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Use of intraoperative ultrasonography in deciding the extent of surgical decompression for Chiari Malformation Type I

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Abstract

Introduction: The optimal surgical treatment for Chiari Malformation Type I continues to be controversial with procedures ranging from posterior fossa bone decompression alone to release or resection of the cerebellar tonsils. Intraoperative ultrasonography can be a useful tool in deciding the least invasive procedure so as to reduce the complications and improve the efficacy of the procedure.

Methods: A retrospective study over a period of seven years of patients who underwent surgery for Chiari Malformation type I under the guidance of intraoperative ultrasonography was done. The patients underwent three types of surgery based on the adequacy of decompression seen as free flow of CSF through foramen of magendie and free movement of cerebellar tonsils on intraoperative ultrasonography placed over the duramater: Group 1: foramen magnum decompression alone, Group 2: foramen magnum decompression and lax duraplasty and Group 3: foramen magnum decompression with lax duraplasty and release or resection of cerebellar tonsils. Outcome of the types of surgery were determined by the clinical outcome at post-operative six months and two years.

Result: A total of 10 patients: four males and six females were included in the study. The mean age of the population was 32.7 +/- 14.57 years with range of 42 years. There were three patients in group 1, two patients in group 2 and five patients in group 3. 100% of the patients in group 1, 50% of patients in group 2 and 60% of the patients in group 3 had improved outcome as evidenced by resolution of symptoms and resolution of syrinx at six months and one year postoperative period.

Conclusion: Intraoperative ultrasonography is an effective tool in deciding the extent of surgical decompression for Chiari Malformation Type I. A larger multicentre study would help statistically validate the findings.

Key words: Ultrasonography, Chiari Malformation Type I, foramen magnum

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Introduction

Chiari malformation Type I (CMI) is a serious neurological disorder characterized by herniation of the cerebellar tonsils through the foramen magnum at the cranio-vertebral junction (CVJ) as first described by Hans Chiari more than 100 years ago.[1,2] In some patients, the herniation can lead to the formation of a syrinx or fluid-filled cyst in the spinal cord tissue, causing paraesthesia, muscle weakness, and paralysis. Symptom onset can occur at any age, but in adults symptoms often emerge in the late 20′s or early 30′s.[3] The symptoms arise due to disruption of the natural flow of cerebrospinal fluid (CSF) across the CVJ and the herniation exerting a mass effect on nearby neural tissue.

The pathophysiology of CMI is not well elucidated, however, the prevailing theory focuses on the underdevelopment of the posterior cranial fossa.[3] A study using many of the same subjects found reduced CSF spaces, or crowding, around the cerebellar tonsils.[4] Despite this supporting evidence, the small posterior cranial fossa theory does not address the issue of asymptomatic cases of tonsillar herniation being far more prevalent than symptomatic ones

The optimal surgical treatment for Chiari Malformation Type I continues to be controversial with procedures ranging from posterior fossa bone decompression alone to release or resection of the cerebellar tonsils.[5] Various procedures have been advocated such as removal of cervical lamina, duraplasty, cerebellar tonsil ablation or syringo-subarachnoid shunts. Complications related to cerebellar ptosis and risk of re-adhesion are more with invasive procedures. Other complications include CSF leakage, pseudomeningocele formation, infection, headache, prolonged recovery, neurological injury, and even death.[6,7] Some authors argue that only bone decompression is enough while others support maximal decompression by duraplasty and tonsillar shrinkage to reach the ultimate goal of restoring normal CSF dynamics at the cranio-vertebral junction (CVJ).

Intraoperative ultrasonography (USG) can be a useful tool in deciding the extent of surgery needed to achieve adequate decompression by documenting adequacy of decompression of internal structures like CSF and cerebellar tonsils.[8] Since the goal of surgery in Chiari Malformation Type I is adequate decompression which can be confirmed by flow of CSF at the CVJ, ultrasonography in the intraoperative period can help to confirm adequate flow of CSF at the foramen of magendie, thereby minimizing the surgical invasiveness.

The objective of our study was to determine the role of intraoperative ultrasonography in deciding the extent of resection in Chiari Malformation Type I and to correlate the extent of resection with the functional outcome of patient.

