



**AVOIDING FUSION IN EARLY ONSET SCOLIOSIS
MORGAN STANLEY CHILDREN'S HOSPITAL
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ANNOUNCER: Welcome to Morgan Stanley Children's Hospital of New York-Presbyterian. Over the next hour, you'll see an expert discussion of VEPTR and growing rods. Patients with early onset scoliosis can develop severe, complex spinal deformity that distorts and reduces the volume of the thorax. The resulting condition can compromise respiratory function and be life-threatening. In just moments, you'll learn how advances in the use of growing rods and VEPTR are providing better outcomes than traditional spinal fusion. OR-Live makes it easy for you to learn more. Just click on the "Request Information" button on your Web cast screen and open the door to informed medical care. Now let's join the doctors.

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MICHAEL G. VITALE, MD, MPH: Hi, and welcome to our Web cast, brought to you live from the Morgan Stanley Children's Hospital of New York-Presbyterian. I'm Michael Vitale, chief of pediatric spine surgery here at Morgan Stanley Children's Hospital of New York. And over the next hour, we'll be speaking about ways to avoid fusion in young children with early onset scoliosis. We'll be detailing some of the newly available techniques that allow us to grow the spine and allow growth of the lungs. These new techniques promise significantly improved outcomes for children with early onset scoliosis. Through the use of live video, we'll detail some of the techniques that are now available. Let me introduce now the rest of our team here. To my right is Dr. David Roye, chief of the division of pediatric orthopedic surgery here at the Morgan Stanley Children's Hospital of New York. With over 25 years of experience, Dr. Roye is a key part of our surgical team. To Dr. Roye's right is Dr. Michael Bye. Dr. Michael Bye is a pulmonologist in the department of pulmonary medicine here at Morgan Stanley Children's Hospital who has a passion in the care and treatment as well as the elucidation of new research in treatment options for children with early onset scoliosis. To Dr. Bye's right is Dr. Lawrence Bodenstein, a member of the department of general surgery, an important part of our team in the operating room who assists in the treatment of young children with early onset scoliosis. We're going to be speaking a little bit about a multidisciplinary approach to treating early onset scoliosis. The fundamental issue is how best to treat the young child with scoliosis. In this presentation you'll learn about progressive, complex deformities of the spine and thorax. And the fundamental question is what is the best way to optimize the outcome in young children with scoliosis. Here you see a two-year-old with a rapidly progressive spinal curvature. More importantly, this child is developing a significant problem in their thorax, a significant problem in their chest wall. They have rib fusions and multiple congenital anomalies. Treatment options for this child have dramatically improved over the last five to ten years, and we're going to be speaking about this over the next 45 minutes. Orthopedics means "straight child," and it's gratifying as a surgeon to obtain a straight spine in a child with a spinal deformity. But really we now understand that we need to be looking at much broader outcomes than just x-rays. We need to, to some extent, forget about the x-rays and to think more about the children

globally, to think about their lungs, to think about their thorax, to think about their quality of life. Newly available treatment options allow us to do just that. We know a lot about the natural history of scoliosis. For the average teenage or adolescent girl who comes in with idiopathic scoliosis, outcomes are excellent. There's no difference in mortality between children with adolescent scoliosis and the normal population, as this graph shows. This is really not the case in children with juvenile idiopathic scoliosis. It's been shown that children with juvenile scoliosis start to have a difference in life expectancy at about 40 years of age. And taken one step further, in children who develop scoliosis as infants, life expectancy is significantly altered with a significant difference, two times the death rate by the age of 40. Our goal is to change this natural history to make these curves coincident rather than separate. And newly available options allow us significant hope at doing just that. I'm going to ask Dr. Bye now to speak about lung growth at an early age and about what we know about lung function and its relationship with scoliosis. Dr. Bye?

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MICHAEL R. BYE, MD: The slide on the right reminds us that children grow lung tissue until about 8 years of age, although much of that occurs in the first two years of life. In fact, if you're born at term, you have about 20 million alveoli, and by the time you reach your max, you've grown to about 300 million. So there is a significant amount of growth that occurs over those first eight years, and especially over the first two years. As pediatric pulmonologists, we think of the chest wall primarily as a bony box that protects the lung. But if, as we'll see over the course of the next hour, that box is too small or too crooked, that lung will not be able to grow properly and adequately, and therefore will not be able to function as well as it should, and that's what Dr. Vitale and his group are all about. This is a compendium of a few of the studies that have been done showing improved pulmonary outcome in children who have had early intervention with their chest wall anomalies. These are data from our own institution, the Morgan Stanley Children's Hospital of New York-Presbyterian, and these are patients, 20 patients who were studied at about 13 years of age, and they had had their surgery at about 6 years of age as a mean. So many of them had already reached a lot of their lung growth. What we see here, looking at their pulmonary functions in adolescence when they should have reached their max, is that the FVC, or forced vital capacity, the vital capacity is you take a maximal breath in and you blow out completely, so it's the air that you have available to use. And we see that this group had about 70 percent of predicted, with some patients going as low as 39 percent. And if we look at total lung capacity, take a deep breath and how much is in there, they're at 87 percent but down as low as 57 percent. Now, if you get a 39 or a 57 on a math test, you're going to have to do better and take it over. By the time you reach these lung functions at this age, you don't have that chance anymore.

