

ROLE OF MAGNETIC RESONANCE IMAGING (MRI) IN EVALUATION OF SPINAL DYSRAPHISM

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ABSTRACT

Background: Spinal Dysraphism includes a spectrum of congenital disorders caused by incomplete or abnormal closure of the neural tube during early embryogenesis. As a result, fusion of the midline spinal elements is either absent or incomplete. MRI is an excellent imaging modality for visualizing the spinal cord of patients of all ages. It is the imaging modality of choice for defining complex spinal dysraphism. Most attractive features of MRI that made it superior to conventional radiography and CT myelography are its ability to image the cord directly without the use of contrast or ionising radiation, absence of bone artefacts and its multiplanar capability. **Methods:** A Cross-sectional Observational study was done in 32 patients. Patients who were diagnosed or provisionally diagnosed as cases of spinal dysraphism, irrespective of age and sex, based on the Clinical profile and imaging profile as preliminary findings on radiographs/Ultrasonography and incidentally detected cases on either Radiographs, CT, USG or MRI were included in our study. **Results:** In the pre CT era the combination of a precise clinical pattern with x ray images of the spine could prompt a suspicion of Spinal Dysraphism. Ultrasonographic evaluation for spina bifida includes both spinal and cranial imaging. Computed tomography better showed osseous abnormalities associated with the spinal congenital malformations. MR imaging is considered as the primary imaging modality in the evaluation of the paediatric spinal canal. Congenital anomalies as well as neoplastic, inflammatory, and traumatic disorders can be reliably evaluated with MR imaging. **Conclusions:** MR imaging clearly reveals an excellent tissue differentiation, especially of lipomatous tissue. The reproducible and comprehensive section planes, as well as the relative operator-independence are also well appreciated facts. MRI is a single safe, non-invasive and quick method of describing the gamut of findings in patients with spinal dysraphism.

KEYWORDS: Magnetic Resonance Imaging, Spinal Dysraphism**INTRODUCTION**

Spinal dysraphism in India is a major public health problem with added problems resulted from lack of medical personnel and diagnostic facilities. Spinal Dysraphism includes a spectrum of congenital disorders caused by incomplete or abnormal closure of the neural tube during early embryogenesis. As a result, fusion of the midline spinal elements is either absent or incomplete.^[1]

It is a generic term encompassing a wide spectrum of congenital anomalies with the common feature of faulty fusion of embryonic dorsal midline structures including spinal cord, its skeletal investment and all the tissue layers of the back.

Spinal Dysraphisms are reported in 0.3 to 0.4/1000 live births in western society. The incidence has significantly decreased over the last 5 decades, all over the world;

however, fall in incidence is much lesser in developing countries with poor socio-economic status, educational background and financial condition of general population. In India, spinal dysraphism is still a major public health problem with added problems resulted from lack of medical personnel and diagnostic facilities. The estimated incidence of spinal dysraphism in India is 1 to 3/1000 live births.^[2] Open spinal dysraphism presents as a swelling over the back which can be noticed at birth. Symptoms appear mainly due to CSF leak and the exposed spinal cord. Sensorimotor deficits, sphincter dysfunction and meningitis are its direct consequences.

Occult spinal dysraphism is not as evident on birth as is open spinal dysraphism and patients are usually neurologically intact at the time of birth. However these cases can be suspected at birth itself as most of these cases have some dorsal cutaneous stigmata like subcutaneous mass, skin dimple, hemangiomas, hairy patches, tail like appendage, denuded skin patch or scar

like patch over the back at birth.^[3] if missed at birth, later on it can present as symptoms like bed wetting, improper toilet habits, back pain, abnormal gait etc.

