

CHIARI MALFORMATION AND SYRINGOMYELIA

A Handbook for Patients and their Families

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Dedicated
To
Marcy Speer, Ph.D.

The SM/CM community is very grateful to the doctors who contributed articles and to Dr. Ulrich Batzdorf's leadership as editor. Unfortunately, one contributor, Dr. Marcy Speer, did not live to see the book published.

Dr. Speer, ASAP Board member, Medical Advisory Board member, and Research Committee Chair, lost her battle with breast cancer on August 4, 2007. Dr. Speer was the Director of the Duke Center for Human Genetics, Chief of the Division of Medical Genetics, and an internationally recognized researcher in neural tube birth defects including Chiari malformations.

Dr. Speer will be remembered for many exceptional scientific contributions, but also for her caring and giving spirit.

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CHIARI MALFORMATION AND SYRINGOMYELIA

ULRICH BATZDORF, M.D.

DEFINITIONS

Chiari Malformation (also known as Arnold Chiari Malformation)

As used today, Chiari Malformation (CM) implies descent of the cerebellar tonsils through the largest opening at the base of the skull (foramen magnum) into the upper cervical (neck) region. Normally the cerebellar tonsils lie within the skull (Fig.1). In persons with CM, the tonsils descend downward to the level of the first, and sometimes even the second (C1, C2), cervical vertebra (Fig. 2).

The term “malformation” may not be entirely accurate. It is certainly not used in quite the same way as we think of malformations of the heart in newborns, cleft palate, clubfoot or spina bifida. When Professor Hans Chiari first described CM over 100 years ago, this distinction was not clear. Today we believe that in most people, descent of the tonsils occurs because the space in the skull in which the cerebellum with its two tonsils (right and left) is housed, is too small for the growing brain; thus, the tonsils “escape” through the foramen magnum. Only a very small number of patients with CM have a truly “malformed” skull. This is usually not apparent from looking at the person, but subtle differences in angles and length of individual bones making up the mosaic of bones we call the skull, exist. Such differences in skull bones similarly can crowd the cerebellum and cause the tonsils to “escape” through the foramen magnum. These conditions include platybasia (literally a flat, rather than angled, skull base) and basilar invagination, in which the cervical spine pushes upward into the bone at the base of the skull like the stem of a mushroom. Sometimes the bone is less hard than usual.

Syringomyelia

Syringomyelia (syrinx = a tube; myelia from the Greek=spinal cord) is a cyst containing fluid within the substance of the spinal cord (Fig. 3). Except for the very rare cysts associated with spinal cord tumors, the fluid in these cysts is the same as normal cerebrospinal fluid (CSF). Because of the effect of normal body activity such as coughing and straining, true syringomyelia (SM) cavities have a tendency to enlarge over a period of time, often years. It is emphasized that many fluid cavities within the spinal cord may not in fact be a syrinx, in spite of a similar appearance. Such entities have been termed persistent central canal, hydromyelia or benign syrinx. These are believed not to enlarge over time.

Spina Bifida

Spina bifida is a true birth defect that occurs because the normal development of the spine (growth from the right and left sides to join in the midline of the back) is incomplete, leaving a bony opening. The spinal cord, lying deep to the bone, may also be involved in this birth defect. The opening is often covered by skin and may not be visible to the naked eye (spina bifida occulta). With more significant abnormalities as may be seen in newborns, the opening is covered by some of the membranes over the spinal cord such as the arachnoid (see below) to form a meningocele, which may leak spinal fluid. Occasionally the spinal cord and nerve roots may protrude through this opening. This is called a myelomeningocele. Leg weakness, numbness and bladder and bowel control problems may be present. Meningoceles and myelomeningoceles require surgical repair in infancy. They are generally associated with hydrocephalus, requiring shunting of the brain ventricles, the cavities containing spinal fluid that are normally present in the brain.

Central Canal

When a baby develops in the uterus, there are many stages of growth. The growth of the brain and spinal cord is exceedingly complex and goes through different phases. At one phase in the development of the spinal cord, there is a tiny slit running the full length of the spinal cord (the central canal). We do not know exactly why this slit develops in the human embryo. We do know that it gradually disappears with age, sooner and more completely in some people than in others. MR scanning allows us to identify such slits, and with modern technology we occasionally still see a remnant of this central canal in adults of all ages. This imaging finding may be very similar to that of a syrinx, although typically these cavities are slender and trail into a fine point at each end (Fig. 4). They should be referred to as hydromyelia rather than syringomyelia.

Syringomyelia was probably first described in post mortem specimens in the 16th Century, although the term syringomyelia was coined in 1824 by a French anatomist and physician, Estienne.

The relationship between descent of the cerebellar tonsils and spinal cord cysts was defined by the work of Cleland and Chiari in 1883 and 1891, respectively. Spine trauma as a cause of syringomyelia was probably first observed in 1880 by Strümpell. Credit for describing the classical clinical syndrome with the “dissociated” sensory loss (loss of pain and temperature perception, while light touch and position sense are preserved) belongs to Gowers (1886), who also noted the tendency for these sensory changes to develop first over the shoulder region. The relationship of spina bifida to syringomyelia dates to the work of Russell and Donald (1935).

Milestones in the development of our understanding of these conditions are the work of Gardner (1959), who recognized the dynamic nature of spinal fluid pulsations. He postulated that the spinal fluid is driven into the central canal of the spinal cord through an opening at the apex of the fourth ventricle, called the obex. The surgical procedure he advocated including plugging of this opening. Williams (1986) identified the partial CSF obstruction (“cranio-spinal pressure dissociation”) and

also postulated that normal physiologic forces, including pulsations of spinal fluid, may act to promote enlargement of a syringomyelic cavity, once formed. His theories led to the development of decompressive surgery as a means of opening up partial obstructions of the spinal fluid spaces and thereby prevent filling of the syrinx cavities.

The development of magnetic resonance imaging (MRI) and other technological advances has given rise to a better understanding of spinal fluid dynamics. Oldfield, (1994) has proposed the piston theory, whereby the cerebellar tonsils are thought to act as miniature pistons driving fluid from the space surrounding the spinal cord into the spinal cord itself. The treatment implications of this theory remain the same as Williams’: relieving the constriction that allows the tonsils to behave like miniature pistons by decompression, i.e. widening the fluid-filled spaces.

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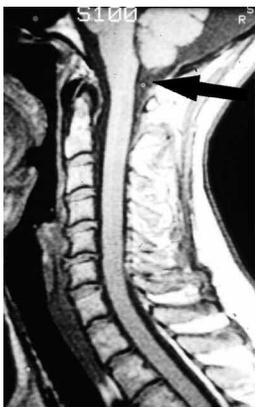


Figure 1

MRI scan (T1W) of a normal posterior fossa and cranio-cervical junction. The cerebellar tonsils are rounded and lie just above the level of the foramen magnum. The arrow points to a small pocket of CSF just below the rounded tonsils; the size of this pocket varies among healthy people. The tonsils lie just above the bone edge, showing as a dark, curved linear structure just to the right of the tonsils.



Figure 2

MRI scan (T1W) of a patient with Chiari malformation. The arrow points to the wedge-shaped, pointed cerebellar tonsils which extend to the lower margin of the C 1 lamina (the dark ovoid area just below the arrow tip). The tonsils have slightly deformed the adjacent brainstem but the spinal cord is normal in appearance; there is no syringomyelic cavity.



Figure 3

MRI scan (T1W) of a patient with Chiari malformation and an extensive syringomyelic cavity. The wedge-shaped tonsils extend to the lower edge of the C 1 lamina; they are slightly less pointed than those shown in Fig. 2. The syrinx cavity distends the spinal cord from the fourth cervical vertebra downward.



Figure 4

Cervical spine MR scan with a typical slit-like fusiform cavity, representing an area of residual central canal.



HOW DID I GET THIS?

ULRICH BATZDORF, M.D.

SPINAL FLUID CIRCULATION AND PATHOPHYSIOLOGY

Syringomyelia really is the result of an abnormality in the circulation of CSF. CSF is normally produced when blood filters through small tufts of tissue called the choroid plexus that lie within the ventricles of the brain. We normally produce about one-third ounce (20 to 25 cc) of this watery, clear fluid every hour, day and night. The fluid surrounds, and thereby cushions, our brain and spinal cord. The fluid is not lost from the body (urine for example is lost), but re-circulates. This means it is taken back into the bloodstream at the same rate at which it is produced, and overall is replaced four to five times each day. Other tufts of tissue, called arachnoid granulations, filter this fluid back into the blood.

The pathway of this internal circulation of spinal fluid is through the subarachnoid space, the arachnoid (spider web-like) being a very thin membrane between the outer, relatively firm dura, and the inner membrane, the pia (pia: tender). At least one possibility is that SM develops because partial blockages (or obstructions) of the subarachnoid space make it easier for the fluid to go through the surface of the spinal cord and into the cord itself, rather than slowly seeping through the blockage. When the fluid enters the cord, it migrates along the spaces surrounding normal blood vessels and then collects inside the cord to form a syrinx cavity.

The driving force for the circulation of CSF is not only the fact that it is produced at one place and removed at another, but normal pulsations of blood and of breathing are indirectly transmitted to the fluid and help propel it along.

TYPES OF CHIARI MALFORMATION AND SYRINGOMYELIA

A. Chiari Malformation

Professor Chiari first described the abnormalities that we now refer to as CM around 1890. His observations were all made on stillborn babies or newborns, and he classified the abnormalities he observed by the severity of tonsillar and cerebellar descent, Type I being the least severe, Type IV the most severe. Today we have a much clearer understanding of these conditions, in no small measure due to the advent of MRI scanning. We are aware of the fact that so-called CM III and IV are indeed true and severe brain malformations and infants with these problems generally do not survive very long after birth. Thus, in practical terms, we see mostly 1) CM I, also sometimes called “adult” type, although it occurs in children as well as in adults; 2) CM II, which occurs exclusively in individuals who had spina bifida defects at birth that required repair in infancy and often also required shunting for hydrocephalus.

Most physicians regard CM I and CM II as very distinct and different conditions, with different treatment approaches. CM may occur with or without an associated syringomyelic cavity, and we do not know exactly why a syrinx cavity develops in some patients and not in others. A group of children have recently been identified who have SM without detectable descent of the cerebellar tonsils, leading to the expression of “Chiari Zero.” This observation underlines the concept that it is really partial blockage of CSF circulation, of whatever cause, rather than tonsil descent, specifically, that may cause SM.

B. Syringomyelia

As discussed above, in almost all conditions in which SM develops, there is at least a partial blockage of the normal CSF circulation. We may divide these situations into two general types, based on anatomy:

1. Abnormalities of the base of the skull, or craniovertebral junction (CVJ). This is the CM discussed above, in which descended cerebellar tonsils act like wedges or partial plugs obstructing the free flow of CSF from the skull (where the fluid cushions the brain) to the spinal canal (where the fluid cushions the spinal cord). Moreover, since these tissue plugs (the tonsils) themselves pulsate with the heart beat and breathing, they may act as miniature pistons to drive fluid from the “water” jacketing (CSF) around the spinal cord into the spinal cord itself.



Figure 1

MRI scan (T1W) of patient who sustained a severe spinal cord injury and C 3-4 spine injury in a diving accident. Note the syrinx cavity which extends both above and below the level of spine injury. The patient became quadriplegic immediately after his injury and subsequently developed additional problems related to syringomyelia.

2. Abnormalities entirely within the spine. Obstructions to the normal flow of spinal fluid around the spinal cord most commonly occur when the arachnoid membrane thickens or develops partitions. This occurs most commonly in the following situations:
 - a. After spinal injury. The injury may be mild or severe, and is not necessarily associated with nerve damage at the time of the injury (Fig. 1).
 - b. After spinal infections, such as meningitis. The resulting condition is called arachnoiditis.
 - c. After spinal surgery, when excessive scar tissue develops.
 - d. In the presence of arachnoid partitions present from birth, such as arachnoid cysts or diverticula (blind pouches).
 - e. With some tumors of the spinal cord that are large enough to interfere with the normal circulation of CSF.

Rarely, after the injection of a foreign substance into the spinal canal.

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WHAT SYMPTOMS CAN I EXPECT TO ATTRIBUTE TO CHIARI MALFORMATION AND SYRINGOMYELIA?

ULRICH BATZDORF, M.D.

As discussed in this primer, CM may exist alone or be accompanied by SM. On the other hand, SM that develops after spinal injury or infection of the linings of the spinal cord and brain (meningitis) is not accompanied by CM. This is sometimes referred to as primary spinal SM. In patients who develop SM after spinal injuries (posttraumatic SM), it may be difficult to distinguish neurological symptoms due to the syrinx cavity from those due to the spinal cord injury itself.

Similarly, it is important to think of symptoms produced by the CM separately from those due to the syrinx cavity within the spinal cord. The list of symptoms that might be seen in patients with these disorders is long and only the more commonly encountered symptoms are listed here. Some lists of symptoms generated by well meaning patients may or may not be helpful. Some relatively common nonspecific symptoms of other illnesses may overlap with somewhat similar symptoms of CM or syrinx patients and might lead individuals to become unnecessarily concerned about a condition they do not, in fact, have. It is also important to realize that no one patient with one of these problems necessarily has all of the symptoms listed; one or another symptom may predominate.

