Breaking Barriers for Cerebrospinal Fluid Flow in Chiari Malformation Type I: "What and How Much Is Enough?" A Retrospective Analysis of 74 Cases

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Abstract

Introduction: Chiari malformation type I is a collection of hindbrain abnormalities, for which natural history of the disease process is not clear. The challenge is to identify which patients will benefit most from posterior fossa decompression.

Objectives: To identify important surgical implications that most likely benefit patients with Chiari malformation type I by analyzing and reviewing various operative interventions in these patients with appropriate symptoms and then following their course.

Subjects & Methods: Retrospective analysis of 74 operated Chiari malformation type I with syrinx adult patients was done.

Results: No definite pattern of progression in natural history of disease was noted. Most of the patients who were symptomatically stable for months to years presented with recent rapid progression. The most common symptom was suboccipital pain. The most common finding was lower extremity weakness. On clinical presentation basis, patients were divided into three categories: foramen magnum compression syndrome, central cord syndrome, and cerebellar syndrome. Most patients in our study fall in first category. Foramen magnum decompression with atlas posterior arch removal and sometimes partial C2 laminectomy depending on extent of tonsillar descent as well as augmentation duraplasty was done in most patients. Improvement was seen in foramen magnum compression syndrome group more significantly.

Conclusions: Individualized surgical techniques for breaking the barriers of cerebrospinal fluid (CSF) flow in Chiari malformation type I with syrinx to restore normal CSF dynamics across craniocervical junction provide the pragmatic solution. The trend is toward balance between optimum wide decompression as compared with long craniocaudal decompression and preserving normal integrity.

Keywords
- Chiari malformation type I
- syrinx
- foramen magnum decompression

Introduction

Chiari malformation (CM) type I are becoming more commonly faced by neurosurgeons with increased uses of magnetic resonance imaging (MRI). This complex entity is a collection of hindbrain abnormalities having long medical history. Several classic reviews have been attempted to clarify a variety of complex associated issues. Despite the volumes of publications on the subject, the literature review shows that the natural history of this disease process has not been established. Abnormal cerebrospinal fluid (CSF) dynamics across craniovertebral junction and frequent spinal cord cavitation are among the prominent facets of disease process.

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The clinical examination is crucial in the diagnosis, and the challenge is to identify which patients will benefit most from posterior fossa decompression. Controversy regarding operative intervention continues in patients without a syrinx, and surgical indications may vary among surgeons, especially for subjective symptoms such as headache. Management is further complicated by variability in technical aspects of decompression. In our study, complete surgical experience of 74 patients who were operated upon for adult presentation CM type I with syrinx was retrospectively analyzed. We set out to identify important surgical implications that most likely benefit patients. By analyzing operative intervention in patients with appropriate symptoms and then following their course, we hope to outline the present status of this entity.

Subjects and Methods

This is a retrospectively analyzed study of 74 patients (mean age: 28.8 years, range: 6–58 years, 55 males and 19 females) who were managed surgically for adult CM type I from 1998 to 2016. The inclusion criteria were symptomatic patients with appropriate clinical findings and MRI consistent with CM type I. Patients included in this study were followed in postoperative period for at least 18-month duration. Patients in whom another cause was established (trauma, arachnoiditis, and tumor), atlantoaxial dislocation, and those who lost follow-up were excluded from study. Control group consisted of the patients with CM type I in long-term follow-up in outpatient department (OPD) who opted for conservative management. Because of this, statistical comparison was not possible between these two groups.

Pain in the suboccipital or cervical region was the predominating symptom in most patients (56%). Average duration of symptoms clearly related to CM type I was 4.2 years (range: 2 months to 15 years). In current era of MRI, latency is reduced. The clinical profile of patients is summarized in (►**Table 1**). The radiologic investigations included MRI craniovertebral junction with screening of the spine and dynamic computed tomography (CT) of the craniovertebral junction in patients with evidence of skeletal anomalies on plain X-ray. The extent of tonsillar herniation in relation to foramen magnum and atlas (C1) arch was meticulously measured in millimeter in all cases along with screening for syrinx. There was associated syrinx in 47 (64%) cases (►**Fig. 1**). The rostrocaudal extent and maximum diameter of syrinx were clearly documented on preoperative scans as a baseline for subsequent follow-up. The summary of radiologic findings is presented in (►**Table 2**).

Surgical management was standard foramen magnum decompression with C1 arch removal and partial axis (C2) laminectomy, depending on extent of tonsillar descent in all the cases (►**Fig. 2**). A wide pericranial graft was routinely harvested from occipital region in all the cases. Intradural manipulation was individualized in form of adhesiolysis (16%) extrapial coagulation of tonsillar tip (13%), and in early phase of study plugging of obex (4%) was also performed. Augmentation duraplasty was done using either pericranial graft (83%) or synthetic graft (8%) (►**Fig. 3**). Fibrin glue reinforcement of suture line was done in 62% patients. Intervention for syrinx without craniovertebral decompression in form of syringostomy (two patients) and syringoplural shunt (two patients) was done in early phase of study. The surgical procedures performed are summarized in (►**Table 3**). The outcome analysis was based on postoperative clinical improvement and reduction in diameter of syrinx on postoperative MRI and categorized into improved, unchanged, or worsened.

Table 1 Presenting signs and symptoms of patients having Chiari malformation type I

Signs and symptoms	No. of cases	%
Suboccipital pain	42	56
Weakness	34	45
Hand atrophy	26	35
Spasticity	24	32
Hyperactive deep tendon reflexes	32	43
Numbness (≥ 1 limbs)	32	43
Loss of temperature sensation	30	39
Posterior column loss	24	31
Facial numbness	2	3
Unsteadiness	30	40
Nystagmus	35	47
Tinnitus	5	7
Deafness	2	3
Cerebellar signs	20	27
Horner's sign	5	6

Results

Out of 74 patients included in study, mean age was 28.8 years (range: 6–58 years). The majority of the included patients were in second or third decades (43%), with male preponderance (55 males and 19 females). Average duration of symptoms clearly related to CM type I was 4.2 years (range: 2 months to 15 years). In our study, no definite pattern of progression in natural history of disease was noted. Most of the patients who were symptomatically stable for months to years presented with recent rapid progression. The most common symptom was suboccipital pain, brought on by neck movements or Valsalva's maneuver. Ten patients had headache as the only complaint, and one-half of the patients were initially labeled as functional. The most common finding included lower extremity weakness, hyperreflexia, and spasticity with atrophy in upper extremities (►**Fig. 4**). More than one-third of patients had sensory abnormalities in the form of decreased pain and temperature sensation in the upper limb and decreased proprioception in the lower limb. Nystagmus was present in 35 (47%) patients, which was of downbeat type. Other cerebellar signs were present in 20 (27%) patients and Horner's sign in 5 (6%) patients. Cranial nerve involvement in form of facial numbness was present in two (3%) patients, tinnitus in five (6%), and deafness in two (3%) patients.

