

High Resolution Ultrasound in Children with Spinal Dysraphism: Role Revisited

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A B S T R A C T

Introduction: Spinal dysraphism is an umbrella term which encompasses anomalies resulting from derangement in the embryogenic cascade during early fetal life (2nd-6th gestational weeks) resulting in incomplete fusion of neuro-osseomesenchymal structures of the spine. It is a common birth defect with significant morbidity and mortality in children and a prevalence rate ranging from 1.9-66.2 per 10,000 live births in South-East Asian region. Present study was done with a detailed evaluation of the role of high resolution ultrasound in children over a wide age group with spinal dysraphism.

Material and methods: We did a prospective study over a period of 2 years from October 2017 to October 2019 in the Department of Radiodiagnosis and Paediatric Surgery, JN medical College, AMU Aligarh. Clinical examination followed by imaging was carried out. All the patients were subjected to high resolution ultrasound of spine initially, followed by MRI the same day. Seven objective predetermined imaging parameters and final diagnosis of each patient on both imaging modalities were compared statistically for sensitivity, specificity, positive predictive value and negative predictive value.

Result: There were fifty patients and were divided into four age. Most patients were in the age group of 0-3 months and amongst these ~70% cases were of open spinal defects. Myelomeningocele was the most common abnormality amongst open defects (~38%) and Lipo-myelomeningocele was the most common defect amongst closed defects (~24%). Seven most relevant imaging parameters were chosen for comparison in each patient. As compared with MRI, across all 4 age groups, on high resolution ultrasound, 3 of 7 imaging parameters viz, abnormalities in posterior elements of vertebra, position of conus medullaris and spinal cord morphology whether split or normal showed more than 90% sensitivity & about 100% specificity, another 3 parameters including expansion of ventral subarachnoid space, filum terminale morphology and fatty mass in and around the defect site with any intraspinal extension showed ~80-90% sensitivity and ~100% specificity and 1 parameter, central canal diameter (normal/dilated) showed <75% sensitivity and 100% specificity of detection on HR-USG.

Conclusion: The overall diagnostic sensitivity of high resolution USG spine for spinal dysraphism was high across 0-14 years of age group and at par with MRI in the age group of 0-3 months. Due to the high sensitivity and specificity observed for infants in 0-3 months age group, HR-USG would suffice as a diagnostic modality for early operative decision. MRI should be reserved only where USG is difficult to perform practically as a bulky sac, ruptured myelomeningocele and suspected complex anomalies. This would be highly cost effective and expedite management of cases coming from lower socio-economic strata.

Keywords: Congenital, Spinal Dysraphism, High Resolution Ultrasonography, MRI

INTRODUCTION

Spinal dysraphism encompass serious congenital defects involving the neural development at fault in the early embryonic life. The estimated incidence of spinal dysraphism is about 1-3/1000 live births.¹⁻⁴ Incidence is on decline in the developed nations due to better antenatal care (pre-conceptional folic acid) and early antenatal detection.^{5,6,7} After birth, most of the cases are detected in early infancy due to grossly abnormal phenotype, however, many present late even in adolescence. In developing countries in the low socio-economic strata, cases may present late with sensorimotor disturbances or dysfunctional sphincter mechanism of

bladder/bowel warranting investigation. Tortori-Donati et al, broadly categorised it into open spinal dysraphism (OSD) where the neural placode is exposed to external environment and closed spinal dysraphism (CSD) where it is skin covered.⁸ There are four abnormalities under OSD - myelomeningocele (most common ~ 98% cases) and the other 3 relatively rare - myelocele, hemimyelomeningocele, hemimyelocele. Nearly all OSD are associated with Arnold Chiari (II) malformations. CSD is a relatively heterogeneous group subclassified on presence/absence of subcutaneous mass overlying the defect. CSD with a subcutaneous mass includes - lipo-myelomeningocele, lipomyelocele,

terminal myelocystocele and a meningocele. CSD without any subcutaneous mass includes simple dysraphic states - intradural lipomas, filar lipomas, dermal sinuses, persistent terminal ventricle, and complex dysraphic states - Diastematomyelia, Neuroenteric cyst, Caudal agenesis and segmental spinal dysgenesis.⁹