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MICHAEL G. VITALE, MD, MPH: Dr. Bye's study of lung function adds to a growing weight of evidence showing significant problems with lung function in children treated with early fusion. Here is an example of a young child who presents with a significant curvature at an early age, before the advent of the new technologies that we're discussing now. This child underwent an early fusion, a relatively straight spine, but unfortunately, a short spine with lung function only 20 percent of normal and a thoracic spine the size of a newborn. This child unfortunately went on to an early demise because of pneumonia. These are the outcomes that we can avoid by avoiding early fusion, and new technologies should allow us to do this. This problem has been described by Dr. Robert Campbell in Texas as thoracic insufficiency syndrome, and in fact, thoracic insufficiency syndrome is a known entity now that's described as the inability of the thorax to support normal respiration or lung growth. Thoracic insufficiency syndrome, or TIS, can be a primary chest wall disorder, as you see in children with Jeune syndrome, or Jarcho-Levin syndrome, for example, or it can be secondary as a result of chest wall disorders or scoliosis. We have children with spinal muscular atrophy and other known muscular disorders who develop lung problems as a

result of secondary thoracic insufficiency. Finally, thoracic insufficiency can be acquired, and early fusion in fact results in acquired or iatrogenic thoracic insufficiency syndrome.

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MICHAEL R. BYE, MD: The lung and the chest wall interact quite extensively during respiration, during breathing. And if the chest wall is insufficient, then we can look at and see both consequences in the short term and in the long term. One of the things that is important to the lung is the ability to cough properly. You get a cold, you're going to have to cough to get rid of secretions, and if you can't do that, then you're at risk for developing asthma -- I'm sorry, pneumonia. Probably asthma too. If that happens and the chest wall cannot perform adequately, then, as Dr. Vitale mentioned in the last young lady, the risk of pneumonia and death from pneumonia becomes quite high. The lung and the chest wall both need to increase their ventilation when you're exercising. You put greater demands on the chest, you create more carbon dioxide that you have to get rid of, you need more oxygen. And if the chest wall and the lung are not in sync, are not able to do that, then the child will at least have diminished exercise tolerance and may even get to the point where he or she cannot exercise adequately. There are some patients where the chest wall is so twisted that the airway becomes twisted. And in fact, some years ago, probably some decades ago, I reported a child with the Jarcho-Levin Syndrome whose trachea actually made a 90-degree turn within the chest wall, and that child did not do well. The increased airway resistance didn't allow the child to breathe adequately, and the child didn't do well in the long term -- actually didn't have a long term. Pulmonary hypertension is the result of inadequate amounts of oxygen delivered to the blood. That's a common problem in children with thoracic insufficiency. And when these children develop their pulmonary hypertension, they are then at risk for developing what's known as core pulmonality, or increased stress on the right heart, with eventually high degrees of mortality. Failure to thrive with poor weight gain is the result of the insufficient lung tissue, the inability to breathe adequately, and using more calories to breathe. If you use more calories to breathe, then you don't have enough calories to grow, and that's where that failure to thrive with poor lung growth comes in. The two major jobs, as we know, of the lung are to get rid of the carbon dioxide and to bring the oxygen in. And we've already talked about how pulmonary hypertension can result from the hypoxemia. If the lung tissue is inadequate to support the normal breathing, the normal metabolic demands of life, then the carbon dioxide levels will increase, and eventually that will cause an acidosis which cannot be overcome and then require the child to undergo chronic mechanical ventilation. And lastly, of course, in the long run, as we showed a couple of slides ago, the concern is that if the chest wall is insufficient, is too small or too crooked, then that lung will not grow adequately and we'll not only see some of these problems in the short but in the long term, resulting in the increased mortality rates that Dr. Vitale talked about.

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MICHAEL G. VITALE, MD, MPH: So in 2008, there are options for avoiding early fusion. While early fusion was the traditional treatment of young children with scoliosis, we now, for all the reasons described and all the reasons understood, we now understand that young children and little children are really not just little adults, that we need to treat these children differently. Early fusion is really not the preferred approach, and new strategies have evolved which allow growth of the spine. There are several growth strategies which we'll detail in the minutes to follow, strategies that allow growth of the spine and growth of the thorax. We're speaking specifically about growing rods and the VEPTR. Additionally, there are technologies that are just on the horizon that promise significantly improved outcomes. One of these is the Shilla technique. This is the example of just such a technique shared with us by Dr. David Skaggs in Los Angeles. This is a technique where a very limited fusion is done just at the apex of the curve. The spine then by the attachment through a special type of screw mechanism can grow away from this rod. This allows growth of the spine and I think will be a significant advance in the treatment of young children. Another

