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Original Article

Stealth cranioplasty in symptomatic adult chiari 1 malformations: Experience at Bangabandhu Sheikh Mujib Medical University

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ABSTRACT

Background: The surgical management of symptomatic adult Chiari malformation type 1 (CM1) with or without syringomyelia (SM) continues to be a dilemma considering the outcomes.

Objectives: The study aimed to compare the clinical outcomes between stealth cranioplasty (SC) and one of the most commonly practiced standard procedures, posterior fossa decompression with duraplasty (PFDD). Clinical outcomes between SC and another procedure posterior fossa decompression (PFD) were also compared.

Methods: This comparative cross-sectional study was carried out on 37 males and 16 females symptomatic adult CM1 patients, ranging from 18 to 47 years of age from June 2019 to May 2021. Clinical outcomes were assessed, compared, and analyzed in terms of changes in clinical symptoms and signs, chicago chiari outcome scale (CCOS) score, and occurrence of complications.

Results: Of the 53 patients, 23, 19 and 11 underwent SC, PFDD, and PFD, respectively. There were no significant post-operative changes in symptoms and signs among groups except changes in limb weakness between SC and PFDD (P=0.004). Considering average CCOS score, SC performed better only than PFDD (P=0.003), while category-wise SC was better than both PFDD (P=0.004) and PFD (P=0.010). Considering complications, the PFDD group had a significantly higher rate of complications than the SC group (P=0.001), while there was no significant difference in the rate of complications between the PFD and SC groups.

Conclusion: SC was found to have better clinical outcomes than the PFDD and PFD groups as a technique.

Keywords: chiari malformation type 1, syringomyelia, stealth cranioplasty, chicago chiari outcome scale

INTRODUCTION

The basic pathology of chiari 1 malformation (CM1) remains controversial and somewhat not well understood. Despite enormous research and discussions, there still remain difficulties in defining the terminology, nosology, etiology, and treatment of CM1 and the outcome remains dubious as ever since the first description by Hans Chiari in 1891.[1,2] CM1 is described as caudal displacement of the cerebellar tonsils below the level of the foramen magnum with or without syringomyelia. Radiologically, based on magnetic resonance imaging (MRI), CM1 is defined as descent of the cerebellar tonsils > 5 mm beyond the foramen magnum in adults.[2-4] The prevalence has been estimated to be 0.24%–3.6% of the population. However, the annual incidence of symptomatic surgical candidates is substantially low, merely 0.06%. Two peaks of ages of incidence are observed, between the ages of 8 and 9 years in children and between 41 and 46 years in adults.[5,6]

The most recognized theory regarding the pathophysiology of CM1 with frequently accompanying syringomyelia is stated to be the discrepancy between small posterior fossa and

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HIGHLIGHTS

1. **Different symptoms and signs of CM1 have different rates of improvement and vary widely.**
2. **Complications following CM1 surgery vary and depend on surgical techniques.**
3. **A common management protocol for CM1 which would be a 'gold standard' or 'one-size-fits-all' with uniform outcome seem challenging.**
4. **Stealth cranioplasty can be a good option for CM1 surgery with good clinical outcomes and avoiding complications.**

overcrowded neural structures leading to cerebellar ectopia. Newer theories emphasize on inadequate CSF flow around the craniocervical junction and hampering the CSF dynamics between the cranial and spinal compartments.[3, 5, 7, 8] Heterogeneity of manifestations poses a major challenge. Myriad of manifestations and findings can be seen, especially in presence of other associated disorders. Neurological signs are mostly due to involvement of the brainstem, spinal cord and cerebellum.[7, 9]

Since the first attempt of surgery in 1930, most patients with CM1 show benefits through different types of surgical procedures that are practiced today.[10, 11] However, the surgical techniques for symptomatic CM1 are diverse and outcomes are debatable, especially in presence of syringomyelia and other associated disorders. Owing to the diversity in clinical and image findings, it is difficult to set any definite guideline for the choice of surgical techniques and thus, numerous techniques have been innovated.[8, 9, 12] With the basic goal of decompressing the posterior fossa and restoring CSF flow and dynamics, most of the common surgical procedures comprise a suboccipital craniectomy with the removal of the posterior arch of C1. Yet, there are arguments regarding the extent of bone removal and additional measures taken along with it.[13, 14] Per operatively, the dura, the arachnoid, and the cerebellar tonsils can be managed in several ways. Options for the dura include keeping it intact with removal of the constricting band only, dural scoring, resection of the outer layer of the dura, opening the dura and leaving it open, and performing a duraplasty.[15-17] Arachnoid manipulation varies from leaving that intact or opening and resecting it.[15, 16, 18] Different surgeons handle tonsils differently by not touching them, dissecting to separate them, shrinking by bipolar coagulation or subpial resection.[15, 19, 20]

