Hindbrain herniation syndrome

The hindbrain herniation syndrome, often referred to as the Chiari I malformation, is a disorder that traditionally has been defined as downward herniation of the cerebellar tonsils through the foramen magnum more than 4–5 mm on sagittal magnetic resonance imaging (MRI).

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).

Herniation of the hindbrain occurs when the lowest parts of the cerebellum and sometimes part of the medulla are moved downwards through the foramen magnum, a pressure difference acting across the foramen magnum moulding the tissues into a plug. It is suggested that the clinical course in both adults and babies with spina bifida may be explained by the hindbrain hernia acting as a valve.The term 'Chiari Type I deformity' is commonly used for an abnormality in which the tonsils and lowermost parts of the cerebellar hemispheres are prolapsed through a normal foramen magnum. Acute herniation may occur as a result of space-occupying lesions. Chronic herniation may be morphologically identical although it tends to be more severe. Sometimes it will produce few symptoms which often may be delayed so that the original causative lesion may not be apparent. Causes include bone softening, tumour, or previous meningitis. Birth injury is probably the commonest cause of the deformity, which presents clinically in adults.In infants with severe forms of spina bifida a hindbrain herniation is present. This abnormality may be called 'Chiari Type II deformity' or Arnold-Chiari deformity and is an intra-uterine abnormality in which the fourth ventricle and medulla are grotesquely herniated before they are properly developed and the foramen magnum is enlarged.The commonest clinical presentation of Chiari Type I deformity is syringomyelia, which is usually not diagnosed until adult life. Other presentations include syringobulbia, headache, oscillopsia, attacks of giddiness, lower cranial nerve palsies, and ataxia. Particularly characteristic are cough headache and cough syncope. Syringomyelia and syringobulbia in particular may be irreversible by the time they are diagnosed. Nevertheless, surgical decompression may be successful in relieving symptoms of headache, cough syncope, and long-tract compression; most cases of syringomyelia show some improvement and in others progression of the disease is arrested. Operative techniques for hindbrain herniation are discussed.Chiari Type II deformity is probably responsible for the progression of hydrocephalus after birth in the majority of babies with spina bifida. Measurement of pressure in the cerebrospinal fluid above and below the foramen magnum shows that intermittent pressure difference is commonly present at times of neurological deterioration. Surgical decompression of the hernia in adults allows correction of the valvular effect, which may be monitored by pressure measurements. In babies the associated hydrocephalus is usually so gross that it requires separate treatment, but pressure monitoring may be of value in assessing the state of the disease [1)].

Treatment

Today, the problem seems not to be in evaluating the anatomical extent of the caudal herniation of the cerebellum, but in determining which patient should be considered for operative intervention and the extent of the surgery. Chiari I patients are presenting at younger ages, occasionally with irritability as their only symptom. Should all of these children be submitted to an operation? Chiari type 2 malformation patients are now operated on with the first detectable symptom or evidence of a syrinx, and yet medullary dysfunction from the Chiari II malformation remains the leading cause of death in treated myelomeningoceles today. Our knowledge of the natural history of the untreated conditions and the increased safety of the operation has made surgical intervention a much more viable option for this group of patients [2)].

Case series

The Management of Myelomeningocele Study (MOMS) demonstrated that fetal myelomeningocele closure results in improved hydrocephalus and hindbrain herniation when compared to postnatal closure.

Flanders et al. from the Children's Hospital of Philadelphia reported on the outcomes of a single institution's experience in the post-MOMS era, with regard to hydrocephalus absence and hindbrain herniation resolution.

Primary outcomes included cerebrospinal fluid (CSF) diversion status and hindbrain herniation resolution. Families were contacted via telephone for outcome information if care was transitioned to outside institutions. Univariate/multivariable analyses were performed using several prenatal and postnatal variables.

Results: From January 2011 to May 2016, data were reviewed from families of 62 postnatal and 119 fMMC closure patients. In the postnatal group, 80.6% required CSF diversion compared to 38.7% fetal cases ($P < .01$). Hindbrain herniation resolution occurred in 81.5% fetal repairs compared to 32.6% postnatal ($P < .01$). In the fetal group, fetal/premature neonatal demise occurred in 6/119 (5.0%) patients. There was a 42.0% decrease (95% CI -55.2 to -28.8) and 48.9% increase (95% CI 33.7 to 64.1) in risk difference for CSF diversion and hindbrain herniation resolution, respectively, in the fetal group. On univariate analysis for both groups, prenatal atrial diameter, frontal-occipital horn ratio, and hindbrain herniation resolution were significantly associated with the absence of clinical hydrocephalus. The treatment of hydrocephalus was significantly delayed in the fetal group compared to the postnatal group (10 mo vs 13.8 d).

This study demonstrates the benefits of fMMC closure with regard to cerebrospinal fluid dynamics [3)].

Worley G, Rocque BG. Letter: Detailed Analysis of Hydrocephalus and Hindbrain Herniation After Prenatal and Postnatal Myelomeningocele Closure: Report From a Single Institution. Neurosurgery. 2020 Nov 16;87(6):E727. doi: 10.1093/neuros/nyaa445. PMID: 34791467 [4)].

Flanders TM, Heuer GG, Adzick NS. In Reply: Detailed Analysis of Hydrocephalus and Hindbrain Herniation After Prenatal and Postnatal Myelomeningocele Closure: Report From a Single Institution. Neurosurgery. 2020 Nov 16;87(6):E728. doi: 10.1093/neuros/nyaa446. PMID: 34791464 [5)]

[1)]
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Flanders TM, Heuer GG, Adzick NS. In Reply: Detailed Analysis of Hydrocephalus and Hindbrain Herniation After Prenatal and Postnatal Myelomeningocele Closure: Report From a Single Institution. Neurosurgery. 2020 Nov 16;87(6):E728. doi: 10.1093/neuros/nyaa446. PMID: 34791464.