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DISCOVERY SCIENTIFIC SOCIETY

Chiari malformations: a review of intervention methods

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ABSTRACT

Treatment of the four Chiari Malformations has varied throughout its short diagnostic history. As one of the more common congenital and acquired cerebral defects, current medical publishing has failed to provide an exhausting report of the morbidity. Most studies are unable to provide a complete image of known therapeutic methods for tackling this comorbidity. Engaging authors choose to write on the general representation of the disease instead of focusing on more specific fields that have not been explored to date. This review prioritises the latest advances in Chiari malformation surgery. The study focuses on the present knowledge of the disease, with special focus on current methods of treatment, including invasive or non-invasive strategies. Appropriate treatment of the condition can reduce the general mortality rate and provide well-being for those born with deformities.

Keywords: Chiari malformations, Arnold Chiari Malformation, cerebrospinal fluid, congenital disabilities, syringomyelia, syrinx, tonsil herniation, spina bifida, neurosurgery

1. INTRODUCTION

Chiari Malformations were classified in the year of 1883 by pathologist Hans Chiari. They represent a significant global issue due their frequency among congenital disorders and associated high mortality and disability rate. CMs are uncommon disorders; however, their diagnosis is becoming more frequent since access to modern imaging techniques has improved (Cociasu et al., 2016). Throughout the flow of time, surgeons and medical professionals would go great distances to treat CM successfully. As the vast majority of CM cases are born deformities, only a quarter of them are diagnosed at birth, with the vast majority diagnosed in adults and late teens. CM can coexist with supposedly unrelated malignancies such as fibrous dysplasia and as part of McCune-Albright syndrome (MAS) (Pan et al., 2018). Choosing a proper therapuetic approach can become more difficult with underlying syndromes as well as in incidental discovery. Decision-making in chronic tonsillar herniation (CTH) in adolescents is complex because many cases are diagnosed incidentally (Vinchon, 2019). Their clinical manifestations vary depending on their individual history and arising complications. While many

newborns will gain from interventional surgery, a conservative approach is better sought out for many. CM are complex disorders that require early recognition and optimal interdisciplinary partnerships to provide extraordinary care for the affected child and parents unit (Fons & Jnah, 2021). This review aims to present the current interventional and non-interventional methods of Chiari malformations and to encourage further research into the topic.

