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VERTICAL EXPANDABLE PROSTHETIC TITANIUM RIB (VEPTR) TECHNIQUE FOR THE TREATMENT OF PROGRESSIVE EARLY ONSET SCOLIOSIS

İLERLEYİCİ ERKEN BAŞLANGIÇLI SKOLYOZLARIN TEDAVİSİNDE VERTİKAL EKSPANDABL PROSTETİK TİTANYUM RİB (VEPTR) TEKNİĞİ

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SUMMARY:

The progression of early onset scoliosis has a negative effect on the visceral organs, respiratory system, and normal spinal growth, and so treatment should be begun as soon as possible. Reports have shown that after early fusion of the spine to treat early onset scoliosis, respiratory function decreases. Spinal fusion also prevents the development of the vertebrae.

Robert Campbell and Melvin Smith have developed expansion thoracostomy and VEPTR instrumentation for thoracic deficiency syndrome associated with congenital scoliosis, rib fusion, and hypoplastic chest wall deformity.

The VEPTR system, a non-fusion technique to correct deformity, is used in children with multiple spinal abnormalities to reduce the spinal deformity, increase the respiratory function, and allow lung growth.

This technique allows growth of the chest cage and the spine by lengthening and expanding the narrowed hemithorax. This technique seems to directly treat thoracic insufficiency syndrome. It also indirectly fixes scoliosis without fusion.

Complications are seen at a high rate, as repeated surgical procedures are required for the use of VEPTR. The patients should have sufficient skin, subcutaneous adipose and muscle tissue for the implementation of the VEPTR system. If the appropriate conditions are not present, complications are likely to be faced when the VEPTR system is applied.

Key words: Early onset scoliosis, VEPTR, indications, surgery technique, complications

Level of evidence: Review article, Level V

ÖZET:

Erken yaşta başlayan skolyozun ilerlemesi, organ ve solunum sistemlerinin gelişimi ile normal spinal büyümeyi kötü yönde etkileyeceği için erken dönemde tedaviye başlanması gerekmektedir. Yapılan çalışmalarda gösterilmiştir ki erken başlangıçlı skolyozların erken dönemde spinal füzyonu sonrası akciğerin fonksiyonları negatif yönde etkilenmektedir. Aynı zamanda spinal füzyon vertebra gelişimini de engellemektedir.

Konjenital skolyoz ve kot füzyonuna, hipoplastik göğüs duvarı deformitesi ve erken başlangıçlı skolyoza eşlik eden torasik yetmezlik sendromu tedavisi için VEPTR ve ekspansiyon torakostomisini Robert Campbell ve Melvin Smith geliştirmiştir. Spinal füzyon yapılmadan deformiteyi düzeltmeye yönelik bir teknik olan VEPTR sistemi, toraksın görevini yapamadığı, akciğerin büyümesini ve solunumunu desteklemesi için multipl etiyolojiye sahip spinal deformiteli çocuklarda kullanılmaktadır. Bu sistem daralmış hemitoraksı genişleterek ve uzatarak göğüs kafesinin ve vertebranın büyümesine izin vermektedir. Bu teknik direk torasik yetmezlik sendromunu tedavi ediyor gözükmektedir. Aynı zamanda indirekt olarak spinal füzyon oluşturmaksızın skolyozu düzeltmektedir.

VEPTR' ın kullanımında tekrarlayan cerrahi işlemler gerektiği için komplikasyon oranları yüksek oranda görülmektedir. VEPTR sisteminin uygulanması için hastaların yeterli cilt dokusu, subkutan yağ dokusu ve kas dokusuna sahip olmaları gerekmektedir. Eğer uygun koşullar yoksa, VEPTR sisteminin uygulanması sonucu olası komplikasyonlarla karşı karşıya kalınabilir.

Anahtar Sözcükler: Erken başlangıçlı skolyoz, VEPTR, endikasyonlar, cerrahi teknik, komplikasyonlar

Kanıt Düzeyi: Derleme, Düzey V

INTRODUCTION:

Early onset scoliosis (EOS) begins to form in the first five years of life. EOS includes many etiologies (congenital, neuromuscular, idiopathic, various syndromic scoliosis, etc.). When early onset scoliosis is not treated, severe cardiopulmonary problems can occur over time. Accurate detection of the cause of EOS will help to direct the treatment[1].

When children under 5 years of age with scoliosis are not treated, this has been shown to be related to growth abnormalities and pulmonary complications[1]. Scoliosis formation at an early age negatively affects lung development, as lung development continues until the age of 8[14,19,21]. The primary effects are on the development of the pulmonary artery and alveoli. In EOS, a major problem is ventilation defects caused by pulmonary vascularity and defects in lung development[27].

Normal longitudinal growth of the vertebrae occurs at the end plate[4]. In congenital scoliosis, severe progression of deformity is caused by growth at the concave edge, the lack of end plates, hypoplasia of hemivertebrae, the presence of unilateral bar and rib fusions, and progression of growth at the convex side[24,33].

In thoracic congenital scoliosis, associated fused ribs can prevent pulmonary development. Limited growth of the thoracic vertebrae and weak function of the thoracic cavity can cause thoracic insufficiency syndrome[23]. Pehrsson et al. reported that the mortality of untreated infantile and juvenile scoliosis increases in the fourth and fifth decades[29]. Treatment should be carried out at an early age, because the progression of the deformity, beginning at an early age, negatively affects visceral and respiratory development and normal spinal growth[3]. The traditional treatment method for progressive congenital scoliosis is spinal fusion. In recent studies, lung function has been shown to be negatively affected after spinal fusion of congenital scoliosis at an early age in children. Pulmonary functions are also negatively affected by the fusion localization, the levels included in the fusion and the presence of rib anomalies[5,22,39]. Spinal fusion also limits vertebral development[24].

Meanwhile, spinal fusion cannot prevent restrictive lung development. A new technique called VEPTR, which includes no fusion, can be used in the treatment of these patients. This new technique seems to directly treat thoracic insufficiency syndrome. It allows growth of the rib cage and the vertebrae by expanding and extending the narrowed hemothorax. It indirectly treats congenital scoliosis without spinal fusion (Figure-1)[7,10].

