

ORIGINAL ARTICLE

Outcome of Posterior Fossa Decompression and Duroplasty in Adult Chiari Malformation

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ABSTRACT

Objective: To evaluate outcome of posterior fossa decompression and duroplasty in patients with Chiari Malformation 1 (CM1).

Methods: This retrospective study was conducted at Neurospinal and Cancer Care Institute, Karachi, from March 2010 to March 2019. After diagnosis with the help of Magnetic Resonance Imaging (MRI) cervical spine, all patients with CM I, having headache, neck pain, limbs numbness and syringomyelia were included. Information was collected regarding demographic characteristics, presenting complaints, and post-operative outcome. Chicago Chiari outcome scale was used to assess the outcome.

Results: Of 91 patients, the mean age of the patient was 31 ± 6.7 years. There were 58 (63.7%) males and 33 (36.3%) females. C2 laminectomy was performed in 10 (11.0%) patients while C1 assimilation in 3 (3.3%) patients. Hydrocephalus was found in 12 (13.1%) patients. The outcome of posterior fossa decompression (PFD) showed that functional outcome was found higher, i.e., 46 (50.5%), followed by excellent outcome 25 (27.5%), impaired outcome 15 (16.5%), and incapacitated in 5 (5.5%). The comparison of outcome with baseline characteristics showed that most of the patients with age >30 years had excellent and functional outcome, i.e., 16 (28.1%) and 26 (45.6%) respectively. Five (8.6%) females had incapacitated outcome. Moreover, headache, syringomyelia, and swallowing difficulty were observed somewhat equal in each outcome.

Conclusions: PFD surgery with duraplasty can improve the clinical symptoms. Decompressive surgery in patients with duraplasty can significantly improve the patient condition while late presentation may affect the clinical outcome.

Keywords: Chiari Malformation Decompression, Chicago Chiari Outcome Scale, Duraplasty, Posterior Fossa

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INTRODUCTION

Chiari Malformation 1 (CM1) is a hindbrain structural abnormality that can cause cerebellar tonsils to herniate through the foramen magnum (FM) may show symptoms in adolescence or early adulthood. However, the pathophysiology may relate to intracranial pressure (ICP), this famous neurosurgical pathology is named after Professor Hans Chiari, and he described four (1 to 4) types, CM type 1 has an incidence of 1:1000–5000.^{1,2} Early diagnosis is of utmost importance, so magnetic resonance imaging cervical spine is one of the prime diagnostic tools which helps in Documentation of cerebellar tonsils below the FM > 5 mm to an average of 12 mm. Available surgical options are cranial/cervical decompression, with/without fusion, and duraplasty or

other viable options depending on patient to patient.³ Surgical decompression of sub-occipital bone plays an optimal role and helps in the relief of the symptoms of the extent of tonsillar herniation prolong symptoms can impact the final outcome.⁴

Restoration path of the circulation of cerebrospinal fluid (CSF) in the craniovertebral junction is an optimal target.^{5,6} It may sometimes be associated with tumor-causing posterior fossa herniation.⁷ It is reported that the problem clinical manifestations of this condition is located at the level of the FM and contents of posterior fossa volume and this may cause an imbalance in CSF flow at the craniovertebral junction this may impact traditional approach of treatment.⁸ Moreover, even though what recent theories suggest a well-liked method for treatment for Chiari malformation,

without craniocervical destabilization is subsequent decompression of the fossa with or without duraplasty.⁹ So much work is done, but still, it is controversial that amount of bony removal, in the use of duraplasty procedure, the need for subarachnoid dissection and role of tonsillar shrinkage.¹⁰

The rationale of this study is to share the experience of cases operated at our hospital showing the results of surgery of Chiari-I malformation cases with duraplasty and C1 laminectomy. Suboccipital craniectomy is a common surgical technique for duraplasty diagnoses, such as C1 laminectomy.

