

## Prospective Evaluation of Role of MRI in Suspected Spinal Dysraphism and Its Management

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**Abstract:** Spina bifida literally means “spine in two parts” or “open spine”. Spinal dysraphism involves a spectrum of congenital anomalies resulting in a defective neural arch through which meninges or neural elements are herniated, leading to a variety of clinical manifestations. They are divided into aperta (visible lesion) and occulta (with no external lesion). Meningocele, myelomeningocele, lipomeningomyelocele, myeloschisis and rachischisis are the usual names associated depending on the pathological findings. Meningocele by definition involves only the meninges with no neural involvement; others have variable extent of neural involvement. The spina bifida aperta is usually associated with skin defect with an impending risk of CSF leak constituting “open defects,” whereas the occult forms have normal skin cover. Both forms demand different approaches in their management. The clinical importance of occult lesion has grown tremendously in the recent years.

**AIMS:** To demonstrate the spectrum of MRI findings in spinal dysraphism and its management.

**MATERIALS AND METHODS:** Prospective evaluation of 66 patients with suspected spinal dysraphism on 1.5 T MRI.

**RESULTS:** Age of the patients ranged from 17 days to 13 years. Commonest clinical presentation was swelling in the back. Open spinal dysraphism, in which lumbosacral myelomeningocele was most common presentation. Diastematomyelia was the next most common presentation. Associated abnormalities like hydrocephalus, Arnold–Chiari, syrinx, hydronephrosis were commonly encountered in open defects. We have one case of open spinal dysraphism with osseous hamartoma. MRI with its multiplanar capabilities and superior soft tissue contrast, allows detection of detailed anatomy and characterization of congenital spinal disorders.

**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.

**Keywords:** MRI, Spinal Dysraphism, Lumbar Myelomeningocele, Diastematomyelia, Open Spinal Dysraphism with Osseous Hamartoma, Tethered Cord, Dermal Sinus.

Date of Submission: 17-04-2018

Date of acceptance: 05-05-2018

### I. Introduction

Congenital abnormalities of the spine and spinal cord are referred to as spinal dysraphisms. Spinal dysraphisms can be broadly categorized into open and closed type. This can be grouped as open if the overlying skin is not intact causing leakage of cerebrospinal fluid and occult if the defect is well covered with full thickness skin. The estimated incidence of spinal dysraphism is about 1–3/1000 live birth. About 55-70% of neural tube defects occur in females. 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. The surgical outcome may be better if intervention occurs before the age of 3 years.<sup>1</sup>

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. Magnetic Resonance Imaging (MRI) is the examination of choice because of its better diagnostic performance, excellent soft tissue characterization and importance in presurgical planning.<sup>2</sup>

### II. Materials And Methods

We prospectively evaluated 66 patients with suspected spinal dysraphism and who had undergone MR imaging of the spine from May 2015 to September 2017 were included in the study after informed consent. This study was conducted in the department of radiology, Prathima Institute of Medical Sciences, Karimnagar, Telangana, India.

**Inclusion Criteria:** Patients who presented with various neurological problems and skin stigmata suspicious of spinal dysraphism.

**Exclusion Criteria:** Patients with metallic implants and patients with claustrophobia were excluded from the study. In all patients clinical history and antenatal history were recorded. Sedation was given in required patients.

MRI was performed on a 1.5 Tesla electromagnet (General Electrical Medical Systems). The pulse sequences included T1WI, T2WI using spin echo and Short Tau Inversion Recovery sequences. Axial and sagittal T2WI were obtained with TR of 3000ms and TE of 120ms. Sagittal T1WI were obtained with TR of 60ms and TE of 30ms. Sagittal and coronal STIR images were obtained with TR of 3000ms and TE of 40ms. Images were obtained with an interslice gap of 5.2mm, slice thickness of 4mm and a matrix size of 512x512. On MRI, imaging findings in vertebrae, spinal cord and soft tissues were noted.<sup>3</sup>

