

# The 18-cm Thoracic-Height Threshold and Pulmonary Function in Non-Neuromuscular Early-Onset Scoliosis

## A Reassessment

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**Background:** Thoracic spine height is cited as a crucial outcome measure in the treatment of early-onset scoliosis (EOS) because of its reported relationship to pulmonary function tests (PFTs). An 18-cm threshold has been proposed, although this single parameter might be overly simplistic for cases of different etiologies and deformity magnitude. We aimed to reevaluate pulmonary function in patients undergoing corrective surgery, assessing the role of residual scoliosis as well as spine elongation

**Methods:** Patients undergoing EOS correction with a minimum of 5 years of follow-up since initial treatment were evaluated. Standard spirometry (forced vital capacity [FVC], forced expiratory volume in 1 second [FEV1]) was correlated to deformity magnitude and T1-T12 height. Patients were compared by age at first surgery (<5 or ≥5 years), final thoracic height (≤18 or >18 cm), and percentage of predicted pulmonary function (<60% or ≥60%).

**Results:** Twenty-nine patients (15 congenital, 11 syndromic, and 3 idiopathic cases) were tested at a mean of 8.5 years following initial surgery. Twenty-two patients (mean initial age, 4.8 years) had growth-sparing instrumentation, and 7 patients (age, 5.1 years) had definitive fusion performed. Age at initial surgery was not associated with a difference in PFT results at the time of follow-up, and both age groups had ominously low percentages of predicted pulmonary-function volumes (50% to 55%). Only 18 of the 29 patients achieved a T1-T12 height of >18 cm. Those with a thoracic height of ≤18 cm had similar percentage-of-predicted spirometry results at the time of follow-up as those with greater thoracic height, possibly because of increased deformity correction. Only 14 of 29 patients had spirometry of ≥60% of predicted volume at the time of follow-up. These 14 had slightly smaller curves and slightly greater T1-T12 heights but significantly better spirometry results than the 15 subjects with <60% of predicted volume. For those with a T1-T12 height of ≤18 cm, the residual Cobb angle negatively correlated with spirometry results. In those with a final T1-T12 height of >18 cm, spirometry did correlate with thoracic height, especially when residual deformity was ≥60°.

**Conclusions:** Regardless of thoracic height of ≤18 or >18 cm, with residual curves of >50°, pulmonary function was ominously low in fully half of the patients, raising doubt about the value of this threshold as an EOS outcome parameter.

**Level of Evidence:** Prognostic Level IV. See Instructions for Authors for a complete description of levels of evidence.

Respiratory impairment from thoracic insufficiency syndrome (TIS) is considered the most serious morbid condition affecting patients with progressive early-onset scoliosis (EOS). Such children, arguably <6 years of age, have a substantial risk of pulmonary morbidity, resulting from the spine deformity itself or from ineffective surgical management. Historically, untreated “infantile-onset” scoliosis is associated with increased mortality, compared with later-onset (e.g., adolescent) scoliosis, because of respiratory failure<sup>1-4</sup>. However, early

fusion can also be ineffective in both arresting the progression and maintaining adequate pulmonary function<sup>5-8</sup>. Campbell et al. identified patients at risk due to spinal and thoracic malformations and due to inhibition of thoracic growth following early spinal fusion<sup>9</sup>. So-called growth-sparing treatment has evolved as an attempt to elongate the spine and/or chest wall while simultaneously controlling deformity, thus mitigating the pulmonary consequences of curve progression and early growth-arresting fusion.

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A frequently cited study<sup>6</sup> of respiratory consequences of early thoracic fusion has been interpreted as defining a longitudinal thoracic height (T1-T12) of 18 cm as a threshold below which long-term pulmonary function, as measured by the percentage of predicted normal volume for forced vital capacity (%FVC), would be significantly impaired and associated with TIS. This threshold was based on the normal thoracic spine height of a 5-year-old child<sup>10</sup> and on outcomes for patients who had predicted FVC of <50%—an amount approaching the 43% of predicted value historically associated with pulmonary failure and early mortality<sup>11</sup>—and thoracic height of <18 cm<sup>6</sup>. This threshold was recently investigated in a multicenter study as a minimum goal of distraction-based growth-sparing surgery<sup>12</sup>.

