International Journal Dental and Medical Sciences Research
Volume 4, Issue 4, July-Aug 2022 pp 708-717 www.ijdmsrjournal.com ISSN: 2582-6018

Imaging Spectrum of Congenital Spinal Cord Anomalies- Aprospective Observational Study

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Submitted: 10-08-2022 Accepted: 22-08-2022

ABSTRACT
Neural tube defects are most common birth anomalies second after congenital heart diseases[1].
Spinal dysraphisms are subtype of neural tube defects with abnormalities of spinal cord development during 2nd and 6th gestational weeks and show incomplete closure of mesenchymal, osseous, and nervous tissue. Clinically they can range from going unnoticed to being discovered because of the associated genitourinary, gastrointestinal or musculoskeletal involvement[1]. This study helps to understand the imaging features of various congenital spinal cord and spinal anomalies knowledge of which is important to clinical radiologists in day to practice.

KEY WORDS-Spinal dysraphism, diastometamyelia, meningoencephalocele, lipomyelomeningocele.

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 types. The estimated incidence of spinal dysraphism is about 1–3/1000 live births[2].About 55-70% of neural tube defects occur in females.Early detection and prompt neurosurgical correction of spinal dysraphism may prevent upper urinary tract deterioration, infection of dorsal dermal sinuses, or permanent neurologic damage.

Spinal neuroimaging, has the important role of determining the presence of spinal dysraphic lesion so that appropriate surgical treatment can be instituted in a timely manner.Magnetic resonance imaging (MRI) because of its better diagnostic performance, good anatomic delineation excellent soft tissue characterization helps in detection of congenital anamolies of spine and its associatons and guides in presurgical planning[1,3].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.

AIMS AND OBJECTIVES

1. To evaluate clinically suspected cases of congenital spinal cord anomalies.
2. To describe the radiological and imaging findings and other associated anomalies.

EMBRYOLOGY

- **First stage** – Gastrulation - 2nd to 3rd week.

Anomalies such as caudal regression syndrome, diastometamyelia, neurenteric cyst and dorsal enteric fistula.

- **Second stage** – Primary neurulation - 3rd – 4th week.

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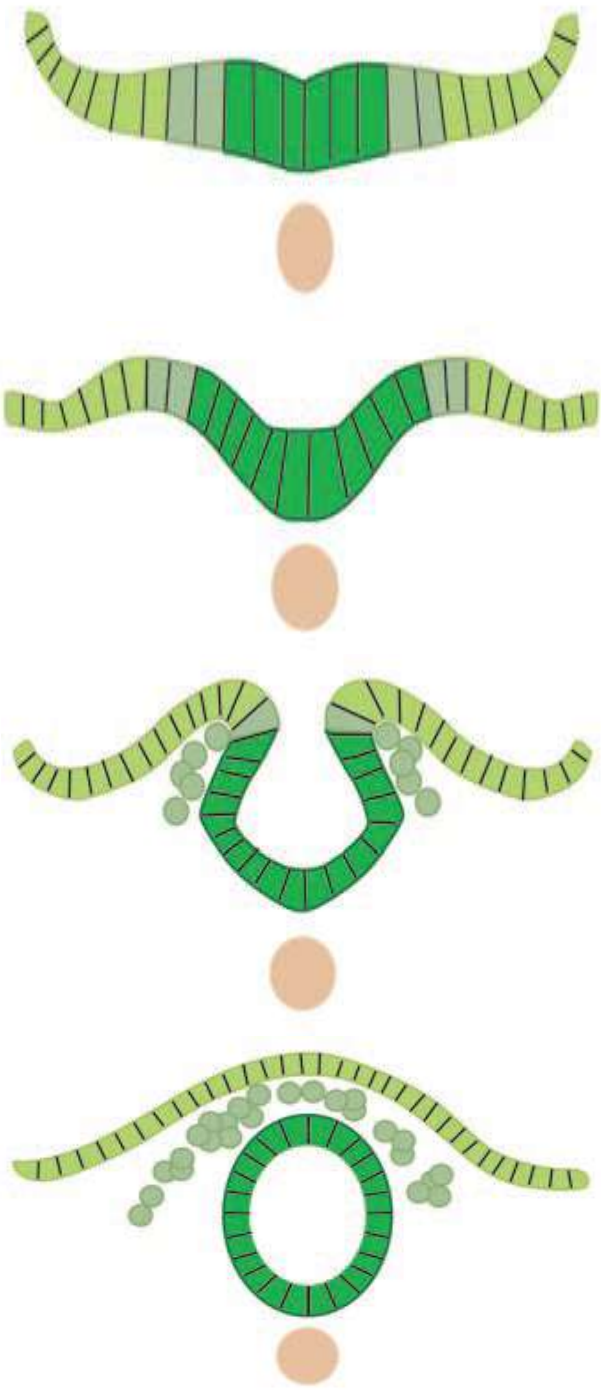

