

**VERTICAL EXPANDABLE PROSTHETIC TITANIUM VEPTR RIB IMPLANT  
RAINBOW BABIES & CHILDREN'S HOSPITAL  
CLEVELAND, OH  
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ANNOUNCER: Today, orthopedic surgeons at Rainbow Babies & Children's Hospital in Cleveland, Ohio, will demonstrate the implantation of a vertical expandable prosthetic titanium rib in a pediatric patient. The VEPTR device is used to treat children who have thoracic insufficiency syndrome. TIS occurs in young children with chest wall malformations often associated with scoliosis. This can restrict lung growth, cause breathing complications, and even lead to early death. The VEPTR is designed to mechanically stabilize and distract the thorax and expand the ribcage to improve breathing and allow for more lung space. During the webcast, you may send questions to the surgeons at any time. Just click the MDirectAccess button on the screen. For more than a century, Rainbow Babies & Children's Hospital has been dedicated solely to the care of children and is one of the most renowned pediatric medical centers in the country.

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GEORGE THOMPSON, MD: Good evening. I'm George Thompson. I'm the director of pediatric orthopedics at Rainbow Babies & Children's Hospital, and I'm the current president of the Scoliosis Research Society. I want to welcome you to our program tonight, which we're going to be presenting the surgical procedure of inserting a VEPTR device for the treatment of thoracic insufficiency syndrome and chest wall deformities. I'm joined tonight by my colleague, Dr. Douglas Armstrong, who is also a pediatric orthopedist and a specialist in pediatric spine deformity. The VEPTR stands for Vertically Expandable Prosthetic Titanium Rib, and it's a relatively new device. It comes in three different styles, one of which is designed for rib-to-rib insertion, one from spine to rib, and the other one, which is the bottom one you'll see on -- on the slide, is to go from the pelvis to the rib. All these can be utilized in children with a variety of diagnoses who have chest wall deformities and/or thoracic insufficiency syndrome. This device was first developed by Dr. Robert Campbell from San Antonio, Texas. Dr. Campbell's a pediatric orthopedist who has spent most of his entire career in the development of this project. It has now been approved by the -- by the FDA and is now available for use in a very limited number of children with specific diagnoses. The -- Doug, do you have any comments you'd like to make?

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DOUGLAS G. ARMSTRONG, MD: Just to remind people that they can e-mail us questions by pressing the MDirectAccess button that you'll see on your screen and you can also get information about referrals.

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GEORGE THOMPSON, MD: All right. We'd love to hear from you, so any questions you have, please feel free to send them to us. And as we go between segments, we'll talk about your questions and try and give you as much information as possible. The patient tonight is a -- is currently a 10-year-old boy who was born with a congenital diaphragmatic hernia. And he's had this repaired twice, and this is not uncommon

because as a child grows, the synthetic material needs to be replaced in order to accommodate growth. This was performed by going through his chest and then working on the top of the diaphragm. And what happened is that he's had two pair of ribs that have fused together, and this resulted in a lateral tether. And that tether has ended up causing him to have a slowly but progressive spinal deformity. If you'll notice the -- the first pair of radiographs on the slides is that in March of 2003, he actually had a 21-degree left thoracic curve, but by July of this past year, this had already progressed to 37 degrees. And our studies had shown that he actually had the two pair of fused ribs, and I hope these project well for you to see them, but they are adjacent to each other, they're right in the midaxillary line, so they were having a significant effect on his overall spinal growth. It was a lateral tether, about as far away from the spine as you could get. But as he was growing, the ribs were not being allowed to separate, and therefore they were tethered and he was developing a progressive spinal deformity. These were shown to Dr. Campbell, who had not encountered a child with this particular problem in the past but felt he would be an excellent candidate to undergo the VEPTR procedure. This just shows his bending film to indicate that the tether was relatively stiff, and this prevented any spontaneous improvement in the curvature when he bent away from the deformity. This is a relatively standard radiograph, or x-ray, that's taken when we assess patients preoperatively to help us determine where the levels of instrumentation are going to be and what type of result to anticipate based on his deformity. This is the -- at the time of the surgery, here he is positioned on the operating-room table. He's on his left side, so he's facing away from us at this point. It is an L-shaped type of incision that allows us to mobilize the entire shoulder and its associated musculature. Once this is moved away, we're right on top of the chest wall and can proceed with the insertion of the -- of the VEPTR device. So let's go to the operating room, and we'll look at some of the highlights of this surgical procedure. We've already made the incision at this point, and the musculature has been mobilized and the shoulder is -- has been elevated, so we're looking right down on the chest wall. And I think you can appreciate that his head is away from us and the -- and the trunk is -- the child is laying on their left side. We are dividing the intercostal muscles at this time, and this is between the -- the lower of the two pair of fused ribs. It's important that good mobilization be achieved so that we can actually correct the chest wall deformity. So the muscles are divided using electrocautery all the way from the spine out to the sternum, which is in the front. And as we begin to develop mobility, the spreader that you see being inserted at this time aid in opening the ribs and separating them and allowing us to have better visualization, and it makes it much easier to divide the intercostal muscles. As we get more and more relaxation, we open the devices further and further. Now, what you're seeing in the base between the retractors there is the lung. In most cases, the lung would collapse and fall away, but because of the previous surgeries, the lung is relatively scarred and it stayed inflated for the most part through the entire procedure. Doug, any comments about the procedure at this point?

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DOUGLAS G. ARMSTRONG, MD: No, it seems that you've got quite a bit of separation between the ribs already. How much separation were you aiming for between the ribs, ultimately?

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GEORGE THOMPSON, MD: Well, I think at the end of the procedure, you'll see that we actually have about four to five centimeters through two thoracotomies. This was a tremendous amount of incision-- of relaxation and expansion of the chest wall we were able to -- to achieve.

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DOUGLAS G. ARMSTRONG, MD: The curvature wasn't too big just now, but the child was only 10, and he had a -- this was the same side that he'd had a diaphragmatic hernia on, so his lung would've been already compromised from birth. What do you think would have happened to his lung if we just waited and watched for a few more years?

