Standard and magnetically controlled growing rods for the treatment of early onset scoliosis

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Abstract: Distraction based spinal instrumentation represents the most common and standard surgical technique to correct early onset scoliosis (EOS), i.e., scoliosis which has been diagnosed before the age of 10 years. Surgical treatment of EOS aims at controlling spinal deformity while maintaining spinal growth which is mandatory for the development of normal lung capacity. To achieve these goals the spinal instrumentation needs to be distracted to facilitate spinal growth during treatment. Distraction can be obtained by repeated surgical lengthenings (traditional growing rods, TGRs) or using magnetically controlled growing rods (MCGRs), which can be lengthened using external remote controller on an outpatient basis. The outcomes of TGR instrumentation for EOS are well described with follow-up until skeletal maturity: normal spinal growth can be maintained, 40–50% of the scoliosis can be corrected, but there is an over 50% risk of complications including deep wound infection, rod failure, and instrumentation pull-out. MCGR instrumentation may reduce the risk of wound related complications, provides similar deformity correction, but may not provide as much spinal growth. Metallosis around the instrumentation necessitates MCGR removal and definitive final instrumented fusion at the end of growth friendly management. Even severe EOS can be treated using distraction based spinal instrumentation.

Keywords: Early onset scoliosis (EOS); traditional growing rods (TGRs); magnetically controlled growing rods (MCGRs); complications; spinal fusion

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Introduction

Early onset scoliosis (EOS) is defined as a scoliosis, which occurs before the age of 10 years (*Figures 1,2*) (1,2). If EOS is left untreated or if treated by spinal fusion resulting into a short trunk and spinal height, EOS is associated with increased morbidity and even mortality (3-5). EOS is a heterogeneous disorder and therefore a widely accepted classification has been published (1). This classification includes age, etiology (congenital or structural, neuromuscular, syndromic and idiopathic), magnitude of scoliosis and kyphosis as well as rate of deformity progression. Growth-friendly management is indicated for progressive scoliosis (4,6,7). EOS may progress rapidly during early growth and, thus, early diagnosis and referral to a pediatric spine unit is necessary (1,7).

Spinal instrumentation is needed in the surgical treatment of EOS to correct scoliosis and to allow spinal growth, which is necessary for normal development of lung capacity (8-11). Skaggs *et al.* (12) have provided a classification for growth-friendly instrumentation used in the surgical management of EOS. They divided the type of instrumentation into distraction based (e.g., growing rods) (*Figures 3-5*), growth-guidance (e.g., Luque-Trolley or Shilla), and compression based (e.g., stapling or tethering).

This review will focus on the surgical treatment of EOS using distraction based spinal instrumentation either traditional growing rods (TGRs) or using magnetically controlled growing rods (MCGR).

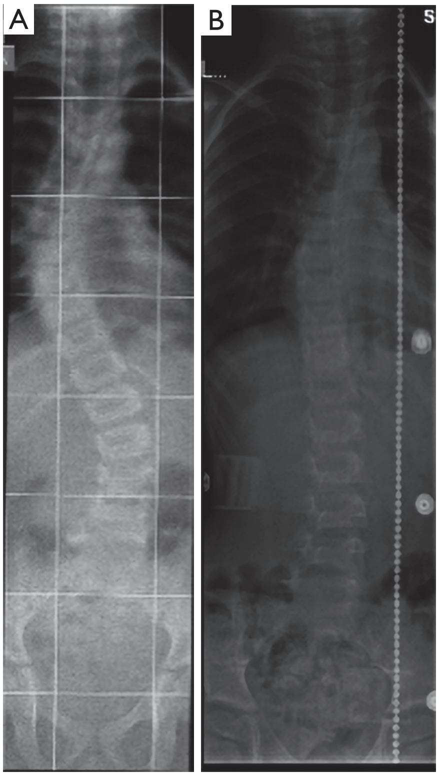

Figure 1 Two-year-old boy with syndromic 50-degree early onset scoliosis (Marfan syndrome). (A) Standing posteroanterior spinal radiographs before casting; (B) 1-year follow-up at the end of casting, radiograph taken in a brace.

