Part 1: Chiari Malformation and Syringomyelia
Overview

Chiari (pronounced key-AR-ee) malformation is a condition in which the lower part of the brain, called the cerebellum, herniates through the skull and down into the spinal canal (Fig. 1 and 2). The herniated tissue compresses the brainstem and blocks the normal flow of cerebrospinal fluid (CSF). The blockage can then cause a buildup of fluid in the spinal cord (syringomyelia) or in the brain (hydrocephalus). Chiari is often misdiagnosed because of the variety of bony and soft tissue abnormalities that compress the cervical spinal cord, brainstem, cranial nerves, or blood vessels, resulting in a wide array of possible symptoms. Symptoms include headache, neck pain, dizziness, arm numbness or weakness, sleep problems, fatigue, etc. Headache in the back of the head that worsens with coughing, sneezing, or straining is a hallmark symptom.

Chiari affects children and adults. Treatment options depend on the type of malformation and severity of symptoms. If symptoms are mild, regular monitoring and medications can be effective. However, symptoms typically progress and worsen over time. Surgery may be recommended to remove a part of the skull bone and create space for the cerebellum and brainstem. An accurate diagnosis and prompt treatment are important to prevent permanent injury to the nervous system. A neurosurgeon with expertise in Chiari can use imaging studies to confirm the diagnosis and recommend a treatment for each unique Chiari case.

Types of Chiari malformations

- **Chiari type I**, the most common, affects both children and adults (Fig. 2). Because the back of the skull is too small or deformed, a crowding of the brainstem, cerebellum, and tonsils occurs. As the tonsils push out of the skull opening (foramen magnum), they press onto the spinal cord and block CSF flow. Chiari I sometimes is found with a fluid-filled cyst (syrinx) in the spinal cord. Symptoms, which may not appear until late childhood or adulthood, include severe headache, neck pain, imbalance, dizziness, swallowing problems, numbness in the hands, depression, fatigue, and sleep problems.

- **Chiari type 0**, a newly identified form of Chiari, describes the absence (or a “zero” herniation) of the tonsils below the foramen magnum. Yet Chiari 0 includes the presence of both symptoms and a syrinx in the spinal cord. This new type is under study and controversial.

- **Chiari type II** is present at birth and affects infants. It occurs with the birth defect myelomeningocele, a form of spina bifida. When the spinal canal does not close before birth, some of the spinal cord protrudes like a sac from the baby’s back. Both the brainstem and tonsils are pulled down into the spinal canal, blocking CSF flow in the brain and causing hydrocephalus. Symptoms can include trouble swallowing and gagging, high-pitched breathing, weak cry, arm weakness, and developmental delays. This type is correctly called Arnold-Chiari malformation.

- **Chiari type III** affects infants and is a rare but severe herniation that involves the cerebellum. It can develop with the birth defect encephalocele, a fluid-filled sac at the back of the baby’s neck.

- **Chiari type IV** affects infants. This rare and often fatal malformation occurs when the cerebellum does not develop properly.
Chiari I malformation

Chiari type I malformation, the most common, affects both children and adults. The condition begins with the underdevelopment of the fetal skull forming during pregnancy. During childhood, the brain continues to grow and the skull hardens. However, the small size or shape of the Chiari skull is mismatched to the size of the brain. Thus, a crowding of the brainstem, cerebellum, and tonsils occurs. Crowding pushes the cerebellar tonsils through the skull’s opening (foramen magnum), where the spinal cord exits. The tonsils put pressure on the spinal cord, blocks CSF flow, and results in the Chiari signs and symptoms. Sometimes a fluid-filled cyst (syrinx) develops within the spinal cord.

Chiari I is seen on MRI scans in people of all ages. Its incidence was earlier estimated to affect 1 in every 1,000 births. Now with increasing use of diagnostic imaging, physicians note that Chiari may be far more common. Patients typically seek medical attention in their 20s and 30s. Three times more women than men are affected. Genetic studies show that Chiari may cluster in some families.

Bone deformity
The skull is formed from eight bones that fuse together along suture lines. Of interest in Chiari are the occipital bone (where the skull and spinal column connect) and the top two spinal bones (C1 and C2 vertebrae). In the middle of the occipital bone lies the foramen magnum, an opening where the spinal cord connects to the brainstem.

Inside the skull are three areas: the anterior, middle, and posterior fossas. The posterior fossa contains the brainstem, cerebellum, and cranial nerves IV-XII (Fig. 3A).

