The effect of a vertical expandable prosthetic titanium rib on shoulder balance in patients with congenital scoliosis

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Abstract

Purpose To evaluate the effect of a vertical expandable prosthetic titanium rib (VEPTR) on shoulder balance in patients with congenital scoliosis.

Methods Fifteen patients had a thoracic congenital scoliosis. The Cobb angles of the thoracic primary curves were measured. Preoperative measurements of the coracoid height difference (CHD; expressed in millimeters) and the clavicular tilt angle difference (CTAD; expressed in degrees) were performed for all patients. All of the patients were treated with VEPTR open wedge thoracostomy.

Result There was a statistically significant improvement in thoracic Cobb angle. At the end of follow-up there was a significant improvement in CHD; all but three of the patients had CHD < 9 mm. Preoperatively, CTAD was 8.9°. It improved to 4.4° postoperatively and significantly decreased to 3.9° by the end of follow-up. Complications included device migration (one patient), infection (one patient), and pedicle screw loosening or displacement (two patients).

Conclusion VEPTR is able to produce a modest intra-operative correction of shoulder imbalance.

Keywords Congenital scoliosis · VEPTR ·
Shoulder balance · Thoracic insufficiency syndrome

Introduction

The prevalence rate of congenital scoliosis is thought to be approximately 1 in 1,000 live births [1]. Vertebral

anomalies causing congenital scoliosis may be caused by a failure of formation, by failure of segmentation, or by a combination of these two factors, resulting in a mixed deformity [2]. Congenital malformations of the ribs and chest wall, as well as anomalies of the scapulae, are often found in association with congenital deformities of the spine. Campbell et al. [3] stated that in cases of congenital scoliosis with extensive rib fusion in growing children, the concave hemithorax can act as a powerful lateral tether to further unbalance the growth of the spine, which is already being deformed by asymmetrical vertebral growth.

The surgical concept of expansion thoracoplasty and stabilization with a vertical expandable prosthetic titanium rib (VEPTR) implant is based on the expansion of the thorax by rib distraction on the concave side of the curve, resulting in the indirect correction of the curve [3–5]. Spinal fusion may be necessary once skeletal maturity is reached and thoracic spinal growth is no longer an issue. However, to date and to our knowledge, the effect of VEPTR on shoulder balance has not been evaluated.

Many radiographic parameters have been used to assess shoulder balance. Coracoid height difference (CHD) measures the height difference between the coracoid processes by tracing a horizontal line at the upper margin of each and measuring the difference in height between these lines [6]. CHD has the highest correlation with clinical shoulder balance [7]. It has been suggested that thoracic 1 tilt (T1 tilt; the angle between the horizontal line and the line through the upper end plate of T1) is mildly correlated with shoulder balance [6, 8]. The clavicular tilt angle difference (CTAD) is a new radiological parameter that has been created in order to assess shoulder balance. It is the difference between the angles formed by the bisection of the lines of the proximal portions of the clavicle with the horizontal [7] (Fig. 1).

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Fig. 1 The clavicular tilt angle difference (CTAD) is the difference between the angles formed by the bisection of the lines of the proximal portions of the clavicle with the horizontal

Materials and methods

Fifteen patients had a right-side congenital scoliosis (thoracic in nine patients and thoracolumbar in six patients) associated with thoracic insufficiency syndrome. The patients (11 girls and 4 boys) had a mean age of 6.4 years (range 5.3–8.4 years) at the time of surgery. Eleven patients had fused ribs and the cause of congenital scoliosis was either unsegmented bar or congenital thoracic hemi-vertebra. They were accepted for surgery only after consulting a multidisciplinary team consisting of pediatric orthopedic surgeons, a pediatric surgeon, a pediatric pulmonologist, a pediatric anesthesiologist, and the pediatric intensive care team. The common denominator for all patients was the small stiff thorax that could not provide any volume that would allow the growth of the lungs, along with minimal secondary breathing mechanisms due to fused or malformed ribs. All of the parents had observed that the right shoulder was apparently more elevated than the left one. Protocol studies included preoperative standing anteroposterior (AP), including both shoulders and lateral radiographs of the entire spine; AP bending films of the spine; and cervical spine films. The patients were asked to stand up straight with their arms by their sides against a vertical bucky; the median sagittal plane should be at right angles to the film and coincident with the midline of the bucky, while the legs should be fully extended and the feet should be slightly apart in the true anatomical plane. A preoperative three-dimensional CT scan was done to assess the osseous vertebral anomalies. A preoperative MRI of the spinal cord was performed to rule out spinal cord abnormality. One patient had untethering of the cord before VEPTR surgery. The Cobb angles of the thoracic primary curves were measured. Preoperative measurements of CHD

(expressed in millimeters) and CTAD (expressed in degrees) were performed for all patients. Patients who had CHD values of >9 mm and CTAD values $>4.5^\circ$ were considered to have clinical shoulder imbalances of >10 mm [7].

