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Shilla growth guidance compared with magnetically controlled growing rods in the treatment of neuromuscular and syndromic early-onset scoliosis

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Introduction

Early-onset scoliosis (EOS) is a challenging deformity of a paediatric spine occurring before the age of 10[1,2] that may lead to poor respiratory and cardiac development, severe progressing deformity, and overall poor outcomes if left untreated[3]. The main goal of the treatment of EOS is to allow growth of spine to ensure the sufficient thoracic development while correcting the deformity. Thoracic height of at least 18 cm has been reported as satisfactory for pulmonary function[4]. Surgical treatment is indicated for progressive and severe deformities.

Surgical treatment of EOS with ‘traditional’ growing rods (TGR) is a distraction-based method requiring surgical lengthening procedures every six months and is linked with high complication

rate, which increases with repeated procedures[5,6,7]. Repetitive surgical lengthenings have a negative effect on patient reported quality of life[8,9,10]. Magnetically controlled growing rods (MCGR) allow non-surgical lengthenings and is reported to have lower rate of complications[11,12,13]. Although lacking the planned surgical procedures, MCGR requires regular hospital visits. MCGR also requires sufficient patient size and straight spinal area to mount the actuator.

The pedicle screw trolley instrumentation (Shilla) presents a distraction free instrumentation by passive growth guidance using fixed apical segment and sliding screw heads to provide the deformity correction[14]. Shilla instrumentation requires minimal observation without planned surgery during the treatment. Shilla and growing rods have been reported with comparable results[15,16,17].

In this study, we evaluated the surgical and health-related quality of life (HRQoL) outcomes of consecutive patients with syndromic or neuromuscular EOS treated with either Shilla or MCGR instrumentation.

We hypothesized that patients with syndromic or neuromuscular EOS might benefit from the reduced implant size in Shilla instrumentation and from the reduced need of outpatient visits in terms of HRQoL.

Materials and methods

Institutional review board approval was obtained from both of the participating study centers.

Patients

The current study was conducted as a dual-center retrospective series of consecutive syndromic or neuromuscular EOS patients under the age of 10 years treated with either Shilla (n = 13) or MCGR

(n = 18) instrumentation between 2010 and 2018 (Table I). **The indication of operative treatment was a major curve of minimum 45 degrees irrespective of the curve type in both study groups. All of our patients had a minimum 15 kg as their body weight.** Of the patients treated with Shilla, eleven had neuromuscular EOS (6 spinal muscle atrophy type II (SMA II), one each myelomeningocele, multiple developmental disabilities, a progressive central nervous disease, and a developmental disability), and two syndromic EOS (one with Edwards' syndrome, one with arthrogryposis). Of the patients treated with MCGR, 12 had neuromuscular EOS (five had unspecified developmental disability, one had SMA II, and one had congenital muscular dystrophy) and six had syndromic EOS (two neurofibromatosis I, two Marfan syndromes, one Coffyn-Lowry syndrome, one Prader-Will syndrome).

Surgical techniques

The Shilla procedures were performed through a midline incision by a senior pediatric orthopaedic surgeon. Apical segments were exposed subperiosteally to place six monoaxial screws on the apex curvature and to perform the apical fusion[18]. Locking screw caps were used in the apical segment to hold the rod in position. Polyaxial screws were used as growth guidance screws, four of which were placed both cephalad and caudal from the apex curvature using the Wiltse paraspinal approach. Shilla caps allow free longitudinal movement of the rods enabling growth. We used 4.5 mm (n = 7) and 5.5 mm (n = 6) Stainless Steel rods.

Magnetically controlled growing rods were inserted through a proximal thoracic and distal midlumbar incisions by the senior and another experienced pediatric orthopaedic surgeon. Patients were treated with dual rod constructs (normal and offset) with 90 mm actuator[19]. Four to six proximal anchors and four distal anchors were inserted, and fusions were performed on both

foundations. **MCGRs were elongated on a three-month interval during the distraction period. The planned lengthening was 3 mm on both sides. The elongation was controlled using ultrasound. In case of failed lengthening, the procedure was repeated once on the outpatient visit.**

Data collection

Data collection was retrospective. Times points analyzed were following: 1) pre-operative visit on the previous day from the initial surgery, 2) after index surgery, 3) pre-definitive visit before final fusion, and 4) after final fusion. We collected the following clinical data: age, sex, specific diagnosis, duration of follow-up, number of surgical procedures, and complications. Complications were categorized wound related, implant-related, alignment related, neurological, or other.

