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The Chiari disease and cervical syringomyelia: can transoral decompression be the ethiological therapy?

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Hindbrain herniation syndrome, or Chiari malformation Type I (CM-I), occurs frequently with craniovertebral junction (CVJ) abnormalities when there is reduction in the posterior fossa volume. Syringomyelia is often present Chiari malformations are often associated with spinal deformities, including scoliosis. Studies have suggested a causal relation between syringomyelia and scoliosis. Posterior fossa dorsal decompression (PFDD) is typically performed but has adverse results when ventral bone abnormality exists. Patients who fail to respond to standard surgical management often have complex anomalies of the craniovertebraljunction and brainstem compression, requiring reduction and occipitocervical fusion. According to Goel, patients with basilar invagination can be categorized into two groups based on the presence (Group A) or absence (Group B) of clinical and radiological evidence of instability of the CVJ. Standard radiological parameters described by Chamberlain can be used to assess the instability of theCVJ. The pathogenesis and clinical features in patients with Group A basilar invagination appeared to be related to mechanical instability, whereas it appeared to be secondary to embryonic dysgenesis in patients with Group B basilar invagination. In Group II, on the other hand, the assembly of the odontoid process, anterior arch of the atlas and the clivus migrated superiorly in unison resulting in reduction of the posterior cranial fossa volume, which was the primary pathology in these patients. The Chiari malformation or herniation of the cerebellar tonsil is considered to be a result of reduction in the posterior cranial fossa volume. Two paediatric patients harbouring CVJ dysembriogenetic pattern (Basilar invagination type B, Chiari disease with and without syringomyelia underwent our observation.

Pat #1 13 yrs harbouring Chiari, syringomyelia and basilar impression type and pat #2 6 yrs harbouring Chiari and basilar impression Type 2 underwent double staged anterior transoral decompression and posterior instrumentation and fusion without direct Chiari and syrinx decompression. Chiari disease disappeared at discharge and syringomyelia wihtin 6 months.

In our experience neurological improvement, Chiari and syringomyelia resolution can occur using only ventral cervicomedullary junction decompression in patients with basilar invagination and basilar impression. This is likely due to the relief of neural encroachment and reestablishment of CSF pathways and represent the ethiologial therapy.