SYMPTOMS RELATED TO CHIARI MALFORMATION

1. Headache, particularly precipitated by coughing, straining, sneezing, etc. (Valsalva maneuvers)
2. Balance problems, which may impair walking
3. Dizziness
4. Eye symptoms, most commonly to-and-fro movements of the eyes, called nystagmus
5. Change in quality of the voice
6. Swallowing problems
7. Sleep disturbances

SYMPTOMS RELATED TO SYRINGOMYELIA

1. Motor
 - a. Muscle weakness and atrophy, particularly in hands and arms
 - b. Increased muscle tone (stiffness or spasticity) in arms and/or legs
 - c. Abnormal curvature of the spine (scoliosis)
2. Sensory
 - a. Decreased feeling in hands and arms. Depending on extent and level of syrinx cavity, legs may also be affected. Sensation involved may be pain perception, temperature perception or position sense.
 - b. Exaggerated sensation (hypersensitivity) in limbs, particularly arms
3. Pain
 - a. Midline pain over the spine, particularly the thoracic area
 - b. Burning pain in arms, over trunk and rarely, legs
 - c. Joint pain, particularly in shoulders

4. Sphincter Problems
 - a. Urinary incontinence, sometimes with spasticity of bladder
 - b. Fecal incontinence
 - c. Male impotence
5. Autonomic symptoms
 - a. Dysreflexia: wide swings in blood pressure, often accompanied by profuse upper body sweating
 - b. Drooping of one eyelid
 - c. Syncope (fainting or near-fainting), which is relatively rare

It is not uncommon for some of these symptoms to be worse on one side of the body.

DO I NEED TREATMENT?

We live at a time when MR scans are easily available (though not inexpensive). As a result, many more patients are diagnosed by MR scan as having a CM, or SM with a CM or SM due to some other problem. The question comes up whether the condition requires treatment. Since there is no effective treatment other than surgery, the question really is whether the person should have surgery. A closely related question is “what would happen if I wait or if I don’t have surgery?”

The decision whether surgery should or should not be considered should not be based only on the imaging (MR) studies, or any other diagnostic studies; it should be based on the patient’s symptoms and whether the patient is worsening.

In medical writing, the course of a particular illness, when no specific treatment is provided, is called the natural history. While we understand that both tonsillar descent (CM) and SM have the potential for worsening, there are no large studies that allow doctors to predict the course of either of these problems. Even if there were such general data, they would be difficult to apply to any one person. The decision whether surgery should or should not be considered should not be based only on the imaging (MR) studies, or any other diagnostic studies; it should be based on the patient’s symptoms and whether the patient is worsening. The availability of MR scans allows us to follow patients with periodic scans, and if there is imaging evidence of progression, i.e. progressive

descent of the tonsils and/or progressive lengthening or widening of the syrinx cavity, the doctor should take this evidence of progression into consideration, along with objective evaluation of the patient by neurological examination.

Because MR scanning is such a readily available diagnostic tool, doctors see an increasing number of patients with borderline abnormalities such as cerebellar tonsils just a few millimeters below the foramen magnum, or a syrinx cavity just a little bigger than “a slit,” which represents a residual central canal. Again, the decision whether or not to recommend surgery should be based on the patient’s symptoms and neurological findings. If there is any doubt about the significance of the findings on imaging studies, the studies should be repeated at a later time and a decision should be deferred. In some instances special studies, such as cardiac-gated MRI CSF flow studies, known widely as CINE studies, or myelography (in patients with primary spinal SM) may be helpful in clarifying the significance of borderline findings on the initial studies.

WHAT CAN BE DONE TO MINIMIZE PROGRESSION?

In previous times, we would hear of patients whose SM symptoms came on abruptly after coughing or sneezing. We now have some understanding of why this happens and generally recommend that patients with SM avoid straining or coughing if they have not been treated for the condition. Coughing and straining, i.e. Valsalva maneuvers, may cause further descent of the cerebellar tonsils and enlargement of a syrinx cavity. Cough medicine should be taken when needed and constipation should be avoided. Untreated patients with SM should not do weight lifting; if they find themselves in a situation where they must do some lifting, it is best to breathe with the mouth slightly open. These same precautions apply to patients with CM, i.e. similar types of straining or breath holding may bring on or aggravate headaches.

Patients with CM should avoid roller coasters before and after decompressive surgery. The unusual and high G force would tend to pull the cerebellum and the tonsils in a downward direction.

There are many types of exercises such as swimming, that can be done safely by patients with SM and with CM. A stationary bicycle provides a possibility for cardiovascular exercise, even for patients with balance problems.

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HOW COMMON ARE CHIARI MALFORMATION AND SYRINGOMYELIA? ARE THERE GENETIC CAUSES?

MARCY C. SPEER, PH.D.

Early studies suggested that 1/18,000 individuals were affected with SM (Small, Sheridan, 1966) although this estimate was most likely low because it was based on autopsy studies rather than direct assessment by magnetic resonance imaging (MRI). The best way to determine how common a condition like SM might be is by using what is called a prospective approach, whereby a large group of individuals is tested regardless of whether they have symptoms or not, and the number of affected individuals in this group is counted and divided by the total number of individuals studied. This approach has never been attempted for SM or CM because it is an expensive and labor-intensive approach; furthermore, CM and SM are relatively “new” conditions since the most accurate way to diagnose them is through MRI, a fairly new technology.

Therefore, we attempted to estimate how common SM might be by using what some might call a “back-door” approach; we identified the common causes of SM (CM II, associated with spina bifida, post-traumatic SM, spinal cord tumors, arachnoiditis, and CM), determined their frequency in the population and how often SM was associated with them, and added up the estimates. While reasonable estimates of most of the conditions were available, we had no good estimate of how frequent CM I, the most common cause of SM, was and therefore we had to estimate that as well. Based on these estimates, we determined that between 182,075 and 234,631

Common questions adults ask when diagnosed with any type of condition are: “Is this genetic?” “Can I pass it on to my children?” When a couple has a child who is diagnosed with CM I, one of the first questions typically asked is “Can this happen in my future children, too?”

Americans are affected with SM; or in other words, somewhere between 1/1,172 - 1,1510 (Speer, Enterline et al, 2003). This estimate is much higher than the 1/18,000 based on the early autopsy series.

Another important question is how common is CM I (regardless of whether SM is associated with it)? In a recent report, physicians and scientists from Johns Hopkins University studied over 22,000 brain MRs (Meadows, Kraut et al, 2000). This series is the largest reported to date, and CM I (defined conservatively as cerebellar tonsillar herniation equal to or greater than 5 mm) was identified in 1/1,280 individuals. This study might be criticized since it was performed at a major medical center and individuals with unusual symptoms might be referred to such places more frequently. Scientists will argue that it underestimates the true frequency (asymptomatic individuals with CM I wouldn't be included in this since the study only included individuals who had brain MRs for some reason, and were therefore symptomatic) and that it overestimates the true frequency (a referral bias because this is a major medical center).

Even though neither of these studies is perfect and both can be criticized, it is remarkable that both lead to very similar results.

IS IT GENETIC?

Common questions adults ask when diagnosed with any type of condition are: “Is this genetic?” “Can I pass it on to my children?” When a couple has a child who is diagnosed with CM I, one of the first questions typically asked is “Can this happen in my future children, too?” Studying the genetic component of a condition like CM I (with or without syringomyelia) can help answer these questions. Genetic studies can also help identify causes for the condition by learning what genes are involved, how they work, and why changes in these genes lead to CM I/SM.

Recently, the clustering of this condition in families (familial aggregation) has been established without question and this is the first step in proving that a condition has a genetic basis.

Over the past 15 years, many scientists have reported the occurrence of CM I/SM in multiple members of families. Recently, the clustering of this condition in families (familial aggregation) has been established without question (Milhorat, Chou et al, 1999; Speer, Enterline et al, 2003) and this is the first step in proving that a condition has a genetic basis. Many unanswered questions affecting the clinical utility of these findings remain, including what proportion of non-traumatic CM I cases demonstrate familial aggregation.

Further evidence supporting a genetic basis in at least some cases of CM I/SM comes from twin studies: when one member of an identical twin set has CM I/SM, the other member has CM I/SM more frequently than a fraternal, (non-identical) twin, and CM I/SM can co-occur with other conditions that are known to have a genetic basis (Speer, Enterline et al, 2003). When considered together, all these data are consistent with a genetic cause for at least some CM I/SM cases.

It is important to recognize that most other conditions in which CM I/SM can also occur are rare (for instance, achondroplasia, Goldenhar syndrome) and altogether probably account for less than 1% of syndromic cases. However, when CM I/SM is known to be associated with another genetic condition, the chance for it to be passed on to other relatives can be straightforward to estimate. Thus, it is important for individuals who are newly diagnosed with this condition, and especially parents of

newly diagnosed children, to learn from their physician whether he or she thinks the condition may be associated with a known genetic condition, in which case referral to a medical geneticist for detailed evaluation of the genetic condition may be important.

Except in unusual circumstances, we are still unable to answer the question of whether or not any person affected with CM I/SM will pass it on to a child. We can confidently say that at least some cases have a genetic basis, and if relatives of an affected individual start to have symptoms of CM I/SM, diagnostic testing may be indicated.

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HOW CHIARI MALFORMATION AND SYRINGOMYELIA ARE DIAGNOSED

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In the 21st Century, the diagnoses of Syringomyelia (SM) and Chiari Malformation (CM) have become more frequent, in part due to the improvements in brain and spine imaging by MRI. MRI is a radiology study that does not use x-rays, but instead applies magnetic fields to image any part of the body. To date there are no scientific studies that show exposure to magnetic fields from MRI is dangerous to a person's health, provided the patient doesn't have certain kinds of metallic implants. The definition and detail provided by MRI is unsurpassed by any other technology



Figure 1

MRI of a child with Chiari I Malformation and Syringomyelia

currently available and is the "gold standard" for the diagnosis of a CM or SM. (Figure 1) Patients with SM and CM will undergo an MRI of their brain and spine during their evaluation. MRI is a more expensive test than a CAT scan (CT), but it provides the physician the best chance of making an accurate diagnosis.

An MRI requires a patient to lie very still in a tunnel-shaped tube for approximately 20-30 minutes. This can be very difficult for some patients, especially if they are claustrophobic and fear confined spaces. Such patients should request sedation, such as valium or another anti-anxiety drug, prescribed by their doctor prior to entering the "tunnel."

Children less than five years of age often need IV or oral sedation to lie still. Young children should be scanned only at MRI

centers that can provide nurses or physicians experienced in monitoring these during the sedation process. In the case of infants who need general anesthesia, one should seek a hospital experienced in providing general anesthesia to pediatric patients.

An MRI of the brain and spine will tell whether or not a patient has CM and/or SM, or any other abnormality of the brain that may cause similar descent of the cerebellar tonsils approximately 3-5mm or more below the opening in the bottom of the skull. [1, 2] A physician is also looking for other potentially treatable causes of CM, such as hydrocephalus. (Figure 2) Hydrocephalus is a rare finding in Chiari I malformations but a very common finding in Chiari II malformation, such as patients with myelomeningocele/spina bifida. Hydrocephalus is a condition in which there is too much cerebrospinal fluid (CSF) in the brain, causing the fluid-filled cavities in the brain (ventricles) to enlarge and thus compress the brain. Other causes of tonsillar descent include mass lesions in the brain such as brain tumors or craniofacial abnormalities. Fortunately, these other causes of Chiari malformation are relatively rare.

A "screening" MRI of the spine is important because it can establish the diagnosis of SM, which is associated with CM I in 10-60% of patients in recent surgical series. SM, which is a cavitation in the spinal cord, can be associated with CM, in both children and adults. (Figure 3) A screening spine MRI should be taken in every patient with CM. Children with an unusual scoliosis (curvature

Figure 2

An MRI of a child with hydrocephalus on the left and a normal MRI of the brain on the right. Note the enlarged ventricles on the left compared to the normal size ventricles on the right (white arrows).

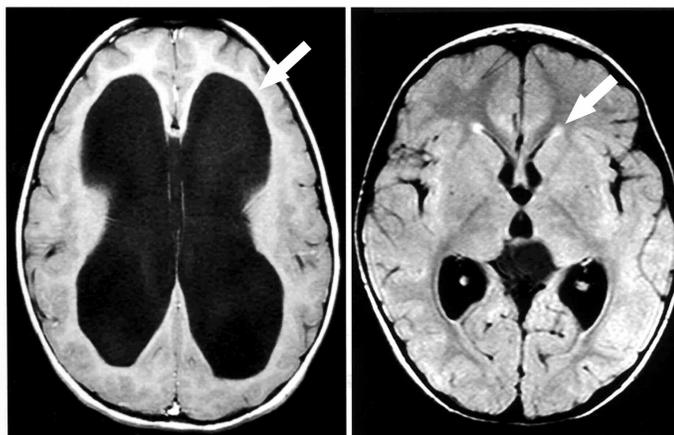




Figure 3

Syringomyelia in a child with scoliosis and Chiari I. The arrow points to the holocord syrinx.



Figure 3a
Preoperative MRI



Figure 3b
Postoperative MRI



Figure 4

Plain X-ray shows a child with scoliosis secondary to his CM and SM.

