

LABELING

Synthes Spine

DRAFT PACKAGE INSET

**IMPORTANT INFORMATION
ON THE SYNTHES VERTICAL EXPANDABLE
PROSTHETIC TITANIUM RIB SYSTEM**

4/04

GPxxxx-A, Rev1

IMPORTANT: Prior to use, the physician should be trained in the surgical procedure recommended for the use of this device.

Humanitarian Device: The Vertebral Expandable Prosthetic Titanium Rib (VEPTR) is authorized by Federal law for use in the treatment of Thoracic Insufficiency Syndrome, the inability of the thorax to support normal respiration or lung growth, in skeletally immature patients. The effectiveness of this device for this use has not been demonstrated.

CAUTION: U.S. federal law restricts this device to sale by or on the order of physician with appropriate training or experience.

INDICATIONS FOR USE

The VEPTR is indicated for treatment of Thoracic Insufficiency Syndrome in skeletally immature patients. Thoracic Insufficiency Syndrome is defined as the inability of the thorax to support normal respiration or lung growth. Thoracic Insufficiency Syndrome is considered to be a rare condition.

For the purpose of identifying potential Thoracic Insufficiency Syndrome patients, the categories in which Thoracic Insufficiency Syndrome patients often fall are as follows:

- Flail Chest Syndrome
- Rib fusion and scoliosis
- Hypoplastic thorax syndrome, including, but not limited to:
 - Jeune's syndrome
 - Achondroplasia
 - Jarcho-Levin syndrome
 - Ellis van Creveld syndrome

DESCRIPTION

The VEPTR devices are attached perpendicularly to the subject's natural ribs and lumbar vertebra or hip. This mechanically stabilizes the chest wall and enlarges the thorax to improve respiration and lung growth. Once the VEPTR is in place, its design allows for expansion, anatomic distraction, and replacement of component parts through less

invasive surgery. The VEPTR device is comprised of a combination of the following titanium component(s):

- Superior Cradle
- Inferior Cradle
- Cradle End Half
- Extended Cradle End Half
- Rib Sleeve
- Cradle Lock
- Distraction Lock
- Lumbar Extension
- Low Profile Lamina Hook
- Sacra Ala Hook
- Connector
- 2mm Ti Rod

The VEPTR is available for assembly in three configurations

- Cradle to Cradle
- Cradle with Lumbar Extension
- Cradle to Ala Hook

The Cradle to Cradle Assembly is available in two different radii (approximately 220mm radius and 70mm radius of curvature) and various lengths. The other two assemblies are available in 220mm radius only. The semicircular end of the inferior and superior cradles are provided in different angles (0°, 30° right, and 30° left) to accommodate subject anatomy, and are connected to the cradle end half by a cradle lock. These portions of the construct encase the natural rib(s). The cross-sections of the proximal ends of the rib cradles are “T-shaped” for enhanced strength. The superior cradle and inferior cradle/lumbar extension attach to the rib sleeve by inserting a distraction lock at each end. The rib sleeve is the central section of the construct. The rib sleeve serves as a track into which the cradles slide. A through hole in the rib sleeve is aligned with one of the blind holes on the superior and inferior cradle. The distraction locks are inserted into aligned sets to holes at both ends of the construct. The position of the inferior cradle assembly along the rib sleeve depends on the desired length of the overall rib prosthesis construct. All of these configurations are required to accommodate the wide variety of anatomical deformities encountered by the clinician in treating Thoracic Insufficiency Syndrome patients.

Cradle to Cradle Assembly

The Cradle to Cradle Assembly consists of 9 pieces: a superior cradle, an inferior cradle, a rib sleeve, two cradle end halves, two cradle locks and two distraction locks. These assemblies are available in 70mm and 220mm radii versions. The Cradle to Cradle

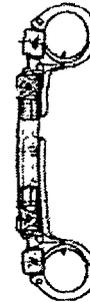
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Assembly is normally used when the patient has Thoracic Insufficiency Syndrome due to fused/missing ribs, severe scoliosis, and/or hypoplastic thorax.

Cradle to Cradle Assembly
70 mm Radius

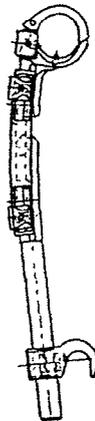


Cradle to Cradle Assembly
220 mm Radius

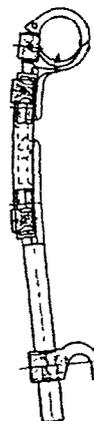


Cradle with Lumbar Extension Assembly

The Cradle with Lumbar Extension Assembly consists of 8 pieces: a superior cradle, a rib sleeve, a lumbar extension, a cradle end half, a cradle lock, two distraction locks and a low profile lamina hook. This is available in the 220mm radius version only. The Cradle with Lumbar Extension Assembly is indicated for use where no lower ribs exist or when the scoliotic curve extends into the lumbar region of the spine.



Lumbar
220mm



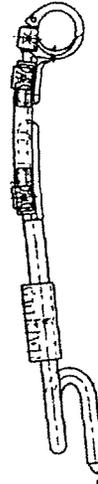
Extension Assembly
Radius Only

Cradle to Ala Hook Assembly

The Cradle to Ala Hook Assembly consists of 9 pieces: a superior cradle, a rib sleeve, a lumbar extension, a cradle end half, a cradle lock, two distraction locks, an extension connector and an ala hook. The assembly is available in 220mm radius only. The Cradle

to Ala Hook Assembly is indicated for use when attachment of the lower portion of the assembly to the hip is necessary.

Ala Hook Assembly
220mm Radius Only



The superior and inferior cradles are available in 0 degree, 30 degree right and 30 degree left configurations to suit the anatomy. The inferior cradles and rib sleeves come in 10 lengths for the 220mm radius version and 8 lengths for the 70mm radius version. The lumbar extensions are available in 8 lengths to allow for anatomical variations and device expansion to accommodate patient growth. The ala hooks and the low profile lamina hooks are available in left and right hand versions to allow placement on the appropriate side of the spine. The cradle end half is available in two configurations: 1) the standard version which is capable of capturing 1 rib and 2) the extended version which can capture 2 or more ribs.

All components are manufactured from titanium alloy, Ti-6Al-7Nb (ASTM F1295), with the exception of the sacral ala hook and 2mm rod, which are manufactured from commercially pure titanium, TiCP4 and TiCP1, (ASTM F67), respectively. These materials have a long history of safe use as an implantable material.

Associated manual instrumentation utilized for the implantation of these components is available for the distraction, insertion, expansion and removal of the VEPTR.

CONTRAINDICATIONS

The VEPTR device should not be used under the following conditions:

- Inadequate strength of bone (ribs/spine) for attachment of the VEPTR
- Absence of proximal and distal ribs for attachment of the VEPTR
- Absent diaphragmatic function
- Inadequate soft tissue for coverage of the VEPTR

- Age beyond skeletal maturity for uses of the VEPTR
- Age below 6 months
- Known allergy to any of the device materials
- Infection at the operative site

WARNINGS AND PRECAUTIONS

Humanitarian Device: Authorized by Federal law for use in the treatment of Thoracic Insufficiency Syndrome in skeletally immature patients. The effectiveness of this device has not been demonstrated.

Patients implanted with the VEPTR should not be braced. The VEPTR device is designed to allow for thoracic cavity growth and the restrictive nature of a brace would not help the condition, but defeat its purpose.

Patients may require additional wound protection to prevent inadvertent rubbing or bumping of the wound.

Patients with a diagnosis of spina bifida should have an occlusive dressing over the wound site to keep the site dry.