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GEORGE THOMPSON, MD: Well, I don't think anything would have happened to his lungs because we had a preoperative computed tomography, and the -- had pretty good space available for his lungs. I think this would have been a pure spinal deformity as a sequelae to his fused ribs had we done nothing. And you -- as you saw from the preoperative radiographs, he was already progressing at a fairly significant rate.

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DOUGLAS G. ARMSTRONG, MD: Well, it's interesting because most of the patients that -- or many of the patients that are -- for which a VEPTR is indicated have an associated congenital deformity of their spine, but in this case there didn't seem to be any deformities of the spine that he was born with.

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GEORGE THOMPSON, MD: That's right. As Dr. Campbell had told us, this was a very unusual indication for VEPTR, and he had personally not even seen this type of -- of case before.

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DOUGLAS G. ARMSTRONG, MD: There's really no alternative to it because it -- any treatment of the spine would not address the primary deformity, which was in his chest wall.

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GEORGE THOMPSON, MD: That's correct. And that was his feeling as well. I know it's hard to visualize right now, but we're actually doing a second release, and this is between the lower pair of fused ribs and the next normal rib. So we're doing a double thoracotomy at this point, and it will be done exactly the same way as the first. It's completely released from the spine out very close to the costal margin or the sternum. And you can get an appreciation of that now by looking, you know, at that kind of thickened band there, which is actually two ribs that are fused together at that particular location. And here we are, we're moving anterior at this point, just releasing the muscles, freeing up that -- that first segment. And care is being taken to avoid any potential injury to the -- the lung. But it was amazing, the amount of -- of scar tissue that was present and also amazed at how much mobility, and you'll see that again, and the audience will see it as well, towards the end of the operative procedure.

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DOUGLAS G. ARMSTRONG, MD: I suppose some people might ask, you know, why couldn't we go and just separate the ribs and see how they grew.

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GEORGE THOMPSON, MD: Well, that was another question that I had actually had, and Dr. Campbell has found that if you try just to divide the rib, they frequently will re-form, and there's a lot of scarring, and you don't get quite as much opening as you do if you go into the more normal musculature of the chest wall.

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DOUGLAS G. ARMSTRONG, MD: You see the same thing once in a while in people who've had to have surgery for -- at a very young age for cardiac or an esophageal anomaly. Do you think the procedure might be indicated for -- for that?

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GEORGE THOMPSON, MD: Well, I do, you know, and that's the -- that's the typical post-thoracotomy scoliosis. And I'm sure, although those people typically don't have fused ribs, they do have fibrosis of muscles which are still acting somewhat as a lateral tether. And that's why I think we see that type of -- the high incidence of spine deformity in children who've had a thoracotomy despite the fact they've never had a -- you know, a spinal abnormality before.

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DOUGLAS G. ARMSTRONG, MD: The procedure's being used now for people having scoliosis but now -- but are not born with anything wrong with their bones, the ones you and I would call infantile scoliosis --

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GEORGE THOMPSON, MD: Yeah, early onset scoliosis.

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DOUGLAS G. ARMSTRONG, MD: Do you think that there's any patients that you've seen that would -- that that would be useful for or that -- that's a pretty controversial area, isn't it?

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GEORGE THOMPSON, MD: It is. And, you know, to quote Dr. Campbell, he said, "This is not a spine-lengthening device," although we certainly get the secondary benefit of lengthening the spine, just as we are going to do today. But this is -- this was devised primarily as a chest-wall-deformity correcting device. And I know nationally, in the few centers that are doing this, some of them are using it as a growing rod system. And for the audience, a growing rod system is when we insert a rod into the spine for children -- very young children -- with a severe spinal deformity who have failed conservative measures. We lengthen the spine with a rod just as we would in the older child who we're going to do a fusion. But in this case, we don't do any fusion. And the concept is similar in that we're going to go back at six-month intervals and lengthen the device so we have internal support for the spine while the child is still growing. And this allows us to maximize lung development and while at the same time to maximize spinal growth, you know, for the child. It's a very difficult problem, not an uncommon problem. We have about 60 children here at Rainbow that have had or currently have growing rod systems in place. And I know, Doug, you have a few of those yourself since you've joined us here.

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DOUGLAS G. ARMSTRONG, MD: Yes. It's been a very -- very good program for people with those selected indications that you've developed over the years.

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GEORGE THOMPSON, MD: All right. So, Doug, that's the exposure thus far. And it -- it took -- as you were there, it took quite a while for us to get that open and to get the ribs -- you know, get the rib spreaders in place and begin to get the spine mobilized, so...

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DOUGLAS G. ARMSTRONG, MD: It's interesting that we were able to develop the intervals with relative ease. And there really was very little blood loss or, you know, trauma to the patient, although it looks like a very big dissection, but really not a lot of difficulty for a patient who's relatively healthy to begin with.

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GEORGE THOMPSON, MD: Yeah. Well, it was a big dissection, but the blood loss was actually quite small. And -- which is, for an incision this big, you'd think that our blood loss would be high, but it was not. Well, let's return to the operating room and look at the next segment if we could, please. These are rib spreaders that have been inserted in both thoracotomies. And I think you're beginning to get an appreciation at this time as to how much we have spread this chest open already. Remember,

