. Focus on the symptoms that have improved, and have patience with those symptoms that remain. Keep a symptom diary to track your progress over time.

What are the results?
The results of your decompression surgery depend on the severity of the Chiari malformation and the extent of any previous brain and nerve injury before treatment. Eighty five to 95% of patients experience major relief of symptoms [2]. However, patients may continue to have residual symptoms from syringomyelia. If injury in the spinal cord has already become permanent, surgery won’t reverse the damage.

Exertional headache and neck pain respond well to decompressive surgery as do most of the brainstem signs (e.g., swallowing problems, facial pain/numbness, voice changes, tinnitus, eye problems, dizziness). Recovery of sleep problems, memory, and spinal cord signs (e.g., numbness or tingling in hands and feet, muscle weakness) take longer and may not completely return to normal.

Figure 8. MRI s before and 1 year after surgery showing restored CSF flow (blue line) around the tonsils and disappearance of the syrinx (yellow arrow) in the spinal cord.

Decompression surgery may allow the syrinx to drain on its own. Follow up is needed to monitor CSF flow and the syrinx site. This progress is evaluated at 1 year with cine MRI (Fig. 8). For any residual symptoms, you and your doctor will discuss possible options to determine the best care.

Recurrence of compression or obstruction of CSF flow is rare.

What are the risks?
No surgery is without risks. General complications of any surgery include bleeding, infection, blood clots, stroke, reactions to anesthesia, and death (rare). Specific complications related to a Chiari decompression craniectomy and duraplasty may include:
• Risk of head and neck pain is variable.
• Cerebrospinal fluid (CSF) leakage is the escape of CSF that flows around the brain. This usually takes the form of a squishy pocket of fluid or drainage from the incision. If leakage is suspected, apply a pressure dressing over the incision and contact your surgeon immediately. If the leak continues, surgical repair may be necessary. New closure techniques and use of biologic glue reduces the risk of CSF leak.
• There is a risk of pseudomeningocele, an abnormal collection of cerebrospinal fluid (CSF) under the tissues of the neck. The collection may resolve on its own; however notify your surgeon if this occurs.
• Nerve or brain damage may cause permanent disability.

Q&A: chiari surgery
Q: Why do some surgeons remove bone from the vertebra in the neck?
A: The amount of tonsillar herniation is a determining factor to remove the arch of the C1 vertebra (e.g., someone with 4mm herniation versus someone with 20mm herniation). If a person is young or athletic, we always try to avoid disturbing the C1 vertebra. There have been cases where a person can develop craniospinal instability years after surgery, either by a neck injury or the natural aging process. Some surgeons prefer to shrink the tonsils rather than remove the bone of C1.

Q: Will more of my brain sag out of my head if you remove bone from the skull?
A: No. The bone removal is in the very middle of the skull to allow the tonsils more space. The cerebellar hemispheres are supported by bone along the undersides of the skull. Cases of cerebellar slumping (cerebellar ptosis) that you may have heard about are rare complications caused by too much bone removal.

Q: Does my syrinx need to be drained with a shunt?
A: No. Years ago placing a shunt into the syrinx cavity was common, but long-term results and problems with shunt clogging have made this technique uncommon and used only for special cases. Adequate decompression of the brainstem and fourth ventricle will allow CSF flow and pressure to normalize and should eventually lead to disappearance of the syrinx on its own.

Q: Why do some surgeons open the dura and some do not?
A: Sometimes bone removal alone is enough to relieve the compression and restore CSF flow (especially in children). Surgeons may use ultrasound to test the movement of CSF and determine if opening the dura and sewing a patch (duraplasty) is necessary. However, in adults the dura is less pliable, so a graft is sewn to enlarge the space. The technique is similar to a tailor letting out the waistband on a pair of pants. While avoiding dura opening may decrease the risk of CSF leak, inadequate decompression may increase the risk of a poor result and lead to reoperation.

Q: Is there a difference in dura patch material?
A: Many patch materials are available, ranging from autologous (patient’s own) tissue to a variety of cadaveric and synthetic materials. We prefer to use patch material obtained from the pericranium (tissue overlying the skull) or a synthetic collagen material.

Q: Does Chiari recur after surgery?
A: True anatomical recurrence of Chiari I is rare. There are many reasons why some patients consider their surgery unsuccessful or “failed.” Technical failure of the surgery means obstructed CSF flow at the foramen magnum. Failure of some symptoms to resolve does not always mean failure of the surgical repair. A cine MRI study is used to evaluate CSF flow.

Scarring of tissue, inadequate removal of bone, new neck or head trauma, increased brain pressure, and tethered cord can be causes of recurrence. Surgical complications such as cerebellar slumping or spinal instability can also cause recurrence.