ADVERSE EFFECTS OF THE DEVICE ON HEALTH

OBSERVED ADVERSE EVENTS

Adverse effects were reported on the study Case Report Form (CRF) or directly to Synthes by telephone or monitoring report in advance of completion of the CRF. This application contains all adverse effects known to Synthes, from the time the device was first implanted in April 1989 to July 2003.

Treatment-emergent events were reported for the entire length of study follow-up (mean follow-up for the feasibility study was 84.8 months, mean follow-up for the multicenter study was 22.6 months). A total of 29 (88%) of 33 patients reported a total of 182 adverse effects while a total of 68 adverse effects considered to be related to the device were reported in 21 (64%) patients. Eleven (11) adverse effects of life-threatening intensity were reported for 2 patients and 13 adverse effects of fatal intensity were reported for 4 patients. These adverse effects were not considered to be device-related by the investigators.

A total of 119 (56%) of 214 patients in the multicenter study reported a total of 356 adverse effects while a total of 141 adverse effects considered to be related to the device were reported in 71 (33%) patients. Twelve (12) adverse effects of life-threatening intensity were reported for 9 patients and 16 adverse effects of fatal intensity were reported for 8 patients. These adverse effects were not considered to be device-related by the investigators.

Most of the device related adverse effects were device migrations. Device migrations included asymptomatic migrations through the proximal ribs, hook migrations distally through the vertebral lamina and also includes dislodgements of the spinal hooks. The migration is a localized “disattachment” from bony points of attachment to the spine or proximal ribs and not a migration inside the chest. Many of the device migrations occurred after subjects had been implanted with the VEPTR for two or more years.

SUMMARY OF CLINICAL INVESTIGATIONS

Objective

A prospective, multi-center clinical trial of the VEPTR device was conducted in the United States to determine the safety and effectiveness of the device in the treatment of TIS. All patients enrolled into the study were treated with the VEPTR device and served as their own controls.

Inclusion and Exclusion Criteria

Eligible patients had a primary diagnosis of TIS with a thoracodorsal malformation classified in one of the following categories:

Category I: Flail Chest Syndrome, including congenital chest wall defect, acquired surgical chest wall deficit due to tumor resection, surgical separation of conjoined twins, traumatic flail chest.

Category II: Congenital Constrictive Chest Wall Syndrome, including rib fusion or hypoplastic thorax syndrome; rib fusion with progressive thoracic scoliosis without vertebral anomalies; rib fusion with secondary chest wall constriction by progressive thoracic congenital scoliosis; hypoplastic thorax syndrome; Jeune’s syndrome (asphyxiating thoracic dystrophy); achondroplasia; Jarcho-Levin syndrome (lethal autosomal short-trunk dwarfism); Ellis van Creveld syndrome (chondroectodermal dysplasia).

Category III: Progressive spinal deformity (scoliosis or kyphosis) without rib anomaly, e.g., progressive scoliosis of neurogenic or congenital origin.

Patients were 6 months of age or older, up to the age of skeletal maturity, depending on the diagnostic category.

Clinical Trial Design

This was a prospective, single-treatment arm study conducted in two phases: a single investigational site feasibility study and a multi-center pivotal trial.

Patient Population and Demographics

A single site feasibility study with thirty three (33) patients and a multi-center, prospective study at seven (7) sites with two hundred twenty four (224) patients were performed. A total of two hundred fifty seven (257) patients were studied, but ten (10)

patients were excluded from the analysis due to the absence of baseline data. Enrolled patients at each site received the VEPTR device assembly according to disease pathology and anatomical requirements. For the purposes of reporting the results, the study population was divided into four diagnostic categories: Flail Chest, Rib Fusion, Hypoplastic Thoracic Syndrome, and Progressive Scoliosis.

Study Phase	Diagnostic Category				Total
	Flail Chest	Rib Fusion	Hypoplastic Thoracic Syndrome	Progressive Scoliosis	
Feasibility	6	19	6	2	33
Multi-Center	8	75	87	44	214
Totals	14	94	93	46	247

	Diagnostic Category				Total
	Flail Chest	Rib Fusion	Hypoplastic Thoracic Syndrome	Progressive Scoliosis	
N	14	94	93	46	247
Male (%)	8 (57.1%)	49 (52.1%)	43 (46.2%)	24 (52.2%)	124 (50.2%)
Female (%)	6 (42.9%)	45 (47.9%)	50 (53.8%)	22 (47.8%)	123 (49.8%)
Age, mean (years)	5.1	3.7	3.3	5.2	3.9
Age range (years)	0.0-15.0	0.0-14.0	0.0-15.0	0.0-12.0	0.0-15.0

Evaluation Schedule

Clinical examinations were performed at each surgical procedure and at the post-operative follow-up visits at 4 months (± 2 months), 8 months (± 2 months), 12 months (± 2 months), 16 months (± 2 months), 20 months (± 2 months), 24 months (± 4 months), and annually thereafter (± 4 months). At each follow-up visit, patients had general physical examinations, measurements of sitting and standing height, chest and abdominal circumference (inspiration and expiration), vital signs, weight, Assisted Ventilation Rating (AVR) (an outcome measure specifically developed for this investigation), Quality of Life Assessment (QOL) (Child Health Questionnaire for children ≥ 5 years, or Infant/Toddler Health Questionnaire for children < 5 years), capillary blood gases, oxygen saturation (pulse oximeter), pulmonary function tests (in children > 7 years without

developmental delay), and radiographs (for measurements of thoracic dimensions and Cobb angles).

As the patients experienced normal growth and/or as the spine and thorax required further correction, the study device would require expansions or replacement of the components to increase the overall size of the device. As a guideline, children with scoliosis or flail chest syndrome were to be scheduled for expansion of the device when the Cobb angle increased by 5° or greater. Children with hypoplastic thoracic syndrome were to be scheduled for device expansion approximately every 6 months.

Surgical Procedures

After the initial VEPTR surgical procedure, patients were expected to undergo multiple surgical procedures to expand, replace and remove the VEPTR as part of the normal course of treatment in order to further correct chest wall deformities and accommodate for growth. For the 214 Multi-Center patients, there were 1,051 follow-up surgical procedures, an average of nearly 5 follow-up surgeries per patient. Approximately 75% of these subsequent surgeries were device expansions.

	Diagnostic Category				Total
	Flail Chest	Rib Fusion	Hypoplastic Thoracic Syndrome	Progressive Scoliosis	
Multi-Center, n	8	75	87	44	214
Total procedures (%)	26	339	592	94	1051
Expansion (%)	19 (73.1)	253 (74.6)	442 (74.7)	71 (75.5)	785 (74.7)
Replacement (%)	0	49 (14.5)	78 (13.2)	14 (14.9)	141 (13.4)
Removal (%)	3 (11.5)	9 (2.7)	2 (0.3)	2 (2.1)	16 (1.5)
Other (%)*	4 (15.4)	28 (8.3)	70 (11.8)	7 (7.4)	109 (10.4)

* Other surgical procedures include device re-seating or repositioning, partial or total removals, revision of components, and implantation of extensions or additional components, wound debridements, drainages, delayed wound closures, incision and drainages, and dressing changes, non-orthopedic procedures including aspiration of pleural effusions, lymph node biopsies, suture removals, tracheotomy closure, laryngoscopy, Porta Cath insertion, inguinal hernia repair, pulmonary lobectomy, bronchoscopy, and gastric tube replacement procedures.

Patient Accountability

There were 33 patients enrolled in the feasibility study and 224 patients enrolled in the multi-center study. Data from ten patients were not available at the time of database closure and were not included in the analysis. Thus, 214 patients from the multi-center study were analyzed.

Of the 247 patients enrolled in either study, 12 patients died and 2 patients withdrew, leaving 233 patients. Within one year of database closure, 215 patients had evaluations, 5 were lost to follow-up, 5 were seen at other study sites after database closure, 3 were seen at an IRB-suspended site, 3 did not require further surgery, 1 lived in New Zealand, and 1 was transferred to another site.

