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Original Article

Radiology Section

Role of Magnetic Resonance Imaging in the Evaluation of Spinal Dysraphism

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ABSTRACT

Introduction: Spinal dysraphism includes spectrum of congenital fusion anomalies of one or more dorsal midline structures including skin, subcutaneous tissue, vertebrae, meninges and neural tissue. It is due to incomplete midline closure of the bony and neural spinal tissues. Magnetic resonance imaging (MRI) is now considered to be the imaging modality of choice.

Aim: To assess the role of MRI in the evaluation of spinal dysraphism and to assess the spectrum of lesions of spinal dysraphism.

Materials and Methods: The study was performed on 30 patients suspected of spinal dysraphism, over the time period from November 2016 to May 2018 in the Department of Radiodiagnosis in Victoria Hospital and Bowring and Lady Curzon Hospital attached to Bangalore Medical College and Research Institute, Bengaluru. All patients underwent MR imaging examination performed on a SIEMENS magneto 1.5-T MR system in Victoria hospital. Radiological characteristics and clinical features were studied. Descriptive and inferential statistical analysis was made. Results on categorical measurements are presented in Number (%) and results on continuous measurements are presented on Mean±SD (Min-Max).

Results: The ages of patients included in the study were in the range of 4 months-11 years with maximum number of patients were in the age group of 1-5 years (~43.3%). Female preponderance was noted. Congenital spinal lesions without subcutaneous masses (43.3%) were more common than the lesions with subcutaneous masses (30%). Vertebral anomalies were the most common spinal anomalies in patients with congenital spinal lesions followed by spina bifida, tethered cord, scoliosis, syrinx and diastematomyelia. Of all the vertebral anomalies, spina bifida was the most common (73.3%). Lumbosacral spine was most common site of involvement in 11 cases (36.65%).

Conclusion: MRI does not involve ionizing radiation, has no biological risk, and avoids intrathecal injection of contrast media, it offers several advantages in the evaluation of children with suspected spinal dysraphism and help in accurate diagnosis.

Keywords: Diastematomyelia, Neural placode, Tethered cord

INTRODUCTION

Spinal dysraphism includes spectrum of congenital fusion anomalies of one or more dorsal midline structures including osseous, mesenchymal and nervous tissue [1,2]. Clinical description of spinal dysraphism was made by Lichtenstein BW and James CC et al., [3,4].

Early imaging approaches were based on the use of conventional Radiography, Ultrasonography (USG) and Computerized tomography (CT) [5,6]. MRI of spinal dysraphism is first described by Barnes PD et al., [7]. Spinal dysraphism is one of the most common congenital disorders associated with significant mortality and morbidity. The estimated incidence of spinal dysraphism is about 1–3/1000 live births [8]. About 55-70% of neural tube defects occur in females. Variations in prevalence based on race, ethnicity, gender and region have also been reported [9,10]. The prevalence of neural tube defects (including anencephaly and spinal dysraphisms) has been on the decline during the last 25 years as a result of antenatal screening and folic acid supplementation [10]. Majority of the closed spinal dysraphic states are asymptomatic at birth. They are suspected in the presence of high risk cutaneous markers, or when these children present with neurological deficit later in life [11].

MRI is the assessment of choice because of its better investigative performance, exceptional soft tissue characterization and importance in presurgical planning [8]. MRI is the imaging modality of choice in the diagnosis and characterization of spinal dysraphism. Rest of the modalities play a supplemental role. Antero-posterior and lateral plain radiographs are a must for evaluation of the vertebral column [12,13].

