



Use of Growth Rod Systems for Management of Early Onset Scoliosis in Cerebral Palsy: A Systematic Review

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ABSTRACT

AIM: To review the literature for the role and outcome of growing rod surgeries in patients with cerebral palsy associated neuromuscular scoliosis.

MATERIAL and METHODS: A systematic search was conducted according to Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines. Electronic literature search was conducted of PubMed and Embase databases. Patient demographics, type of growing rod used, lengthening and complications were analyzed from the included studies.

RESULTS: A total of 11 studies with poor overall study quality (Level of evidence IV, V) were included in the study. A total of 181 patients with mean age 6.8 ± 1.3 (5-13 range) years at index surgery and mean follow-up of 3.02 ± 1.3 (2-5.8 range) years were included in the study, with a female preponderance. The most common curve and instrumentation was thoraco-lumbar and conventional dual growing rods respectively. All studies showed improvement in Cobb's angle and pelvic obliquity. There was better improvement in pelvic obliquity if pelvis was included in instrumentation. Wound related complications (34.6%) were most commonly noted.

CONCLUSION: Overall growing rod construct has shown questionable outcomes in cerebral palsy patients with scoliosis in terms of the complication rate observed although allowing growth of the spinal column with regular lengthenings. Magnetic controlled growth rods hold a bright promise for the future considering its ability to maintain correction as well as the lower rate of complications. The benefits and risk of immediate fusion with respect to growth sparing surgeries should be considered before the decision.

KEYWORDS: Cerebral palsy, Early onset scoliosis, Growth rods, Guided growth, Growing rod

INTRODUCTION

Cerebral palsy (CP) is frequently associated with postural deformities like scoliosis, pelvic obliquity and windswept hip. Scoliosis in these patients can be managed by either non operative measures such as supportive bracing, optimization of seating position, Botulinum toxin injection, intrathecal Baclofen or surgical management. Although in adolescents with progressive curves or curves $>50^\circ$, surgical management forms the mainstay of treatment but there is no

definite consensus among surgeons for the optimum mode of treatment in growing skeleton. In individuals in advanced age, definitive spinal fusion can be done without much concern. Problems arise in individuals with immature skeleton, in which there is a need to take care of patient's growth potential, thoracic cavity volume, pelvic obliquity and progressive curve. All of these factors have a bearing on the patient's quality of life. Growth rods which have been used in variety of diagnoses for early onset scoliosis, presents an opportunity to address all these issues simultaneously.

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Various systems under broad category of “growing rods” are currently available. Growth guiding techniques are divided into distraction based such as traditional growth rods (TGR), Vertical expandable prosthetic titanium rib implant (VEPTR), tension based like staples or based on the principle of guided growth like Luque trolley and Shilla system (14,29,30). Recent advances have used magnetically controlled growth rod (MGR) systems such as MAGEC (MAGnetic Expansion Control; Nuvasive, San Diego, USA) which employ external magnetic distraction of implanted spinal rods(30). Use of growing rods improves Cobb’s angle, achieves spinal growth while maintaining the correction, improves space available to lung in patients with thoracic curves and can even help correct pelvic obliquity with constructs extending up to the pelvis (1,2,18). There are currently no cumulative studies on the use of growing rods in scoliosis in patients with CP and the current study aims to evaluate and analyze the literature regarding indications and outcomes of growing rod constructs in CP patients with scoliosis.

■ MATERIAL and METHODS

Search Strategy

A literature search using PubMed and Embase was carried out in May 2022 according to the Preferred Reporting Items for Systematic Reviews and Meta-analysis (PRISMA) (Figure 1). The study protocol was registered on PROSPERO (CRD42022346486). Search terms used were ‘cerebral palsy’ and ‘early onset scoliosis’ for the study population and ‘growth rod’, ‘growing rod’, ‘magnetically controlled growth

rod’, and ‘guided growth’ for the interventions. The research was replicated using the MeSH terms. The aim was to keep the search strategy fairly general to include a large number of studies in the initial search. The search was not limited by the year of publication or the level of evidence. Similar articles were searched and all bibliographies were checked to look for any other eligible studies.