there are two incisions between the -- there's an incision between the two pair of fused ribs and then there's -- the bottom incision is between the lower pair of fused ribs and the next normal rib. But those have already been spread anywhere from four to five centimeters each. This is what's called the cradle, and there's an upper and lower portion to it. And this actually will completely encircle and encase the rib. There's some little prongs in there that keep it from sliding out on the rib because the ribs are on an angle, and so we want to make sure that it stays in place and does not migrate laterally. So they slide together, and then there is a clip. And you can see that first hole where the clip will go, and there's another receiving hole underneath there, and then the little prong there will accept the VEPTR device itself, which is where the lengthenings will take place. This is Dr. Campbell having assembled the upper one here. Here we go with the -- with the device itself. This is the main portion of the VEPTR, which is a male/female component that will slide on itself, and this is how we elongate this device to accommodate growth. I think we're going to see that here at the bottom in just a moment. And you can see him pulling it a-- see him pulling it apart at this time. And another clip will go in with each lengthening that will hold it in the -- in the current position. There it is, completely inserted into each other. There's a round stem, and it just shows you the elongation, how it can happen, pointing to the holes. There's a side view of it. This is very flat, and this is designed specifically to be as -- to be as flat as possible to avoid any -- any elevation of the skin and to minimize the possibility of having postoperative wound necrosis. All right. I happen to have a couple of the devices here, and I hope we can get a close-up and you can really see how this is -- it's very flat, has a very low profile, and easily slides on itself. And you can see the series of holes where, as each elongation occurs and a new clip is applied, the device can get longer and longer, thereby allowing the child's spine -- and in this case, the chest wall -- to be sequentially lengthened. This is going to preserve his lung function, allow for maximum lung development, and hopefully in this case is going to keep him with a very minimal spinal deformity by the time he reaches maturity. This is the lower cradle, and I don't know if you can appreciate these very sharp prongs, but these will engage into the rib, and because it goes in on an angle, it's important there be prongs so that this thing doesn't begin to slide out and begin to lose its fixation point and the effect that it has been causing on both the chest and the spine itself. And then of course, the upper cradle has prongs and it really captures on to the rib and -- and gets a nice purchase.

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DOUGLAS G. ARMSTRONG, MD: And with time, the rib will actually increase in size around the device, won't it?

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GEORGE THOMPSON, MD: Yes, it can. The interesting thing, for this one, we actually used the upper pair of fused ribs in which to put this, and we have very thick segment in which to be pushing against, so I think our -- the chance of having any hypertrophy or even worse, having this migrate through the bone, is going to be fairly -- fairly small. And by the way, this is a -- this is a spine/rib construct, and you can contrast that to this one, which is a rib-to-rib construct. And I don't know if you can look at the model here, but we also have a pelvis-to-rib construct. This would only be used for children who do not walk in which we need a very long construct in order to support the spine as well as the chest wall itself. But it fits over the pelvis and then attaches to the ribs in the same manner as you saw on the other two.

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DOUGLAS G. ARMSTRONG, MD: You generally need two devices for each rib.

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GEORGE THOMPSON, MD: It's usually two on each side, but there have been cases in which one was used. It depends on what the -- what the ideology is and what we're trying to accomplish.

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DOUGLAS G. ARMSTRONG, MD: So you have to lengthen both of the devices at each -- second procedure.

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GEORGE THOMPSON, MD: Yes. All right, well, let's go back to the operating room and carry on with the case. And here we are back, showing you the exposure. Up close to the spine, this upper portion -- of the upper portion of the pair of fused ribs, and we're just, again, creating a slot in which to put the cradle. And again, you see him just doing a very limited dissection at that point. There are dissecting tools that allow us to create a track beneath the muscle, but not -- but on top of the bone in order to properly seat the cradle. And that's what these tools are being used for now, we're creating a slot in which to put the upper fixation device. You had some e-mails there, Doug?

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DOUGLAS G. ARMSTRONG, MD: Yes, we do. We have a question from Chris Thompson. The question is: what is the risk of infection for this procedure, particularly with the lung being exposed?

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GEORGE THOMPSON, MD: Well, the risk for infection is probably based on the ideology of -- of the diagnosis. If you have a normal, healthy child, the risk for infection will be probably less than 1%, which is the same as it would be for -- for a spine procedure. Just because a lung is exposed does not increase the risk for infection significantly over a standard procedure. Now, if you end up having a very prominent rod and the skin over that is too tight, and the skin necrosis, then a secondary infection will be a real issue. But I think in the absence of having metal exposed through necrosis, the infection rate has been about 0.5 to 1%.

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DOUGLAS G. ARMSTRONG, MD: I don't think that they've had many infections throughout the country since they've started using this.

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GEORGE THOMPSON, MD: No, I'm not aware of any. I'm aware of the ones with skin necrosis more than I am those that were just for primary procedures.

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DOUGLAS G. ARMSTRONG, MD: Another question was: is there any way that this procedure can be carried out in a minimally invasive fashion?

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GEORGE THOMPSON, MD: No, not at this time, it cannot. As you'll be able to tell as you go through the remainder of the procedure, there's a very extensive exposure necessary. Now the one thing that is minimally invasive is when we go back to lengthen these devices, this is performed as an outpatient through a very limited incision. And that is minimally invasive, but this is -- this is a fairly significant procedure, although the blood loss, as you can tell, is not very high with this. It goes through some very important tissue planes which have a lot of normal motion of muscles and soft tissues and is not -- for the length of the incision, there's not near as much blood loss as you would think. What's happened now, we've actually joined the upper and lower cradles, and this is a compression device that's pushing them together, and they're getting ready to put the clip that will make the upper cradle a single circular unit, and that's what's going on at this time. And they're just tapping it into place right now. It's a special holder and a little tool that will seat the clip.

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DOUGLAS G. ARMSTRONG, MD: And a different sort of rib spreader in there right now.

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GEORGE THOMPSON, MD: Yes. As you noticed, the first set were just lamina spreaders, and now these are regular rib spreaders that will help, you know, minimize the force on each rib, but it's a much smoother construct that kind of circles the rib.

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DOUGLAS G. ARMSTRONG, MD: Now, do you use spinal-cord monitoring routinely? I'm sure somebody would want to know.

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GEORGE THOMPSON, MD: Yeah, I -- we use spinal-cord monitoring here because we are going to have an effect on the spine. And it's important that we make sure that we're not putting too much stretch on the spinal cord and the peripheral nerves. Interestingly, one of the major neurological problems with the VEPTR has been more the brachial plexus and the upper extremity rather than the lower extremity, and that's because some of these constructs are started very high in the chest wall, and they've actually put too much pressure on the brachial plexus as it exits through the foramina and are coming down, you know, adjacent to the neck and shoulder.

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DOUGLAS G. ARMSTRONG, MD: Well, it's very clo-- once you're up near the second rib, it's -- you're pretty close to the brachial plexus.

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GEORGE THOMPSON, MD: That's right.

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DOUGLAS G. ARMSTRONG, MD: There was one other question about the cost for the procedure: what's the total cost including hospital, surgery, anesthesia, and hardware? I don't know if we can answer that.