The value of the threshold as a crucial outcome measure may have been overstated<sup>13</sup>. Details of the original study<sup>6</sup> suggest several limitations: the majority of the 28 cases involved congenital scoliosis, calling into question the applicability of this threshold for other etiologies since patients with congenital scoliosis have decreased vital capacity compared with those with idiopathic scoliosis with the same degree of thoracic involvement and Cobb angle<sup>14</sup>. The majority of patients had in situ fusion with little or no curve correction attempted. Consequently, pulmonary outcomes likely reflected function in cases with large residual curves, which likely restrict lung function more profoundly in the congenital population than just the 1 dimension—thoracic height—might predict. Additionally, 5 (31%) of the 16 patients with T1-T12 height of

<18 cm actually had %FVC of >60%, possibly diminishing the invariability of this threshold.

The purpose of the current study was to reexamine the appropriateness of the threshold in a recent cohort of patients who underwent corrective procedures (definitive fusion or growth-sparing) before the age of 9 years with pulmonary function tests (PFTs) at a minimum of 5 years later. Our hypothesis was that patients with thoracic height of >18 cm would have better PFT results than those with thoracic height of ≤18 cm. A second hypothesis was that a corrected Cobb angle would correlate more closely to PFT results than thoracic height. A third hypothesis was that surgery before the age of 5 years would result in a shorter thoracic height and poorer PFTs than if surgery occurred at 5 years of age or older.

### Materials and Methods

Following university institutional review board approval, patients <9 years of age who had corrective surgery for EOS between 2004 and 2014 with complete follow-up data (minimum of 5 years) were identified. Patients with neuromuscular diagnoses, whose PFT results might be affected by muscle weakness, and those with skeletal dysplasias, whose thoracic height and arm span would be uninterpretable for comparison with individuals without dysplasias, were excluded, as were patients with primary pulmonary diseases.

Age at initial surgery and at most recent follow-up, diagnosis, and type of surgery (growth-sparing or definitive fusion) were recorded. Radiographic parameters were measured prior to

TABLE I Thoracic-Height Subgroups

	T1-T12 Height at Follow-up*		P Value
	≤18 cm (N = 11)	>18 cm (N = 18)	
Index Cobb (deg)	86.7 (61.6-129)	60.5 (24.6-102.6)	<b>0.001</b>
Index T1-T12 height (cm)	10.5 (8.2-13.4)	16.02 (10.7-22.1)	<b>&lt;0.001</b>
Index T6 chest depth (cm)	6.8 (4.6-10.1)	7.30 (5.0-10.30)	0.280
Age at index surgery (yr)	3.53 (1.34-6.08)	5.80 (2.69-8.71)	<b>0.005</b>
Time between surgery and follow-up (yr)	8.87 (5.44-12.84)	8.31 (5.31-12.52)	0.653
Age at follow-up (yr)	12.40 (8.66-15.96)	14.11 (9.47-18.22)	0.080
Final Cobb (deg)	53.02 (29-79)	45.07 (21-76.6)	0.225
Final T1-T12 height (cm)	15.1 (10.2-17.8)	21.46 (18.6-25.0)	<b>&lt;0.001</b>
Final T6 chest depth (cm)	8.5 (4.8-12.0)	8.87 (6.7-14.8)	0.719
Final FVC, actual (L)	1.12 (0.49-2.19)	1.91 (0.51-3.37)	<b>0.001</b>
Final FVC, % of predicted	48.45 (22-96)	56.78 (15-78)	0.200
Final FEV1, actual (L)	1.01 (0.36-2.01)	1.5 (0.45-2.44)	<b>0.009</b>
Final FEV1, % of predicted	49.82 (21-104)	51.61 (16-79)	0.544
△ Cobb (final – index) (deg)	–33.65 (–62.2[–8.4])	–15.47 (–61.6-10.8)	<b>0.019</b>
% correction of Cobb	–38.0 (–68.2[–13.6])	–22.0 (–67.5-23.4)	0.138
△ T1-T12 height (final – index) (cm)	4.59 (1.0-8.3)	5.45 (–2.3-10.8)	0.445
△ T6 chest depth (final – index) (cm)	1.7 (–2.47-5.4)	1.57 (–1.2-4.50)	0.928

\*The values are given as the mean and range.

the index procedure and at the most recent follow-up and included coronal-plane Cobb angle, T1-T12 height, and T6 thoracic depth, measured from the dorsal edge of the sternum to the ventral edge of the T6 body. PFTs utilizing standard spirometry were performed at the most recent follow-up, at a minimum of 5 years from the initial surgery, to determine actual FVC and forced expiratory volume in 1 second (FEV1) as well as the percentage of predicted normal volume (%FVC, %FEV1) for each test. Arm span was used to calculate the percentage of predicted volumes.

The radiographic and PFT data were then compared for each of the following patient groupings: <60% or ≥60% of predicted pulmonary function, T1-T12 height of ≤18 cm or >18 cm, and age at initial surgery of <5 or ≥5 years.