Surgical procedure

All of the procedures were performed as described by Campbell et al. [9] with some modifications. The VEPTR was inserted in a prone position. A thoracostomy incision was made between the medial border of the scapula and the midline spine proximally, and was gently curved anteriorly in a J shape 4 cm distal to the tip of the scapula. The number and locations of expansion thoracostomies employed were dictated by the location of the hemithoracic constriction, the bending films, the location of the fused ribs, and secondarily by the location of the spinal deformity or anomaly. Where possible, the thoracostomies were planned so as to divide fused ribs (osteotomy in the potential interval between the fused ribs) but not to isolate groups of fewer than two or three ribs for fear of devitalizing the intervening rib segments. One to two thoracostomies were performed from the interval between the rib heads to the costochondral junction anteriorly. Once the proximal ribs above the open wedge thoracostomy were oriented transversely, there was usually adequate lengthening of the constricted hemithorax with indirect correction of the scoliosis. The upper and lower rib attachments for the VEPTR device were prepared extrapleurally around the ribs, taking care not to place the device above the second rib so that the brachial plexus was protected. For the rib to spine hybrid construct, our modified technique included the use of a pedicle screw (USS system) instead of a laminar hook (this modification was used in eight patients, two of them after the failure of the distal laminar hook during follow-up). The pedicle screw was inserted under image guidance through a 2 cm incision lateral and parallel to the midline of the lumbar spine over the chosen lumbar pedicle. A rib to spine VEPTR construct was applied in eleven patients, and double constructs (rib to rib and rib to spine) were applied in four patients. All of the patients underwent postoperative pediatric intensive care monitoring and respiratory therapy. Standing postoperative X-rays and measurements were taken one week after surgery, as there was no significant pain that could affect the global posture of the patient.

All of the patients were regularly followed up in the outpatient clinic every six months. Routine device expansion was performed in outpatient surgery every six months and the completely expanded VEPTR sleeve was replaced when necessary. The mean follow-up time was 28.9 months (range 24–34 months). A standing

anteroposterior including both shoulders and a lateral X-ray of the entire spine were done at each visit. All digital X-ray images were stored in a Picture Archiving and Communication System (PACS) workstation. All digital radiographs were viewed and measured by the first author using PACS software.

For statistical analysis, a two-tailed Student's *t*-test was performed, where significance was defined as $P < 0.05$.

Results

Clinically, all of the families reported an improvement in the appearance of the shoulders and an increase in the time spent playing and performing sporting activities.

The average primary thoracic curve was 49.8° (range $25\text{--}75^\circ$) before surgery. It decreased to an average of 35.6° (range $15\text{--}55^\circ$) immediately after surgery, and was an average of 30.2° (range $15\text{--}55^\circ$) at final follow-up, with a significant average correction of 19.6° ($P < 0.05$) at the final follow-up (Table 1). CHD averaged 21 mm (range 12–43 mm) before surgery, 11.2 mm postoperatively (range 2–20 mm), and was significantly decreased ($P < 0.05$) to 7 mm (range 2–18 mm) at final follow-up. CTAD averaged 8.9° (range $5\text{--}36^\circ$) before surgery and decreased to 4.4° ($P < 0.05$) (range $2\text{--}6^\circ$) postoperatively. At final follow-up, it had significantly decreased to 3.9° (range $1\text{--}14^\circ$) (Table 2; Figs. 2, 3).

Table 1 Thoracic Cobb angle

Patient number	Preoperative thoracic Cobb angle	Postoperative thoracic Cobb angle	Thoracic Cobb angle at final follow-up
1	25	20	17
2	60	37	52
3	36	23	19
4	37	22	32
5	28	15	14
6	54	40	40
7	41	33	25
8	66	46	30
9	58	39	30
10	75	55	41
11	66	53	44
12	67	52	33
13	44	32	27
14	47	36	25
15	43	32	24
Mean	49.8	35.6	30.2*

* Significant ($P < 0.05$)

Complications in our study were mainly related to the distal fixation point in the rib to spine VEPTR construct. Two patients had dislodgement of the laminar hook, which had been observed during follow-up (replacements were done with pedicle screws). Problems with pedicle screws had been reported in two patients. One of them had radiological loosening and a positional change. The second patient had complete displacement of the screw; revision was done with the insertion of two pedicle screws (Fig. 4). None of the patients had any neurological complications. One patient had dislodgement of the superior cradle, which had been reinserted during lengthening. One patient had a wound infection and was treated with debridement and i.v. antibiotics.

