Surgical Treatment of the Intraspinal Rib Head Dislocation in Children With Dystrophic Scoliosis Secondary to Type 1 Neurofibromatosis

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Background: The purpose of this study was to explore the surgical treatment of intraspinal rib head dislocation (IRH) in children with dystrophic scoliosis secondary to type 1 neurofibromatosis (NF1-DS).

Methods: From 2006 to 2019, 32 of 128 patients with NF1-DS were found to have IRH and enrolled in this study. There were 19 boys and 13 girls with an average age of 8.8 ± 2.6 years. Patients were divided into 2 groups: group A ($n = 25$) without IRH resection and group B ($n = 7$) with IRH resection. The intraspinal rib proportion (IRP), apical vertebra rotation, apical vertebral translation, main thoracic curve Cobb angle, trunk shift and thoracic kyphosis, lumbar lordosis, and sagittal vertebral axis were measured before and after the operation. Spinal injury was graded based on the American Spinal Injury Association (ASIA) Impairment Scale.

Results: The study group had a total of 42 IRH. The mean follow-up duration was 46.1 ± 28.7 months. The preoperative IRP in both groups was similar ($35.5 \pm 14.3\%$ vs. $31.2 \pm 15.3\%$, $P = 0.522$). The postoperative IRP was lower in group B ($18.5 \pm 11.2\%$ vs. 0%, $P = 0.002$). The IRP in group A decreased from preoperative ($31.2 \pm 15.3\%$) to postoperative ($18.5 \pm 11.2\%$) ($P < 0.05$). There was no significant difference in the apical vertebra rotation, apical vertebral translation, main thoracic curve Cobb angle, trunk shift, thoracic kyphosis, lumbar lordosis, and sagittal vertebral axis between the 2 groups before surgery and after surgery. Four patients with nerve injury caused by the IRH had full neurological recovery postoperatively. All patients were ASIA grade E at the last follow-up.

Conclusions: The surgical treatment of IRH in children with NF1-DS should be determined on the basis of the presence of preoperative neurological symptoms. This study supports the practice of correcting spinal deformities only in patients with mild or no spinal cord injury. If there are obvious neurological symptoms, IRH resection is necessary to relieve spinal cord compression to recover nerve function.

Level of Evidence: Level III.

Key Words: children, neurofibromatosis, dystrophic scoliosis, rib head, spinal canal

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Type 1 neurofibromatosis (NF1), also known as von Recklinghausen disease, is characterized by peripheral nervous system involvement and may be associated with skeletal deformities, of which spinal deformities are the most common (incidence: 10% to 60%).[1,2] Spinal deformities can be divided into dystrophic and nondystrophic types.[3] The main manifestations of dystrophic scoliosis secondary to type 1 neurofibromatosis (NF1-DS) are short and sharp scoliosis segments accompanied by wedge changes, severe vertebral rotation, wider pedicle spacing, and enlarged intervertebral foramina, which may be combined with "pencil-like" changes to the ribs and rib head dislocation.[4] In 1986, Flood et al[5] first reported that patients with NF1-DS could have intraspinal rib head dislocation (IRH), which can compress the spinal cord and cause nervous system injury. The necessity of surgical management for NF1-DS–associated IRH is based on the presence of neurological symptoms. While patients with neurological symptoms caused by IRH require IRH resection, it is unclear whether there is a need for IRH correction for asymptomatic patients.[6–8] There is a paucity of evidence regarding IRH correction because this condition is relatively rare, and few cases have been reported. In this single-center retrospective chart review, we explored the clinical and radiographic characteristics and surgical management of IRH in children with NF1-DS.

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METHODS

Participants

This study was approved by the institutional review board. A total of 128 patients with NF1-DS were retrospectively identified in our hospital from September 2006 to December 2019. Patients were included if they had dystrophic thoracic scoliosis secondary to NF1, evidence of IRH on preoperative computed tomography (CT) or magnetic resonance imaging (MRI), received surgical intervention, had complete preoperative and postoperative imaging (anteroposterior and lateral radiographs of the spinal column, CT, or MRI), and > 12 months' follow-up. Patients with encephalopathy, peripheral neuropathy, and additional space-occupying lesions in the spinal canal were excluded.

Patients were classified as group A: not undergoing IRH resection; and group B: undergoing IRH resection. One patient (no. 26) did not have the IRH removed while growing rods (GRs) were in place. However, the IRH was removed in the final posterior spinal fusion (PSF); therefore, this patient was included in group B. Group A comprised 25 patients, and group B comprised 7 patients (Table 1).