METHODS

This retrospective study was conducted at Neuro Spinal & Cancer Care Institute, Karachi, from March 2010 to March 2019. The study was started after taking the ethical approval from the ethical review committee of the institute (Ref#1459/2013).

Inclusion criteria were all adult patients of either gender diagnosed with the help of magnetic resonance imaging (MRI) cervical spine showing >5mm tonsillar herniation, associated with headache, numbness of the limbs, neck pain, deficit related to CM1 or syringomyelia. Patients operated previously for the same disease, cervical trauma, and operated cases of brain tumor were excluded.

All operations were performed by an experienced neurosurgeon, whereas posterior fossa decompression (PFD) with duraplasty and C1 laminectomy was completed under general anaesthesia. The patient lay in a prone position with Mayfield in positional fixation. The skin, subcutaneous tissues, and occipital and paraspinal muscles were cut with a midline incision extending from occipital protuberance to spinous C3 process. The lower portion of suboccipital craniectomy was made about 3 cm x 3 cm. Spinous C1 process and laminectomy were completed when the dura was opened in Y shape. Incision was made to open arachnoid matter and CSF was drained from cisterna magna. The cervical spinal cord was observed, CSF was drained from cisterna magna, adhesions were removed and duraplasty was done with the fascia, wound was closed layer by layer.

The researchers evaluated the results of type I decompression and duraplasty of Chiari malformation. Complete neurological history and examination was done. MRI cervical spine was used as a diagnostic tool. Decompression, C1 laminectomy and Duraplasty was done after (PFD) done in all the cases. A one-year medical record of all patients were retrieved to see the

Variables like age, gender, presenting complaints, syrinx and post-operative outcome were noted. Chicago Chiari outcome scale was used to assess the outcome.^{11,12}

Data were entered and analyzed by using version 26.0 of the statistical package of social sciences (SPSS) software. Mean \pm SD were computed for quantitative variables like, age while frequency and percentages were computed for categorical variables like, gender, C1 assimilation, C2 laminectomy and hydrocephalus. Cross-tabulation was made between outcome variables and baseline characteristics like age, gender, clinical features, and presenting complaints. The p-value of ≤ 0.05 considered as significant.

RESULTS

Of 91 patients, the mean age of the patient was 31 ± 6.7 years. There were 58 (63.7%) males and 33 (36.3%) females. C2 laminectomy was performed in 10 (11.0%) patients while C1 assimilation in 3 (3.3%) patients. Hydrocephalus was found in 12 (13.1%) patients.

The outcome of PFD showed that functional outcome was found higher, i.e., 46 (50.5%), followed by excellent outcome 25 (27.5%), impaired outcome 15 (16.5%), and incapacitated in 5 (5.5%). The comparison of outcome with baseline characteristics showed that most of the patients with age >30 years had excellent and functional outcome, i.e., 16 (28.1%) and 26 (45.6%) respectively. Five (8.6%) females had incapacitated outcome. Moreover, headache, Syringomyelia, and Swallowing Difficulty were observed somewhat equal in each outcome. (Table 1)

Valsalva headache was present at baseline in 91 (100%) patients while the improvement in headache was observed in 87 (95.6%) patients. Neck pain was present at baseline in 82 (90.1%) patients. Of these 82 patients with neck pain, post-procedure findings showed improvement in 74 (90.2%) patients. Syringomyelia was observed in 36 (39.5%) patients at baseline which was improved in 30 (32.9%) patients. Ataxia/disarticulation was present at baseline in 48 (52.7%) patients which were improved in 40 (83.3%) patients, limb numbness present in 78 (85.7%) patients at baseline which were improved in 56 (71.7%) patients, swallowing difficulty was present at baseline in 30 (32.9%) patients which were improved in 25 (83.3%) patients. (Figure 1)

CSF leak and superficial infection were the only complications observed, i.e., 7 (7.7%) and 3 (3.3%) patients respectively. The summary of the patients reported complications is reported in detail in table 2.