On the basis of clinical presentation, patients were divided into three categories: foramen magnum compression

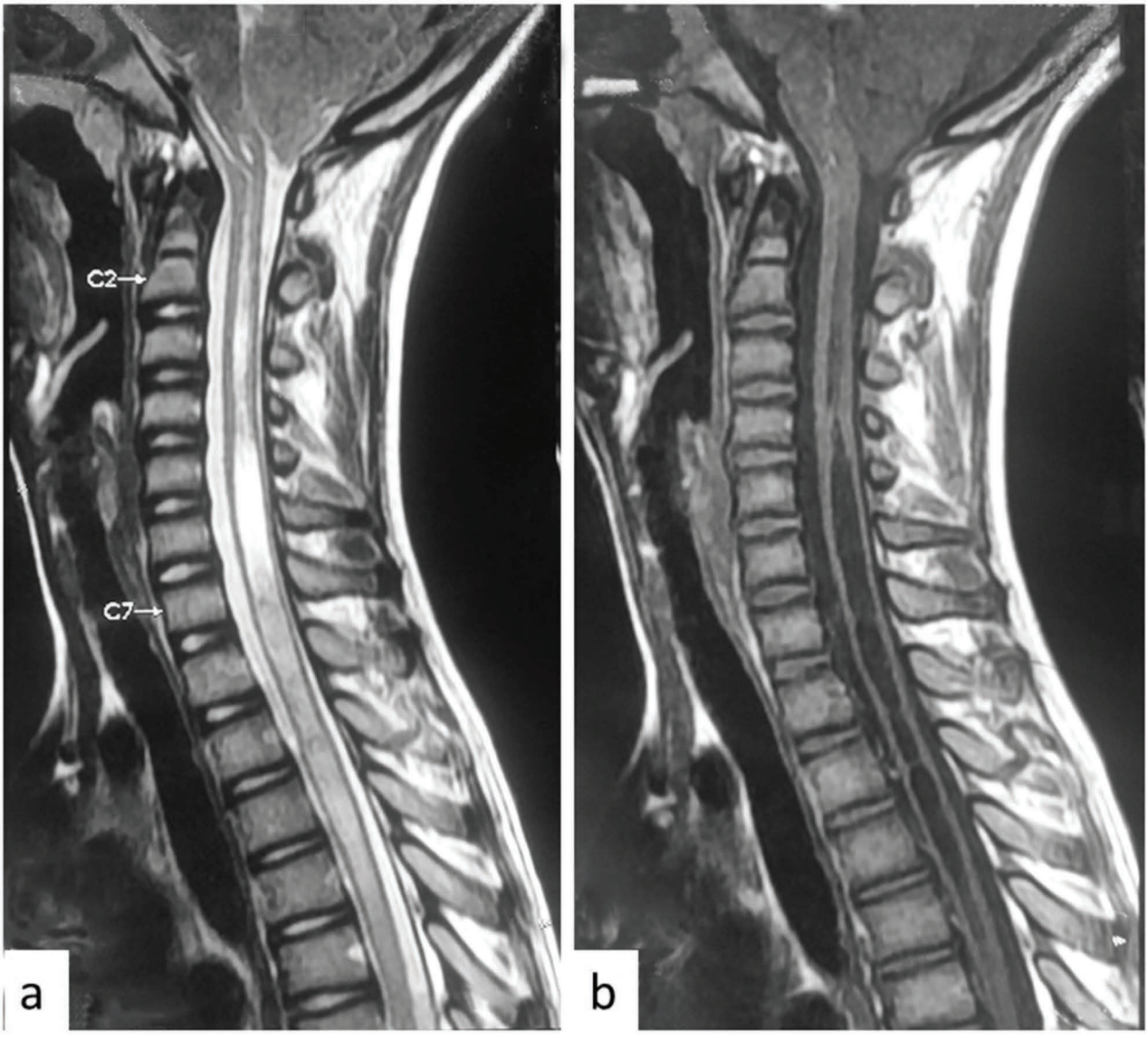

Fig. 1 Magnetic resonance imaging (MRI) of the spine showing Chiari malformation type I with syrinx formation in cervical cord in **(a)** T2- and **(b)** T1-weighted sequence.

Table 2 Radiologic details of Chiari malformation type I and associated bony anomalies

Radiologic features	No. of cases	%
Tonsillar herniation		
Foramen magnum	5	6
C1 arch	31	42
C2 lamina	27	36
C3 lamina and below	11	16
Skeletal anomalies		
Basilar invagination	18	25
Atlanto–occipital assimilation	15	20
Fused cervical vertebrae	9	12
Syringomyelia (Cine–mode magnetic resonance imaging)	47	64

syndrome (suboccipital pain, weakness, spasticity, hand atrophy, hyperactive deep tendon reflexes, facial numbness, and Horner's syndrome as predominant presenting features), central cord syndrome (dissociative sensory loss, segmental lower motor weakness in upper extremity, and long tract signs), and cerebellar syndrome (unsteadiness, nystagmus, and other cerebellar signs). Most patients in our study fall in the category of foramen magnum compression syndrome. MRI study showed tonsillar descent up to C1 arch in 31 (42%) patients, C2 lamina in 27 (36%), and subaxial spine (C3) or below in 11 (16%) patients. The tonsillar descent was meticulously calculated in millimeter in all the cases. There was associated syringomyelia in 47 (64%) patients, principally in the cervical cord. Entire rostrocaudal extent of syrinx was analyzed. Maximum diameter in millimeter was measured and documented as baseline for objective assessment of improvement in postoperative follow-up period. Associated bony anomalies such as basilar invagination, atlanto-occipital assimilation,

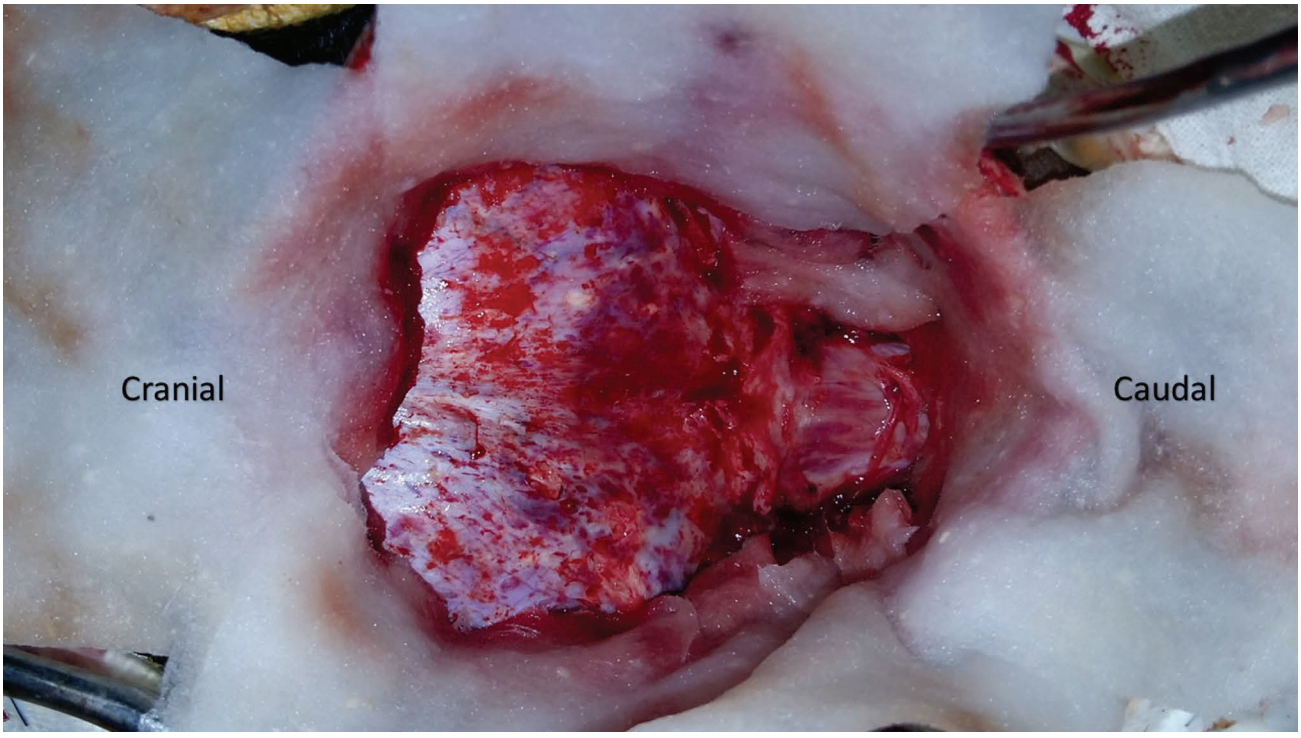

Fig. 2 Exposure after standard foramen magnum decompression, atlas posterior arch removal, and partial axis (C2) laminectomy, depending on extent of tonsillar descent in MRI of the spine.

