

# Role of MRI in the Evaluation of Spinal Dysraphism

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## **Abstract:**

### **Aim:**

1. To study the role magnetic resonance imaging (MRI) for the evaluation of spinal dysraphism.
2. To identify and classify the different spectrum of lesions of spinal dysraphism.

### **Materials and Methods:**

The above study was retrospective, done for a period of 15 months from July 2013 to September 2014. Clinical and radiological findings of 20 cases suspected as Spinal dysraphism were studied and identified different spectrum of dysraphism and classified them. Clinical evaluation of 20 patients was studied and following characteristics MRI findings were evaluated.

**Results:** A total of 20 cases (09 males and 11 females)

## **I. INTRODUCTION:**

Spinal and spinal cord malformations are collectively named spinal dysraphisms. Spinal dysraphism is one of the most common congenital disorders associated with significant morbidity and mortality with estimated incidence of approximately 0.05 to 0.25 per 1000 births [1]. Spinal dysraphisms are categorized into open spinal dysraphisms (OSDs), in which there is exposure of abnormal nervous tissues through a skin defect, and closed spinal dysraphisms (CSD), in which there is a continuous skin coverage to the underlying malformation. Open spinal dysraphisms basically include myelomeningocele and other rare abnormalities such as myelocele and hemimyelo(meningo)cele. Closed spinal dysraphisms are further categorized based on the association with low-back subcutaneous masses. Closed spinal dysraphisms with mass are represented by lipomyelocele, lipomyelomeningocele, meningocele, and myelocystocele. Closed spinal dysraphisms without mass comprise simple dysraphic states (tight filum terminale, filar and intradural lipomas, persistent terminal ventricle, and dermal sinuses) and complex dysraphic states. The latter category further comprises

with age range from 2 months to 45 years were studied in the Department of Radiodiagnosis, Bangalore Medical College and research institute, Bangalore. All the patients underwent Multiplanar MRI examination. Two cases had myelomeningocele, 2 cases had lipomyelomeningocele, 1 case had lipomyelomeningocele, 2 cases had caudal agenesis, 3 cases had tethered cord, 2 cases had diastematomyelia, 1 case had dorsal dermal sinus, 1 case had filar lipoma, 1 case had intradural lipoma and 3 cases had segmental spinal dysgenesis

### **Conclusion :**

1. MRI is the modality of choice for the evaluation of spinal dysraphism.
2. MR classification of spinal dysraphism helps in surgical planning and prognostication.

**Key words:** Congenital, Dysraphism, MR imaging, Notochord, Spinal cord.

defects of midline notochordal integration (basically represented by diastematomyelia) and defects of segmental notochordal formation (represented by caudal agenesis and spinal segmental dysgenesis).

The present article is aimed at documenting various Magnetic resonance imaging findings in the investigation of this complex group of disorders. The article stresses to improve their diagnoses and to increase the amount and accuracy of information that is made available to clinicians involved in the management of children harbouring these disorders.

## **II. MATERIALS AND METHODS**

The above study was retrospective, done for a period of 15 months from July 2013 to September 2014. Clinical and radiological findings of 20 cases suspected as Spinal dysraphism were studied and identified different spectrum of dysraphism and classified them. Relevant history age, sex, birth history, developmental history, presenting complaints and radiological findings were recorded.

The MRI was performed with Siemens 1.5 Tesla and acquisition of images done. One of the biggest challenges of paediatric neuroimaging is the acquisition

of high-quality diagnostic images, as it requires the infant or child to keep still for a long period of time (sometimes nearly 1 h). Neonates under 2 months old (corrected age, if premature) are scanned during natural sleep induced by food, comfort and warmth (feed and wrap), often best after a period of sleep deprivation. The majority of our patients (approximately 70%) require sedation or a general anaesthetic[2].

Cardiorespiratory monitoring with MR-compatible equipment is essential in all infants, whether sedated or not, and all sedated or anaesthetized children [3]. The structures we need to examine in children are generally small and we aim for a maximum slice thickness of 5 mm in the brain and 3 mm in the spine. The slice thickness is reduced to 3 mm when acquiring images of the pituitary gland and orbits.

