Dorsal dermal sinus in children

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Learning objectives

To describe dorsal dermal sinus (DDS), the associated spinal findings and its complications.

Cutaneous stigmata around DDS are easily detected in infants and should initiate a radiological work up of the spine, to include a spinal ultrasound examination in neonates and in children < 6 months.

MRI is more suitable in children > 6 months due to the ossification of the posterior elements.

Background

Cutaneous stigmata over the spine above the level of the natal cleft are associated with a dorsal dermal sinus (DDS).

DDS is associated with a range of abnormalities and its presence increases the risk of local and intra-spinal infection.

Findings and procedure details

The incidence of DDS is approximately 1 in 2500 live births (1).

DDS is part of a wide-ranging spectrum of abnormalities encompassed by the term "closed spinal dysraphism". The term dysraphism refers to incomplete closure of the neural tube during the process of neurulation.

A closed defect results when there is complete cutaneous coverage of the defect.

DDS is a consequence of focal, incomplete separation of the cutaneous ectoderm from the neural ectoderm between the third and eighth week of gestation (2, 7).

DDS is covered by stratified squamous epithelium and extends a variable depth from the skin to the spinal fascia, dura mater, cauda equina or spinal cord (Fig 1).
Clinical findings

Infants are referred to radiology following a postnatal check and the discovery of a cutaneous dimple or stigmata such as skin discolouration, patchy hair at the base of the spine, localised cutaneous swelling or a discharging dimple.

DDS is located above the natal cleft and characteristically has a tract, which is orientated in a cephalic direction (4)(Fig. 2).

Most children with DDS are neurologically normal at birth and become symptomatic later if not surgically corrected.

The most common dimple is sacrococcygeal dimple also known as a sacral or coccygeal dimple or pit. It is usually located in the natal cleft and has a horizontal or caudal tract. Importantly, a sacrococcygeal dimple is not associated with intradural pathology (5). Fig 3-4.

A neurologic deficit can develop due to the presence of a tethered cord (Fig 5), growing tumour, spinal malformation or the onset of infection (Fig. 9, 10).

**Neurologic symptoms and signs** include: Gait disturbance, motor weakness, sensory change, abnormal reflexes, decreased sphincter tone and resultant difficulty with bowel and bladder function.

Imaging

DDS is most frequently located in the lumbosacral region and is less commonly found in the thoracic, cervical or rarely occipital regions.

The most commonly encountered anomalies are tethered cord, inclusion tumours (epidermoid and dermoid), intraspinal lipomata and split cord malformation. Ascending tract infection can result in epidural and subdural abscess formation and meningitis.
Inclusion tumours such as dermoid and epidermoid are found in association with DDS in 50% of cases (3).

**Ultrasound**

On ultrasound (US) examination DDS tract appears slightly hypoechoic and is sometimes difficult to visualise with US. DDS tract is more easily demonstrated within the anechoic CSF filled subarachnoid space as an echogenic structure (6).

**MRI**

MRI is the diagnostic imaging modality of choice for visualisation of the DDS, and also clearly delineates any associated intraspinal abnormalities.

*Our recommended MRI protocol is:*

- Sagittal and axial T1 and T2 weighted images (WI) (3 mm thick slices without gap)
- Sagittal T1 or T2 WI with fat suppression.
- Diffusion weighted imaging in cases of possible epidermoid tumour.
- In cases of spinal infection contrast enhanced images are required.

DDS tract is seen as hypointense structure on both T1 and T2 weighted sequences, traversing through the hyperintense subcutaneous fat (Fig. 6). Complete visualisation of the entire extent of the DDS is important for preoperative assessment and to aid surgical planning (7).

An associated *dermoid* is a well-defined midline structure and has a signal intensity equal to that of fat on T1 and T2 weighted images.

An *epidermoid* is an off-midline structure, with signal characteristics similar to cerebrospinal fluid. A hyperintense signal on diffusion-weighted imaging (DWI) is diagnostic (Fig. 7, 11, 12, 13) (8).