The most commonly performed imaging in all such cases is MRI spine which has its advantage of giving excellent soft tissue characterisation without using any ionising radiation.¹⁰ Ultrasound with similar advantages despite being cheap and widely available has not been adequately exploited for this purpose. The role of ultrasound spine in children has resurfaced since the advent of high frequency ultrasound probes making high resolution imaging possible. Recently; we are witnessing a tremendous rise in the health care costs. In a country like India, primary health care delivery, in practice, is not equipped to handle patients of congenital defects adequately. Imaging facilities like MRI are mostly not available at peripheral centres and any delay in diagnosis only increases the chances of complications, morbidity and mortality.¹¹

This generates a demand to objectively quantify the sensitivity, specificity, positive predictive value and negative predictive value parameters of high resolution USG against the existing gold standard modality MRI, in terms of not just the final diagnosis, but also in detecting specific pointers to each diagnosis. Very few studies exist as of today that explore and compare the sensitivity and specificity parameters of detection of these imaging pointers towards the final diagnosis on high resolution USG with MRI. In our study, a fresh approach of comparing statistical significance of specific pointers to diagnosis on high resolution USG across a wide age group of children from neonates to 14 years of age was done.

MATERIAL AND METHODS

Due clearance from the institutional ethics committee for this study was obtained. Fifty patients of either sex, suspected of spinal dysraphism based on clinical features, in the age range 0-14 years were then evaluated during the two year study period (October 2017-October 2019). Patients with previous surgery for spinal dysraphism or any contraindication to MRI (ferromagnetic implants etc.) were excluded from the study group.

A high resolution ultrasound was performed using Mindray DC-8/ Siemens ACCUSON X700 ultrasound machine with high frequency linear probes (5-12 MHz). A calm and quiet child after adequate feed was allowed to fall in sleep naturally and then taken to USG room. The child was placed preferably prone on a pillow to relax the normal lumbar and cervical curves. Sometimes, a decubitus position was also used.² Due to the lack of ossification of the posterior arch of the spine in infants, ultrasound waves are able to penetrate and image the spinal canal structures. Gradually as the posterior spinous elements start to ossify in infants after 6 months of age, reduction is brought in image quality. But an inherent defect in the posterior spinal osseo-cartilaginous elements gives a persisting acoustic window through which ultrasound beams can very well penetrate for visualisation of

spine at any age.^{12,13} By angulating the probe cranially and caudally from directly above the defect, significant portions of spine could be imaged (3-4 vertebral segments above and below defect site). Counting of the vertebra from sacrum (S4-lowermost ossified sacral vertebra at birth) upwards and then from 12th rib (T12 level) downwards is done to reach to the exact level of defect. Alternatively, lumbo-sacral junction is identified using sacral angulation as an aid.¹⁴ Necessary gray scale images were obtained in axial, sagittal and parasagittal views at site of abnormality and then whole spine was screened. The images were taken from directly above the defect wherever possible (except in cases of rupture where a relatively safe parasagittal view under strict asepsis, was the only view taken). Brain screening wherever possible was performed (in children <18 months through unfused anterior fontanelle). Asepsis was maintained by the use of single use sterile gel pouches and sterile probe covers in cases of open defects and in cases of rupture. After the USG examination sterile gauze with normal saline was used to clean the area. Bactigrass dressing was applied if needed. Use of copious amount of jelly to provide acoustic window with minimal pressure over defect site was frequently practiced.

This was followed by MRI spine of the patient on the same day on 1.5 Tesla MRI scanners MAGNETOM AVANTO FIT Siemens. Most patients required one or the other form of sedation (oral triclofos-chloral hydrate/IV midazolam). Body coils for spine imaging and head coil for brain imaging were used.

