

Original Article (SPINE)

## Chiari Malformation with and without Syringomyelia: Surgical Technique and Outcome in 88 Adult Patients

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### ABSTRACT

**Objective:** This study identified the relationship between posterior fossa craniectomy, expansion neuroplasty, and radiological appearances in patients with Chiari malformation with and without clinical syringomyelia with the surgical outcomes in an attempt to correct the lesion.

**Materials & Methods:** Eighty-eight patients with Chiari malformation (CM) were included in the study where 70 had associated syringomyelia. All underwent posterior fossa craniotomy, expansion duroplasty without fiddling with cerebellar tonsils. Patients were evaluated at 1 month, 3 months, and 12 months. The MRI studies were done at 12 months when symptomatic relief and radiological findings were evaluated and matched.

**Results:** Most of the patients were young adults between the age range of 25 – 40 years. The most common complication was pseudomeningocele (5.68%) formation followed by CSF leak (4.54%). Patients with a longer history of Chiari malformation or syrinx-related symptoms and signs had partial relief in symptoms and signs. The poor outcome as expected was seen in patients with atrophic changes in upper limbs and hypertonia in lower limbs, especially in patients with loss of joints position sense and poor balance. Patients showed maximum improvement in headaches both suboccipital as well as generalized. Syringomyelia was decreased in size in 49 patients and remained unchanged in 21. Dysesthesias were improved in 31 patients.

**Conclusion:** Clinical improvement was related to the expansion of the posterior fossa and subarachnoid cistern and reduction in the size of the syrinx. Surgical decompression of the posterior fossa should create adequate space for its contents and reduce the syrinx cavity. The relationship between symptomatic improvement and radiological findings is not always linear.

**Keywords:** Chiari Malformation, Tonsillar Herniation, Syringomyelia, Duroplasty.

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## INTRODUCTION

Chiari malformations are structural defects in the skull base and cerebellum. It is further subdivided into four main types (I, II, III & IV) These malformations frequently occur in combination with other pathological conditions such as myelomeningocele, hydrocephalus, and/or Syringomyelia. Hindbrain inferior migration occurs in Chiari Malformation reported by Van Houweninge Grafrdijk 1932<sup>1</sup> and Penfield & Coburn 1938.<sup>2</sup> Chiari, I, characterized by one or both pointed (not rounded) cerebellar tonsils that project 5mm below the foramen magnum, measured by a line drawn from the basion to the opisthion (McRae Line).<sup>2</sup> Chiari II consists of brainstem herniation and a towering cerebellum in addition to the herniated cerebellar tonsils and vermis due to an open distal spinal dysraphism/myelomeningocele.<sup>3</sup> Chiari III involves herniation of the hindbrain (cerebellum with or without the brainstem) into a low occipital or high cervical meningocele.<sup>3</sup> Chiari IV is now considered obsolete.<sup>4</sup> Neurological signs and symptoms can arise from 2 mechanisms: direct compression of neurological structures against the surrounding foramen magnum and spinal canal or the development of Syringomyelia or syringobulbia. The obstruction of cerebrospinal fluid (CSF) outflow eventually results in syrinx formation.

Fluid-filled cavities (syrinx) develop within the spinal cord or brainstem, resulting in neurologic symptoms as the cavity expands. This condition is more and more diagnosed in the adult population since the easy availability of MRI, in patients with symptoms of headache and neckache. Lichtenstein in 1943<sup>3</sup> discovered the associated syringomyelia with Chiari Malformation. Different surgical techniques have evolved over the years, but still, there is no uniformity among the neurosurgeons claiming good results.<sup>4</sup> Common to all techniques is an expansion of posterior fossa and foramen magnum allowing free flow of CSF from Cranial to spinal subarachnoid spaces<sup>5-9</sup> which gives

symptomatic improvement. A linear relationship between clinical improvement and radiological change in the postoperative periods has not been established in any published study.<sup>4</sup> This study was carried out to correlate the clinical outcome of post fossa decompression and duroplasty with radiological improvement and to establish the relationship of clinical status with radiography.

## MATERIAL & METHODS

### Study Design & Setting

This prospective study was carried out between January 2002 to Dec. 2018. Eighty-eight patients were included in this study including 68 males and 20 females. The sample was selected through the nonprobability sampling technique. This study was carried out at multiple institutions including Lahore General Hospital, Services Hospital Lahore, Surgimed Hospital & Mayo Hospital Lahore. All patients had posterior fossa decompressive craniectomy and duroplasty. This study was conducted to evaluate the effects of this procedure on clinical symptoms and signs, post-operative MRI appearance, and its relation to the improvement of symptoms and signs.

### Inclusion Criteria

All patients of both genders between the age limit of 25 – 55 years of age with MRI-diagnosed Arnold Chiari malformation with or without syringomyelia were included in this study.

