Scoliosis, Bracing, and Bone Health in PWS presented by Dr. Harold Van Bosse, MD (2026)

Dr. Harold Van Bosse | 2026 FPWR Conference

Edited presentation transcript

This transcript covers Dr. Harold Van Bosse's presentation on movement, foot alignment, bone health, hip development, and spinal deformities in Prader-Willi syndrome (PWS), followed by audience questions. It has been edited for clarity and readability, with filler words, repeated phrases, and nonessential interruptions removed. Clinical approaches and research findings are presented as discussed at the conference. References to photographs, diagrams, and X-rays refer to the presentation slides.

This presentation is educational and does not replace individualized guidance from your healthcare team.

Introduction

MODERATOR:

Welcome to Musculoskeletal Issues in PWS with Dr. Harold Van Bosse. My name is JT, and I'm the president and chair of FPWR Canada. I have identical twin boys who are both in braces. They're upstairs taking a break from the conference.

In this session, Dr. Van Bosse will discuss common orthopedic challenges, including flat feet, bone fragility, hip dysplasia, and scoliosis.

Dr. Harold Van Bosse is a pediatric orthopedic surgeon in the United States with a longstanding relationship with the Prader-Willi syndrome community. Through a patient 25 years ago, he joined the community of specialists and families caring for children with PWS. Orthopedic challenges include delayed milestones, flexible flat feet, hip dysplasia, and spinal deformities.

Please welcome Dr. Van Bosse.

DR. VAN BOSSE:

Thank you very much. It's good to see everybody. Before I start, I noticed yesterday while going through my talk that I misspelled Prader in the fancy ribbon in the bottom-left corner. Since I've moved back to the East Coast, we'll say we swallow our Rs!

We're going to talk about several things, but first let's set the groundwork.

Why PWS needs its own orthopedic approach

What do you know about Prader-Willi syndrome? It was first described by John Langdon Down, the same person who described Down syndrome. He saw an adolescent girl about 150 years ago. It wasn't until about 70 years later that Prader, Labhart, and Willi described a series of patients in the literature. Like a middle child, Labhart got dropped, and it became Prader-Willi rather than Prader-Labhart-Willi syndrome.

Another 25 years later, researchers realized it involved chromosome 15 and identified a microdeletion. The articles became interesting because they discussed children with chromosome 15 microdeletions and children with apparently normal chromosome 15s. Back then, they could only look at the whole chromosome. Much later, they recognized other ways PWS could occur.

PWS has taught people a lot about genetics. An area is missing from the father's chromosome. If that same area is missing from the mother's chromosome, you get a different condition, Angelman syndrome. The area may be deleted, as researchers saw in the 1980s. In some of those with supposedly normal chromosomes, there were actually two copies of the mother's chromosome 15 rather than one from each parent.

Then there is a third type, a methylation disorder, in which the chromosomes look normal and there is one from each parent, but the father's chromosome does not turn on where it should. This is the only kind that can run in families to a significant degree.

It is rare, occurring in about one in 10,000 to one in 30,000 births. Compare that with conditions my orthopedic colleagues see: cerebral palsy is about three in 1,000 and spina bifida about seven in 10,000. Those are uncommon, but not on the same scale as PWS.

Unfortunately, rarity can lead to generalized treatment: "I don't know PWS, but it looks a little like Down syndrome, so let's treat it the same," or, "I have children with cerebral palsy or delayed walking, so we'll do what we do for them."

We need to recognize that children with PWS follow different rules. Think of a child with Prader-Willi syndrome who has scoliosis, rather than a child with scoliosis who also has this Prader-Willi thing. We need the conditions in the right perspective.

The five things we'll discuss are all closely related to hypotonia.

Hypotonia and developmental milestones

Hypotonia is one of the first signs of PWS: reduced movement in utero, less kicking, and often a need for cesarean delivery. In newborns, we see poor sucking and a weak cry. As children grow, we see poor head and postural control and delayed milestones.

In general, we think of milestones as taking about twice as long. If a typically developing child sits at six months, a child with PWS may sit at 12 months. If a typical child walks at 12 months, a child with PWS may walk at two years or a little later.

The first thing we can do is therapy, therapy, therapy from the start: physical and occupational therapists working with the child, and speech therapists helping with feeding issues.