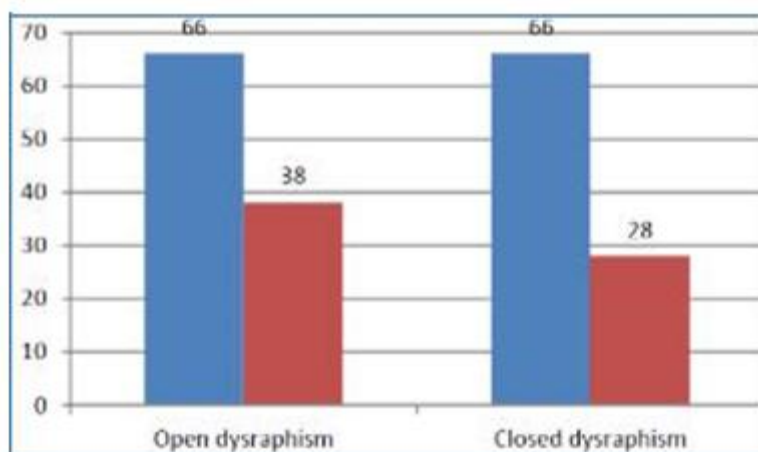
### III. Observations and Results

66 patients who presented with various neurological problems and skin stigmata suspicious of spinal dysraphism were referred to our department in Prathima Institute of Medical Sciences and underwent Magnetic Resonance Imaging of the spine in a time period from May 2015 to September 2017 were included in our study. Age of the patients ranged from 17 days to 13 yrs. Most of the children are below 1 yr. of age. Of the 66 patients, 40 were female patients and 26 were male patients. The below table shows the gender distribution of spinal dysraphism.

GENDER	NUMBER	PERCENTAGE
Males	26	39.4%
Females	40	60.6%

*Table 1: Gender Distribution*

Of the 66 patients, 38 are open spinal dysraphisms and 28 are closed dysraphisms.



*Fig. 1: Types of Spinal Dysraphism*

Children with suspected spinal dysraphism can present with various clinical features such as swelling on the back, dimple, hemangioma, dermal sinus, lower limb weakness, and bladder and bowel incontinence

S.No	Clinical Feature	Number	Percentage
	Swelling in the back	51	77.2%
	Hypertrichosis	6	9%
	Dimple	3	4.5 %
	Lower	14	21.2%
	Lower weakness	6	9%
	Urinary incontinence	4	6%
	Dermal sinus fecal incontinence	2	3%

*Table 2: Clinical Presentation of Spinal Dysraphism*

Myelomeningocele followed by diastematomyelia were the commonest presentations in our study. One case was cervical myelomeningocele with osseous hamartoma.

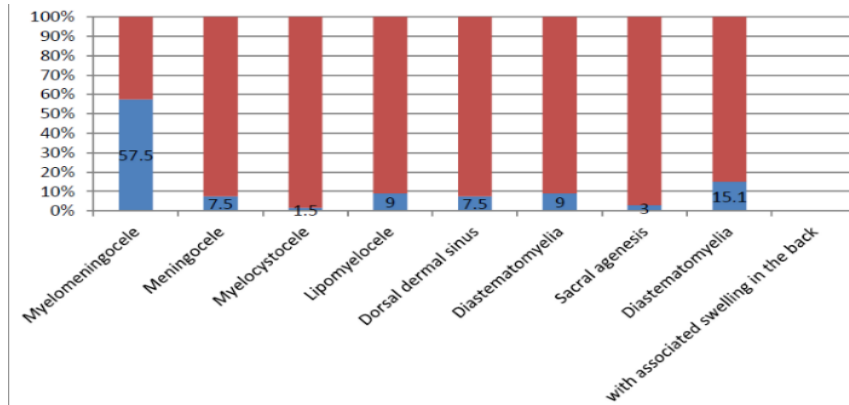


Fig. 2: Different Types of Dysraphism

Location of myelomeningoceles of the 38 patients in our study, 4 were noted in the cervical region, 2 in the dorsal region, 5 in the lumbar region, 7 in the sacral region, 20 in lumbosacral region and 2 in dorsal region.

