The effect of vertical expandable prosthetic titanium rib on growth in congenital scoliosis

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Abstract

Aims: In the treatment of thoracic insufficiency syndrome, the main aim is to maintain spinal and thoracic growth in order to continue respiratory functions. Vertical expandable prosthetic titanium rib (VEPTR) device application is a method of choice especially in the congenital cases with a thoracic deformity. In our study, we evaluated the effect of VEPTR on growth in congenital scoliosis.

Materials and Methods: Four female patients in whom VEPTR was applied were retrospectively evaluated. Anteroposterior (AP) and lateral Cobb angles that were measured preoperatively and during the last control, space available for lung (SAL), T1-S1 and T1-T12 distances, coronal and sagittal balances were compared.

Results: Four female patients in whom VEPTR was applied were retrospectively evaluated. AP and lateral Cobb angles that were measured preoperatively and during the last control, SAL, T1-S1, and T1-T12 distances, coronal and sagittal balances were compared.

Conclusions: VEPTR may provide a good correction, and we observed a growth in the spine height and SAL following the treatment of congenital deformities. Long-term, multicenter, prospective studies that compare the spinal height, respiratory functions, the severity of the deformity, and the spinal balance are required in order to evaluate the efficacy of VEPTR.

Key words: Congenital scoliosis, rib deformity, thoracic insufficiency syndrome, vertical expandable prosthetic titanium ribs

INTRODUCTION

The treatment algorithm of early onset scoliosis (EOS) is not clearly standardized due to the wide spectrum of etiologies. Although previously spinal fusion was recommended in the treatment of progressive spinal deformities in the cases with early ages, recently early spinal fusion is reported as a last choice of treatment due to the negative effects on thorax and the respiratory system.[1,2] Furthermore, an increase of morbidity and decrease of life expectancy are observed in the cases that are not treated.[3] The treatment options described for the treatment
of EOS are observation, bracing, casting, rib-based distraction instrumentation-vertical expandable prosthetic titanium rib (VEPTR; Synthes Spine Co, West Chester, PA), spine-based distraction instrumentation (growing rods), growth guidance (Shilla technique and Luque-Trolley), growth modulation (anterior vertebral body stapling), and spinal fusion.[4-8] There is a variety of implant options and choice of instrumentation levels.[9]

VEPTR technique was first described by Campbell et al. as a nonfusion, growth-friendly implant applied with or without expansion thoracostomy in the patients with severe EOS or thoracic insufficiency syndrome (TIS). Congenital idiopathic or EOS associated with a constrictive chest wall deformity is addressed by VEPTR.[10] The surgical techniques for the application of VEPTR have been described in detail so far.[4,12-14] At the beginning, it was developed to maximize the chest wall expansion, and it was suggested in the treatment of severe congenital spinal deformities and thoracic deformities with secondary TIS due to rib fusions in concert with congenital scoliosis.[15] VEPTR expansion thoracoplasty with an opening wedge thoracostomy may be required in the patients that volume depletion deformity is progressed.[16-19] VEPTR may be applied in patients aged from 6 months and up to skeletal maturity.[11] VEPTR treatment has a complication rate of 9-21% and the complications are rib fractures, proximal migration of the device, repetitive surgical interventions, and wound problems.[8,11]

The purpose of our study was to evaluate the overall results of the VEPTR treatment on the growing spine; and the midterm results of the curve correction, thoracic height, space available for the lung (SAL), coronal and sagittal balance, in the patients with congenital scoliosis accompanied by a thoracic deformity.

### MATERIALS AND METHODS

VEPTR was applied in 4 patients with EOS and severe thorax deformity in the years 2009 and 2010. All of the patients were evaluated with anteroposterior (AP) and lateral radiographies of the spinal column in the pre- and post-operative periods. Cobb’s angle in both planes, sagittal and coronal balance (C7-plumb line), SAL, and spinal height (T1-T12 and T1-S1 distances) were measured.[4,10,20-23] The whole spinal column was evaluated with three-dimensional (3D) reconstructed computed tomography (CT) and magnetic resonance imaging in the preoperative period. All of the patients were classified according ‘classification of EOS (C-EOS)” described by Vitale et al. and Williams et al.[24,25] Two patients were classified as C4N and the others were classified as C3N. Congenital scoliosis types were evaluated as 3 failures of segmentation cases and 1 failure of formation [Table 1].