If a child isn't walking by about 16 or 18 months, I start thinking about braces. Weak ankles and knees make standing difficult. If we build a foundation from the ground up, children can be more stable and get up and around.

Some people worry that braces label the child, or say we need tough love to get them moving. My clinic's mantra is: get them walking any way possible, then work on points for style.

These are examples of an ankle-foot orthosis, an AFO. The ankle is solid, which matters because children with PWS tend to collapse through their ankles.

A therapist accustomed to working with children with cerebral palsy may suggest a hinged brace. But those children may be trying to get up on their toes, whereas our children collapse through their feet. We want more stabilization: a brace without hinges.

Walking and balance in adults

I give this talk often, usually focusing on children. I was asked to include adults because some families here have much older loved ones.

Let's discuss the gait cycle, which you practiced walking into this room. The double-stance phase is when you put one foot down and are about to pick the other up. An important part is push-off, the power phase of walking. Runners know how pushing off the toes propels you forward.

People with PWS tend to have walking speeds and patterns similar to those of an obese adult: shorter steps, reduced speed between steps, and more time with both feet on the ground. They also have reduced knee and ankle motion, keeping them relatively stiff.

Push-off is poor, and sometimes the foot collapses so much that there is hardly any push-off. This relates to reduced calf strength. The plantar flexor muscles are the ones that get you up onto your toes. Ultrasound studies comparing adults with and without PWS show a smaller gastrocnemius muscle in those with PWS.

Flat feet also make gait less efficient. Several things work together to reduce walking efficiency.

What can we do? Exercises to strengthen the ankles, including spending time walking on the heels and toes to strengthen the muscles involved, and work on balance.

Balance is complex. We use the inner ear, vision, and proprioception: knowing where body parts are in space. Even with your eyes closed, you can touch your nose.

People with PWS typically rely heavily on their vestibular system, the inner ear. The idea is to integrate the other systems so balance is more efficient and walking feels more comfortable.

Flat feet and orthotic support

Let's look more closely at pes planus, or flat feet. From behind, the heel rolls to the outside of the ankle. The outer border of the foot curves, and the arch comes down toward the ground.

The issue isn't just the absence of an arch. It's that the heel and the front of the foot are both off to the side.

Flat feet occur in roughly 40% of people with PWS, from a combination of loose ligaments and low muscle tone. The ankle muscles cannot control the heel as well, resulting in poor push-off. That makes walking and running less efficient.

Children just beginning to walk may be more cautious. You may see a high-guard position, with hands up in case they fall.

Besides therapy, we can consider bracing when flat feet are significant, to improve alignment and efficiency. There are two devices to discuss: the supramalleolar orthosis, or SMO, and the UCBL orthosis.

I'm a much bigger fan of the UCBL than the SMO, and here's why. The SMO comes above the ankle to try to prevent the heel from rolling. But the extensions above the ankle would have to be very tight to prevent that rolling, and then they would be uncomfortable.

The bigger issue is the front of the foot. If the heel and forefoot are off to the side, you need a device that pushes the forefoot over to control it. An SMO tends not to be full foot length.

We want a device that rolls the heel underneath the ankle and corrects the outer border of the foot, molded to push the foot over so the whole foot lines up better.

In adults, hypotonia combined with significant flat feet can greatly reduce push-off. Try walking on the inner border of your own foot and notice how difficult it is. Your balance is off, and you tend to widen your stance.

Bracing may be an option. If the flat foot is very severe, surgery to correct foot position may be considered. That doesn't happen often, but it may be appropriate for some people.

Bone mineral density and fracture awareness

Our third topic is bone mineral density, or fragile bones. Bone contains a latticework of proteins filled with calcium and other minerals that make it thicker and stronger.

You've probably heard the terms osteopenia and osteoporosis. Looking at a bell curve of bone density, the distinction involves how far the measurement falls below the average, with osteoporosis beyond two and a half standard deviations.

It's been difficult to establish the prevalence. In an earlier family survey, only 9% reported thinking their loved one had osteoporosis. But surveys of fracture history found figures between 30% and almost 45%, raising greater concern about fragile bones.

One study noted that people with PWS have higher pain thresholds. A fracture could be present for several days before medical attention was sought. Some people were walking on fractured ankles.