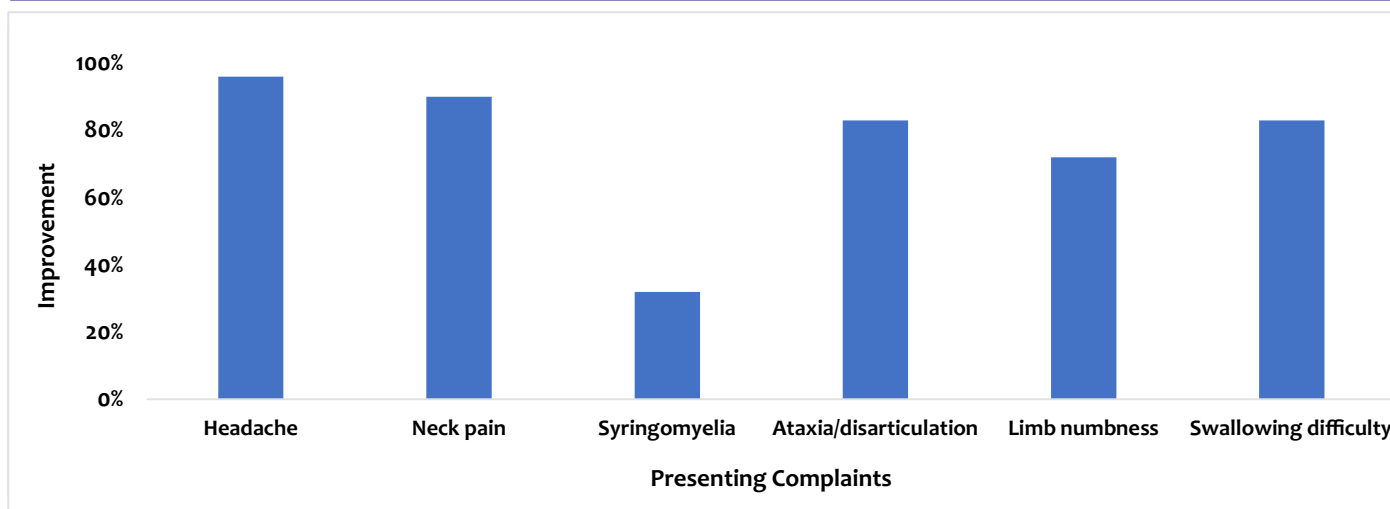


Figure 1: Improvement in complaints of posterior fossa decompression

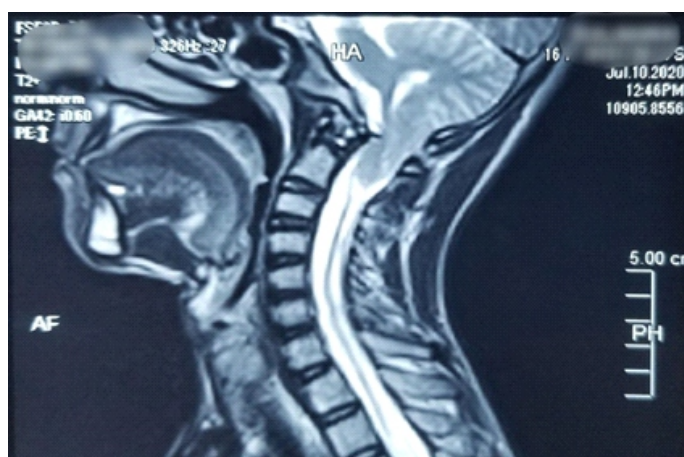


Figure 2: Showing herniation of brain in spinal cord plus associated syrinx

DISCUSSION

The findings of the current study have shown that the success rate with the duroplasty was around ninety five percent, with follow up to one year while in a study by Kennedy BC, et al,¹³ more than ninety per cent of patients improved with good clinical outcome, with a follow-up of thirty-two weeks. Similar to our study they did not had any significant complications, and the average stay was around two days and partial C-2 laminectomy was an independent risk factor that is associated with reoperation compared to our study in which we performed C2 laminectomy based on extension of syrinx.

