Original Article

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Evaluation of Occult Intraspinal Anomalies in Congenital Scoliosis Deformity; Clinico-radiological Study

Raviraj A. Shinde[1], Tanvi R. Shinde[1], Ajit A. Shinde[1], Omkar R. Shinde[1]

Abstract

Introduction: *In the management of congenital scoliotic deformity, it is very important to rule out presence of intraspinal anomaly before planning surgical management. The aim of the present study is to document the incidence of intraspinal anomalies in congenital scoliosis deformity and) to identify clinical predictors for intraspinal anomalies in these patients. We analyzed 106 patients with congenital scoliosis clinically and radiologically to identify these factors associated with intraspinal anomalies. The incidence of intraspinal anomalies reported in this study is 44.34%, tethered cord was most common anomaly seen in 21 out of 47 (44.68%) patients. 11 out of 13 (84.62%) patients with neurological deficit had intraspinal anomaly suggesting it is the most significant clinical predictor for the presence of intraspinal anomaly. Others are male gender, thoracolumbar curve, defects of segmentation and mixed defects and neurocutaneous markers. Patients with these clinical factors should be investigated thoroughly for presence of intraspinal anomalies before planning surgical management.*

Keywords: *congenital scoliotic deformity, intraspinal anomaly, neurological deficit*

Introduction

The vertebral column and spinal cord are closely related anatomically and developmentally, hence it is common to see an intraspinal anomaly associated with congenital spinal deformity. It has been previously noted that in congenital spinal deformity cases, where the neural axis and the vertebral column are being formed in close relation to time and location, malformation in spinal column can be commonly associated with anomaly in the spinal cord, nerve roots or covering membranes[1].

Thus in congenital spinal deformity, intraneural anomalies such as tethering of cord, syrinx, meningomylocele, Arnold chiari malformation etc. are observed. With increasing use of MRI neural axis abnormalities are increasingly being found, which is especially helpful in patients without neurological presentation[2,3,4].

Although all of the detected anomalies may not require active intervention, pre-operative detection is important in patients who are undergoing correction as there is a risk of neurological deficits if they are not addressed prior to deformity correction[3,5,6,7]. It is therefore important to investigate neural axis abnormalities prior to corrective surgery of congenital deformities particularly scoliosis. MRI, being the most sensitive investigation at present to study the anatomy of the neural structures, will be the investigation of choice to detect these anomalies.

With the development of magnetic resonance imaging (MRI) neural axis abnormalities are increasingly being found in patients with presumed "idiopathic" scoliosis where scoliosis was the presenting sign of an otherwise asymptomatic neural axis abnormality[8]. The routine use of MRI is necessary in patients with congenital scoliosis deformities. Several clinico-radiological features have been identified in these scoliotic patients who are known to be associated with a high incidence of such anomalies.

Our aim was to document the incidence of intraspinal anomalies in congenital scoliosis deformity and study the association of intraspinal anomalies with patient symptoms, clinical signs and curve characteristics to establish which of these indicators best predict the finding of clinically relevant abnormality of the central nervous system

Materials and Methods

This is a retrospective study between January 2007 to December 2017 total of 152 children with spine deformity were screened and involved in further investigation and management. Out of these 152 children 106 were having congenital scoliosis deformity and were included in the study. Children with syndromic spine anomalies were excluded.

We documented history, clinical findings and neurological examination, for every patient, whole spine radiographs including bending views were evaluated along with whole spine MRI scans for all the patients. For each patient screening of brain using T2 saggital and axial images was done for any abnormalities in the hind brain, brainstem, spinal cord, nerve roots and the covering membranes and the dedicated MRI brain was done for abnormal cases. The presence and type of anomaly in the MRI was correlated with patient's symptoms, clinical signs and curve characteristics, which are identified as the 'indicators' for intraspinal anomalies.

[1]Asst. Professor, Dept. of Orthopaedics, Grant Medical College and Sir J. J. Hospital, Mumbai.

