



# Clinical outcomes of anterior correction and reconstruction for neurofibromatosis-associated severe cervical kyphotic deformity

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Received: 8 January 2018 / Accepted: 28 May 2018 / Published online: 9 July 2018  
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## Abstract

**Purpose** To assess the clinical outcome of anterior correction and reconstruction for severe cervical kyphotic deformity due to neurofibromatosis type 1 (NF-1).

**Methods** In this study, we reviewed a series of seven patients who underwent anterior procedures for correction of NF-1-associated cervical kyphotic deformity. After continuous preoperative skull traction, all patients received anterior corpectomy and fusion (ACCF), anterior discectomy and fusion (ACDF), or combined ACCF and ACDF for surgical correction and reconstruction. Pre- and postoperative local and global Cobb angles, correction rate, sagittal vertical axis (SVA), and T1-slope were assessed by X-ray. Japanese Orthopaedic Association (JOA) score, JOA recovery rate, visual analog scale (VAS), and Neck Disability Index (NDI) scores were recorded to assess the outcome.

**Results** Kyphosis was corrected successfully in all patients in terms of local and global Cobb angles ( $P < 0.05$ ), with a correction rates of 83.1% (range, 66.0 to 115.5%) and 88.6% (range, 61.1 to 125.0%), respectively. JOA scores of patients were improved from preoperative 10.4 (range, 6 to 14) to postoperative 15.4 (range, 14 to 17), with a recovery rate as 77.6% (range, 66.7 to 100%). NDI scores were reduced from preoperative 25.1 (range, 13 to 35) to postoperative 8.7 (range, 5 to 12). VAS scores were reduced from preoperative 7.0 (range, 4 to 9) to postoperative 2.3 (range, 1 to 3).

**Conclusion** This study has demonstrated that anterior correction and reconstruction is an alternative option for the treatment of NF-1-associated severe cervical kyphosis when deformity is localized, flexible, or fixed.

**Keywords** Japanese Orthopaedic Association score · Kyphosis · Neurofibromatosis

Neurofibromatosis type 1 (NF-1) is a multi-systematic disease frequently associated with musculoskeletal abnormalities especially spinal deformities. Although cervical kyphosis due to NF-1 is relatively rare in clinical practice, the risk of severe neurological dysfunction including myelopathy and life-threatening paralysis is higher than that in patients with thoracolumbar deformities [1]. Only a few of studies have

reported the surgical treatment for this challenging condition, and there is a lack of related guidelines for the treatment in the literature.

In cases of NF-1-related cervical kyphosis, dystrophic changes including scalloping of vertebral bodies have been shown to contribute to the pathogenesis and progression of deformity [2]. Ventral compression of spinal cord by the dystrophic vertebral bodies may cause directly neurological damage. Kyphotic changes in cervical alignment may also increase the mechanical tension of spinal cord and cause stretch-mediated myelopathy [3]. Anterior decompression and spinal column reconstruction can be achieved via an anterior only approach with appropriate preoperative managements [4–6].

In this paper, we introduce a series of seven patients with severe cervical kyphotic deformity who were treated via the anterior-only approach for the correction and analysis of the clinical and radiological outcomes.

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## Materials and methods

### Clinical data

From January 2007 to October 2015, seven patients with cervical kyphotic deformity due to NF-1 were treated in our institution. They included five males and two females who ranged in age from 16 to 61 years with a mean of 33.1. All patients had a clear history of NF-1 presented with typical café-au-lait spots and neurofibroma. Of them, three patients had a history of surgical resection of neurofibroma. Clinical symptoms and signs including neck pain, myelopathy (motor and sensory deficits of extremities, gait disturbance, hyperreflexia and pathological reflexes, bowel/bladder symptoms, and sexual dysfunction), and radicular symptom were recorded (Table 1).

Pre- and postoperative neurological functions were assessed by the Japanese Orthopaedic Association (JOA) score. The JOA scoring system consists of seven categories: motor function of fingers, shoulder and elbow, and lower extremity; sensory function of upper extremity, trunk and lower extremity; and function of the bladder, with the minimum total score being 0 and the maximum total score being 17 [7]. Severity of neck pain was assessed by the 10-point visual analog scale (VAS). A rating of 0 point meant no pain, and 10 points was “very severe pain” [8].

