



SPECTRUM OF FINDINGS OF SPINAL DYSRAPHISM ON MRI

Radiodiagnosis

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ABSTRACT

Introduction: Spinal dysraphism or spina bifida is a congenital anomaly which includes a heterogeneous group of anomalies resulting from incomplete midline closure of osseous, mesenchymal and nervous tissue. These conditions are usually diagnosed prenatally, at birth, or in early infancy; however, some may be discovered in older children or adults. The incidence of neural tube defects ranges between 1.0 and 10.0 per 1,000 births worldwide, with almost half of these cases falling under the category of spinal dysraphism and the other half consisting of anencephaly cases. The prevalence of neural tube defects (including anencephaly and spinal dysraphism) has been on the decline during the last 25 years as a result of folic acid supplementation. Although plain radiographs, Ultrasonography (primarily in antenatal detection), Myelography, and post-myelography computed tomography are helpful in the diagnosis of spinal abnormalities, Magnetic resonance imaging (MRI) has made the diagnosis of spinal dysraphism easier, faster, and more accurate, thereby enhancing the possibility of early and case-tailored treatment, mainly by its multiplanar imaging and tissue characterization capabilities.

Aims and objectives : 1. To assess the role of magnetic resonance imaging (MRI) in the evaluation of the spine and spinal cord malformations.

2. To study the MRI characteristics of the spine and spinal cord malformations.

Material and Methods: Patients who were suspected to have spinal malformations on the clinical background were subjected to MRI examination on 1.5 Tesla superconducting magnet, GE Medical systems, using Phased array spine coils. The findings of MRI spine were assessed and analysed.

Results: In the present study out of 30 patients, spinal dysraphism was most commonly seen in females, with female accounting for 21 cases and males accounting for 9 cases with female to male ratio of 2.3:1. Most cases of spinal dysraphism were seen in 1st decade of life [21 cases, 70%] followed by 2nd decade [7 cases, 23%] and rest of the 2 cases (7%) are seen in the 3rd decade. In the present study of 30 cases, 21 cases were closed dysraphisms accounting for 70% of total cases, whereas 9 cases were open type accounting for 30% of total cases. In open spinal dysraphism, meningocele is the most common ones [8 cases, 88.8%] followed by hemimeningocele [1 case, 11.1%]. Out of 22 cases of closed spinal dysraphism, diastematomyelia was the most common abnormality present in 8 cases [followed by spinal lipomas [7 cases, 25.9%]

Conclusion: Basic knowledge of embryogenesis of the spine and spinal cord is essential to understand the wide spectrum of abnormalities seen in spinal dysraphism. A rational approach focusing on a correlation among clinical, embryological, and neuroradiological data greatly facilitates the diagnosis and MRI is the single best safe modality for complete prognostication and for surgical planning.

KEYWORDS

Spinal dysraphism, MRI, Open spinal dysraphism, Closed spinal dysraphism, Meningocele, Hemimeningocele, Diastematomyelia, Arnold chiari-malformation.

INTRODUCTION:

Spinal dysraphism or spina bifida refers to a spectrum of heterogeneous congenital anomalies characterized by an incomplete fusion of the midline mesenchymal structures, bony or neural elements. These conditions are usually diagnosed prenatally, at birth or in early infancy; however, some may be discovered in older children or adults. The worldwide incidence of neural tube defects ranges between 1.0 and 10.0 per 1,000 live births, with almost half of these cases falling under the category of spinal dysraphism and the other half consisting of anencephaly cases. The prevalence of neural tube defects (including anencephaly and spinal dysraphism) has been on the decline during the last 25 years as a result of folic acid supplementation. Spinal cord development can be summarized into three basic embryologic stages - gastrulation, primary neurulation and secondary neurulation. Abnormalities in any of these stages can lead to spine or spinal cord malformations. Spinal dysraphism can be broadly categorized into open and closed types. In an open spinal dysraphism (OSD), there is a defect in the overlying skin and the neural tissue is exposed to the environment. In a closed spinal dysraphism (CSD), the neural tissue is covered by skin. Closed spinal dysraphism can be further subcategorized on the basis of the presence or absence of a subcutaneous mass. These patients present with various clinical manifestations which include cutaneous stigmata, neurological symptoms, bowel and bladder symptoms and orthopedic symptoms. Thus the evaluation and attention of these patients is not limited only to the primary lesion but also the systems outside the nervous system.

