Mid-long-term surgical outcomes of posterior only instrumented spinal fusion procedure for early-onset scoliosis in type 1 neurofibromatosis

Jianguo zhang (jianguozhangpeking@outlook.com)
Peking Union Medical College Hospital https://orcid.org/0000-0001-9773-530X
Siyi Cai
Peking Union Medical College Hospital
zhenyao Li
Peking Union Medical College Hospital
Guixing Qiu
Peking Union Medical College Hospital
Jianxiong shen
Peking Union Medical College Hospital
hong zhao
Peking Union Medical College Hospital
Yu Zhao
Peking Union Medical College Hospital
Yipeng Wang
Peking Union Medical College Hospital

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Abstract

Background The pediatric neurofibromatosis type 1 (NF-1) patients' mid-long-term surgical outcomes of posterior spinal fusion is rarely reported, and the orthopedic maintenance effect is not clear. This study aims to evaluate the mid-long-term surgical outcomes of posterior only instrumented spinal fusion for early-onset scoliosis (EOS) in NF-1 patients.

Methods A retrospective review was performed on a cohort of 10 patients diagnosed as NF-1 with EOS from 2008 to 2014 in our hospital. This study included four male and six female NF-1 patients with a mean age of 7.8 years old when they underwent posterior only instrumented spinal fusion for their EOS. General clinical data, operation-related data were reviewed, and the dystrophic progression of EOS was evaluated during the follow-up.

Results The mean follow-up duration was 54 months (24 to 88 months). All patients underwent posterior only instrumented spinal fusion at 1 stage. The primary curves of EOS were thoracic in 9 cases, and only one patient had lumbar scoliosis. Preoperative major curve was significantly corrected (from 66.1 to 31.1 degrees). However, the average major curve deteriorated significantly to 40.1 degrees on average at the end of follow-up. The T1-S1 distance increased 2.77cm on average and increasing at 0.6cm/year during the follow-up.

Conclusions For the surgical treatment on EOS in NF-1 patients, although with relatively short segments involvements,posterior only fusion was not good way to treat such patients, especially the maintenance of orthopedic effect was not satisfactory.

Introduction

Neurofibromatosis type 1 (NF–1) is an uncommon neurocutaneous disorder caused by autosomal dominant mutations on chromosome 17q11.2. It was first described by von Recklinghausen in 1882, and it is characterized by a variety of typical manifestations featuring the deterioration of skin, bones, arteries, peripheral nerves, and central nervous system. The incidence of NF–1 is from 1/4000 to 1/5000 in epidemiology.(1) Ten to sixty percent of the NF–1 patients share the symptom of early onset spinal deformity, dystrophic or non-dystrophic.(2) Typical NF–1 dystrophic scoliosis has a short, sharp curve and it can be recognized by having 3 or more characteristic dystrophic features such as rib penciling, vertebral scalloping, wedging, rotation and spindling of the transverse process. Considering that the dystrophic NF–1 scoliosis has a tendency of curve progression which will result in severely poor pulmonary function and trunk height loss, aggressive treatment is recommended to NF–1 patients with dystrophic scoliosis.(3–5) When a final fusion is performed, almost half of the time additional are fused and the growing rod technique that would implicates the longer segments of the spinal. [6] However, due to the short and sharp curve of NF–1 patients with dystrophic scoliosis, some surgeons had suggesting to use the short segments fusion technique would provide improved postoperative motor function and activity of daily living finally.

Anterior-posterior fusion has been widely used to treat dystrophic scoliosis of NF–1 patients.(6, 7) Recently, the posterior only instrumented fusion was reported to be used to treat NF–1 dystrophic scoliosis since it could avoid the challenges in the anterior approach caused by the extensive plexiform tumors and this approach had an excellent short-term result being reported. (8, 9) Only Li's study included patients with a minimum age of 8 years of EOS (mean age 13 years), but the exact number was not stated. However, whether posterior spinal fusion will get a better mid-long term outcome in patients at a young age (age<10Yrs) is rarely reported.[11]

This study aims to evaluate the mid-long-term surgical outcomes of posterior fusion instrumented only surgeries for NF–1 patients who have dystrophic early onset scoliosis (EOS).

Methods

Patients

This study was approved by the institutional review board, and the consent form from each patient was obtained. The NF–1 diagnosis was conducted by using established diagnostic criteria.[8,12] The inclusion criteria, patients' population, Surgical Procedures, and data collection are described in this section. All Patients met the diagnosis of dystrophic EOS. Among all the 94 patients who received surgeries in the study period from March 2008 to March 2014, 26 patients with dystrophic EOS underwent the initial surgery when they were under ten years old.