A more recent study using DEXA scans found osteopenia in 54% of young adults and osteoporosis in 14%. This is more prevalent than we might think.

Bone health in childhood and adolescence

There are conflicting data about childhood and adolescence. In young children, bone mineral density is either similar to or slightly lower than in children without PWS.

Growth hormone seems to improve bone mineral density if started very early, another reason to start early. Normal-weight children with PWS may have lower bone mineral density than children without PWS of the same weight.

My thinking is that heavier children put more weight across their bones, stimulating mineral deposition. That does not mean I think children with PWS should be heavy; it's simply what we see in the literature.

One interesting study found that an exercise program lasting only 24 weeks increased spinal bone mineral density in children with PWS. That's direct information supporting keeping children active. I don't know that there's one particular exercise; it may be more about setting aside a particular time each day for exercise that strengthens bones.

During adolescence, bone mineral density gradually decreases despite growth hormone. The theme is that growth hormone doesn't help bone mineral density after early childhood.

This may relate to sex hormones. Pubertal development is incomplete, and the sex hormones that help strengthen bones at this stage are not released in the usual amounts.

Treatment considerations include growth hormone started early, vitamin D, calcium, activity, and awareness.

If a child limps or uses an arm differently after a fall or accident, get them checked, including an X-ray, rather than assuming they'll shake it off.

For surgical planning, I need to remember that the bones aren't normal. We may need to do things differently for spine surgery to succeed.

Bone health in adulthood

Osteoporosis appears more prevalent in males than females with PWS, the reverse of the usual pattern in the general population. Fractures were more common in males as well.

Growth hormone in adults did not appear to increase bone mineral density, although it improved bone architecture. That's an interesting distinction. As we all age and lose bone density, the bone's architecture can compensate for a time by becoming more efficient. But starting growth hormone in adulthood does not help bone strength.

Sex hormone treatment plus growth hormone appears to help bone mineral density and muscle strength in women with hypogonadism, meaning they don't make as much sex hormone as expected.

The literature in men is sparse. One study suggested a decline in bone mineral density, possibly because replacement was insufficient. Endocrinologists can explain testosterone replacement and concerns about behavioral changes much better. There is a balance to strike.

A quick word about vitamin D: the lowest normal level is about 20 nanograms per milliliter. The average level reported in people with PWS is 9 nanograms per milliliter, about half that lower limit. Many consider 30 nanograms per milliliter a healthy normal.

Low levels seem primarily related to low vitamin D intake. In people with greater fat mass, some vitamin D may be stored in adipose tissue instead of circulating and being available to bone. They may need more vitamin D than is typical.

Vitamin D is also important for maintaining our ability to think. I'm on vitamin D, my wife is, and my whole family is. I think pretty much everyone in the United States should be on it. It's hard to take too much vitamin D, unlike vitamin A, which can be overdosed easily. I think it's a good supplement even if levels have been normal.

For adults, recommendations include screening for low growth hormone and continuing treatment if needed; screening for hypogonadism in adolescence and considering sex hormone replacement; and getting a DEXA scan every two to five years to follow bone health. That information can help the endocrinologist decide whether further treatment is needed. Vitamin D, and probably calcium, are ongoing considerations.

Hip dysplasia and hip development

On the left is a normal adult hip. On the right is someone with PWS whose hip dysplasia went unrecognized until maturity.

The red line shows the sourcil, the eyebrow-like roof above the femoral head. It should be rounded with good coverage on the outside. The green arrows show the distance between the inner side of the ball and the socket. That space is small on the left, but wider on the right. The ball has migrated slightly outward; that's subluxation.

Hip dysplasia is developmental. At about eight weeks of gestation, the pelvis and hips are one block of cartilage. A line develops between what will become the ball and socket. In a sense, the hip is most stable during pregnancy.

Children with PWS don't move as much in utero. That movement helps stimulate the socket to form around the ball. Less movement means less stimulation and can lead to abnormal hip development.

We worry less about the immediate effect in the child than the effect in adulthood. Dysplasia can lead to early arthritis because the joint's cartilage is overloaded and begins to break down.