For the feasibility study, the 2-year, 3-year, and 5-year follow-up rates for those time points or greater were 93.5%, 96.6%, and 89.7%, respectively, and for the multi-center study, 85.7%, 95.8%, and 95.0%, respectively.

Effectiveness Data

- **Assisted Ventilatory Rating (AVR) Outcomes**

Standard pulmonary function test measurements, such as *forced expiratory volume* (FEV), *maximal voluntary ventilation* (MVV), *residual volume* (RV), and *total lung capacity* (TLC), were not feasible in this population because most patients were less than 7 years old and/or developmentally delayed and were unable to follow directions required for these tests. Therefore, the Assisted Ventilatory Rating (AVR) was used to assess treatment effectiveness. AVR outcomes were determined relative to preoperative baseline score. AVR scores were defined as follows:

- +0: room air
- +1: supplemental oxygen
- +2: night ventilation
- +3: part-time ventilation or CPAP
- +4: full-time ventilation

The AVR outcomes demonstrated improvement or stabilization in 84.4% of patients for the feasibility study and 93.4% of patients for the multi-center study, or 92.0% of patients overall. Each of the diagnostic categories demonstrated improvement or stabilization AVR outcomes.

Table 4					
AVR Outcomes					
	Diagnostic Category				Total
	Flail Chest	Rib Fusion	Hypoplastic Thoracic Syndrome	Progressive Scoliosis	
Feasibility	3 (50.0)	17 (94.4)	5 (83.3)	2 (100.0)	27 (84.4)
Multi-Center	7 (100.0)	62 (92.5)	71 (91.0)	29 (100.0)	169

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					(93.4)
Combined	10 (76.9)	79 (92.9)	76 (90.5)	31 (100.0)	196 (92.0)

- **Thoracic dimensions**

The goal of treatment with VEPTR was to equilibrate the height of each individual hemithorax and maintain this correction with each expansion of the devices. Table 5 shows the number and percentage of the subjects who met this goal of treatment, allowing growth of the thoracic spine and increase in the hemithoracic height and volume.

- **Cobb Angle**

The Cobb angle is a measurement of the patient's spinal curvature. A decrease in Cobb angle represents an improvement. For this study, maintenance was defined as stabilization (± 5 degree change from baseline) or improvement (>5 degree reduction from baseline) of the Cobb angle. The Cobb Angle outcomes for this study ranged from are noted in Table 5.

	Flail Chest	Rib Fusion	Hypoplastic Thoracic Syndrome	Progressive Scoliosis
Multi-Center, n	8	75	87	44
Thoracic Ht Outcomes	4 (80.0)	54 (91.5)	56 (84.8)	24 (77.4)
Hemithoracic Ht (Initial Side) Outcomes	4 (80.0)	50 (86.2)	58 (90.6)	23 (88.5)
Hemithoracic (Secondary Side) Ht Outcomes	3 (60.0)	42 (72.4)	52 (80.0)	16 (59.3)
Hemithoracic Width (Initial Side) Outcomes	3 (60.0)	48 (81.4)	54 (83.1)	20 (74.1)
Hemithoracic Width (Secondary Side) Outcomes	4 (80.0)	45 (76.3)	53 (81.5)	15 (53.6)
Cobb Angle Outcomes	5 (100.0)	51 (83.6)	47 (73.4)	24 (80.0)

Safety Analysis

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Twenty-nine of 33 patients in the feasibility study had 408 adverse effects, while 119 of 214 patients (56%) in the multicenter study had 1,051 adverse effects. Respiratory problems such as pneumonia and dyspnea and other conditions, such as fevers, were frequently encountered during the study. These adverse effects are categorized into the following groups:

	Feasibility		Multi-Center	
	Events¹	Patients²	Events¹	Patients²
	408	33	1051	214
Device-Specific	37	16 (48%)	52	34 (16%)
-Device Migration	25	14 (42%)	49	34 (16%)
-Device Failure	13	7 (21%)	6	5 (2%)
-Device Other	1	1 (3%)		
Body as a Whole	9	5 (15%)	47	29 (14%)
-Abscess	1	1 (3%)	13	8 (4%)
-Infection	4	2 (6%)	11	10 (5%)
-Infection, bacterial			4	4 (2%)
-Infection, fungal			2	2 (1%)
-Pain			6	5 (2%)
-Cellulitis	1	1 (3%)		
-Pain, headache	1	1 (3%)		
-Pain, back	1	1 (3%)	2	2 (1%)
-Pain, chest			1	1 (0%)
-Pain, neck			1	1 (0%)
-Fever	1	1 (3%)	3	3 (1%)
-Injury			3	3 (1%)
-Necrosis			1	1 (0%)
Respiratory			16	11 (5%)
-Pneumonia			6	6 (3%)
-Effusion			3	3 (1%)
-Pneumothorax			3	3 (1%)
-Atelectasis			1	1 (0%)
-Respiratory Disorder			1	1 (0%)
-Respiratory Distress Syndrome			1	1 (0%)
-Acidosis, respiratory			1	1 (0%)
Skin and Appendages			13	7 (3%)
-Dermatitis			11	5 (2%)
-Healing, abnormal			1	1 (0%)
-Rash, vesicular			1	1 (0%)
-Injury, accidental	1	1 (3%)		
Metabolic and Nutritional	5	3 (9%)	4	4 (2%)
-Healing, abnormal	5	3 (9%)	4	4 (2%)

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Nervous			4	3 (1%)
-Neuropathy	1	1 (3%)		
-Transient Spinal Cord	1	1 (3%)		
-Convulsion			1	1 (0%)
-Hypokinesia			1	1 (0%)
Urogenital	1	1 (3%)		
-Infection, urinary tract	1	1 (3%)		

1 Number of individual adverse events

2 Number of patients experiencing an adverse event

There were 4 intraoperative complications reported for the feasibility and multi-center studies (1.9% of all patients), including a technical error in device placement; a dural laceration, and pressure on the brachial nerve. Sixteen feasibility patients, or 48%, experienced 37 device-specific adverse events, and 34 multi-center patients, or 16%, experienced 52 device-specific adverse events. These device-specific adverse events included device fractures, device migrations, and other device-related adverse events. Device migrations occurred frequently—25 migrations in the feasibility study and 49 migrations in the multi-center study. They were more common with the cradle-to-lumbar extension and cradle-to-sacral ala because these two configurations undergo greater flexion, extension, rotation and lateral bending forces. The cradle to cradle assemblies are primarily subjected only to the forces of respiration because they function rib to rib. Device migration describes the shift of the superior rib cradle proximally into the rib of attachment, or the distal hook migration through the lamina causing dislodgement, or “disattachment.” Some of these reported device migrations “through” bone may actually be reactive rib bone growing around the superior cradle giving the appearance of device migration. Some cradles actually erode through the bone and emerge superior to the rib into the surrounding muscle.

There were 13 device fractures in 7 of 33 patients in the feasibility study, but only 6 device fractures in 5 of 214 patients in the multi-center study. When the total number of actual surgical procedures (initial surgeries, expansions and replacements) are considered, the rate of device fractures is 3.3% in the feasibility study (13 events in 398 procedures), and 0.5% in the multi-center study (6 events in 1,140 procedures). There were 50 procedure-related infections in the 1,538 surgical procedures for the feasibility and multi-center studies (3.3%).

During the course of this 14-year study, there were 12 deaths among the 257 patients enrolled in the study, 4 in the feasibility study and 8 in the multi-center study. None of the deaths were determined to be related to the study device by the investigators.