### Statistical Analysis

Statistical analyses were completed in SPSS Statistics (version 24; IBM). The initial analysis of continuous variables, such as Cobb angle, age at index surgery, T1-T12 height, and T6 depth, were first examined for normality with a Shapiro-Wilk test, and a t test and Mann-Whitney test were used for 2-group comparisons as appropriate. Pearson correlation coefficients were calculated to assess the relationship between postoperative pulmonary and radiographic outcomes. Contingency tables were used to describe frequency distributions of categorical variables, such as classification of EOS<sup>15</sup> and index treatment. Significance was set at  $p < 0.05$ .

### Source of Funding

No external funds were received in support of this research.

### Results

Twenty-nine patients (mean Cobb angle, 72°; range, 25° to 129°) met the inclusion criteria: 15 patients had a congenital diagnosis; 11, syndromic; and 3, idiopathic. Syndromic diagnoses included Marfan syndrome, neurofibromatosis, and arthrogryposis (2 cases each) and osteogenesis imperfecta, Prader-Willi syndrome, Dubowitz syndrome, Pierre Robin syndrome, and DiGeorge syndrome (1 case each). Twenty-two patients (mean age at initial surgery of 4.8 years; range, 1.3 to 8.7 years) underwent growth-sparing treatment, while 7 patients (mean age of 5.1 years; range, 3.1 to 7.5 years) had definitive fusion as the initial treatment. Mean follow-up from the time of the initial surgery was 8.5 years (minimum, 5 years; range, 5.4 to 12.8 years).

### Age at Initial Surgery (<5 or ≥5 Years)

Seventeen patients (mean age, 3.5 years) were <5 years of age at the initial surgery, while 12 patients (mean age, 6.9 years) were ≥5 years of age. The older group had had a greater index T1-T12 height (mean, 16.1 versus 12.4 cm;  $p = 0.02$ ), although the index Cobb angles and T6 thoracic depth did not differ. Follow-up Cobb angles and thoracic depth did not differ, while T1-T12 height was again greater in the group ≥5 years of age (mean, 20.7 versus 17.9 cm;  $p = 0.06$ ). This increased thoracic

TABLE II Percentage of Predicted Pulmonary-Function Subgroups

	% of Predicted Pulmonary Function at Follow-up*		P Value
	<60% (N = 15)	≥60% (N = 14)	
Index Cobb (deg)	71.4 (24.6-129)	69.47 (42-102.6)	0.913
Index T1-T12 height (cm)	12.88 (8.2-19.8)	15.04 (8.8-22.1)	0.127
Index T6 chest depth (cm)	6.91 (4.6-10.30)	7.33 (5.0-10.1)	0.407
Age at index surgery (yr)	4.36 (1.34-8.06)	5.56 (1.55-8.71)	0.089
Time between surgery and follow-up (yr)	9.61 (5.31-12.84)	7.35 (5.44-11.48)	<b>0.036</b>
Age at follow-up (yr)	13.97 (8.72-18.22)	12.91 (8.66-15.82)	0.169
Final Cobb (deg)	53.36 (23.8-79)	42.43 (21-64)	<b>0.067</b>
Final T1-T12 height (cm)	17.95 (10.2-21.8)	20.21 (12.8-25.0)	<b>0.049</b>
Final T6 chest depth (cm)	8.74 (4.8-14.8)	8.71 (6.7-11.1)	0.743
Final FVC, actual (L)	1.23 (0.49-2.18)	2.01 (1.28-3.37)	<b>0.002</b>
Final FVC, % of predicted	39.13 (15-56)	69.14 (56-96)	<b>&lt;0.001</b>
Final FEV1, actual (L)	1.03 (0.36-2.03)	1.62 (1.24-2.44)	<b>0.001</b>
Final FEV1, % of predicted	36.93 (16-56)	65.93 (34-104)	<b>&lt;0.001</b>
△ Cobb (final – index) (deg)	-17.99 (-62.2-10.8)	-27.04 (-61.6-7)	0.214
% correction of Cobb	-20.7 (-68.2-23.4)	-35.8 (-67.5-12.3)	<b>0.067</b>
△ T1-T12 height (final – index) (cm)	5.07 (0.4-8.88)	5.17 (-2.3-10.8)	0.983
△ T6 chest depth (final – index) (cm)	1.83 (-2.47-5.4)	1.39 (-1.2-4.94)	0.284

\*The values are given as the mean and range.

