## Pictorial review

# Spinal ultrasound in infants

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**Abstract.** This paper discusses the indications for spinal ultrasound, including its advantages and disadvantages compared with spinal MRI. The features and ultrasound findings both in normal infants and in those with spinal dysraphism are reviewed.

Spinal ultrasound (SUS) is becoming increasingly accepted as a first line screening test in neonates suspected of spinal dysraphism (SD) [1, 2]. SUS can be useful in detecting tumours, vascular malformations and cases of trauma; however, as the indication in most cases is to exclude SD, this article will concentrate on this group of abnormalities [2–4]. The advantages of SUS are not only a diagnostic sensitivity equal to MRI [2] but that, unlike MRI, SUS can be performed portably, without the need for sedation or general anaesthesia. In addition, MRI is highly dependent on factors affecting resolution, including patient movement, physiological motion from cerebral spinal fluid (CSF) pulsation and vascular flow, factors that do not affect SUS [5]. New generation high frequency ultrasound machines with extended field of view capability now permit imaging of high diagnostic quality in young babies.

SD refers to abnormalities with imperfect fusion of the midline neural and bony structures. It is the most common congenital central nervous system abnormality, with myelomeningoceles occurring in up to 2 per 1000 live births in some studies, although the incidence in the Western world is now likely to be lower [5, 6]. In infants with occult SD, early diagnosis may be useful, as SD may lead to distortion of the spinal cord and nerve roots with growth, resulting in neurological sequelae in the lower extremities, the lower urinary tract and the gastrointestinal tract. Because some paediatric neurosurgeons believe that early surgical correction of SD may avoid these sequelae, early diagnosis of SD could be important [7, 8].

## How to do it and what to look for

SUS was first proposed in the early 1980s but it has taken some time to enter routine clinical

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practice [9, 10]. The examination is performed using a 7.5–10 MHz linear probe in both the sagittal and axial plane along the entire spine. Ideally the patient is laid prone on a pillow with the neck flexed for craniocervical junction imaging, but imaging with the patient in the decubitus position is also feasible. The examination can be performed in the Ultrasound Department or on the ward with a portable machine. No sedation is necessary.

SUS is possible in the neonate owing to a lack of ossification of the predominantly cartilaginous posterior arch of the spine [11, 12]. The quality of ultrasound assessment decreases after the first 3–4 months of life as posterior spinous elements ossify, and in most children SUS is not possible beyond 6 months of age. However, the persisting acoustic window in children with posterior spinal defects of SD enables ultrasound to be performed at any age [11, 12].

The normal neonatal spinal cord is displayed on ultrasound as a tubular hypoechoic structure with hyperechoic walls (Figure 1). The central canal is hyperechoic, the so-called central echo complex [13]. The subarachnoid space surrounding the cord is hypoechoic. The caudal end of the spinal cord corresponds with the conus medullaris, which continues into the filum terminale. The cauda equina is seen as echogenic linear structures surrounding a hyperechoic filum terminale. The vertebral bodies are seen as echogenic structures ventral to the spinal cord. Particular features to note are:

(i) The level of the conus medullaris. In term infants the tip of the conus medullaris normally lies above the mid level of the L2 vertebral body although there is a large range of normality (from T10/11 to L2/3) [14]. In pre-term infants the tip of the conus lies between L2 and L4, *i.e.* the level of the conus moves proximally with age [15]. A low-lying cord may imply tethering.

(ii) The position of the cord in a dorsal/ventral

or anterior/posterior orientation. In normal infants the cord lies a third to half way between the anterior and posterior walls of the spinal canal. If the cord lies more posteriorly, tethering should be suspected (Figure 2).

(iii) The presence of normal pulsatile movement of the cord and nerve roots (tethering leads to an absence of pulsatility).

(iv) The thickness of the filum terminale (normal 2 mm or less [16]).

Viewed axially, the spinal cord is hypoechoic and round or oval-shaped, lying within the anechoic subarachnoid space (Figure 3a). The spinal cord is fixed by dentate ligaments that pass laterally from the spinal cord. Echogenic nerve roots are seen below the level of L2 (Figure 3b).

