Imaging findings in spinal dysraphisms

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Learning objectives

- To review the normal and pathologic embryological spinal cord development.
- To describe the clinical-radiological classification system of spinal dysraphisms.
- To understand the role and the limits of the different modalities of imaging in the pre and postnatal diagnosis.
- To illustrate the modalities of writing a report.
Background

Spinal dysraphisms is a broad term given to a group of anomalies where there are malformations of the spine and the spinal cord. They arise from defects occurring in the early embryological stages of the three basic steps of spinal development. There is often abnormal fusion of the midline embryonic neural, vertebral and mesenchymal structures.

- The incidence is approximately 1 per 1,000. The defect is located in the lumob sacral region in 90% of cases, in the thoracic region in 6-8%, and in the cervical vertebrae in 2-4%. There is a defect of the vertebrae that may be posterior, anterior (rarely) or, in rare cases, consists of a splitting of the vertebral body.

Their etiology is multifactorial: genetic, environmental factor and folic acid deficiency.

The diagnosis is either prenatally, at birth, in the early childhood based on ultrasound, CT with a particular focus on MRI for imaging complexes abnormalities.
Findings and procedure details

A selection of patients that present different forms of spinal dysraphism, to choose the most appropriate modality of imaging that involves considering many factors.

Imaging of the bony spine requires methods different from those used to image the spinal canal and its contents. The age of the patient also influence the choice of modality.

Investigation of the spinal canal and its contents are best performed by MRI; that technique provides more information than myelography or CT in defining spinal cord anatomy.

The best way to image skeletal anomalies is by means of plain radiography, but they provide little information of the associated malformations of the spinal cord and its coverings. Though this modality has now been more or less replaced by CT.

Ultrasonography is limited to the neonatal and fetal period, fetal ultrasonography is increasingly used as a primary screening tool usually at about 18 weeks' gestational age, to diagnosis early open forms and planify an appropriate treatment to prevent infections.

1- Embryological spinal cord development:

Spinal development can be summarized in three basic embryologic stages. The first stage is gastrulation and occurs during the second or third week of embryonic development.

Gastrulation involves conversion of the embryonic disk from a bilaminar disk to a trilaminar disk composed of ectoderm, mesoderm, and endoderm.

The second stage in spinal development is primary neurulation (weeks 3-4) in which the notochord and overlying ectoderm interact to form the neural plate. The neural plate bends and folds to form the neural tube, which then closes bidirectionally in a zipperlike manner.

The final stage of spinal development is secondary neurulation (weeks 5-6). During this stage, a secondary neural tube is formed by the caudal cell mass. The secondary neural tube is initially solid and subsequently undergoes cavitation, eventually forming the tip of the conus medullaris and filum terminale by a process called retrogressive differentiation. Abnormalities in any of these steps can lead to spine or spinal cord malformations.

Failure of primary neurulation leads to open dysraphism posing the risks of CSF leakage and exposure of neural placode. Defects at the secondary neurulation result in occult
dysraphism connecting the epidermis and the mesenchymal tissues, leading to variety of anomalies and tethered cord.

Deficient development or function of the notochord result in disorders of notochordial development that are various and complex. These malformations show a partial overlap with disorders of secondary neurulation (e.g. caudal regression syndrome). Following malformations may be encountered: "split notochord syndromes”; neurenteric cyst or fistula and segmental disorders of notochordal development.

2-Clinical-neuroradiological classification of spinal dysraphisms

- Spinal dysraphisms can be broadly categorized into open and closed types. In an open spinal dysraphism, there is a defect in the overlying skin, and the neural tissue is exposed to the environment. In a closed spinal dysraphism, the neural tissue is covered by skin. Closed spinal dysraphisms can be further subcategorized on the basis of the presence or absence of a subcutaneous mass.