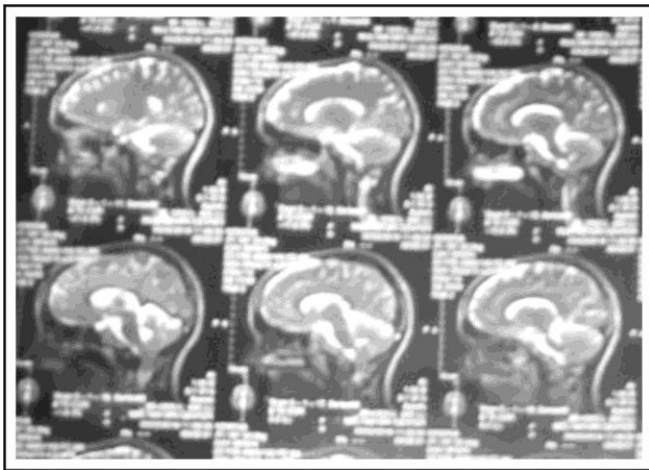
### Exclusion Criteria

Redo cases & those unfit for general anesthesia were excluded from the study.

### Data Collection

Eighty-eight patients were included in this study with a 68 males and 20 females (Ratio 3.4:1). Seventy patients had associated syringomyelia and 18 had only Chiari Malformation. Diagnosis

of the patient was based on clinical and radiological criteria. Postoperatively patients were examined at 1, 3, and 12 months, only if there were no new symptoms and signs which were compared with the checklist of symptoms and signs prepared at the time of admission. The follow-up period was one year after which patients were discharged to be followed up in out-patients when they had any complaints.



**Figure 1:** A 23 years old female presented with ataxia, basal cranial nerve involvement & nystagmus. MRI showed herniation of cerebellar tonsils below the C1 arch.

### Surgical Technique

All Patients underwent posterior fossa decompressive craniectomy, excision of the posterior rim of the foramen magnum, and expansion duroplasty with pericranial or fascia lata graft. In patients where Cerebellar tonsils were descending up to C2, laminectomy of C1 and partial laminectomy of C2 were done keeping the major muscular attachments to the spinous process of C2 vertebrae intact.

Occipital Craniectomy was done to the extent that Cerebellar Hemisphere was pulsating with the heartbeat, visible through the dura mater. Then dura mater was opened in "Y" shaped fashion so that cerebellum is not tight, pulsating the brain, and CSF is found moving across the foramen magnum. No Fiddling was done with

cerebellar tonsils, and arachnoid band, but it was made sure that no obstruction was present at the Craniocervical junction so that there was a free flow of CSF.

Pericranium or fascia lata was used for expanding the dura mater to give more space for the cerebellum and attached to a Y-shaped incised dura mater. Bipolar diathermy was avoided at the dural edges to prevent avascularity and shrinkage which could hamper water-tight dural closure. Duroplasty was done with 4/0 vicryl and watertight closure was confirmed with Valsalva maneuver. The graft and suture line was covered with surgical and 2 ml of fresh blood was sprinkled to have a punctuation of sealing effect.

### RESULTS

#### Age Distribution

Most of the patients were young adults between the age range of 25 –40 years.

**Table 1:** Age Distribution.

Age	Frequency	Percentage
25 – 40 Years	81	92
41 – 55 Years	7	8

#### Gender Distribution

In this study 68 (77%) were females and 20 (33) males.

#### Post-Operative Complications

The most common complication was pseudomeningocele (5.68%) formation followed by CSF leak (4.54%) (Table 2).

#### Table 3: Clinical outcome with improvement

It was noted that pulsatility in the cisterns is the key to having symptomatic & clinical impairment. Patients with a longer history of Chiari malformation or syrinx-related symptoms and

signs had partial relief in symptoms and signs. The poor outcome as expected was seen in patients with atrophic changes in upper limbs and hypertonia in lower limbs, especially in patients with loss of joints position sense and poor balance. Patients showed maximum improvement in headaches both suboccipital as well as generalized. The second most commonly improved symptom was balanced (**Table 3**).

**Table 2:** Post Operative Complications.

Complications	Frequency	% age
Pseudomeningocele.	5	5.68
CSF Leakage	4	4.54
Occipital Neuralgia	3	3.40
Superficial wound infection	3	3.40
Worsening of motor function	1	1.14
Dysphagia	1	1.14
Pneumonia	1	1.14
Total	18	20.45

**Table 3:** Clinical outcome of cases with improvement.

Symptoms	1 Month n (%)	3 Months n (%)	12 Months n (%)
<b>Symptoms</b>			
1 Suboccipital headache	79 (90%)	70 (80%)	70 (80%)
Generalized headache	75 (85%)	75 (85%)	75 (85%)
<b>Tiredness</b>			
2 Upper Extremity	35 (40%)	35 (40%)	35 (40%)
Lower Extremity	35 (40%)	35 (40%)	35 (40%)
3 Dysarthria	53 (60%)	53 (60%)	53 (60%)
<b>Signs</b>			
1 Balance	62 (70%)	66 (75%)	62 (70%)
2 Nystagmus	53 (60%)	57 (65%)	57 (65%)
3 Fascial Sensory deficit	35 (40%)	35 (40%)	35 (40%)
4 Impaired gag	35 (40%)	40 (45%)	40 (45%)
5 Numbness of Limbs	35 (40%)	35 (40%)	35 (40%)
6 Tone	18 (20%)	18 (20%)	18 (20%)
7 Weakness	44 (50%)	44 (50%)	44 (50%)
8 Atrophy	0	0	0

### Syringomyelia Size and Its Effect on The Clinical Outcome

In our study, 70 patients had syringomyelia on pre-operative MRI. Syringomyelia was decreased in size in 49 patients and remained unchanged in

21. Dysesthesias were improved in 31 (63%) patients (**Table 4**). There existed a significant difference (p-value: 0.0430) between outcomes: improved, unchanged, and worsened.

