

# CRANIOCERVICAL DECOMPRESSION-A PROCEDURE OF CHOICE FOR SYRINGOMYELIA WITH ASSOCIATED CHIARI MALFORMATION SHOWING GOOD LONG TERM FOLLOW UP RESULTS

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## ABSTRACT

**Objective:** To determine the immediate and late outcome of craniocervical decompression in patients having syringomyelia (SM) associated with Chiari malformation (CM). Study design: An interventional study. Place and duration: Lahore General Hospital, Lahore. From July, 2011 to December, 2014.

**Methodology:** All patients presenting with sign and symptoms of syringomyelia (SM) and having radiological evidence of syrinx associated with Chiari malformation (CM) were included. Their short term clinical and the long term radiological improvement were assessed post-operatively, 1 month, 3 months, 6months and 1 year.

**Results:** We analysed laminoplasty results in 30 patients who had syringomyelia along with Chiari malformation in our department and who were operated in the period of July, 2011 to December 2014. In all the patients we performed C1 and eventually C2 laminectomy to achieve proper craniocervical decompression. Cerebellar tonsils were coagulated and the dura was left open. We concluded that there was gross improvement in patients having signs and symptoms due to Chiari malformation. Patients presenting with symptoms of syringomyelia improved as well but the reflexes in the upper limbs of such patients remained diminished. In our study there were improvement of 60% to 90% in the sign and symptoms by this procedure.

**Key words:** syringomyelia, Chiari malformation, surgical treatment.

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## INTRODUCTION

Syringomyelia is a disorder of formation and enlargement of fluid filled cavities within the spinal cord. Ollivier d'Angers in 1827 used the syringomyelia term to describe pathological dilatation of the central canal, after the Greek word<sup>1</sup>. Stillings in 1859, used hydromyelia for central canal dilatation<sup>2</sup>. There are three theories of pathogenesis, 'Gardner water hammer theory' in which an obstruction at the foramen of Magendie produces this process<sup>3</sup>. William's theory in which difference between ICP and spinal pressure causes a valve like action at foramen magnum, the malformation of the hindbrain prevents the high CSF pressure from caudally dispersing. This pressure gradient between the brain and spinal cord draws CSF caudally into the syrinx<sup>4</sup>. Oldfield's theory proposed that when cerebellar tonsils move caudally during systole, they build a piston effect in

subarachnoid space of spinal cord<sup>5</sup>. In 1891, Hans von Chiari described three rhombencephalon anomalies which were labelled as Chiari's description of cerebellar malformations that were accompanied with syringomyelia<sup>6</sup>. In the Chiari type I malformation, 57-65% of patients develop syringomyelia while, in the Chiari type II malformation, 20-90% of patients usually develop syringomyelia<sup>6</sup>. The first clinical relationship of this was made by Portal in 1804 who observed that the sensory loss and paralysis of the limbs were progressive. In 1882 Schultze explained the classic dissociated sensory syndrome of loss of pain and temperature with sparing of proprioception and light touch. Symptomatic progression is usually gradual<sup>7</sup>.

## METHODOLOGY

This study was conducted in the Department of Neurosurgery Unit II, Lahore General Hospital from period of July, 2011 to December, 2014. Careful detailed history and clinical examination revealed that all the patients had sign and symptoms of syringomyelia and radiological evidence of CM and syrinx formation. The sign and symptoms included headache, dysesthesia, subjective weakness, sensory loss, oscillopsia, paresthesia and dysphagia which persisted for more than 6 months. On MRI T1 and T2 weighted images there was CM having descent of the cerebellar tonsils 3 to 6 mm below foramen magnum associated with syringomyelia. After an informed consent and anesthetic evaluation all the patients were subjected to craniocervical decompression with leaving the dura open.

### Surgical technique:

We performed this surgery under general anesthesia, patients were placed in prone position in 3 pin Mayfield head fixator. The position of the head was kept elevated at 15 degrees and care taken of pressure points by padding. Proper head shaved from occiput to the upper portion of the neck was performed. Linear incision was made about 7 cm above theinion to the cervical area downwards based on the inferior position of the cerebellar tonsils. In this incision pericranium was also opened. For proper exposure and later closure, the occipital scalp was undermined. After opening the fascia suboccipital craniectomy was performed, followed by laminectomy of C1 and C2 based on the intradural pathology and extent of tonsillar descent. The dura was opened in Y- shaped fashion and temporarily sutured avoiding damage to the occipital sinus. With

the help of a microscope the arachnoid was opened and visible adhesions were excised. The pericranium was sutured water tight with 4-0 prolene. The integrity of the closure was established by Valsalva manoeuvre and any areas of leakage were repaired. Wound was closed in layers restoring normal anatomy. No drain was used.