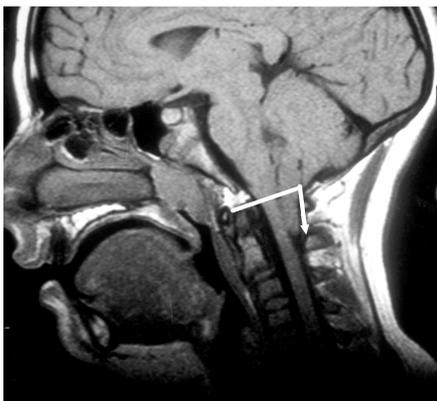


Figure 5

An MRI in a child with a Chiari I Malformation, showing 10mm tonsillar descent (white arrow) below the foramen magnum (white horizontal line)

of the spine) may come to a physician’s attention when the scoliosis rapidly progresses on exam and/or plain x-ray films. The early and timely treatment of a Chiari malformation in a child with progressive scoliosis can yield a good result, in terms of halting the progression of the spine deformity. [3] (Figure 4) These children should also have an MRI of the spine to assist in ruling out SM, which may be contributing to the progressive spine deformity. Patients who have had a traumatic spine injury in the past and who begin to note worsening of their symptoms or deterioration of their neurological function should have an MRI of the spine to assess whether or not they have SM.

The imaging diagnosis of CM is often, but not always, straightforward. However, these studies should always be interpreted by a physician in the context of the patient’s symptoms and neurological exam. An MRI is a powerful tool, but it is just one objective test used by the treating physician to secure the diagnosis and direct appropriate treatment. Thus, surgical treatment of CM and SM requires an experienced neurosurgeon to put the patient’s symptoms together with the neurological examination, prior to recommending surgery. The experienced neurosurgeon looks at the three tools available to him/her before recommending surgical intervention. Each tool provides the patient with confidence in the diagnosis, just as the presence of each of the three legs in a three-legged stool provides improved stability to the stool. The three legs for appropriate diagnosis and treatment of CM and SM are: 1) the patient’s history of the specific characteristic symptoms that brought them to the neurosurgeon, 2) the patient’s examination that shows signs consistent with CM and/or SM and 3) a head and spine MRI demonstrating the characteristic anatomy of CM and/or SM. One leg alone or two legs of this three-legged stool does not uniformly provide the neurosurgeon with enough data to recommend surgery with certainty.

Descent (herniation) of the cerebellar tonsils on the MRI 3-5 mm or more below the level of the foramen magnum (skull opening) constitutes the generally accepted radiological diagnosis is of CM. [2, 4-8] When tonsillar descent is 5 mm or more and there is associated SM of the spinal cord, the diagnoses of CM/S are easily secured. Patients with CM and tonsillar descent of more than 5 mm often show compression and deformation of the tonsils. In fact, the tonsils may be peg-like and also may compress the brainstem and spinal cord, which helps confirm the diagnosis of CM. (Figure 5) However, patients may have tonsillar descent of less than 5 mm, without SM. In these patients, symptoms such as persistently severe headache at the back of the head that are aggravated by exercise or straining help improve the certainty of the diagnosis of a symptomatic CM.

There are certainly “gray” areas in the diagnosis of CM, which engender controversy on this subject and may confuse

both physicians and patients. A very small number of patients may have minimal or almost no tonsillar descent on the MRI, but may have SM. This is called “Chiari 0”. The original description of this entity included 5 patients, each with a syrinx but no hindbrain herniation. Chiari 0 patients may also have all the symptoms of a CM I. (Figure 6) When this extremely rare and relatively new diagnosis is entertained, a patient may benefit from a posterior fossa decompression [9] that results in subsequent collapse of associated SM. (Figure 7)

In CM patients or in any patient suspected of a CM, a special test called a cine-MRI (or cine MRI CSF flow study) can be performed to help improve the certainty of their diagnosis. The cine MRI is a dynamic picture of the brain that shows the movement of CSF around the brain in the region under question, the cerebellar tonsils. It also shows the piston like movement of the cerebellar tonsils. It is essentially a movie of all the MRI pictures taken on a patient. The series of MRI pictures is arranged in a movie format that shows the obstruction of movement of CSF caused by the peg like cerebellar tonsils. In addition, the cine MRI in a CM patient will show the piston like movement of the cerebellar tonsils in the posterior fossa obstructing the normal flow of CSF through the foramen magnum from the brain into the space around the spinal cord. It takes slightly more time for the patient than a standard MRI, and requires special computer software and physicians with experience viewing and

interpreting this type of MRI. Many but not all MRI facilities possess the ability to perform a cine MRI. In some patients, the cine MRI may show a “tight” posterior fossa and obstruction or diminished flow of CSF flow at foramen magnum. (Figure 8) Since all the variations of the operations to treat a CM I attempt to correct the “tight” or small posterior fossa and improve CSF flow in the back of the cerebellum, the cine MRI is yet another modern tool that helps guide the physician and improve the certainty of the diagnosis. [4, 6, 10] After a successful decompression, the cine-MRI often shows return of near normal CSF flow behind the cerebellum. (Figure 9) Not all neurosurgeons use the cine MR for diagnosis. Some institutions use the cine MRI to follow patients postoperatively. In the successful postoperative state for a patient, the cine MRI shows improvement in the CSF flow compared to the obstructed preoperative situation. In patients whose operation for CM and/or SM has failed or just has not improved all the symptoms, a cine MRI is a very useful tool to figure out whether obstruction to CSF flow or compression of the brain persists. [4]

A full spine MRI should be requested in a patient with CM and/or SM. Some physicians prefer that the brain and spine study be performed with contrast (gadolinium) to improve the quality of the exam. The neurosurgeon is trying to rule out a mass lesion such as a tumor (rare) or a tethered cord (less uncommon). A tethered cord is a congenital



Figure 6

A child with severe breathing problems and syringomyelia with minimal tonsillar descent, consistent with the diagnosis of Chiari 0.

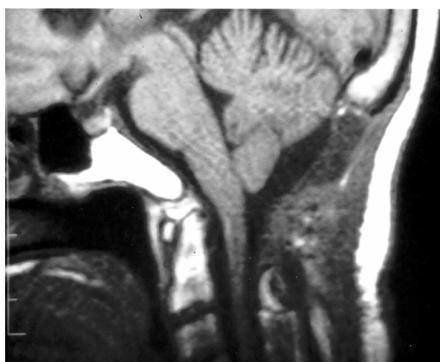


Figure 7

Same child as in Figure 6 but after decompression in which the syrinx collapsed and the patient improved clinically.



Figure 8

Cine MRI: On the left, no posterior flow (arrow) in a patient before decompression of their Chiari I malformation



Figure 9

Cine MRI, return of CSF flow represented by the white space behind the cerebellar tonsils after decompression (white arrow)

condition in which the spinal cord ends too low in the spinal canal, i.e., below the mid body of lumbar level 1 instead of above that level. (Figure 10) The mechanical stretch from a true tethered cord causes stretch on the brain thus contributing to the descent of the cerebellar tonsils. Tethered cord is a rare and controversial cause or associated finding in patients with CM. The treatment of such patients often entails cutting the filum terminale which is the connection



Figure 10

MRI in a Chiari I malformation patient with a tethered cord due to a sacral lipoma. Note the spinal cord goes down to the sacral body (far beyond the normal L1 level) and is attached to a fat mass called a lipoma tethering it to that location like an anchor.

holding [11, 12] the cord too tightly, prior to treating the Chiari malformation.

Another test called a myelogram occasionally may be used in the diagnosis of SM. Although this test has been largely replaced by the MRI, it still may be useful in patients with

difficult to treat SM, or in patients who cannot have an MRI due to metallic implants such as certain artificial joints. A myelogram is an invasive test performed by a skilled radiologist, who injects contrast into the CSF space around the spinal cord, usually by a lumbar puncture. The test is safe but can be uncomfortable. The patient often receives local anesthetic and sometimes IV sedation prior to performing the lumbar puncture. The contrast agent is water-soluble and mixes with the CSF and goes everywhere the CSF flows. X-rays as well as a CT scan are performed after the contrast agent is injected to see how and where the contrast agent has flowed. This test can show where there is a block of contrast agent and thus CSF flow, and what structure is causing this block. Thus, the myelogram and CT scan pictures can guide surgical treatment of this area of flow obstruction. Scarring of the nerve roots (arachnoiditis) can also be diagnosed by this test, or by MRI, and may be of use in deciding whether or not a patient with isolated SM may benefit from surgery.[8, 13]

In summary, if a physician is considering the diagnosis of CM and/or SM, an MRI of the brain and spine is indicated. The interpretation of this test by an experienced radiologist and neurosurgeon is essential prior to any surgical treatment. In addition, these tests must always be viewed in the context of the patient's complaints and neurological deficits in order to decide whether or not surgery is required.

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SURGICAL TREATMENT OF THE CHIARI MALFORMATION WITH OR WITHOUT SYRINGOMYELIA

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FACTORS

As previously discussed, the factors that your doctor takes into consideration in planning an operation for CM with or without syringomyelia would be:

1. The presence or absence of hydrocephalus (increased fluid within the cavities of the brain).
2. Whether or not bony abnormalities exist at the base of the skull, such as basilar invagination or reduction in the size of the posterior fossa, and whether or not there is distortion of the CSF space at the foramen magnum. (Fig. 1A, 1B).
3. The stability of the cranio-vertebral junction (CVJ). This may require imaging of this region with the head moved in the flexed (leaning forward) position and the extended (leaning backward) position, if the doctor is suspicious of instability.
4. The presence of SM or its extension into the brainstem, called syringobulbia.
5. Whether a previous operative procedure was performed, such as a posterior fossa decompression, and whether any syrinx shunt operations were performed.

These factors will guide your physician in making a surgical judgment.

GOALS OF SURGERY

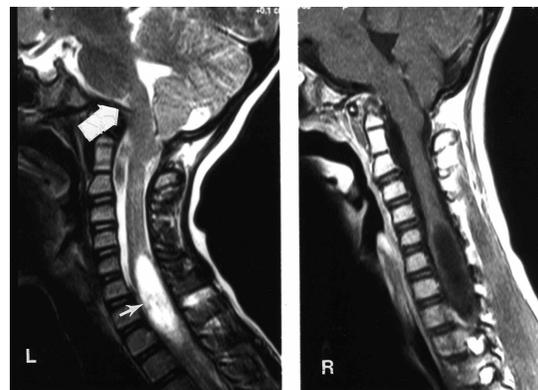
It is more or less recognized that the operative procedure for CM and SM is a posterior fossa (back of skull) craniotomy (opening) with possible removal of the upper-most portion of C1 (cervical laminectomy of the first cervical vertebra). The aim of the operation is to return the cerebrospinal fluid (CSF) circulation to as close to normal as possible, thus achieving relief of symptoms; correction of the impaction or compression of the brainstem by the descending cerebellar tonsils, and deflation or shrinkage of the syrinx. In addition, one of the goals of the operation is prevention of recurrent problems and to arrest progression of problems in the future. Hence, the factors described above come into play, such as instability and presence of the bony abnormalities as well as the potential for scarring.

IMAGING PRIOR TO SURGERY

Your physician may require x-rays of the neck and the skull to visualize the anatomy and stability. As discussed in the previous chapter, the MRIs may be a standard type, or may also include one which is done with the neck in flexed and extended positions, or may evaluate the CSF flow (cine scan).

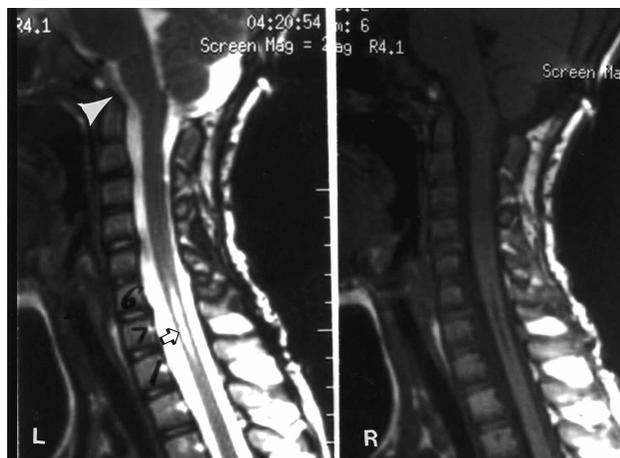
If hydrocephalus is present, this requires treatment prior to embarking on any further management. Fortunately, this occurs in only a very small number of patients. In the absence of hydrocephalus, the physician then looks to see whether there are bony abnormalities at the base of the skull or upper cervical spine.

Figure 1A



Preoperative study of CM-I with bony abnormalities. Composite of mid-sagittal MRI of posterior fossa and cervical spine (back of head and neck) in T2-weighted (left) and T1-weighted (right) modes. The large white (L) points to the abnormal bone invaginating into the brain stem. There is a syringomyelia (SM); white arrowhead.

Figure 1B



Postoperative study of same patient in Figure 1A. An anterior transoral removal of the offending bony problem has been made (arrowhead). This is a composite of mid-sagittal MRI of posterior fossa and cervical spine in T2-weighted (L) and T1-weighted (R) modes. Note that the SM (open arrow) has shrunk.