RISK PROBABLE BENEFIT ANALYSIS

TIS is a life threatening condition that affects a small population of children (less than 4000 occurrences per year in the United States). TIS can be seen in any three (3) of the following general diagnostic categories:

- Flail Chest Syndrome
- Rib fusion and scoliosis
- Hypoplastic thorax syndrome

The patient population of this study is a heterogeneous mixture with respect to underlying cause (genetic, congenital, or acquired), severity of symptoms, patient growth, and other medical conditions concurrent with TIS.

Treatment with the VEPTR device has been shown to maintain or improve the AVR in 92.0% of the patients, and the patient survival rate in the VEPTR clinical trial was 95.1%, whereas this condition is frequently terminal with non-surgical treatment. With the VEPTR, allowed growth of the thoracic spine and lungs while controlling severe scoliosis.

Each child with TIS presents a unique combination of factors that dictate the breadth of treatment required. These factors include age, gender, and length of follow-up, diagnosis, overall health and individual growth pattern. Depending on the presenting condition of the patient, any number of risks may be associated with the implantation and maintenance of the VEPTR device. The adverse events experienced in the VEPTR clinical study were presented in Table 6. Consideration also needs to be given to the age of the patient at initial implantation, the numerous other congenital anomalies these patients can have, and their activity levels. In addition, there are numerous factors that predispose these patients to wound infections, and the use of a prophylactic pre-operative and post-operative antibiotic regime and protective bandages at the operative site may decrease the wound infection rate.

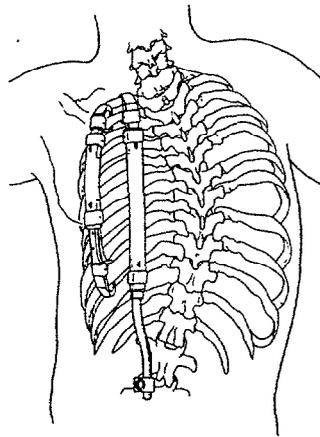
The probable benefits associated with patients implanted with the VEPTR device outweigh the risks present for this patient population.

SYNTHES® Spine

Vertical Expandable Prosthetic Titanium Rib (VEPTR)

Humanitarian Device: The Vertebral Expandable Prosthetic Titanium Rib (VEPTR) is authorized by Federal law for use in the treatment of Thoracic Insufficiency Syndrome (TIS) in skeletally immature patients, the inability of the thorax to support normal respiration or lung growth. The effectiveness of this device for this use has not been demonstrated.

Caution: Federal Law restricts this device to sale by or on the order of a physician.



Read this entire booklet carefully before your child receives the VEPTR device.

- Keep this booklet, as you may need to read it again.
- If you have more questions or do not understand the information provided in this booklet please ask your child's doctor before your child has the first operation.

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Glossary

ABD™: A type of large, thick bandage sold under this particular brand name that will help protect the wound.

Cadaver donor: Someone who has died and donated their body because they wanted to help save other peoples lives.

Centimeter (cm): A centimeter is a unit of length. (2.54 cm is equal to (1) one inch.)

Chest wall: Hollow space that surrounds the lungs.

Diaphragm: Section of your body that is located between your chest and hips.

Distraction and Expansion: These two words are used for describing how the device is made longer.

Feeding Tube: A tube will be placed into a patient's throat and that goes down to their stomach to help food reach the stomach.

Feasibility Study: A clinical study to find out if a new medical device does or does not work. It is usually happens before a bigger clinical study is started.

General Anesthesia: Is being put completely to sleep for an operation.

Flail Chest: A condition of big spaces between the ribs causing poor protection for the lungs and heart.

Hypoplastic Thorax: A condition that leaves children with a very small rib cage with little room for the lungs to grow and expand.

Incision: Cutting of body tissue.

Kerlex™: A type of thick white gauze-like wrap sold under this particular brand name that will help cover the wound.

Magnetic Resonance Imaging (MRI): A test that uses a large magnet to take pictures of your body.

Malnutrition: A condition that is caused by poor eating habits or the body's inability to take them in and use them. It is important to give your child the proper amounts of foods to encourage good skin healing.

Millimeter (mm): A millimeter is a unit of length. (25.4 mm is equal to one (1) inch.)

Occlusive op site™: A sticky saran wrap like covering sold under this particular brand name that directly lays over the wound helping to keep the wound site dry

Paralysis: A complete or partial loss of movement in body part (s).

Pneumonia: Infection of the lungs.

Progressive Scoliosis: A medical condition of a crooked or bent spine that is getting more crooked (bent) as time goes on.

Rib fusion: A medical condition of the ribs being stuck together.

Scoliosis: A medical condition of the spine being very crooked (bent).

Spina Bifida: A defect of the spinal cord that you are born with that does not allow the arches on your individual vertebra to close together.

Thoracic Insufficiency Syndrome (TIS): TIS has been explained as the chest not being able to support normal breathing or lung growth, and is considered to be a rare condition.

Thorax: The area of the chest area that is made up of the spine, the ribs and the breastbone.

Trachea: The main tube by which air passes to and from the lungs. This is a very important tube for our breathing.

Tracheotomy: Temporary opening of your child's windpipe made by an incision in the neck.

Ventilator: A mechanical device that helps you with your breathing.

VEPTR (Vertical Expandable Titanium Rib): Is a titanium device that is curved to fit the spine and is placed in an up and down position. It can be made longer as your child grows. This helps the spine become straighter and allows the lungs to grow and to fill with enough air to breathe. The attachments (*hooks or cradle caps*) can be attached to the ribs or to the spine. The device is used to keep the ribs separated after the operation.

Vertebra: One of the many bony parts that make up your child's spine.

X-ray: Photograph that is taken to see the bones inside your body.

INFORMATION ON THE VERTICAL EXPANDABLE PROSTHETIC TITANIUM RIB (VEPTR)

1. DESCRIPTIONS, PURPOSE AND SURGICAL PROCEDURE OF THE VEPTR DEVICE

1.1 WHAT IS THE VEPTR DEVICE?

The VEPTR device is a metal rod curved to fit the spine and is placed in an up and down position. It can be made longer as your child grows. This helps the spine become straighter and allows the lungs to grow and to fill with enough air to breathe. The attachments (*hooks or cradle caps*) can be attached to the ribs or to the spine. The device is used to keep the ribs separated after the operation.

The device is made of titanium because of its strength and ability to stay inside your child without causing a bad reaction. Titanium is a strong metal used for building skyscrapers and dental implants. It can join with bone. Titanium does not stop your child from having a Magnetic Resonance Imaging (*MRI*).

This design is a result of ten years of research.

1.2 WHAT IS THE PURPOSE OF THE VEPTR DEVICE?

The VEPTR device is indicated for treatment of Thoracic Insufficiency Syndrome (TIS) in skeletally immature patients. TIS has been defined as the lack of ability of the chest to support normal breathing or lung growth, and is considered to be a rare condition. Thoracic Insufficiency Syndrome or TIS describes some of the abnormalities of the thorax, such as

- *flail chest*, which means there are big spaces between the ribs causing poor protection for the lungs and heart
- *rib fusion with scoliosis*, which means the ribs are stuck together and the crooked spine that is getting more crooked (bent)
- *hypoplastic thorax*, which means a very small rib cage with little room for the lungs to grow and expand

Your child suffers from Thoracic Insufficiency Syndrome (TIS) because the *thorax*, which is the area made up of the spine the ribs, and the breastbone, fails

to support normal breathing and lung growth. If your child's chest cannot grow, their lungs cannot grow and life-threatening breathing problems can develop.

All of the above conditions could cause your child to have trouble breathing. Your child may eventually need a machine that will help his or her breathing (*ventilator*).

The VEPTR device has been made to give the rib cage room to grow, in children like yours that suffer from chest wall and/or spine defects. While your child's natural ribs run across their chest, the VEPTR device is placed up and down to give their chest more space. The device is used to rebuild your child's chest by making it larger, longer or more normal in shape and size.