Within these 26 patients, patients who underwent the anterior-posterior fusion (3 cases), one stage posterior osteotomy with short segment fusion (2 cases), growing rod instrumentation (8 cases) and those who were with less than 2-year follow-up information (3 cases) were excluded. 10 NF–1 patients with dystrophic EOS (aged at 7.8±2.1 years，four males and six females) were included in this study finally.

Data collection

General clinical data including age at initial surgery, sex, BMI, neurology statements (ASIA) and follow-up duration were collected. Collected surgery related data included the number of involved surgical segments, the type of anchor instrumentation (hook, screw or hybrid), instrumentation intensity, diameters of the rod, bone grafting strategy (material and location), intraoperative neurophysiology monitor, operation time, blood loose and blood transfusion.

Radiographic Features

The radiographic data included the major coronal curve, the sagittal curve of T2-T5, T5-T12 and T10-L2, the lengths of T1-S1 and T1-T12 and space available for the lung (SAL). The proximal and distal junctional kyphosis were measured at the time of post-operation and final follow-up. 8 cases have been found dural ectasia on preoperative MR image and 2 cases had paraspinal tumors or plexiform neurofibromas located close to the scoliotic curve. The dystrophic changes were reevaluated at the last follow-up through radiographing.

Pulmonary function

Every patient underwent preoperative pulmonary function testing and preoperative FEV1 and FVC were compared with the FEV1 and FVC at the last follow-up. Percent- predicted values were derived from Stanojevic equations.

Surgical Procedures

The preoperative traction was not used. All patients were preferred to be treated with screw-based instrumentation. In order to avoid the limitation of height development and abnormal lung function caused by the fusion of too many segments, we refer to the fusion range of adolescent idiopathic scoliosis. The range of spinal fusion is from the upper end vertebra to the lower stable vertebra. When the pedicle was not large enough or failed in fixation, hybrid with the hook was also used. Correction of deformity was performed by a combination of rod derotation and sequential *in situ* translational reduction, with or without *in situ* bending of the rod.

The curve contained 4.8 (4 to 5) segments on average; the mean fusion segment number was 8 (4 to 13). Implant density means ratio of the total number of anchor points of the internal fixation to the number of fusion segments. In our study, t

the average implant density was 1.2 (0.9 to 1.6). Three patients were operated with hybrids of pedicle screws and hooks, among them two patients were operated with two hooks and one patient with one hook. Five patients used the connector, among them four patients with one connector and one patient with two connectors. All patients used titanium rods. The diameter of the rods used was 5.5mm except for two cases in which the rod diameter was 4.5mm.

Additional correction maneuvers, including appropriate compression and distraction, were performed to provide 3D correction of the deformity. The posterior elements were decorticated, and bone grafts were placed on the decorticated bed using autogenous local bone grafts in combination with allogeneic bone grafts.

Mean blood loss was 580 ml (200–2000ml). Three patients were given transfusions of 506 ml on average. The two patients who have the most blood loss in our observation received the concave side para-spinal tumor resection during the procedure. Hard braces were used after surgeries for 6–8 months in all cases.

Statistical Analysis

Statistical analyses were performed with SPSS (IBM, New York, USA), version 22.0 for Windows. Independent samples t-test was performed when comparing two groups; if the variances were not equal, the Wilcoxon test was applied. One-Way ANOVA was used to compare the means between three groups.

Results were presented as mean (±SD), unless otherwise indicated. The differences were considered significant when $P<0.05$; P values between 0.05 and 0.10 were considered the difference in trends.

Results

Significant coronal curve correction but poor maintenance

Ten NF–1 patients (four males, six females) with dystrophic scoliosis were included in this study. Detailed characteristics of each patient were described in Table 1. The mean age of patients at the initial surgery was 7.8 years (4.2 to 9.6 years). The average follows up duration was 54 months (24 to 88 months). There were 9 cases of thoracic scoliosis and one case of lumbar scoliosis.

Table 1. Clinical and radiographic data of 10 NF–1 patients with early-onset Scoliosis treated by posterior only fusion

The major curve was significantly corrected from 66.1±16.22 degrees on average (range: 43 to 90.3 degrees) to 31.1±14.6 degrees on average (range: 13.4 to 51.2 degrees) (P = 0.00) after the initial surgery. However，the average major curve at the final follow-up increased significantly to 40.95±16.01 degrees (range: 17 to 70.3 degrees) compared to the initial result (P = 0.001). The coronal curve correction rate was 53.8±14.66% after the initial correction, which fell to 38.9±15% at the last follow-up. The difference was statistically significant (P = 0.001). The mean rotation of apex vertebrate was corrected from 2 (range: 1 to 3) preoperatively to 1.8 (range: 1 to 3) after initial surgery and was maintained at 1.9 (range: 1 to 3) at the last follow-up.

