Early-Onset Scoliosis in Cerebral Palsy

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Freeman Miller

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Abstract

There is a group of children with cerebral palsy who developed very-early-onset scoliosis before the age of 7. This scoliosis may be associated with hip dislocation or asymmetric hip contractures as the primary driving force of the scoliosis. In children with hip dislocation or asymmetric contractures and scoliosis under the age of 7 years, first the hip problems should be addressed. If the hip problems are corrected and the patient has been monitored with increasing scoliosis curve size and stiffness, then surgery should be considered. The surgical options to consider include short apical fusion, a growing rod construct using either MAGEC rod or a classic growing rod construct. Growing rods have reported very high complication rates; however, there are no reports at this time of the MAGEC rod use in children with spasticity. Children between the ages of 7 and 9 years whose scoliosis becomes large and stiff can be monitored with radiographs every 6 months. When the scoliosis curve reaches 90° or becomes very stiff, complete spine (T1 to pelvis) fusion is recommended. Children who

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develop early-onset scoliosis tend to have severe medical problems and generally are fragile with most of them having seizures and requiring gastrostomy tubes for feeding. Approximately a quarter of the children require tracheostomies. The mortality in this group of patients is approximately 25% 10 years after the spine fusion. This mortality is not related to the spinal fusion or surgery but due to the many underlying medical problems.

Keywords
Cerebral palsy · Early-onset scoliosis · Scoliosis · Growing rods · Kyphosis · Spine fusion · Pelvic obliquity

Introduction

The term early-onset scoliosis has been defined by the Scoliosis Research Society as occurring in children before the age of 10. This is the age that scoliosis starts to develop in children with cerebral palsy with the most common onset time between the ages of 8 and 10. Therefore this definition includes many of the children with CP. The vast majority of children with cerebral palsy including those with gross motor function classification system (GMFCS) levels IV and V have spines that are very flexible although they tend to lean over and have the appearance of scoliosis before the age of 8–10. On careful physical examination however, these spines tend to be very flexible, and they can just as easy side bend to the opposite direction. This concept of early-onset scoliosis in CP should specifically focus on those children who developed a very significant structural curve which is felt to be due to structural spinal deformity, not a collapsing flexible curve. It is also important to make sure the scoliosis is not being driven by infrapelvic pelvic obliquity causing a compensatory scoliosis. This compensatory scoliosis is really part of the syndrome of asymmetric hip contractures often with hip subluxation or dislocation and should be seen as part of that treatment algorithm. In these children, the hips need to be addressed first, and then the scoliosis almost always will stabilize or even have significant correction (Case 1). The group of children who classify as early-onset scoliosis are those in whom the pelvic obliquity is not being driven by deformities at the hips, but they have a fixed deformity of the spine which may be causing the pelvis to tilt. Often a component of the scoliosis in young children includes a significant kyphosis which needs to be monitored. Many of these early-onset scolioses also are not the typical long C-shaped curves that are more commonly seen with the adolescent developing scoliosis of cerebral palsy. The group of children who develop true early-onset scoliosis with cerebral palsy often are part of congenital syndromes or have congenital syndromic features. This makes this group of children very heterogeneous and in many ways quite difficult to classify. The goal of this chapter will be to review the various presentations that developed in early-onset scoliosis in children with cerebral palsy.

Natural History and Etiology

Early-onset scoliosis in children with cerebral palsy should be classified into those children who really have the very-early-onset scoliosis that this is significant fixed structural deformity before the age of 7 years. Children with this extreme early-onset pattern of scoliosis tend to be very rare. In my experience they only occur in approximately 1 out of every 200–400 children with scoliosis. The pattern of scoliosis in these very young children tends to be sharper, and more stiff scoliosis curves often with significant kyphosis and often involving more the thoracic spine than the typical long curve are seen in adolescents with cerebral palsy scoliosis (Case 2). There is a second group of children whose scoliosis develops early defined as those between the ages of 7 and 10. Many of these children are at the upper end of this age range and in fact fall into the typical pattern of cerebral palsy scoliosis development. There are some children who are very small at that upper end who may be similar to the very-early-onset type. The diameter of the chest wall continues to grow, and over time the ribs tend to grow down over the pelvis; however, this does not seem to cause any problem (Case 3). This

usually occurs in child fused before age 9; however, in some children fused at a later age, this may still become evident. The life expectancy of these children with cerebral palsy and very-early-onset scoliosis fused before the age of 9 years is more limited than the children who developed scoliosis later. In one review the mortality rate was 28% after 10 years and predicted to be 50% at 15 years (Sitoula et al. 2016). It is our belief that the cause of the increased mortality is due to the severity of the cerebral palsy and not due to the early spine fusion; however, there is no definitive evidence to support this.

Early-onset kyphotic deformity may also occur, almost always it is a flexible collapsing kyphosis in sitting, and when the child lies supine, they will be flat or near flat. It is only in the very rare case that this becomes stiff before the age of 9 or 10 years. Paying attention to good wheelchair adjustment especially keeping the lap tray high will tend to be able to manage this until adolescent growth. If the curve is becoming stiff in a very young child under age 7 years, it is always associated with scoliosis in my experience. The management then follows the scoliosis protocol. Symptomatic hyperlordosis is never been seen in my experience in the young child.