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GEORGE THOMPSON, MD: Well, I'm not sure, either, because it's -- that's a variable question and it's based a lot on what the insurance is going to -- to allow. I know this family told me tonight that the implants alone were about \$40,000. Now, that doesn't mean what's going to be paid, but that's what the -- the bill cost has been. And that doesn't include surgeon's fee, doesn't include the hospitalization, so, you know, I'm sure you're looking, you know, \$60-70,000, and that would just be a guess, but again, what the insurance pays, what the hospital accepts, these are all, you know, different. All right. Well, Doug, what do you think so far?

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DOUGLAS G. ARMSTRONG, MD: Well, it's pretty exciting.

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GEORGE THOMPSON, MD: Yes. I think -- I've been involved with VEPTRs now for about three or four years, although this is the first one we have actually utilized here at Rainbow, but I've been helping Dr. -- helping Dr. Campbell as far as FDA issues were concerned and billing issues as well, so it's -- it's been a long, arduous process for him, and we're just now -- like many other surgeons in North America -- are beginning to utilize this -- this device.

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DOUGLAS G. ARMSTRONG, MD: It was in development by -- for 14 years or so, wasn't it?

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GEORGE THOMPSON, MD: Yes, it was.

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DOUGLAS G. ARMSTRONG, MD: It's not something that was just brought in out of the basement and to the patient.

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GEORGE THOMPSON, MD: No. And I think the ones that we've already shown tonight are fourth or fifth generation, you know. They've been through many different design modifications over -- over the years before we've come to the one that we are currently using.

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DOUGLAS G. ARMSTRONG, MD: And in fairness to the company, the -- that designed this, they were very -- there were really no other companies that were willing to take it on and manufacture it, and I -- I was told -- I'm sure you were, too -- that the company that's manufacturing it really is probably not going to make a lot of money from it, and they really don't expect that they're going to be able -- be able to see that as one of their --

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GEORGE THOMPSON, MD: I think what you're saying is absolutely true. This is a device for a very limited, specific number of children. And, you know, companies make money off of, you know, large-volume sales, and that's not going to happen. Dr. Campbell, the inventor of this procedure, has only done about 250 of these, so you can -- and that's taken him 15 years to do that many, so the indications are few and far between. All right, let's go back to the operating room and we'll see the next segment. We're exposing the spine at this point, and this is about L2 or L3. And did not use an x-ray to determine exactly where we were. It was more based on the visual alignment between the spine and where the upper cradle is. So this -- this incision is just off the midline of the spine, that's dead center in the back, right as the spinous processes. And the spinous processes are those little bumps that you feel in your back. Each -- each vertebra has its own spinous process. And we're feeling for those, we're going to expose them, and then we're going to take the muscle off just one side. And that will be the right side, which is the same side the deformity is -- is on. And again, it's a combination of sharp dissection as well as electrocoagulation, and we're going to expose two spinous processes because we need to be over the top of them -- of the lower one so we can actually insert the -- the hook. Exposing the inc-- making the incision a little bit longer here just for better exposure. And we use electrocautery to divide the fascia, which is the covering of the muscle as well as to stop any bleeding from the skin edges. And then we begin to separate the muscle from the -- the bone. And in this case, we're talking about the spinous process and the laminae, which is the backside of each vertebra. And there's one -- there's a hemilamina on each side. And you'll see this more clearly as the video progresses. And here we are now just starting to divide the muscle right from the bone. And it's a very avascular plane, so there's not much bleeding once you get to this level.

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DOUGLAS G. ARMSTRONG, MD: While we're watching this, there was a question about whether the device would ever be removed or whether it would remain implanted for life, and what happens when the patient is going through a peak -- peak growth, peak -- big growth spurt?

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GEORGE THOMPSON, MD: Well, I think the one thing that we've learned about growing rods is that the best results occur when we lengthen the spine every six months irregardless of whether the curve has progressed, and so if you're doing it at six-month intervals, you should not have any problems as far as peak growth is concerned. I think the question of whether or not you take this device out or you exchange it when you've become mature or whether you go to a standard spine

device is an intriguing question because as we get to the end of this segment today, I -- we're going to talk a little bit about what we're going to do for this patient, because this is not a spine deformity -- a primary spine deformity, and we can maintain his spine relatively straight until maturity, I'm not sure he needs to have anything done to his spine. We very well may take this out. The majority of children who have had VEPTRs inserted have associated spinal deformity, and so at the very end of growth, they do undergo a fusion and either leaving the VEPTR in and using it as the primary spinal implant device or they exchange it out to a more standard device. But I think that's variable based on what the diagnosis is and exactly what -- you know, what type of deformity the patient has.

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DOUGLAS G. ARMSTRONG, MD: Well, it's interesting in this case that they're truly -- truly acting as an internal brace and that an external brace would have been ineffective or even harmful to him because of the pressure on the chest wall with his growth.

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GEORGE THOMPSON, MD: Right. Well, this patient we did not even treat with a brace after surgery, and he's already returned to relatively normal activities at three months after surgery.

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DOUGLAS G. ARMSTRONG, MD: Ordinarily would not brace somebody after this sort of procedure, do we?

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GEORGE THOMPSON, MD: Not usually because that may have a detrimental effect on their breathing capability. So the whole purpose of this device is to -- is to accentuate lung development, so we don't want to do anything else that may have an adverse effect on lung development.

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DOUGLAS G. ARMSTRONG, MD: There's one other question about the indications from someone who has a very small baby with 105-degree curve that seems to have failed -- failed Risser casting. Risser casting, if you could explain about that, but in this case, it would seem that they would like to know if it's useful for a severe infantile idiopathic curve.

