

Original Article

Managing Chiari I Malformations; 5 Year Experience

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Abstract

Introduction: Chiari I malformations present with variety of symptoms. There are four types of Chiari malformations described in literature. There are two more types of Chiari introduced later; Chiari 0 and Chiari 1.5. Routine use of MRI has led to frequent identification of the Chiari malformation. Management of Chiari malformation has evolved with time. Asymptomatic patients can be managed conservatively with regular clinical and radiological follow up. Surgical management includes decompression of foramen magnum. This creates a space so that circulation of CSF is unimpeded. We analyzed patients who underwent surgical management at our centre in last 5 years.

Methods: Retrospective analysis of all patients operated in Neurosurgery department of TUTH (Tribhuvan University Teaching Hospital) from 2012 July to 2017 July was done. Data was collected from hospital records and follow up was taken from outpatient department. Demography, associated syringomyelia, improvement in clinical symptoms and radiological improvement were analyzed. Minimum follow up of 1 year was included.

Results: A total of 37 patients were available for analysis. Male to female ratio was 15:22. Major presentation was occipital headache sensory impairment and atrophy of upper limb due to syringomyelia. Clinical and radiological improvement was seen in all patients except 1 patient who developed post operative pneumonia and succumbed. Morbidity in the form of wound infection in 2, Pseudo meningocele was present in 4 patients and CSF leak in 3 patients. Resolution of syrinx was observed in 34 patients except in 3 patients who required a syringopleural shunt in follow up.

Conclusion: Management of Chiari I malformation is relatively safe. In this era of diagnosis of more and more asymptomatic patients, clinical judgement is essential.

Keywords: Chiari I Malformation, Decompression, Syringomyelia, Syringopleural shunt.

Introduction

Hans Chiari in 1895 described 4 types of Chiari malformations. Contributions in literature were available from Arnold, Cleland and Nicholas Tulp as well.

	Diagnosis
Chiari I	Tonsillar herniation below foramen magnum rim > 5mm Associated hydrocephalus Associated syringomyelia
Chiari II	Herniation of tonsils plus brain stem and 4 th ventricle Always with myelomeningocele May have hydrocephalus May have syrinx
Chiari III	Herniation of cerebellum and brainstem into occipital encephalocele (to be distinguished from high occipital encephalocele) Poor prognosis
Chiari IV	No herniation Cerebellar aplasia/hypoplasia Posterior fossa cyst
Chiari 0	No herniation Syringomyelia improving with posterior fossa decompression
Chiari 1.5	Herniation of tonsils plus some part of brainstem

The estimated prevalence in the United States of type I CMs is less than one percent with a slight female predominance. ¹Type I CMs can be seen incidentally in approximately 1% to 4% of patients undergoing brain or cervical spine magnetic MRI studies. ²Most cases of Chiari I are congenital. Various hypothesis like small posterior fossa due to development abnormality in paraxial mesoderm has been stated. Although the exact etiology is unknown, this condition is thought to be secondary to insufficiency of the paraxial mesoderm after neural tube closure with underdevelopment of the occipital somites.

Morphometric analysis revealed reductions in the posterior cranial size and volume. ³Skull base abnormalities are seen in approximately 50% of type I CM cases, (i.e., basilar invagination, retroflexed odontoid, platybasia etc.)⁴

According to Goel, the pathogenesis of CM with or without associated basilar invagination and/or syringomyelia is primarily related to atlantoaxial instability and that the surgical treatment in these cases should be directed toward atlantoaxial stabilization and segmental arthrodesis.⁵

Acquired type I CMs can occur when there is a significant cerebral spinal fluid (CSF) pressure gradient across the craniocervical junction, i.e., CSF leakage or lumboperitoneal shunts can produce negative downward pressure gradients leading to the development of a type I CM. In addition, conditions associated with raised intracranial pressure, such as hydrocephalus and Idiopathic Intracranial Hypertension can promote downward pressure gradient.