**Spine:** MRI is the modality of choice for imaging the intraspinal components of the paediatric spine. Prior to ossification of the posterior elements of the spinal column, US has been shown to be a valuable screening tool; however, infants with an abnormal sonogram or who have a neurological abnormality in the context of a normal sonogram, still require MR imaging [4].

Our standard spine imaging includes sagittal, fast spin-echo T1- and T2-W sequences (3-mm-thick slices). Both axial T1-W and T2-W images are acquired through any abnormality. Unlike most adult spine imaging protocols, groups of axial images through disc levels are not applied because degenerative disc disease is rare. Children with scoliosis and/or suspected spinal dysraphism routinely have axial T1-W images through the conus and filumterminale to detect lipomas of the filumterminale that may not be visible on sagittal imaging.

Clinical-neuroradiological classification of spinal dysraphisms

- Sagittal T1-weighted image (WI) : craniospinal axis, delineate vertebral body marrow, cord size, lipomatous tissue.
- Sagittal, axial and coronal T2WI: cord parenchyma, delineate cerebrospinal fluid (CFS), extradural interface, associated intraspinal masses, fetal evaluation..
- Gradient echo (GRE) in cervical region.
- Sagittal and axial planes. Coronal planes are useful in diastematomyelia.

### III. DISCUSSION

#### Spinal Cord Development: Embryology

Spinal dysraphism includes wide spectrum of congenital disorders caused by incomplete or abnormal closure of neural tube during early embryogenesis. Due to this, fusions of midline spinal elements are incomplete or absent. These defects give rise to open or close neural tube defect.

#### Classification of spinal cord dysraphism:

MRI provides an accurate and non-invasive method for evaluation of Spinal dysraphism, making it the modality of choice. The excellent contrast resolution, wide field of view and multiplanar images allow evaluation of entire spinal cord, contents of back mass, detect cord tethering, associated syringomyelia, chiari malformations and other abnormalities. Spinal dysraphisms can be broadly categorized into open and closed types [5, 8]. 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 [9].

#### Open spinal dysraphisms:

Myelomeningocele · Myelocele  
· Hemimyelomeningocele · Hemimyelocele

**Closed spinal dysraphisms With subcutaneous mass : Lumbosacral**

lipomas with dural defect — Lipomyelomeningocele — Lipomyelocele

· Terminal myelocystocele · Meningocele

Cervico-thoracic · Myelocystocele

skin-covered myelomeningocele (limited dorsal myeloschisis)

· Meningocele

**subcutaneous mass Simple dysraphic states** · Intradural lipoma

· Filar lipoma

· Tight filum terminale

· Persistent terminal ventricle · Dermal sinus

Complex dysraphic states

Disorders of midline notochordal integration

a) Diastematomyelia b) Neurenteric cysts

c) Dorsal enteric fistula

Disorders of notochordal formation a) Caudal agenesis

b) Segmental spinal dysgenesis

## 1. OPEN SPINAL DYSRAPHISMS

### 1.1 Myelomeningocele and myelocele:

Myelomeningoceles and myeloceles are caused by defective closure of the primary neural tube and are characterized clinically by exposure of the neural placode through a midline skin defect on the back. Myelomeningoceles account for more than 98% of open spinal dysraphisms [5]. Myeloceles are rare. Open spinal dysraphisms are often diagnosed clinically, so imaging is not always performed. When imaging is performed, the main differentiating feature

between a myelomeningocele and myelocele is the position of the neural placode relative to the skin surface [7]. The neural placode protrudes above the skin surface with a myelomeningocele (Fig. 2) and is flush with the skin surface with a myelocele (Fig. 3). The lesions usually appear hypointense on T1W images and hyperintense on T2W images similar to CSF. They have complex signal intensity when there is hemorrhage or if the cyst becomes infected.