Treatment Options for Early-Onset Scoliosis

The treatment of early-onset scoliosis is focused at preventing severe and progressive curve progression to the point where restrictions on respiratory function and gastrointestinal function. There is a general concern in the treatment of early-onset scoliosis to preserve spinal growth potential because spinal growth potential can impact on the development of pulmonary function especially on lung volume. In the children with cerebral palsy whose physical function usually is extremely limited because of their very severe neurologic disability, the concern for preserving pulmonary volume may be less important, and furthermore these children have normal potential for lateral chest wall growth. They do not have the restrictions that many congenital syndromes have with regard to lateral chest wall growth. The option to consider in treating early scoliosis is first observation. When the scoliosis is initially identified, observation should always be the first choice. Sometimes these curves will be fairly substantial; however, they will remain stable when the child is not growing very fast.

Therefore, one can monitor them with x-rays every 4–6 months to either document progression or document that the curve is stable. Other options for treating early scoliosis include bracing and casting which are more commonly used for idiopathic early-onset scoliosis. These options have very limited possibility in the children who are medically extremely fragile as the ones with cerebral palsy almost always are. The primary option is surgical treatment which includes early fusion and growing rod constructs.

Early Short Fusion as an Option

Occasionally, children develop a spinal curve that is very stiff with a severe magnitude as early as age 3–5 years. These curves may approach 90° in magnitude and may become very stiff, making orthotic management difficult. Orthotic management also usually fails because the children have difficulty tolerating the orthosis. If the curve is in the thoracic area only, a limited anterior and posterior fusion in the thoracic area using sublaminar wires or pedicle screw construct is the recommended treatment (Cases 2 and 5). The goal of this treatment is to stabilize the sharp curve section and then extend the fusion to the pelvis in case the curve deteriorates as the children grow. This extension will at least allow children to gain height through growth from the lumbar vertebrae. In one child, this growth continued substantially, and a scoliosis in the lumbar spine has not developed; therefore, no additional treatment was required (Case 4). In another example, growth continued for 6 years, and then a rather severe curve extending into the lumbar spine and pelvis developed, requiring a revision surgery. Substantial height was gained, however, and treatment was successful (Case 5). Another option is to implant a spinal rod that is fixed with sublaminar

wires but does not have a fusion. It is hoped that children will continue to get taller, growing off the superior end of the rod; however, our success with the concept has not worked well. The growth phenomena has been documented to occur in children with muscular dystrophy who were not fused and who continued to grow into their adolescent years (Miller et al. 1992)

Growing Rod Constructs

There has been increased interest in the use of growing rod constructs over the past 10–15 years. These systems usually have fixation at one end often in an area of short fusion and then have an extending rod connecting these areas. Every 4–6 months, the rods are distracted. This allows the spine to grow and to correct the spinal deformity over time. Because these systems require frequent surgeries, they have a very high rate of complications especially in the complex child with early-onset CP scoliosis. One report of 28 growing rod patients noted a complication rate of 84% and mortality rate of 18%. CP was not separately reported (Phillips et al. 2013). One study has reported on a series of 27 children with CP with an acceptable outcome; however, only 25% had completed treatment, and there was already a 30% deep wound infection rate (McElroy et al. 2012). The newer version of the growing rod construct is the MAGEC rod which is magnetic motor that allows lengthening without the need for doing additional surgery. At this time there are no reports specific to children with CP. This might have the potential for using the growing concept; however, the problems of weak bones and spasticity will likely continue to make this a high complication procedure (Case 7).

Spinal Deformity in Very Small Children Who Are Older

Growth inhibition in children with severe neurologic disability may be significant with children being only 15 kg in weight at 10 years of age but also having severe scoliosis. These children should be instrumented and fused, but instead of using the regular 6.5-mm-diameter Unit rod as used in the normal larger child, the thinner 5.0-mm Unit rod should be used. The large Unit rod can be used in most children up to 15 kg in size; however, it is extremely difficult, as the rod gets shorter and because of its severe stiffness, to be able to manage it in the small thin osteoporotic pelvis. The smaller Unit rod is available up to 330 mm in length and is much easier to use and has sufficient strength for these small children. The thinner rod should not be used in taller children because of the risk of rod fracture and the development of pseudarthrosis, which would subsequently require a revision. The use of small screw and rod systems is another option.

Another area related to the very small child is the increasing interest in growth attenuation or suppression of young children 3–5 years old, so they will remain small in size and stature to make care easier. This means giving growth and androgenic hormones to drive them through puberty when they are 3–5 years old and stopping their growth. One of alleged benefits of this treatment is that it will prevent scoliosis. This benefit has not been confirmed due to this practice still being relatively infrequent. We have not seen a child that developed scoliosis after this treatment.

Recommendation for Early-Onset Spine Fusion in Children with CP

Children less than age 7 years who develop a structural scoliosis over 60° which has substantial stiffness fall into the very young group. Always the first additional review should be to evaluate the hips, and if there is hip dysplasia or asymmetric contractures driving an infrapelvic pelvic obliquity, this should be addressed first (Case 1). If the scoliosis is flexible, almost always the scoliosis will improve after correcting the hips. This improvement will be temporary, and the curve will typically start the normal increase again at 9–10 years of age. If the hips are normal and the spine is developing increasing stiffness as documented with multiple examinations over 1 year, intervention is indicated (Case 6). During

this year of observation or till progression is clearly documented, body jacket bracing can be attempted if the child can tolerate. When progression is documented to be structural by also having increased stiffness, consider surgical intervention. If the child is relatively healthy and has adequate bone density, a growing rod construct, maybe a MAGEC rod, can be considered with recognition of the likely high complication rate. If the child is medically fragile and/or has severe bone fragility, a short segment fusion at the apex of the curve can be performed (Case 2).