Q: Why do some people have multiple surgeries?
A: Some patients have had an inadequate decompression to restore normal CSF flow. Either the bone removal was too small or the dura was not opened to adequately expand the space. Delayed post-operative scarring may also lead to repeat surgery. Cine flow and MRI studies play a major role in determining the need for reoperation.

Sources & links
If you have questions, please contact Springfield Neurological and Spine Institute at 417-885-3888.

Links
www.asap.org
www.conquerchiari.org
www.ednf.org
csfinfo.org
www.chiariconnectioninternational.com
Spina Bifida Association

Sources
cerebrospinal fluid (CSF): a clear fluid produced by the choroid plexus in the ventricles of the brain. CSF bathes the brain and spinal cord, giving them support and buoyancy to protect from injury. cerebrospinal fluid (CSF) leak: the fluid surrounding the brain can escape through a hole in the dura lining the skull; may require surgery to patch the leak.
cranectomy: surgical removal of a portion of the skull.
decompression: opening or removal of bone to relieve pressure and pinching of nervous tissue. dura mater: the outer protective covering of the brain.
dural patch: (also called a dural graft, dural substitute, duraplasty) a piece of tissue used to close or extend the dura mater in surgery. Material may come from the patient's scalp (autologous), cow, collagen, or synthetic.
Ehler-Danlos syndrome (EDS): a rare genetic defect in collagen that affects connective tissue (e.g., joints, skin, blood vessels). Collagen is a protein, which acts as a "glue" in the body, adding strength and elasticity to connective tissue. There are six types of EDS. Types I - III cause joint hypermobility; joint dislocations and scoliosis are common.
fibromyalgia (FM): a chronic pain illness characterized by widespread musculoskeletal aches, pain, and stiffness; soft tissue tenderness; fatigue; and disturbs sleep, memory and mood. Many people with fibromyalgia also have tension headaches, temporomandibular joint (TMJ) disorders, irritable bowel syndrome, anxiety, and depression.
glossopharyngeal neuralgia: a painful disorder of the ninth cranial nerve (glossopharyngeal nerve). Irritation of this nerve causes intense pain on one side of the throat near the tonsil area that can radiate to the ear.
hydrocephalus: an abnormal build-up of cerebrospinal fluid usually caused by a blockage of the ventricular system of the brain. Increased intracranial pressure can compress and damage brain tissue.
intracranial pressure (ICP): pressure within the skull. Normal ICP is 20mm HG. intrathecal space: the space surrounding the spinal cord through which cerebral spinal fluid (CSF) flows; also called the subarachnoid space.
multiple sclerosis: a chronic degenerative disease in which the body's immune system eats away at the protective sheath (myelin) surrounding nerves in the brain and spinal cord. The symptoms often come and go and vary widely depending on the affected nerve fibers.
myelopathy: a broad term referring to spinal cord dysfunction of any cause. Some processes that lead to myelopathy include transverse myelitis, injury, arthritis, vascular malformation, vertebral fracture from osteoporosis infection or malignancy, or syrinx an enlarged cyst within the spinal cord.
platybasia: a malformation of the occipital bone (clivus), literally flattening of the skull base. It may be developmental in origin or due to softening of the skull base bone, allowing it to be pushed upward as in basilar invagination. It is associated with other congenital bone abnormalities, such as fusion of the first cervical vertebrae to the skull (atlas assimilation).
pseudomeningocele: an abnormal collection of cerebrospinal fluid (CSF) that communicates with the CSF space around the brain or spinal cord. Unlike a meningocele, the fluid has no surrounding membrane but is contained in a cavity within the soft tissues.
pseudotumor cerebri: elevated pressure within the skull due to buildup of cerebrospinal fluid. Symptoms of headaches, vision problems, and ringing in the ears mimic a brain tumor, but no tumor is present. Most often occurs in women who are obese.
scoliosis: an abnormal side-to-side curvature of the spine.
shunt: a drainage tube to move cerebrospinal fluid from inside the ventricles of the brain into another body cavity (e.g., abdomen).
syringomyelia: a chronic progressive disease of the spinal cord caused by an obstruction of normal cerebrospinal fluid (CSF) flow that redirects the fluid into the spinal cord to form a syrinx.
syrinx: a cavity filled with cerebrospinal fluid (CSF) that expands and elongates over time, destroying the center of the spinal cord.
tethered cord syndrome: a condition of the spinal cord caused by an abnormal attachment or "tether" of the cord to the bones of the spinal canal. The spinal cord gets stretched and can become damaged. The filum terminale is a fibrous thread which connects the very bottom of the spinal cord to the coccyx bone.
trigeminal neuralgia: a painful disorder of the fifth cranial nerve (trigeminal nerve). Irritation of this nerve can cause intense pain that usually affects one side of the face usually in the forehead, cheek, jaw, or teeth.
ventriculoperitoneal (VP) shunt: a tube placed in the ventricle of the brain to drain excess cerebrospinal fluid into the abdomen.