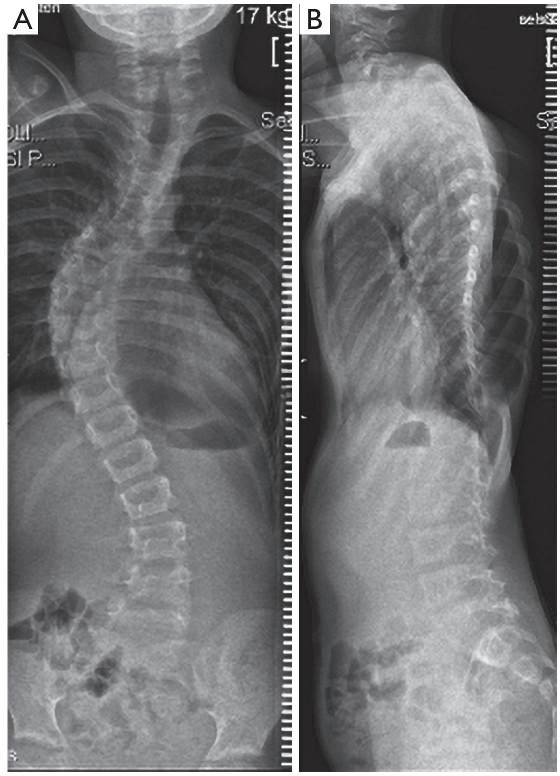

Figure 2 The same boy at the age of 5 years and 8 months before surgery. (A) Standing posteroanterior spinal radiograph; (B) lateral spinal radiograph. Radiographs show 65-degree thoracic scoliosis with hypokyphosis.

Indications for growing rod surgery

EOS can be treated with casting, bracing or surgery (4,7,13,14) (*Figures 1-5*). Serial casting is indicated for progressive infantile scoliosis (scoliosis diagnosed before the age of 3 years) (13) (*Figure 1*), while growth-friendly surgery is indicated when scoliosis is progressing to over 50° and conservative management has failed (4,6,7,14) (*Figures 2-5*). Progressive and non-progressive idiopathic infantile scoliosis are differentiated by using the rib-vertebra angle difference (RVAD) (15). A RVAD of 20° or more is typical of progressive infantile scoliosis. A clear indicator for progressive infantile scoliosis is the apical rib head in phase two. In this stage, the rib head overlaps the corresponding apical vertebral body (15).

Growing rod surgery is associated with high risk of complications (6,16,17) (*Figures 4,5*). Bess *et al.* (16) found a minimum one complication in 81 (58%) out of 140 children treated using a growing rod instrumentation over a minimum 5-year follow-up. Older age and subfascial instrumentation decreased this risk, while every surgery increased the risk of complication by 24%. Postponing growing rod surgery 1 year reduced the risk of complications by 12%. Therefore, Fletcher *et al.* (18) have popularized delaying tactics in the surgical management of EOS. Spinal deformity can be controlled by using cast or brace, and in this manner surgery can be postponed several years and thus reducing risks of repeated surgery (*Figure 1A,B*). Further growth of spine results into larger bony structures, which allow bigger implants and therefore more solid spinal instrumentation. This reduces risks of implant related complications but also the need for repeated procedures. Halo-gravity traction can be used to facilitate larger curves to be amenable for casting under general anaesthesia (18). The upper limit for delaying tactics to be an EOS of 90 degrees or more (19,20). A severe curve above 90 degrees at the time on initial management results into large residual curve and increases significantly the risks of complications (19).

