Combined Halo Gravity Traction and Dual Growing Rod Technique for the Treatment of Early Onset Dystrophic Scoliosis in Neurofibromatosis Type 1

Enjie Xu, Rui Gao, Heng Jiang, Tao Lin, Wei Shao, Xuhui Zhou

OBJECTIVE: **To determine the safety and effectiveness of the combined halo gravity traction and dual growing rod technique in achieving and maintaining scoliosis correction while allowing spinal growth.**

METHODS: **From January 2014 to July 2017, 11 patients with dystrophic neurofibromatosis type 1 (NF1)—associated scoliosis, including 7 men and 4 women, underwent combined halo gravity traction and dual growing rod technique procedures. Diagnoses were all dystrophic NF1-associated scoliosis. Patients with a Cobb angle of major curve >60° and flexibility of spine <30% were included in our research. Analysis included age at the time of treatment, levels of instrumentation, number and frequency of lengthening, lengthening distance, and complications. The changes in Cobb angle of scoliosis and T1-S1 length of spine over the treatment period were measured by radiographic evaluation.**

RESULTS: **The average age of treated patients was 7.2 years (range, 5—9 years). Growing rods were lengthened every 6 months through exposure. The mean number of times of lengthening was 3.9 (range, 3—5). The distance of each extension was 1.6 cm (range, 1.0—2.0 cm). The Cobb angle was corrected 41.7% on average after traction, 48.4% after initial surgery, and 53.3% at the last follow-up. T1-S1 length increased 3.4 cm (range, 1.2—5.1 cm) on average over a mean treatment period of 2.2 years, with an average of 1.5 cm/y (range, 0.5—2.3 cm/y). During the treatment period, complication of hook dislodgement occurred in 1 of 11 patients (9.1%).**

CONCLUSIONS: **The combined halo gravity traction and dual growing rod technique can safely and effectively correct NF1-associated scoliosis. This is an ongoing study that requires long-term follow-up.**

INTRODUCTION

Neurofibromatosis (NF) is an autosomal dominant genetic disorder characterized by aberrant proliferation of neural crest cells. Typically, NF is divided into 2 clinical types: neurofibromatosis type 1 (NF1) and NF type 2. NF1 is the most frequent one, and also termed von Recklinghausen disease or peripheral NF. Scoliosis is the most common skeletal manifestation of NF1. The prevalence of scoliosis in patients with NF1 is between 10% and 64%.[1,2] Furthermore, scoliosis in NF1 is usually classified into 2 basic types (nondystrophic and dystrophic) based on the natural history and curvature characteristics. Dystrophic curves often occur in early age and highly progress because of dystrophic changes such as vertebral wedging, vertebral rotation, and rib penciling, which lead to worse prognosis.[3-10] Progressive spinal deformities in early life present serious health risks for children.[11,12]

Generally, the use of nonoperative methods, such as a brace, are not effective for dystrophic curves,[4,13,14] and early and aggressive surgical intervention is strongly recommended.[6] There are 2 main surgical options available called the fusion and nonfusion techniques. The nonfusion technique is a better choice to achieve spinal growth postoperatively in very young children. One of the nonfusion options is a growing rod, which achieves deformity correction and maintains it during the treatment period while allowing continued spinal growth.[15-17] Many published reports indicate that the growing rod technique is an effective treatment for progressive scoliosis in early onset patients.[16-18] Recently, the dual rod technique has become the preferred method.[19-21]

Key words
- Dystrophic scoliosis
- Early onset
- Growing rod
- Halo gravity
- Neurofibromatosis type 1
- Traction