At times, the surgeon may also request a CT scan in different positions to look for bony abnormalities at the base of the skull or upper cervical spine. When such abnormalities produce bony compression and symptoms related to this may be relieved with positioning of the head or with traction, the operative procedure would be a decompression of the region of the foramen magnum and potentially a fusion of the skull to the upper cervical spine. If on the other hand the bony abnormality cannot be corrected with head position or with neck traction, the compression still needs to be relieved and your physician may elect to perform surgery from a front approach or from a side approach. The anterior, or lateral, decompression is then followed by the traditional operation from behind for the associated "Chiari" problem, and possibly a fusion.

In the absence of bony abnormalities, the presence of CM, with or without syringomyelia (85% of patients), would require suboccipital craniectomy (enlarging the foramen magnum by creating an opening at the back of the skull) to relieve the pressure, to give more room for circulation of CSF and to relieve direct pressure on the brainstem. Many times this may be associated with removal of the back portion of the first cervical vertebra (C1 laminectomy). The need for doing an operation inside the covering of the brain and spinal cord is called an intradural procedure. Many neurosurgeons feel that this is essential to ensure the outflow of CSF from around the brain into the spine as well as to get ride of any scar tissue. Placing a dural graft creates a more generous space for the fluid to circulate.

The surgeon sometimes uses ultrasound during the operation to assist with the detection of any further abnormalities and also to see the flow of CSF as well as its pulsations. This also gives an indication of the descent of the cerebellar tonsils and the extent of compression. The ultrasound can help to visualize relationships between the bone as well as the brainstem and the cerebellum. Some neurosurgeons will use an operating microscope. It is the surgeon's decision whether or not to shrink the cerebellar tonsils. A dural graft may be obtained from pericranium (a layer of deep scalp tissue just outside the skull), from the covering of neck muscle or muscle from the thigh called fascia lata, or even a substitute material such as Gore-Tex.

The surgeon then closes the wound in a layered fashion. In a small number of patients, a posterior fusion procedure may be required. Whether or not an internal shunt is placed would be at the discretion of the treating physician and the abnormalities encountered.

WHAT CAN I EXPECT IMMEDIATELY FOLLOWING SURGERY?

Your neurosurgeon may consider using a long-acting local anesthetic into the muscles and the nerves in the neck to delay the onset of pain. Pain control is also

obtained by using intravenous (IV) medications that go directly into the IV tubing. This may be patient-controlled and is usually done in a manner in which an overdose cannot occur. At times, the surgeon may recommend a soft collar for comfort. Medications such as a muscle relaxant (Robaxin) and pain medication taken by mouth may be prescribed postoperatively. Ice packs and a collar for comfort may be recommended.

It is important to stay away from strenuous physical activities for at least three months to allow for proper healing of the neck musculature. This would include heavy lifting, avoidance of contact sport activities, using a trampoline and avoiding roller coaster rides. It is important to make sure that one has regular and soft bowel movement; straining can be avoided by the use of gentle laxatives and increased fluid intake. Some physicians may also wish to have the patient avoid bending from the waist.

It is important to stay away from strenuous physical activities for at least three months to allow for proper healing of the neck musculature. This would include heavy lifting, avoidance of contact sport activities, using a trampoline and avoiding roller coaster rides.

In children, scoliosis can be a presenting symptom in at least 15-20% of patients. In such a situation, the

If symptoms do recur, this generally happens within the first two years. Many times the recurrence may be due to scar tissue formation, or late development of instability or changes that can occur as a result of the opening of the envelope (dura) of the brain.

scoliosis brace should be worn three to four weeks after the surgery.

FOLLOW UP

This is essential. It is important to be able to pick up problems such as recurrence of symptoms. If symptoms do recur, this generally happens within the first two years. Many times the recurrence may be due to scar tissue formation, or late development of instability or changes that can occur as a result of the opening of the envelope (dura) of the brain. In children, it is essential to periodically obtain an MRI because of continued growth of the skull in all directions.

A postoperative MRI may be requested by the treating surgeon within the first few days and again at periodic intervals to assess the syrinx and its shrinkage or collapse. It is important to recognize that even though the patient may get relief of symptoms, the syrinx cavity may not deflate (Fig. 2A, 2B).

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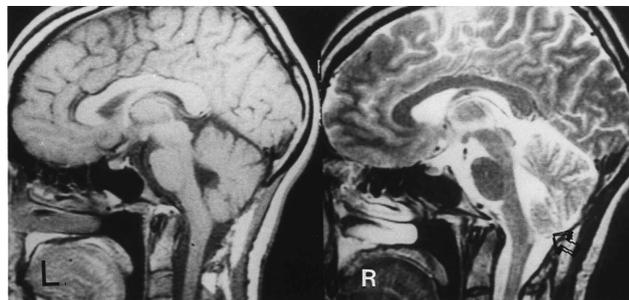
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Figure 2A



Preoperative MRI of brain and upper cervical spine in a patient with CM-I. There is downward location of the cerebellar tonsils (open arrow) below the rim of the foramen magnum (small black arrow).

Figure 2B



Postoperative MRI images of the same patient as in Figure 2A (after posterior fossa decompression surgery). Composite of T1-weighted (L) and T2-weighted (R) images. Note the ascended tonsil region and the restoration of CSF around the brain stem and cerebellum (open arrow).



TETHERED CORD AND PRIMARY SPINAL SYRINGOMYELIA

BARTH A. GREEN, M.D.

There is a common consensus among contemporary clinicians treating syringomyelia (SM) that almost all cases involve an alteration or blockage of normal cerebrospinal fluid (CSF) flow dynamics (Figure 1). CSF is produced from blood by a vascular plexus in the brain (choroid plexus) and circulates through and around the brain and spinal cord, coming up against gravity, to be absorbed back into the blood over the top of the brain by a drainage system (arachnoid granulations). In the case of the Chiari Malformation I (CM I), most often this blockage occurs at the base of the skull and can result in CSF being forced into the spinal cord creating a cystic cavity (syrinx). This condition is called SM, which has been well described earlier.

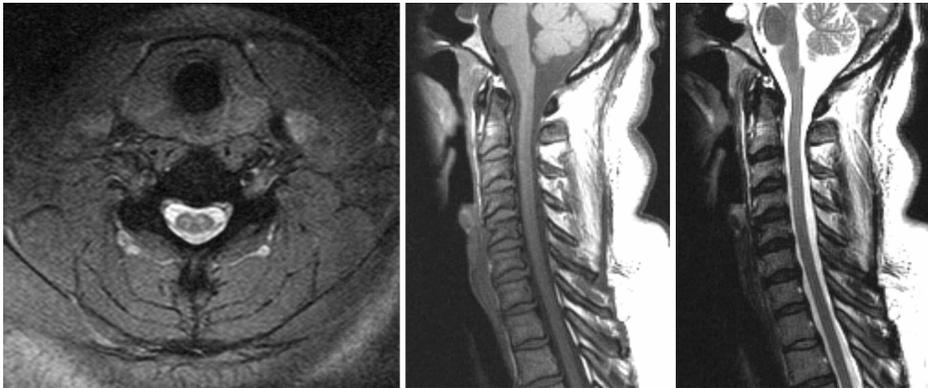


Figure 1

Normal spinal cord MRI images

Other causes of blockage of normal CSF flow dynamics within the spinal canal include trauma, tumors, infections, hemorrhages, postoperative complication and complications following spinal injections, as well as congenital causes. The most common cause is posttraumatic SM, which can occur after spinal column and/or spinal cord injuries. Patients who have had meningitis or other infections, including intraspinal abscesses, or arachnoiditis (intradural scarring), and patients with spinal column or spinal cord tumors that block the CSF flow also can develop these cysts. All these conditions have one common characteristic: blockage of normal CSF flow circulation.

Often, the spinal cord is actually stuck to the lining of the spinal sac (meninges). If the spinal cord is adherent, or stuck, either toward the back of the sac (dorsal) or the front of the sac (ventral) or to the side (lateral) or in any combination, it is termed a tethered (stuck or adherent) spinal cord. Tethered Cord Syndrome (TCS) has been used to describe patients whose spinal cord is adherent to the outer meninges (Figure 3) (arachnoid or dura mater) which are the envelopes of soft tissue – membranes – surrounding the spinal cord and nerves in the spinal canal. Tethering blocks the flow of CSF almost like a boulder or dam in a river and can create “eddy currents” which can force CSF into the spinal cord tissue and is often associated with SM. Tethering may be associated with very small, almost microscopic, collections of CSF, called microcystic changes or myelomalacia (Figure 2), or in other cases with large cysts or cavities i.e., true SM (Figure 4).

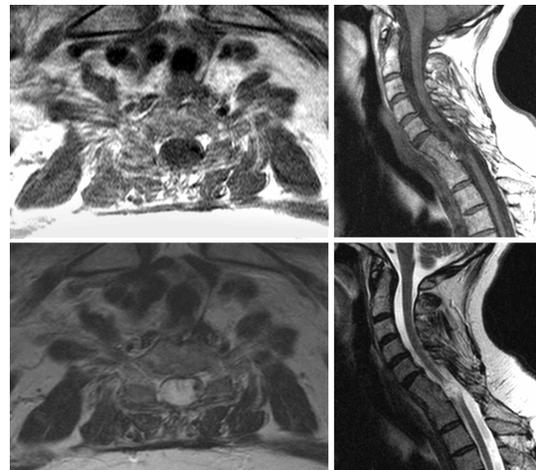
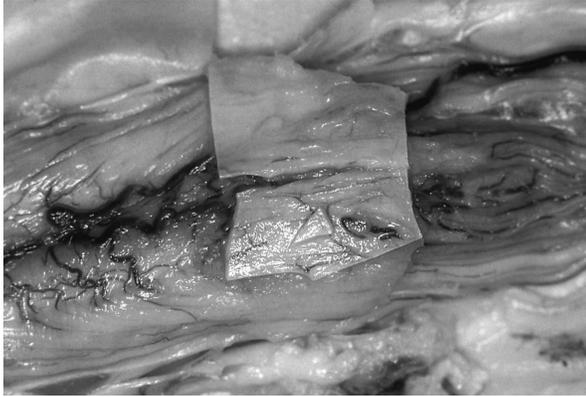


Figure 2

MRI images of spinal cord injury with myelomalacia (microcystic changes) and tethering C7.

Figure 3



Spinal cord tethered to dura.

In either case, these syrinx or myelomalacic changes may be associated with various signs and symptoms of neurological dysfunction. The signs and symptoms of SM, which can occur with or without tethering, include: *weakness* (loss of motor function), numbness (loss of sensory function), *pain*, which can either be local or radiating (radicular), or more diffuse (deafferent = hot, burning, hypersensitivity, electrical, pins and needles, etc), *increased muscle tone* (spasms or spasticity), *autonomic dysreflexia* (widely varying blood pressures, increased spasticity, sweating and face flushing, etc), *hyperhidrosis* (abnormal or profuse sweating), *sphincter dysfunction* (change in bowel, bladder or sexual function), or *Horner's Syndrome* (unequal pupils and droopy eyelid, usually seen in cervical and upper thoracic spinal cord cysts or tethering). These signs and symptoms can appear individually or in any combination and may be unilateral (on one side of the body), bilateral (on both sides of the body), may alternate from side to side or be positionally related.

Historically, the treatment for SM was to shunt or divert the spinal fluid from within the spinal cord to either

the peritoneal (abdominal) cavity or the pleural (chest) cavity with a small silastic tube or catheter (shunt). Today the approach used in most cases is to treat the cause, not the result, of tethering in a manner similar to that used in CM I/SM. In CM I/SM patients we re-establish normal CSF fluid flow dynamics at the base of the skull and upper cervical spine surgically; over the following weeks and months, there is a regression, and often collapse, of the syrinx. In cases of SM other than from a CM I, we now primarily untether the spinal cord through a laminectomy (removal of the dorsal, or back part, of the spinal column) to expose the most frequent areas of tethering of the spinal cord to the meninges, which are dorsal and lateral (back and sides of the spinal canal). Using the operating microscope, ultrasound (sonar) and electrical monitoring (Motor and Sensory Evoked Responses), we are able, in most cases, to safely release the adhesions or tethering and re-establish CSF flow surgically. Most often, the surgeon will use a dural substitute or a patch-like graft from a tissue bank (allograft) or from fascia or connective tissue taken from the patient's leg (autograft). In some cases, a synthetic dural substitute may also be used. Using a dural graft creates a tent-like widening of the CSF space (subarachnoid space) at the level of injury, surgery or tethering, and allows the re-establishment of more normal CSF flow dynamics and the regression of the microcystic (myelomalacic) or syringomyelic (larger cyst) cavity without the need for a shunting procedure.

