CASE REPORT

Decade-Long Progress of a Patient with Early Onset Scoliosis Treated with Serial Casting, Traditional Growth Rods and Posterior Spinal Instrumentation with Fusion – a Case Report

Kashfullah Khalid, MBBS; Mohd Hisam Muhamad Ariffin, MD, MS Orth; Azmi Baharuddin, MD, MS Orth

Research performed at University Kebangsaan Malaysia Medical Center, Kuala Lumpur, Malaysia

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Abstract

Early-onset scoliosis poses unique challenges for spinal and pediatric surgeons, as well as clinicians. Without intervention in pediatric patients, the spinal curvature may worsen, leading to abnormal thoracic cage growth, leading to abnormal lung development and poor respiratory tolerance. Treatment aims to correct the deformity while preserving the spinal mobility during growth. This is important because the growing spine will allow the child to achieve optimal height, a healthy thoracic cage, and cardiopulmonary system development. We can divide treatment modalities into observation, serial casting, bracing, and surgery. We are presenting a decade long progress report of a patient who initially underwent treatment with serial casting, growing rods, and definitive fixation for early-onset scoliosis. We strive to illustrate the complexities of early-onset scoliosis treatment by progressing from bracing to the application of a growth-friendly rod system, and finally to definitive posterior instrumentation.

Level of evidence: IV

Keywords: Early-onset scoliosis, serial bracing, growth rods, rod failure, spinal fusion

Introduction

Scoliotic deformity (spinal curvature >10 degrees) in patients ages 10 and below is considered to be early-onset scoliosis (EOS).[1] EOS could be idiopathic, but it is mainly non-idiopathic, with etiology due to congenital vertebral anomalies and neuromuscular or syndromic disease.[2] Young patients with major spinal deformities during rapid growth spurts are at risk of developing thoracic insufficiency syndrome, especially those with syndromic or congenital scoliosis.[3] Different syndromes have different specific defining characteristics, but those with scoliosis often follow similar treatment algorithms, leading to their grouping under the term syndromic scoliosis.[4] Both incorrect formation (hemivertebra) and segmentation failure (unsegmented bar) can cause birth defects in the spine, leading to scoliosis. Neuromuscular disorders such as cerebral palsy, spinal muscular dystrophy, or Friedreich's ataxia led to muscle tone imbalance without the presence of previous congenital or structural abnormalities.[5] The idiopathic group refers to patients with EOS who have no identifiable causal factors or associated diseases.[4,6] This group of patients often receives a diagnosis of EOS based on clinical and radiological findings, but the cause remains unknown, potentially multifactorial in origin.[4,6] Consensus agrees that EOS curvature progresses with time; hence, patients with earlier onset will have worse final curvature and prognosis.[7-9] Untreated, abnormally large spinal curves make it difficult for the lungs to expand and for alveolar cells to multiply.[9] They also hurt the heart and lungs, leading to cardiopulmonary failure.[9]

Case presentation

In 2013, we received a referral for a 5-year-old girl with thoracolumbar scoliosis. To relieve her respiratory symptoms, we treated her underlying asthma with inhalers. Furthermore, the child was born prematurely at 32 weeks and was underweight at birth. Otherwise, she has no syndromic disease. Initial whole spine radiographs showed Cobb's angle of 61 degrees over the thoracic region (T4-T11) and 100 degrees over the lumbar region (T11-L4) [Figure 1]. Clinically, she has a left-sided thoracolumbar hump when bending forward, with the right shoulder tilted downwards upon standing upright [Figure 1].

Corresponding Author: Kashfullah Khalid, Department of Orthopaedics and Traumatology, University Kebangsaan Malaysia Medical Center, Kuala Lumpur, Malaysia

Email: kashfullah86@yahoo.com

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Serial casting

We conducted serial casting from 2013 to 2017. Measurements obtained after each cast application controlled the spinal curvature progression [Table 1].