In the surgical treatment, the collapsed thoracic region was distracted in the concave side of the deformity. Opening wedge thoracostomy was performed in 2 patients with severe rib fusions. VEPTR was applied on the ribs proximally and was applied on the ribs, the spinal column or pelvis distally. In all patients, VEPTR was applied unilaterally. Expansions were performed with a mean interval period of 8.4 months (range: 7.3-9.2). The surgical procedures performed, the complications, the number, and the amount of the lengthening were noted [Table 2].

### RESULTS

All patients were female. The mean age was 53 months (range: 20-79) at the time of the surgery. The mean follow-up period between the first surgery and the last control was 5.8 years.
The mean implantation period was 5.2 years (range: 3.4-6), the mean age in the last control was 10 years (7-12). The mean AP Cobb’s angle of the major curves was 79.5° (range: 62-98°) preoperatively and was 57.5° (range: 32-79°) during the last control. The mean correction was found as 27.7%. In the sagittal plane, the mean thoracic kyphosis angle was measured as 28.5° (range: 16-42) preoperatively, and 44.2° (range: 22-60°) during the last control, the mean lordosis angle was measured as 42.2° (range: 36-53°) preoperatively, and 45.7° (range: 30-53°) during the last control. An increase in both kyphosis and lordosis was detected. The mean SAL was 7.4 (range: 4.8-9.8) cm in the concave side and 9.1 (range: 7-12) cm in the convex side preoperatively. During the last control, it was measured as 12.4 cm (range: 9.3-13.6) in the concave side and 15 cm (range: 13.6-16.5) in the convex side. The mean increases were 67.5% in the concave side and 64.8% in the convex side. Respiratory functions were not evaluated in the preoperative period. The mean force vital capacity (FVC) was measured as 59% (range: 45-70) and the mean forced expiratory volume in 1 s (FEV1) was measured as 62.25% (range: 48-73), during the last control. At this time, the respiratory pattern was restrictive in 3 patients; the other patient could not be evaluated in term of respiratory pattern. The mean T1-T12 and T1-S1 distances were measured as 11.5 (range: 8.8-14) cm and 21 (range: 15.4-23) cm preoperatively. During the last control, these parameters were measured as 17.5 cm (range: 14-20) and 31.5 cm (range: 27.5-33.7), respectively. The total mean length was 6 cm (T1-T2) (52.1%) and 10.5 cm (T1-S1) (50%), and the annual length observed was 1 cm (T1-T2) and 1.8 cm (T1-S1). In the coronal plane, the C7 plumb line was (+) in all of the patients preoperatively, and neutral coronal balance was obtained in 2 of the patients postoperatively. In the sagittal plane, the C7-plumbline was (+) in 2 patients, (−) in 1 patient, and neutral in 1 patient preoperatively. After the treatment, neutral sagittal balance was obtained in 4 patients. The mean number of the surgical procedures in the patients was 8 (range: 7-9). The mean number of the expansions was 6.5 (range: 6-7) and the mean number of the unforeseen surgical procedures was 1.5 (range: 1-2). The mean number of the surgical procedures was 1.6/year. The complications were migration of the device in 3 cases, wound problems in 2 cases and rod fracture in 1 case [Table 2].

In the first patient, after 3.4 years following 9 surgical interventions, due to autofusion, VEPTR has lost its effect on the deformity for lengthening and correction, and it was removed. The total follow-up period was 5.8 years, and surgery is planned to correct the remaining deformity. In the second patient, although she was a walking child, we have performed rib-to-rib VEPTR and rib to pelvis iliac hook (Dunn - McCarthy s-hook) due to distal fixation problems. As rib-to-rib expansion was made 2.7 years later, the hook between the rib and pelvis was replaced with a single growing rod. The rod was removed and replaced with a magnetically controlled growing rod (MCGR; MAGEC Ellipse Technology, Irvine, CA.) The total follow-up period of this patient was 5.4 years [Figure 1].