In cases of diastematomyelia, bony spur may be seen. Radiographs are used as screening examinations to guide the further imaging work-up. Ultrasonography is useful in the antenatal diagnosis of spinal dysraphism and is also of some use in the neonate and infant [14,15]. During the first year of life it becomes progressively less useful as ossification of posterior elements proceeds. Prenatal ultrasonography can detect the open widened neural arch, with flared laminae, can show the meningomyelocele sac and detect hydrocephalus and associated cranial anomalies. Direct ultrasonography of the sac in children using high frequency transducers gives information about the contents of the sac. But, for complete and detailed information, an MRI scan is often required. In cases of split cord malformation, CT is useful in demonstration of the bony spur. Prior to the advent of MRI, myelography and post-myelogram CT were used. MRI gives a non-invasive and accurate method to evaluate spinal dysraphism, thus making it the modality of choice. The excellent contrast resolution, wide field of view and multiplanar images help evaluate the entire spinal cord, contents of the back mass; detect cord tethering, associated syringomyelia. For the demonstration of syrinx and associated pathologies like dermoid and epidermoid cyst, T2W images are helpful [13,16]. Patients with spinal dysraphism can have multiple spinal anomalies. For example, a patient with myelomeningocele may have associated chiari malformation or syringohydromyelia. Fetal MRI may be used as complimentary modality to USG for the antenatal diagnosis of spinal anomalies and associated hydrocephalus [14,15].

MATERIALS AND METHODS

The present observational study was performed on 30 patients suspected of spinal dysraphism based on clinical examination, over the time period from November 2016 to May 2018; in the Department of Radiodiagnosis in Victoria Hospital and Bowring and Lady Curzon Hospital attached to Bangalore Medical College and Research Institute, Bengaluru. The permisson to conduct the study was obtained from Institutional Ethical Committe via letter number BMC/PGS/289/2016-

17 Newborn-12 years old suspected to have spinal dysraphism and patient diagnosed to have physical abnormality caused by congenital lesions of spinal cord and cauda equina were included and patients with anencephaly and patients with physical abnormality caused due to spinal neoplasms, Friedreich's ataxia, cerebral palsy, old poliomyelitis and local conditions of the feet were excluded.

All patients included in the study or their guardians were explained of the procedure and informed consent was taken from them. All patients were screened for clips, cochlear implants etc. All MR imaging examination was performed on a SIEMENS magneto 1.5-T MR system. Neonates under 2 months of age were scanned during natural sleep. Majority of our patients (approximately 70%) required sedation [17]. Cardio-respiratory monitoring with MR compatible equipment is necessary in each and every infant, whether sedated or not [18]. For imaging, the intraspinal components of paediatric spine, MRI is the imaging modality of choice. The standard spine imaging included sagittal, fast spin-echo T1- and T2-W (weighted) sequences.

Both axial T1-W and T2-W images were acquired. Group of axial images through disc level were not applied, unlike most adult spine imaging protocols, because degenerative disc disease is uncommon. Children with suspected spinal dysraphism and/or scoliosis routinely had axial T1-W images to detect lipomas of the filum terminale that may not be visible on sagittal imaging.

STATISTICAL ANALYSIS

Descriptive and inferential statistical analysis was used. Categorical measurements are presented in Number and percentages (%) and results on continuous measurements are presented in Mean±SD (Min-Max).

RESULTS

The ages of patients included in the study were in the range of 4 months-11 years. Maximum numbers of patients were in the age group of 1-5 years which consisted of 13 patients, accounting for ~43.3% [Table/Fig-1].

Age in years	No. of patients	%
0-1 year	8	26.7
1-5 years	13	43.3
>5 years	9	30.0
Total	30	100.0

[Table/Fig-1]: Age distribution of patients studied.

Out of 30 patients, 21 were female and 9 were male with strong female preponderance, Female: Male ratio 2.3:1 [Table/Fig-2].

Gender	Age in years			Total
	0-1 year	1-5 years	>5 years	
Female	7 (87.5%)	10 (76.9%)	4 (44.4%)	21 (70%)
Male	1 (12.5%)	3 (23.1%)	5 (55.6%)	9 (30%)
Total	8 (100%)	13 (100%)	9 (100%)	30 (100%)

[Table/Fig-2]: Gender distribution of patients studied.

Swelling in the back was the most common clinical presentation. Out of 30 patients, 15 had swelling in the back accounting for 50% of all cases [Table/Fig-3].