Inclusion and Exclusion Criteria

The inclusion criteria for final selection were; 1) Patients with cerebral palsy, 2) growth sparing surgeries using growing rods and, 3) written in English language. Systematic reviews, meta-analysis, reviews and conference presentations were excluded. Two authors initially reviewed all the extracted studies based on the titles and abstracts. If it did not provide enough information to decide on inclusion or exclusion, then full texts were obtained and reviewed by the two researchers. In case no consensus was attained by the two authors regarding the inclusion of the study, a third senior author decided about the inclusion.

Data Extraction and Analysis

Information extracted from the selected studies was tabulated in an MS Excel spreadsheet. Demographic details such as patient age at index surgery, sex, level of scoliotic curve, Cobb’s angle was recorded. Treatment related aspects such as the method of growing rod (single rod, dual rod, VEPTR, etc.), number and duration of lengthening, final fusion done or not, were tabulated. The follow-up data of the operated patient in terms of the duration of follow-up, amount of lengthening

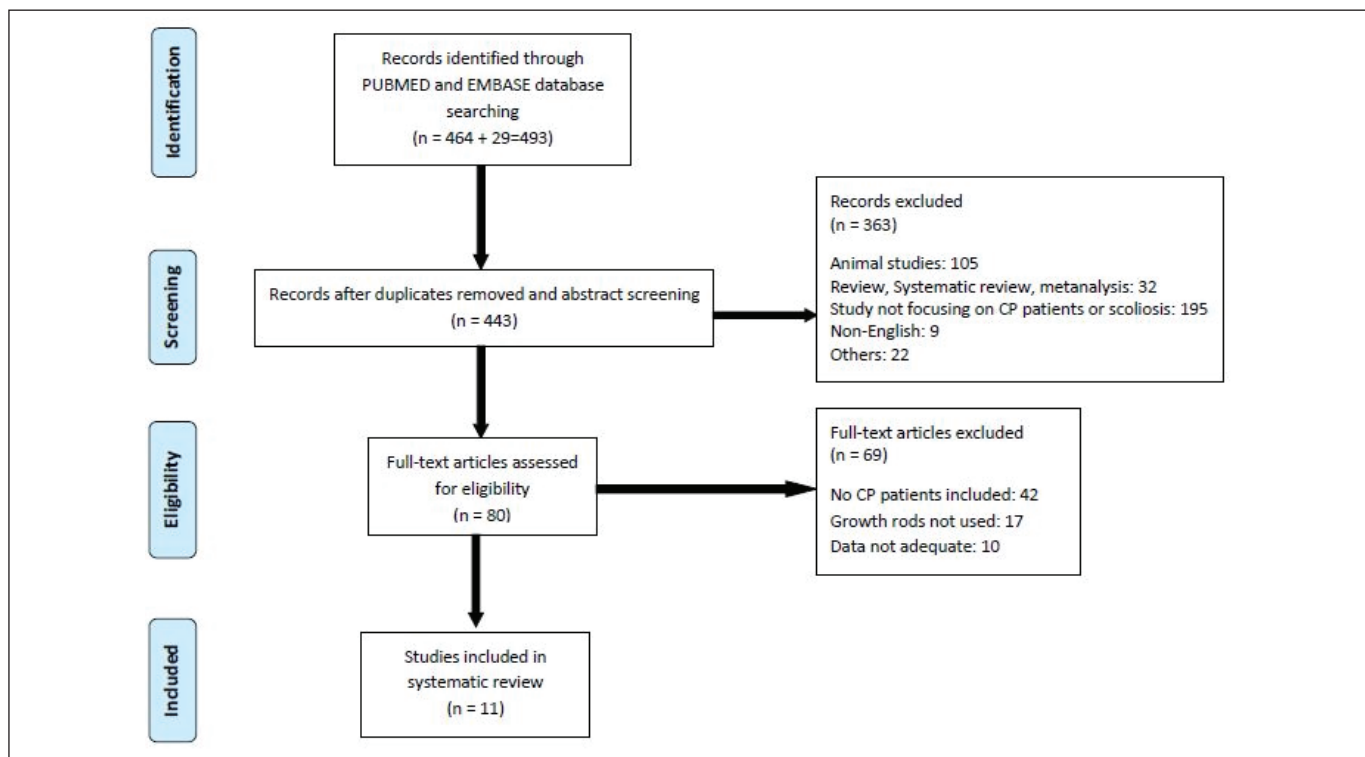


Figure 1: PRISMA Chart showing the selection of studies for inclusion in the systematic review.

obtained, Cobb's angle, pelvic obliquity, T1-S1 length, T1 tilt, space available for lung ratio and any complications was included wherever available. All of the extracted data was analyzed using IBM SPSS 27.0 software. Descriptive analysis using means, SD and ranges (minimum and maximum) across the included studies were performed.

■ RESULTS

Selection of Studies and Demographic Data

The total number of abstracts initially screened was 443 which led to a final selection of 11 articles meeting the inclusion and exclusion criteria to be included in this systematic review (Figure 1) (1,2,7–10,18,19,22,26,33). Of these, only 2 studies focused specifically on role of growing rods in CP (18,26). The studies were retrospective in nature (Level of Evidence IV) except 2 studies which were Level III (8,26). Data was extracted from rest of the articles specific to CP. A total of 181 patients with a mean age of 6.8 ± 1.3 (5-13 range) years at index surgery were included (Table I). There was a female preponderance among the patients (93/160, 58.1%). The patients had a mean follow up of 3.02 ± 1.3 (2-5.8 range) years.

