23. Posterior Fossa Congenital Malformations

Chiari malformations are commonly encountered congenital abnormalities. Type 1 malformations are typically asymptomatic, although they may present with headaches, cerebellar signs, or lower cranial nerve symptoms. In addition to a congenital etiology (the main cause), type 1 malformations may result from elevated intracranial pressure and conditions causing basilar invagination (e.g. Paget's disease of bone). Figure 23.1 A demonstrates the principle feature of a type 1 malformation—the herniation of wedge-shaped cerebellar tonsils (black arrow) below the foramen magnum. In this patient, the tonsils descend almost to the level of the posterior arch of C1. Herniation to the C1 level is seen in approximately two-thirds of patients, while extension to the C3 level may occur in up to one-fourth of cases. Tonsils descending less than 5 mm below the foramen magnum are rarely of clinical significance. Figure 23.1 B demonstrates another case of Chiari type 1 with the tonsils (black arrow) extending below the level of the posterior C1 arch. In this case, the malformation is associated with dilatation of the central canal of the cord, hydromyelia or (a more general term) syringohydromyelia (asterisk)—a longitudinally extending CSF-filled cavity which can be seen in this disorder. Hydromyelia demonstrates high SI on T2 and low SI on T1WI. Other associated findings include hydrocephalus (in symptomatic patients) along with osseous abnormalities such as vertebral fusion, spina bifida occulta, and fusion of C1 to the occiput. The corpus callosum and quadrigeminal plate in Figure 23.1 A, B are normal, ruling out a Chiari type 2 malformation.

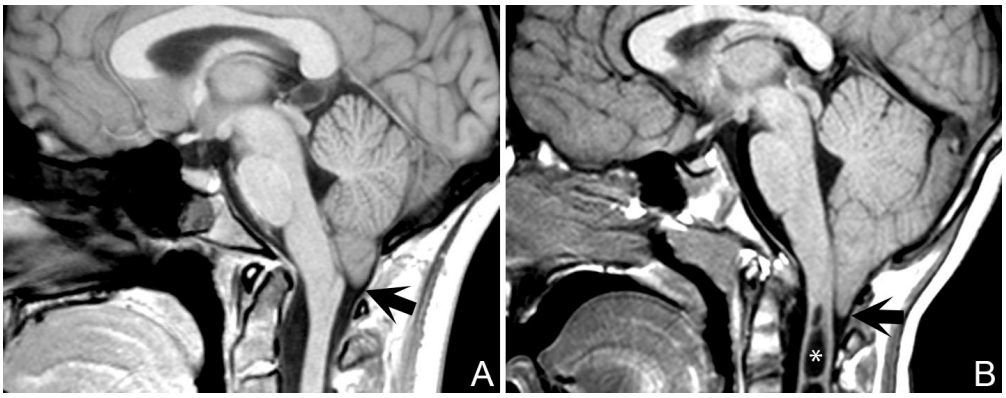

Fig. 23.1

Type 2 malformations are almost uniformly associated with a myelomeningocele—the most severe variant of spina bifida in which both the spinal cord and intact meninges herniate at the lumbosacral midline. Detection of these defects at birth often leads to the diagnosis of Chiari type 2. In about half of cases, cervical or lumbar hydrosyringomyelia may also be

present. As demonstrated in Figure 23.2 A, type 2 malformations involve herniation below the foramen magnum of not only the cerebellar tonsils but also potentially the vermis, brainstem, and fourth ventricle. In severe cases, the medulla may kink, folding over itself at the cervicomedullary junction to rest posterior to the spinal cord. In Figure 23.2 A-1, a portion of the cerebellum herniates through the foramen magnum, but only a smaller portion (the so-called "peg") protrudes through the C1 ring (Fig. 23.2 A-1). Compression of the vermis and the fourth ventricle may occur—giving a slit-like appearance to the latter (Fig. 23.2 A-2). The midbrain colliculi of type 2 malformations (Fig. 23.2 A-3) are typically fused resulting in a beak-like appearance of the tectum. Complete or partial (Fig. 23.2 B-4) agenesis of the corpus callosum is seen in one-third of cases. The massa intermedia—bridging the third ventricle and connecting the two lobes of the thalamus—is frequently enlarged in these patients (Fig. 23.2 B-5). The insertion of the tentorium may also be lower than normal (Fig. 23.2 B-6). As seen in Figure 23.2 C-7 the cerebellum may take on a towering appearance in coronal views with an exaggerated vertical orientation of the folia.

