

Mini-Symposium: Medical and surgical aspects of chest wall deformities

Thoracic malformation with early-onset scoliosis: Effect of serial VEPTR expansion thoracoplasty on lung growth and function in children

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SUMMARY

The effect on pulmonary function of serial VEPTR expansion thoracoplasty was studied longitudinally in anesthetized children with spondylothoracic dysplasia using a special mobile unit. The median age of 24 children at the start of surgery was 4.6 years (1.8–10.8) and most exhibited a moderate-to-severe restrictive lung defect. After a median of 3.2 years (1.0–6.5), their forced vital capacity (FVC) was found to have increased by an average of 11.1%/year. The rate of increase was greater in children who were younger than 6 years at the start of the study than in older children (14.5% versus 6.5%, $p < 0.01$). The average specific respiratory system compliance (C_{rs}) was mildly-to-moderately decreased at the start, and over the study it decreased on average to 56% of the initial value in spite of clinically successful expansion thoracoplasty and lung growth, indicating increasing stiffness of the thorax with growth.

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INTRODUCTION

Surgical interventions for infants and children with thoracic and spinal malformations in early-onset scoliosis have focused almost exclusively on the correction of spinal deformity. Such treatments commonly involved spinal fusion, which stunts growth of the spinal column and limits the thoracic volume and lung growth.^{1,2} More recently, Campbell et al. have focused their attention on the interrelationship between the spine, thoracic wall and lungs with growth, especially in children with combined malformations of the thorax and spine (spondylothoracic dysplasia).³ They have developed vertical expandable prosthetic titanium ribs (VEPTR) for serial expansion thoracoplasty to control the three-dimensional progression with body growth of both spine and chest wall deformities, and resultant respiratory insufficiency, which they have termed thoracic insufficiency syndrome (TIS).⁴

Using the VEPTR technique, Campbell et al. have reported growth of the concave side of the thorax along with clinical improvements in the respiratory status of afflicted children.^{4,5} It has been assumed that the lung volume would also increase with serial expansion of the concave side of the hemithorax. However, direct measurements of lung volumes and pulmonary function in these patients have been limited to a small number of older children.² Furthermore, the longitudinal effects of serial VEPTR thoracoplasty on respiratory function have not been reported in the literature.

Previously, we have developed a mobile unit to perform a variety of pulmonary function tests (PFTs), using both passive and forced deflation techniques, in anesthetized or heavily sedated infants and children in the operating room or intensive care unit.⁶ In our preliminary report, we described the results for 41 serial PFTs in 10 children (aged 2–10 years) in the operating room under general anaesthesia immediately before VEPTR thoracoplasty. The study methods and techniques used are detailed below, and the effects of VEPTR on lung volume and respiratory mechanics described in this preliminary report as well as more recent follow-up study results in a larger patient population are summarized.

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STUDY DESIGN

Pulmonary function tests

Bedside PFTs were performed following the induction of general anaesthesia and intubation with a cuffed endotracheal (ET) tube by means of passive and forced deflation techniques in patients in a supine position.⁶

Passive deflation technique

A three-way slide valve and pneumotachograph were inserted between the anaesthesia delivery circuit and the ET tube. Lungs were partially inflated to the end-inspiratory airway pressure (P_{aw}) of 10 cmH₂O and were then allowed to deflate passively via a pneumotachograph to the end-expiratory volume (functional residual capacity [FRC] or relaxation volume [V_r]), by shifting the slide valve and opening the airway to atmospheric pressure. Respiratory system compliance (C_{rs}) was determined by dividing expiratory tidal volume (V_T) by P_{aw} of 10 ($C_{rs} = V_T/P_{aw}$).

