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DEFORMITY

Complications in Pediatric Spine Surgery Using the Vertical Expandable Prosthetic Titanium Rib

The French Experience

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Study Design. Multicenter retrospective study of 54 children.

Objective. To describe the complication rate of the French vertical expandable prosthetic titanium rib (VEPTR) series involving patients treated between August 2005 and January 2012.

Summary of Background Data. Congenital chest wall and spine deformities in children are complex entities. Most of the affected patients have severe scoliosis often associated with a thoracic deformity. Orthopedic treatment is generally ineffective, and surgical treatment is very challenging. These patients are good candidates for VEPTR expansion thoracoplasty. The aim of this study was to evaluate the potential complications of VEPTR surgery.

Methods. Of the 58 case files, 54 were available for analysis. The series involved 33 girls and 21 boys with a mean age of 7 years (range, 20 mo–14 yr and 2 mo) at primary VEPTR surgery. During the follow-up period, several complications occurred.

Results. Mean follow-up was 22.5 months (range, 6–64 mo). In total, 184 procedures were performed, including 56 VEPTR implantations, 98 expansions, and 30 nonscheduled procedures for different types of complications: mechanical complications (*i.e.*, fracture, device migration), device-related and infectious complications, neurological disorders, spine statics disturbances. Altogether, there were 74 complications in 54 patients: a complication rate of 137% per patient and 40% per surgery. Comparison of the complications in this series with those reported in the literature led the authors to suggest solutions that should help decrease their incidence.

Conclusion. The complication rate is consistent with that reported in the literature. Correct determination of the levels to be instrumented, preoperative improvement of nutritional status, and better evaluation of the preoperative and postoperative respiratory function are important factors in minimizing the potential complications of a technique that is used in weak patients with complex deformities.

Key words: congenital scoliosis, chest wall deformities, respiratory insufficiency, VEPTR, complications.

Level of Evidence: 4

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Treatment of progressive, congenital chest wall and spine deformities in children is a real challenge. This is because of the broad variety of causes and also to the fact that correct management of these difficult cases involves several factors: reduce the deformities or prevent the progression, improve function, and yet not compromise normal growth of the spine and the rib cage.

Very few technical solutions meet these requirements. Orthopedic treatment using a brace or a body cast has its utility and its limitations. Several techniques using distraction spinal instrumentation without fusion,[1–3] expandable thoracoplasty, or spine-to-rib-cage distraction[4,5] have been described. Studies on the results of growing rod techniques have been published; the main drawbacks of these techniques were the risk of premature spinal fusion and the difficulty to perform an arthrodesis at the end of growth.[6,7]

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The vertical expandable prosthetic titanium rib (VEPTR) device has been developed quite recently to address the above-mentioned problems.[8–13] As there is not much published material on VEPTR, we thought it would be of interest to review retrospectively all the children treated with VEPTR since the introduction of the device in France in 2005.

MATERIALS AND METHODS

This retrospective study is based on the results of the French VEPTR series involving patients treated between August 11, 2005, and January 5, 2012. Data were provided by 10 hospital teams.

As there were many different causes, each patient was a special case in itself. Each year, the surgeons from all the involved centers attend formal meetings during which indications are discussed.

Of the 58 case files, 54 were available for analysis. Four files were excluded from the study because of lack of information. The series involved 33 girls and 21 boys with a mean age of 7 years (range, 20 mo–14 yr and 2 mo) at primary VEPTR surgery. Mean follow-up was 22.5 months (range, 6–64 mo). Causes of thoracic and spinal deformities are presented in Table 1.

All the 54 patients were managed using the technique described by Campbell *et al*[9,13] with some modifications in specific cases. As regards the surgical approach, a midline incision was used in 20 patients, the pleura was stripped in 3 patients, and the VEPTR expansion thoracoplasty was associated with vertebral osteotomy for correction of kyphosis in 1 patient.

The number of devices and their respective position were determined on the basis of radiographs and sometimes on the basis computed tomographic scans (for localization of the constricted area of the hemithorax, fused ribs, and spinal deformity).

Twenty-two patients had previously undergone spine or rib cage surgery: distraction instrumentation had been used in 11 patients (growing rod, rib distractor or plating system for thoracic expansion); local fusions had been performed in 5 (epiphysiodesis, multilevel fusion, hemivertebra resection); arthrodesis associated with distraction instrumentation had been used in 3; and 3 patients had been operated for neurosurgical indications (medullary tumor in 2, and Chiari malformation type 1 in 1).