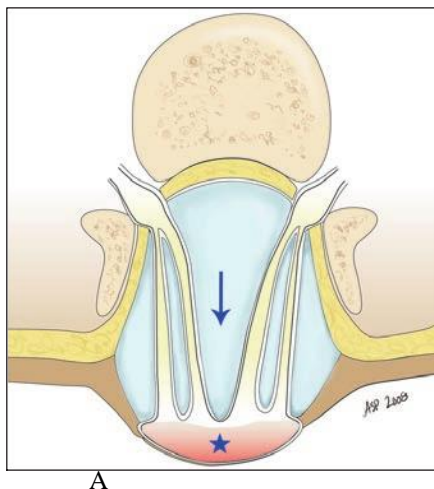
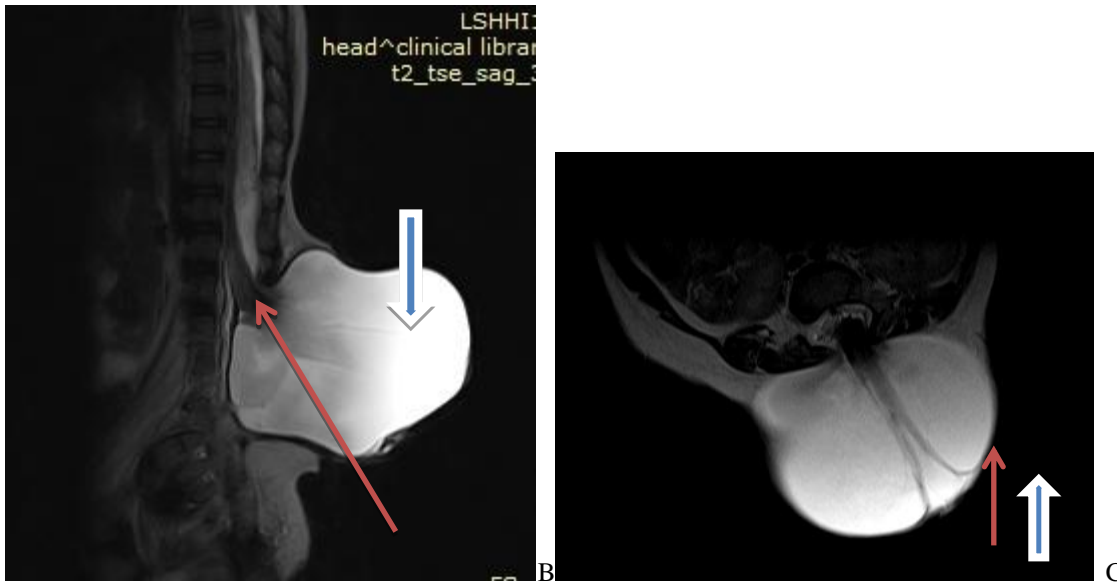


Fig. 2: Myelomeningocele.

A, Axial schematic of myelomeningocele shows neural placode (star) protruding above skin surface due to expansion of underlying subarachnoid space (arrow). B, Axial T2-weighted MR image in 3 year old girl shows neural placode (red arrow) extending above skin surface due to expansion of underlying subarachnoid space (white arrow), which is characteristic of myelomeningocele. C, Sagittal T2-weighted MR image from same patient as in B with myelomeningocele shows neural placode (white arrow) protruding above skin surface due to expansion of underlying subarachnoid space (black arrow).



### 1.3. Hemimyelomeningocele and hemimyelocoele:

Hemimyelomeningocele and hemimyelocoeles can also occur but are extremely rare [9]. These conditions occur when a myelomeningocele or myelocoele is associated with diastematomyelia (cord splitting) and one hemicord fails to neurulate.