Forced deflation technique

After the initial measurement of C_{rs} with passive deflation, the lungs were slowly inflated three times to total lung capacity (TLC) at $P_{aw} = 40$ cmH₂O to eliminate pre-existing airway closure and to establish a constant volume history. From TLC, lungs were deflated within seconds to residual volume (RV) by quickly shifting the slide valve and exposing the airway to a large negative pressure reservoir (-40 cmH₂O). Simultaneously, a maximum expiratory flow-volume (MEFV) curve was displayed on an X-Y screen. The procedure was repeated several times to ensure reproducibility of the MEFV curves. From the MEFV curves, maximum expiratory flow (MEF) at 25% and 10% of forced vital capacity (FVC) from RV (MEF₂₅, MEF₁₀ or FEF₇₅, FEF₉₀) were obtained.⁶

Physiologically, MEFs for small lung volumes are determined by dynamic compression of intrathoracic airways, are independent of the degree of expiratory effort (in clinical settings) and are extremely sensitive indices of relatively small airway resistance, independent of resistance in large extra- and intra-thoracic airways.^{6,7} For children older than 6 years, the predicted values of FVC and MEFs were taken from data reported by Schoenberg et al.⁸ For MEFs and C_{rs} in younger children, normative values developed at the Pulmonology Laboratory of the Children's Hospital of Pittsburgh were used. Arm span instead of height was used for computation in children with scoliosis.

Anaesthetic management

Anaesthesia in most patients was induced with inhalation anaesthesia, and patients were intubated orally with a cuffed ET tube and an intermediate-acting muscle relaxant was given. They were mechanically ventilated throughout the surgical procedures and were monitored with standard anaesthesia monitoring. Anaesthesia was maintained with relatively low concentrations of inhaled anaesthetics or continuous intravenous infusion of propofol, together with intravenous opioid supplementation.

Surgical procedure

The VEPTTR was inserted with the patient in the lateral decubitus position on the convex side of the thorax with continuous monitoring of somatosensory evoked potentials (SSEP) in all cases. An incision was made from the upper paravertebral region (T2–T3), extending to beneath the scapula in a curvilinear fashion. The chest wall was exposed and the upper and lower rib attachments for the titanium rib were prepared around the rib

without entering the pleural space. For the insertion of rib-to-spine hybrid VEPTTR for patients with severe scoliosis, a separate incision was made and the lamina of the lower vertebra was exposed. In patients with congenital rib fusion, the ribs were separated anteriorly to the junction of the spine and a wedge thoracoplasty was performed. The gap in the chest wall, if present, was filled with a porcine graft.⁴ The chest tubes were inserted and the thorax closed. The patients recovered in the paediatric intensive care unit over several days. The surgical procedure was repeated approximately every 6 months for the vertical expansion of the thorax with VEPTTR, aiming for a total expansion of 10 mm. In some children with bilateral thoracic hypoplasias, the second set of VEPTTR was inserted in the contralateral thorax.

Patients in the preliminary study

In the preliminary report,⁹ data from 10 children with spine and chest wall deformities with early-onset scoliosis with TIS were included. The age range was 1.8–9.8 years (median 4.3) at the initial PFTs. One child had Jarcho–Levine syndrome and two others had associated congenital heart disease. Each child had a minimum of two separate bedside PFTs under general anaesthesia prior to the insertion or expansion of VEPTTR. The time span between the first and the last test was 7–33 months (median 23). A total of 41 PFTs were performed. The study was approved by University of Pittsburgh Institutional Review Board (IRB) and informed consent was obtained from the parents.

Patients in the follow-up study

As of November 2007, 24 children (15 girls and 9 boys) with an age range at the initial tests of 1.8–10.8 years (median 4.6) had undergone serial VEPTTR thoracoplasties (2–12 times). Six of these children had VEPTTR insertions in both hemithoraces. The interval between the first and last tests varied between 12 months and 5.6 years (median 2.7 years). There were 151 PFTs performed. All children had the initial diagnosis of TIS with early-onset scoliosis (congenital, neuromuscular and infantile with failed previous treatment) together with fused ribs (4), congenital heart disease (4), VATER association (3), tracheoesophageal fistula (3), tethered spinal cord (4), Jarcho–Levine syndrome (2), Noonan syndrome (1), Conradi–Hünermann chondrodysplasia (1), Williams syndrome (1), Goldenhar syndrome (1) and myelomeningocele or their combinations. Four patients had undergone anterior or antero-posterior partial spinal fusions prior to VEPTTR insertion. One patient was ventilator dependent via a tracheostomy, and another was on continuous positive airway pressure (CPAP) and was oxygen dependent.