In one case, the preoperative T5-T12 kyphosis angle was not recognizable because of the low quality of the film. After analyzing all the available data, it was concluded that the change of T5-T12 kyphosis angle from pre-operation (42.9±21.96 degrees) to post initial operation (28.08±11.58 degrees) was not statistically significant (P = 0.08). This was also the case for the change of T5-T12 kyphosis angle from initial post operation to the last follow up (follow-up

T5–12, 34.55±14.74，P = 0.09). There was no significant difference between the average values of preoperative, initial postoperative and the last follow-up T1-S1 kyphosis angle.

Continued spinal growth during the growth phase

The T1-S1 length after the operation increased by 2.77±1.0 cm on average. During the follow-up, the T1-S1 length increased at 0.6±0.30 cm per year. The mean T1-T12 distance was increased from 21.4±3.29 cm to 23.3±2.96 cm, and the lengthening velocity was 0.4±0.46cm per year. The T1–12 growth velocity was significantly inhibited by fusion operation compared to that of the T1-S1 (P = 0.007). SAL changed from 0.987 (range: 0.854 to 1.145) to 1.008 (range: 0.932 to 1.16) and was maintained at 1.035 (range: 0.961 to 1.16).

No neurological complication but high alignment complication

Three cases had perioperative complications on records (1 urinary tract infection, one ileus, 1 wound superficial infection) (Table 2).

Table 2. Clinical data and surgical information on 10 NF–1 Patients with Early-onset Scoliosis Treated by posterior only fusion

During the follow-up, the incidence of the alignment complication was relatively high. The fusion block of 7 patients deteriorated more than 10 degrees. Besides the two existing cases, there was a new case of lumbar curve development in the third case. In terms of sagittal alignment, the thoracic-lumbar kyphosis of one case deteriorated over 20 degrees. (Fig. 1).

Figure 1. Case 6: A. B. Preoperation. C. D. Postoperation. E. F. The adding on phenomenon is obvious at the 5-year follow-up, which is attributed to at the growth of the anterior column of the fusion segments

However, not all the patients who had got the alignment problem suffered the revision operation. In one case, the patient had the severe thoracic-lumbar kyphosis deterioration, and the rod in convex side slipped from the distal screw track got a revision surgery by putting the rod back to the screw track, replacing the cap, adding a trans-connector and augmenting the distal fusion area with allograft. (Fig. 2). In another case, the two proximal anchor screws of the convex side were pulled out, the instrument of this side was taken off, and the fusion range was extended proximally.

Figure 2. Case 2: A. B. Preoperation. C. D. Postoperation. E. F. The patient had severe thoracic-lumbar kyphosis deterioration at the 5-year follow-up, and his rod on the convex side slipped from the distal screw track. The posterior fusion only was hard to restrict the growth ability of the fusion segments even used the pedicle screws.

Except for the two cases mentioned above, there was another case in which a patient had one proximal screw dislodged radiographically. As there was no symptom, the revision operation was not taken.

There was no neurological complication (transient or permanent neurological deficiency) observed.

Pulmonary Functions

The FVC% changed from 91.23±16.07% to 95.29±11.40% at the last follow-up without statistical difference (P = 0.057). The forced expiratory volume in one second (FEV1) was not significantly improved at the last follow-up (preoperative 93.0±13.09%, postoperative 97.48±9.48%, P = 0.405) (Table 3).

Table 3. Summary of clinical data, body height and pulmonary function on 10 NF–1 patients with early-onset scoliosis treated by posterior only fusion or traditional growing rods

Discussion

In the present study, we evaluated the mid-long term (average 4.5 years follow-up) clinical outcome after posterior only instrumented fusion surgery for NF–1 patients with dystrophic EOS. The mean preoperative major curve was corrected from 66.1±16.22 degrees (range: 43 to 90.3 degrees) to 31.1±14.6 degrees (range: 13.4 to 51.2 degrees) (P = 0.00). However, the average major curve at the final follow-up fell back to 40.95±16.01 degrees (range:17 to 70.3 degrees). The T1-S1 length increased by 2.77±1.0 cm on average after surgery and increased at a speed of 0.6±0.30 cm per year. However, the incidence of the alignment complication was relatively high during the follow-up.

