CLINICAL ARTICLE

Neurofibromatosis Type 1 with Severe Dystrophic Kyphosis: Surgical Treatment and Prognostic Analysis of 27 Patients

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Objective: The aim of the present study was to explore the surgical treatment and prognosis of 27 cases of neurofibromatosis type 1 with severe dystrophic kyphosis.

Methods: We performed surgical treatment for scoliosis and kyphosis caused by dystrophic curves at Peking Union Medical College Hospital, Beijing, China from December 2015 to December 2017. The study included 21 patients with moderate to severe kyphosis, 12 males and 9 females, with an average age of 14.95 ± 6.05 years. All patients had kyphosis angles greater than 70° and had more than four skeletal developmental defects. A total of 6 patients with severe kyphosis, 2 males and 4 females, with an average age of 12.5 years, had more than five skeletal developmental defects with a kyphosis angle greater than 90° or a lumbar kyphosis angle greater than 40°. According to the patient's own situation, we adopted a low-grade surgery scheme (grades 1 or 2) or a high-grade surgery scheme (grades 3–6). The low-grade surgery was mainly lower articular surface resection or pontodestomy, and the high-grade surgery was mainly apical vertebral body or upper discectomy. All patients were followed up to determine their prognosis.

Results: Statistical analysis showed that there was a significant difference in preoperative and postoperative scores between the two groups ($P < 0.05$), and scoliosis correction showed that surgical treatment had a significant effect on scoliosis kyphosis. The mean follow-up time was 66.7 months. Follow-up results showed that 50% of complications after internal fixation were related to high-level surgery. Complications included displacement of the titanium cage, removal of the lamina hook, formation of pseudoarthrosis, and internal fixation failure (with a rate of 7.7%–14.3%). In contrast, there were no associated symptoms for low-grade surgery. In addition, the results showed that gender, age, extent of resection, height, and body mass index had no significant effect on preoperative, postoperative, and prognostic indicators of patients ($P > 0.05$).

Conclusion: Early identification of dysplastic scoliosis-related deformities plays an important role in surgical planning and prognosis, and low-level surgical procedures are more favorable for patients' prognosis.

Key words: Dystrophic curves; Neurofibromatosis type 1; Spine deformity

Introduction

Neurofibromatosis (NF) is the most common single-gene hereditary disease in humans[1]. It is a disease of the crest cells and may originate from vascular diseases involving the neuroectoderm, mesoderm, and endoderm[2]. Its clinical manifestations are common in neurofibroma and schwannoma. It may occur in any organ system of the body, mainly involving the skeleton, skin, and soft tissue. NF has two different clinical forms: NF1 and NF2[2]. The gene mutation mechanisms of NF1, NF2, and NF are presented in diagrams in Figs 1, 2, 3, and the process of NF spine changes is shown in Fig. 4. NF1 is the most common form of NF. It has a wide range of effects and can be seen in all races. Globally, the incidence of NF1 is approximately 1/3000–1/4000[2–5]. The heredity of the disease is autosomal

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Orthopaedic Surgery

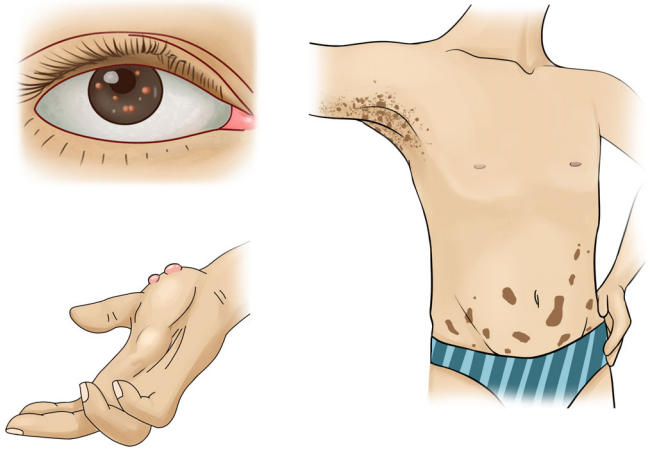

Fig. 1 The gene mutation mechanism of neurofibromatosis type 1.

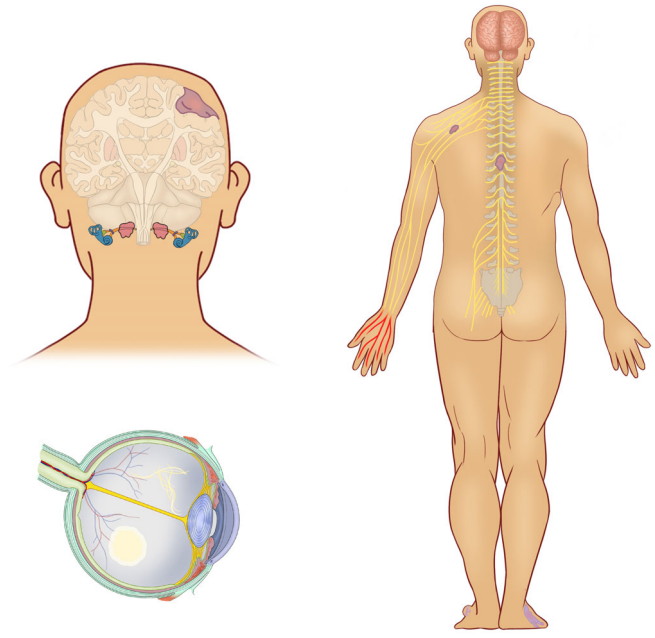

Fig. 2 The gene mutation mechanism of neurofibromatosis type 2.

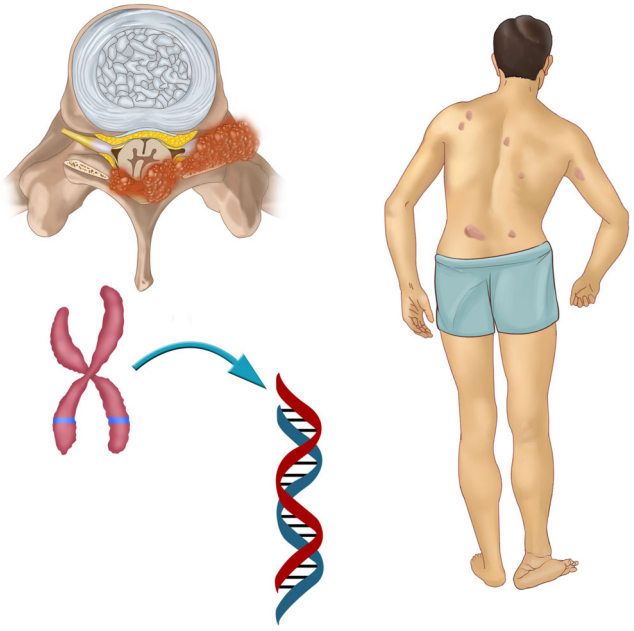

Fig. 3 The gene mutation mechanism of neurofibromatosis.

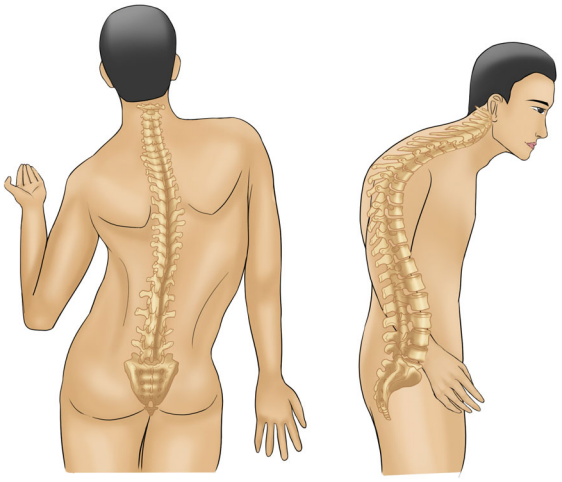

Fig. 4 The process of neurofibromatosis spine changes.

dominant inheritance, with a high exogenous rate. However, approximately 50% of cases are caused by *de novo* mutations[6–8].

Neurofibromatosis type 1 is located in the 17q 11.2 region of the long arm of human chromosome 17 and was cloned in 1990[9–12]. The gene, which spans approximately 280 kb, contains 57 constitutive exons and four alternative splicing exons. It can encode a length of 11–13 kb mRNA, including a 3′ terminus 3.5-kb untranslated region[13]. The formation of neurofibroma in NF1 patients is related to the loss of expression of NF1. Because *NF1* is a large gene with a span of 350 kb, its complexity reflects many genetic rules.

The spontaneous mutation rate of *NF1* is 1×10^{-4}. It is one of the most frequent gene loci in humans and the spontaneous mutation rate is approximately 100 times higher than that of single gene hereditary diseases. *NF1* mutations include chromosome aberrations, multiple exon deletions, insertions, termination mutations, amino acid substitutions, intron mutations, and 3′-untranslated region mutations. Loss of neurofibromin expression leads to tumorigenesis and development of NF1-related tumors, including malignant peripheral nerve sheath tumors, pheochromocytomas, malignant myeloid dysplasia, and benign neurofibromas[14]. It has

been found that 90% of the new mutations in sporadic cases occur on the chromosomes of paternal origin[15].

Coronal spinal decompensation in NF is generally classified into non-dystrophic and dystrophic types. The prognosis and treatment of columnar scoliosis curves largely depends on the presence of dystrophic characteristics. The most common manifestation of scoliosis is seen in the thoracic region, which presents heterogeneous changes. The imaging features of the non-dystrophic type include scallop-like degeneration of the vertebral body, pencil-like ribs, severe apical rotation, enlarged intervertebral foramen, and dysplasia of the vertebral arch (Fig. 5). As a general rule, the more severe the dystrophic changes identified in the vertebral bodies, the higher the likelihood that the scoliotic curvature will deteriorate. Studies on the evolution of NF1 spinal deformity have shown that when three or more developmental abnormalities occur, the risk of curve progression is significantly increased in 85% of patients[16,17].

There are different surgical options for non-dystrophic scoliosis kyphosis and dystrophic scoliosis kyphosis depending on the type of patient. For instance, on-dystrophic scoliosis is treated with a posterior pedicle screw system for three-dimensional correction and bone graft fusion. The posterior pedicle screw system is also used for three-dimensional correction and bone graft fusion. Posterior apical osteotomy and pedicle screw internal fixation are used if kyphosis deformity is rigid. Anterior paravertebral neurofibroma resection, bone grafting in the apical region, and post-stage fixation and orthopaedic surgery are used in cases of deformity with paravertebral neurofibroma.