The self-reported neck disability of patients was measured by Neck Disability Index (NDI) scores. The NDI consists of ten items: neck pain intensity, self-care, lifting, reading, headache, concentration, work, driving, sleeping, and recreation. For each item, 6 detractor statements ordered for gradually increasing of severity from 0 (no problem) to 5 (worst problem). The total NDI score ranges from 0 to 50 [9].

### Imaging evaluation

All patients received cervical X-ray (anteroposterior/lateral plain radiographs and dynamic flexion/extension

radiographs), thin-cut computerized tomography (CT) scan, and magnetic resonance imaging (MRI) pre- and postoperatively. The global and local cervical curvatures were measured on lateral plain radiographs using the Cobb method [10]. Global Cobb angle was defined as the angle between inferior endplates of C2 and C7 vertebrae, and the local Cobb angle was defined as the angle of the kyphotic segments [11]. Positive values were assigned to the lordotic curvature and negative values to the kyphotic curvature. Dynamic flexion/extension radiographs were taken to evaluate the overall flexibility of the cervical spine. Fixed kyphosis was defined by less than 50% reduction of deformity on dynamic flexion-extension radiographs and segmental ankyloses visualized on CT scan [12]. Sagittal balance of the cervical spine was assessed by sagittal vertical axis (SVA) and T1-slope [13]. Further investigation of the intraspinal contents and compressive pathological features was obtained on MRI for all patients.

### Pre- and intraoperative skull traction

Prior to surgery, all the patients underwent continuous skull traction for six to ten days starting with 4 kg and gradually increased to the maximum weight as one eighth of the body weight. During the traction period, bedside cervical X-ray was taken for every three days to assess the correction of kyphosis.

For the intraoperative traction, the patient was positioned supine on the surgery table and underwent general anesthesia with adequate muscle relaxant. A shoulder pillow was placed beneath the scapula to allow a slight neck extension. Intraoperative skull traction (with one eighth of the body weight) was performed to maintain the neck extension in all the patients under the monitoring of MEPs and SSEPs. In most cases, a partial correction could be obtained by position placing and continuous traction under of general anaesthesia and muscle relaxation.

**Table 1** Demographic and surgical data of patients

Case	Age (years), sex	Chief complain	Flexibility	Surgery procedures	Operation time (min)	Blood loss (ml)
1	16, male	Progressive cervical kyphosis with a history of resection of intraspinal tumor	Flexible	C3–C4 ACCF + C2/3, C4/5 ACDF	155	125
2	35, male	Paresis of upper limbs and gait disturbance	Fixed	C5–7 ACDF	120	300
3	49, male	Numbness of limbs and gait disturbance	Fixed	C5–6 ACCF	135	100
4	17, male	Weakness of limbs and gait disturbance with a history of resection of neurofibroma in the postauricular region	Fixed	C5–6 ACCF + C4/5 ACDF	150	135
5	25, male	Neck pain and numbness of the right arm	Fixed	C5 ACCF + C6/7 ACDF	180	215
6	29, female	Neck pain and gait disturbance	Flexible	C5–6 ACCF	145	185
7	61, female	Weakness of limbs and numbness of arms	Fixed	C5–6 ACCF	135	200

## Surgical procedures

Exposure was made via the classical anterior Smith-Robinson approach. Corpectomy was usually designed to be performed at the dysplastic vertebrae. After placement of Caspar pins perpendicular to the anterior plane of the vertebrae above and below the identified segments, intervertebral discs were resected to release the anterior structure. The Caspar distractor and intervertebral spreader were used simultaneously to gradually dilate the space. The great part of dysplastic vertebrae was resected with a set of rongeurs. The Caspar pins were distracted under the continuous skull traction to produce lordosis. The position of pillows beneath neck and shoulders was adjusted to help to extension of the cervical spine. The titanium mesh cage filled with the autogenous bone graft along with the pre-curved plate was used for reconstruction (Fig. 1). The bone graft was harvested from local vertebrae or autogenous ilium. In some cases with severe and extensive kyphosis, corpectomy combined with additional 1- or 2-level discectomies and fusion was performed to provide a multi-level correction (Fig. 2).