Address of Correspondence:
Dr. Raviraj A. Shinde,
1404, Pawanputra Housing Society, Opposite Global Hospital, Near Shirodkar highschool, Near KEM hospital, Parel, Mumbai
E-mail: drravirajshinde@gmail.com

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Table 1: Sex wise distribution of intaspinal neural anomalies.

Intraspinal anomaly	Females	Males	Total cases
Tethered Cord	15	6	21
Diastomatomyelia	7	3	10
Syringomyelia	12	6	18
Arnold Chiari syndrome	5	3	8
Meningomyelocele	2	1	3
Miscellaneous	1	1	2

Table 2: Incidence of intraspinal anomaly in different patterns of scoliosis in sex wise distribution of curve.

Region	Females	Males	Incidence Intraspinal anomaly
Cervical	5	3	0
Cervicothoracic	6	3	1
Thoracic	15	6	4
Thoracolumbar	35	12	35
Lumbar	12	3	7
Lumbosacral	3	1	0
Cervico thoracolumbosacral	2	0	0
Total	78	28	47

Results

In this study of total 106 children, 78(73.58%) were females and 28(26.42%) were males depicting clear female preponderance. The mean age of presentation was 9.82 years (3 years to 16 years). Amongst these 82(77.36%) children had Congenital scoliosis while 24(22.64)% had additional Kyphotic element. Right sided curve was predominantly seen in 52.83% (56/106) followed by Left sided curve in 39.62%(42/106) while 8/106 (7.55%) children had double curve. Thoracolumbar Curve was the most common curve seen in 47/106 (44.34%) followed by Thoracic curve 21/106 (19.81%) and lumbar curve 15/106 (14.15%). Failure of Formation was most common deformity seen in 54/106 (50.94%) followed by Mixed type of defects in 31/106(30.18%) and Failure of Segmentation was least common 21/106 (19.81%).

Intraspinal abnormalities were present in 47/106(44.34%) children and tethered cord was the most common anomaly seen in 21/47(44.68%). Syringomyelia 18/47(38.30%) and Diastomatomyelia 10/47(17.02%) were the other common anomalies. 15/47(91.91%) children had multiple intraspinal anomalies. Out of 47 children, 30 were females (38.46%) while 17 were males (60.71%) [Table 1].

Out of these 47 cases, 21 were with left sided curve, 19 were having right sided curve, 7 with double curve. Thoracolumbar curve had highest number of intraspinal abnormality 35/47 (74.49%) followed by Lumbar 7/15(46.67%) and Thoracic 4/21 (19.05%) curve [Table 2] [Figure 1].

Failure of segmentation was most frequently associated with intraspinal anomaly 17/21(80.95%), trailed by mixed deformities 20/31(64.52%) and failure of formation 25/54(46.30%)[Table 3].

Neurocutaneous markers were seen in 8 children, tuft of hair(3/8) and café au lait spots (3/8) being most common. Amongst these 6 children had intraspinal anomaly [Figure 2].

Neurological derangement was present in 13/106 children. Neurological examination revealed motor deficit in 10/106 (9.43%), sensory deficit in 5/106(4.71%), abnormal reflexes in 13/106(12.26%), bowel bladder involvement in 7/106 (6.60%). Only two of these patients were not having intraspinaly anomaly but both were cases of kyphoscolisis with Cobb angle of 900 at T10 in one case and 600 at T12 in second case [Table 4].

Discussion

As the embryonic development of vertebral column and spinal cord is closely related, it is not uncommon to have coexistent anomalies[1]. As intraspinal anomalies are not always detected by clinical examination they can be overlooked. Identifying these anomalies are of prime importance in the treatment of congenital scoliosis as they need to be managed beforehand to avoid dreadful complications.

In our study, the mean age at presentation is 9.82 years which is higher as compared to the study of Neeraj Gupta et al but similar to Erfani et al[2,9]. Female predominance (M:F - 2.78) for congenital scoliosis is similar to previous

Figure 1: 9 year old female child with left congenital scoliosis at thoracolumbar region with tethered cord at lumbar level.