Surgical Method

The surgical strategy was based on the child's age, the severity of the spinal deformity, growth potential, and development potential. A pedicle screw system or screw-hook mixed system was used for PSF. Fourteen patients underwent 1-stage PSF, and 18 patients received GRs (15 patients with 2 GRs and 3 patients with a single GR), 3 of whom had undergone PSF. The spinal canal was not opened during GR implantation and lengthening. Ten patients underwent halo-gravity traction (HGT) for ∼4 weeks preoperatively to increase spinal flexibility. The IRH was removed in 7 patients: 6 cases of 1-stage PSF and 1 case of GR lengthening. All procedures were performed with neurophysiological monitoring, namely recording the somatosensory evoked potentials and motor evoked potentials.

IRH resection technique: After the pedicle screw placement, the IRH segments were confirmed, and the corresponding convex lamina and articular process were removed. The rib was stripped to 1 cm outside the spinal canal and then cut. Next, the IRH was carefully extracted, and the bone wax was applied to the cut end of the rib.

Radiographic Measurements

(1) Coronal and sagittal plane parameters were based on anteroposterior and lateral radiographs of the spinal column taken preoperatively, 3 months postoperatively, and at the last follow-up.
 (a) Coronal plane parameters: main thoracic curve Cobb angle; trunk shift (TS): the horizontal distance between the C7 plumb line and the center sacral vertical line; coronal plane imbalance, defined as TS > 20 mm; and apical vertebral translation: the horizontal distance between the center of the apical vertebra and the center sacral vertical line (Fig. 1A).
 (b) Sagittal plane parameters: thoracic kyphosis (TK; T2-T12); lumbar lordosis (LL; T12-S1); and sagittal vertebral axis (SVA): the horizontal distance between the C7 plumb line and the plumb line of the posterior superior angle of the sacrum. Sagittal plane imbalance was defined as SVA > 40 mm.

(2) CT parameters: All NF1-DS patients in our hospital require a CT scan and preoperative 3-dimensional reconstruction. The following parameters were measured on axial CT at the area of most severe transection by the IRH using a picture archiving and communications systems workstation:
 (a) Intraspinal rib proportion (IRP; Fig. 1B): the proportion of the IRH length to the spinal canal transverse diameter. Line x longitudinally bisected the vertebral body. Line c was the distance between the rib head tip and line x. Line d was the distance between the most concave area of the osseous spinal canal and line x. IRP was calculated as: $\text{IRP}=(d-c)/2d$ when lines c and d were on opposite sides of line x or $(d+c)/2d$ when lines c and d were on the same side as line x.
 (b) Apical vertebral rotation (Fig. 1C): defined by the angle formed between the line longitudinally bisecting the vertebral body and the line connecting the midpoint of the sternum and the midpoint of the posterior edge of the spinal canal.[9]

(3) MRI parameters: Distance between the spinal cord and intraspinal rib head dislocation (DSCIRH; Fig. 1D), defined as the length measured from the rib head tip to the convex edge of the spinal cord.

(4) Correction rate was defined as: (preoperative value−postoperative value)/preoperative value×100%.

(5) Spinal injury was graded based on the American Spinal Injury Association (ASIA) Impairment Scale using the following categories.[10,11]

A = Complete: No sensory or motor function preserved in the sacral segments S4-S5; B = Sensory Incomplete: Sensory but not motor function is preserved below the neurological level and includes the sacral segments S4-S5, AND no motor function is preserved > 3 levels below the motor level on either side of the body; C = Motor Incomplete: Motor function is preserved below the neurological level, and more than half of key muscle functions below the single neurological level of injury have a muscle grade <3 (grades 0 to 2); D = Motor Incomplete: Motor function is preserved below the neurological level, and at least half (half or more) of key muscle functions below the neurological level of injury have a muscle grade ≥ 3; E = Normal: Sensory and motor functions are normal.

We define ASIA D as mild spinal cord injury, ASIA A, B, and C as obvious spinal cord injury.