- Open spinal dysraphisms:
  - Myelomeningocele
  - Myelocele
  - Hemimyelomeningocele
  - Hemimyelocele

- Closed spinal dysraphisms (With subcutaneous mass):
  - Lipomyelomeningocele
  - Lipomyelocele
  - Meningocele
  - Myelocystocele
  - Skin-covered myelomeningocele (limited dorsal myeloschisis)

- Closed spinal dysraphisms without subcutaneous mass
  
  Simple dysraphic states
  - Intrudal lipoma
  - Filar lipoma
  - Tight filum terminale
  - Persistent terminal ventricle
  - Dermal sinus

- Complex dysraphic states
  1. Disorders of midline notochordal integration
     a) Diastematomyelia
     b) Neurenteric cysts
     c) Dorsal enteric fistula
  2. Disorders of notochordial formation
     a) Caudal agenesis
     b) Segmental spinal dysgenesis
3-Imaging modalities

Prenatal

Foetal ultrasonography and in utero MRI have increasingly enabled the identification of various spinal pathologies during early stages of gestation.

This can be important to distinguish the open spinal dysraphisms for delivery planning decisions regarding pregnancy interruption, and in some cases, preparation for in utero surgery. Patients with closed neural tube defects have a better postnatal prognosis, including superior bladder functionality and lower risk of scoliosis, than the open neural tube defect.

With the advent of fetal therapy including surgery, accurate prenatal diagnosis of open and closed spinal dysraphisms becomes critical in appropriate counselling and perinatal management.

Foetal ultrasonography:

Advances in prenatal US including high frequency linear transducers and three dimensional imaging can provide detailed information concerning spinal anomalies.

Fetal spine is visible since 9 Weeks of gestation, correctly accessible in the 12 week, which allow us to make an early diagnosis of most malformations.

In week 22, the spine is well studied catching some missed diagnosis or discrete forms of dysraphism.

Prenatal sonographic detection of open spinal dysraphisms(OSD) is reported to be 80-90 %.

Direct US finding for OSD (spinal):

Common signs:

- Absence of fusion of vertebral posterior processes on the midline.

- Presence of neural tissue in the cyst.

Sagittal images demonstrate the defect by defining the lack of overlying soft tissues and absent posterior line (formed by the ossification centers of the posterior elements of the vertebrae) at the level of the defect. They are also useful to evaluate spinal curvatures that may exagered with large spinal defects.
In the coronal plane, the central line (corresponding to the ossification centers of vertebral bodies) is usually difficult to find and the two external lines (corresponding to the posterior elements of the vertebrae) are more separate than the adjacent normal ones.

Axial planes, demonstrate splaying of the ossification centers pertaining to the lateral processes; the neural canal exposed posteriorly.

The presence of a sac certainly helps OSD diagnosis with the visual extension of the subarachnoid space through the bony defect.

**Indirect US finding for OSD (brain):**

There is 100% association between OSDs and the Chiari II malformation, a congenital hindbrain anomaly characterized by:

- A small posterior fossa.
- Caudal displacement of the vermis, brainstem and fourth ventricle toward the cervical subarachnoid space.

This association is explained by the downward displacement of the brain secondary to cerebrospinal fluid leakage through the spinal defect into the amniotic cavity and hypotension in the subarachnoid spaces, leading to caudal displacement of the brain with obstructive hydrocephalus providing indirect signs for OSD with large prevalence during second trimester and subsequently were developed first trimester indirect sonographic markers to screen dysraphism at an earlier stage, similarly to second trimester:

- Tonsillar herniation.
- Large foramen magnum.
- Obliteration of cistern magna
- Distortion of the cerebellum
- Reduction of the fourth ventricle (v4)
- Venticulomegaly
- Retraction of frontal bones

**Other associations:**

- Collosal dysgenesis, tectal beaking, subependymal heterotopias (better visualized by MRI).

- Feet malposition.

After the 12th week US can find
**Banana sign:**

- Obliteration of the cisterna magna.
- Small size and abnormal shape of the cerebellum.

**The lemon sign:**

- Abnormal shape of the frontal bones due to scalloping.

**Hydrocephalus**

**Closed spinal dysraphism:** Uncommon and rarely diagnosed prenatally. If associated with a subcutaneous mass may be identified prenatally and can be mistaken for an OSD if not carefully evaluated.

**Fetal MRI:** (Fig. 1)

Fetal MRI is well accepted as complementary image tool to confirm the suspected spinal anomalies and even detecting malformations that were not detected at the prenatal ultrasonography. It is used to better delineate involvement of neural elements associated with osseous spinal anomalies.

