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Resolution of Syringomyelia After Posterior Fossa Decompression with and Without Duraplasty in Chiari Malformation-1

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ABSTRACT

Background: Chiari malformations (CMs) encompass a range of clinicopathological conditions that exhibit diverse etiology, pathophysiology, and clinical characteristics. This study was done to compare the resolution of syringomyelia after posterior fossa decompression with and without duraplasty in Chiari malformation-1 (CM-1).

Methods: This Quasi-experimental study was conducted at the Department of Neurosurgery, Mardan Medical Complex, Mardan, from January to November 2023. A non-probability, consecutive sampling technique was adopted. Thirty-two children of either gender, aged below 16 years, presenting with CM-1, and planned to undergo either posterior fossa decompression with duraplasty (PFDD) or posterior fossa decompression without duraplasty (PFD) were analyzed. Duration of procedure (minutes) estimated intra-operative blood loss (ml) and post-surgical complications were noted. Outcomes in terms of resolution or improvement in syringomyelia along with syrinx diameter were compared after 6 months. Qualitative variables were compared using chi-square or Fisher's exact test, and quantitative variables were assessed using independent sample t-tests with $p<0.05$ considered significant.

Results: Out of 32 children analyzed, 18(56.3%) were boys while the mean age was 11.4±4.2 years. There were 18(56.3%) patients who underwent PFDD while the remaining 14(47.7%) had PFD performed. Syringomyelia resolution (88.9% vs. 92.9%, p=0.7024) were relatively similar. Duration of surgery (2.4±0.7 hours vs. 3.6±1.4 hours, p=0.0035) and hospitalization (29.5±8.5 hours vs. 40.5±9.4, p=0.0018) were significantly less among children who underwent PFD when compared to PFDD.
Conclusion: Foramen magnum decompression with and without duraplasty in Chiari malformation-1 showed similar clinical outcomes and both were effective in reducing the syrinx size associated with syringomyelia.

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Keywords: Etiology, Foramen Magnum, Hospitalization, Spinal Cord, Syringomyelia.

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INTRODUCTION

Chiari malformations (CMs) encompass a range of clinicopathological conditions that exhibit diverse etiology, pathophysiology, and clinical characteristics[1]. These involve different levels of hindbrain herniation through the foramen magnum. Professor Hans Chiari (1851–1916) established a classification system consisting of four tiers to categorize these conditions. His classification was primarily derived from over 40 autopsies he conducted during his time as a pathologist in Prague[2].

Chiari malformation Type-1 (CM1) is a congenital condition characterized by a notable protrusion of the cerebellar tonsils through the foramen magnum. The occurrence of CM1 can often be detected through radiographic evidence, specifically magnetic resonance imaging (MRI), with a prevalence of around 4% in children[3]. Although some individuals with CM1 may not experience any symptoms, the narrowing of the posterior fossa frequently results in the compression of the cerebellum and the corticomedullary junction, leading to various manifestations, including headaches and compromises to the brainstem[4]. Posterior fossa decompression for CM-1 has remained the primary therapeutic approach for individuals, both children and adults, who exhibit symptoms related to the malformation and require surgical intervention due to their severity[5].

In children, surgery for CM-1 is a frequently performed neurosurgical procedure, yet there is a lack of agreement among medical professionals regarding the preferred surgical approach. Several posterior fossa decompression techniques yielding positive results have been documented for pediatric CM-1[6]. There is significant debate surrounding the necessity of dural opening for sufficient decompression. Surveys indicate that approximately 75% of pediatric neurosurgeons routinely perform dural opening, while others base their decision on various clinical, radiographic, or intraoperative factors[7].

The absence of comparative local literature on "posterior fossa decompression with duraplasty (PFDD)" versus "posterior fossa decompression without duraplasty (PFD)" for treating CM-1 in children highlights a significant gap in research. Conducting comparative studies exploring the efficacy, outcomes, and potential advantages or disadvantages of these surgical approaches locally could provide valuable insights into optimizing treatment strategies for CM-1 in pediatric populations within the specific context of our region or locality. So, the present study was planned and aimed to compare the resolution of syringomyelia between PFDD and PFD in the treatment of CM-1 in children.