Children with cerebral palsy between the ages of 7 and 9 years old fall into the technical definition of early-onset scoliosis; however, these children can almost always be managed with wheelchair seating adjustment and comfort orthotics until the sitting scoliosis reaches 90°. Almost all curves in this age range stay flexible until around 90°. The whole spine should then be fused. Most of the children who are developing scoliosis in this early time also tend to be very medically fragile, and some may have hip subluxation. If there is a structural scoliosis and a dislocated hip, it usually means the pelvic obliquity has a combined infra- and suprapelvic pelvic obliquity. In these cases it is better to wait and correct the spine and then 6 months later address the hips (Case 6).

Children with cerebral palsy who are over 10 years old even if they are small in their body size should be considered for full spine fusion when the scoliosis approaches 90° or the stiffness makes it impossible to bend the child to the midline (Case 3). These are the indications for the child who has significant remaining growth, not for the individual who is skeletally mature (▶ Chap. 118, "Surgical Treatment of Scoliosis Due to Cerebral Palsy").

Long-Term Outcome of Early Spine Fusion

We have reviewed our children who were fused between 4.4 and 9.9 years of age with a mean of 8.3 years. They were reviewed at a mean 10-year follow-up. These were very medically involved children with 94% having seizures, 88% having gastrostomy tubes for feeding, and 27% having permanent tracheostomies. There was 28% mortality at 10 years of follow-up. None of the deaths were related to the spinal surgery or due to restrictive lung disease (Sitoula et al. 2016). This review suggests that this is a very fragile group of children, and we need to also focus on quality of life. For this review we did not have any quality of life measures, but parents report children being more comfortable and better able to sit after the spinal surgery.

Conclusion

Early-onset scoliosis in children with CP tends to fall roughly into two groups, the very early group with deformity before age 7 years and those 7–9 years of age. For the early group, clinically monitoring to document progression in curve size and magnitude is the first treatment. The 7–9-year-olds are almost always similar to the older children with CP and can be managed with monitoring followed by fusion at approximately 90° scoliosis.

Cases

Case 1 Clarissa

Clarissa, an 8-year-old girl with severe spastic quadriplegia, presented for a second opinion concerning her progressive scoliosis. Her parents were most concerned about her increasing problems with sitting, which they perceived came primarily from her scoliosis. She had been prescribed a spinal orthosis to help with sitting and control her scoliosis. She was fed orally and was small for her age but appeared well nourished. She was taking Tegretol for seizure control and had not had a seizure for 6 months. She was a dependent sitter and had minimal function in her hands. On physical examination she was noted to be diffusely spastic with mild shoulder contractures. The spine

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had a flexible scoliosis, and the hips were limited to 10° of abduction on the left side and 50° of abduction with some limited adduction on the right side. The knees had a popliteal angle of 60° bilaterally, and the feet were controlled with solid ankle-foot orthotics with minimal fixed deformity. Observation of her sitting demonstrated rather poorly adjusted chest laterals, as she was hanging over the lateral on the right side. A radiograph of the spine demonstrated 48° of scoliosis (Fig. C1.1), and the right hip was dislocated, and the left hip appeared to be abducted in the classic wind-blown deformity (Fig. C1.2). Based on this assessment, it was concluded that she had a primary infrapelvic pelvic obliquity due to the spastic hip disease. It was recommended to her parents that she have a repair of the hips by bilateral femoral shortening derotation, varus osteotomy, adductor muscle lengthening, and peri-ilial pelvic osteotomy (Fig. C1.3). Following this procedure, she could sit much better until age 12 years when her sitting again deteriorated, and the pelvic obliquity now was caused by suprapelvic pelvic obliquity coming from a progressive 74° scoliosis (Figs. C1.4 and C1.5). This was corrected with a Unit rod instrumentation, and she was again comfortable as a sitter (Fig. C1.6). This case demonstrates the importance of making the correct diagnosis of the pelvic obliquity, because correcting the spine will not help treat the symptoms of infrapelvic pelvic obliquity and vice versa. When in doubt, the spine should be corrected first if there is a significant scoliosis.