A variety of bone abnormalities can occur in patients affected by Chiari:

- The posterior fossa may be smaller than normal. If too small, the effects can be crowding of the brainstem and cerebellum, as well as herniation of the tonsils through the foramen magnum (Fig. 3B).
- Sometimes the occipital bone is misshaped or thickened.
- Basilar invagination is a condition where the top of C2 (odontoid) pushes upward into the foramen magnum. This defect can narrow the foramen magnum and crowd the brainstem and cerebellum causing Chiari-like symptoms.
- Scoliosis is a bone deformity that causes curvature of the spine. There is a high rate of scoliosis associated with Chiari and syringomyelia, especially in children.
- Ehlers-Danlos syndrome (EDS) is a connective tissue disorder that may increase the incidence and severity of Chiari. EDS causes joint hypermobility and loose/unstable joints. In an area called the craniocervical junction, strong ligaments attach the C1 and C2 vertebrae to the skull, allowing movement of the head. For someone with both Chiari and EDS, extra testing and precautions are taken to ensure the connection between the spine and skull is intact. Spinal fusion surgery may be needed to support the neck and skull.
**Cerebellum herniation**
The cerebellum is the lower part of the brain located in the posterior fossa. On the underside of the cerebellum are two tonsils (Fig. 4). The cerebellum primarily coordinates body movement. It maintains muscle tone and balance. The cerebellum is also involved in cognitive functions (e.g., attention, language), memory, and learning. Signs of cerebellum problems include loss of coordination, unstable walking (gait), trouble with speech, and difficulty with eye movement and swallowing.

In Chiari, the cerebellar tonsils are stretched as they push through the foramen magnum into the spinal canal (Fig. 5). This results in compression of the brainstem and spinal cord. The extent and progression of this tonsillar herniation can be seen on MRI.

**Brainstem compression**
Acting as a relay center, the brainstem connects the cerebrum and cerebellum to the spinal cord. The brainstem performs many automatic functions such as breathing, heart rate, body temperature, wake and sleep cycles, digestion, sneezing, and coughing. Ten of the 12 cranial nerves originate in the brainstem.

In Chiari, compression of the brainstem and cranial nerve nuclei can occur. Patients may experience problems with sleeping (pons), breathing (medulla), swallowing (CN IX), facial pain or numbness (CN V), hearing loss (CN VIII), irregular heartbeat (X), and digestion (X).

**Cerebrospinal fluid blockage**
Cerebrospinal fluid (CSF) is a clear, watery-like liquid that flows within and around the brain and spinal cord. CSF helps keep the brain buoyant and cushions it from injury. This fluid is produced inside the ventricles by the choroid plexus and is constantly being absorbed and replenished.

The CSF flows through the ventricles and out into the space between the brain and skull (subarachnoid space) and down into the spinal canal (Fig. 6). A large amount of CSF fluid lies at the back of the cerebellum in an area called the cisterna magna. From the fourth ventricle, CSF drains into the cisterna magna. From there, it circulates down into the spinal canal and around the spinal cord. CSF also flows from the fourth ventricle down the central canal inside the spinal cord.

As the heart beats, CSF flows into the brain. This is normally balanced by CSF then flowing from the brain into the spinal compartment. In a Chiari malformation, this balanced flow is disrupted. The obstructed CSF begins to force its way like a water hammer through the foramen magnum. Pushing the tonsils down even farther, it exerts pressure on the brainstem. The increasing pressure compromises normal functions of the brain and/or spinal cord and a myriad of symptoms occur. The excess CSF can collect and enlarge either the ventricles in the brain (hydrocephalus), or form a cyst in the spinal cord (syringomyelia).
Syringomyelia

Syringomyelia (pronounced sir-RING-o-my-elia) is the development of a fluid-filled cyst (syrinx) within the spinal cord. When cerebrospinal fluid (CSF) flow is obstructed and collects within the spinal canal, it can eventually form a syrinx. The syrinx expands and elongates over time, then damaging the center of the spinal cord. The compressed nerve fibers inside the cord cause a wide variety of symptoms. Problems affect the arms or legs, or affect feeling, strength, or balance.

Symptoms

- Loss of sensitivity, especially to hot and cold
- Sensory loss in a "cape" distribution (over the tops of the shoulders)
- Numbness and tingling in hands and feet
- Muscle weakness and spasticity
- Headaches (due to Chiari malformation)
- Pain in neck, arms and back
- Loss of bowel and bladder control
- Scoliosis (curvature of the spine)

Causes

Cerebrospinal fluid normally flows in a pulsatile manner throughout the subarachnoid space surrounding the spinal cord and brain. Blockage of flow or changes in pressure can cause CSF to collect inside the central canal — a narrow channel down the middle of the spinal cord.