Abbreviations and Acronyms
NF: Neurofibromatosis
NF1: Neurofibromatosis type 1

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Table 1. Clinical Data

Patient Number	Sex	Age at Treatment (years)	Follow-Up Time (years)	HGT Period (weeks)	Upper Foundation Anchors	Lower Foundation Anchors	Number of Times of Lengthening	Average Lengthening Interval (month)	Distance of Lengthening (cm)	Instrumental Levels	Complication
1	F	7	2.00	3	S	S	3	6.0	1.0	T2-L4	None
2	M	6	2.50	4	S	S	4	6.0	1.2	T2-L3	None
3	M	7	2.75	8	S	S	5	6.0	2.0	T2-L3	None
4	F	9	2.00	5	S	S	3	6.0	2.0	T2-L3	None
5	F	7	2.00	4	S	S	4	6.0	1.2	T2-L5	None
6	F	5	2.50	3	S	S	4	6.0	2.0	T2-L3	None
7	F	7	2.25	3	S	S	4	6.0	1.5	T3-L3	None
8	F	7	2.50	4	H	H	5	6.0	2.0	T2-L3	*
9	M	9	2.00	3	S	S	4	6.0	1.5	T2-L4	None
10	F	7	2.00	3	S	S	4	6.0	1.5	T3-L3	None
11	F	8	2.00	4	S	S	3	6.0	2.0	T2-L3	None

HGT, halo gravity traction; F, female; S, screw; M, male; H, hook.
*Hook dislodgement.

Although the dual growing rod technique is an effective treatment method for early onset spinal deformity, the rapid correction of rigid deformities may increase the risk of neurologic complications. In addition, there are extreme corrective forces during the rapid correction period which make fixation difficult.[22] We support the use of preoperative halo gravity traction to aid in the correction of such deformities to overcome these problems. Curve correction can be done gradually and provides a good condition for subsequent surgery.

The purpose of this paper is to present the overall clinical experience with the treatment of combined halo gravity traction and the dual growing rod method for dystrophic NF1-associated scoliosis.

MATERIALS AND METHODS

Study Design and Patients Characteristics

A consecutive series of patients who underwent halo gravity traction prior to spinal deformity surgery for early onset dystrophic NF1-associated scoliosis in our institution from January 2014 to July 2017 were reviewed. Inclusion criteria were patients with dystrophic NF1-associated scoliosis who were <10 years of age at onset, with a major curve >60° and a stiff spine with flexibility <30%, and who planned to undergo combined halo gravity traction and dual growing rod technique. All patients had >2 years of follow-up (range, 2–2.75 years) (Table 1).

With institutional review board approval, all medical records and radiographs were retrospectively reviewed. The clinical information included age at the time of treatment, sex, diagnosis, prior treatment, surgical information (levels of instrumentation, number of times of lengthening, and lengthening intervals), and complications. Side-bending radiographs were taken before treatment to evaluated spine flexibility. All upright posteroanterior and lateral radiographs were available to calculate the Cobb angle before treatment, after halo gravity traction, after surgery (within 2 weeks after the initial surgery), at each lengthening period, and at latest follow-up. T1-S1 length was measured from the middle of the upper end plate of T1 to the middle of the upper end plate of S1. All postoperative radiographic measurements were calibrated and corrected for magnification to represent actual change. Two observers measured each radiograph independently.

Preoperative Halo Protocol

Halo application was performed under conscious sedation with local anesthesia. A total of 6–8 pins were placed and tightened for all patients. The halo was positioned slightly above the equator of the skull, eyebrows, and superior aspect of the pinnae. Halo gravity traction was started at 2–3 kg and increased gradual until 50% of body weight was reached. Patients who had difficulty tolerating traction were kept at the lowest weight tolerable and advanced as they became more comfortable. All patients achieved goal traction (50% of body weight) by the third or fourth week. Traction was maintained at all times, even sleep, using a mobile wheelchair apparatus and a traction frame used while moving (Figure 1). Patients can move in the wheelchair in daily life. If they need to interrupt traction because of going to the toilet or for other reasons, they can untie the leash and walk on their own, and their walking will not be affected. Patients were monitored and evaluated daily for pin site complications or neurologic changes. Pins were torqued weekly by the surgical team.

