

# Scoliosis in Congenital Muscle Disorders

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## How to view the Webinar on Scoliosis:

Click the following link to view the webinar again on You Tube, or copy and paste into your browser search:

[http://www.youtube.com/watch?feature=player\\_embedded&v=ZBK8lo0Fz7M](http://www.youtube.com/watch?feature=player_embedded&v=ZBK8lo0Fz7M)

## Questions & Answers

### Relating To Pre-Operative Care and Evaluation:

*What is the optimal time for someone with a congenital myopathy/dystrophy to have scoliosis surgery? I have read varying articles some state it is better to have surgery sooner – ie around 20 - 30 degrees, and other state between 30 and 60 degrees?*

To some extent it depends on the specific diagnosis, but in general if the curve has progressed beyond 30 degrees in the young child surgery should be considered as an option. Variables would include flexibility, rib hump, trunk balance and the overall function of the child. The point is that given today's growing techniques curves should not be allowed to progress. In the near skeletally mature child one should not wait until the curves cannot be nearly fully corrected.

*How can families prepare and be pro-active in optimizing their child's health prior to surgery?*

There are several areas to emphasize. First is nutrition. Increasing calorie intake in any way that is feasible should be attempted. Even installing a PEG or similar device may be indicated after consultation with the team. Second is to maximize pulmonary function with a period of pulmonary "rehabilitation" prior to the surgery. Preparing the child and the family for the stress of the surgery is also key to a good outcome.

*What are some things that should be avoided in a child with mild to moderate scoliosis to prevent the curve from worsening in terms of activities, behavioral things (like slumping), therapies that are contraindicated, etc? I am interested in Dr. Roye's suggestions for PT and other beneficial activities for a child with mild to moderate scoliosis with nemaline myopathy that is walking. Aquatics, hippotherapy, etc?*

I can't think of activities that should be avoided in kids with CMD and mild to moderate scoliosis nor can I think of contraindicated therapies. Maintaining truncal alignment is important and within the capacity of the child sitting and standing posture should be maintained along with flexibility. Hands on manipulative therapies to maintain flexibility, aquatics to maintain trunk strength and flexibility and hippo therapy to work on core strength are all good. However, some kids are not going to be strong enough to maintain that posture particularly as the curve and weakness progresses – at that point the child may need bracing and/or surgery.

***How often should a child with neuromuscular disease who has mild scoliosis get back xray and see the orthopedic surgeon?***

Most of the consensus groups have hit upon every six months with x-rays as the right frequency. This is particularly true for the child who is between the ages of 7-8 and 14 when the late childhood and early adolescent growth spurt will make progression of the curve more likely. Basically, once scoliosis has been discovered it should be followed twice a year. Obviously, if progression has been documented there may be a need for more frequent follow up with the orthopaedic surgeon.

***What are current indications for bracing?***

Bracing is probably never definitive treatment of CMD scoliosis. With that in mind, we will start bracing in children with curves over 30 degrees or so if they tolerate the brace. We also will use the brace as a functional aid – helping with sitting posture and alignment and enhancing upper extremity function by limiting leaning. We want to delay surgery if we can as we know the older the child is when the first surgery is performed the better in terms of complications. Very early aggressive bracing can create chest wall deformity, causing modeling of ribs and exacerbating funnel chest, another reason to think about growing rods in that group of kids.

***Is casting more efficacious than bracing in slowing progression of scoliosis?***

We use casting in young children but generally not in kids with CMD. Children with CMD do not respond any better to casting than bracing and there are issues with the casting. It requires general anesthesia and it is difficult to ascertain the effect the cast will have on respiratory effort when the child is asleep. We have used casting on some kids with neuromuscular disease who have no respiratory impairment and more or less normal tone.

***My 11 year old daughter has Collagen 6 Myopathy and has developed a 50 degree + curvature in her spine. She is scheduled to undergo a Shilla procedure in the next few months. We are doing various things to prepare her for this surgery. One of the items is attempting to stretch her hip flexors where she has considerable contractions. I'm concerned about what happens if we are not able to get those flexors completely stretched.***

***How will this affect her mobility and what other options other than a stretching regime do we have? Is casting available or advisable for hips?***

Stretching her hips is important. Almost all of our procedures reduce somewhat the lumbar lordosis thus bringing the pelvis forward. This will accentuate any existing hip flexion contractures. My usual approach is to treat preoperatively with stretching

and see how the surgery affects the contracture. In most cases we are able to restore satisfactory sitting and positioning with a conservative approach and more therapy after the surgery. If there is too much hip flexion for comfortable positioning after she has recovered from the spine surgery and has had therapy, hip flexor release surgery needs to be entertained. I have not found casting of hips in older kids effective. It is poorly tolerated and really disabling.

