A critical analysis of the Chiari 1.5 malformation.

OBJECT: Although the traditional nomenclature used to describe hindbrain hernias is useful, there are certainly patients in whom these morphological entities do not strictly apply. One such group is composed of patients with the more recently described Chiari 1.5 malformation in which a Chiari I malformation is seen in combination with brain-stem herniation through the foramen magnum. In an attempt to elucidate further the best surgical strategy and to refine the descriptive terminology, the authors retrospectively analyzed all cases at their institutions in which this form of hindbrain herniation was diagnosed.

METHODS: The authors reviewed the database for all patients in whom Chiari 1.5 malformation had been diagnosed. Each patient had undergone a posterior fossa decompressive surgery. Magnetic resonance images were evaluated for the extent of caudal descent of the brainstem, amount of tonsillar ectopia, inclination of the odontoid process, and any brain or brainstem abnormalities. Clinical presentations and postoperative results were correlated to the aforementioned radiological findings. Twenty-two patients were identified. The obex was a mean 14.4 mm inferior to the foramen magnum, and the medulla exhibited a flattened appearance in the midsagittal plane in all patients. Syringomyelia was documented in 50% of the cases. The cerebellar tonsils were found to lie at C-1 and C-2 in nine and 13 patients, respectively. The mean angulation of the odontoid process was 84.4 degrees. No abnormalities or caudal descent of the midbrain or pons was identified. Eighteen patients experienced resolution of preoperative
symptoms. Persistence of syringomyelia prompted a second posterior fossa operation secondary to progressive scoliosis in 13.6% of the patients.

**CONCLUSIONS:** No single sign or symptom was found to be peculiar to the Chiari 1.5 malformation, although all patients in whom this diagnosis was established had undergone a posterior fossa decompressive surgery. A significant number (13.6%) of patients required repeated operation for persistent syringomyelia. Neurosurgeons may wish to consider that many patients may not respond as well to posterior fossa decompressive surgery especially if syringomyelia is present.

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