Case 2

Hammie is a girl with severe GMFCS V spastic quadriplegia who has seizures, has

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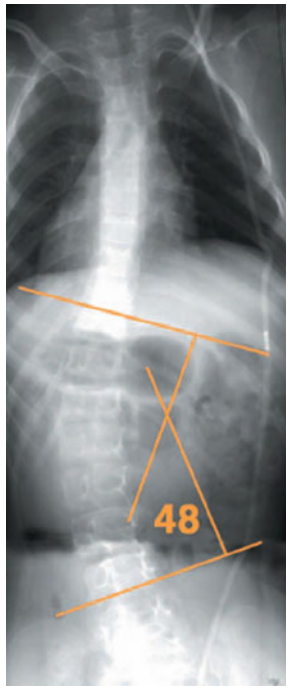

Fig. C1.1

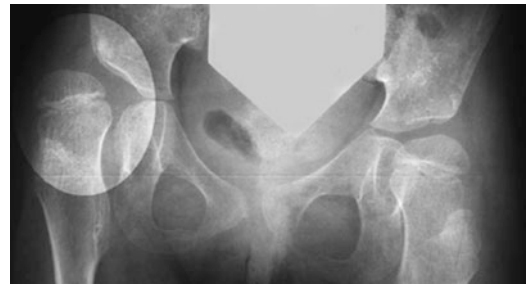

Fig. C1.2

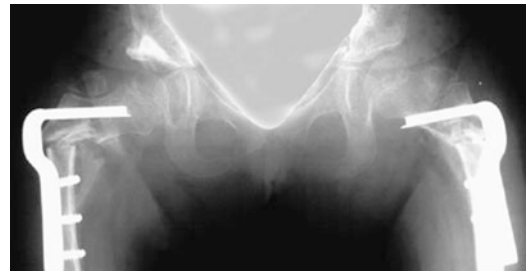

Fig. C1.3

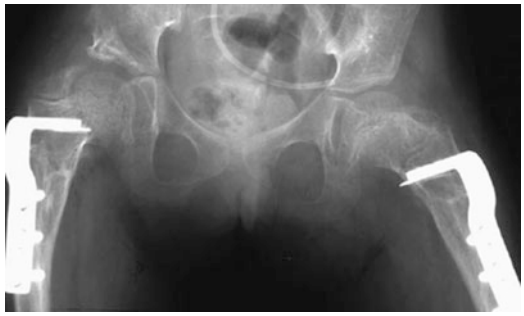

Fig. C1.4

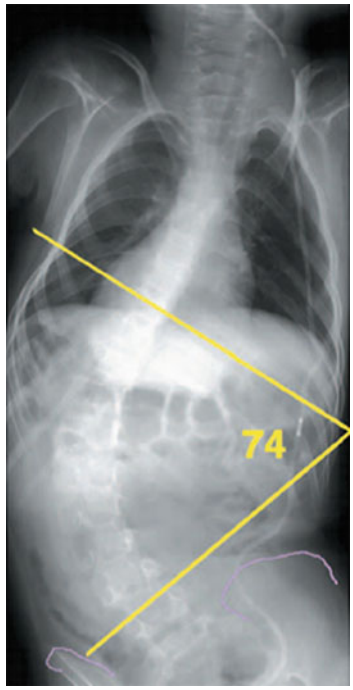

Fig. C1.5

severe respiratory impairment, and has severe osteoporosis. She has had multiple fractures and has been treated with pamidronate. By age 6, she developed a severe thoracic scoliosis (Fig. C2.1). We elected to treat this with a localized thoracic spine fusion using screws at every level because of the osteoporosis (Fig. C2.2). We continued to follow her until the

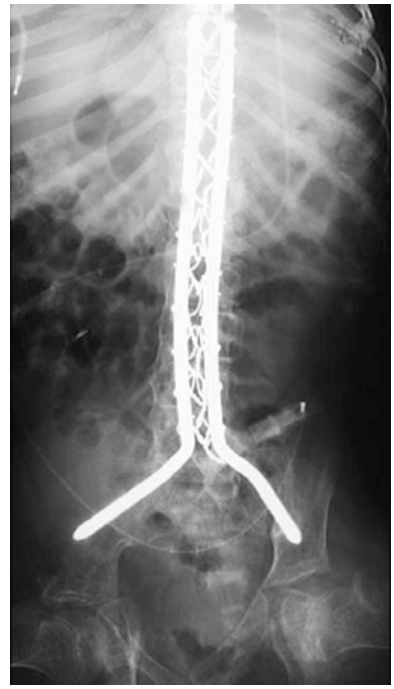

Fig. C1.6

scoliosis in the lumbar spine became so severe that she had difficulty sitting at age 10 (Fig. C2.3). At this time the spinal fusion was extended to the pelvis with correction of the pelvic obliquity (Fig. C2.4).

Case 3

Georg is now a 21-year-old man with GMFCS V quadriplegic CP who had early-onset scoliosis, developing a severe curve with pelvic obliquity at age 8. This was treated with a whole spine fusion, and during the following 12 years, he has completed his growth with a significant amount of circumferential chest growth. This has resulted in the ribs growing out over the pelvis (Fig. C3.1), but because the spine is fused, there is not enough movement against the pelvis to cause pain. He has

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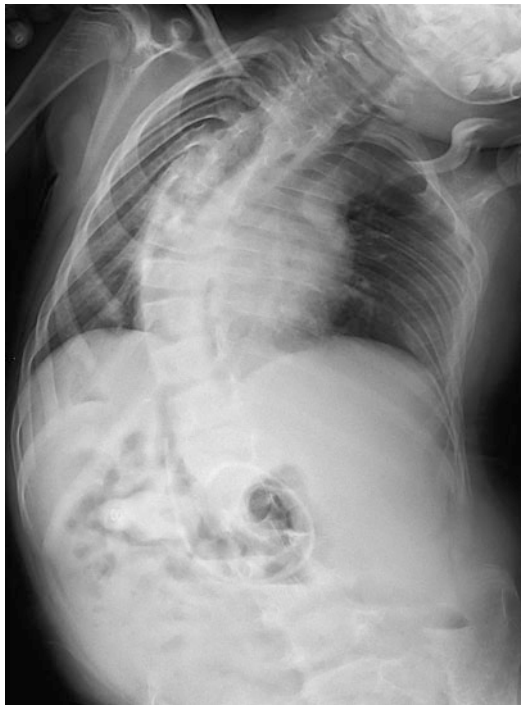

