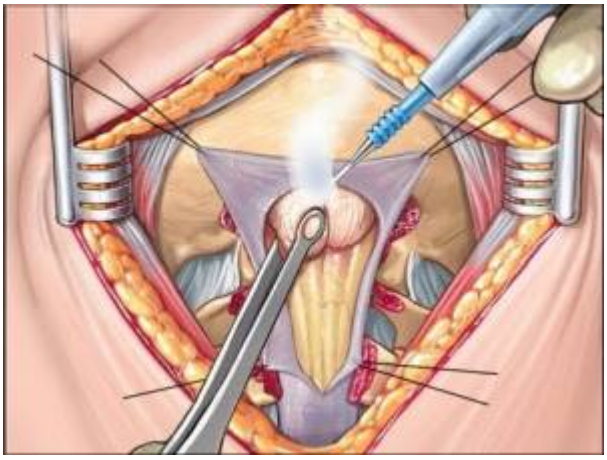
Posterior fossa decompression for Chiari malformation

- A comparison of prone versus sitting position for the surgical treatment of Chiari malformation type I in children
- Resolution of tension pseudomeningocele complicating foramen magnum decompression for Chiari I malformation after ventriculoperitoneal shunt: A case report
- Rehabilitation in a child with Chiari II malformation, lumbosacral meningomyelocele, achondroplasia and impaired respiratory regulation - a case report and literature review
- Syringohydromyelia, syringobulbia and syringocephaly associated with Chiari - I malformation: A case report
- Clinical outcome of different surgical approaches for symptomatic Chiari malformation without syringomyelia: a 13-year retrospective study
- Surgical Treatment of Basilar Invagination without Evident Atlantoaxial Instability (Type B) - A Systematic Review
- Adult Chiari Type I Malformation Presenting as Emergent Acute Respiratory Insufficiency: A Case Report and Literature Review
- Clinical phenotypes among patients with familial forms of Chiari malformation type 1



The most frequently performed operation is posterior fossa decompression (suboccipital craniectomy), with or without other procedures (usually combined with a dural patch grafting and cervical laminectomy of C1, sometimes to C2 or C3). Options for grafts: same incision (pericranium), a separate incision (e.g. or fascia lata), and allograft (avoided by many authors because of dissatisfaction with the ability to provide water-tight closure and because of infectious risks).

Goals of surgery: decompress the brain stem and reestablish the normal flow of CSF at the craniocervical junction.

The most effective therapy for patients with Chiari type I malformation/syringomyelia is surgical decompression of the foramen magnum, however there are non-surgical therapy to relieve neuropathic pain: either pharmacological and non-pharmacological. Pharmacological therapy use drugs that act on different components of pain. Non-pharmacological therapies are primarily based on spinal or peripheral electrical stimulation. It is important to determine the needs of the patients in terms of health-care, social, educational, occupational, and relationship issues, in addition to those

derived from information aspects, particularly at onset of symptoms. Currently, there is no consensus among the specialists regarding the etiology of the disease or how to approach, monitor, follow-up, and treat the condition. It is necessary that the physicians involved in the care of people with this condition comprehensively approach the management and follow-up of the patients, and that they organize interdisciplinary teams including all the professionals that can help to increase the quality of life of patients [1)].

The brainstem width and cervical cord volume showed a modest increase after PFD surgery, although standard deviations were large. A reduction in compression after PFD surgery may allow for an increase in neural tissue dimension. However, clinical relevance is unclear and should be assessed in future studies with high-resolution imaging [2)].

Despite decades of experience and research, the etiology and management of Chiari type 1 deformity (CM-I) continue to raise more questions than answers. Controversy abounds in every aspect of management, including the indications, timing, and type of surgery, as well as clinical and radiographic outcomes.

A review of recent literature on the management of CM-I in pediatric patients was presented by Alexander et al., along with the experience in managing 1073 patients who were diagnosed with CM-I over the past two decades (1998-2018) at Children's National Medical Center.