About 10% of children with NF–1 develop scoliosis that predominantly involves the cervical and thoracic spine.[13] Dystrophic scoliosis with NF–1 has a high risk of rapid progression.(5) The progression of the dystrophic curve can be neither stopped nor relieved by corset therapy.(10) Therefore, the aggressive surgical treatment of dystrophic scoliosis in NF–1 patients was widely recommended.[3,15]

Traditionally, the combined anterior and posterior fusion was supported by most authors and was recognized as the most reliable method.[16–18] The clinical outcome of dystrophic scoliosis patients treated with posterior-only fusion by using hooks and rods demonstrated that the pseudarthrosis rate was high and curve progression was common.(11) Recently, some good results of posterior only fusion surgeries in NF–1 patients with dystrophic scoliosis have been reported.[9,10, 19] In this study, we reported our results about the posterior only fusion procedure in NF–1 patients with EOS.

In addition to our study, there are currently only two articles specifically described the effect of fusion procedure to treat EOS in NF–1 patients Greggi et al.(12)reported that NF–1 EOS patients underwent posterior fusion if the thoracic kyphosis was less than 50°, or underwent anterior-posterior spinal fusion surgery if the thoracic kyphosis was 50° or more. In their study, the correction rate on average was 60%, slightly higher than our results. And the follow-up results were good as well, showing no significant progress. Ryoji Tauchi and his colleagues applied the anterior-posterior fusion surgery techniques to all EOS patients before their age reached 10 and the orthopedic effect was more noticeable. The main curve was corrected from 71.2° to 24.1° (66.15%). After a follow-up averaged on 14 years, the result showed that there was no significant progress in scoliosis. Their correction rate was superior to our study in both groups, which might be related to better anterior release due to the combination of the anterior and posterior spinal surgery. Ryoji Tauchi also performed tumor resection at the anterior concave side in combination with the anterior and posterior approach, rib support and bone graft, accounting for the possible reason for the fact that his surgery maintained good correction rates at follow-up(13).

In contrast to Greggi's criteria for grouping patients with kyphotic angles, we used posterior orthopedic fusion procedures for all patients and did not separate the patients by degree of kyphosis or deformity disposal. In our patients with a kyphotic angle greater than 50°, the initial postoperative correction rate was 44.5%, the last follow-up correction rate was 31.1%, and the loss rate was 13.4%. In patients with a kyphotic angle of less than 50°, the initial postoperative correction rate was 67.5%, the last follow-up correction rate was 43.8%, and the loss rate was 23.7%. Patients with a more than 50° kyphotic angle had both a lower initial correction rate and a lower final follow-up correction rate than those patients who had a lower kyphotic angle of 50° or less. But the two groups were similar in terms of subsequent orthopedic maintenance. Compared with the anterior and posterior fusion, simple posterior only orthopedic surgery may not be able to satisfy orthopedic maintenance in NF–1 patients with EOS, while simple posterior orthopedic fusion in patients with larger kyphosis may not even meet the needs of initial orthopedic surgery.

Five years of clinical follow-up showed that crankshaft occurred in 6 of 10 patients and the Cobb's angle of fusion segments increased by 10°. By group comparison, we also found that the incidence of the crankshaft was significantly higher for children who were under seven years of age than that of patients who were 7 to 10 years old. Using a single posterior approach to orthopedic fusion surgery was at very high risk of the crankshaft, which was consistent with the previous non-surgical observation of such patients(2).

In addition, some scholars have found that higher density of pedicle screw placement can help to improve the postoperative correction rate of EOS in NF–1patients(9, 14). The lack of O-arm at the early stage of our study, at that time, we can only apply ordinary fluoroscopy. Which made it difficult to place the pedicle screws. The average ratio of fixed/surgical segments was 0.69. However, we found that there were three patients whose fixed/surgical segment ratio was above the average, reaching 63.91% on average. The correction rate at the last follow-up was 48.47%, and the rate of loss was 15.44% for these three patients. For the other patients, the average postoperative correction rate was 49.4 %, the final follow-up correction rate was 34.78%, and the correction loss rate was 14.62%. Perhaps the insufficiency of screws placed on the spine resulted in the insufficiency of the correction force, which led to the poor initial orthopedic effect on the patients included in the current study. However, lacking enough screws did not significantly affect the result of the surgery in terms of the prevention of scoliosis progress.

It is generally accepted that scoliosis caused by NF–1 is relatively stiff and the preoperative traction may contribute to the improvement of orthopedic effect. However, no specific study on this problem has been published yet. Tauchi et al. had given HALO traction to scoliosis patients with a Cobb degree of 80° or more, while Greggi did not use preoperative traction(12). For most patients with severe scoliosis, preoperative traction can improve the orthopedic effect, but it also increases the medical costs and the distress of patients. And poor compliance was found in patients with EOS due to their young age. Therefore, further evidence is needed to conclude the effect of preoperative traction for EOS in NF–1patients.