ANATOMY, DEVELOPMENT AND PATHOPHYSIOLOGY OF THE THORAX:

An interaction between the vertebrae, the rib cage and the lungs is normal, and the ribs–vertebrae–lungs should be considered as a complex and handled as an elastic structural model[17]. The thorax has a complex and dynamic structure. Strong respiration is provided by the contraction of the diaphragm, the expansion of the rib cage and the support of the cranium, cervical vertebrae and pectoral girdle.

The thorax is composed of the sternum, ribs, thoracic vertebrae and diaphragm. The diaphragm shows continuity with the abdominal muscles.

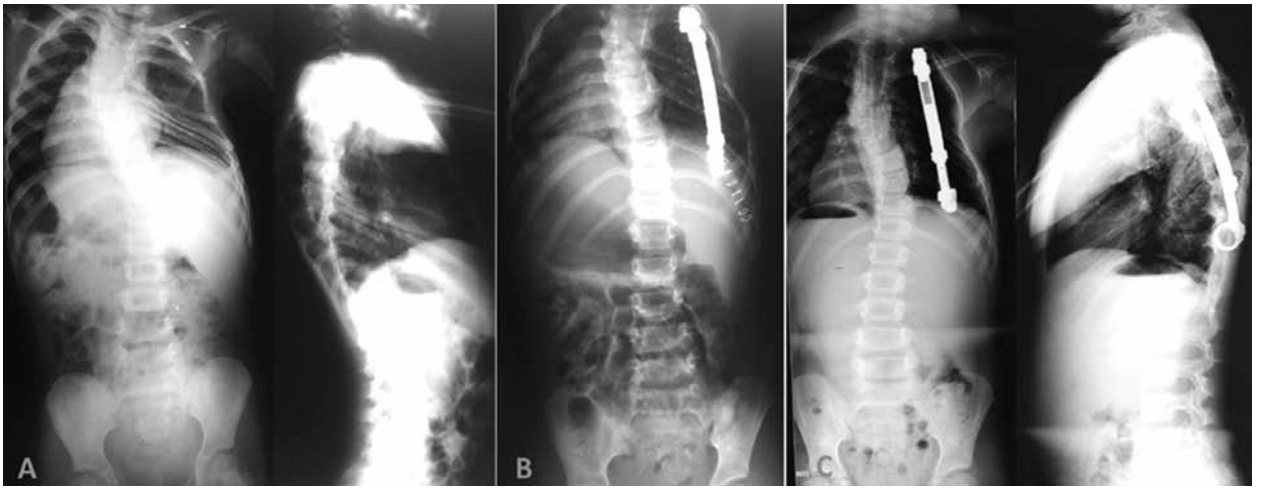

Figure-1. a. Preoperative anteroposterior and lateral X-rays, **b.** postoperative anteroposterior X-ray, **c.** anteroposterior and lateral X-rays after the follow-up period.

The adult thorax narrows until the first rib level, and progresses as a form of ellipsis by laterally expanding in the coronal plane at the eighth and ninth rib levels. The anterior wall of the thorax is at 50% of the level of the sternum thoracic vertebra. While the first rib connects to T1, the eleventh and twelfth ribs connect to their vertebral bodies. The second to ninth ribs connect to the site between vertebral bodies. The direction of the rib tubercles shows differences at the facet joints. The upper seventh rib-transverse process facet joint is directed vertically, and the other distal facet joints are directed horizontally. The eleventh and twelfth ribs do not articulate with their own vertebral transverse processes[2].

The growth of the thorax is complicated. Its geometric structure and functions change over time. The thorax volume consists of 6.7% of the final volume at birth, 30% at 5 years of age and 50% at 10 years of age. It fully completes development at 15 years of age. While the remaining volume to be developed is 70% at 5 years of age, it is about 50% at 10 years of age[17].

The transverse structure of the thorax after birth is circular and narrow. In adulthood, it becomes elliptic. In babyhood, the ribs have transverse planes and all respiration is carried out by the diaphragm. After the age of 4, the angle of the ribs directs downwards, and the anteroposterior diameter of the chest begins to decrease. It fully changes by the age of 10. This new structure of the rib cage contributes to active respiration of the ribs[17,28]. The gain in thorax circumference is 24 cm from 0–5 years old. This slows down between the ages of 5 and 10, gaining 9 cm. This becomes 23 cm between the ages of 10 and 18, peaking at the age of 10[17].

The development of the thorax and the lungs are parallel to each other. 85% of the alveoli develop after birth[15]. Proliferation of alveolar cells occurs at 0–2 years and continues until age 8. After that, lung growth continues by alveolar hypertrophy[10,17]. This continues until the growth of the rib cage stops. The functional residual capacity (FRC) is 30 cc at birth, reaching 3000 cc at adulthood. The lungs weigh 60 g at birth, becoming 750 g in adults[25].

While the diaphragm provides an 80% increase in the vital capacity of lung expansion in adults, rib cage expansion contributes 20%[2]. The thoracic vertebral height is 26.5 cm in females and 28 cm in males at skeletal maturity[17]. When this height reduces due to early fusion or congenital deformities, the lung volume will decrease due to a decrease in the thoracic volume[2]. In a study including patients who received early fusion, Karol et al. stated that a risk of severe restrictive lung disease is present when the height of the vertebrae in the thoracic region is 22 cm or less[22].

Normally, the thorax has two volumes. The first is the stable volume and the second is the variable volume. The volume is dependent on the depth and width of the rib cage, and the thoracic vertebral height. The thoracic vertebrae are positioned posterior to the rib cage as an important support. The diaphragm and the secondary respiratory muscles provide the dynamic structure of the thoracic volume, the variable volume of the lung[10].