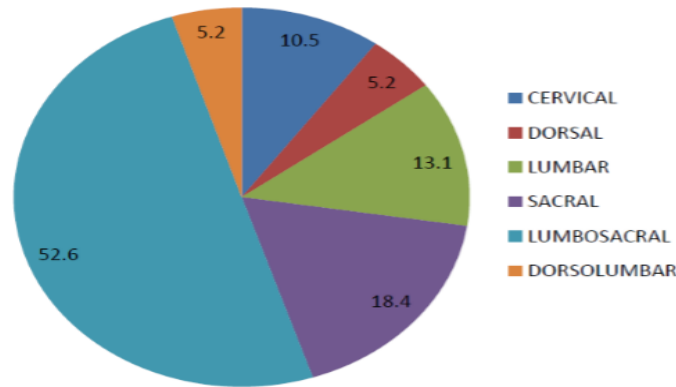


Fig. 2: Location of Myelomeningoceles

Of 66 patients, 16 Diastematomyelia patients were detected. In type 1, two hemicords are encased in respective dural sacs and separated by a bony septum. In type 2, the hemicords are encased in a single sac and separated by a fibrous septum.

S.No	Type of Diastematomyelia	Number	Percentage
1.	Type-1	4	25%
2.	Type-2	12	75%

Table 3: Type of Diastematomyelia

Open defects were associated with abnormalities such as hydrocephalus, Arnold-Chiari, syrinx.

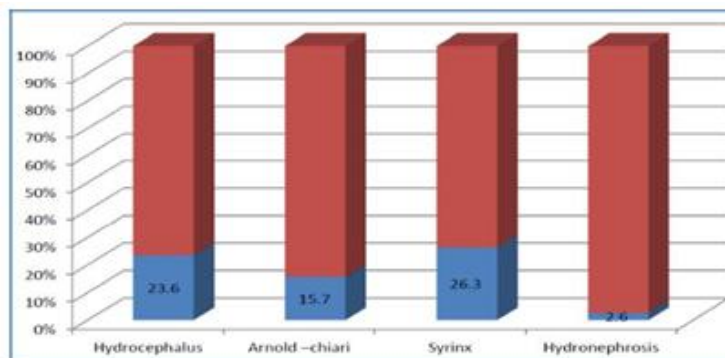
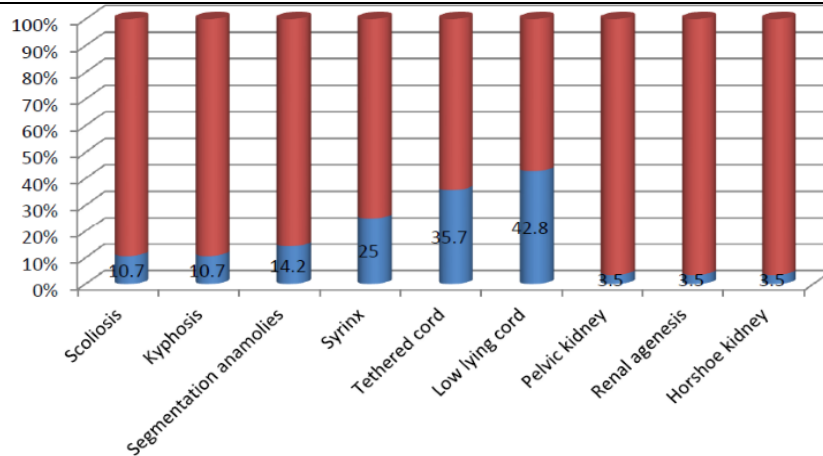