In Tauchi and coworkers' study, augmentation and bone graft surgery could maintain good orthopedic effect if dura mater or tumor growth were developed[20]. Follow-up data demonstrated that patients received an average number of 1.5 intensive procedures. This may also be a reasonable and applicable way to maintain the good orthopedic effect. In our study, patients did not undergo an MRI assessment during follow-up duration, so it is not clear if the progress of scoliosis, was related to an intraspinal deformity. Based on the published knowledge, it may be necessary to be examined in long-term follow-up for NF–1 patients with EOS.

The average fusion segments we performed was 8.1 while it was 13.1 in Tauchi's study[20]. It suggested that fewer fusion segments in our study led to the poor orthopedic maintenance. In our study, the majority of spine fusion ranges were from upper end vertebrae to lower stable vertebrae, which was adequate for patients with AIS and NF–1 patients with non-dystrophic scoliosis, but insufficient for NF–1 patients with dystrophic EOS. For EOS patients, height retention is one of the factors we must take into account, because maintaining spinal length is critical factor in allowing adequate lung development, which is a major goal of EOS treatment.The average length of T1-S1 in our patients was 34.5 cm and 39.8 cm preoperatively and at the last follow-up, respectively. There was a 5.3 cm growth. The length of T1-T12 was 21.4cm preoperatively and 24.5 cm at the final follow-up, resulting in a 3.1 cm growth. In Tauchi's study, the average preoperative and final follow-up of T1-S1 were 30.7 cm and 36.2 cm respectively, resulting in a 5.5 cm growth. The preoperative and final follow-up of T1-T12 length was 18.8 cm and 21.9 cm, respectively, resulting in a growth of 3.1cm. Compared to Tauchi's study, our surgery was not superior in preserving trunk length. This might be due to the shorter follow-up time, 4.5 years in our study and 14 years in Tauchi's study, and the fact that patients in our study have the potential to continue their torso growth.

In this study, there was no significant improvement in lung function relative to preoperative lung function at the patient's last follow-up, probably because the patient's preoperative lung function impairment was not significantly associated. On the other hand, the early fusion of the spine did not cause damage to the patient's lung function, which may be due to the shorter fusion segment we chose.

In this study, higher complication rates were associated with posterior-only fusion surgery during the perioperative period and the follow-up, such as crankshaft phenomenon, scoliosis progress, and adding-on phenomenon. A possible reason for these complications may be the overload of the posterior fixation system. Internal fixation complication that occurred at the interface between the implant and the bone was associated with the presence of osteoporosis in neurofibromatosis. There was no neurological complication (transient or permanent neurological deficiency) observed. Considering the facts that the complication rate of anterior-posterior approach is as high as 64%, the probability of perioperative pulmonary dysplasia is as high as 45.4% and lung injury and dural tear are also seen [20], we suggest that posterior fusion surgery may be a good alternative way to get a better postoperative outcome and reduce the postoperative complications.

Until now, none of the current approaches to treat EOS in NF–1 patients has found a balance between maintaining good orthopedic outcomes and reducing complications and patient distress. Will non-fusion technology be the new direction for the treatment of NF–1patients with EOS. Jain et al. (15)used the growth-bar technique on 14 NF–1 patients with EOS in 5 centers and performed an average follow-up of 54 months. It was shown that the correction rate at the final follow-up was 51% and the annual spine length increase was 1.1 cm. Considering the correction rate and retaining of the space to grow, it is undoubtedly worth the wait. Of course, there are disadvantages of growth rod technology as well such as the high incidence of complications associated with internal fixation. Although further observation on non-fusion technology to treat NF–1 patients with EOS is needed, it is undoubtedly that this is another promising new choice to treat the disease.

Conclusions

We studied a group of EOS in NF–1 patients who had undergone simple posterior orthopedic internal fixation fusion surgery at Peking Union Medical College Hospital for an average follow-up of 4.5 years and found that the orthopedic effect of posterior only fusion method was not good way to go, especially for the maintenance of orthopedic effect. For the surgical treatment of NF–1patients with EOS, the ideal goal of treatments should be to maintain the patients' height as much as possible while achieving good orthosis and its maintenance in patients, reducing the risk of surgery and complications at the same time.