Syringomyelia is identified in 30-85% of patients. (Figure 1,2,3) There are many hydrodynamic theories to explain the formation of syringomyelia.⁶

Abnormal and increased pulsatile motion of the cerebellar tonsils “tonsillar pistoning” can produce selective obstruction of CSF flow during systole. The increased systolic CSF waves are then transmitted to the spinal subarachnoid space and drive the CSF into the central canal of the spinal cord through engorged perivascular and interstitial spaces and lead to syrinx formation.^{7,8}



Figure 1 Sagittal T 1 W image showing tonsillar herniation below C1 posterior arch and associated T1 low signals inside the cervical cord suggestive of Syrinx

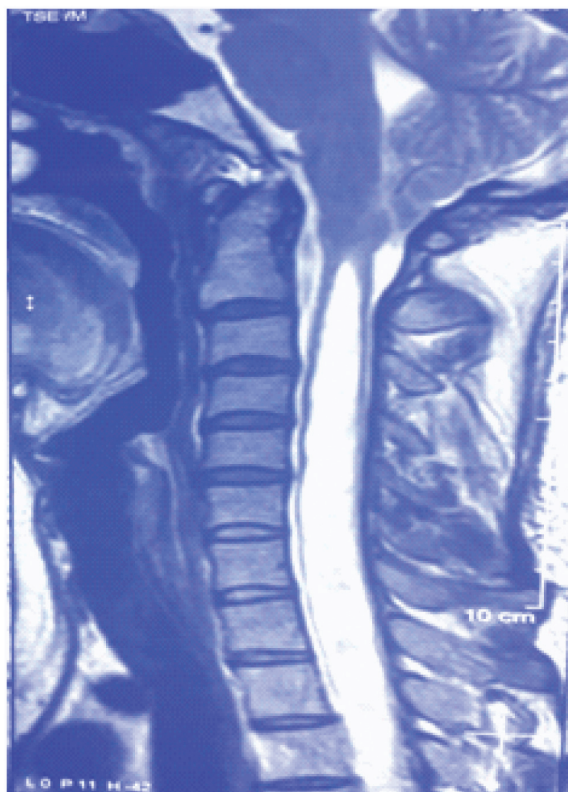


Figure 2 Sagittal T2W image showing tonsillar herniation and T2 high signal in cervical cord suggestive of syrinx



Figure 3 Post op T1 W image showing marked decrease in cord swelling and syrinx as compared to pre op.

Clinical presentation

Chiari I malformation can present with various symptoms. Occipital headache and neck pain are the most common symptoms in adults. Patients often present with headaches aggravated by “Valsalva maneuvers” during coughing and sneezing or strain. Symptoms are based on the structural and functional (impaired “CSF-dynamics”) pathology associated with CM1, which often leads to a wide spectrum of focal and non-focal findings in the clinical and neurological presentation, making it difficult to diagnose.

MRI of the brain with craniocervical junction generally detects tonsillar descent. Mc Rae’s line (basion-opisthion line) is generally taken as standard. The shape of the tonsil (peg-like) may be more important than the descent alone. The presence of hydrocephalus and other lesions causing secondary Chiari has to be ruled out before any surgical intervention. In situations where there is confusion regarding CSF flow around the foramen magnum, CINE MRI will be useful. (figure 4) This can also be useful in follow up to see if we have achieved adequate decompression. Other associated radiologic anomalies occur infrequently and include most commonly atlanto-occipital assimilation, platybasia, basilar invagination, and fused cervical vertebrae. Despite the less frequent occurrence, these changes should be sought because they can lead to cervical instability. “Complex Chiari” malformations (Figure 5, 6) may be defined as cerebellar tonsil herniation combined with one or more of the following radiographic findings: brainstem herniation through the foramen magnum (Chiari 1.5 malformation), medullary kink, retroflexed odontoid, abnormal clival-cervical angle, occipitalization of the atlas, basilar invagination.⁹

Some authors are of the opinion that each patient with Chiari should get a flexion extension x ray of CVJ to rule out instability. If Chiari is detected, then we need to screen the spine to check for associated syrinx formation.