Although plain radiographs (x-rays) are helpful in the diagnosis of spinal abnormalities, sometimes patients with significant underlying disease can have post-myelography computed tomography are superior to all other techniques, but they are limited in use because of their invasive nature and recognized risks. Myelography and post-myelography computed tomography are recommended for those cases in which MRI is equivocal or incomplete. Ultrasonography is primarily useful in the antenatal diagnosis of spinal dysraphism and is

used as screening modality in the neonate and infant. There are substantial limitations in B-mode sonography such as imprecise soft tissue characterization, direct contact scanning cannot be performed on patients with exposed neural tissue or fragile meningeal sacs without incurring substantial risk and sonographic penetration of dense scar is poor. Ultrasound becomes progressively less useful as ossification of the posterior elements proceeds during the first year of life.

Magnetic resonance imaging (MRI) is a non invasive technique and has made the diagnosis of spinal dysraphism easier, faster and more accurate, thereby enhancing the possibility of an early and case-tailored treatment, mainly by its multiplanar imaging and tissue characterization capabilities. This procedure can provide a definitive diagnosis without the hazards of ionizing radiation or intrathecal injection of contrast media.

Clinico-radiological classification of spinal dysraphism

Open spinal dysraphisms	
Myelomeningocele	
Meningocele	
Hemimyelomeningocele	
Hemimyocele	
Closed spinal dysraphisms	
With subcutaneous mass	Without subcutaneous mass
Lumbosacral	Simple dysraphic states
Lipomyelocele	Posterior spina bifida
Lipomyelomeningocele	Lipoma
Meningocele	• Intradural
Terminal myelocystocele	• Intramedullary
	• Filum terminale
Cervical	Tight filum terminale
Meningocele	Abnormally long spinal cord
Myelocystocele	Persistent terminal ventricle
Myocele	
	Complex dysraphic states
	Dorsal enteric fistula
	Neurenteric cysts
	Split cord malformations
	• Diastematomyelia
	• Dileptomyelia
	Dermal sinus
	Caudal regression syndrome
	Segmental spinal dysgenesis

AIMS & OBJECTIVES:**The main objectives of this study are :**

1. To assess the role of MRI in the evaluation of spine and spinal cord malformations.
2. To study the magnetic resonance imaging characteristics of spine and spinal cord malformations.

MATERIALS & METHODS:

Study design: Hospital based prospective study

Study sample: 30 cases

Duration of Study: 2 years (2018 to 2020)

INCLUSION CRITERIA:

All clinically suspected cases of spine and spinal cord malformations.

EXCLUSION CRITERIA:

All post-operative cases

Patients who have contraindications for MRI.

EQUIPMENT AND SEQUENCES:

All MR examinations were performed using 1.5 Tesla superconducting magnet, GE Medical systems, using a phased array spine coils.

Patient position: Supine position.

Sequences: Sagittal and axial images of cervical, thoracic and lumbar regions were obtained with following sequences:

1. Sagittal T1 weighted images (TR: 427ms, TE: 30ms): Craniospinal axis, delineate vertebral body marrow, and lipomatous tissue.
2. Sagittal and axial T2 weighted images (TR: 3050ms, TE: 110ms): Cord parenchyma, delineate cerebrospinal fluid, extradural interface, and associated intraspinal masses.
3. STIR (TR: 3375ms, TE: 42ms): to confirm the presence of fat and other pathologies.

In addition coronal images were used to assess segmentation disorders with hemivertebrae or diastematomyelia. Multi-planar reconstruction was used for proper depiction of tiny structures such as a very small fistula tract.

CLINICAL SIGNS AND SYMPTOMS:

Most patients with spinal anomalies present either with external manifestations of the anomaly or with the tight filum terminale (tethered cord) syndrome. First step in the evaluation of patient with dysraphism is thorough clinical assessment. The child's back should be evaluated to look for presence/absence of skin covering, back mass and cutaneous stigmata of dysraphism. Cutaneous markers have been reported in nearly 50–80% of patients. These lesions are usually already evident at birth, which can point to the underlying pathology.