Declarations

Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this article

Abbreviations:

EOS: Early-Onset Scoliosis

NF–1: Neurofibromatosis type 1

FEV1: The forced expiratory volume in one second

PF: posterior only fusion

GR: traditional growing rod

FU: Follow-up

Pre-O: Preoperative

Post-O: postoperative

MC: Major curve

CP: crankshaft phenomenon

PI: Post the Initial surgery

The forced expiratory volume in one second

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Tables

Table 1. Clinical and radiographic data of 10 NF-1 patients with early-onset Scoliosis treated by posterior only fusion

No	SEX	Age	FU（Mon）	MC	Pre-O MC(cobb)°	Post-O MC(cobb)°（%）	FU MC(cobb)°（%）	Pre-O T5-12 kyphosis (cobb)°	Post-O T5-12 kyphosis (cobb)°(%)	FU T5-12 kyphosis (cobb)°(%)	Pre-O T12-S1 Lordosis (cobb)°	Post-O T12-S1 Lordosis (cobb)°(%)	FU T12-L1 Lordosis(cobb)°(%)	Pre-O T10-L2 Lordosis (cobb)°	Post-O T10-L2 Lordosis (cobb)°(%)	FU T10-L2 Lordosis(cobb)°(%)	Pre-O T1S1（cm）	Post-O T1S1 (cm)	FU T1S1 (cm)	T1S1 gain/year(cm)	Pre-O T1-12（cm）	Post-O T1-12(cm)	FU T1-12(cm)	T1 gain/y
1	F	9.1	24	L1-4	51.5	25(51%)	41.2(20%)	16.5	41.8(-153%)	39.4(-139%)	58.0	66.6(-15%)	59.8(-3%)	-5.0	5.8(216%)	5(200%)	33.6	35.1	37.9	1.4	22.1	22.2	25.4	[illegible]
2	M	7.2	61	T10-L2	70.0	21(70%)	39(44%)	22.9	27.6(-21%)	14.2(38%)	52.7	45.8(13%)	40.6(23%)	2.9	1.3(55)	12.5(-331%)	30.2	31.8	34.1	0.4	19.6	20.3	22.1	[illegible]
3	M	8.3	88	T5-9	88.0	55(38%)	70.3(20%)	47.7	32.3(32%)	38.5(19%)	49.9	17.5(65%)	43.1(14%)	9.2	15.3(-66%)	1.9(79%)	36.1	39.5	43.2	0.5	22.1	24.7	25.6	[illegible]
4	F	4.2	35	T6-9	43.0	27.1(37%)	27.4(36%)	60.8	24.8(59%)	34.9(43%)	48.4	35.5(27%)	58.2(-20%)	24.1	7.5(69%)	10.9(55%)	38.7	41.9	43.9	0.7	24.9	26.9	27.8	[illegible]
5	M	7.1	49	T5-9	83.3	45.3(46%)	57.7(31%)	70.5	35(50%)	62.7(11%)	66.0	22.6(66%)	66.2(0%)	3.1	12.7(-310%)	4.2(-35%)	36.0	39.5	41.9	0.6	20.4	23.8	24.4	[illegible]
6	M	4.1	34	T7-11	60.9	30.8(49%)	41.4(32%)	26.0	20(23%)	30.9(-19%)	47.6	46.8(2%)	54.2(-14%)	1.0	1.5(-50%)	6.5(-550%)	26.7	29.0	31.4	0.8	16.7	18.4	19.7	[illegible]
7	F	9.2	67	T6-10	59.6	13.4(78%)	27.2(54%)		6.2	20.6	52.5	44.7(15%)	62.7(-19%)	23.0	5.3(77%)	8(65%)	42.5	44.6	46.7	0.4	26.8	27.4	29.6	[illegible]
8	F	9.5	68	T5-8	90.3	51.2(43%)	54(40%)	56.8	46.3(18%)	50.7(11%)	46.4	43.5(6%)	45.6(2%)	8.0	6.5(19%)	7.4(7.5%)	30.8	34.3	36.4	0.4	16.7	20.4	20.6	[illegible]
9	F	9.6	62	T5-9	58.9	27.4(53%)	34.3(42%)	66.5	26.8(60%)	34.6(48%)	65.7	39(41%)	43(35%)	6.7	2(70%)	18.4(-175%)	32.4	37.0	39.4	0.5	20.5	23.4	24.0	[illegible]
10	F	9.6	52	T8-12	55.0	15(73%)	17(69%)	18.0	20(-11%)	19(-1%)	30.5	43.8(-44%)	44.7(-47%)	9.2	0.9(90%)	4.9(47%)	38.0	40.0	43.2	0.7	24.0	25.1	25.6	[illegible]

PF: posterior only fusion; GR: traditional growing rod; FU: Follow-up; Pre-O: Preoperative; Post-O: postoperative; MC: Major curve;

Table 2.