Evoked potentials were monitored during primary VEPTR surgery in 36 patients.

To compare our results with those reported in the literature, we have analyzed and summarized the data from 21 articles (entered in PubMed Central)[13–33] dealing with VEPTR complications (Table 2).

Because of the fact that a number of preoperative data were missing, the respiratory function and the growth of spine and rib cage could not be assessed in this study.

RESULTS

In total, 184 procedures have been performed, including: 56 VEPTR implantations (2 patients with Jeune syndrome received 2 devices in 2 stages), 98 expansions in 45 patients,

TABLE 1. Causes

Patient	Sex	Date of Birth	Cause
1	F	09/24/2004	Neurofibromatosis type 1 (Von Recklinghausen)
2	M	06/06/2003	Malformative: failure of segmentation and vertebral formation (hemivertebra and fused ribs, associated lumbar scoliosis)
3	F	07/02/2004	Malformative: failure of segmentation and vertebral formation (hemivertebra, unsegmented bar, abnormal rib number)
4	F	07/06/2000	Malformative: failure of segmentation and vertebral formation (unsegmented bar and block vertebrae)
5	M	11/08/2007	Malformative: failure of segmentation and vertebral formation (hemivertebra and puzzle-like thoracic vertebrae)
6	F	04/25/2005	Thoracogenic scoliosis: rib fusions secondary to pleuropulmonary disease
7	F	02/12/2004	Malformative: spondylocostal dysostosis
8	F	07/24/1998	Prader-Willi syndrome
9	F	10/23/2006	Malformative: failure of segmentation and vertebral formation (hemivertebra and rib fusion)
10	M	10/24/2007	Malformative: spondylocostal dysostosis
11	F	08/13/2004	Malformative: failure of segmentation and vertebral formation (hemivertebra and butterfly vertebra)
12	F	07/12/2001	Neuromuscular: LGMD2A (limb-girdle muscular dystrophy type 2A)
13	M	03/22/2003	22q11 microdeletion
14	F	07/02/1997	Thoracogenic scoliosis: rib fusions secondary to surgical treatment of diaphragmatic hernia
15	M	02/01/1992	Malformative: spondylocostal dysostosis
16	F	03/08/2005	Neurological: total absence of myelin sheaths (amyelinization of uncertain Cause)
17	F	06/28/2006	Neurological: grade 2 spinal cord oligodendroglioma. Flaccid paraplegia after surgical treatment

(*Continued*)

TABLE 1. (*Continued*)

Patient	Sex	Date of Birth	Cause
18	M	07/30/2003	AMC
19	F	08/04/1995	Juvenile scoliosis (left thoracic)
20	F	11/30/1999	Neurological: CP with axial hypotonia and spastic quadriplegia
21	M	11/16/1994	Thoracogenic scoliosis: rib fusions secondary to surgical treatment of diaphragmatic hernia
22	M	07/11/1999	AMC
23	M	11/27/2006	Malformative: failure of segmentation and vertebral formation (hemivertebra and fused ribs)
24	F	03/30/2001	Malformative: failure of segmentation and vertebral formation (chest wall breakdown, hemivertebra, fused ribs)
25	F	12/08/2005	Mixed: neurological and malformative (thoracolumbar block vertebrae and congenital myopathy related to *DPM2* gene mutation)
26	M	02/28/1999	VATER syndrome (with fused ribs)
27	F	11/14/2001	Malformative: failure of segmentation and vertebral formation (with fused ribs)
28	F	01/10/2005	Malformative: failure of segmentation and vertebral formation (hemivertebra and fused ribs)
29	F	04/02/1999	Neurological: CP
30	F	07/28/2000	Neurological: type II SMA
31	F	02/29/2004	Malformative: failure of segmentation and vertebral formation (block vertebrae)
32	F	01/29/2004	Neurological: left brachial plexus injury
33	M	05/15/2006	VATER syndrome
34	M	09/14/2006	Malformative: failure of segmentation and vertebral formation (puzzle-like vertebrae and rib fusion)
35	F	11/25/2004	Malformative: failure of segmentation and vertebral formation (puzzle-like vertebrae, hemivertebrae, unsegmented bar, fused ribs)
36	M	07/26/1998	AMC
37	F	01/11/2001	Malformative: spondylocostal dysostosis (tethered cord at T3–T4)
38	M	08/01/1997	Neurological: syringomyelia, Chiari Syndrome—type 1
39	M	12/06/2000	AMC
40	F	04/04/2005	Malformative: spondylocostal dysostosis
41	F	02/16/2000	Neurological: CP
42	M	09/01/1999	Jeune syndrome
43	F	07/31/2004	Malformative: spondylocostal dysostosis (with associated diastematomyelia or split cord malformation)
44	F	02/25/2005	Neurological: flaccid paraplegia secondary to excision of a neuroblastoma at T3-T9
45	M	06/03/2007	Jeune syndrome
46	F	03/20/2008	Malformative: failure of segmentation and vertebral formation (puzzle-like vertebrae, fused ribs)
47	F	01/04/2007	Progressive idiopathic scoliosis
48	M	01/04/2000	Infantile kyphoscoliosis
49	F	01/08/2007	Malformative: spondylocostal dysostosis
50	F	11/07/1998	Thoracogenic scoliosis: secondary to surgical treatment of diaphragmatic hernia
51	M	04/07/2001	Malformative: Klippel-Feil syndrome
52	M	05/22/2009	Malformative: Spondylocostal dysostosis
53	F	11/24/2000	Malformative: failure of segmentation and vertebral formation (puzzle-like thoracic vertebrae)
54	F	12/25/1998	Mixed: malformative (fused ribs and costal agenesis, vertebral malformation, chest wall defect) and neurological (myelomeningocele)

