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Effect of Growing Systems on Hyperkyphotic Deformities

Greggi T[1], Maredi E[1]*, Vommaro F[1], Lolli F[1], Martikos K[1], Giacomini S[1], Di Silvestre M[1], Baioni A[1], Scarale A[1], Morigi A[2] and Bacchin MR[2]

[1]*Spinal Deformity Surgery Department, Rizzoli Orthopaedic Institute, Italy*
[2]*Anesthesiology Department, Rizzoli Orthopaedic Institute, Italy*

Abstract

Study Design: Retrospective mini case series, single centre.

Objective: To report the efficacy of growing spine distraction-based implants in the treatment of hyperkyphotic and kyphoscoliotic early-onset deformities during initial surgery and lengthening.

Backround: Growth-sparing implants, such as growing rod and VEPTR-like systems, are distraction-based systems involving repetitive lengthening procedures, which mean that hyperkyphosis may be a relative contraindication in the treatment of early onset deformities. The role of growing implants in the treatment of coronal deformities is now acknowledged, but there are very few studies on the effect of both primary surgery and several lengthening procedures on sagittal balance.

Methods: Twenty paediatric patients affected with kyphoscoliosis and surgically treated with growing systems were retrospectively reviewed. Etiology was heterogeneous; there were 10 males and 10 females, aged 7 yrs on average. The dual growing rod technique was used in 9 cases, VEPTR in 11. Preoperative main thoracic scoliosis averaged 64° (range, 10° to 100°) and thoracic kyphosis 71° (60° to 90°),67° in patients with Growing Rods and 77° in those with VEPTR with a history of EOS (Early Onset Scoliosis). At Follow-up ranging from 6 months to 7 years, 31 lengthening procedures had been performed (1.9 per patient). For the purpose of this study, patients were divided into two groups: Growing-Rod Group (GR-group) and VEPTR-like-Group (VL-group); preoperative and postoperative degrees of scoliosis and kyphosis were measured, as well as final results at follow-up.

Results: A significant decrease in scoliosis and kyphosis was observed during initial surgery, then a significant loss of correction occurred during the FU period, first on coronal and then on sagittal plane, both in GR-group and in VL-group; however, in the VL-group the loss of correction in terms of kyphosis was significantly higher than in the other group. In particular, after initial surgery, in GR-group thoracic kyphosis was corrected from 67° to 44°, whereas in VL-group from 77° to 60°. After the lengthening procedures, a loss of correction occurred: in GR-group, thoracic kyphosis increased from 44° to 50° ($p<0.05$), whereas in VL-group from 58° to 68°. 15 minor complications occurred in 8 patients and revision surgery was performed in 7.

Conclusion: Growing implants can be safely used in the treatment of EOS, even in the presence of hyperkyphosis. Distraction procedures inevitably led to the loss of some correction on sagittal plane which was observed at follow-up and was higher in the VL-group. In any case, the final result was mostly related to the correction of kyphosis achieved during initial surgery and in any case the loss of correction was always lower than the first correction obtained. When the cantilever maneuver is performed during initial surgery, growing rods seem to grant a better sagittal plane restoration compared to VEPTR. The complication rate turned out to be a little higher (30%) than the rate observed after general surgical treatment of EOS, thus confirming an increase in complication rate when hyperkyposis is present; the most frequently encountered complication was proximal failure.

Keywords: Early onset deformities; Growing rod; VEPTR; Kyphosis; Hyperkyphosis

Introduction

Early onset scoliosis (EOS) presents treatment challenges to the surgeon. Indications for surgery include failure of conservative treatment, confirmed progression of the deformity and pulmonary disease.

Nowadays, distraction-based systems are probably the most commonly used surgical management method for EOS and their efficacy is demonstrated by the correction achieved on coronal plane, as well as effective curve maintenance and spinal growth over time. The most commonly used surgical techniques without fusion are growth-sparing procedures, such as rib- (VEPTR) [1] and spine- (GROWING ROD) based devices, as well as the "growth modulation" procedure with staples, the Shilla technique and the surgical tethering of the spine [2]. "Growth sparing" techniques have ensured the best results in young patients.