Follow-up

Patients with GR were followed up 6 months postoperatively, and the interval of lengthening was determined based on growth rate and imaging, usually once every 9 to 12 months. Patients who underwent PSF were followed up every 3 months for the first year after surgery and then every

TABLE 1. Date of Intraspinal Rib Head Dislocation in Children With Dystrophic Scoliosis Secondary to Type 1 Neurofibromatosis Included in the Study

	Case No.	Sex	Age (y)	Curve Pattern	Apical Vertebra	Plane of Intraspinal Rib	DSCIRH (mm)	Treatment	ASIA Grade	Complication
Group A	1	F	10.6	D	T5	T5	6	PSF	E	
	2	M	6.3	L	T7	T7	6	GR (single)	E	Screw loosening
	3	F	6.3	L	T7	T7	7.5	PSF	E	
	4	F	8.2	D	T9	T9	4.8	PSF	E	
	5	M	6.8	R	T8	T7	3	GR (dual)/PSF	E	Screw loosening
	6	M	5.3	R	T7	T7	6	GR (single)/PSF	E	Screw loosening broken rod
	7	M	7.7	R	T8	T8	1.8	HT, GR (dual)	E	
	8	M	9.1	R	T9	T10	3.4	GR (dual)	E	
	9	M	6.4	L	T8	T8	5	GR (dual)	E	
	10	F	7.1	L	T6	T6	4	GR (single)	E	
	11	M	5.9	R	T6	T6	6	GR (dual)	E	Bolt loosening, broken rod
	12	M	9.5	R	T8	T7	5	GR (dual)	D	
	13	M	11.4	R	T6	T6	5	PSF	E	
	14	F	9.9	R	T8	T7	5	PSF	E	DJK
	15	M	8.3	D	T4	T4	5.5	HT, GR (dual)	E	
	16	F	7.5	L	T5	T6	2.5	HT, GR (dual)	A	
	17	M	11	D	T5	T5	4	PSF	E	
	18	M	8.4	D	T4, T9	T3/T4, T9	2.6/2.7/3	HT, GR (dual)	E	
	19	M	11.6	L	T7	T6/T7	6.5/4	PSF	E	
	20	M	13.3	L	T5	T4/T5/T6	4/4.3/4	PSF	E	
	21	F	6	R	T5	T5	8	HT, GR (dual)	E	
	22	F	9.8	D	T8	T7	6	HT, GR (dual)	E	
	23	F	4.6	R	T10	T10	4.5	GR (dual)	E	
	24	F	5	D	T5	T4/T5/T6	5.2/3.3/3	HT, GR (dual)	D	
	25	M	8.7	R	6	T6/T7	6/5	GR (dual)	E	
Group B	26	F	6.5	L	T8	T9	3	GR (dual)/PSF	E	
	27	F	9.5	D	T5	T5	3	HT, PSF	E	
	28	M	11.8	D	T4	T5	1.1	PSF	D	Adding-on
	29	F	8.9	R	T5	T4	5	HT, PSF	E	Adding-on
	30	M	11.5	R	T6	T7	3	AR, PSF	E	Adding-on
	31	M	10.8	R	T5	T4	4.8	HT, PSF	E	Adding-on
	32	M	16.3	R	T6	T5/T6/T7	2/0/1.6	PSF	C	Adding-on

Adding-on indicates adding-on phenomenon; AR, anterior release; ASIA, American Spinal Injury Association; D, double thoracic curve; DJK, distal junctional kyphosis; DSCIRH, distance between the spinal cord and the intraspinal rib head dislocation; F, female; GR, growing rod; HT, halo traction; L, light thoracic scoliosis; M, male; PSF, posterior spinal fusion; R, right thoracic scoliosis.

other year thereafter. At each follow-up visit, patients were offered radiographs and CT scans.

Statistical Analysis

The data were analyzed using IBM SPSS 19.0 statistical software (IBM Corp., Armonk, NY). All measurement data are expressed as mean ± SD. Paired *t* test was used to analyze and compare the imaging results of patients preoperatively, postoperatively and at the last follow-up. Independent sample *t* test was used to compare groups A and B. Statistical significance was defined as a *P*-value <0.05.

RESULTS

A total of 32/128 (25%) NF1-DS patients with IRH were included in this study. Patients comprised 19 boys and 13 girls, with an average age of 8.8 ± 2.6 years. The patients' characteristics are described in Table 1. There were 8 patients with a left thoracic curve, 15 patients with a right thoracic curve, and 9 patients with double thoracic curves; 19 patients had TK. We identified 42 IRHs, all of which were located on the convex side of the main thoracic curve and in the adjacent segments to the convex apical vertebra. There were 21 (50%) apex IRHs, 9 (21%) apex+1 (distal) IRHs, and 12 (29%) apex−1 (proximal) IRHs; 26 patients had 1 IRH, 2 had 2 IRHs, and 4 had 3 IRHs.