RESULTS

Preliminary results

At the initial PFTs, there were moderate-to-severe restrictive defects in most children, with an average FVC of $69.2 \pm 17.4\%$ of predicted value (range 36–101%); as a group, FVC was significantly decreased ($p < 0.001$).⁹ At the last PFTs following serial VEPTTR thoracoplasties, there were definite signs of lung growth, as indicated by significant increases in FVC at an average annual rate of $19.6 \pm 6.0\%$ ($p < 0.01$), or about 73% of normal growth. On the other hand, in terms of % predicted values, FVC changed minimally (from $69.2 \pm 17.4\%$ to $70.3 \pm 11.3\%$ predicted; $p > 0.1$). Specific compliance (C_{rs}) was abnormally decreased ($C_{rs} < 1.0$ mL/cmH₂O/kg) in some patients, indicating abnormally stiff chest walls. The mean C_{rs} was near the lower normal limit at the initial tests (1.2 ± 0.44 /kg) and was

not significantly different at the last tests ($1.0 \pm 0.41/\text{kg}$; $p > 0.1$). As has been reported previously,^{10,11} a number of patients in our study also showed clinical improvements in their quality of life. Complications were relatively infrequent and were without major morbidity, and there was no mortality (see below). Figs. 1 and 2 are the chest radiographs of one child before and after the insertion of VEPTRs.

Based on this preliminary study, serial VEPTR thoracoplasties appeared to be a promising approach for the surgical treatment of TIS, by correcting or attenuating the progression of scoliosis from the uneven growth of the hemithorax. However, a number of questions remained. Would the increase in lung volume keep up with body growth with serial expansions of the hemithorax over the years? What is the optimal age at which to start the application of VEPTR? More specifically, is VEPTR effective even when the initial thoracic expansion is done in children aged 6 or even 10 years, when morphological and structural development of lung tissues has long been completed? In the preliminary study, no significant difference could be detected in the degree of improvement in FVC between the younger and older children. The purpose of the ongoing follow-up study in these children with TIS has been to answer these questions.

Results from the follow-up study

The mean FVC at the initial tests, based on arm span, was $71.9 \pm 21.6\%$ (range 40–103%) with apparent overestimation in at least three patients due to their unusually short arm spans (e.g. dwarfism). Eight patients had an FVC 60% below predicted, indicating severe restrictive defect.

Over the duration of the study, FVC increased at an average of $11.1 \pm 9.6\%/ \text{year}$ ($p < 0.001$) (Fig. 3; Table 1). Patients were divided into two groups, based on their age at the initial VEPTR thoracoplasty. There was a significant difference in the degree of annual increase in FVC. In those in whom initial surgery was performed before the age 6.0 years ($n = 16$), the mean increase in FVC was $14.7 \pm 8.5\%/ \text{year}$,



Figure 1. Chest radiograph of a 3-year-old child with spondylocostal dysplasia and congenital scoliosis before the initial insertion of VEPTR. The Cobb angle (T3–T12) is 45° .



Figure 2. Chest radiograph of the same child as in Fig. 1, now 7 years old, with VEPTR in the left hemithorax. The child underwent eight VEPTR expansion thoracoplasties, including two device replacements over the 4 years. The shorter device is placed laterally between the third and tenth ribs, while the second 'hybrid' device spans between the second rib and the L2 vertebral body. The Cobb angle (T3–T12) is decreased to 38° .

whereas in the group in whom the VEPTR was initially inserted after 6.0 years of age ($n = 7$), the average increase was only $6.5 \pm 5.5\%/ \text{year}$ ($p < 0.01$).

