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Royal Medical Services (RMS), Amman, Jordan

Corresponding author: Asem Almajali, MD, FACS. Orthopedic and Spine surgeon. KHMC. Royal Medical Services (RMS), Amman, Jordan. Phone: +962772540705. E-mail: Almajali_asem@yahoo.com. ORCID ID: http//www.orcid.org/0000-0002-5350-1824.

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Early Childhood Scoliosis Management by Vertical Expandable Prosthetic Titanium Rib (VEPTR): Experience of Royal Medical Services (RMS)

Asem Almajali, Mohammad Obeidat, Omar Bashmaf, Raed Wagokh, Bassam Harahsheh, Raed Alzaben

ABSTRACT

Introduction: Scoliosis could develop at the childhood age and progress beyond skeletal maturity. An early spinal fusion arrests growth of the spine and thorax, risking the development of secondary thoracic insufficiency syndrome. Vertical expandable prosthetic titanium rib (VEPTR) is a fusionless technique aiming to correct the deformity with preservation of growth potential. **Aim:** To show our experience and results regarding the use of VEPTR in children with scoliosis in regard to coronal profiles(length and deformity angle), spinal growth, and the complications we faced during the follow-up of two years after the index procedure. **Methods:** A retrospective analysis of prospectively collected data of a case series. Forty child with scoliosis of different etiologies. Their primary diagnoses were neuromuscular scoliosis in 13, Juvenile idiopathic scoliosis in12, Congenital Scoliosis in 8, syndromatic patients 5 and 2 with Arthrogryposis. All 40 patients received percutaneous rib-to-pelvis or rib to vertebra or rib to rib VEPTR implantation between January 2016 and January 2018. None of them needed blood transfusion. They underwent 56 primary implantation, 16(40%) bilateral system and 24(60%) unilateral followed by lengthening procedure in a period of 4-6 months. The patients were assessed based on mechanical measures, that is, the radiographic improvement of their scoliosis, spinal height, and sagittal and coronal correction, which are measured and compared preoperatively, immediately postoperatively and at two years follow up, complication encountered during this period are also counted. **Results:** The average initial correction in Cobb angle immediately after the index surgery was 14.4° (5°-26°) and the average final correction of Cobb which is measured after the last expansion procedure (Cobb angle of the major curve measured after last expansion minus initial preoperative Cobb angle of the major curve) was 7.3° (12%). The average of preoperative coronal T1-S1 length was 25.6 cm with an average initial correction achieved immediately after implantation of VEPTR of2.8 cm (1.2-5.1cm) which is 10.9%, and the average coronal length gain at 2 years follow up was 5.7 cm (3.7-9.8cm) that is 22.2%. Complication occurred in 18 of our patients (45%). **Conclusion:** Early results of VEPTR for childhood scoliosis are encouraging. Follow-up till skeletal maturity will best determine future indications.

Keywords: Early Onset Scoliosis (EOS), VEPTR, Major curve.

1. INTRODUCTION

Early-onset scoliosis (EOS) is defined as scoliosis of any etiology which develops before the age of 10 years (1-10). Untreated EOS can result in short trunk and thoracic cavity decreased capacity, which may significantly affect pulmonary function as lung alveoli development continues until the age of 8-10 years (1-3, 11). Progressive EOS if remained untreated, and causes cardiac disease, respiratory problems and, higher rates of mortality if compared treated patients (8, 10).

Healthy children without spinal deformities reach25- 30% of chest capacity and up to 70% of T1-S1 height by the age of 5-6 years (1, 2). The coronal height from T1-S1 continues to grow at a slower rate (9mm/y) between the age of 6 and 10 years and then increases (16-20 mm/y) between the age of 10 years and skeletal maturity Spinal fusion is better to be avoided in young children to prevent long-term complications related to fusion arrested spine growth and subsequently small thoracic volume (14). Alternatively, fusionless procedures are now commonly used in to limit the progression of scoliosis while enhancing thoracic growth (1, 2, 4, 10, 11). The growing rods and rib-based distraction devices [vertical expandable prosthetic titanium rib (VEPTR) have been developed to correct the scoliosis and to increase spine height during early childhood (3, 7).

