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Chiari I Malformation



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Pathophysiology

Displacement of Cerebellar Tonsils

Dice with Displaced Silver-bell Tassels

Chiari I malformation is a downward displacement by more than four millimeters of cerebellar tonsils through the foramen magnum. It occurs during fetal development. Chiari I malformation differs from Chiari II malformation, in which displacement involves the cerebellum and medulla. One structure involves Chiari I, and two in Chiari II.

Through Foramen Magnum

Foreman Magnum

Chiari I malformation and Chiari II malformation together pass through the foramen magnum. The foramen magnum is one of the several oval or circular openings in the base of the skull.

Congenital

Present-from-birth

Chiari I malformation is a disease that is present from birth.

Signs and Symptoms

Asymptomatic in Childhood

Thumbs-up Kid

Chiari I malformation is typically seen as asymptomatic in children with this disorder. It is commonly found in teenagers or young adults.

Symptomatic in Adults

Symptoms in Adult

Symptomatic Chiari I malformation is typically seen in adult patients, who may present with headache and cerebellar symptoms.

Headaches

Head-egg-lump

Headaches are the most common symptom experienced by patients. These can be located in the back of the head or neck and worsen with sneezing and coughing.

Ataxia

A-taxi

Patients can experience ataxia which may cause difficulty with walking and balancing. A loss of coordination, when it occurs, is due to a loss of muscle control in the arms and legs.

Diagnosis

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MRI

M-R-eyes Machine

Magnetic resonance imaging, strong magnetic fields, and radio waves are used in MRIs to produce detailed images of the inside body. An MRI of the brain can show low-lying and elongated cerebellar tonsils displaced below the level of the foramen magnum. It can detect the displacement of the cerebellum and brainstem displacement, syringomyelia, and craniocervical junction abnormalities.

Association

Syringomyelia

Syringe-spine

Syringomyelia is a cystic cavity in the central canal of the spinal cord that often develops in association with Chiari malformations or after trauma to the spine.

Treatment

Supportive Therapy

Supportive IV Bags

Supportive therapy may include analgesics, physical therapy, and a form of psychotherapy where patients are supported with listening and managing the symptoms. Education may also include information on how to avoid activities that increase intracranial pressure (ICP), e.g., lifting heavy objects and jumping.

Surveillance

Surveillance-camera

Surveillance is a detailed observation. Clinical and MRI surveillance is used in asymptomatic patients with no syringomyelia or CSF flow obstruction. A repeat MRI needs to be done if there's a new sign or symptom related to Chiari I malformation.