Images were obtained using FOV of 200-250 mm in infants and young children and 250-350 mm in older children (10-14 years) with slice thickness of 3 mm, slice interval of 0.5-1.0 mm, T1WI: TR/TE/Flip Angle = 600-700/9-12/90° and T2WI: TR/TE/Flip Angle = 3500-4000/90-110/150°. Sagittal, axial and coronal images of spine were obtained. First, in turbo spin-echo sequences -T1WI and T2WI were taken in the sagittal plane followed by T2WI in axial plane at defect site and coronal plane STIR images of whole spine to look into spinal curvature abnormality and spinal segmentation dysgenesis. Then, T2WI-(3D -Space seq. focussed at defect site), HASTE MR Myelography, T2 axial brain screening (for associated Arnold Chiari II malformations) and any additional sequences (if needed) were obtained.

Findings of each modality were recorded in separate proformas for each patient. After going through various previous studies conducted on this subject, we identified 7 objective imaging parameters that are very well appreciated on both high resolution spinal ultrasound and MRI in children of spinal dysraphism and decided to compare them across 4 age groups (0-3 months, 3 months-1 year, 1-5 years and 5-14 years) and also the final diagnosis given on each modality.

Seven imaging parameters used for comparison are -

1. Abnormalities in posterior elements of vertebrae
2. Expansion of ventral subarachnoid space
3. Position of conus medullaris
4. Spinal cord morphology (normal/split)
5. Central canal diameter (normal/dilated-syrinx)
6. Filum terminale morphology

7. Fatty mass in and around defect site with any intraspinal extension.

These imaging parameters were objectively looked for. Wherever the finding could not be assessed / was ambiguous on either of the imaging modalities, it was recorded as such. Final diagnosis on each modality at the end of imaging was also recorded in proformas to be compared later on.

STATISTICAL ANALYSIS

The descriptive statistical approach was used to describe findings using numbers, percentage, and arithmetic mean. Comparison between USG and MRI findings with MRI as gold standard imaging modality was done calculating sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV).

RESULT

Out of 50 cases included in our study 28 (56%) were male and 22(44%) females with a male to female ratio of 1.2:1. The mean age of presentation was 1 year and 8 months. Maximum number of patients presented in the age group of 0-3 months (44%) followed by patients between 3months to 1 year (24%). Together nearly 68% cases presented before 1 year of age and 32% cases presented after 1 year of life. Most commonly observed clinical abnormality was the presence of a sac in the back region (58%) and the second most common was the presence of a subcutaneous bulge/mass in the back (30%). Complaint of urinary / bowel incontinence was present in 07 cases (14%) and lower limb weakness was present in 06 cases (12%). Cutaneous stigmata like sacral dimple was found in 3 cases (6%) and a case each of hyperpigmented patch, tuft of hair with a dermal sinus, skin tag (2% each) in lumbar region was seen. The most common site of abnormality was lumbosacral region, in 26 cases (52%), followed by dorso-lumbar region in 14 cases (28%), with sacral region involved in 08 cases (16%) and cervical the least in only 02 cases (04%). Using Tortori-Donati et al clinico-radiological classification⁸, amongst the 50 cases under study, 26 cases (~52%) were of closed defect and 24 cases (~48%)were of open type. Using MRI as the gold standard imaging modality, amongst the closed defects the most common finding was lipomyelomeningocele in 12 out of 26 cases (~46%). Figure 1 (a &b) show MRI images of a case of lipomyelomeningocele with corresponding high resolution ultrasound images Figure 1 (c&d). Meningocele was the predominant finding in 05 (~19.2%) of closed defects [Figure 2-a,b]. Lipomyelocele was

found in only 02 cases of the closed type (~7.6%). We did not encounter any case of a terminal myelocystocele during our