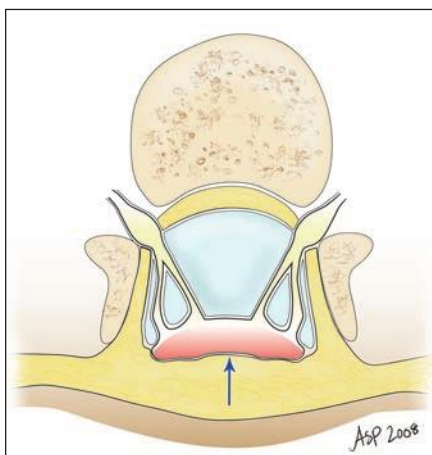
## 2. CLOSED SPINAL DYSRAPHISMS

### 2.1. Closed Spinal Dysraphisms With a Subcutaneous Mass

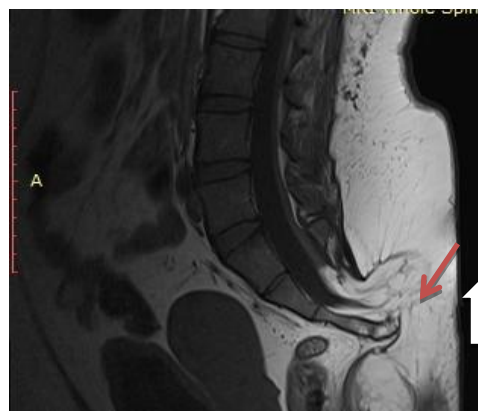
#### 2.1.1 Lipomas with a dural defect:

Lipomas with a dural defect include both lipomyelocoeles and lipomyelomeningocele. These

abnormalities result from a defect in primary neurulation whereby mesenchymal tissue enters the neural tube and forms lipomatous tissue [10]. Lipomyelocoeles and lipo-myelomeningocele are characterized clinically by the presence of a subcutaneous fatty mass above the intergluteal crease. The main differentiating feature between a lipomyelocoele and lipomyelomeningocele is the position of the placode-lipoma interface [9]. With a lipomyelocoele, the placode-lipoma interface lies within the spinal canal (Fig. 3). With a lipomyelomeningocele, the placode-lipoma interface lies outside of the spinal canal due to expansion of the sub-arachnoid space (Fig. 4). MRI imaging shows mass appears as hyperintense on T1W images and shows fat suppression.



A

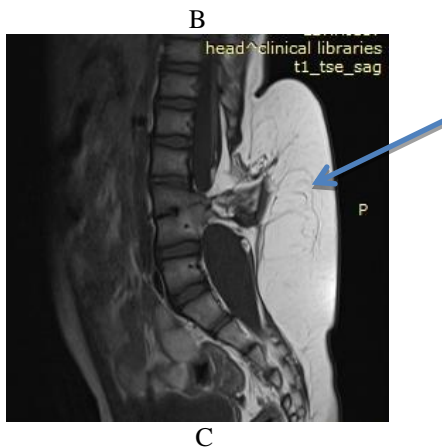
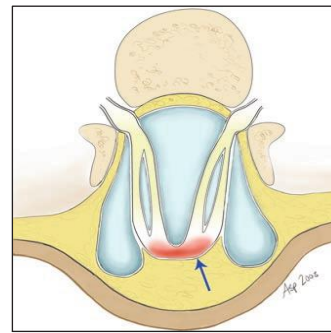


B

**Fig. 3: Lipomyelocoele.**

A, Axial schematic of lipomyelocoele shows placode–lipoma interface (arrow) lies within spinal canal.

B, Sagittal T1-weighted MR image with lipomyelocoele shows subcutaneous fatty mass (red arrow) and placode–lipoma interface (white arrow) within spinal canal.