In one case with severe ankylosis, a two-stage anterior release and fusion was performed. At the first stage, the patient

underwent an anterior procedure to release the contractual soft tissue and resect the pre vertebral osteophyte and degenerated intervertebral discs. After wound closure, the patient received continuous skull traction for seven days. During the second stage, anterior correction and intervertebral fusion with poly-ether-ether-ketone (PEEK) cage via primary incision was performed.

Postoperatively, all patients were prescribed to wear an individually customized sternal-occipital-mandibular-immobilizer brace for eight weeks to maintain the correction and support bone healing.

## Follow-up observations

Clinical and radiological follow-ups were performed two, six, and 12 months postoperatively and annually thereafter. All follow-up assessments were performed by two orthopaedic surgeons who were blinded to the surgical procedure.

During the follow-up period, cervical X-rays including anteroposterior, lateral, and dynamic radiographs were performed. Global and local Cobb angle of cervical spine, SVA, and T1-slope were measure as described previously.

The correction rate of Cobb angle was calculated as:

$$(\text{Postoperative Cobb angle} - \text{preoperative Cobb angle}) / \text{Preoperative Cobb angle} \times 100\%$$

A correction rate greater than 100% means that the cervical spine was corrected from a kyphotic curvature to a lordotic one.

NDI and VAS scores were recorded to assess the neck function. The JOA scores and JOA recovery rate were used to assess the improvement of neural function. The recovery rate was evaluated by Hirabayashi's formula [14]:

$$(\text{Postoperative JOA score} - \text{preoperative JOA score}) / \text{Preoperative JOA score} \times 100\%$$

## Statistical analysis

Statistical comparisons were performed with a chi-square test for categorical parameters and unpaired or paired *t* tests for continuous variables. *P* values of <0.05 were considered statistically significant.

## Results

### Surgical procedure

All procedures were performed by one senior surgeon. Average operation time was 145 minutes (range, 120 to

180 minutes) with blood loss of 180 ml (range 100 to 300 ml). Three patients underwent anterior cervical corpectomy and fusion (ACCF); three patients received ACCF hybrid with anterior cervical discectomy and fusion (ACDF), and the remaining patient received two-stage ACDF.