### **Questions & Answers** **Relating To Surgery:**

*I have been reading about Universal Clamping which I understand has been given the green light in USA for patients with neurological scoliosis. The papers on Universal Clamping are very positive in terms of better correction, a faster procedure with less blood loss, fewer fixing points, fixing load spread (good for patients with osteoporosis), and gives a more natural resulting posture. Why would a surgeon choose the more traditional method of rods/screws/sublaminar wiring over this new technique, other than the fact it is new and they may not be so familiar with it yet?*

We use the Universal Clamp as another tool in correction of neurogenic (and some idiopathic cases as well) scoliosis along with screws and hooks. (The clamp requires the use of rods.) It is not a panacea and we do not use clamps in isolation. In our hands it does not improve operative time or blood loss (it takes me approximately 2-3 minutes per level for a screw and about 4-5 for the sublaminar clamp). Read reports of new instrumentation critically – someone is trying to sell it and will tend to exaggerate the good parts and deemphasize the negative parts.

*I have read that fixing is recommended to T4 (shoulder blades height) – in patients with DMD. But I know patients who have been fixed from the base to the top of their spines. Why is this, and what is the minimal amount of ‘fixing’ that a patient can have to correct their scoliosis?*

In patients with DMD a long fusion is indicated. As the child weakens, they will progress curves above or below the unfused area – particularly the lumbar area. If not otherwise contraindicated I will usually recommend fusing from the pelvis to the upper thoracic spine – T4 to T2 depending on the details of the curve and the spine anatomy.

*In congenital muscular dystrophy, some children develop extension neck contractures, what is the indication to perform fusion from neck (cervical region) through to pelvis?*

Those indications are rare. Because the Cervical spine becomes stiff as the paravertebral muscles become fibrotic fusion is rarely indicated.

***In congenital muscle disease where paraspinal muscles are involved and scarred, is there an increased risk of bleeding during surgery?***

In general children with neuromuscular disease have an increased risk of bleeding and transfusion particularly during fusion operations where the exposure is more extensive and more bone is exposed. In CMD the tiny muscles in the blood vessel walls are affected by the disease and are less capable of clamping down which may be part of the problem. In addition these children are usually osteoporotic and osteoporotic bone bleeds more.

***I have a thirteen year old boy with merosin-deficient cmd. He's 145cm tall and weighs 30 kgs. He is wheelchair bound and ventilator dependent during the night. His vital capacity is about 23%. He has a brace from 2006 and he's quite comfortable with it. He has a significant scoliosis (65%, "S" curve) and his lordosis is affecting his breathing capacity. One surgeon wants to do the arthrodesis in January. The other one wants to put growth rods and wait until the end of puberty.***

***Is there a role for growing rods in a 13 year old? How does the low vital capacity play into the decision about the proper approach?***

I think there is a place for growing rods in 13 year old kids. Skeletal maturity and potential growth (particularly of the anterior spine) is more important than calendar age. Many patients with MD CMD are slow growing and slow maturing. It is hard to say without seeing him and the x-rays, but I would likely seriously consider growing rods. In addition, the growing system is a little less surgery than the definitive surgery and may turn out to be all that he needs. I would hope that either approach may help his vital capacity somewhat, by preventing "collapse" and the encroachment of abdominal contents on the diaphragm.

***What is the difference between VEPTR surgery and spinal fixing – and what considerations will the surgeon make before deciding which procedure to use on a patient?***

VEPTR and growing rod surgery are performed to correct the spine deformity without fusing the spine and stopping growth. So in kids with a lot of growth left, these techniques make sense as they allow continued chest wall and lung development. The child who is at our near skeletal maturity does not need that extra burden of care (lengthening of the rods periodically) so an operation to correct the scoliosis and fuse the spine makes sense. There are definitely grey areas where growing systems may be indicated and fusion may be indicated; the factors that are considered include the amount of growth, general health and nutrition, the prognosis of the specific illness, family support systems etc.