Clinical data and surgical information on 10 NF-1 Patients with Early-onset Scoliosis Treated by posterior only fusion

No	Fusion or involved level	Anchor sites (Hooks)	Anchor sites nearby the apex level (upper 1/lower 1)	Rod diameter (mm)	Intraoperative blood lost(mm)	Transconnector (numbers)	Complications		
							Perioperative complication	Alighment complication	Implant related
1	T11-L5	6(2)	N	5.5	500	1		the curve deteriorated to 37.6°at 2 years FU and worsen continuously	
2	T9-L3	11	Y	5	400	0		the curve deteriorated to 55°at 5 years FU and the thoracilumbar kyphosis deteriorated from 7.8°to 27°	1 distal screw slipped at 5 years FU
3	T3-12	10(4)	N	5.5	500	1	urinal infection	CP, MC increased 15.3°	
4	T6-9	4	N	4.5	400	1		decompensated lumbar curve.	1 screw dislodgement
5	T4-11	8	N	5.5	400	1	intestinal paralysis	CP, MC increased 13.4°	2 screws pulled out
6	T7-11	9	Y	4.5	400	0		CP, MC increased 11.4°, decompensated lumbar curve.	
7	T5-11	8	N	5.5	300	0		CP, MC increased 13.8°, decompensated lumbar curve.	
8	T2-L1	12(2)	N	5.5	3000	2			
9	T2-L2	15	N	5.5	700	0			
10	T6-L1	11	Y	5.5	400	0	superficial infection		

CP: crankshaft phenomenon; MC: Major Curve; Postop: postoperative; PI: Post the Initial surgery

Table 3.

umary of clinical data, body height and pulmonary function on 10 NF-1 patients with early-onset scoliosis treated by posterior only fusion or traditional growing rods

No	SEX	Age	FU（Mon）	Pre-Height(cm)	FU-Height(cm)	Pre-O T1S1（cm）	FU T1S1 (cm)	Pre-% FVC (Present/ Predicted)	FU-% FVC (Present/ Predicted)	Pre-% FEV1 (Present/ Predicted)	FU-% FEV1 (Present/ Predicted)	Pre-ASIA score	FU-ASIA score
1	F	9.1	24	126	149	33.6	37.9	85.4	91.2	106.3	105.0	112	112
2	M	7.2	61	110	136.5	30.2	34.1	87.5	90.0	104.2	91.4	112	112
3	M	8.3	88	120	148.2	36.1	43.2	98.9	99.1	103.1	104.2	112	112
4	F	4.2	35	85	105.0	38.7	43.9	77.0	90.2	81.2	92.5	108	110
5	M	7.1	49	118	147.1	36.0	41.9	90.6	91.6	82.0	86.4	112	112
6	M	4.1	34	120	140.5	26.7	31.4	121.2	120.4	108.3	107.5	112	112
7	F	9.2	67	131	152.6	42.5	46.7	88.4	90.4	87.4	92.1	112	112
8	F	9.5	68	147	166.9	30.8	36.4	62.0	77.2	69.0	76.4	107	107
9	F	9.6	62	134	163.0	32.4	39.4	106.9	104.1	98.3	99.1	112	112
10	F	9.6	52	142	167.4	38.0	43.2	94.4	98.7	90.2	93.1	112	112

FU: Follow-up; Pre-O: Preoperative; Post-O: postoperative;

Figures

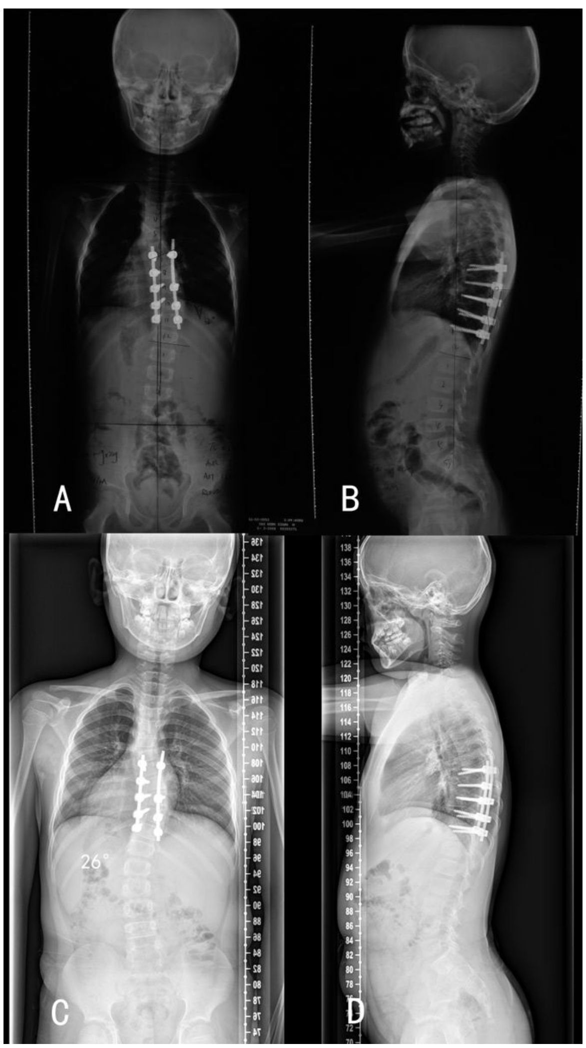

Figure 1

Case 6: A.B. Preoperation. C.D. Postoperation. E.F. The adding on phenomenon is obvious at the 5-year follow-up, which is attributed to at the growth of the anterior column of the fusion segments

