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Treatment for paraplegia due to severe kyphoscoliosis associated with neurofibromatosis type 1 via halo-pelvic traction: a case report

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Abstract

Background A sharply angular thoracic deformity, commonly seen in dystrophic scoliosis associated with neurofibromatosis type 1, can compress the spinal cord and potentially cause neurological impairment. However, managing paraplegia due to severe kyphoscoliosis associated with neurofibromatosis type 1, coupled with low body mass index and extremely severe kyphoscoliosis, presents a significant challenge.

Case presentation A 13-year-old girl of Mongolian ethnicity with severe dystrophic kyphoscoliosis associated with neurofibromatosis type 1 presented with paraplegia and dyspnea. Preoperative radiograph imaging revealed the presence of a thoracic kyphosis and scoliosis, with a Cobb angle of 150° and 130°, respectively. A two-stage strategy was devised, comprising halo-pelvic traction and spinal fusion with pedicle screws. The neurological deficit showed gradual improvement and ultimately complete recovery during the distraction phase. The curve decreased to an acceptable level, and posterior pedicle screws were implanted and fused without osteotomy. Postoperatively, the hunchback was no longer visible. There were no complications associated with halo-pelvic traction. At the 3-year follow-up, the correction angle and trunk balance were well maintained.

Conclusion It is possible that neurological deficit resulting from severe scoliosis may be reversed following the correction of the spinal curvature. The application of halo-pelvic traction generates substantial corrective forces, facilitating the correction of severe spinal deformities in a gradual and secure manner. A two-stage treatment strategy for patients with severe kyphoscoliosis in neurofibromatosis type 1 may offer an alternative approach to correcting the severe curve while avoiding the potential complications associated with a rapid, one-stage correction.

Keywords Dystrophic kyphoscoliosis, Neurofibromatosis type 1, Paraplegia, Halo-pelvic traction

Background

Neurofibromatosis type 1 (NF1) is a rare autosomal dominant disorder with an approximate morbidity of 1/3000 [1–3]. The disorder presents with a range of distinctive lesions, including café-au-lait spots, neurofibromas, Lisch nodules, and skeletal disorders [1, 4, 5]. The most frequently observed skeletal abnormality is scoliosis, which can be classified as either dystrophic or nondystrophic in individuals with NF1. Nondystrophic scoliosis is comparable to the more common idiopathic scoliosis,

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and the recommended treatment strategy is applicable in both cases [1, 4]. In contrast, dystrophic scoliosis is a progressive condition, characterized by the presence of sharply angulated short curvatures, rib penciling, elongated and attenuated pedicles, and widened spinal canal and foramen [6, 7]. Early surgical intervention has been recommended for dystrophic scoliosis, although progression may occur despite fusion [8, 9].

We report the case of a girl of Mongolian ethnicity who presented a sharply angular thoracic dystrophic kyphoscoliosis associated with NF1, paraplegia, and poor pulmonary function, with excellent clinical outcome. An approved consent form was signed by the patient and her family before any testing was performed. Approval was secured from the Institutional Review Board.

Case presentation

In 2014, the senior author initially encountered a 13-year-old girl of Mongolian ethnicity with a known diagnosis of NF1-related scoliosis. The diagnosis of dystrophic kyphoscoliosis associated with NF1 was established when the patient was 5 years old, yet no further treatment was initiated at that time. The scoliosis curve progressed gradually throughout the patient's growth. Bilateral lower extremity paresis had emerged 2 months prior to the patient's presentation to the senior author. The patient's mother was diagnosed with neurofibromatosis without scoliosis. She exhibited low self-confidence and was reluctant to engage in social activities.

On admission, the patient presented with paraplegia at the T12 level and dyspnea. The results of the physical examination indicated the presence of dystrophy, multiple café-au-lait spots, and myelopathy (Fig. 1). Preoperative radiographic examination revealed the presence of thoracic kyphosis and scoliosis, with a Cobb angle of 150° and 130°, respectively(Figs. 2, 3). It is possible that the paraplegia is a result of compression caused by severe kyphoscoliosis rather than displacement of the ribs (Fig. 4). A halo-pelvic traction apparatus was fitted to the patient (Fig. 5) as a provisional measure while the patient was unable to tolerate corrective surgery to release the compression of the spinal cord. Gradually, distraction (5 mm each week) was carried out after the patient had adapted to the apparatus. Consequently, the neurological deficit status was meticulously monitored throughout the distraction process.