## RESULTS

In our study of 30 patients, 21 were males and only 9 were females. All patients presented in the age range of 18 to 58 years (mean of 32 years). All patients had at least 6 months of symptoms and showed radiological evidence of syringomyelia associated with CM on MRI scans.

In early outcome mortality was nil, the post-operative course was smooth and unremarkable. In all our patient there was a slight rise in temperature of about 1000 F for the first 24 hours. 11 patients had post-operative nausea and headache which required administration of analgesics and anti-emetics for 48 hours. Symptoms of pain and paraesthesia improved post operatively and there were improvement progressively with physiotherapy in the first 1 to 2 months but somehow the 13 patients presenting with hyporeflexia of the upper arms didn't have improvement in this symptom.

Follow up MRI were performed in all patients at 3 to 6 months after surgery in all the patients and repeated in 3 patients after 1 year. In all cases MRI showed syrinx collapsed in 3-12 months follow up with persistent shrinkage with longer at longer MRI studies and increase in CSF space surrounding the medulla and cerebellar tonsils and spinal cord. Cerebellar tonsil peg like shape also transformed to a round normal architecture in most of the patients.

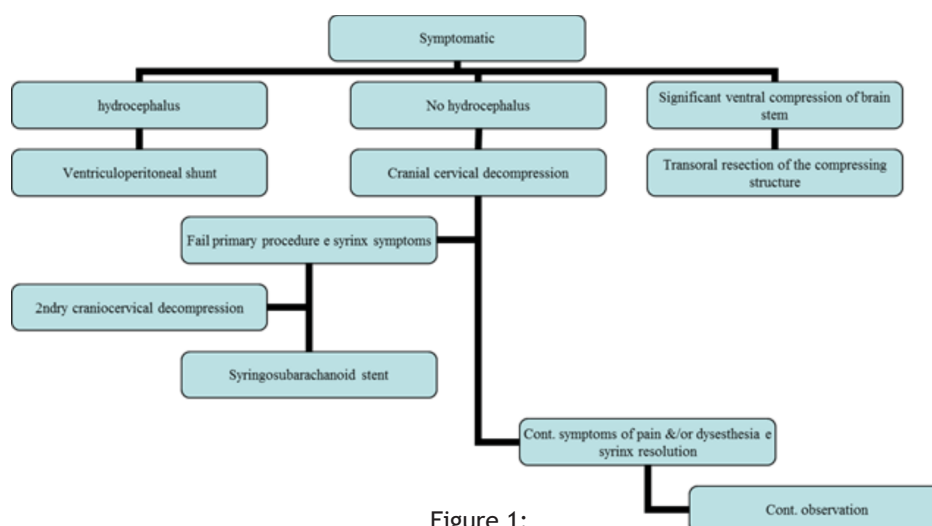


Figure 1:

Figure 2: MRI craniocervical junction. A, B. sagittal section T1 and T2 images showing cerebellar decent and syringomyelia. C. showing CSF with in the cord- Syringomyelia.

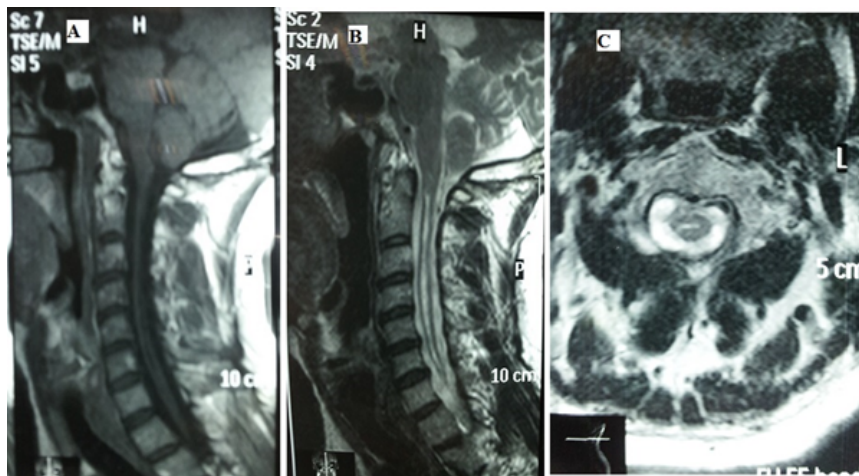


Figure 3:

