ECRONICON OPEN ACCESS

EC ORTHOPAEDICS
Research Article

Analysis of Natural History of Curve Progression in Congenital Scoliosis

Shyamasunder N Bhat*, Praveen Cheedella, S P Mohanty and Chandreyee Duttagupta

Department of Orthopaedics, Kasturba Medical College, Manipal University, Manipal, India

***Corresponding Author:** Shyamasunder N Bhat, Department of Orthopaedics, Kasturba Medical College, Manipal University, Manipal, India.

Received: March 27, 2017; **Published:** May 04, 2017

Abstract

Background: Vertebral anomalies causing an imbalance in the longitudinal growth of the spine are called as Congenital scoliosis. It can be associated with systemic abnormalities which have implications on treatment. With its relentless progression, congenital scoliosis presents a unique problem whose management needs special attention. We sought to trace the natural history of congenital scoliosis in our patients by observing the patterns of presentation, deformity, curve progression and incidence of associated anomalies.

Materials and Methods: Sixty patients with a median age of 7 years, presenting with congenital scoliosis between January 2006 and September 2012 and with a minimum of one year follow up on record, were included in our cohort study. Comparisons were made using initial radiographs with erect radiographs taken at the end of a minimum of one year prospective follow up.

Results: The evaluation of progression was done using Curve Progression Index (CPI). Variation pattern of CPI with vertebral anomalies and curve patterns were statistically insignificant (p = 0.352). Sixteen patients (26.7%) had general anomalies.

Conclusion: Decisions in managing congenital scoliosis are based on the natural history of progression of the curve, clinical and radiological assessments are necessary. Detection of diastematomyelia and spinal cord anomalies becomes relevant in choosing appropriate management. Knowledge of the natural history of congenital scoliosis helps to anticipate the progression of the curve and prevent deterioration.

Keywords: *Congenital Scoliosis; Natural History; Curve Progression*

Introduction

Failure of formation or segmentation of any part of vertebral column can result in the development of scoliosis [1-3]. Scoliosis is a triplanar deformity with lateral, anteroposterior and rotational components. The main types according to Scoliosis Research Society (SRS) are Idiopathic (80%), Neuromuscular (6%), Congenital (12%) and Secondary to neoplasm (2%) [4,5].

Congenital scoliosis occurs due to anomalies of vertebrae causing an imbalance in the longitudinal growth of vertebral column [5]. It is often recognized at birth, but some can remain undetected. It is more rigid and is frequently associated with kyphosis and can be associated with intraspinal, cardiovascular, genitourinary and other abnormalities [6-8]. When the abnormally formed vertebrae are identified, the curve should be classified as congenital, even if the deformity appears during adolescence [9].

Several theories like osseous metaplasia, failure to ossify of annulus fibrosus or persistent notochord have been considered for these vertebral anomalies [10]. During somitogenesis, TBX6 gene plays an important role [11].

Natural History

The natural history of the congenital scoliosis needs to be understood because deformities with steady progression are to be treated appropriately. Congenital scoliosis is usually very rigid and resistant to correction. Usually, 25% congenital scoliosis don't progress, 50% progress slowly and 25% progress rapidly. Early spinal fusion is essential to prevent the development of severe curves in patients with a tendency for progression. The degree of scoliosis depends on the type of anomaly and the overall growth potential of the individual. In the presence of a vertebral anomaly, there is an imbalance resulting in localized unilateral longitudinal growth leading to increased spinal deformity as a child grows. The failure of formation and segmentation usually has significant sequele in spinal growth during childhood. Winter [12] and later McMaster [13] reported that the rate of deterioration and the severity of scoliosis could be predicted by curve location and the type of anomaly.

The deformity progresses rapidly during the first two to five years of life and later during the adolescence which is on average between 12 - 15 years in boys and 10 - 13 years in girls [14,15]. This is the time that the condition is often diagnosed. As is in the idiopathic variety, congenital scoliosis with rapid progression is common in girls [16]. An article by Pahys JM and Guille JT reviews the natural history of congential scoliosis with latest classification and treatment options. Magnetically controlled growing rods are being used nowadays in several studies with small groups of congenital scoliosis [17].

Co-existent Anomalies

These patients can have co-existent fused or absent of ribs and can also have involvement of non-skeletal organs. Congenital scoliosis patients have congenital heart defects in 10 - 15% of cases. About 20 - 40% of them have renal anomalies. Restricted pulmonary function in patients with severe scoliosis is of concern because of hypoplastic lung development [10].

Diagnosis

The Cobb's method of measurement by the Scoliosis Research Society has been adopted [18].