**Table 4:** Syringomyelia Size and its effect on the clinical outcome

Syringomyelia Size (70 Patients)	Improved Dysesthesia & Numbness n (%)	Unchanged Dysesthesias & Numbness n (%)	Worsened n (%)	Chi-Square	P value
Reduced (49)	31 (63%)	13 (27%)	5 (10%)	6.29	0.0430 (significant result)
Unchanged (21)	7 (33%)	12 (57%)	2 (10%)		

## DISCUSSION

Old field et al<sup>8</sup> pointed out that the shape & descent of tonsils may change once the decompression procedure is performed adequately so the term malformation may not be true. The development of the posterior fossa when the hind brain is normal in size leads to over grading of its contents and descent of the cerebellar tonsils.<sup>9</sup> Heiss et al<sup>10</sup> demonstrated that an increase in pulse pressure in the subarachnoid space of the cervical region leads to the formation of syringomyelia & its progression. Poor absorption of extracellular fluid in the cord may be an important factor in syrinx formation.<sup>11</sup>

The majority of patients have settled with decompression craniectomy and duroplasty without fiddling with the cerebellar tonsils.<sup>12,5</sup> Breaking the arachnoid band, adhesiolysis, Coagulating or resecting the tonsils leaves necrotic tissue which can increase the risk of infection and new adhesion formation<sup>10,7</sup>. Bentson et al<sup>10</sup> has reported that breaking of arachnid band & coagulation or resection of tonsils is mandatory for good results but we never resected tonsils. We have always been vigilant for the free pulsatile flow of CSF from the cervical cavity to the spinal canal this keeps the tonsils away from the cord, cervical roots, and brain stem.<sup>13</sup>

In the study symptoms and signs of dysarthria, dysesthesia, poor gag, and swallowing and balance were commonly found. Obese ladies have more complaints of headache & exertional symptoms in their extremities. Patients with atrophic changes in upper limbs also showed an inability to do daily work and had poor results. In this group of patients, the clinical presentation was similar to that reported in other papers.<sup>6,14-17</sup>

Postoperatively headache improved in most of the patient's balance and dysarthria also improved in a good number of patients (60 – 70%) dysesthesia, tone and weakness did not show improvement more than 40 – 50% and

atrophy has shown no improvement. With time from 1 month to 12 months, some further deterioration was noticed which was not picked up on MRI. All patients had some improvement after the surgical procedure but significant improvement in symptoms and signs was observed in patients in whom syrinx was reduced in size. Reduction in syrinx size and improvement in clinical outcome was not found related to the size of subarachnoid space on MRI which was quite contrary to the findings reported by John R Bentson et al.<sup>4</sup> We have done a procedure to restore the free and pulsatile flow of CSF across the Craniocervical junction<sup>7,8,10,18</sup> this gives adequate space for the tonsils to stay away from the brain stem, upper cervical cord, and roots and does not merit any fiddling with cerebellar tonsils.

## CONCLUSION

Chiari Malformation should be diagnosed early to treat early for better outcomes. The optimal surgical procedure is decompressive occipital craniectomy, widening of the foramen magnum, and expansile duroplasty of adequate size tailored for the patients as per radiological findings. The relationship between symptomatic improvement and radiological findings is not always linear.

## Conflict of interest

Nothing to disclose.

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## Additional Information

**Disclosures:** Authors report on conflict of interest.

**Ethical Review Board Approval:** The study was confirmed to the ethical review board requirements.

**Human Subject:** Consent was obtained by all patients/participants in this study.

**Conflict of Interest:**

In compliance with the ICMJE uniform disclosures form, all authors declare the following:

**Financial Relationships:** all authors have declared that they have no financial relationship at present or within the previous three years with any organizations that might have an interest in the submitted work.

**Other Relationships:** all authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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### **AUTHORS CONTRIBUTIONS**

<b>Sr.#</b>	<b>Author's Full Name</b>	<b>Intellectual Contribution to Paper in Terms of:</b>
1.	Muhammad Irfan	1. Study design and methodology.
2.	Samra Majeed	2. Paper writing and data calculations.
3.	Samra Majeed, Muhammad Irfan	3. Data collection and calculations.
4.	Muhammad Hammad Nasir, Jahangir Khan	4. Analysis of data and interpretation of results etc.
5.	Ajmal khan, Azam Niaz	5. Literature review and referencing.
6.	Anjum Habib Vohra	6. Analysis of data and quality insurer.