Dysplasia and dislocation are different. Dislocation means the hip is outside the joint at birth. This occurs in about one in 1,000 typical births and can occur in PWS, but it is uncommon. Dysplasia is the more frequent concern.

Depending on the research, hip dysplasia is reported in 8% to 30% of children with PWS, compared with about 1% to 2% of the general population. Rates appear higher in those with UPD than deletion. Hypotonia is a major risk factor, including during pregnancy. Dysplasia appears less common in children receiving growth hormone.

What an FPWR supported hip study found

Early arthritis may lead to hip replacement. We wanted to know how big a problem this was in adults with PWS, and FPWR sponsored a database study.

Hospitals across the United States report to a database. We can identify hip replacements and how many were in people diagnosed with PWS, then do some simple math.

Across an 11-year period, there were 3.1 million hip replacements. Only 39 involved people with a PWS diagnosis. Most of those people were younger than 50, compared with the usual age over 60. So those who needed replacement tended to need it early.

One in about 80,000 replacements involved someone with PWS. Compared with a PWS prevalence of one in 30,000 at the rarer end of estimates, that suggests people with PWS were about two to two and a half times less likely to need a hip replacement.

How does that fit with frequent hip dysplasia? One explanation is remodeling.

Remodeling and when hips need intervention

Here is a five-and-a-half-year-old whose hips initially look quite concerning. The socket edge is angled upward instead of beginning to cover the ball. In a child without PWS, we'd generally be planning surgery because we wouldn't expect remodeling after about four years.

But children with PWS follow different rules. A year and a half later, the hip looks better. By age 10, it is essentially normal. There is a remarkable ability to continue remodeling, which makes observation worthwhile.

Here is another child at 13 months with sockets that don't yet match the expected shape. With observation, they pretty much normalize.

That isn't always the case. We need to watch for subluxation, with the ball moving out of the socket. This child had severe dysplasia and beginning subluxation, so we did a procedure to aim the ball farther into the socket, hoping to stimulate more normal socket growth. I don't have follow-up for this child.

Some centers use newborn hip ultrasounds. Differences in the timing of assessment partly explain why one study finds 30% dysplasia and another 8%. If you look in infancy, you find more. Six months later, there may be less because of remodeling. As we continue watching, the amount can decline.

If dysplasia is diagnosed early, the response may be bracing, often a Pavlik harness, which holds the hips bent so the ball sits deeply in the socket while lying down.

My concern is that a child already moving little may move even less in a brace. Movement is important for hip formation, metabolism, and development. I would rather avoid bracing in these situations and allow movement.

Children with PWS also often lie in a frog-like position for the first months, already positioning the femoral head deeply in the socket.

My preference is careful observation. I don't usually obtain hip images until about one year, which I'll discuss further.

Understanding scoliosis and kyphosis

Scoliosis is the side-to-side curve seen on an X-ray. Viewed from the side, the normal spine has kyphosis in the thoracic region, where the rib cage is, and lordosis in the lower back. Too much is called hyperkyphosis or hyperlordosis.

Typical children with scoliosis may have a fairly flat thoracic spine from the side, called hypokyphosis. We sometimes describe this as lordoscoliosis.

By contrast, this child with PWS has significant kyphosis along with scoliosis: kyphoscoliosis. I selected the X-ray to make the point, but that pattern is more typical of PWS, probably because hypotonia allows the body to collapse into itself somewhat.

By the end of growth, about 60% to 70% of children with PWS will have developed a curve. About a quarter develop one before their fourth birthday, compared with fewer than 1% of typically developing children. Adolescence is the second major period of onset; about 50% develop a curve during that age range. In typical children, the overall rate is about 2%.

About 15% of children with PWS need surgery. Knowing that 60% to 70% may develop a curve but only about 15% require surgery provides some reassurance. Still, many need back surgery, and reported complication rates have been as high as 56%.

Why screening matters

Missed curves concern me. This four-year-old looks fairly normal, but already has a 30-degree curve, well within the range where we'd want treatment. Here is an 18-month-old standing a little unusually, perhaps with slight asymmetry, but the curve is over 100 degrees.

That's why I favor automatic X-ray screening. Visible clinical signs can be less apparent in PWS.