Illustration of primary neuralation: Notochord (circle) interacts with overlying ectoderm to form neural plate (dark green), which then blends to form neural tube that ultimately closes in zipper like fashion.

The neural tube separates from the surface ectoderm by a process called disjunction.

Premature disjunction - lipomyelocele, lipomyelomeningocele and intradural lipomas

Incomplete disjunction -dermal sinus

- **Third stage** -secondary neurulation - 5 - 6wk

Aberrations in this process lead to anomalies such as filar lipoma, tight filum terminale and persistent terminal ventricle

CLASSIFICATION

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Spinal dysraphisms can be broadly categorized into open and closed types[1].
Clinico neuroradiological classification of spinal dysraphisms :

1. OPEN SPINAL DYSRAPHISM

- Myelomeningocoele
- Hemimyelomeningocoele
- Myelocoele
- Hemimyelocoele

2. CLOSED SPINAL DYSRAPHISM

A. With subcutaneous mass

- Lipomyelomeningocoele
- Lipomyelocoele
- Meningocoele
- Myelocystocoele

B. Without subcutaneous mass(simple dysraphic states)

- Intradural lipoma
- Tight filum terminale
- Dermal sinus
- Persistent terminal ventricle

Complex dysraphic states

1. Disorders of notochord integration

- Diastometamyelia
- Neuroenteric cyst
- Dorsal enteric fistula

2. Disorders of notochord formation

- Caudal agenesis
- Segmental spinal dysgenesis

MATERIALS AND METHODS

It is a Prospective Observational study.
Study sample: 52 patients
Study period: July 2021 to July2022
Study area:Department of Radiodiagnosis,

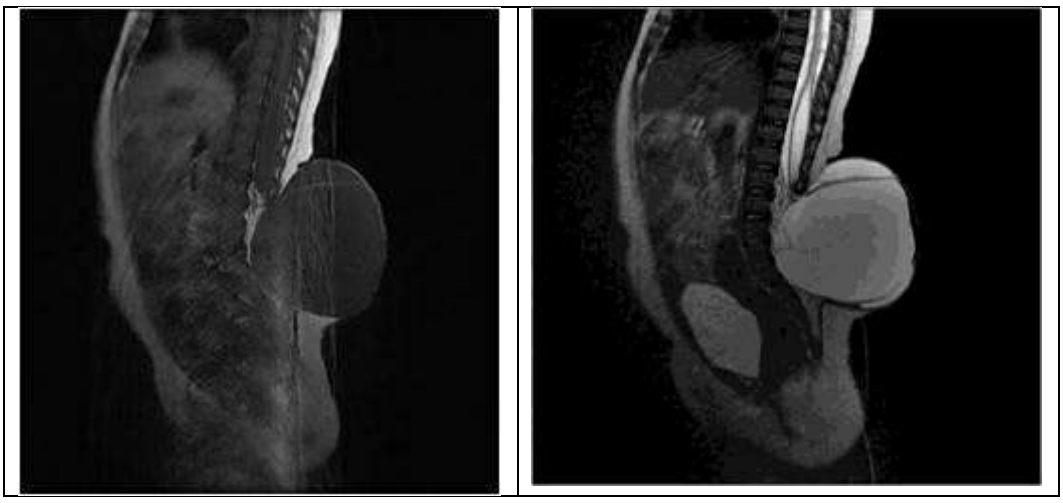
Osmania medical college
Study equipment: MRI was performed on a 1.5 Tesla AVANTO (Siemens Ltd).

- **Inclusion criteria:** patients who presented with various neurological problems and skin stigmata suspicious of spinal dysraphism. Both closed and open types of spinal dysraphism are included in the study.
- **Exclusion criteria:** patients with metallic implants and patients with claustrophobia.
- In all patients clinical history and antenatal history were recorded. Sedation was given in required patients.