AMC indicates arthrogryposis multiplex congenita; CP, cerebral palsy; SMA, spinal muscular atrophy.

VATER indicates vertebral anal tracheal renal-radial syndroma

and 30 nonscheduled procedures for complications. The average number of expansion procedures was 1.8 per patient (range, 0–6). Expansions were performed at a mean interval of 10 months (range, 4–35 mo). Mean follow-up was 22.5 months (range, 6–64 mo).

TABLE 2. Table of the Meta-Analysis Results Relative to Each of the Sections of Complications

Reference	Fracture	Migration	Device-Related	Infection	Skin Lesion	Neurological	Statics	Other	Total No. of Complications
13 (27 patients)		11		3	4	3		1	22 (81.5% per patient)
14 (36 patients: 19 treated with VEPTR)	31			18		5		20	74 (45 in VEPTR group: 237%)
15	No figures (description of potential complications)								
16 (97 patients) dealing with infection only				19 (in 16 patients)					19.6% infection per patient
17 (23 patients)	5		2 failures	6	10		(6 not taken into account by the author	(5 not taken into account by the author)	23 (100% per patient) (148% per patient, altogether)
18 (37 patients)	19			11				3	33 (89% per patient)
19 (6 VEPTR)	3	1		1	1	2	1	1	10 (166% per patient)
20 (20 patients)		5	1	7		1		2	16 (80% per patient)
21 (26 patients)		5		12		9		10	36 (138% per patient)
22 (15 patients)		3		1					4 (26% per patient)
23 (9 VEPTR)		2				1			3 (33% per patient)
24 (299 patients) dealing with neurology only						8 (+6 changes in SEP)			2.7% neurological complications
25 (10 patients)	1							5	6 (60% per patient)
26 (24 patients)		12		1	1				14 (58% per patient)
27 (11 patients)		5		1					6 (54% per patient)
28 (14 patients)	3	7		4	1	1			16 (114% per patient)
29 (22 patients)		7		1					8 (36% per patient)
30 (31 patients)	2	11		3		5		3	24 (77.5% per patient)
31 (14 patients)	4	7		2			11		24 (171% per patient)
32 (43 patients) dealing with mortality and comorbidity only								8	8 (19% morbidity/mortality)
33 (12 patients)	4	5	2				1	6	18 (150% per patent)

VEPTR indicates vertical expandable prosthetic titanium rib.