## Patient selection: who should be investigated for SD

Spinal MRI is a limited resource and cannot be used as a screening tool. Ward et al [17] suggested that only patients with a clinically demonstrable neurological abnormality, such as increased tone or hemiplegia, should be investigated for SD using MRI, as they found that only those with clinically demonstrable neurological defects demonstrated an abnormality on MRI. Other authors concur with this suggestion. Schwend et al [18] found that all patients who needed neurosurgical intervention could be identified by abnormal neurological findings on clinical examination.

SUS has the advantage over MRI that it is a less restricted resource and can therefore be used as a screening tool in all babies suspected of SD. SD may be suspected if the infant has: (i) a neurological deficit; (ii) cutaneous stigmata, such as a haemangioma, sacral pit or tuft of hair; or (iii) other abnormalities associated with SD such as cloacal exstrophy [19] or anorectal or sacral abnormalities and agenesis [12, 20]. Current Royal College of Radiologists guidelines are that all neonates with a hairy patch or sacral dimple should undergo SUS [21]. However, while more than 90% of patients with occult SD have a cutaneous abnormality over the lower spine [22], a cutaneous marker may have a low yield in predicting the presence of a clinically significant abnormality. In a recent review of 200 SUS examinations performed over an 11-year period, SD was found in less than 1% of cases when a cutaneous marker was the only clinically detected abnormality [23].

The diagnostic value of SUS has been shown to be equal to MRI [2]. Rohrschneider et al [2] found that SUS exactly correlated with MRI in 32 out of 38 cases. In five cases, SUS depicted the main abnormality but MRI gave additional information. Wherever SUS was normal, MRI was also normal, *i.e.* SUS had a sensitivity of 100%. Current literature therefore suggests that SUS may be used as a primary screening tool, with MRI being performed in any case where SUS revealed an abnormality [1, 2, 24].

## Embryogenesis

The defects of SD occur in the first 8.5 weeks of pre-natal life, as the fetal nervous system develops. The neural tube and the subsequent spinal cord arise from ectodermal cells. The surface ectoderm separates from the neural tube, with mesoderm coming to lie between the neural tube and the ectoderm. The mesoderm forms the bony spine, meninges and muscle. Incomplete separation of the neural tube from the ectoderm may result in cord tethering, diastematomyelia or a dermal sinus. Premature separation of the cutaneous ectoderm from the neural tube can result in abnormal mesenchymal elements such as lipomas, forming between the neural tube and skin [16]. If the neural tube fails to fold and fuse in the midline, defects of posterior SD such as a myelomeningocele occur. Finally, disorders of the distal cord (so called "caudal cell mass") lead to fibrolipomas of the filum terminale.

## Classification of SD

SD can be classified into non-skin-covered, skin-covered and occult abnormalities (see Table 1) [5].

### Low cord without tethering (Figure 4)

In a term infant, the conus should lie above the level of mid L2. The clinical significance of a low cord unaccompanied by any other abnormality is unknown [7].

**Table 1.** Classification of common types of spinal dysraphism (SD)

| |
|---|
| ***SD without a back mass (SD occulta)*** |
|   low cord |
|   tethered cord |
|   diastematomyelia |
|   anterior sacral meningocele |
|   spinal lipoma |
| ***SD with a skin-covered back mass*** |
|   lipomyelomeningocele |
|   myelocystocele |
| ***SD with a non-skin-covered back mass*** |
|   myelomeningocele/myelocele |

Adapted with kind permission from Byrd SE, Darling CF, McLone DG, Tomita T. MR imaging of the pediatric spine. Magn Reson Imaging Clin N Am 1996;4:797–833.

## Tethered cord syndrome (Figure 2)

In tethered cord syndrome, the conus is low, the spinal cord is often displaced dorsally, there is a lack of normal spinal cord pulsatility as well as thickening of the filum terminale (>2 mm) [6]. This thickening may be fibrous or lipomatous in nature.