CLASSIFICATION OF SPINAL DYSRAPHISM

Open Spinal Dysraphisms: not covered by intact skin

Myelocele flushed with skin surface	Neural placode
Myelomeningocele protrudes above skin surface	Neural placode
Hemimyocele with diastematomyelia	Myelocele associated
Hemimyelomeningocele associated with diastematomyelia	Myelomeningocele

Closed/Occult Spinal Dysraphisms: covered by intact skin

With a subcutaneous mass

Lipomyelocele interface within the spinal canal	Placode–lipoma
Lipomyelomeningocele interface outside of the spinal canal	Placode–lipoma
Meningocele	Herniation of CSF-filled sac lined by dura
Terminal myelocystocele herniating into posterior meningocele	Terminal syrinx
Myelocystocele herniating through posterior spina bifida	Dilated central canal

Without a subcutaneous mass

Simple dysraphic states	
Intradural lipoma dural sac	Lipoma within the
Filar lipoma thickening of filum	Fibrolipomatous
Tight filum terminale shortening of filum	Hypertrophy and
Persistent terminal ventricle within conus medullaris	Persistent cavity
Dermal sinus fistula between neural tissue	Epithelial lined and skin surface

Complex dysraphic states

Dorsal enteric fistula bowel and skin surface	Connection between
Neurenteric cyst of dorsal enteric fistula	More localized form
Diastematomyelia (Split cord)	Separation of cord into two hemicords
Caudal agenesis agenesis of spinal column	Total or partial
Segmental spinal dysgenesis segmentation anomalies	Various

METHODS

This Cross-sectional Observational study was conducted in the Department of Radio-diagnosis at P.G.I.M.E.R &

Dr. Ram Manohar Lohia Hospital, New Delhi from 1st November 2012 to 31st March 2014. Approval from hospital and Institutional Ethical Committee was obtained prior to initiation of the study. Patients who were diagnosed or provisionally diagnosed as cases of spinal dysraphism, irrespective of age and sex, based on the Clinical profile and imaging profile as preliminary findings on radiographs/Ultrasonography and incidentally detected cases on either Radiographs, CT, USG or MRI were included in our study. A written informed consent was taken from all patients. A detailed history was taken with complete physical and systemic examination of the patient. Relevant biochemical investigations were done wherever required.

Clinical Evaluation: A detailed history was taken with emphasis on duration of presence of lump, presence or absence of leak, fever, sensory or motor deficit, bladder or bowel disturbances, vomiting, difficulty in vision, enlarged head size etc. in cases of open spinal dysraphism and h/o of presence of any cutaneous stigmata, deformity of spine, back pain, any neurological deficit or bladder bowel disturbances in case of closed dysraphism cases. A complete general physical examination, local examination and neurological assessment of the patients were done.

Ancillary radiological investigations: X ray whole spine A-P and Lateral views were done in all clinically suspected cases, however X ray P-A view was done in place of A-P view in open or closed spinal dysraphism cases presenting with large back mass, which made the patient positioning difficult for the routine A-P film. Oblique views were taken to assess the anomalies involving posterior elements of vertebrae, when suspected on A-P/ P-A or lateral view. Imaging evaluation of spinal dysraphism was tailored based on the patient presentation and physical examination as well as clinical suspicion.

Ultrasonography (HR USG): Ultrasound of spine was performed as an initial imaging modality in patients having low clinical suspicion of spinal dysraphism and those in which bony defects present in the vertebrae could be used as an acoustic window.

Computed Tomography: CT was performed only in cases where bony anomalies could not be assessed in detail on plain radiograph and MR scanning. CT was performed on Philips Brilliance 40- slice Multi- detector CT scanner.

MRI spine was performed using State of Art Siemens 1.5 Tesla Magnetom Symphony MR system using appropriate phased array coils according to the specific protocol. The MR sequences included: T1_spin echo, T2_ spin echo, T2 FLAIR,STIR (fat suppression), Gradient Sequences (GRE) for blood and calcification, Special cord sequences.

RESULTS

The present study was conducted in the Department of Radiodiagnosis, PGIMER, Dr. RML Hospital, New Delhi. A total of 32 cases who were diagnosed or provisionally diagnosed as cases of spinal dysraphism were included in our study. The study group underwent a clinical imaging workup including detailed history, general and neurological examination, followed by MR imaging. The diagnosis of these congenital

malformations was made on the basis of MR scans in conjunction with classical clinical findings. The **youngest patient** in our study was a 15 days male child diagnosed as Myelomeningocele and **eldest one** was a 28 years female diagnosed as a case of Dorsal dermal sinus. A patient of **occipital encephalocele** presented as closed dysraphic defect. **Myelomeningocele** accounted for majority in our cases (Figures 1 & 2).