Sl. No	Clinical features	Number	Percentage (%)
1	Swelling in back	15	50
2	Hyper trichosis	1	3.33
3	Sacral dimple	1	3.33
4	Lower limb weakness	4	13.33
5	Urinary incontinence	6	20
6	Dermal sinus	3	10

[Table/Fig-3]: Cutaneous lesions distribution of patients studied.

The [Table/Fig-4] shows different types of spinal dysraphism in relation to age distribution of the patients. The [Table/Fig-5] shows different types of spinal dysraphism in relation to age distribution of the patients. Spinal dysraphism can involve any segment of spine. The most common site was the lumbosacral region accounting for 36.66% of all cases [Table/Fig-6].

Variables	Age in years			Total (n=30)
	0-1 year (n=8)	1-5 years (n=13)	>5 years (n=9)	
Spina bifida	8 (100%)	9 (69.2%)	5 (55.6%)	22 (73.3%)
Myelomeningocele	5 (62.5%)	2 (15.4%)	1 (11.1%)	8 (26.7%)
Myelocele	1 (12.5%)	1 (7.7%)	0 (0%)	2 (6.7%)
Lipomyelomeningocele	1 (12.5%)	5 (38.5%)	3 (33.3%)	9 (30%)
Lipomyelocele	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Diastematomyelia	2 (25%)	4 (30.8%)	4 (44.4%)	10 (33.3%)

[Table/Fig-4]: Different type of spinal dysraphism distribution in relation to age distribution of patients studied.

Variables	Age in years			Total (n=30)
	0-1 year (n=8)	1-5 years (n=13)	>5 years (n=9)	
Filar lipoma	0 (0%)	1 (7.7%)	3 (33.3%)	4 (13.3%)
Dorsal dermal sinus	0 (0%)	3 (23.1%)	0 (0%)	3 (10%)
Syrinx	6 (75%)	12 (92.3%)	4 (44.4%)	22 (73.3%)
Tethered cord	8 (100%)	9 (69.2%)	6 (66.7%)	23 (76.7%)
Sacral agenesis	0 (0%)	3 (23.1%)	3 (33.3%)	6 (20%)
Vertebral segmentation anomalies	2 (25%)	7 (53.8%)	3 (33.3%)	12 (40%)
Scoliosis	3 (37.5%)	8 (61.5%)	3 (33.3%)	14 (46.7%)

[Table/Fig-5]: Different type of spinal dysraphism distribution in relation to age distribution of patients studied.

Sl. No	Site involvement	Number	Percentage (%)
1	Cervical	1	3.33
2	Thoracic	2	6.66
3	Dorsolumbar	3	10
4	Lumbar	10	33.33
5	Lumbosacral	11	36.66
6	Sacral	3	10

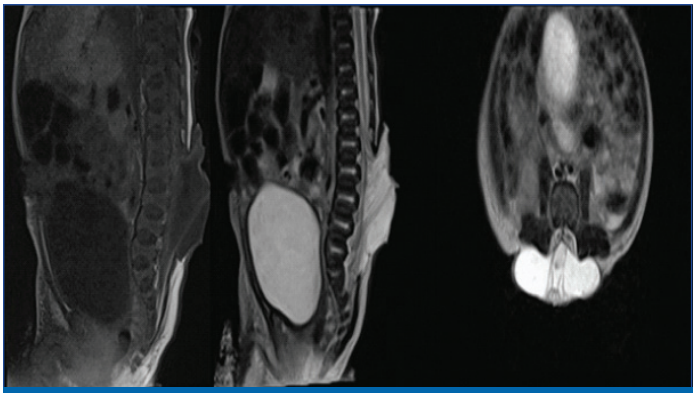
[Table/Fig-6]: Site of involvement distribution of patients studied.