TABLE 3. Characteristics of Complication-Free Patients

Patient	Pathology	Type of Construct	Number of Expansions	Follow-up (mo)	City
1	Neurofibromatosis type 1	2 VEPTR rib-to-pelvis	1	17	Besançon
5	Mosaic thoracic spine and hemivertebra T5	2 VEPTR rib-to-rib rib-to-lamina	1	6	Lille
7	Spondylocostal dysostosis	1 VEPTR rib-to-lamina	1	6	Brest
9	Hemivertebra and rib fusion	1 VEPTR rib-to-rib	4	36	Rennes
11	Failure of segmentation	1 VEPTR rib-to-lamina	1	10	Rennes
14	Thoracogenic scoliosis	2 VEPTR rib-to-rib rib-to-lamina	1	9 (death unrelated to VEPTR)	Nancy
15	Spondylocostal dysostosis	2 VEPTR rib-to-rib rib-to-lamina	3	64 (growth completed, hardware removal requested)	Nancy
17	Neurological (paraplegia after tumor resection)	2 VEPTR rib-to-pelvis	0	6	Nancy
23	Block vertebrae and fused ribs	1 VEPTR rib-to-lamina	2	33	Montpellier
24	Puzzle-like vertebrae rib fusion chest wall breakdown	2 VEPTR rib-to-rib rib-to-lamina	3	34	Toulouse
31	Block vertebrae	1 VEPTR rib-to-lamina	1	6	Toulouse
34	Puzzle-like thoracic vertebrae rib fusion	1 VEPTR rib-to-lamina	5	40	Toulouse
35	Puzzle-like thoracic vertebrae lumbar hemivertebra rib fusion	1 VEPTR rib-to-rib	1	20	Marseille
37	Spondylocostal dysostosis	1 VEPTR rib-to-rib	1	35	Marseille
41	CP	1 VEPTR rib-to-rib	1	20	Marseille
49	Spondylocostal dysostosis	1 VEPTR rib-to-lamina	0	6	Marseille
53	Costovertebral malformation	1 VEPTR rib-to-pelvis	0	24	Marseille
54	Costovertebral malformation fused ribs chest wall defect	1 VEPTR rib-to-rib	2	41 (arthrodesis scheduled 01/12)	Marseille

VEPTR indicates vertical expandable prosthetic titanium rib; CP, cerebral palsy.

Seventy instrumentations of varying types have been used in the 56 VEPTR procedures: 23 rib-to-rib constructs, 27 rib-to-lumbar lamina constructs, and 20 rib-to-ilium constructs). Thirty-eight patients received a standard construct using a single VEPTR device and 16 received 2 VEPTR devices (a bilateral rib-to-ilium VEPTR construct in 6 patients, a construct consisting of 1 rib-to-rib and 1 rib-to-lamina VEPTR in 7 patients, a construct consisting of 1 rib-to-lamina and 1 rib-to-ilium VEPTR in 1 patient, and a bilateral rib-to-rib construct in 2 patients).

The implants and attachment sites used (*i.e.*, rib, lamina, and iliac crest) depended on the chest wall and spine deformities encountered. As no 2 cases are alike, it was just impossible to design a standardized protocol. Nevertheless, all the centers involved used the same implants.

Seventy-four complications occurred in 36 patients, which means that one-third of the patients (18) in our series had no complication at all. Therefore, 66.7% of the patients experienced at least one complication (mean, 2.05 complications per patient) (Table 3).

Overall, we found a complication rate of 137% per patient and 40% per surgery.

Mechanical Complications

There were 9 proximal rib fractures and 3 lamina fractures in 11 patients (Tables 4, 5). Fractures occurred in 20.4% of the patients; the overall fracture rate was 22.2% per patient. There were 15 anchors migrations in 11 patients (20.4% of the patients). The overall migration rate was 27.8% per patient.

Device-Related Complications

Only one device-related complication occurred (1.8% of the patients), which resulted from jamming of the sliding mechanism (Table 6).

TABLE 4. Bone Fractures Due to VEPTR

Patient	Pathology	Fracture	Time Elapsed Between VEPTR Surgery and Fracture	Time Elapsed Since Last Expansion	Management
6	Thoracogenic scoliosis	Superior rib	Intraoperative		Nonabsorbable bone suture
10	Spondylocostal dysostosis	Superior rib	16 mo	8 mo	Repositioning during a nonscheduled procedure
13	22q11 microdeletion	Right superior rib	12 mo		Repositioning during a scheduled expansion procedure
		Right superior rib	44 mo	10 mo	VEPTR removal and fusion of T5–T10 (for junctional kyphosis)
18	AMC	Superior rib	9 mo		Ring placed in parallel alignment during a scheduled expansion procedure
30	Type II SMA	Superior rib	3.5 mo		Ring placed in parallel alignment during a nonscheduled procedure
39	AMC	Superior rib	Intraoperative		0
47	Congenital scoliosis	Superior rib	1 wk		VEPTR replaced by a growing rod
48	Infantile kyphoscoliosis	Superior rib	13 mo		Replacement during a scheduled expansion procedure
2	Failure of segmentation with fused ribs	Lamina	3.5 mo		Attached to another lamina during a nonscheduled procedure
20	CP	Lamina	2 mo		Replaced by an iliac hook during a nonscheduled procedure
33	VATER syndrome	Lamina	18 mo	4 mo	Attached to another lamina during a scheduled expansion procedure

VEPTR indicates vertical expandable prosthetic titanium rib; CP, cerebral palsy; SMA, spinal muscular atrophy; AMC, arthrogryposis multiplex congenita.