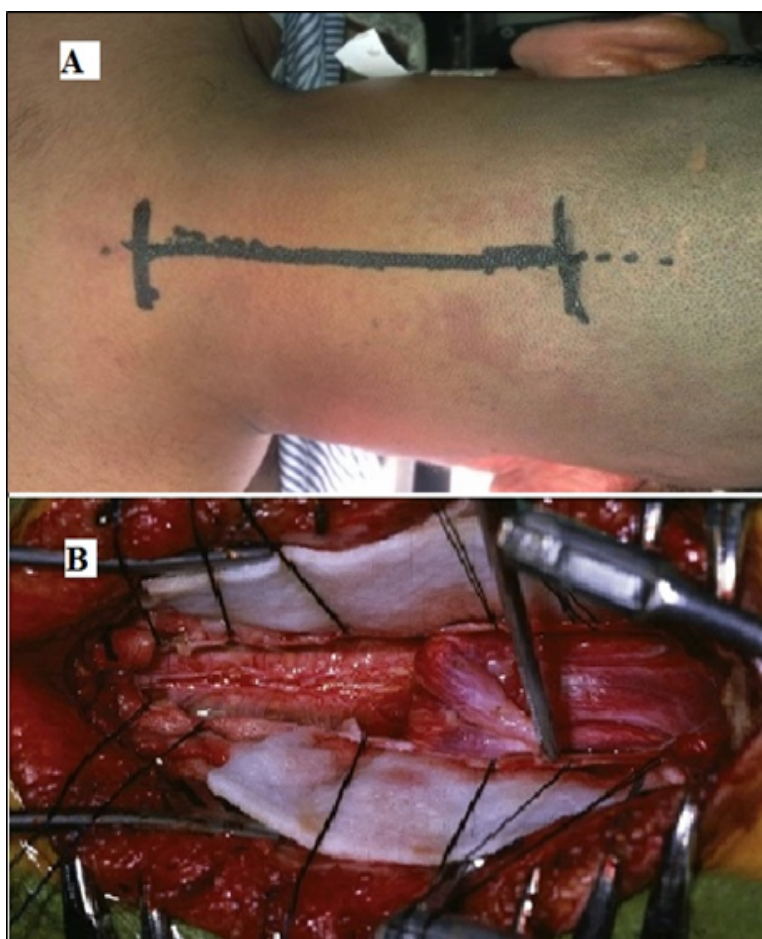


Figure 4:

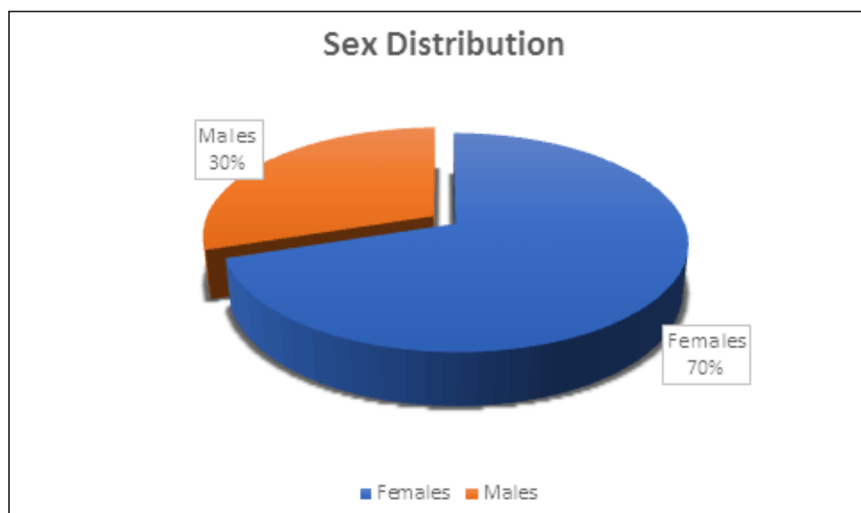


Table-1: Early improvement in sign and symptoms of SM and CM Post operatively

Sign and symptoms	No. of Patients	
	24 hours after surgery	48 hours after surgery
Headache	11	11
Nausea	11	11
Fever	19	0
Paraesthesia	30	15
Hyporeflexia	13	13
Mortality	0	0

Table-2: Late Outcome in terms of Sign and symptoms

Sign and symptoms	No. of Patients	
	3 months after surgery	6 months after surgery
Headache	0	11
Nausea	0	11
Fever	0	0
Paraesthesia	4	1
Hyporeflexia	13	12
Mortality	0	0



Figure-5: A, Post Op. MRI Sagittal view T1weighted image and B, T2 weighted image showing regression of CM and SM.