Studies have report results regarding surgical treatment and the prognosis of scoliosis and kyphosis. Jochen *et al.* reported 53 cases of scoliosis correction using video-assisted thoracoscopic surgery. The Cobb angle of the thoracic curve before surgery was 69.5° on average, and the average correction rate was 68%, but the incidence of pseudoarthrosis after thoracoscopic surgery was higher[18]. Betz *et al.* studied the safety and feasibility of 58 adolescent congenital scoliosis patients using the U-nail technique to correct scoliosis. They used a thoracoscopic technique to place a U-nail on the convex side of the technique to correct scoliosis. After a follow-up period of more than 1 year, they found that the Cobb angle was 35° preoperatively and 37° postoperatively, with complications including hemothorax, chylothorax, and postoperative pancreatitis[19]. At present, there is controversy at home and abroad about the use of U-nail technology to control the treatment of scoliosis, particularly given the lack of long-term follow up. Multivertebral wedge osteotomy has achieved good clinical results in the treatment of severe scoliosis kyphosis. Guille *et al.*[20] reported on 14 patients who underwent wedge osteotomy of the vertebral body. All 14 had successful surgeries, with an average correction rate of 86%. Maruyama *et al.* reported follow-up data on the treatment of scoliosis by multivertebral wedge osteotomy, with an average follow-up time of 9 years. The results showed that the Cobb angle was 64° preoperatively and 48.2° postoperatively on average, with few systemic complications occurring[21]. At present, there is no mature technology for the surgical treatment of scoliosis kyphosis. Improvements in surgical treatment and reductions in complications are necessary. In addition, the surgical treatment of scoliosis kyphosis, especially that caused by NF1 in adolescence, requires long-term observation.

This experiment adopts different levels of multivertebral osteotomy surgery to treat scoliosis kyphosis. In this experiment, according to the patient's own

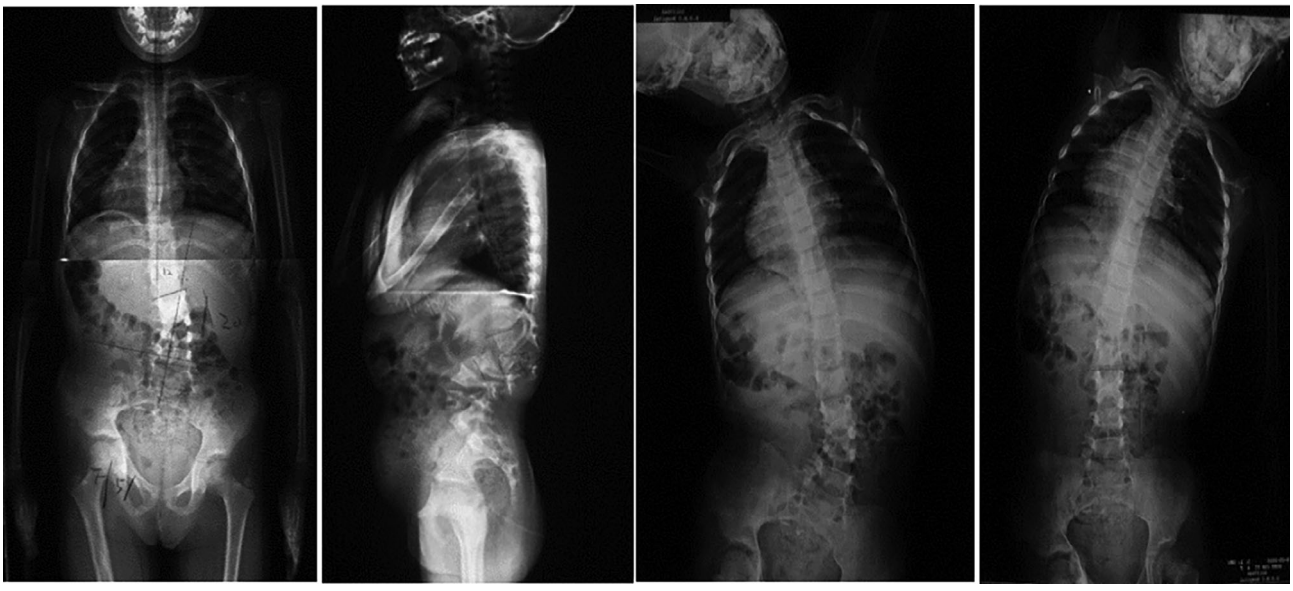

Fig. 5 Common imaging features of non-dystrophic xx.

conditions, different surgical options were adopted, including low-grade surgery (grade 1 or 2) and high-grade surgery (grade 3–6); low-grade surgery was mainly lower articular surface resection or pontine fusion, and high-grade surgery was mainly apical vertebral body or upper discectomy. In addition, the difference in preoperative and postoperative scores between low-grade surgical programs and high-grade surgical programs were compared to determine the effectiveness of scoliosis correction. Finally, according to the follow-up data, we compared the prognosis of patients who underwent low-grade surgery with that of patients who underwent high-grade surgery, and we compared the incidence of complications between the low-grade surgery group and the high-grade surgery group. The effectiveness of and differences between low-grade surgery and high-grade surgery were evaluated in the treatment of scoliosis, to determine the best surgical program for the treatment of scoliosis kyphosis.

Patients and Methods

Subjects

This study was approved by the Ethics Committee of Peking Union Medical College Hospital, Beijing, China (Approval No. ZS-952). Twenty-seven NF1 patients with malnutrition curve were collected.

Inclusion Criteria and Exclusion Criteria

The inclusion criteria were as follows: (i) patients younger than 20 years; (ii) patients diagnosed with idiopathic scoliosis and kyphosis, and undergoing orthopaedic surgery for idiopathic scoliosis; (iii) the kyphosis angle was greater than 60°, or a complete follow-up record for lumbar kyphosis was available; and (vi) patients agreed to this study and provided informed consent.

Exclusive criteria were: (i) patients over 18 years old; (ii) patients who had other spinal diseases or had a previous history of spinal surgery; (iii) patients who were followed up for less than 1 year; and (iv) patients who did not provide informed consent.

Patients and Typical Cases

Twenty-one patients with moderate or severe kyphosis had a kyphosis angle greater than 70°; these cases were accompanied by more than four kinds of skeletal development defects. Six patients with severe kyphosis had a kyphosis angle greater than 90° or a lumbar kyphosis angle greater than 40°; these patients had more than five kinds of skeletal development defects. An 18-year-old patient had scoliosis of 110° and kyphosis of 70°, with visible paravertebral tumors (Fig. 6). A 5-year-old female patient had scoliosis of 20° and kyphosis of 82° (Fig. 7). A 13-year-old female patient had scoliosis of 116° and kyphosis of 110° (Fig. 8).

Surgical Procedure

The operative methods varied according to the severity and nature of kyphosis in different patients. In evaluating spinal

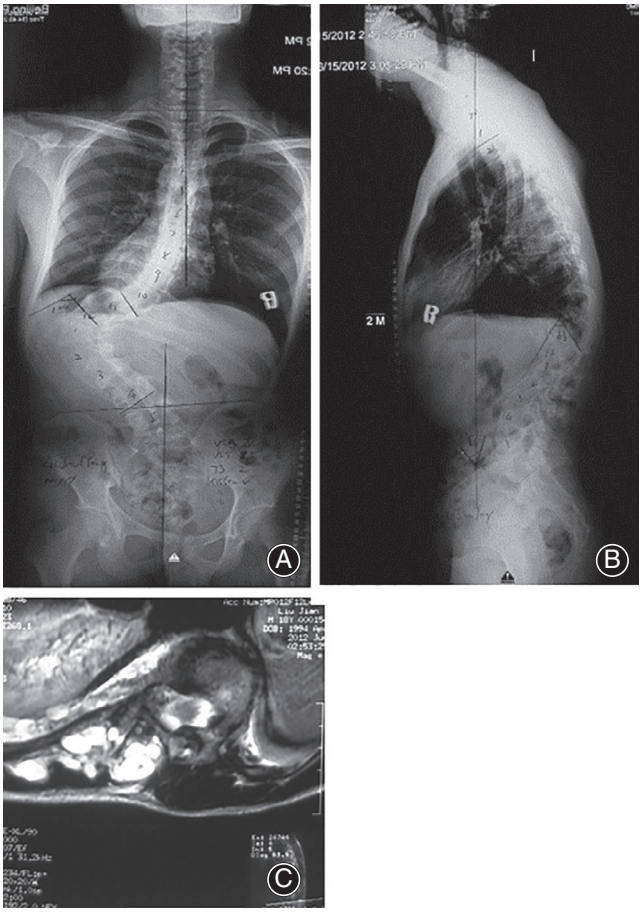

Fig. 6 Imaging picture of the patient.

deformities, in addition to the degree of kyphosis, the integrity and mobility of the anterior and posterior columns of the spine also play a key role in determining appropriate treatments. According to the different levels of operation, the specific operation was as follows:

1. General anesthesia was used in all patients. The patients were placed in prone position and the abdomen was suspended. All operations were performed by the same group of doctors.

2. The deformed segment was exposed through a median incision at a preoperatively determined surgical site, and pedicle screws were placed at the corresponding segment.

3. Deformed vertebrae were identified after full exposure, and corresponding segments of lamina were removed to fully expose the spinal cord. If an inferior facet resection or a Ponte osteotomy (grade 1 or 2) was performed, the facets of the vertebrae near the apical area were resected and the deformity was corrected by putting the rod into the screw using a cantilever. If a grade 3 or 4 resection was performed, the apical vertebrae were resected; if the trans pedicle (grade 3) or upper disc was also resected, we put the precontoured rod to the screws and corrected the kyphosis. If vertebral

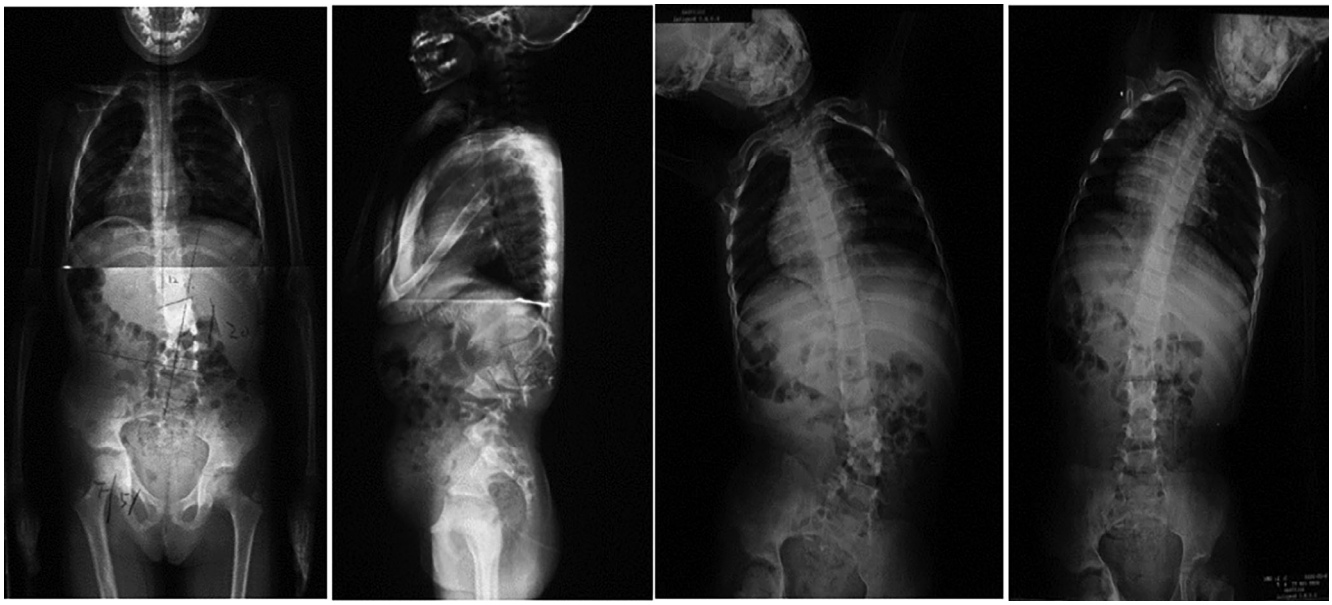

Fig. 7 Imaging pictures of a 5-year-old female patient.