Technique of Growing Rod Used

The most common location of the apex of the curves in the patients was thoracolumbar region. Dual rod traditional growth system was the most common instrumentation used (103/181, 56.9%). The instrumentation spanned an average of 15.2 ± 1.4 vertebrae. The other growing rod systems used were VEPTR (n=35), MCGR (n=35), single rod system (n=4), Luqué trolley technique using L- or U-shaped rods fixed to the spine by segmental sublaminar wires without fusion (n=2), and self-growing rods with gliding connectors (n=2) (7,9,18,19,26,33). The most common anchors used were hooks or pedicles proximally (T2-T4 commonly) and pedicle screws distally if ilium is not included and modified Galveston technique if pelvis is included.

Outcome

The Cobb's angle in the coronal plane improved (85 ± 20 degrees to 49 ± 20 degrees, 83 ± 22 degrees to 60 ± 25 degrees) at final follow up in the two large studies (18,26). Similarly, pelvic obliquity (27 ± 19 to 14 ± 13 degrees), T1-S1 length (increase of 7.9 cm) and space available for lung ratio (0.86 ± 0.29 to 1.04 ± 0.19) improved at the final follow up (18,26). Inclusion of pelvis led to improved correction of pelvic obliquity with similar correction of the Cobb's angle (18). Sun et al. noted that the primary curve correction was better in MCGR (46 ± 18 degrees) as compared to traditional growth rods (58 ± 26 degrees) and VEPTR (72 ± 24 degrees) at final follow up (26). Also, the maintenance of correction between immediate postoperative period and final follow up was superior in MCGR ($0 \pm 26\%$) as compared to traditional growth rods ($-49 \pm 88\%$) and VEPTR ($-62 \pm 78\%$). Dede et al. showed a gain of 17 mm in T1-12 and 54 mm in T1-S1 length over a 60 month follow up using self-growing rods with gliding connectors in a single patient (7). Hasler et al. reported an

improvement of T1 tilt of 55 degrees in a patient where VEPTR had been used (9).