We don't treat scoliosis just because of appearance. A very large curve can affect breathing and the heart. Chest deformity compresses the lungs, making it harder to get air and blood into them and harder for oxygen exchange to work efficiently.

Over time, compressed lung blood vessels make it harder for the heart to pump blood into the lungs. The heart becomes overworked, potentially leading to cor pulmonale and heart failure.

These effects occur with very large curves, over 80 or 90 degrees. But pulmonary function tests may already show reduced respiratory reserve around 60 degrees.

I want routine X-rays once children can sit well independently. If you prop a child who cannot yet sit, they collapse and the curve may look larger. If you take images lying down, gravity is removed and the curve may look smaller.

Independent sitting is usually around one year. That's also when I usually obtain the first hip X-rays.

Curves under 25 degrees are generally observed. Curves under 10 degrees aren't called scoliosis; scoliosis is a curve over 10 degrees. Above 25 degrees, we start treatment.

We're looking at the long game: what the curve looks like at maturity. If it is under 40 degrees at the end of growth, 95% of the time it won't get larger. Over 50 degrees, 95% of the time it will continue to enlarge in adulthood. Between 40 and 50 degrees is the gray zone.

Early care and positioning

Prevention is our first goal: screening, physical therapy, then casting for infants with larger curves, bracing, and ultimately surgery when needed.

There used to be concern that growth hormone could start or worsen scoliosis. We're much more comfortable now that this isn't the case and that its effects are beneficial.

In a survey about 20 years ago, we tried to correlate growth hormone use with curves that eventually required surgery. Each month's delay in starting growth hormone was associated with a 7% increase in the risk of needing scoliosis surgery. That may not sound large month to month, but it accumulates.

Another preventive consideration is delaying unsupported upright sitting. We all want to sit babies on our laps to play. But children with PWS develop arm and leg strength before trunk strength. If we sit them upright before they can support themselves, they may collapse into what we call a hypotonic slouch. We worry this could start or worsen a curve.

I ask families to use a high chair that tilts back about 30 degrees. That allows feeding, looking around, and interacting without an upright slouch. I wait until children can pull themselves into sitting before allowing more independent sitting.

The first screening X-ray is around one year. If the spine is straight, I may wait a year for the next image. If there's the beginning of a curve, I'll repeat it in three, four, or perhaps six months to see whether it worsens.

Many children get stronger and resolve their early curve, which is good news. But I want that information early.

Therapy starts from the beginning, emphasizing trunk strength and sensory integration so children use the balance systems we discussed. During crawling, I want them to become strong at crawling. Again, limb strength may develop before trunk strength.

I emphasize tummy time while awake and playing, not for sleeping. Holding the head up strengthens the neck and back. Getting frustrated and trying to roll over is exercise. Give them opportunities to work on those movements.

Spine casting in young children

If a young child develops a significant curve, we discuss casting. I want the child already sitting because learning to sit in a cast is difficult. Once they can sit, they can learn to walk in the cast; they're quite good at that.

The oldest child I've cast was almost seven. Casting takes place under anesthesia. Under age two, we change the cast every two months; between two and three, every three months; and over three, every four months. It is a commitment.

We stop when we've reached a goal: the curve is small enough to come out of the cast, or improvement has plateaued. Sometimes casting is a way to delay surgery while a child grows.

This is a casting table. Traction across the back of the head and jaw, and across the pelvis, allows access around the body. Scoliosis isn't just a side-to-side curve; it also involves rotation. While applying the cast, we try to derotate the chest as well as correct the lateral curve.

Rotation pushes some ribs backward and others forward, creating a prominence. Infants have less rotation, one reason scoliosis can be missed in them. We haven't explained that fully, but know to watch for it.

We wrap and mold the plaster, then make the appropriate cutouts.

Results from casting and the importance of follow up

We reported on our first 34 casting patients. The criteria included a curve over 25 degrees and beginning casting by age five. We found three broad groups.

The first we called the "cured" group, about a third of patients. We reduced curves below 20 degrees, from an average of 44 to 17 degrees over about a year and a half.

About half were in the "braced" group. We improved the curve but couldn't get it under 20 degrees and reached a plateau. We then switched to full-time bracing. These curves started larger, averaging 55 degrees, and improved to about 35 over a little more than two years.

