Original Article

ISSN (O): 2347-338X; ISSN (P): 2347-3371

MR Imaging Spectrum of Spinal Dysraphism: A Study from South India

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Abstract

Background: Congenital anomalies of spine carry significant mortality and morbidity. Hence, they must diagnosed with great accuracy and at an earliest possible point of time. This study was undertaken to study the spectrum of MR imaging findings in patients with suspected spinal dysraphism regarding detection, accurate localization, non-invasive exploration of complex anatomy/pathological process and possible associations. **Subjects and Methods:** A total 40 cases attending the department of Radiology of JJM Medical College constituted the sample size. All the patients were subjected for non-contrast Magnetic resonance imaging in shortest possible examination time. The data thus collected was analyzed. **Results:** About 32.5% of the study were aged between 1 – 5 years. Moreover, in only 5 (25 %) cases the parents of the patients had history of consanguineous marriages. About 70% of the patients had neurological manifestations and 75% had cutaneous manifestations. A wide range of abnormalities were seen with myelomeningocele found in 65% of the patients, lipomyelomeningocele in 17.5% and 17.5% patients had diastematomyelia. Associated tethering of the cord is seen in 40% of the cases, while syrinx was noted in 10% patients, 5% patient showed cerebellar tonsillar herniation with cervical syrinx, 12.5% patients showed diffuse syringomyelia involving whole spine, 2.5% patient showed cervico-thoracic septated syrinx extending to medulla oblongata superiorly. **Conclusion :** MRI is an excellent imaging modality of choice for defining complex spinal dysraphism and associated abnormalities.

Keywords: Magnetic resonance imaging, Spinal dysraphism, Myelomeningocele, Syrinx, Congenital anomalies.

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Received: 06 October 2021 **Revised: 17 November 2021** **Accepted: 25 November 2021** **Published: 24 December 2021**

Introduction

A wide variety of congenital anomalies of spine which are mainly due to faulty midline fusion of the embryonic neural tube is often termed as spinal dysraphism.[1] This disease carries significant mortality and morbidity. The estimated incidence of spinal dysraphism is about 1 – 3/ 1000 live births.[2] The literature available had shown that, about 55 – 70% of the neural tube defects occur in females. The disease is known to vary with the race, ethnicity, gender and region.[3] Thorough antenatal screening and supplementation of the folic acid to the pregnant women had shown decline in the prevalence during last 25 years.[4] Most of the closed spinal dysraphisms remain asymptomatic during birth and can be suspected only in presence of cutaneous markers or manifestation of neurological deficit in later stages of life.[5,6]

The studies have shown that Magnetic resonance imaging has better investigative performance, noninvasive soft tissue characterization and emerged as an important tool in surgical planning than other imaging modalities.[7] The aim of present study is to demonstrate the spectrum of MR imaging findings in patients with suspected spinal dysraphism regarding detection, accurate localization, non-invasive exploration of complex anatomy/pathological process and possible associations.

Subjects and Methods

Patients referred from pediatrics department who presented with skin stigmata and various neurological problems having a suspicion of spinal dysraphism. A total of 40 patients were evaluated over a period of 8 months. All the patients underwent plain MRI of spine using 1.5Tesla PHILIPS ACHIEVA machine. Routine sequences used are T1&T2 – SAGITTAL, T1&T2 AXIAL, STIR-CORONAL, MR MYELOGRAM.

High spatial and contrast resolution images were obtained in shortest possible examination time. T-1 and T-2 weighted pulse sequences were acquired in sagittal as well as axial planes. T-1 weighted pulse sequences were used for evaluation of entire craniospinal axis helping to delineate the vertebral body marrow space, cord size and contour. T-2 weighted sequences were used to highlight the lesions in the

cord parenchyma as well as to delineate cerebrospinal fluid and extradural interface. Both T-1 and T-2 weighted images were helpful when lipomatous component was present as it gives high signal intensity on both sequences. Descriptive statistics were used to present the data.