Campbell and Smith defined 3D deformity of the thorax by evaluating in three planes (coronal, sagittal and transverse). They separated volume loss due to thorax deformity into three types. In Type 1, volume loss at the deformity site develops due to hypoplasia of the unilateral thorax and the lack of rib and exotic scoliosis (complex early onset spinal deformity with lordotic and advanced rotation). In Type 2, there is a volume insufficiency due to hypoplasia of the unilateral thorax, congenital scoliosis, rib fusion, and vertebral, anal, cardiac, renal, esophageal and extremity abnormalities. In Type 3, there is volume loss due to global thoracic hypoplasia (Jeune's syndrome)[12].

Day et al. defined respiratory deficiency in congenital scoliosis[16]. Campbell et al. defined thoracic insufficiency syndrome (where the thorax cannot support normal respiration and lung growth) in children with volume loss due to deformity. This situation occurs because the rib cage cannot support lung growth and normal respiration [10,11].

Mechanical problems causing respiratory deficiency in early onset scoliosis are thoracic volume loss, rigid thorax, anomaly of chest wall muscles and inhibition of diaphragm muscles. Thoracic vertebrae can be shorter in congenital scoliosis. Congenital chest wall anomalies include large rib fusion and a small hemothorax, and result in a decrease in the final thorax volume. Lordosis and rotation of scoliosis narrow the thoracic region at the convex site. Distortion of rib fusion and the rib cage limits expansion by causing rigidity in the thorax. In severe thoracolumbar deformities, thorax collapse and kyphosis can cause thoracic insufficiency by inhibiting the diaphragm[20].

While lung volume in the hemothorax decreases at the convex site due to rotation of thoracic vertebrae in EOS, as this is a rigid structure, movement at this site negatively affects tidal volume. The aim of EOS surgery is to obtain the best pulmonary function which is stable for the patient's lifetime, by correcting the thorax form and function and allowing its development. Although there is currently no ideal technique for this, technological improvement will allow this to be obtained in the future[2].

VERTICAL EXPANDING PROSTHETIC TITANIUM RIB (VEPTR):

Robert Campbell and Melvin Smith developed VEPTR and expansion thoracostomy for the treatment of thoracic insufficiency syndrome associated with congenital scoliosis, rib fusion,

hypoplastic chest wall deformity and early onset scoliosis [9,11].

This technique with expansion thoracostomy became common after it was used with an establishment from rib-to-rib and/or rib-to-pelvis, and successful results were obtained (Figure-2)[9,20].

VEPTR is a technique that aims to correct deformity without fusion. Sufficient tissue, subcutaneous fat and muscle tissue are required for application of the VEPTR system. The VEPTR technique aims to correct lung capacity and function by expanding the thorax and controlling progression of the deformity. The system directly repairs reduced thorax volume and 3D thoracic insufficiency, and indirectly repairs scoliosis with repetitive extensions after thoracostomy[7,26].

The Food and Drug Administration (FDA) approves the use of the VEPTR system for diagnoses including flail chest syndrome with thoracic insufficiency syndrome (TIS), narrowed chest wall syndrome associated with scoliosis and rib fusion, Jeune's syndrome, achondroplasia, Jarcho-Levin syndrome, hypoplastic thoracic syndrome associated with Ellis–van Creveld syndrome, and neurologically- and congenitally-originated progressive scoliosis without rib anomalies. In tomography, a thorax "windswept" deformity in the apex of the curve is present, which is the basis of narrowed chest wall syndrome. If this situation is present in infantile idiopathic scoliosis and early onset syndromic scoliosis, the VEPTR system can be used in these patients[11].

Campbell[8,9] listed the indications and contraindications of the VEPTR system in his article as follows:

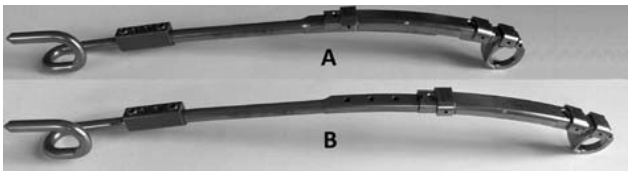

Figure-2. a. Hybrid system (showing Dunn-McCarthy hook used as distal attachment), **b.** elongated hybrid system.

Indications

1. Abnormal vertebrae in apex of rib and concave hemithorax with three or more fusions and progressive deformity.
2. Progressive insufficiency syndrome, thorax that cannot support lung growth or normal respiration.
3. More than 10% loss of height of concave hemothorax when both hemithoraces are compared (current lung area under 90%).
4. Child must be at least 6 months of age.

Contraindications

1. If soft tissue cover is not sufficient.
2. If normal body index is lower than 25%.
3. The presence of disease (cardiac disease, pulmonary disease, medical problems) that cannot handle general anesthesia, or a lack of tolerance for repetitive surgery.
4. The presence of active pulmonary infection.
5. The absence of diaphragm function.
6. If the rib bone stock is insufficient for attachments (severe osteogenesis imperfecta etc.).
7. Severe rigid kyphosis (over 50°).

Relative Contraindications

1. In the absence of ribs in the superior (for placement of attachment).

2. Fusion history in vertebrae in the thoracic region.

If there is a large degree of curvature and/or a rapid progression risk for congenital deformities, surgery is considered[40].

The VEPTR system is used for children who have spinal deformities with multiple etiologies, and whose thorax cannot function, to support lung growth and respiration[11]. The strongest alternative treatment for these diseases is the growing rod technique[38]. Congenital scoliosis with no severe chest deformity can be treated with VEPTR or the growing rod. In severe chest deformities, the VEPTR system is the first choice after thoracostomy[40].

PLANNING BEFORE SURGERY:

Planning should begin by determining whether there are respiratory problems. It should be learned primarily whether there are episodic pneumonia attacks, whether the patient requires extra oxygen, and whether support with a ventilator or CPAP (continuous positive airway pressure) has been required during pneumonia attacks in the history.

It should be analyzed whether there was any prematurity, bronchopulmonary dysplasia, tracheobronchial malacia or intrinsic lung disease in the birth history. The age at surgery and fusion levels of any prior spinal surgery should be determined.