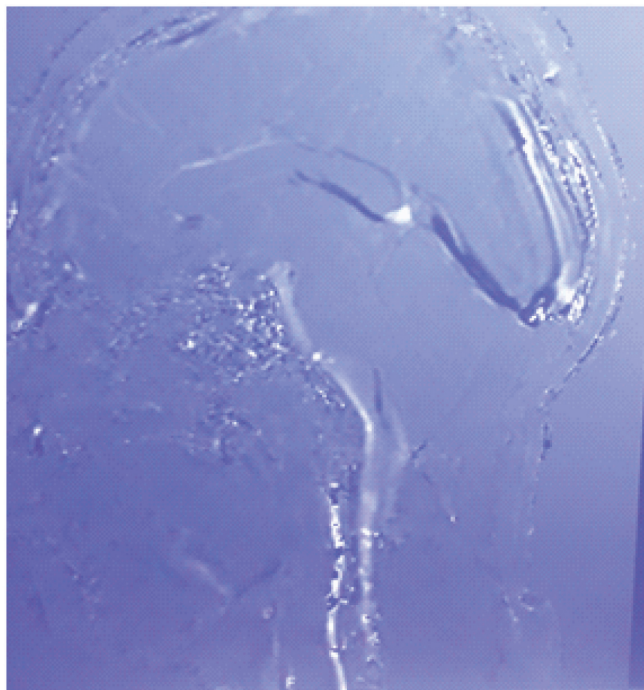


Figure 4 CINE MRI showing CSF flow across the foramen magnum suggestive of adequate decompression

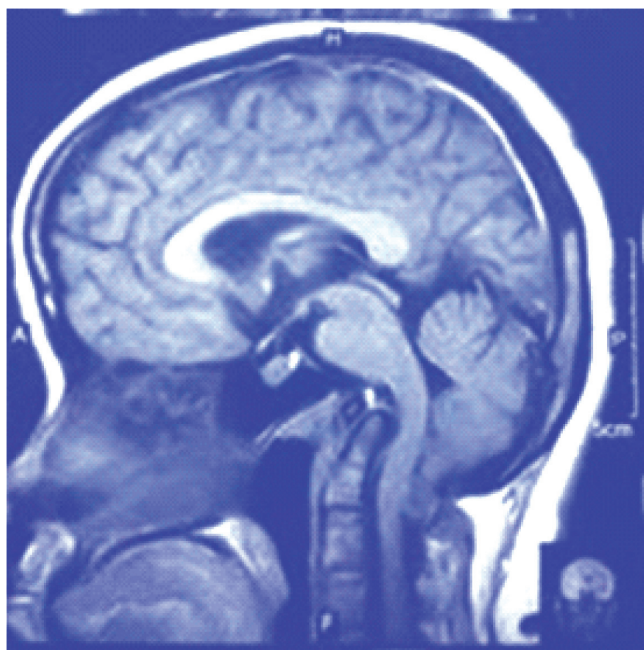


Figure 5 Complex Chiari showing medullary kinking and retroflexed odontoid

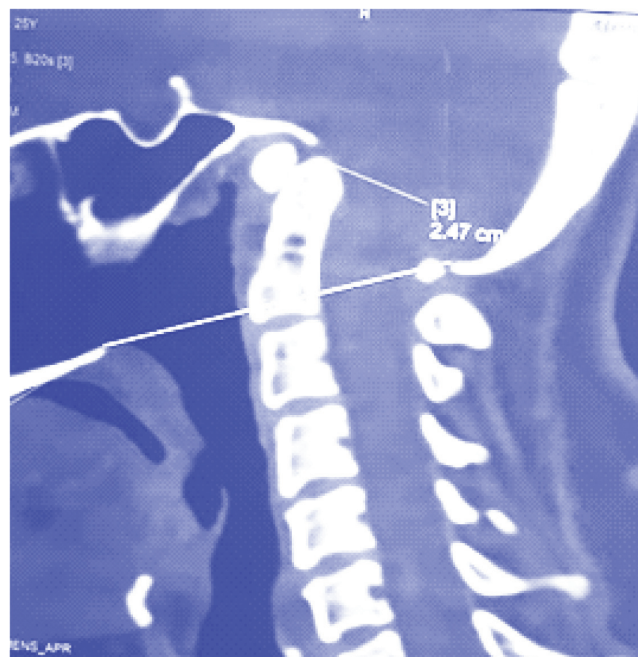


Figure 6 Note the clival agenesis and odontoid well above the Chamberlain Line (hard palate – opisthion line) - Complex Chiari

Management: Symptomatic Chiari with or without syrinx formation generally undergo surgical intervention. However, in asymptomatic patients with Chiari and syrinx, Nishizawa et al observed and found that only few patients needed intervention in follow up and concluded that small, asymptomatic syringes can be safely followed with serial examinations and imaging.¹⁰

Adequate decompression of occipital bone and posterior arch of C1 is generally required. This can be added variably with outer dural layer opening, duroplasty, tonsillar coagulation and opening the obex to see free CSF flow. Recently, interest has been increasing in including ultrasound in observing tonsillar movement after bony removal and avoiding dural opening if tonsillar movements are normal.