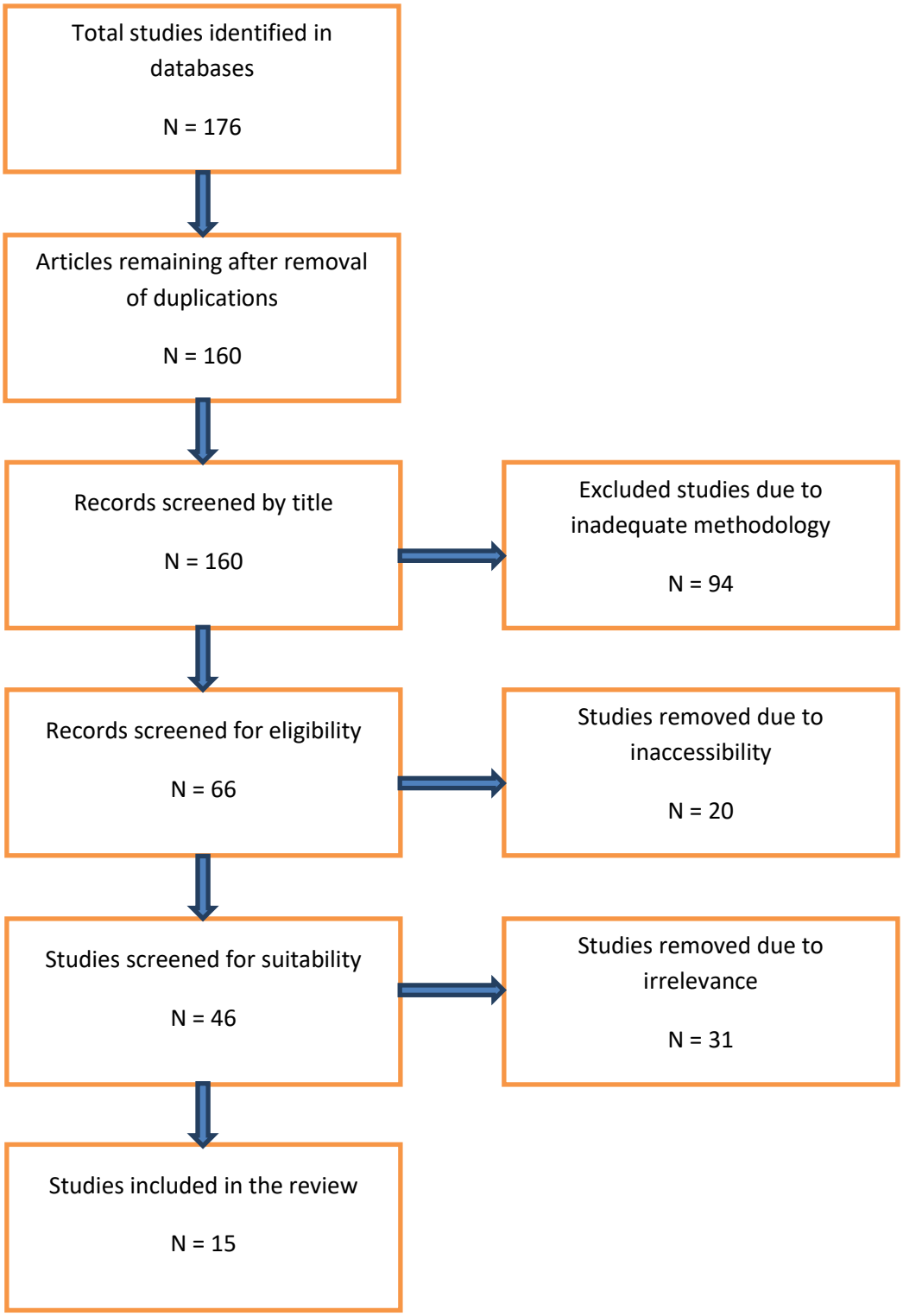

Figure 1 PRISMA flow chart diagram of the study selection process

2. REVIEW METHODS

Our research would not have been possible without the prior engagement of the medical community and their sacrifices. We dedicate this paper to them. This paper provides a comprehensive review of existing literature on Chiari Malformation interventions. Keywords like "Chiari Malformations", "Arnold Chiari Malformation", and "congenital disabilities" were used to search readily available medical sources like PubMed, ScienceDirect, Springer and Google Scholar. The review included articles published between July 2001 to April 2025. The review included clinical research, randomized controlled trials, meta-analyses, observational studies, and systematic reviews. Excluded were publications written in languages other than English, and studies focused on ineffective interventional methods (Figure 1).

3. RESULTS AND DISCUSSION

Chiari malformations are structural abnormalities of the hindbrain where the cerebellum improperly descends into the brainstem. It is pressed through the foramen magnum at the base of the skull and into the spinal canal. CM occurs when the skull is of abnormal shape, often misshapen or of smaller size. While the precise cause has not yet been discovered, it has been linked to genetic factors. Other causes include irregularities during fetal development, injuries, hydrocephalus and a tethered spinal cord. CM can be divided into four types depending on their gravity and anatomical differences.

In CM1, the most commonly occurring type, the cerebellar tonsils protrude through the foramen magnum to the upper cervical canal. Hindbrain anomalies are typically diagnosed in children and young adults. Symptoms typically begin following an increase in pressure build-up during coughing and sneezing. CM1 often coexists with syringomyelia. CM2 is known as the Arnold-Chiari malformation. In ACM both the cerebellum and the brainstem stretch into the brain canal. Symptoms result from coexisting hydrocephalus, corpus callosum agenesis and underdevelopment of both halves of the cerebellum. Cerebellar-tonsillar herniation and kinking set even larger pressure on the spinal canal.

CM3 bypasses the foramen magnum posteriorly, resulting in an encephalocele and spina bifida. The fourth ventricle protrudes into the cervical canal, a rarer congenital condition than the prior. Severe neurological defects appear earlier, causing distress and quickly becoming life-threatening.

CM4, the rarest of them all, involves the cerebellum being underdeveloped. The position of the cerebellum remains anatomical, however, results in other irregularities. The surrounding tissue of the spinal cord is flawed, resulting in the spinal cord's exposure and the emergence of spina bifida.

