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Original Article

Occult intraspinal abnormalities and congenital scoliosis

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Abstract

BACKGROUND: Congenital scoliosis occurs because of either the failure of formation or the failure of segmentation or both. Evaluation of the incidence and the types of occult intraspinal abnormalities in congenital scoliosis is the subject of this study.

METHODS: During a period of 29 years, 103 patients with congenital scoliosis were studied. MRI was used in 46 patients, myelography or CT myelography was used in 64 patients and both MRI and myelography or CT myelography were used in 7 patients for intraspinal abnormalities.

RESULTS: In the MRI group, among the 46 patients, 19 patients (41.3%) had intraspinal abnormalities consisting syringomyelia in 9 (19.5%) diastematomyelia in 8 (17.4%), tethered cord syndrome in 6 (13%), low conus in 5 (10.8%) and diplomyelia in 3 (6.5%) of the patients. In the myelography group, among the 64 patients, 17 (26.5%) had intraspinal abnormalities and diastematomyelia was the most common one found in 14 (21.8%) patients.

CONCLUSIONS: Intraspinal abnormalities are frequent in congenital scoliosis. Syringomyelia may be associated with congenital scoliosis. In congenital scoliosis, rib fusion may be an indicator of intraspinal abnormalities in MRI. A significant difference between clinical findings and intraspinal anomalies ($P<0.05$) was noted. Moreover, we believe that total spinal MRI with coronal, sagittal and axial views is a valuable tool in determining the intraspinal abnormalities in congenital scoliosis. This method is highly recommended for detection and neurosurgical intervention before corrective surgeries.

KEY WORDS: Congenital scoliosis, intraspinal abnormalities, diastematomyelia.

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Congenital scoliosis occurs because of either the failure of formation or the failure of segmentation or both. Congenital scoliosis most frequently occurs in the first 8 weeks of the prenatal development. During this time, the bony elements of the spine are forming and the neuraxis is completing its infolding and it closes the neural tube. These events are closely related and any intrauterine event that causes congenital scoliosis could be associated with an occult intraspinal anomaly. The subject of this study was to measure the incidence and the types of occult intraspinal abnormalities in congenital scoliosis. Because of the relationship between neural and vertebral development, one could expect to find a high incidence of intraspinal abnormalities in association with congenital scoliosis. These intraspinal abnormalities may not be recognized because the skin overlying the spine usually appears normal and the associated neural abnormalities are mild or absent [1].

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Methods

This was a retrospective study of patients with congenital scoliosis to find incidence of intraspinal abnormalities from 1975 to 2004. All the patients were evaluated by clinical examination, plain X-ray, routine myelography or MRI studies, especially in preoperative period. During the last 5 years, MRI has been performed on all patients while previously myelography or CT myelography had been the procedure of choice. Both procedures have been performed on seven patients (figure 1). All patients with meningocele, meningomyelocele and neurofibroma, and all the patients with no myelography or MRI study were excluded. Finally, 103 cases remained in the study. X^2 test was used for statistical analysis. A significant level of 0.05 was chosen for all the statistical tests.

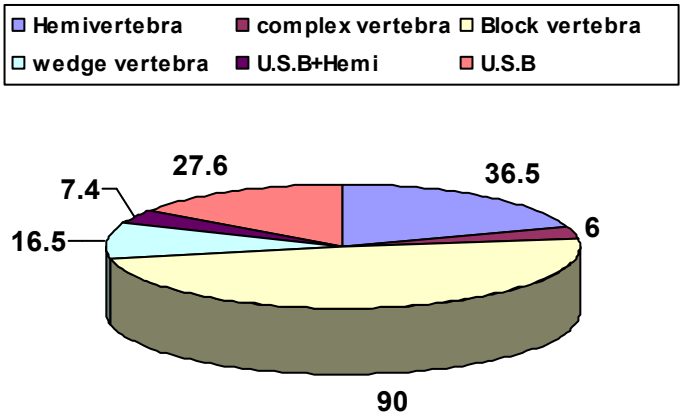

Figure 1. Distribution of Vertebral anomaly in 103 patients with congenital scoliosis. (USB: Unsegmented bar)