## Diastematomyelia (Figure 5)

Diastematomyelia consists of partial or complete sagittal clefting of the spinal cord into symmetrical or assymetrical hemicords, which usually re-unite caudally. Each segment has a central canal, and dorsal and ventral nerve roots. In 50–60% of cases the hemicords are contained in a single dural sac and arachnoid covering without a dividing spur (type A) [6]. In 40–50% of cases (type B), each hemicord has a separate dural and arachnoid covering with a spur or septum between, which may be bony, cartilaginous or fibrous in nature. SUS in the axial plane identifies the two hemicords and also the spur, which is usually echogenic. Diastematomyelia occurs most commonly in the thoracolumbar region. 50–75% of patients have cutaneous stigmata at the site of diastematomyelia [16]. Diastematomyelia is commonly associated with tethering in around 75% of patients and syringo-hydromyelia in around half of patients. Both these abnormalities must therefore be looked for and excluded in the presence of diastematomyelia.

## Anterior sacral meningocele (Figures 6a–c)

In anterior sacral meningoceles, there is herniation of a dural sac containing CSF through a bony sacral or coccygeal defect. The meningoceles may be isolated or associated with neurofibromatosis or Marfans syndrome. Patients may be asymptomatic or may present with bladder/bowel dysfunction or a pelvic mass. Plain radiographs often show a sacral defect. SUS demonstrates an anechoic meningocele anterior to the sacrum. MRI confirms that it is fluid-filled with signal isointense to CSF. The meningocele may be unilocular or multilocular. Associated cord tethering and lipomata should be looked for [16].

## Spinal lipoma (Figures 7a–d), including lipomyelomeningocele (Figure 7e)

Spinal lipomas comprise 20–50% of occult SD [16]. Lipomas are highly echogenic and easily discernible on ultrasound [13]. They are classified into the following groups: (i) lipomyelocele or lipomyelomeningocele (84%); (ii) fibrolipoma of the filum terminale (12%) often associated with a tethered cord (see Figures 7a–d); or (iii) intradural lipoma (4%) [25]. Unlike most other forms of SD, which are commonest in the lumbar spine, spinal lipomas are most commonly found in the cervical and thoracic spine. They can be large and may cause local compression. Spinal lipomas are usually dorsal and subpial in situation. Where they are associated with myeloceles and myelomeningoceles, the lipomatous mass is in continuity with a spinal cord that has not fused in the midline. In these cases the lipoma extends dorsally through a spina bifida defect and is flush with the skin (lipomyelocele) or herniates beyond the skin (lipomyelomeningocele, associated with an enlarged dorsal subarachnoid space; Figure 7e). 50% of patients with lipomyeloceles or lipomeningoceles present with a lumbar soft tissue mass and 50% present with lower limb neurology or bladder symptoms, often in later childhood [8].

## Myelocystocele

Myelocystoceles are rare subtypes of myelomeningoceles where the central canal dilates and herniates through the posterior spinal bifida defect. The mass is covered by skin. Hydromyelia is present in all cases. On ultrasound, characteristic flaring of the anechoic central canal as it enlarges into a cystic mass is seen. Continuity with the central canal is the key feature in differentiating it from myelomeningocele [16].

## Meningocele and myelomeningocele (Figures 8a–c)

These are two of the most common SD abnormalities, occurring in up to 2 of 1000 live births [25]. They arise from localized failure of fusion of the neural folds dorsally. In meningocele there is herniation of distended spinal meninges but not neural tissue through a dysraphic spine. In myelomeningocele, portions of the spinal cord and nerve roots lie within the sac. These anomalies occur most commonly at the lumbosacral level. Ultrasound shows an anechoic, lobulated meningocele in continuity with a tethered, low lying spinal cord. Myelomeningoceles and myeloceles are both associated with cord tethering [5, 16].