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GEORGE THOMPSON, MD: Well, as I said earlier, Dr. Campbell feels this is not primarily a -- you know, a growing rod system. This is a chest wall system. So as a consequence, you know, I would probably favor more of a spinal implant rather than a VEPTR for that particular child, but I would have to see the child, look at the x-rays, you know, and see exactly, you know, what was -- what was going on, you know, with the child. 105-degree curve, as you know, is really a huge curve, and -- and I can tell you, our success at Rainbow with bracing has been very poor, but our casting program, the Risser cast, has been relatively successful in buying time. It certainly has not been definitive. Let me go back to the video for a moment. We've seen that the hook has gone in and is now being lined up to -- to accept the VEPTR device. And here's a subcutaneous tunnel that's being created from where that upper cradle is down to the spinal hook. And this will allow the path from which the device to -- to go through. To aid in its passage, we'll put a little rubber catheter that will attach to the device and allow it to be kind of pulled easily down this -- this slot in the subcutaneous tissues. But to answer the question about congenital scol-- or infantile scoliosis, that child at 19 months with 105-degree curve needs some type of treatment relatively soon because that child will have significant lung problems if not -- not appropriately treated. And with growth, the deformities in the vertebrae,

secondary just to the malrotation, will result in, you know, significant malformations of each vertebral body. Do you have any comments about that?

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DOUGLAS G. ARMSTRONG, MD: Well, I completely agree. It's a very, very difficult situation, and really, in many of these cases, I -- the best we can aim for is some -- some control. You can't -- you can't really aim for getting the spine perfectly straight, but we -- it's really very urgent to get that kind of --

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GEORGE THOMPSON, MD: Right. And I know from my own personal experience here at Rainbow, the children that have done the best are those that we treated early. We did not let them get to the huge curves of 80, 90, 100 degrees. The children that we were able to see when their curves were 30, 40 degrees saw them fail the other forms of conservative management and then put a growing rod system in actually did the best compared to those that presented late with the big deformities. All right, let's go back to the operating room. Now, here's the device. We're measuring the length. And the stem can actually be cut to accommodate -- they come in a variety of sizes, but it can also be cut to accommodate the appropriate length for the child. There's the rod being cut. And that's the lower round stem that I showed earlier. Now it's being attached to the -- to the catheter, which is just a plastic tube, and it just slides very easily down the tunnel. And then the plastic tube will be removed. Now you can see, the device is in place, and it will need to be attached both to the spinal hook and to the upper cradle. And this is what's going to be done at this time. The hook's been put back into place and there's a slot that the rod will go through, which has a set screw that then can be tightened to it. And the rod is pushed down through that slot until we have enough room in order to attach the VEPTR to the upper cradle. And this is what's happening at this point. These are just tools that help stabilize both the cradle as well as the VEPTR so they can be brought together and joined. And once they're properly aligned, a clip will be attached that will actually hold the cradle and the VEPTR device in proper alignment. So everything is basically in place at this time. And what we have to do now is there goes -- I think they're going to put the clip on the upper part. That joins the VEPTR to the upper cradle. And that's in place. And now he's going to put on this lengthening -- well, he's going to trim a little extra bone right now just to make sure there's no impingement. But then there will be a lengthening device attached down at the spinal hook that will allow the VEPTR to be linked or to be appropriately tensioned. Now you have to remember, in this particular case, the rib spreaders are in place, so this -- the chest wall has already been opened maximally, and so we're going to -- they're going to tension that, and you'll see that in the next segment, but that will then maintain the alignment of the chest wall once those retractors are removed. So, Doug, we have some more e-mails?

00:36:48

DOUGLAS G. ARMSTRONG, MD: Just a question as to the adjustments that will be need-- that might need to be made as the children grow up, but I think you addressed that when you said that it'll be lengthened every six months.

00:37:02

GEORGE THOMPSON, MD: Yes, it's lengthened every six months, but depending on the age of the child, the device could actually ultimately become too small, and when that happens, you have to replace the -- the VEPTR with a longer one, and that's a little bit larger operation. That's not quite as simple as just this -- as the lengthening of the device. But depending on the age of the child, that will happen. And like we were talking about that one child that was infantile idiopathic at 19 months, you know that child will require several, you know, system changes, no matter what type of system is utilized.

00:37:35

DOUGLAS G. ARMSTRONG, MD: One of the concerns, of course, is pain management after this. Was that a big issue for -- was pain a difficult problem for this child?

00:37:44

GEORGE THOMPSON, MD: They're fairly uncomfortable for the first week because of the elongation that's taken place. Now we're back to the video right now, and you can see we're putting the -- this is just a wrench device that's going to go on that's going to tighten the -- the set screw that will therefore join the -- join the rod to the hook. Now before it's finally tightened, we're going to actually do some distraction, and you'll see that coming up here in just -- just a moment. We're just now getting the device locked together. And that's going down at this point. Just getting it just ready to be elongated so that it can then be -- can undergo its final tightening. Now I think -- here comes the spreader, and this actually grabs on to the end of the rod and goes around the hook. And then when it's squeezed together, you're actually pushing up on the rod, you're lengthening the rod in the interval between the upper cradle and the hook itself, and that just appropriately tensions the device into place. And this is just another part of the -- the center or set screw that's being removed right now. But this is very strong, and this will prevent this from settling over time. All right, now there's the first VEPTR device in, and this is a rib/spine device. You can see it in place, and now the -- the rib spreaders have been removed for the most part. They're going to put one in just for a little bit longer while we put the second one in, but this will help hold the spine open while the VEPTR goes in, so you don't have to put a lot of pressure on the VEPTR itself with any initial lengthening. It's kind of already done by these devices here. Now the upper cradle's in place. And remember, this is going around a pair of fused ribs, so it looks a lot thicker than what it would be if it was a single rib.

00:40:12

DOUGLAS G. ARMSTRONG, MD: Very stable fixation.

00:40:14

GEORGE THOMPSON, MD: Yes. Those little prongs really help hold it in place and keep it from wanting to slide laterally. Here they are measuring the device to see which one of the systems we're going to be using for this location. And there it is going in place. But this one doesn't have the stem because it's a rib-to-rib device, as I showed earlier. And it's got the upper part of the cradle in there, so now we're going to get that fixed into place. And now the lower portion of the cradle will be applied in a very similar manner that we did putting the cradle on the upper part of the first device. This is the little slot being made between the intercostal muscles between -- around a single rib, you'll notice it's a lot smaller at this particular location. And a little tunnel will be made with the dissector so that device can be easily passed. This is just going from the top. They dissect -- they work from both the top and the bottom until the dissectors are actually touching, there's an adequate space to put the cradle in. and now here comes the cradle into place. Kind of goes in sideways and is rotated, slid together, and then there will be another little clip. And here comes the clip right now that will join the segments -- the two segments together. Now, we don't have to lengthen this at this time because this is already being held in proper alignment by those rib spreaders that you see in place. And that is basically it. You can see the are-- you can see the areas where lengthening will take place, and that's where the other little holes are. Well, that's kind of the completion of how the two VEPTRs went in, a combination of both rib/spine and rib-to-rib VEPTRs were utilized, and as we talked earlier, that's a fairly standard construct for this type of chest wall deformity. Or chest -- or fused rib constructs.