We did duroplasty in all patients which resulted in improvement of syrinx while the resection of tonsils was not done in any patient as it was not required. In another study by Zhao JL, et al,¹⁴ they favored a study for duroplasty to improve results from their study and did not recommend Resection of tonsils, so it depends

on institutional practice as well as surgeon preference. In our study, no patient developed meningitis while CSF leak was reported and was managed surgically and conservatively varying patient to patient. In contrast, in a study by Chen J, et al,¹⁵ the most common complication was aseptic meningitis and associated more with PFD, and duraplasty that are based on positive CSF studies on lumbar puncture and negative culture is treated with steroid, this may be due to difference in number of patient plus all our patient were managed with double antibiotics in intensive care unit for day.

In our study, we did not performed tonsillectomy in any patient, it is not that it was not our policy but did not came across any of such situation while for syrinx we did duroplasty plus c1 laminectomy and in eleven percent patients we did C2 laminectomy compared to Liu B, et al,¹⁶ performed surgery in all cases successfully, and confirmation was done by intraoperative ultrasonography, and follow-up was 20 months. Clinical success was seen in eighty five percent patients who have no significant variations between the two classes. In group A, ninety percent patients were labelled as a "good" outcome compared to eighty seven percent patients in a group. They also performed tonsillectomy. We performed the procedure on the elective list after proper diagnosis and taking pre and post-operative measures. Surgery was done within 24 hours of hospitalization. Three patients with acute symptom regression, five percent reported an improved syrinx duration and breadth and more rostral syrinx expansion relative to non-acute CMI patients.¹⁷ In the study of Tosi U, et al,¹⁸ they showed that in contrast to the MRI post-surgery did not had significant changes based on syrinx but patient clinical condition improved.

According to findings, it was observed that patients

Table 1: Comparison of outcome with baseline characteristics (n=91)

Variables	Total	Outcome			
		Excellent (n=25)	Functional (n=46)	Impaired (n=15)	Incapacitated (n=5)
Age, years					
≤30	34	9 (26.5)	20 (58.8)	5 (14.7)	0 (0)
>30	57	16 (28.1)	26 (45.6)	10 (17.5)	5 (8.8)
Gender					
Male	33	5 (15.2)	21 (63.6)	7 (21.2)	0 (0)
Female	58	20 (34.5)	25 (43.1)	8 (13.8)	5 (8.6)
Clinical Features					
C2 laminectomy	10	2 (20.0)	7 (70.0)	0 (0)	1 (10.0)
C1 Assimilation	3	0 (0)	3 (100)	0 (0)	0 (0)
Hydrocephalus	12	2 (16.7)	4 (33.3)	4 (33.3)	2 (16.7)
Presenting Complaints					
Headache	91	25 (27.5)	46 (50.5)	15 (16.5)	5 (5.5)
Neck Pain	81	4 (40.0)	5 (50.0)	1 (10.0)	0 (0)
Syringomyelia	35	13 (23.2)	29 (51.8)	11 (19.6)	3 (5.4)
Ataxia/Disarticulation	48	10 (23.3)	24 (55.8)	8 (18.6)	1 (2.3)
Limb Numbness	77	5 (35.7)	7 (50.0)	1 (7.1)	1 (7.1)
Swallowing Difficulty	61	17 (27.9)	27 (44.3)	12 (19.7)	5 (8.2)

All data presented as number (%)