The "controlled" group consisted of children we knew would need surgery but who were too young. For growing children, we need a construct that can expand, which requires a patient and bones of sufficient size.

We've also learned that we may have only about five years during which the spine can be expanded. Repeated manipulation makes it stiffer, until eventually it cannot stretch further.

If expandable implants are placed very early, we reach that limit while the child is still young. Starting at age five may take us to 10; holding off until seven may take us to 12, with a larger spine and chest.

Here is a child in the "cured" group. At 18 months, he had a 36-degree curve. We obtained excellent correction in the first cast, with further improvement through successive casts. Two and a half years after finishing, there was hardly any curve. Six years after casting, he still looked good.

At nine, a curve began returning during the second vulnerable period, adolescence. It eventually became large enough that we recently needed surgery.

Does that mean casting was for nothing? His parents would say absolutely not. He had years without a brace, years without worrying about surgery, and reached the end of growth before needing surgery. They were happy with the timeline.

This child in the controlled group had a 106-degree curve at 18 months, clearly needing eventual surgery but far too small. The first cast reduced it by about half. We kept it around 50 to 60 degrees over time.

Her parents kept her involved in activities, including hikes. They obtained a special dry suit so she could swim. When she reached five, I said she was old enough for surgery. They asked, "What's the rush? Why not wait longer?" They taught me that while casting is a commitment, some families are happy to continue if it delays surgery.

I like to say casting is survivable. Here is a photograph a patient sent showing five casts, a brace, and a nice correction.

Bracing and its limits

Bracing is used for treatable curves over about 20 to 25 degrees. Braces generally prevent progression; casting can make a curve smaller.

When someone asks why we can't simply brace a two-year-old, the distinction is that the curve may stay the same or worsen in a brace, whereas casting can reduce it.

Weight control matters for a well-fitting brace. My protocol for 25- to 30-degree curves is a nighttime bending brace that bends out the curve and helps maintain flexibility. Above 30 degrees, I add a different daytime brace designed for standing and correction.

That differs from the common approach of either a nighttime brace or a full-time brace worn day and night. I've found separate daytime and nighttime braces work well. The nighttime brace isn't designed for walking; it bends the body to the side.

Here is a ten-year-old with a 32-degree curve. Four years later, at skeletal maturity, we had limited progression to about three degrees per year. She was in the gray zone, and I think we avoided surgery.

Another child had a curve over 40 degrees at ten, but responded so well that three years later the curves were under 25 degrees. That was more improvement than expected.

But there's no silver bullet or crystal ball. Another ten-year-old started with curves in the 30s and looked good in her brace. Despite that, the curves progressed rapidly and surgery became necessary.

Goals of scoliosis surgery

Surgery is indicated for curves that are, or will be, over 50 degrees at the end of growth. We aim to improve side-to-side alignment, but in PWS, alignment from the side is particularly important.

Even in typical scoliosis, a straight appearance on the front X-ray isn't enough. Poor alignment from the side can cause trouble in adulthood. There are rules for the sagittal, or postural, profile. People with PWS follow different rules.

We hold the correction with rods and screws. My first patient with PWS came to me around age one to one and a half. This was about 25 years ago, when I didn't yet know the condition. I tried different braces and got him to five or six, but he developed a very large curve.

We used a manually expandable implant. Rods were anchored to the upper and lower spine and overlapped through a connector. Twice a year, we returned to the operating room, loosened the connector, lengthened the rods, and tightened it again.

At 16, he had a 50-degree curve, which wasn't bad. We "graduated" him by leaving the implants in place without a formal fusion. He did relatively well.

Expandable rods and completing growth

Technology has changed. This ten-year-old had an 83-degree curve, clearly requiring surgery. We used magnetically controlled growing rods. There are several types, but the idea is an internal actuator that allows external lengthening.

In this example, a magnet inside the expanded part of the rod turns when an external rotating magnet is applied. That lengthens the rod. After several lengthenings, the curve had improved. The clear segment at the yellow arrows shows where the rod telescoped out.

This is the external remote controller. Lengthening is comfortable and does not require returning to the operating room. It can be done four times a year rather than twice, making changes less abrupt and generally well tolerated.

Here is another child treated with older manually expandable implants. After years, she had finished growing and couldn't be lengthened further. We removed the rods through small upper and lower incisions and replaced them without a formal fusion.

