Chiari Malformation

- The endoscopic treatment of fourth ventricle outlet obstruction: Report of two children and systematic review
- Impact of Spina Bifida on Sleep Quality: Current Insights
- The Pathogenesis of Chiari Malformation and Syringomyelia: A Case Report and Systematic Review of Current Theories
- Research process, recap, and prediction of Chiari malformation based on bicentennial history of nomenclature and terms misuse
- Management of persistent syringomyelia in patients operated for Chiari Malformation Type 1
- Prenatal diagnosis and postnatal outcome of closed spinal dysraphism
- Bilateral Vocal Cord Abduction Paralysis after C1 Laminectomy in a Child with Type 1 Arnold Chiari Malformation
- Non-invasive intracranial pressure waveform analysis in Chiari Malformation type 1: A pilot trial

The Chiari malformations were first described in 1891 by Hans Chiari, Austrian pathologist (1851-1916).

Two pathologists, Julius Arnold (1835-1915) and Hans Chiari (1851-1916), described a complex clinical and pathological condition involving deformity of the cerebellum and brainstem, in children.

Although Hans Chiari made significant and meaningful contributions to our understanding and classification of hindbrain herniations, others have also contributed to this knowledge. One figure who has been lost to history is Otto Mennicke 1).

While our knowledge of the Chiari malformations continues to improve through the efforts of clinical and basic science researchers, surgeons, and patients, our current understanding of these entities represents a monumental improvement in patient care over a relatively short time period 2).

**Definition**

Chiari malformations are now defined as a spectrum of hindbrain abnormalities involving the cerebellum, brainstem, skull base, and cervical cord.

**Epidemiology**

Together with basilar invaginations, Chiari malformations represent the most common craniocervical junction malformations seen in adults.
Classification

Chiari Malformation Classification.

Etiology

Analysis of unique and powerful Utah genetic resources allowed the identification of 38 strong candidate Chiari malformation predisposition gene variants. These variants should be pursued in independent populations. One of the candidates, a rare HOXC4 variant, was identified in 2 high-risk CM pedigrees, with this variant possibly predisposing patients to a Chiari phenotype with craniocervical kyphosis 31.

Diagnosis

The diagnosis of Chiari malformation typically involves a combination of medical history assessment, physical examination, and medical imaging. Here is an overview of the diagnostic process for Chiari malformation:

Medical History and Symptoms Assessment: The initial step in diagnosing Chiari malformation is a thorough review of the patient’s medical history. The healthcare provider will ask about the patient's symptoms, including headache patterns, neck pain, neurological symptoms, and any other relevant information. It's essential to provide a detailed account of your symptoms, their onset, duration, and any factors that exacerbate or alleviate them.

Physical Examination: A neurological examination will be conducted to assess any signs of neurological dysfunction. The healthcare provider will check for abnormalities in reflexes, muscle strength, coordination, and sensation. Specific neurological signs associated with Chiari malformation may be present.

Imaging Studies:

MRI (Magnetic Resonance Imaging): The gold standard for diagnosing Chiari malformation is an MRI of the brain and cervical spine. An MRI can provide detailed images of the brain, cerebellum, and the spinal cord. It can reveal the presence of a Chiari malformation, its type, and any associated complications, such as syringomyelia (a condition where fluid-filled cavities form within the spinal cord). Additional Tests: Depending on the clinical presentation and findings, additional tests may be ordered to evaluate specific symptoms or complications. These tests may include:

CT (Computed Tomography) Scan: Sometimes, a CT scan may be used in conjunction with MRI to provide additional information. Electroencephalogram (EEG): If seizures or abnormal brain activity are suspected, an EEG may be performed. CSF Flow Studies: In some cases, cerebrospinal fluid (CSF) flow studies using MRI or other techniques may be conducted to assess CSF circulation. Differential Diagnosis: Chiari malformation shares some symptoms with other medical conditions, such as migraines, tension headaches, or other neurological disorders. The diagnostic process may include ruling out these other potential causes through a comprehensive evaluation.

Consultation with Specialists: Depending on the findings and severity of Chiari malformation, patients
Chiari Malformation may be referred to a neurosurgeon or other specialists for further evaluation and management.

It's important to consult with a healthcare professional if you suspect you have Chiari malformation or are experiencing symptoms that could be related to this condition. Early diagnosis and appropriate treatment can help manage symptoms and prevent complications associated with Chiari malformation. Treatment options may include medication, physical therapy, or in some cases, surgical intervention to alleviate pressure on the brainstem and spinal cord.