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) was developed based on a trial of a Steinmann pin chest wall prosthesis that was placed in 1987 in an 8-month-old ventilator dependent infant with sever Thoracic Insufficiency syndrome (TIS) and sever rib cage deformity (13) then. The concept of thoracic insufficiency syndrome soon became the central guiding for VEPTR principle (2, 3, 9).

How does the VEPTR work?

When the spine curves, it pushes on the lungs so your child cannot breathe deep enough and the lungs cannot grow enough.

VEPTR: The VEPTR device is a metal rod curved to fit the back of the chest and spine which is placed in an up and down position. It can be expanded as the child grows (13).

Spreads the ribs out so the chest is shaped more naturally that Helps the lungs have enough space during the 1st 8 -10 years while the alveoli are developing to grow and fill with enough air to breathe (12, 14).

Can be expanded as the child grows so it still fits right (a surgeon will need to adjust it from time to time) (1, 3, 10).

It is fusionless technique for the spine during the growing period of the child, so you maintain the proportional growth of the non-fused spine and the limbs.

Despite those many advantages, a concern has been raised that serial posterior distraction surgeries may progressively lead to less spine growth with time (4, 8). The repetitive surgical lengthening procedures and the high rate of complications is another disadvantage for this implant which we will discuss in our study (6, 9, 12, 13).

2. AIM

The aim of this study is to show experience and results regarding the use of VEPTR in children with scoliosis in regard to coronal profiles(length and deformity angle), spinal growth ,and the complications we faced during the follow-up of two years after the index procedure.

3. MATERIALS AND METHODS

This is a retrospective analysis of prospectively collected data of case series study to examine the outcome of rib-based distraction (VEPTR) in patients with EOS with or without rib abnormality at 2-year follow-up.

VEPTR implant surgery in all cases was done by the same surgical team at Royal Rehabilitation center (RRC) of Royal Medical Services (RMS), Using percutaneous technique (5, 7) in all cases using Rib To Rib connection (1 case 2.5 %), Rib To Lumbar spine using pedicle screw or sub laminar hock (9 cases 22.5%) or Rib to Pelvis using iliac hock(30 cases 75%) (Figure1).

Some of the patients mandate bilateral implant (16 cases 40%) but the majority required only unilateral implant (24 cases 60%).the ribs used to implant the proximal rib hocks and ring were the 2nd and 3rd ribs and in complicated rib fractures and cut through we reimplant the ring in lower ribs.

The study enrollment started in January, 2016 and ended in January, 2018. Total number of the enrolled patients was 40 patients, 26 females (65%) and 14 males (35 %), the average age was 5.4 years(4.1 years to 7.9 years) All of them had completed 2-year follow-up at the time of data analysis. The underlying cause of the deformity in our patients is shown in Table 1.

All patients underwent regular surgical expansion procedure in time interval of 4-5 months; some required exchange of the sliding bar to a longer one during the expansion surgery because it reached its maximum limit.

Some of our patients required revision surgeries in between expansion surgeries because of complication which we will discuss later.

Whole spine radiographs (standing or sitting) with both anterior-posterior (AP) view and lateral view were performed at each clinic visit (preoperative, direct postoperative, and on regular follow-up visits).

The radiographs were assessed by the study group to measure the major and secondary curve scoliosis angle sand maximum spine kyphosis. Spinal (T1-S1) coronal spine height (measured between the upper border of T1 and the lower border of S1 vertebrae) as well as the major curve Cobbs angles and sagittal spine length (SSL) were measured on the AP and lateral spine X ray views, respectively. Some patients did not have sufficient lateral radiographs for accurate measurement of kyphosis and SSL; therefore, we depended only on their AP X-rays and their values (Figure 2).

The end point of our study was the radiographic findings at the 2-year follow-up after VEPTR implantation surgery. The End result changes was calculated as the difference between X ray finding and measurement at 2 years follow up and the initial X ray findings and measurement before and after the index procedure, Percentage of change at 2 years was calculated.

