
Scanning Technique and Anatomy

In the neonate, vertebral ossification is not complete. Hence in the first half of infancy before the spinous processes ossify and fuse, it is possible to image the spinal canal from a dorsal view. Later in life a paramedian approach can be used when the spinous processes are more ossified and lead to posterior acoustic shadowing hence precluding imaging in the midline sagittal plane. A high frequency 7–12 MHz linear array transducer is used. Images are obtained in sagittal/longitudinal and axial/transverse planes. Typically a sagittal and axial cine clip at the level of the conus in rest is obtained to document spinal pulsations due to Cerebrospinal Fluid (CSF) pulsation. Pulsation is restricted in tethered cord. However pulsation is best seen a couple weeks after birth. Typically scanning is done in the prone position in a well-fed infant. Having the caregiver hold the baby

in prone position just after feeding increases the chances of an easy motion free exam [1].

The lumbar vertebrae can be labeled by various methods. One method is to assume that the last rib bearing vertebra is T12, another to assign the last square shaped ossified vertebra as S5 and yet another uses the lumbosacral junction as L5-S1 with the vertebra at the end of the lumbar lordosis being L5. When counting the sacral and coccygeal bodies note that the coccygeal vertebrae have a central ossification center compared to the square shaped ossification of the sacral vertebrae. All these methods are an approximation. Usually two or more of these criteria are used to determine the lumbar levels.

Normal Sonographic Findings

The cord appears hypoechoic to almost anechoic. The central canal of the spinal cord is visible as two echogenic lines in the center of the cord. Some believe this to be the interface between the anterior white matter commissure and median fissure [2]. The filum terminale, which is the fibrotic continuation of the spinal pia, below the conus may have a small cyst called the filar cyst as a normal variant [3]. The filum terminale is identified as an echogenic line that is thicker and straighter than the surrounding cauda equina nerve roots. The filum terminale is normally 1–2 mm thick and moves with CSF pulsations (Fig. 2.1).

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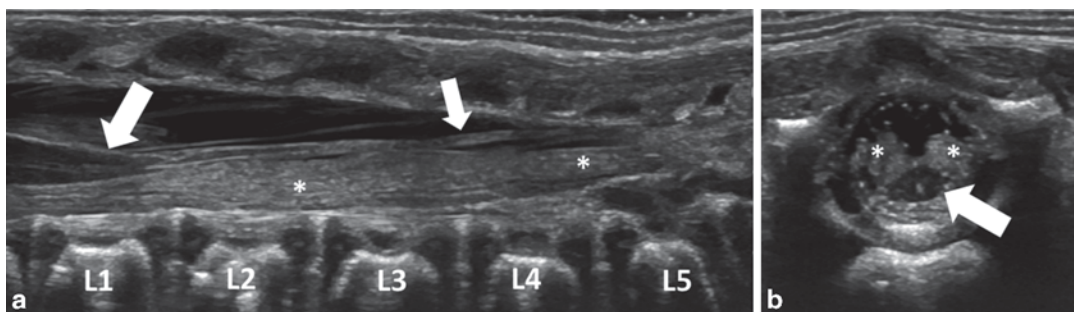


Fig. 2.1 Normal ultrasound of the spine. Sagittal (a) and axial (b). High-resolution ultrasound image of an infant on prone position. The conus medullaris (arrow), the

filum terminale (small arrow) and the cauda equina nerve roots (*)

The position of conus is above the L2-3 disc in a normal term infant. Some accept up to the upper border or midbody of L3 to be within normal limits, if the filum is normal. Ventriculus terminalis is persistent focal dilation of the central canal that is limited to the distal cord. It is a normal variant [3]. If there is question about the position of the conus, placing a radiographic skin marker at the level of the conus under ultrasound guidance and taking a subsequent radiograph of the spine to determine the vertebral level can be performed.

Simple coccygeal dimples or pits, which are shallow, 5 mm or smaller in diameter, located within 2.5 cm cephalad to the anal verge and without any associated suspicious skin lesions, are not associated with an increased risk for spinal dysraphism (Fig. 2.2) [4–6]. No imaging is required for simple coccygeal pits.