00:43:06

DOUGLAS G. ARMSTRONG, MD: The challenge will be to maintain coverage over the devices as the patient grows older.

00:43:11

GEORGE THOMPSON, MD: Yeah, that's absolutely the key. And we're going to -- we'll show in a little bit the closure of this -- of this wound, which is -- which is challenging. And there was a little bit of stretching of the soft tissues that have to go on prior to doing the closure in order to make sure that VEPTR is completely covered by healthy muscle with a minimal tension on it so that there'd be no tendency for that muscle to necrose and become in the subcutaneous tissue and perhaps even erode through the skin. Do we have any more e-mails?

00:43:41

DOUGLAS G. ARMSTRONG, MD: There's one comment from a -- a parent who says that their child would not be alive if it weren't for the VEPTR, although it was very expensive. And she's -- she hadn't had the opportunity to see the procedure before.

00:43:57

GEORGE THOMPSON, MD: Yeah. All right, we have another segment to review at this point, so let's go back to the operating room. Here is -- the device is in, the retractors have been removed, and I think you saw them shake the child to show you how -- how strong and stable this construct is. Here we are measuring the intervals that have been opened. You have to appreciate that all that area where the lung was exposed was at one time, you know, completely covered with by the chest wall, and this is how much we've elongated this patient. And I would estimate that's eight to ten centimeters, if not more that we've been able to achieve, you know, just with the insertion of these two devices.

00:44:40

DOUGLAS G. ARMSTRONG, MD: Are you going to have to close the -- cover over the chest wall with Marlex or any --

00:44:46

GEORGE THOMPSON, MD: No, we elected not to, and the -- I know in a primary VEPTR where the lung would collapse, he has used some type of device to prevent adhesions from occurring, but this one, as you can see, the lungs did not even fall -- the lung did not fall away and there was already a lot of scarring, so he didn't feel this would really be very helpful in this particular case, so it was just closed primarily. But I hope the audience can appreciate how much lengthening has occurred. That -- where you see the -- that second VEPTR, that's almost in your -- right underneath your armpit, the midaxillary line. That is a tremendous amount of lengthening that has gone into that -- that chest wall. And that -- because of the effect on the ribs, it has a secondary effect on the spine itself. There's another view of it a little bit farther away. And it's a -- it's a formidable procedure to go through.

00:45:44

DOUGLAS G. ARMSTRONG, MD: Most of the time, these patients are very unwell with a restrictive element to their lung disease, and it -- do you usually see a lot of lung problems in the immediate postoperative period with these people, or...

00:46:01

GEORGE THOMPSON, MD: I think that, Doug, that depends on the diagnosis. This -- this boy did not, but he was otherwise a normal child with normal -- relatively normal lung function, normal activity level. Children who have other syndromes, who are not as healthy, are much more likely to have significant problems. In fact, many of these children spend a week or two in the pediatric intensive care unit after surgery before they are healthy enough to be on the floor and ultimately discharged home.

00:46:29

DOUGLAS G. ARMSTRONG, MD: Well, by spreading open the ribs like that, are you able to measure a difference in the lung capacity at some point afterward -- after the procedure?

00:46:39

GEORGE THOMPSON, MD: Yeah, sometimes that does occur. All right, well, let's go to the last segment, and we'll see the closure of this -- of the wound. Now that the VEPTR's been inserted, the wound will be closed, but what's happened is that the chest wall has been so elongated it's difficult to reapproximate the tissues. So what's happening now is that we are stretching the soft tissues and the muscles about the shoulder by grabbing the tissues with a towel and then pulling gently to achieve as much elongation as possible. This will allow us to close the muscle cuff with the least amount of tension and prevent the possibility of having the wound problems postoperatively. You can see the towel around the muscle that allows us to be able to hang on to the wet tissues and then to stretch them. At this point, the muscles about the shoulder are being reapproximated over the VEPTR device, and good wound closure is critically important because you want this to be well covered to prevent any possibility of -- of erosion of the soft tissues over the -- the VEPTR device. It's not possible to close the muscles -- the intercostals muscles between the ribs because they've been spread so far open. As you recall from the previous segment, this has been lengthened approximately eight or nine centimeters. So this is left open. Occasionally, if you had -- not had previous thoracotomy, you might consider covering the lung tissue with a -- with material to prevent scar-tissue formation. This would allow the lung to have a gliding surface over which to slide if we did not have as much scar tissue formation. However, in this particular case, there's quite a bit of scar tissue already from the previous surgeries, and we did not feel this would be -- would be helpful for this patient. So the muscle cuff is being repaired with absorbable sutures at this time. This typically takes quite a while because it's a fairly large incision. And it's very important that we get a very good closure in order to prevent the muscles from separating postoperatively, and this would be done in layers. Following wound closure, we put the patient back in a more normal position on the table and we take an x-ray, or radiograph, to see what we've achieved, also to make sure the lung is fully inflated. Chest tube is typically necessary in order to keep the lung inflated until the soft tissues begin to heal. You'll notice on this postoperative x-ray that there's been considerable improvement in the spine, and this is achieved strictly by releasing the tether of the two pair of fused ribs and then the insertion of the VEPTR device. You'll notice the one closest to the spine is a spine/rib construct while the one that is most lateral or to the side is a rib-to-rib construct. But in concert together, we have reduced this curve from 37 degrees to 10 degrees. Now, as he grows, we anticipate the curve will slowly recur because there's not going to be much separation of the ribs with growth on the right-hand side, so as he grows, the curve will slowly recur. So the plan will be to go back at six-month intervals and to lengthen the VEPTR devices. And this will allow us to maintain maximum correction of the spine while at the same time allowing him to have relatively normal growth. Most children who have this device in require a final fusion. I'm not certain at this time if he will require a fusion because we're not treating a purely spinal deformity, we're treating a chest wall deformity. So when he reaches maturity, the devices may be able to be removed and he'll have normal spinal mobility. Now let's meet the family. Well, I'd like to welcome everybody to the Iacobucchi family. We have Mary and we have Anthony here tonight. Anthony was the patient you have seen having surgery, and that was just four months ago. So, Anthony, we've had some questions tonight. In fact, I have one here from the United Kingdom wanting to know about pain and physical therapy afterwards.