## Myelocele

In myeloceles, the spinal cord is openly exposed to the skin surface with the central canal lying open. There is no skin closure in the midline. This occurs most commonly in the lumbosacral region. Surgical repair is performed within 48 h of birth because of the high risk of infection. Pre-operative imaging is rarely performed and should be avoided owing to the risk of infection [25].
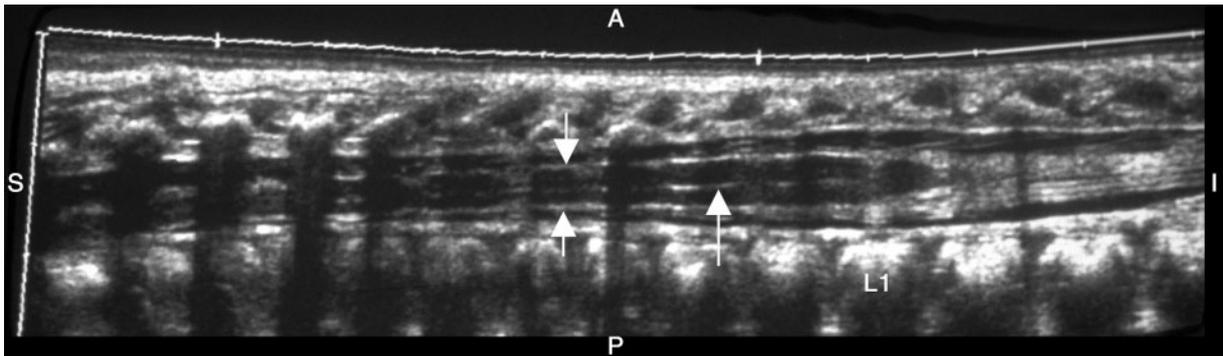
**Figure 1.** Normal sagittal neonatal spine on an extended field of view ultrasound. The L1 vertebral body is indicated. Note the position of the cord in an anteroposterior direction as well as the echogenic nerve roots extending beyond the tip of the conus medullaris. The spinal cord lies between short arrows. Hyperechoic central canal (long arrow). A, anterior; P, posterior; S, superior; I, inferior. (Reproduced with kind permission from Dick EA, de Bruyn R, Patel K, Owens CM. Spinal ultrasound in cloacal exstrophy. Clin Radiol 2001;56:289–94.
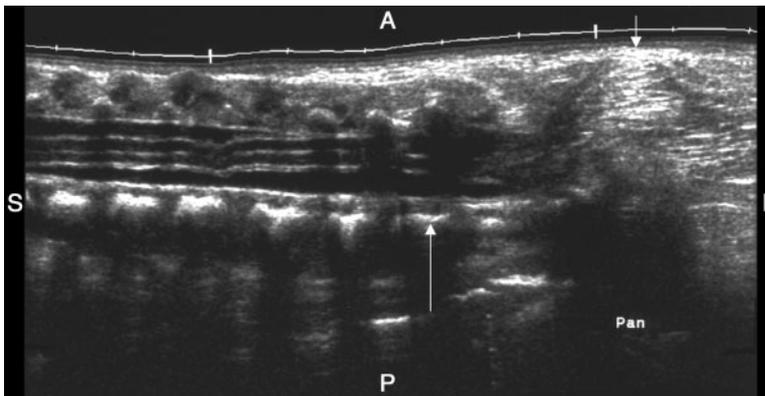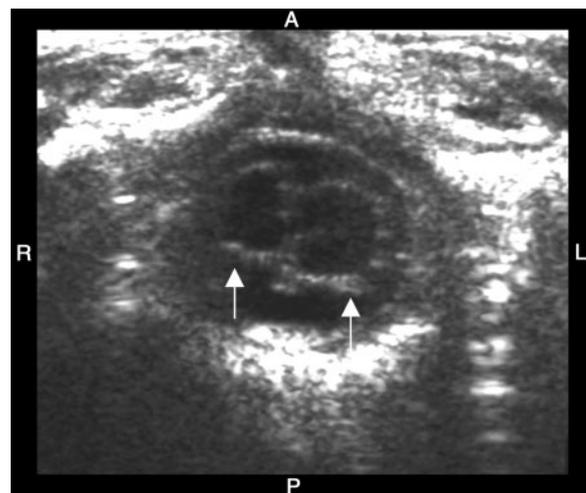


**Figure 2.** Sagittal spinal ultrasound showing tethering of the cord with dorsal displacement. The conus medullaris lies at the level of L5 (long arrow). Cutaneous sacral dimple is seen (short arrow).