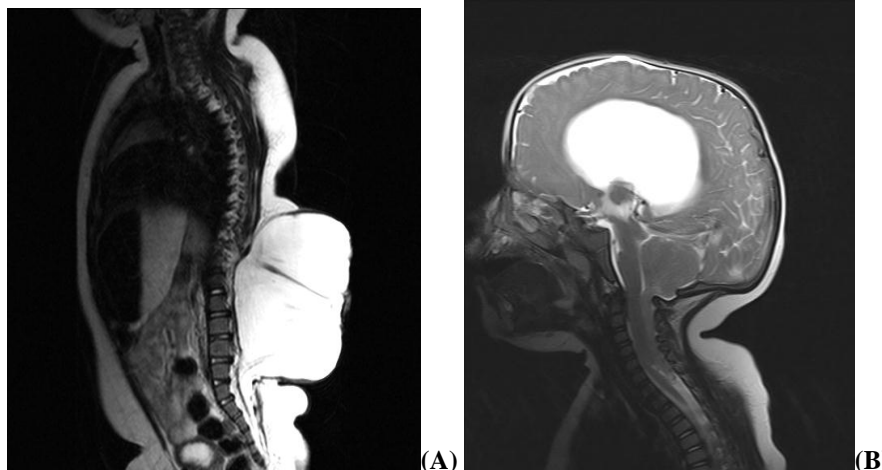


Figure 1 Myelomeningocele (A): T2W Sagittal image of spine of the patient showing a large myelomeningocele sac involving dorsal, lumbar and sacral region as well.(B): T2W Sagittal image of brain of the same patient showing cerebellar herniation, slit like fourth ventricle, tectal beaking and other features of Arnold Chiari type-II malformation.

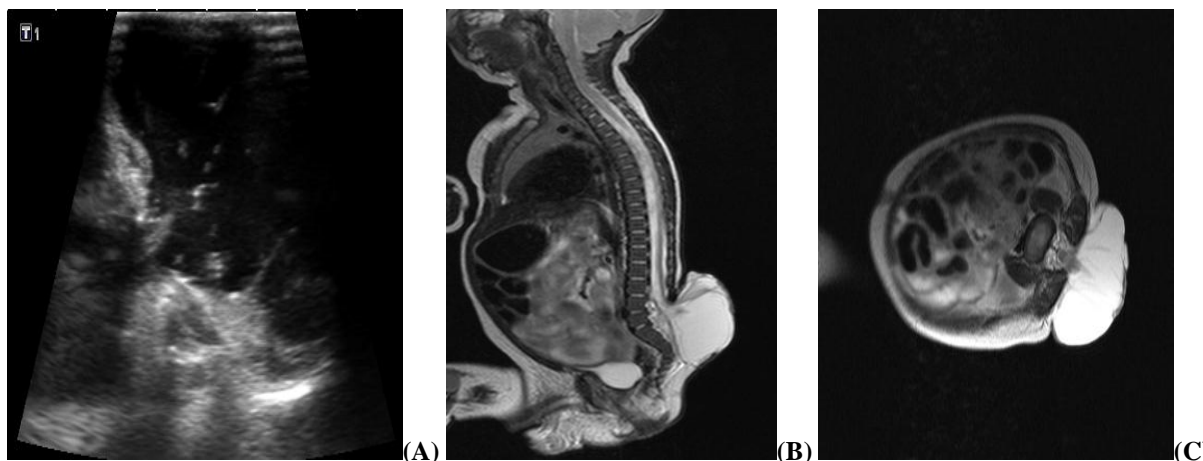


Figure 2. Myelomeningocele sac with neural placode and nerve roots (A) showing Ultrasonographic Images (B) Sagittal image and (C) Axial image

Maximum no. of patients presented within 1 yr. of age. Incidence of **Myelomeningocele** in our study was **17 (53.12%)**. Dysraphism involved different parts of the spine. The defect was more commonly seen in **14(43.75%) males** in our present study, which mostly presented within first 12 months of life. The most common location for the defect was dorsolumbar, lumbosacral and sacral spine. Each location contributed to **4(23.52%)** patients respectively. The patients with myelomeningocele had associated anomalies of brain such as Chiari malformation II (9 cases = 52.94%) & hydrocephalus (4 cases = 23.52%).