Several studies have confirmed that hyperkyphosis increases the incidence of implant-related complications, most commonly rod fracture [3], when growing rods are used. Even with VEPTR-like constructs, high kyphosis has proved to be difficult to be managed and the kyphotic effect of the lengthening procedure, ascribable to the implant design [4], has been observed. Above all, hyperkyphosis is associated with a higher incidence of complications, of which the most common is anchor mobilization [5]. Hyperkyphosis therefore poses challenges to the spine surgeon and very few studies have been published on sagittal plane alignment in paediatric patients.

The aim of our review is to describe the effect of growing spine systems, such as Dual Growing Rods and VEPTR-like constructs, on

***Corresponding author:** Elena Maredi, via Pupilli 1 40136 Bologna (BO) Italy, Tel: +39 3486650558; Fax: 0516366188; E-mail: elena.maredi@gmail.com

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sagittal plane alignment in the treatment of early onset hyperkyphotic deformities.

Material and Methods

Twenty paediatric patients affected with kyphoscoliosis and surgically treated with growing systems were reviewed. Aetiology was heterogeneous: idiopathic scoliosis (5 cases), congenital scoliosis (4 cases), scoliosis in arthrogryposis (3 cases), and scoliosis in trisomy 8 (1 case), scoliosis in spondylocostal dysplasia (1 case), scoliosis in Prader-Willi syndrome (1 case), scoliosis in Escobar syndrome (1 case), kyphosis in Morquio disease (1 case) and kyphosis in Pott disease (1 case). Ten males and 10 females aged 7 (range, 4 to 11 years) on average were assessed. All patients except for the idiopathic cases were evaluated from the genetic point of view to establish a clear diagnosis and evaluate comorbidity.

A brain and spinal cord MRI was performed to exclude myeloradicular malformations, such as Arnold-Chiari malformation, syrinx or tethered cord. In the literature, the incidence of syrinx in scoliotic patients varies from 25% to 85%; on the other hand, 15% of the “idiopathic” scoliosis present with myeloradicular deficit [6].

In addition, patients underwent spirometry, *cardiac ultrasonography* to exclude cardiac abnormalities and a*bdominal ultrasound* to exclude abdominal malformations, as well as a neuropsychiatric and neurological (or neurosurgical) evaluation. A CT-scan evaluation was performed only to check for congenital deformities. All patients underwent standard X-ray examination. Dual growing rods and VEPTR-like systems were used in 9 and 11 cases, respectively.

The growing-rod surgical technique includes preparation of short foundation with proximal hooks or screws and distal screws, and two pre-bent submuscular rods; when VEPTR is used, a rib-spine construct is prepared with a proximal anchor placed in one or two ribs and a laminar hook as distal anchor. Only the main curve was treated.

The lengthening procedure for both techniques was performed every 6-8 months in the operating room [7]. All surgical procedures, both initial surgery and lengthenings, were carried out with SSEP and MEP neuromonitoring.

Preoperative main thoracic scoliosis was 64° Cobb (range, 10° to 100°) and thoracic kyphosis 71° Cobb (range, 60° to 90°). Patients were divided into two groups: Growing-Rod Group (GR-group) and VEPTR-like-Group (VL-group); preoperative and postoperative range of scoliosis and kyphosis, as well as final result at follow-up, was assessed. In particular, kyphosis was 67° in patients instrumented with growing rods and 77° in VEPTR patients affected with EOS regardless of aetiology. Follow-up (FU) ranged from 6 months to 7 years and a total of 31 lengthening procedures (1.9 per patient) were performed. Results are expressed as the mean ± standard deviation for parametric values. The *t* test for two dependent means was used to determine whether there was a significant difference between different follow-up intervals in Cobb's angle for kyphosis and scoliosis in each group. The comparison between groups was descriptive because given the rarity of the pathology it was not possible to reach a sufficient number of patient to perform a statistical evaluation.