All children with scoliosis at an early age should be evaluated with ultrasonography for renal pathologies, magnetic resonance for intraspinal pathologies and echocardiography for cardiac pathologies[8].

SURGICAL TECHNIQUE:

The VEPTR system should be placed into the concave hemithorax along the posterior axillary line. In the VEPTR system connecting rib-to-rib, the attachments should be placed at the upper and lower ribs. A hybrid VEPTR system, on the other hand, might consist of rib attachment on the upper side, and laminar hook, pedicular screw, or Dunn-McCarthy rod (placed in the middle 1/3 of the iliac crest) on the lower side.

A hybrid VEPTR system is placed medial to the rib-to-rib VEPTR system. The placement of the second system provides both mechanical support and scoliosis correction. As there is sufficient soft tissue cover in children older than 18 months, both a rib-to-rib VEPTR system and a hybrid system can be placed[9] (Figure-3).

Patients should be placed on the operating table in the lateral decubitus position with the concave hemithorax upwards. The upper extremity at the shoulder should be removed from the area of surgery, providing that does not require more than 90° of flexion. A modified thoracotomy skin incision is used. A skin incision is drawn in the cranial direction starting from 1 cm lateral to the upper thoracic vertebral middle line to the caudal. The skin incision should be extended in an L shape by turning from 4 cm below the scapula tip in the caudal direction to the anterior. Then, the rhomboid and trapezius muscles should be divided in two by electrocautery.

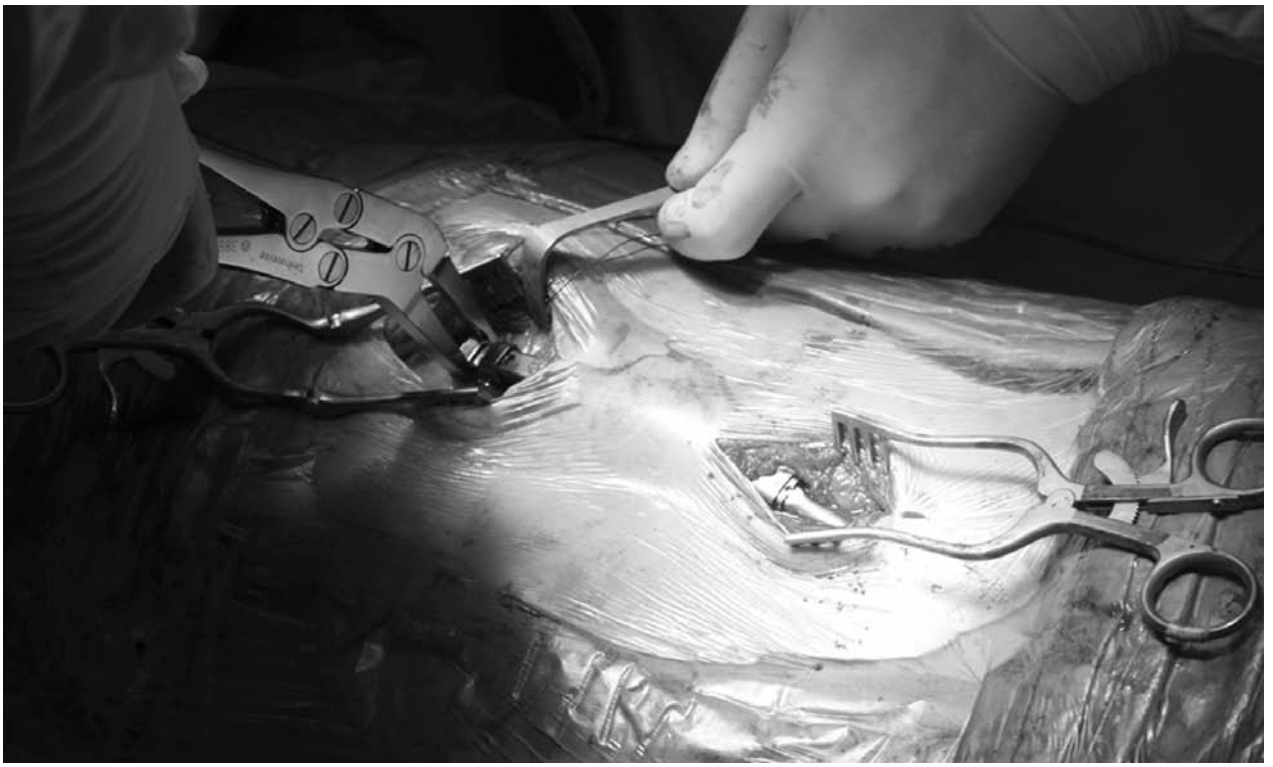

Figure-3. Hybrid system (the placement of the extension clip is seen in the proximal part).

The scapula is retracted towards the superior, and by accessing the interval between the scapula and chest wall by blunt dissection, a space is created in the proximal for the VEPTR system to be placed next to the ribs. The posterior and middle scalene muscles and the neurovascular bunch located in the anterior of the scapula can be seen. Anterior to those muscles is a dangerous area, while posterior is a safe area. The system should not be set to the first and second costa, as this has a risk of pressing the neuromuscular bundle.

Since it will trigger fusion, the periosteum of the ribs should not be opened and the vascularity of the ribs should be protected. By accessing the intercostal muscle tissue below the rib with a 1 cm incision, the place where the attachment will be placed should be prepared. The periosteal part just below the rib is exposed and the pleural part is avoided. The rib to which the attachment will be placed should not be thicker than 1 cm. If the rib is weak, as this creates a risk that the attachment will migrate, two ribs are placed together in a single attachment. Since migration of the attachment to the cephalad part might put the brachial plexus at risk, the first rib should never be used. The place for the cephalad attachment should be prepared as adjacently as possible to the transverse process tip from the transverse side, as this is advantageous. The attachment is locked after placement. The place for attachment in the inferior part is also similarly prepared. After the placement of the attachments, an attachment lock is placed using pliers[9].

Then, if there are missile ribs in the apex part of the thoracic area in the concave part, open wedge thoracostomy should be administered to this crinkled area.