IMAGING PROTOCOL

The pulse sequences included

- T1WI (axial ,sagittal)
- T2WI (axial,sagittal ,coronal)
- STIR (sagittal , coronal)
- Images were obtained with an interslice gap of 5.2mm slice thickness of 4mm and a matrix size of 512 x 512.
- On MRI, imaging findings in vertebrae, spinal cord and soft tissues were noted.



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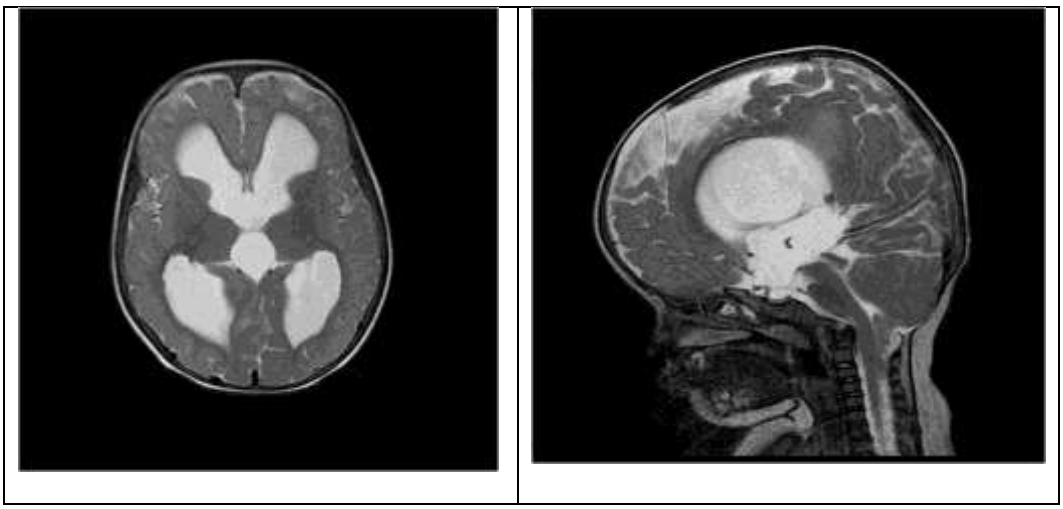

Figure1. T1 and T2 sagittal and axial MRI showing spina bifida with lumbar myelomeningocele with tonsillar herniation in a 1year old female.

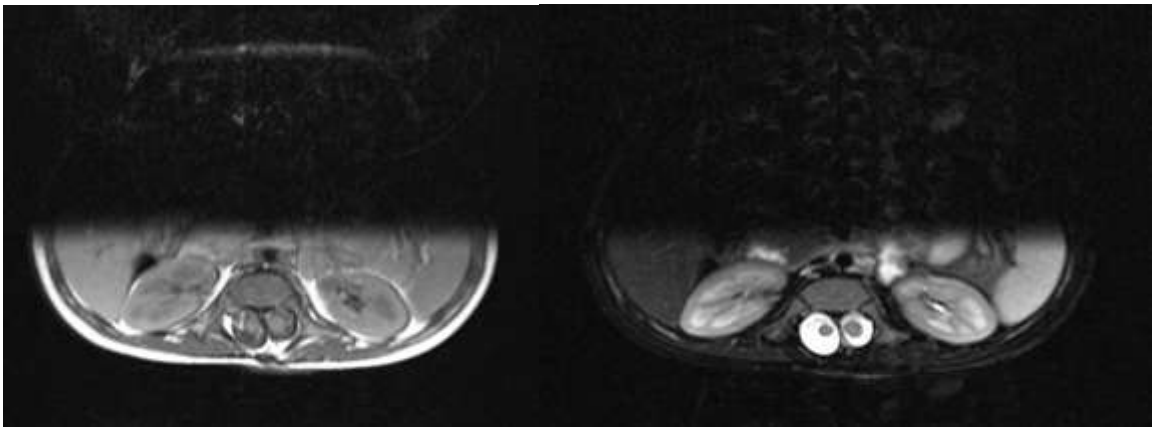

Figure 2. Axial T1 and T2 MRI showing diastometamyelia with a fibrous septum in a 12year old female who presented with tuft of hair in the back since birth.