Results

A significant decrease in scoliosis and kyphosis was observed after initial surgery, followed by a significant loss of correction during the FU period, on coronal and on sagittal plane and in both groups (GR- and VL-groups), although the loss of correction in terms of kyphosis was significantly higher in the VL-group than in the other.

After initial surgery, scoliosis improved from a mean value of 64° to 42° Cobb and correction was maintained till final follow-up when it averaged 43.4°: the correction rate achieved with growing systems was 34,4% on coronal plane.

Regarding thoracic kyphosis, it was corrected from a mean value of 71° (range, 60° to 90°) to 52° (21° to 80°) ($p<0.05$). In some cases treated with growing rods, it was corrected from 67° to 44° ($p<0.05$), with a correction rate of 34,3%, whereas it ranged from 77° to 58° ($p<0.05$) with a mean correction rate of 24,7% in those cases instrumented with VEPTR.

It is important to note that after 31 lengthening procedures (1.9 per patient), a loss of correction was seen on sagittal plane at final follow-up: thoracic kyphosis increased from a mean value of 52° to 59° ($p<0.05$). In the GR-group the loss of correction ranged from 44° to 50° ($p<0.05$), which means a loss of 12% of sagittal correction, whereas in the VL-group it ranged from 58° to 68° ($p<0.05$), i.e. a loss of correction of 15% on sagittal plane. Results are shown in Tables 1 and 2.

Fifteen minor complications were encountered in 7 patients (38.8% of the cases) and revision surgery was required in 7 cases (22% of all surgeries): proximal junctional failure with screw loosening or PJK was observed in 3 children with a severe hyperkyphosis (GR-group) and in 3 children with rib fracture (VEPTR group); all of them were youngest children of the seriesand presented with syndromic features, all treated during the lengthening procedure; the remaining patient had the most severe angular hyperkyphosis of the series and experienced growing rod fracture; complications, such as protrusion of instrumentation through the skin or pain, were seen in all of the 7 children. No major complication or infection occurred in this series of patients.

Discussion

Early onset scoliosis (EOS) poses treatment challenges to the surgeon. Children with congenital deformities often present with congenital malformations (renal, cardiac and gastrointestinal anomalies) that can compromise their quality of life. Patients with these diseases have been found to have a higher mortality rate than patients with adolescent scoliosis (AIS) [8], regardless of curve magnitude (Cobb method). Therefore, early treatment is recommended and mandatory, not only in “early onset [9-11], but also in “late onset“[12] deformities.

Historically, treatment options for early deformities have included initial management with casting and bracing.

Several studies have demonstrated that, generally speaking, this type of aggressive deformities does not respond to conservative treatment, which remains the first approach for every scoliosis [13].

Distraction-based fusionless instrumentation systems have been used since 1960s: Harrington [7] first described a fusionless procedure with a single rod; then, Moe et al. [14] modified the technique using subcutaneous rod passage and reducing subperiosteal dissection; in Italy, Faldini et al. presented the “end fusion technique”, and finally,

Mean	PREOP	POSTOP	F.U.	MeanCorrection
Scoliosis	64°	42°	43,4°(P>0.05)	34,4°(P<0.05)
Kyphosis	71°(60-90)	52(21-80)	59°(P<0.05)	20%

Table 1: Results of scoliosis and kyphosis.

KYPHOSIS	PREOP	POSTOP	F.U.
G.R.	67°	44°	50° (P<0.05)
VEPTR	77°	58°	68° (P<0.05)

Table 2: Results of G.R. and VEPTR.

Akbarnia described surgical indications for growing rod treatment using two vertebral anchors and two connecting rods through a subfascial tunnel [15]. Ever since this technique was first used, the complication rate turned out to be high and in the literature it is reported to range from 29% to 48% [9].

The VEPTR (Vertical Expandable Prosthetic Titanium Rib) was designed to treat respiratory insufficiency in young children affected with congenital scoliosis associated with rib fusion; afterwards, surgeons expanded indications for VEPTR to include early onset scoliosis, when the patient's young age could not grant solid vertebral anchoring (rib ossification before the vertebra) [16,17].