This area should generally be in the superior part of the apex of the curvature. Osteotomy is performed by protecting the pleura from the transverse process of the vertebra to the thorax in the costachondral joint. This line is generally joined by the fibrosis band. It is possible to open this line withelectrocautery. Another alternative is to open this line with Kerrison equipment. The distractor is placed between the ribs on this osteotomy line and distraction is applied slowly. After obtaining enlargement, the system is locked. If a space of more than 3 cm is formed after osteotomy, the VEPTR system can be attached to the rib with a cable for the stability of the system[9].

A hybrid system is generally preferred for thoracolumbar scoliosis of children older than 18 months. The canal diameter should be evaluated before placing the hook at the lumbar area. The place for the inferior attachment is prepared among the paraspinal muscles by opening a second longitudinal incision from the lateral of the caudal lumbar vertebra. The hook is generally placed into the neutral vertebrae (generally L1, L2 or L3). The longer the hybrid system the better, meaning that placement distal to the T12 enlargement is preferable. A hybrid rod is attached lordotically by curving in the valgus in the inferior. Then, a rib-to-rib VEPTR system is placed. The superior attachment should be attached generally to the third rib providing that this is not upwards of the third costa, and the inferior attachment should be attached to the T10 rib, which is generally stable. First the rib-to-rib system and then the hybrid system is distracted. An autograft can be placed to the edges of the ribs containing the hook and the attachment[9].

In the hybrid system, in cases when there is a defect in the inferior, especially in the lumbar vertebral lamina with meningocele, a Dunn-McCarthy hook is placed. The middle or posterior iliac apophysis is cut with a transverse incision. After placement of the Dunn-McCarthy hook, providing that it will contain the iliac crest, the apophysis is sutured again[9].

A drain is placed into the area without closing the skin or subcutaneous tissue. When the amount coming out of the drain falls to <20–25 ml/day, it can be removed. There is no need for brace use in the postoperative period. The patient is allowed to resume normal activity approximately 1.5 months after surgery. The extension/expansion process is performed at 4–6 month intervals. Approximately 5–10 mm of distraction is administered by making an incision to the area where the system is locked for extension[9].

The first expansion is performed under general anesthesia six months after implantation. Maximum correction should be provided in the first expansion. After providing the expansion by removing the VEPTR system lock after accessing it with a 3 cm short incision, the lock is replaced. The patients are discharged from hospital 24 hours after the expansion process. Correction is performed in the next expansion with growth, but correction of the chest and spinal deformation is not as successful as in the first expansions. All expansions should be repeated every six months. With regular expansions, fewer implant-related complications have been reported[8,40].

The use of the VEPTR system is suggested until the skeleton reaches maturity and lung enlargement is provided. When sufficient

development is obtained, a fusion process is administered by removing the system[8].

POSTOPERATIVE FOLLOW-UP:

The patients are generally removed from intubation. The functional mechanism of the respiratory system changes acutely with the thoracic reconstruction of VEPTR. Some patients might tolerate extubation the day after surgery. In those patients, more than 30% hematocrit provides the optimum oxygen carrying capacity. To prevent acute pulmonary edema development, the fluid intake should be restricted. The drain should be removed when the amount draining falls below 20 cc in a 24-hour period. After 24 hours, when the chest tube has less than 1 cc/kg drainage, it is removed[8].

If a respiratory problem occurs in the patient after the removal of the drain and the chest tube, an emergency chest tube should be placed by considering the formation of pleural effusion and lung compression[8].

The patients are mobilized as much as possible. Use of a brace is avoided, as it compresses the thorax[8].

COMPLICATIONS:

With VEPTR, as with other techniques, repetitive surgeries are required, meaning it is a technique of intermediate severity with a high rate of complications[38].

Whichever surgical method is chosen for the treatment of spinal deformities, it is accompanied by comorbidity. Deformities with congenital abnormalities are generally accompanied by cardiac pathologies, renal pathologies, gastrointestinal pathologies, repetitive respiratory system problems, and a failure to thrive. These problems increase the postoperative complication risk[13,36].

Some of the most common complications are skin infection, skin necrosis, loss of skin and subcutaneous tissue over the implant and contact of the implant with the outside. When there is an infection around the VEPTR system, treatment initially includes irrigation, sufficient debridement, and intravenous antibiotics followed by oral antibiotics, without removal of the instrumentation. According to the results of bacterial cultures, the antibiotics may be changed. Sometimes vacuum assisted closure (VAC) can be used. The edges of the skin can be loosened with surgical exploration to give a 5 mm gap, and the wound edges sutured to each other with prolene (Figure-4)[8,36].

Smith et al. reported that they implemented 678 VEPTR procedures (including implementation, revision and elongation) on 97 patients, and in 16 patients they encountered 19 infections (2%). 13 of the 19 infections encountered were superficial, while six were deep infections. For 15 patients, the pathogen was *S. aureus*.

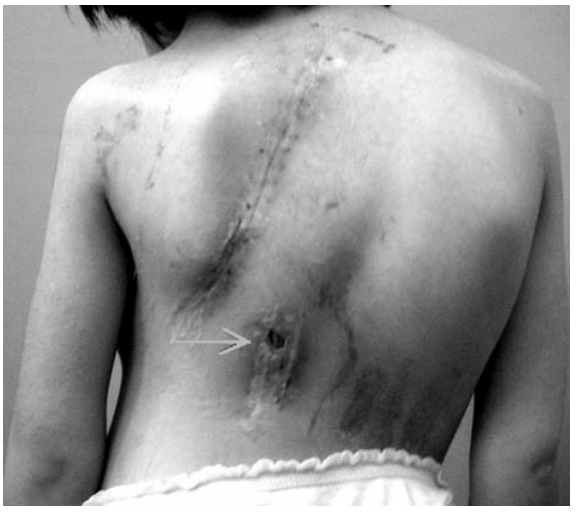

Figure-4. Arrow showing the opening on the inferior attachment and infection.