Respiratory system compliance (C_{rs}), expressed as a unit per body weight (specific compliance) was severely decreased ($C_{rs} < 1 \text{ ml}/\text{cmH}_2\text{O}/\text{kg}$) in 10 children in the first test. During the course of serial expansion thoracoplasties, specific compliance decreased further in most patients. In the most recent tests, the average C_{rs} had decreased to $56.0 \pm 22.0\%$ of the value in the initial study ($p < 0.01$) (Table 1).

Outcome of patients with respiratory failure

Six tracheostomized patients needed supplemental oxygen with continual or nocturnal mechanical ventilation prior to VEPTR

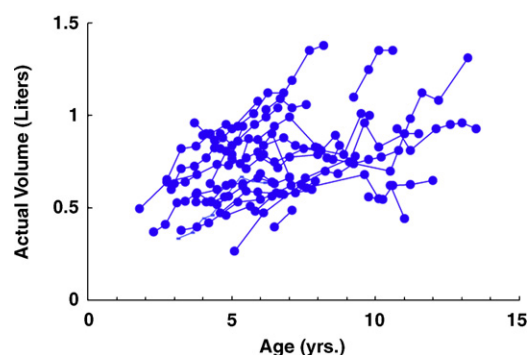


Figure 3. Longitudinal changes in forced vital capacity (FVC) in actual volume with serial VEPTR expansion thoracoplasties in 24 children with TIS versus patient age at each test. There was a significant increase in FVC over time ($p < 0.001$).

Table 1
Longitudinal changes in respiratory function

	FVC (% increase/year)	FVC (% of predicted)	C _{rs} (ml/cmH ₂ O/kg)
Initial	100	71.9 ± 21.6	1.19 ± 0.51
Last		66.3 ± 16.2	0.65 ± 0.30
% change	+11.1 ± 9.86	−7.25 ± 2.4	−44.0 ± 22.0
p	<0.001	<0.05	<0.001

FVC, forced vital capacity; C_{rs}, respiratory system compliance.

expansion thoracoplasty. They have shown different degrees of improvements. Two children were completely weaned off mechanical ventilation and nocturnal oxygen insufflations. Of the remaining four children, three were being weaned off oxygen and remained only on night-time CPAP (BiPAP). Only one patient still required continuous mechanical ventilation, although the mother reported that the child could stay off the ventilator longer, appeared more comfortable and had better oxygen saturation.

DISCUSSION

Growth and development of the spine, thorax and lungs in early childhood

A condition of thoracic wall abnormalities with progressive early-onset scoliosis in infants and young children is a challenging disease both medically and surgically. If untreated, the progression of thoracic deformity would impede the growth of thoracic and lung volumes and would compromise respiratory function and pulmonary gas exchange, leading to respiratory insufficiency, known as TIS.⁴ More than in any other age group, spinal fusion in young children – which until recently was the primary surgical treatment of choice for intervening in the progression of scoliosis – would result in a short trunk and stunted growth of the thorax and lungs.¹² For a better understanding of the effect of defects in the thorax and progressive scoliosis on the development of the lung and pulmonary function, it is essential to know the normal development and growth patterns of the thorax, spine and lungs during infancy and early childhood.