She was already very stiff and autofusing. Avoiding a formal fusion meant avoiding a larger operation and greater blood loss.

Timing a formal fusion

What about children who have finished growing and have curves over 50 degrees?

Once I know an out-of-brace X-ray would show more than 50 degrees, I may follow the curve with in-brace images. If the brace keeps it under 50, that can buy time for growth, physiological and emotional maturity, and family preparation.

Sometimes, after a year, a family says they're ready and want to schedule surgery the following summer. Choosing a date makes it easier for everyone, including the child. It's never an emergency.

Here is a 67-degree curve and the implants we use: a rod on either side, with screws anchored from the back into the vertebrae. We aim for good side-to-side correction, but the side view is more important. This child had more kyphosis than scoliosis, shown before and after surgery.

Protecting the incision after surgery

Children with PWS need special consideration. Infection risk is higher, closely related to skin picking. We need multiple barriers to protect the incision.

With my first patient, I knew about skin picking and warned the family, but wasn't yet putting children in postoperative braces. He didn't reach the incision with his hands; instead, he rubbed his back against a doorjamb like a bear against a tree. The incision broke down and became infected. We had to remove the metal and wait a year before replacing it. This is serious.

Now, after surgery I use a vacuum dressing to manage any drainage. Because it is sealed and connected to a pump, breaking the seal triggers an alert. If the child gets a hand under it, everyone knows.

Five days after surgery, I fit a brace. Unlike the corrective brace used beforehand, this protects the incision and prevents access. It stays on except for bathing.

Families tell stories of stepping away during a shower for only a minute, distracted by a phone, cooking, or another child, and finding the child has reached the incision. Families are increasingly aware of the need for protection.

Preserving comfortable spinal alignment

Side-to-side correction matters, but front-to-back alignment is especially important. Children with PWS position themselves differently. If the implanted rods are too straight, the spine may pull away above or below them.

At the top, this is proximal junctional kyphosis; at the bottom, distal junctional kyphosis. In effect, the person says, "You've straightened me, but I don't want my head there. I want it over here."

On this skeleton, a plumb line dropped from the upper cervical spine passes just in front of the sacrum, near the center of the hips. In the individual with PWS, the line falls farther forward relative to the sacrum.

We want to preserve that comfortable alignment. That means tailoring the upper part of the construct. I generally avoid extending it too high, leaving an area where the person can transition into a comfortable position.

This exaggerated forward-head posture is shown here. We must avoid correcting someone to what we think is natural if it isn't natural for them, because they may compensate above or below the rods.

This child came from Hawaii after a spinal fusion. She developed a problem above the construct, the rods were removed, and the kyphosis worsened. We could see where she wanted her head positioned from the plumb line. We corrected the deformity while preserving that position.

Anesthesia breathing and bowel considerations

Complications can include anesthesia issues. Anesthesiologists are becoming more familiar with PWS, but an advance discussion helps ensure the team knows a child with PWS is coming and is prepared.

I ask the pulmonologist to evaluate sleep apnea and whether CPAP or BiPAP may be needed after surgery.

Gastrointestinal complications also matter. Constipation is common in PWS. After surgery, everyone has a period of ileus, when the bowel slows, and this can be worse in PWS.

What makes it more challenging is that a child may come out of the operating room asking for a hamburger. That has happened to me. We need to advance the diet carefully because progressing too fast can lead to major problems, including gastroparesis.

One child remained hospitalized a month. Each time he began recovering, someone would give him something like chicken nuggets, his bowel function would worsen, and we'd have to start over.

There is also a higher risk of spinal cord issues following surgery in children with PWS. It's not a very high risk, but higher than in typical children, and we don't yet know why.

Sometimes onset is delayed. The child may wake with normal leg movement but begin stumbling a day or two later when therapy gets them walking. We need to stay alert and keep investigating why this happens.

Growth hormone and scoliosis

A little history: growth hormone was first used in Turner syndrome. People with Turner syndrome also have a scoliosis risk, and there was concern growth hormone worsened it. Orthopedic surgeons were advised to stop growth hormone if a curve developed.

That thinking carried over to PWS. For a long time, people believed a child with PWS and scoliosis should stop growth hormone.

