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Chiari Malformation

What is the Chiari malformation?
(Kee-AR-ee)
The Chiari I Malformation is considered a congenital malformation, although there have been some reported cases of an acquired form. It is characterized by a small or misshapen posterior fossa (the compartment in the back of the skull), a reduction in cerebrospinal fluid pathways and a protrusion of the cerebellar tonsils through the bottom of the skull (foramen magnum) into the spinal canal. The tonsils would normally be round but often become elongated as they protrude down the spinal canal. Diagnosis can be difficult because not all patients will have the classical sign of deeply herniated tonsils.

Since the advent of MRI, the incidence of the Chiari I Malformation has risen dramatically. MRI is safe and painless and currently the most reliable means available for diagnosing Chiari Malformations. Chiari Malformations are also known as herniation of the cerebellar tonsils, cerebellar ectopia, hindbrain herniation and Arnold-Chiari malformations.

A German pathologist, Professor Hans Chiari, first described abnormalities of the brain at the junction of the skull with the spine in the 1890's. He categorized them in order of severity, types I, II, III, and IV.

The Chiari type II Malformation is usually found in children with spina bifida or myelomeningocele. Not only is part of cerebellum unusually low and lying below the bottom of the skull, but the brain stem can be malformed in several ways. Types III and IV represent gross herniations of the cerebellum and are very rare.

What are the symptoms?
Many people with the Chiari I Malformation experience no symptoms. When symptoms are present, they usually do not appear until adolescence or early adulthood, but can occasionally be seen in young children. The majority of patients complain of severe head and neck pain. Headaches are often accentuated by coughing, sneezing or straining. Patients may complain of dizziness, vertigo, disequilibrium, muscle weakness or balance problems. Often fine motor skills and hand coordination will be affected.

Vision problems can also occur. Some patients experience blurred or double vision, difficulty in tracking objects or a hypersensitivity to bright lights. Physical examination may reveal nystagmus (involuntary eye movements). Other symptoms include tinnitus (buzzing or ringing in the ear), hearing loss or vocal cord paralysis. Patients may have difficulty swallowing, frequent gagging and choking and, in some cases, sleep apnea may be present.

The Chiari I Malformations may also be associated with other disorders such as hydrocephalus (build up of fluid in the ventricles of the brain) or Syringomyelia.

Syringomyelia is a disorder in which cerebrospinal fluid enters the spinal cord, forming a cavity known as a syrinx. It is recommended that patients diagnosed with a Chiari Malformation have the entire spine imaged to rule out the presence of a syrinx, since it may be a consideration in treatment and prognosis.

Is there a treatment?
Surgical procedures to enlarge the posterior fossa are considered a treatment option for patients with the Chiari I Malformation. Techniques are quite diversified amongst neurosurgeons, and patient responses vary greatly. A successful surgery will alleviate pressure on the neural elements and may result in an improvement of symptoms.

The decision to treat a Chiari Malformation surgically requires careful consultation between patient and physician. Factors to be considered are the patient's current neurological condition and progression of symptoms over a period of time.

Is this condition hereditary?
Research into the risk of inheritance for the Chiari I Malformation is still in its early stages. In some families, more than one member has been documented to have the Chiari I Malformation. Familial recurrences are suggestive of a possible genetic component of the condition, but unfortunately there is no conclusive answer to the question of inheritance at this time. It is currently recommended that only those relatives experiencing symptoms commonly associated with the Chiari I Malformation need undergo investigational procedures.

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