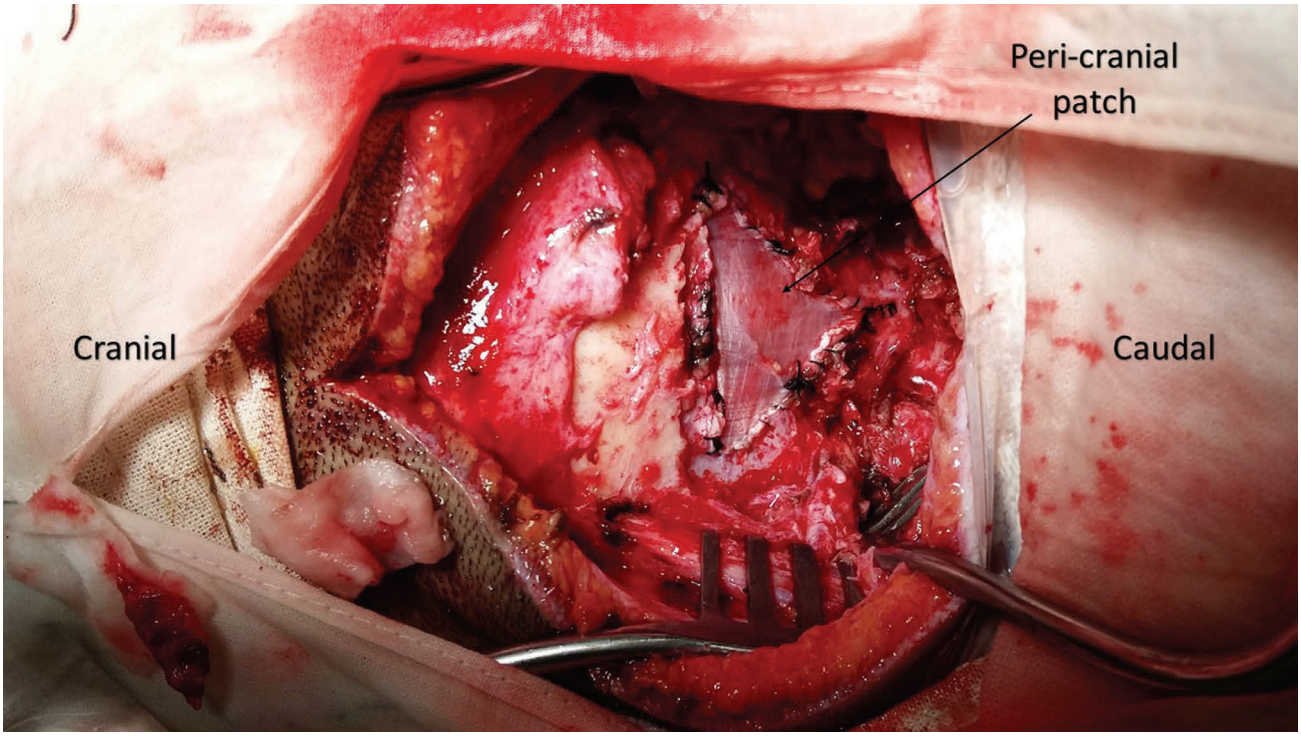

Fig. 3 Augmentation duraplasty using pericranial patch after bony bridge decompression.

and fusion of the cervical vertebrae were seen in 20 (27%) patients. In all these subset of patients, dynamic CT of the craniovertebral junction was done to rule out atlantoaxial dislocation (►**Fig. 5**a, b). Foramen magnum decompression with C1 arch removal and partial C2 laminectomy depending on extent of tonsillar descent was done in most patients. The extent of suboccipital craniectomy was up to medial margins of occipital condyles in the initial phase of this study, followed by suboccipital craniectomy limited to decompression of entire posterior surface of tonsils in the recent phase of

this study (►**Figs. 6**, **7**). The dense constrictive band causing intradural constriction was seen in 22 (30%) patients in whom intradural exploration was done (►**Figs. 6**, **8**). Adhesiolysis was done in 12 (16%) patients. In severe tonsillar ectopia occluding the foramen of Magendie, extrapial coagulation of tonsillar tip was done in 10 (13%) patients. In nine (12%) patients, a thin membrane was found covering the obex that on piercing. free flow of CSF was established. In all cases of intradural manipulation, choroid plexus in the fourth ventricle was visualized, and free flow of CSF into subarachnoid space was ensured. In the early phase of this study, in three (4%) patients, an occluding muscle plug was placed in dilated central canal at the obex to interrupt its hydrodynamic continuity with the fourth ventricle. This obex plugging procedure was abandoned later due to more neurologic complications and change in concept of syrinx pathophysiology. Augmentation duraplasty with autologous pericranial graft was done in 64 (83%) patients, and synthetic graft in 6 (8%) patients. Reinforcement of suture line with fibrin glue was performed in 48 (62%) patients. Intervention for syrinx was done in four (6%) patients (►**Table 4**). Postoperative complications included CSF leak in nine patients and wound infection in three patients, which were tackled with uneventful recovery. With the experience of previous decompressive procedures addressing to basic pathophysiology of CSF dynamics in syringomyelia, surgical strategy evolved from more aggressive approach into simpler and least invasive approach in the recent study. At 6-month follow-up, 42 patients had improvement, and 19 had stabilization in clinical course with worsening of symptoms seen in 13 patients (►**Table 5**). Significant improvement

Table 3 Surgical details

Surgical Methods	No. of cases	%
Foramen magnum decompression		
Suboccipital craniectomy	70	95
C1 laminectomy	42	55
C2 partial laminectomy	15	20
Intradural exploration		
Adhesiolysis	12	16
Extrapial coagulation of tonsillar tips	10	13
Plugging of obex	3	4
Duraplasty		
Pericranial graft	64	83
Synthetic graft	6	8
Reinforcement by fibrin glue	48	62
Intervention for syrinx		
Syringostomy	2	3
Syringoplural shunt	2	3

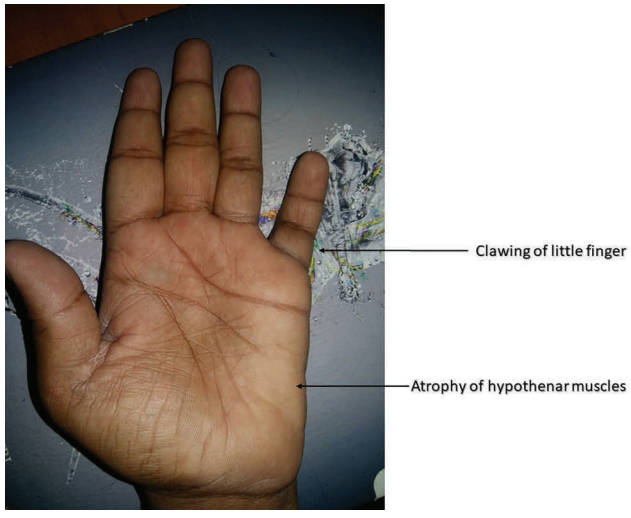

Fig. 4 Clinical photograph of hand muscle atrophy in Chiari malformation type I with syrinx.