Methods: Retrospective analysis of all patients operated in Neurosurgery department of TUTH (Tribhuvan University Teaching Hospital) from 2012 July to 2017 July was done. Data was collected from hospital records and follow up was taken from outpatient department. Demography, associated syringomyelia, improvement in clinical symptoms and radiological improvement were analyzed. Minimum follow up of 1 year was included.

Management protocol

We operate on symptomatic patients and all patients with Chiari associated with a syrinx. Any patient with asymptomatic Chiari is kept under regular follow up with regular MRI. Most of our surgical strategy is focused on enlarging the foramen magnum and maintaining CSF flow across the foramen Magendie. No procedure for syrinx is done at the time of foramen magnum decompression. We repeat MRI with CSF flow study at 1 year. If there is persistent sizable symptomatic syrinx despite an adequate space in region of foramen magnum, we go for syringopleural shunt.

Results

There were 40 patients who underwent surgery for Chiari I in the time period. At least 1 year follow up was available for 37 patients. There were 15 male patients and 22 female patients.

Out of 37 patients available for analysis, fifteen were male and 22 patients were female. The youngest patient operated was 15 years of age and eldest was 65 years with mean age of 32 years.

Symptoms	Frequency
Occipital headache	28
Tingling/numbness	25
Atrophy	10
Weakness	10
Intractable hiccups	1
Nystagmus	1
Scoliosis	5

Major presentation was occipital headache and tingling and paresthesia of the extremities. Clinical and radiological improvement was seen in all patients except 1 patient who developed post operative pneumonia and succumbed. Morbidity in the form of wound infection in 2 and Pseudo meningocele was present in 4 patients. CSF leak occurred in 3 which was managed successfully with lumbar drain. Resolution of syrinx was observed in many patients except in 3 patients who required a syringopleural shunt in follow up.

Surgical technique	Number of patients
FMD+ duroplasty+tonsillar coagulation	10
FMD+duroplasty	25
FMD+duraplasty+syringopleural shunt	2

Complications	Frequency
Pseudomeningocele	4
CSf Leak	3
Wound infection	2
Pneumonia	1

Radiological presentation

Focal cervical syrinx	5
Cervicodorsal syrinx	18
Holocord syrinx	5
Scoliosis	5
CVJ bony abnormality	3

Neurological recovery was seen in all patients. Residual numbness was present in almost 50% patients. Syrinx did not resolve in 3 patients who underwent Syringopleural shunt after 2 years. Patients with complex Chiari who underwent decompression only are under regular follow up and have no clinical or radiological deterioration.

Discussion

We had a predominance of female in our series of patients. The average female: male ratio has been 1.3 to 1.7 in various studies. One reason for this predominance might be that women, particularly in the adult population, present more commonly with headache syndrome, which in turn may prompt more frequent MRI diagnostics. Meadows et al. questioned whether the female predominance is the result of more frequent initial cerebellar ectopia or a faster progression of symptoms later in life.^{11,12,13}

The mean age of presentation in our series was 32 years. In a large analysis, the mean and median ages in the pediatric patient population were both 8 years, whereas in the adult population, they were 41 years. The mean and median ages in the combined studies were 33 and 35 years, respectively.¹⁴

As a clinical entity, the Chiari type I malformation is well known for its highly variable clinical presentation

and pernicious onset. Patients may have only minor symptoms for months or years. Headache is an almost invariable complaint. Although several discrete types of headache occur in patients with Chiari type I malformation, ranging from short “cough- induced” to continuous headaches, there are no discerning characteristics unique to Chiari malformations, nor is there any obvious relationship between such pain and the extent of tonsillar herniation or the presence of syringohydromyelia.¹⁵