ACMII	Age in years			Total
	0-1 year	1-5 years	>5 years	
Negative	4 (50%)	12 (92.3%)	9 (100%)	25 (83.3%)
Positive	4 (50%)	1 (7.7%)	0 (0%)	5 (16.7%)
Total	8 (100%)	13 (100%)	9 (100%)	30 (100%)

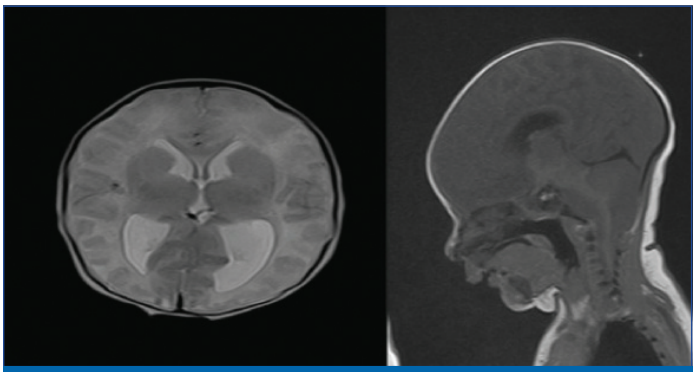
[Table/Fig-7]: Arnold chiari malformation type II (ACMII) distribution in relation to age distribution of patients studied.

There were total 5 cases of Arnold chiari malformation type II, 4 in 0-1 age group and 1 in 1-5 years age group [Table/Fig-7]. Few examples are illustrated in [Table/Fig-8-16].

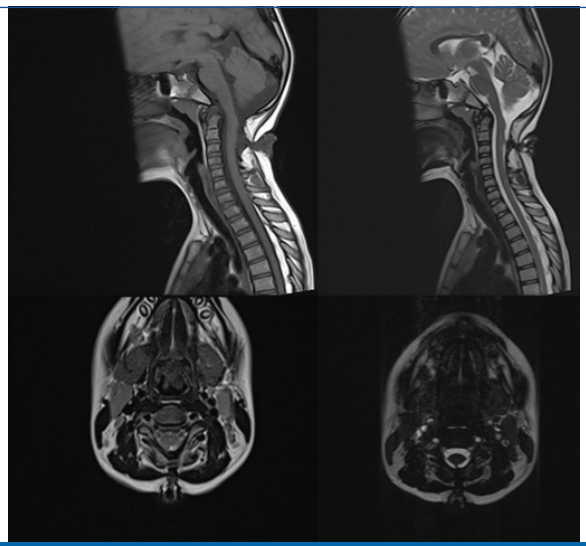
Of the 30 patients, 10 cases were of open spinal dysraphism with 8 cases presenting as myelomeningocele among which 5 cases were associated with Arnold chiari malformation II. Lumbosacral spine was most common location of myelomeningocele (5 cases) followed by 1 cervical and 1 sacral and 1 lumbar. Twenty patients were closed spinal dysraphism. Congenital spinal lesions without subcutaneous masses (43.3%) were more common than the lesions with subcutaneous masses (30%). Out of 10 cases of diastematomyelia, 6 were type 1 (60%) and 4 were type 2 (40%). Lumbosacral spine was most common site of involvement in 11 cases (36.65) followed by lumbar (33.3%) and sacral spine (10%) and dorsolumbar spine (10%).



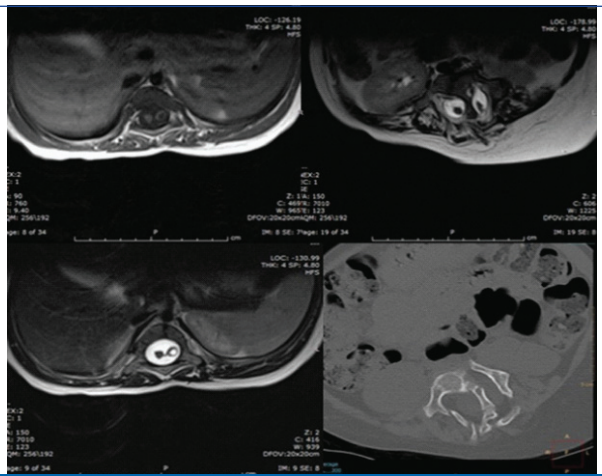
[Table/Fig-8]: Sagittal T1 and T2 W, Axial T2 W MR Images showing lumbosacral myelomeningocele.



