Patients with syringohydromyelia without hindbrain herniation that responded to posterior fossa decompression have been described (so-called "Chiari zero malformation"). Conversely, 14% of patients with tonsillar herniation > 5 mm are asymptomatic (the average extent of ectopia in this group was 11.4 ± 4.86 mm).

In 1998, a group led by Dr. Jerry Oakes [1] identified five patients with syringomyelia and no evidence of tonsillar herniation (i.e. no Chiari I malformation). MR imaging of the entire neuraxis ruled out other causes of a syrinx (e.g. tumor, post-traumatic). Ultimately, abnormal CSF flow at the posterior fossa or foramen magnum was identified as the suspected cause. The label "Chiari 0" was used to categorize these patients by virtue of the fact that they behave as though the afflicted patients have fourth ventricular outlet obstruction - and at surgery, frequently do have physical barriers to CSF movement but do not have caudal displacement of the cerebellar tonsils beyond a point that could be considered pathological. All underwent a posterior fossa decompression and duraplasty identical to the technique used to treat patients with a Chiari I malformation. No attempt was made to directly treat the syrinx. Significant syrinx and symptom resolution was observed in all patients. Interestingly, two patients who did not respond to previous shunting of their syrinx experienced long-term improvement only after the posterior fossa decompression. Their response to surgery suggests "Chiari-like" pathophysiology is present in the absence of tonsillar herniation. This contention is supported by the identification of a crowded foramen magnum in two patients, multiple arachnoid adhesions in two others and a 4th ventricular arachnoid veil in one. Each of these findings can detrimentally alter CSF flow. In this study, cine-MRI failed to accurately identify patients most likely to benefit from surgery.

Morphological studies of the posterior fossa in the Chiari 0 group have found the sagittal anteroposterior diameter of the foramen magnum to be greater than controls and inferior descent of the obex (i.e. caudal displacement of the brainstem) was found to be three standard deviations below normal [2]. These findings suggest caudal displacement of the medulla and compensatory enlargement of the foramen magnum may result from a small posterior fossa.

More recently, Kyoshima et al. [3] have reported four patients with what they termed a "tight cistern magna" and syringomyelia. These patients did not have hindbrain hernias and each underwent posterior fossa decompression with improvement of the syrinx in three. We would consider these patients as examples of the Chiari 0 malformation.

It is emphasized that this subgroup represents a very small cohort found within the Chiari malformations. In addition, the original authors considered the diagnosis of Chiari 0 only in patients with syringomyelia of no apparent cause. Careful patient selection is critical when making the diagnosis of Chiari 0 malformation. Without an obvious Chiari I malformation, other etiologies of a spinal syrinx must be conclusively ruled out. Only then can one reasonably expect to ameliorate the clinical course of these patients with a posterior fossa decompression.

1)
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