**TABLE III Correlations: PFTs with Cobb and T1-T12 Height at Final Follow-up (N = 29)**

	Final Cobb		Final T1-T12 Height	
	R Value	P Value	R Value	P Value
Final FVC, actual	-0.351	0.062	0.634	<b>&lt;0.001</b>
Final FVC, % of predicted	-0.328	0.083	0.218	0.256
Final FEV1, actual	-0.263	0.168	0.504	<b>0.005</b>
Final FEV1, % of predicted	-0.180	0.351	0.035	0.858

height was not associated with better actual or percentage of predicted FVC and FEV1 volumes. Thus, our third hypothesis concerning poorer PFT results in the age group of <5 years was not confirmed; however, ominously, both groups had low % FVC and %FEV1 values (50% to 55%).

#### Thoracic Height ( $\leq 18$ or $>18$ cm)

Only 18 of 29 patients achieved a T1-T12 height of  $>18$  cm at the time of follow-up (Table I). The 11 patients with a measurement of  $\leq 18$  cm were younger at initial surgery (mean, 3.5 versus 5.8 years;  $p = 0.005$ ). Although the  $\leq 18$ -cm group had larger initial Cobb angles (mean,  $86.7^\circ$  versus  $60.5^\circ$ ;  $p = 0.001$ ) and shorter initial thoracic height (mean, 10.5 versus 16 cm;  $p < 0.001$ ), there was no difference in Cobb angles at the time of follow-up (mean,  $53^\circ$  versus  $45^\circ$ ), possibly because of more effective surgical correction (mean,  $-33.6^\circ$ ) for the patients in the  $\leq 18$ -cm group. The difference in thoracic height at the time of follow-up remained significant (mean, 15.1 versus 21.5 cm;  $p < 0.001$ ). Although actual FVC and FEV1 volumes were significantly greater in the  $>18$ -cm group, the %FVC and %FEV1 did not differ, again possibly because of better correction in the  $\leq 18$ -cm group. Thus, our first hypothesis—that a T1-T12 height of  $>18$  cm would produce better follow-up PFT results—was confirmed only for the actual FVC and FEV1 volumes.

#### PFTs (<60% Versus $\geq 60\%$ of Predicted Volume)

Only 14 of 29 patients had  $\geq 60\%$  of predicted pulmonary function at the time of follow-up (Table II). These 14 patients had significantly higher actual and percentage of predicted pulmonary-function volumes than the 15 patients with  $<60\%$  of predicted pulmonary function. Unexpectedly, there was no difference between the 2 groups regarding age at initial surgery, index Cobb angle, or index thoracic height or depth. The only differences between the 2 groups at the time of follow-up were a slightly greater percentage of deformity correction, slightly smaller residual deformity, and slightly greater thoracic height in the  $\geq 60\%$  group. Our hypothesis that residual deformity and degree of correction would correlate with better PFT results was modestly confirmed, as was the hypothesis concerning thoracic height just mentioned. The percentages of predicted pulmonary-function volume at the time of follow-up in the  $<60\%$  group were ominously low, ranging from 15% to 56% (mean, 37% to 39%).

#### Correlation of Radiographic Parameters and PFTs

We assessed actual and percentage of predicted pulmonary function in relation to radiographic parameters at the time of final follow-up using a Pearson correlation test (Table III). For the entire cohort, the T1-T12 height demonstrated a moderate correlation ( $r = 0.5$  to  $0.63$ ) to actual FVC and FEV1 volumes but no correlation to percentage of predicted volumes or the residual deformity. However, when the cohort was divided into those with a final curve measurement of  $<60^\circ$  or  $\geq 60^\circ$  (Table IV), the patients with larger residual curves ( $n = 8$ ) showed good correlation ( $0.66$  to  $0.80$ ) between actual and percentage of predicted volumes and T1-T12 height. For curves of  $<60^\circ$  ( $n = 21$ ), there was no correlation between PFT values and curve magnitude or thoracic height.

When dividing the patients into groups with T1-T12 height of  $\leq 18$  or  $>18$  cm (Table V), the actual FVC and FEV1 volumes of the  $\leq 18$ -cm cohort ( $n = 11$ ) were negatively correlated with residual Cobb angle, while the  $>18$ -cm cohort showed no such correlation. Furthermore, for the  $>18$ -cm cohort, there was only very modest correlation between PFT parameters and thoracic height, and no correlation with residual deformity.

#### Subanalysis of Congenital and Syndromic Diagnoses

The 15 congenital and 11 syndromic cases were analyzed separately to determine a possible effect of diagnosis on the outcomes. All congenital cases involved segmentation anomalies over at least 6 segments, with or without rib fusions. Eight patients had a T1-T12 height of  $\leq 18$  cm at the time of follow-up, and 7 had a height of  $>18$  cm. Cases with  $>18$ -cm height had smaller index curves and greater index T1-T12 heights, and these differences were maintained at the time of follow-up, but no differences in PFT outcomes were demonstrated (Table VI).