Results

This research consisted of 103 cases of congenital scoliosis, 73 females (71%) and 30 males (29%), ranging from younger than one to 22 years old (mean: 9.5 years). Positive physical findings including neurological deficits, skin manifestations and foot deformities were noted in 23.3% of the patients. Plain X-ray revealed the following congenital vertebral anomalies: hemivertebra (36.5%), unilateral unsegmented bar and contralateral hemivertebra (7.4%), block vertebra (6%) and complex group (6%) (figure 1). The magnitude of curves varied from a minimum of 20° to a maximum of 112° with a mean of 55°. The left side curve was observed in 51% of the patients while the right side curve was observed in 49% of them. In addition to scoliotic deformity, kyphosis was noted in 26% of the cases. Fused ribs were noted in 21.4% of the patients. By the MRI study, we found a significant association between rib fusion in congenital scoliosis and intraspinal abnormalities ($P<0.04$). In the46 patients who were studied by MRI, the intraspinal abnormalities were noted in 19 patients (41.3%). Most of them had more than one intraspinal abnormality. These abnormalities consisted of a tethered cord syndrome in six patients (13%), syringomyelia in 9 (19.5%), diastematomyelia in 8 (17.4%), diplomyelia in 3 (6.5%), and low conus in 5 (10.8%) of the patients (figure 2). Sixty-four patients underwent myelography and five of them had additional CT myelography. Occult intraspinal abnormalities were detected in 17 patients (26.5%). These abnormalities consisted of a diastematomyelia in 14 patients (21.8%), tethered cord syndrome in 2 (3.1%), low conus in 1 (1.5%) and diplomyelia in 2 (3.1%) patients. No Syringomyelia was detected (figure 3). A neuro-

surgical intervention for intraspinal abnormalities was performed in 80% of the patients. Other associated anomalies in organs other than the spine were present in 26.2% of the patients. Genitourinary anomalies were the most common anomaly (table 1).

Table 1. Distribution of other associated congenital anomalies with congenital scoliosis.

Genitourinary anomaly	4.9%	Sacral agenesis	1.9%
Congenital heart disease	4.9%	Hip dysplasia	2.9%
Klippel-Feil syndrome	3.9%	Chest and rib deformity	2.9%
Torticollis	3.9%	Upper extremity anomaly	1.9%
Sprengel's deformity	3.9%		

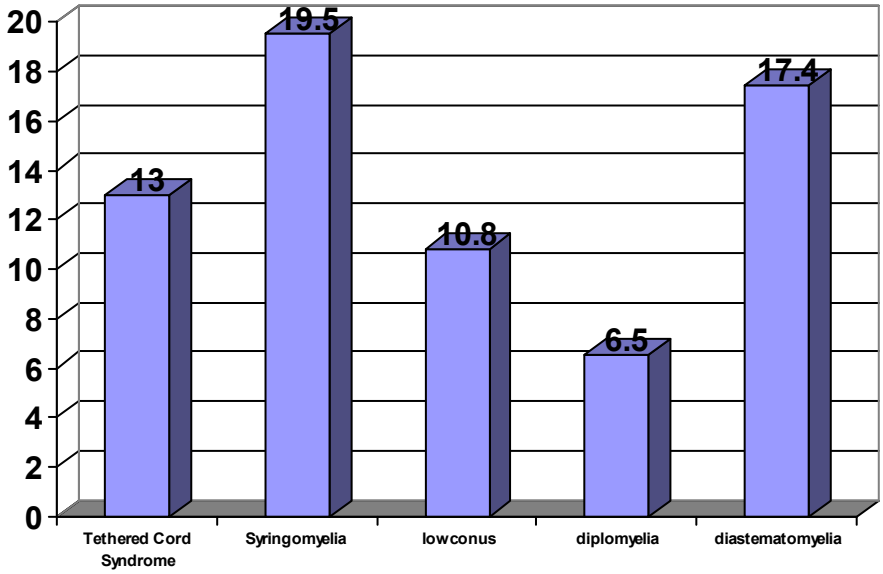

Figure 2. Findings of MRI study among 46 patients with congenital scoliosis.