Nowadays, distraction-based systems have become the most common surgical procedures and their efficacy is confirmed: they ensure correction on coronal plane while allowing continued spinal growth. In case of severe deformities, spinal fusion is limited to very short segments because of the acknowledged risk of crankshaft phenomenon and inhibition effect on spinal and thoracic growth [12]. The most commonly used surgical techniques without fusion are growth-sparing procedures, such as rib- (VEPTR) [3] and spine- (GROWING ROD) based devices, as well as the "growth modulation" procedure with staples, the Shilla technique (McCarthy et al) and the surgical tethering of the spine [4]. "Growth sparing" techniques have ensured the best results in young patients.

Several studies have confirmed that hyperkyphosis increases the incidence of implant-related complications, most commonly rod fracture [5], when growing rods are used.

It is important to control deformity correction to avoid the cost of short fusion (adding-on) and prevent PJK (rod pre-contouring, UIV> T2) or the use of hooks as proximal anchors. Even with VEPTR-like constructs, high kyphosis has proved to be difficult to be managed and the kyphotic effect of the lengthening procedure, ascribable to the implant design [6], has been observed. Above all, hyperkyphosis has turned out to be associated with a higher incidence of complications, among which the most common is anchor mobilization [7]. The use of the longest possible instrumentation and strong proximal fixation are recommended to ensure control of deformity correction.

According to Lenke Classification of AIS, kyphosis is defined as normal, when it measures between 10° and 40°, whereas hyperkyphosis exceeds 40° [18]. Hyperkyphosis poses a challenge to the spine surgeon: theoretically speaking, sagittal deformities place more stress on the rod and foundation, above all in the proximal area, which means a higher potential for failure than in non-kyphotic deformities [5,19]. Few studies have been published on sagittal plane alignment in paediatric patients. Although kyphoscoliosis is considered a contraindication to treatment with growing spinal implants, it is widely recognized that children with severe progressive deformities need early treatment to help lung development and improve quality of life [20]. In addition, very few studies are available in the literature on growing spinal implants and kyphosis.

According to the data available, kyphosis does not seem to affect clinical outcome and complication rate, when growing rods are used [21].

However, a more recent review has reported an increased incidence of "implant-related" complications in case of thoracic kyphosis > 40° and rod fracture [5] was the most frequently encountered one.

Therefore, it is important to control deformity correction to avoid the cost of short fusion (adding-on) and prevent PJK (rod pre-contouring, UIV> T2) or the use of hooks as proximal anchors, which seem to grant a better follow-up in terms of proximal mobilization.

As far as VEPTR-like constructs are concerned, the greater difficulties in controlling 'high' kyphosis are clearly stated in the literature and are related to the difficulties in pre-bending the rod.

The kyphotic effect of lengthening procedures, due to the kyphotic rod contour [6], has also been clearly described; subsequently, the longest possible instrumentation (>2nd rib), a strong proximal fixation (4 ribs) and the use of the new VEPTR device for better rod contouring in high kyphosis are all recommended to ensure control of deformity correction.

Referring to the literature, our attention was focused on two issues: the high rate of complications and the loss of correction at follow-up, as shown in Figures 1-4. The complication rate we have registered (38.8%) is obviously a high rate but it is comparable to that reported in the literature (58%) [9]; moreover, all of the complications encountered were minor and were all treated during lengthenings.

In addition, our series included more complications of syndromic type experienced by the youngest children who were affected with the most severe hyperkyphosis.

In our experience, the best ways to reduce the number of complications seem to be the following:

- Delay surgery as much as possible to develop a better ossification, the patient's age playing a key role;

Figure 1: Severe kyphoscoliosis in arthrogryposis, male 4 yrs. Preoperative clinical picture (Case 1).

Figure 2: 3.5-year FU. Postoperative clinical picture after 3 lengthenings and two implants (Case 1).