Treatment

Differences in treatment emerge between classification and the severity of symptoms. Treatment of all cases is considered individually. The priority for most clinical cases is the normalization of cerebrospinal fluid flow at the base of the skull and decompression (Friedlander, 2024). One of the reasons to disregard surgical interference overall in symptom-less cases is the absence of the syrinx. However, the presence of the syrinx does not necessary involve intervention. In such cases the syrinx may resolve without any surgical intervention (Mazzola and Fried, 2003).

Surgical treatment in asymptomatic patients proceeds differently. Asymptomatic patients are only considered for decompressive surgery by 9% of pediatric neurosurgeons (Alden, 2001). Brainstem and cranial nerve dysfunction, hydro-syringomyelie and kyphosis directly linked to CM-1 are all indications from general surgery (Siasios et al., 2012). Current medical knowledge allows us to consider various surgical approaches for the same pathology. The most influential approaches are listed and described as follows. Conservative courses of action are under consideration as their prevalence are the topic of debate regularly.

Prenatal Surgical Closure

While associated with additional fetal and unnecessary maternal risks, early fetoscopic myelomeningocele correction can prevent further fetal underdevelopment if operated on. Mental development and motor score at age 30 months can be greatly improved when undertaking surgery early on in-utero (P=0.007) (Adzick et al., 2011), thus decreasing the overall need for later hospitalization. The surgery is typically undertaken between 19 and 26 weeks of gestation, closing the opening in the spine before the spinal cord matures often has better results than operating after birth. It is performed under general anesthesia, after uteral incision. Only then it’s possible to remove the protrussion of the meninges and seal the spinal cord. While maternal deaths are uncommon, however, not nonexistent. Complications of pregnancy post-surgery include placental abruption and oligohydramnios.

A study showed the need for cerebrospinal fluid shunting was significant at 68% within 12 months, yet diametrically lower compared to the post-natal surgical group, standing at 98% (relative risk, 0.70; 97.7% confidence interval [CI], 0.58 to 0.84; P<0.001) (Adzick et al., 2011). This method is primarily used in the treatment of CM2, specifically the Arnold-Chiari malformation.

Obex plugging

Closing off the central canal at the obex is a method often performed in conjunction with posterior fossa decompression. It reduces pressure on the spinal cord, allowing symptoms to deescalate. The obex is a structure of triangular nature found deep at the bottom of the fourth ventricle. In this procedure, it is closed off with a muscle plug (or related material) to decompress the spinal column further. CM-1 and acquired CM-1 have been positively affected by this treatment, with an outcome of even 70% improvement in overall points granted for treatment. Additionally, no deaths occurred during this method of treatment (Siasios et al., 2012).

Cervical laminectomy

A procedure directly resulting in decompression of the spinal cord, cervical laminectomy is undertaken in symptomatic as well as asymptomatic patients. Removing a segment of the C1 vertebrae relieves pressure set by the skeletal structure. It alleviates symptoms by restoring the proper flow of CSF. Patients undergoing cervical laminectomy with tonsil herniation between 11-14mm showed improvement in two clinical groups of 80% and 92% (Hwang et al., 2023). Both clinical groups observed improved syringomyelia, and no complications of surgical nature were present in either cohort. The decrease in the volume of the syrinx post-surgery was observed regularly with values averaging between 5.9% - 7.5% per month (Hwang et al., 2023). Cervical laminectomy is frequently critiqued of being an instable intervention with multiple studies showing evidence of radiographic instability post-surgery. However, there is no proof of clinical instability as no patient proved to show clinical findings with the need for cervical fixation following surgery (Lam et al., 2008).

Posterior Fossa Decompression with or without duraplasty

Removing the segment of the occipital bone directly posterior to the foramen magnum, part of the posterior fossa, results in widening the space for the herniated tonsils and brainstem. Reduction of constriction allows for the anatomical flow of CSF, resulting in the regression of symptoms. The surgery can be performed with or without duraplasty. Duraplasty is the surgical expansion of the dura post-surgery. Currently minimally invasive PFD to the craniocervical junction is performed to avoid spinal biomechanic adjustments (Agudelo-Arrieta et al., 2023). Duraplasty included the use of autologous grafts of cervical fascia and non-autologous grafts. No complications are observed as well as no significant differences noted between the two methods.