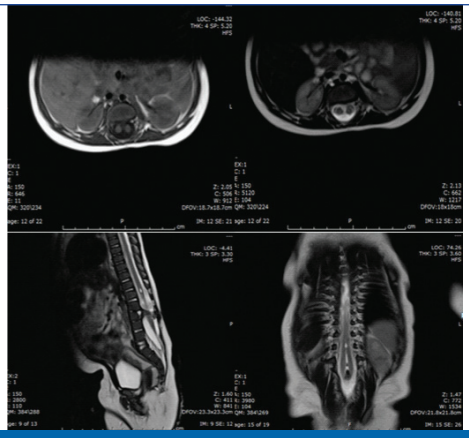
[Table/Fig-9]: Axial T2 W and Sagittal T1 W MR Image showing hydrocephalus and cerebellar tonsilar herniation (ACMII).



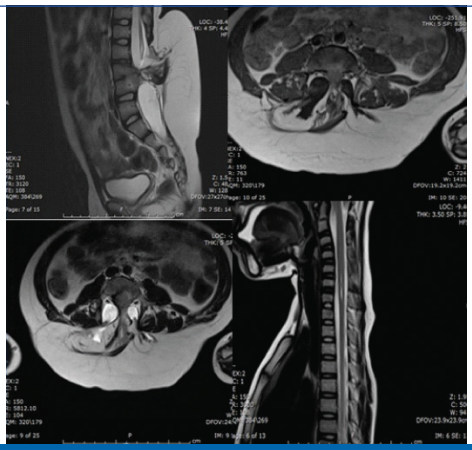
[Table/Fig-10]: Sagittal and Axial T1 and T2 W MR Images showing cervical myelomeningocele.



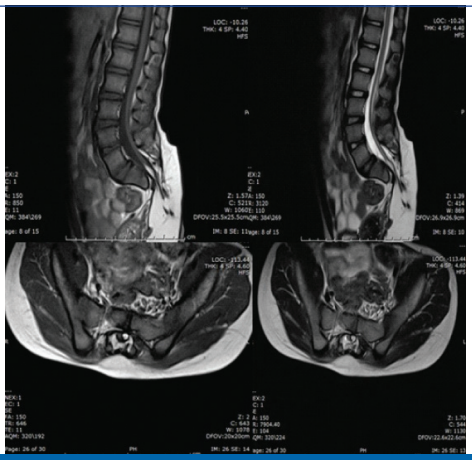
[Table/Fig-11]: Axial T1 and T2 W MR Images and axial CT image showing Type I diastematomyelia with dorsolumbar lipomyelomeningocele with syrinx in left hemicord.



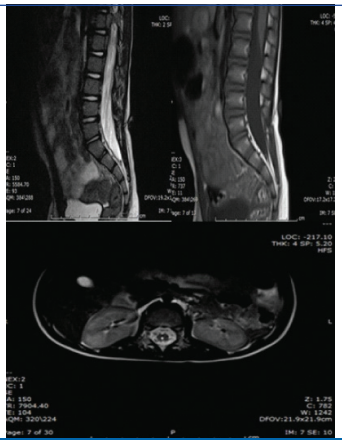
[Table/Fig-12]: Axial T1 and T2 W, sagitall T1 W and COR T2W MR Images Type 1 diastematomyelia with tethered cord and spina bifida in lumbosacral spine.



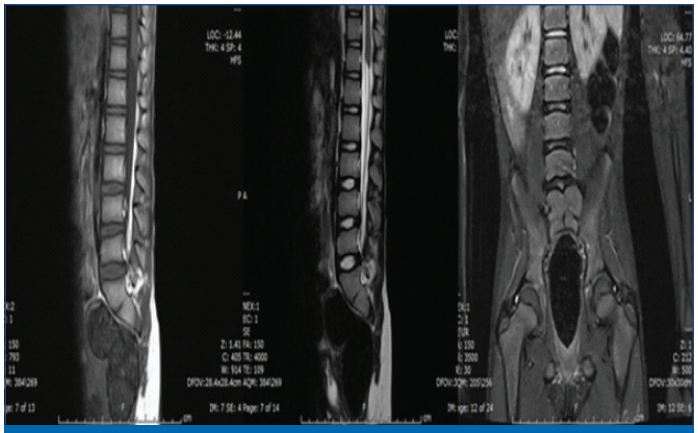
[Table/Fig-13]: Sagital T2 W and AXILA T1 and T2 W MR Images showing lumbar LPMMC with type I diastematomyelia with tethered cord and syrinx.