Classification of cutaneous markers based on index of suspicion of underlying spinal dysraphism.

High index of suspicion	Low index of suspicion
Hypertrichosis	Telangiectasia
Dimple (large and >2.5 cm from the anal verge)	Capillary malformation (PWS)
Acrochordons (true/pseudo tails)	Hyperpigmentation
Lipomas	Melanocytic nevus
Hemangiomas	Sacral dimple (<2.5 cm from anal verge)
Aplasia cutis or scar	
PWS - Port wine stain	

The syndrome of tight filum terminale, also known as the tethered spinal cord syndrome, denotes a complex of neurologic and orthopaedic deformities. This clinical syndrome may be associated with diastematomyelia, spinal lipoma or with a short, thick filum terminale; frequently conus medullaris is in a low position. Patients may present with new onset of symptoms at any age (As late as eighth decade).

Adults present more often with pain, possibly because of accompanying degenerative changes and less commonly with incontinence of urine, weakness and scoliosis. Indeed, it has been suggested that tethering of the spinal cord accelerates disc

degeneration.

RESULTS:**Table 1 : Gender-wise Distribution Of Spinal Dysraphism**

Sex	No. of patients	Percentage(%)
Male	9	30
Female	21	70
Total	30	100

In the present study spinal dysraphism was most commonly seen in females, with female accounting for 21 cases and males accounting for 9 cases with female to male ratio of 2.3:1.

Table 2 : Age-wise Distribution Of Spinal Dysraphism

Age group(years)	No. of patients
0-10	21
11-20	7
21-30	2
31-40	0
41-50	0
>50	0
Total	30

In the present study, most cases of spinal dysraphism were seen in 1st decade of life [21 cases , 70%] followed by 2nd decade [7 cases , 23%] and rest of the 2 cases (7%) are seen in the 3rd decade.

Table 3 : Distribution Of Spinal Dysraphism According To Its Type (clinico-radiological Classification)

Type of spinal dysraphism	No. of patients	Percentage
OSD	9	30
CSD	21	70

OSD-open spinal dysraphism, CSD-closed spinal dysraphism.

In this study of 30 cases, 21 cases were closed dysraphisms accounting for 70% of total cases, whereas 9 cases were open type accounting for 30% of total cases.

Table 4 : Distribution Of Abnormalities In Open Spinal Dysraphism (osd)

Type of OSD	No. of cases	Percentage(%)
MMC	8	88.9
Myelocele	0	0
HMMC	1	11.1
HMC	0	0
Total	9	100

MMC-mylomeningocele , HMMC-hemimeningomyelocele, HMC-hemimyocele

In this study of open spinal dysraphism, meningocele were the commonest ones [8 cases, 88.8%] followed by hemimeningomyelocele [1 case, 11.1%]. No cases of myelocele/hemimyocele were seen

Table 5 : Distribution Of Abnormalities In Closed Spinal Dysraphism (csd)

Spinal anomalies	No. of cases
DMM	8
LMMC	1
LMC	2
MC	2
Flp	4
LP	3
TFT	1
PTV	2
DDS	1
NEC	2
Total	26

DMM-diastematomyelia, LMMC-lipomeningomyelocele, LMC-lipomyelocele, MC-meningocele, Flp-filum terminale lipoma, LP-intradural lipoma, TFT-tight filum terminale, PTV-persistent terminal ventricle, DDS-dorsal dermal sinus, NEC-neurenteric cyst

In this study, out of 21 cases of closed spinal dysraphism,

diastematomyelia was the most common abnormality present in 8 cases] followed by spinal lipomas [7 cases, 25.9%]

In the present study out of 30 cases, about 24 cases (80%) were diagnosed antenatally and 6 cases (20%) were not diagnosed antenatally.

In this study, cutaneous markers were seen in 40.9% of cases [9 cases] of closed spinal dysraphism. Hypertrichosis and subcutaneous lipoma each constituted 33.3% [4 cases], haemangiomas constituted 16.6% [2 cases], dermal sinus and port wine stain constituted each of 8.3% [1 case].