00:51:29

ANTHONY IACOBUCCHI: It was a lot of pain.  
00:51:31  
GEORGE THOMPSON, MD: How long did your pain last?  
00:51:33  
ANTHONY IACOBUCCHI: A couple days.  
00:51:35  
GEORGE THOMPSON, MD: Just a couple of days?  
ANTHONY IACOBUCCHI: Mm-hmm.  
00:51:38  
GEORGE THOMPSON, MD: How was it when you went home?  
00:51:40  
ANTHONY IACOBUCCHI: I felt a little sore.  
00:51:43  
GEORGE THOMPSON, MD: Did you have trouble sleeping at night, or were you able to sleep pretty well?  
00:51:46  
ANTHONY IACOBUCCHI: I had a little trouble sleeping.  
00:51:48  
GEORGE THOMPSON, MD: All right. But you were able to be up and about during the daytime?  
00:51:50  
ANTHONY IACOBUCCHI: Mm-hmm.  
00:51:51  
GEORGE THOMPSON, MD: How long did it take you to get your shoulder motion back?  
00:51:57  
ANTHONY IACOBUCCHI: A couple weeks.  
00:51:58  
GEORGE THOMPSON, MD: A couple of weeks. All right, now you're not wearing any brace or anything now, are you?  
00:52:02  
ANTHONY IACOBUCCHI: Mm-mm.  
GEORGE THOMPSON, MD: All right. And are you back to pretty normal activities, or are you still taking it easy?  
00:52:07  
ANTHONY IACOBUCCHI: I'm still taking it easy.  
00:52:09  
GEORGE THOMPSON, MD: What do you mean by that? Are you playing any sports?  
00:52:13  
ANTHONY IACOBUCCHI: No.  
00:52:14  
GEORGE THOMPSON, MD: Can you throw a ball?  
00:52:15  
ANTHONY IACOBUCCHI: Yeah.  
00:52:16  
GEORGE THOMPSON, MD: All right. Do you play catch with Dad sometimes?  
00:52:18  
ANTHONY IACOBUCCHI: Yeah.  
00:52:19  
GEORGE THOMPSON, MD: All right. Mary, how was your experience in the hospital here at Rainbow Babies & Children's Hospital?  
00:52:24

MARY IACOBUCCHI: Well, as you know, we've had a lot of experience here at this hospital, and, you know, the staff and yourself and your staff, it was just incredible how -- because this was so new to us and, you know, for Anthony, we've never experienced any procedure like this, so we really, going into it, didn't know what to expect. And the staff was just so informative and just told us exactly, you know, what we can expect. And Connie and everybody, you know, "This is what to expect after surgery and, you know, the weeks following surgery." And of course the experience here was, as always, was tremendous. You know, everybody just took such good care of Anthony and our whole family, so we feel very, very blessed to be at this hospital.

00:53:08

GEORGE THOMPSON, MD: All right. Are you looking forward to your first lengthening?

00:53:12

ANTHONY IACOBUCCHI: Yeah.

GEORGE THOMPSON, MD: Yeah? That's not going to happen for a while just yet. Doug, do you have any comments for the family?

00:53:19

DOUGLAS G. ARMSTRONG, MD: No, I can't believe he's the patient. He looks so good.

00:53:22

GEORGE THOMPSON, MD: Yeah. And --

00:53:24

MARY IACOBUCCHI: Yeah, he's -- he's doing great.

00:53:25

GEORGE THOMPSON, MD: I know when you look at his back, you don't really even see the implants. There's one little area, especially the one that's, you know, farthest out that you can just see a little rise in the tissue, but that doesn't cause you any pain, Anthony?

00:53:37

ANTHONY IACOBUCCHI: Mm-mm.

GEORGE THOMPSON, MD: Are you aware it's there?

00:53:40

ANTHONY IACOBUCCHI: No, not really.

00:53:41

GEORGE THOMPSON, MD: If you put your hand back there, do you know you're feeling it, or...

00:53:43

ANTHONY IACOBUCCHI: Yeah.

GEORGE THOMPSON, MD: But otherwise, you can move your arm, do all your schoolwork, everything is just fine?

00:53:49

ANTHONY IACOBUCCHI: Mm-hmm.

00:53:50

MARY IACOBUCCHI: I mean, I think, really, if you see him with just his everyday clothes on, you would never even know that he ever even had anything done. You know? He's real thin, so you can kind of see, you know. But we're working on that.

00:54:03

GEORGE THOMPSON, MD: Well, the thin children are the ones that we are most concerned with about having problems because they're the ones most likely to have the implants visible and maybe have too much pressure on -- on the skin. Now, for a normal, healthy kid such as Anthony, that's unlikely to happen, but certainly some of the compromised kids, it's been a problem.

00:54:21

MARY IACOBUCCHI: Right.

GEORGE THOMPSON, MD: So...

00:54:23

MARY IACOBUCCHI: I think the first, you know -- I mean, this was definitely, like I say, a different procedure than any of the other ones he had had because most of the ones have been through his abdomen and his chest, so this was definitely something different, and the pain level was definitely higher, you know, for him, so - - which as a parent, you don't ever want to see, but...

00:54:42

GEORGE THOMPSON, MD: Well, I think we've all noticed over -- over time that, you know, these elongation procedures, although some of the muscles are cut, others are under a lot of tension, they're being stretched, and it does take a period of time for the muscles to accommodate to that stretch phenomena that we have induced by the implant itself.

00:54:58

MARY IACOBUCCHI: Yeah, I know. I'm sure that -- you know, I didn't watch the video, but I'm sure that it was an extremely invasive procedure, and knowing that, I knew that the recovery was going to be, you know, difficult for him, but I -- we kept on him every day to keep up with the stretching and the rotating the arm and everything. We didn't -- we didn't let him slack, so...