By positioning the patient up on one side or the other for several weeks after surgery (only while in bed), we have achieved a better overall outcome and less recurrent tethering. In rare cases in which untethering fails as the first line of therapy, shunts can still be safely performed with diversion of the spinal fluid from within the spinal cord to another cavity or compartment in the body (chest or abdominal cavity) as described earlier. The shunt can be placed either with open surgical exposure into the abdomen or chest, or through a cannula (large needle) as

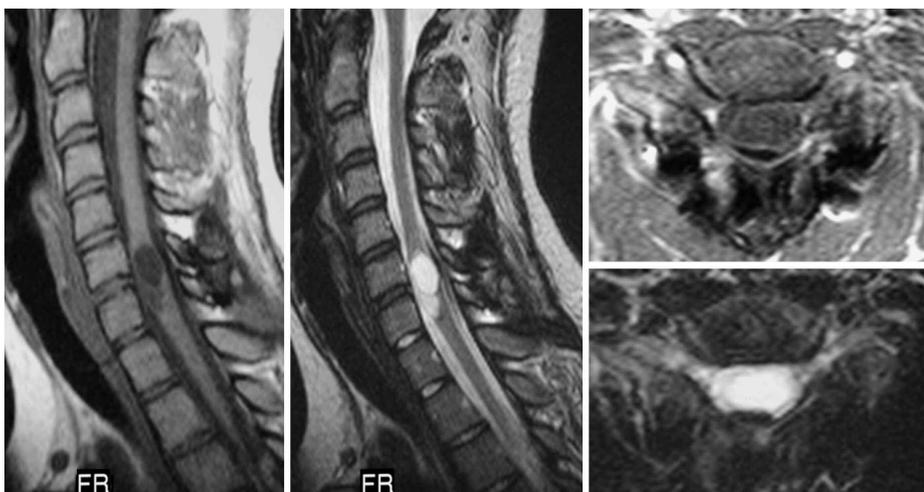


Figure 4

Spinal cord syrinx (confluent) C6 and myelomalacia (microcystic) C7.

a means of insertion. Often in cases of diffuse arachnoiditis (scarring) or multiple failed operations, shunting is necessary. In some patients this technique is modified by placing a short piece of the silastic catheter (shunt) into the cyst and bringing it out just a fraction of an inch and placing it into the spinal fluid (subarachnoid) space. This is sometimes called a stent rather than a shunt, but in reality, it is shunting the spinal fluid from the syrinx cavity into the spinal fluid space surrounding the cord rather than into the abdomen or chest. Another term for this procedure is a syringo-subarachnoid shunt.

As with any major surgery, the treatment of SM or of a tethered cord can be associated with complications. There are risks that can occur with any general anesthetic, that range all the way from death to other anesthetic complications. The neurological complications of spinal cord surgery could include paralysis or weakness, loss of sensation, bowel, bladder or sexual dysfunction, as well as infection, hemorrhage, etc. These risks are usually in the few percent range, with the most common problems being CSF leak, recurrence of tethering and reformation of a cyst or SM. Unfortunately, recurrence is not a rare situation even in the experience of the most skilled surgeons, and even when patients follow the medical advice for postoperative activity. If tethering or cyst formation recurs, treatment may require revision or repeat surgery. The majority of patients can expect to stop the progression of their symptoms with surgery. However, often there is a reversal of some, and less commonly of all of the preoperative signs and symptoms. As mentioned previously, some patients may be unchanged or may even worsen with surgery. Any patient undergoing an operation for tethered cord or SM must be aware of these issues. For example, almost every patient who has a shunt placed in the spinal cord experiences some increased numbness, often patchy and reversible, although it may be permanent. This occurs from just making a tiny incision with the aid of a microscope into the spinal cord to place the shunt tube (often less than 1/10 of an inch in diameter). However, when one realizes that the spinal cord is like an electrical cable with millions of wires or fibers going

through it, it becomes understandable how even the smallest incision can cause temporary or permanent damage. For these reasons, shunts are usually placed in the back of the spinal cord where one is less likely to lose motor strength or pain and temperature sensation. The loss of crude touch-type sensation or position sense are more likely to be experienced. Depending on which surgeon you talk to or which article you read, failure of these operations may result in patients requiring re-operation or losing function permanently.

It is imperative that every patient feels comfortable with their surgeon and his/her credentials and experience in this area so that the surgical procedure becomes a team effort.

It is imperative that every patient feels comfortable with their surgeon and his/her credentials and experience in this area so that the surgical procedure becomes a team effort. The patients and their families or significant others should have full knowledge and understanding of all the pros and cons, alternatives and risks. When surgery is performed with the surgeon and patient working together as a team in every sense, it can result in the best possible outcome for everyone involved.

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WHAT CAN I EXPECT FROM SURGERY?

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The surgical management of the Chiari malformation and syringomyelia may differ significantly. They, as well, are associated with significant overlap regarding both symptoms and surgery. In order to understand what an individual undergoing surgery for either or both of these entities can expect, it behooves us to divide the surgical process into: (1) the events that occur before surgery, (2) the surgery itself, and (3) the events that occur following surgery.

PRE-SURGICAL CONSIDERATIONS

Prior to surgery for the Chiari malformation or syringomyelia, the patient and the patient's family and friends should expect a lengthy discussion about indications for the procedures and be actively involved in the decision-making process. This process has been traditionally termed "the informed consent process." It should perhaps be entitled the "informed decision-making process." The surgeon informs the patient about the issues at hand to the best of his/her ability. Then, the patient and the patient's family and friends participate in an ongoing dialogue with the surgeon regarding the pros and cons of surgery and the strategies for surgical intervention.

First and foremost, it should not be assumed that the presence of either a Chiari malformation or the presence of syrinx constitutes, in and of itself, an indication for surgery. The progression of a syrinx on imaging studies, significant symptoms, or progression of symptoms, in the presence of the anatomical findings consistent with a Chiari malformation and/or syrinx constitute the indication constellation for surgery. Patients should understand this concept and should "interrogate" their surgeon regarding this decision-making process. Realistic expectations regarding outcome, weighed against risk, can then be understood and considered preoperatively (i.e. before surgery is undertaken).

The Surgical Procedure

From the patient's perspective, surgery is painful and is associated with some risks. Assuming that no neurological complications of surgery ensue, the other major risks of surgery include leakage of spinal fluid, pseudomeningocele formation (spinal fluid that has leaked from the spinal sac but is contained under the skin), bleeding and infection. Almost all (but not all) surgical procedures for Chiari malformation and syringomyelia are performed in the prone (face down) position. Most surgeons use skull fixation during Chiari surgery in order to immobilize the operative site. This may cause the patient to have some pain at sites (usually three) where the pins of the skull fixation device have penetrated the skin and attach to the skull during surgery. The patient can expect to be unaware of this device, since it is applied after the patient is asleep and is removed before the patient emerges or awakens from general anesthesia. The incision for the Chiari malformation is usually located in the lower part of the back of the skull and the upper part of the neck in the midline. The incision for a syrinx can be located at any point in the posterior neck or upper back, depending on the location of the syrinx cavity. Although both operations may be painful, Chiari malformation surgery is usually associated with a greater amount of pain due to muscle retraction and the dissection required to perform the surgery.

After Surgery and the Postoperative Period

The postoperative period can be divided into several phases: (1) hospital phase, (2) the first months after surgery, and (3) the long-term period. Each period is discussed separately.

Postoperative In Hospital Course

During this period, pain is usually the most significant issue confronting most patients undergoing Chiari malformation or syringomyelia surgery. Neurological complications obviously may ensue, as well as spinal fluid leakage, bleeding and infection. Most other complications are relatively infrequent and are usually unique to the specific situation at hand. They are therefore difficult to discuss in general terms. As stated in the previous paragraphs, pain is the most significant issue during the first several days following surgery. Narcotic pain medications and muscle relaxants are often used to manage pain. The patient and family must understand that the surgeon or other physicians caring for the patient cannot overmedicate for fear of complications. A drowsy patient who is over sedated with pain medication is at risk for developing pneumonia, and does not get out of bed and walk. This patient is therefore prone to the complications associated with bed rest. The treating physicians must carefully titrate (adjust the level) of medication to balance the patient's comfort with safety. It is imperative that family, friends and the patient understand and appreciate the balancing act that the physician must perform.

The First Month Following Surgery

During the first month following surgery, the patient is still in the initial phase of recovery. Usually, if the operation is successful, neurological and symptomatic improvements ensue. Pain subsides relatively rapidly during this timeframe (usually over a two- to three-week period). Of note, some patients have persistent surgical pain that may last for several weeks or months following surgery. They must be prepared to deal with such consequences of surgery. During this period, patients should maintain a modestly active lifestyle. Heavy lifting is not recommended at this time. Walking and appropriate rest is encouraged. It is during this time that some complications may still arise, such as leakage of spinal fluid or infection. Drainage of purulent fluid from the wound or signs of infection, such as a painful, puffy erythematous (reddened) wound, should cause significant concern and be reported to the treating physician as soon as possible.

The Long Term

Many people are concerned about whether or not they will end up in a wheelchair, "like so many others." The vast majority of patients who have surgery for Chiari malformation have only one operation and do very well with their neurological symptoms. It is much less common for people to experience a gradual and progressive downhill course. This can obviously occur in complicated cases, but is an exception, rather than the rule.

Patients may initially improve from a neurological perspective and then subsequently deteriorate. Neurological or symptomatic deterioration can be multifactorial. There may be no structural cause identified (therefore no surgical treatment indicated) or other surgical pathology identified, such as basilar impression, cerebellar ptosis (sinking of the cerebellum and posterior fossa contents into the hole created at the time of the previous Chiari surgery), blockage of a shunt if a shunting procedure was performed for syringomyelia, etc. These potential causes should be pursued, as guided by the treating surgeon. The optimally informed patient, friends and family can help the surgeon most effectively by understanding the proposed strategy for diagnosis, work up and subsequent treatment.

"Don'ts"

In general, there are several don'ts following Chiari or syringomyelia surgery or, for that matter, regarding patients with Chiari malformation or syringomyelia who have not undergone surgery.



Any activity that causes a significant strain on the nervous system and spinal axis, or any activity in which a high impact can be expected, such as roller coasters, skydiving, rugby or football, expose the nervous system and the supporting structures (spine and skull) to excessive and potentially harmful forces. In general, these activities should be avoided. Patients should avoid straining for a bowel movement and may have to take laxative medication, particularly in the first few weeks after surgery, when they may still be on significant pain medication (narcotic pain medication often causes constipation) and may not have resumed their normal mobility. Patients should also avoid extensive coughing, and may need to take cough medicine or even consult their physician if coughing persists. These precautions regarding lifting and straining are particularly important during the first few months (typically three) after surgery.

WHY DOESN'T EVERYONE USE THE SAME TREATMENTS?

It is very difficult for many people to understand the variety of treatments recommended by physicians. This is perhaps no more glaringly obvious than it is with the Chiari malformation and syringomyelia. One neurosurgeon may recommend no surgical treatment, another may recommend surgery, and yet another may recommend surgery utilizing a significantly different approach. How can this be? First of all, medicine is not as

Two patients with apparently identical problems, treated in an identical way, by the same surgeon, may have widely different responses to surgery. One may improve and the other may deteriorate and worsen. As a result, surgeons frequently have different opinions regarding the management of complex and even simple medical disorders.

analytic as one would like, or even expect. There is significant variability from patient to patient, related to their own unique characteristics and the variety of pathologies present. Two patients with apparently identical problems, treated in an identical way, by the same surgeon, may have widely different responses to surgery. One may improve and the other may deteriorate and worsen. As a result, surgeons frequently have different opinions regarding the management of complex and even simple medical disorders.

Two well informed neurosurgeons can differ significantly regarding the management of a complex problem based on their overall and particularly their most recent experiences. Patient outcomes and complications clearly influence the decision-making process. Medical literature, colleagues, mentors and teachers from medical school and surgical training also heavily influence surgeons. This composite of experiences and influences, combined with the physician's interpretation of the patient's problem, cause the surgeon to develop a clinical plan that may be unique and that may differ widely from that of another surgeon.

Differences in scholarly opinions are not unique to medicine and not unique to neurosurgeons. In fact, it is not unique to the decision-making process regarding the Chiari malformation and syringomyelia. Therefore, it is not surprising a surgeon may be conservative, and not recommend surgery for a patient with minimal symptoms, while another surgeon may recommend an aggressive surgical approach. Some surgeons may recommend the use of intraoperative ultrasound and others may not. Some may recommend opening the arachnoid and resecting or

coagulating the tonsils, while others may rarely do so. Some may recommend shunting a syrinx while others may not.

Although situations exist in which there are clearly "wrong" or "right" answers regarding management decisions for a patient, there often lies between them significant "shades of gray." As time passes (years, decades and centuries), much more will be known about the management of such complex disorders as Chiari malformation and syringomyelia. Perhaps decision-making with this newly acquired knowledge will provide more objective and well-defined algorithms for patient management. It is probable, however, that there will always be controversy and difference of opinion among scholars. Political scholars centuries from now will continue to differ regarding opinions on political and legal issues. Religious scholars will continue to differ regarding religious and philosophical issues. Informed consumers will continue to differ regarding the most appropriate purchase. Most certainly, neurosurgeons will continue to differ regarding management of even the simplest of problems, let alone the management of Chiari malformation and syringomyelia.

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PAIN IN CHIARI MALFORMATION AND SYRINGOMYELIA

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DEFINITIONS

What is Pain?

The International Association for the Study of Pain defines pain as: “An unpleasant sensory and emotional experience associated with actual or potential tissue damage or described in such terms.” Pain by its nature is unpleasant. There are a number of physiologic processes (see figure 1), neural pathways, and chemicals that mediate pain. Pain is also an emotional experience. If pain was not an emotional experience, all that pain would be is a complex interaction of nerves and chemicals. Thinking in evolutionary terms, we probably have pain systems within us so that we can prevent damage and harm to ourselves. Although this is not the case in CM or SM, sometimes patients will describe pain when we cannot find anything wrong with testing. Thus, by this definition we can ask, “Can a patient still have pain without any identifiable cause?” If the patient says they have pain, the answer is yes.