Few years back, we developed a surgical technique, the "Stealth Cranioplasty (SC)".[21-23] We addressed some aspects of pathophysiology to overcome some limitations related to outcomes seen in the traditional surgical approaches by combining and modifying some of the techniques together, particularly to reduce complications and recurrence by performing a smaller craniectomy, arachnoid preserving linear durotomy and duraplasty by investing layer of the deep cervical fascia, a titanium mesh cranioplasty in the shape of the cockpit of a stealth bomber and finally, dural graft tenting with the titanium implant. However, a full proof procedure is yet to come.

In this study, we compared our surgical technique, the SC with the most common and standard surgical technique of the posterior fossa decompression with duraplasty (PFDD) in terms of changes in clinical symptoms and signs, scoring of chicago chiari outcome scale (CCOS) and occurrence of complications. We also compared the results of these parameters in our technique with those in another commonly practiced and standard surgical procedure, the posterior fossa decompression (PFD).

METHODS

Patient selection

After obtaining approval from the institutional review board (IRB) of Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh, patients were recruited for this cross-sectional observational study from June 2019 to May 2021. All consecutive adult symptomatic chiari malformation patients with or without syringomyelia having a minimum three-month post-operative follow up were evaluated. Patients aged >18 years having preoperative diagnosis of CM1 confirmed by tonsillar descent >5 millimeter with or without syringomyelia having symptoms and signs related to CM1 who had undergone any of the three surgical procedures, two commonly practiced standard procedures of PFDD and PFD, and the SC that we introduced, were included in the study. Patients having associated anomalies other than CM1 like basilar

invagination, atlanto-axial dislocation or hydrocephalus, not having a minimum of three-month follow up clinically and radiologically, and not giving consent to take part in the study, were excluded.

Procedural details

For surgery, informed written consent was obtained from the patients or their legal guardians. Three procedures were performed depending on surgeon's choice. Posterior fossa decompression (PFD) included suboccipital craniectomy (3 cm X 3 cm approx.) and C1 laminectomy with removal of the dural band when present. In posterior fossa decompression with duraplasty (PFDD), best possible watertight duraplasty with pericranium or fascia lata was accomplished following 'Y' shaped dural opening after the suboccipital craniectomy and C1 laminectomy. During the dural opening, it was the surgeon's choice to open the arachnoid or not. However, the tonsils were not disturbed on any occasion. In brief, the stealth cranioplasty (SC), which we described earlier in details, comprised of 3 cm X 3 cm suboccipital craniectomy, C1 laminectomy, midline linear arachnoid preserving durotomy, duraplasty with superficial layer of the deep cervical fascia, pre-shaped 5 cm X 5 cm titanium mesh cranioplasty shaped into the cockpit of a Stealth bomber fixed with screws and tacking of the duraplasty with the titanium mesh.

All the patients were evaluated three months post-operatively for changes in symptoms like occipital headache, neck pain, arm pain, limb weakness, wasting, paraesthesia and gait disturbance as well as changes in neurological signs like weakness of limbs, wasting, dissociated sensory loss, gait disturbance and cerebellar signs. Patients were evaluated by CCOS to measure outcome and complications were assessed by observing infection, cerebellar sagging, CSF leak, hydrocephalus or pseudomeningocele.

Statistical analysis

Parametric data were expressed as mean (standard deviation, SD) and compared using t-test and ANOVA. Non-parametric data were expressed as medians, and categorical variables were compared using Chi-square test. All statistical analysis was performed using SPSS 23 software. A *P* value <0.05 was considered to be statistically significant.

RESULTS

Overview

This study was conducted on 53 adults symptomatic CM1 patients with or without syringomyelia where 11, 19 and 23 patients underwent PFD, PFDD and SC, respectively. Average age of the 37 male and 16 females enrolled in the study was 30.4 (7.5) years, respectively ranging from 18 to 47 years of age. All the patients were followed up both pre- and post-operatively to assess outcome clinically with history and neurological examination as well as observing the complications.

