41. Congenital Abnormalities

A number of different congenital conditions can involve the lumbar spine. Caudal regression refers to the absence of sacrococcygeal vertebrae with or without lumbar involvement.

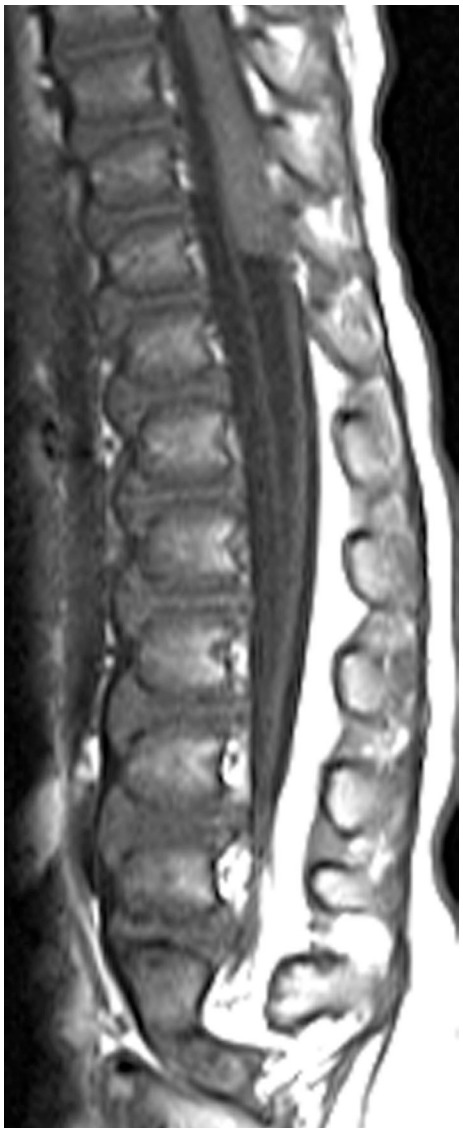

Fig. 41.1

The typical appearance on MR of caudal regression is illustrated in Figure 41.1. On this sagittal T1WI portions of the sacrum are clearly absent and the conus demonstrates a wedge or hatchet-shaped terminal portion. Associated anomalies include renal dysplasia, pulmonary hypoplasia, and neuromuscular weakness. Two groups have been described depending on whether the conus terminates rostrally (group 1) or caudally (group 2 - with the conus medullaris elongated and tethered) to the inferior portion of L1, the latter exhibiting more frequent neurologic dysfunction. Such an abnormally low-positioned conus is referred to as a tethered cord, an entity with many additional (and more common) causes. These include a tight filum terminale, an intradural lumbosacral lipoma, and diastematomyelia. The typical tethered cord patient presents with progressive neurologic dysfunction. Most lesions are repaired at birth (being suspected due to clinical findings, specifically a lumbar neural tube closure defect), but re-tethering may occur, as was the case of the lesion illustrated in Figure 41.2 A, B. Here, two adjacent sagittal T2WI demonstrate a re-tethered cord: the cord gradually tapers until reaching the end of the thecal sac without a distinctly identifiable conus—a typical appearance. Whenever a tethered cord is present, a reason for the tethering must be sought out. In this case a concomitant Type 2 Chiari malformation (see Chapter 25) is to blame, demonstrating classic tectal beaking as well as partial callosal agenesis in the sagittal T1WI of Figure 41.2

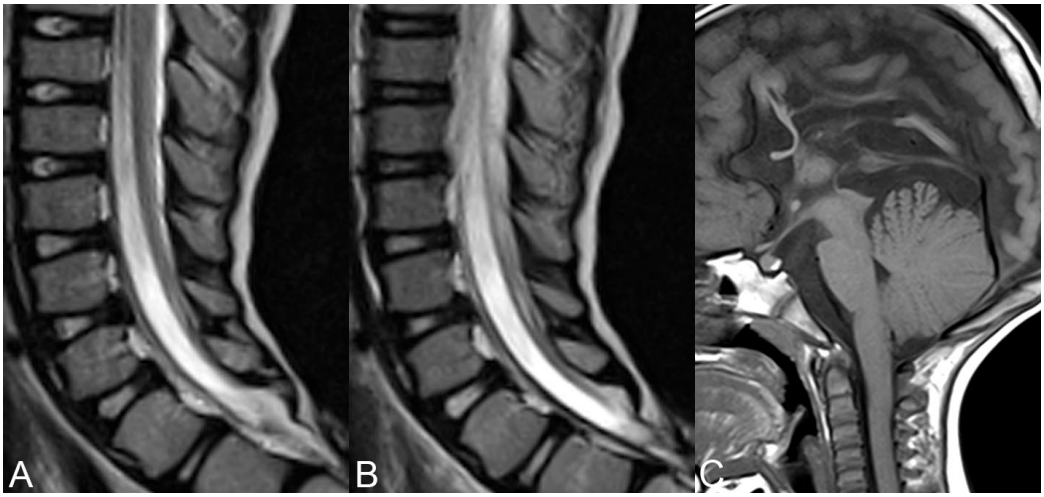

Fig. 41.2

C. Chiari 2 malformations are almost always associated with a lumbar meningomyelocele (Figure 41.3), often with a tethered cord as illustrated. In contrast to spina bifida occulta,