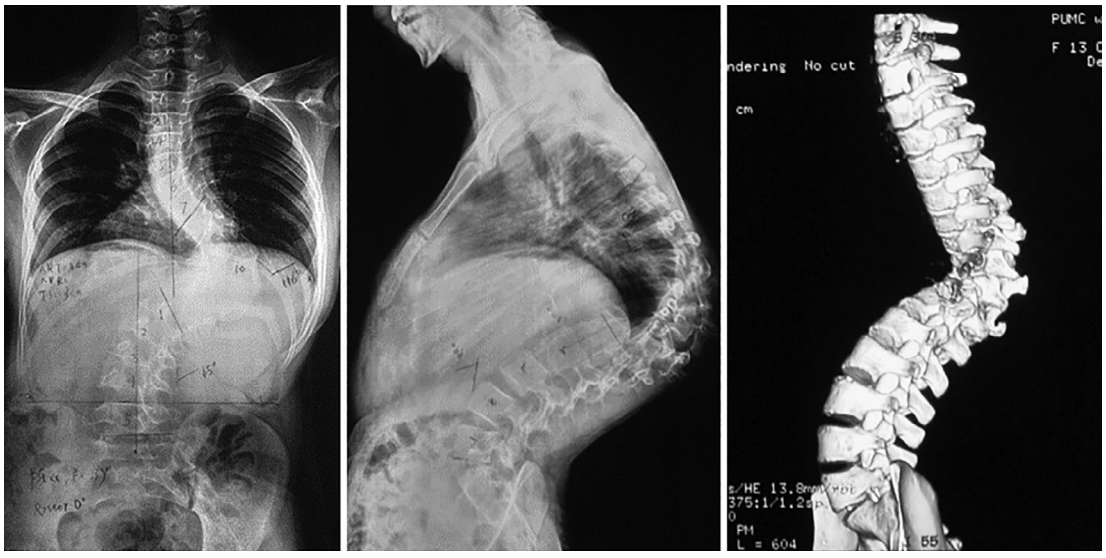

Fig. 8 Imaging picture of a 13-year-old female patient.

column resection or two-column vertebrate resection (grade 5 or 6) was performed, a relatively short rod was temporarily installed on the pedicle screw above and below more than two levels to provide stability during posterior vertebral column resection.

4. Deformity correction was gradually performed through compression and *in situ* contouring of the resected vertebral column using a precontoured permanent rod. In the maneuver, the surgeon would carefully confirm whether the central canal and the exiting nerve root were compressed, paying attention to IOM changes during correction of the deformity. The defect gap was stuffed with a titanium mesh cage or a bone strut to achieve bony fusion. After the final fixation procedure, posterior and posterolateral fusion was performed with autografts and allografts.

5. All patients received routine antibiotic therapy and neurotrophic therapy postoperatively.

Surgical Procedure in Typical Cases

The surgical procedures for 18-year-old male patients were as follows: T11–L2 was treated by facet osteotomy (grade 1) and T8–L5 by joint fixation and fusion. The treatment of 5-year-old female patients was L2 wedge vertebrectomy, mesh cage reconstruction, and T11–L5 fixation and fusion.

The 13-year-old female patient underwent T12 grade 4 osteotomy and T4–L4 fusion.

Detection of Spinal Nerve Function with Frankel Classification

Frankel classification of spinal cord injury: Class A, complete loss of sensory and motor function below the level of injury; Class B, loss of only partial sensation and voluntary movement; Class C, sensory presence, movement without practical value in the residual part; Class D, sensory and motor presence, but with manifestations and signs of nerve injury; Class E, normal.

Follow Up and Clinical Efficacy Evaluation

Postoperative outcomes and the prognosis of all patients were evaluated. All patients were followed up once a year after surgery. The average follow-up time of all patients was 66.7 months. Postoperative and prognostic follow-up-related indicators were *T1S1 distance (cm) (coronal)*, *T1S1 distance (cm) (sagittal)*, *major curve (Cobb) (coronal)*, *C7CSVL (cm) (coronal)*, *apical vertebra translation (cm)*, *kyphosis angle (Cobb)*, *sagittal vertical axis (SVA)*, and *postoperative spinal nerve function (Frankel grade)*.

T1S1 distance (cm) (Coronal) *and* T1S1 distance (cm) (Sagittal)

The average increase in T1S1 distance can be used to judge early-onset scoliosis and the effectiveness of surgical treatment. Patients with scoliosis and kyphosis should have a significantly larger T1S1 distance after surgery.

C7CSVL (cm) (Coronal)

In evaluating imaging parameters of coronal imbalance, the horizontal distance between the C7 plumb line (C7PL) and the center sacral vertical line (CSVL) is often used to evaluate the overall balance of the spine. When the vertical distance of C7PL offset CSVL line exceeds 2 cm, there is coronal imbalance.

Apical Vertebra Translation (cm)

Apical vertebra translation (*AVT*) is a common index for evaluating scoliosis deformity and is important for guiding the formulation of surgical strategies. When C7PL overlaps with CSVL, the *AVT* is the horizontal distance from the midpoint of the apical vertebra (or disc) to the CSVL of scoliosis; when C7PL does not overlap with CSVL, the *AVT* of the thoracic curve is the horizontal distance from the midpoint of the apical vertebra (or disc) to the C7PL, and the *AVT* of the thoracolumbar curve and the lumbar curve is the horizontal distance from the midpoint of the apical vertebra (or disc) to the CSVL. The *AVT* to CSLV/C7PL was negative on the left and positive on the right.

Sagittal Vertical Axis

Sagittal vertical axis (*SVA*): it is usually determined using the C7 plumb line method; that is, the vertical distance between C7PL and the upper back corner of the S1 endplate is measured. SVA > 5cm is defined as sagittal imbalance.

Major Curve (Cobb) (Coronal)

The scoliosis was measured by Cobb method when the angle was more than 10°. The significance of the common Cobb method is: negative for <10°, positive for >10°, >25° needs to be treated with a brace, and >40° needs to be treated with surgery.

Kyphosis Angle (Cobb)

Local kyphosis was measured using the Cobb method, and a kyphosis angle >10° was considered as kyphosis. The significance of the common Cobb method is: negative for <10°, positive for >10°, >25° needs to be treated with a brace, and >40° needs to be treated with surgery.

Postoperative Spinal Nerve Function (Frankel Grade)

Franker spinal cord injury grading is a criterion for evaluating the severity of spinal cord injury. Grade A indicates complete disappearance of sensation below the level of injury; grade B represents only residual sensation; for grade C, sensation exists and the residual part has no practical value for movement; for Grade D, sensorimotor exists and indicates nerve injury; Grade E indicates normal.

Statistical Analysis

SPSS 19.0 (SPSS, USA) statistical software was used for statistical processing. Count variables were expressed by frequency (%) and Fisher's exact probability method was used for comparison between groups. Metrological variables were expressed as mean, standard deviation, or median (interquartile range). Comparisons between the two groups were normal using Student's *t*-test, and non-normal using the Wilcoxon two-sample test. The comparison of detection indexes and the comparison of effects between groups at three time points before, after, and during the follow-up period were corrected by repeated measures of variance analysis and the Bonferroni method. The difference was statistically significant ($P < 0.05$).

Results

Basic Situation of Surgery

Patients were grouped according to surgical grade, with grades 1 and 2 included in group 1 and grades 3, 4, 5, and 6 in group 2 (Table 1). All patients completed the operation successfully. The operation time, intraoperative bleeding volume, and postoperative hospital stay are shown in Table 1. There were significant differences in surgical grade, intraoperative bleeding volume, and length of hospital stay between the two groups. The operation time, the intraoperative bleeding volume, and the hospital stay of the low-grade surgical treatment group were lower than those of the high-grade surgical treatment group. Therefore, the higher the level of surgery, the more complex the treatment

TABLE 1 Comparison of operation time, intraoperative bleeding volume, fixed segment, and postoperative hospital stay between two groups

(I) group	Number	Operative time (min)	Intraoperative bleeding volume (mL)	Postoperative hospital stay (days)
1.00	16	292.5 ± 80.3 (240–360)	1037.143 ± 330 (400–2500)	13.5 ± 2.0 (11.0–18)
2.00	11	362.5 ± 40.3 (303–387)	3690 ± 1580 (2600–5000)	20.6 ± 5.1 (11.0–26)
F		0.759	2.845	2.905
P		0.036	0.042	0.037

The significant level of mean difference is 0.05.

process, the greater the surgical trauma, and the longer the postoperative recovery time.

Postoperative Spinal Nerve Function (Frankel Grade)
Patients were grouped according to surgical grade, with grades 1 and 2 included in group 1 and grades 3, 4, 5, and 6 in group 2. In group 1, 4 of the 16 patients improved to grade D before and after surgery, 7 patients improved to grade E after surgery, and 5 patients improved to grade E after surgery. In group 2, 8 of 11 patients improved to grade D preoperatively and 3 patients improved to grade E postoperatively.

Effectiveness Testing of Therapeutic Surgical Programs
All patients were grouped according to the results of preoperation, postoperation, and follow up. There was 1 group before operation, and there were 2 groups after operation and 3 groups after follow up. The results were analyzed by pairwise comparison. The detection indexes were *T1S1 distance (cm) (coronal)*, *T1S1 distance (cm) (Sagittal)*, *major curve (Cobb) (Coronal)*, *C7CSVL (cm) (coronal)*, *AVT (cm)*, *kyphosis angle (Cobb)*, and *SVA* (*sagittal vertical axis*).

T1S1 distance (cm) (Coronal) ***Results***
There were significant differences among the three groups of data ($P = 0.028$). Comparing different groups in pairs, the results showed that there was no significant difference between groups 1 and 2 ($P > 0.05$), and there was significant difference between groups 1 and 3 ($P < 0.01$) (Table 1). There was also no significant difference between groups 2 and 3 ($P > 0.05$).