TABLE 1 Post-operative change in symptoms in posterior fossa decompression with duraplasty (PFDD) and stealth cranioplasty (SC)

Symptoms	Surgery		Change in symptoms		*P**
		Total	Improved	Remained same	
		n (%)	n (%)	n (%)	
Occipital headache	PFDD	13 (68.4)	8 (61.5)	5 (38.5)	0.287
	SC	10 (43.5)	9 (90.0)	1 (10.0)	
Neck pain	PFDD	17 (89.5)	11 (64.7)	6 (35.3)	0.823
	SC	18 (78.2)	12 (66.7)	6 (33.3)	
Arm pain	PFDD	6 (31.6)	3 (50.0)	3 (50.0)	0.413
	SC	2 (8.7)	1 (50.0)	1 (50.0)	
Paresthesia	PFDD	16 (84.2)	4 (25.0)	12 (75.0)	0.764
	SC	23 (100.0)	8 (34.8)	15 (65.2)	
Limb weakness	PFDD	15 (78.9)	2 (13.3)	13 (86.7)	0.004
	SC	21 (91.3)	14 (66.7)	7 (33.3)	
Gait disturbance	PFDD	4 (36.4)	1 (25.0)	3 (75.0)	0.806
	SC	5 (21.7)	2 (40.0)	3 (60.0)	
Wasting	PFDD	10 (100.0)	0 (0.0)	10 (100.0)	-
	SC	16 (100.0)	0 (0.0)	16 (100.0)	

*Chi-square test

TABLE 2 **Post-operative change in symptoms in posterior fossa decompression (PFD) and stealth cranioplasty (SC)**

Symptoms	Surgery		Change in symptoms		*P**
		Total	Improved	Remained same	
		n (%)	n (%)	n (%)	
Occipital headache	PFD	6 (54.5)	5 (83.3)	1 (16.7)	0.698
	SC	10 (43.4)	9 (90.0)	1 (10.0)	
Neck pain	PFD	10 (90.9)	5 (50.0)	5 (50.0)	0.646
	SC	18 (78.0)	12 (66.7)	6 (33.3)	
Arm pain	PFD	1 (9.0)	0 (0.0)	1 (100.0)	0.662
	SC	2 (8.0)	1 (50.0)	1 (50.0)	
Paresthesia	PFD	9 (81.8)	1 (11.1)	8 (88.9)	0.368
	SC	23 (100.1)	8 (34.8)	15 (65.2)	
Limb weakness	PFD	10 (90.9)	3 (30.0)	7 (70.0)	0.125
	SC	21 (91.0)	14 (66.7)	7 (33.3)	
Gait disturbance	PFD	2 (18.1)	0 (0.0)	2 (100.0)	0.887
	SC	5 (21.2)	2 (40.0)	3 (60.0)	
Wasting	PFD	9 (100.1)	0 (0.0)	9 (100.0)	-
	SC	16 (100.1)	0 (0.0)	16 (100.0)	

*Chi-square test was done.

Pre-operative data

Pre-operatively, out of 53 patients in three groups, 29 (54.7%) patients had occipital headache, 45 (84.9%) had neck pain, 9 (17.0%) had arm pain, 48 (90.6%) had paraesthesia, 46 (86.8%) had weakness of limbs, 35 (66.0%) had wasting and 11 (20.8%) had gait disturbance. Pre-operative neurological signs comprised of weakness of limbs in 46 (86.8%), dissociated sensory loss in 47 (88.7%), gait disturbance in 12 (22.6%), wasting in 35 (66.0%) and cerebellar signs in 3 (5.7%) patients.

Post-operative data

Regarding symptoms among the groups, none of the symptoms improved significantly except only the weakness of limb which improved significantly (*P* 0.004) in SC group than the PFDD group **(TABLE 1 and 2)**. None of the neurological signs improved significantly in any of the groups. In addition, no patients having wasting and cerebellar signs improved in any of the groups **(TABLE 3 and 4)**.

The post-operative average CCOS score in SC group (13.0 ± 1.8) was higher than the PFD (11.6 ± 1.1) and PFDD (11.4 ± 1.3) groups which was significant only between PFDD and SC (*P*=0.003). Assessing post-operative outcome with CCOS score based on the type of surgery showed most of the patients in PFD (81.8%) and PFDD (78.9%) groups achieved functional outcome (CCOS 9-12) while most of the patients in SC group (65.2%) achieved excellent outcome (CCOS 13-16).

TABLE 3 **Post-operative change in neurological findings in posterior fossa decompression with duraplasty (PFDD) and stealth cranioplasty (SC)**

Symptoms	Surgery		Change in symptoms		*P**
		Total	Improved	Remained same	
		n (%)	n (%)	n (%)	
Limb weakness	PFDD	15 (78.9)	2 (13.3)	13 (86.7)	0.073
	SC	21 (91.3)	10 (47.6)	11 (52.4)	
Dissociated sensory loss	PFDD	16 (84.2)	4 (25.0)	12 (75.0)	0.92
	SC	22 (95.7)	7 (30.4)	15 (65.2)	
Gait disturbance	PFDD	4 (21.1)	0 (0.0)	4 (100.0)	0.631
	SC	6 (26.1)	2 (33.3)	4 (66.7)	
Wasting	PFDD	10 (52.6)	0 (0.0)	10 (100.0)	-
	SC	16 (69.7)	0 (0.0)	16 (100.0)	
Cerebellar signs	PFDD	1 (5.3)	0 (0.0)	1 (100.0)	-
	SC	2 (8.7)	0 (0.0)	2 (100.0)	