About 65% of patients with Chiari I develop syringomyelia, which can cause irreversible damage to the spinal cord. In Chiari type 0, patients have no herniation of the tonsils but do have a syrinx in the spinal cord that causes symptoms (Fig. 7). Chiari 0 is sometimes called "borderline" Chiari. This variant supports the clinical finding that symptoms are caused by CSF flow obstruction but do not directly relate to the size of tonsillar herniation.

Syringomyelia can also form as a complication of spine injury, meningitis, spinal cord tumor, arachnoiditis, arachnoid cysts (Fig. 8), or a tethered spinal cord. Symptoms may appear years after the initial injury.

Treatment

If syringomyelia is caused by a Chiari malformation, surgery will likely be recommended. A neurosurgeon will remove bone at the back of the skull to enlarge the opening. The dura overlying the tonsils is opened and a patch is sewn to expand the space, similar to letting out the waistband on a pair of pants. In most cases, this surgery can restore the normal flow of CSF and may allow the syrinx to drain on its own.

If syringomyelia is caused by a lesion, treatment may involve removing a tumor or scar tissue. In cases of a tethered spinal cord, surgery may be performed to release the attachment on the cord at the filum terminale.

Another treatment option is drainage of the syrinx. A neurosurgeon can surgically insert a drainage system called a shunt. The shunt includes a flexible tube with a 1-way valve that directs the fluid out of the syrinx and in the desired direction. One end of the tubing is placed in the syrinx. The other end is placed outside the spinal cord. The shunt remains inside the spine after surgery. However, shunts pose risks and often become clogged or dislodge. Repeated surgeries may be necessary.
Chiari I signs and symptoms

Chiari I symptoms vary from person to person and do not necessarily relate to the size of tonsillar herniation. Some people with large herniations have no symptoms (asymptomatic). Yet others with small herniations have severe symptoms. When symptoms are present, they are often vague or nonspecific. Symptoms are caused by disruption of the CSF flow and compression of nervous tissues.

Because the brainstem is responsible for most body functions, Chiari causes all kinds of strange symptoms. People may experience symptoms that range from headache to irritable bowel. The five most common symptoms are:

1. Posterior headache on exertion with neck pain (70%)
2. Hoarseness or swallowing problems
3. Sleep apnea
4. Weakness or numbness in an extremity
5. Balance problems

People with Chiari I often develop symptoms during their teen or early adult years. The disorder is also seen in young children and older adults. In some cases, a head or neck injury from a car accident or sports injury triggers the onset of symptoms. Some patients have had Chiari triggered by a spinal tap or epidural anesthesia.

Listed below are Chiari symptoms grouped by the body area affected:

**Headache**
- Pressure-like headaches that start at the back of the skull and may radiate behind the eyes.
- Headaches that worsen with physical strain, coughing, sneezing, or bending forward (Chiari headaches are often mistaken for migraines)

**Pain / spine problems**
- Neck pain, pain across shoulder blades
- Chest pain
- General body pain
- Curvature of the spine (scoliosis) may be present with syringomyelia
- Joint hypermobility (Ehler-Danlos syndrome)
- Tethered cord

**Balance / ear problems**
- Ringing or buzzing in the ear (tinnitus)
- Dizziness, spinning (vertigo)
- Imbalance, clumsiness
- Trouble walking (gait)
- Hearing loss

**Eye problems**
- Blurred or double vision
- Sensitivity to bright light
- Spots or floaters in your vision
- Jerking eye movements (nystagmus)
- Difficulty tracking or following objects with your eyes

Chiari I symptoms in infants / children:
- Trouble feeding and swallowing
- Excessive drooling
- Noisy breathing (strider), especially with crying
- Apnea (stop breathing spells)
- Irritability, head banging and nighttime awakening (signs of headache)
- Stiff neck
- Poor arm strength (trouble crawling)
Sleep problems
- Snoring
- Sleep apnea
- Fatigue
- Inability to fall or stay asleep (insomnia)

Face and throat problems
- Difficulty swallowing, choking and gagging
- Facial pain, numbness or tingling
- Hoarseness, change in voice
- Chronic cough

Problems in arms and legs (spinal cord signs)
- Numbness or tingling in arms / hands / legs
- Weakness in arms / hands / legs
- Poor hand coordination
- Loss of feeling in arms / hands
- General body weakness

Thinking (cognitive) problems
- Trouble speaking, word finding
- Trouble thinking
- Problems with memory and concentration
- Depression or mood changes
- Nervousness or anxiety

Other problems
- Nausea and vomiting
- Abdominal pain
- Frequent urination
- Irregular heart beat, palpitations
- Passing out episodes, syncope