Major Curve (Cobb) (Coronal) ***Results***
Major curve was different between groups 1 and 2, and the average of the two groups was 48.52° smaller than that of group 1 ($P < 0.001$). The difference between groups 1 and 3 was extremely significant, and the average of the three groups was 48.16° smaller than that of group 1 ($P < 0.001$). However, there was no significant difference between the two groups and the three groups ($P > 0.05$), indicating that the surgical program had a certain effect (Table 2).

Kyphosis Angle (Cobb) ***Results***
There were significant differences between groups 1 and 3 ($P < 0.001$), 2 and 3 ($P < 0.001$), kyphosis angle (Cobb) in group 2 and 3 were 55.35° and 51.99° smaller than 1, respectively. There was no difference between groups 2 and 3 (Table 3).

Sagittal Vertical Axis ***Results***
The differences between groups 1 and 2 were significant ($P < 0.05$), and the sagittal vertical axis of groups 2 and 3 was 3.14 cm and 1.89 cm larger than that of group 1, respectively. In addition, there was no significant difference between groups 2 and 3 ($P > 0.05$) (Table 4).

There were no differences in *T1S1 distance (cm) (sagittal)*, *AVT (cm)*, and *C7CSVL (cm) (coronal)* between groups 1 and group 2, with *P*-values of 0.154, 1.806, and 0.692, respectively. There were also no significant differences in the three indicators between groups 1 and 3, with *P*-values of 0.063, 1.807, and 0.598. There was no difference between groups 2 and 3, and the *P*-values were 0.645, 0.911, and 0.598, respectively. Based on a comprehensive analysis of all the above indicators, the surgical treatment plan for patients is effective.

Detection of Treatment Outcome in Different Grades of Surgical Schemes

Preoperative and Postoperative Test Results of Indicators
Patients were grouped according to surgical grade, with grades 1 and 2 included as group 1 and grades 3, 4, 5, and 6 as group 2. Differences between the two groups are displayed in Table 5. There were significant differences between *major curve* and *C7CSVL* ($P < 0.05$). There was no difference in the other indicators between the two groups ($P > 0.05$), reflecting that there was no significant difference in the treatment results of different grades of surgical treatment schemes.

Follow-Up Data and Preoperative Data Result Detection
There were no significant differences in all the indicators between the two groups in follow-up data and preoperative data ($P > 0.05$) (Table 6). This indicates that there is no

TABLE 2 Difference analysis of three groups of data with dependent variable *T1S1distanceS*

					95% confidence interval	
(I) group	(J) group	Mean deviation (I–J)	Standard error	Significance	Lower limit value	Upper limit value
1.00	2.00	−3.49000	1.98144	0.087	−7.5213	0.5413
	3.00	−5.52500*	1.98144	0.009	−9.5563	−1.4937
2.00	1.00	3.49000	1.98144	0.087	−0.5413	7.5213
	3.00	−2.03500	1.98144	0.312	−6.0663	1.9963
3.00	1.00	5.52500*	1.98144	0.009	1.4937	9.5563
	2.00	2.03500	1.98144	0.312	−1.9963	6.0663

Group 1, preoperative data; group 2, postoperative data; group 3, follow-up data.; The significant level of mean difference is 0.05.

TABLE 3 Difference analysis of three groups of data with dependent variable *major curve (Cobb)*

					95% confidence interval	
(I) group	(J) group	Mean deviation (I–J)	Standard error	Significance	Lower limit value	Upper limit value
1.00	2.00	48.51500*	6.01897	0.000	36.4527	60.5773
	3.00	48.15667*	6.18391	0.000	35.7638	60.5495
2.00	1.00	−48.51500*	6.01897	0.000	−60.5773	−36.4527
	3.00	−0.35833	6.18391	0.954	−12.7512	12.0345
3.00	1.00	−48.15667*	6.18391	0.000	−60.5495	−35.7638
	2.00	0.35833	6.18391	0.954	−12.0345	12.7512

Group 1, preoperative data; group 2, postoperative data; group 3, follow-up data.; The significant level of mean difference is 0.05.

TABLE 4 Difference analysis of three groups of data with dependent variable *kyphosis angle*

					95% confidence interval	
(J) group	(I) group	Mean deviation (I–J)	Standard error	Significance	Lower limit value	Upper limit value
1.00	2.00	55.35000*	6.48321	0.000	42.3676	68.3324
	3.00	51.99000*	6.48321	0.000	39.0076	64.9724
2.00	1.00	−55.35000*	6.48321	0.000	−68.3324	−42.3676
	3.00	−3.36000	6.48321	0.606	−16.3424	9.6224
3.00	1.00	−51.99000*	6.48321	0.000	−64.9724	−39.0076
	2.00	3.36000	6.48321	0.606	−9.6224	16.3424

Group 1, preoperative data; group 2, Postoperative data; group 3, follow-up data.; The significant level of mean difference is 0.05.

difference between low-grade surgical treatment and high-grade surgical treatment. During follow up, 50% of internal fixation-related complications were found in patients undergoing high-level surgery. For the high-level surgical protocol, follow-up data showed that 2 patients had titanium cage displacement and fusion failure, 1 patient had osteotomy space fusion failure, 1 patient had neurological symptoms aggravation, and 1 patient had distal coronal decompensation. Pseudoarthrosis also occurred after follow up for high-grade surgical treatment, and the failure rate of internal fixation ranged from 7.7% to 14.3%. No associated complications were found in the prognosis of low-grade surgical protocols (Table 7).

Different Factors (Age, Sex, Body Mass Index, Resection, Height) Were Used to Analyze the Difference of Each Index

The differences of each index were analyzed to determine whether the different age, sex, BMI, and height had an impact on the preoperative, postoperative, and prognostic indicators of the patients (Tables 8, 9, 10, 11, 12). The results showed that gender, age, resection, height, and BMI had no

TABLE 5 SVA difference analysis of three groups of data with dependent variable *SVA*

					95% confidence interval	
(I) group	(J) group	Mean deviation (I–J)	Standard error	Significance	Lower limit value	Upper limit value
1.00	2.00	−3.14400*	0.86625	0.001	−4.8786	−1.4094
	3.00	−1.89550*	0.86625	0.033	−3.6301	−0.1609
2.00	1.00	3.14400*	0.86625	0.001	1.4094	4.8786
	3.00	1.24850	0.86625	0.155	−0.4861	2.9831
3.00	1.00	1.89550*	0.86625	0.033	0.1609	3.6301
	2.00	−1.24850	0.86625	0.155	−2.9831	0.4861

Group 1, preoperative data; group 2, postoperative data; group 3, follow-up data.; The significant level of mean difference is 0.05.; SVA, sagittal vertical axis.

TABLE 6 Difference analysis of index in different surgical

Index	Sum of squares	Degrees of freedom	Mean square	*F*	Significance
T1S1distanceC T1S1distanceS	36.038	1	36.038	2.526	0.138
	11.098	1	11.098	0.488	0.501
Apical vertebra translation	105.450	1	105.450	2.475	0.133
Kyphosis angle	69.621	1	69.621	0.184	0.673
SVA	7.698	1	7.698	0.494	0.491
Major curve	1836.584	1	1836.584	5.343	0.034
C7CSVL	41.429	1	41.429	11.230	0.004

The significant level of mean difference is 0.05.

TABLE 7 Difference analysis of index in different surgical

Index	Sum of squares	Degrees of freedom	Mean square	*F*	Significance
T1S1distanceC T1S1distanceS	10.133	1	10.133	0.242	0.632
	10.268	1	10.268	0.587	0.461
Apical vertebra translation	0.066	1	0.066	0.007	0.936
Kyphosis angle	62.795	1	62.795	0.147	0.706
SVA	3.336	1	3.336	0.205	0.656
Major curve	192.200	1	192.200	0.191	0.667
C7CSVL	2.190	1	2.190	0.775	0.390

The significant level of mean difference is 0.05.

significant influence on the preoperative, postoperative, and prognostic indicators of patients ($P > 0.05$).

Postoperative and Follow-Up Results of Typical Cases

An 18-year-old male patient with scoliosis T11–L3 110° and kyphosis T11–L2 70° before the operation had scoliosis 51° and kyphosis 14° after the operation. The comparison before and after the operation is obvious, and shows that the operation was effective (Fig. 9).

A 5-year-old female patient who had scoliosis L1–L4 20° and kyphosis L1–L3 82° (Fig. 7) before the operation had kyphosis L1–L3 6° after the operation. However, she suffered from titanium cage displacement and vertebral hook removal 4 years after the operation, resulting in loss of bone (Fig. 10). We then performed revision surgery on the patient. Seven years after surgery (2 years after the first revision), the patient had a fracture of the left sacral canal screw; we then performed a second revision, 9 years after surgery, and 4 years after the first revision. (Figs 11, 12, 13). A 13-year-old female patient had preoperative scoliosis L1–L4 20° and kyphosis L1–L3, 82°, and kyphosis L1–L3 6°after surgical treatment. The initial treatment was effective. However, the internal fixation rod broke 3.4 years later (Fig. 14). We performed a revision operation. Postoperative images are shown in Fig. 15.