By early detection, before any gross deformity becomes evident, prophylactic surgery may prevent a major problem later. Knowledge of the natural history of the deformity helps anticipate the progression of the curve and prevent deterioration. A simple operation to balance the growth is preferred to a complex multi- stage surgery required later for correction.

This study was undertaken with the aim of analysing the natural history of congenital scoliosis by observing the presentation patterns, the rate of curve progression and incidence of associated anomalies.

Materials and Methods

Study Design: Cohort study.

Study Approach: Ambidirectional, with both prospective and retrospective phases.

Sample: In total, 320 patients with scoliosis presented to our department of Orthopaedics between January 2006 and September 2012, out of which 60 were congenital scoliosis with one year follow up on record.

Inclusion Criteria: Congenital scoliosis patients who were followed up for a minimum period of one year after presentation.

Exclusion Criteria: Patients with other types like idiopathic scoliosis, neuromuscular scoliosis and secondary scoliosis.

Procedure: Institutional ethical committee approval was sought before the study commencement. Patients meeting the inclusion criteria were evaluated radiologically and clinically after obtaining consent and their previous medical records were reviewed. Age at time of presentation, gender and type of curve were noted. Radiological assessment was done by anteroposterior and lateral views from C5 to L5. Skeletal maturity was assessed by Risser sign. Neurological status at presentation and associated intraspinal anomalies were looked for in Magnetic Resonance Imaging. Echocardiography and Ultrasound abdomen were done to look for cardiac and renal anomalies respectively. Congenital vertebral anomalies present at levels in spine other than those involved in the scoliotic curve were ignored.

Follow Up: Radiographs were obtained at six months intervals and compared. The evaluation of progression was done using Curve Progression Index (CPI).

Curve progression index (CPI) = $\frac{(x-y)}{z}$

Where, (x = final Cobb's angle); (y = initial Cobb's angle); (z = number of years)

Statistical Analysis

The variation of curve progression index with vertebral anomalies and type of curve was analysed using Kruskal-Wallis test. Cobb's angle at the 1st visit and last visit were measured by two observers and Intra-class correlation coefficient (ICC) was used to assess the agreement between them. ICC was found to be 0.99 for Cobb's angle measured (p-values < 0.001), indicating a good agreement between the measurements of two observers.

The analysis was performed using software SPSS version 15.0.

Results

Sixty cases of congenital scoliosis were observed for curve progression before definitive treatment for the period January 2006 to September 2012. Girls (38) with congenital scoliosis were more than boys (22) with a female: male ratio of 3:2.

The median age at presentation in the study group was seven years with a range of 1 year to 15 years. Of these, 43.33% were below five years of age whereas 28.33% were between 6 and 10 years, 25% were in 11 - 15 years age group and 3.33% were in 16 - 20 years age group.

Type of curves: Majority were thoracolumbar type (TL) (n = 27) followed by thoracic (T) (n = 18). Remaining were cervicothoracic (CT) (n = 11) and lumbar (L) (n = 4). Right sided curves (34) were more than left sided curves (26).

Types of anomaly: Based on Winter and Mac Ewen [4] classification of congenital scoliosis, curves were analysed. Hemi-vertebra was most commonly observed, followed by wedge, unilateral unsegmented bar and block vertebrae.

Median curve progression: In single hemivertebra, the maximum rate of curve progression index was found in the thoracic region (median CPI 8), followed by lumbar and cervicothoracic curves (median CPI 2.5). In multiple hemivertebrae curve progression index was almost same as that of single hemivertebra with maximum in the thoracic region (median CPI 6) and minimum in thoracolumbar region (median CPI 2).

In wedge vertebra, curve progression index (CPI) was found to be more than double in lumbar curves (median CPI 4) as compared to thoracic and thoracolumbar curves (median CPI 2). Curves having unilateral unsegmented bar with contralateral hemivertebrae had a very high curve progression index, especially in thoracolumbar curve pattern (median CPI 15). Block vertebrae had minimum curve progression index (median CPI 1.5) and there was no major variation of CPI among different types of curves.

The CPI for different types of vertebral anomalies is shown in Figure 1 and Table 1.