Fig. C2.1

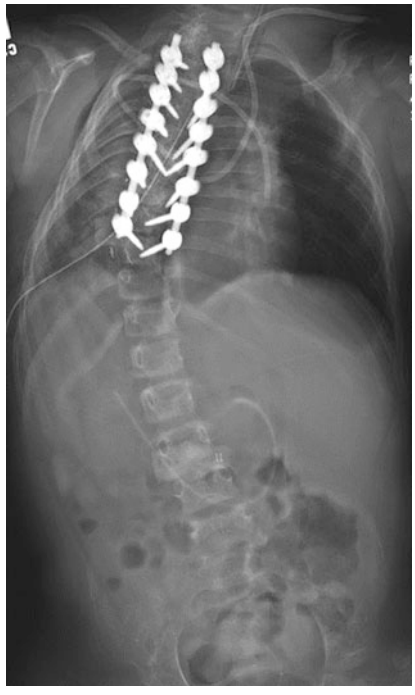

Fig. C2.2

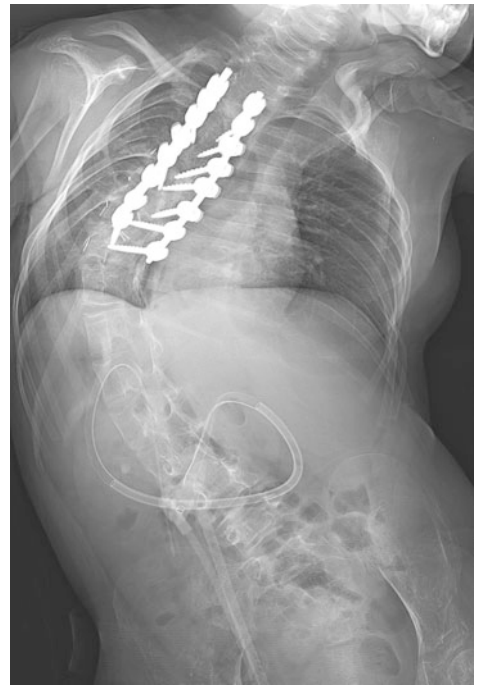

Fig. C2.3

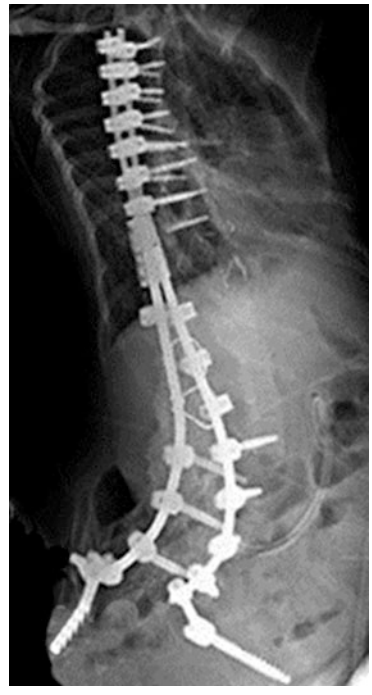

Fig. C2.4

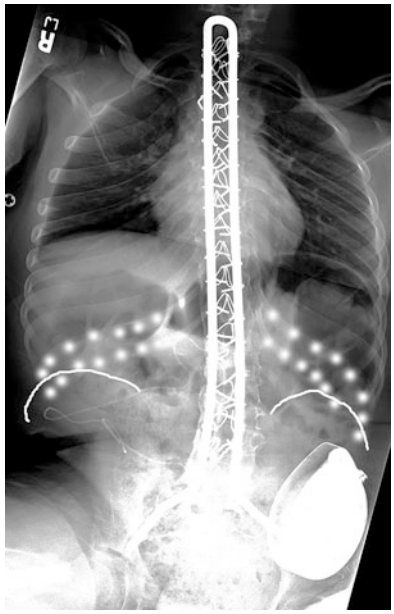

Fig. C3.1

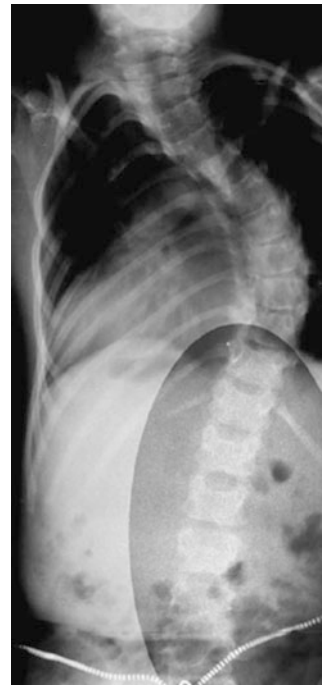

Fig. C4.1

also had no appreciated respiratory impairment or other effect of this short spine.