technique which is just on the horizon and we're just embarking on here at Morgan Stanley Children's Hospital is anterior growth modulation. Through growth modulation, we feel that it will be possible to stop the curve from developing and in fact allow the curve to reverse itself. This is done by the insertion of staples using a thoroscope, a camera inserted into the chest, with one such example shown here. In the next minutes, we'll speak specifically about growth strategies, growing rods and the VEPTR. Growing rods are rods that are implanted that are lengthened serially over time. This does not involve extensive fusion. This allows us to delay definitive fusion, and through the use of lengthening, expand the spine, expand the thorax, allow lung growth, and control the curve. More recently available is the VEPTR, or the vertical expandable prosthesis titanium rib. This is a new device which has in the last five to ten years shown very positive results in a select group of patients that not only treats the curvature but allows us to treat the chest wall deformity and the thoracic insufficiency that is so often present in these children. Results of growing rods have been generally positive. A number of authors have in fact shown that the use of growing rods allows us to control large curves to expand the thorax, to allow the thoracic spine to grow with some but relatively acceptable complications. The question is, in 2007, based on what we know, how can we improve on our experience to date? And I think that we can. This is an example of a recent case where Dr. Bodenstern and I performed an expansion thoracoplasty. The approach here is the approach towards a young child with scoliosis and fused ribs. We will go in and surgically separate the ribs, insert the VEPTR, which is the device seen on the right, allowing the ribs to expand. This is an example of what the VEPTR looks like. The VEPTR is made by the Synthes Corporation and is an expansion device made of titanium that inserts on ribs above and below the deformity. We have a model of one such spine and one such device here. And here you see the insertion of two VEPTR devices on a model of a young spine. In many cases, the device attaches only to the ribs, avoiding the spine entirely, and at other times we do attach the spine. One of the things that's particularly interesting and dramatic is the effect of insertion of the VEPTR on the lung. The slide on the right in fact shows how the lung bulges out to meet the environment after it's been constricted within a chest cavity for a long period of time. The approach to both growing rods and VEPTRs involves serial lengthening, and we'll go ahead and lengthen these growing rods at regular four to six-month intervals over time. As we discussed, either a traditional growing rod or a VEPTR device can be inserted either to the spine, here an example shown inserting into the lumbar spine, or to the pelvis. And going back to our presentation, the slide to the right here shows the VEPTR inserted down to the pelvis on both sides. The use of a VEPTR or a growing rod allows us to completely avoid the spine, completely avoid fusion, and really promises to allow maximum growth of this child's spine over time.

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Based on both our experience at our institution and the experience in growing rods to date, we feel there are a number of general principles which promise optimum outcomes and success in the use of growing rods. We feel that dual rods, the use of two rods, are particularly important, that stable foundations based on pedicle screw constructs allow us a more solid area for support. We feel that long constructs allow us to share the load, share the stress of these constructs over time. Sagittal plane, or the lateral x-ray, is really critical. And the use of the VEPTR or appropriate growing rod constructs allows us to maintain the normal sagittal curvature that's so important to growth and function. It's clear now that in order to optimize outcomes, lengthening needs to be done frequently at four to six-month intervals. It's a key part of the procedure and a part that our general surgeon here, Dr. Bodenstern, is a particular help with, is the meticulous attention to soft tissue closure. Because many of these kids have issues with nutrition and soft tissue coverage, we really emphasize the need to really close these kids in a way so that we minimize the chance of wound problems and the chance of infection. And again, whenever possible in the use of growing constructs, we want to avoid the spine so that we don't have any fusion whatsoever.

Even before we get into the operating room, several things allow us to optimize outcomes. And in terms of making these procedures as safe as possible, appropriate electrophysiological monitoring is really key. Somatosensory evoked potentials, transcranial motor evoked potentials, and appropriate upper and lower extremity monitoring is really critical, and we're very fortunate here at the Morgan Stanley Children's Hospital of New York at having a great team that allows us to monitor very specifically very young children. Appropriate imaging is key. Some of these children have complex deformities that involve not only a very disordered spine but the thorax. X-rays give us some information, but we're able to obtain CT models. And here you see a CT scan in the middle and then an actual physical model on the right that we actually can have constructed that we take into the operating room that allows us to guide the surgery. On the bottom you see an expansion thoracoplasty being performed by Dr. Bodenstern and myself, which really mimics the three-dimensional model. In the next minutes, we will be detailing with the use of live video both the traditional growing rod technique and the vertical expandable titanium rib construct. The first case involves a 5-year-old girl. As many of these kids, she was in the fifth percentile for weight. As Dr. Bye alluded to, many of these kids have nutritional issues because they're working so hard to breathe because of their diminished lung function. This young child had a tetralogy of Fallot, a complex cardiac problem. She also had significant gastrointestinal problems and a very rapidly progressive scoliosis. These are the x-rays of this young child initially and then one year later. And here you see the rapid evolution of her scoliosis. Dr. Roye will now take us through the actual surgical procedure of this young child. She ended up having a growing rod implanted. And here you see a dramatic improvement of not only her spine but the space available for her lung, her scoliosis and the room available for her breathing. Dr. Roye?

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DAVID P. ROYE, Jr., MD: We have a choice of incisions, which we're going to see going forward. We can do a long midline incision or multiple incisions. When we do the long midline incision, however, we don't approach the spine, we simply use it to help guide the rod and to allow for future surgeries and lengthenings. But the incisions will be dictated by the placement of the lengthening device itself. The -- this exposure video shows us approaching, doing a meticulous subperiosteal approach to that lumbar foundation. We need an area that's completely exposed so that we can get a solid fusion in that area where we want it. Above that level we don't want to approach the bone at all, so we'll go paramedian to allow for those areas to escape fusion, to escape any bone encroaching on that area to cause the area to fuse that's beyond our foundation technique. Our lumbar screw placement is typical lumbar screw placement that spine surgeons are quite familiar with. We would want to see the anatomy very well to see the facet joint, to use the middle of the transverse process as our cephalad-caudal guide, and the medial lateral starting point is determined by the joint. The lumbar screw wants to be small enough for the pedicle, appropriate, but we want it to be as long as possible for good fixation. So we want to make sure that we measure very carefully to maximize length and not to, of course, exceed length and go through the body interiorly.