Chiari is often misdiagnosed because of the variety of bony and soft tissue abnormalities that compress the cervical spinal cord, brain stem, cranial nerves, or blood vessels, resulting in a wide array of possible symptoms. In fact, the diagnosis is often delayed until symptoms become severe or persistent. However, accurate diagnosis and an effective treatment plan are important to prevent further injury to the person's nervous system.
Diagnosis and testing

The complex symptoms of Chiari I malformation can mimic other diseases—often leading to misdiagnosis and delay in treatment. At times, Chiari I is mistaken for fibromyalgia, chronic fatigue syndrome, migraine, multiple sclerosis, mental disorder, depression, sinus disease, trigeminal neuralgia, or other neurologic disorders. Some people wait for years before a diagnosis is made. Yet, an accurate diagnosis and plan of treatment are important to prevent permanent injury to the nervous system. The average patient suffers symptoms for 3 to 7 years before a diagnosis is made.

There is no specific test to confirm Chiari. Rather, a diagnosis is made by assessment of the patient’s symptoms, neurological exam, and MRI findings (i.e., tonsillar herniation, bone deformity, CSF blockage, syrinx).

A complete medical history and physical exam can determine if your symptoms relate to Chiari or another problem. A neurological exam detects problems with cranial nerves, such as gag reflex, facial numbness, hoarseness, double vision, tremors, and vision problems. You may be referred to an eye (ophthalmologist) or ear (otolaryngologist) specialist, or to undergo a sleep evaluation.

If Chiari is suspected, the doctor will order one or more imaging tests to confirm the diagnosis. Diagnostic tests may include:

MRI (magnetic resonance imaging) scan is a noninvasive test used to evaluate the brain, spinal cord, and surrounding CSF. MRI can identify the extent of cerebellar herniation (Fig. 9). The herniation can reach to the level of the first two vertebrae (C1 and C2) of the cervical spine. Herniation of the tonsils is often measured in millimeters (mm) below the foramen magnum. The classic definition of Chiari I is a herniation of more than 5 mm below the foramen magnum. However, the size of herniation seen on MRI does not closely correlate with symptoms. Someone without herniation may have severe symptoms while another with a large 20-mm herniation may have no symptoms.

Today the diagnosis of Chiari I is based on symptoms and CSF blockage. MRI of the neck and thoracic spine can detect abnormal collections of CSF within the spinal cord (Fig. 10). This fluid-filled cavity (syrinx) can be seen surrounded by stretched tissues of the spinal cord.

Cine MRI scan is a special MRI study used to observe CSF flow (Fig. 11). With each heartbeat, CSF is forced out of the ventricle of the brain, into the cisterna magna, and down the spinal canal. When the heart relaxes, the CSF flow reverses. The movie-like cine MRI captures the fluid movement. The test can determine if, and by how much, a Chiari is blocking the back-and-forth flow of CSF between the brain and spine.

CT (computed tomography) scan is used to view the bony skull base and spinal column. It can detect thickened bone or previous trauma. CT can also be used to more clearly see bone abnormalities, such as basilar invagination or Ehler-Danlos syndrome.

X-rays of the neck may be taken in flexion and extension to view the bony vertebrae. These images can help your doctor identify any instability at the craniocervical area.

Figure 9. An MRI of the brain shows the cerebellar tonsils (arrow) herniating through the foramen magnum (yellow line).

Figure 10. An MRI of the neck shows a collection of CSF in the spinal cord (yellow arrow) called a syrinx.

Figure 11. A cine MRI movie shows a blockage of CSF flow (white) behind the tonsils and pistoning (up and down movement) of the tonsils.
Treatment options

Treatment options vary depending on severity of symptoms, the extent of tonsillar herniation, and the presence of other conditions, such as syringomyelia, hydrocephalus, and disorders of the skull and spine.

Observation (watch and wait)
If a person has mild or no symptoms, monitoring by regular check-ups and periodic MRI scans may be recommended. Some people experience headache relief with anti-inflammatory or pain-relieving drugs.

Follow these self-care tips to minimize neck strain in daily activities:
• Ice packs for 20 minutes can help relieve neck and shoulder pain.
• Get at least 8 hours of sleep and use a good pillow.
• Have a sleep study and evaluation for sleep apnea. A CPAP (Continuous Positive Airway Pressure) machine can greatly improve your sleep quality and reduce fatigue.
• If you are overweight, shed extra pounds to reduce the strain on your arms and legs and help with numbness/tingling sensations.
• Eat a healthy diet, include fiber, and drink plenty of water.
• Stay active with low-impact activities, such as walking, cycling, or water aerobics.
• Play cards, crossword or Sudoku puzzles to sharpen your thinking.
• Tai Chi or yoga can help stretch and tone muscles, improve balance, and reduce stress. Avoid poses that aggravate your symptoms.

