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Modified Surgical Technique for Chiari Malformation

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Abstract Craniectomy with or without cervical laminectomy is a well-recognized treatment option for patients with symptomatic Chiari malformation. However, performing craniectomy removes the protection of the suboccipital cranial bone while laminectomy may limit cervical spine movement. We undertook a retrospective study of seventeen patients who underwent conventional craniectomy and cervical laminectomy or cranioplasty alone for correction of Chiari malformation. The objective was to present cinical outcome and complications when the Chiari malformation was corrected by conventional craniectomy/ laminectomy or craniotomy alone. In the craniotomy group with eleven patients, the inside of the suboccipital bone was resurfaced prior to replacing it thereby creating space for the cerebrospinal fluid to flow after adhesionolysis and careful resection/ shrinkage of subpial tonsils without performing laminectomy. Craniectomy and C1-C2 laminectomy was done in six patients as per standard procedure. At a variable followup period of 10 years after the surgery, we did not observe any difference in clinical outcome between the two groups. The resolution of syrinx was 54.5 % in the craniotomy group as compared to 66.7% in the craniectomy/ laminectomy group, while it persisted unchanged in one patient in each group. CSF leak was noted in 2 patients in each group. Reduction of symptoms like headache, neck pain or paresthesia showed reduction in intensity in all patients except paresthesia that persisted in 1 patient of the craniotomy group. The finding of this study suggests that performing craniotomy alone is as effective as craniectomy/ laminectomy in terms of outcome and relief of signs and symptoms of Chiari malformation.

Keywords: *chiari malformation, craniotomy, craniectomy, cervical laminectomy*

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1. Introduction

Chiari malformations (CM) comprise of congenital hindbrain abnormalities that alter the structural relationship between the cerebellum, brainstem, the upper cervical cord, and the bony cranial base. It has been graded as type I-IV [1,2]. Type I is the most prevalent form. In this, the cerebellar tonsils are displaced below the level of the foramen magnum, and has a prevalence rate of 0.1-0.5% while in Type II ("classic" Chiari malformation or Arnold-Chiari malformation), both the cerebellum and the brain stem are displaced into the foramen magnum [3]. The resultant symptoms of Type I and II CM are secondary to an alteration of cerebrospinal fluid (CSF) flow dynamics between the cranial and spinal compartment at the level of the foramen magnum. To re-establish the CSF flow, decompressive suboccipital craniectomy with or without cervical laminectomy has been the mainstay of surgical treatment [4,5,6]. Type III and IV CM are rare.

Over the last 10-year period, one of the neurosurgical units in our institution has modified the surgical treatment for CM from craniectomy and cervical laminectomy to craniotomy alone. In this case series, we critically analyze and present patient outcome of one of the unit performing craniotomy alone as against patients undergoing conventional craniectomy and C1-C2 laminectomy under another unit for surgical correction of CM over a 10-year period.

2. Material & Methods

After obtaining permission from Hospital Ethical Issues Committee to use patient data for publication, we retrospectively analyzed records of all patients undergoing corrective surgery for CM at our institution over the last 10-years (2005-2016, January). A total of seventeen patients underwent corrective surgery for CM during this period. Of these patients, eleven underwent craniotomy and six craniectomy/ laminectomy. All patients were noted to have undergone a thorough clinical examination and MRI prior to surgery. All patients, including parents of the pediatric patients, were explained the advantages and possible complications of the procedure before obtaining written consent.

Of the eleven patients undergoing craniotomy alone, nine had Type I while the other two had Type II CM with a nearly similar type distribution in the craniectomy/ laminectomy group (five patients with Type I and 1 with Type II CM). Details of patient age, sex and perioperative status in the craniotomy and craniectomy/ laminectomy

groups have been summarized in Table 1 and Table 2 respectively.

Table 1. Clinical characteristics of the patients and post craniotomy status.