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MICHAEL G. VITALE, MD, MPH: And I'll just say that one of the advances in recent years has been the introduction of pediatric systems, of systems for young children with small stature that allow us to utilize screws of appropriate size and length for pediatric deformity. In the last years we really have seen a significant innovation in terms of options available for pediatric spine deformity.

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DAVID P. ROYE, Jr., MD: That's certainly true, Dr. Vitale. The thoracic screw technique is again the usual one: meticulous exposure so that we have the topographical anatomy. We do recommend in our service to removing the facet with an osteotome, although there are other techniques. And that osteotome technique we will demonstrate here, that allows for

accurate placement in the pedicle of the appropriate-sized screw. And again, not approaching subperiosteally any of the other structures surrounding that area that we want for our foundation. You can see here, using the osteotome to create that dissection and to expose the anatomy that we need by moving the facet. Once those solid foundations have been established, through appropriate placement of those thoracic screws, we then have an opportunity to push hard on those foundations and to allow correction of the deformity.

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MICHAEL G. VITALE, MD, MPH: As the previous video just demonstrated, it's really key to pay careful attention towards anatomical placement of your screws. Particularly in surgical cases where you have relatively limited exposure, an anatomical understanding of appropriate screw placement is critical. In the previous video, you saw, for example, that at the T2 pedicle, we are starting right at the midline of the transverse process. And each of the starting points for pedicle screws are well documented and well demonstrated. It's really important to understand screw placement at each of the different levels and screw location in order to be able to obtain a solid foundation, which is so important in these growing constructs.

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DAVID P. ROYE, Jr., MD: After these foundations have been established, we have to get a rod in. So that's accomplished by tunneling the rod. We are not approaching the spine through that midline incision, as we said earlier, but rather passing this long clamp from the distal incision through the muscle and above the spine, of course, and out the upper incision, or the upper dissection, as you see here on the video. At this point we can then pass back through this tunnel we've created a chest tube which will allow the safe passage of the rod from distal to proximal. We want to measure that axial connector. We know it has to fit between T10 and L1. That's the straight segment. Here we are with the rod placement on this portion of the video. And again, you can see that the tunneling and the chest tube make that a safe passage and avoid damage to the chest wall -- or penetration of the chest wall. There is a -- we want to carefully contour the rod after we've determined the length. We want to have an appropriate sagittal contour in particularly the lumbar lordosis, although keeping in mind that the segment that's in the lengthener has to be straight. If we contour the lordotic segment into the area that's supposed to fit into the end-to-end connector, it won't move properly in that segment. Of course, the appropriate kyphotic correction above is also -- is equally important.

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MICHAEL G. VITALE, MD, MPH: And I just want to emphasize how important the sagittal contour, the lateral contour, of our instrumentation is. I think we've learned from experience over time that some of the problems with growing rod constructs had to do with an inadequate amount of lumbar lordosis. With the tension towards lumbar lordosis and solid foundations using polyaxial pedicle screws, I think many of these problems in the past are diminished or completely avoided.

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DAVID P. ROYE, Jr., MD: When that rod is placed, we are just about ready to do the correction and lengthening. Here you can see the rod sliding in and out of the connector, which is then placed. And that distal portion of the rod is going to be fixed in the screw heads. There's a -- in general, we will start the process of the tensioning and lengthening by tightening the screws in the upper portion of the spine, the upper foundation, after we've mated the rods within the connector. In the end, we want to leave a little bit of space between the ends in order to achieve lengthening and subsequent lengthenings, which you're going to see later. The screw fixation is tightened on that rod to prevent proximal migration as the lengthening is started and the correction is obtained. There's a -- we want to have enough tension in that rod. We don't want to disrupt the early foundations. Obviously the fusion's not occurred acutely, and so care has to be taken not to overlengthen on that first try. We want to make sure that we're bone grafting extensively in these areas

to -- again, in just the area of the foundation, just where you've placed those screws, we want a solid, well-decorticated bone and bone graft to create a solid foundation so that the lengthening can proceed without failure of those fixation points. The closure is really critical, and maybe I'll ask Dr. Bodenstern to comment on the closure techniques for these multi-operative patients.

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LAWRENCE BODENSTEIN, MD PhD: Sure. As you see here, you'll see in the later video, closure's obviously crucial to get coverage over the device, and these devices, some of them are fairly bulky, especially for small children. If you have a 200-pound teenager, that's usually not a problem. If you have a skinny little 3-year-old who may have some nutritional problems, it can be more of a problem. They have very little subcutaneous tissue, they have very small musculature, and so we use several techniques. As I said, we'll illustrate those later. One of which is that we create a flap so the device lies underneath the flap and lives underneath the flap of tissue rather than where the incision is. The other thing we do is we go through a shuttered or a staggered incision through the various layers, so the incision through the skin subcutaneous tissue and the muscle are not laying in the same plane, therefore there is not a direct shot from the outside world down to the chest wall, so it's more difficult to get to the device. And finally, we use this tunneling method. If we don't need to access the chest between the top and the bottom, then you can create a tunnel, which obviously would leave much less incision which is at risk to breaking down. Sometimes, however, you have to actually do a thoracoplasty or work on the chest, in which case you have to make a full-length incision, but if possible, the tunneling method is a great advantage.