Fig. 3: Open Defects with Associated Abnorma



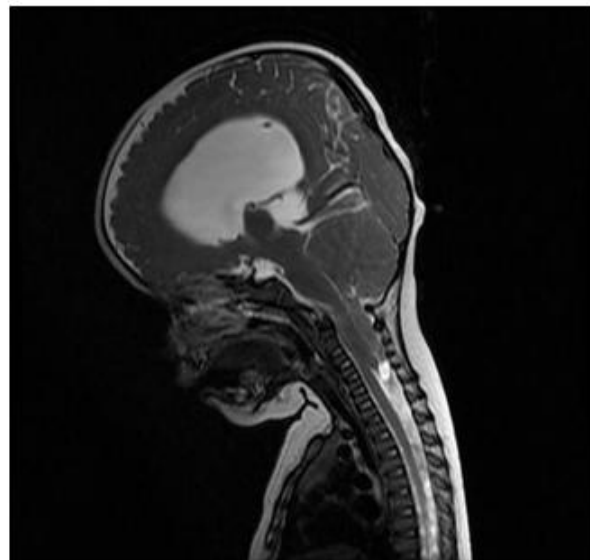
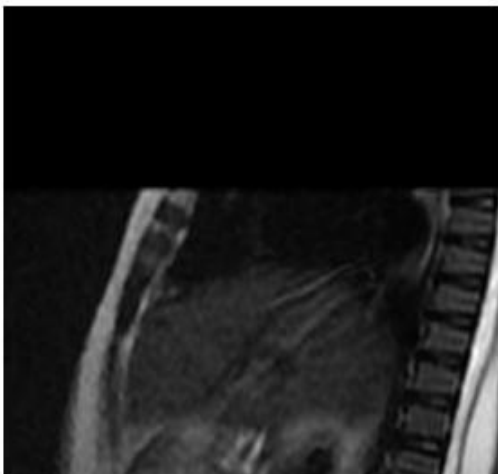
**Fig. 4: Closed Defects with Associated Abnormalities**

Closed defects were associated with abnormalities such as segmentation anomalies, syrinx, scoliosis, tethered cord.

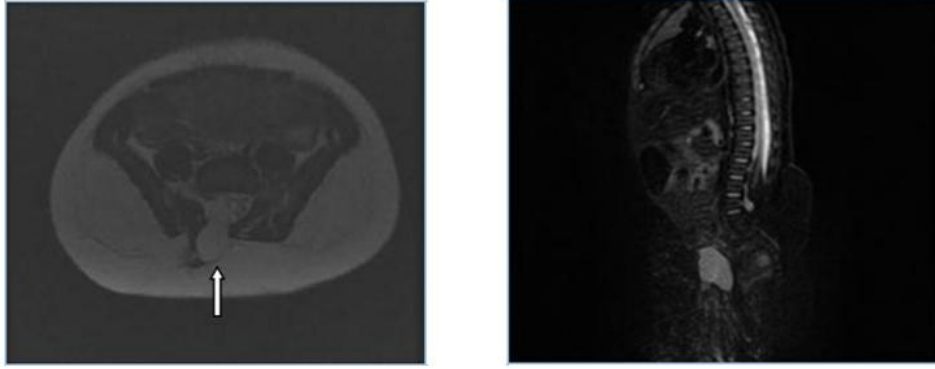
Tethered cord can result from variety of conditions. In our study, 10 cases are associated with closed spinal defects and 4 cases are due to surgery for myelomeningocele.

S.NO	TETHERED CORD	NUMBER	PERCENTAGE
1.	Associated with closed defects	10	71.5%
	Post Surgery	4	28.5%