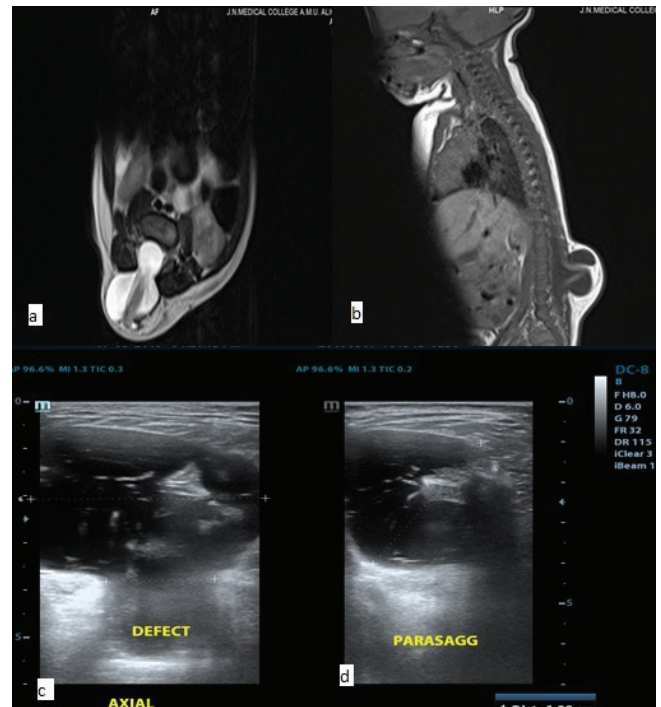


Figure 1: (a,b): T2 weighted axial and T1 weighted sagittal images showing herniated spinal cord and neural elements in a case of lipo-myelomeningocele. (c,d): Transverse and parasagittal views of high resolution ultrasound spine in the same patient.

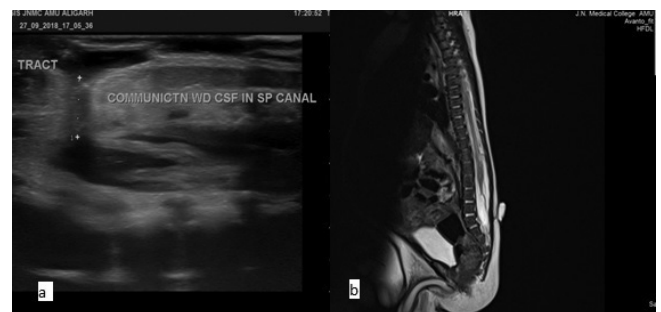


Figure-2: Sagittal high resolution USG image at L5 level of spine shows anechoic CSF containing sac with small communication with spinal canal via defect in posterior element of L5 suggestive of posterior meningocele .T2 weighted sagittal MRI spine in the same patient confirming the findings of USG.

Imaging parameter	HR-USG +	MRI +	Sensitivity (of HR-USG)	Specificity (of HR-USG)	PPV	NPV
Posterior element defects in vertebral canal	46	48	95.8	100	100	50
Expansion of ventral SAS	36	43	83.7	100	100	50
Splitting of cord	12	13	92.3	100	100	97.36
syringohydromyelia	11	15	73.3	100	100	89.74
Low lying conus	32	35	91.4	100	100	83.33
Filar abnormalities	31	35	88.5	100	100	78.94
Fatty mass at defect site	165	19	84.2	100	100	91.17

Table-1: Showing various imaging parameters in all age groups

study. Amongst those with closed defects presenting without a subcutaneous mass we observed 03 cases of dorsal dermal sinus (~11.5%) and one case each of a lipoma, a Neuroenteric cyst and an internal meningocele.