Surgical Technique

The cephalad and caudal exposures for insertion of anchors are subperiosteal. The remaining area of exposure is subcutaneous or subfascial to prevent premature unwanted fusion (autofusion). The selection of the anchor sites is based on the location and type

Figure 1. A modified wheelchair and a patient who received halo gravity traction. (**A**–**C**) Side, front, and back of the modified wheelchair, respectively. (**D**) A patient with dystrophic neurofibromatosis type 1–associated scoliosis received halo gravity traction.

of the curve and the child's age. Generally, in patients with dystrophic conditions, longer instrumentation is preferred. The choice of screw or hook depends on availability of posterior elements and anatomic variations. We prefer pedicle screws which appear to add additional stability to the construct. Placing bone grafts at the exposed levels of foundation allows fusion of these segments and adds stability at the anchor sites.

The titanium rods are 5.5 mm in diameter. They are cut into 2 segments, 2 for each side, and contoured for sagittal alignment. At the foundation levels, the rods are connected to the anchors before all the screws are tightened. There should be 2 rods connected to each foundation. The proximal and distal rods are then connected by a tandem connector on each side. The connectors are attached to the rods by first sliding their proximal sides and placing the distal sides on the thoracolumbar junction with minimal adverse effect on the sagittal contour of the spine (Figure 2). The blood loss of these surgical patients was about 150–300 mL.

In our study, a thoracolumbosacral orthosis is used for postoperative patients in the first 3 months when the fused foundation sites are solid.

Lengthening is performed every 6 months in our patients, and the distance of each extension is 1–2 cm. The tandem connector is partially exposed through a small midline incision. The set-screws at one end of the connectors are loosened, and traction is accomplished between the 2 rods by placing a special distractor inside the tandem connector. The set-screws are tightened subsequently. Traction can also be accomplished between the rod and the tandem connector on either side by using the rod holder and regular distractor. Lengthenings are stopped when no further traction can be achieved. A final fusion surgery is performed when the distance of the last extension is <0.5 cm. Magnetic resonance imaging was applied to determine the location of the tumor and to minimize bleeding during surgery by avoiding touch with the tumor.

RESULTS

Patient Data

The patients with a Cobb angle of major curve >60° and flexibility of spine <30% were included in this study, and subsequently underwent preoperative halo gravity traction. There were 7 boys and 4 girls in our study, and all were diagnosed with dystrophic NF1-associated scoliosis. The mean age at surgery was 7.2 years (range, 5–9 years). Patients underwent preoperative halo gravity traction for 4 weeks (range, 3–8 weeks).

The anchors were all screws in 10 patients and all hooks in 1 patient. The location of the dual rods was subfascial in all patients. The rods were changed in 2 patients during the planned