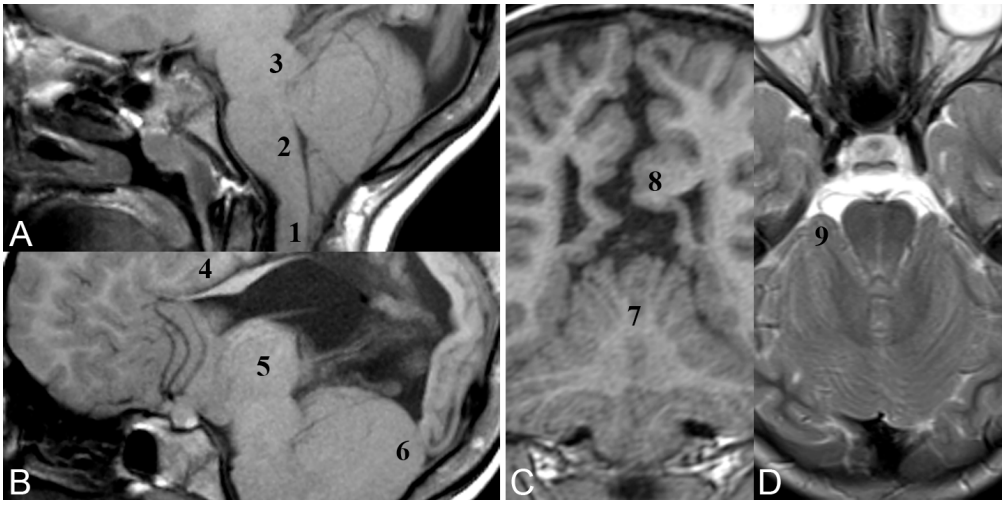

Fig. 23.2

Complete or partial absence of the falx (Fig. 23.2 C-8) may lead to an interdigitated appearance of the gyri. In addition, the gyri are often thin and numerous (stenogyria), not to be confused with polymicrogyria (in which the MRI appearance is grossly smooth) which is not associated with Chiari malformations. Anterior displacement of the cerebellar hemispheres relative to the pons may also occur in Chiari type 2 (Fig. 23.2 D-9). In extreme cases, the hemispheres may touch anterior to the pons, enveloping the brainstem. Hydrocephalus is also common with features including inferiorly pointing frontal horns of the lateral ventricles, large atria, and a prominent suprapineal recess of the third ventricle. Chiari type 3 malformations share the features of Chiari type 2 but are also associated with

a cervical-occipital encephalocele. These may herniate through osseous defects in the posterior vertebral elements or occipital skull.

Dandy-Walker is another congenital malformation of the posterior fossa, the differential considerations for which will be discussed in Chapter 24. In contrast to Chiari malformations, the fourth ventricle is enlarged in Dandy-Walker, communicating with a posterior cyst-like structure. This structure may expand the posterior fossa, elevating the tentorium and the torcular herophili (the confluence of superior, straight, and transverse sinuses). The partial or even complete absence of the cerebellar vermis is an important diagnostic clue. Associated embryonic dysgenesis of the foramens of Magendie and Luschka may alter CSF flow resulting in hydrocephalus—a frequent finding and important prognostic factor. Dandy-Walker may be associated with other intracranial defects—agenesis of the corpus callosum, cortical heterotopias, polymicrogyria, and brainstem lipomas. Extracranial abnormalities include craniofacial and cardiac septal malformations along with polydactyly.