METHODS

This comparative study was performed at the Department of Neurosurgery, Mardan Medical Complex, Mardan, Pakistan, from January 2023 to November 2023. We analyzed children of either gender, aged below 16 years and presenting with CM-1 during the study period, and planned to undergo either PFDD or PFD. Children having Chiari malformations other than type 1 were excluded.
Children who had undergone previous surgical interventions for Chiari malformations or those who had coexisting neurological conditions or disorders that could confound the outcomes or complicate the interpretation of the study results were also not included. Cases missing any of the planned follow-ups were excluded. Non-probability, consecutive sampling technique was adopted. Approval from the "Ethical Research Committee" of Mardan Medical Complex was acquired (481-38 MTI/MMC). Informed and written consents were acquired from parents/guardians of all study participants. A total of 32 children fulfilled inclusion and exclusion criteria during the study period as CM-1 is not a very common entity in our healthcare setting. Allocation to both treatment groups was made non-randomly, based on the treating surgeon's discretion.

At the time of enrollment, demographic information like age and gender were noted. Presenting symptoms and pre-surgery magnetic resonance imaging (MRI) findings were documented (Figure 1). The diameter of the syrinx and cord was determined using sagittal and axial spine images. The axial image at the level of the syrinx's maximum diameter was used for quantitative assessment of the syrinx diameter. Follow-up measurements were obtained at the corresponding vertebral level. Changes in syringomyelia size on postoperative MRI scans were evaluated.

The surgical treatment criteria consisted of confirmation of CM1 with syrinx through MRI imaging and the presence of symptoms related to the condition. A standard suboccipital craniectomy measuring 3

cm × 3 cm was performed on all patients, either with or without duraplasty. A consistent bony decompression was carried out for all patients. The decision to perform a specific surgical procedure, whether PFD without duraplasty or PFDD, was based on the individual surgeon's preference. Duration of procedure (minutes) and estimated blood loss (ml) were noted. Post-surgical complications during the hospital stay were noted and managed as per standard protocols. Length of hospitalization (days) was noted. Pre-surgery, syrinx diameter (mm), and cord diameter (mm) were measured. Patients were asked to follow up after 2 weeks, 8 weeks, and 12 weeks. Outcomes in terms of resolution or reduction in syringomyelia along with syrinx diameter were compared after 6 months (24 weeks) of follow-up between both management approaches. A proforma was formatted to record all study data.

Data analysis was performed using "IBM-SPSS Statistics" version, 26.0. Percentages were shown for qualitative variables. Mean and standard deviation were computed for numeric variables. Qualitative variables were compared using chi-square or Fisher's exact test, and quantitative variables were assessed using independent sample t-tests. A significance level of $p < 0.05$ was considered.

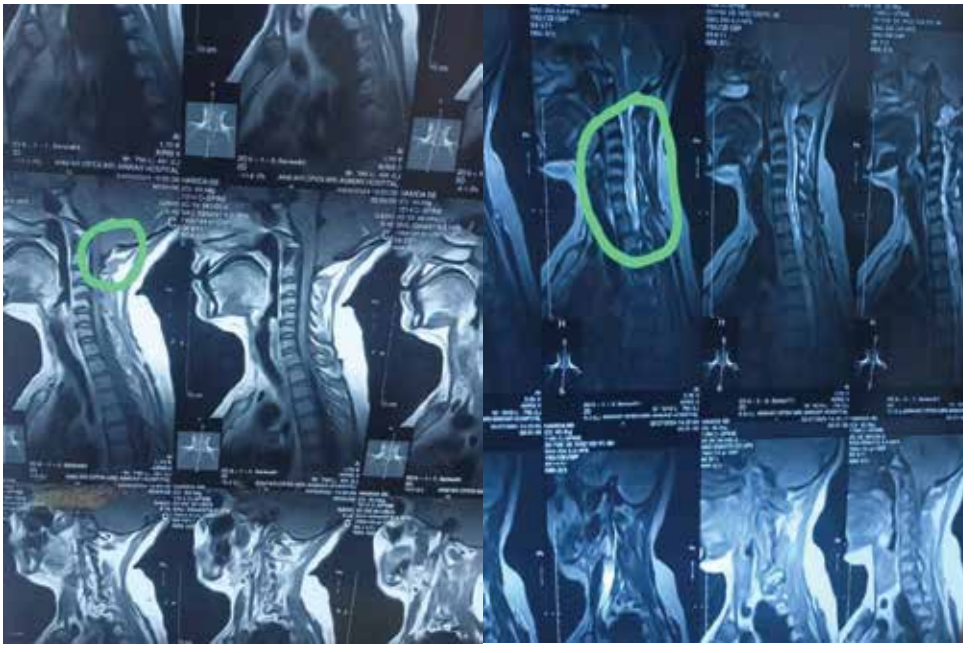

Figure 1: Pre-operative MRI images showing tonsillar herniation with syringomyelia