Results

Table 1: Baseline characteristics of the patients

Characterist		Frequency	Percent
Age group	Less than 1 year	9	22.5
	1 – 5 years	13	32.5
	5 – 10 years	11	27.5
	11 – 15 years	6	15.0
	More than 15 years	1	2.5
H/O	No	38	95.0
	Yes	2	5.0

guineous marriage

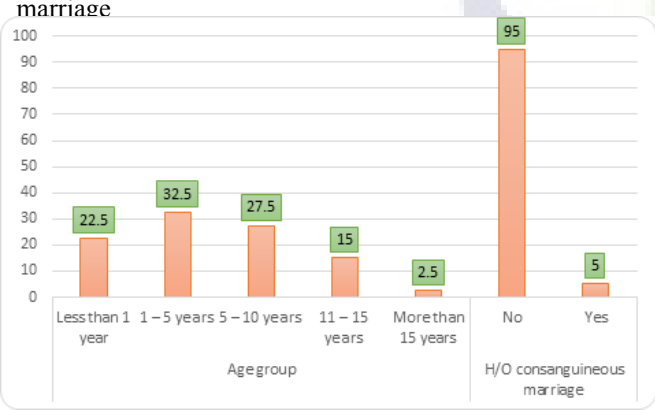

Chart 1: **Baseline characteristics of the patients**

This study had shown that, about 32.5% of the study were aged between 1 – 5 years. Moreover, in only 2 (5 %) cases the parents of the patients had history of consanguineous marriages.

This study had shown that, 70% of the patients had neurological manifestations and 75% had cutaneous manifestations. About 45% of the patients had midline swelling.

A wide range of abnormalities were seen with myelomeningocele found in 65% of the patients, lipomyelomeningocele in 17.5% and 17.5% patients had diastematomyelia. Associated tethering of the cord is seen in 40% of the cases, while syrinx was noted in 10% patients, 5% patient showed cerebellar tonsillar herniation with cervical syrinx, 12.5% patients showed

Table 2: Symptoms in the study group

Symptoms	Frequency	Percent
Neurological manifestations	28	70.0
Cutaneous manifestations	30	75.0
Midline swelling	18	45.0

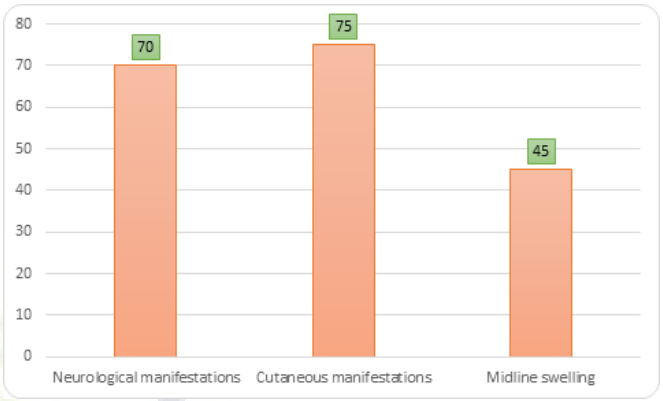

Chart 2: **Symptoms in the study group**

Table 3: Diagnosis and MRI findings of the study group

		Frequency	Percent
	Diastematomyelia	7	17.5
	Lipomyelomeningocel	7	17.5
	Myelomeningocele	26	65.0
MRI	Tethering	16	40.0
	Syrinx	4	10.0
	Herniation	2	5.0
	Diffuse syrinx	2	5.0
	Scoliosis	5	12.5
	Hemi – vertebrae – butterfly vertebrae	1	2.5

diffuse syringomyelia involving whole spine, 2.5%) patient showed cervico-thoracic septated syrinx extending to medulla oblongata superiorly. Scoliosis is seen 12.5% patients and other Vertebral anomalies like hemi-vertebrae/butterfly vertebrae are found in 2.5%. Only one patient had low lying tethered cord associated with other congenital anomalies like lumbar hernia and absent ribs which was diagnosed as a rare lumbo-costo-vertebral syndrome. All the patients showed one or multiple suspected intraspinal anomalies on MRI spine.