S. No.	Age (yr), Sex	Type of malfor-mation	Sign/ symptom	Early postoperative complications	Late postoperative improvement/ complications	Period past surgery
1	13, M	Type I	Decreased light touch sensation left side of body sparing face. Numbness, patchy loss of temperature sensation and left sided weakness	Nil	Significant improvement in terms of weakness and paresthesia	10 yr
2	5, F	Type II	Incidental due to seizure investigation.	Nil	Remained static	9 Yr
3	35, F	Type I with syrinx	Neck pain with radiation to right arm, weakness on right side, upper limb power (4/5) with reduced pinprick sensation.	Nil	MRI shows improvement in preoperative syrinx.	8 Yr
4	41, F	Type I	Neck pain with upper limb radiation, impaired sensation, cerebellar signs, dysthesia.	Nil	Improvement in neck pain and syrinx.	8 Yr
5	3, M	Type I	Headache, irritability.	Immediate postoperative localized collection that resolved spontaneously	Improvement in headache.	7 Yr
6	19, M	Type I with syrinx	Right upper limb weakness (4/5) extending to right sided hemiparesis, nystagmus, and unsteady gait.	Nil	Improvement in syrinx, persistence of paresthesia.	3 Yr
7	41, M	Type I with syrinx	Left sided neck pain and numbness/ paresthesia in upper limb.	Immediate nil but later re-admitted with CSF leak that stopped with conservative treatment.	Persistent syrinx, but improvement in neck pain and paresthesia.	4 Yr
8	30, M	Type I with syrinx	Neck and right shoulder pain with local tenderness.	Nil	Improvement in symptoms and syrinx.	2 Yr
9	34, F	Type I with syrinx	Left occipital, shoulder and inter-scapular pain. Decreased touch and pressure sensation over C5-T1 distribution.	Nil	Clinically improved and syrinx reduced.	8 months
10	17, M	Type I	Occipital headache, horizontal nystagmus.	Nil	Improvement in nystagmus and headache.	8 months
11	12, F	Type I with syrinx	Incidental due to investigation for skull osteoma. No deficits.	Nil	Continues to be asymptomatic	1 month

Table 2. Clinical characteristics of the patients and post craniectomy/ laminectomy status.

S. No.	Age (yr), Sex	Type of malfor-mation	Sign/ symptom	Early postoperative complications	Late postoperative improvement	Period past surgery
1	10, M	Type I with lumbar myelo-mening-ocele	Quadriparesis, dysmetria, cerebellar signs.	Nil	Postoperative improvement.	Lost to follow up
2	44, M	Type I with syrinx	Neck pain with radiculopathy, unsteady gait, numbness.	CSF collection at wound site.	Postoperative improvement	5 yr. Persist-ence of syrinx, advised shunt but patient reluctant
3	38, F	Type II with syrinx	Left sided body pain and numbness. Sensory impairment in C5-C8 area, local neck tenderness.	Postoperative CSF leak managed with assurance and lumbar drain.	Gradual improvement	2 yr
4	5, F	Type I with hydroce-phalus with cranios-ynostos-is	Cloverleaf skull with breathing difficulty, Piffer syndrome.	CSF flap collection managed with shunt, suturectomy.	Postoperative improvement	2 yr
5	28, F	Type I with syrinx	Neck pain, nystagmus.	Nil	Postoperative improvement	1 yr
6	10, M	Type I with syrinx	Deformity back with paresthesia in limbs.	Nil	Postoperative improvement.	9 months

Surgical procedure was performed in all patients in the prone position with a head elevation of 20-30°. Head was held in a Mayfield 3-pin frame. Patient neck was flexed at the craniocervical junction and extended at the cervicothoracic junction for a safe and easy access to the foramen magnum. Surgery was performed via a midline incision that extended from 2-3 cm above the inion to the second cervical vertebra. The inion, the midline of the occiput down to the foramen magnum, the posterior arch of C1 and the upper aspect of C2 lamina was carefully exposed. In the craniotomy group, a 2.5 cm by 3 cm suboccipital bone was lifted. Next, dura was opened that was later repaired using Lyoplant, an absorbable dura mater substitution from bovine collagen (Aesculap AG, Tuttlingen, Germany). No attempt was made to perform laminectomy of C1 and C2 vertebrae in this group. During the surgery, adhesionolysis along with careful resection/ shrinkage of subpial tonsils was performed using cautery till fourth ventricle was visible so as to achieve free flow of CSF from foramina of Magendie, Luschka and spinal