Figure 1. Clinical pictures and initial presenting radiograph images of the whole spine

Table 1. Comparison of Cobb's angles during serial casting (2013-2017).

Serial Casting				
Session	**Age**	**Cobb's Angle Measurement (degrees)**		**Kyphotic Angle Measurement**
		Thoracic	**Lumbar**	
Initial	4	61	100	56
1	5	32	61	51
2		36	82	48
3	6	49	63	46
4		50	69	41
5	7	53	83	35
6		55	92	37
7	8	56	76	40

Posterior instrumentation with Traditional Growth Rods (TGR)

The patient's scoliosis progressed as she grew. At 8 years old, we decided to perform surgery, which involved instrumenting the patient's spine with traditional growing rods. The patient was brought to the operating theatre to lengthen the growing rods every six (6) months. The angular measurements taken before the surgery, immediately after the surgery, and after the lengthening procedure are shown [Table 2; Figure 2].

Table 2. Angular measurements during traditional growing rods.

Posterior Instrumentation with growth rods				
Session	**Age**	**Cobb's Angle Measurement (degrees)**		**Kyphotic Angle Measurement**
		Thoracic	**Lumbar**	
Initial	8	56	76	24
Post Op		55	73	33
1	9	51	67	25
2	10	44	54	20
3	11	49	58	19

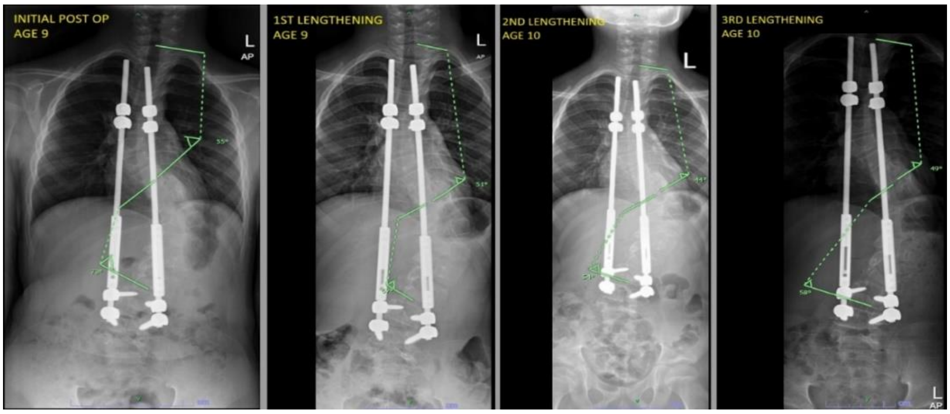

Figure 2. Whole spine AP radiographs during traditional growth rods lengthening

The patient, who was 12 years old, came to us a few months after the third lengthening procedure, experiencing an abrupt increase in pain and a deterioration in their spinal deformity. Repeated whole spine radiography and computer tomography studies revealed a fracture of bilateral growth rods [Figure 3].