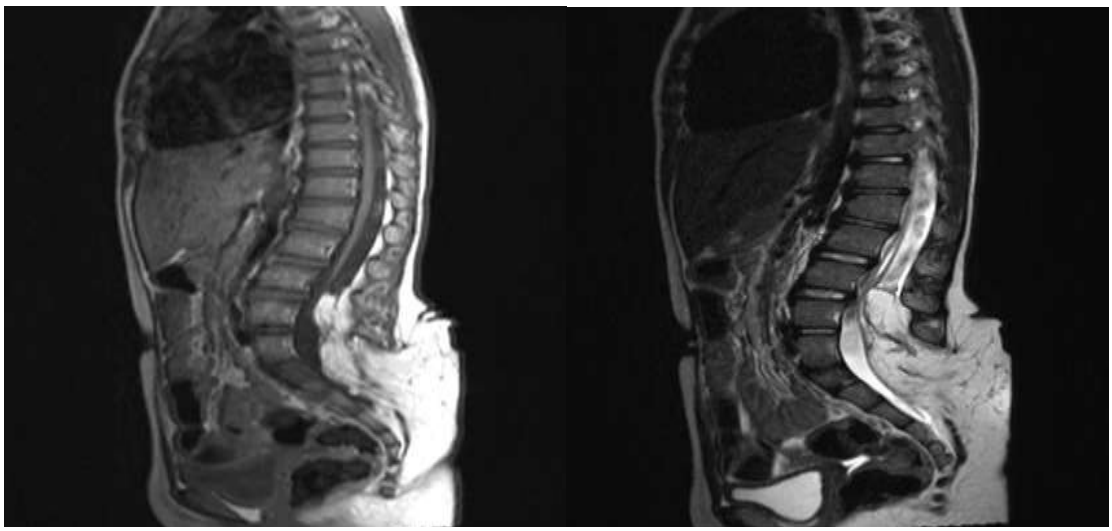

Figure 3. sagittal T1 and T2 MRI showing spina bifida with lipomyelomeningocele in a 7year old male.

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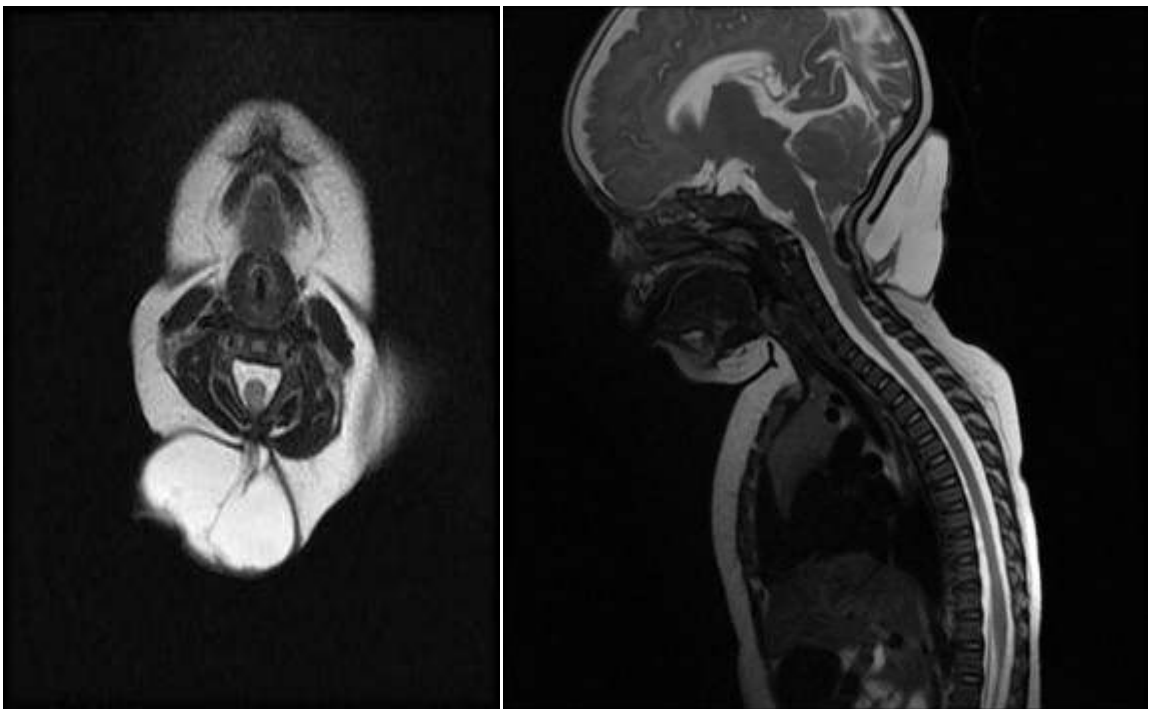


Figure 4.Axial and sagittal T2 MRI showing cervical myelomeningocele in a 2month old female.