Table 2: Summary of the patients reported complications

S. No	Age, years	Gender	C2 Laminectomy	Hydrocephalus	Headache	Neck Pain	Syringomyelia	Ataxia/Disarticulation	Limb Numbness	Swallowing Difficulty
CSF Leak (n=7)										
1	26	M			✓	✓		✓	✓	
2	28	F	✓		✓	✓		✓	✓	✓
3	19	F	✓	✓	✓	✓	✓		✓	✓
4	21	M			✓	✓	✓	✓	✓	
5	32	M			✓	✓		✓	✓	✓
6	32	M			✓	✓				✓
7	32	M			✓	✓	✓	✓	✓	
Superficial Infection (n=3)										
1	19	M			✓	✓			✓	✓
2	40	M			✓	✓	✓	✓	✓	
3	38	M			✓		✓		✓	✓

without syrinx had a better outcome than with syrinx that may be due to the compression to cord for a significant period of time plus syrinx itself taking some time to resolve. Kumar A, et al.¹⁹ Out of 19 patients in whom syrinx diameter decreased on postoperative MR imaging, Surgical decompression with or without duraplasty does not have a statistically significant

relationship in the degree of syrinx resolution. Surgical decompression without duraplasty provides the benefits of surgical decompression while avoiding the complications of intradural techniques.

As per study findings, headache was present in all patients while the improvement in headache was observed in ninety six percent patients. Neck pain was

present in ninety percent patients. Syringomyelia was observed in forty percent patients and improved in thirty three percent patients. Ataxia/disarticulation was present in fifty three percent patients which were improved in eighty three percent patients, limb numbness present in eighty six percent patients at baseline which were improved in seventy two percent patients, swallowing difficulty was present at baseline in thirty three percent patients which were improved in eighty three percent patients, While in the study of Yahanda AT, *et al*,²⁰ in their study showed that even after use of different grafts for dural closure autograft has minimal complication as compared to other materials similarly in our study we only used the autografts rather than any other material as it is low cost and has minimal complication rate.

In the current study, we did not had tonsillectomy of any patient as we did not find any need to do so while according to Edilen CO, *et al*.²¹ Cerebellar tonsillectomy with suboccipital decompression and duroplasty have better impact in providing the cure in CM I cases. And early diagnosis and treatment essential to for effectiveness of treatment.

In the current study, we had 91 patients, thirteen percent patients had hydrocephalus, and a Ventriculoperitoneal (VP) shunt was placed. Valsalva headache was in all patients with duration resolved in ninety six percent patients, neck pain was in ninety-one percent patient resolved in 74 ninety percent cases, and syrinx was in forty percent cases improved in 30 eighty three percent cases. While of the 25 patients examined, sixty eight percent of Botelho RV, *et al*,²² evaluated the diagnosed with sleep apnea. The mean number of respiratory events decreased after surgery from 180.70 to 69.29; the mean number of obstructive events decreased from 107.37 to 60.58, and the mean number of central events decreased from 38.45 to 8. We did not observe sleep apnea while difficulty in swallowing improved in seventy seven percent out of thirty four percent. Similar to our study there are many studies that support the concept of duroplasty with different modification to earn better results for the patient.^{23,24}

The limitation of the study was that we did not had a comparative study between with and without duroplasty which can be significant plus we had limited follow up and we had single centre study. Furthermore, we had enough patients in our study that we can conclude that most symptoms can be improved with duroplasty and in a symptomatic patient, surgery should be planned soon enough to minimize the deficit duration.

CONCLUSION

Posterior fossa decompression with duroplasty can improve patient symptoms clinically plus it improves the symptomatic condition of the patient while the delayed surgery may affect the clinical outcome.

ETHICAL APPROVAL: The study was approved by Ethical Review Committee of the Neuro Spinal & Cancer Care Institute, Karachi (Ref #1459/2013).

AUTHORS' CONTRIBUTION: AK: Study design and methodology. Manuscript writing. ABS: Referencing and data calculations. Analysis of data and interpretation of results. ASMH: Literature review and manuscript writing and data analysis.

CONFLICT OF INTEREST: Authors report no conflict of interest.