The pulmonary alveoli derive from the primitive terminal air sacs (or saccules) relatively late in gestation (32 weeks) and are not uniformly present until 36 weeks' gestation.¹³ At full-term birth, the number of terminal air spaces (most of which are saccules, rather than alveoli) is estimated to be between 20 and 50 millions. Alveolar formation accelerates after birth and by 18 months of age is complete, the number having increased 10-fold to 400–500 millions,¹³ along with several additional airway branchings (generations) of around 23 of the mature lung before terminating at the alveoli.¹⁴

The thoracic wall during early infancy is extremely compliant and consists of the cartilaginous rib cage and poorly developed respiratory muscles. Sustained tension of the inspiratory muscles keep the thorax rigid enough to prevent reductions in FRC when awake. Relaxation of the inspiratory muscles during rapid eye movement (REM) sleep decreases FRC significantly. It is important to note that both general anaesthesia and muscle relaxants abolish thoracic muscle tone, resulting in marked reductions in FRC, significant airway closure and atelectasis.^{9,15} By 9–12 months of age, the development of the thoracic muscles makes the chest wall more rigid and FRC becomes increasingly stable, while the structural development of lung parenchyma with increasing elastic fibres and collagens continues at least for several more years.¹⁶ By 5 years of age, both FRC and vital capacity (VC) have increased 10-fold from the newborn period.⁹

The rapid development and growth of the lung parenchyma is paralleled by the corresponding growth of the rib cage and spine or vice versa. Two-thirds of an adult's sitting height is achieved by 5 years of age.¹² Correspondingly, the growth velocity of the T1–L5 segments of the spine is greatest between birth and 5 years of age (1.4 cm/year). This period is followed by a marked deceleration of growth velocity between 5 and 10 years of age (0.6 cm/year), after which growth again picks up but at a slower pace toward the teenage years (1.2 cm/year).¹² Overall, the length of the thoracic spine doubles between full-term birth and skeletal maturity.^{1,12} Thoracic volume at birth is about 6% of adult volume, increasing to about 30% by 5 years and 50% at 10 years. Final thoracic volume is achieved by 15 years of age in both males and females.¹² These percentages roughly correspond with those for lung volumes.¹⁷

Surgical approaches: a historical review

The surgical treatment of children younger than 5 years of age with early-onset scoliosis has been challenging. The progressive curvature of scoliosis and thoracic deformity, if left untreated, would significantly affect the development and growth of the lungs and impede pulmonary function, and may result in hypoventilation, hypoxaemia and eventually pulmonary hypertension and cor pulmonale in some patients.⁴ Spinal fusion, the standard treatment option for scoliosis in adolescents, has limited use in much younger children because of its adverse effects on the development and growth of the spine, thorax and lungs.¹

Instrumentation without arthrodesis has been used in an attempt to prevent the progression of scoliosis, by placing a single distraction rod on the concave side of the spine connected to hooks on the upper and lower vertebrae.¹⁸ Although longitudinal results were not reported, Harrington believed that children younger than 10 years of age can be managed with instrumentation alone, whereas older children should have spinal fusion combined with the rod.¹⁸

Using Harrington rods with orthotic support from the Milwaukee brace postoperatively, Moe et al¹⁹ reported the correction of curve progression and an average increase of 2.9 cm in the length of the instrumented spinal segments (as compared with a predicted growth of 4.5 cm) in all 20 patients studied, with notable decreases in the magnitude of the Cobb angle²⁰ in some of these patients. Complications, however, were high (50%), including hook dislodgement from the rod and dislocation from the lamina and rod breakage. In a more recent report of serial spinal instrumentations in young children with the Moe technique, but who eventually underwent spinal arthrodesis, the Cobb angle progression improved or arrested over the course of treatment in approximately two-thirds of the patients; in the remaining patients, including those with neuromuscular diseases, curve progression continued by an average of 33%. The overall growth rate of instrumented but not fused spinal segments was 0.8 mm/segment/year.²¹

In 1977, Luque and Cardoso developed the technique of segmental spinal instrumentation (SSI) using the Harrington rod without arthrodesis.²² Luque subsequently replaced this rod with an L-shaped rod, known as the Luque trolley, and reported a growth of immobilized segments of 4.6 cm, with 78% curve correction in 47 children with paralytic scoliosis.²³ Follow-up studies of the Luque trolley technique by others, however, demonstrated that the subperiosteal exposure and sublaminar passage of wire caused scarring and weakening of the lamina, which made revision and later spinal fusion more difficult;²⁴ and the growth of the instrumented spinal segments was far less than was originally reported.²⁵ More recently, Blakemore et al reported

periodic lengthening with a submuscular rod with limited apical fusion in 29 young children with severe scoliosis (Cobb angle $>70^\circ$) with the application of the Milwaukee brace postoperatively.²⁶ There was an initial improvement in the spinal curvature with reduction of the Cobb angle from an average of 66° to 38° , but subsequent slight deteriorations. The growth of the spinal segments involved was not reported.¹