Chiari malformation II was observed in 9 (52.94%) patients (7 males & 2 females with 8 of them presenting within 1st year of their life) and all these patients were cases of myelomeningocele. Spinal Cord was tethered to the sac in all cases of myelomeningocele.

Out of 32 patients in our study, incidence of **myelomeningocele** was **17(53.12%)**. In our study group of 32 patients, the incidence of dermal sinus was **1(3.12%) female** patient. Cutaneous stigmata of **hair tuft and skin dimple** was present. In **Lipomyeloceles/Lipomyeloschisis**, the lipoma attaches

to the open neural placode within the confines of the spinal canal.



Figure 3: Lipomyelocoeles (A) shows a child presenting with a skin covered lower back mass.(B) T1W Sagittal image of lumbosacral spine of the same child showing features of Lipomyelocoele with low lying tethered cord.

In cases of **Type I split cord malformations** the hemi cords were in different dural sheath with osteocartilaginous spurs. While in cases of **Type II split cord malformations** the hemicords were in the same dural sheath with absence of osteocartilaginous spur. Incidence of meningocele in our study was **6(18.75%)**. One case of **anterior sacral meningocele** was associated with caudal regression syndrome. Two **(6.25%)** cases with **tight filum terminale** were observed in our study. Both of them were observed in female patients. None of the cases presented as an isolated anomaly, one of them was associated with diastematomyelia and the other one with caudal regression syndrome. We encountered 25(78.12%) cases of **tethered cord** in our study. All of them were tethered posteriorly. Thirteen (40.62%) cases of **hydrosyringomyelia** were noted in our study.

DISCUSSION

Developmental disorders of the paediatric spine are a group of congenital malformations commonly referred to as spinal dysraphism. These malformations are characterized by incomplete or absent fusion of midline mesenchymal, bony, and neural structures.^[4] Embryologically, a detailed cascade of events of spinal cord development occurs through the three consecutive periods of gastrulation (2-3 weeks), primary neurulation (3-4 weeks), and secondary neurulation (5-6 weeks) which result in the proper formation of both the musculoskeletal and neural elements of the spine. Alterations in these embryologic steps can result in one or more congenital abnormalities of the spine.

Before the advent of magnetic resonance (MR) imaging the diagnosis of the spinal anomalies especially the occult one was relied on strong clinical suspicion coupled with a specifically tailored radiologic examination with X rays, myelography and/or computed tomography (CT). But now Magnetic resonance imaging (MRI) has emerged to be the imaging modality of choice in these cases.^[5] Spinal real time sonography is

inexpensive, easily available and can be used as a screening modality in cases in which an acoustic window is present.^[6] Computed tomography better demonstrates osseous abnormalities but the radiation dose and lack of Multiplanar capabilities did not make CT as the investigation of choice for spinal evaluation.^[7]

MAGNETIC RESONANCE IMAGING

At present, MRI is an excellent imaging modality for visualizing the spinal cord at all ages. A clear advantage of MR imaging is the excellent tissue differentiation, reproducible and comprehensive section planes, as well as the relative operator-independence. In our study, our aim in the evaluation of spinal dysraphism was to obtain the complete information of the anomaly with high spatial and contrast resolution.

MRI PROTOCOL: In the past, MR imaging of spine was largely confined to sagittal plane only and secondarily if required images in the axial and coronal plane were also obtained. In our imaging protocol we performed T1W spin echo sequence in sagittal and axial plane, T2W spin echo pulse again in sagittal & axial planes, T2w sagittal scan with extended field of view to include craniovertebral junction and brain and an additional T2W coronal scan. MRI was found to provide the most complete information of the anomalies.