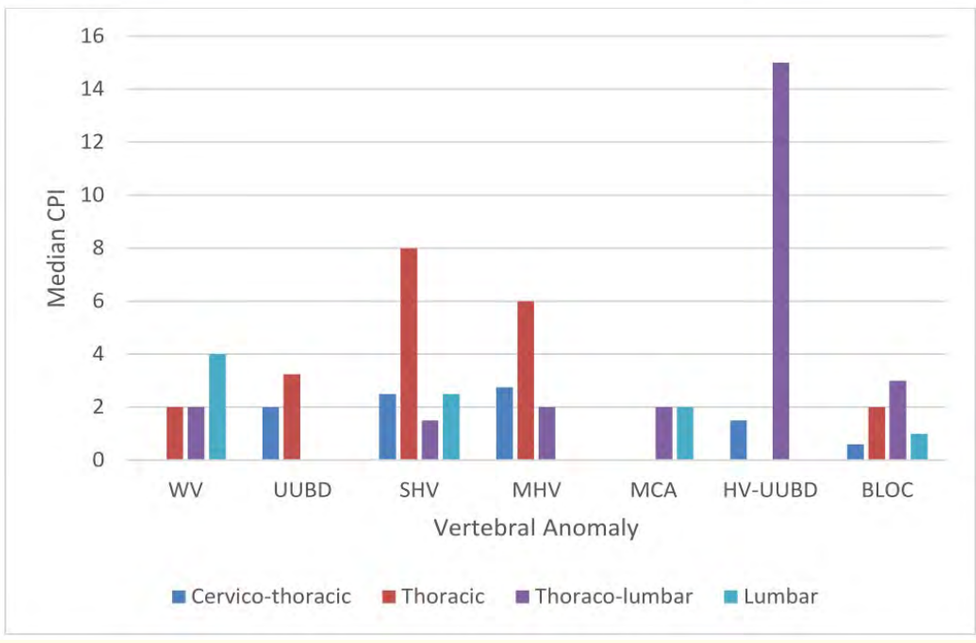

Figure 1: *Curve progression index with type of vertebral anomaly and type of curve.*

WV: Wedge vertebra; UUBD: Unilateral unsegmented bar; SHV: Single hemivertebra; MHV: Multiple hemivertebrae; MCA: Multiple congenital anomalies; HV-UUBD: Unilateral unsegmented bar with hemivertebra and BLOC: Block vertebra

Type of Curve	Vertebral Anomaly with Median CPI						
	Wedge Vertebrae	**Unilateral unsegmented bar with contralateral hemivertebrae**	**Single hemivertebra**	**Multiple hemivertebrae**	**Mixed congenital anomalies**	**Hemivertebrae with unilateral unsegmented bar**	**Block Vertebrae**
Cervico-thoracic	-	2	2.5	2.75	-	1.5	0.6
Thoracic	2	3.25	8	6	-	-	2
Thoraco-lumbar	2	-	1.5	2	2	15	3
Lumbar	4	-	2.5	-	2	-	1

Table 1: *Vertebral anomaly with median CPI and type of curve.*

The median CPI was found to be more among girls (4.0) as compared to boys (3.6). The variation pattern of CPI with vertebral anomalies was found to be statistically not significant by using Kruskal-Wallis test (p = 0.352). The median difference in curve progression was found to be insignificant using the Kruskal-Wallis test (p = 0.35).

Assessments done for general anomalies showed 44 (73.3%) congenital scoliosis patients had no other anomalies while the remaining 16 (26.7%) had various anomalies (Table 2).

	Frequency	Percent
None	44	73.3
Congenital taliped equino varus	3	5.0
Congenital vertical talus	2	3.3
Torticolis	2	3.3
Flexion contracture hip	1	1.7
Local gigantism	1	1.7
Anotia	1	1.7
Developmental dysplasia hip	1	1.7
Pneumoencephalocele	1	1.7
Poly neuropathy	1	1.7
Radial club hand	1	1.7
Syndactyly	1	1.7
Tibial hemimelia	1	1.7
Total	60	100.0

Table 2: *Associated general anomalies.*

Discussion

Among different types of scoliosis, congenital scoliosis with rigid refractory deformity and continuous progression presents a unique problem and needs special attention. Congenital scoliosis has its origin from embryonic malformation and is not very common [19]. The modification of Kuhn and Hormel's classification by Winter., *et al*. [12] is used for treatment and prognostication at present. Being a developmental abnormality, it can accompany other systemic abnormalities which may have a bearing on treatment.

We evaluated our cases of congenital scoliosis for the presentations, patterns of deformity, and curve progression. It is necessary to understand the normal growth of spine with the patho-anatomy of different types of congenital scoliosis. Normally longitudinal growth of the spine is the total of growth occurring at end plates of the vertebra which occurs equally on either side so that spine grows straight. A congenital vertebral anomaly can cause growth imbalance due to the rate of growth on one side of the spine or deficiency in either the number of end plates. The lateral curve that results is of severity proportion to the degree of growth imbalance [13]. As the probability of progression of scoliosis depends on age at presentation, sex, location or pattern of curves and underlying vertebral anomalies it becomes relevant to evaluate patients clinically and radiographically [6,13]. Our study observed that congenital scoliosis was more prevalent in girls than boys (3:2), which is similar to observations made in the literature [13].