Amongst the open type the most common defect was a myelomeningocele which was present in 19 out of 24 cases (~79%). Overall the most common defect was a myelomeningocele which was present in ~38% of all cases of spinal dysraphism. We found many cases with overlapping features where clinically, open defect was apparent but imaging revealed association with underlying closed defect. Following Tortori-Donati et al classification⁸ the patients were initially categorised as closed or open defect based on the clinical examination findings. Subsequently, we labelled these findings as an associated defect. This highlights the important concept of origin of these defects as simultaneous arrest of development occurring at multiple stages and involving multiple processes leading to overlapping features.¹⁵ Tortori-Donati et al classification was proposed for making management decisions easy, aiming at correction of the defect rather than identifying a strict embryological event at fault, which is more the concern of anatomists.⁹ 12 cases of Diastematomyelia (types 1 and 2) were seen in isolation or associated with other defects (open/closed) [Figure 3-a,b,c,d]. Spinal segmental dysgenesis was seen in 05 out of total 50 cases as an associated finding. A filar cyst was seen in a case of posterior meningocele. Filar thickening and tight filum terminale were encountered in many cases as an associated finding. We did not encounter any case of caudal regression syndrome during our study. High resolution USG also revealed most of the findings till 1 year of age and

certain other findings in higher age groups. The findings on comparison between ultrasonography and magnetic resonance imaging, in the 4 age groups have been tabulated in Tables 1. Objectively comparing 7 imaging parameters on HR-USG vs MRI we have tabulated following observations and enlisted the calculated sensitivity and specificity of detection of each finding on HR-USG as against gold standard imaging modality of MRI. All imaging parameters aiding final diagnosis were detected with very high (>95%) sensitivity and specificity on HR-USG as against MRI in 0-3 months age group. 5/7 parameters were detected with >88% sensitivity and 100% specificity in the age group of 3 months-1 year. Thus overall, HR-USG showed great detection rates for all findings under 3 months and for most findings under one year of age as against MRI.

DISCUSSION

Studies in northern part of India suggest a male preponderance of spinal dysraphism. Most cases present in young age and mean age of presentation lies in the first decade of life.¹⁶ We also noted a similar trend in our study, with male to female ratio of 1.2:1 and mean age of presentation 1 year 8 months. Almost 44% cases presented in the 0-3 months age group and ~68% patients presented as infants. Closed spinal defects (~52%) were commoner than open spinal defects (~48%).

Overall, in our study, when we compared final diagnosis given on each modality, ultrasound fared well with 36 cases (~72%) showing complete agreement between final diagnosis on HR-USG and MRI. Amongst the 14 cases in disagreement, 10 (~20%) showed partial agreement, with MRI detecting few additional details. Thus, ~92% cases could be diagnosed on USG alone using structured and objective approach towards identifying key abnormal pointers towards diagnosis. Only 4 cases (8%) were showing completely different results and also significant additional abnormality on MRI affecting patient management directly like exact extent of a dorsal dermal sinus or lipomatous tissue within spinal canal in older children, segmentation anomalies and resultant scoliosis, associated anomalies like-ARM / bladder exstrophy etc. For the detection of segmentation anomalies and degree of scoliosis a standard two view radiograph of spine could suffice in most cases, considering the minute radiation exposure and low costs. For exact delineation of anatomy in complex cases we can always upgrade to a higher available modality -MRI. We did not encounter any cases of CRS in our study, however all previous studies point towards poor sensitivity of USG for its detection and thus MRI should be the investigation of choice in such complex dysraphic states.^{17,18} Majority of case load of patients of spinal dysraphism is shared by MMC (19 cases) and lipomyelomeningocele (12 cases) accounting for a total of ~62% cases in our study, both of which showed comparable detection at both imaging modalities in our study.