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MICHAEL G. VITALE, MD, MPH: It's been recently shown by Dr. David Skaggs and company in Children's Hospital of Los Angeles that 75% of the children who are candidates for this type of procedure are in the fifth percentile of weight, the lowest fifth percentile of weight for their age. As Dr. Bye pointed out, many of these kids have a very high energy cost of breathing. They spend so much of their effort just oxygenating their lungs. So it's really critical to pay attention towards soft tissue closure, as Dr. Bodenstern pointed out. This is the result of our patient, our case example before and after the initial implantation of a growing-rod system. In this case, we used the Stryker 4.5 millimeter vitalium growth system. Vitalium is a new alloy that has more strength for a given size than other constructs that potentially avoids rod bending and rod breakage, which has been reported in the past. Our initial x-rays go from 61 to 26 degrees, but as importantly, we'll continue to expand this rod over time, continue to expand the thorax, expand the space available for the lung, and allow lung growth during this critical time of development for this young child. Our next case will involve the VEPTR, or the vertical expandable titanium rib device. This is a 2.5-year-old with congenital diaphragmatic hernia treated successfully at this institution. Unfortunately, she developed -- very rapidly, in the course of just two years, a very significant curvature with two curves measuring nearly 100 degrees. This slide shows her curvature before surgery and then after implantation of the VEPTR device. The vertical expandable prosthesis titanium rib is made by Synthes spine and is a newly available option that allows us to expand the thorax, straighten the spine, and allow the lungs to grow. Here you see that the curves originally measured 88 and 91 degrees. After the initial implantation of the VEPTR, we have that down to 58 and 66 degrees, and then after our first lengthening recently, down to 51 and 55 degrees. Maybe more importantly, however, what you really see is her dramatic increase in the space available for the lung with the thoracic length going from 216 millimeters beforehand to 256 millimeters right after surgery and 266 millimeters after lengthening. This is a dramatic increase in the space available for the lung, which is much more than can be attributed to just normal growth in a six-month period of time. In this example, you see that we're nowhere near the spine where it's attached to ribs proximally and the pelvis distally. And again, Dr. Roye is going to detail the surgical

technique in dealing with large curves with a VEPTR in children with early onset scoliosis and thoracic insufficiency syndrome.

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DAVID P. ROYE, JR, MD: So I wanted to talk a little bit again about those principles of successful growing rods, dual rods in general to share forces, avoid the early thoracic fusion, and long extra-spinal constructs to allow for some motion to be maintained in the spine and to prevent that fusion from occurring. The -- the frequent lengthening is also an extremely important part of this concept. We need to lengthen every four months in young children, children less than 3, and then as the kids get over 3 or 4, we can probably cut back to every six months. The concept of waiting for the curve to progress prior to lengthening is probably not the right one. We should be looking to have regularly scheduled lengthenings to allow maximal growth of the spine.

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MICHAEL G. VITALE, MD, MPH: In fact, Dr. Robert Campbell, who is the innovator behind the VEPTR device showed clearly that pulmonary function improved the most in those children who were treated the earliest. So a key part of our thinking now is to treat these kids before significant spinal deformity and thoracic insufficiency develops. Now we're going to speak a little bit about the surgical technique of the implantation of a VEPTR using a video of a recent case.

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DAVID P. ROYE, JR, MD: So here we are with the child in the operating room. We've outlined the iliac crest, and we're going to localize that Dunn-McCarthy hook on the iliac crest medial to the point of maximum height of the ilium. That would allow it to be more stable and to tend to move medially as opposed to moving laterally. About a 4 centimeter extensile incision is made. That incision is enough to allow exposure of the iliac crest and successful development of pockets for the placement of the Dunn-McCarthy hooks. Proximally, we identify the rib that we want to connect to. That's done with careful x-ray control and planning. Of course, that's planned prior to surgery, but then in the operating room, we would use fluoroscopy. We're going to do a single midline incision. It's going to be located in the same area that perhaps we would need to have an incision for definitive fusion of the spine. We would tend to make that incision to coincide to these often severe curves to make sure that when the time came, it would be -- it would be appropriately placed. However, even though that vertical incision is being outlined over the spine, we're not going to look at the spine, we're not going to make any kind of approach to the spine because we don't want the spine to fuse or to be impeded even by soft-tissue scarring. So here the surgeon is making an incision over the ilium and preparing to place the Dunn-McCarthy hooks. We do a careful extraperiosteal dissection of the ilium. We want to maintain those growing cells to try to prevent migration of the Dunn-McCarthy hook into the pelvis, and that's probably best accomplished by maintaining blood supply in periosteum over the bone. An incision can be made in the growth cartilage of the ilium to allow the Dunn-McCarthy hook to settle onto the pelvis itself. And here you can see that we're developing those pockets that are needed on the medial and lateral aspect of the ilium to those pockets that will allow the Dunn-McCarthy hook to be placed. We want that hook to the largest portion -- deepest portion of the hook to be placed inside the pelvis and with this blunt dissection, we're continuing to develop that. But we want that deep part to be included in the pelvis. Do you want to make a comment on that, Dr. Vitale?

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MICHAEL G. VITALE, MD, MPH: Yeah, I think that's a really key point. It's a point that we've learned. The -- we have a couple options for placing that pelvic cradle, and by placing that largest portion of the cradle anterior to the pelvis in a so-called reversed position, it allows us to get the instrumentation more anteriorly, allowing better maintenance of lumbar lordosis. Here's an example of the cradle going in in just that position. This allows better outcomes in a sense of some of the issues that some of these kids, particularly very active,

have had with postoperative pain. So again, the reverse position allows us better lordosis on some of these children.