Sagittal & axial T1 & T2 W scans were adequate for complete diagnosis predominantly in all cases. Axial scans were used to assess the exact level of conus medullaris and to measure the thickness of filum at L5-S1 level in cases of tight filum terminale. Addition of **whole spine scanning** added information about other multiple associated anomalies like Syringomyelia in 13(40.62%) cases where the syrinx was above/below the dysraphic lesion and low lying tethered cord in 1(3.12%) case in which the dysraphic lesion was above the L2 vertebral body level.

Coronal scans were found useful for demonstrating back pressure changes in kidneys & ureters in 3(9.37%) cases, costovertebral anomalies in 3(9.37%) cases, split cord malformations 7(21.8%) cases and extent of syringomyelia in 13(40.62%) cases.

Brain screening revealed other associated anomalies like Arnold Chiari malformation in 9 (28.12%) and hydrocephalus in 4(12.5%) of cases.

The usefulness of the coronal view and the whole spine screening in sagittal view has also been mentioned in previous studies. In year 1999 L. Santiago M et al^[8] concluded that conventional three-plane lumbosacral MR imaging, thick-section sagittal spin-echo T1-weighted localizer sequence followed by thin-section sagittal, axial, and coronal spin-echo T1-weighted sequences provide better information than the fast screening two-sequence protocol included thin-section sagittal and axial spin-echo T1-weighted sequences They also suggested MR imaging of the entire spine in patients with clinical findings above the lumbosacral level or with symptoms and signs that may be due to conditions other than Dysraphic Myelodysplasia despite a negative lumbosacral fast screening MR study.^[8]

Similarly S. Chopra et al^[9] conducted study in year 2001 and suggested to include as much of the brain as possible in order to screen for Chiari malformations and hydrocephalus simultaneously or separately. They also highlighted the usefulness of coronal scans in cases of diastematomyelia. In our study MRI proved to be the best modality for complete evaluation of myelomeningoceles and same was also concluded by Byrd SE et al^[10] who evaluated 755 children with myelomeningoceles radiologically and they found MRI to be the best modality to evaluate the posterior fossa and total spine within 1st year of their life.

Kumar R and Singh SN^[11] also found myelomeningocele as the most common anomaly (in 72% cases) and lumbosacral region (in 38% cases) as the most common site of occurrence of spina bifida in their study on spinal dysraphism in northern India. Lumbosacral dermal sinuses are usually associated with tethering of the spinal cord & low position of the conus in 80% cases^[12]. however tethering of cord was not observed in the patient in our study. E.Schijman^[13] in year 2003 reviewed 22 cases and concluded that CT scan is useful for the evaluation of vertebral bodies and posterior arch abnormalities and spur characteristics in SCMs Shen SH et al^[14] in their study on intradural lipoma suggested that though both CT and MRI can reveal the fat component of the lipoma, MRI is superior to CT in demonstrating its relationship with adjacent normal nerve tissue. Currarino triad^[15] was present in our case that includes anorectal malformation (operated for anal canal stenosis), sacrococcygeal osseous defect (partial sacral agenesis) and a pre sacral mass (anterior sacral meningocele) in our case.

CONCLUSIONS

Most of the spinal cord anomalies occur due to abnormalities of primary neurulation during embryogenesis of spinal cord, i.e. due to Nondisjunction and Premature disjunction. There is wide age group of presentation of spinal dysraphism, however most of the cases of spinal dysraphism presented within 1st year of their life and percentage of cases decreases with age.

MRI is a single safe, non-invasive and quick method of describing the gamut of findings in patients of spinal dysraphism. It excellently demonstrates all the morphological details required to be evaluated in these patients before neurosurgery. Evaluation of newer MR imaging techniques such as spinal cord motion sequences and CSF flow studies for CSF flow dynamics are suggested, the clinical effectiveness of which has not yet been fully determined.

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