Each fetal MRI study will examine the entire fetus including the head, torso, extremities, umbilical cord, placenta and uterus. An exact knowledge of the spinal findings as discovered by US will guide the fetal MRI examination. If necessary, additional dedicated sequences will be focused on the suspected spinal malformation.

Standard fetal MRI is performed at 1.5 or 3 Tesla field strength. Typically, a triplanar (axial, coronal and sagittal to the mother) "scout" sequence (e.g. TRUE-Fast Imaging-with-Steady-state-Precession sequence (TRUE FISP) or a breath-held, heavily T2-weighted thick-slab MR-fetography sequence) is acquired at the start of the study. These sequences allow to position the subsequent high resolution sequences adapted to the position of the fetus. In addition, these fast, non-breath hold sequences give valuable preliminary anatomical information about the fetus and its position. The thick-slab MR fetography sequence is a heavily T2-weighted sequence which covers the entire fetus, preferably in the sagittal plane. The prominent signal contrast between the soft tissues and fluid (e.g. amniotic fluid and cerebrospinal fluid [CSF]) give an excellent, fast overview of the fetal contours as well as of the integrity and shape of the spinal canal. Obvious spinal canal malformations are usually easily identified.
These "scout" sequences are followed by high resolution, dedicated T2-weighted ultrafast Single-Shot-Fast-Spin-Echo or Half-Fourier-Single-Shot-Turbo-Spin-Echo sequences. The single-slice acquisition and the ultrafast acquisition-times (# 500 msec per slice) allow to "picture freeze" the fetus intrauterine.

The T2-weighted sequences have an excellent contrast to noise and signal-to-noise ratio. The fetal central nervous system (CNS) and spinal cord are still very "watery" which renders an optimal MRI signal. In addition, the spinal cord and canal is well depicted because the spinal cord is outlined by the T2-hyperintense CSF. Multiplanar imaging is necessary to study the exact anatomy of the normal and abnormal spinal cord.

T1-weighted imaging is limited because currently no single-shot ultrafast T1-weighted sequences are available. T1-weighted imaging may however be essential to identify T1-hyperintense fat as can be seen in cases of a lipomeningomyelocele, intradural lipomas or in fetuses with sacro-coccygeal teratomas.

The intravenous injection of gadolinium should be avoided. Injected gadolinium will reach the fetal circulation via the placenta and will be excreted into the amniotic fluid by the fetal kidneys. The gadolinium will remain for a prolonged time period in the fetal circulation because the excreted gadolinium will re-enter the fetal circulation multiple times because the amniotic fluid is swallowed by the fetus.

**Plain radiography and CT:**

Are rarely used as an alternative imaging technique for the evaluation of fetal skeletal abnormalities. The use of these ionizing radiation loaded techniques should however be limited as much as possible and limited to the third trimester of pregnancy.

**Post natal**

Additional imaging in the postnatal period can be useful in evaluating the newborn with vertebral anomalies noted on pre-natal imaging.

**Ultrasonography**

It is possible to perform spinal ultrasonography in the newborn, owing to the lack of ossification of the predominantly cartilaginous posterior elements of the spine. Spinal sonography is usually not possible after the age of 6 months except in cases of a persistent posterior spinal defect.

Neonatal spinal ultrasound will show major intraspinal anomalies and tethering.

**MRI**
MRI is helpful in assessing the degree of hindbrain herniation and spinal cord anatomy when ultrasonography is limited. It demonstrates better intraspinal anomalies but generally requires a general anesthetic or heavy sedation which has theoretical deleterious effects in the young child.

When and whether all vertebral anomalies should be evaluated postnatally with MRI for intraspinal anomalies is debated. Certainly any patient with clinical signs of intraspinal anomaly (extremity deformity or bladder function suggestive of neurologic abnormality) or about to have surgery of the spine should have a whole spine MR.

Plain radiography and CT

Are used in the evaluation of skeletal abnormalities. Three-dimensional CT reconstructions may be helpful for complex osseous malformations and various skeletal dysplasias.

4-Congenital spinal anomalies

1-Open spinal dysraphisms

1.1 Myelomeningocele and myelocele:

Myelomeningoceles account for more than 98% of open spinal dysraphisms. Myeloceles are rare.