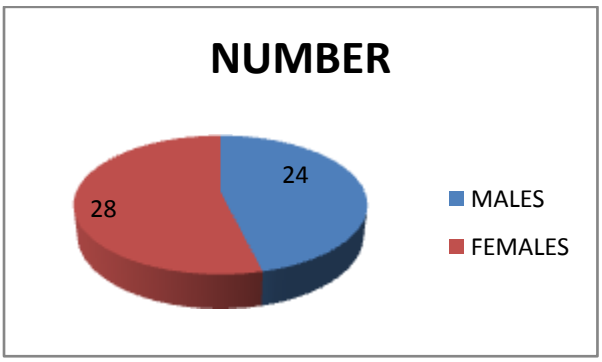
II. OBSERVATIONS AND RESULTS

A total of 52 patients who presented with various neurological problems and skin stigmata suspicious of spinal dysraphism were included in study .

The age of the patients ranged from 3 days to 15yrs. Most of the children are below 1yr of age.
Out of the 52 patients 28 were female patients and 24 were male patients.

Table 1. Gender Distribution

Sl. No	Gender	Number	Percentage
1	Males	24	46.1%
2	Females	28	53.8%



Of the 66 patients, 31 are open spinal dysraphisms and 21 are closed dysraphisms.

Table 2. Type of Dysraphism

Sl. No	Type of dysraphism	Number	Percentage
1	Open dysraphism	31	59.6%
2	Closed dysraphism	21	40.3%

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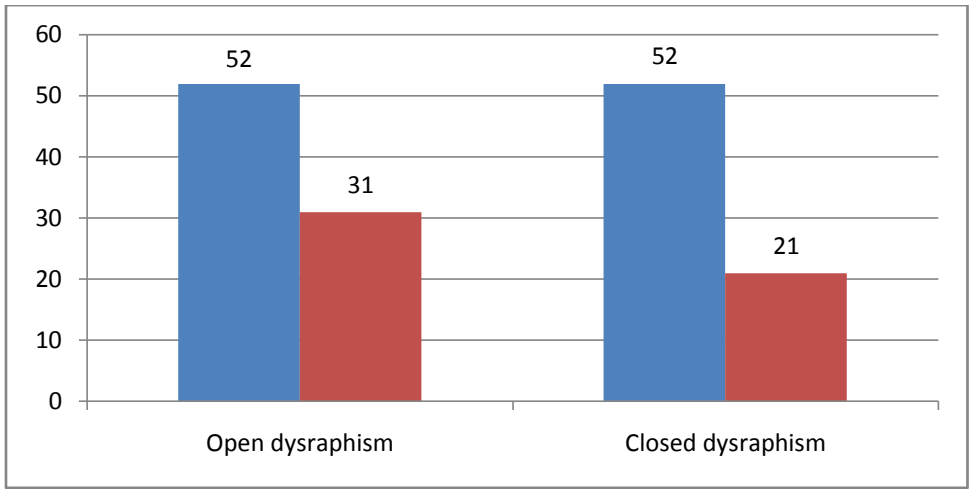
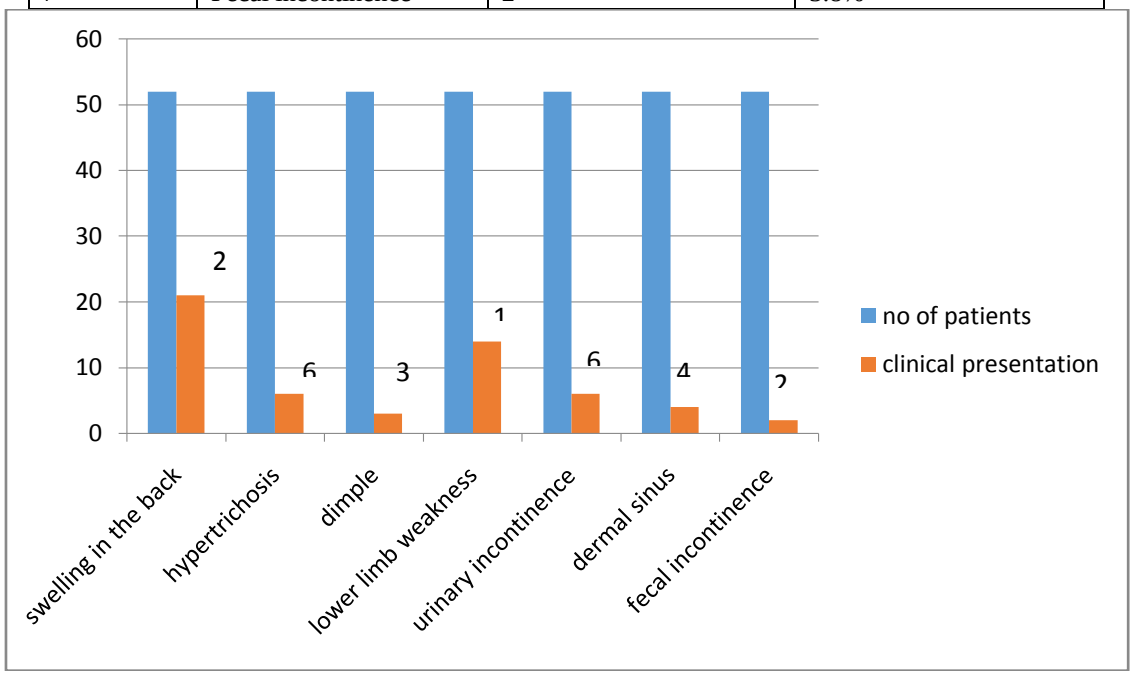