Stenting of the fourth ventricle

Placing a stent from the fourth ventricle to the cervical subarachnoid space allows for the improvement of CSF flow and alleviation of symptoms. The catheter relieves the pressure build-up at the lowest cavity and removes accessory CSF accumulation. Fourth ventricular subarachnoid stenting is possible when PFD does not bring the expected results. As most CM-I patients have associated syringomyelia, CSF outflow obstruction is a fairly common disorder. The vast majority of PFD surgery patients experience alleviation of symptoms; however, persistent syrinx-associated symptomatology does occur occasionally (Han et al., 2023). Such cases may require the placement of the fourth ventricular stent because of the blockage of the fourth ventricular outflow. Of 41 patients requiring a Chiari re-exploration, eight stents have been placed regardless of syringomyelia (Han et al., 2023). Fourth ventricular stent placement serves as the final therapeutic option. By decreasing the syrinx's length, it ends the patient's treatment process.

Conservative approaches

Conservative treatment includes a combination of prescriptive medication, physiotherapy and selective aerobic exercises. Limiting straining physical activities can also benefit health. Due to the risks involved with direct surgery, the option of a conservative approach is ever more standard. Certain studies have found that for selected adult patients with CM-1, non-interventional treatment is favored (ASV=16.7 μL) (Abdallah et al., 2022). This method of non-surgical treatment method was only ineffective in 5.6% of cases, patients having undergone surgery.

The vast majority of asymptomatic cases should be approached conservatively, as in cases without progression, the risk of surgical complications outweighs the benefits. The natural history of those mild asymptomatic cases determines that the decision of

decompression is only possible when considering the patient's severity of symptoms and individual presentation. Regular monitoring of CM progression is always necessary, regardless of the approach.

A second study finds that solely conservative treatment of CM1 is not adequate, particularly in cases with scoliosis and syringomyelia (Rigo et al., 2013). Its only worthy uses are pre-operative and post-operative, as spontaneous regression of CM1 in children is uncommon. Moreover, the authors highly discourage the practice of surgery abstention.

Revision surgery

Reoperations are not uncommon in hindbrain anomalies as they are performed in all four Chiari malformations. The need for reintervention is caused not only by insufficient decompression and surgeon error but also because of secondary herniation and syringomyelia in cases properly handled. Oftentimes, new neurological symptoms were not caused by inadequate decompression as the CSF pressure was within range (Mazzola et al., 2003). Dural grafts have been proven to be the root of symptom recurrence, as well as anterior basilar invagination. Certain studies have found arachnoid cysts undergoing cyst fenestration and shunting to be the reason for revision surgery. Meanwhile, other researchers proved that arachnoid adhesions compress the spinal canal at C1, resulting in a tethered spinal cord. Overall, the restoration of CSF flow is considered the primary goal in decompression and revision surgery (Table 1).

Table 1 Comparison of Treatment Methods of Chiari Malformations

Treatment method	Description
Prenatal surgical closure	Early in-utero surgery prevents further fetal underdevelopment, decreases hospitalization rates and complications.
Obex plugging	Allows for the deescalation of symptoms, reduces spinal cord pressure by closing off the central canal at the obex.
Cervical laminectomy	Surgical removal of cervical vertebrae, results in pressure relief. Post-surgery decrease of syrinx volume observed.
Posterior Fossa Decompression	Removal of a segment of the occipital bone. Minimally invasive, results in constriction reduction.
Stenting of the fourth ventricle	Subarachnoid stenting reestablishes CSF flow, relieves neurological symptoms and decreases the length of the syrinx.
Conservative approaches	Highly controversial methods of questionable success rate. Lower immediate complication risk due to non-surgical approach.
Revision surgery	Performed when insufficient decompression is administered, carried out to restore the proper flow of CSF.

4. CONCLUSION

Specialists continue to question the techniques of surgical approach to Chiari Malformations. They often discuss which method of intervention will bring the highest yield with the lowest amount of complications. As medical arbitration continues to evolve, the prospect of what the future holds is ever more promising, with the following intervention methods surprising all of us.

At the time of submission, no surgical procedure is considered superior to others, as further research is needed on the topic, which is why a conservative approach to Chiari Malformations is considered. However, greater involvement in the topic is required to understand its importance and the applications of conservative actions fully. Hopefully, the future will resolve every problem that therapists currently face, and the proper direction of treatment will be more precise and consistent.

