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## **A case report on result of posterior fossa decompression on syringomyelia in a case of chiary type I malformation**

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**Abstract:** The prime objective in the surgical treatment of Chiari malformation (CM) and/or syringomyelia (SM) is based on the restoration of the normal cerebrospinal fluid (CSF) dynamics at the craniovertebral junction through the creation of a large artificial cisterna magna. In this case a patient came to our hospital with type 1 chiary malformation having large syrinx which underwent posterior fossa decompression by midline sub occipital craniectomy with subpial cerebellar tonsillar resection which after one year of follow up we have found significant resolution of syrinx radiologically.

**Key words:** Syringomyelia SM, Chiary malformation type 1 CMI, Foramen magnum decompression FMD

### **Introduction**

The pathophysiology of syrinx formation and progression is important for surgeons in their selection of a surgical approach for syringomyelia (SM) secondary to Chiari malformation type I (CMI). There is broad consensus on the role of mechanical blockage of cerebrospinal fluid flow at the craniocervical junction in the pathogenesis of SM secondary to CMI [1, 2]. Posterior fossa decompression (PFD) is currently preferred by most surgeons as the standard surgical option in the treatment of SM secondary to CMI [3–6]. This results in decompression of the cerebellar tonsil restores the normal cerebrospinal fluid flow [7]. In addition, a wide variety of surgical procedures as adjuvants to standard PFD, including

syringosubarachnoid shunting, obex plugging and resection of the cerebellar tonsil, have been advocated in previous studies, which potentially induce bias with respect to the evaluation of syrinx resolution. To date, few reports have quantitatively evaluated syrinx resolution after PFD or identified predictive factors for better improvement. In addition to occipital pain and vertigo, dissociative sensory loss is one of the most frequent clinical presentations in children and adolescents with CMI. In this case we have done posterior fossa decompression by midline sub occipital craniectomy with subpial cerebellar tonsil resection in a patient of chiary malformation type 1 with syringomyelia then after one year of follow up there was resolution of syrinx was found radiologically.

## Case report

A 45 year old female presented to our neurosurgery opd with the chief complaints of occipital headache since two years, weakness bilateral upper limb since six months, difficulty in holding objects since six months on examination there was dissociative sensory loss as decrease in pain and temperature sensations up to C7 dermatome with preservation of pressure and joint position sense, power was -4/5 on bilateral upper limb with bilateral hand grip weakness, reflexes were exaggerated on both upper and lower limbs and planter reflex was extensor. On MRI cervical spine showed there was chiary type 1 malformation with large syrinx over crevico dorsal region (Figure 1).

Patient was planned for routine surgery and posterior fossa decompression by midline suboccipital craniectomy with subpial resection of cerebellar tonsil and decompression of the cervical cord was done in sitting position. After one year of follow up now patient have relieved occipital headache with no sensory loss but grip of both hand was weak till now, on MRI cervical spine there was significant reduction in the syrinx was found (Figure 2).

## Radiographic evaluation

The severity of cerebellar tonsillar descent and features of the syrinx were evaluated via measurement of the following indices on the T1-weighted sagittal MRI: the distance of cerebellar tonsillar descent (mm), the configuration of the syrinx, the maximal syrinx/cord (S/C) ratio and length of the

syrinx. The extent of cerebellar tonsillar descent was further classified into three grades [14]: grade I, in which the tonsil descended beyond the foramen magnum but did not reach the C-1 arch; grade II, in which the tonsil reached the C-1 arch; and grade III, in which the tonsil descended beyond the C-1 arch. Syrinx configuration was categorized into distended, moniliform, slender, and circumscribed types [14]. The maximal anteroposterior diameter of the syrinx (S) divided by the anteroposterior diameter of the spinal cord (C) at the same level provided the maximal S/C ratio [15]. Syrinx length was defined as the number of vertebral segments spanned by the syrinx. Syrinx resolution was defined as any demonstrable decrease in maximal S/C ratio or length. Any postoperative decrease in the maximal S/C ratio or syrinx length divided by the preoperative value was calculated as a radiographic improvement rate. Significant improvement of syrinx was defined as any more than 20% decrease in maximal S/C ratio or length on follow-up MRI, and complete resolution was used to describe the syrinx disappearing after PFD.

In our case preop parameters of patient was grade II tonsillar herniation was found, syrinx was moniliformed, syrinx length was 13 vertebra, S/C ratio was 0.86, after operation and 1 year of follow up parameters were no cerebellar tonsils herniation syrinx was slender with length of 13 vertebra, S/C ratio was 0.46. So there was significant improvement in syrinx with radiographic improvement rate was 46.5%.



Pre op MRI



Post op MRI after 1 year follow up

## Discussion

The outcome of PFD for the treatment of SM secondary to CMI has varied widely in previous studies in terms of maximal S/C ratio and syrinx length. Hida et al. [8] reported that 30 of 33 patients with SM secondary to CMI exhibited obvious syrinx resolution after PFD. Depreitere et al. [16] reported that 80% of patients (16/20) showed a favorable result consisting of complete syrinx resolution in 8 patients and obvious syrinx reduction in another 8. Wetjen et al. [12] documented the time course of syrinx resolution after PFD and found that the syrinx resolved significantly over a short period of time after decompression. Significant improvement was also reported in 7 of 7 patients in a study by Feldstein and Choudhri [17] and 11 of 12 in a study by Ghanem et al. [18].

However, low rates of significant improvement have also been reported. In studies by Caldarelli and colleagues and McGirt and colleagues, syrinx resolution after PFD was found only in 50 and 62% of patients, respectively [10,19]. This discordance may be related to broad age range, different surgical procedures, or variability in the definition of significant improvement. In general, pediatric patients have a better response to PFD than adults.

We found significant syrinx improvement after PFD in patients with CMI, particularly within the initial several months after surgery.

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