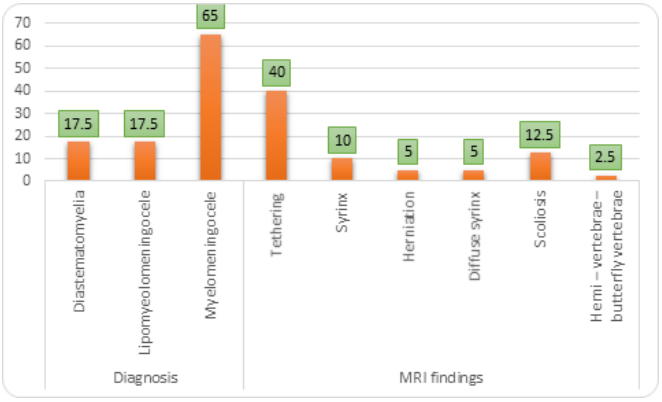

Chart 3: **Diagnosis and MRI findings of the study group**

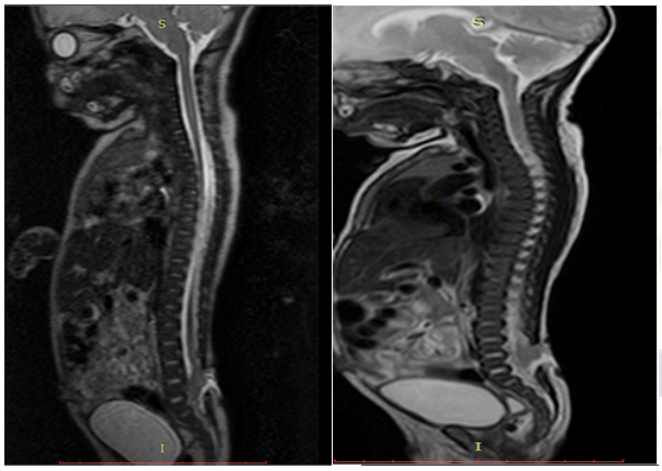

Figure 1: A meningocele is a defect consisting of a herniation of Meningeal tissuethrough a defect in the skull and/or spine. Myelomeningocele is a defectconsisting of a herniation of meningeal tissue and nervous tissue through adefect in the skull and/or spine.

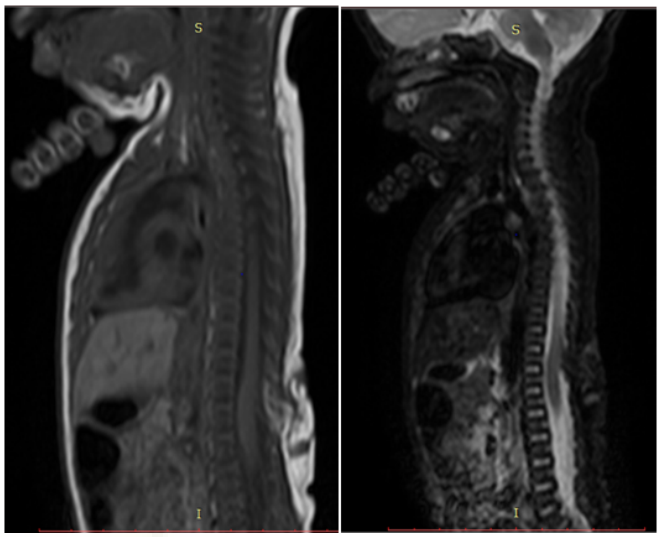

Figure 2: Lipomyelomeningocele consists of a skin covered back mass that containsneural tissues, CSF, meninges and presence of lipoma or lipomatous componentthat extends from the subcutaneous tissue of back through the back mass intothe spinal canal.

Discussion

Spinal dysraphism, or neural tube defect (NTD), is a broad term encompassing a heterogeneous group of congenital spinal anomalies related to improper closure of the caudal neuropore. These conditions include spina bifida Aperta, spina bifida occulta, meningocele, myelomeningocele, Lipomyelomeningocele, myeloschisis, and rachischisis - names given variably according to radiological or pathological findings. These variations can be grouped as open if the overlying skin is not intact, pending leakage of cerebrospinal fluid, and occult if the defect is well covered with full thickness skin. Early detection and prompt neurosurgical correction of occult spinal dysraphism may prevent upper urinary tract deterioration, infection of dorsal dermal sinuses, or permanent neurologic damage.[8]

Several studies have demonstrated that motor function, urologic symptoms, and urodynamic patterns may be improved by early surgical intervention in patients with occult spinal dysraphism. The surgical outcome may be better if intervention occurs before the age of 3 years. Spinal neuroimaging, therefore, has the important role of determining the presence or absence of an occult spinal dysraphic lesion so that appropriate surgical treatment can be instituted in a timely manner. There is no consensus regarding the most appropriate neuroimaging modality to diagnose occult dysraphism. Most of the authors emphasize the importance of spinal magnetic resonance imaging (MRI) examination because of its better diagnostic performance, excellent soft tissue characterization, and importance in presurgical planning. In our study we have emphasized that MRI is the imaging modality of choice to demonstrate the spectrum of findings in patients with suspected spinal dysraphism.[9]