The craniocervical junction can be imaged using the foramen magnum as a sonographic window. It is rarely performed, however it can be used to image the inferior cerebellum and proximal cervical cord [1].

Spinal Dysraphism

Spinal dysraphism is the term used for incomplete fusion of the posterior arch of the vertebrae. Closed spinal dysraphism is covered by skin and in open spinal dysraphism the spinal canal contents are exposed without overlying skin.

Open spinal dysraphism can be of two major types: (1) Myelocele with a flat neural placode

(distal end of the cord) exposed and flush with the surrounding skin; (2) Myelomeningocele where the placode is associated with herniated subarachnoid space and meninges. In both the defect is repaired with approximation of the skin within the first 72 h of birth (Fig. 2.3).

Myelomeningocele is virtually always associated with Chiari II malformation. Chiari II malformation involves a small posterior fossa with downward herniation of cerebellar tonsils. In fetal life ultrasound demonstrates the bifrontal skull narrowing called lemon sign and crowding of cerebellum around the brainstem, called banana sign. In the neonate sonographic scanning of the posterior fossa and via the foramen magnum at the craniocervical junction can demonstrate the herniated cerebellar tonsils lying posterior to the upper

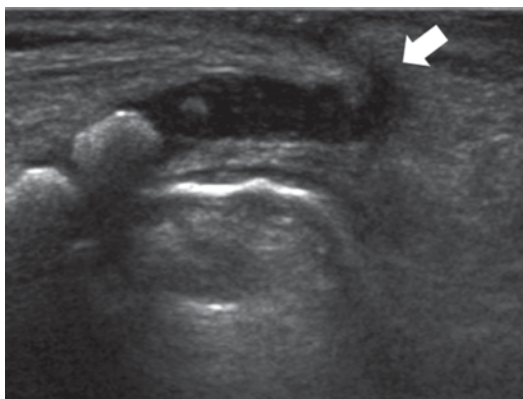


Fig. 2.2 Simple coccygeal dimple. Sagittal ultrasound image of the coccygeal region. Hypoechoic line extending from the skin dimple to the distal coccyx. This finding is considered a normal variant and is not associated with spinal dysraphic anomalies

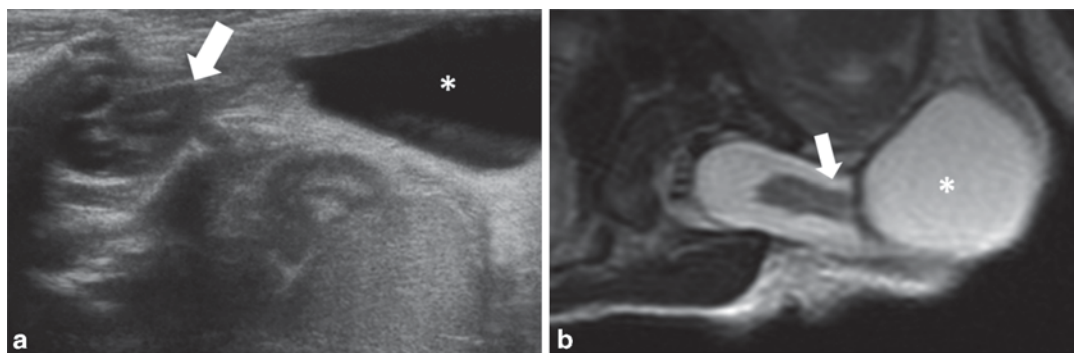


Fig. 2.3 Myelomeningocele. Axial ultrasound (a) and axial T2 MRI weighted image (b). The neural placode

(arrows) and the meninges (*) protruding through the spinal dysraphism

cervical cord. Repair of the cerebellar herniation is only rarely performed in infancy [3]. The cord remains low in position after tethered cord release and closure of the myelomeningocele. It is very difficult to diagnose secondary tethering due to adhering scar tissue after surgery as the imaging findings overlap normal postoperative appearance. Clinical assessment is of utmost importance when re-tethering is suspected.