An accurate and reliable selection of patients based on clinical and neuroimaging findings is paramount for the success of neurosurgical treatment [3)].

Indications

Posterior fossa decompression (PFD) for Chiari malformation type I (CM-I) is indicated in patients with symptomatic hindbrain herniation. The decision to operate is based on clinical presentation, radiological findings, and disease progression.

Indications for Surgery: Symptomatic Chiari Malformation Type I

Debilitating headaches, typically occipital, exacerbated by Valsalva maneuvers (e.g., coughing, straining). Cervicomedullary compression symptoms, including dizziness, dysphagia, dysarthria, sleep apnea, and lower cranial nerve dysfunction. Myelopathic symptoms, such as weakness, spasticity, and balance issues. Chiari Malformation with Syringomyelia

Progressive neurological deficits (e.g., limb weakness, sensory disturbances, bladder dysfunction). Expanding syrinx on serial imaging, even in the absence of severe symptoms. Progression of Symptoms Despite Conservative Management

Worsening clinical picture despite observation. Failure of medical management (e.g., NSAIDs for headaches, physical therapy). Severe CSF Flow Obstruction on Cine MRI

Evidence of impaired cerebrospinal fluid (CSF) dynamics at the foramen magnum. Abnormal or absent posterior subarachnoid space flow. Relative and Contraindications: Asymptomatic CM-I → Usually observed with periodic imaging. Mild symptoms with no progression → Conservative management preferred. Chiari malformation due to secondary causes (e.g., tumors, hydrocephalus) → Address underlying etiology first.

Guidelines

Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Patients With Chiari Malformation: Surgical Interventions

https://www.cns.org/guidelines/browse-guidelines-detail/3-surgical-interventions [4)]

Technique

see Posterior fossa decompression for Chiari malformation surgical technique.

Foramen magnum decompression

see Foramen magnum decompression

The preferred treatment for Chiari type 1 deformity is foramen magnum decompression (FMD), and it is assumed to normalize ICP and craniospinal pressure dissociation.

Observations suggest that anatomical restoration of cerebrospinal fluid pathways by FMD does not lead to immediate normalisation of preoperatively altered pulsatile and static ICP in patients with CMI. This finding may explain persistent symptoms during the early period after FMD [5)].

The purpose of a study was to examine the utility of iMRI in determining when an adequate decompression had been performed.

Patients with symptomatic Chiari I malformations with imaging findings of obstruction of the CSF space at the foramen magnum, with or without syringomyelia, were considered candidates for surgery. All patients underwent complete T1, T2, and cine MRI studies in the supine position preoperatively as a baseline. After the patient was placed prone with the neck flexed in position for surgery, iMRI was performed. The patient then underwent a bone decompression of the foramen magnum and arch of C-1, and the MRI was repeated. If obstruction was still present, then in a stepwise fashion the patient underwent dural splitting, duraplasty, and coagulation of the tonsils, with an iMRI study performed after each step guiding the decision to proceed further.

Eighteen patients underwent PFD for Chiari I malformations between November 2011 and February 2013; 15 prone preincision iMRIs were performed. Fourteen of these patients (93%) demonstrated significant improvement of CSF flow through the foramen magnum dorsal to the tonsils with positioning only. This improvement was so notable that changes in CSF flow as a result of the bone decompression were difficult to discern.

The authors observed significant CSF flow changes when simply positioning the patient for surgery. These results put into question intraoperative flow assessments that suggest adequate decompression by PFD, whether by iMRI or intraoperative ultrasound. The use of intraoperative imaging during PFD for Chiari I malformation, whether by ultrasound or iMRI, is limited by CSF flow dynamics across the foramen magnum that change significantly when the patient is positioned for surgery [6].

C1 laminectomy

see C1 laminectomy for Chiari malformation.

Duraplasty

see Duraplasty for Chiari Malformation.

Complications

Posterior fossa decompression for Chiari malformation complications.