canal (compare Figure 1A from 1B). After the dural repair, the inferior part of the suboccipital craniotomy bone flap that was lifted (Figure 2A) and possibly compresses the neural structures was trimmed along with resurfacing of the inner table and central bony ridges (Figure 2B). Subsequently, this resurfaced and trimmed craniotomy bone was re-placed (Figure 3) and wound closed keeping the bony anatomy intact (Figure 4 A & Figure 4B). In contrast, in the craniectomy/ laminectomy group of patients, suboccipital craniectomy was carried out from the inferior nuchal line to the posterolateral rims of foramen magnum. In this group, bone flap was not replaced and C1 and C2 laminectomy or C1 laminectomy alone was done to restore CSF flow. In one of our patients with craniosynostosis and hydrocephalus, who underwent craniectomy/ laminectomy, VP shunt was placed prior to CM correction.

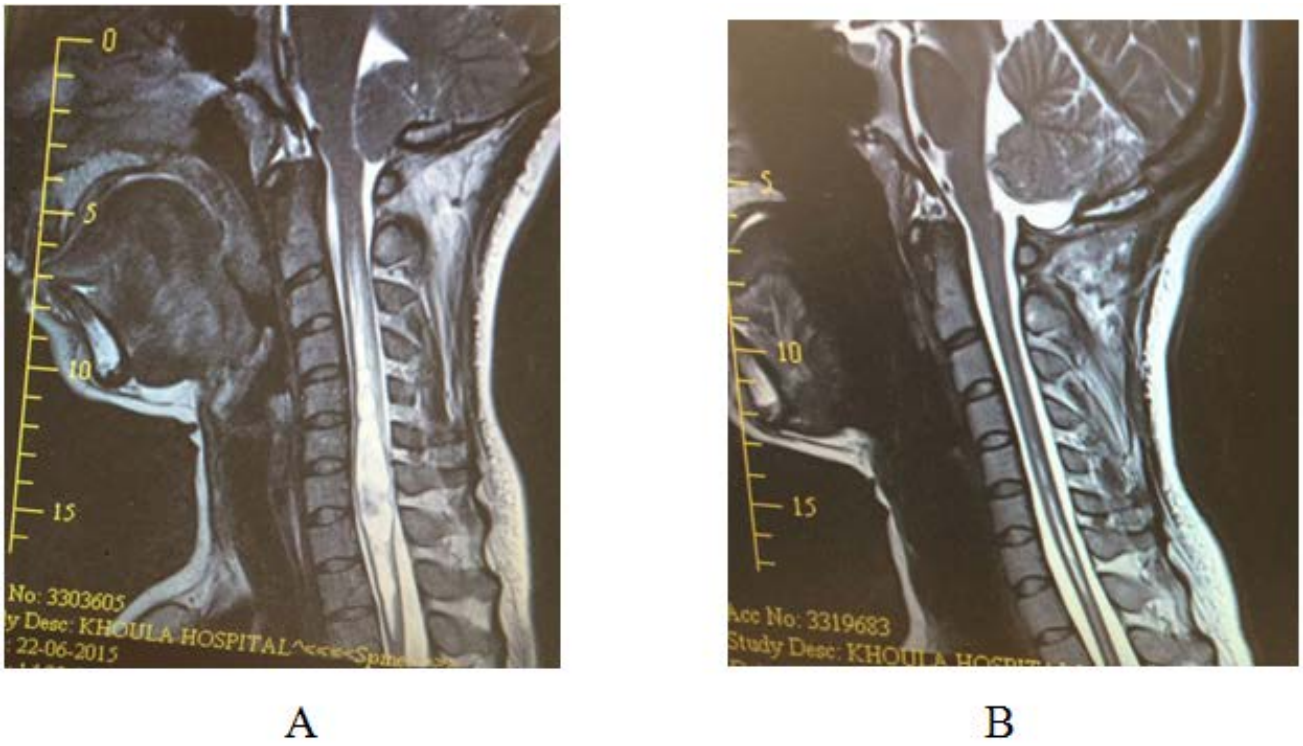

Figure 1. A is the preoperative MRI of brain and upper cervical spine in a patient with CM-I while 1B is the postoperative MRI image of the same patient after posterior fossa decompression surgery showing good restoration of CSF around the brain stem and cerebellum with a much reduced syrinx