**TABLE IV Correlations: PFTs with Cobb and T1-T12 Height at Final Follow-up by Cobb Subgroup**

	Final Cobb			Final T1-T12 Height		
	N	R Value	P Value	N	R Value	P Value
Final Cobb $<60^\circ$						
Final FVC, actual	21	-0.130	0.575	21	0.545	<b>0.011</b>
Final FVC, % of predicted	21	-0.136	0.558	21	-0.040	0.862
Final FEV1, actual	21	-0.020	0.930	21	0.348	0.122
Final FEV1, % of predicted	21	0.004	0.986	21	-0.225	0.326
Final Cobb $\geq 60^\circ$						
Final FVC, actual	8	-0.606	0.111	8	0.795	<b>0.018</b>
Final FVC, % of predicted	8	-0.468	0.243	8	0.750	<b>0.032</b>
Final FEV1, actual	8	-0.621	0.101	8	0.769	<b>0.026</b>
Final FEV1, % of predicted	8	-0.472	0.237	8	0.656	0.077

TABLE V Correlations: PFTs with Cobb and T1-T12 Height at Final Follow-Up by Thoracic-Height Subgroup

	Final Cobb			Final T1-T12 Height		
	N	R Value	P Value	N	R Value	P Value
T1-T12 height ≤18 cm						
Final FVC, actual	11	-0.657	<b>0.028</b>	11	0.078	0.820
Final FVC, % of predicted	11	-0.419	0.200	11	-0.209	0.538
Final FEV1, actual	11	-0.637	<b>0.035</b>	11	0.005	0.989
Final FEV1, % of predicted	11	-0.413	0.207	11	-0.254	0.451
T1-T12 height >18 cm						
Final FVC, actual	18	-0.106	0.677	18	0.520	<b>0.027</b>
Final FVC, % of predicted	18	-0.178	0.479	18	0.450	0.061
Final FEV1, actual	18	0.069	0.784	18	0.402	0.098
Final FEV1, % of predicted	18	0.081	0.750	18	0.350	0.155

Among the congenital cases, 9 patients had <60% of predicted pulmonary function, while 6 had ≥60% (Table VII). In spite of the marked differences in PFT results (actual and percentage of predicted) between the groups ( $p = 0.001$  to  $0.008$ ), the groups did not differ in terms of index Cobb angle or thoracic height, height gained, residual Cobb angle, or thoracic height at the time of follow-up.

Among the syndromic cases, there were 3 patients with a T1-T12 height of ≤18 cm and 8 patients with a height of

>18 cm at the time of follow-up (Table VIII). As in the congenital group, there was a trend toward a smaller index Cobb angle and a greater thoracic height in the >18-cm group. At the time of follow-up, thoracic height remained greater, while residual Cobb angles did not differ ( $48^\circ$ ) because of greater correction in the ≤18-cm group. PFTs showed greater actual volumes in the >18-cm group, but there was no difference in the percentage of predicted values, similar to findings for the entire cohort.

TABLE VI Thoracic-Height Subgroups: Congenital Etiology (N = 15)

	Final T1-T12 Height*		P Value
	≤18 cm (N = 8)	>18 cm (N = 7)	
Index Cobb (deg)	81.81 (61.6-97)	53.46 (24.6-67.6)	<b>0.004</b>
Index T1-T12 height (cm)	10.16 (8.2-13.37)	15.45 (11.22-18)	<b>0.004</b>
Index T6 chest depth (cm)	6.87 (4.6-10.1)	6.99 (6.09-8.1)	0.684
Age at index surgery (yr)	3.49 (1.34-6.09)	5.2 (2.69-8.31)	0.132
Time between surgery and follow-up (yr)	9.05 (5.44-12.84)	8.2 (5.31-12.51)	0.563
Age at follow-up (yr)	12.53 (8.72-14.89)	13.39 (9.47-15.82)	0.355
Final major Cobb (deg)	54.78 (35-75)	37.97 (22-64)	<b>0.049</b>
Final T1-T12 height (cm)	14.54 (10.2-17.2)	21.7 (19.1-25)	<b>0.001</b>
Final T6 chest depth (cm)	8.75 (4.8-12)	8.03 (6.7-10.1)	0.452
Final FVC, actual (L)	1.21 (0.69-2.19)	1.64 (0.51-2.33)	0.105
Final FVC, % of predicted	52.63 (25-96)	53.29 (15-78)	0.728
Final FEV1, actual (L)	1.1 (0.68-2.01)	1.3 (0.45-2.06)	0.643
Final FEV1, % of predicted	54.5 (21-104)	47.43 (16-79)	0.772
Δ Cobb (final - index) (deg)	-27.04 (-43.4[-8.4])	-15.49 (-45.6-7)	0.247
Δ T1-T12 height (final - index) (cm)	4.38 (1-8.3)	6.25 (2.68-8.88)	0.165
Δ T6 chest depth (final - index) (cm)	1.88 (-2.47-5.4)	1.04 (0.1-2.3)	0.246

\*The values are given as the mean and range.