In this study of 30 cases, lumbar spine (8 cases, 25.8%) was most commonly involved followed by lumbosacral spine [7 cases, 22.5%], thoracolumbar spine [6 cases, 19.3%], sacral spine [5 cases, 16.1%], thoracic spine [3 cases, 9.6%] and cervical spine [2 cases, 6.4%] in decreasing order.

In this study spinal curvature abnormalities were seen in 8 cases (26.7%).

In this study, vertebral anomalies were present in 66.66% of cases [20 cases], among which spina-bifida is the most common constituting to 70.8% [17 cases], hemivertebra and block vertebrae each constituted 12.5% [3 cases] and block vertebra constituted 4.1% [1 case].

In this study, most common spinal abnormality encountered was tethered cord [20 cases, 19.2%] followed by spinal bifida [17 cases, 16.3%], diastematomyelia and Arnold Chiari malformation [9 cases, 8.7%], scoliosis [8 cases, 7.7%], and syringohydromyelia [7 cases, 6.7%] in decreasing order of frequency.

DISCUSSION:

The term spinal dysraphism (SD) includes the overall group of defects derived from the maldevelopment of the ectodermal, mesodermal, and neuroectodermal tissues.

Spinal ultrasound (SUS) is becoming increasingly accepted as a first line screening test in neonates suspected of spinal dysraphism. Prior to ossification of the posterior elements of the spinal column, USG 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 [1]. Fetal MRI is used as complimentary modality to USG for the antenatal diagnosis of spinal anomalies and associated hydrocephalus.

Spinal dysraphism is believed to be more common in females than in males. In the present study spinal dysraphism was most commonly seen in females, with female accounting for 21 cases and males accounting for 9 cases with female to male ratio of 2.3:1.

The study by De Wals P et al (2) has shown that, about 55-70% of neural tube defects occur in females. In a study conducted by Kemal Sarica et al (3) out of 47 children enrolled 27 were girls and 20 were boys with male: female ratio was 1.3.

In the present study, spinal dysraphism was more commonly seen in younger age group (0-20 yrs). In the study by Kemal Sarica et al (3) the age range of the children with spinal dysraphism was 2 months to 16 years (mean 6.9 years).

In the study by David Guggisberg et al (4) of 14 cases of spinal dysraphism, lipoma with overlying port wine stain [35.7%] and deviation of gluteal fold [42.8%] were the most frequent lesions associated with occult spinal dysraphism. Spinal lipoma and tethered spinal cord were the most frequent spinal anomalies. Hypertrichosis and subcutaneous lipoma were the most common cutaneous markers noted which accounted for 33.3%. Hypertrichosis was present in 4 out of 9 cases of diastematomyelia [44.4%].

This is in accordance with study done by Miller et al (5) in which hypertrichosis was present in cases of spinal dysraphism in 17 patients out of 43 cases of diastematomyelia [39.5%].

In the present study, lumbar spine (n= 8, 25.8%) is the most commonly

involved followed by lumbosacral spine. Whereas in a study by Kumar R et al (6) lumbosacral region is the commonest site of occurrence of spinal dysraphism followed by lumbar region.

In the present study spinal curvature abnormalities were seen in 26.7% cases [n=8] which is in accordance with the retrospective study by Prahinski et al (7), in which 30% of patients with spinal dysraphism have congenital spinal curvature abnormalities.

In a study by Erfani et al (8) intraspinal abnormalities were present in 41.3% of congenital scoliosis.

By far the most common variety is adolescent scoliosis (9)

In the present study, of all the vertebral anomalies in patients with spinal dysraphism, spina bifida is the commonest (n=17, 70.8%), followed by hemivertebra (n=3, 12.5%) and butterfly vertebra (n=3, 12.5%) and block vertebra (n=1, 4.1%).

In a study by Tortori Donati et al (10), closed spinal dysraphism is more common than open spinal dysraphism [64.2% cases]. In our study of 30 cases, 76.7% cases are closed dysraphisms whereas 23.3% are the open type.

In the present study, meningocele were most common (87.5%) in open type followed by hemimeningocele, while diastematomyelia is the most common in closed type (29.6%).

Meningocele

In the present study meningocele is the most common open spinal dysraphism accounting for 88.9% of the cases.

In cases of meningocele in the present study, the spinal cord showed syringohydromyelia in 1 patient, and tethered cord in 5 patients, although study done by Kumar R, Singh SN et al (11) states that syringohydromyelia is common association in MMC.