The coronal spine height growth was calculated for the total 2-year follow-up period and the primary outcome measure was the major scoliosis curve angles and

Cause	Number of cases	Percentage
Juvenile idiopathic scoliosis	12	30%
Neuromuscular : Myopathies:	6	15%
:cerebral palsy:	5	12.5%
:myelomeningiocele:	2	5%
Congenital scoliosis: multilevel anomalies:	5	12.5%
: kyphosis :	3	7.5%
Arthrogryposis :	2	5%
syndromatic	5	12.5%

Table 1. Demographic distribution of the scoliosis causes in our patients.

Patient	Sex	Age Year	Cause of scoliosis	Major curve	Preoperative Cobbs Angle	Immediate post op Cobbs Angle	Initial correction	No. of distractions	Period of f-up Month	Final coobsangle	Final correction
1.	F	4.7	JIS	T	43°	35°	8°	5	24	38°	5°
2.	M	5.1	NM	T	54°	40°	14°	4	23	46°	8°
3.	F	6.2	NM	TL	62°	39°	23°	5	24	48°	14°
4.	F	5.3	Syndrom	TL	55°	43°	12°	5	24	47°	8°
5.	M	4.6	Cong.	T	48°	40°	8°	5	23	49°	-1°
6.	M	6.3	JIS	T	77°	55°	22°	4	23	64°	13°
7.	F	4.1	NM	L	56°	43°	13°	6	24	49°	7°
8.	F	7.1	Syndrom	TL	44°	31°	13°	5	24	40°	4°
9.	F	6.2	NM	L	66°	40°	26°	5	23	42°	14°
10.	F	7.2	Syndrom	T	75°	61°	14°	5	23	67°	8°
11.	M	5.3	JIS	T	61°	38°	23°	4	23	50°	11°
12.	M	5.6	NM	T	45°	31°	14°	4	25	37°	8°
13.	F	6.1	JIS	TL	61°	46°	15°	6	26	52°	9°
14.	F	4.3	Cong	TL	40°	31°	9°	5	24	40°	0°
15.	M	4.6	Cong	T	49°	36°	13°	5	25	41°	8°
16.	F	4.9	NM	TL	67°	50°	17°	5	24	55°	12°
17.	M	5.8	JIS	T	58°	42°	16°	6	26	50°	8°
18.	F	6.1	JIS	T	70°	48°	22°	6	26	61°	9°
19.	F	7.3	JIS	TL	68°	50°	18°	4	23	58°	10°
20.	F	4.7	NM	L	55°	38°	17°	5	25	45°	10°
21.	F	5.9	JIS	TL	58°	44°	14°	4	23	51°	7°
22.	M	4.1	Cong	T	49°	41°	8°	4	24	50°	-1°
23.	F	7.3	NM	T	70°	53°	17°	5	26	56°	14°
24.	F	5.6	NM	T	55°	43°	12°	5	24	50°	5°
25.	M	6.8	JIS	T	64°	51°	13°	5	26	59°	5°
26.	M	4.9	Cong	TL	49°	41°	8°	4	23	52°	-3°
27.	F	7.9	Syndrom	TL	60°	50°	10°	4	24	55°	5°
28.	F	4.5	Cong	T	44°	32°	12°	6	25	40°	4°
29.	F	5.2	NM	L	50°	°34	16°	6	26	42°	8°
30.	M	5.1	Syndrom	TL	53°	40°	13°	5	24	50°	3°
31.	F	6.1	JIS	T	51°	37°	14°	5	23	44°	7°
32.	F	4.9	Cong	T	40°	35°	5°	4	23	42°	-2°
33.	M	5.3	AG	TL	57°	46°	°11	4	24	52°	5°
34.	F	5.4	NM	TL	70°	48°	22°	5	25	58°	12°
35.	F	4.5	Cong	L	41°	34°	7°	4	23	40°	1°
36.	F	5.4	AG	L	44°	33°	11°	6	26	44°	0°
37.	F	5.0	NM	TL	71°	56°	15°	4	24	60°	11°
38.	M	6.1	JIS	TL	67°	47°	20°	5	24	54°	13°
39.	F	5,8	NM	TL	69°	54°	15°	5	25	58°	11°
40.	M	6.1	JIS	T	49°	32°	17°	4	23	36°	13°
Avg	-	5.6	-	-	56.6°	42.2°	14.4° (25.4%)	4.8	24.1	49.3°	7.3° (12%) P<0.001

