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Long-term Outcome of Early Fusions for Congenital Scoliosis*

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Abstract

Study Design: The current literature on outcomes of early fusions for early-onset scoliosis (EOS) has a short follow-up of 7.7 to 12.5 years, with many patients not at the end of growth. The forced vital capacity (FVC) at follow-up ranged from 40.8% to 64% of predicted. A study was undertaken to evaluate the long-term outcome of these fusions.
Methods: The study is of congenital patients who had a fusion under age 8 years, with the current age being >20 years. A follow-up consisting of radiographs, outcome questionnaires, and pulmonary function tests was performed. Of the 42 patients identified, 20 were traced and 11 agreed to participate, and completed all the data. The average follow-up was 37 years. All the fusions included the thoracic spine, with an average of 10.2 levels fused, of which an average of 8.8 levels were thoracic.
Results: The average scoliosis at presentation was 52°, 56° at surgery and 47° at follow-up. At follow-up, the average FVC was 53% of predicted, with an average PaO_2 of 86 and $PaCO_2$ of 44 mm Hg. The mean T1–T12 length was 20 cm. There was no correlation of the FVC percentage predicted and the proximal extent of the fusion, the T1–T12 length or the number of thoracic levels fused. The average ODI was 23. On the SF36, the average Physical Component *t* score was 50; with the average Mental Component *t* score was 52.
Conclusion: At a long-term average follow-up of 37 years, the patients had a low FVC of 53%, with one on permanent oxygen and 5 with dyspnea. The patients were functioning well at follow-up, but it is unknown what their function would be without surgery or with longer follow-up.

Keywords: Congenital scoliosis; Early fusion; Long-term follow-up

Introduction

Early-onset scoliosis (EOS) has gained prominence with the use of techniques to preserve growth while controlling the scoliosis. There is concern of the pulmonary effects of scoliosis, with recognition of the associated thoracic insufficiency syndrome (TIS) as described by Campbell [1]. Current techniques to control the deformity include growth rods for scoliosis, and VEPTR for scoliosis and TIS [2-8]. The effects of these treatment methods for EOS can only be evaluated with a comparison with untreated cases or alternate treatments like a spinal fusion.

Early fusion of scoliosis has been shown to effect pulmonary function in short-term follow-up studies. Goldberg et al. [9] reported on 21 patients with infantile idiopathic scoliosis who had an anterior and posterior fusion and were followed to over age 20 years. Eleven of the 21 were operated under age 10, with a mean age at surgery of 4.1 years (1.4–7.8) and a mean age at follow-up of 16.6 years (12.6–23.9). They found that the mean forced vital capacity (FVC) was 41% of predicted normal (12% to 67%).

Emans et al. [10] reported on a series of 13 patients with congenital scoliosis patients fused at an average age of 2.5 years (1–4.5) with an average of 6.9 levels fused. Earlier and more extensive thoracic fusion is associated with diminished pulmonary function: At an average of 10.8 years' follow-up, the FVC averaged 62% (32% to 94%), with a younger age at fusion being correlated with decreased pulmonary functions and decreased body height. In addition, a larger number of thoracic levels fused was associated with decreased pulmonary function.

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Karol et al. [11] reported on the pulmonary functions following early thoracic anterior/posterior fusions in non-neuromuscular scoliosis. They reported on 28 of the 54 eligible patients, of whom 20 had congenital scoliosis. The average age at surgery was 3.3 years, with an average of 59% of the thoracic spine fused. At an average age at follow-up of 14.6 years, the average FVC was 58% of predicted normal, with two patients requiring respiratory support. The thoracic height was <18 cm in 16/28 cases, with an inverse relationship between the FVC and the extent of thoracic fusion—the shorter the thoracic spine, the smaller the FVC. In addition, there was a correlation between the proximal level of the fusion and the decreased FVC.

Vitale et al. [12] reported the pulmonary functions on 21 of a series of 62 patients who underwent fusions for congenital scoliosis. Of these, 12 had a thoracic fusion averaging 7.2 levels at mean age of 5.3 years at surgery and 13 years at follow-up. The FVC at follow-up was 64% of predicted normal.

All the studies in the literature have a short follow-up, and the question is, What is the pulmonary function and patient function abilities with a longer follow-up. This study was undertaken to evaluate the long-term pulmonary function and outcomes of early spinal fusions for congenital scoliosis.

Materials and Methods

After IRB approval, the scoliosis database at the center was queried for early fusion with inclusion criteria of 1) congenital scoliosis, 2) fusion under age 8 years, 3) five or more levels of the thoracic spine fused, and 4) current age >20 years. The eligible patients were traced, invited to participate, and then sent a follow-up outcome questionnaire (SF-36), Modified Oswestry low back disability questionnaire (version 2.1A), and a questionnaire on work status and respiratory symptoms (appendix 1), as well as instructed to obtain standing radiographs as well as pulmonary functions including blood gases. The medical records were reviewed retrospectively, with the radiographs being scanned into the PACS system with a 2.5-cm reference disc on the radiograph to allow measurements to be performed with the PACS software. Measurements were made at the following time points—preoperative, postoperative, 1 and 5 years postoperatively, at the end of growth and at follow-up. These included curve measurements as well as T1–T12 height, T1–S1 height, fusion length and the height of the right and left lung space (apex of lung/first rib to dome of diaphragm).

Forty-two patients met the inclusion criteria, and 20 were successfully traced. Of these, 18 agreed to participate in the study, and 11 completed all the required follow-up (radiograph, pulmonary function tests, and questionnaire). There were 7 males and 4 females.

To ascertain if any of the patients not traced were deceased, the Minnesota death records were searched at the

Table 1
Patient information on anomalies, procedure, and curves.

	Age pres.	Age surg.	Anomaly			Other	Procedure		Fusion extent	No. T fused	Curves		
			HV	Bar	FR		P	AP			Initial	2-year	Follow-up
1	0	4.6	y		y		x		T1–L4	12	55°		70°
2	3.5	3.8	y	y	y	KF, S		x	C7–T10	10	71°	50°	74°
3	0	7.1		y	y	D		x	T4–L4	9	77°	42°	8°
4	0.4	1.9		y	y	S, D	x		T3–T8	8	65°		64°
5	4.2	7.6	y					x	T2–T8	8	37°		36°
6	2.2	5.9	y			KF	x		T7–T12	6	40°	40°	10°
7	0.7	1.2	y	y	y	KF	x		T1–T9	8	39°	33°	36°
8		6.4	y	y	y		x		T5–L2	8		42°	54°
9	1.2	5.1	y		y		x		T1–T9	9	50°	56°	61°
10	1	3	y	y	y		x		C4–T7	7	40°	48°	53°
11	1.8	7.7	y	y				x	T1–L3	12	46°	32°	48°
Average	1.7	4.9								8.8	52°	43°	47°

Age pres., age at presentation; Age surg., age at surgery; D, diastematomyelia; FR, fused ribs; HV, hemivertebra; KF, Klippel Feil; No. T fused, Number of thoracic levels fused; S, Sprengels.