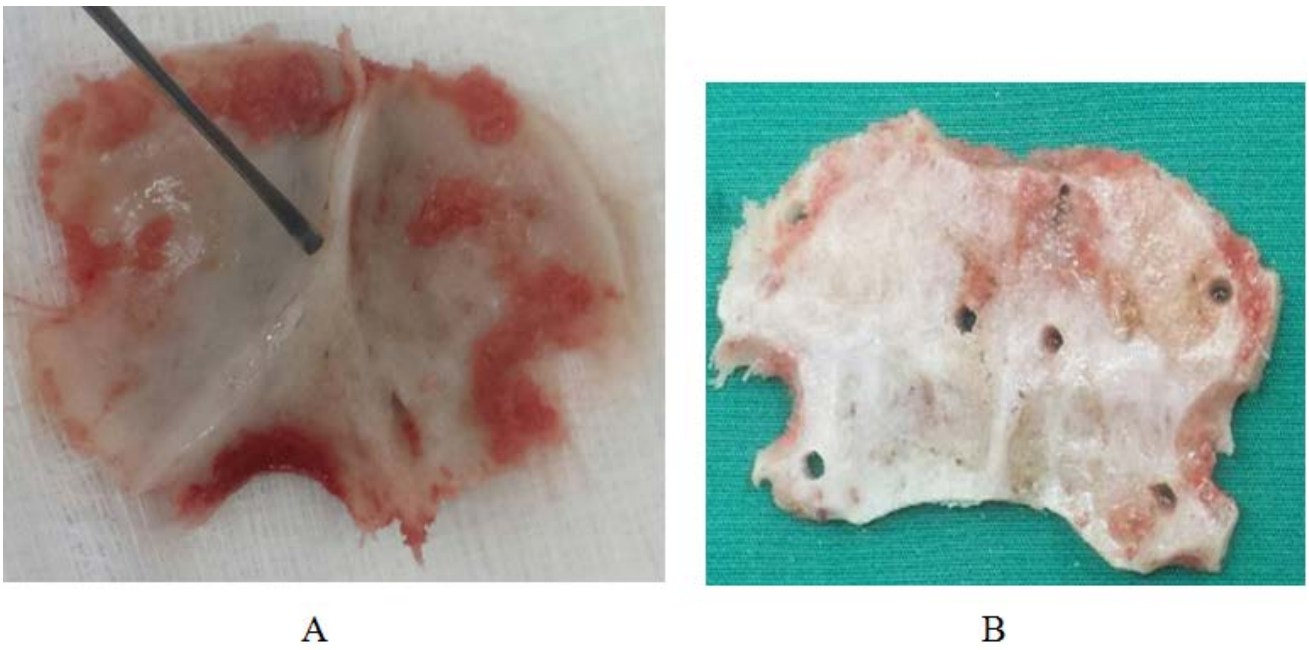

Figure 2. Pre- (A) and post-shaping (B) images of the suboccipital bone

Patients were closely followed up over the years to pick up any recurrence of symptoms at an early stage. In occasional patients, MRI was requested by the treating surgeon to assess the syrinx for its shrinkage or collapse.