Imaging data

Posteroanterior and lateral **upright full spinal radiographs** were evaluated before and after index surgery, before the definitive visit, and after the final fusion by an independent observer (Figures III – V). Coronal major curve was measured with the Cobb technique[20]. Thoracic height was measured from the upper end plate of T1 to the lower end plate of T12. Spinal height was measured from the upper end plate of T1 to the upper end plate of S1. Coronal and sagittal alignment were measured. Lumbar lordosis and thoracic kyphosis were measured with the Cobb technique. We observed for any evidence of anchor loosening or pull-out, or rod failure in the radiographs.

EOSQ-24 questionnaire

EOSQ-24[21] is a disease-specific HRQoL assessment developed for children with EOS and their families. Patients and their parents were asked to complete the EOSQ-24 at the final follow-up visit.

Statistical analyses

Statistical comparisons were performed using chi-square tests for categorial parameters and t test for continuous variables. Mann-Whitney U test was used for comparison of EOSQ-24 domains. Significance was set up at $p<0.05$.

Results

The mean age of patients at index surgery was 6.0 years (range, 2.7 – 9.1) in the Shilla and 6.8 years (range, 2.1 – 10) in the MCGR groups (p=0.164, Table II). Patients treated with Shilla had significantly less surgical procedures (mean 1.4, range 1 – 2) as compared with the MCGR group (mean 2.6, range 1 – 9, p=0.034). In the MCGR group, there was a mean of 7.4 (range, 2 – 15) lengthenings performed. The mean duration of follow-up was 4.0 (2.0 – 9.0) in the Shilla and 3.2 (1.3 – 6.8) in the MCGR group (p=0.093). The mean operative times were 2.6 (2.0 – 4.5) hours in the Shilla and 2.0 (1.4 – 2.8) hours in the MCGR group (p=0.045).

Radiographic Outcomes

The mean preoperative Cobb angle was 64° (range, 39° – 108°) in the Shilla and 58° (range, 45° – 85°) in the MCGR group (p=0.151, Figure I, Table III). This was corrected at the initial surgery to 22° (range, 6.7° – 52°) and 28° (range, 9.8° – 46°) respectively (p=0.095). At the last pre-definitive visit, the mean major curve was 31° (range, 9.4° – 54°) and 30° (range, 16° – 53°) respectively

(p=0.392). This represented a mean correction of major curve of 45% (range, -40° – 90°) in the Shilla and 48% (range, 19° – 76°) in the MCGR group (p=0.383).

Preoperative thoracic height was 165 mm (range, 126 – 226) in the Shilla and 147 mm (range, 91.7 – 180) in the MCGR group (Figure II, p=0.024). This increased to 186 mm (range, 152 – 235) and 166 mm (range, 138 – 190) after the index surgery (p=0.003), and to 207 mm (range, 167 – 256) and 200 mm (range, 165 – 248) at pre-definitive visit (p=0.202). This resulted into an increase of 21 mm (range, 6.0 – 56) and 19 mm (range, 2.0 – 48) after the index surgery (p=0.362), and 22 mm (range, 1.0 – 55) and 34 mm (range, 3.5 – 73) during the distraction period (p=0.042), respectively. The mean annual thoracic height improvement during the distraction period was 6.2 mm (range, 0.1 – 11) and 11 mm (range, 1.9 – 27, p=0.019). Eleven (85%) children in the Shilla group and 17 (94%) in the MCGR group achieved T1-T12 length of ≥18 cm at final follow-up (p=0.202).

Preoperative spinal height was 272 mm (range, 206 – 366) in the Shilla and 245 mm (range, 183 – 291) in the MCGR group (p=0.022). This increased to 306 mm (range, 251 – 372) and 276 mm (range, 236 – 319) after index surgery (p=0.004), and to 332 mm (range, 290 – 406) and 323 mm (range, 282 – 388) at final follow-up (p=0.194). The spinal height increased by a mean of 34 mm (range, 5.4 – 86) in the Shilla and 33 (range, 8.6 – 67) mm in the MCGR group after the index procedure (p=0.432), and 26 mm (range, 5.8 – 63) and 48 mm (range, 3.8 – 83), respectively, during the distraction period (p=0.006). The annual spinal height improvement during the distraction period was 7.2 mm (range, 1.1 – 16 mm) in the Shilla and 15 mm (range, 2.3 – 34 mm) in the MCGR group (p=0.004).