A meningocele, whether in the cervical or more commonly lumbosacral region, contains only CSF-filled sac of dura mater without any neural elements. The cord can be tethered to the periphery of the sac. It is not associated with Chiari II malformation. A terminal myelocystocele is a rare condition where the herniated CSF space communicates with the distal spinal canal. It is associated with more proximal cord syrinx. Terminal myelocystoceles are associated with omphalocele, cloacal exstrophy, imperforate anus, and spinal anomalies (OEIS) complex [7].

Closed spinal dysraphism is covered by skin. Also called as occult spinal dysraphism it is not associated with an increase in maternal serum and amniotic fluid Alpha-feto Protein (AFP) levels. Most commonly it manifests as a midline spinal abnormality on physical examination in the newborn. Bifurcation or asymmetry of the superior gluteal crease, skin covered hairy patch, skin tag, subcutaneous mass or lump (lipoma), abnormally pigmented patch, telangiectasias, hemangiomas, and high sacral dimples may herald an underlying spinal dysraphic anomaly [4, 8].

Spinal lipoma is the term given to a variety of spinal dysraphisms associated with a fatty component. Lipomyelocele is akin to a myelocele, except that the neural placode is covered by a lipoma that is contiguous with the subcutaneous fat. The neural elements lie within the confines of the vertebral canal (Fig. 2.4). In a lipomyelomeningocele the subarachnoid spaces bulges out of the vertebral canal and pushes the

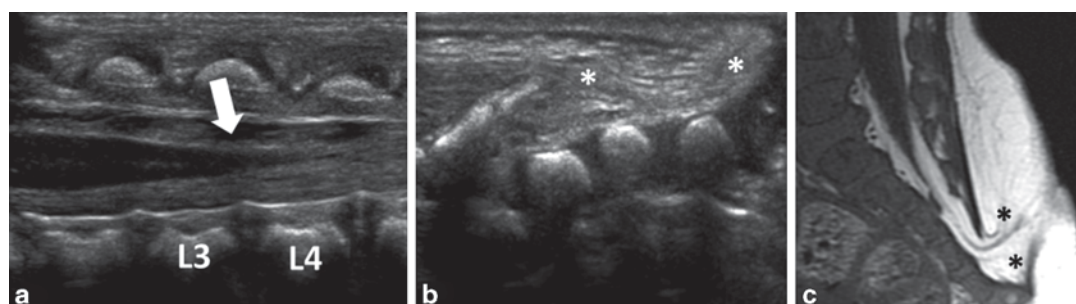


Fig. 2.4 Lipomyelocele. Sagittal ultrasound image (a, b) and sagittal MRI T1 weighted image (c). Low-lying conus medullaris terminating at the level of L4 (arrow) with the

distal spinal dysraphism covered by a lipoma that is contiguous with the subcutaneous fat (*)

