

THE CHIARI MALFORMATIONS

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Dr Hans Chiari¹ first described three hindbrain disorders associated with hydrocephalus in 1891. They have neither an anatomical nor embryological correlation with each other, but they all involve the cerebellum and spinal cord and are thought to belong to the group of abnormalities that result from failure of normal dorsal induction. These include neural tube defects, cephaloceles, and spinal dysraphic abnormalities. Symptoms range from headache, sensory changes, vertigo, limb weakness, ataxia and imbalance to hearing loss. Only those with a type I Chiari malformation may be born grossly normal. The abnormalities are best shown on midline sagittal T1 weighted magnetic resonance imaging (MRI),² but suspicious features on routine axial computed tomographic brain scans (an abnormal IVth ventricle, a “full” foramen magnum, and absent cisterna magna) should be recognised and followed up with MRI.

CHIARI I

This is the mildest of the hindbrain malformations and is characterised by displacement of deformed cerebellar tonsils more than 5 mm caudally through the foramen magnum.³ The brainstem and IVth ventricle retain a relatively normal position although the IVth ventricle may be small and slightly distorted (fig 1). A number of subgroups have been defined.

- ▶ In the first group, intrauterine hydrocephalus causes tonsillar herniation. Once myelinated the tonsils retain this pointed configuration and ectopic position. Patients tend to present in childhood with hydrocephalus and usually with syringomyelia.
- ▶ A second group involves those with associated craniocervical dysgenesis. They usually present later as children or young adults with occipital headaches especially when straining (cough–laugh headache), cranial nerve palsies or dissociated sensory abnormalities secondary to syringomyelia.
- ▶ The third group relate to acquired deformities of the foramen magnum such as basilar invagination. These are usually adults who develop syringomyelia and have headaches and cranial neuropathies.

Although the spectrum of Chiari I malformations is not usually associated with other cerebral abnormalities, syringomyelia is found in 20–70% of patients, depending on the degree and extent of disruption of normal cerebrospinal fluid (CSF) flow between the spine and cranium. Adequate surgical decompression at the foramen magnum and upper cervical spine is the treatment of choice for neuronal dysfunction, symptomatic syrinx or hydrocephalus.⁴ Ventriculoperitoneal shunting may be required for hydrocephalus and syringostomy or syrinx shunting for cord drainage if the craniocervical decompression alone does not relieve the pressure in the syrinx.

CHIARI II

This is a more extensive and complex abnormality than the Chiari I malformation, with infratentorial and supratentorial abnormalities. It affects 0.02% of births, girls twice as often as boys. The cerebellar tonsils, inferior vermis, IVth ventricle, and brain stem are herniated from a shallow posterior fossa through a wide foramen magnum with obstruction to CSF flow at the exits of the IVth ventricle (fig 2). Occasionally the IVth ventricle becomes “trapped” or encysted and will enlarge to appear normal or dilated. There is virtually always a meningocele or meningomyelocele present with some associated hydrocephalus.² Partial or complete agenesis of the corpus callosum is found in most patients, while falx hypoplasia, fused enlarged massa intermedia, colpocephaly, abnormal gyral patterns, and interdigitation of the paramedial gyri across the midline are all associated features. Infratentorially there is beaking of the tectum, petrous bone scalloping, a low torcula, and cervicomedullary kinking. A degree of spinal dysraphism is usually present with a tethered cord and filum lipoma. The abnormality is present at birth and when the meningomyelocele is closed—usually in the first 24 hours—symptomatic hydrocephalus develops. Signs of brain stem compression with swallowing difficulties, stridor, apnoeic spells, a weak cry or arm weakness can

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Figure 1 T1 weighted sagittal MRI showing a Chiari I malformation with cervical–thoracic syrinx in a 25 year old woman with headache and dissociated sensory loss in both arms. There is tonsillar ectopia with the nodulus lying at CV1 level, no cisterna magna, with the brainstem and cerebellum filling the foramen magnum. The IVth ventricle is relatively normal and there is no evidence of hydrocephalus, but a dilated septated syringomyelia is present caused by the disruption of CSF flow at the foramen magnum.



Figure 2 T2 weighted (A) and T1 weighted (B) sagittal MRI showing a Chiari II malformation with a small dysplastic posterior fossa and tonsillar ectopia down to CV3/4 level. There is hydrocephalus with tectal beaking and a dysplastic corpus callosum. T1 weighted sagittal (C) and T2 weighted axial (D) sections through the mid thoracic spine showing the associated meningocele that had been closed at birth but not imaged. The axial section shows the dysplastic cord and the nerve roots merging with a neural placode and fibrofatty mass.

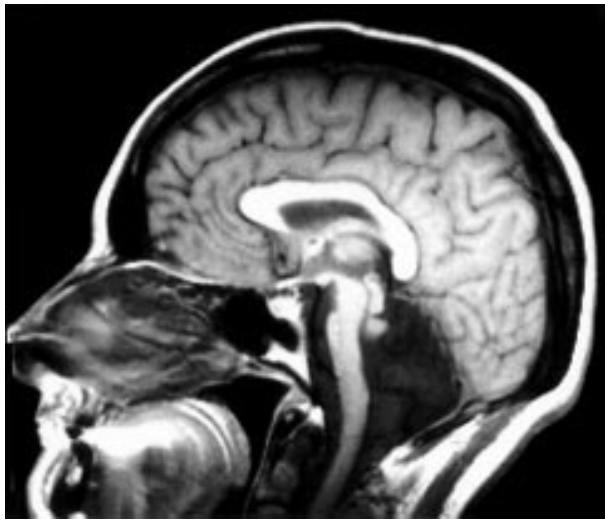


Figure 3 T1 weighted sagittal MRI showing a so called “Chiari IV” or primary cerebellar agenesis. There is a tiny portion of residual quadrangular lobule just caudal to the tectum and a normal sized posterior fossa. No associated meningocele is present.

be found. If presenting as an adult bilateral limb weakness and wasting followed by sensory disturbance are most common, with dysphagia and ataxia being less common. Early symptomatic treatment with shunting and foramen magnum decompression prevents further neurological deterioration.⁵

CHIARI III

Chiari III is very rare. There is herniation of the posterior fossa contents into an associated occipital or high cervical

cephalocele with the other features of a Chiari II malformation.² Patients have severe neurological defects and a poor prognosis.⁶

CHIARI IV

This is controversial and extremely rare.^{2,7} Many authors consider the features of primary cerebellar agenesis as a Chiari IV malformation, but it has to be differentiated from a Chiari II malformation associated with a “vanishing” cerebellum. In primary cerebellar agenesis there are remnants of residual cerebellum—for example, anterior quadrangular lobule—a normal brainstem and a normal sized posterior fossa, and a normal brain and spine (fig 3). The lack of a meningocele virtually excludes a Chiari II malformation.

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