All patients were initially treated with irrigation and debridement with simultaneous intravenous (IV) antibiotics. 13 of the superficial infections were successfully treated with antibiotic treatment, irrigation, or debridement. In the other six patients, two or three of the treatments were used. For these patients, an average of 58 days of IV and 34 days of oral antibiotic treatment was continued. The authors proposed the following strategy in the case of infection: after cultures have been obtained, irrigation and debridement should initially be carried out, followed by closure of the wound when the infection is controlled. IV antibiotic treatment must be continued for 4–6 weeks, until the ESR is normalized, and after that oral treatment should be given for a maximum of six weeks. They reported that they had success without the removal of the implant[35].

In the case of recurrent infections, removal of the implant is suggested. After six weeks of IV antibiotic treatment and wound healing, re-implantation surgery should be performed.

Recurrent surgery and scarring increase the complication risk. In order to decrease complications during elongation, the second incision should not be done directly above the implant[36].

Soft tissue gangrene (skin slough) is a very important complication. In order to prevent this complication, the system must be implanted under the muscle tissue as much as possible. The patient's nutritional status must be corrected as soon as possible. In Campbell's clinic, to solve the nutritional problem, 0.25 mg/kg/day of cyproheptadine hydrochloride (generally a 2 mg dose (1.5 tablets) orally, divided into 2–3 doses a day) was supplemented to children older than 2 years of age, and a gastrostomy tube was placed for patients with severe nutritional problems. The skin was primarily closed after debridement.

When there is recurrent skin thinning, rotational flaps may be required. Tissue expanders placed subcutaneously lateral to the implant prepare the skin tissue, and this skin tissue is transferred to the posterior of the implant to close the defect. Sutures must be removed a minimum of four weeks after surgery. After closure, antibiotic treatment must be continued for at least six weeks. If the skin necrosis is larger than 5 mm, the attachment underneath must be removed[8,9].

A few months after implantation, the location of the rib attachment is filled with bone tissue. New bone tissue is sometimes formed under the attachment. Because of this, migration of the attachment to the hypertrophied rib may be observed. Complete migration is observed three years after the implantation of the system.

Migration of the superior attachment is generally asymptomatic. Migration of the attachment is generally observed under the trapezius. For diagnosis, computerized and 3D tomography is useful. Due to the risk of skin erosion, revision must be done one rib above and one rib below[9]. This can be performed by a small incision during expansion surgery[8].

Rarely, the hook used in hybrid systems may be displaced. A Dunn-McCarthy rod might result in further distal migration. If the migration is greater than 2 cm, it must be repositioned[9]. Unlike the growing rod system, failure of the system and spontaneous fusion are complications that are rarely seen (Figure-5,6)[9,40].

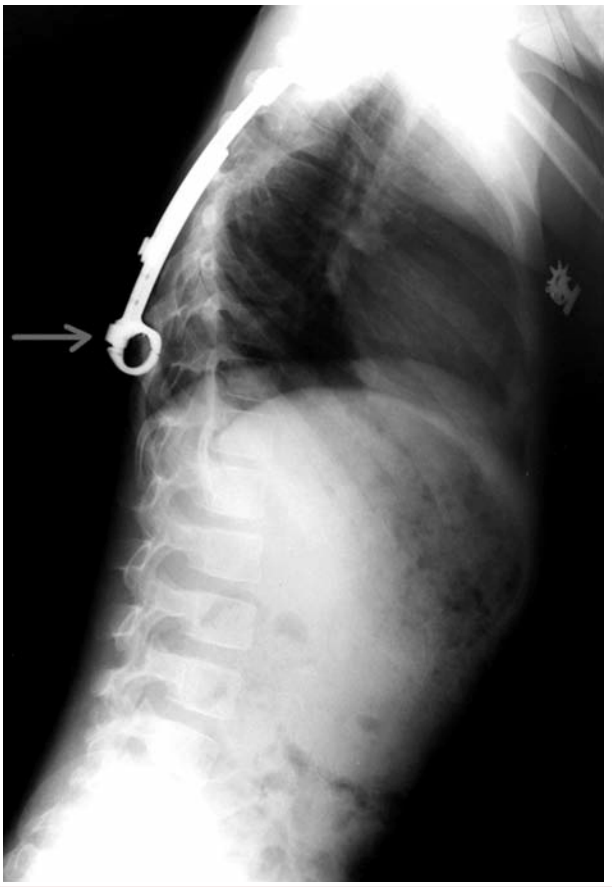

Figure-5. Arrow showing migration of the inferior attachment.

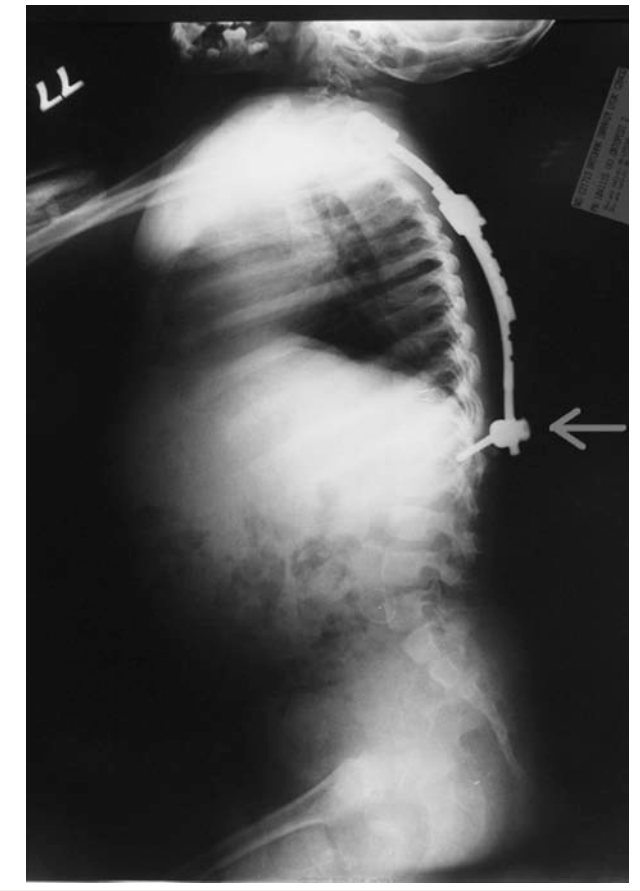

Figure-6. Arrow showing pulling out of the inferior screw.