The mean preoperative thoracic kyphosis was 45° (range, 6.8° – 80°) in the Shilla and 39° (range, 7.4° – 67°) in the MCGR group (p=0.249). This decreased to 28° (range, 12° – 53°) and 25° (range, 3.8° – 45°, p=0.226) after the index surgery, but there was a slight further increase to 31° (range, 13° – 54°) in the Shilla and slight decrease to 22° (range, 3.0° – 38°, p=0.030) in the MCGR at final follow-up.

Complications

Five (38%) patients in the Shilla and six (33%) in the MCGR group had at least one complication during the follow-up (p=0.768, OR 0.8, 95% CI 0.18 – 3.5, Table IV). The total number of complications were five in the Shilla and ten in the MCGR group (p=0.347).

The patients with complications in the Shilla group were as following (n = 1 each): 1) rod perforated trough skin leading to infection and reinstrumentation, 2) anchor failure leading to worsening of balance and requiring revision surgery, 3) broken and detached rod requiring revision surgery, 4) detached rod requiring revision surgery, and 5) post-operative pneumonia. The patients with complications in the MCGR group were as following (n = 1 each): 1) broken distractor mechanism leading to reinstrumentation with TGR, 2) worsened balance and anchor close to spinal cord requiring revision, 3) anchor failure, temporal change in the intra-operative monitoring during revision surgery, 4) reversible changes in the motor evoked potentials during the index operation, 5) anchor and rib connector failure, rod detachment, rod stuck due to metallosis requiring revision, and 6) post-operative pneumonia.

Final Fusion

Four patients (31%) in the Shilla and five (28%) in the MCGR group underwent final fusion (p=0.856). The treatment period after the initial surgery and before the final fusion was 4.5 years in the Shilla and 3.7 years in the MCGR patients (p=0.244). The mean major curve in patients who underwent final fusion was 12° (range, 0° – 21°) in the Shilla and 16° (range, 5.1° – 32°) in the MCGR group (p=0.252), the mean thoracic height was 254 mm and 220 mm (p=0.107), and the mean spinal height was 386 mm and 349 mm (p=0.117) respectively. Metallosis at the time of final

fusion was more pronounced in children undergoing final fusion after MCGR as compared with the children treated using the Shilla instrumentation

Health-Related Quality of Life

EOSQ-24 questionnaires were available from nine (69%) patients treated with Shilla and 15 (83%) with MCGR. There were no statistically significant differences between the study groups (Table V). At the end of follow-up, the highest score of the domains was for pulmonary function in both Shilla group (mean 96, range 88 – 100) and in the MCGR group (mean 86, range 50 – 100), whereas the lowest score in the both groups was for daily living, mean 23 (range, 0 – 63) in the Shilla and mean 48 (range, 0 – 100) in the MCGR group.

Discussion

In this study, we presented a consecutive series of patients with syndromic or neuromuscular early onset scoliosis treated with Shilla instrumentation or magnetically controlled growing rods. The current study is the first comparison between the Shilla growth guidance technique and MCGR on EOS.

Growing rods have become the standard treatment for EOS[5], as repetitive surgical lengthenings expose patients to anesthetics and deep surgical site infections[6] . Continuous distraction with growing rods presents a risk for unintended autofusion, which may lead to limited correction and spinal growth[22]. MCGR addresses some of the challenges of TGR by lacking the repetitive surgical procedures and anesthetics but requires regular outpatient lengthening visits. MCGR has been reported with decreased of complication rate when compared to TGR instrumentation, but do not totally avoid the challenges previously reported[23].