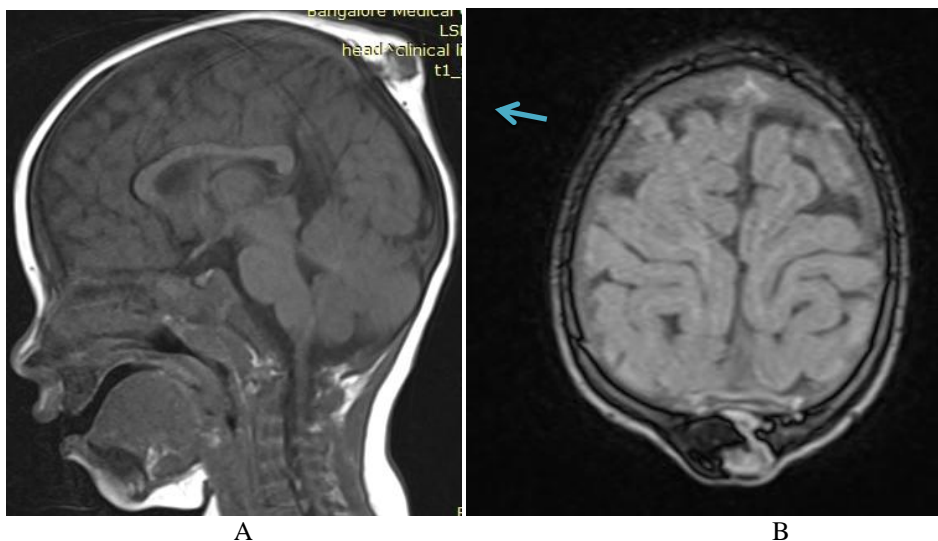


**Fig. 4: Lipomyelomeningocele.**  
 A, Axial schematic of lipomyelomeningocele shows placode–lipoma interface (arrow) lies outside of spinal canal due to expansion of subarachnoid space. B, Axial and C. Saggital T1-weighted MR image in 11 year girl shows lipomyelomeningocele (arrow) showing location of placode–lipoma interface outside of spinal canal due to expansion of subarachnoid space.

**2.1.2. Meningocele:**

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 (Fig. 5). 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 [7]. On MRI imaging shows hypointense mass on T1W image and hyperintense on T2W images.



**Fig. 5: Meningocele.**  
 Sagittal T1-weighted (A) and axial (B) T1-weighted post contrast MR images in 11 month-old girl shows small parietal meningocele (arrows).

### 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 [6]. 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 [12].

### 2.1.4. Myelocystocele:

A nonterminal 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 [13].

## 2.2 Closed Spinal Dysraphisms Without a Subcutaneous Mass:

Closed spinal dysraphisms without a subcutaneous mass can be subcategorized into simple

and complex dysraphic states.

### 2.2.1. Simple dysraphic states:

Simple dysraphic states consist of intradural lipoma, filar lipoma, tight filum terminale, persistent terminal ventricle, and dermal sinus.

### 2.2.2. Intradural lipoma:

Lipoma located along the dorsal midline that is contained within the dural sac (Fig. 6). No open spinal dysraphism is present. Intradural lipomas are most commonly lumbosacral in location and usually present with tethered-cord syndrome, a clinical syndrome of progressive neurologic abnormalities in the setting of traction on a low-lying conus medullaris [7]. On MR imaging, lipoma appears as a mass which follows signal intensity of subcutaneous fat on all pulse sequences i.e. high signal intensity on T1W and FSE T2W images, hypointense on fat saturated images.



**Fig. 6: Intradural lipoma.**

A and B, Sagittal T1-weighted (A) and sagittal T2-weighted fat-saturated (B) MR images show large intradural lipoma (arrows), which is hyperintense on T1-weighted image and hypointense on T2-weighted fat-saturated image.

### 2.2.3. 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 (Fig. 7). Filar lipomas can be considered a normal variant if there is no clinical evidence of tethered-cord syndrome [14, 15].

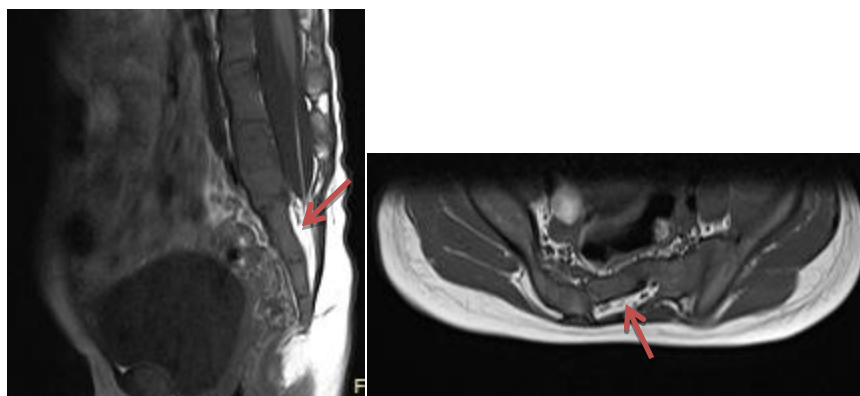


Fig. 7: Filar lipoma. A and B, Sagittal (A) and axial (B) T1-weighted MR images in 4-year-old girl with filar lipoma (arrows), which as characteristic T1 hyperintensity and marked thickening of filum terminale.