Operative Site Infection

Operative site infection and management are presented in Table 7. Eleven infections occurred in 6 patients (11% of the patients); in 7 cases, infection was bacteriologically documented. The infection rate was 20.3% per patient.

Noninfected Skin Lesions

Noninfected skin lesions and management are presented in Table 8. Nine noninfected skin lesions (skin slough and wound dehiscence) occurred in 7 patients (13% of the patients). The rate of skin lesions was 16.6% per patient. This complication was surgically managed in 89% of the cases.

Neurological Complications

Three of these complications involved the brachial plexus (see Supplemental Digital Content Table 9 available at http://links.lww.com/BRS/A836). The neurological complication rate was 5.5% per patient. During the 56 primary VEPTR procedures, evoked potentials were monitored in 36 patients (28 SEPs, 2 NMEPs, 6 combined SEPs/NMEPs). Intraoperative changes in evoked potentials occurred in 3 patients (see Supplemental Digital Content Table 10 available at http://links.lww.com/BRS/A836). There was no monitoring of evoked potentials in 17 patients. In another 2 patients, there was no monitoring of evoked potentials but a wake-up test was performed, with no deficit.

Spine Statics Disturbances

Eight static disorders were noted in 7 patients (4 in the sagittal plane, 2 in the frontal plane, and 2 cases of pelvic imbalance) (see Supplemental Digital Content Table 11 available at http://links.lww.com/BRS/A836). These complications occurred in 13% of the patients, with an imbalance rate of 14.8% per patient.

Other Complications

Other complications included intraoperative pneumothorax in 4 patients, 3 of which occurred during VEPTR placement and 1 during expansion (7.4% of the patients); 4 cases of respiratory insufficiency subsequent to VEPTR surgery: acute respiratory insufficiency in the immediate postoperative period in 3 patients, and chronic respiratory insufficiency resulting from decreased chest wall compliance (due to stiffness of the rib cage caused by VEPTRs) in 1 patient

TABLE 5. Anchors Migrations

Patient	Pathology	Location	Time Elapsed Since Anchor Fixation (mo)	Time Elapsed Since Last Expansion (mo)	Management
2	Failure of segmentation with fused ribs	Proximal rib	20	9	Fixation ring repositioned during a scheduled expansion procedure
		Proximal rib	18	11	VEPTR removal (sagittal imbalance)
4	Failure of segmentation	Lamina	24	11	Hook repositioned during a scheduled expansion procedure (migration caused dural tear)
13	22q11 microdeletion	Proximal rib	6		0
20	CP	Iliac hook	12	2	VEPTR removal (infection)
27	Failure of segmentation with fused ribs	Distal rib	47	9	Replaced by a lamina hook during a scheduled expansion procedure
28	Failure of segmentation with fused ribs	Distal rib	20	7	Replaced by a lamina hook during a scheduled expansion procedure
36	AMC	Lamina	4.5		Repositioning during a scheduled expansion procedure
		Lamina	14		Replaced by an iliac hook during a scheduled expansion procedure
38	Neurological scoliosis	Lamina	4		Repositioning during a scheduled expansion procedure (+ hook secured with bone suture)
		Lamina	13		Replaced by an iliac hook during a scheduled expansion procedure
		Ilium	3		VEPTR removal and posterior fusion of T2–L3
39	AMC	Proximal rib	12		VEPTR removal (infection of skin lesion caused by ring migration)
40	Spondylocostal dysostosis	Proximal rib	7		0
43	Spondylocostal dysostosis	Proximal rib	33	8	Fixation ring replaced during a nonscheduled procedure

VEPTR indicates vertical expandable prosthetic titanium rib; CP, cerebral palsy; AMC, arthrogryposis multiplex congenita.

(see Supplemental Digital Content Table 12 available at http://links.lww.com/BRS/A836). This complication occurred in 7.4% of the treated patients; 2 patients had a lung pathology, and another 2 patients developed a pleuropulmonary disease in the immediate postoperative period (altogether, 7.4% of the patients); 1 pleural fistula (1.8% of the patients) was revealed by an accumulation of sterile fluid under the pleural scar tissue 8 days after an expansion procedure; and 2 patients (3.7%) complained of scapular pain after VEPTR surgery.