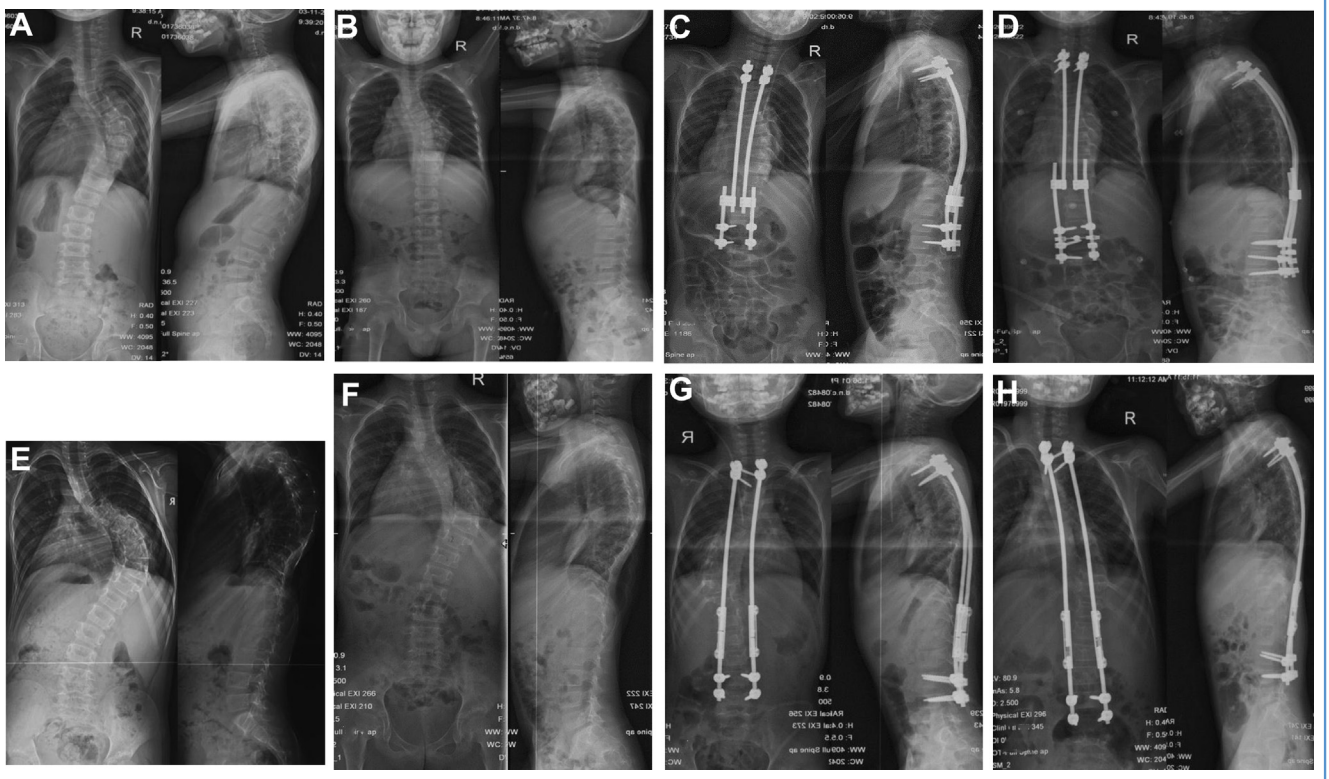

Figure 2. Clinical example of combined halo gravity traction and dual growing rod technique. (**A**–**D**) A boy with early onset dystrophic neurofibromatosis type 1–associated scoliosis. (**E**–**H**) A girl with early onset dystrophic neurofibromatosis type 1–associated scoliosis. (**A** and **E**) Pretreatment, (**B** and **F**) after halo gravity traction, (**C** and **G**) after initial surgery, and (**D** and **H**) at last follow-up.

lengthening procedures. The patients were observed for a minimum of 2 years after initial surgical treatment with an average of 2.2 years of follow-up (range, 2.00–2.75 years). No one in our study performed final fusion because of immature skeleton at the last follow-up. The average number of times of lengthening was 3.9 (range, 3–5) per patient with an interval of 6 months. The distance of each extension was 1.6 cm (range, 1.0–2.0 cm) (Table 1).

Measurement of Curves and Spinal Balance

The scoliosis Cobb angle improved from an average of 72.0° (range, 60°–100°) before treatment to 42.0° (range, 35°–75°) after traction to 37.2° (range, 29°–63°) after initial surgery to 33.6° (range, 27°–40°) at latest follow-up. The percent change of scoliosis Cobb angle was corrected by 41.6% (range, 31.7%–52.6%) on average after traction, 48.4% (range, 37.0%–59.0%) after initial surgery, and 53.3% (range, 40.0%–61.8%) at the last follow-up (Table 2).

Growth

The spinal length obtained was calculated from the amount of elongation of T1-S1 before treatment to latest follow-up. The T1-S1 length increased from 26.6 cm (range, 22.6–29.1 cm) before treatment to 29.9 cm (range, 26.5–33.2 cm) after traction to 32.0 cm (range, 28.0–34.7 cm) after initial surgery (elongation) to 35.4 cm (range, 32.2–39.2 cm) at latest follow-up (growth). In addition to initial elongation averaging 5.4 cm (range, 3.8–6.2 cm), the growth over the treatment period was 3.4 cm (range, 1.2–5.1 cm) over a mean treatment period of 2.2 years, with an average of 1.5 cm/y (range, 0.5–2.3 cm/y) (Table 3).