AB

#### 2.2.4. Persistent 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 [16].

at the level of the cauda equina or conus medullaris (Fig.8). Clinically, patients present with a midline dimple and may also have an associated hairy nevus, hyperpigmented patch, or capillary hemangioma [17]. 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.

#### 2.2.5. 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

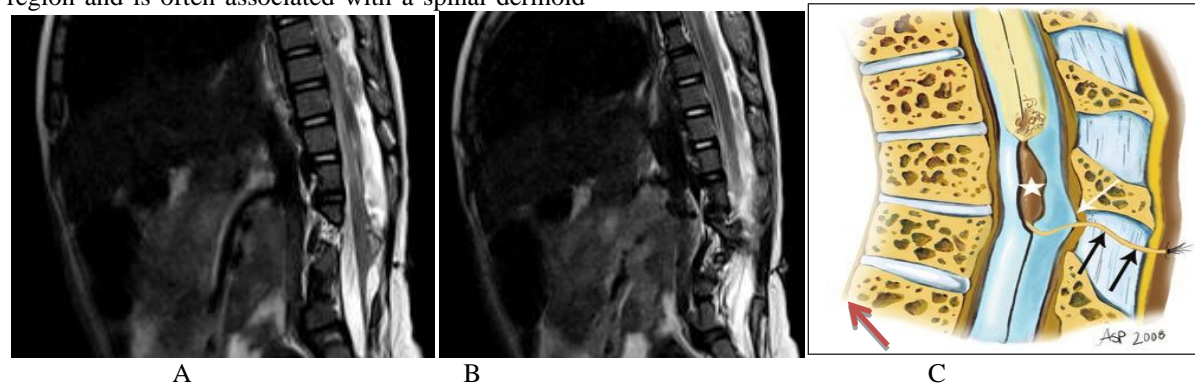


Fig. 8: Dermal sinus : Sagittal schematic (A) and sagittal T2-weighted (B) and (C) MR images in 12-month-old boy shows dermal sinus with tract extending from central canal to skin surface (red arrows). Note tenting of dural sac at origin of dermal sinus (blue arrows).

#### 2.2.6. Complex dysraphic state :

Complex dysraphic states can be divided into two categories: disorders of midline notochordal integration, which include dorsal enteric fistula, neurenteric cyst, and diastematomyelia, and disorders of notochordal formation, which include caudal agenesis and segmental spinal dysgenesis.

#### 2.2.6. Disorders of midline notochordal integration:

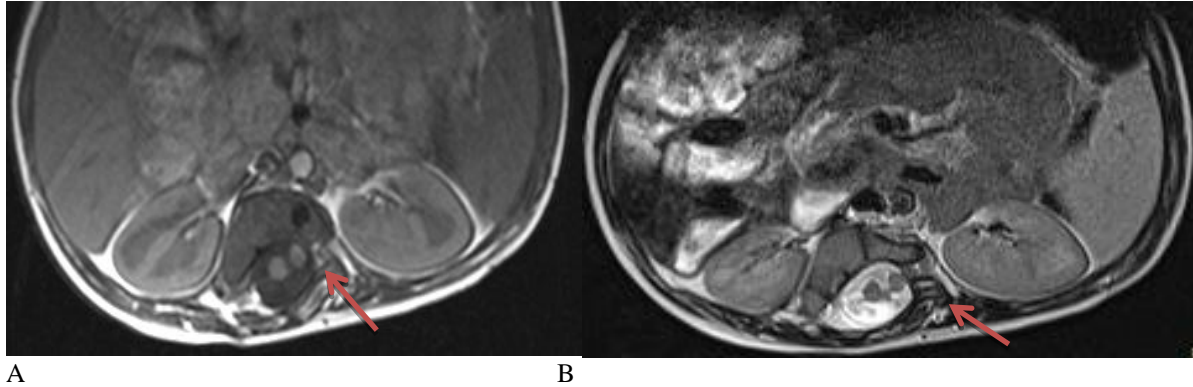
Dorsal enteric fistula and neurenteric cyst: A dorsal enteric fistula occurs when there is an abnormal connection between the skin surface and bowel. Neurenteric cysts represent a more localized form of dorsal enteric fistula. These cysts are lined with mucin-secreting epithelium similar to the gastro-intestinal tract and are typically located

in the cervicothoracic spine anterior to the spinal cord [18].