Dorsal Dermal sinus:

In the present study a dorsal dermal sinus was noted in only one patient (3.7%) of all the CSDs. It was associated with low lying conus and tethered cord. (12)

Meningocele:

It is shown that meningoceles account for 2.5% of all closed spinal dysraphisms and is commonly located in the lumbar and sacral region. (13)

Meningoceles accounted for 9% [n=2] of all the CSDs. One was noted in the lower thoracic region while the other was noted in the sacral region.

Diastematomyelia:

The incidence of diastematomyelia in the present study was 30%. In the study done by Mc Comb JG et al (14) shows the incidence to be 20-40% of all cases.

In a retrospective study by Y. C. Gan et al (15) out of 17 children with diastematomyelia operated during 1989-2004, there were nine girls and eight boys. In the present study, female predominance (n=6, 66.7%) of diastematomyelia is noted with male to female ratio of 1:2.

Lipomyelomeningocele, Lipomyelocele:

Lipomyelomeningocele was detected in 1 patient in the present study and they presented with a midline back mass just above the intergluteal crease.

In other case there were tethered cord and diastematomyelia.

Lipomyelocele was detected in 2 patients in our study and both the cases were associated with cord tethering.

Tethered cord syndrome:

Spinal dysraphic lesions can anatomically tether the conus medullaris and over time it causes progressive cord ischemia and neural dysfunction (16)

In our study, tethered cord was seen in 20 cases accounting for 66% of

cases which is similar to the study done by Nishtar T et al(17) in which tethered cord was seen in 64.7% of patients.

In the present study tethered cord was most commonly associated with myelomeningocele (34%) and followed by arnold chiari malformations(20%) which is slightly different from the study by Khoshhal et al(18) in which tethered cord syndrome the most frequently associated with lipomeningomyelocele (34.3%), followed by myelomeningocele [22.8%] and diastematomyelia [11.4%].

Syringohydromyelia:

In the present study, out of total 7 cases spina bifida [5 cases,18.5%] is the most common associated abnormality with syringohydromyelia followed by tethered cord,diastematomyelia [4 cases each,14.8% each]

In a study done by Koyanagi I et al(19) out of 15 reported cases of syringohydromyelia with spinal dysraphism, 8 patients were occult spinal dysraphism (lumbosacral lipoma) and 7 were spina bifida aperta (meningomyelocele).

Chiari malformation:

In the present study , there were total 9 patients of Chiari malformation out of which 6 were females and 3 were males, with a female to male ratio of 2:1 which is in accordance with Hadley DM(20) who stated that Chiari malformations affect girls twice as often as boys.

All the patients in the present study with open spinal dysraphism had Chiari II malformation which is in accordance with TortoriDonati P et al who reported 100% association of OSDs with Chiari II malformation.(13) In one case of diastematomyelia Chiari I malformation is noted.

CONCLUSIONS AND SUMMARY:

The present prospective hospital based study of 30 patients with a clinical suspicion of spinal cord malformations had a female predominance with the age group of 0-20 years accounting for most(90%) of the cases.

Cutaneous markers were seen in 40.9% cases of the closed spinal dysraphisms with hypertrichosis and subcutaneous lipoma being the commonest cutaneous markers.

Of all the vertebral anomalies, spina bifida was most common followed by hemivertebra and butterfly vertebra in equal proportions.

The most common location for the spinal involvement was lumbar region followed by lumbosacral region.

In the present study closed spinal dysraphisms were more seen than open spinal dysraphism. The most common open spinal dysraphism observed was a meningocele .

The most common closed dysraphism observed in this study is diastematomyelia which had a female preponderance. Diastematomyelia was found to be more commonly associated with vertebral anomalies followed by tethered cord.

The associated abnormalities studied were: Tethered cord, syringohydromyelia and Chiari malformation. Tethered cord was most commonly associated with meningocele followed by diastematomyelia and spinal lipomas.

Syringohydromyelia was most commonly associated with spina bifida, followed by abnormal curvature, diastematomyelia, and tethered cord.