Four patients had spinal cord injury caused by IRH, of which 1 patient (no. 32) was classified as American Spinal Injury Association (ASIA) grade C preoperatively. The spinal cord injury was related to direct compression of the spinal cord owing to the IRH; therefore, the IRH was surgically removed. The other 3 patients (nos 12, 24, and 28) were classified as ASIA grade D, and the IRH was not removed in 2 of these patients (nos 12 and 24). One patient (no. 16) was classified as ASIA grade A, and preoperative imaging showed that the severe spinal cord injury was not directly related to the IRH, but rather to severe dural ectasia and bone resorption which led to spinal dislocation with stenosis. After HGT for 2 months, the patient recovered to ASIA grade D. The IRH was not removed while GRs were in place. Six months postoperatively, the patient was classified as ASIA

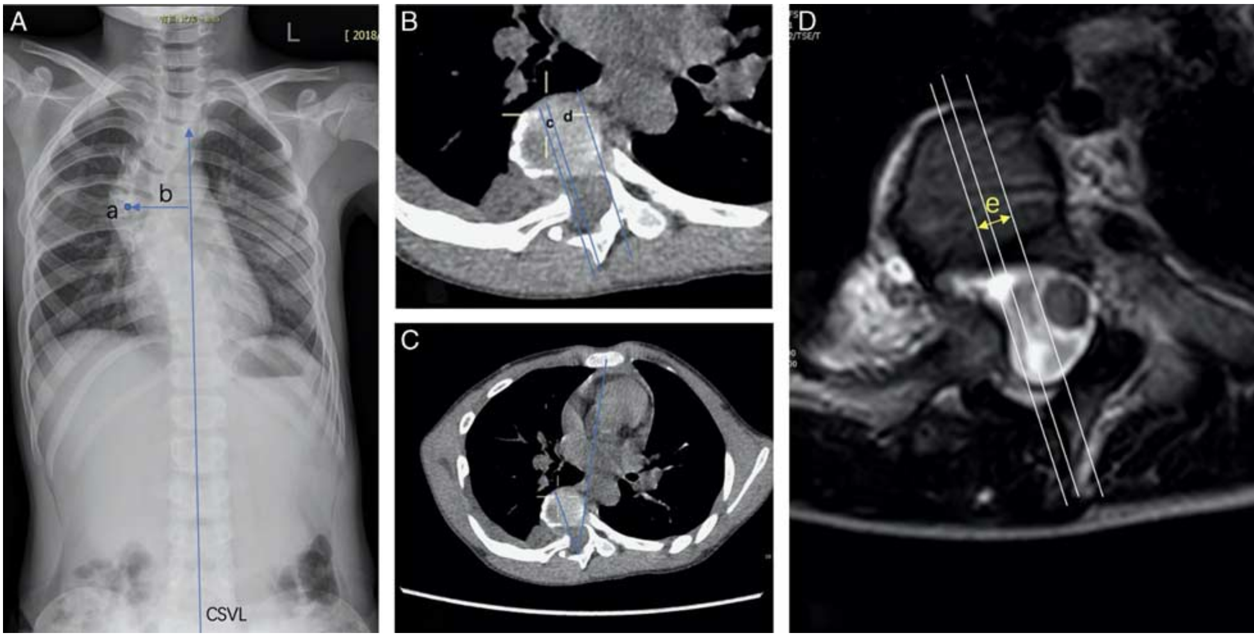

FIGURE 1. Radiographic measurements. A, Apical vertebral translation (line *b*): the horizontal distance between the center of apical vertebra (point *a*) and centre sacral vertical line (CSVL). B, intraspinal rib proportion = (*d*–*c*)/2*d* when lines *c* and *d* were on opposite sides of line *x* (line longitudinally bisecting the vertebral body) or (*d*+*c*)/2*d* when lines *c* and *d* were on the same side as line *x*. C, Apical vertebra rotation: the angle formed between the line longitudinally bisecting the vertebral body and the line connecting midpoint of the sternum and midpoint of the posterior edge of the spinal canal. D, Distance between the spinal cord and the intraspinal rib head dislocation (line *e*): the length measured from the rib head tip to the convex edge of the spinal cord.

grade E. There was no change in clinical symptoms in the remaining patients after HGT for 4 weeks preoperatively. The other 27 patients were classified as ASIA grade E preoperatively. The ASIA grade improved after surgery, and all patients, including the 4 patients with spinal cord injury caused by IRH preoperatively, were classified as grade E at the last follow-up.