Table 2. Results in our study: M: Male, F: Female, JIS: Juvenile Idiopathic Scoliosis,NM:Neuromuscular,Cong:Congenital,AG:Arthrogryposis,Syndrom: Syndromatic ,T: Thoracic, TL: ThoracoLumbar, L: Lumbarand Avg.: Average.

coronal T1-S1 spine height and sagittal T1 –S1 height at 2 years followup, we used the T test comparing two means: preoperatively and final result at two years follow up using Spass program, Successful result was defined if one of these 2 criteria was met:

The patient's scoliosis angles at 2-year follow-up was less than or at least equal to the patient's preoperative scoliosis angles.

The patient's trunk height or spinal length at 2-year follow-up was greater than or at least equal to the patient's immediate postoperative trunk height or spinal length. In contrast, failure result was defined if none of these criteria was met.

4. RESULTS

From the forty patients included in our study, twenty six (65%) were females and fourteen (35%) were female.

Their average age at the time of index surgery for implantation of the VEPTR device, was 5.4 years (4.1 to 7.9years) (Table 1). The causes of the scoliosis in our study group are summarized in Table 1.

The median of the angles of the curves in the pre- and immediate postoperative radiographs, as well as the postoperative correction of the coronal spine height as well as sagittal height, are shown in Table 2. The final Cobb angles, measured after the last distraction performed for each patient, together with the number of distractions and the follow-up time, are shown in Table 3. In this study, the average values in the preoperative, immediate postoperative, and following the last distraction at 2 years follow up were 56.6°, 42.2°, and 49.3° respectively. The patients in our study group underwent an average of 4.8 distractions (4-6) during an average of 24.1 months follow up (23-26 months), which is the time between index surgery of VEPTR implantation and the last distraction performed. The average initial correction in Cobb angle immediately after the index surgery was14.4° (5°-26°) and the average final correction of Cobb which is measured after the last expansion procedure (Cobb angle of the major curve measured after last expansion minus initial preoperative Cobb angle of the major curve) was 7.3° (12%). (Table 2).with a p value of less than 0.001 which is clinically significant.

The average of preoperative coronal T1-S1 length was 25.6 cm with an average initial correction achieved immediately after implantation of VEPTR of2.8 cm (1.2-5.1cm) which is 10.9%, and the average coronal length gain at 2 years follow up was 5.7 cm (3.7-9.8cm) that is 22.2%. (Table 3). with a p value of less than 0.001 which is clinically significant.

Complication occurred in 18 of our patients (45%): 10 patients(25%) had fractured rib and cut through of the proximal hock or ring managed by repositioning procedure not counted in the total number of distractions , 2 patients (5%) had migration of the iliac hock managed by repositioning of the hock and the procedure is not counted in distractions, 2(5%) patients had breakage of the railway end of the sliding bar, which managed by sliding bar exchange, 4 patients (10 %) had superficial infection and wound dehiscence: three of them managed and did very well, and unfortunately one of them developed deep infection which mandate removal of the implant, and this case was mot excluded from our study group because he was almost 23 months post index procedure when infection developed(Table 4) and (Figure 2).

It worth mentioning that none of our patients required blood transfusion, neither during the primary surgery nor during the subsequent expansion procedures.