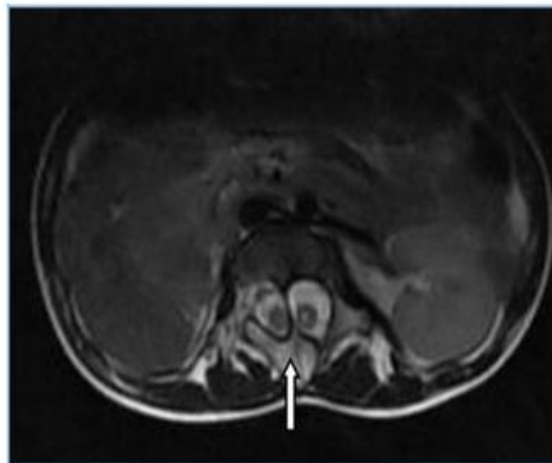
**Table 5: Tethered cord**



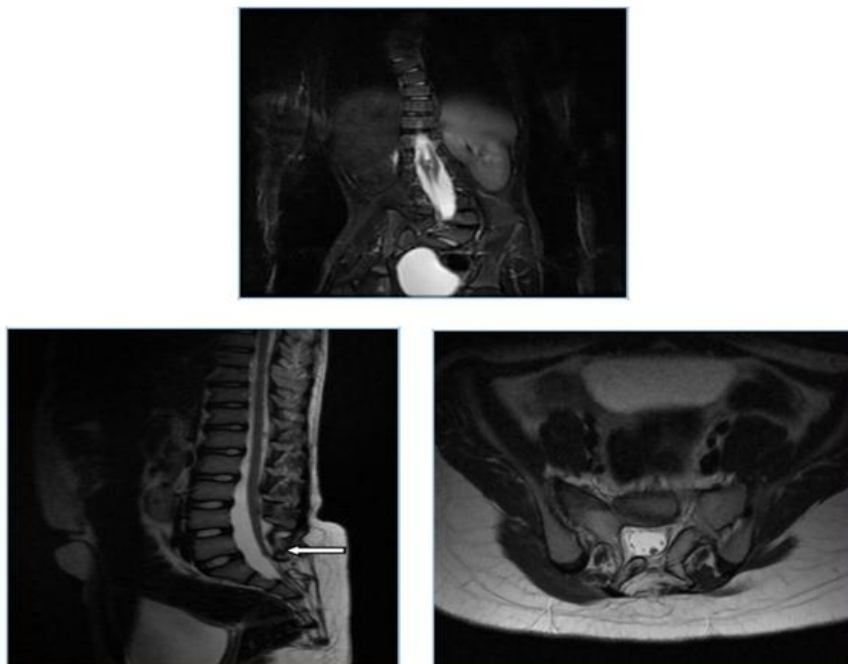
**Fig. 6: Sagittal T2WI – Images showing the Herniation of Meninges and Nerve Roots into the Subcutaneous Plane and Tonsillar Herniation - Lumbar Myelomeningocele with Arnold Chiari Type 2**



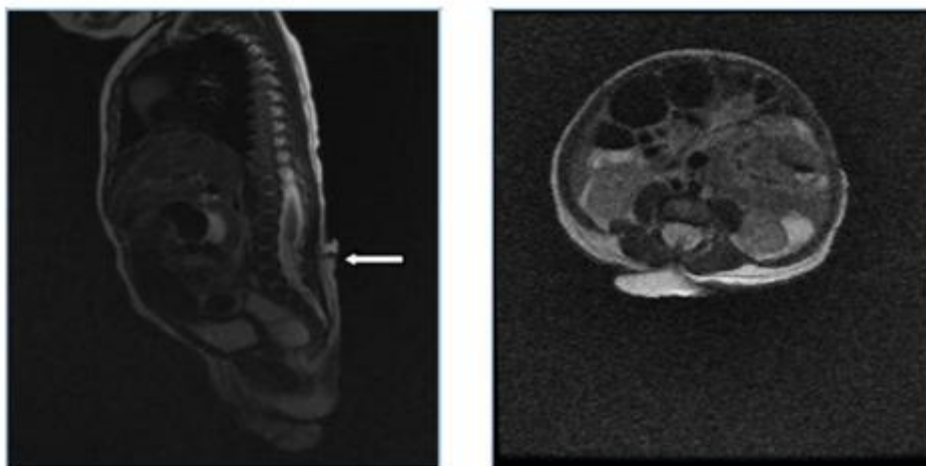
**Fig. 7:** Axial T2W - Images showing Herniation of Nerve Roots and Altered Signal Intensity Mass Noted in Epidural Space showing Continuity with Subcutaneous Fat. Sagittal STIR - Suppression of the Lesion – Lipomyelocele