(a)



(b)
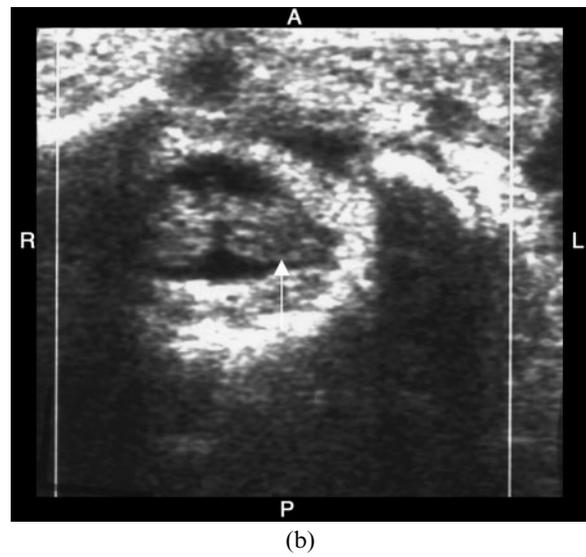
**Figure 3.** (a) Normal axial neonatal spine at the level of T12 showing the dentate ligaments extending laterally from the spinal cord (arrow). (b) Normal axial neonatal spine at the level of L3 showing echogenic nerve roots. Left-sided nerve roots arrowed.
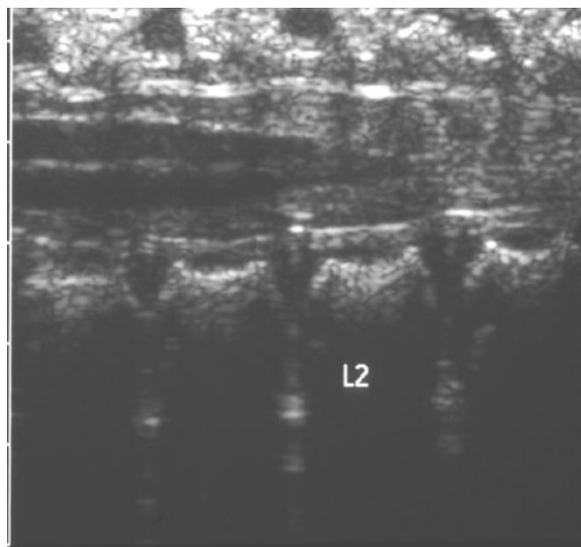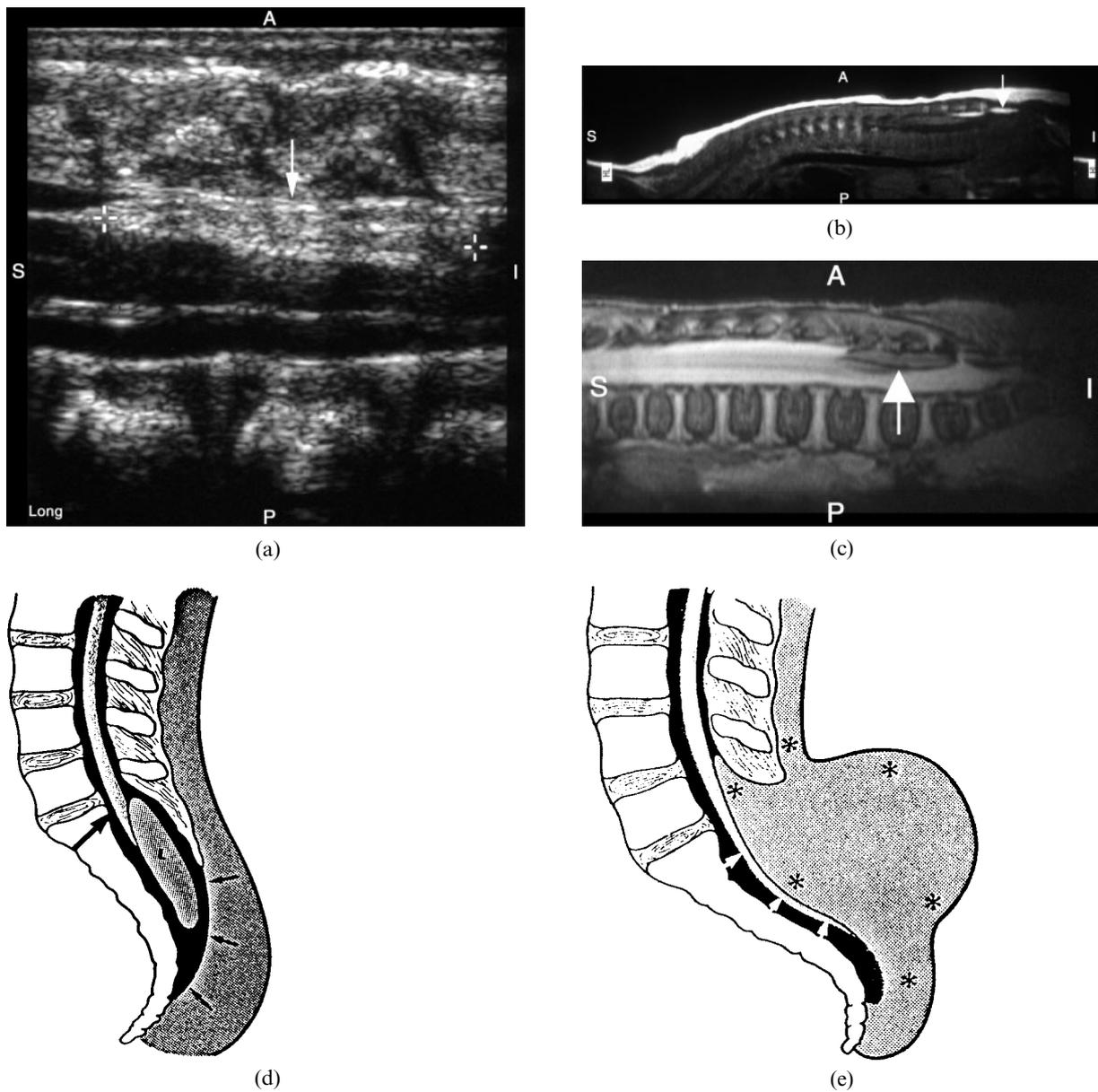