Methods and Materials

We conducted a retrospective case series in the Department of Neurosurgery at Kathmandu Medical College Teaching Hospital. All patients who underwent intraoperative ultrasonography (USG) guided surgery of Chiari Malformation Type I were included in the study. Patients who did not undergo intraoperative USG guided surgery of Chiari Malformation Type and who did not give consent for the study were excluded. All patients with diagnosis of Chiari Malformation Type I based on the clinical and imaging (MRI) findings were admitted. The patients were planned for surgery as per hospital protocol. The surgery was planned in prone position. Posterior midline linear incision was given from inion to level of C2 spinous process. Foramen magnum decompression was done by 2X2 cm posterior fossa craniectomy. Intraoperative ultrasonography was done soon after bone removal to see for the location of cerebellar tonsils. C1 posterior arch removal was done depending upon the location of cerebellar tonsils. Intraoperative ultrasonography was also done to see for the tonsillar movement and free flow of CSF through Foramen of Magendie into the central canal and subdural space. The level of cerebellar tonsils, CSF flow and free excursion of cerebellar tonsils with respiration was looked on USG. If there was adequate tonsillar movement and CSF flow then it meant adequate decompression was done and the surgery was completed. However, if no adequate tonsillar movement or CSF flow seen on ultrasound after bony decompression, then the surgery was proceeded in the form of lax duraplasty or cerebellar tonsillar release/resection and lax duraplasty. The wound was then closed in layers as per treatment protocol. Sutures were removed on the 14th POD. The patients were then observed for resolution of symptoms at the time of discharge, one month, six months and one year period. MRI cervical spine was done at one year postoperative period and at six months if there was no improvement in symptoms. The patients were also observed for need for repeated surgery and for complications like wound gaping, CSF leak, post-operative meningeal infection. The data was analyzed with the help of Microsoft Excel and SPSS version 21. Functional outcome in terms of resolving of symptoms, need for re-operation and complications was calculated. Ethical clearance was taken from the institutional review committee.

Results

A total of 10 patients over a period of seven years (2014-2020) were included in the study according to inclusion criteria. There were four (40%) male and six (60%) female patients. The mean age of the population was 32.7 +/- 14.57 years with range of 42 years. Majority of the patients (seven patients) were in the age group of 21-30 years. Three (30%) patients had presented with symptoms of headache, two (20%) patients had tingling sensation in limbs and five (50%) patients had both symptoms. Three (30%) patients underwent posterior fossa (PF) decompression alone, two (20%) patients underwent posterior fossa (PF) decompression with lax duraplasty and five (50%) patients underwent posterior fossa (PF) decompression with lax duraplasty with release/resection of cerebellar tonsils based on intraoperative USG.

Seven (70%) patients improved symptomatically, three (30%) patients had no change in symptoms as last followed up at one year post surgery and none of the patients degraded. Of the seven patients who improved, three (42.85%) patients had undergone PF decompression alone, one (14.28%) patient had undergone PF decompression with lax duraplasty and three (42.85%) patients had undergone PF decompression with lax duraplasty with release/resection of cerebellar tonsils. All the patients undergoing PF decompression alone improved symptomatically, the decompression which was confirmed intraoperatively with the use of ultrasonography and this shows the effectiveness of ultrasonography in evaluating the decompression. Whereas, the patients who had no change in symptoms, one (33.33%) patient had undergone PF decompression with lax duraplasty and two (66.66%) patients had undergone PF decompression with lax duraplasty with release/resection of tonsils.

Seven (70%) patients had cervical syrinx in preoperative MRI. The syrinx had resolved in all of these seven patients in the post-operative MRI of cervical spine done at one year post surgery period. Of these seven patients, one (14.28%) patient had undergone PF decompression alone, one (14.28%) patient had undergone PF decompression with lax duraplasty and five (71.42%) patients had undergone PF decompression with lax duraplasty with release/resection of cerebellar tonsils. The extent of decompression and manipulation were decided intraoperatively based on the ultrasonographic findings for adequate CSF flow.