Discussion

Specific vertebral anomalies related to congenital scoliosis tend to lead to a malignant curve progression. Of particular concern is the presence of an unsegmented bar, which is associated with severe progression of spinal deformity and has an ominous prognosis if left untreated. Hemivertebral anomalies tend to have a slower progression initially but tend to peak during adolescence [10, 11].

Shoulder balance evaluations are important for spine surgeons that need to make surgical decisions about patients with idiopathic scoliosis. The problem of shoulder imbalance in congenital scoliosis has not gained much attention, probably due to the difficult nature of the deformity, the different lines of treatment [2] (in situ fusion, hemivertebral excision, convex hemiepiphysiodesis), and the greater concern over spine growth.

CHD shows the highest correlation with clinical shoulder balance, the highest intraobserver reliability, and exhibits high interobserver reliability [7]. It is the method of choice for assessing radiological shoulder balance; differences of more than 9 mm correlate with shoulder imbalances of >10 mm [7]. In our study, all of the patients had preoperative a CHD of >9 mm; postoperatively there was significant improvement in CHD, but most of the patients still had CHD >9 mm. At the end of the follow-up period there was significant improvement in CHD; all of the patients had CHD <9 mm except in three cases (10, 15 and 18 mm).

CTAD was recently created as a new parameter for radiological shoulder balance assessment. It has the additional advantage of being the method of choice when the shoulders are not included in the X-ray [7]. CTAD $> 4.5^\circ$ indicates a clinical shoulder imbalance of >10 mm [7]. In our study, the preoperative mean of CTAD was 8.9° , which improved to 4.4° postoperatively. It significantly decreased to 3.9° by the end of the follow-up period. In our study, we

Table 2 Coracoid height difference (CHD) and clavicular tilt angle difference (CTAD)

Patient N	Preoperative CHD	Postoperative CHD	CHD at final follow-up	Preoperative CTAD	Postoperative CTAD	CTAD at final follow-up
1	13	12	2	5	2	2
2	43	9	15	36	5	14
3	12	2	2	5	2	2
4	15	12	10	7	4	1
5	22	2	2	6	3	3
6	30	20	18	8	4	3
7	16	8	4	7	4	3
8	24	13	8	9	5	4
9	18	13	7	7	6	5
10	20	14	9	9	5	4
11	24	12	5	9	6	5
12	19	13	8	8	6	4
13	23	14	6	6	5	3
14	18	12	4	6	5	3
15	19	13	5	6	5	3
Mean	21	11.2	7*	8.9	4.4	3.9*

* Significant ($P < 0.05$)

Fig. 2 **A** Right congenital thoracolumbar scoliosis; the preoperative Cobb angle was 36°, CHD was 12 mm, and CTAD was 5°. **B** At the end of the follow-up period, the Cobb angle was 23°, CHD was 2 mm, and CTAD was 2°

did not use other radiological parameters for shoulder balance assessment such as first rib angle, clavicle–rib cage intersection, or first rib clavicle height because of rib anomalies or rib fusion that are frequently found in congenital scoliosis.

In our study there was significant correction of the preoperative Cobb angle from 49.8° to 30.2° at the end of the follow-up period. The effect of VEPTR in gradual restoring shoulder balance in our study was mainly attributed to the gradual correction of the Cobb angle. That

secondary effect of the VEPTR in maintaining shoulder balance may help when fusion is undertaken after skeletal maturity.

Hell et al. [12] reported three complications encompassing 15 primary VEPTR implantations. These included rib fracture, lumbar hook displacement, and skin breakdown. Emans et al. [13] reported 17 complications over the course of 31 primary expansion thoracostomies with VEPTR implantations. The complications in their study included rib fracture, device migration, deep wound

Fig. 3 **A** Right congenital thoracic scoliosis; the preoperative Cobb angle was 25°, CHD was 13 mm, and CTAD was 5°. **B** At the end of the follow-up period, the Cobb angle was 17°, CHD was 2 mm, and CTAD was 2°

Fig. 4 **A** Displacement of the pedicle screw in a rib to spine VEPTR construct. **B** Revision with distal pedicle screws

infection, rib refusion, and two instances of brachial plexus palsy with recovery. In our study, one patient had dislodgement of the superior cradle that had been reinserted during lengthening. One patient had a wound infection, and was treated with debridement and i.v. antibiotics. Dislodgement of the distal laminar hook was reported in two patients, both of whom had revisions with pedicle screws instead. Our modification in replacing the distal laminar hook with a pedicle screw did not completely solve the problems related to the distal fixation point in the rib to

spine VEPTR construct. Two patients out of eight had displacement of the pedicle screw; revision was done with two distal screws in one of these patients.

In this series of cases of thoracic congenital scoliosis, in addition to improving the measured Cobb angle, VEPTR with expansion thoracostomy provided satisfactory restoration of shoulder balance. The improvement in shoulder balance occurred not just at the initial operation, but also over time with subsequent VEPTR lengthening.

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