There are two fundamental types of pain: *nociceptive* and *neuropathic pain*. Nociceptive pain implies the activation of chemicals (neurotransmitters) in neural pathways that mediate the sensation of pain. We are all familiar with this type of pain from cuts and the dentist. Neuropathic pain implies damage to neural tissue. It does not occur in a normal state. The types of pain that are associated with CM and SM are of the neuropathic type. Neuropathic pain is classically described as lancinating (electric-like), paroxysmal (a sudden severe attack), burning, constant, cramping, and aching. Moreover, neuropathic pain is associated with a number of sensory abnormalities. The definitions of these sensory phenomena are included in Table 1.

There are four physiologic processes that are associated with the development and perception of pain. *Transduction* occurs when active or potential tissue damage causes a release of an algescic (i.e., “pain-causing”) “soup” of painful chemicals that excite or stimulate nerves to send a barrage of electrical information back to the spinal cord. The process of relaying this information to the spinal cord is called *transmission*. At the level of the spinal cord there are a number of complex interactions with secondary, tertiary or quaternary nerve cell bodies. Eventually, the neural pathways for pain cross over to the other side of the spinal cord and ascend to the thalamus (part of the brain) and to higher centers including the cortex, where the process of *perception* occurs. We do not understand much about the process of perception. Various areas of the brain contain chemicals, which actually act as painkillers (analgesics). These nerves send fibers back down the spinal cord to the areas where pain transmission enters the spinal cord. This process of shutting down pain transmission by analgesic substances that are present in our brain is known as *modulation*. Nociceptive and neuropathic pain states can affect any portion of these four physiologic processes.

MANAGEMENT OF PAIN

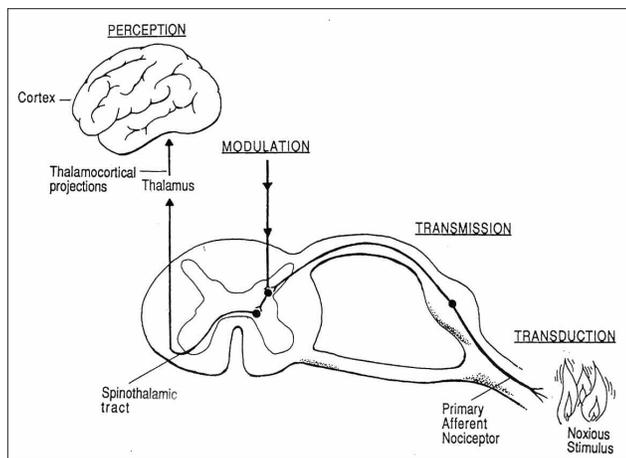
In order to understand how to manage pain, we need to explain further how drugs act on pain mechanisms. Nerves that are involved in pain transmission can become excited or sensitized. When a nerve becomes *excited*, it fires and carries information along its fibers. However, when a nerve becomes *sensitized*, it begins to fire at a lower threshold of stimulation, and it begins to generate a greater magnitude of impulses. This process of sensitization can occur in the peripheral nervous system (i.e., nerves running to the spinal cord) as well as in more central nerve pathways (i.e., spinal cord and higher centers in the brain). Of particular

Table 1

Other Sensory Phenomena

- Hyperalgesia: heightened response to painful stimulus
- Nociception and neuropathic pain
- Dysesthesia: unpleasant, spontaneous or evoked
- Paresthesia: abnormal, spontaneous or evoked
- Hyperesthesia: ↑ sensitivity to stimulus (painful or not)

Figure 1



Four physiologic processes underlying nociception: transduction, transmission, modulation, and perception.

Reprinted from *Postoperative Pain Management*, Ferrante FM, VadeBoncouer TR (eds), p.18, New York, 1993, with permission from Elsevier

interest is the process whereby nerves within the spinal cord or higher centers begin to fire at lower thresholds and with greater magnitude of impulse generation, and even change their connections to other nerves and the areas that they innervate. This process is known as *central sensitization*. Central sensitization is a complex, and frankly, quite beautiful series of chemical, physiologic, and neural interactions. Central sensitization is important because these changes can become permanent. These permanent changes in the firing of nerve cells may be the genesis of chronic pain states, and may be one of the mechanisms for pain in SM.

It is also important to understand that drugs work by binding to receptors that are found on membranes. A good analogy would be a lock and a key. If the right “key” (drug) fits into the “lock” (i.e., receptor), then the lock can be “turned” (i.e., a biologic effect can occur). Sometimes, the same class of drugs can fit into different receptors that are localized in different areas of the nervous system so that different effects are achieved.

When a drug binds to a receptor to “turn the key” to produce a “full” response it is called an *agonist*. Some chemicals attach to receptors to prevent agonists from binding. These agents are called *antagonists*. They have no pain killing activity, but prevent the agonists from causing pain-relief or side effects. Some opioids actually have divergent actions at different receptors, being an agonist at one receptor and an antagonist at another receptor. These agents are referred to as *agonist-antagonists*.

In summary, drugs affect nerves to stop the processes of central and peripheral sensitization by binding to receptors localized on various nerve membranes in different locations throughout the nervous system.

Peripheral Sensitization

Many of the drugs we use to treat neuropathic pain would not seem to be analgesic (painkillers) at first glance. Many of the anticonvulsants can be used to treat neuropathic pain. They work by stabilizing membranes, i.e., they don't allow sensitization or excitation to occur. Thus, in reference to pain, anti-convulsive agents are better called “membrane-stabilizing agents.” They stabilize membranes by preventing the passage of charged elemental particles called ions through pores. These membrane-stabilizing agents have been used for some time. We have older agents such as Phenytoin (Dilantin®) and Carbamazepine (Tegretol®). These older drugs that affect peripheral sensitization usually quiet membranes by blocking the flow of sodium into the nerve cells. This prevents sensitization or excitation from occurring. However, these older membrane-stabilizing agents were associated with a number of side effects, which made their use difficult. Moreover, it took a long time for these drugs to take effect. Newer drugs such as Pregabalin (Lyrica®) do not have these side effects and are better tolerated.

Local anesthetics also block the uptake of sodium into nerve fibers. While the use of local anesthetics is largely confined to injection therapy, a patch has recently been developed that may benefit certain patients with neuropathic pain (Lidoderm® patch). The patch is placed over the areas where there is burning pain. Lidocaine is absorbed from the non-woven felt patches. These patches may be cut to fit the dimensions of the painful areas of skin. Patients cannot use more than three patches continuously for a twelve-hour period. After twelve hours, these patches must be taken off. Used this way, one does not get high levels of local anesthetic in the blood, which could cause ringing in the ears, strange taste sensations, and eventually seizures.

Descending Modulation

There are certain drugs that actually recapitulate how our own inherent analgesic systems work or they affect the analgesic systems in a beneficial manner. Such drugs are the antidepressants and the opioids. Opioids are agents that act like morphine. It is intuitive for the layperson to think that if you give a strong painkiller like morphine or a related drug, it will certainly cause pain relief. Unfortunately, this is not necessarily true with respect to neuropathic pain. Opioids are effective in neuropathic pain, but one has to use much more of them than in other pain conditions. Thus, there is a greater potential for side effects.

Opioids bind to several different receptors; these receptors normally bind to our own naturally produced internal painkiller substances. Opioids are external agents that we administer. Most opioids bind to μ -receptors. This is the usual way that opioids cause pain relief. While

(mu) μ -receptors are found throughout the central nervous system, other receptors called (delta) δ -receptors and (sigma) σ -receptors are actually found in greater numbers in the spinal cord. The binding of opioids to any of these receptors causes pain relief. However, the binding of opioids to μ -receptors can cause the untoward side effects that are sometimes associated with opioids (respiratory depression, slowing of the heart rate, etc.). Binding of opioids to delta-receptors can cause the dysphoric (i.e., “weird” and disliked) feelings that sometimes accompany the pain relief of opioids. Naloxone (Narcan[®]) is an antagonist to the effects of opioids and can block/prevent pain relief as well as reversing the side effects associated with opioids.

recently, most of the serotonin-specific uptake inhibitors (SSRIs) were not effective for neuropathic pain. Two agents, Venlafaxine (Effexor[®]) and more recently Duloxetine (Cymbalta[®]), have been discovered to have analgesic properties for neuropathic pain. These agents are associated with many less side effects than the tricyclic agents. It is interesting to note that at low dosages these new analgesic SSRIs affect only the absorption of serotonin. However, in order to achieve any pain-relieving effect, you must give sufficient dosages so that both the reuptake of serotonin and norepinephrine are affected.

Tramadol (Ultram[®]) is an interesting newer agent because it is an opioid that also partially acts like an antidepressant. Tramadol is a relatively weak opioid that

The layperson usually hears of “nerve blocks” or “epidural steroid injections” that are used to treat various spinal conditions. Because of the nature of the anatomy and the neurophysiology of the pain associated with SM, nerve blocks and steroid injections have little role in treatment.

It is important to note that some opioids can act via another mechanism besides descending modulation to directly prevent/reduce central sensitization. This effect is not mediated through the same receptor interaction (i.e., μ -receptor) for usual pain relief. A different receptor (NMDA-receptor: N-methyl-d-aspartate receptor) is affected that is directly involved in the genesis of central sensitization. Methadone is a good example of a drug that affects the NMDA-receptor. Methadone acts to disrupt central sensitization. If an opioid is to be chosen for the treatment of neuropathic pain, methadone makes a good choice.

The antidepressants exist in old (tricyclic) and newer forms (serotonin-specific reuptake inhibitors). Antidepressants work by preventing the release of chemicals such as norepinephrine and serotonin (examples of our body’s own painkillers) from various nerves in the brain. In essence, they block the reuptake of norepinephrine and serotonin after they are released from nerves. The older agents are referred to as tricyclic because they have a 3-ring organic chemical structure. These agents are associated with many side effects such as: dry mouth, constipation, sedation and sleepiness, and weight gain. Newer agents specifically only affect the re-absorption of serotonin into nerves. Until

is stronger than codeine. Most of its pain killing effects (about 70%) come from modulation of norepinephrine and serotonin, just like the antidepressants.

Central Sensitization

Some of the newest breakthroughs with respect to the management of pain have occurred with respect to the treatment/prevention of central sensitization. From the media, you may be aware of Gabapentin (Neurontin[®]). The great breakthrough with respect to Gabapentin is that it was not more efficacious than previous agents, but it has less side effects. Most side effects associated with Gabapentin are dizziness and sedation. More recently, a new agent, Pregabalin (Lyrica[®]), has become available. Pregabalin works in exactly the same manner as Neurontin[®]. However, Pregabalin is five times more potent than Gabapentin. In the studies that have been done, Pregabalin has about the same side effect profile as Gabapentin. Pregabalin may cause some degree of euphoria.

An exciting new development in the treatment of neuropathic pain is the use of botulinum toxin type A (Botox[®]). It is a protein that is produced by the bacterium *Clostridium botulinum* and is a true

neurotoxin. Botox® was at first released for the treatment of painful hypertrophic muscular conditions such as cervical dystonia. In such conditions, it binds to certain nerve terminals and prevents the release of chemicals (e.g. acetylcholine) that would cause muscular spasm and contraction. However, botulinum toxin has been shown to work by inhibition of central sensitization in neuropathic pain states. There will be more news forthcoming regarding the potential uses of Botox. To date, botulinum toxin type A has not been used to treat the pain associated with SM.

Nerve Blocks

The layperson usually hears of “nerve blocks” or “epidural steroid injections” that are used to treat various spinal conditions. Because of the nature of the anatomy and the neurophysiology of the pain associated with SM, nerve blocks and steroid injections have little role in treatment.

Spinal Cord Stimulation

Spinal cord stimulation involves placement of leads (electrical wires) into the epidural space (the same space that women receive local anesthetics to prevent labor pain). The insertion of a spinal cord stimulator lead is exactly the same as inserting an epidural for the treatment of pain of labor. An electrical current (utilizing a source no greater than a 9-volt battery) is then applied to the various areas of the spinal cord. Spinal cord stimulation works by preventing transmission of electrical stimulation i.e., pain signals, to higher areas of the central nervous system (the brain). It also works by enhancing the effects of certain chemicals (gamma-amino-butyric acid [GABA] and adenosine) in the spinal cord. Spinal cord stimulation may be conceptualized as a “pacemaker” for the nervous system.

Spinal cord stimulation is performed first as a trial. Leads are placed into the spine only via needles through the skin. If the trial is successful, leads may be reinserted and buried under the skin and connected to a battery that is also implanted under the skin. The patient can manipulate how the stimulation feels by adjusting a hand-held programmer over the battery.

Implantable Drug Delivery Systems

For pain that is refractory to usual measures, patients will sometimes be offered implantation of a catheter (tube) that goes directly into the CSF that surrounds the spinal cord. The catheter is connected to a pump that is placed under the skin of the abdomen. Many different drugs (but usually opioids) can be administered through the pump by continuous infusion.

Like spinal cord stimulation, a trial is first performed to see if the patient obtains pain relief. This usually involves

insertion of a temporary catheter into the spine and administration of the drug being considered for use. If the patient obtains pain relief without side effects, he/she is a candidate for the surgical implantation. Unlike with spinal cord stimulation, the patient is not in control of the pump and cannot affect how drugs are delivered. The physician sets the rate of drug delivery. Unlike spinal cord stimulation, the patient must return to the physician at periodic intervals to have the drug replenished in the pump. This is done by inserting a needle through the skin and into the central reservoir of the pump. Refill of the pump must be performed every 3 – 6 months depending upon the kind of drug the patient is receiving and how much drug the patient is receiving.