There were two (20%) patients who had complications in the post-operative period. Of these, one patient had wound gaping who had undergone PF decompression alone and the other patient had wound CSF leak who had undergone PF decompression with lax duraplasty with release/resection of cerebellar tonsils. Both of them were managed with secondary suturing.

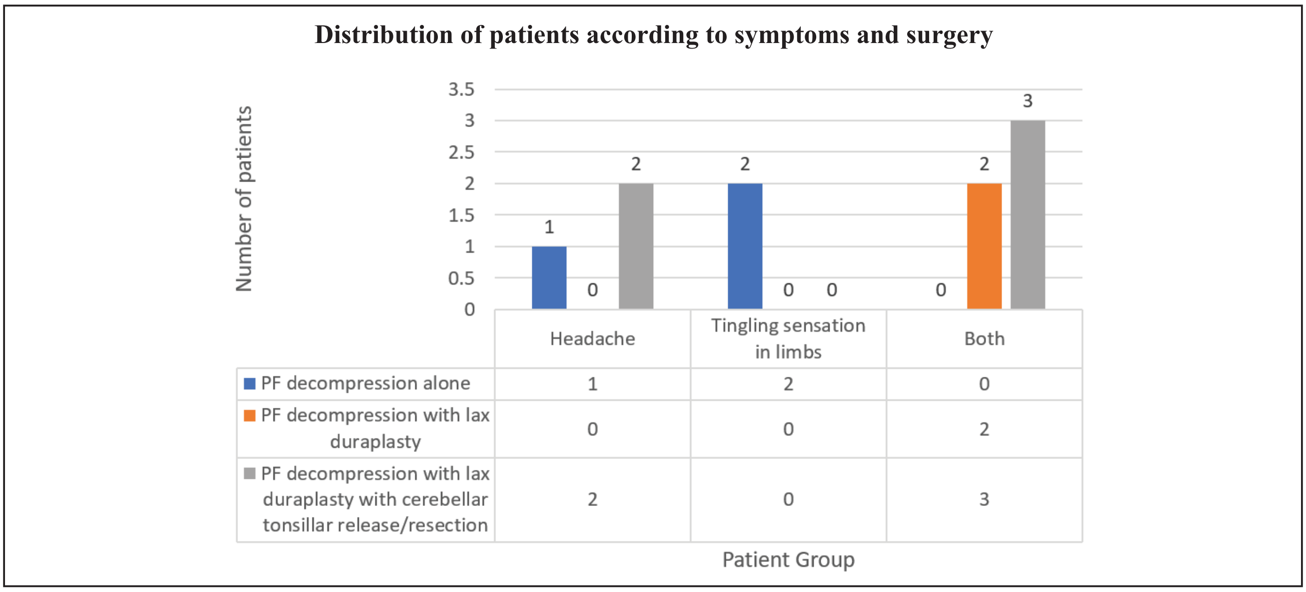