In our study, congenital scoliosis was commonly observed in the thoracolumbar area. These findings are different from an almost similar study done by McMaster and Ohtsuka [13] who found the commonest site to be lower thoracic region (33%) followed by upper thoracic (31%) and thoracolumbar (20%). We also found that right sided curves were more than left sided curves.

Detection of underlying vertebral anomalies and their classification has prognostic significance in terms of the progression of the

deformity. In this study, the commonest type of vertebral anomaly causing scoliosis was multiple hemivertebrae (31%) followed by single hemivertebra (25%). Unilateral unsegmented bar with contralateral hemivertebrae (5.5%) was least common anomaly observed. Comparative literature shows unilateral unsegmented bar (38%) as the commonest and wedge vertebrae as the least common vertebral anomaly (3%) [13]. Johal., *et al*. have done a comprehensive review of embryology, imaging, classification and management of hemivertebrae [20].

We found that the rate of progression and the severity of congenital scoliosis also depends on the site of occurrence. The thoracic region had the maximum curve progression rate with median CPI of 4.5, similar to the study done by Winter., *et al*. [12]. Our study agrees with McMaster and Winter in that the thoracic and thoracolumbar curves usually have the worse prognosis than lumbar curves.

According to literature, the most benign curves occur in the upper thoracic region, whereas according to our study, the most benign ones were in cervicothoracic region with median CPI of 2. The type of vertebral anomaly associated with a higher rate of progression of the curve in our study was unilateral unsegmented bar with contralateral hemivertebra with median CPI of 8, but this cannot be generalized as we did not have a sufficient number of patients with this anomaly in our study. The difference in median CPI with the vertebral anomalies was found to be statistically insignificant ($p = 0.352$).

It is important to recognize unilateral unsegmented bar with contralateral hemivertebrae as described by Nasca., *et al*. [21] as it has worst prognosis of any congenital anomaly. Two patients had scoliosis due to unilateral unsegmented bar with contralateral hemivertebrae with age of presentation at 6 and 9 years respectively. These curves deteriorated at the rate of 15 degrees and 6.5 degrees per year. The poor prognosis associated with this anomaly is so predictable that these curves should be treated immediately without any period of observation. A hemivertebra produces scoliosis by enlarging wedge on the affected side whereas, in cases with unilateral unsegmented bar, there is slower growth on the affected side [13]. Our study had 15 patients with single hemivertebra causing congenital scoliosis.

Neurological involvement at presentation indicates the presence of an occult intraspinal anomaly [22,23]. It is important to exclude underlying spinal cord anomalies too. We had ten patients with underlying intraspinal defects detected by MRI Scan or CT Myelogram. Spinal dysraphism in the form of diastematomyelia was the most common intraspinal anomaly. Winter et al. found diastematomyelia in 4.9% of 392 patients having congenital scoliosis [21]. In an earlier study, McMaster [24] found intraspinal defects in 18.3% of 251 patients with congenital scoliosis. Diastematomyelia was the commonest anomaly (16%). Among all reported series, Blake., *et al*. [16] had the highest incidence of occult intraspinal anomalies (58%).

Congenital Scoliosis is also commonly associated with the anomalies in mesenchymal tissues. They remain asymptomatic and remain undetected [13,25]. Majority of the patients screened for congenital renal abnormalities were found to be normal. Genitourinary anomalies were less in frequency in contrast to the view of McMaster and Winter who stated that these are the commonest. Cardiovascular anomalies were seen in 4 patients. Roth., *et al*. [26] showed that incidence of scoliosis in patients with congenital heart defects was 12%. Associated general anomalies were found in 16 patients (25%). Nasca., *et al*. [21] observed these anomalies in 13.33% of their patients.

Conclusion

The decision making in congenital scoliosis is based on the natural history of progression of the curve. Thorough clinical and radiological evaluation is important. Detection of the vertebral and spinal anomalies becomes relevant in management including surgery.