The main differentiating feature between a myelomeningocele and myelocele is the position of the neural placode relative to the skin surface. The neural placode protrudes above the skin surface with a myelomeningocele (Fig.2) and is flush with the skin surface with a myelocele. The lesions usually appear hypointense on T1W images and hyperintense on T2W images similar to CSF.

1.2. Hemimyelomeningocele and hemimyelocele:

Hemimyelomeningoceles (Fig.3) and hemimyeloceles can also occur but are extremely rare. These conditions occur when a myelomeningocele or myelocele is associated with diastematomyelia (cord splitting) and one hemicord fails to neurulate.

2. Closed spinal dysraphisms

2.1. Closed Spinal Dysraphisms With a subcutaneous Mass

2.1.1 Lipomas with a dural defect:

Lipomas with a dural defect include both lipomyeloceles and lipomyelomeningoceles.
The main differentiating feature between a lipomyelocele and lipomyelomeningocele is the position of the placode lipoma interface. With a lipomyelocele, the placode lipoma interface lies within the spinal canal. With a lipomyelomeningocele, the placode lipoma interface lies outside of the spinal canal due to expansion of the sub-arachnoid space.

2.1.2. Meningocele (Fig.4):

Herniation of a CSF-filled sac lined by dura and arachnoid mater is referred to as a meningocele. The spinal cord is not located within a meningocele but may be tethered to the neck of the CSF-filled sac.

Posterior meningoceles herniate through a posterior spina bifida (osseous defect of posterior spinal elements) and are usually lumbar or sacral in location but also can occur in the occipital and cervical regions. Anterior meningoceles are usually presacral in location but also can occur elsewhere. On MRI imaging shows hypointense mass on T1W image and hyperintense on T2W images.

2.1.3. Terminal myelocystocele:

Herniation of large terminal syrinx (syringocele) into a posterior meningocele through a posterior spinal defect is referred to as a terminal myelocystocele. The terminal syrinx component communicates with the central canal, and the meningocele component communicates with the subarachnoid space. The terminal syrinx and meningocele components do not usually communicate with each other.

2.1.4. Myelocystocele:

A non terminal myelocystocele occurs when a dilated central canal herniates through a posterior spina bifida defect. Myelocystoceles are covered with skin and can occur anywhere but are most commonly seen in the cervical or cervicothoracic regions.

2.2. Closed spinal dysraphisms without a subcutaneous mass:

2.2.1. Simple dysraphic states:

2.2.1.1. Intradural lipoma (Fig.5):

Lipoma located along the dorsal midline that is contained within the dural sac.

Intradural lipomas are most commonly lumbosacral in location and usually present with tethered-cord syndrome. On MR imaging, lipoma appears as a mass which follows signal intensity of subcutaneous fat on all pulse sequences, high signal intensity on T1W and FSE T2W images, hypointense on fat saturated images.
2.2.1.2. Filar lipoma:

Fibrolipomatous thickening of the filum terminale is referred to as a filar lipoma. On imaging, a filar lipoma appears as a hyperintense strip of signal on T1-weighted MR images within a thickened filum terminale. Filar lipomas can be considered a normal variant if there is no clinical evidence of tethered-cord syndrome.

2.2.1.3. Persistant terminal ventricle:

Persistence of a small, ependymal lined cavity within the conus medullaris is referred to as a persistent terminal ventricle. Key imaging features include location immediately above the filum terminale and lack of contrast enhancement, which differentiate this entity from other cystic lesions of the conus medullaris.

2.2.1.4. Dermal sinus:

A dermal sinus is an epithelial lined fistula that connects neural tissue or meninges to the skin surface. It occurs most frequently in the lumbosacral region and is often associated with a spinal dermoid at the level of the cauda equina or conus medullaris.

MRI imaging shows hypointense tract in both T1W and T2W images extending from skin surface to spinal canal. Surgical repair is of great importance because the fistulous connection between neural tissue and the skin surface can result in infectious complications such as meningitis and abscess.

2.2.2. Complex dysraphic state:

2.2.2.1. Disorders of midline notochordal integration:

2.2.2.1.1. Dorsal enteric fistula and neurenteric cyst (Fig.6):

A dorsal enteric fistula occurs when there is an abnormal connection between the skin surface and bowel.