**Figure 4.** Sagittal spinal ultrasound showing a low cord but no tethering. The tip of the conus lies at the lower level of L2 but the cord is not dorsally displaced.
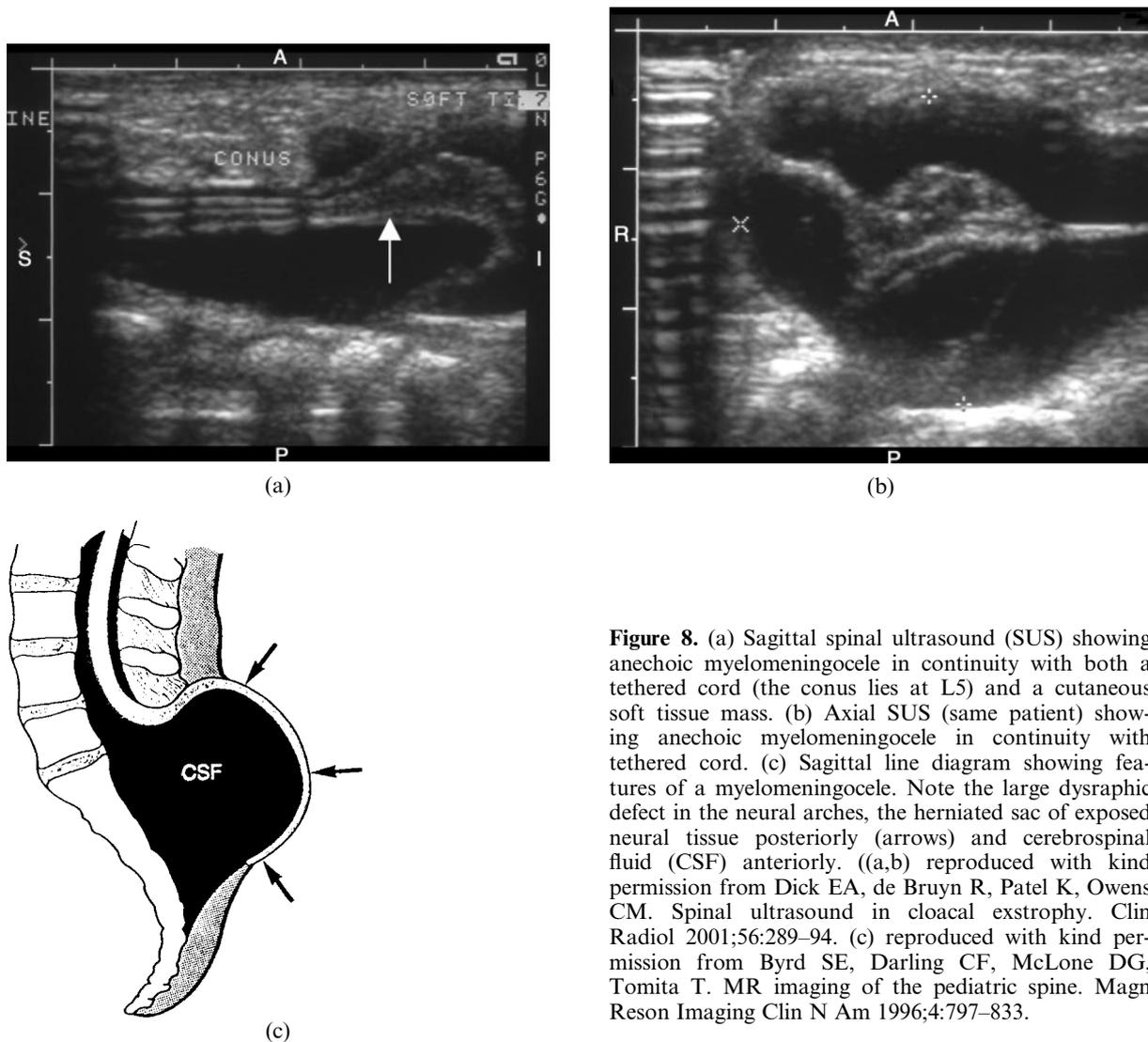