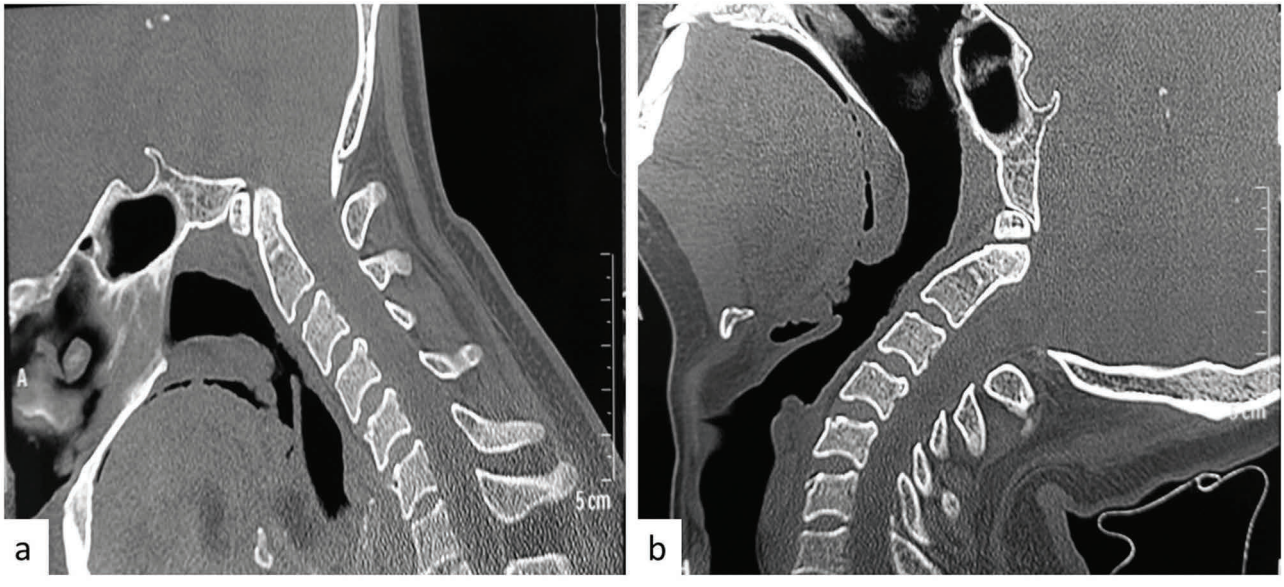

Fig. 5 **(a, b)** Dynamic computed tomography scan of craniovertebral junction to rule out bony instability in Chiari malformation type I with syrinx.

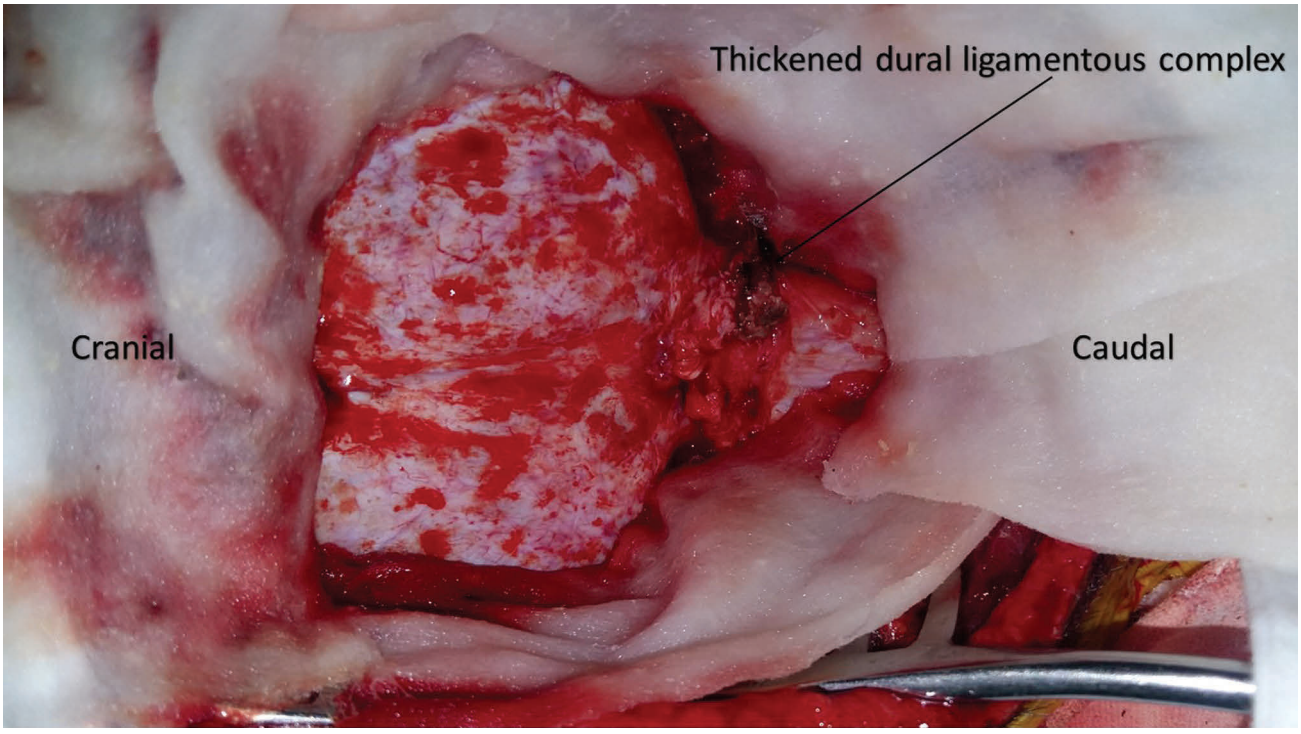

Fig. 6 Intraoperative image showing thickened dural ligamentous complex at the level of craniovertebral junction causing compression in Chiari malformation type I.

was seen in foramen magnum compression syndrome group. Follow-up MRI of the cervical spine in these patients showed marked reduction in syrinx size and well-decompressed foramen magnum with adequate CSF space around the cervicomedullary junction as compared with preoperative MRI (►**Figs. 9**, **10**).

Many patients in our study who initially improved with surgery frequently returned to their presurgical state or sometimes even became worse and continued to deteriorate. Whether this denotes the natural course of the condition in a subset of this patient population is not known. In our study an attempt is done to overcome this bias by analyzing patients with CM type I who opted for conservative management as control group. This comparative analysis, unique in our study, is instrumental to ascertain the true benefits of surgery over what may be the natural history of this condition.

Discussion

The natural history of syringomyelia varies from spontaneous and complete regression to progressive neurologic deficits rightly described as "relentlessly progressive" by Lord Brain.[1] The unpredictable clinical course of syringomyelia causes difficulties and controversies regarding management and continues to pose challenges. There is no currently clear consensus about the optimal therapy, and differing opinions are found in the literature. Another problem, encountered in interpreting the available information, is that a fairly long follow-up is required because syrinx progression may occur slowly over time. However, there is no effective nonsurgical alternative to operative decompression for patients with symptomatic CM type I. The detailed understanding of the underlying pathophysiologic mechanisms is required to identify which patients will benefit most from posterior fossa decompression.