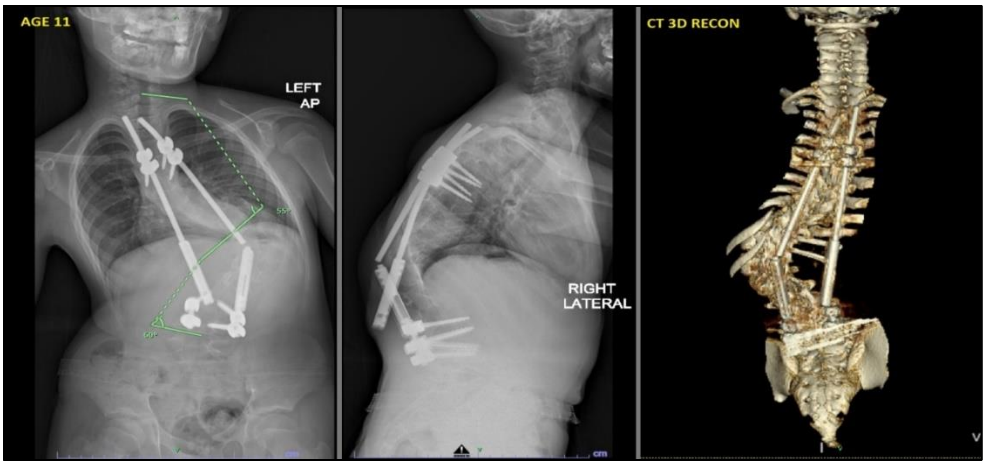

Figure 3. Whole spine radiographs show bilateral growth rod fractures

Removal of growth rods and definitive posterior spinal instrumentation and fusion (PSIF)

We took the patient to the operating room and removed the growing rods, followed by instrumentation and fusion from T5 to L4 vertebrae [Figure 4]. Postoperatively, there was no more soft tissue impalement previously due to fractured growing rods [Figure 4]. From 2020 onwards, we closely monitored the patient for two weeks until the surgical wounds healed, then for six months, and then yearly, using repeat radiographs. The patient is 16 years old and currently attends secondary school [Figure 5].

Figure 4. Post-operative radiographs with clinical pictures pre- and post-operatively after growing rod removal and PSIF

Figure 5. Whole spine radiograph and clinical images at age 16

Discussion

The role of serial casting

Bradford *et al.* proposed mechanical and external force casting to straighten abnormal spinal curvatures in 1863.[10] Mehta later popularized serial casting by using the elongation-derotation-flexion approach[11,12] to correct scoliotic deformity using the patient's natural growth. Children with EOS received casts every 2-3 months at the earliest opportunity to achieve optimal curvature correction of scoliotic deformity.[13,14] Patients with a severe scoliotic deformity of less than 30 degrees could achieve complete correction, according to his findings.[11] Nonetheless, for patients presenting after the age of 2 with more pronounced curvatures, this approach was only capable of diminishing the scoliotic deformity without attaining complete correction.[11] The documented side effects are quite minor and primarily associated with the application of body casts, including skin breakdown or irritation, musculoskeletal discomfort, and intolerance.[14] Nonetheless, the advantages of serial casting stem from its non-invasive nature, its capacity to control spinal curvature progression in a developing child, and the opportunity it provides to delay surgical intervention.[11,14]

Timing to start growing rod treatment

Growth potential and the degree of curvature are two factors that are considered while determining whether or not to perform surgical intervention for early-onset scoliosis.[14,15] To enable the growth of the thoracic contents, particularly the lungs, the procedure is primarily required to fulfill the primary necessity. As a consequence, the obvious indicators include a very young age and prominent curves. Systems based on distraction are utilized frequently to correct EOS surgically. The growing rods and the vertical expandable prosthetic titanium rib (VEPTR) are the two primary kinds that fall under this category.[3,6] Both methods are based on the notion of delivering traction force to the scoliotic spine between proximal and distal anchor points that are connected by expandable rods. These rods go through a process of periodic lengthening as the child develops, which will ensure that the correction is maintained. Recently, S. Wang *et al.* investigated the best time for growing rod system surgery and concluded that it should wait until after the age of 6.[16] The rationale behind the recommendation was that children in the low-age group (below 6 years) have approximately 47% risk of developing implant-related complications such as rod fracture, screw fracture, proximal junctional kyphosis, distal junctional kyphosis, coronal imbalance, and pedicle screw, hook, or rod dislodgement.[16] These complications stem from physiological factors in the younger cohort, including reduced soft-tissue coverage, smaller bone structures, diminished physiological reserve relative to older children during postoperative recovery, and an increased number of lengthening procedures preceding definitive instrumentation.[16]