Children with suspected spinal dysraphism can present with various clinical features such as swelling on the back, dimple, hemangioma, dermal sinus,lower limb weakness,bladder and bowel incontinence.

Table 3. Clinical Features:

Sl.No	Clinical Feature	Number	Percentage
1	Swelling in the back	21	40.3%
2	Hypertrichosis	6	11.5%
3	Dimple	3	5.7%
4	Lower limb weakness	14	20.9%
5	Urinary Incontinence	6	11.5%
6	Dermal sinus	4	7.6%0
7	Fecal incontinence	2	3.8%



Among different types of dysraphism myelomeningocele is the commonest.

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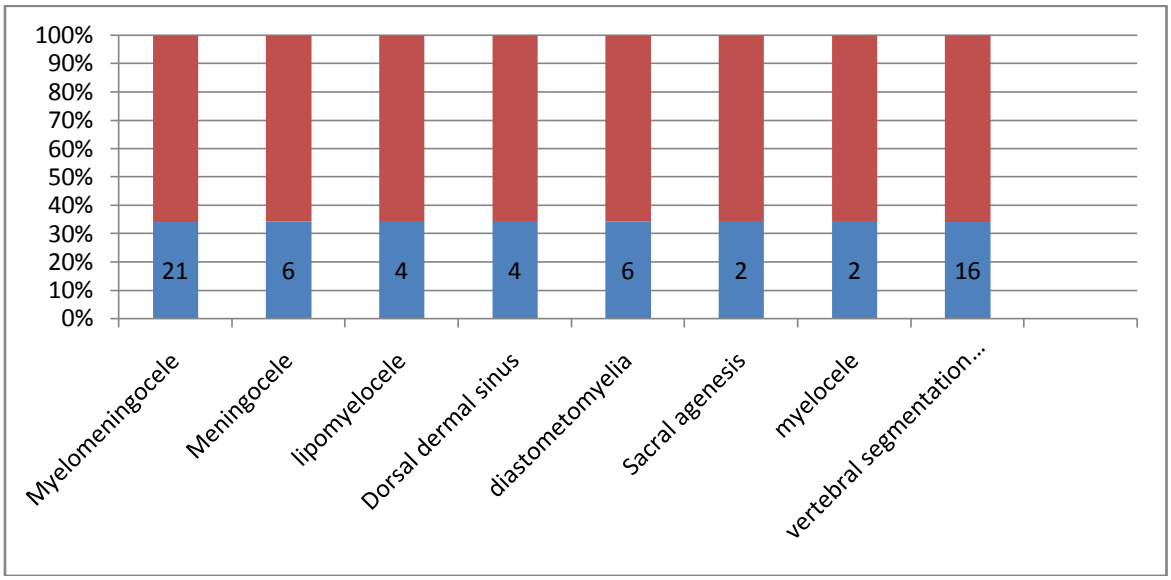
Table 4. Different types of dysraphism