*Chi-square test

TABLE 4 Post-operative change in neurological findings in posterior fossa decompression (PFD) and stealth cranioplasty (SC)

Symptoms	Surgery		Change in symptoms		*P**
		Total	Improved	Remained same	
		n (%)	n (%)	n (%)	
Limb weakness	PFD	10 (90.9)	2 (20.0)	8 (80.0)	0.279
	SC	21 (91.0)	10 (47.6)	11 (52.4)	
Dissociated sensory loss	PFD	9 (81.8)	1 (10.1)	8 (88.9)	0.458
	SC	22 (95.0)	7 (30.4)	15 (65.2)	
Gait disturbance	PFD	2 (18.1)	0 (0.0)	2 (100.0)	0.999
	SC	6 (26.2)	2 (33.3)	4 (66.7)	
Wasting	PFD	9 (81.8)	0 (0.0)	9 (100.0)	-
	SC	16 (69.0)	0 (0.0)	16 (100.0)	
Cerebellar signs	PFD	0 (0.0)	0 (0.0)	0 (100.0)	-
	SC	2 (8.0)	0 (0.0)	2 (100.0)	

*Chi-square test

Comparing the outcome with CCOS category wise, SC group was significantly better than both PFD (*P*=0.010) and PFDD (*P*=0.004) groups **(TABLE 5)**.

TABLE 5 Category-wise post-operative chicago chiari outcome scale in PFDD and SC groups, and PFD and SC groups

Surgery	CCOS*		*P*†
	9-12	13-16	
PFDD vs SC			
PFDD	15 (78.9)	4 (21.1)	0.004
SC	8 (34.8)	15 (65.2)	
PFD vs SC			
PFD	9 (81.8)	2 (18.2)	0.010
SC	8 (34.8)	15 (65.2)	

*4 = Incapacitated outcome; 5– 8 = Impaired outcome; 9 – 12 = Functional outcome; 13 – 16 = Excellent outcome.
†Chi-square test

Post-operative complications were very high in the PFDD group. Nine (47.4%) out of 19 patients in this group had complications in the form of infection, cerebellar sagging, CSF leak, HCP and pseudomeningocele, and five of these patients had CSF related complications. In the PFD group, only two (18.2%) had complications in the form of cerebellar sagging, while in the SC group there was no complication in any form and the findings were significant only between SC and PFDD groups (*P* =0.001).

DISCUSSION

CM1 is recognized as caudal ectopia of the cerebellar tonsils more than 5 mm beyond the foramen magnum. Both clinical and radiological findings are integral parts of the diagnosis and more importantly for decision making in management as well as for assessing the outcome. CM1 is mostly a congenital disorder where transgression occurs around the cranio-cervical junction. Shallow posterior fossa along with disparity of CSF flow and dynamics between the cranial and spinal compartments lead to tonsillar herniation. This disparity of CSF circulation following tonsillar impaction at the foramen magnum as well as discrepancy of CSF circulation from the subarachnoid space into the spinal cord parenchyma are also responsible to bring about the formation of syringomyelia.[2, 3, 6]

Bony decompression is the recognized basic surgery for CM1 as well as for syringomyelia that is well accepted and commonly practiced. With the goal of decompressing the crowded posterior fossa as well as the craniocervical junction, and reestablishing the CSF flow and dynamics there, combination of different surgical techniques has evolved. They range from bony decompression with or without duraplasty; preserving or breaching the arachnoid; not touching, dissecting or resecting the tonsils; dural graft tenting or performing cranioplasties.[15-23] Each of the surgical procedures has their own debatable advantages and disadvantages. All the techniques ultimately are adapted to alleviate the symptoms and signs, manage syringomyelia and reduce complications. This study, is the first of its kind to be conducted at BSMMU. The surgical technique that we innovated is basically a combination of different procedures that are generally practiced with the aim to enlarge the posterior fossa and maintain the newly created space, reestablish CSF flow and dynamics around the foramen magnum, prevent recurrence, and avoid complications. Although the present surgical

techniques yield better results than before, outcomes of different modalities of surgery for CM1 are varied owing to lack of absolute measurement standard. However, our technique, the Stealth cranioplasty, seems to be a good one considering its outcome compared to the traditional standard surgical practices. In this study, we compared the clinical outcomes between the traditional standard procedure of PFDD and our technique of the SC. We also compared the clinical outcomes between another standard procedure of PFD and our innovated technique of SC.