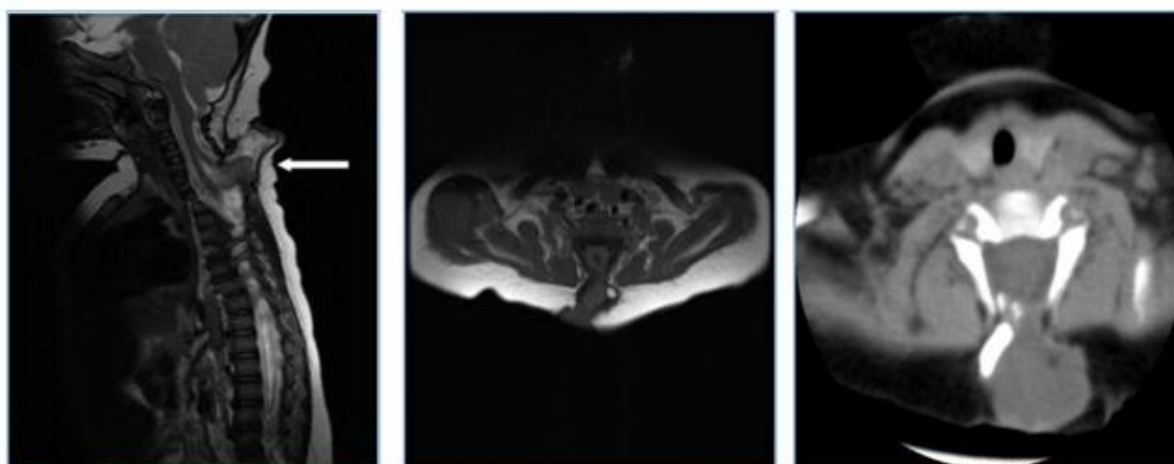
**Fig. 8:** Coronal STIR and Axial T2W Images showing Two Hemicords with Two Dural Sacs with Bony Septum between them with Absent Right Kidney - Diastematomyelia Type 1



**Fig. 9:** Sagittal and Axial T2W Images showing Spina Bifida of L5 and S1 Vertebrae with Low Lying and Tethering of Cord - Spina Bifida with Low Lying and Tethered Cord with Thick Filum



**Fig. 10:** Sagittal and Axial T2WI showing Hypointense Linear Structure Extending from Skin and Subcutaneous Planes at L4 Level into the Dura s/o Dorsal Dermal Sinus



**Fig. 11:** Sagittal T2WI and Axial T1WI showing Herniation of Spinal Cord and Meninges. Axial CT showing Osseous Component - Cervical Myelomeningocele with Osseous Hamartoma

## MANAGEMENT

Management of these children needs multidisciplinary approaches. Complete clinical evaluation and appropriate investigations are necessary. Parents need to be counseled and informed regarding the immediate as well as long-term management strategy.<sup>6</sup>

### Surgical treatment

The aim of surgery is to free the placode from the surrounding abnormal skin and reposition into the spinal canal with reconstruction of the dura and coverings to prevent CSF leak and infection. The surgical technique depends on the size and the level of the lesion. The help of pediatric, orthopedic and plastic surgeons may be necessary. Several attempts for maternal fetal surgeries to improve their outcomes have been made. The role of fetal surgery for myelomeningocele is yet to be proven.<sup>7</sup>

Timing of intervention primarily depends on the clinical condition of the child and the impending risks. Surgery need not be done as a compelling emergency but should be undertaken as soon as it is practical.<sup>8</sup> In case of suspected meningitis or CSF infection or colonization of the wound, prophylactic antibiotics and anticonvulsants form the initial treatment. Child is nursed in an incubator; routine blood counts and serum electrolytes are monitored. Blood grouping and cross matching is done for possible transfusion. Careful assessment of body weight is essential for intraoperative management. The newborn child with myelomeningocele should have saline dressings. It is essential not to use corrosive agents, spirit or antiseptics indiscriminately over the open defects to avoid damage to the underlying exposed neural tissue.