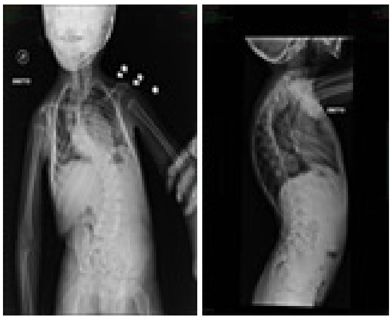

Figure 3: Severe kyphoscoliosis in arthrogryposis, male 3 yrs, Preoperative X-rays (Case 1).

Figure 4: Radiographic pictures at a 3.5-year FU. ostoperative radiographic picture after 3 lengthenings and two VEPTR implants: proximal hooks in fourth and fifth ribs; asymptomatic PJK. (Case 1).

- Precontour the rods (VEPTR2-group experienced a minor rate of anchor breakage and Growing Rod-group had the best precontoured rods);
- Place the proximal anchor as high as possible to avoid risk of PJK and cantilever stress.

With regards to the loss of correction, the results obtained were comparable to those available in the literature [5]:

- A higher loss of correction was found in VEPTR group and it is likely to be ascribable to an initial proximal anchor placement below the bottom of the kyphotic curve
- A minor correction on sagittal plane at initial surgery
- A stronger kyphotic effect of the lengthening procedures ascribable to the rod contour.

The better correction achieved on sagittal plane by growing rod instrumentation grants better results even at follow-up, because the loss of correction is really low (less than 10° Cobb, which is almost inappreciable in a complex spinal deformity) (Figures 5-9).

Conclusion

In conclusion, our results have proved that growing spinal implants can be safely used in the treatment of kyphotic deformities (correction rate of 34.3% in growing rod group and 24.7% in VEPTR group after initial surgery). The complication rate is acceptable if compared to the data reported in the literature.

On account of the number of distraction procedures involved, a loss of correction on sagittal plane is commonly observed at follow-up (6° GR-group and 10° VEPTR-group, on average) but the final result is mostly related to the kyphosis correction obtained during initial surgery; in any case, the loss of correction is always lower than the first correction achieved.

Figure 5: 6-yr-old female affected with kyphosis in Pott disease. Preoperative clinical picture (Case 2).

Figure 6: 6-yr-old female affected with kyphosis in Pott disease Preoperative X-rays (Case 2).

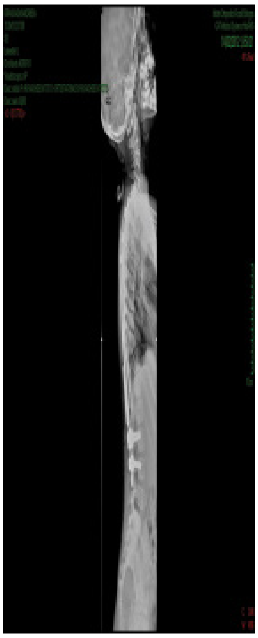

Figure 7: Postoperative x-rays after initial surgery (Case 2).

Figure 8: Postoperative clinical picture afters 3 lengthenings (Case 2).

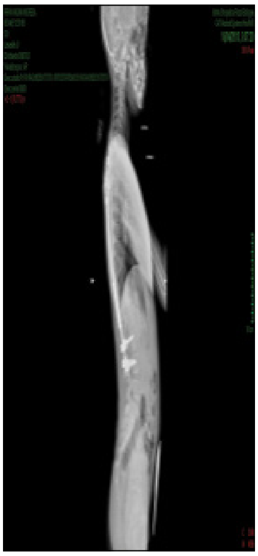

Figure 9: Postoperative radiographic picture after 3 lengthenings (Case 2).

Therefore, the growing rods seem to grant a better sagittal plane restoration compared to VEPTR, not only in the postoperative but also at follow-up, when the cantilever maneuver is performed during initial surgery.

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in 43 patients (13%)… Risk factors: single rods, history of previous fracture, small diameter rods, stainless steel rods, proximity to connectors, ambulatory patients, syndromic diagnosis… No effect: size of scoliosis and kyphosis, n° of lenghthenings, anchor type. International SRS conference.