Figure 1 below shows what a normal chest wall should look like and Figures 2, 3 and 4 show what a chest wall of a child with Thoracic Insufficiency Syndrome may look like.

Figure 1: Normal lung space

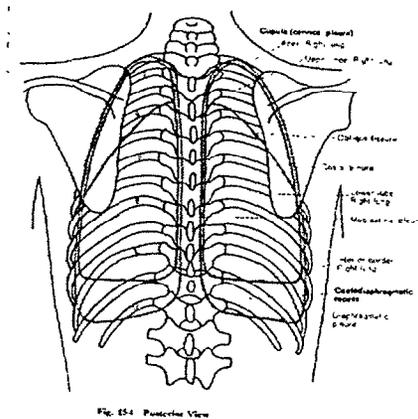


Figure 2: Abnormal lung space

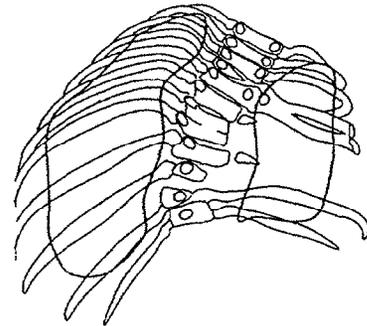
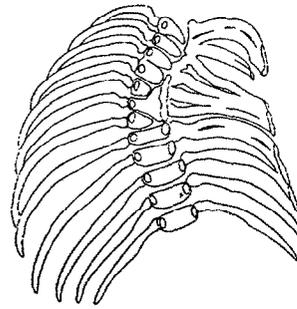
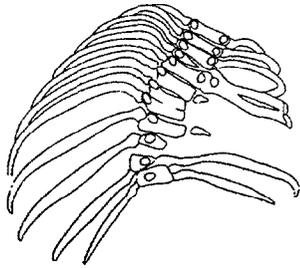


Figure 3: Missing Ribs

Figure 4: Fused Ribs with Scoliosis



The VEPTR implant or device is made of titanium, which is a common metal used for implantation. All of the parts are made of titanium or titanium mixed with a non-metal substance (*titanium alloy*). Both of these materials have a long history of safe use being put into the human body.

The VEPTR device is made up of a combination of the following titanium part (s):

- Superior Cradle
- Inferior Cradle
- Cradle End Half
- Extended Cradle End Half
- Rib Sleeve
- Cradle Lock
- Distraction Lock
- Lumbar Extension
- Low Profile Lamina Hook
- Sacra Ala Hook
- Connector
- 2mm Ti Rod

1.3 VEPTR IS AVAILABLE TO BE PUT TOGETHER IN THREE COMBINATIONS

- The Cradle to Cradle
- Cradle with Lumbar Extension
- Cradle to Ala Hook

Please see Appendix 1 in the back of this packet for a more detailed description of the above mentioned device combinations.

1.4 WHEN SHOULD THE DEVICE NOT BE USED

The VEPTR device should not be used when:

1. There is not enough bone strength of the ribs or the spine where VEPTR would be attached.

2. There are missing ribs nearest and furthest away from where the VEPTR device needs to be placed and attached.
3. The *diaphragm* (section of your body that is located between your chest and hips) cannot work properly.
4. Not enough soft tissue for coverage of VEPTR.
5. In patients that are skeletally mature (about age 14 for girls and age 16 for boys) with problems other than chest wall instability
6. Patients are younger than 6 months of age.
- 7 Your child has a known allergy to any of the device materials
8. Your child has an infection at the operative site.

1.5 WHAT ARE MY CHILD'S RISKS?

Surgery to implant the VEPTR into the chest is considered a major operation.

- The risks that are linked with any surgery of the chest include problems from anesthesia, bleeding, infection, heart or lung problems, pneumonia (infection of the lungs), surgical wound problems (like infection or not healing) and sudden death.
- The risks that are linked with the use of the VEPTR include but are not limited to the following (Also see Appendix 2):
 1. The VEPTR device could bend or break from an accident, during lengthening, or being worn out because of daily activities. This will be checked by taking photos (*x-ray*) and may result in the device being taken out and a new one being put in.
 2. Breakage or movement of the device could cause the device to damage areas like the lung, heart or large blood vessels in or around the chest area that may cause your child to have a surgical repair.
 3. As your child grows and expansion surgeries are done, the VEPTR device(s) could move or loosen from its original placement on the ribs (go through the newly formed bone). This might cause an unscheduled hospitalization and surgery to reposition, take out and/or put in a new implant.
 4. The VEPTR implant can often be felt or even seen under the skin. Your child's skin and muscle around and over the VEPTR device has been stretched. This could cause the skin to break open or become infected. They might need treatment or removal of the implant. A protective device or covering should be worn over the VEPTR implant to avoid any injury to the skin.
 5. Your child's arm has nerves and blood vessels that lie close to the area where the device will be put in. They could become damaged by the surgery to put the device in because all of the muscles on the side of the chest are lifted up and moved to allow for the operation on the ribs, or by your child's daily activities. This may call for more surgery.
 6. There is the possibility that your child's body will reject the metal by the chest wall muscles or other tissues that could result in the removal of the implant.

7. There is a possible risk to the spinal cord as the result of straightening the spine by pushing the ribs apart, which could result in temporary or permanent loss of movement in a body part (*paralysis*).
8. After all surgeries, your child will need to be put on a ventilator to help with their breathing. The ventilator may be used for a long time after surgery because of lung complications and a tracheotomy which is temporary opening of your child's windpipe (*trachea*) may be made to help to stop any long lasting throat irritation.
9. Your child will most likely have some pain following the surgeries.
10. Your child could experience allergic or unexpected drug reactions, like irritability, vomiting, or prolonged drowsiness.

Each child with Thoracic Insufficiency Syndrome comes with a unique combination of medical problems that determine the amount of treatment required. These problems include age, gender (female or male), and how long the follow-up needs to be, diagnosis, general health and individual growth pattern.

1.6 WHAT ARE MY CHILD'S BENEFITS?

Some of the possible benefits to patients receiving the VEPTR are the following:

- More normal growth patterns without spinal growth limitations
- Decreased deformity of the spine
- More room for your child's lungs to grow
- Increased amount of daily activities because of the increase amount of space for air in the lungs
- Decreased need for using ventilators

The probable benefits to the quality of life for your child with the implantation of the VEPTR device outweigh the risks that could be caused with its implantation. Please look at Appendix 3 for more details.

Though some risk is always linked with any surgical procedure, the quality and length of life provided to patients with TIS outweighs the risk that may be caused with the surgical procedures.

1.7 WHAT ARE SOME WARNINGS AND PRECAUTIONS FOR THIS DEVICE?

Patients implanted with VEPTR should not be braced. It is assumed that a brace would squeeze the chest and stop its natural movement. With the VEPTR implant, natural breathing movements are encouraged. Additional bandages over the wound will protect it from inadvertent rubbing or bumping of the wound. If your child has spina bifida, place an occlusive dressing over the wound site to keep the site dry.

The VEPTR device should not be used when your child has any of the following:

1. They do not have enough bone strength of the ribs or the spine where VEPTR would be attached.
2. They have missing ribs nearest and furthest away from where the VEPTR device needs to be placed and attached.
3. Their *diaphragm* (section of your body that is located between your chest and hips) cannot work properly.
4. There is not enough soft tissue for coverage of VEPTR.
5. In patients that are skeletally mature (about age 14 for girls and age 16 for boys) with problems other than chest wall instability.
- 6 Your child is younger than 6 months of age.
- 7 Your child has a known allergy to any of the device materials.
8. Your child has an infection at the operative site.