CONCLUSION

We have attempted to explain many complex physiologic and neurologic processes in terms that are understandable. We have then attempted to show how various medications and procedures can be utilized to treat pain via a mechanistic approach. For pain due to SM, the management of pain is best accomplished via a multidisciplinary approach in which there is access to neurosurgeons, neurologists, rehabilitation specialists and pain management physicians who may act in concert to devise a plan that will help.

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MEDICAL GLOSSARY

- Abduction:** Movement of an arm or leg away from the body.
- Acute:** Having rapid onset, severe symptoms, and a short course. Not chronic.
- Adduction:** Movement of an arm or leg toward the body.
- Adhesions:** Tissue surfaces that are adherent or attached to each other, either loosely or firmly, as a result of wound healing and sometimes inflammation.
- Amyotrophic:** Muscle wasting.
- Anomaly:** A deviation from the average or norm. Anything structurally unusual or irregular i.e., presence of an extra finger or absence of a limb or congenital malformation.
- Anterior:** Pertaining to the front of the body.
- Antiemetics:** Medication to stop or prevent vomiting.
- Apnea:** Cessation of breathing. Skin color changes, pallor and/or cyanosis may ensue. There is a lack of chest wall movement. Can be caused by compression of the brainstem or lower cranial nerves.
- Arachnoid:** Delicate, web-like middle layer of the three membrane layers that cover the brain and spinal cord; arachnoid mater. Named after a spider web.
- Arachnoiditis:** Inflammation of the arachnoid membrane.
- Ascending tracts:** Groups of nerve fibers in the spinal cord that transmit sensory impulses upward to the brain.
- Aseptic:** Sterile, without bacteria; living pathogenic organisms are absent.
- Aseptic meningitis:** Inflammation of the membranes (meninges) that cover the brain and spinal cord. NOT an infection.
- Aspiration:** The act of withdrawing a fluid from the body by a suction device. Inspiratory sucking into the airways of fluid or foreign body, such as vomit.
- Astrocytes:** A type of neuroglial cell that functions to connect neurons to blood vessels.
- Asymptomatic:** Without symptoms, or producing no symptoms.
- Ataxia:** Impaired ability to coordinate the muscles in voluntary muscular movements; symptomatic of any of several disorders of the nervous system.
- Atrophy:** A wasting of tissues or decrease in size of a part of the body because of disease or other influences.
- Atypical:** Not typical.
- Autonomic nervous system:** Portion of the nervous system that functions to control the actions of the visceral organs and skin; its actions are not under voluntary control.
- Axon:** A nerve fiber that conducts a nerve impulse away from a neuron cell body.
- Barium swallow:** An x-ray using barium to view the act of swallowing, the esophagus or stomach. It can show if a person may be aspirating.
- Basal ganglion:** Mass of gray matter located deep within a cerebral hemisphere of the brain; has important functions in automatic movements of the limbs and in the control of muscle tonus.
- Basilar impression:** Upward displacement, particularly of the uppermost part of the cervical spine, into the region of the posterior fossa often producing compression of the brainstem and portions of the cerebellum.
- Bilateral:** Something that occurs or appears on both sides. A patient with equal strength bilaterally means there is equal strength on both sides of the body.
- Brainstem:** The portion of the brain that includes the midbrain, pons and medulla, thalamus and hypothalamus.
- Calamus scriptorius:** Inferior part of the rhomboid fossa; the pointed lower end of the fourth ventricle of the brain. It is shaped like a pen and lies between the restiform bodies.
- Canalization neurulation:** The formation of canals or passages to form the neural tube during the early stages of embryonic development.
- Catheter:** A tube designed for insertion into vessels, canals, passageways or body cavities to permit the injection or withdrawal of fluids or substances. It can also be used to keep a passageway open.

Caudal: Toward the lower end of the spine.

Central canal: The opening or channel normally present through the length of the spinal cord in later fetal life and early infancy. It gradually disappears throughout childhood, but segments of it may remain in adults (see also hydromyelia).

Central nervous system: The part of the nervous system consisting of the brain and spinal cord, which coordinates the entire nervous system of the body.

Cerebellar cortex: The outer layer of the cerebellum.

Cerebellar speech: Abnormal speech patterns seen in people who have a disease of the cerebellum or its connections; a slow, jerky and slurred speech that may come and go or it may be unvaried in pitch.

Cerebellar tonsils: Normal downward extensions of each cerebellar hemisphere.

Cerebellomedullary: Refers to the connections of the cerebellum and the medulla.

Cerebellum: Portion of the brain that lies in the posterior fossa and coordinates skeletal muscle movement.

Cerebral aqueduct: A narrow conduit or passage between the third and fourth ventricles located in the midbrain. CSF moves from the third ventricle through the cerebral aqueduct to the fourth ventricle.

Cerebral cortex: The outer layer of the cerebrum.

Cerebral hemisphere: One of the large paired structures that together constitute the cerebrum of the brain.

Cerebral spinal fluid: Fluid occupying the ventricles of the brain, subarachnoid space of the meninges, and the central canal of the spinal cord.

Cerebrum: Portion of the brain that occupies the upper part of the cranial cavity.

Cervical: The area of the neck made up of seven cervical vertebrae, which are counted from top to bottom. The top is C1, the first cervical vertebra, followed by C2, C3, etc.

Charcot joint: A type of diseased joint associated with varied conditions, syringomyelia among them, which involves disease or injury to the spinal cord. Because normal pain sensation of the joint is impaired, the pain mechanisms that protect the joint are diminished or absent. As a result, the joint may undergo relatively painless severe degenerative changes with deformity.

Chiari malformation: Descent of the brainstem and lower cerebellum through the foramen magnum into the cervical vertebral canal.

Choroid plexus: Mass of specialized capillaries that lie in the ventricles of the brain; these vascular tissue tufts produce cerebral spinal fluid from blood.

Chronic: Long-lasting; a disease having protracted course, not acute.

Cine MRI: Test performed in the MRI scanner that looks at the flow of CSF around the cerebellum and into the spinal canal.

Cisterna magna: Widened area of the subarachnoid space located between the cerebellum and the medulla. It receives CSF from the fourth ventricle through the foramen of Magendie and foramina of Luschka.

Clonus: A series of alternating muscle contractions and partial relaxations that produces a jerking spasm of a limb, most often seen at the ankle, indicative of a brain or spinal cord abnormality involving motor pathways.

CM: See Chiari malformation.

CNS: See central nervous system.

Coele or cele: Related to a cavity or space.

Congenital: Existing at birth, usually refers to certain mental or physical traits, peculiarities or diseases; a more general term than hereditary since congenital includes conditions due to influences arising during gestation.

Contraindicated: A medication or procedure that is not advisable i.e., tetracycline is contraindicated during pregnancy.

Contrast: The difference between two areas in an image; a substance that selectively increases the imaging signal of specific structures such as blood vessels or tumors.

Conus medullaris: The lowest end of the spinal cord.

Cranial nerves: The 12 nerves of the brain that control motor and sensory functions, including swallowing, heart rate, eye movement and smell.

Craniectomy: The excision (removal) of part of the skull.

CSF: See cerebral spinal fluid.

CT scan: A specialized radiographic technique in which many fine x-ray beams converge on one small target area (pixel); a computer calculates the x-ray absorption of tissue in each pixel, and converts this numerical value into a gray scale value; placing pixels of various shades of gray into anatomic arrangement results in an anatomic image.

Cyanosis: Blue or purple color to the skin and mucous membranes resulting from insufficient oxygen in the blood.

Dandy Walker Syndrome: A condition characterized by hydrocephalus in infants associated with an abnormal closure of the foramina of Luschka and Magendie.

Decompression: To relieve or take pressure off.

Diencephalon: Portion of the brain in the region of the third ventricle that includes the thalamus and hypothalamus.

Diplopia: Double vision; occurs when the two eyes are unable to fix (look at) the same point.

Dissociation of sensation: Loss of pain and temperature sensation while light touch sensation is preserved.

Distal: Moving further from the midline or center of the body.

Dorsal: Posterior; pertains to the back of the body or of its parts, such as the spinal cord.

Dura mater: Tough outer layer of the membranes surrounding the brain and spinal cord.

Dysarthria: Speech that is difficult and poorly articulated; this may be caused by damage to the cerebellum or its connections, or to injury to nerves involved in speech.

Dysequilibrium: Inability to maintain proper balance.

Dyesthesia: Alteration in sensation. Sensation of pins and needles, burning pain or unpleasant exaggeration of normal sensation that may occur with or without skin stimulation.

Dysmetria: An inability to accurately control the range or force of muscle movement. Often seen in cerebellar disorders.

Dysphagia: Inability or difficulty in swallowing.

Dysplastic tonsil: Abnormal development of a cerebellar tonsil. Each cerebellar hemisphere has a downward extension called tonsil.

Dyspnea: Labored or difficult breathing resulting in air hunger.

Ectopia: Malposition or displacement of any organ or structure, congenital or acquired.

Edema: An excessive accumulation of fluid within tissues.

Elongated: To make or to grow longer.

Enuresis: Involuntary passage of urine, usually during sleep.

Epidural space: Space between the dura and the bone of the vertebral canal.

Esophagus: Muscular tube extending from the pharynx at the back of the throat to the stomach.

Excision: To cut away a portion.

Extremity: A limb; an arm or leg.

Fascia lata graft: A graft-covering or repair of tissue with fascia, the fibrous membrane that covers muscle over the lateral thigh.

Fasciculations: Involuntary contractions or twitching of groups of muscle fibers; a coarser form of muscle contractions than fibrillation.

Filum terminale: A long, slender filament at the end of the spinal cord.

Foramen: An opening, usually in bone or organ or membrane (plural is foramina).

Foramen magnum: Large opening in the base of the skull through which the spinal cord becomes continuous with the medulla oblongata.

Foramina of Luschka: Openings or passages for CSF on the lateral sides of the fourth ventricle.

Fossa: A depression or cavity within bone or surrounded by bone.

Fourth ventricle: Ventricle (a normal cavity) of the rhombencephalon of the brain; a cavity of irregular tent-like shape extending from the obex upward to its communication with the sylvian aqueduct, enclosed between the cerebellum and the rhombencephalic tegmentum. The ventricle of the brain that lies between the cerebellum and the brainstem, it expresses CSF into the subarachnoid space via the two lateral foramina of Luschka and the single medial foramen of Magendie.

Gait: Manner of walking.

Gliogenous: Of the nature of neuroglia, glia – the tissue that forms the support element of cells and fibers of the nervous system.

Gliosis: Proliferation (growth by reproduction) of the neuroglial tissue in the central nervous system.

Greenstick fracture: A bone break in which the bone is bent but cracked only on the outside of the bend.

Gyrus: One of the convolutions of the cerebral hemispheres of the brain. The upraised ridges of the cerebrum.

Hemiplegia: Paralysis or severe weakness (paresis) of one side of the body, usually due to injury or disease of the brain or spinal cord.

Hemivertebrae: A congenital absence of half of a vertebra.

Horner syndrome: A condition with constriction of the pupil, partial drooping of the eyelid, recession of eyeball back into the socket, and sometimes loss of sweating over the affected side of the face, due to paralysis of the cervical sympathetic nerve trunk. It is often incomplete, i.e. not all the listed manifestations are present in one patient.

Hydro: Water, or collection of watery fluid.

Hydrocephalus: Enlargement of the normal cavities (ventricles) present in the brain. It may result from impairment in outflow of CSF normally produced within the brain ventricles. It may also result from developmental anomalies, infection, injury or brain tumors.

Hydromyelia: Accumulation of fluid in the enlarged central canal of the spinal cord.

Hyper: Prefix meaning above, excessive or beyond.

Hyperreflexia: Increase in action of the reflexes.

Hypo: Prefix meaning less than, below or under.

Hypoplasia: Defective development of tissue.

Hyporeflexia: Decrease in the action of the reflexes.

Hypotonia: Reduced tension, relaxation of arteries; loss of muscle tone.

ICP: See intracranial pressure.

Idiopathic: Pertaining to conditions without clear cause, as of spontaneous origins.

Impulse: A wave of depolarization conducted along a nerve fiber or muscle fiber.

Increased intracranial pressure: An increase in CSF production or blockage of CSF pathways resulting in pressure on the brain. The skull cannot expand to accommodate the pressure, which leads to symptoms such as headache.

Inferior: Situated below something else, pertaining to the lower surface of a part.

Insidious: A disease that develops without recognized symptoms so that the patient is unaware of the onset of the disease.

Interpedicular spaces: Space between the pedicles of the vertebrae.

Invasive procedures: A medical procedure that necessitates entrance into the body as part of the action required. Examples include needles introduced for injections and for lumbar puncture, as well as all surgical procedures.

Ipsilateral: On the same side; affecting the same side of the body. Said of findings (paralysis) appearing on the same side of the body as the brain or spinal cord lesion producing them.

Ischemia: A deficiency of blood in a part of the body.

Klippel Feil syndrome: Congenital anomaly characterized by a short wide neck, low hairline and fusion of two or more cervical vertebrae. The central nervous system may be affected.

Kyphosis: One form of abnormal curvature of the spine. The condition of kyphosis of the thoracic spine commonly called hunchback is an extreme form.