Complications of growing rod systems

Implant-related issues occur at a rate of 20% at index surgery and up to 58% during the treatment period in EOS patients who get instrumentation to fix alignment and deformity issues without fusion.[16] This is because the construct is load-bearing instead of load-sharing. These structures frequently break as a result of the metal or implant experiencing fatigue failure due to continuous cyclical loading.[16,17] Other things that can cause an implant to fail are pedicles that aren't appropriately formed or are not fully developed, anatomy that has changed or isn't fully developed yet, which means that bigger and thicker implant designs can't be used, and poor fixation due to poor bone quality.[17,18] Dual rod TGR constructs are superior to single rod constructs in terms of coronal correction and lengthening; nevertheless, larger constructs are more likely to result in surgery site problems, such as skin breakdown and implant prominence.[17] The strongest construct in pull-out tests is four pedicle screws inserted in two adjacent bodies at each anchor site.[17] However, crosslinks do not appear to improve fixation.[17] Rod fracture is the most common implant-related complication in EOS patients treated with growing rods, and it accounts for 15% of all complications. [17] Our case showed that both growing rod fractures occurred simultaneously. Additional adverse outcomes associated with growth rod systems include junctional failure, pseudoarthrosis, screw retraction, failure to achieve lengthening, wound dehiscence accompanied by implant exposure, infections, secondary lumbar scoliosis, and medical complications such as pulmonary embolism or pulmonary insufficiency.[16-18]

Conversion from growing rods to definitive spinal instrumentation with fusion

The ultimate objective in the treatment pathway for children undergoing growth-friendly procedures is their transition to definitive fusion surgery (often also termed "graduation").[13,15,19] The pertinent question at that juncture is whether the metalwork should remain, be substituted with a permanent fusion system, or be completely removed.[19] Factors affecting the selection of the optimal graduation approach encompass the primary diagnosis, the status of the spine and chest wall post-treatment, and the employed instrumentation.[19] The predominant method employed is the extraction of the metalwork and the introduction of a definitive fusion system that facilitates further rectification of deformities during the fusion process. The date of conclusive fusion remains a contentious issue. Factors such as sagittal misalignment, issues resulting from previous implants, and major curvature deformity that is either unacceptable or worsening support posterior spinal fusion.[15]

Fusion is relevant only after the child has attained adequate spinal growth, thoracic cage volume, and pulmonary function, as it inhibits future spinal development.[13.19] By the age of 10, the majority of children will have completed most of their thoracic spine development.[11,15,19] During puberty, typically occurring between the ages of 11 and 13, the T1-S1 segment retains approximately 7cm of potential growth, comprising 4cm in the thoracic region and 3 cm in the lumbar region.[15] If a growing rod system is employed, more distraction no longer produces significant benefits at this stage (the principle of diminishing returns).[14,15,18] As a result, it is more rational to forgo the minimal remaining development potential and proceed with spinal fusion rather than risk the progression of deformity or complications associated with instrumentation.[15] The patient, aged 12, experienced failure of the TGR hardware, prompting the decision to remove it and construct a new definitive fusion device. Final curvature correction could be achieved during the final posterior spinal fusion surgery.

Conclusion

The nature of the deformity, the underlying etiology, and the existence of medical comorbidities all significantly influence the management of EOS, which can be highly complex. Serial casting and bracing, followed by 'Growth-friendly' surgery utilizing growth stimulation or growth guidance, is the preferred therapeutic approach for children with EOS. These treatments generally serve as a provisional intervention in a developing skeletal system, deferring the requirement for a definitive spinal fusion until a later stage of life. The principal aim is to correct the scoliotic deformity, promote spinal growth, and safeguard cardiopulmonary development. Effective treatment of this complex disease depends on the multidisciplinary team strategy being used. Achieving good treatment outcomes depends on the cooperation among surgeons, pediatricians, respiratory doctors, anesthetists, physiotherapists, occupational therapists, orthotics, nutritionists, nurses, pain management specialists, and psychologists.

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Kashfullah Khalid MBBS [1]
Mohd Hisam Muhamad Ariffin MD, MS Orth [1]
Azmi Baharuddin MD, MS Orth [1]

1 Department of Orthopaedics and Traumatology, University Kebangsaan Malaysia Medical Center, Kuala Lumpur, Malaysia

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