Akbarnia and Marks have developed a technique of dual expandable rods. The rods are placed on both sides of the spinal column and hooks are placed on both upper and lower anchoring sites over two to three spinal levels, and the transverse rod connectors secure the two rods in place. Serial lengthening of the rods is done approximately every 6 months.^{27,28} The results from the multicentre study of this device are encouraging. The average age at initial surgery was 5 years and 5 months and the mean Cobb angle improved from 82° to 38° following the initial surgery, and was 34° after follow-up at 2–9 years. The average growth of the T1–S1 segments was 1.2 cm/year. The space available for the concave side of the lung, as compared to the convex side, increased from 87% to 100%.²⁸ Lung volumes, however, were not measured.

While all of the above techniques are aimed at preventing and correcting the spinal curvature, Campbell et al were the first group to focus attention on the interrelationship of the spine, thoracic wall and lungs with growth in young children with progressive scoliosis, chest wall defects and fused ribs, and resultant respiratory insufficiency, which the authors termed TIS.^{4,5} As described above, they have developed the technique of serial expansion thoracoplasties using VEPTTR to control the progression of both spine and chest wall deformities. Reports of the favourable outcome of this procedure have recently emerged in the literature.^{10,29}

Estimates of thoracic volumes and lung function

Thoracic volume can be estimated from posteroanterior and lateral chest radiographs,³⁰ posteroanterior chest radiographs alone³¹ or, more recently, CT scans. Schlesinger et al demonstrated an excellent correlation ($r > 0.90$) between the radiological estimates and the direct measurements of total lung capacity (TLC) by means of a body plethysmograph in children, provided that the radiographic imaging was consistently done at maximum inspiration, i.e. at TLC.³² Gollogly et al obtained the total parenchymal lung volume by determining the volume of the left and right lungs separately. By adding these up from normal CT scans in 1050 children using the technique of Schlesinger et al,^{32,33} they constructed lung volume versus age scattergrams for boys and girls from birth to 18 years of age, to be used as a reference for the study of the surgical correction of children with spondylothoracic deformities and TIS. Unfortunately, these retrospective measurements were performed without controlling for the lung volume and sedation, which would also significantly affect FRC.^{9,15} Reports on studies in children with TIS using radiological techniques have been scarce.^{2,34} Ramirez et al also reported single standard PFT results in a small number of adults and children older than 7 years of age who showed severe restrictive defect (FVC 17–51% predicted).²

Direct measurements of pulmonary function in children under general anaesthesia

Studies of direct measurements of lung volumes in children with TIS are limited and longitudinal assessments of serial VEPTTR expansions are lacking, because of patients' inability to perform standard PFTs due to their young age as well as physical or mental

handicaps. As stated above, we have developed the instrumentation and technique for performing complete PFTs in anaesthetized and paralyzed children of all ages, regardless of their physical and mental status and maturity. This technique allowed us to study FVC, C_{rs} and MEFV curves, with which highly accurate assessments of lower airway obstruction can be made, independent of events in upper and large intrathoracic airways.^{6,7,17} Using this technique we performed serial PFTs in children with TIS to examine the efficacy of serial expansion thoracoplasty.