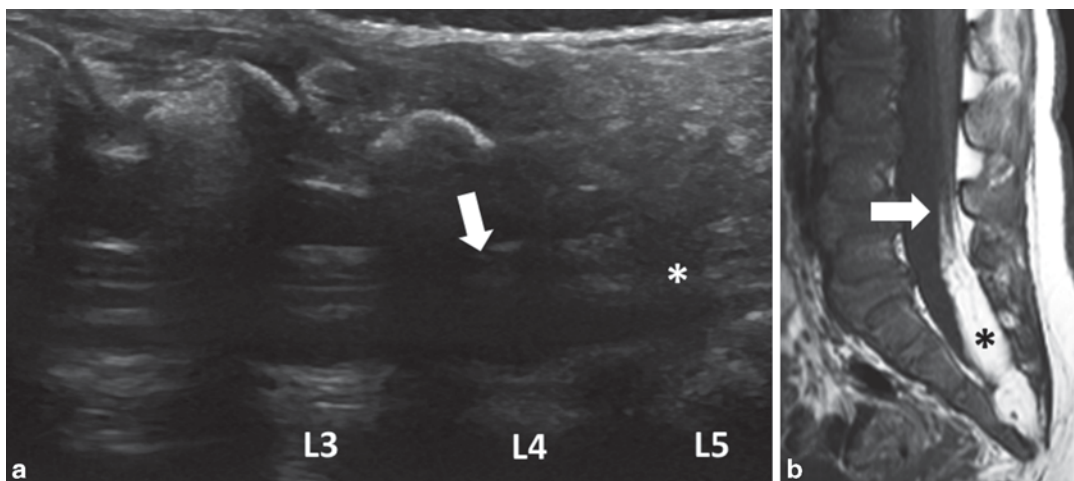


Fig. 2.5 Terminal lipoma. Sagittal ultrasound images of the lumbosacral junction (a) and the sagittal MRI T1 weighted image (b). Tethered cord terminating at the level

of the lumbosacral junction (*arrow*) inseparable from an echogenic mass, consistent with the terminal lipoma seen on MRI T1-weighted image (*)

neural placode and the overlying lipoma as well. The nomenclature of these defects can be easily understood as it represents the layer that is first encountered from the dorsal aspect [1].

Intradural lipoma is a lipoma in the subpial location that is typically attached to the dorsal spinal cord. Intradural lipomas occur more commonly in the thoracic spine, followed by the cervicothoracic junction, and may cause symptoms related to cord compression. The intradural lipomas located in the lumbosacral region, intimately related to the filum terminale, are named terminal lipomas and are often associated with tethered cord (Fig. 2.5). Terminal lipomas are frequently associated with sacral hypoplasia, anorectal malformations, genitourinary malformations, and dorsal dermal sinus.

Lumbosacral dimple: High lumbosacral dimples that are located higher than the gluteal cleft, more than 2.5 cm cephalad to the anal verge, have a higher risk of underlying spinal anomalies and tethered cord. They may represent the opening of dorsal dermal sinuses, which communicate with the underlying spinal canal and dura via a stratified squamous epithelial-lined sinus tract and dysraphic spinous process. Dimples may or may not be associated with hair tufts or hemangiomas.

Tethered Cord

The caudal fixation of spinal cord which leads to stretching of the lower spinal cord and associated neurological dysfunction is the essence of tethered cord (Fig. 2.6). When not treated, the neurological disability may progressively become irreversible. The cord can be tethered in a variety of conditions associated with occult and open spinal dysraphisms. In addition diastematomyelia, filar lipoma, and dorsal dermal sinus may lead to tethering of the cord [3].

Diastematomyelia

Diastematomyelia can be recognized as a split cord which usually reunites distally and may be separated by a bony or fibrous septum. Scanning the entire cord can identify the level of the split and reunited cord.

Findings in Anorectal Malformation

Anorectal malformations may be associated with a variety of spinal anomalies.