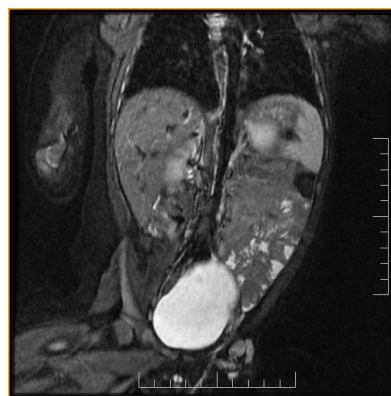
All the cases of open spinal dysraphism were associated with Chiari malformations while only one case of closed spinal dysraphism was associated with Chiari malformation.

IMAGING FINDINGS:

CASE OF MENINGOMYELOCELE: A 11 yr old male child with swelling in the lumbosacral region(L5-S1 level), A) Axial T2 B) Coronal STIR C) Sagittal T2 weighted images shows lumbo-sacral meningocele with herniation of the cauda equina nerve roots above the skin surface with tethered cord seen at L5 level.



A



B

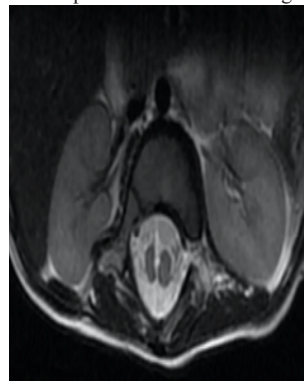


C

CASE OF DIASTEMATOMYELIA:

A 2 year male child with history of kyphoscoliosis,

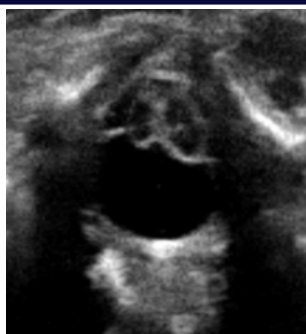
A) Axial T2 weighted image shows two hemicords enclosed in a single dural sac with no intervening septum. B) Coronal STIR image shows hemivertebra from D8 to L1. C) Corresponding ultrasound image shows split cord enclosed in a single dural sac.



A



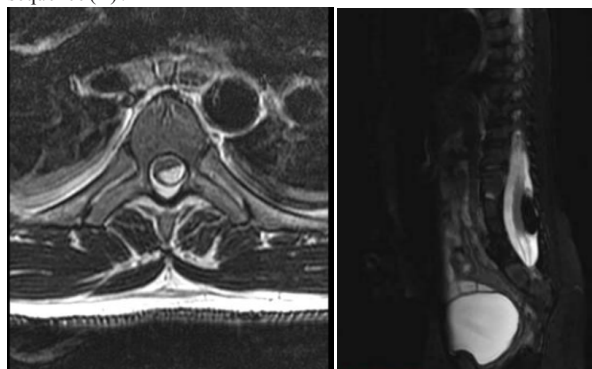
B



C

CASE OF INTRADURAL LIPOMA:

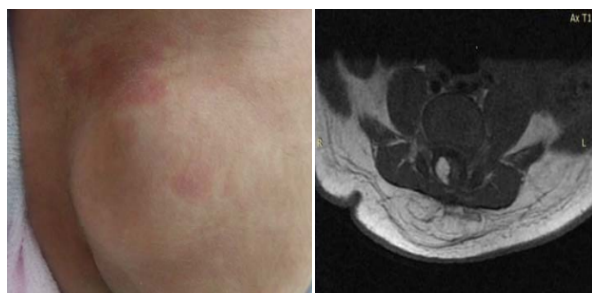
(A) Axial T1 weighted image showing hyperintense lesion posterior to the spinal cord in the intradural location with signal loss on STIR sequence (B).



A

B

CASE OF LIPOMYELOCELE: A 15 year female with A) Subcutaneous swelling and port wine stain at the lumbar vertebral level , B) axial T1 weighted image intraspinal lipomatous tissue s/o lipomyelocele



A

B

CASE OF MENINGOMYELOCELE:

A 30 yr old female patient with swelling in the lumbosacral region A) Sagittal STIR B) Sagittal T2 C) Axial T2 weighted images shows lumbo-sacral meningocele with midline cleft in spinous process at L5 level and herniation of the nerve roots above the skin surface , tethered cord seen at L5 level .



A

B



C

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