***How long does a typical VEPTR or growing rod surgery (non-fused) surgery take?  
How long does a typical scoliosis fusion surgery take?***

Truly depends on the complexity and the number of implants used, but a typical 2<sup>nd</sup> 3<sup>rd</sup> rib to pelvis bilateral implant takes about four hours. The typical definitive surgery procedure is 6-8 hours. Please, don't make a decision based on the time of surgery. In any individual case, the growing system could be faster or slower and the same for the definitive surgery. Typically there is less surgical exposure needed for the

growing rod cases and there is therefore less time needed and less blood loss. The growing systems use fewer implants which also reduces the amount of time needed.

***How painful are the rib fractures that can happen with VEPTR?***

Most VEPTR rib fractures are surprisingly not painful at all. The diagnosis is made by the parent who notes a bump or by the surgeon who sees it on routine x-ray. Occasionally when there is a sudden failure there will be pain, but that is not common.

***If a VEPTR is placed into a 4 year old, how often does the entire VEPTR need to be removed to accommodate for growth or does the original VEPTR last the child til they are greater than 10 years old and can undergo spinal fusion?***

That is a hard question as it really depends on the size of the child from the get go and the rate of growth. If the four year old is small, the size of the implant must be small and therefore will not last as long. Typically, between age 4 and 12 or 13 there would be two “major” changes of the implant. In addition, we do not yet understand what needs to be done for “VEPTR Graduates”. It appears that some may not need definitive fusion. I would also point out that age is not the criteria for definitive fusion – it is skeletal maturity and many of our CMD patients are quite immature at age 10 in which case we would continue lengthening surgery.

***Dr. Roye, you expressed that you feel strongly that children under the age of 10 years should not undergo spinal fusion surgery. Are VEPTRS available globally? What are the downsides of fusing early under age 10 years?***

Growing systems are available globally – you don’t need the VEPTR you need the knowledge and expertise. Before the VEPTR was available in New York, I performed chest wall growing rod surgery with ordinary implants (hooks, rods, connectors) used in spine surgery. These constructs worked very well. I still use them in China where the VEPTR was recently approved, but is too expensive for all but the rich. Fusing immature spines reduces spine growth therefore limiting trunk height, limiting room for the lungs and limiting lung growth. There is a strong literature supporting that early fusion results in bad pulmonary outcomes and worse quality of life.

***Would fixing the spine to the pelvis (to avoid pelvic obliquity) prohibit a patient from ever walking again (with support/equipment)? Is there any half-way measure where pelvis can be fixed, but still allows for a walking motion?***

Most children who are walking before fusion to the pelvis can continue to walk with equipment after the fusion. I no longer list that as a contraindication in my practice, as I see waiting, or fusing short as offering more risk to the patient than the risk of losing some ambulation capacity. I’ve never had a child go off their feet after fusion or instrumentation to the pelvis, but I have had children lose up right function in walking standing and or sitting as they lose balance from progressive curves and pelvic obliquity.

***If pelvic fixation (screws into the pelvic bone) was not done during the original spine fusion surgery, what are the factors that lead to worsening shift of the pelvis, requiring future additional surgery to extend fixation to pelvis?***

Our default setting in neuromuscular scoliosis is fusion to the pelvis in all sitters and in most of the ambulators as the risk of progressive pelvic obliquity is so high. In addition treating it after the original fusion is really hard. The reasons for the progression vary with the diagnosis but basically are that the same factors that caused the scoliosis in the first place are still there after the surgery – low tone or high tone – they will continue to attempt to deform the remaining unfused spine. The lower extremities are rarely symmetrical and exert asymmetrical forces on the pelvis further worsening the prognosis

***Should pelvic fixation (putting screws down into pelvic bone) be done during scoliosis surgery in a teenage (>10 year old) who is still walking?***

This is a controversial topic, and the decision is very much based on the individual, the diagnosis and a long discussion with caretakers and the patient. However, our assessment in general is that the downside risk of progression of pelvic obliquity outweighs the risk of some loss of ambulatory function. The revision surgery for those patients fused short who develop pelvic obliquity is never easy and carries significant risks as well. Progressive pelvic obliquity also threatens ambulation significantly.

### **Questions & Answers** **Relating To Post-Surgery Care:**

***Generally, how long is the recovery period after surgery in patients with congenital myopathy or dystrophy?***

Most kids are in the hospital for 5-7 days. Most are off of narcotic pain relievers by two weeks. Sitting endurance of 3-4 hours is generally reached by 4-6 weeks. It is exceedingly individual and these times are generalizations. Most kids have just about returned to base line ADL by 12 weeks after definitive fusion or original growing system implant. Recovery from lengthening surgeries is about one week.