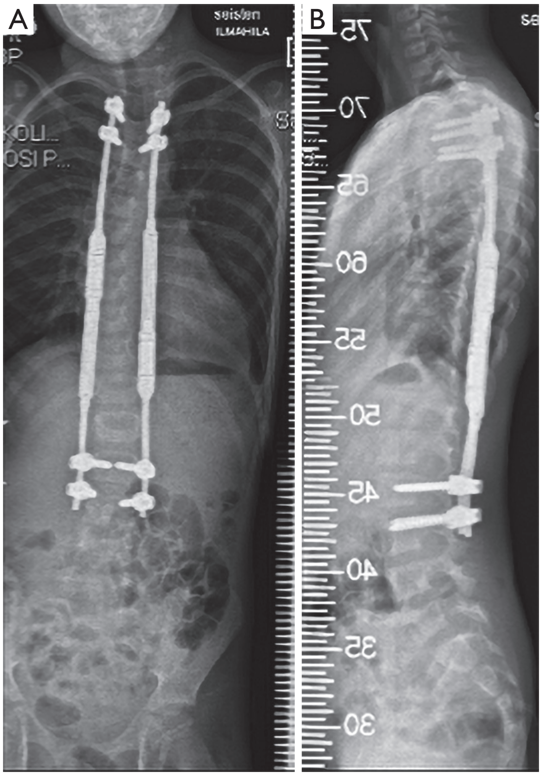

Figure 3 Standing posteroanterior (A) and lateral (B) spinal radiograph after surgery using magnetically controlled growing rods. Fixation points using pedicle screws.

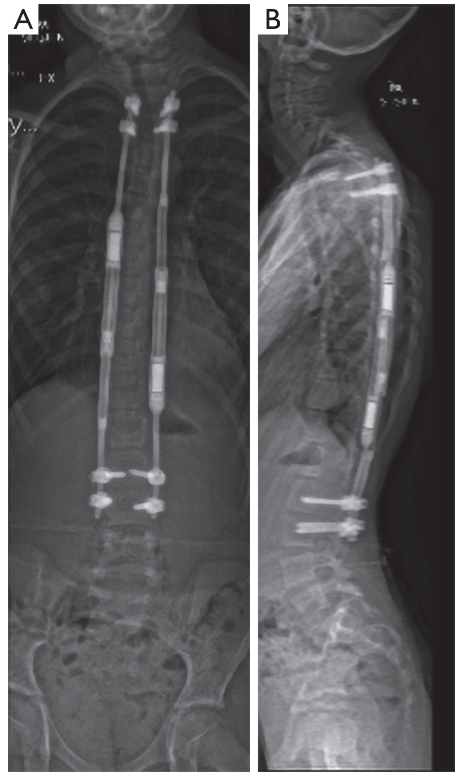

Figure 4 Standing posteroanterior (A) and lateral (B) spinal radiograph 3 years after index surgery at the age of 8 years. Radiographs demonstrates well-aligned and corrected spinal deformity, but the proximal thoracic pedicle screws seem to be pulling out.

Surgical technique and outcomes

Growth-friendly spinal instrumentation options for EOS include growing rods (TGR or MCGR) (4,6,7,21-24), Shilla and other growth guidance systems (25), and a vertical expandable prosthetic titanium rib (VEPTR) (26,27). At skeletal maturity segmental instrumentation and final spinal fusion can be performed (28,29).

Growing rod instrumentation has been the main surgical method to address EOS and several studies have documented follow-up until skeletal maturity with final fusion or observation only after last surgical lengthening (4,6,7,16,17,20,24,28,29). TGR is a growth-friendly surgical management, which requires repeated lengthenings but is associated with a high risk of complications (16,17,20). These include deep surgical site infection, rod fractures and failure of proximal fixation (*Figure 4A,B*). MCGRs are a new distraction-based spinal instrumentation, which allows outpatient construct lengthenings (21-24). This may reduce the risk of deep wound infection (*Figure 3A,B*) (21-24). VEPTR instrumentation is typically reserved for patient with fused ribs (14,26,27).

Both TGRs and MCGRs are used in a sub-muscular, dual-rod fashion (4,7,21-24). Fixation points are exposed using two small separate incisions and the rest of the spine is left untouched (4). Typical construct involves upper thoracic pedicle screws or laminar hooks and pedicle screws in the mid-lumbar spine (*Figure 3A,B*) (7). Rods are tunneled from the upper incision submuscularly to the lower incision. Growing rods provide correction of the spinal deformity using indirect methods: distraction on the concave side and cantilevering on the convexity of the curve resulting into roughly 40% correction of scoliosis (4,6,7). Severe EOS (defined as major curve >90°) remains difficult to correct using growing rods (19,20). Coronal curve correction remains lower (43% in this study) using growing rods than when using high-density pedicle screw instrumentation in children undergoing surgery for adolescent idiopathic scoliosis (>60%). Therefore, a significant residual curve