TABLE 8 Analysis of the difference of each index in patients of different gender

Time	Variable	Group	Sample size (missing number)	Average ± standard deviation	Median	Z	P
Preoperation	*T1–S1 distance (cm) (coronal)*	Female	8 (4)	35.34 ± 3.10	35.81 (32.53,38.30)	1.839	0.066
		Male	8 (1)	31.06 ± 3.52	31.62 (27.17,34.41)		
	T1–S1 distance (cm) (sagittal)	Female	7 (5)	36.78 ± 3.67	37.20 (33.83,40.03)	2.598	0.023
		Male	7 (2)	30.98 ± 4.63	29.07 (27.18,36.81)		
	Major curve (Cobb) (Coronal)	Female	12 (0)	89.65 ± 23.92	97.80 (72.45,104.60)	−0.604	0.546
		Male	9 (0)	84.52 ± 26.34	90.50 (81.10,95.40)		
	C7CSVL (cm) (coronal)	Female	12 (0)	1.28 ± 1.83	1.02 (0.10,1.91)	0	1.000
		Male	9 (0)	0.64 ± 2.26	0.90 (0.50,1.83)		
	Apical vertebrate translation (cm)	Female	12 (0)	5.56 ± 4.55	6.09 (3.35,7.45)	0.769	0.451
		Male	9 (0)	4.11 ± 3.82	5.17 (2.62,6.88)		
	Kyphosis angle	Female	12 (0)	80.79 ± 19.35	85.60 (74.30,92.10)	0.604	0.546
		Male	9 (0)	90.88 ± 29.66	89.00 (71.50,102.40)		
	SVA (sagittal vertical axis)	Female	12 (0)	−1.98 ± 3.85	−2.12 (−3.11,-1.21)	−0.32	0.752
		Male	9 (0)	−1.43 ± 3.86	−2.12 (−2.92,-0.44)		
Postoperation	*T1–S1 distance (cm) (coronal)*	Female	10 (2)	37.49 ± 5.42	36.92 (33.90,41.45)	1.373	0.189
		Male	8 (1)	34.21 ± 4.47	34.78 (30.45,37.88)		
	T1–S1 distance (cm) (sagittal)	Female	10 (2)	38.31 ± 5.58	39.59 (32.49,42.27)	1.392	0.183
		Male	8 (1)	35.19 ± 3.35	35.01 (33.37,37.41)		
	Major curve (Cobb) (coronal)	Female	12 (0)	42.78 ± 20.10	46.75 (28.00,57.20)	1.253	0.226
		Male	8 (1)	31.33 ± 19.91	31.50 (13.00,45.40)		
	C7CSVL (cm) (coronal)	Female	12 (0)	0.86 ± 2.03	1.04 (−1.09,2.09)	−0.579	0.563
		Male	8 (1)	0.53 ± 2.05	0.19 (−0.77,0.83)		
	Apical vertebra translation (cm)	Female	12 (0)	3.70 ± 10.99	0.95 (−1.52,5.12)	0.039	0.969
		Male	8 (1)	1.37 ± 1.93	1.05 (0.28,2.93)		
	Kyphosis angle (Cobb)	Female	12 (0)	26.58 ± 16.46	25.65 (12.15,41.05)	−0.986	0.337
		Male	8 (1)	34.05 ± 16.80	34.70 (20.00,45.45)		
	SVA (sagittal vertical axis)	Female	12 (0)	0.74 ± 2.35	0.90 (−0.35,1.81)	−0.536	0.598
		Male	8 (1)	1.31 ± 2.30	0.90 (0.34,3.49)		
Follow up	*T1–S1 distance (cm) (coronal)*	Female	10 (2)	37.17 ± 6.42	37.65 (34.50,41.84)	0.677	0.511
		Male	9 (0)	35.67 ± 2.73	35.38 (33.80,37.52)		
	T1–S1 distance (cm) (sagittal)	Female	10 (2)	40.40 ± 4.36	40.80 (37.10,43.43)	2.304	0.034
		Male	9 (0)	36.32 ± 3.19	35.40 (34.05,38.14)		
	Major curve (Cobb) (coronal)	Female	12 (0)	45.33 ± 16.72	46.55 (30.05,61.50)	1.854	0.079
		Male	9 (0)	30.93 ± 18.79	33.80 (21.00,42.10)		
	C7CSVL (cm) (coronal)	Female	12 (0)	0.87 ± 0.92	0.55 (0.28,1.68)	−0.107	0.915
		Male	9 (0)	0.38 ± 2.22	0.68 (0.20,1.62)		
	Apical vertebra translation (cm)	Female	12 (0)	1.84 ± 2.92	2.00 (1.40,4.04)	0.036	0.972
		Male	9 (0)	1.94 ± 2.15	2.45 (0.82,3.15)		
	Kyphosis angle (Cobb)	Female	12 (0)	28.90 ± 19.78	29.90 (13.60,41.40)	−1.198	0.246
		Male	9 (0)	38.73 ± 16.88	36.80 (26.00,46.90)		
	SVA (sagittal vertical axis)	Female	12 (0)	0.17 ± 2.33	0.38 (−1.22,1.82)	1.132	0.272
		Male	9 (0)	−1.07 ± 2.68	−2.13 (−2.40,−0.10)		

Discussion

Scoliosis Kyphosis Caused by Neurofibromatosis Type 1

The most common clinical manifestation of *NF1* is six or more café-au-lait spots on the skin of the trunk or limbs, usually apparent in early childhood. The diameter of plaque in adults should be greater than 15 mm and that in children should be greater than 5 mm. In normal individuals, five or more café-au-lait spots are less likely to occur, and freckles like NF1 clusters are rare, so they are useful diagnostically[22]. In addition, there are some common clinical manifestations, such as scoliosis and vertebral dystrophy[23, 24]. Neurofibroma originating from the spinal nerve root can cause pain, muscle atrophy, and weakened tendon reflex; approximately 15% of

TABLE 9 Analysis of various indicators of patients of different ages

Time	Variable	Group	Sample size (missing number)	Mean ± standard deviation	Median	*Z*	*P*
Preoperation	*T1–S1 distance (cm) (coronal)*	<15	9 (3)	32.50 ± 3.74	33.46 (30.93,34.96)	−0.811	0.431
		≥15	7 (2)	34.11 ± 4.19	34.74 (31.40,38.30)		
	T1–S1 distance (cm) (sagittal)	<15	8 (4)	33.34 ± 5.44	35.32 (28.13,37.24)	−0.451	0.660
		≥15	6 (3)	34.60 ± 4.78	33.39 (32.20,37.20)		
	Major curve (Cobb) (coronal)	<15	12 (0)	88.88 ± 29.78	97.30 (76.65,108.00)	0.302	0.766
		≥15	9 (0)	85.54 ± 16.47	85.70 (81.10,96.40)		
	C7CSVL (cm) (coronal)	<15	12 (0)	1.18 ± 2.52	1.27 (0.45,2.09)	−0.853	0.394
		≥15	9 (0)	0.77 ± 1.07	0.72 (0.40,1.48)		
	Apical vertebrate translation (cm)	<15	12 (0)	4.80 ± 4.39	5.21 (2.21,7.09)	−0.169	0.867
		≥15	9 (0)	5.12 ± 4.21	6.47 (3.70,7.50)		
	Kyphosis angle	<15	12 (0)	82.68 ± 21.64	87.25 (71.55,99.45)	0.036	0.972
		≥15	9 (0)	88.37 ± 28.21	85.70 (76.20,89.20)		
	SVA (sagittal vertical axis)	<15	12 (0)	−2.57 ± 4.38	−2.66 (−4.19,-1.81)	1.883	0.060
		≥15	9 (0)	−0.65 ± 2.60	−1.38 (−1.92,-0.44)		
Postoperation	*T1–S1 distance (cm) (coronal)*	<15	10 (2)	35.97 ± 5.94	35.69 (30.49,39.37)	−0.051	0.960
		≥15	8 (1)	36.10 ± 4.39	34.51 (32.81,39.87)		
	T1–S1 distance (cm) (sagittal)	<15	10 (2)	36.41 ± 5.30	35.84 (32.49,40.49)	−0.487	0.633
		≥15	8 (1)	37.56 ± 4.52	37.05 (34.31,40.30)		
	Major curve (Cobb) (coronal)	<15	11 (1)	32.65 ± 23.66	32.00 (8.00,59.80)	−1.38	0.185
		≥15	9 (0)	44.97 ± 13.69	50.90 (36.00,53.70)		
	C7CSVL (cm) (coronal)	<15	11 (1)	0.47 ± 2.05	0.17 (−0.97,1.97)	−0.614	0.547
		≥15	9 (0)	1.03 ± 1.99	0.82 (0.20,1.94)		
	Apical vertebra translation (cm)	<15	11 (1)	3.41 ± 11.44	0.37 (−1.60,3.77)	0.912	0.362
		≥15	9 (0)	1.99 ± 2.74	1.57 (0.30,4.10)		
	Kyphosis angle (Cobb)	<15	11 (1)	27.53 ± 15.93	30.00 (12.10,43.20)	−0.599	0.557
		≥15	9 (0)	32.07 ± 17.98	26.00 (18.00,47.40)		
	SVA (sagittal vertical axis)	<15	11 (1)	1.18 ± 1.43	0.71 (0.30,1.49)	0.416	0.686
		≥15	9 (0)	0.71 ± 3.11	1.09 (−2.92,3.38)		
Follow up	*T1–S1 distance (cm) (coronal)*	<15	10 (2)	36.21 ± 5.68	36.47 (33.80,39.15)	−0.228	0.822
		≥15	9 (0)	36.74 ± 4.32	34.71 (34.50,40.69)		
	T1–S1 distance (cm) (sagittal)	<15	10 (2)	38.83 ± 4.49	38.00 (35.40,42.49)	0.372	0.714
		≥15	9 (0)	38.07 ± 4.29	38.14 (35.27,42.20)		
	Major curve (Cobb) (coronal)	<15	12 (0)	36.95 ± 21.74	38.60 (19.85,55.45)	−0.617	0.544
		≥15	9 (0)	42.11 ± 14.26	41.40 (33.80,54.20)		
	C7CSVL (cm) (coronal)	<15	12 (0)	0.30 ± 1.87	0.52 (−0.02,1.33)	0.817	0.414
		≥15	9 (0)	1.14 ± 0.99	0.62 (0.31,2.03)		
	Apical vertebra translation (cm)	<15	12 (0)	1.82 ± 3.03	2.33 (0.73,3.80)	−0.141	0.889
		≥15	9 (0)	1.98 ± 1.93	1.65 (1.38,3.73)		
	Kyphosis angle (Cobb)	<15	12 (0)	33.83 ± 18.83	36.45 (25.15,44.30)	0.195	0.847
		≥15	9 (0)	32.17 ± 19.90	25.80 (20.00,43.60)		
	SVA (sagittal vertical axis)	<15	12 (0)	−0.36 ± 2.72	−0.38 (−2.27,1.92)	0.008	0.994
		≥15	9 (0)	−0.37 ± 2.33	0.07 (−2.17,0.43)		

patients have optic glioma[25]. In addition, studies have shown that NF1 patients have a higher incidence of mental disorders[26]. Scoliosis is the most common musculoskeletal manifestation in NF, usually in the thoracic region[27]. In contrast, 2%–3% of scoliosis patients with obvious curves had NF. The etiology of spinal imbalance includes local neurofibroma erosion or bone infiltration, primary mesodermal dysplasia, osteomalacia, and endocrine disorders[28, 29]. According to the natural development history of NF1 and the shape of scoliosis, it can be divided into two types: non-dystrophic and dystrophic types. The radiological features and treatment regimen in the non-dystrophic type are similar to those for idiopathic scoliosis.