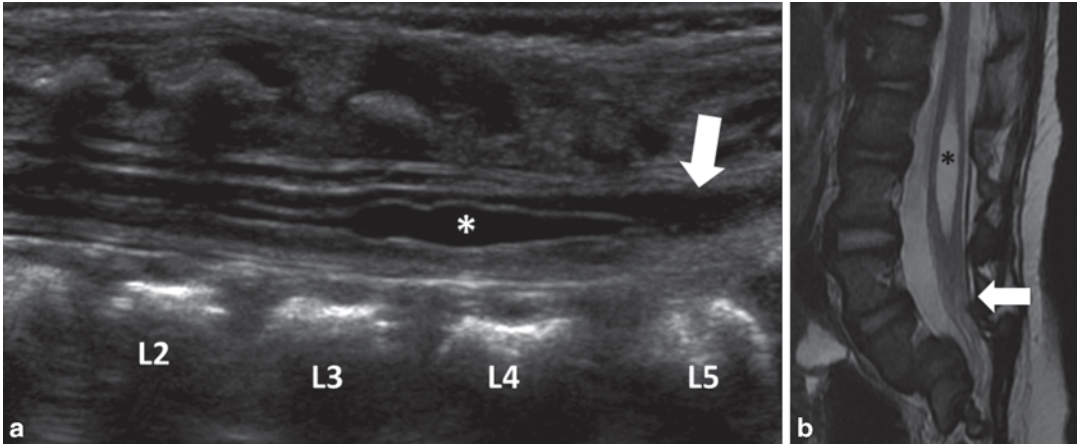


Fig. 2.6 Tethered cord with hydromyelia. Sagittal ultrasound images of the lumbosacral region (a) and the sagittal MRI T2 weighted image (b). Tethered cord terminat-

ing at the level of L5-L1 (arrow) with dilation of the distal ependymal canal, consistent with hydromyelia (*)

Caudal regression syndrome is characterized by insult to the caudal cell mass that gives rise to the coccyx, distal sacrum and the lower lumbar spine. It is more common in infants of diabetic mothers [9]. Anorectal anomalies such as imperforate anus are typically associated. Bladder and

renal anomalies may also be present. The spinal cord in caudal regression type 1 is high ending and blunted (Fig. 2.7). In type 2 caudal regression, which is less common, the cord is low lying and tethered to a fibrolipoma.

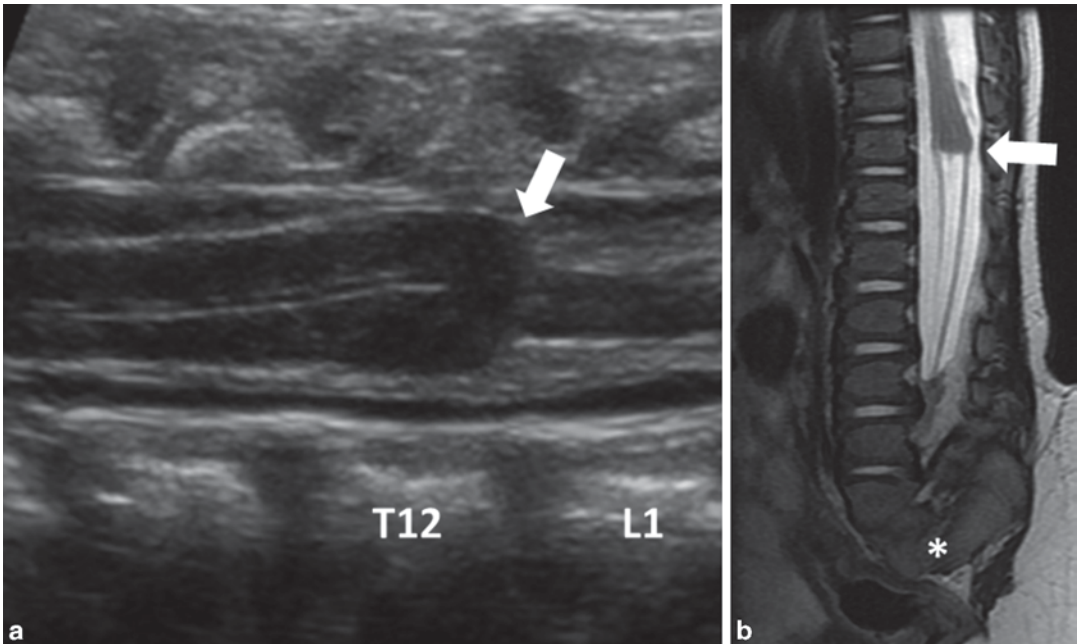


Fig. 2.7 Caudal regression syndrome. Sagittal ultrasound image of the thoracolumbar junction (a) and sagittal MRI

T2-weighted image (b). High ending, blunted distal spinal cord (arrow) and dysplastic sacrum (*)