Following a period of 2 months during which the patient was distracted from the injury, the neurological deficit showed signs of improvement. After nearly 6 months, the patient had made a complete recovery. It was imperative that the patient engage in daily physical exercise and respiratory training throughout the period of distraction. The surgical procedure was scheduled to be

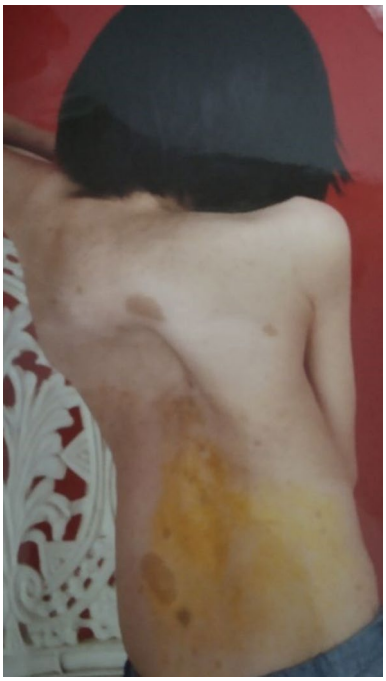

Fig. 1 A preoperative photograph showing multiple neurofibromas and café-au-lait spots

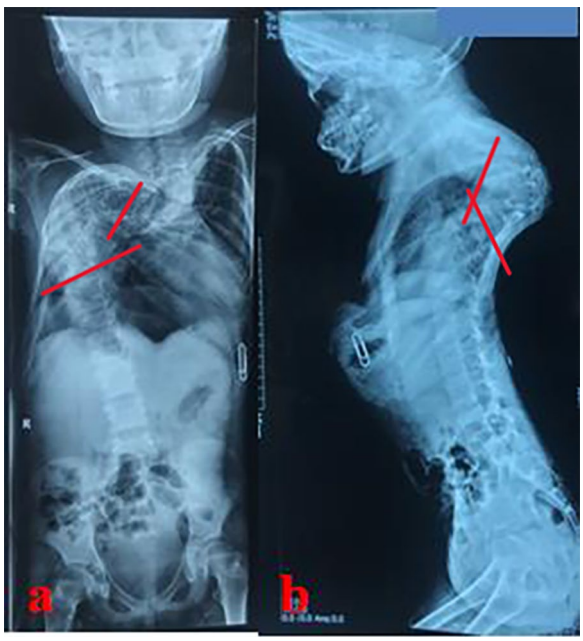

Fig. 2 Anteroposterior (**a**) and lateral (**b**) radiographs demonstrate the sharply angular thoracic scoliosis and kyphsis, with a Cobb angle of 150° and 130°, respectively. The red lines mark the upper and lower end vertebrae of the selected measurement angle