### **Surgical technique**

To obtain successful repair, it is essential to study the surface anatomy and its relationships to the surrounding structures. At the apex of the myelomeningocele usually, the flat neural placode is located and from its edge the remnants of the arachnoid membrane get attached at the nerve root entry zone. From this junction, the nerve roots emerge and exit through neural foramina located ventrally. They are seen through the transparent arachnoid membrane which is fused with the skin at the lateral edges of the lesion. The dura matter which is defective posteriorly is loosely adherent to the underlying soft tissue of the back and densely adherent to the bony structures underneath. Rostrally, the dura forms tube and the neural placode continues into it, which leads to functional spinal cord.

The child is operated under general anesthesia with endotracheal intubation in prone position, with the head and neck placed comfortably. The first step is to isolate the neural placode, while salvaging the nerve roots as much as possible irrespective of their functional status. It is essential that no skin element becomes buried in the repair to prevent the possibility of an implantation dermoid. Vertical midline incision is preferable. In a circumferential fashion, arachnoid is lifted and the nerve roots are identified. This technique is continued all around till the neural placode is completely free. The neural placode can then be inverted and sutured. The dura is dissected from the underlying soft tissue. The dural closure needs to be made water tight with a graft, if necessary. The overlying skin is dissected from underlying fascia and musculature, mobilized and approximated. If necessary, relaxing incisions or flaps may be used to close larger defects. Kyphotic deformity or gibbus can pose special problems. Reigel had advocated resection of the kyphus with primary spinal fusion at the time of surgical repair.<sup>9</sup> When the dysplastic skin is large causing larger skin defects, balloon tissue expanders can be used to enlarge the normal skin area which can then cover the defect.<sup>10</sup>

Postoperative care is equally important. To nurse in prone position, protect the wound from fecal and urinary contamination. Symptomatic hydrocephalus or arnold chiari malformation (ACM) needs to be treated simultaneously. Supportive nutrition and antibiotics are required. Adequate urological treatment can prevent future complications of the upper urinary tract.<sup>11</sup> Early management, maintaining a low intravesical pressure and continuing intermittent catheterization with or without pharmacotherapy, is beneficial. But urological management should ideally be based on and modified by the urodynamic studies.

### **IV. Discussion**

Congenital abnormalities of the spine and spinal cord are referred to as spinal dysraphisms. The purpose of MR imaging in spinal dysraphism is (a) Detection of detailed anatomy (b) For soft tissue characterization of lesion (c) For presurgical planning (d) For early detection and prompt neurosurgical correction of occult spinal dysraphism in order to prevent permanent neurological damage. Our study of role of MRI in evaluation of suspected spinal dysraphism consisted of 66 patients who presented to our hospital with suspected spinal dysraphism and underwent MR imaging of the spine.

The age of the patients in our study ranged from 17 days to 13 yrs. Most of the children are below 1yr of age. In a study by Muhammed Nafees, et al. age of the patients ranged from 16 days old to 37 yrs. old, most of them below 6.4 yrs.<sup>1</sup> In our study, of the 66 patients 40 (66.6%) were female patients and 26 (33.3%) were male patients similar to study by Mohamed Fathy Dawodh et al., in which 18 are female patients and 14 are male patients.<sup>2</sup>

In our study of the 66 patients, 38 (57.5%) are open spinal dysraphisms and 28 (42.4%) are closed dysraphisms. The results were similar to study by Mohamed Fathy Dawodh et al.; 18 are open dysraphism and 14 are closed dysraphism.<sup>2</sup> Children with suspected spinal dysraphism can present with various clinical features such as swelling on the back, dimple, hemangioma, dermal sinus, lower limb weakness, bladder and bowel incontinence. In our study, swelling in the back is the commonest clinical feature which constituted 51 (77.2%). In a study by Mohamed Fathy Dawodh, et al., swelling in the back is the commonest clinical feature which constituted 26 (81.2%). In a study by Kumar R, Singh SN, et al. swelling in the back is the commonest clinical feature which constituted 89 (57%).<sup>2,3</sup>

Among different types of dysraphism myelomeningocele is the commonest. In our study, myelomeningocele constituted 38 (57.5%). In a study by Mohamed Fathy Dawodh, et al., myelomeningocele is the commonest dysraphism which constituted 15 (46.8%).