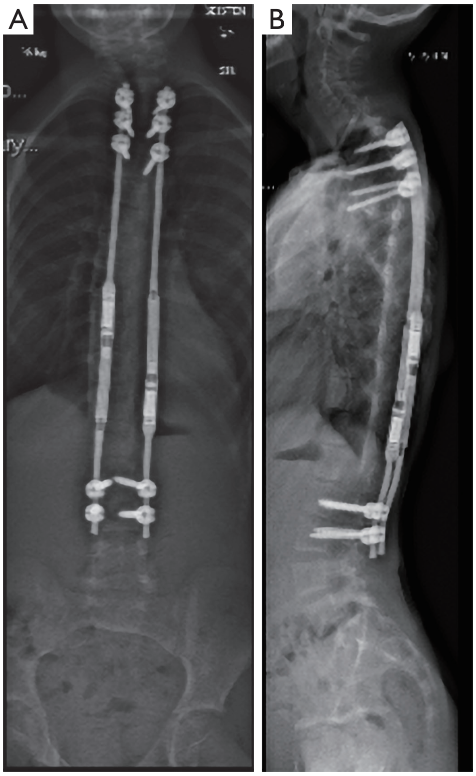

Figure 5 Standing posteroanterior (A) and lateral (B) spinal radiograph at the age of 9 years. Radiographs 6 months after revision surgery, where all fixation points were revised. This boy had a transient intraoperative neuromonitoring change (loss of left sided motor evoked potentials), while revising the proximal anchors. Postoperatively he was neurologically fully intact.

remains at the end of growth-friendly management (4,6,7,30).

In a typical surgical setting a standard MCGR rod (distraction towards caudal direction) is applied on the left side and an MCGR offset rod (distraction towards cranial direction) is applied on the right side (*Figures 3,4*) (21-24,31). This construct allows separate lengthening of the rods and minor adjustment of the coronal balance. By placing two standard MCGR rods may allow both rods lengthening at the same time and thus theoretically providing higher lengthening forces (31).

The initial growing rod surgery provides approximately 50% of the spinal length increase and the lengthenings provide the remaining half in children (4,6,7). In a multicenter investigation, Akbarnia *et al.* (4) followed 23 EOS children (7 with idiopathic, 3 congenital, 13 secondary scoliosis) who were operated using TGRs with a minimum 2-year follow-up. All patients underwent lengthenings every 6 months. The average number of lengthenings per patient was 6.6 and this resulted into T1–S1 growth of 4.6 or 1.2 cm/year. Patients with congenital scoliosis received significantly less spinal length during the index surgery while lengthenings produced similar spinal growth. Distraction of the spine with instrumentation may stimulate growth of the spine, since annual growth of 1.2 cm/year exceeds that of the normal spine. The Growing Spine Study Group (31) has evaluated the spinal height gain over repeated surgical lengthenings (32). A decrease of T1–S1 gain from 10 mm at first lengthening to 6 mm at 7th lengthening occurred, but some gain occurred even after multiple lengthenings, i.e., phenomenon called "law of diminishing returns" (32).

The use of MCGRs is the latest technique allowing non-invasive lengthenings (*Figure 4A,B*) (21-24,31). Bilateral MCGRs are instrumented subfascial to the spine with pedicle screws or hooks to connect the proximal and the distal fixation of the rods. The MCGR contains a magnetically-driven lengthening mechanism. After the index surgery, lengthening can be done without anaesthesia with an external remote controller on an outpatient clinic. It has been suggested that, because there is no need for repeated surgeries, the risk of wound infections would be lower than with TGRs (21-23). MCGRs have been shown to be a safe and effective in children undergoing primary surgery for EOS (21,22). However, patients who have had a TGR and are converted to MCGR seem to achieve less spinal growth (23). In one study, 47% of children operated using MCGR for EOS have required an unplanned re-operation during a minimum 2-year follow-up (21). The most common indications for re-operation were failure of distraction, proximal foundation failure, and rod breakage (*Figure 4A,B*). More frequent distractions (between 1 week and 2 months) were associated with a higher rate of re-operation than distraction frequencies between 3 and 6 months (21).