Shilla instrumentation enables natural vertebral growth and deformity correction without planned surgeries[24,25]. Shilla instrumentation provides control of the apex of the deformity as compared with growing rods (traditional or MCGR). **Our practice has been to use Shilla growth guidance mainly in children with syndromic or neuromuscular early onset scoliosis.**

The main goals of treatment of EOS are preventing the progression of scoliosis and improving the pulmonary function. Thoracic height of at least 18 cm is needed in the treatment of early-onset scoliosis for satisfactory pulmonary function[4]. Previous studies have presented satisfactory deformity correction and thoracic height improvement using Shilla[15,16,17,25,26,27,28]. TGR has been with reported slightly better deformity correction and spinal height when compared to Shilla instrumentation[16]. McCarthy and McCullough reported satisfactory main curve correction and thoracic height outcomes with mean T1-T12 height being 23 cm in their five-year follow-up in patients treated with Shilla, but many of these patients had an idiopathic type of EOS[25]. Another study reported satisfactory thoracic height increase in both Shilla and growing rod groups[17]. In our study there was no significant difference in the major curve correction. Patients treated with MCGR presented with better thoracic and spinal growth during the distraction period, but the additional value of this statistically significant difference remains unclear in this patient group with co-morbidities. At the end of follow-up, 85% patients treated with Shilla and 89% patients treated with MCGR reached the minimum satisfactory requirement of thoracic height.

Shilla instrumentation has been reported to reduce the required surgical procedures when compared to growing rods[15,16,17,25]. One study found no difference in the complication rate between patients treated with Shilla and traditional growing rods[17]. A study with five-year follow-up reported complication rate of 73% in the Shilla instrumentation, including wound infection, progression of the deformity, and implant related complications[25]. The most typical implant related complication reported was rod fracture, following with rod prominence and screw pull-out. In our study, two patients grew off the rods, one had screw pull-out, one had rod perforating trough skin

211 leading to infection after pull-out, and one patient with 5.5 mm rod had a rod fracture. Metallosis
212 has been observed in both caprine models and patients without clinical effects[14,25]. In our study,
213 metallosis was more pronounced in children undergoing final fusion after MCGR as compared with
214 the Shilla group.

215 A recent study reported primary curve migration in significant number of the patients
216 treated with Shilla and incidence of compensatory curves in the longest follow-up of Shilla patients
217 to date[27]. There were no significant compensatory curves in our cohort of patients.

218 To our knowledge, there are no prior studies on the Shilla instrumentation including
219 HRQoL assessments. Difficult etiology, repeated surgical procedures, and regular outpatient
220 clinical lengthening visits have a negative effect on the patient reported quality of life[29,30,31].
221 Children with neuromuscular EOS have been reported with lower quality of life scores than
222 idiopathic children[30]. **Some of the patients had developmental disabilities due to neurologic**
223 **disorders which may prevent the self-assessment of the health-related quality of life. However,**
224 **EOSQ-24 addresses also the caregiver burden, which may improve the quality of life**
225 **information in patients with significant developmental disabilities[32].** There were no significant
226 differences in the domains EOSQ-24, which may be explained by the limited number of patients
227 with EOSQ-24 questionnaires. Based on the current study, children in the Shilla group with
228 syndromic or neuromuscular background avoided multiple outpatient clinic visits. Reduced
229 outpatient clinical visits may be beneficial for the families as children with co-morbidities often
230 require multiple additional visits to other medical specialties, although further research is needed on
231 the subject.

232

233 **Limitations**

Our study was limited for the retrospective design, the lack of baseline EOSQ-24 assessments, and the lack of pulmonary function data. Despite the dual-center approach, our sample size was relatively restricted. All patients with Shilla instrumentation were operated by the senior author. The clinical characteristics and the follow-up time in both of the groups were similar. **Children with syndromic or neuromuscular early-onset scoliosis are a heterogeneous group as some of the neurologic conditions may present as flaccid or spastic subtypes.** The lack of baseline EOSQ-24 assessments prevented us from comparing the change of patient experienced quality of life during the treatment. The restricted sample size of EOSQ-24 answers may have resulted into limited power for evaluating the differences between the groups. **The quality of life assessment may have been limited due to developmental disabilities.** Two children within the Shilla group passed away during follow-up for pneumonia which further limited the number of questionnaires. Twenty-three percent in the Shilla and 28% in the MCGR group underwent final fusion, complicating the evaluation of the definitive outcomes.

Conclusion

According to our findings, Shilla provided similar outcomes in deformity correction in children with syndromic or neuromuscular EOS with similar rate of complications and significantly less surgical procedures when compared to MCGR. MCGR provided slightly better spinal growth during the distraction period. There were no significant differences in the EOSQ24 domains. Shilla instrumentation seems to be useful in the treatment of syndromic or neuromuscular EOS. These benefits are more pronounced in skinny children not fitting the large size of the actuator of MCGR, in children with low-demand (i.e. severe neuromuscular involvement) or in families living at remote locations.