# Spinal Dysraphism: A Three Year Experience at Armed Forces Institute of Radiology and Imaging

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## ABSTRACT

***Objective:*** To review all the patients of spinal dysraphism referred to our center over a three year period in order to identify the most typical neuro-radiological appearances on Magnetic Resonance Imaging.
***Study Design:*** Cross sectional study.
***Place and Duration of Study:*** Armed Forces Institute of Radiology and Imaging, Rawalpindi from Jan 2016 to Dec 2018.
***Methodology:*** MR spine reports of 144 patients of spinal dysraphism were retrospectively analyzed. Age, gender, indication for MRI, operative status, and neuro-radiological features (including site and type of lesion) were recorded for these patients.
***Results:*** Congenital spinal malformations were more frequent among females87 (60.4%) and between 0-20 years 135 (93.9%) of age. Tethered cord 97 (67.4%) was the most common congenital spinal abnormality followed by spina bifida, diastematomyelia, vertebral segmentation anomalies, myelomeningocele, menigocele. Lip-myelomeningocele, lipoma of filum terminale, and sacral agenesis. Frequently observed associated abnormalities included scoliosis 61 (42.4 %), syrinx 47 (32.6%) and dural ectasia 40 (27.8 %).
***Conclusion:*** Congenital spinal malformations are usually complex with variable radiological appearances. Modern high resolution MRI screening is the examination of choice for identification, preoperative evaluation, and long term follow up of such congenital anomalies.

**Keywords:** Diastematomyelia, Myelomeningocele, Spinal dysraphism, Tethered cord.

***How to Cite This Article:*** *Nasir H, Nisar U, Slehria AUR, Iftihar A, Halim A, Nisar S. Spinal Dysraphism: A Three Year Experience at Armed Forces Institute of Radiology and Imaging. Pak Armed Forces Med J 2022; 72(Suppl-2): S150-154. DOI: https://10.51253/pafmj.v72iSUPPL-2.3309*



## INTRODUCTION

Spinal dysraphism refers to a spectrum of congenital malformations due to defective fusion of neural tube during embryological development of spine and spinal cord.[1,2] These spinal anomalies are broadly classified as open and closed types. Open spinal dysraphism is associated with a defect in the overlying skin through which neural tissue is exposed to the environment. In closed form, the neural tissue is covered by intact skin. Both forms are associated with varying degrees of neural tissue involvement.[2,3]

Open spinal dysraphism includes myelocele, myelomeningocele, hemimyelocele, and hemimyelomeningocele. Closed spinal dysraphism is further classified into two groups; those with an associated subcutaneous mass (lipomyelocele, lipomyelomen-ingocele, meningocele, myelocystocele) and those without an associated mass (such as filar lipoma,tethered cord (tight filum terminale), dorsal dermal sinus, diastematomyelia, caudal agenesis, and segmentation anamolies).[1,4]

Spinal dysraphism has an estimated incidence of 1-3/1000 live births.[2,5] The incidence has decreased significantly in the last few decades due to folic acid supplementation in pregnant females, antenatal screening with high resolution ultrasound, and availability of biochemical markers.[2,5] However the health problems in surviving children continue to be a challenge, particularly in the resource-poor developing countries.[6]

Plain X-ray cannot exclude spinal dysraphism. Spinal ultrasonography is possible in the new born, before the age of six months, while the posterior cartilaginous elements have not yet ossified.[7] Sono-graphy can be performed at any age in cases of a persistent posterior spinal defect.[7] Although most live cases are diagnosed at birth or early in the life, some occult forms may not be detected until adulthood.[5,8]

Traditionally,patients with suspected spinal malformations have been evaluated with contrast enhanced computed tomography (CT) and CT myelography. These techniques are invasive with the recognized risk of ionizing radiation.[7]

Magnetic resonance imaging (MRI) is superior to CT myelography and contrast enhanced CT due to its multiplanar capabilities, superior soft tissue characterizationand lack of ionizing radiation.[8] Availability of

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*Received: 24 Sep 2019; revision received: 18 Jan 2020; accepted: 20 Jan 2020*

MRI has facilitated early identification of occult variety, visualization of other associated anomalies in both open and closed forms, understanding of their clinical consequences, and provision of individually tailored management plans.[2,9,10]

The objective of this study was to review all the patients of spinal dysraphism referred to our center over a three year period in order to identify the most typical neuroradiological appearances on MRI.