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REFERENCES

1. Piper RJ, Magdum SA. Chiari 1 malformation and raised intracranial pressure. *Childs Nerv Syst* 2019; 35:1719-25. doi: [10.1007/s00381-019-04232-x](https://doi.org/10.1007/s00381-019-04232-x)
2. Zaffanello M, Sala F, Sacchetto L, Gasperi E, Piacentini G. Evaluation of the central sleep apnea in asymptomatic children with Chiari 1 malformation: an open question. *Childs Nerv Syst* 2017; 33:829-32. doi: [10.1007/s00381-017-3399-9](https://doi.org/10.1007/s00381-017-3399-9)
3. Epstein NE. Definitions and treatments for chiari-1 malformations and its variants: Focused review. *Surg Neurol Int* 2018; 9:152. doi: [10.4103/sni.sni_208_18](https://doi.org/10.4103/sni.sni_208_18)
4. Heffez DS, Golchini R, Ghorai J, Cohen B. Operative findings and surgical outcomes in patients undergoing Chiari 1 malformation decompression: relationship to the extent of tonsillar ectopia. *Acta Neurochir (Wien)* 2020; 162:1539-47. doi: [10.1007/s00701-019-04172-0](https://doi.org/10.1007/s00701-019-04172-0)
5. Zuev AA, Pedyash NV, Epifanov DS, Kostenko GV. Results of surgical treatment of syringomyelia associated with Chiari 1 malformation. An analysis of 125 cases. *Zh Vopr Neurokhir Im N N Burdenko* 2016; 80:27-34. English, Russian. doi: [10.17116/neiro201680127-34](https://doi.org/10.17116/neiro201680127-34)
6. Mozaffari K, Davidson L, Chalif E, Phan TN, Sparks AD, Myseros JS, et al. Long-term outcomes of posterior fossa decompression for Chiari malformation type 1: which patients are most prone to failure? *Child's Nervous System* 2021; 1-8. doi: [10.1007/s00381-021-05280-y](https://doi.org/10.1007/s00381-021-05280-y)
7. Zuev AA, Kostenko GV. Lechenie siringomieli u patsientov s anomalieĭ Kiari [Treatment of syringomyelia associated with Chiari 1 malformation]. *Zh Nevrol Psikhiatr Im S S Korsakova* 2017; 117:102-6. Russian. doi: [10.17116/jnevro201711731102-106](https://doi.org/10.17116/jnevro201711731102-106)