This study had shown that, the spinal dysraphism was diagnosed before 5 years in most of the children in this study. A study by Lee,[10] Kumari MV et al,[11] Nafees M et al also noted similar findings.[12]

This study had shown that majority of the spinal dysraphisms were closed type unlike study by Kumari et al.[11] Myelomeningocele was the common spinal dysraphism in this study. A study by Hosagavi et al,[13] and Kumari et al also

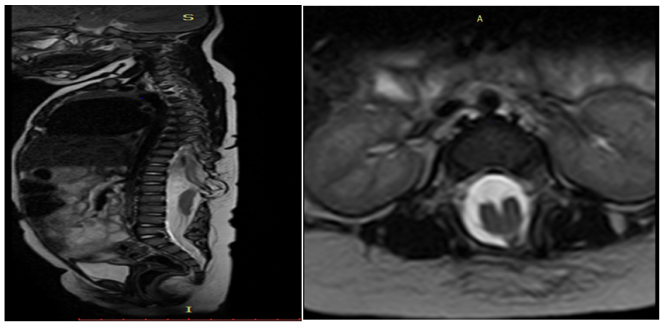

Figure 3: Spinal cord is split into two hemicords, each having one set of dorsal and ventral nerve roots. Each has its own dural sheath. They are separated by fibrous, bony, or cartilaginous septae associated with bony abnormalities such as split or fused vertebrae.

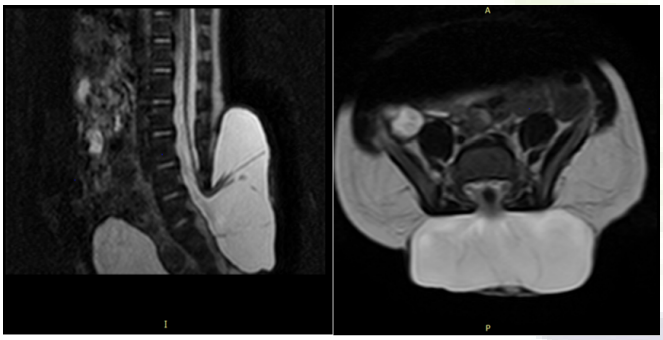

Figure 4: Low lying tethered spinal cord: A condition in which the spinal cord is fastened to an immovable structure, such as a lipoma, vertebra, dura, or skin. Always associated with spina bifida occulta and manifests it as low conus. By the age of 2 months, a conus below L2-L3 is considered abnormal.

shown similar findings.[11]

This study has reported that, the magnetic resonance imaging is the good modality of choice for diagnosis of spinal dysraphisms. A prospective study carried out in 100 cases of suspected occult spinal dysraphic anomalies with MRI provided accurate preoperative information in 91 out of 92 cases (98.9%).[12] Another study revealed that MRI is the imaging method of choice for investigation of complex group of spinal disorders. Comparing with the above-mentioned studies this study revealed that MRI is imaging modality of choice to demonstrate the spectrum of findings in patients with suspected spinal dysraphism.

Conclusion

MRI is an excellent imaging modality for visualizing the spinal cord at all ages and is the imaging modality of choice for defining complex spinal dysraphism and associated abnormalities. Because of its multiplanar imaging and tissue characterization capabilities, magnetic resonance imaging (MRI) has greatly ameliorated the diagnosis of these disorders and has advanced the possibility of earlier treatment. It is considered as an accurate screening modality in the initial diagnosis of occult spinal dysraphism. MRI has replaced other neuroimaging modalities, such as conventional tomography (CT) and CT myelography.

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How to cite this article: Chethan S, Jeevika MU, Gadag G, Patil S. MR Imaging Spectrum of Spinal Dysraphism: A Study from South India. Asian J. Med. Radiol. Res. 2021;9(2):31-35.

DOI: dx.doi.org/10.47009/ajmrr.2021.9.2.8

Source of Support: Nil, **Conflict of Interest:** None declared.