Case 4 David

David, a 5-year-old boy, presented being unable to ambulate with mild lower extremity spasticity, mental retardation, and very poor motor control. A relatively stiff thoracic scoliosis was noted. Because of his young age and relatively straight lumber spine, he was fused only to T12 (Fig. C4.1). Over the next 10 years, he completed his growth to a height of 170 cm, gradually developing the ability to do assisted ambulation in the home. His spine remained straight (Fig. C4.2).

Case 5 Roger

Roger, a 4-year-old boy with severe spastic quadriplegia, presented with his mother with a concern about his increased scoliosis. He also had grand mal seizures with poor seizure control, was a poor feeder, and had gastroesophageal reflux, which was being medically managed. He was scheduled to have a gastrostomy tube inserted. A radiograph demonstrated a 60° very stiff scoliosis. Because he had many gastrointestinal problems, he was a poor candidate for spinal bracing; therefore, we agreed to see him again in 4 months. During that time, he was fed by a gastrostomy tube and had gained 2 kg in the previous 2 months. A spine radiograph showed a scoliosis that had progressed to 80° and was very stiff (Fig. C5.1). He was instrumented from T1 to T12 with sublaminar wires because the majority of the scoliosis was in the thoracic spine (Fig. C5.2). He was then followed for 6 years as he grew, until he again developed

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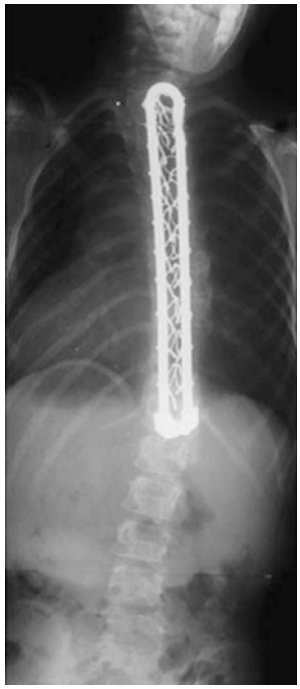