TABLE VII Percentage of Predicted Pulmonary-Function Subgroups: Congenital Etiology (N = 15)

	Final % of Predicted Pulmonary Function*		P Value
	<60% (N = 9)	≥60% (N = 6)	
Index Cobb ( <i>deg</i> )	66.89 (24.6-97)	71.12 (54-95.4)	0.724
Index T1-T12 height ( <i>cm</i> )	11.92 (8.2-17.22)	13.68 (8.8-18)	0.409
Index T6 chest depth ( <i>cm</i> )	6.64 (4.6-8.1)	7.36 (5.66-10.1)	0.442
Age at index surgery ( <i>yr</i> )	3.73 (1.34-7.49)	5.11 (1.55-8.31)	0.239
Time between surgery and follow-up ( <i>yr</i> )	9.33 (5.31-12.84)	7.63 (5.44-11.48)	0.239
Age at follow-up ( <i>yr</i> )	13.06 (8.72-15.2)	12.74 (10.74-15.82)	0.480
Final Cobb ( <i>deg</i> )	50 (23.8-75)	42.33 (22-64)	0.289
Final T1-T12 height ( <i>cm</i> )	16.99 (10.2-21.8)	19.22 (12.8-25)	0.346
Final T6 chest depth ( <i>cm</i> )	8.08 (4.8-12)	8.92 (6.7-11.1)	0.367
Final FVC, actual ( <i>L</i> )	1.09 (0.51-1.85)	1.9 (1.28-2.33)	<b>0.008</b>
Final FVC, % of predicted	39.11 (15-56)	73.67 (56-96)	<b>0.002</b>
Final FEV1, actual ( <i>L</i> )	0.86 (0.45-1.21)	1.7 (1.24-2.06)	<b>0.001</b>
Final FEV1, % of predicted	35.22 (16-48)	75.17 (56-104)	<b>0.001</b>
Δ Cobb (final – index) ( <i>deg</i> )	–16.89 (–37[–0.8])	–28.78 (–45.6-7)	0.11
Δ T1-T12 height (final – index) ( <i>cm</i> )	5.07 (2-8.88)	5.54 (1-8.8)	0.724
Δ T6 chest depth (final – index) ( <i>cm</i> )	1.44 (–2.47-5.4)	1.56 (0.1-4.94)	0.679

\*The values are given as the mean and range.

Among the syndromic cases, 4 patients had <60% of predicted pulmonary function and 7 had ≥60%. Again, there were sharp differences between the groups in percent of predicted volumes (Table IX). However, as with the congenital cohort, there were no differences in radiographic parameters, either index or follow-up, to explain these differences.

## Discussion

The 2008 study by Karol et al.<sup>6</sup> is frequently quoted when discussing indications for growth-sparing surgical procedures for EOS, emphasizing the 18-cm threshold as a goal for successful treatment as measured by %FVC. This threshold was based on the finding that 16 of 28 patients in the study did not achieve this T1-T12 height—the thoracic height of a normal 5-year-old child<sup>10</sup>—and that their mean %FVC was 48.2%, a figure perilously similar to the <43% of predicted vital capacity reported by Pehrsson et al. for adults with eventual respiratory failure<sup>11</sup>. Five to 11 thoracic segments were fused in the 16 subjects, and all with <50% FVC had at least 5 segments fused. This established a principle that growth-arresting fusion of >4 segments of the thoracic spine in patients <9 years of age should be avoided due to the potential for inducing TIS by limiting thoracic spine height.