8. Navarro R, Olavarria G, Seshadri R, Gonzales-Portillo G, McLone DG, Tomita T. Surgical results of posterior fossa decompression for patients with Chiari I malformation. *Childs Nerv Syst* 2004; 20:349-56. [doi:10.1007/s00381-003-0883-1](https://doi.org/10.1007/s00381-003-0883-1)
9. Sabba MF, Renor BS, Ghizoni E, Tedeschi H, Joaquim AF. Posterior fossa decompression with duraplasty in Chiari surgery: A technical note. *Rev Assoc Med Bras* 2017; 63:946-9. [doi:10.1590/1806-9282.63.11.946](https://doi.org/10.1590/1806-9282.63.11.946)
10. Abila AA, Link T, Fusco D, Wilson DA, Sonntag VK. Comparison of dural grafts in Chiari decompression surgery: Review of the literature. *J Craniovertebr Junction Spine* 2010; 1:29-37. [doi:10.4103/0974-8237.65479](https://doi.org/10.4103/0974-8237.65479). PMID: 20890412
11. Abdulghani YS, Elzain MA, Ali AA, Mohamed HH, Basheer BM. Chiari type-I malformation: Clinical features and surgical outcome assessment using Chicago Chiari Outcome Score (CCOS) among Sudanese patients. *Pak J Surg* 2019; 35:325-30.
12. Gallo P, Copley PC, McAllister S, Kaliaperumal C. The impact of neurosurgical technique on the short- and long-term outcomes of adult patients with Chiari I malformation. *Clin Neurol Neurosurg* 2021; 200:106380. [doi:10.1016/j.clineuro.2020.106380](https://doi.org/10.1016/j.clineuro.2020.106380)
13. Kennedy BC, Kelly KM, Phan MQ, Bruce SS, McDowell MM, Anderson RC, et al. Outcomes after suboccipital decompression without dural opening in children with Chiari malformation Type I. *J Neurosurg Pediatr* 2015; 16:150-8. [doi:10.3171/2014.12.PEDS14487](https://doi.org/10.3171/2014.12.PEDS14487)
14. Zhao JL, Li MH, Wang CL, Meng W. A Systematic Review of Chiari I Malformation: Techniques and Outcomes. *World Neurosurg* 2016; 88:7-14. [doi:10.1016/j.wneu.2015.11.087](https://doi.org/10.1016/j.wneu.2015.11.087)
15. Chen J, Li Y, Wang T, Gao J, Xu J, Lai R, et al. Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation type I in adult patients: A retrospective analysis of 103 patients. *Medicine (Baltimore)* 2017; 96:e5945. [doi:10.1097/MD.0000000000005945](https://doi.org/10.1097/MD.0000000000005945)
16. Liu B, Wang Y, Liu S, Zhang Y, Lu D, Chen L, et al. Tonsillectomy with modified reconstruction of the cisterna magna with and without craniectomy for the treatment of adult Chiari malformation type I with syringomyelia. *Acta Neurochir (Wien)* 2020; 162:1585-95. [doi:10.1007/s00701-019-04177-9](https://doi.org/10.1007/s00701-019-04177-9)
17. Almotairi FS, Tisell M. Acute deterioration of adults with Chiari I malformation associated with extensive syrinx. *Br J Neurosurg* 2020; 34:13-7. [doi:10.1080/02688697.2019.1684438](https://doi.org/10.1080/02688697.2019.1684438)
18. Tosi U, Lara-Reyna J, Chae J, Sepanj R, Souweidane MM, Greenfield JP. Persistent Syringomyelia After Posterior Fossa Decompression for Chiari Malformation. *World Neurosurg* 2020; 136:454-61. doi.org/10.1016/j.wneu.2020.01.148
19. 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; 9:587-92. [doi:10.4103/jnrrp.jnrrp_10_18](https://doi.org/10.4103/jnrrp.jnrrp_10_18)
20. Yahanda AT, Simon LE, Limbrick DD. Outcomes for various dural graft materials after posterior fossa decompression with duraplasty for Chiari malformation type I: a systematic review and meta-analysis. *J Neurosurg* 2021; 1(aop):1-4. doi.org/10.3171/2020.9.JNS202641
21. Edilen CO, Takibi OU. Cerebellar tonsillectomy with suboccipital decompression and duraplasty by small incision for Chiari I malformation (with syringomyelia): long term follow-up of 76 surgically treated cases. *Turkish Neurosurg* 2012; 22:274-9. [doi:10.5137/1019-5149.JTN.4634-11.1](https://doi.org/10.5137/1019-5149.JTN.4634-11.1)
22. Botelho RV, Bittencourt LR, Rotta JM, Tufik S. The effects of posterior fossa decompressive surgery in adult patients with Chiari malformation and sleep apnea. *J Neurosurg* 2010; 112:800-7. [doi:10.3171/2009.7.JNS09174](https://doi.org/10.3171/2009.7.JNS09174)
23. Osborne-Grinter M, Arora M, Kaliaperumal C, Gallo P. Posterior fossa decompression and duraplasty with and without arachnoid preservation for the treatment of adult Chiari malformation type 1: a systematic review and meta-analysis. *World Neurosurg* 2021; 151:e579-e598. [doi:10.1016/j.wneu.2021.04.082](https://doi.org/10.1016/j.wneu.2021.04.082)
24. Tam SK, Brodbelt A, Bolognese PA, Foroughi M. Posterior fossa decompression with duraplasty in Chiari malformation type 1: a systematic review and meta-analysis. *Acta Neurochir (Wien)* 2021; 163:229-38. [doi:10.1007/s00701-020-04403-9](https://doi.org/10.1007/s00701-020-04403-9)