Fig. C4.2

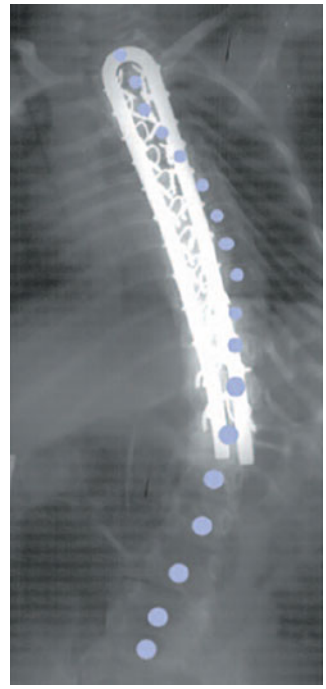

Fig. C5.2

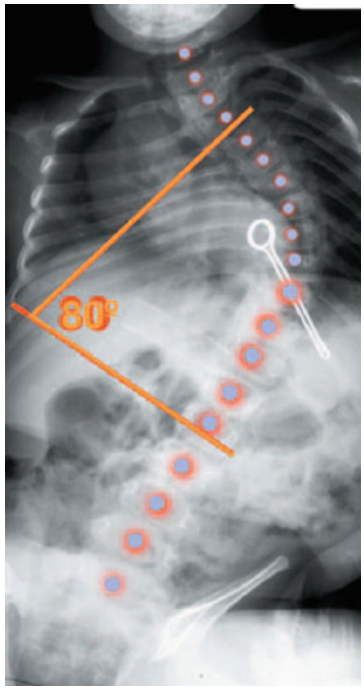

Fig. C5.1

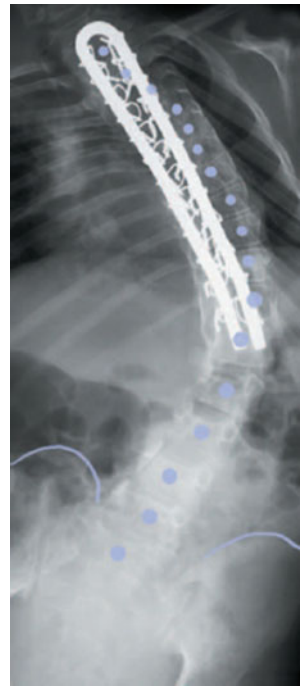

Fig. C5.3

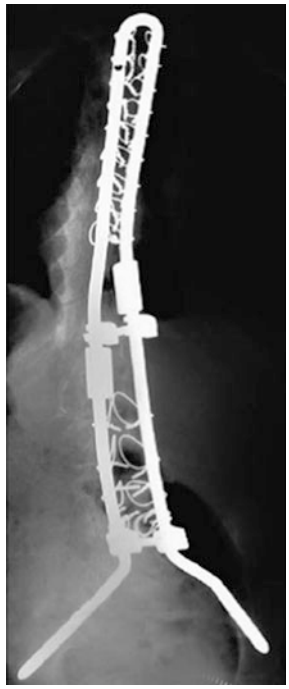

Fig. C5.4

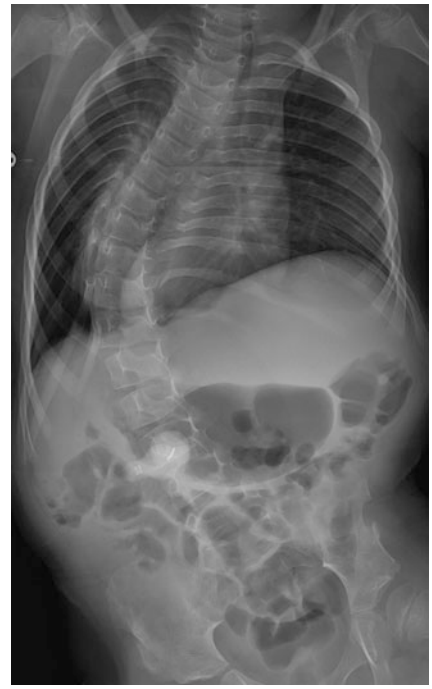

Fig. C6.1

increasing deformity distal to his previous instrumentation (Fig. C5.3). He had an anterior lumbar release and was instrumented to the pelvis, attaching to the proximal rod (Fig. C5.4). He had good trunk balance and had gained height by taking this two-step approach. Because of his seizures and poor feeding history, he would have been a very poor candidate for a subcutaneous growing rod.

Case 6 Buddy

Buddy was a 5-year-old boy with GMFCS V quadriplegia and severe hypotonia who presented for the first visit for a second opinion related to his scoliosis which was 65°, and very flexible (Fig. C6.1). His primary physician recommended growing rods. He did not have a definitive diagnosis for his condition,