The mean preoperative IRP was 32.5 ± 15.6% and the mean DSCIRH was 4.3 ± 1.7 mm. Notably, in 1 case with 3 IRHs (no. 32), the IRH compressed the spinal cord leading to a DSCIRH of 0 mm. Ten patients had paravertebral neurofibromas. The mean follow-up duration was 46.1 ± 28.7 months. Postoperative complications occurred 12 person-times, namely 5 distal adding-on, 2 patients (nos 29 and 30) requiring revision surgery, 3 loosened distal screws, 2 broken rods, 1 loosened distal bolt, and 1 distal junctional kyphosis resulting in revision surgery (Table 1).

A comparison of the patients' clinical and radiographic characteristics is shown in Table 2. The IRP in group A was similar to that in group B preoperatively: 31.2 ± 15.3% versus 35.5 ± 1 4.3%, respectively ($P = 0.522$). After surgery, the IRP decreased in both groups, with a significantly lower IRP in group B than in group A: 18.5 ± 11.2% versus 0%, respectively ($P = 0.002$). There were no significant differences between the groups in preoperative and postoperative apical vertebral rotation, apical vertebral translation, main thoracic curve Cobb angle, TS, TK, LL, and SVA, and their correction rates ($P > 0.05$). The mean main thoracic curve Cobb angle, TS, TK, and LL postoperatively and at the last follow-up differed significantly compared with the respective preoperative values ($P < 0.05$) in both groups. There were no significant differences in the main thoracic curve Cobb angle, TS, and TK at the last follow-up compared with the respective postoperative values ($P > 0.05$) in both groups. The mean LL and SVA postoperatively and at the last follow-up were not significantly different compared with the respective preoperative values ($P > 0.05$).

DISCUSSION

NF1 is an autosomal dominant hereditary disease with an incidence of 1/3000.[12] NF1-DS with IRH is rare, and most previous studies were case reports or case series.[5,13–17] Our study involved the largest number of NF1-DS patients with IRH from a single center to date. Mao et al[7] and Qian et al[9] reported IRH incidences of 15.9% (23/145) and 6.5% (8/124), respectively, compared with the incidence of 25% (32/128) in our study. There is a significant proportion of IRH in NF1-DS, which warrants clinical attention to recognize rib subluxation causing spinal cord injury owing to rib head compression on the spinal cord. CT and MRI are effective to confirm the diagnosis of IRH. However, MRI provides a clearer picture of the relationship between the IRH and the dural sac and can be used for evaluating the degree of intraspinal rib involvement and spinal cord compression. In this study, the indentation of the dural sac was seen in 27 patients, and the dural sac and spinal cord drifted to the concave

TABLE 2. Clinical and Radiographic Characteristics of the Patients in Groups A and B

	Mean ± SD			
	Group A	**Group B**	***t***	***P***
Age (y)				
Preoperative	8.1 ± 2.3	11.2 ± 2.0	3.024	0.005
Follow-up (m)	45.5 ± 29.2	64.0 ± 28.2	1.406	0.170
DSCIRH (mm)				
Preoperative	4.5 ± 1.4	3.6 ± 1.9	−1.387	0.173
IRP (%)				
Preoperative	31.2 ± 15.3	35.5 ± 14.3	0.646	0.522
Postoperative	18.5 ± 11.2*	0*	−3.993	0.002
Correction rate (%)	39.7 ± 22.2	100	4.830	0.024
AVR (deg.)				
Preoperative	36.0 ± 13.4	33.3 ± 10.6	−0.453	0.654
Postoperative	25.9 ± 8.6*	24.3 ± 9.3*	−0.386	0.703
Correction rate (%)	24.1 ± 26.9	26.3 ± 16.1	0.181	0.858
AVT (mm)				
Preoperative	35.4 ± 16.9	27.9 ± 15.9	−1.008	0.320
Postoperative	17.2 ± 9.9*	14.6 ± 9.3*	−0.598	0.553
Correction rate (%)	45.4 ± 32.8	40.7 ± 27.7	−0.444	0.657
Last follow-up	16.4 ± 9.7‡	13.4 ± 7.7‡	−0.719	0.476
Main thoracic curve Cobb angle (deg.)				
Preoperative	68.6 ± 28.3	65.3 ± 19.7	−0.268	0.791
Postoperative	38.1 ± 20.8*	30.8 ± 5.8*	−0.845	0.405
Correction rate (%)	45.9 ± 19.8	48.1 ± 22.5	0.233	0.817
Last follow-up	36.4 ± 20.7‡	35.5 ± 4.7‡	−0.103	0.919
TS (mm)				
Preoperative	15.5 ± 13.8	12.2 ± 4.7	−0.570	0.573
Postoperative	5.7 ± 3.6*	3.5 ± 3.9*	−0.613	0.433
Correction rate (%)	64.7 ± 29.7	77.7 ± 23.2	0.392	0.706
Last follow-up	6.3 ± 3.2‡	5.2 ± 4.8‡	−0.602	0.476
TK (deg.)				
Preoperative	64.0 ± 19.3	48.4 ± 22.5	1.570	0.127
Postoperative	32.7 ± 14.6*	37.0 ± 12.2*	0.667	0.510
Correction rate (%)	21.9 ± 39.2	35.0 ± 33.4	0.752	0.458
Last follow-up	36.5 ± 14.1‡	45.5 ± 12.6‡	1.430	0.163
LL (deg.)				
Preoperative	48.1 ± 11.8	41.3 ± 16.0	−1.181	0.247
Postoperative	43.1 ± 9.2†	40.3 ± 9.8†	−0.649	0.521
Correction rate (%)	5.4 ± 8.4	2.9 ± 7.8	−0.202	0.841
Last follow-up	46.4 ± 8.2‡	41.2 ± 9.7‡	−0.369	0.681
SVA (mm)				
Preoperative	25.5 ± 14.6	37.2 ± 22.2	1.596	0.121
Postoperative	21.8 ± 16.3†	29.2 ± 17.9†	0.985	0.332
Correction rate (%)	19.6 ± 10.1	23.7 ± 15.7	0.229	0.820
Last follow-up	21.3 ± 15.1‡	25.0 ± 16.5‡	0.536	0.596