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DAVID P. ROYE, JR, MD: I think another important issue is you can see from that demonstration that that hook is not solid, that hook gives us a lot of movement or -- to prevent total immobilization. Here's the proximal rib cradle. The rib cradle is -- encircles the rib. It doesn't grab it or hold it. Again, allows motion; the rib can move back and forth through the device. We're demonstrating it here outside the child, and this is obviously -- can't be put in if it's closed already, but the two halves are held together by that closure clip and the rod is then attached to the rib cradle. This is a solid device and is, again, allows fixation but also allows motion. The approach to the proximal segment is paraspinal. The midline is to your left as you look at this picture, and we're going through the lateral spine musculature down to the rib. Once the rib is identified and we would check it on fluoroscopy to make sure that we were identifying the correct rib, the preplan-- the planned rib, I should say. Once the rib is exposed, we then will elevate around the rib using these rib elevators in an attempt to prevent any damage or opening in the pleura. We're not doing a thoracostomy in this patient. This is a bilateral rib to pelvis construct, so we don't want to enter the chest. We want to be careful about providing a safe passage for the superior part of the cradle which has just arrived on the scene. And then the inferior cradle. They are then joined and clipped to be locked together.

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MICHAEL G. VITALE, MD, MPH: And now we're demonstrating a multilayered approach towards the left-side rib construct. As Dr. Roye just mentioned we are generally speaking above the lungs, but occasionally we can get a small rift in the pleura. It's one of the complications associated with implantation of rib devices. Dr. Bodenstein, can you comment about optimum management for a small rift in the pleura in these children? How often do chest tubes need to be utilized, for example?

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LAWRENCE BODENSTEIN, MD PhD: It'd be very rare to have to use a chest tube in that situation. Assuming there's no parenchymal injury to the lung, we would just evacuate the air on closure of the chest wall and the child would probably have a tiny pneumothorax postoperatively that would get very quickly resolved. So it's -- the only time you would really need to use a chest tube is if there was a significant pulmonary parenchymal injury or if there was a large dissection in the chest where you expect there to be a significant amount of fluid evacuat-- being secreted postoperatively. But that's becoming -- the use of chest tube is really becoming more the exception than the rule now.

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MICHAEL G. VITALE, MD, MPH: Yeah, that's absolutely been our experience. In fact, we're generally not using any postoperative drainage at all with these minimally invasive techniques. The one situation where we may in fact use a chest tube is in the formal expansion thoracoplasties, where we're going in, separating fused ribs, coming right down to the chest wall. But generally speaking, we really don't use postoperative drains. Now the fixation is in above, the fixation is in below, and we're showing the tunneling.

00:42:58

DAVID P. ROYE, JR, MD: Right, so again, tunneling, as Dr. Bodenstein said, is a way of saving the kid's skin integrity. It's great, there's no incision at all, so that's even better than multilayered careful closure. That rod is passed using a similar technique that you saw with the growing rod system, using a chest tube, which is our principle use for the chest tube in these cases, as opposed to putting them in to the chest cavity. Here the domino has been hooked to the Dunn-McCarthy hook distally and lengthening has started against a C-clamp. You see that little blue C-clamp above? That's a removable implant that's used to allow distraction of the rod system against the domino. Now we have our three incisions, and Dr. Bodenstein, would you take over the description here as we attempt to close this case?

00:43:52

LAWRENCE BODENSTEIN, MD PhD: Right, well, here are the other methods that we use to prevent visualizing the rod postoperatively, which is if the incision here is in the midline but the rod is placed laterally. So the incision-- so the rod sits under the flap, not directly under the incision. And you can see that the incision through the skin is not in the same position as the incision through the muscle, so we have this staggered layered closure.

00:44:15

MICHAEL G. VITALE, MD, MPH: I'd like to just emphasize one of the points that Dr. -- hold the video there, please. I'd like to emphasize one of the points that Dr. Roye spoke about. Many times after insertion of the original VEPTR, the pelvic fixation clamp is not firmly seated on the pelvis. Rather than expanding the device by attaching that C-clamp and then distracting distally, you can lock on the iliac fixation point and then use your expansion within the system for future lengthenings. We'll usually remove the C-clamp after we've locked down the device. One of the principles of success here, as has been pointed out over and over, is frequent lengthenings. We want to not only keep up with the spine, thorax, and lung growth but actually push it along. Data emerging from our multi-center study in this area shows that in fact after insertion of the VEPTR device, growth on that side exceeds normal growth. We're actually driving growth in that thorax. In order to do that, we need to lengthen frequently, which means that a child needs to come back to the operating room for lengthening. We'll show video now of a lengthening, one such lengthening device. These are often performed as outpatient surgeries through a relatively minimal incision. Dr. Roye.

00:45:33

DAVID P. ROYE, JR, MD: So you can see that we're using the same incision, the old incision, approaching the device, and it's off to the side, away from the skin incision, which is just exactly as we desire. And in lengthening this growing rod system, we're going to expose the end-to-end connector. You can actually usually appreciate a little bursa that forms over it and makes the approach to the lengthener fairly straightforward. And this is a demonstration outside of the body showing that the lengthener has a way that a device can be inserted into it and tension placed and the device lengthened.

00:46:15

MICHAEL G. VITALE, MD, MPH: And here you see the Stryker Xiao 4.5 system, which again is a vitalium system that allows growth across this axial connector.