Different studies, show that most of the adult CM1 patients are affected in their fourth decades and in most of the studies, females outnumbered males.[24-26] The demographic findings were different in our study. The racial difference may have some influence on the dimensions of the posterior fossa and more importantly on the diameter of the foramen magnum which might have caused relatively early manifestations in the patients. For the male dominance, possibility is that males in Bangladesh start to work at an early age and most of them are manual heavy workers which may affect the existing problems to manifest more in them. On the other hand, females in Bangladesh are very reluctant to go to physicians unless the disease process becomes unbearable.

The late presentation with longer duration of symptoms and signs in our study reflects the negligence of patients as well as financial constraints in some cases although more than three years of symptom duration are not uncommon.[25, 27] Duration of symptoms has profound influence over outcome as literatures show that the longer the duration of symptoms, the worse is the prognosis. This stresses the urgency to diagnose and treat at the earliest to prevent or even to reverse the deficits in some cases.[2, 28]

CM1 patients usually present with myriad of symptoms, especially when they have associated SM and heterogeneity of presentations continually pose mystery. Symptoms and signs typically arise from impairment of functions involving the cerebellum, brain stem or spinal cord, alone or in combinations particularly when associated syringomyelia is present. In CM1 patients, most studies labelled short-lived valsalva-type maneuver provoked headache, neck pain, arm pain, paraesthesia, numbness, muscle weakness, and gait disturbance as the most common symptom. Among the neurological findings, weakness of limbs, dissociated sensory loss, gait disturbance, muscle wasting and cerebellar signs are the common ones.[2, 5, 29] Following surgery, post-operative improvements of symptoms and signs are reported to range from 73.6-88.6%.[24, 30] Post-operatively, findings of this study show that almost all the symptoms and signs improved evidently. The better outcome of symptoms and signs were seen in cases of SC than in the other two groups in this series although statistical significance between the groups could not be established. However, direct comparison of surgical outcome with previous studies would be non-judicious as the parameters of evaluation of clinical conditions, types of surgeries and time of follow ups are different in different studies from ours.

CCOS was introduced in 2012 and was validated later as an outcome assessment tool for post-operative CM1 patients.[31, 32] CCOS is being used widely to assess post-operative outcome of CM1 patients and has become a reliable tool.[25, 27, 33, 34] Post-operative outcome can be measured with CCOS in 2 ways; by assessing improvement with the overall score and by categorizing the patient into functional outcome groups based on that score. Most of the studies on adult CM1 patients show that the average CCOS that could be achieved ranges between 13 to 14.5.[25, 28, 33-35] Most of the patients in different series succeeded in achieving excellent outcome on average with good CCOS. The average score or the groups that the patients fall varies depending on several factors like the duration of symptoms, variability of symptoms, abundance of surgical techniques or range of follow up periods.[28, 33-35] Altogether most of the patients in our series had functional outcome considering SC alone, two third of the patients had excellent outcome. In most of the other series, the general outcome with CCOS improved gradually with time and varied noticeably depending on different factors. The overall surprising low number of excellent outcomes in our patients can be attributed to short period of follow up which could have been better if there would be longer follow ups. The relatively better outcome in SC group than the other two groups might be due to the absence of any complications as well as formation and maintenance of more space in the posterior fossa in this group and better maintenance of the CSF dynamics at the craniovertebral junction.

Complications are common following surgery for CM1 as in any other surgery. The rates vary from center to center and are largely dependent on the procedure performed and certain patterns of complications can be recognized depending on techniques. Complication rate varies from zero complication to as high as 33.3% in different series.[25, 27, 30, 33, 34, 36] Common complications include cerebrospinal fluid leak, pseudomeningocele, meningitis, wound infection, hydrocephalus, subdural collections, and often a less common cerebellar slump or ptosis.[26, 27, 30, 34, 37] It is well established that the complication rates, particularly the CSF related ones occur more following procedures that breach the arachnoid, while extradural or arachnoid preserving procedures are evidently better in avoiding CSF related complications.[27, 29, 30, 34, 38]

The high number of CSF related complications in the PFDD group can easily be linked to the breach of the arachnoid during surgery. The cerebellar slumps in the PFD group were most likely due to extra-large craniectomies than usual. The SC group did not have any complications which marks the superiority of this technique over the other two. It is always better to have less complications but it is best to avoid those. We developed our technique initially to avoid recurrence and complications, and it seems this works well although to assess the recurrence longer follow ups are needed. As we performed arachnoid preserving duraplasty, the chances of CSF related complications are automatically eliminated. The tacking of the graft with the implant helps in maintenance of the CSF space better than other procedures. With the smaller craniectomy augmented by the measured stealth cranioplasty, the chance of cerebellar slump is checked as well.