[Table/Fig-14]: Sagittal and Axial T1 and T2 W MR Images showing filum terminale lipoma with tethered cord and syrinx.



[Table/Fig-15]: Sagittal T1 and T2 W, Axial T2 MR Images showing dorsal dermal sinus with syrinx.



[Table/Fig-16]: Sagittal T1 and T2 W, stir coronal MR Images showing sacral agenesis.

DISCUSSION

Spinal dysraphisms are the congenital abnormalities of the spine and spinal cord. In present study, the role of MRI in evaluation of suspected spinal dysraphism was studied. The age of the patients in present study ranged from 4 months to 11 years. Maximum number of patients were in the age group of 1-5 years, accounting for ~43.3%. In a study by Kumari MV et al., age of the patients ranged from 17 days to 13 years [8]. Most of the children are below 1 year of age and by Nafees M et al., age of the patients ranged from 16 days to 37 years and most of them were below 6.4 years [19].

In this study out of 30 patients, 10 (33.33%) had open spinal dysraphisms and 20 (66.66%) had closed dysraphisms. The results were different to study by Kumari MV et al., in which 38 (57.5%) were open spinal dysraphisms and 28 (42.4%) were closed dysraphisms [8] Children of spinal dysraphism present with clinical features like swelling on the back, dermal sinus, haemangioma, dimple, lower limb weakness, and bowel and bladder incontinence. In this study, swelling in the back is the most common clinical feature which constituted 15 (50%) patients. In a study by Kumari MV et al., also swelling in the back was the most common clinical feature which constituted 51 (77.2%) patients [8]. Among different types of dysraphism, lipomyelomeningocele was the most common in closed spinal dysraphism which constituted 9 (30%) and in open spinal dysraphism, myelomeningocele is most common, which constituted 8 (26.6%). In a study by Kumari MV et al., myelomeningocele was the most common dysraphism which constituted 38 (57.5%) [8]. In a study by Nafees M et al., also myelomeningocele is the most common dysraphism which constituted 29 (39.2%) [19].

Location of myelomeningoceles of the 8 patients in this study were: one were noted in the cervical region, 1 in the lumbar region, 1 in the sacral region, 5 in lumbosacral region. In a study by Nafees M et al., also lumbosacral region was the most common location which constituted 38 (51.4%) and in study by Kumari MV et al., it was seen in 20 (52.6%) patients [8,19].

In this study, 10 (33.3%) Diastematomyelia patients were detected, Of which type 2 constituted 40% and type 1 constituted 60%. In a study by Kumari MV et al., 16 (24.2%) Diastematomyelia patients were detected. Of which type 2 constituted 75% and type 1 constituted 25% and in a study by Nishtar T et al., of the 53 patients 2 (4%) Diastematomyelia patients were detected [20].

Open defects are associated with abnormalities such as hydrocephalus, Arnold-Chiari, syrinx. In our study Arnold-Chiari type 2 constituted 5 (16.7%). In a study by Kumari MV et al., Arnold-Chiari type 2 constituted 6 (15.7%) and In a study by Kumar R et al., Arnold-Chiari type 2 constituted 62 (45%) patients [8,21].

LIMITATION

The number of cases included was limited. This small number could have potentially skewed the findings and age group >12 years was not included in this study, which could have resulted missing of cases with delayed presentation.

CONCLUSION

Imaging of spinal dysraphism is convoluted as various different conditions are involved in it which can have variable imaging appearance. An organized approach and MRI can helps in making the correct diagnosis as it does not involve ionizing radiation, is advanced and safe modality for assessing the spinal cord at all ages and defining complex spinal dysraphism.

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