Patient	Preop coronal length (T1-S1) (cm)	Immediate post op coronal length(cm)	Initial correc-tion(cm)	Coronal length at 2 years (cm)	Final coronal length at 2 years(cm)
1.	27.2	30.7	3.5	34.6	7.4
2.	24.3	28.0	3.7	29.5	5.2
3.	31.5	34.2	2.7	36.1	4.6
4.	26.7	29.6	2.9	31.4	4.7
5.	22.0	25.7	3.7	26.9	4.9
6.	30.7	32.3	1.6	36.0	5.3
7.	23.6	26.6	3	31.1	7.5
8.	33.8	35.8	2	38.3	4.5
9.	24.2	27.0	2.8	31.2	7
10.	31.1	32.9	1.8	34.8	3.7
11.	22.8	24.4	1.6	27.0	4.2
12.	26.0	30.1	4.1	33.3	7.3
13.	30.7	33.6	2.9	36.7	6
14.	24.5	26.9	2.4	30.1	5.6
15.	20.4	23.3	2.9	28.0	7.6
16.	19.0	23.4	4.4	25.9	6.9
17.	24.4	27.0	2.6	33.0	8.6
18.	28.5	30.5	2	34.7	6.2
19.	33.7	35.9	2.2	38.1	4.4
20.	26.8	29.8	3	32.1	5.3
21.	24.3	28.6	4.3	30.3	6
22.	18.9	20.8	1.9	24.3	5.4
23.	30.2	33.3	3.1	35.2	5
24.	24.4	27.0	2.6	30.3	5.9
25.	29.9	31.1	1.2	34.4	4.5
26.	19.3	22.9	3.6	23.8	4.5
27.	31.0	33.0	2	36.4	5.4
28.	20.4	22.4	2	26.1	5.7
29.	27.4	31.2	3.8	33.9	6.5
30.	26.2	30.5	4.3	32.4	6.2
31.	32.0	34.8	2.8	38.3	6.3
32.	20.3	22.2	1.9	25.6	5.3
33.	24.2	26.3	2.1	29.2	5
34.	22.9	23.8	0.9	25.5	2.6
35.	20.7	24.0	3.3	26.6	5.9
36.	24.4	25.9	1.5	28.7	4.3
37.	20.3	25.4	5.1	30.1	9.8
38.	24.6	26.8	2.2	28.8	4.2
39.	24.7	27.9	3.2	29.8	5.1
40.	27.3	30.2	2.9	34.2	6.9
Average	25.6	28.4	2.8 (10.9%)	31.3	5.7(22.2%) P<0.001

Table 3. Results in our study: changes in coronal length in our study group

complication	No. of patients
Rib fracture and cut through	10(25%)
Migration of iliac hock	2(5%)
Breakage of sliding bar rails	2(5%)
Infection	4(10%)

Table 4. Complications during follow up period.

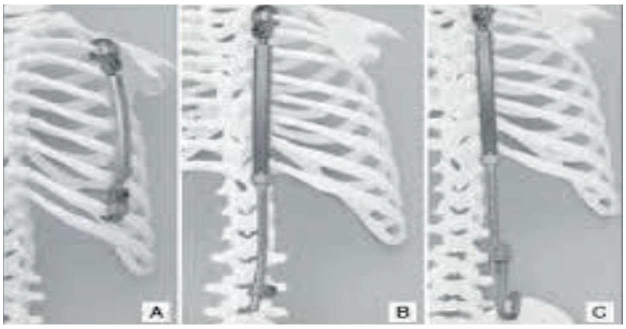

Figure 1. methods of VEPTR Implant: A: Rib To Rib, B: Rib To lumbar Spine and C: Rib to Pelvis (Source: VEPTR II–Vertical Expandable Prosthetic Titanium Rib II; Technique guide; Synthes Spine Co).

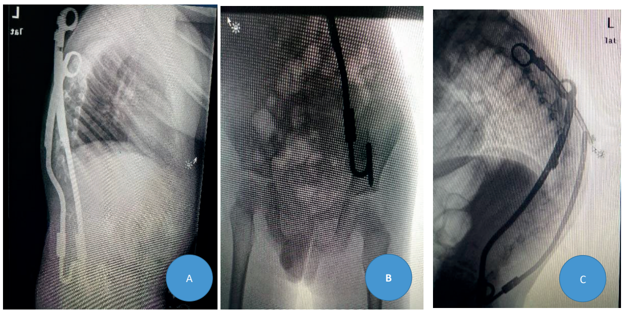

Figure 3. Complications: A-rib fracture cut through ,B-iliac hock migration,C-sliding bar breakage.