Limitations

Due to the COVID-19 pandemic, the study population was smaller than anticipated. Although SC was done by a single surgeon, the other two types of surgeries (PFD and PFDD) were done by multiple surgeons which may have influenced the outcome in those. The follow up period of this study was also short.

Conclusion

Despite the remarkable advents of modern medical science in the past few decades, CM1 continues to be an enigmatic entity. Substantial variations exist in the management of these patients surgically. Rate of post-operative improvement of different symptoms and signs, and development of complications in CM1 patients vary widely depending on surgical techniques. Divergences in techniques of surgery and different lengths of post-operative follow ups make the outcome assessments more complex. It had been and probably will continue to be difficult to stratify patients of CM1 into definite categories based on indications and surgical strategies for CM1. Although SC showed better outcome in terms of CCOS and complications compared to other two most commonly practiced standard procedures PFDD and PFD, a common management protocol for CM1 which would be 'gold standard' or 'one-size-fits-all' with uniform outcome seem rather elusive.

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Author Contributions

- Conception and design: AR, MAR, ASMAO, ANWU
- Acquisition, analysis, and interpretation of data: AR, MAR, MSA, ASMAO, ANWU, MRK
- Manuscript drafting and revising it critically: AR, MAR, MSA, ASMAO, MFR, ANWU, ABMMH, MRK, MH, AHK
- Approval of the final version of manuscript: AR, MAR, MSA, ASMAO, MFR, ANWU, ABMMH, MRK, MH, AHK
- Guarantor accuracy and integrity of the work: AR, MAR, MSA, ASMAO, MFR, ANWU, ABMMH, MRK, MH, AHK

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Conflict of Interest

The authors have no conflict of interest to declare.

Ethical approval

This study was approved by the institutional review board of BSMMU (memo no. 2019/3941 dated 16 April 2019).