Most of our patients (28/37) had headache as the initial symptom which was more on the occipital region and Valsalva induced. Other complaints of pain are often described as diffuse, most frequently in the cervical region and upper extremities. One of our patient presented with unusual symptom of intractable hiccups.¹⁶

A common complication of Chiari malformation is syringomyelia, which occurs in 37% to 75% of Chiari patients.¹⁷ Symptoms of syringomyelia may be present in addition to, or overshadow those of, the Chiari malformation but are almost invariably an ominous prognostic sign.¹⁸ 28(75%) patients in our series had syringomyelia. Most of the syrinx was in cervicodorsal region and 5 (13%) had holocord syrinx.

Similarly, both sensation deficits and weakness are more common in the upper extremities than in the lower extremities. Almost 54% (20/37) patients had limb weakness out of which half of them already had atrophy of hand muscles. (Figure 7)



Figure 7 Left thenar muscle atrophy in a patient with Chiari I malformation and syrinx

In their reviews of pediatric Chiari I publications, Durham and Fjeld-Olenecand Hankinson et al. reported that posterior fossa decompression and dural opening with duroplasty are associated with a lower risk of reoperation but a greater risk of CSF-related complications.^{19,20}

As summarized by Batzdorf the main goal of surgery is: resolving cranio spinal pressure dissociation, restoring subarachnoid spaces and the cisterna magna in the posterior cranial fossa, eliminating and reducing the syrinx, relieving compression of the brainstem, and relieving or eliminating symptoms and signs of CM-I.²¹ Posterior fossa decompression, involving suboccipitalcraniectomy and upper cervical laminectomy (generally posterior arch of C1), is the most frequently performed procedure. Further surgical technique remains highly variable and is usually dictated by patient characteristics and surgeon experience. The dura is usually incised in a standard Y pattern, although some authors have suggested dural opening may be unnecessary.²² Bands and arachnoid adhesions may be freed, although several authors have stressed the importance of avoiding undue manipulation of the subarachnoid space. Manipulation of the tonsils has become a matter of considerable debate. Whereas several authors have suggested some form of manipulation, ranging from simple retraction to bipolar cauterization or resection study has shown any additional benefit to such methods.²³

Our protocol is to involve tonsillar coagulation only in large syrinx and if the tonsils are at or below the level of second cervical vertebrae. We performed tonsillar coagulation in 10/37 patients. In our patients we removed foramen magnum rim and posterior arch of C1 in all patients. Dural opening with duraplasty using pericranial /fascia was performed in all patients.

Opening the dural and arachnoid is bound to increase some chances of CSF leak and pseudo meningocele as seen in our cases (7/37). The utilization of Ultrasound to check for movement of tonsils can obviate the need of dural opening in selective patients as seen in some studies.²⁴

Early surgical techniques attempted to address Chiari and syrinx together, with a decompression at the foramen magnum and shunting procedures for the syringomyelia.

However, many surgeons including our team now prefer to reserve shunting for patients who fail to respond to initial posterior fossa decompression. This strategy has proven effective in many studies, but syrinx cavities managed in this manner may take more time to respond than those treated with techniques focused directly at the syrinx (e.g., shunting procedures).

Outcome: Headache specifically and pain in general appear to respond best. If the patient already has atrophy of muscles, the response is not encouraging as might be expected with loss of alpha motor neurons. Weakness in the absence of atrophy tends to respond well. Scoliosis almost certainly because of its relation to syringomyelia, seems to respond reasonably well to surgery. In contrast, sensory loss has been widely recognized as largely unresponsive to surgery.

Conclusion

Posterior fossa decompression is relatively safe and effective for management of patients with symptomatic Chiari I malformations. Prompt surgical action needs to be undertaken before the patient develops atrophy of muscles. For patients with associated syringomyelia, it seems prudent to deal with the Chiari first and follow the syrinx radiologically. Any syrinx not improving with adequate posterior fossa decompression needs syringopleural/peritoneal shunt. Recent techniques like C1 C2 fixation for management seem to be an overkill and need validation. Asymptomatic patients need regular follow up.

Conflict of Interest: None to declare

Reference

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