00:46:24

DAVID P. ROYE, JR, MD: We suggest that the -- the set screws should be replaced each time you loosen them, remove them and replace them to make sure that there's a good and solid closure and no displacement, no, I should say, wear and tear or asymmetries in the threads of the screws. The other side is also approached in a similar way, and one technique that we use is alternative lengthening. In other words, going from one side to the other, lengthening first the right, then the left, and going back as the tension develops. That avoids putting tensioning to failure, the foundations you have proximally and distally, and can be indeed a very useful technique in promoting maximum correction.

00:47:20

MICHAEL G. VITALE, MD, MPH: If you look closely, the two ends of the rod within the axial connector in the previous example were not touching each other even though this was our first lengthening. And that's because the easiest way to lengthen is to leave three or four millimeters to allow the distractor entry into that growing-rod system. Finally you see the closure, again very careful attention to a very tight, complete, meticulous closure, as has been discussed by Dr. Bodenstern and Dr. Roye. Coming back to our presentation, again you see this child's results to date. Large curve made smaller, further made smaller after a recent lengthening. Significant improvement in space available for the lung, significant improvement in space available for the thorax. Many of our children, after insertion of VEPTR or growing-rod constructs have a significant improvement in lung function. They report fewer respiratory illnesses. There are children, for example, with spinal muscular

atrophy that no longer require ventilatory assistance like a CPAP or even ventilation at night. And it can be very, very gratifying, improving quality of life in some of these affected children with early onset scoliosis. What we've hoped to show today is that early onset scoliosis is in fact a significant challenge. It's not yet a completely solved problem. But there are newly available options that promise much better outcomes to affected children. Above all, we seek to avoid early fusion. There's a wealth of data now really arguing against fusing young children. We need to keep our eye on the whole child. Children are not just small adults, and new instrumentation systems that are made specifically for kids allow us to treat kids differently, allow us to recognize the particular issues of lung growth and thoracic growth. I think that through the use of newly available techniques, we can really promise significantly better outcomes to children with early onset scoliosis. I want to thank my panel here today for participating in our live webcast here at the Morgan Stanley Children's Hospital in New York. I see questions are already coming in from the internet, and after a short close now, we'll take your questions. Thank you very much for joining us today. Okay, so we now have time for a few questions. first question comes in from Missouri for Dr. Bye: what is the ideal time to do this from a lung growth standpoint?

00:50:04

MICHAEL R. BYE, MD: Well, as we saw in one of the earlier slides, the lungs grow up until age 8, yet much of that occurs before age 2, so that if you can repair that chest wall before age 2, you should be able to optimize that child's lung function in the short run and certainly allow for better growth with the lengthenings into the future.

00:50:29

MICHAEL G. VITALE, MD, MPH: Another question: when to do VEPTR, when to do growing rod. Actually a very good question, something we speak about quite often. Dr. Roye, can you address that?

00:50:37

DAVID P. ROYE, JR, MD: Sure. It's a -- not an easy question. There's a -- I think that there are easy parts of it in a sense. The indications for the VEPTR are certainly where there is associated significant chest-wall deformity. So if one has a congenital scoliosis with a fused rib, perhaps multiple levels of congenital anomalies, a lung that's being affected by the lack of development of the ribcage, then the VEPTR is absolutely the right instrumentation. The whole concept of putting that instrumentation on the chest wall, expanding the chest wall while controlling the scoliosis is brilliant and it just works very well. The -- on the other extreme, if there is no chest wall involvement and it's primarily a scoliosis where the ribs are normal and there is apparently fairly normal development of the lung to date, then likely the growing rod would be the right way to go. It's mechanically very stable, it's predictable, we have a long track record. It's the cases in between that are more difficult to judge. We have created another criteria, which is age. Using the VEPTR, in other words rib to spine or rib to pelvis instrumentations in the very young affected children seems to make sense to us because it allows more motion than the growing rods do. So that would be another consideration when choosing the instrumentation. But there are many overlapping indications, and we haven't yet sorted all of those out.

00:52:32

MICHAEL G. VITALE, MD, MPH: A question from Chicago which we'll ask Dr. Bodenstern to address: when should the chest thoracoplasty be done? Dr. Bodenstern is our assistant in the formal chest thoracoplasties as part of the VEPTR. I think it's a great question.

00:52:47

LAWRENCE BODENSTEIN, MD PhD: Well, I think it sort of has some overlap with the question Dr. Roye was asked and answered. If the problem appears to be a primary chest wall problem, then a thoracoplasty may be helpful. I mean, a classic example of that is the asphyxiating thoracic dystrophies like Jeune syndrome, where the defect is really a primary constriction from the chest wall, and then we can deal with the chest wall directly with a thoracoplasty and a VEPTR construct. If the -- if the problem seems to be primarily a spine

problem, then we're less inclined to do a thoracoplasty, but for example, if there are multiple fused ribs and that appears to be a limiting entity in terms of getting appropriate expansion of the chest, then we would proceed with probably a limited thoracoplasty in that case as well.

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DAVID P. ROYE, JR, MD: I would mention also that there's a -- we haven't I don't think directly alluded to this tonight, but we can also with the VEPTR actually substitute for ribs. So in kids with big missing rib segments and paradoxical movements of the chest wall, that's another reason for doing a thoracoplasty and moving ribs to substitute for those areas of loss.

00:54:05

MICHAEL G. VITALE, MD, MPH: Another question really for the entire panel, from Massachusetts: has this improved the prognosis for any specific set of disorders?