Figure 1: Distribution of patients according to symptoms and surgery

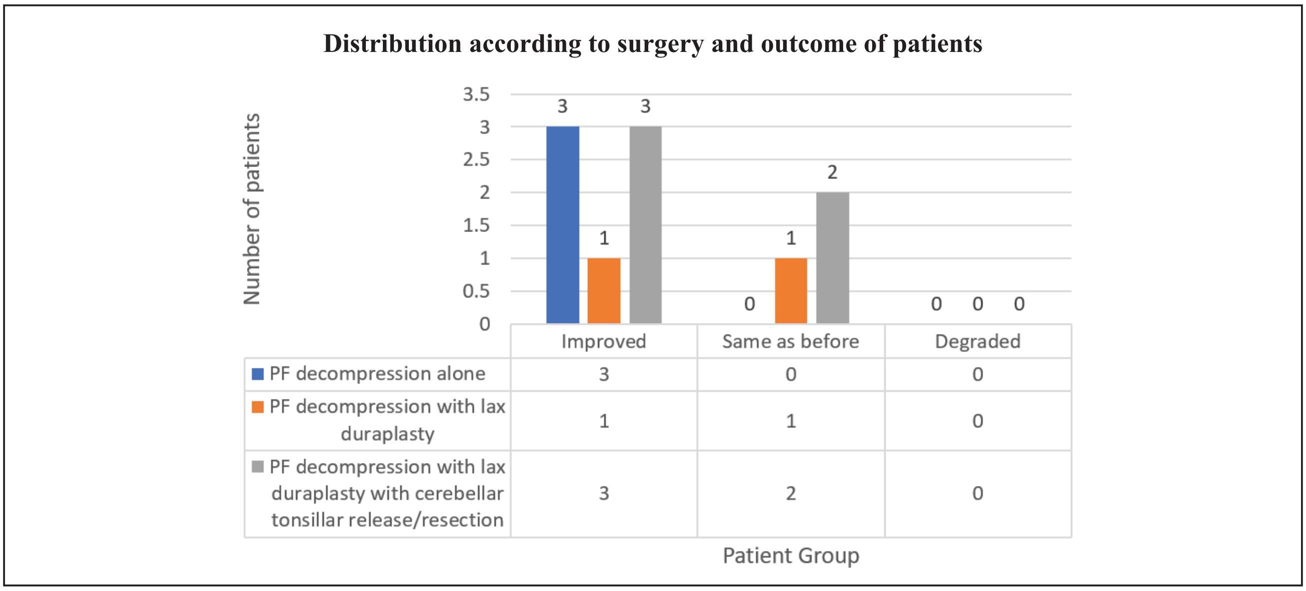

Figure 2: Distribution of patients according to surgery and clinical outcomes

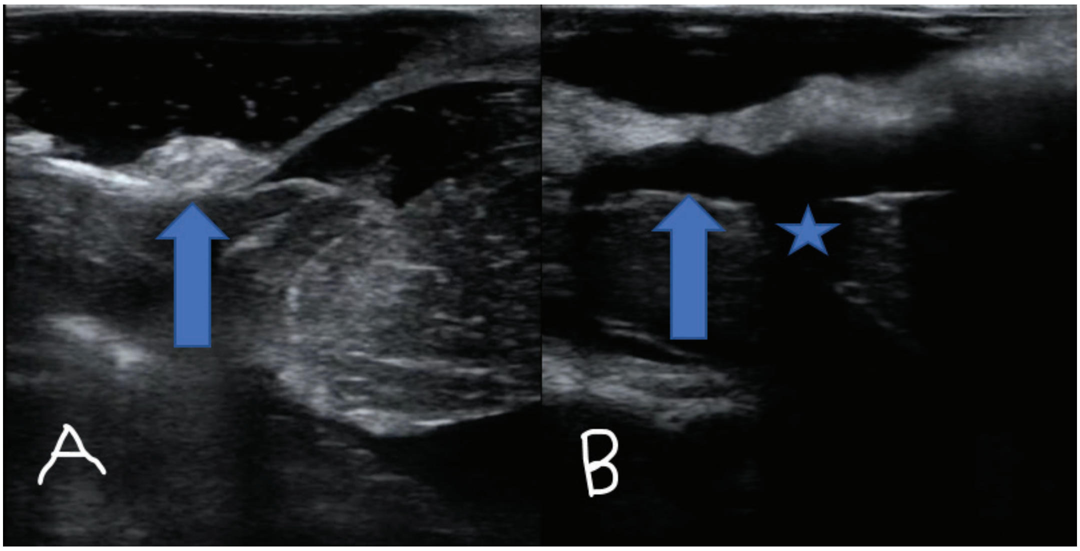

Figure 3: Intraoperative ultrasonographic picture (A) before decompression (B) after decompression. Blue Arrow shows the subdural CSF flow. Blue star shows flow through foramen of magendie