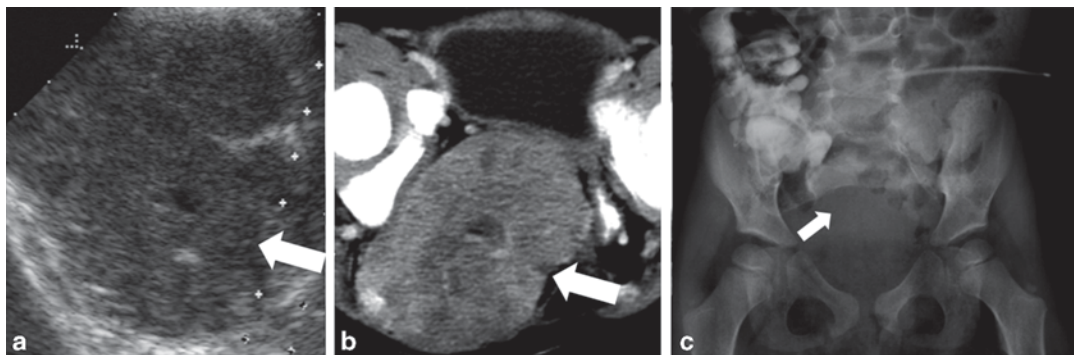


Fig. 2.8 Currarino syndrome. Sagittal ultrasound image of the lumbosacral region (a), axial contrast enhanced CT image (b) and anteroposterior radiograph of the pelvis

(c). Heterogeneous presacral mass (arrow) and dysplastic sacrum (small arrow) in an infant with the anorectal malformation

The syndrome of Currarino is a rare congenital disorder that comprises a triad of dysplastic sacrum, anorectal malformation, and presacral mass (Fig. 2.8). Infants with Currarino syndrome may also have urogenital malformation. The presacral mass is commonly a teratoma, although anterior sacral meningocele or duplication cysts are also possible. The dysplastic sacrum is typically partial and one sided leading to a scimitar shaped sacrum. Agenesis, scalloping, and sacral hypoplasia are also described. Inheritance is autosomal dominant [10].

The presence of cloacal anomalies, such as exstrophy, cloacal malformations and, imperforate anus and ectopic anus are highly associated with spinal cord anomalies and dysraphism. OIES syndrome is associated with cloacal exstrophy and terminal myelocystocele (vide supra).

Neoplasm

The most common spinal tumor of infancy is intraspinal extension of a neuroblastoma. Ultrasound can be used to evaluate the extent of intraspinal tumor and may demonstrate the extent of cord compression [1].

Sacroccoccygeal teratomas form the next most common spinal tumors. They can have an intrapelvic and extrapelvic component. They were classified by Altman into four types that progressively have an increase in the intrapelvic compo-

nent, with minimal intrapelvic extension in type I to predominantly intrapelvic in type IV. Up to 50% are of type I (Fig. 2.9). Sacroccoccygeal teratomas may be mature or immature on pathology and the level of differentiation determines the malignant potential. These masses are typically heterogeneous in echotexture with solid and cystic components. Many are now diagnosed in prenatal life and up to 70% are evident on neonatal exam as a lump or exophytic extrapelvic mass. Diagnosis may be delayed with Type IV sacroccoccygeal teratomas. Delayed diagnosis is associated with a more complex surgery as well as higher incidence of malignant elements on pathology [11].

In adults and older children intraoperative guidance with ultrasound can be useful to delineate tumor from the spinal cord. Sonography is performed after laminectomy is done. A high frequency transducer is used.

Spinal Trauma

Intraspinal hemorrhage after lumbar puncture may be seen as echogenic debris in the subarachnoid space in infants. An epidural or subdural hemorrhage may occasionally be seen as a fluid collection posterior to the cord. For bony fractures and more extensive traumatic insult including traumatic infarcts of the cord, an MRI is more useful. Beyond infancy the value of spinal