**Figure 5.** Axial spinal ultrasound showing diastematomyelia. Both hemicords are arrowed. An echogenic spur lies between the two hemicords.

(a)

(b)



(c)
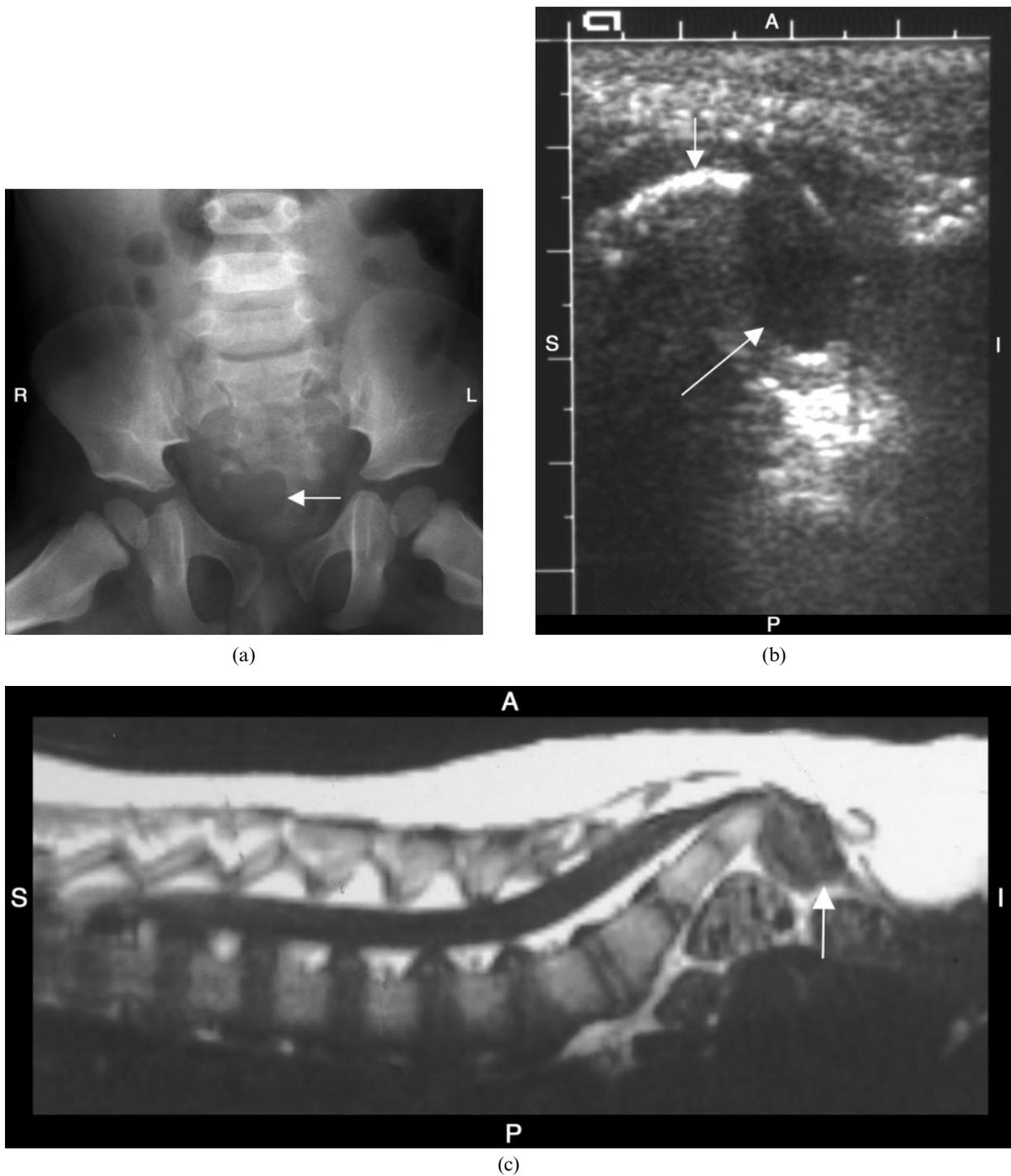
**Figure 6.** (a) Plain radiograph showing partial absence of the right sacrum and coccyx (arrow) in a patient with an anterior meningocele. (b) Sagittal spinal ultrasound (same patient) demonstrates an anechoic unilocular meningocele anterior to the sacrum (long arrow), anterior to the inferior portion of the sacrum (short arrow). (c) Sagittal $T_1$ weighted MRI (same patient) demonstrates a low signal meningocele (arrow) anterior to the inferior portion of the sacrum.