Number of Lengthenings and Final Fusion

The largest series reported a mean of 4.4 ± 2.2 times of lengthenings per patient at an average interval of 11 ± 7 months (18). The lengthenings lasted for about 3.5 ± 1.8 years from the index surgery. Akbarnia et al. followed a protocol of 3 lengthenings at an interval of 6 months (1,2).

In the series by McElroy et al, 8 patients underwent final fusion at 12.3 ± 1.7 years (18). At final follow up post fusion, all parameters (Cobb angle, T1-S1 length and space available for lung ratio) showed improvement except pelvic obliquity as its correction gradually declined but still remained better than preoperative period of index surgery. In the series by Sun et al, 4 patients had undergone final fusion at the last follow up (26). Of these, 3 patients had undergone traditional growth rod and 1 had undergone VEPTR. The correction parameters had reversed as compared to immediate post-operative period but still remained improved as compared to the pre-operative period.

Complications

Wound related complications were most commonly noted (61/176, 34.6%). The complications were either in the form of deep or superficial wound infection or wound dehiscence. Other complications included unplanned surgeries for implant related complications (52/176, 29.5%), transient neuropraxia (1/45, 2.2%) and post-operative pneumonia (1/45, 2.2%). All the implant related complications occurred distally, either at the thoracolumbar junction or at the distal anchor site. No complications of iliac screw loosening had been noted.

The wound infections were noted after an average of 18.6 ± 4 months from the index surgery and after 2.3 ± 1.5 lengthenings. Of the patients with deep wound infection (n=8), 5 were treated with debridement and antibiotics and recovered, while in other 3, it led to abandonment of procedure and early fusion after removal of instrumentation (18). Gram negative bacteria (*Pseudomonas*, *E coli*) were the common pathogens isolated from the wound in case of deep infections.

Sun et al. compared the complications of MCGR and traditional growth rods separately (26). Infections were the commonest cause of unplanned return to operation room in traditional growth rods whereas implant failure was the commonest cause in the MCGR group. The percentage of complications was significantly higher in the traditional growth rod technique (57/86, 66.2%) as compared to the MCGR group (11/34, 32.3%).

■ DISCUSSION

Most of the reviews on natural history of neuromuscular scoliosis are derived from the behavior of the curve in cerebral palsy (20). The development of scoliosis in CP depends on the global disability caused by it. Nearly 50% of children with GMFCS IV-V develop severe scoliosis over their lifetime (21). A Cobb's angle of more than 40 degrees, younger age of onset

Table 1: Details of Studies Included in the Study

Study	Level of evidence	Number of patients	Age at index surgery(Years)	Sex	Scoliosis	Type of Instrumentation	Instrumentation (Y/N)	Lengthening (Interval in months)	Lengthening obtained (cm)	Result	Follow-up	Complication
Akbarnia et al, 2005 (2)	4	1	7.67	F	Thoracolumbar (T3-L4) Cobb-109°	T4-L4, Dual growing rods	Y (Post fusion)	3 (6)	T1-S1 gain- 12.0	Fusion done 2 years after index surgery. Final Cobb- 24°	24.96 months	Instrumentation was extended to pelvis post fusion due to curve progression
Akbarnia et al, 2008 (1)	4	1				Dual growing rods		3 (<=6)			25 months	None reported
Hasler et al, 2009 (9)	4	1	5	M	Left Thoracic curve Coronal Cobb- 100° Sagittal Cobb- 88° Pelvic obliquity- 27°	VEPTR; 1st construct- Rib to Pelvis; 2nd Construct- Rib to lumbar spine	N			Correction of Coronal Cobb- 30° Pelvic obliquity- 15° T1 tilt- 30° Sagittal Cobb increased by 6°	48 months	Upper cradle showed cut through after 1 year. Second load sharing construct applied
Mc Elroy et al, 2012 (18)	4	27	7.6 ± 2.4	M-9 F-18	Mean Cobb angle- 85 ± 20° Mean Pelvic obliquity- 27 ± 19°	Growth Rods: Single- 4 Dual- 23	Y (15 patients)	4.4 ± 2.2 (11 ± 7)	T1-S1 gain-7.9 ± 4.4	Correction of Cobb- 35 ± 23° Correction of Pelvic obliquity- 14 ± 19°	4.8 ± 2.1 years	Deep wound infection-8 Rod exchange-6 Rod fracture-5 Anchor revision-4 Superficial wound infection-2 Anchor dislodgement-2 Wound revision/dehiscence-2 Pneumonia-1
Dede et al, 2015 (7)	5	2			T2-L3, Cobb angle- 70° Pelvic obliquity- 28° Hypokyphotic thoracic curve	Self growing instrumentation with gliding connectors		NA	T1-T12 gain- 14 S1 gain- 23	Coronal Cobb corrected to 47° Pelvic obliquity corrected to 5°	24 months	None
					T8-L5, Cobb angle-33° Pelvic obliquity- 6° Maximum kyphotic angle- 112°	Self growing instrumentation with gliding connectors, T2-L5	Y		T1-T12 gain- 17 S1 gain- 54	Kyphosis corrected to 20° Cobb corrected to 30° Pelvic obliquity corrected to 2°	60 months	1 unplanned surgery to exchange rods after becoming dislodged from connector
Ridderbusch et al, 2016 (22)	4	1	9.2	F	Primary Lumbar Curve	MCGR		4			17 months	None