RESULT

Out of these 32 children, there were 18 (56.3%) boys with a mean age was 11.4±4.2 years. The residential status of 21 (65.6%) children was rural. The most common presenting symptoms were headache, neck pain, and sensory changes noted among 28 (87.5%), 10 (31.2%), and 9 (28.1%) children respectively. Pre-surgery, the mean tonsillar descent was calculated to be 11.2±6.4 mm. There were 18 (56.3%) patients who underwent PFDD while the remaining 14 (47.7%) had PFD performed. Comparisons of demographical, clinical, and pre-surgery characteristics between both management approaches are shown in Table 1.

Table 1: Comparisons of demographical, clinical, and pre-surgery characteristics between study groups

Characteristics		Total (n=32)	PFDD (n=18)	PFD (n=14)	p-value
Gender	Boys	18 (56.3%)	12 (66.7%)	6 (42.9%)	0.2831
	Girls	14 (47.7%)	6 (33.3%)	8 (57.1%)	
Age (Mean±S.D)		11.4±4.2	11.1±4.2	11.8±4.7	0.6602
Residence	Urban	11 (34.4%)	6 (33.3%)	5 (35.7%)	0.8881
	Rural	21 (65.6%)	12 (66.7%)	9 (64.3%)	

Presenting symptoms	Headache	28 (87.5%)	16 (88.9%)	12 (85.7%)	0.7876
	Neck pain	10 (31.3%)	6 (33.3%)	4 (28.6%)	0.7731
	Sensory changes	9 (28.1%)	5 (27.8%)	4 (28.6%)	0.9605
	Ataxia	7 (21.9%)	2 (11.1%)	5 (35.7%)	0.0949
	Bulbar dysfunction	5 (15.6%)	2 (11.1%)	3 (21.4%)	0.4252
	Weakness	5 (15.6%)	3 (16.7%)	2 (14.3%)	0.8540
	Visual complaints	3 (9.4%)	1 (5.6%)	2 (14.3%)	0.4006
Pre-surgery tonsillar descent, mm (Mean±S.D)		11.2±6.4	11.0±6.6	11.5±6.0	0.8265
Pre-surgery Syrinx diameter, mm (Mean±S.D)		6.8±2.3	6.5±2.1	7.0±2.6	0.5515
Pre-surgery cord diameter, mm (Mean±S.D)		11.6±1.5	11.2±1.3	11.9±1.6	0.1820

The mean estimated blood loss during surgery was statistically similar between PFDD and PFD patients (p=0.1373). The mean duration of surgery was significantly less among patients undergoing PFD in comparison to PFDD (p=0.0035). The mean duration of hospitalization was significantly less among patients who underwent PFD vs. PFDD (p=0.0018). The cerebrospinal fluid leak was the most common post-surgery complication reported in 3 (16.6%) patients who underwent PFDD while none of the patients who had undergone PFD experienced cerebrospinal fluid leak (p=0.1086). Revision duraplasty was needed among 1 (5.6%) patient who had undergone PFDD versus 2 (14.3%) with PFD, (p=0.4006). Details about the intra-operative characteristics, duration of hospitalization, and post-surgery complications are shown in Table 2.

Table 2: Comparison of Intra-operative characteristics, duration of hospitalization, and post-surgery complications in both study groups (n=32)

Parameters	PFDD (n=18)	PFD (n=14)	p-value
Estimated intra-operative blood loss in ml (Mean±S.D)	84.52±56.18	56.84±42.95	0.1373
Duration of surgery in hours (Mean±S.D)	3.6±1.4	2.4±0.7	0.0035
Duration of hospitalization in days, (Mean±S.D)	40.5±9.4	29.5±8.5	0.0018
Post-surgery cerebrospinal fluid leak	3 (16.6%)	1 (7.1%)	0.1086
Revision duraplasty needed	1 (5.6%)	2 (14.3%)	0.4006

Assessment at the last follow-up after 6 months post-surgery revealed that syringomyelia resolution or reduction occurred among 16 (88.9%) patients who had undergone PFDD versus 13 (92.9%) with PFD (p=0.7024). Syrinx diameters reduced among patients of both approaches showed that there were no statistically significant differences found (p=0.5181), as shown in Table 2.