1.8 WHAT SHOULD YOU DO BEFORE VEPTR SURGERY TO HELP WITH THE SUCCESS?

- Keep your child away from crowds and anyone who is sick.
- Before surgery, it is important to give your child the proper amounts of foods to encourage good skin healing. Surgery is like a marathon; your child needs to eat very well in the weeks before the surgery.
- Let your Doctor or Nurse know if your child won't eat or if your child is having trouble eating.
- Make sure that your child takes all prescribed medicine and breathing treatments.
- Flu shots are suggested. They are suggested for all children with lung problems.
- Pneumonia shots are suggested if your child is on a ventilator.
- Call your Doctor if your child is sick. If you live far away, **DO NOT** come to the hospital for the surgery unless your Doctor tells you to.
- If your child is not eating, your doctor may prescribe medicine to increase your child's appetite.
- In cases of severe malnutrition or poor eating habits, a tube will be placed into your child's throat and that will go down to their stomach (*feeding tube*). This will help the food reach the stomach and help your child get stronger, but until then surgery may be delayed.

1.9 WHAT HAPPENS DURING THE VEPTR PROCEDURE?

During the first surgery the doctor makes a small cut (*incision*) on your child's body and puts the device in that will make the rib cage longer and larger as your child grows. Your child's reconstructed chest can provide more room for your child's lungs to grow and expand. This surgery will take place with your child being completely asleep during the operation (*general anesthesia*). The doctor makes a small cut in your child's body and then inserts the device that will lengthen and expand as your child grows. There will also be other smaller surgical openings to

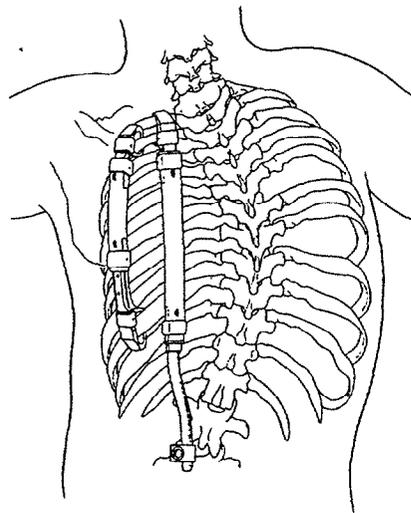
help place the parts of the VEPTR and lock them in place. The expansion surgery normally only requires an overnight hospital stay. There is a full description of the surgery for an expansion and replacement later in this booklet (See page 14).

After surgery, your child will stay in the Pediatric Intensive Care Unit until the doctor feels that your child is ready to be transferred to a regular hospital room. Your child will probably be put on a ventilator for about three days to help them breath.

If your child has problems on both sides of his or her chest and/or spine, another surgery to implant more VEPTR parts may be needed. If additional implantation surgery is necessary, it will be scheduled about three months after the first operation.

Picture 5 shows the VEPTR device after it is implanted with a Rib to Rib and Rib to Spine attachment.

Figure 5: Rib to Rib and Rib to Spine Attachment



If your
would

surgery.

child has fused ribs, his or her x-rays
look something like the Figure 6 before
implanting the device and Figure 7 after

During your child's surgery the following will happen:

- Your child will be placed on their side.
- Their skin will be washed with a liquid that kills germs.
- Then they will be covered with towels, so that only the place where the incision (s) will be done (surgical site) is left uncovered.
- Finally, a standard thoracotomy is performed as described in the first paragraph of this section.

After your child's ribs are exposed, the ribs that are fused together will be gently separated into multiple sections, to look like ribs that had not been fused together, with an instrument called a lamina spreader.

Examples of what is done are in Figure 6 and 7:

Figure 6: Separating fused ribs

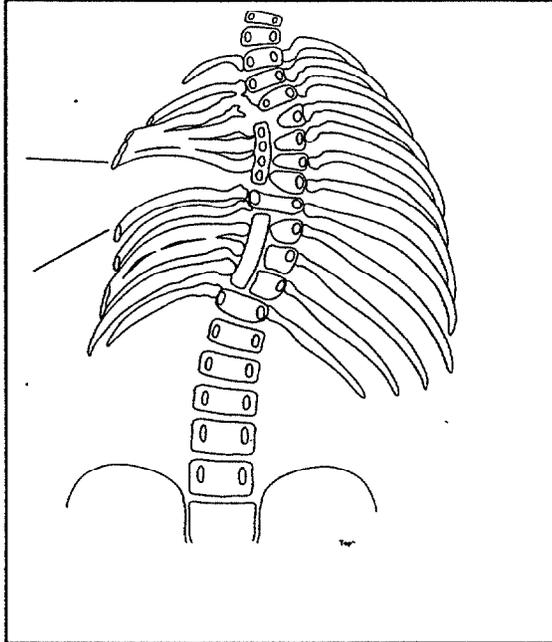
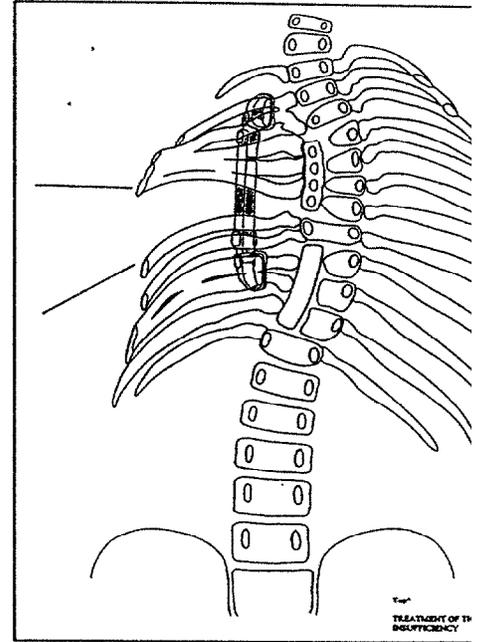


Figure 7: After separation of fused ribs (Rib to Rib Device Attachment)



If separating the fused ribs by the instrument is difficult, then there is a possibility that an additional rib(s) will need to be separated.

A VEPTR device is placed on your child's ribs in back of their body as close to the spine as possible attaching up and down in the same manner as the device used for rib absence. At some point your doctor may suggest that a second titanium rib should be placed closer to under the arm on the same side.

After both devices are completely assembled, they are made to fit snugly without much pressure.

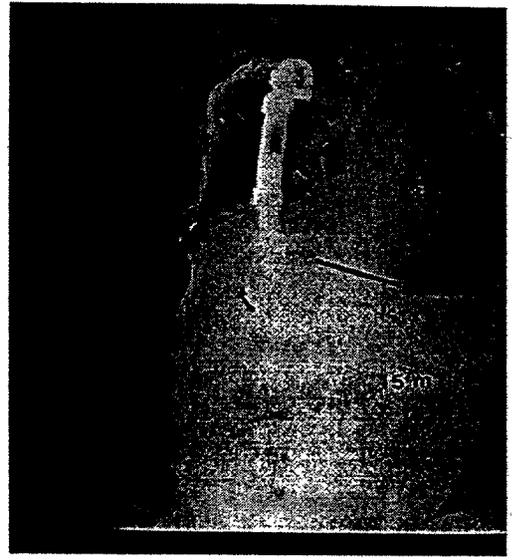
1.10 CASE EXAMPLES:

1.10.1 Case I

Photo 1: Fused Ribs with Scoliosis
(8 month old child) Before Surgery



Photo 2: Seven (7) months after surgery



If your child has a bent spine (*scoliosis*), they will have two VEPTR devices placed. One will attach from the top of the rib cage to the bottom of the rib cage. The other will be shaped to fit from the top of the rib cage to the lumbar spine.