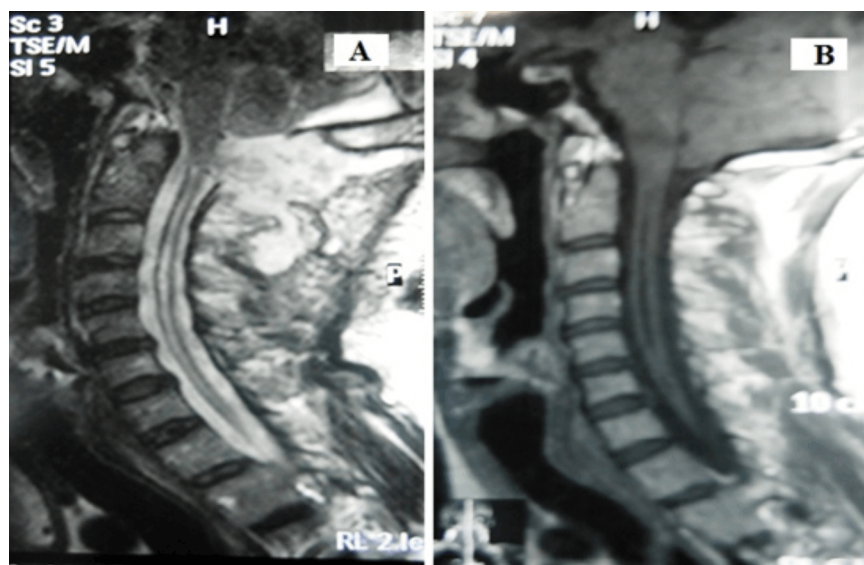


Figure-6: A,B. Post-operative picture of the patient having 04/05 power in upper limbs with hyporeflexia. C, D. Power lower limb 05/05 with normal reflexes.



## DISCUSSION

Different surgical techniques are used by neurosurgeons to treat syringomyelia associated with Chiari malformations. Recently a new technique of decompression is becoming more popular in providing relief from the clinical signs, symptoms and radiologically decreasing the syrinx size<sup>8</sup>. Although it is debatable whether the arachnoid should be opened or not for at the craniocervical junction which can improve the CSF flow. In our study we performed posterior fossa decompression, splitting of the arachnoid along with coagulating of the cerebellar tonsils just to make them retract a bit without causing necrosis. However, in our study, some of the cases required extra bone removal for proper visualization and lyses of arachnoidal adhesions. Even with extra bone removal and dura open there were no complication which was also shown by a study conducted by Ellenbogen RG & Zeidman SM in 2000, they recommended that craniocervical decompression technique even if extra bone removal is required, it should be performed for proper arachnoid lysis. With their technique there had been no CSF leak or infection like meningitis or any pseudomeningocele formation, similarly, no patient developed a new neurologic deficit with a relatively brief follow-up<sup>9</sup>.

In a study conducted by Farhad. M in Oregon during period of 2000 to 2002, patients who underwent dura-splitting craniocervical decompression returned home significantly sooner than patients treated using duraplasty and suffered no complications of surgery. Dura-splitting decompression for pediatric Chiari I malformation is better tolerated by patients and avoids exposure to CSF-related complications compared with duraplasty<sup>10</sup>.

In CM with syringomyelia, suboccipital posterior fossa decompression and atlas laminectomy is the customary surgical method in almost all of symptomatic patients. In a study by Depreitere B in 2000 revealed that 76% of patients had an improvement in the early follow-up period, while 68% in the later follow-up improved, whereas 26% experienced deterioration. Postoperative MR images showed a favourable result in 80%, consisting of syrinx collapse or reduction of syrinx. Numerous clinical trials have reported 95%-97% of improvement in preoperative symptoms of their patients<sup>9,10,11,12</sup>. In our patients we followed them for almost 2 years and their neurological status which improved with laminoplasty remained maintained. The size of their syrinx got diminished and recurrence rate was zero. Many of the neurosurgeons prefer to put a

pericranium patch graft for closing the dura<sup>8,9</sup>. But we had modified this technique by leaving the dura open and after achieving the proper hemostasis, closing the wound layer by layer to prevent CSF leak. This procedure, however, has some benefits first, this technique is relatively easy, secondly, it needs a single incision, with no need of fascia lata or pericranium graft and thirdly it avoids the occipital sinus. This technique has very good clinical and radiologic outcomes, more over the rate of complications are minimal.

Craniocervical decompression is first line of treatment for symptomatic syringomyelia with Chiari malformation. The results of Craniocervical decompression with lysing of arachnoid and leaving the dura open are encouraging in short term and more in long term follow up.

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