00:55:17

GEORGE THOMPSON, MD: All right.

00:55:19

MARY IACOBUCCHI: But he was -- he was a trouper. He -- he's amazing.

00:55:22

GEORGE THOMPSON, MD: All right. And he hasn't had any physiotherapy. That was another question that came tonight. The physical therapy is usually not necessary. In fact, I think our experience at Rainbow is that we have trouble slowing kids down, not getting them going.

00:55:32

MARY IACOBUCCHI: Mm-hmm. I mean, they came to his room, you know, the first few days in the hospital, they did, to help us get him out of bed the first time and everything because that was really difficult, the first time getting out of bed. So they did help us here in the hospital, but then once we went home, they just told us what exercises we should do, and we just followed that routine.

00:55:48

GEORGE THOMPSON, MD: Okay. There was another -- I was going to say, there was another e-mail tonight that -- from a family who's had a VEPTR done, and the cost of the implants, \$138,425, and I was quoting tonight earlier --

00:56:03

MARY IACOBUCCHI: Whoa.

00:56:04

GEORGE THOMPSON, MD: -- that yours were 40, you know. And even that sounds outrageously high.

00:56:07

MARY IACOBUCCHI: Maybe you need to recheck that bill, because maybe it was -- it was 40 each, it could have been 80.

00:56:11

GEORGE THOMPSON, MD: Maybe in -- and that may have been the total bill, you know, but --

00:56:15

MARY IACOBUCCHI: Yeah, yeah. Our total bill was probably somewhere in that range, too, so that sounds --

00:56:19

GEORGE THOMPSON, MD: So just be -- but it still is -- the cost is not the issue. The issue -- those are all worked out through the hospitals and the insurance companies.

00:56:25

MARY IACOBUCCHI: Right.

00:56:26

GEORGE THOMPSON, MD: Because that's not the cost to the families, and I think people need to be aware that that doesn't -- that's not what's going to happen to them.

00:56:30

MARY IACOBUCCHI: Right, absolutely.

00:56:31

GEORGE THOMPSON, MD: Because there are ways of -- of dealing with these types of issues.

00:56:34

MARY IACOBUCCHI: Right.

00:56:36

DOUGLAS G. ARMSTRONG, MD: Anthony -- did Anthony get back to school right away?

00:56:38

ANTHONY IACOBUCCHI: Yeah.

00:56:40

MARY IACOBUCCHI: Yeah, we got back to school. I couldn't believe it. A month -- actually, about almost a month exactly after the surgery, he returned to school. I was very nervous about having him go back to school and just met with all his teachers because he was entering a new school with the bigger kids, a middle school, so I was very nervous. I met with all the teachers and told them what was going on and just said, you know -- and I -- and I had many talks with Anthony before he went back to school, too, about, "if you feel uncomfortable, you know, go see the nurse, she knows to call me." And, you know, we never had any problems, so, you know...

00:57:13

GEORGE THOMPSON, MD: Does he have any breathing problems or anything, Mary, before surgery? As I remember, he wasn't.

00:57:18

MARY IACOBUCCHI: He wasn't really having any breathing problems. The only problems he ever really had -- anything like running out of breath and stuff -- was before the last diaphragmatic hernia repair, which was about three years ago. That was the -- like, a couple months before that surgery, that was when he was having trouble -- getting short of breath and everything, and that was when, you know, Dr. Magnuson decided, you know, we needed to -- to repair that hernia again. But before this surgery, no, he was having no problems with breathing at all.

00:57:44

GEORGE THOMPSON, MD: All right. And do you notice any difference in your breathing ability now? About the same, I would guess.

00:57:50

ANTHONY IACOBUCCHI: Yeah, it was.

00:57:52

GEORGE THOMPSON, MD: You're able -- can you run and have pretty good endurance?

00:57:54

ANTHONY IACOBUCCHI: Yeah.

00:57:56

MARY IACOBUCCHI: Ask his sister. I think he chases her.

00:57:58

GEORGE THOMPSON, MD: All right. I thought that was his girlfriend, but it was your sister, right? You don't think that's funny, do you? Okay. Okay. All right.

00:58:09

MARY IACOBUCCHI: He's got a lot of time for that.

00:58:11

GEORGE THOMPSON, MD: All right. Doug, any final questions?

00:58:13

DOUGLAS G. ARMSTRONG, MD: None that I can think of. You look amazing, Anthony. Congratulations.

00:58:20

MARY IACOBUCCHI: It's tremendous. And thank you to you and your staff and --

00:58:23

GEORGE THOMPSON, MD: Well, we're -- we're always happy to have satisfied patients, and, you know, it's one of the things we pride ourselves on here at Rainbow. We have a -- a tremendous spine program here, we're one of the largest in the country, and we try and do cutting-edge things for anybody who -- who really needs them, and you're an example of that. And we're -- we're appreciative that you selected us, and we were delighted to have him be as successful as he's been.

00:58:46

MARY IACOBUCCHI: I think it was just meant to be, Dr. Thompson. It was just one of those things, you know?

00:58:50

GEORGE THOMPSON, MD: Well, thank you very much.

00:58:52

MARY IACOBUCCHI: Thank you.

00:58:53

GEORGE THOMPSON, MD: So -- well, ladies and gentlemen, we'd like to thank everybody for tuning in tonight to see the insertion of the VEPTR patient -- the VEPTR device in a pediatric patient. I hope this has been informative for you. This is a very cutting-edge procedure that's being done throughout the world, but primarily in North America and in a very limited number of -- of cases. And this is a prime example of what this device can do in the right situations. So I'd like to thank Anthony and Mary and Dr. Armstrong for joining me tonight. We look forward to seeing you again at some point in the future. Thank you very much.

00:59:32

ANNOUNCER: Thank you for watching this webcast featuring the implantation of a Vertical Expandable Prosthetic Titanium Rib from Rainbow Babies & Children's Hospital in Cleveland. If you would like to make an appointment, receive a physician referral, or request more information about the procedure, please click the buttons on the screen.

00:59:57

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