Laminectomy: The surgical removal of the posterior arch (lamina) of a vertebra.

Larynx: Structure located between the pharynx and trachea that houses the vocal cords.

Lateral: Pertaining to the side of the body.

Magnetic resonance imaging: A scanner using magnetic energy that interacts with tissue to yield clear black and white pictures, for example of the brain and spinal cord.

Medial: Toward or near the middle of the body.

Medulla oblongata: Portion of the brainstem located between the pons and the spinal cord.

Meninges: A group of three membranes that covers the brain and spinal cord. Closest to the brain and spinal cord is the pia, then the arachnoid and the outermost covering is the dura.

Meningitis: Infection or swelling of the membranes (meninges) that cover the brain and spinal cord.

Meningo: Refers to the meninges, membranes covering the brain and spinal cord.

Mesencephalon: The midbrain, one of three primitive cerebral sacs from which develop the corpora quadrigemina, the crura cerebri and the aqueduct of Sylvius.

Microgyri: Abnormally small cerebral convolutions.

Morvans chorea type: A condition with irregular uncontrollable movements.

MRI: See Magnetic resonance imaging

Myelo: Refers to the spinal cord.

Myelodysplasia: Defective formation of the spinal cord.

Myelogram: Imaging technique of the spinal cord and nerve roots by use of a radiopaque medium injected into the subarachnoid space, the fluid space surrounding the spinal cord.

Myelomeningocele: Form of spina bifida in which portions of the spinal cord and its membranes protrude through the open space in the vertebral column.

Myelotomy: Surgical incision into the spinal cord.

Necrosis: Death of cells or areas of tissue surrounded by healthy tissue.

Neurovascular bundle: Structure consisting of a group of nerves and blood vessels lying in direct contact with each other.

Nissen fundoplication: An operation of the fundus of the stomach which sutures the fundus of the stomach to the esophagus as a treatment for gastric reflux.

Nuchal rigidity: Muscle stiffness in the back of the neck.

Nystagmus: Constant, involuntary, cyclical movement of the eyeball. Movement may be in any direction; i.e. sideways, up, down or rotatory. May be present continually or only with looking in a certain direction. May be due to congenital conditions, labyrinthine irritability or neurological disease.

Obex: A thin, crescent-shaped band of tissue covering the Calamus scriptorius at the point of convergence of the nervous tissue at the lower end of the fourth ventricle. The point on the midline of the top surface of the medulla oblongata that marks the tail end of the fourth ventricle.

Occipital: The back of the head.

Occipital bone: The cup-like bone at the back of the skull. It houses the occipital lobes of the brain and the cerebellum; its lower edge makes up the back rim of the foramen magnum.

Opisthotonos: Involuntary backward arching of the head, neck or back with stiffening of the entire body.

Papilledema: Swelling of the optic nerve at the point of entrance into the eyeball. Choked disk. In general, considered a sign of increased ICP.

Paraparesis: Partial paralysis affecting the lower limbs.

Paraspinous muscles: Muscles on either side of the spine.

Paresthesia: Abnormal sensation such as numbness, prickling and tingling.

Paucity: Smallness or lower in number.

Peduncle: Stalk-like structures in the brain connecting different functioning areas.

Percutaneous aspiration: Drawing out through the skin.

Peritoneum: The membrane covering the visceral organs and lining the abdominal cavity.

Permeable: Capable of allowing passage of fluid or substances in solution.

Pia mater: The inner membrane of the meninges that encloses the brain and spinal cord.

Platybasia: A developmental anomaly of the skull or an acquired softening of the skull bones so that the floor of the posterior cranial fossa bulges upward in the region adjacent to the foramen magnum.

Pleura: The membranes covering the lungs and lining the inside of the chest cavity.

Pleural space: Space between the lungs and the inside lining of the chest cavity.

Polygyria: Excess of the normal number of convolutions of the brain.

Posterior: Toward the back of the body.

Posterior fossa: Concavity at the back of the skull wherein the cerebellum lies.

Posterior fossa angiogram: A study of the blood vessels of the back of the brain: cerebellum and brainstem.

Prone: Lying horizontal with face down.

Proprioception: The sensory modality allowing awareness of posture, movement and changes in equilibrium and the knowledge of position, weight, and resistance of an object in relation to the body.

Proximal: Closer to the midline or origin; opposite of distal.

Pseudomeningocele: A pocket of cerebrospinal fluid that has formed in an area of previous surgery as a result of an opening in the covering membranes of the spinal cord.

Ptosis: Drooping of the eyelid, often related to the third cranial nerve function; also applied to the drooping of the cerebellum through a large skull opening.

Queckenstedt: A sign or maneuver used for diagnostic purposes. Upon compression of the veins of the neck, unilaterally or bilaterally, CSF pressure measured by lumbar puncture rises rapidly in healthy persons, and falls rapidly when pressure is released. In spinal canal block, the pressure is scarcely affected by this procedure.

Reflex: A rapid automatic response mediated by the nervous system.

Reflux: A return or backwards flow. Regurgitation.

Respiratory distress: Difficulty breathing of any cause, including cardiac, pulmonary and neurological problems.

Reticular formation: Groups of cells and fibers arranged in a diffuse network throughout the brainstem. They fill and connect the tracts that ascend and descend through this area. They are important in controlling or influencing alertness, wakefulness, sleeping and some other reflexes.

Rhombencephalon: Primary division of the embryonic brain that gives rise to the metencephalon and myelencephalon. It includes the pons, cerebellum and medulla oblongata. Sometimes called the hindbrain.

Sagittal: A plane or section that divides a structure into right and left portions.

Scoliosis: A side-to-side curvature of the vertebral column.

Sensory: Pertaining to or conveying sensation (i.e. pain, touch, temperature).

Sheath: A covering structure, usually elongated.

Shunt: Passage constructed to divert flow of fluid when the normal pathways for the fluid are either blocked or inadequate.

Skull series: A group of x-rays taken of the skull from various positions.

Sleep apnea: To stop breathing for brief periods while sleeping.

SM: See syringomyelia.

Somatosensory evoked potentials: An electrophysiological test often used during spinal cord surgery to help determine whether conduction of electrical signals through the sensory pathways of the spinal cord are impaired.

Spasticity: Stiffness or position that is difficult to release voluntarily.

Spina bifida: Failure of the spine to close properly during the first month of pregnancy. In severe cases, the spinal cord protrudes through the back and may be covered by only skin or a thin membrane. When there is no externally evident abnormality, referred to as spina bifida occulta.

Stenosis: A constriction or narrowing of a passage.

Stent: A device used to maintain an opening into a cavity or to hold tissues in place during healing.

Strabismus: Disorder in which the two eyes cannot be directed at the same object; when one eye fixes upon a point (sees an object), the other eye deviates to some other point; vision in the deviated eye is usually suppressed; if not, diplopia results; squint.

Stridor: A harsh sound made during respiration. It is high-pitched and sounds like the howling of the wind. It is due to constriction of the air passages.

Subarachnoid space: The space within the meninges between the arachnoid mater and the pia mater; it is normally filled with CSF.

Subcutaneous tissue: Tissue beneath the skin.

Suboccipital: Area beneath the back of the head; below the occipital bone.

Subperiosteal: Beneath the periosteum (the membrane covering of the bones).

Sulcus: A furrow, fissure or depression, especially of the brain. Many brain sulci have specific names.

Supine: Lying on the back; a position.

Sylvian aqueduct: A narrow canal connecting the third to the fourth ventricle.

Syncope: Fainting, most often the result of inadequate circulation of blood to the brain. Characterized by sudden pallor, coldness of the skin and partial or complete unconsciousness.

Syringo: Prefix used to denote a procedure or process originating in a syrinx cavity (example: syringoperitoneal shunt).

Syringocoele or Syringocoele: The central cavity or canal of the spinal cord continuous with the fourth ventricle of the brain stem; used synonymously with central canal; also used for the cavity in the ectopic cord in a myelomeningocele.

Syringomyelia: Chronic progressive disease of the spinal cord characterized by the development of a fluid-filled cavity or cavities within the spinal cord. Cavitation can occur in any area of the spinal cord. It can involve pathways of the cord that carry impulses of pain and temperature sensations resulting in sensory losses. Pain and paresthesias also occur. Destruction of lateral and anterior gray matter in the cord causes muscular atrophy, spastic paralysis and weakness. Scoliosis is often found in association with syringomyelia.

Syringotomy: An operation to create an opening into a syrinx cavity.

Syrinx: A hollow cavity or tube. In medicine it refers to a fluid-filled cavity within the spinal cord.

Telencephalon: The embryonic endbrain or the anterior division of the prosencephalon from which the cerebral hemispheres, corpora striata and the rhinencephalon develop.

Tentorium: A tent-like structure or part. In the brain the tentorium cerebelli is the fold of the dura mater that lies between the cerebellum and the cerebrum.

Tethered cord: Abnormal attachment and scarring of the spinal cord or its coverings (meninges) can occur as the result of a developmental disorder such as a small mass of fatty tissue, a tight filum terminale or a midline bone spur. Tethering results in loss of normal tiny movements of the spinal cord inside its linings, and may place tension on the cord resulting in cord injury. The spinal cord can also become adherent, i.e. tethered, by scar tissue that results from injury, surgery or disease process.

Thoracic: The area of the back between the cervical and lumbar region comprised of 12 vertebrae.

Tinnitus: A ringing, tinkling or buzzing sound in the ear.

Torticollis: A stiff neck caused by spasms of the neck muscles drawing the head to one side with the chin pointed to the other side. It may be congenital or acquired.

Trachea: Tubular organ that leads from the larynx to the bronchi.

Trachea malacia: Softening of the cartilage of the trachea.

Trophic: Concerning nourishment; applied to a type of nerve believed to control the growth and nourishment of the parts they innervate (supply).

Unilateral: Pertaining to one side.

Ventral: Pertaining to the front of the body or its parts; the belly.

Ventricle: A cavity such as those normally present in the brain that are filled with cerebrospinal fluid.

Ventriculography: An x-ray process used to visualize the size and shape of the brain's ventricles by injecting air or contrast to replace the CSF that normally fills this space.

Ventriculo-peritoneal Shunt: A shunt or tube inserted into the ventricles of the brain attached to tubing that is placed into the abdominal (peritoneal) cavity to drain excess spinal fluid from the brain.

Ventriculostomy: Establishment of an opening performed in the third ventricle to relieve hydrocephalus.

Vermis: The normal midline portion of the cerebellum lying between the two cerebellar hemispheres. Its outer surface appearance reminded early anatomists of a worm.

Visceral: Pertaining to one of the organs found in the skull, chest, abdomen or pelvis (brain, lung, liver, etc.).

Weakness: Inability of muscles to perform their normal function. Weakness of the hands may result in difficulty grasping objects; weakness of the legs may result in difficulty walking; weakness of certain muscles in the pharynx may cause difficulty swallowing.

RESOURCES FOR SUPPORT & INFORMATION

American Chronic Pain Association (ACPA)

Since 1980, ACPA has worked to provide education to help people better manage their pain and live more satisfying, productive lives.

P.O. Box 850
Rocklin, CA 95677-0850
800-533-3231
ACPA@pacbell.net
<http://www.theacpa.org>

American Pain Foundation

Founded in 1997, the American Pain Foundation is an independent nonprofit serving people with pain through information, advocacy, and support.

201 North Charles Street Suite 710
Baltimore MD 21201-4111
1-888-615-PAIN (7246)
info@painfoundation.org
<http://www.painfoundation.org>

American Syringomyelia Alliance Project (ASAP)

Founded in 1988, ASAP provides patient support, information and research funding to improve the lives of persons affected by syringomyelia, Chiari malformation and related disorders while finding the cure.

P.O. Box 1586
Longview, TX 75606-1586
903-236-7079
800-ASAP-282 (272-7282)
info@asap.org
<http://www.asap.org>

Chiari & Syringomyelia Foundation, Inc

CSF's mission is to fund research while offering support and information to individuals affected by Chiari malformation, syringomyelia & related CSF disorders.

29 Crest Loop
Staten Island, NY 10312
718-966-2593
dpoppe@CSFinfo.org
<http://www.csfinfo.org>

Chiari & Syringomyelia Patient Education Foundation

The Foundation works to improve the lives of Chiari and syringomyelia patients through education, awareness, and research. Their Conquer Chiari website is a comprehensive source of information about Chiari Malformation, syringomyelia, and related topics.

320 Osprey Court
Wexford, PA 15090
director@conquerchiari.org
<http://www.conquerchiari.org>

Chiari Connection International

Offers on-line support and help to bring emotional and tangible support and understanding to patients afflicted with Arnold Chiari Malformation and its related conditions.

<http://health.groups.yahoo.com/group/ChiariConnectionInternational/>

Christopher and Dana Reeve Foundation

The Foundation is dedicated to curing spinal cord injury by funding research, and improving quality of life for people living with paralysis through grants, information and advocacy.

636 Morris Turnpike, Suite 3A
Short Hills, NJ 07078
800-225-0292
info@christopherreeve.org
<http://www.christopherreeve.org>

World Arnold Chiari Malformation Association

Staffed by volunteers, WACMA is committed to providing on-line support, current information, and understanding to those affected Chiari malformation & syringomyelia.

chiari-owner@yahoogroups.com
<http://www.pressenter.com/~wacma>