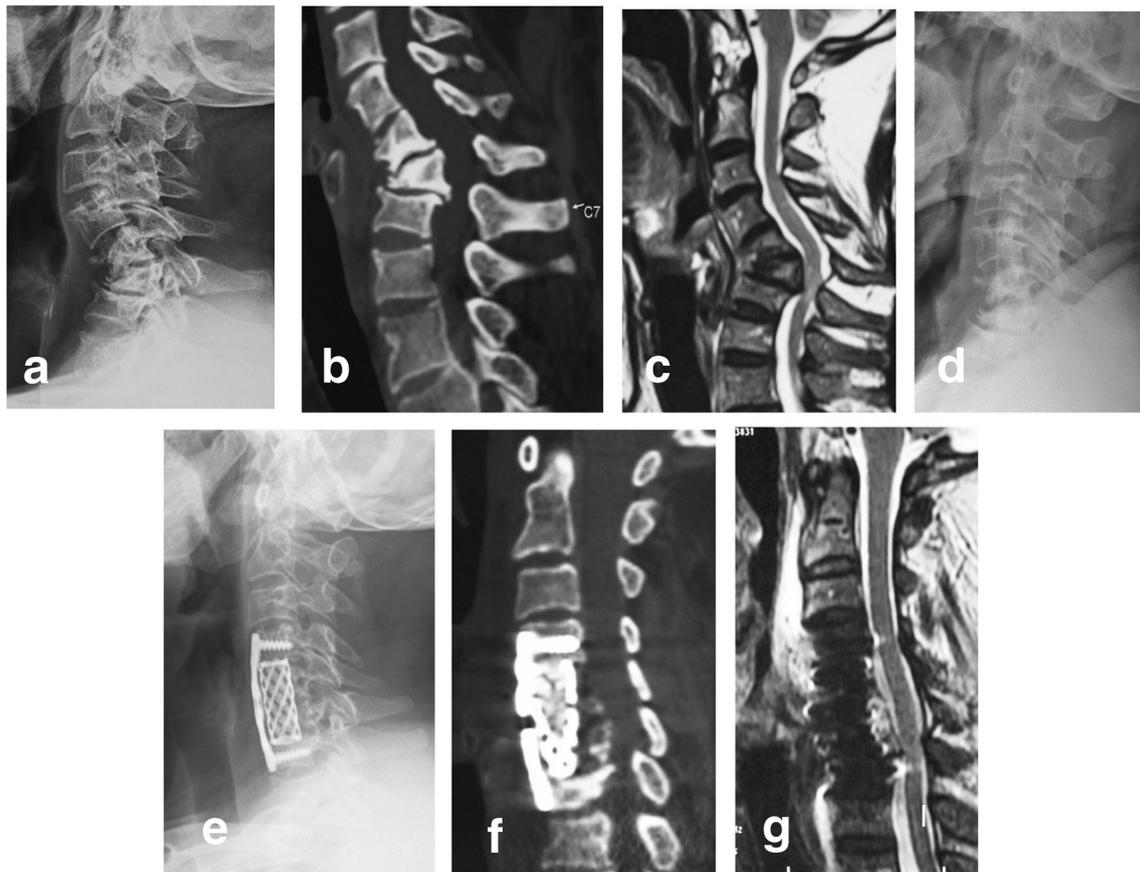
### Clinical outcomes

All patients had achieved satisfactory relief of neurological symptoms. JOA scores of these patients were improved from 10.4 (range, 6 to 14) preoperatively to 15.4 (range, 14 to 17) postoperatively, with a recovery rate of 77.6% (range, 66.7 to 100%).

NDI scores were reduced from 25.1 (range, 13 to 35) to 8.7 (range, 5 to 12). VAS scores were reduced from 7.0 (range, 4 to 9) to 2.3 (range, 1 to 3) (Table 3). Patients with severe preoperative neck pain had achieved effective pain relief at the final follow-up.

### Radiographic outcomes

Correction of kyphosis was achieved in all patients in terms of local and global Cobb angle (*P* < 0.05). The mean preoperative local Cobb angle was  $-67.7^\circ$  (range,  $-35^\circ$  to  $-110^\circ$ ), and the mean preoperative global Cobb angle was  $-39.4^\circ$  (range,  $-20^\circ$  to  $-68^\circ$ ). After continuous traction prior to



**Fig. 1** Case 3, a 49-year-old male, presented with numbness of limbs and gait disturbance due to myelopathy caused by neurofibromatosis-associated cervical kyphotic deformity. Preoperative X-ray (a) and CT scan (b) showing significant scallop of C5 and C6 vertebrae. The local and global Cobb angle was  $-58^\circ$  and  $-20^\circ$ , respectively. MRI shows compression of spinal cord at C6 level (c). After 7-day continuous skull

traction, the local and global Cobb angle reduced to  $-32^\circ$  and  $-12^\circ$  (d). Anterior correction and reconstruction was performed via C5–6 corpectomy and fusion. At the time of 1-year follow-up, the postoperative local and global Cobb angle was  $9^\circ$  and  $5^\circ$ , respectively (e, f). MRI shows thoroughly decompression of the spinal cord (g). JOA score of this patient increased from preoperative 10 to postoperative 16

surgery, the local and global kyphotic angles were reduced to  $-48.6^\circ$  (range,  $-19^\circ$  to  $-70^\circ$ ) and  $-22.4^\circ$  (range,  $-12^\circ$  to  $-38^\circ$ ), respectively. At the time of final follow-up, the postoperative local and global Cobb angles were  $-12.4^\circ$  (range,  $9^\circ$  to  $-20^\circ$ ) and  $-6.9^\circ$  (range,  $5^\circ$  to  $-21^\circ$ ), respectively, and local and global correction rates were 83.1% (range, 66.0 to 115.5%) and 88.6% (range, 61.1 to 125.0%), respectively (Table 2).

Sagittal balance of cervical spine was effectively restored in all patients. SVA was increased from  $-3.0$  cm (range,  $-29.1$  to  $6.6$  cm) to  $5.3$  cm (range,  $-17$  to  $16.5$  cm), and T1-slope was increased from  $-27.9^\circ$  (range,  $-66^\circ$  to  $14^\circ$ ) to  $4.1^\circ$  (range,  $-7$  to  $24$ ) (Table 3).