The acceptance of this threshold in the treatment of EOS has several limitations. Residual deformity magnitude at the time of follow-up PFTs was not reported, an important omission since the negative association of scoliosis magnitude and pul-

monary function has been recognized, most notably in preoperative studies of adolescent idiopathic scoliosis<sup>16,17</sup>. Three-quarters of the 2008 cohort had congenital deformities and were treated by in situ fusion, so that curve correction was minimal, with the pulmonary outcome reflecting what might be expected for large residual untreated deformities. A recent multicenter study<sup>12</sup> assessing achievement of the 18-cm threshold from distraction-based treatment of EOS reported that only 48% of patients with congenital scoliosis achieved the threshold, compared with 78% for all other etiologies, and that those not reaching the threshold had only 4% correction—essentially an in situ fusion outcome. It is not surprising, then, that cases of congenital deformities gaining little or no correction will likely not attain the 18-cm threshold, thus questioning its value as an important outcome measure for this diagnosis. Finally, overlooked when invoking the apparently poor prognosis of a short thoracic height was the finding that 5 (31%) of the 16 patients with a T1-T12 height of <18 cm actually had %FVC of ≥60%,<sup>6</sup> thus calling into question the validity of this “threshold” as a major outcome parameter<sup>13</sup>.

In the current study, we reassessed the threshold in a cohort of 29 patients with EOS who had undergone corrective, rather than in situ, surgery to evaluate effects of both height and deformity correction on pulmonary outcome. T1-T12 height tended to best correlate with actual FVC or FEV1 volumes (as opposed to percentage of predicted volumes) for the entire cohort (Table III), and with most PFT measures (Table IV) in patients with curves of ≥60°. However, the most striking

TABLE VIII Thoracic-Height Subgroups: Syndromic Etiology (N = 11)

	Final T1-T12 Height*		P Value
	≤18 cm (N = 3)	>18 cm (N = 8)	
Index Cobb ( <i>deg</i> )	99.6 (78.6-129)	64.94 (46.2-102.6)	0.066
Index T1-T12 height ( <i>cm</i> )	11.43 (10.79-11.8)	17.03 (10.7-22.1)	0.066
Index T6 chest depth ( <i>cm</i> )	6.64 (6-7.51)	7.18 (5-9.6)	0.540
Age at index surgery ( <i>yr</i> )	3.66 (3.11-4.39)	6.21 (3.73-8.71)	<b>0.025</b>
Time between surgery and follow-up ( <i>yr</i> )	8.39 (5.56-11.58)	8.31 (5.46-11.96)	1.00
Age at follow-up ( <i>yr</i> )	12.05 (8.66-15.96)	14.53 (10.64-18.22)	0.414
Final Cobb ( <i>deg</i> )	48.33 (29-79)	48 (33-60)	0.540
Final T1-T12 height ( <i>cm</i> )	16.57 (14.7-17.8)	21.56 (18.6-24.1)	<b>0.014</b>
Final T6 chest depth ( <i>cm</i> )	7.83 (6.7-9.4)	8.85 (7-9.8)	0.305
Final FVC, actual ( <i>L</i> )	0.88 (0.49-1.31)	2.23 (1.43-3.37)	<b>0.014</b>
Final FVC, % of predicted	37.33 (22-64)	61.25 (45-74)	0.152
Final FEV1, actual ( <i>L</i> )	0.78 (0.36-1.26)	1.74 (1.36-2.44)	<b>0.014</b>
Final FEV1, % of predicted	37.33 (21-70)	56.38 (34-67)	0.413
△ Cobb (final – index) ( <i>deg</i> )	–51.3 (–62.2[–41.6])	–16.94 (–61.6-10.8)	<b>0.041</b>
△ T1-T12 height (final – index) ( <i>cm</i> )	5.14 (2.9-6.41)	4.53 (–2.3-10.8)	0.540
△ T6 chest depth (final – index) ( <i>cm</i> )	1.2 (0.7-1.89)	1.68 (–1.2-2.9)	0.219

\*The values are given as the mean and range.

TABLE IX Percentage of Predicted Pulmonary-Function Subgroups: Syndromic Etiology (N = 11)

