

included short pons, short midbrain/long pons with large cerebellum, and thick, short medulla. *Segmental hypoplasia* involving the pons in 59 patients was associated with microcephaly in 34. *Postsegmentation anomalies* included midbrain enlargement, enlarged quadrigeminal plates, midline clefts, 33 with congenital muscular dystrophies and O-glycosylation defects (10 with Walker-Warburg syndrome, 7 with muscle-eye-brain disease, and 2 with Fukuyama CMD), and 19 with Joubert's syndrome and the characteristic molar tooth malformation. *Associated cortical organization abnormalities* included polymicrogyria and cerebellar hypoplasia with pontine hypoplasia in 11 patients. Disorders involving the cranial nerves usually had no brainstem abnormalities on imaging other than hypoplasia of the affected nerves. (Barkovich AJ, Millen KJ, Dobyns WB. A developmental classification of malformations of the brainstem. **Ann Neurol** Dec 2007;62:625-639). (Respond: Dr Barkovich, Neuroradiology Room L371, University of California at San Francisco, 505 Parnassus Avenue, San Francisco, CA 94143).

COMMENT. Brainstem malformations appear to be more common than generally recognized. This study and proposed classification should alert neurologists and radiologists to the diagnosis of congenital malformation of the brainstem in infants and children with nonprogressive cranial nerve and long tract signs. We can look forward to an anticipated separate account of cerebellar malformations from the same institutions. Intrauterine ischemic atrophy rather than a primary developmental malformation is suggested in some reports of brainstem lesions presenting with congenital apnea and failure of central respiratory drive (Cortez C, Kinney HC. **J Neuropathol Exp Neurol** 1996;55:841-849; Reviewed by Sarnat HB. Recent advances in congenital malformations. In: **Progress in Pediatric Neurology III**, Chicago, PNB Publ, 1997;365-369).

ACUTE BRAINSTEM SYMPTOMS WITH CHIARI TYPE 1 MALFORMATION

Two children who presented with rapidly worsening neurological symptoms attributable to a previously undiagnosed Chiari malformation Type 1 are reported from Children's Hospital, Birmingham, AL. One patient became hypopneic and dysphagic and developed a right hemiparesis in less than a 48-hour period. Another patient presented with a rapidly worsening right hemiparesis, ataxia, and anisocoria. MRI revealed the Chiari 1 in both patients, and a syrinx was also identified in the second patient. Following surgical posterior fossa decompression, symptoms immediately improved. (Wellons JC III, Tubbs S, Bui CJ, Grabb PA, Oakes WJ. Urgent surgical intervention in pediatric patients with Chiari malformation Type 1. Report of two cases. **J Neurosurg: Pediatrics** 2007;107(1). (Respond: Dr W Jerry Oakes, Division of Neurosurgery, Section of Pediatric Neurosurgery, Children's Hospital, Birmingham, AL).

COMMENT. Acute presentation of Chiari malformation Type 1 (CM-1) is rare, especially in children. Chiari 1 should be included in the differential diagnosis of acute onset of brainstem or long tract signs. In a study of CM-1 at the Children's Hospital, Birmingham, UK, abnormalities of the skull base were identified by MRI measurements, indicative of a mesodermal defect. (Sgouros S et al. **J Neurosurg (3 Suppl Pediatrics)** 2007;107:188-192).