***We currently manually lift our son during transfers and he is able to slightly weight bear with our support, for seconds. Will we be able to do this ever again after surgery, or will he always have to be hoisted during transfers?***

He will be able to do weight bearing transfers more easily once recovered from his surgery. For the first six to eight weeks he may need to be lifted – once the operative pain is gone he will at least resume his preoperative transfer ability.

***How soon is it recommended/possible to go swimming after surgery?***

Most kids can be back in the water at about six weeks.

***How soon is it recommended that a child uses their standing frame again after surgery?***

As soon as comfort (pain) allows.

*Is it possible to use powerchairs with adjustable back rests after surgery?*

Absolutely it is. One needs to make sure the chair fits the new “straighter” child.

*What outcome should families and kids with CMD or Myopathy expect from a scoliosis surgery?*

That’s a hard question. It really depends on the type of surgery and the child’s diagnosis. In general with modern surgical science, pediatric intensive care units and the team approach to planning and surgery CMD patients can be successfully treated with an appropriate implant either growing or definitive. What you can expect is that the outcome is much preferable to NOT treating and allowing for progression of the curve and progressive loss of function both in ADL and in respiratory function.

*Is there a benefit to wearing brace after scoliosis surgery?*

For the most part we do not use braces after scoliosis surgery unless we are concerned that our fixation points are poor and we are worried about failure. In reality, I don’t remember the last kid I sent home in a brace after scoliosis surgery either growing or definitive.

**Glossary of Terms:**

**ADL** – Activities of daily living

**Adolescence** – early adulthood / late childhood

**Arthrodesis** – The fusion of bones across a joint space, thereby limiting or eliminating movement. It may occur spontaneously or as a result of a surgical procedure, such as fusion of the spine

**Autogenous Bone** – Bone originating from the same individual; i.e. an individual's own bone

**Autograft Bone** – Bone transplanted from one part to another part of the body in the same individual

**Ambulators** – people who can walk

**Bilateral Implant** – one implant in each rib

**Bone** – The hard tissue that provides structural support to the body. It is primarily composed of hydroxyapatite crystals and collagen. Individual bones may be classed as long, short, or flat

**Bone Graft** – Bone which is harvested from one location in an individual and placed in another individual (allograft bone) or in a different location in the same individual (autogenous bone)

**Bone Marrow** – The tissue contained within the internal cavities of the bones. A major function of this tissue is to produce red blood cells

**Bone Plate** – Usually a relatively thin metal device which is affixed to bone via screws. Bone plates are used to immobilize bones or bone fragments such that healing can occur

**Bone Screw** – A threaded metal device which is inserted into bone. The functions of bone screws are to immobilize bones or bone fragments or to affix other medical devices, such as metal bone plates, to bones

**Brace** – A back support limiting the motion of the spine

**Casting** – A series of plaster casts to try to correct the spine deformity without surgery

**Chest Wall Deformity** - When the chest wall or underlying rib cage appears deformed

**Cobb Angle Measurement** – Calculated by selecting the upper and lower end vertebrae in a curve. Erecting perpendiculars to their transverse axes. At their point of intersection, the angle is measured to indicate the curve's angle

**Contractures** - A condition of shortening and hardening of muscles, tendons, or other tissue, often leading to deformity and rigidity of joints.

**Congenital** – Present at and existing from the time of birth

**Core Strength** – Strong muscle control in the back and stomach, see Trunk Balance

**CNM** – Centronuclear Myopathy, a neuromuscular condition

**CMD** – Congenital Muscular Dystrophy, a neuromuscular condition

**DMD** – Duchenne Muscular Dystrophy, a neuromuscular condition

**Fracture** – A disruption of the normal continuity of bone

**Funnel Chest** - Pectus excavatum (a Latin term meaning hollowed chest) is the most common congenital deformity in children with neuromuscular weakness due to poor pulmonary function

**Fusion** – Union or healing of bone

**Growing Rods** – This operation can allow for continued controlled growth of the spine. This is done through the back of the spine. In general, the curve is spanned by one or two rods under the skin to avoid damaging the growth tissues of the spine. The rods are then attached to the spine above and below the curve with hooks or screws. The curve can usually be corrected by fifty percent at the time of the first operation. The child then returns every six months to have the rods "lengthened" to keep up with the child's growth. This is usually an outpatient procedure performed through a small incision. When the child becomes older and the spine has grown, the doctor will remove the instrumentation and perform a formal spinal fusion operation. In the past, this procedure had a very high complication rate, most of which were related to the instrumentation (hook dislodgement, rod breakage). Modern techniques reduced this complication rate to a reasonable degree.