### Postoperative complications

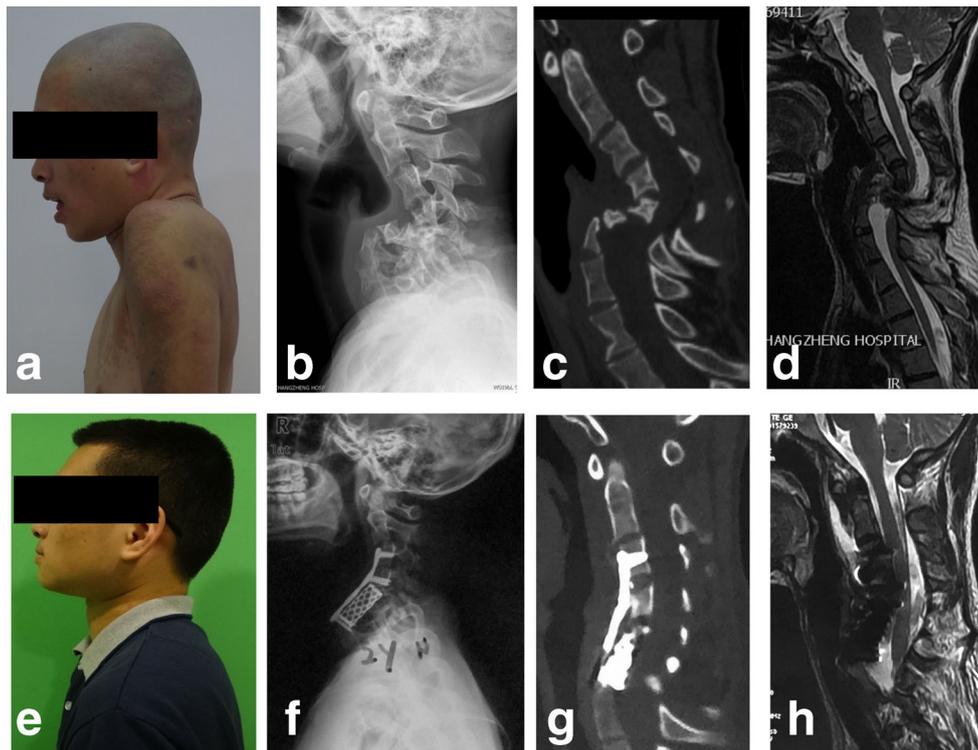
Epidural haematoma occurred in one of the seven cases due to postoperative intermittent bleeding. Emergency evacuation of the haematoma was performed and no neurological deterioration occurred. No neurological complications or surgical site

infection was obtained. At the final follow-up, all seven patients had solid bony fusion. There were no patients with failure of internal fixation.

### Discussion

Neurofibromatosis is a multi-systematic disease in which spinal deformity is commonly involved [10, 15]. Thoracolumbar deformities including scoliosis and kyphosis are frequently reported in literature and the treatment strategies are also more mature than that of cervical deformities [16, 17]. Cervical abnormalities of NF-1 include vertebral scalloping, enlarged neural foramina, defective pedicles, cervical kyphotic deformity, spondylolisthesis, or dislocation. Most cervical deformity cases presented sharply angular kyphosis with a high risk of severe neurological dysfunction which usually requires surgical intervention [2, 3, 18].

Although several surgical techniques have been described in known studies, no clear and generally accepted guidelines