Full-endoscopic technique

Two patients diagnosed with CM were operated on by the full-endoscopic PFD technique. The patients consented to the procedure and to the publication of their image. An endoscope with an oval shaft cross-section with a diameter of 9.3 mm, a working length of 177 mm, a viewing angle of 20°, and a working channel of 5.6 diameters were used. Operative videos were recorded. The surgical steps were easily applied after the clear anatomic landmarks, such as the C1 posterior tubercle and the rectus capitis posterior minor muscles. The patients were followed up for 6 months. Both patients were symptom-free with a significant decrease in Visual Analog Scale score and a good functional outcome assessed by Chicago Chiari Outcome Scale after surgery without any complications.

All the steps of the full-endoscopic technique for PFD described by the authors in their previous human cadaveric study were also feasible on patients with CM [7].

References

1)
Fernández AA, Guerrero AI, Martínez MI, Vázquez ME, Fernández JB, Chesa i Octavio E, Labrado Jde L, Silva ME, de Araoz MF, García-Ramos R, Ribes MG, Gómez C, Valdivia JI, Valbuena RN, Ramón JR. Malformations of the craniocervical junction (Chiari type I and syringomyelia: classification, diagnosis and treatment). BMC Musculoskelet Disord. 2009 Dec 17;10 Suppl 1(Suppl 1):S1. doi: 10.1186/1471-2474-10-S1-S1. PMID: 20018097; PMCID: PMC2796052.

2)
Karamzadeh M, Al Samman MM, Vargas AI, Bhadelia RA, Oshinski J, Barrow DL, Amini R, Loth F. The effect of posterior fossa decompression surgery on brainstem and cervical spinal cord dimensions in adults with Chiari malformation type 1. World Neurosurg. 2023 Sep 30:S1878-8750(23)01382-7. doi: 10.1016/j.wneu.2023.09.112. Epub ahead of print. PMID: 37783305.
3)
Poretti A, Ashmawy R, Garzon-Muvdi T, Jallo GI, Huisman TA, Raybaud C. Chiari Type 1 Deformity in Children: Pathogenetic, Clinical, Neuroimaging, and Management Aspects. Neuropediatrics. 2016 Jun 23. [Epub ahead of print] PubMed PMID: 27337547.
4)
Pattisapu JV, Ackerman LL, Infinger LK, Maher CO, Quinsey C, Rocque BG, Silberstein H, Jackson EM, Jernigan S, Niazi T, Qaiser R, Raskin JS, Vachhrajani S, Bauer DF. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Patients With Chiari Malformation: Surgical Interventions. Neurosurgery. 2023 Oct 1;93(4):731-735. doi: 10.1227/neu.0000000000002635. Epub 2023 Aug 30. PMID: 37646504.
5)
Frič R, Eide PK. Perioperative monitoring of pulsatile and static intracranial pressure in patients with Chiari malformation type 1 undergoing foramen magnum decompression. Acta Neurochir (Wien). 2016 Feb;158(2):341-7. doi: 10.1007/s00701-015-2669-0. Epub 2015 Dec 28. PubMed PMID: 26711284.
6)
Bond AE, Jane JA Sr, Liu KC, Oldfield EH. Changes in cerebrospinal fluid flow assessed using intraoperative MRI during posterior fossa decompression for Chiari malformation. J Neurosurg. 2015 May;122(5):1068-75. doi: 10.3171/2015.1.JNS132712. Epub 2015 Feb 20. PubMed PMID: 25699415.
7)
Dolas I, Unal TC, Yorukoglu AG, Ruetten S, Dolen D, Gulsever CI, Sahin D, Aydoseli A, Sencer A. Full-Endoscopic Technique for Posterior Fossa Decompression in Chiari Malformation. Oper Neurosurg (Hagerstown). 2023 Oct 9. doi: 10.1227/ons.0000000000000875. Epub ahead of print. PMID: 37820101.