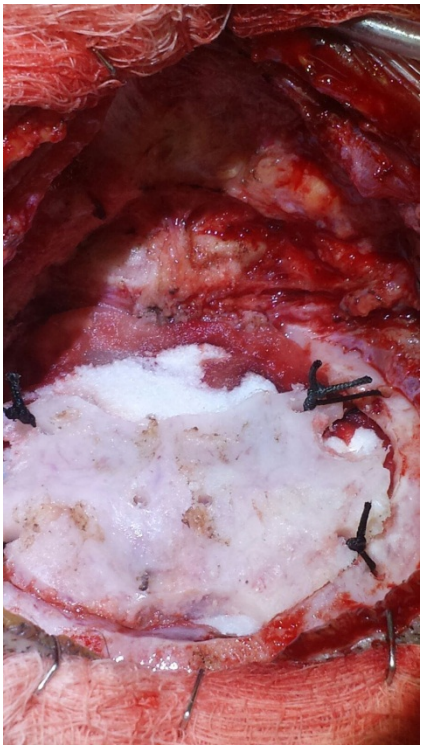

Figure 3. showing the replaced suboccipital cranial bone in the craniotomy group

3. Results

A total of seventeen patients underwent corrective surgery for CM over the last 10 years. Their data were retrieved and subsequently analyzed. The mean age of the patients in the craniotomy group was 22.7 ± 14.0 yr (range 3-41 yr) while it was 22.5 ± 16.4 yr (range 5-44 yr) in the craniectomy/ laminectomy group. Like the age, male to female sex ratio was nearly identical in the two groups. (Table 1 & Table 2).

One patient in each group had Type II CM, while the rest had Type I CM. Syrinx was noted in 54.5 % of craniotomy group of patients as compared to 66.7% in the craniectomy/ laminectomy group. In either group, syrinx persisted in one patient each while it reduced in rest of the patients. Patient with persistent syrinx in the craniectomy/ laminectomy group was advised shunt but the patient refused.

CSF leak was noted in two patients in each craniotomy (18.2%) and craniectomy/ laminectomy group (50%). They either resolved spontaneously or needed a shunt procedure.

Neck and shoulder pain, headache and paresthesia were the common presenting complaints in either group of patient. These symptoms improved in all patients of either group except for one 19-year-old patient in the craniotomy group who showed no improvement in paresthesia.

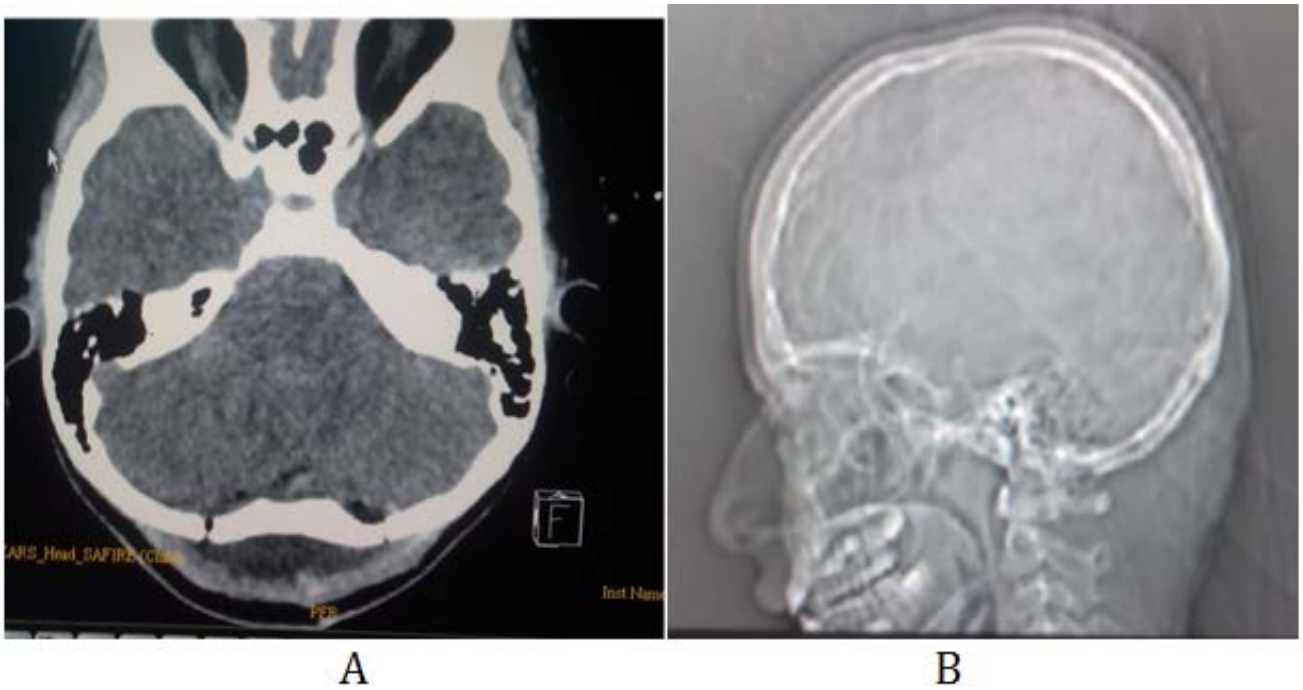

Figure 4. shows replaced suboccipital bone in one of the craniotomy patients with intact C1-C2