**Hip Flexors** - The hip flexors are several muscles that bring the legs and trunk together in a flexion movement

**Hippotherapy** – Treatment with the help of a horse, to emulate walking motion while seated.

**Hook** – For spinal applications, a metallic medical device used to connect spinal structures to a rod

**Hydrotherapy** – Use of warm water pool for pain relief and muscle therapy

**Kyphosis** – An abnormal increase in the normal kyphotic curvature of the thoracic spine, ie a humped shoulders

**Joint** – The junction or articulation of two or more bones that permits varying degrees of motion between the bones

**Lordosis** – An abnormal increase in the normal lordotic curvature of the lumbar spine

**Lumbar** – The lower part of the spine between the thoracic region and the sacrum. The lumbar spine consists of five vertebrae

**MTM** - Myotubular Myopathy, a neuromuscular condition

**Nutrition** – nourishment, food

**Orthopaedic** – Medicine or surgery designed to help correct or ameliorate the discomfort of disorders of the spine and joints

**Osteoporosis** – A disorder in which bone is abnormally brittle, less dense, and is the result of a number of different diseases and abnormalities. (Sometimes referred to as osteoporotic bone bleeds or fractures)

**Paraspinal Muscles** – The muscles which are next to the spine

**Pedicle Screw** – Screws used in spine fusion to fix bone to the support device

**Pelvic Obliquity** – Deviation of the pelvis from the horizontal in the frontal plane. Fixed pelvic obliquities can be attributed to contractures either above or below the pelvis

**Pulmonary** – breathing

**Rehabilitation** – to restore good health or useful life

**Rib Hump** - A rib prominence and/or a prominent shoulder blade, caused by rotation of the ribcage - best exhibited on forward bending.

**Rod** – In spinal applications, a slender, metal implant which is used to immobilize and align the spine.

**Scoliosis** – Lateral curvature of the spine, when a person's spine is curved may look more like an "S" or a "C" than a straight line in xray.

**Shilla Procedure or Technique** – This a technique used that allows for the natural growth of the spine and correction of the spine at the same time. The Shilla is similar to a track and trolley system. The rods are placed but are partially fixated to the spine. This allows the system to “grow with the spine”. Rod breakage can occur and this is a normal outcome. When this happens revision surgery is indicated. This is a newer technique being applied at leading centers across the country in USA. Pedicle Screws are inserted at specific points in the spine. Screws in the center are holding the rods in place. Other screws allow for the spine to move and elongate at either end.

**Spinal Fusion** – A surgical procedure to permanently join bone by interconnecting two or more vertebrae in order to prevent motion (see Arthrodesis).

**Spine** – The flexible bone column extending from the base of the skull to the tailbone. It is made up of 33 bones, known as vertebrae. The first 24 vertebrae are separated by discs known as intervertebral discs, and bound together by ligaments and muscles. Five vertebrae are fused together to form the sacrum and 4 vertebrae are fused together to form the coccyx. The spine is also referred to as the vertebral column, spinal column, or backbone.

**T2 / T4** - By convention, the human thoracic vertebrae are numbered, with the first one (T1) located closest to the skull and higher numbered vertebrae (T2-T12) proceeding away from the skull and down the spine.

**Titanium** – A metallic element used to make surgical implants

**Trunk Balance** – A person's ability to sit unsupported and without falling or leaning over

**Universal Clamp** – a medical device implant to fix the spine. “The *Universal Clamp* Implant offers sublaminar fixation with immediate stability resulting in reduced stress on the bone/implant interface with gradual 3D correction” see <http://emea.zimmer spine.eu/>

**Vertebra** – One of the 33 bones of the spinal column. A cervical, thoracic, or lumbar vertebra has a cylindrically-shaped body anteriorly and a neural arch posteriorly (composed primarily of the laminae and pedicles as well as the other structures in the posterior aspect of the vertebra) that protects the spinal cord. The plural of vertebra is vertebrae

**VEPTR** - Vertical Expandable Prosthetic Titanium Rib

**Vital Capacity or VC** - The greatest volume of air that can be expelled from the lungs after taking the deepest possible breath.



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Please don't hesitate to contact [foyesarah@gmail.com](mailto:foyesarah@gmail.com) if you have any further specific questions for Dr Roye.

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