Complications

During the treatment period from halo gravity traction to last follow-up, 1 of the 11 patients (9.1%) had a complication with hook dislodgement (Table 1). The patient was found by his mother to have an abnormal lump on his waist. Radiographic examination was done in hospital which showed a hook dislodgement at lumber level. A subsequent surgery was performed to remove and replace all hooks with pedicle screws. There was no particular discomfort in this patient who received an unplanned surgery. No other complications occurred in our study, especially autofusion, which exists frequently during growing rod procedures (Figure 3).

DISCUSSION

Rapid correction of rigid deformities increases the risk of neurologic complications and hardware failure.[23] In particular, patients' bone quality may be inadequate to provide a strong corrective force.[7] We recommend preoperative application of halo gravity

Table 2. Flexibility and Cobb Angle Changes During Treatment Period

Patient Number	Flexibility (%)	Cobb Angle						
		Pre-HGT	Preoperation	After Initial Operation	Last Follow-Up	Correction from Pre-HGT to Preoperation (%)	Correction from Pre-HGT to After Initial Operation (%)	Correction from Pre-HGT to Last Follow-Up (%)
1	25	65°	40°	32°	31°	38.5	50.8	52.3
2	20	65°	35°	29°	26°	46.2	55.4	58.5
3	23	78°	37°	32°	30°	52.6	59.0	61.5
4	25	75°	42°	40°	42°	44.0	46.7	48.0
5	22	100°	75°	63°	39°	25.0	37.0	61.0
6	18	62°	35°	30°	27°	43.5	51.6	56.5
7	19	60°	41°	37°	36°	31.7	38.3	40.0
8	26	65°	35°	33°	32°	46.2	49.2	50.8
9	25	70°	36°	34°	32°	48.6	51.4	54.3
10	24	72°	42°	39°	40°	41.7	45.8	44.4
11	22	80°	44°	40°	35°	45.0	50.0	53.8

HGT, halo gravity traction.

traction to aid in the correction of rigid deformities. The primary goal of preoperative traction is to avoid major neurologic risk, while in a controlled and safe way, to obtain correction effectively from surgery in early onset dystrophic NF1-associated scoliosis. Halo spinal traction was introduced for spinal deformity management by Nickel et al.[24] Halo gravity traction is one of the methods of preoperative spinal traction. Halo gravity traction is a safe, well-tolerated method of providing significant corrective forces while still allowing patient mobility.[25] Importantly, halo gravity traction has been said to achieve a gradual correction over a long period, to reduce the complexity of subsequent surgery, and to improve safety.[26,27] Park et al.[28] thought that preoperative traction reduced the risk of neurologic injury, and an alignment corrected spine would make surgery less technically challenging. The effectiveness of preoperative halo gravity traction has been demonstrated in some research. Sink et al.[29] reported 19 children with severe scoliosis who were treated with 6–21 weeks of preoperative halo gravity traction before spinal fusion. The Cobb angle corrected an average of 35% with traction. Rinella et al.[25] reported 33 patients with severe scoliosis treated with

Table 3. T1-S1 Growth Including Elongation and Growth

Patient Number	Pre-HGT (cm)	Preoperation (cm)	After Initial Operation (cm)	Last Follow-Up (cm)	Elongation (cm)	Growth (cm)
1	29.1	33.2	34.7	39.2	5.6	4.5
2	28.7	32.5	34.7	39.0	6.0	4.3
3	27.9	31.4	33.4	35.0	5.5	1.6
4	28.4	30.6	33.8	37.8	5.4	4.0
5	27.4	28.6	32.8	34.0	5.4	1.2
6	26.9	28.4	30.7	34.5	3.8	3.8
7	25.6	30.1	31.8	36.9	6.2	5.1
8	22.6	26.5	28.0	32.4	5.4	4.4
9	26.4	30.1	31.9	34.4	5.5	2.5
10	24.5	28.8	30.2	34.0	5.7	3.8
11	25.4	28.6	30.4	32.2	5.0	1.8

HGT, halo gravity traction.