Apart from the short follow-up period and the small number of expansions performed in these patients, the absence of complications could not be attributed to one or several specific factors (see Supplemental Digital Content Tables 13–15 available at http://links.lww.com/BRS/A836).

DISCUSSION

The learning curve required for each surgical team may indeed partly explain the high complication rate reported in this study. However, this rate has been stable during the whole period considered. This treatment method is only part of the surgical armamentarium to manage severe chest wall and spine deformities.

What follows is a discussion of what the surgeons and institutions in this study did to help reduce that rate of complications, but does not represent any statistically verifiable techniques, only a sharing of a series of "tips" which our group used in navigating through the care and surgery of these difficult patients.

Thereby, VEPTR surgery is associated with different types of complications. These are inherent both to the repetitive surgery approaches and to weakness of the patients who have significant comorbidities. The literature review[13–33] involved 776 patients with a variety of causes just like in our own series. In our study, the complication rate was 137% per patient (mean follow-up, 22.5 mo) and 40% per surgery.

TABLE 6. Device-Related Complications

Patient	Pathology	Complication	Time to Complication	Management
21	Thoracogenic scoliosis	Jamming of the sliding mechanism	7 mo	VEPTR removal

VEPTR indicates vertical expandable prosthetic titanium rib.

As regards mechanical complications, we had 22.2% fractures in our series *versus* 18.4% in the published studies.[14,19,25,28,30,33] A number of factors may contribute to increase the incidence of fracture: osteopenia, malnutrition, bone dysplasia, inappropriate construct with one single VEPTR device, hook that is not positioned perpendicular to the bone surface, severe kyphosis, excessive distraction force, or a pathology associated with severe hypotonia or extreme stiffness (as is the case in arthrogryposis multiplex congenita).

Hook migration is the most common complication associated with VEPTR surgery.[17,23,30,34,35] Obviously, the higher the loads and the longer a patient has an implanted device, the higher the likelihood of migration. In most cases, migration does not adversely affect function.[15] In an attempt to evaluate the migration risk per patient, Campbell and Smith[8] developed a "migration index" (total number of migrations per patient divided by years since implantation) that represented the risk per year of complete migration per patient. In their series, the migration index was 0.09 *versus* 0.15 in our own series. However, this comparison is somewhat biased because we have included incomplete migrations. Analysis of the data from the literature[13,14,17–23,26–31,33] shows a migration

TABLE 7. Operative Site Infection

Patient	Pathology	Mechanism	Germ	Time to Infection (mo)	Management
3	Failure of segmentation	After expansion, fluid accumulation in expansion area	*Staphylococcus aureus meti-S*	6	Lavage, surgical debridement, and antibiotics
		After routine biopsy during expansion procedure	*Staphylococcus aureus meti-S*	15	Antibiotics
		Wound dehiscence 1 mo after expansion procedure	?	33	Lavage, surgical debridement, and antibiotics
		Wound dehiscence, exposed hardware	?	34	VEPTR removal + antibiotics
20	CP	Skin breakdown at the device	*Corynebacterium striatum*	14	VEPTR removal + antibiotics
25	Mixed: congenital myopathy and failure of vertebral formation	Wound dehiscence, exposed hardware (noncontoured rod bulging under the skin)	*Staphylococcus aureus meti-S*	1	Lavage, surgical debridement, and antibiotics
		Local sepsis occurring within 5 mo of first lavage	*Staphylococcus coagulase*	5	Lavage, surgical debridement, and antibiotics
		Local sepsis occurring within 2 mo of second lavage	*Staphylococcus aureus meti-S*	6	Removal of 1 of the 2 VEPTRs + antibiotics
29	CP	Wound dehiscence, exposed hardware	*Staphylococcus aureus meti-S*	6	Removal of 1 of the 2 VEPTRs + antibiotics
39	AMC	Ring protrusion and skin lesion, exposed hardware	?	36	Rib hook removal + antibiotics
42	Jeune syndrome	Wound dehiscence, exposed hardware	?	29	VEPTR removal + antibiotics

VEPTR indicates vertical expandable prosthetic titanium rib; CP, cerebral palsy; AMC, arthrogryposis multiplex congenita; ?, no bacteriological identification.