Once your child stops growing or when he or she has reached skeletal maturity (about age 14 for girls and age 16 for boys) the VEPTR device may no longer be of any benefit and you and the doctor treating your child will decide if the device should be removed. For a child with a spinal deformity, a spinal fusion may be suggested in the future.

You and your child's physician will continue to work together during your child's treatment.

Your child will need to go back to the hospital for surgery to:

- make the device longer
- or replace the VEPTR device at specific times to make room for the growth of your child
- and/or to further correct spinal or chest wall deformity.

These surgeries will be done while your child is under general anesthesia. These surgeries may need to be done as often as every four (4) to six (6) months. When the rod cannot be made longer, the middle piece will be taken out and a longer one will be put back in.

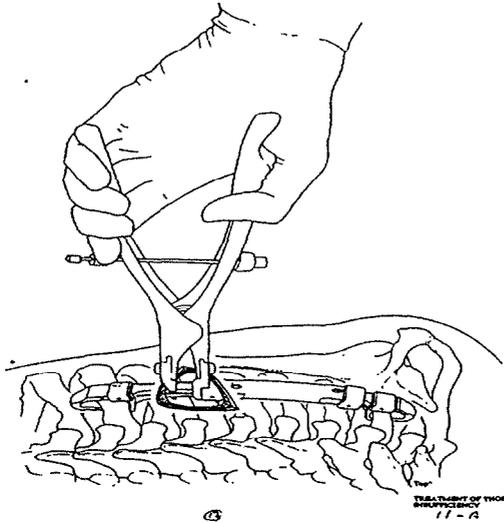
A very small incision is needed to make the VEPTR device longer. The incision only needs to expose the small portion of the device where the expansion takes place.

When your child outgrows the device, the central section or middle part of the device will need to be replaced.

The following is a description of the expansion (*sometimes called distraction*) operation:

As seen in Figure 8, a small incision is made and an instrument called a distraction forcep is put in place. Gentle pressure is used to begin the distraction of the device. When there is a large amount of pressure the pliers are locked and three minutes are allowed to go by to allow the tissue to relax. Next the rod is distracted another 2-3 mm and locked in place for another three minutes to again allow all the tissue to relax. Distraction continues until the device is lengthened 0.5 cm, 1 cm or 1.5 cm, based on the amount of force needed to expand it. Most prosthetic ribs attached only to the chest wall closer to the spine will permit only .5 cm. lengthening per surgery while the prosthesis closer to the underarm generally will stand about 1 cm of lengthening per surgery. The lumbar extensions may be expanded 1 to 1.5 cm, per time space.

Figure 8: Expansion of the VEPTR device



2. POSTOPERATIVE WOUND CARE FOR VEPTR SURGERY

2.1 WHAT KIND OF INCISION WILL MY CHILD HAVE?

Your child will have two kinds of incisions as seen in Figure 9

1. The main incision is long and shaped like the capital letter J.
2. The other incisions are smaller and are used for making the device longer.

Figure 9: Child with a J shaped and smaller incision



Your child will need to have multiple repeat operations to allow for their growth. Over time the attachments to your child's ribs or spine may loosen and may need to be replaced or repositioned. This can be done as a part of a lengthening operation. For the lengthening surgeries, a small incision is required at the lower end of each VEPTR implant. When replacement of your child's VEPTR implant is required a larger incision will be needed. The lengthening operation is brief, but your child will experience mild to moderate pain following these surgeries. At a minimum, overnight hospitalization will be required. Complications such as infection, device breakage and rib fractures are possible risks associated with these procedures.

2.2 WHAT SHOULD I KNOW TO HELP MY CHILD HAVE A SUCCESSFUL SURGERY?

- Make sure that all caregivers, including yourself, wash their hands before working on the wound or bandages to avoid infection.
- Your child's skin and muscles over the VEPTR device had to stretch a lot to cover the expanded chest. Your child may have been born missing muscles in the area operated on or have had previous surgery in this area. New postoperative wounds may be under strain and may be more easily injured.
- New postoperative sites are more at risk to injury and infection
- Your child's wound or incision will be covered with a large, thick bandage

and then a wrap.

- Some children will have a donut sponge on the outside of the bandage for protection so that the child doesn't rub their wound area or bump into anything.
- If your child has a diagnosis of Spina Bifida (a defect of the spinal cord that you are born with that does not allow the arches on your individual vertebra close together), your child will then have a sticky saran wrap like dressing (*Occlusive op site™*) over the wound site to keep the site dry.
- Your child will stay in the hospital for 10 days to 14 days, unless there are complications. Your child's medical condition before surgery will also affect how long your child will be hospitalized.
- The stitches are left in for a minimum of four weeks.
- An antibiotic will be given to prevent infection for 6 weeks after surgery.

2.3 HOW SHOULD I CHANGE MY CHILD'S POSTOPERATIVE BANDAGE?

- All caregivers **MUST** wash their hands before working on the bandages or the wound area to avoid infection.
- When your child is allowed to leave the hospital you will be given written discharge instructions from your Doctor or Nurse about how often to change the wound bandages.
- In your discharge instructions they will let you know what to use as bandages for the site wound.
- Types of Bandages:
 1. The large, thick bandage (*ABD™*) and a thin gauze like wrap (*Kerlex™*) will help protect and cover the wound.
 2. If your child has a diagnosis Spina Bifida, they will have a sticky saran wrap (*Occlusive op site™*) covering the wound directly. This will help in keeping the wound site dry.
 3. In some cases, the bandages will be covered with a donut looking, sponge over the dressing. This will protect the wound from some bumps and irritation to the wound.

2.4 WHAT SIGNS OR SYMPTOMS OF WOUND INFECTION SHOULD I LOOK FOR?

If you observe any of the following signs or symptoms, call your doctor immediately:

- Fever that does not go away
- Fever of 101 degrees or greater
- Redness and swelling at the wound site
- Pain at the incision site that is getting worse not better as the days go by
- Tenderness at the wound site that is increasing or does not go away
- Opening of the wound site, (for example the skin coming apart)
- Liquid drainage coming from wound site

2.5 HOW DO I PROTECT THE POSTOPERATIVE (AFTER SURGERY) WOUNDS FROM INJURY AND INFECTION?

- Make sure that all caregivers, including yourself, **wash** their hands before working on the wound or bandages to avoid infection.
- Don't let your **child pick at the wound**, because their hands carry many forms of bacteria. If they pick at their wound site, they will increase their risk of getting an infection.
- **Keep the wound covered with protective bandage for the following reasons:**
 - 1) New wounds are more likely to get infected.
 - 2) The VEPTR device is usually very close to the outer layer of skin.
 - 3) Provide protection for the wound site from irritation and bumps.
 - 4) The skin and muscles over the device have been stretched, so it is very thin.
 - 5) If rubbed the thin skin covering the wound can easily break (for example when your hands crack in the winter).
- **Don't let your child do things that rub directly against the wound because any rubbing may break the thin skin covering the wound.** The following are two examples:
 - sitting in a hard chair
 - lying on a hard floor
- **If your child returns to school and you see that the chair and/or desk bumps or rubs the wound**, you should ask for padding for the chair or desk or some kind of a protector. If needed, you can ask your Doctor for a prescription.
- **Either avoid taking your child on long car rides in car seats or provide extra padding to protect your child's wound.** The car seat will rub the wound area and may break the already stretched and thinned out skin covering the wound.

3. COMMON VEPTR DEVICE ISSUES WITH GROWTH

3.1 HOW OFTEN WILL MY CHILD NEED LENGTHENING OPERATIONS?

Your child will need to go back to the hospital for surgery to make the VEPTR device longer or to replace the VEPTR device at specific times, to make room for the growth of your child and/or to further correct spinal or chest wall deformity. These operations may need to be performed as often as every four (4) to six (6) months. Your child will be asleep (*general anesthesia*) during the operation (s). When the rod cannot be made longer, the middle piece will be taken out and a longer rod will be put inside your child. This operation will usually need just an overnight stay in the hospital.