Table I: Cont.

Study	Level of evidence	Number of patients	Age at index surgery(years)	Sex	Scoliosis	Type of Instrumentation	Pelvic Instrumentation (Y/N)	Lengthening (Interval in months)	Lengthening (cm)	Result	Follow-up	Complication
Yilmaz et al, 2016 (33)	4	1				MCGR, T7-L3	N	7 (6)	R- 23 L- 31	Final Cobb angle- Thoracic coronal- 39° Lumbar coronal- 16° Lumbosacral- 5° Thoracic sagittal- 19° Lumbar sagittal-48°	46 months	None
Helenius et al, 2018 (10)	4	14				Majority-TGR						Right-sided motor and sensory deficit of lower extremity due to difficulty in placement of T2 right side pedicle screw in one patient
Mehdian et al, 2020 (19)	4	2				Self growing Rod technique		NA		Revision surgery and fusion due to wire or rod breakage		Wire or Rod breakage
Gaume et al, 2021 (8)	4	11	10.1 ± 2.3		Mean Cobb angle- 69 ± 23.42° Mean Pelvic Obliquity- 23 ± 16.86°	OWSER	Y	NA	T1-S1 gain- 7.45 ± 3.1	Cobb Correction- 36.18 ± 19.84° Pelvic Obliquity Correction- 17.18 ± 13.32°	3 years	Lack of Rod Expansion- 2 Surgical Site Infection- 2 Central Venous catheter infection- 2 Pyelonephritis- 1
Sun et al, 2022 (26)	3	120	7.2 ± 2.1	M-53, F-67	Mean Cobb angle- 83 ± 22° Mean Kyphosis- 56 ± 27°	TGR- 52 VEPTR- 34 MCGR- 34			T1-S1 gain: TGR- 25.3 ± 4 to 33.4 ± 5.7 VEPTR- 25.1 ± 4.1 to 32.2 ± 4.2 MCGR- 25.1 ± 2.7 to 33.4 ± 4.2 T1-T12 gain: TGR- 16.7 ± 2.8 to 21 ± 3.4 VEPTR- 15.9 ± 3.3 to 19.6 ± 3.4 MCGR- 16.6 ± 2.5 to 21 ± 3.3	Cobb Correction: TGR- 82 ± 24 to 58 ± 26° VEPTR- 80 ± 22 to 72 ± 24° MCGR- 90 ± 19 to 48 ± 18° Kyphosis Correction: TGR- 52 ± 24 to 53 ± 25° VEPR- 62 ± 29 to 57 ± 26° MCGR- 57 ± 29 to 42 ± 15°	5.8 ± 3.1 years	Total: 87 TGR- 42 VEPTR- 34 MCGR- 11 Most common complication in TGR and VEPR groups was deep infection while in MCGR group was implant failure in first 2 years and implant failure and deep infection at final followup

and non-ambulatory status are significant risk factors for rapid progression of scoliosis (19). Larger curves (>50 degrees) have been shown to progress at twice the rate compared to smaller curves (<50 degrees) (28). Curves have also been seen to progress even after skeletal maturity in quadriplegics.