| Table 2: Preoperative and postoperative values of the evaluation criteria |
|---|---|---|---|---|---|---|---|---|
| **Initial surgery** (months) | **Follow-up** (years) | **Implant duration** (years) | **Last control** (years) | **Number of complication interval** (months) | **Average lengthening interval** (months) | **Number of lengthening** | **Average lengthening interval** (months) | **Number of complication** |
| 45 | 5.8 | 5.4 | 5.4 | 6.6 | 6.0 | 6.0 | 6.0 | 6.0 |
| 20 | 5.4 | 6.6 | 6.6 | 6.0 | 7.0 | 7.0 | 7.0 | 7.0 |
| 68 | 5.0 | 5.0 | 5.0 | 5.0 | 5.0 | 5.0 | 5.0 | 5.0 |
| 79 | 6.0 | 6.0 | 6.0 | 6.0 | 6.0 | 6.0 | 6.0 | 6.0 |

TK: Thoracic kyphosis, LL: Lumbar lordosis, SAL: Space available for lung, FVC: Force vital capacity, FEV1: Forced expiratory volume in 1 s, AP: Anteroposterior
third patient, VEPTR was applied due to congenital scoliosis and thorax deformity, and the total follow-up period was 6 years. The device was regularly expanded. VEPTR was removed also in the last patient. This patient was the oldest one in the group. After 2.9 years following the first procedure, maximum spinal growth was obtained, and expansion was effectless anymore. Hence, the device was removed. Selective thoracic posterior instrumentation and fusion were performed. The total follow-up period was 6 years. Opening wedge thoracostomy was repeated in the 2 patients with TIS, due to fibrosis that developed between the ribs during the expansions. The mean time between the expansions was 8.4 months (range: 7.3-9.2). Intraoperative neurophysiological monitoring was used only in the patient having MCGR together with congenital bar excision.

DISCUSSION

There is a variety of surgical options in the treatment of EOS. Surgical treatment has some disadvantages such as technical difficulties, heterogeneous patient population (Congenital, idiopathic, neuromuscular, and syndromic) and unapproved, off-label application of the implants. As described in the original technique by Campbell and Hell-Vocke after standard thoracotomy incision in the posterior thorax, fibrous tissues between the fused ribs are released with or without, then VEPTR device is placed (as Rib-Rib, Rib-Spine, or Rib-Pelvis Hybrid). It is attached to proximal ribs in the proximal side and to distal ribs or lumbar vertebrae or iliac crest in the distal side. The device is expanded every 6 months. We also performed thoracotomy in our 2 patients in order to decrease thorax compression.

Lung growth is essentially completed by the age of 8 years with maximum growth occurring before 5 years of age. The best treatment strategy for EOS is to open the thorax before 5 years of age and keep the spine flexible before puberty. As it shortens, the thoracic spine precocious arthrodensis of this segment has negative effects on thoracic growth and lung development. Karol et al., reported the thoracic height under 18 cm in 16 of the 28 patients included in their study. For this group, the mean predicted FVC was 48.2% as an evidence of severe restrictive pulmonary disease, in the same study, in 8 patients, thoracic height was reported to be between 18 and 22 cm, the mean predicted FVC was reported to be 63%, and finally, in 4 of the 28 patients, thoracic height was measured between 22 and 28, these values were considered normal for this age group, and the mean FVC was found as 85.2%. It is shown that the shortness in the thoracic spine leads to a smaller FVC and probably to pulmonary restrictive disease.

Preoperative evaluation of the pulmonary volume and the pulmonary function tests are essential. In young patients, the pulmonary volume can be measured with 3D-CT imaging. In the children in whom VEPTR was applied without the preoperative evaluation of the pulmonary function tests, the increase of the pulmonary volume may be displayed using CT.