### 2.2.7. Diastematomyelia :

Separation of the spinal cord into two hemicords is referred to as diastematomyelia. The two hemicords 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 [19] (Fig. 13). Diastematomyelia can present clinically with scoliosis and tethered-cord syndrome. A hairy tuft on the patient's back can be a distinctive finding on physical examination [20]. MRI shows split cord as hypointense on T1W and hyperintense on T2W images.



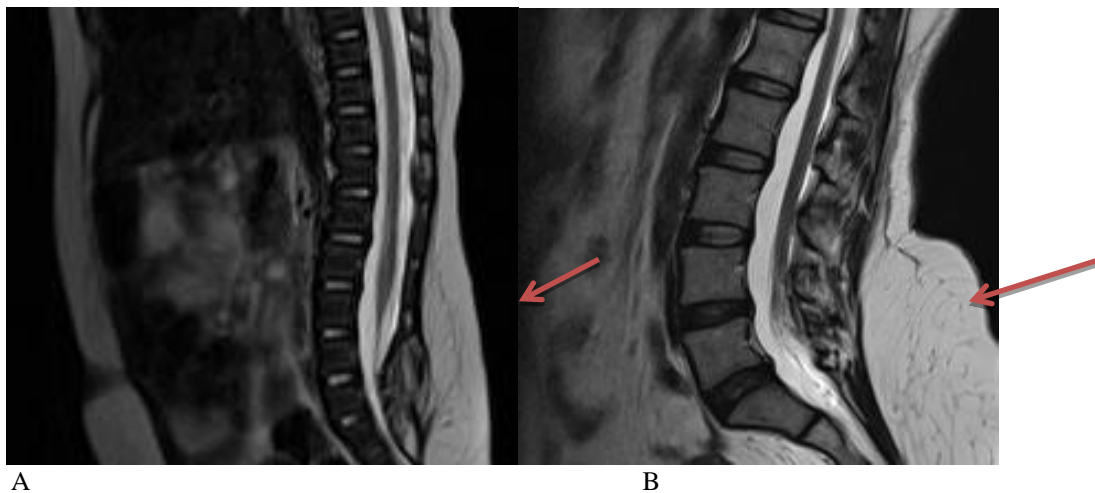
**Fig. 9: Type 2 Diastematomyelia.**

A and B, Axial T1-weighted and T2-weighted MR images in 12 month old boy show splitting of distal cord into two hemicords within single dural tube, which is characteristic for type 2 diastematomyelia.

### 2.2.8. Disorders of notochordal formation: Caudal agenesis , Tethering cord:

Refers to total or partial agenesis of the spinal column (Fig. 10) 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, 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 [21]. 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.