**Fig. 2** Case 4, a 17-year-old boy (a), presented with severe cervical kyphotic deformity and myelopathy (including paresis of the limbs, gait disturbance, and bladder dysfunction). Preoperative X-ray and CT showed significant dystrophic changes in cervical spine with a local and global Cobb angle of  $-110^\circ$  and  $-68^\circ$  (b, c). MRI shows significant spinal cord compression (d). After 9-day continuous skull traction, the

patient underwent anterior correction and reconstruction via combined corpectomy and discectomy and fusion. At the time of 2-year follow-up, the postoperative local and global Cobb angle reduced to  $-20^\circ$  and  $-21^\circ$ , respectively (e–g). The spinal cord was decompressed sufficiently (h). The neurological function was improved significantly, with the JOA score which increased from preoperative 6 to postoperative 15

are currently available for the treatment of NF-associated cervical kyphosis. Most authors preferred to use combined anterior-posterior procedures for thorough decompression and circumferential reconstruction [1, 19–21]. The posterior-only approach was also reported in some studies [1, 22]. Helenius et al. [1] used the posterior-only approach in 9 of their 22 cases of NF-associated kyphosis, with a mean correction rate of 58%, and the anterior-posterior approach in the

remaining 13 cases, with a mean correction rate of 83%, and they therefore concluded that the anteroposterior approach was an optimal option. Ma et al. [19] reported seven patients who underwent anteroposterior correction with satisfied outcomes and therefore believed that a more aggressive anteroposterior spinal fusion procedure was the most reliable surgical strategy for cases with a kyphotic angle greater than  $50^\circ$ . Despite the reliable correction rate, surgical invasion and

**Table 2** Local and global Cobb angles of cervical spine

Case	Local Cobb angle					Global Cobb angle				
	Preoperative	Post-traction	Postoperative	Correction degree	Correction rate (%)	Preoperative	Post-traction	Postoperative	Correction degree	Correction rate (%)
1	-71	-58	-24	47	66.2	-41	-34	-15	26	63.4
2	-65	-51	-11	54	83.1	-38	-16	2	40	105.3
3	-58	-32	9	67	115.5	-20	-12	5	25	125.0
4	-110	-70	-20	90	81.8	-68	-24	-21	47	69.1
5	-82	-65	-21	61	74.4	-47	-38	-9	38	80.9
6	-35	-19	-2	33	94.3	-26	-12	4	30	115.4
7	-53	-45	-18	35	66.0	-36	-21	-14	22	61.1
Mean	-67.7	-48.6	-12.4	55.3	83.1	-39.4	-22.4	-6.9	32.6	88.6

**Table 3** Symptomatic data and sagittal balance parameters of patients

Case	JOA score		Recovery rate (%)		NDI score		VAS		SVA (cm)		Tl-slope	
	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative
1	11	15	66.7	8	35	8	9	3	5.4	12.4	-15	12
2	9	14	62.5	8	21	8	7	2	2.1	9.4	-27	-6
3	10	16	87.7	5	13	5	4	1	4.3	3.5	-14	5
4	6	15	81.8	12	37	12	9	3	-13	-3.4	-66	24
5	14	17	100	9	28	9	8	2	-29.1	-17	-23	-3
6	12	16	80.0	11	26	11	7	2	2.4	15.6	-19	4
7	11	15	66.7	8	16	8	5	3	6.6	16.5	-31	-7
Mean	10.4	15.4	77.6	8.7	25.1	8.7	7.0	2.3	-3.0	5.3	-27.9	4.1

postoperative complications of posterior procedure should not be ignored. Apart from the prolonged operation time and considerable blood loss, the posterior procedure may also cause damage to the nuchal muscle and bony structure, leading to iatrogenic kyphosis and axial pain [19, 20]. Additionally, postoperative complications including C5 palsy, surgical site infection, graft dislocation, severe pneumonia, and even pulmonary embolus [1, 21, 23] may have negative effects on the prognosis.

The single anterior approach was described in very few cases and suggested only in moderate and flexible kyphosis [19, 24]. Choksey and Modi [24] presented a case of 13-year-old boy with 46° kyphosis who underwent a multi-level anterior corpectomy with fibular cortical grafting and plate fixation, showing a satisfactory correction outcome and full neurological recovery postoperatively.

Kyphotic deformity associated with NF-1 occurred more frequently with short and sharp dysplastic curves than with long curves resembling the idiopathic ones. The apex of kyphosis was more likely to be obtained at the segments with the dystrophic vertebrae [3, 21]. The anterior approach allows for direct access to the ventral pathological structures. Resection of the dystrophic vertebral bodies and degenerated discs along with multi-level distractions and instrumental reconstruction allows for adequate restoration of alignment. For severe and extensive cases, we usually chose corpectomy combined with 1- or 2-level discectomy for multi-level distraction and to provide additional anchor points for correction and stabilization.