Table 2: Comparison of Post-Surgery Outcomes after 6 months of follow-up (n=32)

Outcomes	PFDD (n=18)	PFD (n=14)	p-value
Syrinx resolved or reduced	16 (88.9%)	13 (92.9%)	0.7024
Syrinx diameter, mm, (Mean±S.D)	4.8±1.8	4.4±1.6	0.5181

DISCUSSION

Syrinx is a common observation among CM1 cases, and its presence can significantly impact the clinical presentation and prognosis. Individuals with syrinx often exhibit more pronounced neurological symptoms and signs of myelopathy[8,9]. It is widely recognized that CM1 is the primary underlying cause of syringomyelia, and the progression of syringomyelia is a major contributor to the neurological symptoms experienced by these patients[10]. Consequently, one of the primary objectives of surgical treatment for individuals with CM1 is to achieve improvement or resolution of syringomyelia.

The present study revealed that syringomyelia resolution (88.9% vs. 92.9%, p=0.7024) was relatively similar among children who underwent posterior fossa decompression with or without duraplasty for the resolution of syringomyelia in CM-1 children. But, duration of surgery (2.4±0.7 hours vs. 3.6±1.4 hours, p=0.0035) and hospitalization (29.5±8.5 hours vs. 40.5±9.4, p=0.0018) were significantly less among children who underwent PFD when compared to PFDD. No local data exists regarding the effectiveness of PFDD versus PFD for the resolution of syringomyelia in CM1 children. Our findings are consistent with what has been described previously where Lee A et al from the USA reported that patients undergoing PFDO had a shorter duration of surgery (1.5 vs. 2.8 hours, p<0.001) and duration of hospitalization (2.1 vs. 3.3 days, p<0.001)11. Post-surgery syrinx imaging found that resolution rates were relatively similar (p=0.26). The authors concluded that due to higher morbidity rates and the economic burden of PFDD, PFD can be adopted as a valuable 1st-line approach among the majority of CM1 cases[11].

Over the past few years, several series of investigations have been published, examining the outcomes of extradural decompression for CM1 compared to the more invasive decompression with duraplasty. The primary focus of these studies has been on operative morbidity, rates of postoperative complications, and the need for repeat surgery due to persistent disease[12-14]. Many of the researchers have consistently found that posterior fossa decompression is linked with reduced operative morbidity, shorter hospital stays, and lower rates of complications related to cerebrospinal fluid (CSF)[15,16]. However, there have been conflicting findings regarding the symptomatic outcome, with some researchers, including a meta-analysis, suggesting higher rates of reoperation for persistent disease associated with PFD[16,17]. It is worth noting that the evidence regarding the difference in symptomatic outcome is mixed, and the meta-analysis did not find statistically significant results[18-20]. Moreover, certain findings indicate higher rates of syrinx improvement in patients undergoing PFDD, which may indicate the potential benefit of avoiding extradural decompression in patients with syringomyelia[21-23].

Prior studies encountered notable limitations, notably the absence of well-matched treatment groups concerning demographic factors, symptoms, and radiographic findings[24]. Additionally, their reporting methods for clinical outcomes lacked clear definitions and did not rely on validated measures[25]. In response to these shortcomings, the present study aimed to rectify these gaps by offering a balanced comparison of treatments, emphasizing clinical outcomes and improvements in syrinx conditions. Being a single-center study conducted on a relatively small sample size is one of the limitations of this study. Non-randomized approach's absence of long-term follow-up data warrants further research.

CONCLUSION

Resolution of syringomyelia after foramen magnum decompression with and without duraplasty in Chiari malformation-1 showed similar clinical outcomes. Duration of surgery and hospitalization were significantly less among children who underwent foramen magnum decompression without duraplasty for the resolution of syringomyelia in Chiari malformation-1. Both surgical approaches were effective in reducing the syrinx size associated with syringomyelia. The addition of duraplasty did not significantly enhance the overall clinical benefits of the decompression surgery.

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ETHICAL APPROVAL

Approval from the Institutional Ethical Research Committee was acquired (481-38 MTI/MMC).

PATIENT CONSENT

Informed and written consents were acquired from parents/guardians of all study participants.

AUTHORS CONTRIBUTION

All authors contributed equally.

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