Table 3: Correlation of type of deformity and intraspinal anomalies

Type of deformity	Intraspinal Anomalies	Percentage
Failure of Formation	25	46.30%
Failure of segmentation	17	80.95%
Mixed deformity	20	64.52%

Figure 2: Neurocutaneous markers

studies[2,10,11,12]. Thoracolumbar region is most commonly seen similar to other studies of Gupta et al as well as Mohanty and Kumar but it is inconsistent with that of Mac Master and Ohtsuka study which shows predominance of lower thoracic curve[9,10,13]. In the current study, Failure of formation is the most common vertebral anomaly seen in 50.94% cases, Mixed deformity was seen in 30.18% while Failure of segmentation was seen in 19.81%. These results are consistent with some reported results in other studies[3,9].

There has been previous studies which has reported incidence of intraspinal anomaly in the range of 15-47% [9,13]. In the current study, the overall incidence of intraspinal abnormality was 44.34%. In the study of Mohanty and Kumar, this incidence was 15% while in MacMaster series reported it to be 18.3% [1,13]. The lower incidence of intraspinal anomalies in the studies may be because Myelography was used as investigation method, which is considered to be less sensitive to detect intraspinal anomalies. In Gupta et al series the incidence was 47% while in Erfani et al. series it was 41.3% when MRI was used as investigation of choice[2,9]. Similar to previous studies tethered cord was the most common anomaly seen in this study [3,4,7,9,14]. In MacMaster series and Mohanty and Kumar series Diastematomyelia was the most common anomaly observed while in Erfani et al series is it was syringomylia [1,2,13]. This variable result may be due to heterogenous population included in the study.

The incidence of intraspinal anomalies was higher in left sided curve as compared to right sided curve but without any statistical significance. Although the incidence of scoliosis is more in females, in our study it was observed that males were more prone to have intraspinal anomalies($p<0.05$) which was in contrast to study by Lie et al. which showed intraspinal anomalies were more common in Females[15]. We also found that Thoracolumbar curve was most commonly associated with intraspinal anomalies. As regards to the type of vertebral anomaly, defects of segmentation and mixed defects were more commonly associated with the intraspinal anomaly as seen in previous series of MacMaster and Gupta et al [1,9]. In the present study 11 out of 13 (84.62%) cases with neurological impairment had intraspinal anomaly, also 6 out of 8(75%) cases with neuroutaneous markers had intaspinal anomaly.

Table 4: Summary of clinicoradiological factors associated with intraspinal anomaly

Clinical indicators	Incidence of intraspinal anomaly	Percentage
Male gender	17/28	60.71%
Thoracolumbar curve	35/47	74.49%
Defects of segmentation	17/21	80.95%
Mixed defects	20/31	64.52%
Neurocutaneous Markers	6/8	75%
Neurological impairment	11/13	84.62%

These observations are in line with findings of study by Rajasekaran et al. which showed 75% with cases with neurological impairment and 66% cases with Neurocutaneous markers had intraspinal anomaly.16 Also, in Gupta et al. series 83% patients with Neurocutaneous markers had intaspinal anomaly[9]. In contrast to these findings, MacMaster and Bradford series did not find any correlation between Neurocutaneous markers and intraspinal anomaly [1,7].

Main drawback of our study is being a retrospective study, the data collected to predict clinical indicators is insufficient. The other drawback is sample size is smaller and to establish clinical predictors larger sample size is recommended.

Conclusion

Patients with congenital scoliosis should be thoroughly investigated clinically and radiologically before planning surgical management. MRI is the investigation of choice to identify intraspinal anomalies. The overall incidence of intraspinal anomaly in this study is 44.34%, while in patients without any neurological manifestation is 33.96% which is considerably high. Hence preoperative MRI is recommended for the best surgical outcome in every patient. Male gender, Thoracolumbar curve, Defects of segmentation and mixed defects, presence of neurocutaneous markers and neurological deficit are associated with higher risk of intraspinal anomaly and should be given special attention.

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