## METHODOLOGY

This cross sectional study was performed at Armed Forces Institute of Radiology and Imaging, Rawalpindi after approval from institutional review board (IERB approval certificate number 001). All spinal MRI studies performed over three year period between January2016 and December2018 and relevant radiology reports were screened for features of spinal dysraphism.

**Inclusion Criteria:** Patients with congenital spinal anomalies were included in the study.

**Exclusion Criteria:** Non-consenting patients were excluded**.**

A total of 144 patients with congenital spinal anomalies were evaluated using universal sampling method.MR imaging was performed on 1.5T GE,1.5T Toshiba or 3.0T Seimens with the patient lying supine during the procedure. Imaging was done with an FOV of 300-500, matrix size of 256 x 256 and a slice thickness of 3-4 mm.Sagittal T1-and T2-weighted sequences were performed for all cases. These sequences can be performed with or without fat suppression; fat suppressed sequences being more helpful in the evaluation of fatty lesions (such as, intradural lipoma, lipoma of filum terminale). Axial acquisitions with the angle of axial sections parallel to vertebral discs were also acquired.All images were viewed on high resolution monitors and were read by certified radiologists with several years' experience in spinal MRI.

Data was evaluated retrospectively for all patients. Age, gender, indication for MRI, operative status, and neuroradiological features (including site and type of lesion) were recorded.Based on the presence or absence of intact overlying skin, patients were divided into two categories: open spinal dysraphism, and closed spinal dysraphism.

Data was entered and analyzed using SPSS v20.0. Descriptive statistics were used to describe the basic features of the data and associated anomalies.

## RESULTS

The mean age of patients was 6.5 ± 9.94 (range 01 month-90 years) with 57 (39.6%) males and 87 (60.4%) females. 14 (9.7%) patients had a surgery performed on them previously. Congenital spinal malformations were more frequent1 35 (93.9%) between 0–20 years of age.

Dysraphic spinal anomalies without subcutaneous masses 84 (58.3%) were more common than congenital lesions withsubcutaneous masses 60 (41.7%). Over all, tethered cord was the most common congenital spinal lesion observed in 97 (67.4 %) patients followed by spina bifida, diastematomyelia, vertebral segmentation anomalies, and myelomeningocele (Figure-1).

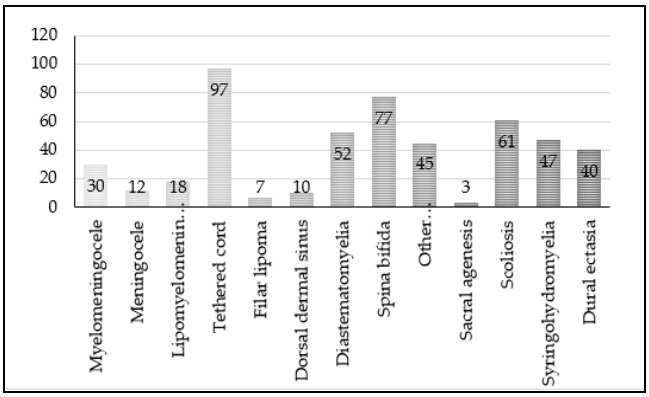


**Figure-1: Distribution of congenital anomalies in patients with congenital spinal lesions. Lumbar spine was the commonest location for congenital spinal lesionsfollowed by thoracolumbar, isolated thoracic and isolated sacral involvements.**

Scoliosis was the most common associated spinal abnormality. 61 (42.4%) of the cases with congenital malformations were associated with spinal curvature abnormality while 83 (67.4 %) were without any curvature abnormality. Other frequently observed associated abnormalities included syringohydromelia 47 (32.6 %) and dural ectasia 40 (27.8 %).

Among the 30 cases of myelomeningocele, 25 (83.3%) were associated with tethered cord, 20 (66.7%) with scoliosis, 5 (16.7%) with diastematomyelia, 4 (13.3%) cases of dural ectasia and 3 (10.0%) with syrinx formation. Lipomeningocele was associated commonly with tethered cord (83.3%), scoliosis (38.9%), and dural ectasia (38.9%). Similarly, most common associations with meningocele were tethered cord (66.7%) and scoliosis (66.7%).

The commonest vertebral anomaly was spina bifida (53.5%). Spina bifida was commonly associated

with tethered cord (76.6%), syringohydromyelia (46.8%), myelomeningocele (31.2%), diastematomyelia (28.6%), and dural ectasia (27.3%) (Figure-2).