A *tethered cord* is diagnosed when the conus medullaris is seen to lie below the L2 vertebral body (Fig. 5). The filum terminale may also demonstrate thickening (8).
DDS with *lipomyelocele* is diagnosed when placode-lipoma connection lies in the spinal canal. Subcutaneous fat extends through bone defect into the spinal canal and attaches to the cord (Fig. 14, 15, 16) (4).

**Role of radiography**

AP and lateral radiographs of the lumbosacral spine have a role in evaluation of possible DDS in the child older than 18 months.

Radiographic findings include:

- Bifid laminae
- Increased interpeduncular distance
- Vertebral segmentation anomalies (hemivertebrae, fused vertebrae) and subsequent scoliosis (Fig. 8).

Radiography has a limited role in children younger children than 18 months due to the presence of an immature skeleton.

**Differential diagnoses:**

Differential diagnoses include: Sacrococcygeal dimple and pilonidal sinus.

- **Sacroccocygeal dimples** (also known as sacral or coccygeal dimple or pits) are located in the natal cleft, and have horizontal or caudal direction of the tract.

They are not associated with intradural pathology.

Ultrasound is the method of choice for evaluation of the sacroccocygeal dimple. MRI of a sacroccocygeal dimple is indicated in the case of abnormal ultrasound findings or atypical symptoms (9).

- **Pilonodal sinuses** are not connected with spinal canal. They occur at the coccygeal level and are usually seen in patient’s greater than 15 years of age.
Management

Surgical resection of DDS and release of tethered cord is indicated prophylactically to prevent development of neurologic complications and infection.

Even when MRI imaging shows normal findings DDS located above the gluteal crease should be surgically explored to its termination (5).

Infections are treated with antibiotic therapy as per local protocol. Abscesses may require neurosurgical intervention.

Images for this section:

Fig. 2: XR L/S spine: Levels of DDS and sacro-coccygeal dimple
Fig. 3: US, axial plane: Blind ending sacro-coccygeal dimple.
Fig. 1: DDS illustration
**Fig. 4:** US, sagittal plane: Low sacro-coccygeal dimple
Fig. 5: MRI, Sagittal T2 WI and STIR: Hypointense DDS, low conus medullaris, tight filum terminale

Dorsal dermal sinus at the level S1/2 with a low lying conus medullaris

* Asterisk is positioned over the Dorsal Dermal Sinus

Hypo-intense structure traversing the bright fat (far left image) is the sinus tract of the DDS.
Fig. 6: MRI, sagittal T2, STIR and T1 WI: Lipomyelocele and DDS
Fig. 8: MRI, coronal T1WI: hemivertebrae, DDS
Fig. 7: MRI Sagittal T1 WI, DWI, ADC: Occipital DDS and epidermoid
Fig. 9: MRI, sagittal T2 WI: DDS and spinal canal abscess
Fig. 10: MRI, sagittal T1-w fat suppressed post contrast images: DDS and spinal canal abscess

Fluid filled thecal sac and avid contrast enhancement of the DDS, thecal sac and intradural structures. Conus medullaris and cauda equina are not discernible.
Fig. 11: MRI sagittal planes: DDS, epidermoid and filum terminale lipoma
Fig. 12: MRI T1 and T2 WI, axial plane: DDS, epidermoid and filum terminale lipoma
**Fig. 13:** US sagittal plane: Epidermoid shown as an anechoic cyst in spinal canal
Fig. 14: MRI sagittal T2 WI: Hypointense DDS, bone defects, subcutaneous and intraspinal lipoma with placode.
Fig. 15: MRI, Axial T2 WI: DDS with lipomyelocele
**Fig. 16:** MRI and US in the same patient with DDS and Lipomyelocele. US(right)image: subcutaneous lipoma, DDS is not shown.
**Conclusion**

**Conclusion:**

DDS is located above the natal cleft and has a high rate of associated abnormalities and complications. The presence of cutaneous stigmata or DDS above the natal cleft should initiate appropriate imaging, with surgical referral.

DDS should be differentiated from a sacrococcygeal dimple that is found within the natal cleft and is not associated with intraspinal pathology.

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