(a)



(b)



(c)



(d)



(e)

**Figure 7.** (a) Sagittal spinal ultrasound showing lipoma of the filum terminale (arrow). (b) Sagittal $T_1$ weighted MRI (same patient) demonstrates high signal lipoma (arrow). (c) Sagittal $T_2$ weighted MRI (same patient) demonstrates lipoma of intermediate signal, and confirms that the cord is tethered, extending to level L5. (d) Sagittal line diagram showing features of lipoma of the filum terminale. The lipoma (L) tethers a low lying spinal cord (large arrow). The intradural lipoma is separated from subcutaneous fat of the back by a tissue plane (small arrows). (e) Sagittal line diagram showing features of lipomyelomeningocele in the lumbosacral region. There is a large dysraphic defect in the neural arches and a skin-covered herniated sac composed of fat that is contiguous with the subcutaneous fat (asterisks) and a low lying tethered spinal cord (arrows). ((a–c) reproduced with kind permission from Dick EA, de Bruyn R, Patel K, Owens CM. Spinal ultrasound in cloacal exstrophy. Clin Radiol 2001;56:289–94. (d,e) reproduced with kind permission from Byrd SE, Darling CF, McLone DG, Tomita T. MR imaging of the pediatric spine. Magn Reson Imaging Clin N Am 1996;4:797–833.)

(a)



(b)



(c)

**Figure 8.** (a) Sagittal spinal ultrasound (SUS) showing anechoic myelomeningocele in continuity with both a tethered cord (the conus lies at L5) and a cutaneous soft tissue mass. (b) Axial SUS (same patient) showing anechoic myelomeningocele in continuity with tethered cord. (c) Sagittal line diagram showing features of a myelomeningocele. Note the large dysraphic defect in the neural arches, the herniated sac of exposed neural tissue posteriorly (arrows) and cerebrospinal fluid (CSF) anteriorly. ((a,b) reproduced with kind permission from Dick EA, de Bruyn R, Patel K, Owens CM. Spinal ultrasound in cloacal exstrophy. Clin Radiol 2001;56:289–94. (c) reproduced with kind permission from Byrd SE, Darling CF, McLone DG, Tomita T. MR imaging of the pediatric spine. Magn Reson Imaging Clin N Am 1996;4:797–833.

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