Pathophysiologic Basis of Clinical Manifestations of Disease

The CM type I consists of caudal displacement of cerebellar tonsils into the upper cervical canal, causing direct compression of the cerebellum or medulla at foramen magnum, which was the basis of suboccipital headache and neurologic signs and symptoms arising from the cerebellum and medulla. The full foramen magnum potentially compresses the herniated cerebellar tissue and restricts normal CSF flow across the craniovertebral junction, which increases the CSF movement in the spinal canal resulting in syrinx formation. The spinal cord damage starts centrally and spreads centrifugally to involve other spinal cord structures. Characteristically, the decussating fibers of spinothalamic tract conveying pain and temperature sensation are compromised initially. This results in loss of pain and temperature in "vest like" bilateral distribution with the preservation of soft touch sensation and proprioception, characteristically described as dissociation of sensory loss. With forward extension of disease process, the anterior horn cells become involved at the level of lesion resulting in segmental lower motor neuron weakness. Similarly lateral extension results in Horner's syndrome, and dorsal extension causes involvement of posterior column.

Selection of Candidates for Surgery

The likelihood of improvement of symptoms with surgery should influence the decision to recommend surgery.

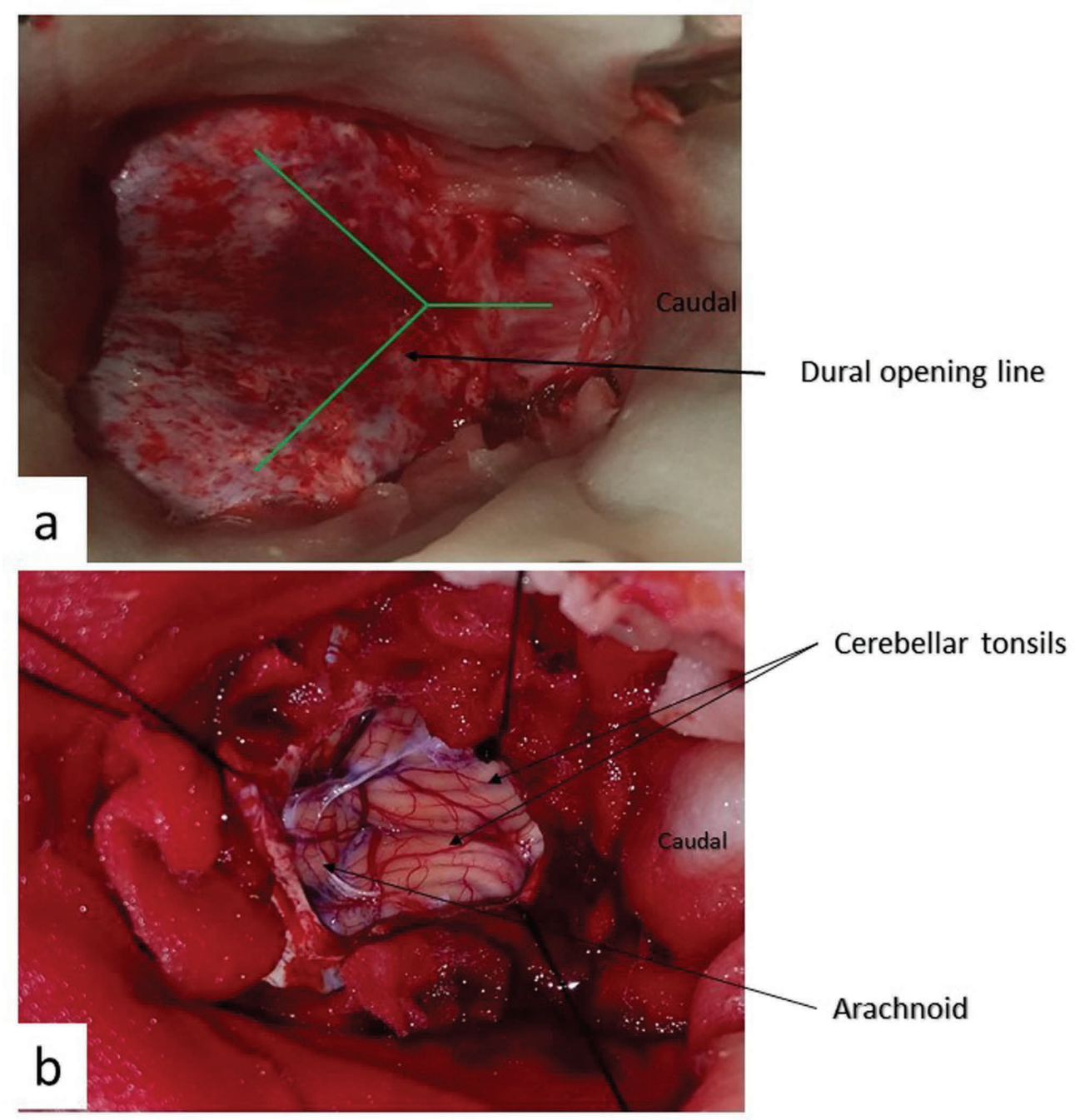

Fig. 7 Intraoperative images showing **(a)** dural incision line across the craniovertebral junction (in green) and **(b)** exposure of both tonsils beyond foramen magnum and arachnoid band for adhesiolysis.

All the clinical manifestations of syrinx associated with CM type I do not respond equally to surgery. However, early diagnosis and surgical treatment of syringomyelia are essential to arrest progressive myelopathy and prevent further loss of neurologic function. Many authors[2] have recognized the prognostic value of certain clinical variables including clinical syndromes. Suboccipital headache caused by tonsillar impaction respond well to adequate decompressive therapy. Pyramidal tract manifestations and spinothalamic sensory loss improve as pressure of the cysts on these pathways is reduced. Weakness and atrophy of the hands show little improvement because of destruction of corresponding anterior horn cells. Similarly dysesthetic pain that is a form of denervation dysesthesia due to destruction of ascending spinal pathways with thalamic projection responds poorly to decompressive therapy. Each patient must be judged individually keeping in mind that progression of symptoms can sometimes occur rather abruptly. Because some of the neurologic deficits tend to become fixed once developed, most patients with Chiari and a distended syrinx must be candidates for surgery. CM presenting with lower cranial nerve involvement or symptoms of direct brainstem compression must also be candidates for surgery.[3]