*$P < 0.05$ compared with preoperative value.
†$P > 0.05$ compared with preoperative value.
‡$P > 0.05$ compared with postoperative value.
AVR indicates apical vertebral rotation; AVT, apical vertebra translation; DSCIRH, distance between the spinal cord and the intraspinal rib head dislocation; IRP, intraspinal rib proportion; LL, lumbar lordosis; SVA, sagittal vertebral axis; TK, thoracic kyphosis; TS, trunk shift.

side of the spinal canal and away from the IRH. The median DSCIRH was 4.3 ± 1.7 mm preoperatively. This may explain why most patients in this study had no spinal cord injury, as in the example shown in Figure 2. Only 4 patients had spinal cord injury caused by IRH. One patient (no. 32; Fig. 3) had lower extremity neurological symptoms owing to the rib head directly compressing the spinal cord. The other 3 patients had no direct compression of the spinal cord but had positive preoperative neurological signs in both lower extremities. The reason for the neurological signs may be that the spinal cord drifted to the concave side of the spinal canal, which led to impingement of the concave apical pedicle.

The management of IRH in children with NF1-DS is controversial. Most surgeons believe that for patients with no neurological symptoms or only mild signs of spinal cord injury direct correction of the thoracic scoliosis and neurophysiological monitoring are safe and effective.[6–9] For patients with neurological symptoms and signs of spinal cord compression by the IRH on imaging, the IRH should be removed.[15,16,18] In this study, 7 patients underwent IRH excision (included in group B), of which 1 patient (no. 32) was classified as ASIA grade C, 1 patient (no. 28) as ASIA grade D, and the other 5 patients as ASIA grade E (nos 26, 27, 29, 30, and 31). Six of these patients had no neurological symptoms or only mild spinal cord injury. On the basis of previous protocols, IRH removal was performed to reduce the risk of intraoperative neurological complications.[5,19] With evolving evidence, our hospital gradually adopted conservative management for IRH for patients with no neurological symptoms or only mild spinal cord injury preoperatively.

Overall, the characteristics of the 2 treatment groups were very similar. Group B comprised patients who underwent PSF, while most of the patients in group A (18/25) were young patients who underwent GR implantation. The IRP in group A decreased from 31.2% ± 15.3% preoperatively to 18.5 ± 11.2% postoperatively, and the correction rate was 39.7 ± 22.2%. Thus, the IRH reduced passively outside the spinal canal after the scoliosis was corrected. Similarly, Mao et al[7] reported that 14.8% of the IRHs in their study invaded the concave half-circle of the spinal canal and that the percentage decreased to 3.7% postoperatively. Cai et al[8] reported that the median percentage of the spinal canal occupied by the intraspinal rib was significantly lower at the 1-year follow-up compared with the preoperative value (28.6% vs. 23.1%, respectively). Although the spinal cord shifted from the concave side to the convex side with surgical correction, this did not cause spinal cord injury. Instead, the symptoms of 3 patients in group A with mild spinal cord injury caused by the IRH were relieved after surgery and at the last follow-up. Therefore, the spinal cord was not compressed by the IRH directly. Instead, IRH removal in this group may have increased surgical complexity and the risk of surgical complications, such as spinal cord injury.[13,20] In addition, the preoperative and postoperative LL and SVA values were not significantly different between the treatment groups, suggesting that RH resection has little effect on scoliosis correction in NF1-DS. However, if the IRH causes severe neurological symptoms, it must be removed for nerve recovery. For example, patient No. 32 was classified as ASIA grade C preoperatively. The IRH was excised, and with neurological rehabilitation, the patient's neurological symptoms recovered completely 6 months postoperatively.