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REFERENCES

1. Arnautovic A, Splavski B, Boop FA, Arnautovic KI. Pediatric and adult Chiari malformation Type I surgical series 1965-2013: a review of demographics, operative treatment, and outcomes. J Neurosurg Pediatr. 2015 Feb;15(2):161-77. doi: 10.3171/2014.10.PEDS14295.
2. McClugage SG, Oakes WJ. The Chiari I malformation. J Neurosurg Pediatr. 2019 Sep 1;24(3):217-226. doi: 10.3171/2019.5.PEDS18382.
3. Alden TD, Ojemann JG, Park TS. Surgical treatment of Chiari I malformation: indications and approaches. Neurosurg Focus. 2001 Jul 15;11(1):E2. doi: 10.3171/foc.2001.11.1.3.
4. Barkovich AJ, Wippold FJ, Sherman JL, Citrin CM. Significance of cerebellar tonsillar position on MR. AJNR Am J Neuroradiol. 1986 Sep-Oct;7(5):795-9. PMID: 3096099.
5. Holly LT, Batzdorf U. Chiari malformation and syringomyelia. J Neurosurg Spine. 2019 Nov 1;31(5):619-628. doi: 10.3171/2019.7.SPINE181139.
6. Luciano MG, Batzdorf U, Kula RW, Rocque BG, Maher CO, Heiss J, Martin BA, Bolognese PA, Ashley-Koch A, Limbrick D, Poppe DJ, Esposito KM, Odenkirchen J, Esterlitz JR, Ala'i S, Joseph K, Feldman RS, Riddle R; Chiari I Malformation Common Data Element Working Group. Development of Common Data Elements for Use in Chiari Malformation Type I Clinical Research: An NIH/NINDS Project. Neurosurgery. 2019 Dec 1;85(6):854-860. doi: 10.1093/neuros/nyy475.
7. Ciaramitaro P, Ferraris M, Massaro F, Garbossa D. Clinical diagnosis-part I: what is really caused by Chiari I. Childs Nerv Syst. 2019 Oct;35(10):1673-1679. doi: 10.1007/s00381-019-04206-z.
8. Grangeon L, Puy L, Gilard V, Hebant B, Langlois O, Derrey S, Gerardin E, Maltete D, Guegan-Massardier E, Magne N. Predictive Factors of Headache Resolution After Chiari Type 1 Malformation Surgery. World Neurosurg. 2018 Feb;110:e60-e66. doi: 10.1016/j.wneu.2017.10.070.
9. Sadler B, Kuensting T, Strahle J, Park TS, Smyth M, Limbrick DD, Dobbs MB, Haller G, Gurnett CA. Prevalence and Impact of Underlying Diagnosis and Comorbidities on Chiari 1 Malformation. Pediatr Neurol. 2020 May;106:32-37. doi: 10.1016/j.pediatrneurol.2019.12.005.
10. Tubbs RS, Oakes WJ. The Chiari malformations: a historical context. The Chiari Malformations. 2013:5-11. doi: 10.1007/978-1-4614-6369-6_2.
11. Wilkins RH, Brady IA. The Arnold-Chiari malformations. Arch Neurol. 1971 Oct;25(4):376-9. doi: 10.1001/archneur.1971.00490040102013.
12. Siasios J, Kapsalaki EZ, Fountas KN. Surgical management of patients with Chiari I malformation. Int J Pediatr. 2012;2012:640127. doi: 10.1155/2012/640127.
13. Abd-El-Barr M, Groff MW. Less is more: limiting the size of posterior fossa decompressions in Chiari I malformations. World Neurosurg. 2014 May-Jun;81(5-6):706-7. doi: 10.1016/j.wneu.2013.07.116.
14. Massimi L, Frassanito P, Bianchi F, Tamburrini G, Caldarelli M. Bony decompression vs duraplasty for Chiari I malformation: does the eternal dilemma matter? Childs Nerv Syst. 2019 Oct;35(10):1827-1838. doi: 10.1007/s00381-019-04218-9.
15. Batzdorf U, McArthur DL, Bentson JR. Surgical treatment of Chiari malformation with and without syringomyelia: experience with 177 adult patients. J Neurosurg. 2013 Feb;118(2):232-42. doi: 10.3171/2012.10.JNS12305.
16. Bonney PA, Maurer AJ, Cheema AA, Duong Q, Glenn CA, Safavi-Abbasi S, Stoner JA, Mapstone TB. Clinical significance of changes in pB-C2 distance in patients with Chiari Type I malformations following posterior fossa decompression: a single-institution experience. J Neurosurg Pediatr. 2016 Mar;17(3):336-42. doi: 10.3171/2015.7.PEDS15261.
17. Isu T, Sasaki H, Takamura H, Kobayashi N. Foramen magnum decompression with removal of the outer layer of the dura as treatment for syringomyelia occurring with Chiari I malformation. Neurosurgery. 1993 Nov;33(5):845-9; discussion 849-50. doi: 10.1097/00006123-199311000-00009.
18. Zhang L, Yi Z, Duan H, Li L. A novel autologous duraplasty in situ technique for the treatment of Chiari malformation Type I. J Neurosurg. 2017 Jan;126(1):91-97. doi: 10.3171/2016.1.JNS152161.
19. Radmanesh A, Greenberg JK, Chatterjee A, Smyth MD, Limbrick DD Jr, Sharma A. Tonsillar pulsatility before and after surgical decompression for children with Chiari malformation type 1: an application for true fast imaging with steady state precession. Neuroradiology. 2015 Apr;57(4):387-93. doi: 10.1007/s00234-014-1481-5.
20. Rocque BG, Oakes WJ. Surgical Treatment of Chiari I Malformation. Neurosurg Clin N Am. 2015 Oct;26(4):527-31. doi: 10.1016/j.nec.2015.06.010. Epub 2015 Aug 4. PMID: 26408062.
21. Rahman A. "Stealth Cranioplasty" for Adult Chiari Malformation Type 1: A Philosophical Journey of Innovation, Adaptation, and Evolution. Neurosurgical Procedures-Innovative Approaches: IntechOpen; 2019. DOI: 10.5772/intechopen.89472.
22. Rahman A. Role of Cranioplasty in Management of Chiari Malformation. Neurosurgical Procedures-Innovative Approaches: IntechOpen; 2020. doi: 10.5772/intechopen.90055.
23. Rahman A, Rana MS, Bhandari PB, Asif DS, Uddin ANW, Obaida ASMA, Rahman MA, Alam MS. "Stealth cranioplasty:" A novel endeavor for symptomatic adult Chiari I patients with syringomyelia: Technical note, appraisal, and philosophical considerations. J Craniovertebr Junction Spine. 2017 Jul-Sep;8(3):243-252. doi: 10.4103/jcvjs.JCVJS_76_17.
24. Jussila MP, Nissilä J, Vakkuri M, Olsén P, Niinimäki J, Leinonen V, Serlo W, Salokorpi N, Suo-Palosaari M. Preoperative measurements on MRI in Chiari 1 patients

fail to predict outcome after decompressive surgery. Acta Neurochir (Wien). 2021 Jul;163(7):2005-2014. doi: 10.1007/s00701-021-04842-y.