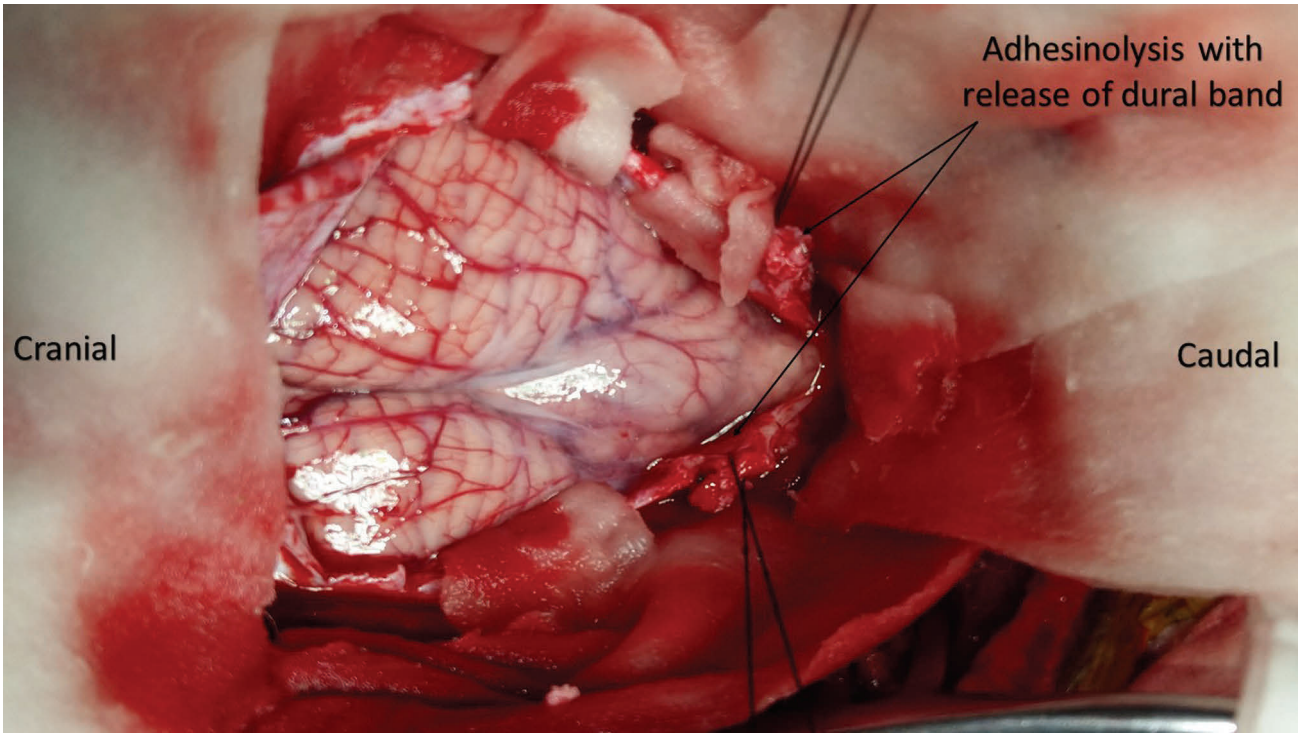

Fig. 8 Intraoperative image showing cutting of dural-ligamentous complex band at the level of craniovertebral junction.

Table 4 Surgical outcome at 6 months of follow–up

Surgical procedure	Total	Better	Worse	No change
1. Foramen magnum decompression with duraplasty	58	35	9	14
2. Foramen magnum decompression with intradural exploration and duraplasty	12	6	4	2
3. Intervention for syrinx	4	1	0	3

Table 5 Outcome in long–term follow–up of 74 cases grouped in categories

Outcome	Foramen magnum compression group	Central cord syndrome group	Cerebellar syndrome group
Improved	31	5	6
Unchanged	3	12	4
Worse	4	6	3
Total	38	23	13

Surgical Techniques Followed, Its Correlation to Underlying Pathophysiologic Mechanisms and Recommendations

Improved understanding of the pathophysiology of syringomyelia encourages the design and implementation of procedures directed toward eliminating the obstruction of CSF pathways, that is, strategies designed to reverse the pathophysiologic process underlying syringomyelia. The purpose of the operation is to enlarge the bony area of the craniocervical junction and expand the dura, surrounding the brainstem to effectively open the CSF pathways at the foramen magnum for providing effective and lasting treatment of syringomyelia with low morbidity. The specific surgical steps in this operation continue to undergo modification as surgeons attempt to identify the optimum treatment. This is usually dictated by patient characteristics and surgeons experience.

The management strategy follows a “top down” rule.[4] Treatment always begins by addressing hydrocephalus, if it is present. A ventriculoperitoneal shunt placed in patients with hydrocephalus may relieve both cerebellar ectopia and dilatation of the central canal as well. In 1950, Gardner and Goodall at the Cleveland Clinic recognized the association of the CM type I with syringomyelia.[5] They postulated that the outlets of the fourth ventricle were occluded by the CM type I and that a water-hammer pulsation was directed from the fourth ventricle, through the obex and into the central canal of spinal cord, leading to pulsatile expansion of the central canal to form the syrinx. To reverse this process, Gardner performed the surgical procedure that removed the bone from the posterior aspect of the foramen magnum, opened the fourth ventricle to the subarachnoid space, and plugged the obex.[5] In the 1970 Logue introduced a less invasive alternative

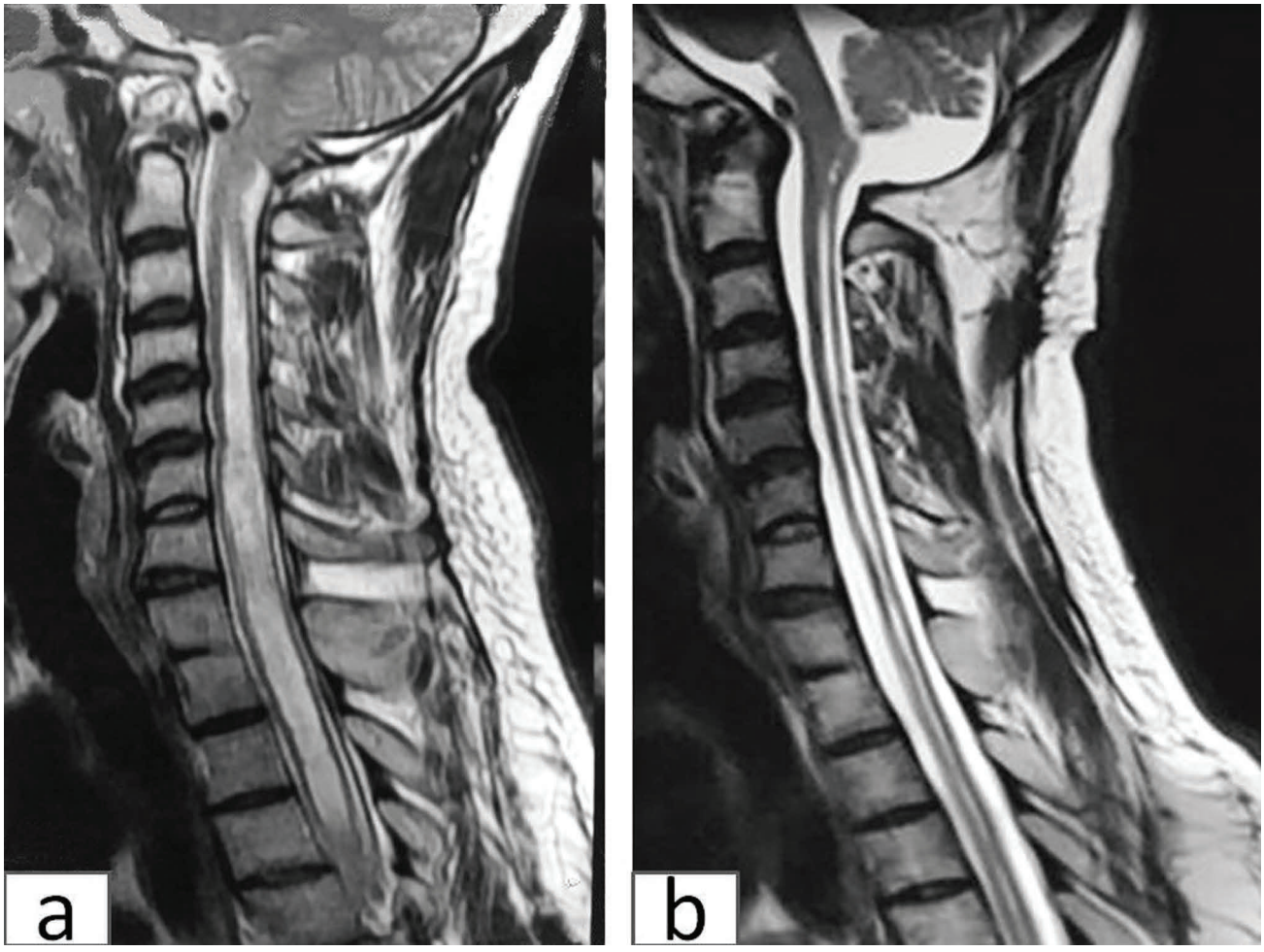

Fig. 9 Comparative MRI sagittal image of the cervical spine. **(a)** Preoperative and **(b)** follow-up at 6 months showing adequate foramen magnum decompression and continuation of craniospinal CSF space all around the cervicomedullary junction as well as marked reduction in syrinx diameter size.