In the surgical management of NF1-DS, accurate pedicle screw placement combined with moderate correction is a prerequisite for spontaneous reduction of the RH

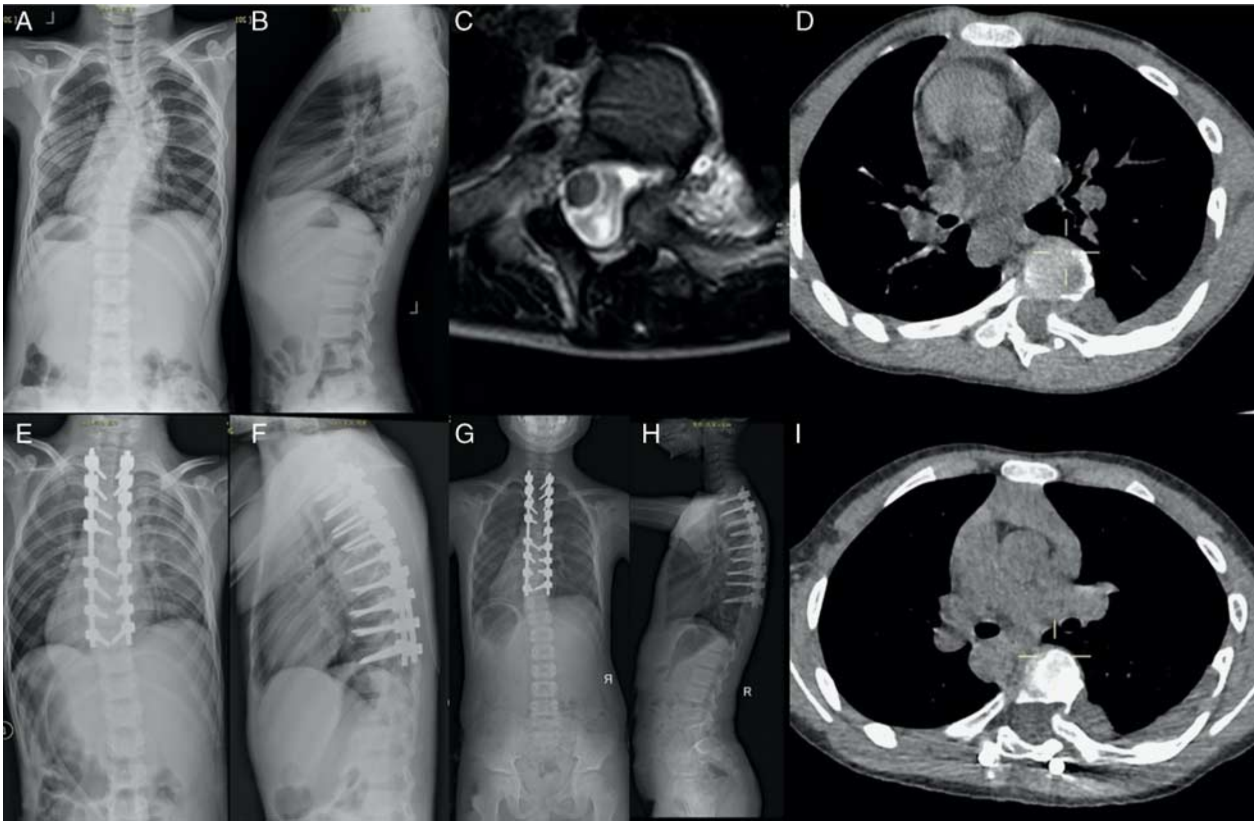

FIGURE 2. A 11.4-year-old male (no. 13) with right thoracic scoliosis, and the apical vertebra was at T6. He was preoperative American Spinal Injury Association (ASIA) grade E. A and B, Preoperative anteroposterior and lateral radiographs of the spinal column. C and D, Preoperatively the ribs were protruding into the spinal canal from the right intervertebral foramen in the T6 plane, and the rib heads did not cross the center line of spinal canal. The distance between the spinal cord and the intraspinal rib head dislocation was 5 mm, and the intraspinal rib proportion was 31.1%. E and F, Postoperative anteroposterior and lateral radiographs of the spinal column. The intraspinal rib head dislocation (T6R) was not removed intraoperatively. G and H, Post-operative 50 months' follow-up anteroposterior and lateral radiographs of the spinal column. I, The intraspinal rib head dislocation was passively reduced after the operation, and the postoperative intraspinal rib proportion was 21.6%.