Based on our preliminary report involving 10 children with serial VEPTTR thoracoplasties, we found the following:

1. VEPTTR expansion thoracoplasty is a safe procedure with extremely low morbidity and no related mortality.
2. At the first PFT, most patients demonstrated moderate-to-severe restrictive lung defect, as evidenced by abnormally decreased FVC, below the predicted values.
3. With serial VEPTTR thoracic expansions, FVC continued to increase in absolute terms. In the majority of patients, the average increase in FVC appeared to keep up with body growth, since FVC, expressed as a percentage of predicted values at the last test, did not differ significantly from the first test during an average time span of nearly 2 years. Most children and their parents reported improvements in respiratory function as well as quality of life (two children were weaned from the ventilator or CPAP, while another was weaned from supplemental oxygen). It was, however, difficult to assess the extent of improvement, since there was no control group for comparison, other than historical cases of deterioration without surgery.
4. In the majority of patients studied, compliance (C_{rs}) of the respiratory system (lungs and thorax), adjusted for body size (i.e. specific compliance), was moderately to severely reduced, indicating that the stiff respiratory system did not change significantly over the duration of the study.
5. Based on the pattern of rapid growth velocity of the thoracic spine and postnatal growth in thoracic and lung volumes during the first 5 years of life, as discussed above, the benefit of VEPTTR expansion thoracoplasty should be greatest when the procedure starts earlier in life and definitely before 6 years of age, as Campbell et al have suggested.⁴ Based on the preliminary data in 10 children, however, we did not find a significant difference in the degree of improvements between those whose first VEPTTR implant was made before versus after 6 years of age. A larger database was required for further analysis.

From the expanded database on 24 patients, including over 150 PFTs with longer observation periods (median duration of 4.6 versus 1.9 years), we have now found somewhat different results of the effect of VEPTTR thoracoplasty. Again, the incidence of complications was infrequent: of 151 VEPTTR thoracoplasties, there were 12 dislocations or dislodgement of the VEPTTR (nine dislocations on the rib and three on the spine), one localized infection and one skin erosion, which were resolved without sequelae.

We have confirmed our previous findings that the lung volume (FVC) does increase with body growth in absolute terms. However, the extent of increase in FVC is limited; lung growth did not keep up with the rate of body growth, as evidenced by falling % predicted values of FVC with time (see Table 1).

As for the efficacy of VEPTTR vis-à-vis the age of children at the first surgical intervention, we now found a clear-cut difference between the younger and older children. Surgical intervention was much more effective when it was initiated before 6 years of age. The annualized percentage increase in FVC before 6 years of age was more than twice that of the older age group.

Another significant finding, and the one which concerns us most, is the deterioration of respiratory system compliance (C_{rs}) over time. In spite of general increases in FVC, C_{rs} , even in absolute terms, did not increase or even change in most of the patients studied. It is important to note that C_{rs} adjusted for body size markedly and significantly decreased over time. This finding may well become a potential limitation to expansion thoracoplasty with VEPTR. Further longitudinal study is needed to confirm these findings.

CONCLUSION

The effect of surgical intervention in children with thoracic and spinal malformation and progressive early-onset scoliosis has been reviewed, with emphasis on the effect of serial expansion thoracoplasty with VEPTR and its longitudinal effect on lung volume and respiratory mechanics, which were studied under general anaesthesia at the time of surgery using a specially constructed mobile PFT device. As expected from clinical impressions and indirect measurements, most children with TIS of varying aetiologies with or without fused or defective ribs and thorax demonstrated moderate-to-severe restrictive lung defects with a very stiff respiratory system, primarily due to restriction by the thoracic wall. In general, the technique of serial VEPTR expansion thoracoplasty carries relatively low surgical risks, and results in clinical improvements and significant increases in lung volume in most patients, as evidenced by increases in FVC over time. The effect of VEPTR thoracoplasty appears to be far better when the initial surgical intervention is before 6 years of age. The rate of lung growth with expansion thoracoplasty, however, appears to be less than that based on body growth. Decreasing respiratory system compliance over time, an indication of increasing stiffness of the chest wall with growth, is a major concern for the long-term effect of surgical intervention.

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