Scoliosis associated with thoracic or thoracolumbar kyphosis is the most common spinal deformity in CP (30). Two distinct types of curves occur in CP with group 1 curves occurring in patients with good ambulatory function and less severe deficit like monoplegia or hemiplegia (16). The pelvis is levelled in such curves and they usually behave on the lines of idiopathic scoliosis. Group 2 curves develop in severely affected individuals who are usually non-ambulatory and have a distinct component of pelvic obliquity with the typical long C-shaped curves as seen in neuromuscular scoliosis. Pelvic obliquity usually develops either due to extension of curve distally or asymmetrical contractures at the hip joint (15). Apart from scoliosis, sagittal deformities like thoracolumbar kyphosis can develop in quadriplegics with prevalence of around 7% (23). These also develop in patients with generalized hypotonia, hamstring spasticity, hamstring contracture and post-operative rhizotomy patients. Hyperlordosis of lumbar spine may also develop in patients with flexion contractures of hip or post-operative rhizotomy patients secondary to multilevel laminectomies.

Trunk instability during the growing years leads to asymmetric bone growth leading to progression of the curve as well as impediment of head control and hand function. Current evidence points to use of brace at a very early age when the patient is not able to sit upright without support or Cobb's angle is more than 20 degrees and it may be in the form of a lumbar or thoracolumbar spinal orthosis. The child is then closely followed up clinically and radiologically (24). The brace should have an appropriate fit, with tight fit at the pelvis, spacer between pelvis and ribs and adequate space at the chest region to allow for normal breathing. In curves less than 45 degrees, conservative treatment has been seen to provide correction in the range of 35-40% at final follow up (24).

Surgery has traditionally been indicated in patients with Cobb's angle more than 40 to 50 degrees with focus on sitting tolerance and additional aims to be improvement in ease of doing activities of daily living, ease of care and social interaction (11). Fusion was performed traditionally to halt the curve progression and obtain a level pelvis over a level spine. But fusion at age less than 10 years would affect the final height and the thoracic volume. In such patients, growth friendly constructs are an option which is being increasingly accepted globally.

Growth friendly constructs in the management of scoliosis in cerebral palsy has shown promising results because it preserves the growth as well as provides a degree of correction in the meantime (18). Dual rod traditional growth system was the most common instrumentation used. The most common anchors used were hooks or pedicles proximally (T2-T4 commonly) and pedicle screws distally if ilium is not included, and modified Galveston technique if pelvis is included. Since a substantial number of these patients have pelvic obliquity, it is

imperative to instrument the pelvis in such cases if an improved spinal alignment is expected. For curves with more than 15 degrees of pelvic obliquity, it is recommended to instrument pelvis even if the child is skeletally immature (12). Moreover, even if the improvement in pelvic obliquity is minimal (27 to 14 degrees), it should nevertheless be done to prevent the progress of the obliquity. More correction of the pelvic obliquity in large, less flexible curves can be improved by an anterior release along with posterior spinal fusion (3). Apart from dual rods, other rarer implant system used were single rod system, Luqué trolley technique using L- or U-shaped rods fixed to the spine by segmental sublaminar wires without fusion, self-growing rods with gliding connectors, VEPTR and MCGR. The data on these rarer methods are very scarce to comment on its role in CP. In this review, T1-S1 length increased by 7.9 ± 4.4 cm) at final follow up. In another systematic review on early onset scoliosis, TGR have shown maximum increase in T1-S1 length per year (10mm per year), followed by MCGR (9mm per year) and VEPTR (5mm per year) (32).