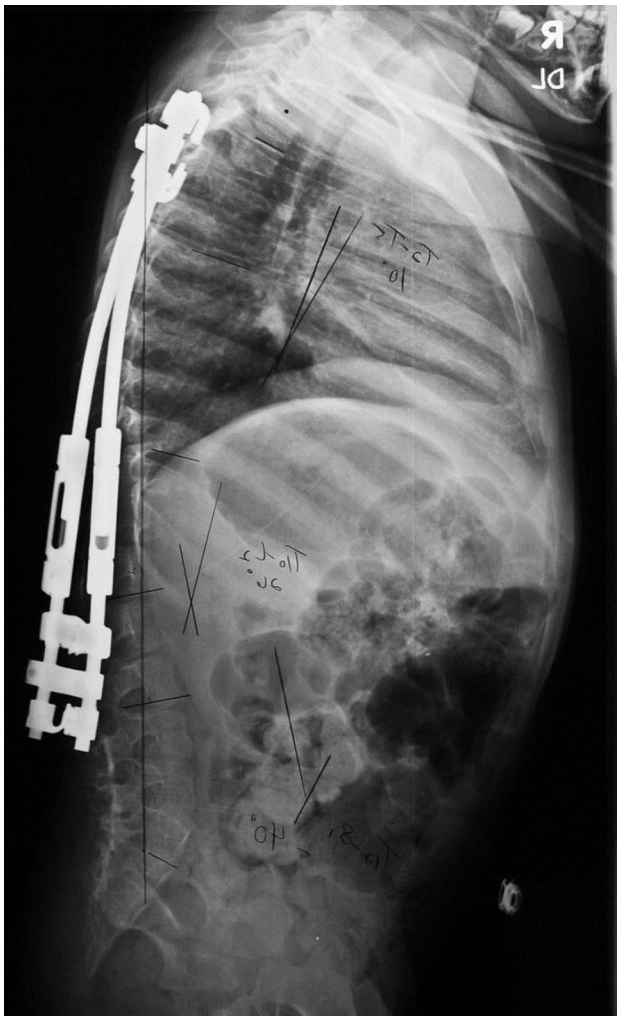

Figure 3. Only 1 complication occurred in our patient series. The radiographic examination of this patient showed a hook dislodgement at lumber level.

halo gravity traction. They suggest that preoperative halo gravity traction is a safe, well-tolerated method to improve the surgical results of patients with severe scoliosis. Sponseller et al.[26] found that the patients with preoperative traction less frequently underwent a vertebral body resection. In our patients, the average correction of major coronal curves because of preoperative traction was 41.6%, and the lower incidence of complications also suggests that preoperative traction can reduce the complexity of surgery and improve its effectiveness. However, complications do exist while using halo gravity traction, including pin loosening, superficial, and deep pin tract infections, deep intracranial lesions, cranial osteomyelitis, and intradural and extradural infections.[25,30-32] Complications such as temporary cranial nerve palsy, respiratory distress, triceps palsy, and brachial plexus palsy are also found in some cases.[25,33] In contrast with other reported case series, we did not see any traction-related complications in our patients. This result may be because of the short duration of traction or subtle changes in examinations that were not reports.