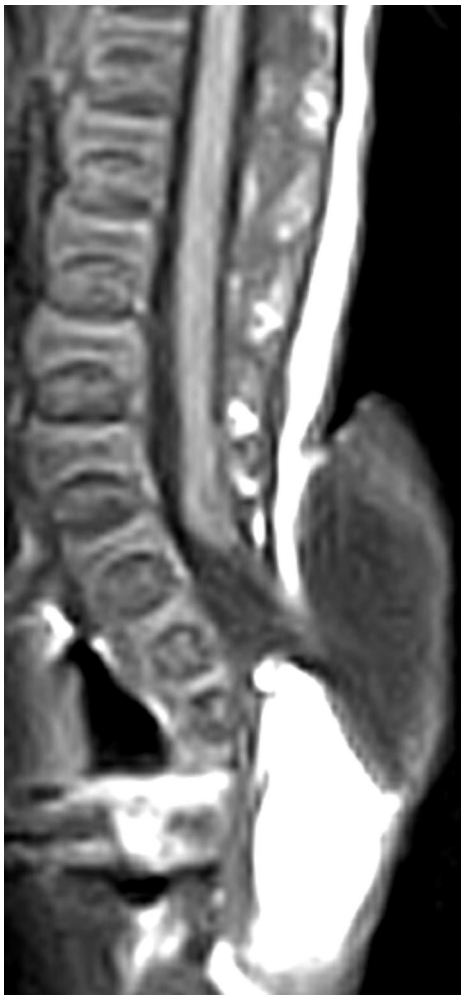

Fig. 41.3

meningomyeloceles consist of not only a posterior arch defect but also herniation of the meninges and neural structures through this defect. Here, the midline sagittal T1WI reveals a CSF-filled sac posteriorly in the lower lumbar region communicating with the normal thecal sac. Although not visible on this image, a single nerve was identified within this fluid-filled sac. The spinal cord extends to at least the level of the lumbosacral junction and dysraphic posterior osseous elements are present from L4 to S1. Abundant fatty tissue inferior to the defect manifests as high SI on T1WI. Note also the relative hypointensity of the vertebral bodies to the intervertebral disks, typical for an infant (see Chapter 50). Following repair of a myelomeningocele re-tethering may occur: there is a limited amount of midline skin and dura, complicating closure over the cord and thecal sac and allowing adherence of the cord to the closure site. In post-operative re-tethering, the posterior cord will not be

visible at the level of closure and the posterior subarachnoid space absent at this level as well. Dorsal dermal sinuses also fall within the spectrum of congenital abnormalities of the spine. In this lesion, a midline epithelium-lined tract, with CSF-like SI characteristics, extends from the thecal sac to the skin surface. Half of such patients have an associated dermoid or epidermoid tumor at the tract's termination, and the tract will enhance in the presence of infection—a common complication. Lipomyelomeningoceles are an additional consideration and are similar to myelomeningoceles except the lipoma is firmly attached to the dorsal surface of the neural placode (cord terminus) which herniates through the dysraphic spinal canal, the lipoma merging with the subcutaneous fat. Lipomas may also occur in the absence of dural defects, as illustrated in Figure 41.4 A-D. Here, the lipoma appears essentially isointense to the vertebral bodies on (A) T2WI but demonstrates high SI consistent with fat on (B) T1WI. Confirming the presence of fat, the lesion loses SI on (C) CE FS T1WI and, as would be expected in a lipoma, does not enhance. Notable in all of these images, and best demonstrated in the axial T1WI of Figure 41.4 D is the undisrupted passage of nerve roots through the lipoma. As such these lesions are typically asymptomatic, not requiring treatment. This appearance may be confused with fatty infiltration of the filum terminale—a common incidental finding occasionally associated with a tethered cord.

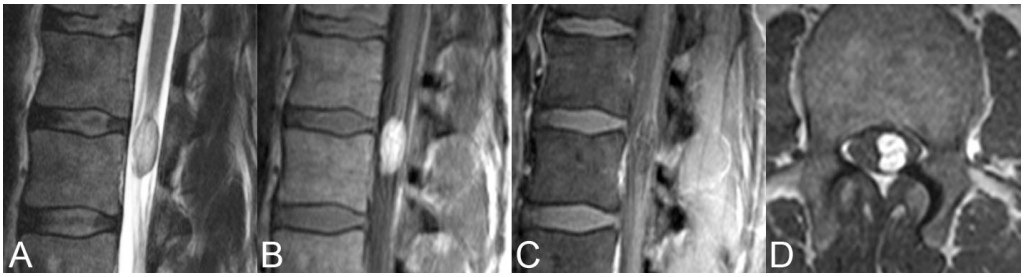

Fig. 41.4