to Gardner's procedure.[6] His procedure consisted of simple bony decompression and expansion of the dura with tissue graft and avoidance of opening of the arachnoid membrane and entrance into the subarachnoid space or fourth ventricle. His group performed a clinical study comparing Gardner's procedure with their procedure of simple decompression and duraplasty and demonstrated that there was no difference in syrinx resolution between the procedures, although the Gardner's operation resulted in higher complication rate.[6] Therefore, simple and least invasive craniocervical decompression and duraplasty without opening arachnoid membrane that eliminates the pathophysiologic mechanisms of syringomyelia was preferred in most of the cases in the later phase of our study.

A wide triangular pericranial graft harvested from the occipital region at the beginning of procedure was our standard institutional practice. The foramen magnum and arch of axis was exposed in the entire width of the dura, and more lateral exposure was avoided due to risk of vascular injury. It was our institutional preference to preserve muscle attachments and lamina of C2 intact to minimize postoperative pain and instability with the exception of 11 cases in which tonsillar descent was below C2. There is controversy regarding the extent of decompression required to alleviate symptoms. The importance to find answer to question "how much is enough" in our study is magnified in patients with CM type I because a significant proportion of these patients have small posterior fossa volume. Data analysis in the initial phase of this study revealed greater belief in wider foramen magnum decompression even up to extent of partial resection of medial margins of occipital condyles. The distance between the midpoints of the condyles is approximately 4.5 cm.[7] Resection of occipital condyles results in overt instability. In experimental studies when 50% of a condyle was resected, the range of motion was found to be increased by 153% during flexion extension, by 40.8% during lateral bending, and by 28.1% during axial rotation. Condylectomy was also associated with an increase in rotation at C1–C2, although not significantly when less than 75% of the condyle was resected.[8] Apart from craniovertebral instability, oversize craniectomy can result in cerebellar ptosis due to herniation of tonsils and vermis with resultant adherence to overlying graft leading again to obstruction of CSF flow and reappearance of syringomyelia. Samii and Klekamp[9] have advised that the size of the craniectomy should be limited to the width of the spinal canal and not to extend further

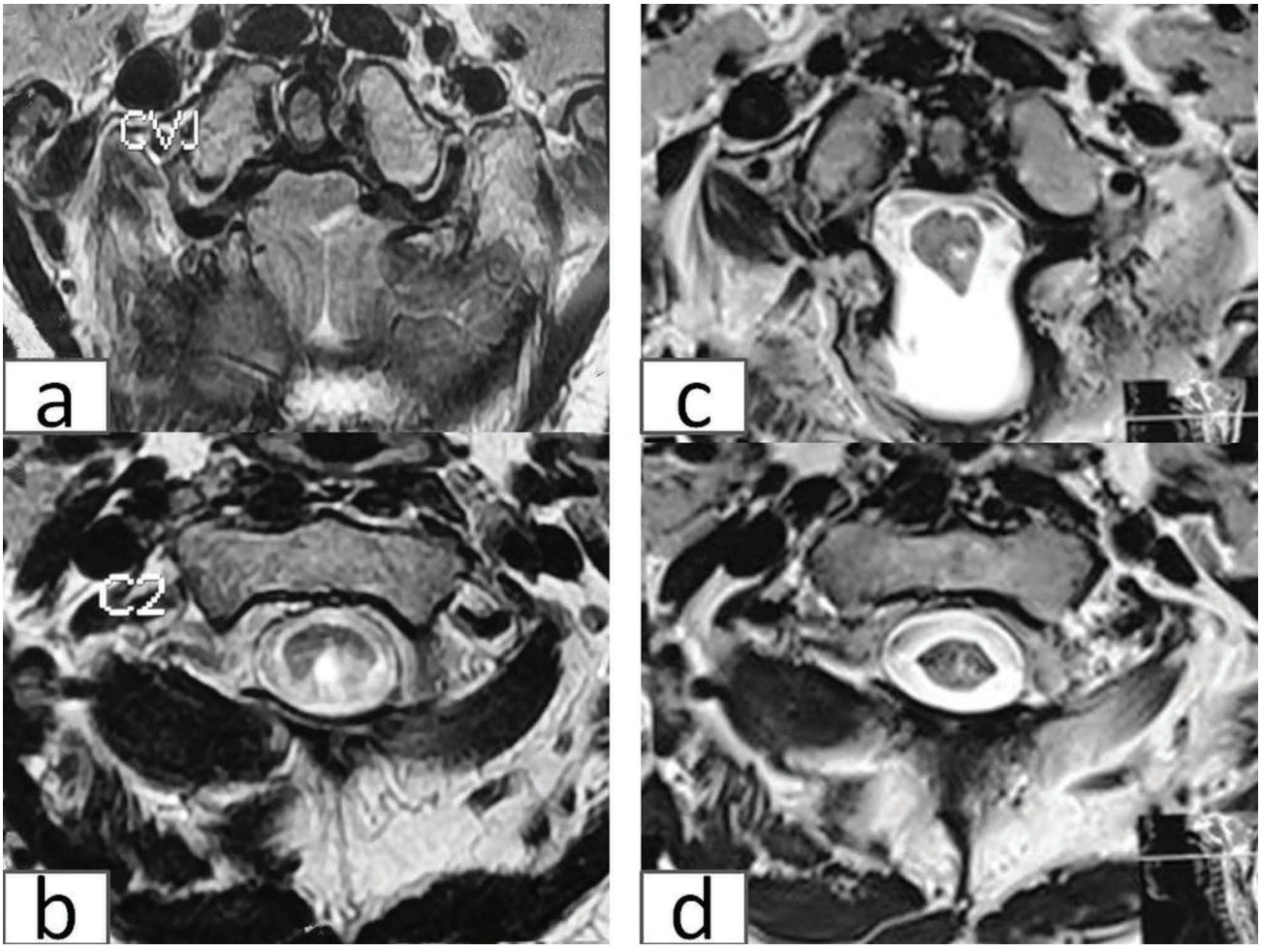

Fig. 10 Comparative MRI axial image of the cervical spine. **(a, b)** Preoperative and **(c, d)** follow-up at 6 months showing adequate foramen magnum decompression and continuation of craniospinal CSF space all around the cervicomedullary junction as well as marked reduction in syrinx diameter size.

upward than 2 cm from the rim of the foramen magnum. There was no significant difference in outcome when more conservative approach of limited removal of the bone to completely decompress the entire posterior surface of the cerebellar tonsils was advocated. In this study, associated bony anomalies were present in 20 (27%) patients in whom the occipital bone was considerably flattened with rostral tilt at the foramen magnum and assimilation of the arch of atlas. In patients with CM type I, the bones of the skull base often are underdeveloped in 25% cases, resulting in skeletal anomalies such as basilar invagination (25–50%), Klippel-Feil syndrome (5–10%), and atlanto-occipital assimilation (1–5%) that results in reduced volume of posterior fossa.[10] In all of those patients, our institutional preference was to do preoperative CT study of the craniocervical junction that was having important implications in operative strategy for bony decompression. Grabb et al have described the entity of ventral brainstem compression in pediatric and young adult patients with Chiari I malformations.[11] They have observed ventral cervicomedullary encroachment by the odontoid and its investing tissues into the rostral spinal canal resulting in flattening and distortion of the ventral brainstem in 28% of patients in their study. Significant ventral compression was defined as greater than 9 mm of reclination of the odontoid process from a line connecting the basion to the posterior aspect of the body of the axis. Their recommendation for patients with measurements of 9 mm or greater, is the reduction in ventral brainstem compression before posterior fossa decompression. In our subset of associated 25% basilar invagination, all have less than 9 mm of reclination of the odontoid process from a line connecting the basion to the posterior aspect of the body of the axis, so we have done only posterior decompression for CM with syrinx associated with basilar invagination.