In severe EOS a preoperative halo traction is a useful additional corrective maneuver, since it has been shown to reduce thoracic kyphosis more effectively than anterior spinal discectomy in EOS, which reduces forces on the spinal instrumentation (33,34). Preoperative halo traction has not been associated with severe complications in severe EOS (19).

Complications associated with growing rods include fixation point failure, rod fracture, autofusion, and increased risk of deep wound infection (16,17). Growing rods for severe EOS have also been associated with a relatively high risk of neurological deficits (5%) (19) The main mechanisms of the neurological deficits have been: (I) correction and distraction of the spine during initial surgery; (II) pedicle screw pull-out during follow-up; and (III) difficulties in placing thoracic pedicle screws during revision surgery (*Figure 5A,B*) (19). To reduce the risk of upper thoracic pedicle screw related complications following recommendations have been suggested: in case of difficulties in obtaining a solid pedicle screw fixation at index surgery, consider using laminar or rib hooks; a minimum of two pairs of pedicle screws should always be used, although this may not fully prevent pull-out related neurologic complications; Revision of fixation points should be regarded as major surgery and the use of intraoperative monitoring is recommended (19).

Comparison of traditional and MCGRs

TGRs are lengthened surgically typically at 6-month interval (6). MCGRs can be lengthened more often, but more frequent distractions than 3 months appear to be associated with increased risk of instrumentation failure (21) and therefore most centers have elected to lengthen these devices at 3-month interval.

Noordeen *et al.* (35) have measured the forces needed to distract TGRs during surgery. Based on stiffening spine and surrounding soft tissues these forces increase during the treatment and after 6th lengthening forces up to 650 Newtons (N) are required to provide the 10 mm lengthening of the instrumentation. MCGR provides a force of 230 N until the safety mechanism begins to stall or clicking (23,36). The maximal force generated by MCGR decreases by 8% when the rod is lengthened up to 40 mm (36). Recently, explanted MCGR have been investigated (37). The duration the rods were *in vivo* was negatively correlated with the force produced on testing. After 38 months' use *in vivo*, explanted MCGR did not produce any force (37). This suggests these rods should be exchanged within 3 years after implantation (*Figure 4A,B*). The forces needed to keep up lengthening of spine may explain, why TGRs seem to provide more spinal growth than MCGR instrumentation does. This difference seems also clinically relevant in revision cases where TGRs should be given a priority (23). TGRs are also indicated in patients who need repeated MR images for medical follow-up, as MCGR prevents high quality MR images of the of spinal cord and perivertebral structures (38). Such follow-up might be needed, e.g., in the follow-up of children with neurofibromatosis.

TGR surgery with repeated surgical lengthenings results into autofusion and stiffening of the spine (39). The additional correction and spinal length obtained at final fusion has therefore remained very limited (28). Jain *et al.* (29) have reported that patients with acceptable spinal deformity and balance at the end of TGR management may just be followed up instead of final fusion. In contrast, patients with severe EOS at the beginning of growth friendly management may benefit from final fusion in terms of better spinal deformity correction and spinal length obtained (20).

Metallosis around the MCGR instrumentation has been reported in revision cases (40). This appears to be the result of micro-movement between the rod and the housing part of the device and consists of metal fragments of Titanium. As this metallosis appears to be clinically significant the MCGR instrumentation needs be removed at the end of growth friendly management and a definitive spinal instrumentation performed at the end of this management. Preliminary findings suggest that instrumented segments experience similar stiffness as with TGR thus limiting further correction and length gain during the final fusion (31).

Conclusions

Growing rods represent the standard and well documented surgical technique to address EOS even in its severe forms. TGRs allow reasonable correction of scoliosis and maintain adequate spinal growth, but are associated with relatively high risk of complications and unplanned re-operations due to repeated surgical lengthenings. MCGRs are currently widely used, but few patients have been followed up to skeletal maturity. Lengthenings on an outpatient basis represent a more advancement in the treatment of EOS, but more information is needed on the risk of metallosis during and other complications during treatment. Halo-gravity traction is a useful adjunct in severe EOS. TGR management results into stiffening and autofusion of the spine and therefore not all patients need final fusion, while patients with MCGRs should be treated by explantation of these rods and segmental spinal instrumentation and fusion at the end of growth friendly management.

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Footnote

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Ethical Statment: The author is accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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