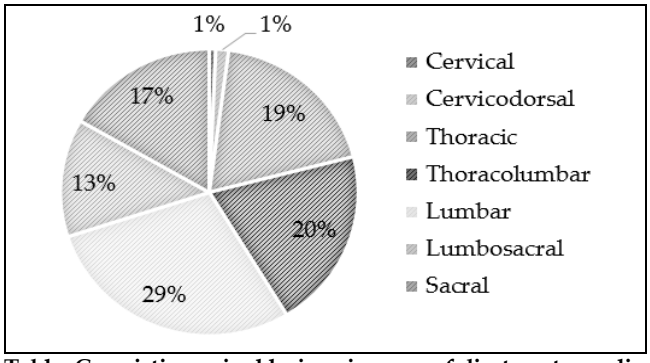


**Table: Co-existing spinal lesions in cases of diastematomyelia.**

Among the patients with diastematomyelia, Type II (63.5%) was more common than Type I (36.5%).Split cord malformations were more common in lumbar spine (38.5%) followed by thoracolumbar (36.5%) and isolated thoracicinvolvement (17.3%). The two types of diastematomyelia were also compared for co-existing spinal lesions (Table).

**Table: Co-existing spinal lesions in cases of diastematomyelia.**

| Co-existingSpinal Lesions | Diastematomyelia | |
|---|---|---|
| | Type I (n=19) | Type II (n=33) |
| Tethered cord | 18 (94.7%) | 25 (75.8%) |
| Spina bifida | 9 (47.4%) | 13 (39.4%) |
| Segmentation anomalies | 11 (57.9%) | 12 (36.4%) |
| Syringohydromelia | 6 (31.6%) | 13 (39.4%) |
| Myelomeningocele | 2 (10.5%) | 3 (9.1%) |
| Meningocele | - | 3 (9.1%) |
| Lipomyelomeningocele | - | 4 (12.1%) |

Dorsal dermal sinus was noted in 10 patients; 8 were associated with vertebral segmentation anomalies and 4 with scoliosis.[7] cases of lipomatous filum terminale were reported; spina bifida of the lumbosacral region was noted in 6 and vertebral segmentation abnormalities in 5 of these subjects. Among the three cases with sacral agenesis, a coexisting tethered cord was reported in one and vertebral segmentation anomaly in two subjects.

The comparison ofcongenital spinal lesions among the two genders revealed that vertebral segmentation anomalies (75.6%),distematomyelia (69.2%), tethered cord (62.9%), spina bifida (55.8%), lipomyelomeningocele (72.2%), and myelomeningocele (56.6%) were more common among females. Equal number of cases with meningocele and dorsal dermal sinus were observed among the male and female subjects.

## DISCUSSION

Spinal dysraphism is a group of congenital spinal malformations with a wide range of neororadiological features.[9] The malformations range from mild asymptomatic disorders such as small bony defect in the spinous process of fifth lumbar or first sacral vertebra (spina bifida occulta) to complex dysraphic states with multiple coexisting lesions.[4]

Congenital spinal anomalies are relatively more common among the female population as reported in studies conducted in India,[7] China,[10] and Turkey[12]. Our demographic analysis also showed a female preponderance for most anomalies except for cases of meningocele and dorsal dermal sinus. A higher proportion of patients with meningocele and dorsal dermal sinus had a male gender.Most cases of congenital spinal malformations present early in life either with a neurocutaneous stigmata or with a neurological deficit,[13,14] as shown in a study by Huang *et al* in which majority of the cases presented under 6 years.[12] Another study by Kumar *et al* reported mean age in their study as 5.7 years.[13] In our study, congenital spinal disorders were also more frequent in younger age group with 93.9% patients under 20 years of age. We observed lumbar spine as the most common location for congenital spinal lesions. This was consistent with previous literature which also reports lumbarand lumbosa-crals-pine as the common location for most congenital spinal disorders.[10,15].

Tethered cord is visualized as a thickened filum terminal with a low lying conus medullaris (below second lumbar vertebra). Tethered cord was the most common congenital spinal lesion according to our results followed byspina bifida, and diastematomyelia. Ramacharya and coworkers,[7] reported the occurrence of tethered cord to be 68.18%, Kumar and colleagues,[13] noted it to be 65% and while Ujala and associates,[14] found it to be 75%.