3.2 WHAT HAPPENS DURING MY CHILD'S GROWTH?

The following are signs and symptoms that your child has outgrown the device.

- Complaints of mild discomfort in the device area
- Worsening of your child's curvature or bent spine
- Your child may actually feel that the device has loosened or drifted
- A bump under the skin in the operative site area
- You could see the device coming through the skin or close to the skin surface
- Your child complains that they have heard a pop in the back area (this is usually only with very demanding activity)

3.3 ARE MY CHILD'S ACTIVITIES LIMITED?

- After the first month, there are no limitations in your child's daily activities
- If your child has a device with a lumbar extension or an Ala Hook Assembly, we strongly encourage that he or she not play in violent sports (such as wrestling, weight lifting, football, etc.). There have been times when the device has broken or moved from the place where it was attached during surgery as a result of children playing in the above rough sports.

3.4 WHAT HAPPENS AT THE END OF GROWTH?

Once your child stops growing or when he or she has reached skeletal maturity (about age 14 for girls and age 16 for boys) the VEPTR device may no longer be of any benefit to your child. You and the doctor treating your child will decide if the device should be removed. We strongly suggest that parents discuss all options and expectations associated with the end of growth needs for this device with their child's doctor.

3.5 HOW LONG CAN I EXPECT THE DEVICE TO WORK?

Once your child stops growing or when he or she has reached skeletal maturity (about age 14 for girls and age 16 for boys) the VEPTR device may no longer be of any benefit to your child. You and the doctor treating your child will decide if the device should be removed. For children with a spinal deformity, a spinal fusion may be needed.

We strongly suggest that parents discuss all options and expectations related with the end of growth needs for this device.

3.6 WHEN AND HOW SHOULD I CONTACT MY CHILD'S DOCTOR?

The doctor and nurse's numbers should be listed.

You should call your child's doctor when:

- If your child has a fever of 101 degrees or greater
- If you notice redness or swelling at or around the wound site
- If your child has trouble eating
- If your child complains of pain that does not go away
- If the pain medication does not appear to be working
- If your child is having difficulty breathing

3.7 WHAT ALTERNATIVES TO THIS PROCEDURE ARE AVAILABLE?

Thoracic Insufficiency Syndrome (TIS) is a life-threatening condition found in children. A mechanical device that helps you with your breathing (*ventilator*) can be used as a non-surgical treatment in some patients; however, this is just treating the symptom and not treating the cause of the condition.

Currently available surgical treatments for chest wall defects include use of:

- a plastic sheet of fixed size that will protect your child's chest wall area
- artificial (*prosthetic*) ribs made of plastic that are a fixed size, meaning they cannot be made longer as your child grows.
- if your child is old enough, sections of ribs from someone who has died (*cadaver donor*) or autograft (sections split from your own ribs on the normal side)

Fused Ribs

- Requires splitting the fused ribs apart, then putting a spacer to stop the ribs from sticking together again or growing back together.
- Past treatment for patients with underdeveloped chests has involved splitting the breastbone and inserting a spacer to hold it apart.

Scoliosis

- Can be treated with an operation that is called a surgical fusion with instrumentation (meaning they put in growing rods), but these methods do not take care of the malformations of the chest and fusion (means unable to move) will prevent growth.

4. ADDITIONAL INFORMATION

The company that makes and sells this product is

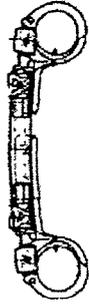
SYNTHES® Spine
1230 Wilson Drive
West Chester, PA 19380
1.800.295.1278 (toll free)

Appendix 1: VEPTR Combinations

Ala Hook Assembly

is primarily used when your child has Thoracic
of fused/missing ribs, severe scoliosis, and/or

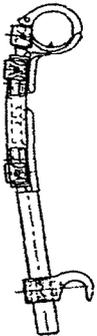
ly Cradle to Cradle Assembly



Ala Hook Assembly

Ala Hook Assembly is made up of 8 pieces. The Cradle
to Cradle Assembly (Figure 7) is indicated for use where no lower ribs
goes into the lower part of your back (lumbar region)

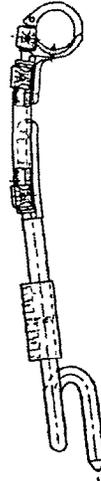
nb,



Ala Hook Assembly is made up of 9 pieces. The Cradle to Ala Hook
end of the end section of the device to the hip is
bottom ribs or strong enough lumbar bones to which
to help even out your child's hips if one side is
him/her to sit up while in a wheel chair.

Using instruments that help
and made longer in a move
and then locked back into p
child's rib cage is increase
allow their lungs to grow.

Ala Hook Assembly



Using instruments that help put the implant in place, the end section can be unlocked and made longer in a movement similar to flat curtain rods sliding inside each other and then locked back into place. By making the implant longer, the space in your child's rib cage is increased, and there will be more space within your child's chest to allow their lungs to grow.

Appendix 2: Risks linked with the use of the VEPTR device

What are the results of clinical studies with the VEPTR device?

The goals of VEPTR surgery in the clinical trials are the following:

- 1- To even out the height of each individual half side of the chest and then try to keep this change with each expansion (lengthening of the rod(s)) of the devices.
- 2- A decrease in Cobb angle (a measurement used to help figure out the degree of a scoliotic curve) represents an improvement. If possible, a lessening in the Cobb angle is the goal of both scoliosis surgery and expansion using VEPTR.

The VEPTR device was evaluated in a single site feasibility study with thirty three (33) patients and a multi-center, prospective study done at seven (7) sites with two hundred twenty four (224) patients. A total of two hundred fifty seven (257) patients were studied, but ten (10) patients were excluded from the analysis due to the absence of baseline data. Enrolled patients at each site received the VEPTR device assembly according to their specific disease and body dimension requirements. Patients experienced no life-threatening or fatal adverse effects that were considered to be device related.

There were 4 intraoperative complications reported for the feasibility and multi-center studies (1.9% of all patients), including a technical error in device placement; a dural laceration, and pressure on the brachial nerve. Sixteen feasibility patients, or 48%, experienced 37 device-specific adverse events, and 34 multi-center patients, or 16%, experienced 52 device-specific adverse events. These device-specific adverse events included device fractures, device migrations, and other device-related adverse events. Device migrations occurred frequently—25 migrations in the feasibility study and 49 migrations in the multi-center study. They were more common with the cradle-to-lumbar extension and cradle-to-sacral ala assemblies (these two configurations undergo greater flexion, extension, rotation and lateral bending forces than the cradle to cradle assemblies, which are primarily subjected only to the forces of respiration because they function rib to rib). Device migration describes the shift of the superior rib cradle proximally into the rib of attachment, or the distal hook migration through the lamina causing dislodgement, or “disattachment.” Some of these reported device migrations “through” bone were actually bone growing around the superior cradle giving the appearance of device migration. Some cradles actually eroded through the bone and emerged superior to the rib into the surrounding muscle.

There were 13 device fractures in 7 of 33 patients in the feasibility study, and 6 device fractures in 5 of 214 patients in the multi-center study. When the total number of actual surgical procedures (initial surgeries, expansions and replacements) are considered, the rate of device fractures were 3.3% in the feasibility study (13 events in 398 procedures) and 0.5% in the multi-center study (6 events in 1,140 procedures). There were 50 procedure-related infections in the 1,538 surgical procedures for the feasibility and multi-center studies (3.3%).

During the course of this 14-year study, there were 12 deaths among the 257 patients enrolled in the study, 4 in the feasibility study and 8 in the multi-center study. None of the deaths were determined by the investigators to be related to the study device.