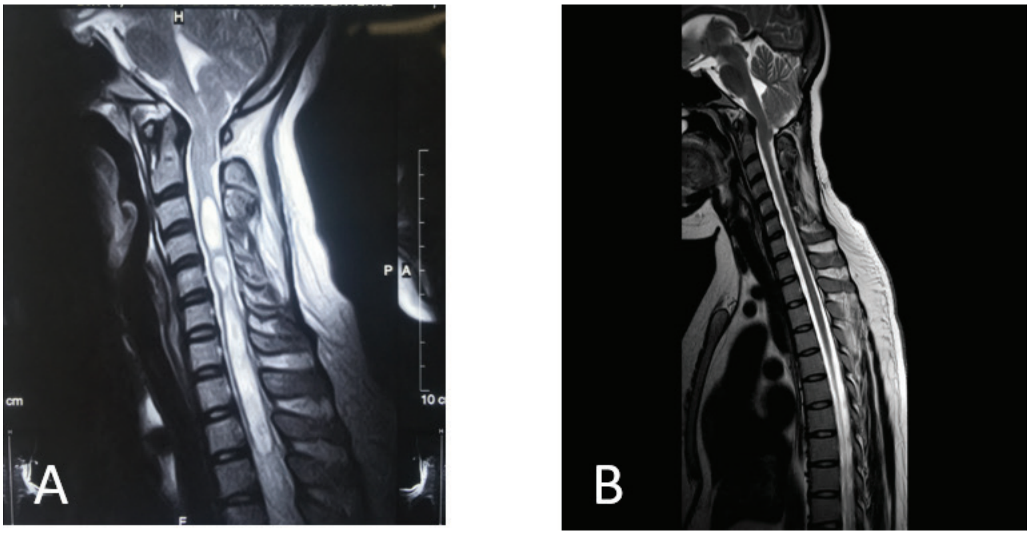

Figure 4: MRI craniovertebral junction showing resolving syrinx (A) Preoperative (B) Postoperative

Patient Number	Age (years)	Sex	Symptoms	Surgery	Syrinx in preoperative period	Syrinx at 1 year postoperative period	Clinical Outcome at 1 year post surgery period
1	28	Male	Headache and tingling sensation in B/L upper limbs	PF decompression with lax duraplasty with resection of cerebellar tonsils	Present	Resolved	Same as before
2	56	Female	Headache	PF bony decompression only	Absent	Absent	Improved
3	21	Male	Headache	PF decompression with lax duraplasty with release of cerebellar tonsils	Present	Resolved	Improved

4	24	Female	Tingling sensation in B/L upper limbs	PF bony decompression only	Present	Resolved	Improved
5	24	Female	Tingling sensation in B/L upper limbs	PF bony decompression only	Absent	Absent	Improved
6	63	Female	Headache	PF decompression with lax duraplasty with release of cerebellar tonsils	Present	Resolved	Same as before
7	25	Female	Headache and tingling sensation in B/L upper limbs	PF decompression with lax duraplasty with resection of cerebellar tonsils	Present	Resolved	Improved
8	28	Male	Headache and tingling sensation in B/L upper limbs	PF decompression with lax duraplasty	Present	Resolved	Same as before
9	25	Male	Headache and tingling sensation in B/L upper limbs	PF decompression with lax duraplasty	Absent	Absent	Improved
10	33	Female	Headache and tingling sensation in B/L upper limbs	PF decompression with lax duraplasty with release of cerebellar tonsils	Present	Resolved	Improved

Table 1: Summary of patient data

Discussion

The goal of surgery in Chiari Malformation Type I is adequate decompression with pulsatile flow of CSF around the CVJ.[9] There are various theories suggested for the surgical treatment of symptomatic Chiari Malformation Type I. These theories stress the importance of CSF dynamics near the CVJ. Various studies suggest that Chiari Malformation Type I results from a mesodermal defect after the closure of the neural folds. This leads to underdevelopment of the basichondrocranium thus resulting in a small posterior fossa and overcrowding of neural elements which ultimately leads to abnormal CSF dynamics at the CVJ.[10-12] Williams B et al proposed the "cranial-spinal pressure dissociation theory". He stated that CSF flow obstruction in patients with Chiari Malformation Type I at the CVJ due to the herniated cerebellar tonsils results in a differential pressure gradient between the cranial and spinal cavities. This significant pressure differential occurs during the systolic cardiac cycle or during routine activities such as sneezing and coughing. This effect increases the impact of tonsils at the CVJ which drives CSF into the Virchow–Robin and interstitial spaces, thus leading to the formation of a syrinx and subsequent neurological symptoms.[13,14] Similarly, Gardner WJ et al stated that an incomplete embryonic opening of the fourth ventricle outlet leads to communication between it and the central canal which contributes to a "water hammer" effect into the central canal with each arterial pulse. This continued process leads to formation of syrinx.[15,16]