Personal considerations

Worth considering is the impact of the circumference of the foramen magnum; however, no notable scientific correlation is determined between the size and prevalence of the four Chiari syndromes.

Author's Contributions

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All authors have read and agreed with the published version of the manuscript.

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Not applicable.

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Conflict of interest
The authors declare that there is no conflict of interest.

Data and materials availability
All data associated with this study will be available based on the reasonable request to corresponding author.

REFERENCES

1. Abdallah A, Rakip U. Conservative treatment of Chiari malformation Type I based on the Phase-Contrast Magnetic Resonance Imaging: a Retrospective study. World Neurosurg 2022;163:e323–34. doi: 10.1016/j.wneu.2022.03.126
2. Adzick NS, Thom EA, Spong CY, Brock JW, Burrows PK, Johnson MP, Howell LJ, Farrell JA, Dabrowiak ME, Sutton LN, Gupta N, Tulipan NB, D'Alton ME, Farmer DL. A Randomized Trial of Prenatal versus Postnatal Repair of Myelomeningocele. N Engl J Med 2011;364(11):993–1004. doi: 10.1056/nejmoa1014379
3. Agudelo-Arrieta M, Henao-Romero S, Mendez-Gutierrez A, Sardi JP, Berbeo ME, Diaz-Orduz R. Minimally invasive approach to decompression for chiari malformation type 1. J Minim Invasive Spine Surg Tech 2023;8(1):64–73. doi: 10.21182/jmisst.2023.00647
4. Alden TD, Ojemann JG, Park TS. Surgical treatment of Chiari I malformation: indications and approaches. Neurosurg Focus 2001;11(1):1–5.
5. Cociasu I, Pastia M, Davidescu I, Buraga I, Popescu BO. Chiari malformation type i – case report and review of literature. Romanian J Nurol 2016;15(3):138–42. doi: 10.37897/rjn.2016.3.6
6. Fons K, Jnah AJ. Arnold-Chiari Malformation: Core concepts. Neonat Network 2021;40(5):313–20. doi: 10.1891/11-t-704
7. Friedlander RM. Congenital and acquired chiari syndrome. N Engl J Med 2024;390(23):2191–8. doi: 10.1056/nejmra2308055
8. Han RK, Medina MP, Giantini-Larsen AM, Chae JK, Cruz A, Garton ALA, Greenfield JP. Fourth ventricular subarachnoid stent for Chiari malformation type I–associated persistent syringomyelia. Neurosurg Focus 2023;54(3):E10. doi: 10.3171/2022.12.focus22633
9. Hwang JK, Park EK, Shim KW, Kim DS. Effectiveness of C1 laminectomy for chiari malformation Type 1: posterior fossa volume expansion and Syrinx-Volume decrease rate. Yonsei Med J 2023;64(3):191. doi: 10.3349/ymj.2022.0506
10. Lam FC, Irwin BJ, Poskitt KJ, Steinbok P. Cervical spine instability following cervical laminectomies for Chiari II malformation: a retrospective cohort study. Childs Nerv Syst 2008;25(1):71–6. doi: 10.1007/s00381-008-0694-5

11. Mazzola CA, Fried AH. Revision surgery for chiari malformation decompression. Neurosurg Focus 2003;15(3):1–8. doi: 10.3171/foc.2003.15.3.3
12. Pan KS, Heiss JD, Brown SM, Collins MT, Boyce AM. Chiari I Malformation and basilar invagination in fibrous dysplasia: Prevalence, mechanisms, and clinical implications. J Bone Miner Res 2018;33(11):1990–8. doi: 10.1002/jbmr.3531
13. Rigo M, Janssen B, Campo R, Tremonti L. Conservative treatment of juvenile with Chiari I malformation, syringomyelia and scoliosis. Two case reports. Scoliosis 2013;8(S1). doi: 10.1186/1748-7161-8-s1-o52
14. Siasios J, Kapsalaki EZ, Fountas KN. Surgical Management of Patients with Chiari I Malformation. Int J Pediatr 2012;2012:1–10. doi: 10.1155/2012/640127
15. Vinchon M. Surgery for Chiari 1 malformation: the Lille experience. Childs Nerv Syst 2019;35(10):1875–80. doi: 10.1007/s00381-019-04242-9