5. DISCUSSION

Scoliosisis not uncommon problem in Jordan, and one entity is Early Onset Scoliosis (EOS) which is defined as that starting prior to the age of 10 years (8, 10). EOS is a spinal deformity with different etiologies in the growing child. Management of such cases is challenging (2, 4). with limited treatment options, but whatever the treatment option is our aim is to avoid the problems associated with spinal fusion if done in this age group, and all treatment options aims to allow adequate development of the lung in an adequate lung space and to avoid the crankshaft phenomenon and the problems of dissociated growth of the trunk and the limbs. All options to manage these case is depending on the growing rod option to avoid the fusion of the spine until the lungs are well developed and the patient is reaching adolescence and adequate length.

VEPTR as a treatment option for these cases, It was initially conceived as a way to maximize thoracic volume in children with thoracic insufficiency syndrome, With its success in controlling these deformities (10, 11) its use was expanded to the correction of scoliotic deformities in younger patients by doing repetitive distractions performed in 4-6 months intervals until you reaches adequate growth of the trunk and skeletal maturity, and at that point the final spinal fusion is performed. Our case series is of40 patients, the average preoperative Cobb angle was56.6°, reached an average immediate postoperative Cobb anglewas42.2°, with an average initial correction of approximately 14.4°which is about 25.4% correction after VEPTR implantation, an average of 4.8 surgical distractions were performed in an average follow up period of 24.1 months, the average final Cobb angle was 49.3°, with an average correction of 7.3 ° or12%. It is clear that the initial correction achieved after implantation was greater than those at the end of the periodic distractions and in most patients there was less correction with follow up, especially for patients with congenital entity, which is not bad if you know that it is good to stop progression of the scoliotic curve mainly and achieving whatever correction you can until you perform the definitive spinal fusion at adulthood (9, 10).

Since the period of follow up in this study is considered to be short we need to conduct more studies about longer time of follow up to assess the maintenance of correction until adequate lung growth (1).

VEPTR has a high rate of complications (2, 6, 9).which are mainly mechanical and infectious, are similar to those resulting from the implantation of growing rods even in terms of the complication rate (1, 6). As already mentioned, in our study we had a complication rate of 45%, with the problems of the loosening of the implant due to rib fracture, migration of the hocks and infection of the surgical site being the most common. This rate of complication in our study matches the rates of complications for VEPTR worldwide: Hasler et al, this rate was 40%., Lucas et al.12 also observed a complication rate of 40% and In their study (1) the intraoperative planning of the site of the incision and the avoidance of incisions directly over the implant during implantation of VEPTR and in the following distractions, care with the molding of the implant to avoid sagittal imbalance and prominence of the metal, will definitely reduce the rate of complications (12).

6. CONCLUSION

Early Onset Scoliosis is a common problem worldwide, this deformity which occur before the age of 10 years can badly affect the development of the alveoli and function of the lung and heart as well, meanwhile spinal fusion surgeries at this age is not recommended because of the same negative effect on the lung and the length of the patient trunk and the balance of the spine.

Aiming to avoid those problems in skeletally immature patients, the Growing Rods is the most accepted method of treatment of such a cases, one known modality is Vertical Expandable Prosthetic Titanium Rib (VEPTR) which is based on rib anchor proximally and distal pelvic or spinal anchor with expandable mid part in a surgical procedure regularly aiming in keeping spine and thoracic cavity to grow until the adolescence.

This method according to our study and other studies showed good results in initial spinal deformity correction and in preventing the progression of the scoliotic deformity in growing children and maintaining acceptable final scoliotic deformity correction at the time of final spinal fusion.

Despite the high rate of complications associated with VEPTR, it can be avoided and reduced with good preoperative planning and good surgical techniques.

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