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Clinical treatment and diagnostic criteria for Chiari I and Chiari 0 malformation in pediatric patients

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Introduction: Chiari I and Chiari 0 malformations often have clinical and radiological overlap. We performed this study for better insight in clinical and radiological criteria for classifying two groups of patients.

Methods: This study includes all patients with diagnosed Chiari I or Chiari 0, treated at our clinic from 2002 – 2013. The diagnosis was established according to clinical and MR criteria. Patients with tonsillar herniation less than 5 millimeters below the line basion-opisthion were diagnosed as Chiari 0, and those with 5 or more millimeters as Chiari I. Patients with Chiari II malformation (n=21) were excluded from this study.

Results: In our series of 89 pediatric patients with Chiari malformation (age ranging from 7 months to 18 years), 45 Chiari I and 23 Chiari 0 patients were diagnosed. Among them, 27 Chiari I and 12 Chiari 0 patients were symptomatic. A total of 23 operations in 19 patients were performed, among them 2 in Chiari 0 patients. In 4 Chiari I patients operated with bone only decompression, a reoperation with expansile duraplasty was required. In some patients with disturbed anatomy of the craniocervical junction, there was a diagnostic difficulty to classify them properly in absence of precise neuroradiologic criteria for the point of opisthion when thick occipital bone is present.

Conclusion: Clinically asymptomatic patients with Chiari I and Chiari 0 malformation have to be carefully followed by clinical and MR examinations. At operation, we suggest to perform expansile duraplasty, regardless the age of the patient or degree of tonsillar descent, especially if tonsils are peg like due to heavy compression. The "5-millimeter criteria" for neuroradiological diagnosis of Chiari has to be reevaluated, in order to avoid some patients from being underdiagnosed and untreated.