TABLE 10 Difference analysis of each index in patients with resection

Time	Variable	Group	Sample size (Missing number)	Mean ± standard deviation	Median	*Z*	*P*
Preoperation	*T1–S1 distance (cm) (coronal)*	No	7 (3)	35.37 ± 4.12	36.87 (33.85,38.30)	1.907	0.057
		Yes	9 (2)	31.51 ± 2.89	31.60 (30.93,33.46)		
	T1–S1 distance (cm) (sagittal)	No	5 (5)	36.10 ± 4.94	37.20 (34.28,37.34)	1.259	0.232
		Yes	9 (2)	32.65 ± 4.89	32.50 (28.84,36.81)		
	Major curve (Cobb) (coronal)	No	10 (0)	93.01 ± 12.73	97.80 (85.70,102.30)	1.028	0.322
		Yes	11 (0)	82.40 ± 31.52	85.00 (54.60,112.00)		
	C7CSVL (cm) (coronal)	No	10 (0)	1.73 ± 1.79	1.58 (0.65,2.13)	1.514	0.130
		Yes	11 (0)	0.35 ± 2.02	0.72 (−0.20,1.31)		
	Apical vertebrate translation (cm)	No	10 (0)	6.37 ± 1.88	6.50 (5.25,7.30)	1.602	0.134
		Yes	11 (0)	3.63 ± 5.32	3.00 (0.87,7.50)		
	Kyphosis angle	No	10 (0)	80.53 ± 20.77	86.15 (76.20,89.20)	−0.387	0.699
		Yes	11 (0)	89.28 ± 27.20	85.50 (71.50,102.40)		
	SVA (sagittal vertical axis)	No	10 (0)	0.04 ± 3.71	−1.64 (−2.40,2.94)	1.584	0.113
		Yes	11 (0)	−3.37 ± 3.15	−2.14 (−5.16,-1.50)		
Postoperation	*T1–S1 distance (cm) (coronal)*	No	8 (2)	38.14 ± 5.17	37.95 (34.18,41.57)	1.627	0.123
		Yes	10 (1)	34.34 ± 4.72	34.51 (30.49,39.23)		
	T1–S1 distance (cm) (sagittal)	No	8 (2)	38.90 ± 5.36	39.21 (34.35,43.44)	1.61	0.127
		Yes	10 (1)	35.35 ± 4.01	35.01 (33.34,39.10)		
	Major curve (Cobb) (coronal)	No	9 (1)	46.22 ± 16.30	51.50 (35.00,59.80)	1.672	0.112
		Yes	11 (0)	31.63 ± 21.60	32.00 (8.00,50.90)		
	C7CSVL (cm) (coronal)	No	9 (1)	0.74 ± 1.46	0.82 (−0.59,1.97)	0.03	0.976
		Yes	11 (0)	0.71 ± 2.41	0.20 (−1.31,1.94)		
	Apical vertebra translation (cm)	No	9 (1)	1.93 ± 3.45	1.57 (0.42,4.68)	0.988	0.323
		Yes	11 (0)	3.46 ± 11.28	0.30 (−1.54,2.08)		
	Kyphosis angle (Cobb)	No	9 (1)	25.33 ± 16.48	21.30 (14.50,35.50)	−1.035	0.314
		Yes	11 (0)	33.04 ± 16.61	38.90 (14.00,44.20)		
	SVA (sagittal vertical axis)	No	9 (1)	0.99 ± 2.06	1.09 (0.37,1.49)	0.041	0.968
		Yes	11 (0)	0.95 ± 2.56	0.56 (−0.60,3.59)		
Follow up	*T1–S1 distance (cm) (coronal)*	No	9 (1)	36.03 ± 5.86	36.15 (34.50,40.69)	−0.346	0.734
		Yes	10 (1)	36.84 ± 4.26	36.12 (33.80,39.15)		
	T1–S1 distance (cm) (sagittal)	No	9 (1)	38.51 ± 3.44	39.02 (36.98,39.40)	0.038	0.970
		Yes	10 (1)	38.43 ± 5.13	36.77 (34.05,42.49)		
	Major curve (Cobb) (coronal)	No	10 (0)	45.77 ± 14.56	46.25 (35.10,59.40)	1.607	0.124
		Yes	11 (0)	33.15 ± 20.55	33.80 (18.70,51.50)		
	C7CSVL (cm) (coronal)	No	10 (0)	0.77 ± 0.69	0.55 (0.31,1.04)	−0.106	0.916
		Yes	11 (0)	0.56 ± 2.13	0.75 (−0.25,2.03)		
	Apical vertebra translation (cm)	No	10 (0)	2.78 ± 1.24	2.55 (1.65,3.73)	1.631	0.127
		Yes	11 (0)	1.08 ± 3.19	1.91 (−1.83,4.10)		
	Kyphosis angle (Cobb)	No	10 (0)	27.71 ± 15.35	29.90 (17.00,36.80)	−1.274	0.218
		Yes	11 (0)	38.03 ± 20.98	39.20 (22.90,51.20)		
	SVA (sagittal vertical axis)	No	10 (0)	0.25 ± 1.90	0.38 (−0.10,1.34)	1.08	0.294
		Yes	11 (0)	−0.93 ± 2.92	−1.23 (−2.92,1.54)		

The King and Lenke classification for coronal deformities brought about uniformity of thought processes and discussions, and the classification method is suitable for scoliosis[30, 31]. The classification of kyphosis is incomplete if it is based solely on the sagittal balance of the patient without considering flexibility, the size of the sagittal curve, or the integrity of anterior and posterior columns. Rajasekaran *et al.* proposed the classification of kyphosis, and these factors are important in determining the type of osteotomy[32]. In view of the severity of kyphosis caused by dystrophic curves, we have adopted different surgical procedures.

TABLE 11 Analysis of various indicators of different BMI patients

Time	Variable	Group	Sample size (Missing number)	Mean ± standard deviation	Median	*Z*	*P*
Preoperation	*T1–S1 distance (cm) (coronal)*	<20	11 (3)	32.06 ± 3.76	31.84 (27.30,34.96)	−1.882	0.081
		≥20	5 (2)	35.71 ± 3.15	36.87 (33.46,38.30)		
	T1–S1 distance (cm) (sagittal)	<20	10 (4)	32.20 ± 4.37	33.02 (28.84,36.81)	−2.279	0.042
		≥20	4 (3)	38.08 ± 4.33	38.62 (34.85,41.32)		
	Major curve (Cobb) (coronal)	<20	14 (0)	90.33 ± 26.56	95.90 (78.30,104.00)	−1.007	0.314
		≥20	7 (0)	81.70 ± 20.26	85.00 (66.60,101.90)		
	C7CSVL (cm) (coronal)	<20	14 (0)	1.09 ± 2.34	1.07 (0.50,2.04)	−0.597	0.550
		≥20	7 (0)	0.83 ± 1.16	0.72 (−0.20,1.68)		
	Apical vertebrate translation (cm)	<20	14 (0)	4.58 ± 4.72	5.48 (3.00,6.88)	−0.544	0.593
		≥20	7 (0)	5.66 ± 3.16	6.52 (2.62,7.59)		
	Kyphosis angle	<20	14 (0)	87.03 ± 27.62	87.80 (71.60,100.10)	0.504	0.620
		≥20	7 (0)	81.29 ± 16.40	85.50 (76.20,89.20)		
	SVA (sagittal vertical axis)	<20	14 (0)	−1.84 ± 3.40	−2.23 (−3.06,-1.50)	0.858	0.391
		≥20	7 (0)	−1.55 ± 4.71	−1.38 (−3.15,0.40)		
Postoperation	*T1–S1 distance (cm) (coronal)*	<20	12 (2)	35.64 ± 5.73	34.66 (31.11,39.30)	−0.446	0.662
		≥20	6 (1)	36.82 ± 4.14	37.26 (33.90,39.94)		
	T1–S1 distance (cm) (sagittal)	<20	12 (2)	36.84 ± 5.55	35.84 (32.92,41.38)	−0.099	0.922
		≥20	6 (1)	37.09 ± 3.56	37.05 (35.27,40.08)		
	Major curve (Cobb) (coronal)	<20	13 (1)	38.68 ± 22.00	35.00 (22.00,59.80)	0.141	0.890
		≥20	7 (0)	37.30 ± 18.41	42.00 (24.00,51.50)		
	C7CSVL (cm) (coronal)	<20	13 (1)	1.03 ± 2.19	0.45 (−0.95,1.97)	0.93	0.365
		≥20	7 (0)	0.16 ± 1.54	0.35 (−1.42,1.25)		
	Apical vertebra translation (cm)	<20	13 (1)	3.29 ± 10.52	0.37 (−1.54,3.77)	0.713	0.476
		≥20	7 (0)	1.80 ± 2.60	1.57 (−0.98,4.10)		
	Kyphosis angle (Cobb)	<20	13 (1)	32.56 ± 16.64	35.50 (21.30,44.20)	1.106	0.283
		≥20	7 (0)	24.01 ± 16.18	18.00 (9.80,41.40)		
	SVA (sagittal vertical axis)	<20	13 (1)	1.21 ± 1.86	1.24 (0.37,2.13)	0.616	0.545
		≥20	7 (0)	0.53 ± 3.04	0.06 (−2.92,3.98)		
Follow up	*T1–S1 distance (cm) (coronal)*	<20	12 (2)	35.50 ± 4.87	35.77 (33.11,38.63)	−1.11	0.283
		≥20	7 (0)	38.10 ± 5.00	37.52 (34.50,41.84)		
	T1–S1 distance (cm) (sagittal)	<20	12 (2)	38.42 ± 4.40	38.00 (34.73,42.35)	−0.061	0.952
		≥20	7 (0)	38.55 ± 4.44	38.14 (35.27,42.92)		
	Major curve (Cobb) (coronal)	<20	14 (0)	40.84 ± 21.54	46.50 (24.00,61.20)	0.571	0.574
		≥20	7 (0)	35.81 ± 11.67	36.00 (25.00,41.60)		
	C7-CSVL (cm) (coronal)	<20	14 (0)	0.48 ± 1.85	0.52 (0.21,1.62)	0.41	0.682
		≥20	7 (0)	1.03 ± 0.83	0.62 (0.31,2.02)		
	Apical vertebra translation (cm)	<20	14 (0)	1.66 ± 3.05	2.33 (−0.46,4.35)	−0.187	0.852
		≥20	7 (0)	2.34 ± 1.12	1.91 (1.42,3.73)		
	Kyphosis angle (Cobb)	<20	14 (0)	39.59 ± 18.12	38.00 (32.30,51.20)	2.509	0.021
		≥20	7 (0)	20.16 ± 13.24	20.00 (10.20,26.00)		
	SVA (sagittal vertical axis)	<20	14 (0)	−1.05 ± 2.35	−1.22 (−2.40,0.42)	−1.88	0.076
		≥20	7 (0)	1.00 ± 2.37	0.43 (0.07,3.63)		

Dystrophic Scoliosis Treatment Plan

Dystrophic scoliosis is uncommon but much more resistant to treatment[33]. Many methods and techniques for the treatment of spinal deformities have been reported. Dystrophic curves should be treated actively, because even after spinal fusion, dystrophic curves still have a strong development trend[34]. Untreated dysplastic curves, especially in individuals between the ages of 6 and 18 years, continue to deteriorate. Passive observation of dystrophic curves throughout childhood is unrealistic. Dystrophic scoliotic curves of less than 20° should be closely observed every 6 months to determine any sudden rapid development to provide prompt surgical

