

# Symptomatic Chiari Malformation

People with SB frequently have an Arnold Chiari Malformation

Persons with Myelomeningocele have abnormalities that are not limited to their spinal cord. These include the development of hydrocephalus (in up to 85% of patients) and the development of symptoms related to the area where the brain and spine join (Chiari malformation).

## What is a Chiari malformation?

Over a hundred years ago, Professor Chiari, an Austrian pathologist studied a group of patients who had a portion of the brain displaced into the neck. He graded these malformations in order of severity. Type I (the mildest form) occurs when the lowest part of the cerebellum, called the cerebellar tonsils, descends a short distance into the cervical spine.

## What type occurs in SB?

Chiari Type II or Arnold-Chiari malformation, is a more severe form in which the cerebellar vermis and some portion of the brain stem descend into the cervical spine. Many changes of the brain are associated with this abnormality. Although some degree of this malformation is present in the vast majority of persons with Spina Bifida, it appears to be symptomatic in about one of three individuals. This 33% may be a falsely low figure in that, as we follow patients for longer periods of time, adolescents and young adults appear to have some propensity to develop clinical progression. The development of symptoms related to the Chiari malformation is, in part, related to the severity of the malformation.

When brain tissue is below the level of the fourth cervical vertebrae (C4), the likelihood of serious

symptoms occurring from this abnormality is high. Conversely, minor degrees of descent of the brain well above the level of C3, are less likely to have clearly recognized progressive symptoms. Symptoms tend to come from one of three areas of the central nervous system: cerebellum, lower brain stem and spinal cord. The symptom complex is quite different with each of these and appears to be predictable to some degree, based on the patient's age when symptoms are first noticed.

## What are the symptoms?

Newborns and young infants with symptoms tend to present with difficulty swallowing that leads to:

- poor feeding
- weak or poor cry
- inspiratory stridor (noise on breathing in), frequently exacerbated by crying, arching of the head, and possibly facial weakness, among others.

When severe, the symptoms may result in insufficient breathing to maintain life. The symptomatic Chiari malformation is currently the leading cause of death in the Spina Bifida population, particularly in early life.



A child or adolescent may develop stiffness or spasticity of the arms or hands. This stiffness may be so severe that the upper extremities will not work smoothly. If these symptoms are progressive, they may make the upper extremities useless to the patient. Associated with the stiffness may be loss of feeling or sensation in the hands or arms. This loss may not be to all types of sensation, but may be restricted to certain types, such as the loss of pain or loss of temperature sensation.

The third area that can be affected is the cerebellum. This portion of the brain, located in the back of the head, controls balance and coordination. These symptoms are the least likely to occur and may be the most difficult to pinpoint- especially in the younger child.

### **What tests are done for Arnold Chiari malformation?**

There is variability in how medical centers perform testing of brainstem function. With the advent of magnetic resonance imaging (MRI), the evaluation of structural abnormalities of the cervical spine has become greatly simplified. Although other methods are available, MRI provides extremely high-resolution images of the area of concern with little to no risk to the patient. Not only can the position of the brain be obtained, but even cysts or other problems within the spinal cord can be assessed. The MRI scan can also be used to evaluate the flow of cerebrospinal fluid (CSF) around the back part of the brain.

Additional examinations, which test the integrity of the functional aspects of the brain stem and spine, are sometimes helpful. These include a CO<sub>2</sub> breathing test that reflects the function of the brain's breathing center, somatosensory evoked response (timing of electrical signals that connect the arms and legs with the brain), brain stem evoked response (ability of the brain to process sounds) and others.

### **What is the “shunt first” approach to care?**

Of primary importance in patients suspected of being symptomatic from their Chiari malformation is to ensure normal intracranial pressure prior to any consideration of intervention for the Chiari malformation. This means that the patient should have her/his shunt evaluated first. The “shunt first” approach cannot be over emphasized. (See SBA's Hydrocephalus information sheet.) If there is any question concerning shunt function, the system should be surgically inspected. Once it is determined that the intracranial pressure is normal, and that the shunt is working properly, the next question to be answered is whether the patient's symptoms are life threatening and/or progressive. Most patients with radiographic evidence of a Type II Chiari malformation do not need surgery and appear not to be progressive. If serious symptoms are present, consideration is given to surgical intervention.

### **What about surgery?**

The purpose of the operation is to unroof the cervical spine over the abnormally low brain and to reestablish normal movement of the CSF.

The operation is relatively safe in experienced hands but should be performed by a surgeon especially trained and experienced in operations on children's brain malformations. The technical excellence necessary to perform this operation is high and there is no substitute for significant experience.

The alternative strategy of further conservative care for patients with symptomatic Chiari malformation demands special comment. It is currently believed that the majority of Spina Bifida patients do not demonstrate rapid clinical progression. Opinions vary as to whether an individual patient should have surgery or simply be clinically monitored. Generally, the more the patient's progression is felt to be secondary to their Chiari malformation, the more universal is the acceptance of operative intervention. Serial examinations of the individual are essential in the evaluation of this problem and again, fix the shunt first!