TABLE 8. Noninfected Skin Lesions

Patient	Pathology	Skin Lesion	Time to Lesion (mo)	Management
3	Failure of segmentation	Wound dehiscence	7	Flap coverage
		Wound dehiscence	21	Surgical scar revision
		Wound dehiscence	32	Flap coverage
13	22q11 microdeletion	Wound dehiscence (secondary to sterile hematoma)	62	Surgical scar revision
22	AMC	Skin breakdown, wound dehiscence	1	VAC followed by a sliding flap
29	Encephalopathy	Skin breakdown at the device	6	VAC followed by a sliding flap
42	Jeune syndrome	Skin breakdown at the device	34	Left VEPTR removal
43	Spondylocostal dysostosis	Skin breakdown at the device with brace on	1	Local wound care
46	Failure of segmentation with fused ribs	Skin breakdown at ring	3	Ring removal

VEPTR indicates vertical expandable prosthetic titanium rib; AMC, arthrogryposis multiplex congenita.

rate of 39% per patient. This is much closer to our own results (27.8%) that are based on a mean follow-up of only 22.5 months.

Device-related complications are rare. In our series, there was one single device-related complication (1.8% of the patients) that resulted from jamming of the sliding mechanism. The results reported in the literature[14,17,20] are less favorable, with a device-related complication rate of 9% per patient.

Infection and skin lesions are frequent VEPTR complications. Analysis of the data from the literature[13,14,16–22,26–31] shows an infection rate of 22.3% per patient. Smith and Smith conducted a study on VEPTR infections[16] in which they reported an infection rate of 19.6% per patient and 2.8% per surgery. In our series, the VEPTR infection rate was 20.3% per patient and 6% per surgery. But, in our series, only 11% of the patients had VEPTR infection *versus* 14.7% in the literature[13,14,16,17,19,22,26–31] and 16.5% in the Smith series.[16]

Noninfected skin lesions also are quite common in this type of surgery. The results reported in the literature show a 17.8% rate per patient[13,16,17,19,26,28] *versus* 16.6% in our series. In 89% of the patients, this complication was surgically managed (using flaps in one-third of the cases).

In the literature, 6.6% of the patients have been reported to have neurological complications,[14,19,21,23,24,28,30] including brachial plexus injury (2.6%), Horner syndrome (0.7%), lower extremity neurological injury (1.2%), pain, or neurological deficit in unspecified territory (2.1%). In our series, there were 5.5% neurological complications, all of which involved essentially the brachial plexus. Brachial plexus injuries may result from nerve compression occurring in the costoclavicular region of the thoracic outlet (between the first rib and the clavicle)[36] or from direct trauma sustained during VEPTR placement. Spinal cord injuries or damage to the dura mater may occur when treating rib fusions by a release that extends to the spine, or during lamina hook placement.

Skaggs *et al*[24] evaluated the efficacy of neurological monitoring in VEPTR surgery in 299 patients. Results clearly showed that the incidence of potential complications (proven neurological complications and intraoperative changes in evoked potentials) warranted intraoperative neurological monitoring of upper and lower extremities during VEPTR placement or replacement surgery and not during expansion procedures (except in patients who had changes in evoked potentials during primary VEPTR surgery).

Regarding spine statics disturbances, VEPTR has been shown to reduce lumbar lordosis when the distal anchor was placed in a more distal position. On the contrary, it preserves thoracic kyphosis,[15] but sequential expansions may result in increased kyphotic curve due to the increasing radius of curvature of the rods. Upper thoracic hyperkyphosis cannot possibly be corrected by VEPTR because no anchor can be placed proximal to the second rib. Lower hyperkyphosis is correctable by using a construct with a significant lever arm. Pelvic imbalance and frontal plane disorders are essentially due to an asymmetrical construct. Therefore, they can be easily corrected by modifying the initial construct. In our series, 14.8% of the patients had spine statics disturbances. The results reported in the literature show worsening of upper thoracic kyphosis or development of junctional kyphosis in 13.8% of the patients,[17,19] frontal imbalance in 8.7%,[17] and loss of pelvic stability in 12.5%.[17,18,33]

Additional complications that were not found in our series have been reported in the literature in isolated cases: ectopic bone formation, allergy to device material, costovertebral dislocation, and esophageal rupture[37] (multiple reoperations for esophageal atresia ended up in rupture of the esophagus during VEPTR expansion thoracoplasty).

What Can Be Done to Minimize the Incidence of Such Complications?