Avoid these activities if you have a Chiari, with or without syringomyelia:
• High-velocity chiropractic manipulation that can worsen the herniation and aggravate the spinal cord.
• Cervical traction.
• Trampolines, roller coasters, scuba diving, and other activities that apply G forces to the neck.
• Contact sports to avoid include football, soccer (heading the ball), diving, running, weight lifting, etc.
• Constipation and straining during bowel movements. Straining can cause formation or worsening of a syrinx. A high fiber diet, plenty of water, and stool softeners can help, especially if you take narcotic pain relievers (Vicodin, Percocet).
• Lumbar punctures (spinal taps) and epidurals can be dangerous for a person with Chiari. Ask your doctor to review this important literature on these procedures with respect to increasing herniation. Or discuss with your neurosurgeon.

Childbirth (bearing down and pushing) can also increase cerebellar herniation and formation of a syrinx. Make sure your OB/GYN is aware of your Chiari and tell your neurosurgeon if you become pregnant.

It is important for patients to closely monitor their symptoms. Some patients find it helpful to keep a symptom diary. By keeping track daily of how you feel and what you do, you may be able to find patterns, identify triggers, and notice subtle changes over time. Bring the symptom diary to each appointment to help you communicate more clearly with your doctor. Knowing what symptoms you experience most, and to what degree, can help shape your diagnosis and treatment. If your symptoms worsen or if any new ones develop, call your neurosurgeon’s office.

Download a symptom diary at www.mayfieldchiaricenter.com
Surgery
In patients with moderate to severe symptoms or with a syrinx, surgery is advisable. The goals of surgery are to stop or control the progression of symptoms caused by herniation of the cerebellar tonsils, and relieve compression of the brainstem.

In patients with a syrinx, the goal of surgery is to prevent or control progression. Symptoms related to the obstruction of CSF should decrease as flow normalizes.

- **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. 13). The surgeon opens the dura overlying the tonsils and sews a patch to expand the CSF space, similar to letting out the waistband on a pair of pants.

  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.

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

- **Shunting** is used to reroute CSF. The shunt includes a flexible tube with a 1-way valve that directs the fluid out in the desired direction. For a syrinx in the spinal cord, one end of the tubing is placed in the syrinx. The other end is placed outside the spinal cord. For hydrocephalus, one end of the tubing is in the ventricle of the brain. The other end is placed in the abdomen (called a ventriculoperitoneal shunt). A shunt remains inside the body after surgery. However, shunts pose risks and often become clogged or dislodge. Repeated surgeries may be necessary.

- **Transoral decompression** is a surgical procedure to treat basilar invagination. The surgery is performed through the mouth and to the back of the throat to remove an abnormal odontoid bone (C2 vertebra).

Figure 13. Posterior fossa decompression surgery removes bone and creates more space for the brainstem and cerebellum.
Resources

If you have more questions, please contact the Mayfield Chiari Center at 800-325-7787 or 513-221-1100.

Links
American Syringomyelia Alliance Project, www.asap.org
Conquer Chiari, www.conquerchiari.org
Chiari & Syringomyelia Foundation, www.csfinfo.org
Spina Bifida Association, www.spinabifidaassociation.org

Glossary
basilar invagination: (basilar impression) a rare condition in which the upper portion of the second cervical vertebra (C2) migrates upward and back into the intracranial space. It can be associated with other conditions, such as rheumatoid arthritis, Chiari malformation, syringomyelia, C1-2 instability, or congenital abnormalities.
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.
cervical: the neck portion of the spine made up of 7 vertebrae.
cranietomy: 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. Chiari is often misdiagnosed as fibromyalgia.
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.
herniate: to protrude through the wall of the cavity in which it is normally enclosed.
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 20 mm HG.
intrathecal space: the space surrounding the spinal cord through which cerebral spinal fluid (CSF) flows; also called the subarachnoid space.

Marfan's syndrome: a genetic disorder in which patients develop skeletal defects in long bones, chest abnormalities, curvature of the spine, and circulatory defects.

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. Chiari is often misdiagnosed as MS.

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.

Paget's disease: also known as osteitis deformans, a bone disease in which normal bone is destroyed and then replaced with thickened, weaker, softer bone. This weaker bone easily bends and deforms. Most often affects the pelvis, thoracic and lumbar spine, skull, femur, tibia, fibula, and clavicles.

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.

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).

subarachnoid space: the space between the pia and arachnoid mater of the brain and spinal cord that contains cerebrospinal fluid (CSF).

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.