Neurenteric cysts represent a more localized form of dorsal enteric fistula. These cysts are lined with mucin secreting epithelium similar to the gastrointestinal tract and are typically located in the cervicothoracic spine anterior to the spinal cord.

2.2.2.1.2. Diastematomyelia (Fig.7):

Separation of the spinal cord into two hemicords is referred to as diastematomyelia. The two hemi-cords are usually symmetric, although the length of separation is variable. There are two types of diastematomyelia. In type 1, the two hemicords are located within
individual dural tubes separated by an osseous or cartilaginous septum. In type 2, there is a single dural tube containing two hemicords, sometimes with an intervening fibrous septum. MRI shows split cord as hypointense on T1W and hyperintense on T2W images.

2.2.2.2. Disorders of notochordal formation:

2.2.2.2.1. Caudal agenesis, Tethering cord:

Refers to total or partial agenesis of the spinal column and may be associated with the following: anal imperforation, genital anomalies, renal dysplasia or aplasia, pulmonary hypoplasia, or limb abnormalities. Caudal agenesis can be categorized into two types. In type 1 (Fig. 8 and 9), there is a high position and abrupt termination of the conus medullaris. In type 2, there is a low position and tethering of the conus medullaris.

Sagittal T1W MRI images are excellent for evaluation, level, and shape of conus and cord tethering which appears hypointense on both T1W and T2W images.

2.2.2.2.2. Segmental spinal dysgenesis:

The clinical-radiologic definition of segmental spinal dysgenesis includes several entities: segmental agenesis or dysgenesis of the thoracic or lumbar spine, segmental abnormality of the spinal cord or nerve roots, congenital paraparesis or paraplegia, and congenital lower limb deformities. Three-dimensional CT reconstructions can be helpful in showing various vertebral segmentation anomalies. MRI imaging is evaluated on Sagittal, Axial, and Coronal planes of vertebral column helpful in detection of segmental anomalies.
**Fig. 1:** Prenatal MRI in 29 years old with 26 week gestation foetus. A-Sagittal T2 weighted MR image ; B- axial T2 weighted MR image . Show a large lombosacral myelomeningocele associated with Chiari II and hydrocephalus .

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Fig. 2: A and C, Sagittal T2 and T1-weighted MR image in 9 days new born. B- Axial T2-weighted MR image from the same patient shows neural placode (yellow arrow) extending above skin surface due to expansion of underlying subarachnoid space, which is characteristic of myelomeningocele.

Fig. 3: Hemimyelomeningocele A - Sagittal T2 weighted MR image in 3 days new born. B- Axial T2-weighted MR image. C- Axial CT image.

Fig. 4: Meningocele A and B, Sagittal T1 and T2-weighted MR image in 9 days new born. C- Axial T2-weighted MR image from the same patient.
Fig. 5: An 8 years old boy with hair tuft. A - Sagittal T2-weighted , B sagittal T1-weighted C sagittal T1-weighted fat-saturated MR images show intradural lipoma (arrow), which is hyperintense on T1 and T2 weighted image; hypointense on T1-weighted fat-saturated image.
**Fig. 6:** Neuroenteric cyst A 9 years old boy with scoliosis. A, B sagittal T2 and T1 WI; C-Focused image on the tajet. 02 cavitis communicating with a tajet.

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**Fig. 7:** A 6 years old girl with scoliosis Type 1 Diastematomyelia. A -coronal T2-weighted MR image; B-axial T2-weighted MR image show splitting of the cord into two hemicords with an intervening bony spur, which is characteristic for type 1 diastematomyelia.

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Fig. 8: A 9 days new born A, B sagittal T2 and T1 WI Abrupt termination of the conus medullaris.

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Fig. 9: Same patient in Fig.8 Kidneys in horseshoe
Conclusion

Spinal dysraphism results from defective closure of the neural tube early in fetal life and anomalous development of the caudal cell mass. Some forms of spinal dysraphism may cause progressive neurologic deterioration and exact an enormous emotional and economic toll on families and health care systems in both developed and developing countries.

Imaging has a big role in early diagnosis and the prognosis of the different forms.
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