The role of MRI in diagnosing and planning management in patients of spinal dysraphism is established beyond doubt and so this imaging modality holds the position of being the current gold standard. But there are identified practical problems associated with imaging young patients with MRI which become troublesome for patients and clinicians

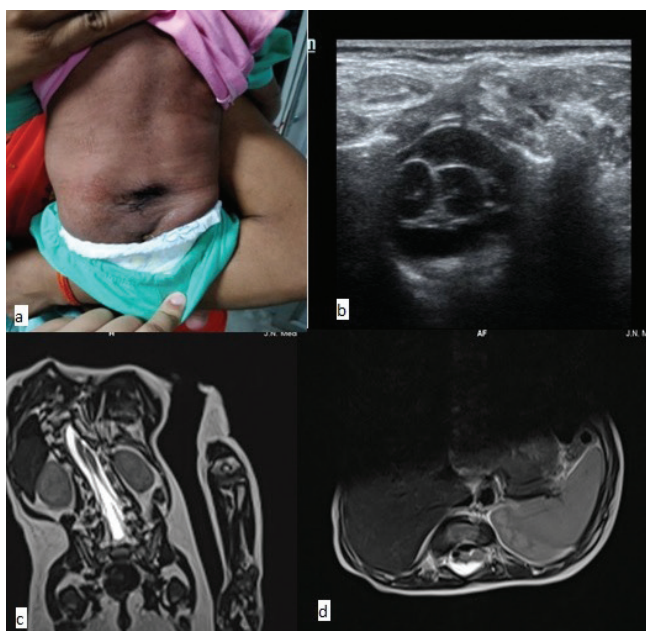


Figure-3: (a,b): High resolution ultrasound image of the patient of closed spinal tube defect with splitting of lower spinal cord into two hemi-cords within a single dural sac with no fibrous/bony bar - type 2 diastematomyelia. (c,d): Coronal and axial T2 weighted MRI spine shows splitting of the terminal part of spinal cord into two hemi-cords without any intervening septa s/o type 2 diastematomyelia

at times. These include time constraints, motion artefacts degrading image quality, requirement for anaesthesia, post sedation care and cost constraints.¹⁸ In a study by Ranjan et al¹⁸, the average time taken to scan a patient of spinal dysraphism was 26.96 ± 5.99 min by MRI and 10.62 ± 1.99 min on USG of spine and this difference was found highly significant ($P < 0.0001$). Real-time imaging is not possible with MRI. USG can show normal movement and pulsatility of the cord and nerve roots, which is absent in a tethered cord and this real time assessment cannot be done with standard static spinal MRI images.¹⁸ This is very important when assessing for tethered cords and tight filum terminale. Also, universal availability of MRI is a concern especially in the primary health care system of India. The problems faced with MRI examination paves way for higher utility of USG in these cases. There have been various studies analysing the role of spinal ultrasound in neonatal and infantile age group for varied reasons. Amongst these the most commonly studied was the role of USG spine particularly, in cases of occult spinal dysraphisms.^{19,20,21} Use of high resolution ultrasound has various advantages over MRI - Lesser requirement of general anaesthesia with less complications related to anaesthesia and post anaesthetic care, possibility of varying section planes individually to demonstrate fine longitudinal structures, less motion artefacts as compared to MRI, possibility of real time assessment of spine.^{5,18} Two main reasons due to which spinal dysraphism can be ideally evaluated using USG: Firstly, dysraphic states generally present early in neonatal or infantile age wherein the vertebral column is unossified providing good acoustic window. Secondly, the non-fusion of vertebral arches in these defects again provides an acoustic window into the spinal canal rendering intraspinal structures visible on USG even in older children.²²

CONCLUSION

Development of newer high resolution ultrasound probes providing better resolution for superficial structures makes the use of high resolution ultrasound in imaging spinal dysraphic states an accepted and established practice. There are many benefits in high resolution real time sonographic assessment of spine in screening infants for spinal dysraphism. Whenever need arises MRI is available to solve the problem. Due to the constraints of the present state of primary health care delivery in India, high resolution ultrasound of spine can prove a boon, increasing early diagnosis and referral, significantly reducing health care costs, thus, providing an opportunity for early surgical management in the limited resources available. Hence, there is a need for revamping the training in spinal ultrasound amongst radiologists to standardise its use for better patient outcomes.

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