Expert experience and published papers showed growth hormone is, for the most part, helpful: improving metabolism and muscle strength. If anything, it may be modestly protective against scoliosis.

The same was later recognized in Turner syndrome: growth hormone doesn't cause scoliosis. As discussed earlier, delaying its start was associated with an increased risk of surgery in our survey. Only about 15% need surgery overall, but the delay increases individual risk.

I don't see a reason to stop growth hormone simply because a child has scoliosis.

Spine monitoring in adults

Recent studies have examined adult scoliosis. Not all curves require surgery. One found scoliosis in about 80% of adults with deletion-type PWS and a little under 60% in the other group. Those are high numbers.

Thoracic kyphosis appears to worsen by only about three degrees per decade. In a person who has finished growing with a large kyphosis, that rate helps us weigh whether intervention is needed.

Bone health remains important. A study of adults averaging 31 years old found vertebral fractures in 3%, resembling the compression fractures seen in older adults, where a vertebra collapses somewhat on the side view.

Other study findings make it difficult to know how to reduce scoliosis rates. Overall rates have remained similar regardless of childhood or adult growth hormone supplementation, sex hormone supplementation, sex, BMI, or UPD versus deletion.

We recommend regular spine assessments in adults, with X-rays every few years as appropriate to see whether things are worsening. I think about older adults in the general population who develop scoliosis in their 60s or 70s and may need surgery for quality of life. The PWS population may need that consideration as it ages, too.

My overall care strategy

For newborns and infants, I do a full clinical evaluation of the spine and hips without routine X-rays, checking that the hips feel located and everything looks appropriate. If there is an obvious spinal curve, I obtain an X-ray.

Otherwise, I emphasize stimulation, supervised tummy time, physical and occupational therapy, and avoiding unsupported upright sitting until children can pull themselves into sitting. In the meantime, use a feeding chair reclined about 30 degrees.

Once children can sit independently, I obtain the first spine X-rays, then at least yearly images until age four. There is a relatively quiet period from four until adolescence, perhaps age eight or ten. Then we monitor closely again because more curves develop during that period.

Treatment options include casting, generally for children under five; bracing for curves under 50 degrees; nonfusion spinal instrumentation, or NFSI, meaning expandable implants, for children under ten; and fusion for those around 12 or older.

The sitting milestone is when I begin X-rays. AFOs are worth considering when a child isn't walking by 16 to 18 months, to get them moving, exploring, and strengthening their muscles.

Thank you for your attention. We're a little over time.

Audience questions on feeding chairs

MODERATOR:

Thank you, Dr. Van Bosse, for an informative session on orthopedic challenges in PWS. We're a little over time, but can take a couple of questions.

AUDIENCE QUESTION:

Does the entire chair need to tilt back 30 degrees? Some feeding chairs maintain the hips and knees at 90 degrees and tilt the whole seat, while reclining a typical high chair opens the angle at the hips. Does that difference matter?

DR. VAN BOSSE:

I don't think it makes a difference. I wasn't aware of chairs that tilt back in space, but I like those better. I do.

Audience questions on active therapy and DMI

AUDIENCE QUESTION:

There is traditional PT and OT, and now DMI therapy, which is integrative and challenging. In my experience, some children receiving it stand and walk earlier. Have you noticed a difference, including in their spines or the need for casting?

DR. VAN BOSSE:

I haven't seen data on that. I'm not familiar with it. Can you tell us more?

AUDIENCE SPEAKER:

I'm a speech therapist. Many of my families participate in DMI. It is very dynamic and uses the whole body. There are videos of children being held upright, working on wooden planks, balancing, and practicing standing movements even before they walk.

In my opinion, some stand earlier and do better with it. I wondered whether that differs from more traditional therapy focused on the ground or stretching, and whether earlier standing changes spinal development or the time spent crawling.

DR. VAN BOSSE:

I don't know about effects on the spine, but I think more active therapy is better than purely passive therapy. I agree with you on that.

Completely anecdotally, some children I've seen with larger curves were active at younger ages. That makes it difficult to understand the relationship. I think infantile curves relate to hypotonia, yet some of these children were moving earlier than a typical child with PWS and still had large curves. There's a lot we still need to tease out.