and avoidance of recurrence.[13] However, in NF1-DS, the dysplastic pedicle is too thin for pedicle screw placement, and there is a high pedicle screw misplacement rate because of low bone mineral density, the biomechanical properties of pedicle screw loosening after placement, and because the screw channel is susceptible to change and postoperative instrumentation loosening or displacement.[21] Ten (31.25%) patients sustained 12 instrumentation-related complications (IRCs) in our study. Yao and colleagues[22,23] reported that 17/59 (28.8%) patients sustained IRCs and that mean age below 9 years, TK > 50 degrees, and GR implantation are risk factors for IRC after surgical treatment of NF1-DS. In the current study, 18 patients (56.3%) were younger than 9 years of age, 20 patients (62.5%) had TK > 50 degrees, and 18 patients (56.3%) were treated with GRs. Most patients had risk factors for IRC; therefore, the incidence of IRC in this study was slightly higher than that reported in previous studies. To reduce the IRC incidence, we use pedicle screws for fixation as often as possible. If this is not possible, we use a lamina hook or transverse process hook instead and avoid leaving too many segments uninstrumented, especially in the apical vertebral area. In addition, autogeneic or allogeneic bone grafting can be performed at the instrumentation site to enhance stability. Recently, we have used preoperative CT to create a 3-dimensional model, which has improved the efficiency and accuracy of pedicle screw placement. In 2020, we introduced an intraoperative navigation robot. The detailed mechanics behind curve progression, adding-on, and proximal junctional kyphosis are not completely understood. However, the potential growth ability and paraspinal neurofibromatosis invasion might be 2 mechanisms.[22] We found no obvious dystrophic bony changes of the vertebral body and additional space-occupying lesions within the fusion segments of the GR or PSF, whereas adjacent segments were seen in the patients who sustained the complication of adding-on phenomenon postoperatively. Thus, we recommend that the choice of upper and lower instrumentation levels involve considering the extent of dural ectasia and dystrophic anatomy in neurofibromatosis deformities. Furthermore, the fusion segment can be appropriately extended to the distal normal vertebrae.

FIGURE 3. A 16.3-year-old male (no. 32) with right thoracic scoliosis, and the apical vertebra was at T6. He was preoperative American Spinal Injury Association (ASIA) grade C. A and B, Preoperative anteroposterior and lateral radiographs of the spinal column. C and D, Preoperatively the ribs were protruding into the spinal canal from the right intervertebral foramen in the T5, T6, T7 plane, and the rib heads crossed the center line of spinal canal. The distance between the spinal cord and the intraspinal rib head dislocation was 0 mm, and the intraspinal rib proportion was 68.5%. E and F, Postoperative anteroposterior and lateral radiographs of the spinal column. The intraspinal rib head dislocation (T5, T6, 7R) were removed intraoperatively. G and H, Postoperative 36 months' follow-up anteroposterior and lateral radiographs of the spinal column. Partial rib regeneration was found. Function recovered to ASIA grade E after 6 months postoperatively.

We monitored somatosensory evoked potentials, and motor evoked potentials intraoperatively in this study to reduce the risk of spinal cord injury. No new neurological injury or aggravation of neurological symptoms occurred postoperatively in any patient.

This study has several limitations. First, without postoperative MRI, it is not possible to accurately measure the changes in DSCIRH before and after surgery, making it difficult to evaluate safety regarding the spinal cord. Furthermore, 18 patients received GRs, and only 3 underwent subsequent PSF. However, it is unclear whether the other 15 individuals eventually required PSF because these patients were lost to follow-up. Finally, we calculated the IRP through CT images of patients, aiming to be used as a quantitative indicator of the severity of the rib head into the spinal canal, and also to conduct statistical comparison and analysis of important imaging parameters for patients before and after surgery, which has certain clinical value. However, the clinical value of IRP in guiding the choice of surgical treatment really needs further study and analysis.

Overall, the surgical treatment of IRH in children with NF1-DS should be determined on the basis of the presence of preoperative neurological symptoms. This study supports the practice of correcting spinal deformities only in patients with mild or no spinal cord injury. If there are obvious neurological symptoms, IRH resection is necessary to relieve spinal cord compression to recover nerve function.

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