Sl.No	Type of dysraphism	Number	Percentage
1	Myelomeningocele	21	40.3%
2	Meningocele	6	11.5%
3	Lipomyelocele	4	7.6%
4	Dorsal dermal sinus	4	7.6%
5	Diastometamyelia	6	11.5%
6	Sacral agenesis	2	3.8%
7	Myelocele	2	3.8%
8	Vertebral segmentation anomalies	14	26.9%



Location of myelomeningocele: Of the 21 patients in our study, 1 was noted in the cervical region, 2 in the dorsal region, 4 in the lumbar region, 6 in the sacral region, 8 in lumbosacral region.

Table 5. Myelomeningoceles at different locations of spine

Sl. No	Location	Number	Percentage
1	Cervical	1	4.7%
2	Dorsal	2	9.5%
3	Lumbar	4	19.0%
4	Sacral	6	28.5%
5	Lumbosacral	8	38.0%

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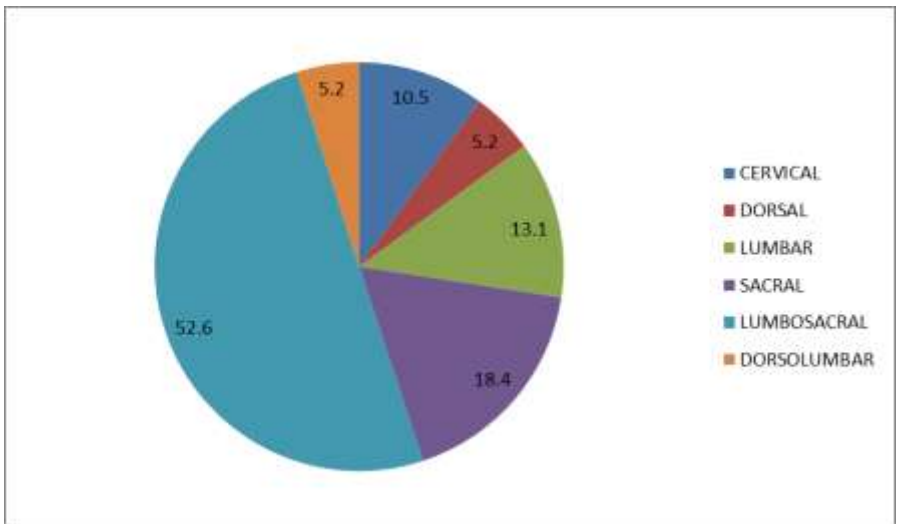
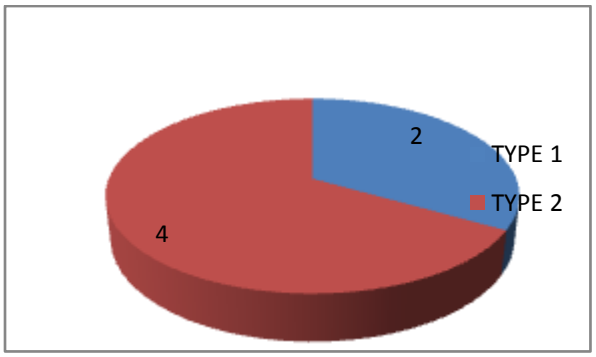


Of 52 patients, 4 Diastometamyelia patients were detected, of which 2 were of type 1 and 4 were of type 2. Two types of Diastometamyelia are noted.

- 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.

Table 6. Type of Diastometamyelia

Sl No	Type of Diastometamyelia	Number	Percentage
1	**Type-1**	**2**	**33.3%**
2	**Type-2**	**4**	**66.6%**



Open defects are associated with abnormalities such as hydrocephalous, Arnold-chiari , syrinx.