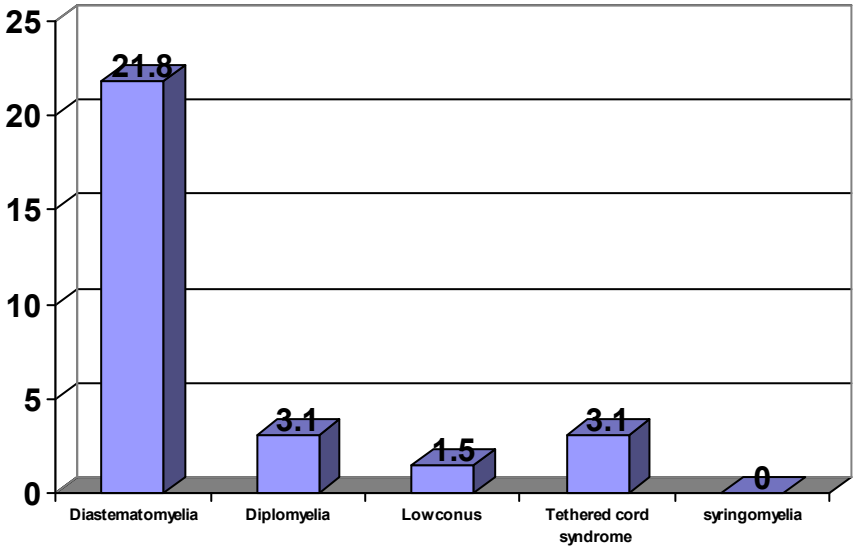

Figure 3. Findings of myelography and CT myelography among 64 patients with congenital scoliosis.

Discussion

Congenital scoliosis is a developmental spinal deformity originated from the embryonic malformation of vertebrae [2]. The age of presentation peaked first at the age of 2 and then again at the age of 8 to 13 which was similar to the literature [3]. Female preponderance (female to male ratio of 2.43) was similar to all the previous reports [3-5]. We also did not find any association between sex identity and intraspinal abnormalities. Intraspinal abnormalities in congenital scoliosis are more frequent than what has been suggested previously. Intraspinal abnormalities were diagnosed by MRI in 19 out of 46 patients (41.3%) with congenital scoliosis, and they were diagnosed in 17 out of 64 patients (26.5%) by myelography or CT myelography. Occult intraspinal abnormalities were also accompanied by skin abnormalities and neurological findings. However, these stigmata did not always indicate structural intraspinal abnormalities. The association of clinical and physical findings (skin manifestations, neurologic deficits, foot problems) and intraspinal abnormalities were confirmed (P = 0.01 and P = 0.03 by myelography and MRI, respectively). Prahinski-JR has reported a poor correlation between the findings on the physical examinations, plain radiograms and the subsequent occult intraspinal abnormalities on MRI [6]. In series of McMaster [1] and Bradford [7] the association of hemivertebra with contralateral bar and intraspinal abnormalities was confirmed. Intraspinal abnormalities had no association with the types of congenital scoliosis in our series. The same result was reported by Shahcheraghi et al [8]. We found a relationship between the fusion of the ribs and positive MRI findings for occult intraspinal abnormalities (P<0.04). Eight out of 19 patients with positive MRI had rib fusion (42%) in comparison with 4 out of 27 patients (14%) with negative MRI findings. Rib fusion was suggestive for higher curve progression rate especially in the lower thoracic area [8]. Overall, occult intraspinal abnormalities were detected in 41.3% of our MRI cases. The result is comparable with that of Bradford (38%) and Prahinski-JR series (30%) [6,7]. Surprisingly, the presence of unsuspected syringomyelia was observed in 9 (19.5%) patients with congenital scoliosis in MRI group. This was not described in the series of Blake et al [9] and McMaster [1] but, in the series of Bradford it is mentioned 4 out of 42 (9.5%) and in Prahinski-JR series 4 out of 30 patients (13.3%). Nearly similar results about other intraspinal abnormalities were observed in our study. In myelography and CT myelography group of 64 cases, 17 patients (26.5%) were positive for occult intraspinal abnormalities. In this group, diastematomyelia was the most common anomaly which included 14 patients (21.7%). This finding was similar to that of McMaster series (16.3%). Other findings were similar to the literature [1,5,10].

Conclusion

Intraspinal abnormalities are frequent in congenital scoliosis. Syringomyelia may be associated with congenital scoliosis. In congenital scoliosis rib fusion may be an indicator of high prediction of intraspinal abnormalities in MRI. A significant difference between clinical findings and intraspinal anomalies was noted. Moreover, we believe that total spinal MRI with coronal, sagittal and axial views is a valuable tool in determining the intraspinal abnormalities in congenital scoliosis; and it is highly recommended for detection and neurosurgical interventions before corrective surgeries.

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