In our study, the mean thoracic spinal height was measured as 17.5 cm (range: 14-20) at the end of final control. The thoracic spine height was under 18 cm in 2 patients (14 and 15 cm),
and over 18 cm in 2 patients (19.6 and 20 cm). The increase in the thoracic spine length was approximately 6 cm (T1-T12) and the increase in the thoracic-lumbar spine was 10.5 cm (T1-S1), which was about 1 cm/year (T1-T12) and 1.8 cm/year (T1-S1). We were not able to evaluate pulmonary functional tests preoperatively because of the noncooperation of the children. During the last control, the mean FVC was measured as 59%, the mean FEV1 was measured as 62.25%, and the respiratory pattern was mild-restrictive in the patients except for incompatible one. The spinal height and the pulmonary function test values of the patients were measured under the normal physiological values that was expected to be in 9.5 years which is the mean age during the last control.

Pelvic anchors may be required during VEPTR application. Pelvic hooks (Dunn - McCarthy s-hook) and screws can be used for this purpose. Despite the easy application technique, migration of the pelvic hooks is not rare, and the revisions are difficult. It is especially preferred in nonambulatory children. It is also useful in pelvic obliquity. The distraction of the pelvic hooks causes sagittal imbalance in ambulatory children. The pelvic approach is performed on just lateral of muscle erector spine. Reverse pelvic hook application is preferred in order to shift sagittal axis anteriorly. However, in case of migration of the implant, removing it may be challenging. We also performed pelvic hook application in one of our patients in whom spinal hook application was not possible. When spinal fixation became possible as the patient grew, we replaced the pelvic hook with a single growing rod system using pedicle screws. Later, in the same patient, due to a broken rod, it was changed with MCGR and excision was performed in the unsegmented congenital bar located in the lumbar area. In the other 3 patients, we performed rib-to-rib and rib-to-laminar hook fixation for the distal anchor point. For proximal anchor points, we performed rib-to-rib fixation in all of the patients.

The main complications of VEPTR application can be listed as migration, infection, and brachial plexus paralysis. With the new C-EOS described by Vitale et al., (C-EOS), VEPTR complications, proximal anchor insufficiency, and the patients with high risks are predicted. As a solution, multi-level fixation with 4 hook anchors is suggested. According to the Sankar et al., standard dual growing rods had complication rate of 2.3/patient; hybrid growing rods with rib anchors proximally and spine anchors distally had complications rate of 0.86/patient; and VEPTR had complications rate of 2.37/patient. The complication rate in growing spine surgery is uniformly high but varies by implant type, with a trend toward fewer complications in hybrid constructs. Rigid cranial and caudal constructions may cause the risk of insufficiency. VEPTR becomes less effective with the increased number of lengthenings. The complication rate of VEPTR is defined as 73%, and the neurological deficit is encountered in one case. VEPTR anchor insufficiency is shown often in especially neuromuscular cases, in the cases with high angled curves (51-90°) and in the hyperkyphotic cases. Psychosocial effects of repetitive surgeries performed during VEPTR application are evaluated, and it is found out that psychosocial scores decrease as the number of surgeries increase. The challenges of this treatment, as demonstrated by previous studies, include the demands of multiple procedures, skin problems, and device. We also had to perform unexpected surgical procedures due to complications. Lamina/rib hook migration, lamina/rib fracture, compression of the implant on the skin, insufficient wound closure, deep or superficial infections, and rod fractures were the complications we encountered. We had to perform a mean number of 1.5 (range: 1-2) additional surgical procedures for implant revision or debridement. We also realized that as the number of the lengthening and the follow-up period increased, the efficacy of VEPTR decreased.

Even though severe congenital spine deformity limits the normal spinal and thoracic growth, with VEPTR application, after a follow-up period of about 5.8 years, we obtained a well-balanced spinal column, correction of the deformity, and increase in the spinal height and SAL, in the patients in whom TIS is expected to develop. VEPTR treatment proved to be useful for the expected growth to be realized.

The limitation of our study is a low number of the cases and the lack of the preoperative evaluation of the pulmonary function tests. VEPTR may be a good choice of first line treatment at the beginning when a secure and reliable fixation is not feasible in the spinal column. Long-term, multicenter, prospective studies with more patients that compare the spinal height, respiratory functions, the severity of the deformity, and the spinal balance are required in order to evaluate the efficacy of VEPTR.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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