25. Lei ZW, Wu SQ, Zhang Z, Han Y, Wang JW, Li F, Shu K. Clinical Characteristics, Imaging Findings and Surgical Outcomes of Chiari Malformation Type I in Pediatric and Adult Patients. Curr Med Sci. 2018 Apr;38(2):289-295. doi: 10.1007/s11596-018-1877-2.

26. Perrini P, Anania Y, Cagnazzo F, Benedetto N, Morganti R, Di Carlo DT. Radiological outcome after surgical treatment of syringomyelia-Chiari I complex in adults: a systematic review and meta-analysis. Neurosurg Rev. 2021 Feb;44(1):177-187. doi: 10.1007/s10143-020-01239-w.

27. Rangari K, Das KK, Singh S, Kumar KG, Bhaisora KS, Sardhara J, Mehrotra A, Srivastava AK, Jaiswal AK, Behari S. Type I Chiari Malformation Without Concomitant Bony Instability: Assessment of Different Surgical Procedures and Outcomes in 73 Patients. Neurospine. 2021 Mar;18(1):126-138. doi: 10.14245/ns.2040438.219.

28. Koechlin NO, Abuhusain HJ, Gunawardena M, Auschwitz TS, Teo C. Symptomatic Outcome after Bone-only Suboccipital Decompression in Adult Patients with Chiari Type I Malformations in the Absence of Hydromyelia or Hydrocephalus. J Neurol Surg A Cent Eur Neurosurg. 2017 Jul;78(4):344-349. doi: 10.1055/s-0037-1599841.

29. Nikoobakht M, Shojaei H, Gerszten PC, Shojaei SF, Mollahoseini R, Azar M. Craniometrical imaging and clinical findings of adult Chiari malformation type 1 before and after posterior fossa decompression surgery with duraplasty. Br J Neurosurg. 2019 Oct;33(5):481-485. doi: 10.1080/02688697.2019.1617407.

30. Zhao JL, Li MH, Wang CL, Meng W. A Systematic Review of Chiari I Malformation: Techniques and Outcomes. World Neurosurg. 2016 Apr;88:7-14. doi: 10.1016/j.wneu.2015.11.087.

31. Aliaga L, Hekman KE, Yassari R, Straus D, Luther G, Chen J, Sampat A, Frim D. A novel scoring system for assessing Chiari malformation type I treatment outcomes. Neurosurgery. 2012 Mar;70(3):656-64; discussion 664-5. doi: 10.1227/NEU.0b013e31823200a6.

32. Yarbrough CK, Greenberg JK, Smyth MD, Leonard JR, Park TS, Limbrick DD Jr. External validation of the Chicago Chiari Outcome Scale. J Neurosurg Pediatr. 2014 Jun;13(6):679-84. doi: 10.3171/2014.3.PEDS13503.

33. Feghali J, Marinaro E, Xie Y, Chen Y, Li S, Huang J. Family History in Chiari Malformation Type I: Presentation and Outcome. World Neurosurg. 2020 Oct;142:e350-e356. doi: 10.1016/j.wneu.2020.06.238.

34. Walker-Palmer TK, Cochrane DD, Singhal A, Steinbok P. Outcomes and complications for individual neurosurgeons for the treatment of Chiari I malformation at a children's hospital. Childs Nerv Syst. 2019 Oct;35(10):1895-1904. doi: 10.1007/s00381-019-04201-4.

35. Gilmer HS, Xi M, Young SH. Surgical Decompression for Chiari Malformation Type I: An Age-Based Outcomes Study Based on the Chicago Chiari Outcome Scale. World Neurosurg. 2017 Nov;107:285-290. doi: 10.1016/j.wneu.2017.07.162.

36. Geng LY, Liu X, Zhang YS, He SX, Huang QJ, Liu Y, Hu XH, Zou YJ, Liu HY. Dura-splitting versus a combined technique for Chiari malformation type I complicated with syringomyelia. Br J Neurosurg. 2018 Aug 27:1-5. doi: 10.1080/02688697.2018.1498448.

37. Shetty J, Kandasamy J, Sokol D, Gallo P. Clinical deterioration despite syringomyelia resolution after successful foramen magnum decompression for Chiari malformation - Case series. Eur J Paediatr Neurol. 2019 Mar;23(2):333-337. doi: 10.1016/j.ejpn.2019.01.003.

38. Kumar A, Pruthi N, Devi BI, Gupta AK. Response of Syrinx Associated with Chiari I Malformation to Posterior Fossa Decompression with or without Duraplasty and Correlation with Functional Outcome: A Prospective Study of 22 Patients. J Neurosci Rural Pract. 2018 Oct-Dec;9(4):587-592. doi: 10.4103/jnrp.jnrp_10_18.