TABLE 12 Analysis of various indicators of patients with different heights

Time	Variable	Group	Sample size (missing number)	Mean ± standard deviation	Median	*Z*	*P*
Preoperation	*T1–S1 distance (cm) (coronal)*	<1.5	9 (3)	31.70 ± 3.97	31.84 (27.30,33.85)	−1.897	0.079
		≥1.5	7 (2)	35.13 ± 3.03	34.96 (31.60,38.30)		
	T1–S1 distance (cm) (sagittal)	<1.5	8 (4)	32.18 ± 5.41	30.79 (28.01,37.24)	−1.543	0.149
		≥1.5	6 (3)	36.15 ± 3.68	35.55 (33.83,37.20)		
	Major curve (Cobb) (coronal)	<1.5	12 (0)	85.13 ± 26.83	91.95 (79.70,100.75)	0.391	0.696
		≥1.5	9 (0)	90.54 ± 22.11	96.40 (75.00,104.00)		
	C7-CSVL (cm) (coronal)	<1.5	12 (0)	1.01 ± 2.51	0.81 (0.40,1.98)	0.036	0.972
		≥1.5	9 (0)	1.00 ± 1.13	1.23 (0.65,1.68)		
	Apical vertebrate translation (cm)	<1.5	12 (0)	4.83 ± 2.43	5.21 (2.81,7.09)	−0.119	0.908
		≥1.5	9 (0)	5.08 ± 6.01	6.47 (3.70,7.59)		
	Kyphosis angle	<1.5	12 (0)	86.46 ± 31.18	87.25 (71.55,101.40)	0.323	0.751
		≥1.5	9 (0)	83.32 ± 10.83	85.70 (76.20,89.20)		
	SVA (sagittal vertical axis)	<1.5	12 (0)	−2.18 ± 4.49	−2.27 (−4.19,-0.02)	0.746	0.456
		≥1.5	9 (0)	−1.17 ± 2.68	−1.90 (−2.32,-1.38)		
Postoperation	*T1–S1 distance (cm) (coronal)*	<1.5	10 (2)	34.97 ± 4.57	34.78 (31.72,39.37)	−0.97	T1–S1 distance(cm)(coronal)
		≥1.5	8 (1)	37.35 ± 5.85	36.77 (32.50,41.57)		
	T1–S1 distance (cm) (sagittal)	<1.5	10 (2)	35.82 ± 4.30	35.01 (33.34,37.90)	−1.084	0.294
		≥1.5	8 (1)	38.31 ± 5.45	39.80 (33.90,41.88)		
	Major curve (Cobb) (coronal)	<1.5	11 (1)	33.03 ± 22.40	35.00 (8.00,50.90)	−1.278	0.217
		≥1.5	9 (0)	44.51 ± 16.49	51.50 (32.00,54.60)		
	C7CSVL (cm) (coronal)	<1.5	11 (1)	0.46 ± 2.19	0.17 (−1.31,1.97)	−0.66	0.518
		≥1.5	9 (0)	1.06 ± 1.78	0.82 (0.35,1.94)		
	Apical vertebra translation (cm)	<1.5	11 (1)	1.20 ± 2.42	0.42 (0.25,3.77)	−0.076	0.939
		≥1.5	9 (0)	4.69 ± 12.55	1.47 (−1.54,5.55)		
	Kyphosis angle (Cobb)	<1.5	11 (1)	31.57 ± 18.13	35.50 (12.10,44.20)	0.587	0.565
		≥1.5	9 (0)	27.12 ± 15.16	21.30 (14.50,38.90)		
	SVA (sagittal vertical axis)	<1.5	11 (1)	1.39 ± 2.15	0.71 (0.30,3.59)	0.906	0.377
		≥1.5	9 (0)	0.46 ± 2.46	1.09 (−0.60,1.49)		
Follow up	*T1–S1 distance (cm) (coronal)*	<1.5	11 (1)	35.18 ± 5.24	35.38 (32.42,37.52)	−1.353	0.194
		≥1.5	8 (1)	38.22 ± 4.19	39.69 (34.51,41.27)		
	T1–S1 distance (cm) (sagittal)	<1.5	11 (1)	36.87 ± 3.59	36.52 (34.05,39.02)	−2.079	0.053
		≥1.5	8 (1)	40.68 ± 4.40	42.35 (38.12,43.18)		
	Major curve (Cobb) (coronal)	<1.5	12 (0)	33.40 ± 19.15	34.45 (21.35,46.50)	−1.711	0.103
		≥1.5	9 (0)	46.84 ± 15.81	51.50 (41.40,61.20)		
	C7CSVL (cm) (coronal)	<1.5	12 (0)	0.63 ± 2.06	0.86 (−0.03,2.12)	−0.32	0.749
		≥1.5	9 (0)	0.70 ± 0.61	0.47 (0.31,0.75)		
	Apical vertebra translation (cm)	<1.5	12 (0)	1.73 ± 2.51	2.21 (1.37,3.02)	0.107	0.915
		≥1.5	9 (0)	2.10 ± 2.76	1.65 (1.38,4.35)		
	Kyphosis angle (Cobb)	<1.5	12 (0)	36.19 ± 21.84	35.40 (24.45,49.05)	0.86	0.401
		≥1.5	9 (0)	29.01 ± 14.01	25.80 (18.00,39.20)		
	SVA (sagittal vertical axis)	<1.5	12 (0)	−0.51 ± 3.11	−1.40 (−2.66,2.72)	−0.317	0.755
		≥1.5	9 (0)	−0.18 ± 1.51	0.33 (−1.21,0.43)		

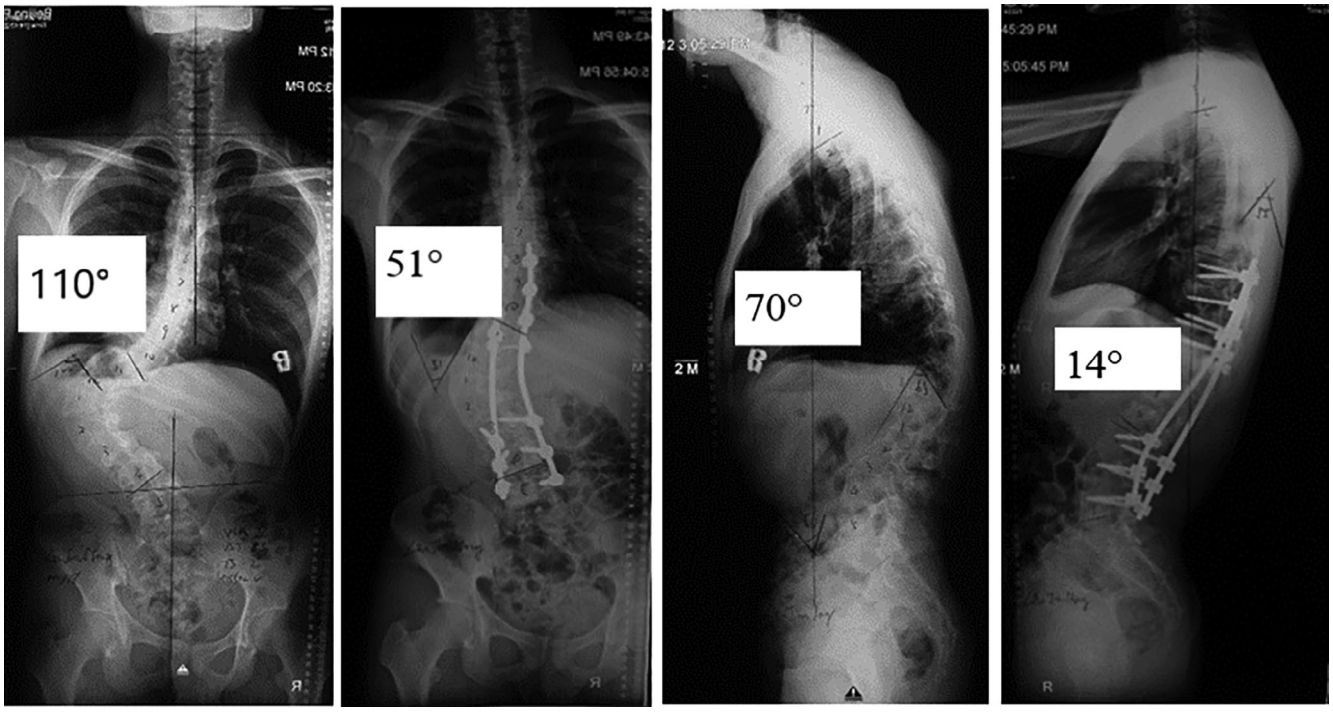

Fig. 9 Preoperative and postoperative comparison of an 18-year-old male patient.

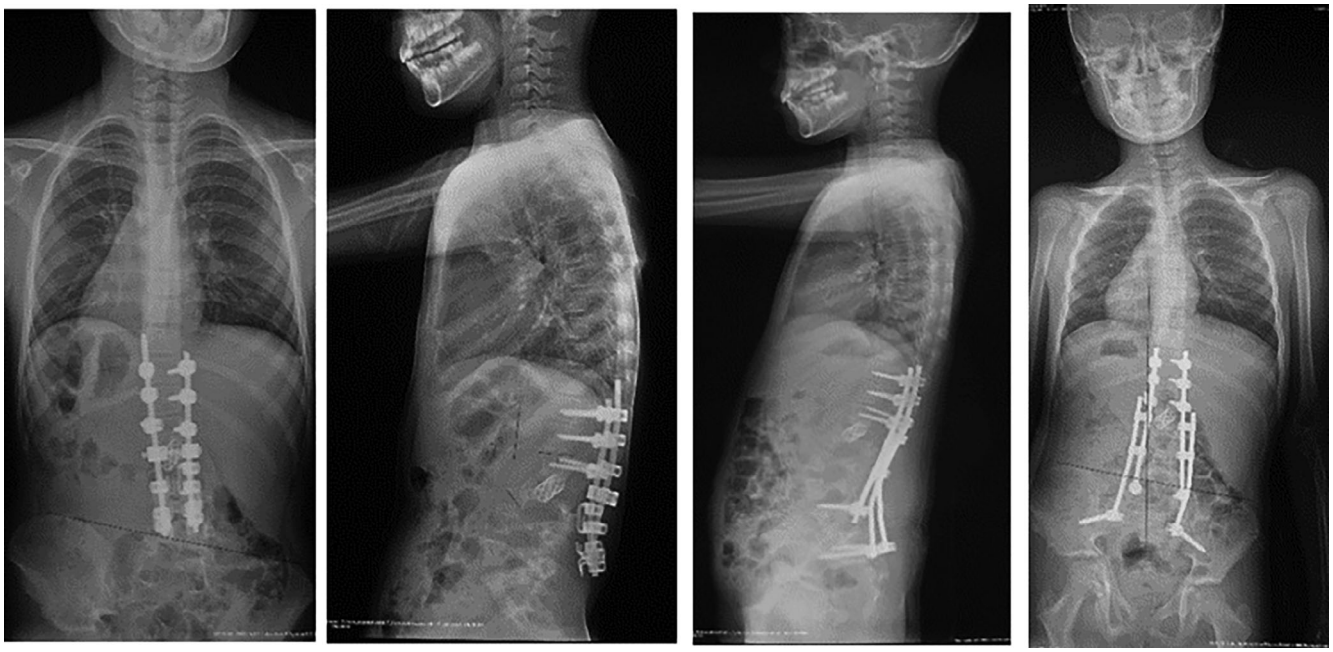

Fig. 10 Imaging picture of 5-year-old female patient 4 years after operation.