Compliance with Ethical Standards

Ethical Approval: The procedures performed in this study were in accordance with the ethical standards of the institutional ethics committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent: Informed consent was obtained from all participants included in the study. The authors certify that they have obtained all appropriate patient consents. The patients have given their consent for their clinical information and images to be reported in the publication. They also understand that all efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

Bibliography

1. Hungerford DS. "Spinal deformity in adolescence: Early detection and non-operative treatment". *Medical Clinics of North America* 59 (1975): 1517-1525.
2. Mehta MH. "Pain provoked scoliosis". *Clinical Orthopaedics and Related Research* 135 (1978): 58-65.
3. Erol B., *et al*. "Etiology of congenital scoliosis." *The University of Pennsylvania orthopaedic Journal* 15(2002): 37-42.
4. Mac Ewen GD. "Classification and Terminology of Scoliosis". *Clinical Orthopaedics and Related Research* 93 (1973): 10-22.
5. Mohanty SP and Kumar N. "Patterns of presentation of Congenital Scoliosis- An analysis". *Journal of Orthopaedic Surgery* 8.2 (2000): 33-37.
6. McMaster MJ. "Congenital Scoliosis the Paediatric Spine- Principles and Practice". Raven Press Ltd., New York (1994): 227-244.
7. Mac Ewen GD., *et al*. "Congenital Scoliosis with Unilateral Bar". *Radiology* 90.4 (1968): 711-715.
8. Winter RB. "Congenital Scoliosis". *Clinical Orthopaedics and Related Research* 93 (1973): 75-94.
9. Shands AR and Eisberg HB. "The incidence of scoliosis in the state of Delaware. A study of 50,000 mini films of the chest made during a survey for tuberculosis". *Journal of Bone and Joint Surgery-American* 37.6 (1955): 1243-1249.
10. Blake NS., *et al*. "Spinal cord abnormalities in congenital scoliosis". *Annales de Radiologie* 29.3-4 (1986): 237-241.
11. Chen W., *et al*. "Progress and perspective of TBX6 gene in congenital vertebral malformations". *Oncotarget* 7.35 (2016): 57430-57441.
12. Winter RB., *et al*. "Congenital scoliosis. A study of 234 patients treated and untreated". *Journal of Bone and Joint Surgery-American* 59 (1968): 15-47.
13. McMaster MJ and Ohtsuka K. "The natural history of congenital scoliosis. A study of two hundred and fifty-one patients". *Journal of Bone and Joint Surgery-American* 64.8 (1982): 1128-1147.
14. Dimeglio A. "Growth in pediatric orthopaedics". *Journal of Pediatric Orthopaedics* 21.4 (2001): 549-555.
15. Bunch WH and Patwardhan AC. "Scoliosis, Making Clinical Decisions". CV Mosby Co, St. Louis 735 (1989): 50-67.
16. McMaster MJ and David CV. "Hemivertebra as a cause of Scoliosis". *Journal of Bone and Joint Surgery-British* 68.4 (1986): 588-590.
17. Pahys JM and Guille JT. "What's new in congenital scoliosis?" *Journal of Pediatric Orthopaedics* (2016).
18. Lonstein JE. "Idiopathic Scoliosis". In: Lonstein JE, Winter RB, Bradford DS, Ogilvie J (Eds) Moe' Textbook of Scoliosis and other Spinal Deformities, 3rd Ed. WB Saunders Co, Philadelphia (1995): 219-256.
19. Ulrich HF. "Experiences with congenital scoliosis". *Clinical Orthopaedics and Related Research* 7 (1956): 163-170.
20. Johal J., *et al*. "Hemivertebrae: a comprehensive review of embryology, imaging, classification, and management". *Child's Nervous System* 32.11 (2016): 2105-2109.

21. Nasca RJ., *et al*. "Progression of Congenital Scoliosis due to Hemivertebra and Hemivertebra with bars". *Journal of Bone and Joint Surgery-American* 57.4 (1975): 456-466.

22. Gillespie R., *et al*. "Intraspinal anomalies in congenital scoliosis". *Clinical Orthopaedics and Related Research* 93 (1973): 103-109.

23. Winter RB., *et al*. "Diastematomyelia and congenital spine deformities". *Journal of Bone and Joint Surgery-American* 56.1 (1974): 27-39.

24. McMaster MJ. "Occult intraspinal anomalies and congenital scoliosis". *Journal of Bone and Joint Surgery-American* 66.4 (1984): 588-601.

25. Carman DL., *et al*. "Measurement of scoliosis and kyphosis radiographs. Intraobserver and interobserver variation". *Journal of Bone and Joint Surgery-American* 72.3 (1990): 328-333.

26. Roth A., *et al*. "Scoliosis and congenital heart disease". *Clinical Orthopaedics and Related Research* 93 (1973): 95-102.

Volume 6 Issue 3 May 2017