In a study by Muhammed Nafees, et al. myelomeningocele is the commonest dysraphism which constituted 29 (39.2%).<sup>2,1</sup>

Location of myelomeningoceles of the 38 patients in our study, 4 were noted in the cervical region, 2 in the dorsal region, 5 in the lumbar region, 7 in the sacral region, 20 in lumbosacral region and 2 in dorsal region. Among these lumbosacral region is the commonest location, which constituted 20 (52.6%). In a study by Muhammed Nafees et al., lumbosacral region is the commonest location which constituted 38 (51.4%). In a study by Mohamed Fathy Dawodh et al., lumbosacral region is the commonest location which constituted 11 (73.3%).<sup>1,2</sup>

In our study of the 66 patients, 16 (24.2%) Diastematomyelia patients were detected. Of which type 2 constituted 75% and type 1 constituted 25%. In a study by Taahira Nishtar et al., of the 53 patients 2 (4%) Diastematomyelia patients were detected.<sup>4</sup>

Open defects are associated with abnormalities such as hydrocephalus, Arnold-Chiari, syrinx. In our study hydrocephalus constituted 9 (23.6%), Arnold-Chiari type 2 constituted 6 (15.7%). In a study by Kumar R, Singh SN, et al., hydrocephalus constituted 71 (49%), Arnold-Chiari type 2 constituted 62 (45%).<sup>3</sup>

We had a rare case of cervical myelomeningocele with osseous dysraphic hamartoma. Zuppani HB, et al. reported a case of lipomyelocele with osseous dysraphic hamartoma in a child.<sup>5</sup>

A prospective study at NUR Research Centre Institute of Nuclear Medicine and Allied Sciences, Lucknow Marg, Delhi, Dept. of Neurosurgery, GB Pant Hospital New Delhi, was carried out in 100 cases of suspected occult spinal dysraphic anomalies with MRI in order to determine its diagnostic efficacy as the initial imaging modality. MR imaging provided accurate preoperative information in 91 out of 92 cases (98.9%). It is concluded that MRI is an excellent primary diagnostic tool, together with a plain radiograph for complete preoperative evaluation of mid-line spinal anomalies. Comparing with the above mentioned studies, our study revealed that MRI is imaging modality of choice to demonstrate the spectrum of findings in patients with suspected spinal dysraphism.

## V. Conclusion

MRI features of 65 patients with suspected spinal dysraphism were studied and imaging findings were interpreted.

1. The age of the patients in our study ranged from 17 days to 13 yrs. Most of the children are below 1 yr. of age.
2. Female predominance was noted in our study, which constituted 66.6%.
3. Open defects (57.5%) predominated in our study than closed defects (42.4%).
4. Swelling in the back is the commonest clinical feature in our study, which constituted 51 (77.2%).
5. Myelomeningocele is the commonest type of dysraphism, which constituted 38 (57.5%).
6. In myelomeningoceles, lumbosacral region is the commonest location which constituted 20 (52.6%).
7. In our study, 16 (24.2%) Diastematomyelia patients were detected. Of which type 2 constituted 75% and type 1 constituted 25%.
8. Open defects are associated with abnormalities such as hydrocephalus, Arnold-Chiari syrinx. Other spinal cord abnormalities such as tethered cord, low lying cord, segmentation anomalies, scoliosis, kyphosis were well delineated in our study.
9. Most of the myelomeningoceles in our study are associated with abnormalities.

Hence, MRI with its multiplanar capabilities and superior soft tissue contrast, allows detection of detailed anatomy and characterization of congenital spinal disorders.

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.

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Deepthi T "Prospective Evaluation of Role of Mir in Suspected Spinal Dysraphism and Its Management. IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17, no. 5, 2018, pp 21-28.