During implantation of the VEPTR system, brachial plexus lesions can occur. Brachial plexus injuries can happen when the upper rib attachment is placed too high and too laterally. More frequently observed is acute thoracic outlet syndrome, when the upper rib attachment migrates to the superior, and during coverage of the system the scapula muscles are pulled down caudally[20].

Spontaneous rib fusion may be observed after the initial thoracostomy, and expansion thoracostomy may be needed. Regular elongations can decrease these complications[40].

While there is no treatment required for pleural tears of 1–2 cm, larger tears must be sutured. When large pleural defects are present, a chest tube must be placed. The chest tube is removed when the drain flow decreases to less than 1 ml/kg/day[9].

DISCUSSION:

Early onset scoliosis affects lung function and leads to pulmonary hypoplasia by preventing lung development, while late period scoliosis affects lung function through disturbing the chest wall mechanics[10].

Davies et al. examined lung parenchyma of children with kyphoscoliosis at autopsy and found pulmonary hypoplasia and reduction in the number of alveoli of the children whose scoliosis began before the age of 8. They observed

that the shape of the lungs takes the form of the thorax, and deformity compresses the lung. They reported that the major effect of this compression is a reduction in alveoli. They explained the cause of death of three out of four patients as cardiopulmonary failure[14].

In 2008, Karol et al. evaluated pulmonary function after an average follow-up of five years, after the application of thoracic fusion to 28 patients without neuromuscular etiology before the age of 9. As a result of early fusion, the thorax anteroposterior diameter of these patients remained short. The patients who received fusion of more than four levels in the proximal thoracic region (stretching to T2), and with congenital grade abnormalities in this region, were found to be at the highest risk for respiratory failure[22].

Muirhead et al. evaluated the vital capacity, total lung capacity and gas volume (FEV1) values of patients with major thoracic curvature and with congenital and idiopathic scoliosis at 13.3 years of age. They found that the lung function, in particular, was worse in infantile idiopathic scoliosis patients, in patients with congenital scoliosis, and with more deformity[27]. Lung function was found to be worse in the group with the same Cobb angle but more thoracic anomalies[16].

As seen in all studies performed, although there is a decline in lung function in congenital or idiopathic scoliosis, greater decreases in lung function are seen in early-period deformity, especially of a congenital origin. In congenital scoliosis, particularly with thoracic insufficiency syndrome, attacks of atelectasis and pneumonia occur extensively. As a result, chronic respiratory failure occurs[10].

Early fusion, especially at the proximal part of the thoracic spine, disturbs the thorax morphology and blocks the thoracic volume. For normal lung development and spinal and thorax growth, it is necessary to protect the elastic structure of the spine. Cambell and Akbarnia developed changes to the treatment of EOS. The current popular techniques (the growing rod technique or VEPTR) prevent failure of thoracic spinal deformities, and allow the lungs and vertebrae to develop. Spinal fusion inhibits the development of the lungs by stopping vertebral growth and thorax development. Early fusion should be avoided, and if it becomes necessary then short-segment fusion is more appropriate[3,17]. After early fusion, shortness of body develops due to the prevention of vertical growth, and so a disproportionate anatomy occurs (a short body and longer lower extremities)[38].

The VEPTR technique allows the development of the lungs by expanding the thorax. At the same time, the VEPTR technique aims to fix lung capacity function and control the progression of deformity. While VEPTR and expansion thoracostomy directly fix the three-dimensionally decreased volume of the thorax and thoracic insufficiency, they indirectly fix scoliosis[7,26]. This can be best understood using computed tomography[38]. Campbell et al. reported an effective technique for surgical treatment of cases with grade fusion and thoracic insufficiency syndrome as well as congenital scoliosis. They reported that they provided rigid thoracic scoliosis correction and extension of the shrunken concave hemithorax, and as a result, the lungs expanded and the tidal volume also increased[7]. Emans et al. reported that spinal deformity was reduced, growth of the vertebrae continued, and lung development was permitted for patients treated with VEPTR and expansion

thoracotomy[20]. Smith et al. evaluated the lungs of congenital scoliosis patients who received the VEPTR system using computed tomography and found an increase in lung volume in both hemithoraces (even on the side that was not instrumented)[35].

More frequent breaths are taken with a lack of pulmonary function in thoracic insufficiency syndrome, and as a result, more energy is required. The nutritional status of most patients is poor, with a weight ranking below the 5% percentile. Skaggs et al. published that fixing pulmonary function by use of the VEPTR system contributed to weight gain of the patients[26].

The normal growth of the spine depends on the uniform distribution of the load on the last discs in the vertebral body[17,18]. According to the Heute-Volkmann principle, asymmetric loads lead to increased deformity and abnormal growth[37]. The growth of the spine is greatest between the ages of 0–5. While the growth in the T1–S1 distance between the ages of 0–5 is 2.2 cm/year (increase in T1–12 distance: 1.1 cm/year), the growth in the T1–S1 distance between the ages of 5 and 10 is 1.1 cm/year (increase in T1–12 distance: 0.8 cm/year). Campbell et al.[7] identified the growth in the T1–12 distance as 0.8 cm/year in grade-fused and congenital scoliosis patients who received the VEPTR system after expansion thoracostomy. In another study, Campbell et al. published the growth of the T1–12 distance as 0.71 cm/year in patients with congenital scoliosis who received surgery. Campbell et al. monitored patients with congenital scoliosis treated with VEPTR and grade fusion using computed tomography. They also determined the growth at the concave edge of the unilateral unsegmented bar[7,11].