Early onset dystrophic NF1-associated scoliosis progresses rapidly in young age and generally with a poor response to nonoperative methods. Spinal fusion surgery in young children will stunt the growth of trunk and thorax, resulting in decreased vital capacity and lung development.[23] Nonfusion techniques, such as a growing rod, are considerable methods to correct spinal deformity while allowing for continued spinal growth. Harrington[15] originally reported the growing rod technique in 1962. Moe et al.[17] subsequently developed the technique for the treatment of progressive curves in young children. Several modifications were introduced subsequently to improve results. Studies of the single growing rod technique can be used to evaluate the effective by these methods. Klemme et al.[16] reported 67 children with progressive scoliosis who were treated with a program of incremental traction spinal instrumentation without fusion supplemented by full-time external orthotic support. During the treatment period, curve magnitude improved from an average of 67° (range, 38°–118°) at initial instrumentation to 47° (range, 19°–88°) at definitive fusion. Unfused spinal segments grew an average of 3.1 cm (range, 0.0–10.2 cm) over a mean treatment period of 3.1 years (range, 0.5–6.6 years). A recent study conducted by Acaroglu et al.[34] retrospectively evaluated 12 patients who underwent single rod instrumentation without fusion, noting that the patients had an improvement in the degree of spinal curvature but increased vertebral rotation. The single growing rod technique was improved to the dual rod growing technique subsequently to get better outcomes. Recent reports have indicated that the dual rod method had a better therapeutic effect in correcting adolescent idiopathic scoliosis compared with the single rod construct.[35] Fewer implant and crankshaft complications occurred in the patients treated with the dual rod technique compared with previous reports of the single rod technique.[36,37] Akbarnia et al.[18] found that patients who underwent the dual rod technique after initial elongation from surgery had a growth that was very close to the normal growth of the spine. The duration of treatment was longer than previously reported series, and the complication rate was relatively lower than other reports significantly.[37]

Final spinal fusion surgery will be performed when maximum spinal growth is accomplished. The most common indications for final surgical fusion were satisfactory axial alignment and balance, no rod fractures within the previous 2 years, and little length gained with the latest lengthening procedure.[38] No one in our study performed the final fusion because of immature skeleton at the last follow-up. In addition, the correction of curves did not exceed 60% even in the last follow-up. We think it is caused by severe spinal deformity and stiff spine of the patients in our study.

The complication rate in our patients is 9.2%, probably because of the small sample size we studied. It is evident that there is a high rate of complications associated with the growing rod technique. Blakemore et al.[39] reported 29 patients who underwent single submuscular Isola rod with a 24% complication rate. Mineiro and Weinstein[36] studied 11 children who were treated with growing rods. There were 1.5 complications per patient. Bess et al.[40] reported on 140 patients with early onset scoliosis

who underwent growing rod treatment between 1987 and 2005. Eighty-one of the 140 patients (58%) had a minimum of 1 complication. Klemme et al.[16] reported multiple complications in a group of 67 patients with severe early onset scoliosis receiving spinal instrumentation without fusion. Hook dislocations occurred on 21 occasions, with 16 involving the cephalad hook. In 2 cases, dislocation was associated with documented laminar fracture. Rod fracture was seen 12 times. Mineiro and Weinstein[36] questioned the worthiness of growing rod techniques. Fifty-three surgical procedures were performed in 11 patients before the definitive spinal instrumentation and fusion, and 17 complications were reported (an average of 1.5 per patient), including hook dislodgment, rod failure, deep infection, and superficial infection.

Interestingly, a complication reported frequently in other studies called autofusion did not occur in our patients, which suggests preoperative halo gravity traction may play a role in reducing this complication. We imagine this phenomenon is because of subfascial rods installation, thereby reducing injury in immature bone. Gradual correction before surgery also provide good conditions for decreasing disturbance in paraspinal muscle during surgery. In addition, our patients did not show significant pain during the traction process. This may be because the patient can move freely in the wheelchair during treatment, which may improve the patient's quality of life, or because the force of traction increased slowly. The small number of patients may also be a cause.

This study is limited by its retrospective nature and its small sample size. Further investigation on this subject, or a multicenter study, may help to elucidate any such correlation.

CONCLUSIONS

The combined halo gravity traction and dual growing rod technique is safe and effective. It performs traction at the first stage to provide a better surgical condition and maintains correction obtained at initial surgery while allowing spinal growth to continue. It provides an increased duration of treatment period and has an acceptable rate of complication compared with previous reports using the growing rod technique alone. However, this is an ongoing study that requires long-term follow-up to confirm our interim findings.

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