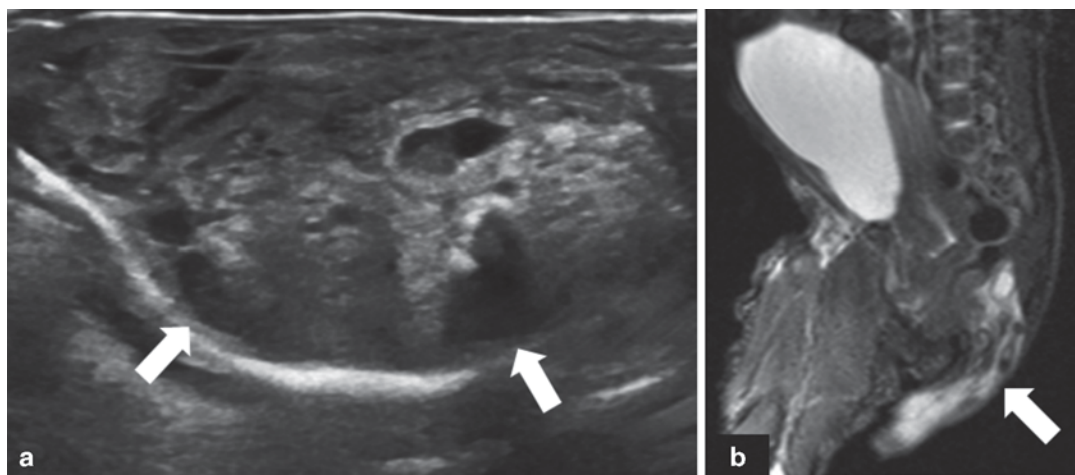


Fig. 2.9 Sacrococcygeal teratoma. Sagittal ultrasound image (**a**) and sagittal MRI T2-weighted (**b**) of the perineal region. Exophytic, heterogeneous mass with solid

and cystic components (*arrows*), consistent with type I sacrococcygeal teratoma

sonogram is rapidly reduced. It remains useful for evaluating posterior spinal collections and seromas after surgery [1].

With the recent focus on child abuse, a study by Edelbauer et al. suggests use of spine sonography in less than 6-month-old infants for evaluating subdural collections. Spinal trauma with ligamentous injuries and cord insult are described with non-accidental trauma. Spinal subdural collections are also common in the presence of subdural haemorrhage in the cranium. Ultrasound may in future prove to be a valuable adjunct in evaluating extra-axial collection in the spinal canal in this patient group [12].

Prenatal Diagnosis

During second trimester routine screening fetal ultrasound scans the skin overlying the spine is evaluated for integrity. The spinous processes form a linear echogenic line. The absence of skin and the spinous process herald the presence of an open neural tube defect. The splayed ossification centers lateral to the defect is seen on axial imaging. When the defect is covered by a thick sac, it may represent skin covering of a closed neural tube defect. Indirect signs of a myelomeningocele in the brain in the form of frontal notching

and small posterior fossa can be seen. Closed spinal dysraphisms are generally occult and difficult to diagnose prenatally unless associated with a lipomyelomeningocele that protrudes significantly away from the canal. Sporadically some of these have been diagnosed prenatally. Visualizing the level of the conus prenatally is challenging, however a significantly low-lying conus below L3 may point towards underlying occult spinal dysraphism. Anterior sacral meningocele and sacrococcygeal teratomas are diagnosed prenatally in a large percentage of patients. Smaller defects, particularly closed spinal dysraphisms and intradural lipomas may not be apparent on prenatal scanning [13].

Summary

Vertebral ossification is not complete in the first half of infancy and sound beam transmits through the cartilage. This combination makes imaging the spinal canal with sonography possible. Sonography is usually the first imaging exam to be performed in infants because of its simplicity, low cost, and lack of deleterious effects. Another advantage of sonography over most imaging modalities is its capacity to visualize the motion of the filum terminale, which is restricted

in tethered cord. In this chapter, the ultrasound techniques, the indications and the imaging findings of the most common disorders involving the infant's spine are reviewed.

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**Diagnostic and Interventional Ultrasound in Pediatrics
and Pediatric Surgery**

Scholz, S.; Jarboe, M.D. (Eds.)

2016, XV, 287 p. 278 illus., 134 illus. in color.,

Hardcover

ISBN: 978-3-319-21698-0