For better surgical correction, continuous preoperative cervical traction is usually suggested [25]. Preoperative traction helps partially correct kyphosis and release the cervical structure to make the rigid kyphosis flexible and provide an easier ventral approach [1]. Combined with cervical traction, correction can be obtained gradually, which makes the surgical procedure less difficult with less operative risk. In cases of NF-1-associated kyphosis, a smaller traction force is usually suggested due to the fragility of the dystrophic cervical spine [21]. In the present study, we recommend initiating the traction from 4 kg and then increasing the weight gradually to the maximal one eighth of the body weight. Intraoperative skull traction was used to maintain neck extension and ease the surgical procedure. In addition, adjusting patients' position with pillows beneath the head and scapula during the surgery can provide an extensional traction force, thus facilitating the correction procedure. In our series of cases, the patients presented with various degrees of kyphosis from 35° to 110°, preoperatively. After the anterior-only decompression and reconstruction, the neurological symptoms were relieved satisfactorily in all cases, with a mean correction rate of 83.1%.

Iatrogenic neurological injury during corrective surgical manoeuvres is one of the most severe complications in the treatment of NF-1-associated cervical kyphosis [21], probably due to the increased compressive force on the apex of

kyphosis, stretch stress on the spinal cord, or tethering of nerve root during restoration of the cervical lordosis [1, 26]. Changes in neural functions should be observed carefully via intraoperative neuro-monitoring to achieve a gradual and safe correction [27]. Although greater kyphosis correction is usually correlated with an improved neurological outcome, the risk of neural injury associated with aggressive correction is significant. Therefore, expected degree of correction should be carefully designed and weighted between the benefits of possible neurological improvement and the accompanying risk. According to this study and the results reported in previous papers [1, 11, 28–30], we are prudent to suggest that a correction rate about 80% should be an adequate and safe degree of correction.

Difficulty to obtain solid bone fusion due to the poor bone quality of patients with neurofibromatosis is another main concern [21]. Most previous studies recommended circumferential instrumental fixation or autogenous bone grafting such as fibula strut grafting [1]. In our series, we carefully assessed the bone quality of the segments for placing the internal fixation system and bone graft. In cases with massive bone defect, autogenous iliac grafts were used. An individually customized sternal-occipital-mandibular-immobilizer brace was also used to confirm the postoperative bone healing.

Proximal junctional kyphosis (PJK) is a long-term complication that has been reported in several studies [1]. The occurrence of PJK may be due to newly developed vertebral erosion or continued growth of posterior elements, which may lead to surgical revision in case of a neural defect. Helenius et al. [1] suggested that instrumentation should span all dystrophic and kyphotic segments to prevent junctional issues. In our series, we performed corpectomy on dysplastic segments and anterior fusion on kyphotic segments. Although no junctional kyphosis was observed, long-term prognosis still needs to be assessed.

Despite the significant advantages of anterior approaches, the indications should be seriously considered. Previous studies suggested that the anterior-only strategy should be used when the deformity was flexible or fixed without ankylosis of the facet joint [24]. In our series, release via preoperative skull traction proved to increase the flexibility and partially reduce the cervical kyphosis, even in some fixed cases. Therefore, we suggest re-assessing the flexibility and reducibility after continuous skull traction and carefully consider the indications of different approaches. In cases of localized kyphosis due to dystrophic changes of vertebral bodies, correction can be achieved and well maintained via anterior resection of pathological structure and reconstruction. For these cases, the anterior-only approach seems to be an alternative option. However, meticulous long-term follow-up observation should be abided in any case to discover the sign of progressing vertebral erosion.

**Funding** This study is supported by a grant from National Natural Science Foundation of China (81501918).

No relevant financial activities outside the submitted work.

## Compliance with ethical standards

The manuscript submitted does not contain information about medical device(s)/drug(s).

**Conflict of interest** The authors declare that they have no conflict of interest.

**Ethical approval** Our research was approved by the ethics department of Changzheng Hospital, Shanghai. We have consensus with all participants. We also followed the Declaration of Helsinki and relevant policies in China.

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