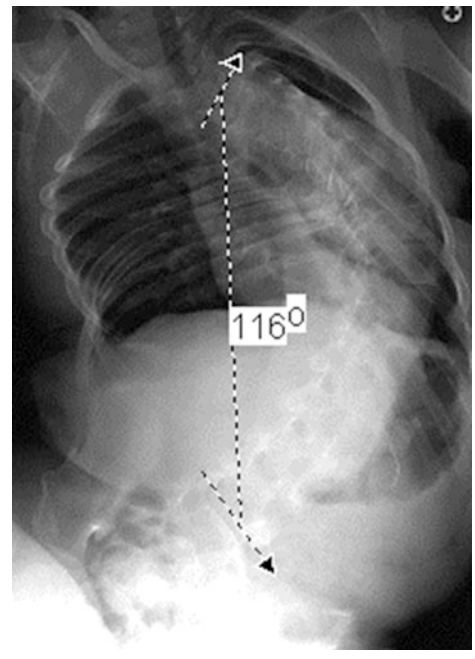

Fig. C6.2

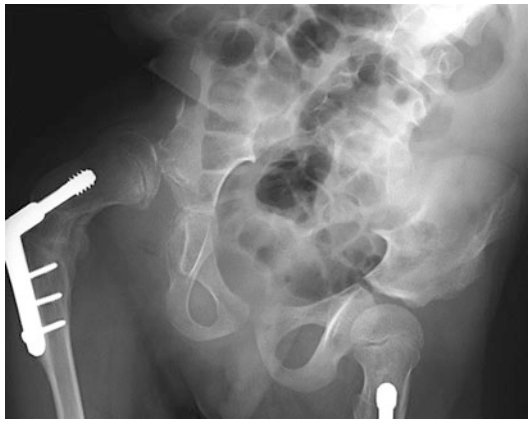

Fig. C6.3

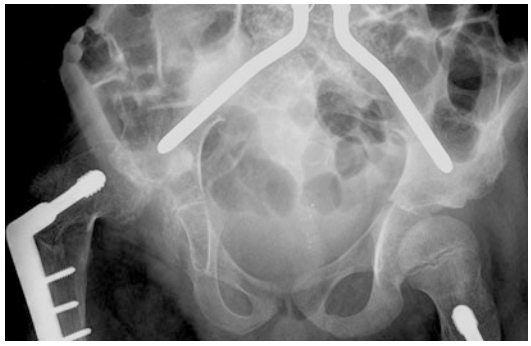

Fig. C6.4

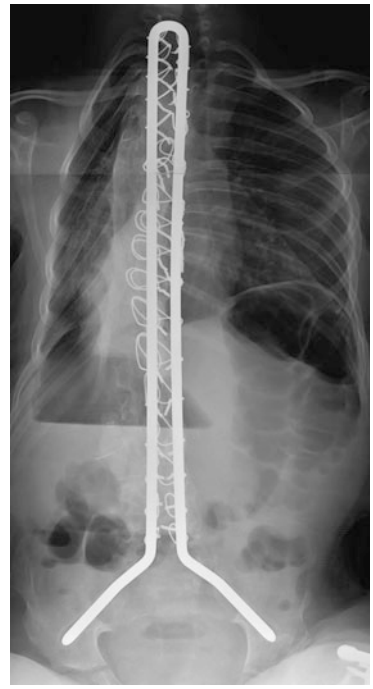

Fig. C6.5

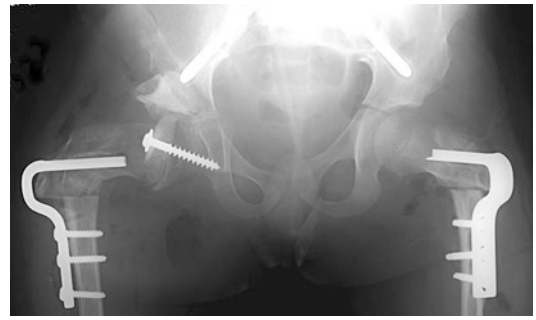

Fig. C6.6

and by parental history, he has been in his current condition since birth. There has not been any neurologic change; therefore, we presume it is a static condition although most likely some underlying metabolic problem considering the severe hypotonia. He has recently under gone varus femoral osteotomy for correction of hip subluxation. Our recommendation was for a well-fitting wheelchair with the goal of waiting until he is not able to sit comfortably. The family chose this approach, and 4 years later at age 9, his scoliosis reached 116°, and his seating became very difficult (Fig. C6.2). By this time his right hip had dislocated again (Fig. C6.3), and we intentionally waited to reconstruct the hip until the severe suprapelvic pelvic obliquity was corrected by the spine fusion (Fig. C6.4). At this time a spinal fusion was performed with excellent correction (Fig. C6.5). After the spine fusion, the right hip became painful, likely due to increased movement and stress caused by the fused spine. Six months after the spinal fusion, hip reconstruction was performed (Fig. C6.6). He has done well with a 4-year follow-up.

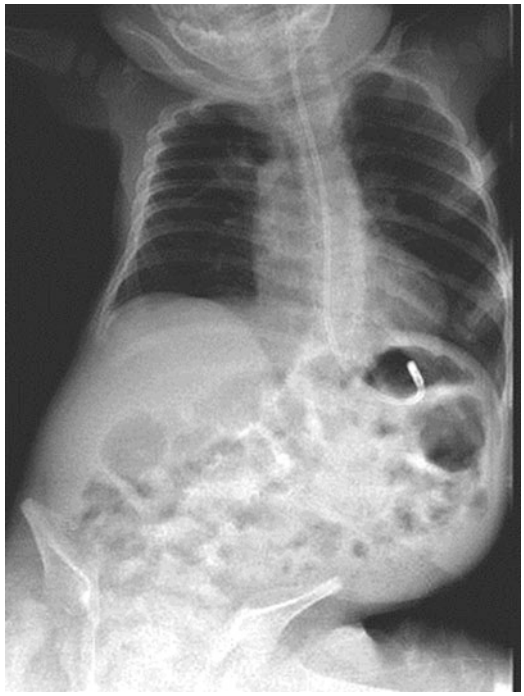

Fig. C7.1

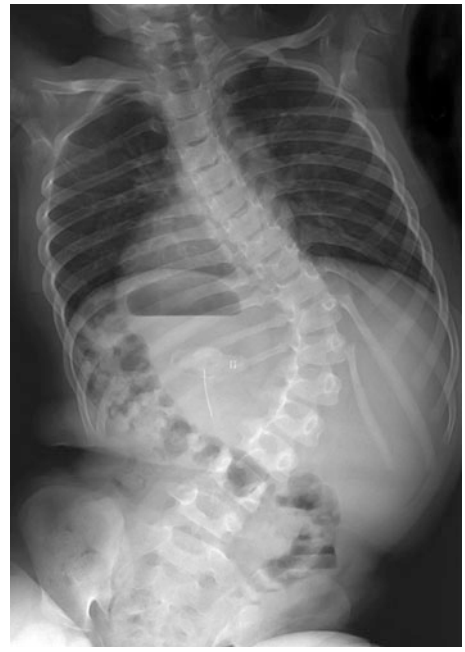

Fig. C7.2

Case 7 Shem

At age 1 year, Shem has severe quadriplegic pattern CP and was already developing scoliosis (Fig. C7.1). He was managed with wheelchair adjustments and orthotics until age 6 years (Fig. C7.2). Because of increasing problems with seating, he was implanted with a growing rod system (Fig. C7.3). Over the next 4 years, he developed several deep wound infections following lengthenings, requiring complete rod removal at age 10. An attempted spinal fusion resulted in reactivation of the infection, again requiring rod removal, and he is currently 11 years old with a 60° scoliosis, which will very likely increase as his growth continues. He also developed skin breakdown from use of spinal orthosis, requiring treatment of the decubitus. This case is representative of the severe complications frequently encountered in children with severe

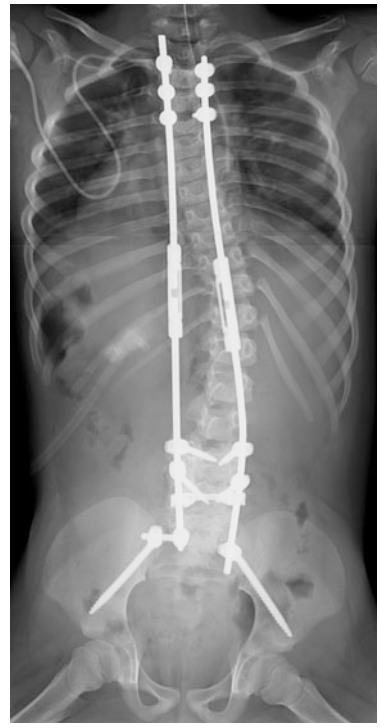

Fig. C7.3

quadriplegic pattern CP utilizing growing rods. The use of the magnetic lengthening rods may decrease these risks; however, there is still not enough experience to document this.

Cross-References

- Cerebral Palsy Spinal Deformity: Etiology, Natural History, and Nonoperative Management
- Surgical Treatment of Scoliosis Due to Cerebral Palsy

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