4. Discussion

There are several approaches to the management of CM ranging from conservative to surgical decompression. Conservative treatment is usually reserved for patients with Chiari I malformations who have minimal or equivocal symptoms without syrinx. For symptomatic patients, decompressive surgery is performed that includes suboccipital craniectomy, C1 laminectomy, dural opening, and duraplasty. In patients where macroscopic assessment shows that restoration of the CSF flow behind the tonsils and lower brainstem is not possible, intra-arachnoid exploration is performed. A C2 laminectomy is generally reserved for patients with tonsillar herniation below the midpoint of the C1-C2 interspace. Some other surgical variations include incising the outer leaf of the dura and leaving the inner leaf intact, tonsillar shrinkage, and fourth ventricular shunting. When duraplasty is undertaken, a variety of material have been used that includes: pericranial and fascia lata autografts; pericardial, fascia

lata, and dural allografts; bovine pericardium; and synthetic patches [4,5,6].

The conventional approach to surgical correction of CM is craniectomy with or without laminectomy. However, suboccipital craniectomy obviates the protection of an intact cranium against trauma, predisposes dural adhesion to soft underlying tissue and also places psychological stress to the patient. To circumvent these deficiencies, one of our neurosurgical units practices craniotomy alone without resorting to craniectomy and C1-C2 laminectomy during surgical correction of CM. The result of this retrospective study shows that the outcome is similar in both groups of patients.

The resurfacing of the suboccipital bone prior to replacement serves the dual purpose of increasing the space within it and secondly, easing the compressive forces once it is put in place. We did not note any complications that could be attributed to the replacement of suboccipital bone or avoiding laminectomy.

The more common surgical complications following conventional craniectomy/ laminectomy for CM includes CSF leakage, formation of pseudomeningocele, brainstem infarction, meningitis and cerebellar sag [5]. In the craniotomy group of our patients, one patient developed localized collection at operative site while the other was noted to have CSF leak. Both responded to conservative management. In contrast, two out of six patients in the craniectomy groups developed CSF leak. One of them was managed by placing lumbar drain while another had a CSF collection at the operative site that responded to conservative approach. No other early complications were noted in this series.

The incidence of syrinx varies between 30-70% [5]. We noted syrinx in 58.8% (10/17). Nine out of ten patients with syrinx showed gradual resolution. None of our patients were re-explored for persistence or aggravation of syrinx.

It has been observed that complete or near complete resolution of symptoms may be noted in 68% patients while 12% have mild to moderate residual deficits and 20% show no improvement following surgery [7]. Generally good results are obtained for headache or neck pain, followed by cerebellar symptoms. In this series, all patients had improvement in their headache and neck pain while one patient had persistent paresthesia in the craniotomy group. However one 5-year-old female child had CM without symptoms. CM was an incidental finding while she was being investigated for seizure. This is not unusual as Type I CM patients with <6 mm displacement of cerebellar tonsils may be asymptomatic in 70-95% [8]. This patient had a tonsillar displacement of nearly 8 mm. She underwent craniotomy for CM but her seizures remained static with no improvement in the postoperative follow up period.

This retrospective case series has three limitations. First, there was no uniformity in the clinical and radiological follow up of patients after surgery in the two groups. This varied as per consultant's instructions rather than a protocol. Second, the duration of follow up was variable (two month to ten years) as we selected all patients operated within a ten-year period. Third, we did not compare the results statistically, as the sample size was small and non-proportionate.

In conclusion, results of this retrospective case series show that restoration of normal CSF dynamics from the fourth ventricle to the subarachnoid space and relief of brainstem compression in patients with symptomatic CM can be safely achieved by craniotomy alone without resorting to craniectomy/ laminectomy. Replacing the suboccipital bone after reshaping its interior or avoiding laminectomy does not affect the clinical outcome postoperatively in terms of improvement in sign and symptoms of CM.

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