treatment. For patients with scoliotic deformities measuring 20–40° and with less than 50° of kyphosis, posterior spinal arthrodesis should be performed using segmental fixation with multiple sublaminar wires. When the dystrophic scoliotic curve exceeds 40°, we recommend anterior/posterior spinal fusion and intervertebral bony fusion. Both procedures can be performed under the same anesthetic session. When thoracic kyphosis exceeds 50°, most authors choose anterior/posterior spinal arthrodesis, which is considered the most reliable surgical procedure[35]. Studies have shown that

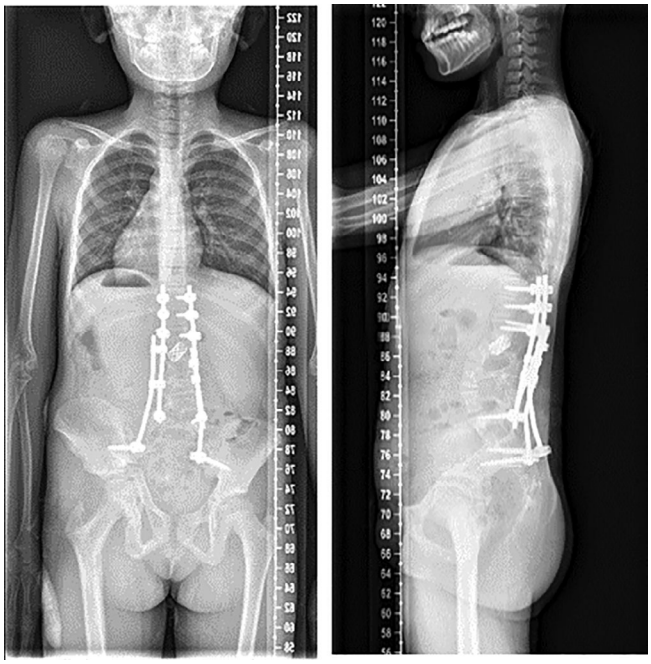

Fig. 11 Imaging picture of a 5-year-old female patient 1 year after revision.

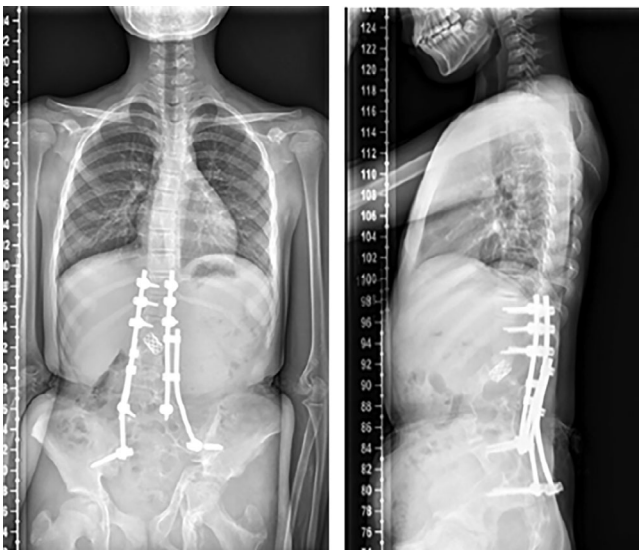

Fig. 12 Imaging picture of a 5-year-old female patient 2 years after revision.

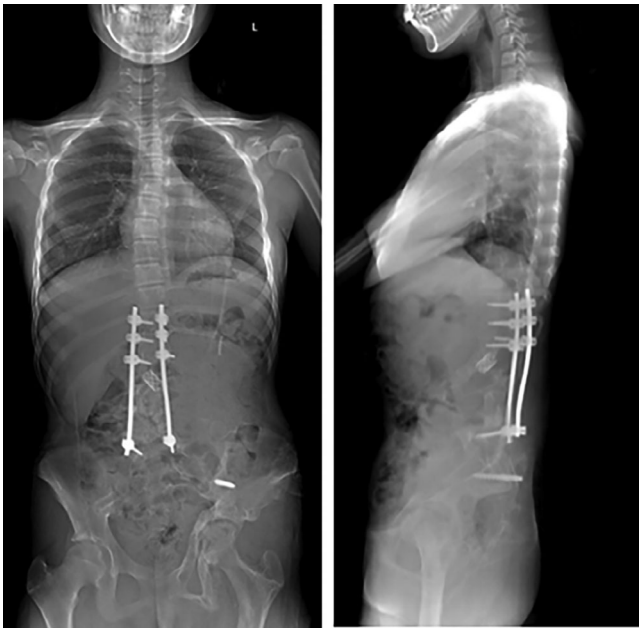

Fig. 13 Imaging picture of a 5-year-old female patient 4 years after revision.

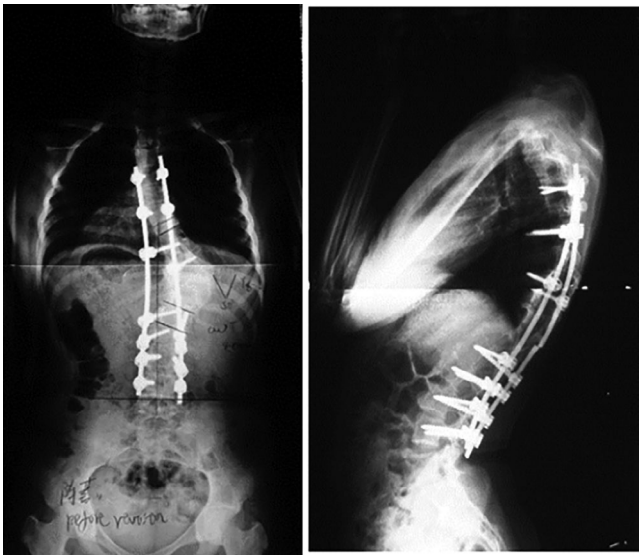

Fig. 14 Images of a 13-year-old female patient 4 years after surgery.

hemivertebrectomy for scoliosis can achieve satisfactory orthopaedic results without major impact on the growth and development of the spinal canal and vertebral body[36]. In this experiment, we used different surgical schemes with various grades, but all patients underwent corpectomy first, and then lateral fusion of autologous and allograft after fixation. The treatment effect is significant; all patients have significant differences before and after surgery; spinal nerve function has also improved significantly, and there is no difference between the different levels of surgical programs.

Treatment of Kyphosis

Rajasekaran classified kyphosis into three types. For type I patients (both anterior and posterior columns being intact), surgery is determined by the need to restore sagittal balance.

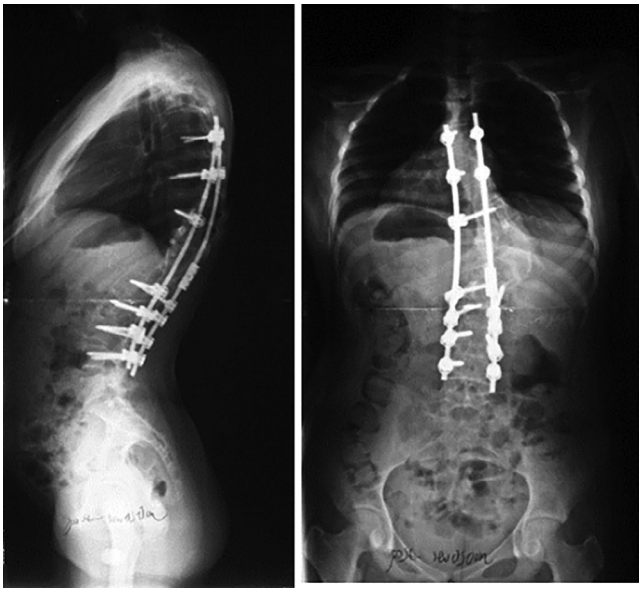

Fig. 15 Image of a 13-year-old female patient after revision.

Activity of the intervertebral disc space is an important consideration for planned osteotomy. Bridge osteotomy can be used in this case. Vertebral resection is usually not performed and complete correction can be achieved. Kyphosis due to loss of one column was classified as type II. In the case of loss of the anterior column and integrity of the posterior column, if kyphosis is less than 60°, bridge osteotomy or intervertebral disc osteotomy can be performed without vertebral body resection, and asymmetric osteotomy can be performed in the case of coronal imbalance[37]. Simple posterior column loss can be corrected by pedicle subtraction osteotomy or intervertebral disc osteotomy without vertebral resection. In patients with type III kyphosis, both anterior and posterior columns are missing, and curve progression is unstable or flexion collapses. When the deformity exceeds 60°, the progress of flexion collapse is faster, which leads to dislocation of multiple facet joints[38]. Osteotomy of the intervertebral disc can be used for deformities of 60°, and vertebral resection is usually used. Generally, the loss of the anterior column is much more serious in type III kyphosis patients than that of the posterior column. Pedicle decompression or intervertebral disc osteotomy requires removal of the obtuse wedge. When the posterior column closes, this can lead to severe spinal cord shortening or posterior hernia. Kawahara *et al.* demonstrated neurological deficits caused by anterior spinal artery kinking after acute spinal cord shortening[39]. According to the specific conditions and severity of scoliosis and kyphosis in different patients, different surgical procedures were performed.

In this experiment, most of the patients had moderate or severe scoliosis and kyphosis. We also carried out graded surgical treatment. The results showed that the treatment efficiency of different grades of surgical treatment was similar, without significant difference. There were no significant differences in the follow-up data. There were some complications in high-level surgical treatment, so it was relatively low-level surgery is more recommended.

Neurofibromatosis type 1 is more likely to cause peripheral neurilemmoma, so the treatment of tumors also needs to be actively addressed. Kahn *et al.* treated NFI-induced tumors in 2014. Twenty patients received radiotherapy, and the 5-year survival rate of all patients was 43.7%. This provides a basis for future NF1 surgery[40]. In addition, with the identification of NF1 and the production of murine strains, it is possible to treat NF1 with molecular and cellular pathophysiology[41]. In this experiment, 1 patient had a paravertebral tumor, so the tumor needed to be removed during surgical treatment.

Recently, some scholars have suggested that in the treatment of scoliosis deformity in children, the correction of spinal curvature should not be simply pursued, and internal fixation devices should be avoided as far as possible to limit the development of thoracic and respiratory issues in children[42]. Nonfusion scoliosis orthopaedics are believed to preserve spinal growth while correcting deformity and promoting normal development of cardiopulmonary function. The present study has some limitations, including that the data from follow-up studies were not comprehensively tested and the sample size was small.

Conclusion

In conclusion, spinal deformities in patients with NFI are still an important diagnostic and therapeutic challenge. Morphological classifications based on spinal defects, flexibility, and curve size show a high degree of consistency among observers and can be used to determine the appropriate osteotomy for NF1 patients. Classification of NF1 patients according to scoliosis and kyphosis is helpful to guide the selection of osteotomy for deformity correction. The therapeutic effect of low-grade surgical treatment in this experiment is ideal. New medical and behavioral interventions are emerging to improve the quality of life of patients. Much progress has been made, but many challenges remain. We need a collaborative and interdisciplinary approach to develop more effective treatments.

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