Although conservative decompression was sufficient in most patients, few subsets of patients having thick dural band and significant tonsillar descent required intradural approach. Arachnoid adhesions are not directly visualized in conventional CT and MRI. However, preoperative cinewave MRI, intraoperative visualization of movements of tonsils seen through dura, and intraoperative ultrasound may help in deciding need for intradural approach. The dura was opened in midline at the C1 level with incision extended superiorly to split below foramen magnum to create Y-shaped dural opening. In all the cases in which intradural exploration was done, endpoint was visualizing

choroid plexus of the fourth ventricle and free flow of CSF into subarachnoid space. In cases of severe tonsillar ectopia, extrapial coagulation of tonsillar tip was preferable to facilitate free flow of CSF from the foramen of Magendie. Capacious duraplasty using previously harvested pericranial graft with reinforcement of suture line using fibrin glue to prevent CSF leakage was our routine institutional practice. Older procedures of intervention for syrinx without bony decompression done in earlier part of our study were discontinued due to high recurrence rate and poor outcome. Overview of the literature about craniectomy size, C1 arch removal, duraplasty, and tonsillar resection and arachnoid dissection are listed in ►**Tables 6** to **9**, respectively.[10,12–20]

Conclusion

Chiari malformation type I presents a difficult challenge to neurosurgeons due to its unpredictable clinical course and controversies regarding management. Our understanding of managing this entity is still evolving, but innovative surgical techniques for breaking barriers of CSF flow to restore normal CSF dynamics across the craniocervical junction provide the pragmatic solution. The trend is toward balance

Table 6 Literature overview related to craniectomy size in Chiari malformation type I

Author	No. of patients	Children/Adults	Extent of bone removal
Erdogan et al (2010)[10]	27	Both	Decompressive suboccipital craniectomy, at least 3 cm above foramen magnum, width of 3 cm
Gurbuz et al (2015)[12]	39	Both	Decompressive suboccipital craniectomy, at least 3 cm above foramen magnum, width of at least 4 cm
Kennedy et al (2015)[13]	156	Children	Suboccipital craniectomy
Kumar et al (2014)[14]	1	Adult	Suboccipital craniectomy
Mutchnick et al (2010)[15]	121	Both	Wide suboccipital craniectomy
Rehman et al (2015)[16]	21	Adult	Suboccipital craniectomy
Furtado et al (2011)[17]	20	Children	Midline suboccipital craniectomy, individualized according to age
Hoffman and Souweidane (2008)[18]	40	Both	Suboccipital decompression, superior extension of ~1.5–2 cm, lateral extension to the lateral most aspect of the foramen magnum and cervical spinal canal

Table 7 Literature overview related to C1 arch removal in Chiari malformation type I

Author	No. of patients	Children/Adults	C1 arch removal
Erdogan et al (2010)[10]	27	Both	Total C1 laminectomy (removal of the atlantooccipital ligament and dural scarring or bands on the dura outside)
Gurbuz et al (2015)[12]	39	Both	Total C1 laminectomy
Kennedy et al (2015)[13]	156	Children	Total C1 laminectomy and incision of the atlantooccipital ligament in all cases; additional C2 partial laminectomy in 12 cases
Kumar et al (2014)[14]	1	Adult	C1 laminectomy (3 cm wide)
Mutchnick et al (2010)[15]	121	Both	Total C1 laminectomy and careful resection of dural bands
Rehman et al (2015)[16]	21	Adult	C1 laminectomy in all cases, additional C2 laminectomy in 2 cases
Furtado et al (2011)[17]	20	Children	C1 laminectomy in all cases, additional C2 laminectomy in 1 case
Hoffman and Souweidane (2008)[18]	40	Both	Extent of cervical laminectomy determined by degree of tonsillar descent

Table 8 Literature overview related to duraplasty in Chiari malformation type I

Author	No. of patients	Children/Adults	Materials	Results
Abla et al (2010)[19]	Review of literature	Both	Autologous and nonautologous	Duraplasty essential because of creating a cisterna magna where one was not previously present; no superiority of neither autologous nor nonautologous graft; pericranium is preferred if possible
Erdogan et al (2010)[10]	*n* = 27 (15 FMD with duraplasty, 12 only FMD)	Both	Y–shaped opening and dural grafting with cadaveric dura	No statistical postoperative differences a dura opening recommended in cases of any suspicion about maintaining CSF flow in the posterior fossa
Gurbuz et al (2015)[12]	*n* = 39 (21 duraplasty, 18 nonduraplasty)	Both	n.m.	No statistically significant difference for surgical results, but in regression of postoperative syrinx size a in patients with syrinx, tonsillar herniation > 10 mm, and symptom duration < 36 mo, duraplasty considered to be a more reliable choice despite a slightly higher rate of complications
Hoffman and Souweidane (2008)[18]	*n* = 40	Both	Autologous pericranial tissue	PFD with duraplasty is safe and appropriate
Kennedy et al (2015)[13]	*n* = 156 (nonduraplasty)	Children	n.m.	Dura opening recommended for patients with rapid progression of neurologic deficits, scoliosis with syrinx, craniovertebral instability requiring fusion, and if preoperative MRI suggests that partial C2 laminectomy will be necessary
Mutchnick et al (2010)[15]	*n* = 121 (56 nonduraplasty, 64 duraplasty)	Both	Y–shaped incision extending caudal past the foramen magnum with a generous pericranial patch, covered with Tisseel	Clear benefits to most children without duraplasty, but recurrence is slightly higher than with duraplasty
Rehman et al (2015)[16]	*n* = 21	Adults	n.n.	PFD with duraplasty best treatment option
Furtado et al (2011)[17]	*n* = 20	Children	With pericranium or artificial dura	PDF with duraplasty preferred

Abbreviations: CSF, cerebrospinal fluid; FMD, foramen magnum decompression; MRI, magnetic resonance imaging; n.m., not mentioned; PFD, posterior fossa decompression.

Table 9 Literature overview related to tonsillar resection and arachnoid dissection in Chiari malformation type I

Author	No. of patients	Children/Adults	Intervention done
Erdogan et al (2010)[10]	27	Both	Opening of thick arachnoid layers and resection of thick arachnoid bands between the tonsils can be necessary to obtain CSF passage
Furtado et al (2011)[17]	20	Children	Dense subarachnoid bands were released, and tonsils were shrunk with bipolar cautery until free egress of CSF was seen from the foramen of Magendie
Guyotat et al (1998)[20]	75	Both	Better outcome in patients treated by PFD and additional tonsil resection

Abbreviations: CSF, cerebrospinal fluid; PFD, posterior fossa decompression.

between maximum decompression and preserving normal integrity.

Informed Consent

Informed consent was obtained from all individual participants included in this study.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names or initials will not be published and due effort will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of Interest

There are no conflicts of interest.

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