	Final % of Predicted Pulmonary Function*		P Value
	<60% (N = 4)	≥60% (N = 7)	
Index Cobb ( <i>deg</i> )	78.6 (46.2-129)	71.99 (50.4-102.6)	0.850
Index T1-T12 height ( <i>cm</i> )	14.56 (10.79-19.8)	16.04 (10.7-22.1)	0.450
Index T6 chest depth ( <i>cm</i> )	6.83 (6.2-7.51)	7.14 (5-9.6)	0.850
Age at index surgery ( <i>yr</i> )	4.72 (3.49-6.26)	5.97 (3.11-8.71)	0.257
Time between surgery and follow-up ( <i>yr</i> )	10.58 (8.03-11.96)	7.05 (5.46-9.43)	<b>0.023</b>
Age at follow-up ( <i>yr</i> )	15.3 (11.52-18.22)	13.02 (8.66-15.37)	0.089
Final Cobb ( <i>deg</i> )	52.5 (29-79)	45.57 (33-60)	0.571
Final T1-T12 height ( <i>cm</i> )	18.88 (17.2-20.3)	20.96 (14.7-24.1)	0.131
Final T6 chest depth ( <i>cm</i> )	8.6 (7.4-9.4)	8.56 (6.7-9.8)	0.924
Final FVC, actual ( <i>L</i> )	1.4 (0.49-2.18)	2.13 (1.31-3.37)	0.257
Final FVC, % of predicted	36 (22-51)	65.43 (61-74)	<b>0.008</b>
Final FEV1, actual ( <i>L</i> )	1.27 (0.36-2.03)	1.6 (1.26-2.44)	0.705
Final FEV1, % of predicted	37 (21-56)	59.29 (34-70)	<b>0.023</b>
△ Cobb (final – index) ( <i>deg</i> )	–26.1 (–62.2-10.8)	–26.41 (–61.6-4.6)	0.850
△ T1-T12 height (final – index) ( <i>cm</i> )	4.32 (0.4-6.41)	4.92 (–2.3-10.8)	1.000

\*The values are given as the mean and range.

finding of this study was the ominously low PFT results for fully half of the cohort (15 subjects with <60% of predicted FVC or FEV1, Table II), and the equally ominous ~50% of predicted mean FVC or FEV1 regardless of the T1-T12 height (Table I) or the age at which surgery was performed (<5 or ≥5 years). Such low values for the percentage of predicted pulmonary function would be expected to cause notable eventual pulmonary morbidity<sup>11</sup>.

The effect of curve correction on pulmonary outcome, however modest, was noted by the negative correlation of actual FVC and FEV1 volumes and residual Cobb angle in subjects with a T1-T12 height of ≤18 cm (Table V), and by the slightly smaller mean Cobb measurement (42°) and better percent correction (36%)—as well as greater mean T1-T12 height (20.2 cm)—in the group with ≥60% of predicted pulmonary function (Table II). In subjects with ≤18 cm of T1-T12 height, the percentages of predicted volume were no different from those with >18-cm height, possibly because of comparatively better curve correction, to ~53°, in the shorter thoracic-height group (Table I). The fact remains, however, that while better correction and greater T1-T12 height were associated with larger actual FVC and FEV1 volumes, the %FVC and %FEV1 remained ominously low, at 48% to 57%.

Recently, 16 patients from the original 2008 cohort were reexamined at an average of 23 years (minimum, 18 years) of follow-up<sup>18</sup>. Pulmonary-function deterioration with the passage of time affected essentially every patient at a higher rate than expected because of aging<sup>19,20</sup>. Ten of the 16 had thoracic height of ≥18 cm, but only 3 had %FVC exceeding 60%, and those 3 patients each had had >86% FVC when tested in 2008. The remaining 7 patients with a thoracic height of ≥18 cm had follow-up %FVCs varying from 38% to 50%—thus, the 18-cm threshold did not appear to be protective of what was believed to be sufficient %FVC in the original study. The remaining 6 of 16 subjects had T1-T12 height of <18 cm with ominously low %FVC of 14% to 36%, and the subsequent death of 1 patient with 14% FVC emphasized just how ominous these values are. It suggests that achieving the 18-cm threshold, even with ~50°

residual curve, does not reliably stabilize the long-term pulmonary prognosis for the patient with EOS undergoing thoracic fusion before the age of 9 years.

This study's limitations, in addition to those mentioned in the introduction reflecting limitations of the 2008 study, include being retrospective and having a small number of subjects with diverse etiologies, including few idiopathic cases, although in analyzing the congenital and syndromic subgroups separately, we found no important differences when comparing these more homogeneous groups to the overall cohort. No patients had pretreatment baseline PFTs, nor do data exist for a comparative cohort of subjects with ≤18-cm thoracic height but with minimal or no scoliosis. With regard to congenital scoliosis, others have shown the difficulty in achieving an 18-cm thoracic height<sup>12</sup>, suggesting that deformity correction in such cases may be crucial to maintaining pulmonary function at ≥60% of predicted, a rather modest immediate goal of EOS treatment (Tables I and V).

Considering the documented deterioration in pulmonary function over a minimum of 18 years of follow-up<sup>18</sup>, it seems likely that %FVC or %FEV1 of <60% prior to maturity is unlikely to avoid respiratory morbidity from TIS, regardless of T1-T12 height exceeding 18 cm. Thus, it is our belief that this frequently quoted threshold height cannot be invoked as a main indication for growth-sparing surgical treatment. ■

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