Emans et al.[20], in an article published in 2005, reported the growth of the thoracic region for patients with congenital scoliosis who received surgery as 1.2 cm/year. Emans achieved similar growth to normal growth. Motoyama et al. achieved a spinal growth greater than the normal growth rate[26]. In our clinic, after an average follow-up period of 2.2 years for the patients who received the VEPTR system with various etiologies, we identified the increase in the T1–S1 distance as 1.53 cm/year. These studies support the suggestion that the VEPTR technique permits the growth of the spine.

Although cervicothoracic or cervical scoliosis is rarely seen, this can be seen, depending on the vertebral malformations. In the early period, in the absence of pain, a painful process may occur with the progression of the deformity. Commonly in the advanced stages, head tilt, torticollis, shoulder asymmetry, and restriction of the neck movement can be seen. The status of the cervical tilt is important for patients in terms of problems that may develop in the future. Campbell et al. evaluated whether there was any improvement in the cervical tilt of patients with congenital scoliosis and a severe cervicaltilt who received the VEPTR system. They stated that there was a slight improvement in the cervical tilt of 14 patients with congenital scoliosis and more than 10° of tilt, the horizontal shoulder angle of 57% of the patients improved an average of 10°, and there was an improvement in the head and trunk decompensation[6].

Samdani et al. detected an improvement in T1 tilt and head shift of patients treated with the VEPTR system[30].

In another study, implant problems were seen at high rates after the establishment of the VEPTR system in patients with a low percentile[34].

A study by Sankar et al. showed that the VEPTR system did not significantly affect the formation of BMI or kyphosis complications[31].

Campbell et al.[11] encountered 52 major complications in 22 patients with congenital scoliosis in a study in 2004 (2.36 complications per patient). Sankar et al.[31] reported that 45 complications occurred in 19 patients in the VEPTR group (11 congenital, eight neuromuscular, one idiopathic) in a study in 2010 (2.37 complications per patient).

In a study by Shultz et al[32], since only infantile idiopathic scoliosis patients were included, the complication rate was not shown to be high. In our clinical series of 12 patients (six congenital, three idiopathic, two syndromic and one neuromuscular), 26 complications occurred in ten patients (2.17 complications per patient).

In Emans's study, they observed attachment migration eight times, pelvic fixation loss once, brachial plexus palsy twice, grade fusion repeat twice, deep wound infection twice, and grade fracture twice[20].

In our 12 patients, the complications that occurred during an average follow-up period of 2.2 years were similar to those published in other studies (superior grade attachment migration six times, hook dislocation five times, deep wound infection five times, inferior grade attachment migration four times, screw dislocation twice, lamina fracture once, crankshaft phenomenon once, loosening of McCarthy rod once, and superficial wound infection once) (Figure-7).

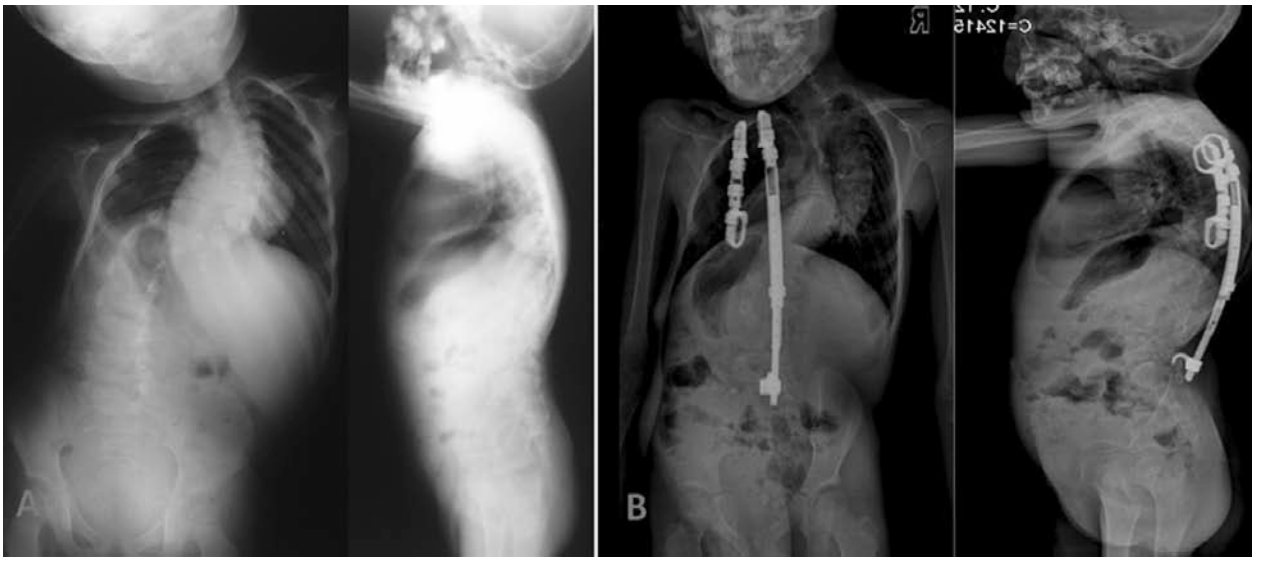

Figure-7. a. Preoperative posterior-anterior and lateral radiographs, **b.** Posterior-anterior and lateral radiographs after the follow-up period.

A common major complication in other studies, as well as in our own, is superior grade attachment migration [11,20]. Schulz et al. supported the attachment with a Mersilene band to reduce the superior grade attachment complications and reported that they encountered less migration[32].

The VEPTR system in EOS seems to support the development of the chest, the growth and development of lung function, and spinal correction and growth. In the VEPTR system, a series of operations are required to provide expansion and to improve the correction. As a

result of this, the risks of infection and implant failure increase. A further series of operations may be required to solve these complications.

Despite recent technical advances, the surgical treatment of early onset scoliosis is still complex and shows a high complication rate. Additionally, these children are at high risk due to comorbidity, and the treatment period is very long. All the extensions are performed under anesthesia.

As a result, patients suitable for the use of the VEPTR system should be selected well. The family should be informed about the treatment, the duration of the treatment, and possible complications.

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