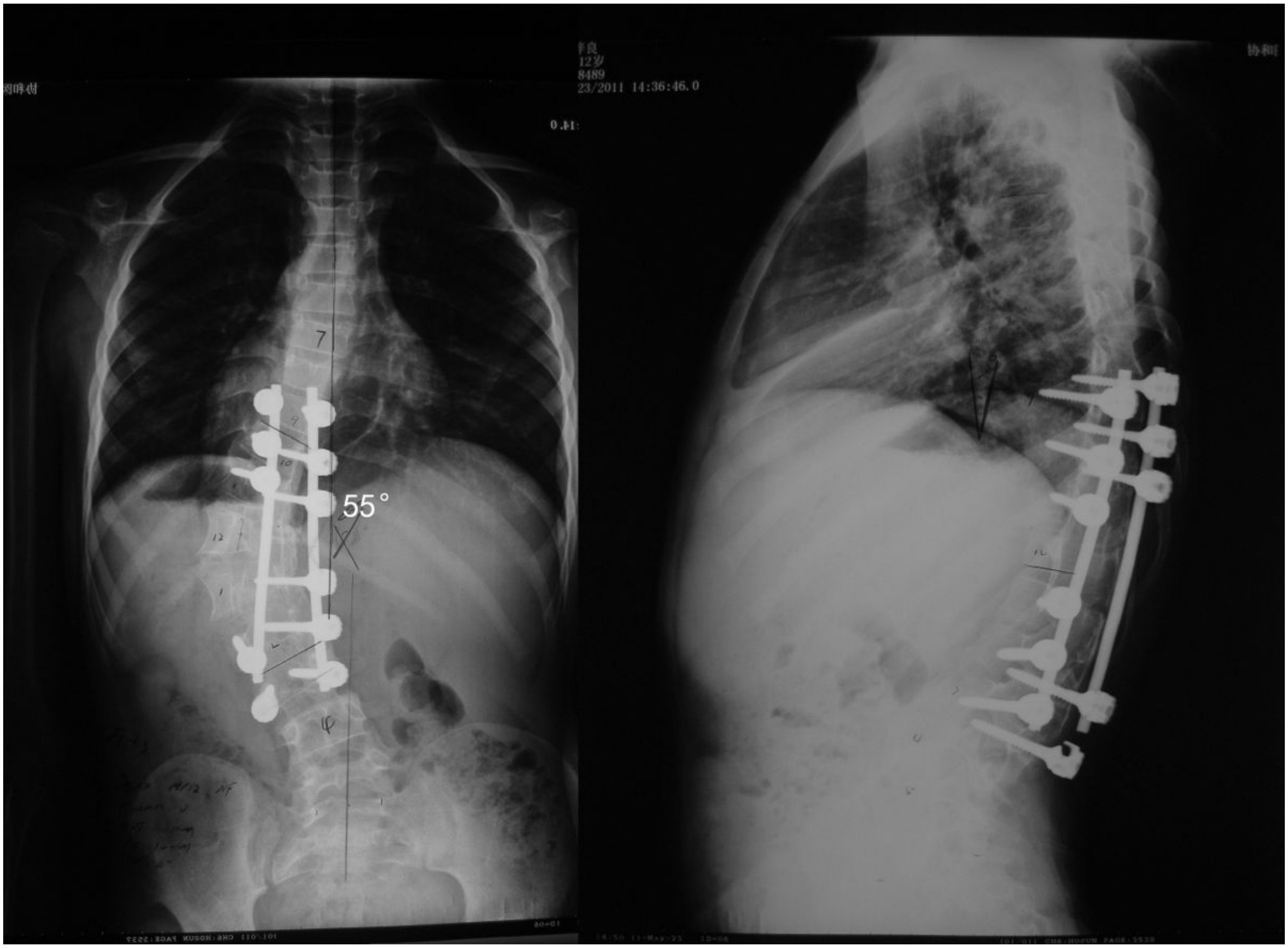

Figure 2

Case 2: A.B. Preoperation. C.D. Postoperation. E.F. The patient had severe thoracic-lumbar kyphosis deterioration at the 5-year follow-up, and his rod on the convex side slipped from the distal screw track. The posterior fusion only was hard to restrict the growth ability of the fusion segments even used the pedicle screws.