Table 7. Closed defects with associated abnormalities

Sl. No	Abnormality	Number	Percentage
1	Scoliosis	3	10.7%
2	Kyphosis	3	10.7%
3	Segmentation anomalies	4	14.2%
4	Syrinx	7	25%
5	Tethered cord	10	35.7%

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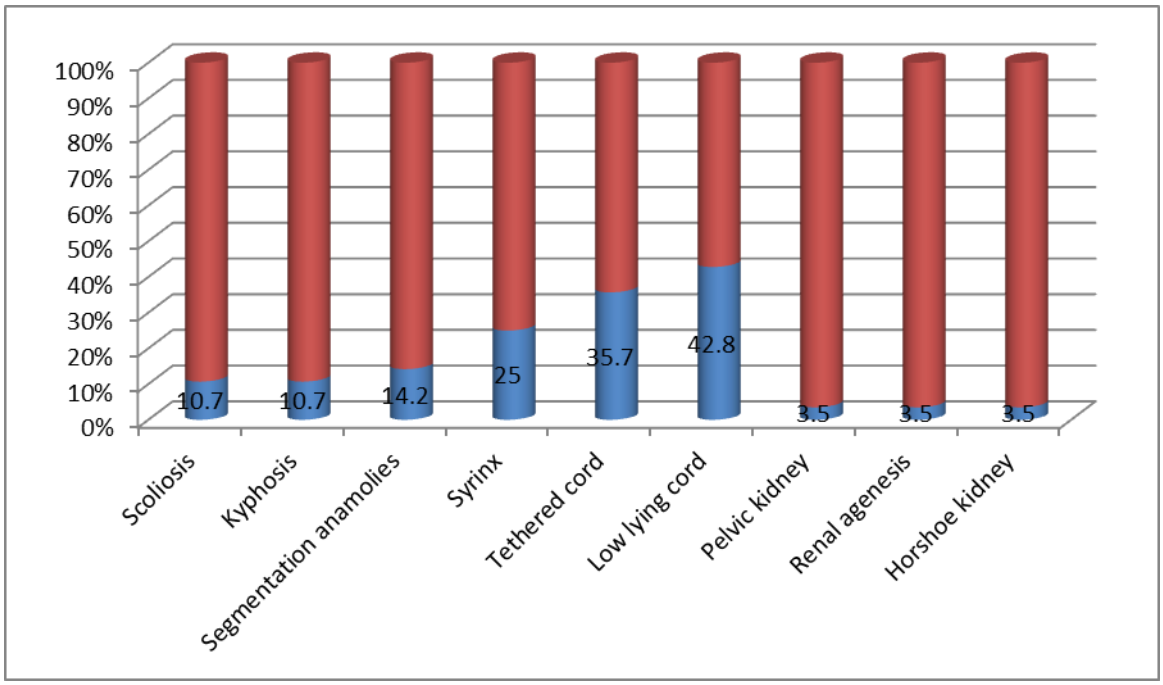
6	Low lying cord	14	42.8%
7	Pelvic Kidney	1	3.5%
8	Renal agenesis	1	3.5%
9	Horseshoe kidney	1	3.5%



Of the 52 cases in our study 40 cases were operated. M.R.I findings were compared with surgical findings.

Table 8. MRI findings in correlations with surgical findings

Sl No	Type of dysraphism	Total no of cases	Number of cases operated	Showing surgical correlation	Lost to follow up
1	Myelomeningocele	38	33	33	5
2	Meningocele	5	4	4	1
3	Myelocystocele	1	1	1	-
4	Lipomyelomeningocele	6	4	4	2
5	Diastometamyelia	6	3	3	3
6	Dorsal dermal sinus	5	4	4	-
7	Sacral agenesis	2	-	-	2
8	Tethered cord	3	3	3	-

III. CONCLUSION

The age of the patients in our study ranged from 3 days to 15yrs. Most of the children are below 1yr of age.Female predominance was noted in our study which constituted 53.8%. Open defects (59.6%) predominated in our study than closed defects (40.3%). Swelling in the back is the commonest clinical feature in our study which constituted 21 (40.3%). Myelomeningocele is the commonest type of dysraphism which constituted 21 (40.3%).In Myelomeningoceles, lumbosacral region is the commonest location which constituted 8 (38.0%).Among Diastometamyelia, type 2 predominated in our study (66.6%).Open defects are associated with abnormalities such as hydrocephalous, Arnold-chiari , syrinx. Low lying cord constituted 57.1 % and tethered cord constituted 33.3%.Most of the myelomeningoceles in our study are associated with abnormalities.

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

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