In thisstudy, spina bifida was seen in 77 (53.5%) cases and was commonly associated with tethered cord andsyringohydromyelia.Similar results were observed in a study conducted in India which reported 65% of the spina bifida cases to be associated with low lying cord and 23% with syrinx.[13,16] The incidence of spina bifida reported by another study conducted in India was 46%.[5] Myelomeningocele, myelocele, and lipomyelomeningocele were relatively infrequent in our study.Common associated spinal lesions with all these dysraphic anomalieswere tethered cord and scoliosis.

Diastematomyelia, also known as split cord malformation, is divided into two types; type I is characterized by a duplicated dural sac separated by a midline cartilagenous or osseous spur and each hemicord is located individually in its own dural sac; Type II has a single dural sac containing the two hemicords occasionally with an intervening fibrous septum.[9] In our study, 52 cases of diastematomyelia were reported, an incidence of 36.1%. Similar incidence of 32% has been reported previouslyby Kumar, *et al*,[13] Most common site of lesion was lumbar spine followed by thoracolumbar and isolated thoracicinvolvement. Similar results were reported in previous studies.[10,12] One study reported thoracic spine as the more common site of split.[11] Type II was more common than Type I in current study which corresponds to a previous study conducted in Iran,[10] but was in contrast to few other studies Borkar *et al*,[11] Borcek *et al*,[12] and Kumar *et al*[13] which have reported more cases of Type I diastematomyelia. Co-existing spinal lesions included tethered cord, spina bifida, vertebral segmentation anomalies, syrinx, and myelomeningocele which were same as in literature.[11,12] 12 cases (23.1%) of diastematomyelia had an associated myelomeningocele, meningocele, or lipomyelomeningocele; level of the split in all of these cases was either the same or one to two levels above the level of spina bifida. This finding was consistent with Indian study by Kumar *et al*.[13]

Some of the less commonly observed congenital malformations in our study included dorsal dermal sinus, filar lipoma, and sacral agenesis. Dorsal dermal sinus is defined as epithelium lined tract extending inwards from the skin surface to the thecal sac.[16] A total of 10 cases of dorsal dermal sinus were reported in the current study. Lumbar and sacral spines were most frequent sites of involvement with 4 cases each at these levels. Associated anomalies seen in our study were segmentation disorders and scoliosis. Singh *et al* also reported lumbar spine as the most frequently involved location and scoliosis as the most common associated anomaly.[16] Lipoma of filum terminale is visualized as linear high signal area on T1- and T2-weighted sequences with signal loss on fat saturated sequences.[17] It was present in only 7 cases; 6 (85.7%) of which were seen in association with the spina bifida and 5 (71.4%)with cord tethering. Another series reported the incidence of lipomatous filum terminal in spina bifida cases to be 14.8%[12]. Three (2.1%) cases of sacral agenesis were reported in our analysis. Our results were comparable with the study by Kumar *et al* in which incidence of sacral agenesis was 5%,[13]. An even lower percentage of 1% was reported by Borcek *et al*.[12]

Any additional morphological abnormalities associated with the congenital spinal lesions pose a risk for further neurological deficit and have a significant impact on the postsurgical outcome.[18,19] Therefore a careful search for any such association should be undertaken. Scoliosis, syringohydromelia, and dural ectasia were frequently noted. Scoliosis (42.4%) was the most common associated spinal abnormality and this was consistent with another Turkish study in which incidence of scoliosis with congenital spinal malformations was 44.5%.[17] In the same study, second most commonas sociated abnormality was syringohy-dromelia.[18] Hydromyelia is defined as the dilatation of ependymal lined central spinal canal while syringomyelia refers to cystic dissection of the cord itself. The two are difficult to distinguish radiologically and are often termed together as syringohydromyelia or syrinx. The incidence of syrinx in our analysis was 32.6% and in all cases, syrinx was noted to be cranial to the spinal dysraphic lesion. Similar findings were reported by others.[19]

In our study,[14] patients with previous surgery had presented with progressive neurological deficits. They had been referred to our center for reevaluation of dysraphic spinal lesions and associated pathology before being subjected to a re-surgery.Therefore, it is logical to assume that a thorough screening of the entire neuroaxis before definitive surgery is crucial to adequate management of such cases.

## CONCLUSION

Congenital spinal malformations are usually complex with variable radiological appearances. Modern high resolution MRI screening is the examination of choice for identification, preoperative evaluation, and long term follow up of such congenital anomalies.

**Conflict of Interest:** None.

**Authors' Contribution**

HN: Direct contributino, UN, AURS, AI, AH, SN: Intellectual contribution.

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