00:54:14

MICHAEL R. BYE, MD: Well, I think there are a couple of specific disorders that have sort of been mentioned tonight. One is the Jeune syndrome, also known as asphyxiating thoracic dystrophy. And from its name, you can guess that it doesn't have a very good prognosis. And certainly, years earlier in my career, most such babies had a dismal prognosis and often never got out of infancy. I think if these procedures can offer them a better short-term and then eventually a better long-term life, then that's terrific for these children and their families. Similarly, the Jarcho Levin syndrome not only is crooked but affects the airways as well, and if we can help those babies straighten out, then they can lead much better, much more normal lives. And they don't have to spend the majority of their life in a hospital or in an ICU or on a ventilator.

00:55:05

DAVID P. ROYE, JR, MD: Yeah, I would have two answers to that question. You know, the first answer is a kind of disclaimer, and that is that we don't yet have 50-year follow-up on these patients, right? So if we're talking about prognosis, what we're really talking about is what have we done to improve the quality of life of that patient going forward into adult years? We've already seen from the Knockensum data that we're looking at increasing death rates in these untreated natural history patients in infantile cases at age 20. And in the juvenile cases at age 40. So the 12 years or to 15 years of follow-up we have on the VEPTR is not enough time to let us know really what we've done to change the quality of life. But I can tell you that as a -- someone who's been on the scene, doing spine surgery in children since 1980, that the way that we're treating the chest wall particularly, but also growing the spine in these early cases, is a breath of fresh air to me. It's the first real innovation as far as I'm concerned in deformity surgery in my career.

00:56:22

MICHAEL G. VITALE, MD, MPH: Great. A question from Finland, actually: it's 2 a.m., and I myself am a patient with scoliosis correction about five years ago. I became interested in non-fusion methods because my whole spine is not yet fused. I still have growth left. How long is the operation? What are the most common complications? Is there major blood loss associated with it? From Finland. Dr. Roye?

00:56:50

DAVID P. ROYE, JR, MD: I would love to know how old that person is. There's a -- the operations for the application of a growing rod is -- I would describe it as being less complicated technically actually than an actual fusion or a typical fusion because the dissection is smaller, the blood loss is less, we almost never have transfusions. We -- the complication if you will, the real morbidity comes into the multiple procedures that are needed. The rod implantation initially would take three or four hours and the child would be in hospital for three or four days. Subsequent lengthening operations are done as outpatients, but there are many of them. And we do -- we have seen that particularly in the very young child, these multiple procedures have an effect on the child's psyche. It's hard to

put up with that much intervention. The complications are related to the issues we've already spoken to: closure and skin integrity, infection due to the multiple procedures that we're doing, and the migration of the implants because we're not fusing is common. So unplanned returns to the operating room are fairly frequent, as are the need for treatment of wound problems. I would state, however, that those problems almost never actually impair the outcome. The outcomes continue to be good. it's just troublesome for the patient to go through.

00:58:32

MICHAEL G. VITALE, MD, MPH: Another question from Springfield, Massachusetts, which is certainly a timely question: what is the radiation dose and cancer risk for these CT scans? In fact, researchers at this institution have recently and not so recently pointed out that CT scans carry with it a small increase in risk in lifetime cancer. And as with anything else that we do, we need to weigh risks of any intervention or diagnostic treatment like a CT scan with potential benefits. We do not get necessarily frequent or routine CT scans in all of these kids, but in some kids it's really very helpful in guiding our surgical approach. The other thing to point out is with the use of high-speed helical CT scans that are at this institution and many other leading children's hospitals, the radiation dose can really be controlled to some extent. But I think it's a very good point. For Dr. Roye: can a VEPTR be used in a 2-year-old with an 84-degree curve in the lumbar region without TIS? Can this be done without using the VEPTR chest-wall device? Eighty-four degree curve in a 2-year-old in the lumbar region.

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DAVID P. ROYE, JR, MD: There's -- we do use the VEPTR to treat primary lumbar curves. And the -- what would be done in that situation, likely, would be an attachment to the pelvis and then an attachment to a rib above. We still would go to the rib, it wouldn't be a thoracoplasty. There would be no invasion of the chest wall itself except at the site where the rib cradle was placed. The -- we haven't done the -- a pelvis-to-spine construct because to us it seems to -- if that was going to be the case, we would probably go to a growing rod as opposed to a VEPTR.

01:00:38

MICHAEL G. VITALE, MD, MPH: But I think that's absolutely right. In some children, for example children with spinal muscular atrophy at times or with children with myelodysplasia, their chest -- their scoliosis is such that their entire spine is not necessarily involved. And having the full gamut of options from growing rod to limited fusion to VEPTR allows us to cater the specific implant approach, the specific treatment -- again, implant choices that were not available even five or ten years ago -- to the needs of the patient. I think that the menu of treatment options that we have available certainly allows us some choices and allows us to cater the specific types of treatment to the specific child. In summary, I want to say that in 2007, the options available are much greater than ever before, that we have the ability to promise significantly different and hopefully better results to children with early onset scoliosis. I want to again thank very much the panel for joining me here today, and I want to thank all of you for being with us as part of this live webcast from the Morgan Stanley Children's Hospital in New York. Thanks very much.

01:01:58

ANNOUNCER: This has been an expert discussion of VEPTR and growing rods from Morgan Stanley Children's Hospital of New York Presbyterian. OR-Live makes it easy for you to learn more. Just click on the "request information" button on your webcast screen and open the door to informed medical care.

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