Recent advances in imaging technology support the role of abnormal CSF dynamics in the pathophysiology of Chiari Malformation Type I. Magnetic resonance imaging and phase-contrast studies show compromised subarachnoid space in the dorsal spinal-medullary region and the small posterior fossa volume in patients with Chiari Malformation Type I.[3] Oldfield EH et al demonstrated that expansion of the brain during systole imparts a downward force through the CVJ. This creates an abnormal piston motion of the cerebellar tonsils. This abrupt motion compresses the spinal cord and medulla, forcing CSF into the cord via the lymphatic vessels and resulting in the formation of syrinx.[17]

Various procedures ranging from posterior fossa bony decompression alone to resection/release of cerebellar tonsils have been advocated for restoring the normal CSF dynamics at the CCJ. The more invasive procedures are more associated with complications. Also, there is no defined procedure for adequate decompression. Intraoperative ultrasonography can be useful tool to minimize the extent of surgery and maximize the effect of decompression at the same time.

Isu T et al used intraoperative ultrasonography in seven patients to determine if duraplasty was needed to decompress the posterior fossa. Six of seven patients whose intraoperative ultrasonography studies showed evidence of adequate decompression improved clinically.[18] Yeh DD et al in their series of 130 patients who underwent intraoperative ultrasonography such that the CSF dynamics at the CVJ could be analysed, 40 (31%) patients had adequate decompression with bone removal alone and 90 (69%) had abnormal ultrasonographic findings after bone decompression. Of the 90 patients, 85 patients underwent additional duraplasty and tonsillar shrinkage while the remaining five patients had preoperative comorbid condition due to which additional procedure could not be done inspite of abnormal intraoperative USG findings. At 20 months follow up, 124 patients improved symptomatically. In this series, 31% patients had bony decompression only of which only four patients had failure.[5] In our study, three patients underwent PF decompression alone based on adequacy of decompression confirmed by intraoperative USG and all three patients improved symptomatically at one year post surgery follow up. Also one patient who underwent PF decompression alone had cervical syrinx on preoperative imaging (MRI) which resolved at one year follow up seen on post-surgery imaging (MRI). In another study on the use of intraoperative ultrasonography, Yundt and coauthors found that none of the three children with Chiari Malformation Type I in their study required duraplasty and all improved clinically after bone removal alone. They used the presence of retro-cerebellar space and decreased pulsatility of the tonsils as an index of adequate decompression.[19] In our study, we deemed decompression to be successful when the piston-like activity of the cerebellar tonsil was alleviated and CSF spaces and flow through foramen magnum, were present as seen on intraoperative USG.

All the patients who underwent PF decompression alone after evaluation by intraoperative ultrasonography improved symptomatically. This shows the effectiveness of intraoperative ultrasonography in deciding the adequacy of decompression.

Navarro R et al in their series, had mentioned that 15 patients required further surgical decompression, including duraplasty with/without tonsillar manipulation on the basis of intraoperative ultrasonographic findings and success rates of 72 and 68% respectively were achieved. They mentioned that complications were higher in the duraplasty group.[20] In our study, two (20%) patients had complications: wound gaping and wound CSF leak of which CSF leak was seen in patient undergoing PF decompression with lax duraplasty with tonsillar resection. Both of them were managed with secondary suturing.

Intraoperative ultrasound does help to minimize the extent of decompression required for CM-I. Less invasive surgery means less complications. In our study non-superiority of one procedure over the other allows us to go for lesser invasive procedure based on intraoperative USG. However, this is a retrospective study with a small sample size. A prospective study with a larger sample size is thus recommended to allow for statistical validity. Though USG is an operator dependent modality of investigation which may assumingly create variation in the result; using a protocol based examination with ultrasonographic end-points (as used in this study) shall provide uniformity of study findings.

Conclusion

The option of limiting the extent of surgical decompression to restore normal CSF dynamics across foramen magnum, which can be confirmed on intra-operative ultrasonography, can make surgery less invasive.

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