A complication rate as high as 34.6% was noted in these patients undergoing growth sparing surgery. Patients with greater neurological involvement and increased curve size were at significant risk for postoperative complications (17). Although in a recent meta-analysis, respiratory complications had been the commonest, it was not so in this review which may be due to patient selection bias for growth sparing surgeries in CP (25). Wound related complications (34.6%) were most commonly noted in this review and they occurred after an average of 18.6 ± 4 months from the index surgery and after 2.3 ± 1.5 lengthenings and maybe related to the multiple surgeries. This rate of infection is high as compared to growing rods in idiopathic scoliosis which may be due to increased spasticity of muscles, increased rigidity of curves and comparative reduced care of the patients with cerebral palsy (4,5,13). Anecdotal evidence may suggest use of vancomycin at the surgical site may reduce the incidence of infections (6). Current best evidence points to use of intra-wound antibiotics as well as intravenous antibiotics, frequent posture change on bed and improving nutritional status to reduce infection rates (31). These patients are less prone for mechanical complications (implant failure) as compared to more active patients of idiopathic scoliosis (18).

The major anesthetic concern in such patients is the level of patient's communication ability and cognition, which makes it a challenge for the anesthetist. Many of these patients present with significant co-morbidity and high ASA scores. Patients with scoliosis are more likely to have respiratory compromise and ideally should have preoperative physiotherapy, antibiotic and ICU care to prevent postoperative complications such as secretion retention, atelectasis and lung collapse. Propofol is the induction agent of choice in patients with reactive upper airways, which is common in such patients (27). A gas induction with a head tilt of 20-30 degrees is the only safe option in an uncooperative patient and it can be done in a "veinless" manner. Difficulties in communication can make postoperative pain assessment difficult and postoperative analgesia should be based on "continuous" rather than "on demand" regimens.

The major and compelling limitation of this systematic review is the limited number of cases who underwent growth sparing surgeries for scoliosis in CP. Further, the studies included in this review were heterogeneous to a large extent in terms of age and instrumentation used for these patients. Also, other issues of contention like necessity of pelvic fixation in pelvic obliquity, the ideal age for intervention and ideal instrumentation with predictable results with reduced complications should be studied on a larger scale with larger sample size with an adequate follow up. Only 2 studies in this series dealt with CP exclusively and the rest dealt with neuromuscular scoliosis with CP being one of the causes (18,26). Also, the current literature lacks any high-quality study comparing growth sparing surgeries in idiopathic and cerebral palsy. Most of the studies included in this review did not include any objective evaluation of the outcome on the basis of any score. But this study is the first comprehensive review of growth sparing surgeries for scoliosis in CP.

CONCLUSION

Treatment of scoliosis in CP remains a challenge. Growth sparing surgeries can be an effective method to treat scoliosis in selected patients of CP which would prevent more severe anterior or posterior fusions and would give time for spinal growth before the final fusion. MCGR holds a bright promise for the future considering its ability to maintain correction as well as the lower rate of complications. Complications and risks may be higher as compared to other categories of early onset scoliosis and this long course of treatment till final fusion warrants caution. The benefits and risk of immediate fusion with respect to growth sparing surgeries should be considered before the decision. Dual rod growth instrumentation has been the most commonly used till date. Lengthenings can be done in interval of 6-12 months. Wound related complications in view of repeated surgeries and implant related complications in view of the spasticity remain areas of concern and warrants future biomechanical studies.

AUTHORSHIP CONTRIBUTION

Study conception and design: SB, PK

Data collection: KG, MHS

Analysis and interpretation of results: SB, KG, MHS

Draft manuscript preparation: SB, VR

Critical revision of the article: VR, PK

All authors (KG, SB, MHS, SC, SKS, VR, PK) reviewed the results and approved the final version of the manuscript.

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