First of all, we recommend that this surgery and use of this device should ideally be performed only by surgeons who have training and experience with this instrumentation and performed in centers that also have the experience and facilities to manage these cases and all the possible complications.

Preoperatively, a multidisciplinary care team should evaluate the child's nutritional status and lung function. If it needs to be improved prior to surgical treatment, gastrostomy and noninvasive ventilation can be performed. Magnetic resonance image and computed tomographic scan will allow detection of any medullary and/or vertebral anomaly.

Intraoperatively, make a short skin incision. The incision of the deep muscle layer should not be performed in line with the skin incision but should be slightly offset.[15] In expansion procedures, making an incision that is not straight above the hardware avoids damage to the soft tissue cover.[9,11,13] Therefore, a single midline incision can be used, as is done in several of the centers involved in this study. A midline incision has 2 major advantages: it minimizes the number of scars (but this implies raising a significant musculocutaneous flap), and it allows reuse of the incision in cases where posterior arthrodesis will be performed at the end of growth. Select a rib at least 1-cm thick, and position the anchors perpendicular to the bone surface (as described by Campbell *et al*[9,11,13]), being careful to preserve a musculoperiosteal flap between the device and the rib so as to prevent migration. Secure the hook by placing resorbable suture thread or a bone graft around the laminar hook.[9,11,13] Reduce the loads on the anchors as much as possible.

To avoid direct injury to the brachial plexus, the superior rib hook should not be positioned proximal to the second rib and one should stay medial to the distal insertions of the scalenus medius and posterior muscles (the "safe zone" according to Campbell).[13] To make sure not to compress the brachial plexus by traction on the scapula when closing, evoked potentials must be checked particularly in the ulnar nerve, as well as the oxygen saturation reading obtained by pulse oximetry of the ipsilateral finger, for signs of vessel compression.[13] Monitoring of evoked potentials during VEPTR placement or replacement is critical to prevent neurological complications.[24]

Where long constructs are used, the distal rod can be contoured to preserve the lumbar lordosis. A long bilateral construct with a significant lever arm can improve a lower thoracic hyperkyphosis. Changing the sizes of the rods can correct a thoracic hyperkyphosis that is because of the increased radius of curvature of the rods. Pelvic imbalance and frontal plane disorders can be easily corrected by using several VEPTR devices and adjusting their expansion individually. Imbalance can also be because of a VEPTR losing its efficacy because ribs become fused again; therefore, during expansion procedures, it is important to iteratively seek and resect any fused ribs.[30]

Closure is performed in the normal layered fashion, taking care to achieve good soft tissue coverage, which is critical for VEPTR success. If necessary, one can use the "stretching maneuver" described by Campbell: it consists in applying manual traction on the musculocutaneous flap for a few minutes.[13] Gill *et al*[38] suggest the use of an acellular dermal matrix for prevention and treatment of exposed VEPTR hardware secondary to wound breakdown.

Postoperatively, place a protective device or covering over the VEPTR implant to avoid any injury to the skin. Bracing is not recommended.[13] The role of a multidisciplinary care team is critical, particularly in the resuscitation unit for monitoring of the respiratory function.

We are aware of the fact that the short follow-up period (22.5 mo) in a very young patient population is a weak point in this study. Obviously, another study with a much longer follow-up will have to be performed, when a large number of patients with VEPTR have reached skeletal maturity and are old enough to have a surgical fusion. Finally, we have also warned surgeons against the temptation to follow any new attractive trend.

CONCLUSION

The high incidence of complications noted in this study must be weighed against that of other treatments or the natural progression of these deformities (often very severe) when left untreated. Furthermore, our rate is consistent with that reported in the literature. However, familiarity with the technique on the one hand, and both pre- and postoperative management by a multidisciplinary team on the other are likely to significantly reduce the complication rate. Working in team with anesthesiologists, pediatric pneumologists, and dieticians is indispensable. Preoperatively, the focus must be on improvement of nutrition and lung function in these often weak patients.

➢ Key Points

- ❑ Congenital chest wall and spine deformities in children are complex entities. They result in thoracic insufficiency syndrome that is defined as the inability of the thorax to support normal respiration and lung growth.
- ❑ Orthopedic treatment is generally ineffective.
- ❑ VEPTR been developed to address the treatment of these patients.
- ❑ The incidence of complications must be weighed against that of other treatments or the natural progression of these deformities when left untreated.
- ❑ Evaluations of the preoperative and postoperative respiratory function are important factors in minimizing complications rate.

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