Chiari Type II Malformation

The Chiari Type II malformation is a combination of brain anomalies – most notably a hindbrain herniation – which is only found in people with spina bifida. At the back and bottom of the brain is the cerebellum (see diagram). In the Chiari II malformation the cerebellum is small and thin and along with the brainstem – the part of the brain that joins to the spinal cord – is “squashed” into the upper part of the spinal canal. Although the brain anomalies vary from person to person, almost everyone with spina bifida (myelomeningocele) has the malformation.

The malformation shows up clearly on magnetic resonance imaging (MRI), but because only about a third of children have symptoms, the decision to perform surgery is usually based on the symptoms and their severity, not just the results of an MRI examination.

**CAUSE**

There have been a number of theories about the cause of the Chiari II malformation since its discovery in 1891. Although the matter is by no means settled, the currently accepted theory is that because the cerebrospinal fluid is able to leak out through the open neural tube defect during early foetal development, there is not enough pressure within the ventricular system of the developing brain for proper development. This theory also explains why most people with spina bifida are also born with hydrocephalus.

**SYMPTOMS**

The symptoms can vary quite markedly according to the age of the person and for ease of...
clarification can be divided into two groups: those symptoms in children younger than 2 years of age and those in older children and adults.

For the under 2 group, the most common and potentially fatal symptoms involve respiratory difficulties. Inspiratory stridor, a harsh high-pitched noise on breathing in, is the most noticeable sign. An inability to breathe out properly, often accompanied by cyanosis (turning blue) is also life-threatening. Choking, nasal regurgitation, prolonged feeding time and weight loss are all symptoms of Chiari Type II malformation. Other signs and symptoms are partial paralysis of the upper limbs, nystagmus (squint), weak cry, arching of the neck and lack of muscle tone (floppiness.)

For the group older than 2 years of age, the symptoms are less often a medical emergency than in the younger group. Their symptoms tend to progress more slowly and are less likely to be life threatening. The most common symptoms are weakness in the upper extremities and spasticity. The first noticeable sign may be a loss of dexterity manifesting as deteriorating handwriting and loss of self care skills. Ataxia or loss of coordination of the upper limbs and trunk is also common. Headaches in the back of the head or neck may also be present.

Symptoms in older children and adults can include difficulty swallowing, dizziness, unsteady gate, neck pain and arm weakness.

**TREATMENT**

The treatment for Chiari Type II malformation, if and when it is required, is invariably surgery. The neurosurgeon will want to insure that hydrocephalus or shunt problems are not the cause of the symptoms before operating for Chiari II. When he or she is satisfied that this is the case, then a decompression procedure will be performed.

The aim of this procedure is to reduce the pressure on the brainstem and other brain tissue by cutting away the bone at the bottom of the skull and top of the spinal canal. Because of the abnormal anatomy of the brain with Chiari II, the procedure can be very difficult. The success rate for the procedure is high and improving all the time.

**FOETAL SURGERY AND THE CHIARI II MALFORMATION**

Since the late 1990s, foetal surgery for spina bifida has been performed in 3 centres in the USA. Although a randomised control study is currently underway and the final results of it won’t be available until around 2011, preliminary data seem to indicate that the greatest benefit of this surgery may be the reduction in the severity of the hindbrain herniation.