**Fig. 10: Caudal agenesis, Tethered cord.**

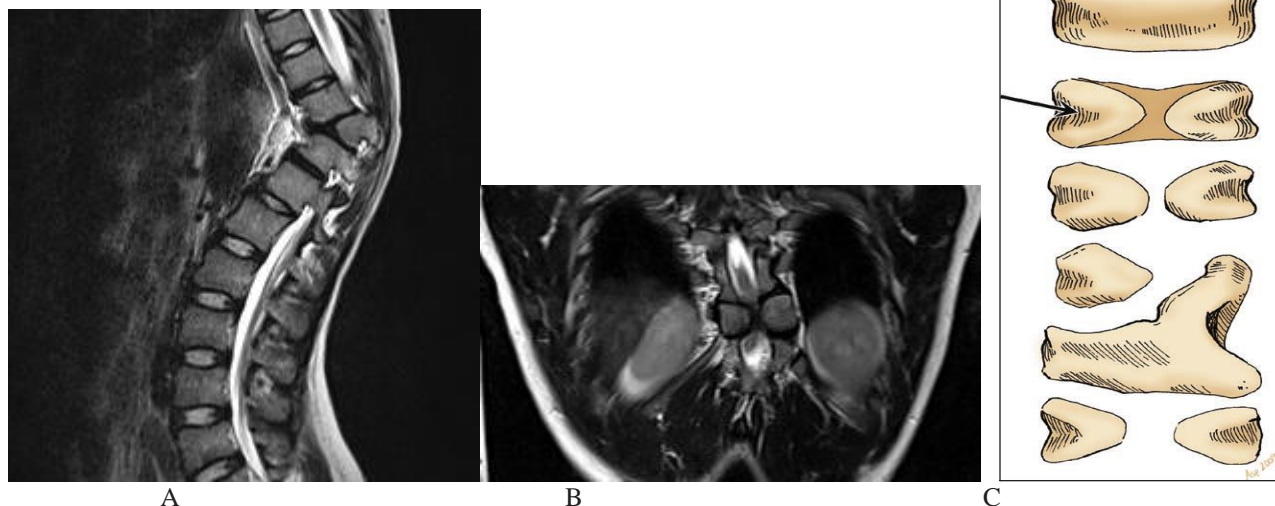
Sagittal T2-weighted (A) MR images of 2-month-old girl showing low lying of conus medullaris and (B) another image showing low position and tethering of the conus medullaris.



**2.2.9. 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 [22] (Fig. 11). MRI imaging is evaluated on Sagittal, Axial and Coronal planes of vertebral column helpful in detection of segmental anomalies.



**Fig.11:Vertebralsegmentationanomalies.**

Sagittal (A) and Coronal (B) reconstruction image in 15-year-old boy showing butterfly hemivertebra with kyphosis and schematic illustration (c) show multiple segmentation anomalies in lumbar spine (superior to inferior beginning at level of arrow): partial sagittal partition, butterfly vertebra, hemivertebra, tripedicular vertebra, and widely separated butterfly vertebra.

In our study of 20 patients with suspected congenital dysraphism werestudied in the Department of Radiodiagnosis, Bangalore Medical College and research institute, Bangalore and classified them into open and closed types.A total of 20 cases (09 males and 11 females) with age range from 2moths to 45 years were diagnosed as having Spinal dysraphism based on clininal and MRI imagings findings and confirmed through open spinal surgery, making 100% sensitivity of MRI imaging in the diagnosis of Spinal dysraphism. Out of these 20 cases, 2 (10%) had open

spinal dysraphism, with 2 cases presenting as myelomeningocele .18 cases (90%) were closed spinal dysraphism of which 2 cases had lipomeningocele, 1 case had lipomyelomeningocele, 2 cases had caudal agenesis, 3 cases had tethered cord, 2 case had diastematomyelia, 1 case had dorsal dermal sinus, 1 case had filar lipoma, 1 case had intradural lipoma and 3cases had segmental spinal dysgenesis.Results are tabulated below.

TYPE	CASE NUMBERS	PERCENTAGE
MYELOMENINGOCELE	2	10
LIPOMENINGOCELE	2	10
LIPOMYELOMENINGOCELE	1	05
DIASTEMATOMYELIA	4	20
CAUDAL AGENESIS	2	10
TEETHERED CORD	3	25
INTRADURAL LIPOMA	1	05
FILAR LIPOMA	1	05
DORSAL DERMAL SINUS	1	05
SEGMENTAL SPINAL DYSGENESIS	3	15

## V. CONCLUSIONS:

Spinal dysraphism is a heterogeneous group of developmental abnormalities in which there is defective closure of neural tube. Congenital malformations of the spine and spinal cord can be complex and variable in imaging appearance. MRI is the single imaging modality in majority of cases and classify them based on the commonly followed clinico-radiological classification proposed by Tortori et al. An organized approach to imaging findings with consideration of clinical and developmental factors allows greater ease in diagnosis.

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