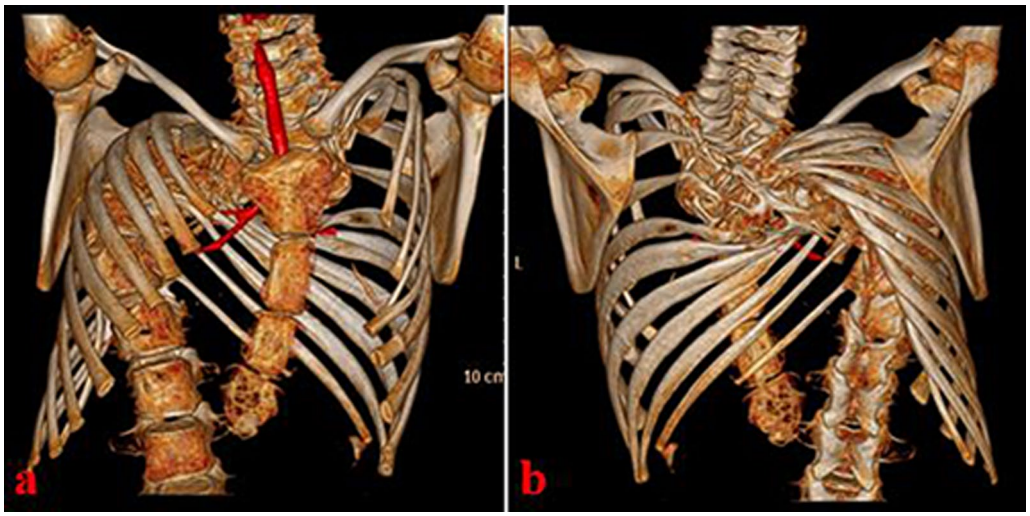

Fig. 3 Anterior (**a**) and posterior (**b**) three-dimensional reconstruction using a computed tomography scan

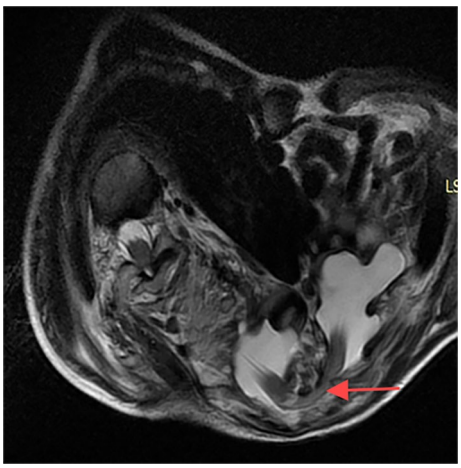

Fig. 4 Thoracic spinal cord on magnetic resonance imaging. The red arrow towards the thoracic spinal cord

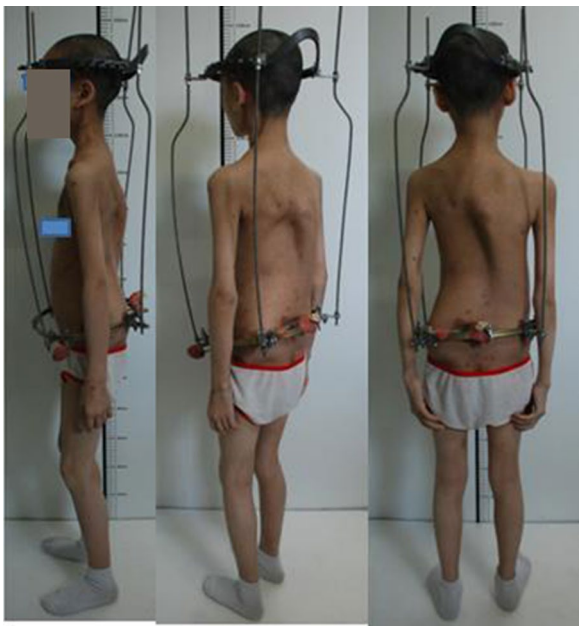

Fig. 5 The huge change in appearance during the distraction

performed only when the patient's respiratory function had reached a sufficient level to avoid the occurrence of severe pulmonary complications during the perioperative period. The combined intervention of respiratory training, physical exercise, and distraction took approximately 1 year to complete. Given the favorable reduction in the Cobb angle (Figs. 6, 7), the decision was made to proceed with the implantation of posterior pedicle screws and fusion without a three-column osteotomy. Subsequent to the surgical procedure, the hunchback was no longer discernible (Figs. 8, 9).

During the treatment, no specific complications were observed in relation to halo-pelvic traction. Subsequently, the patient made a full recovery from the disease. At the 3-year follow-up, the correction angle and trunk balance were both well maintained (Fig. 10).

Discussion

Paraplegia is rarely seen in patients with dystrophic scoliosis in NF1 [10, 11]. The potential risk of paraplegia may be the penetration of ribs into the spinal canal, with or without a traumatic episode at any time [10]. Mukhtar and Letts reported a staged strategy comprising rib excision of the protruding rib initially and

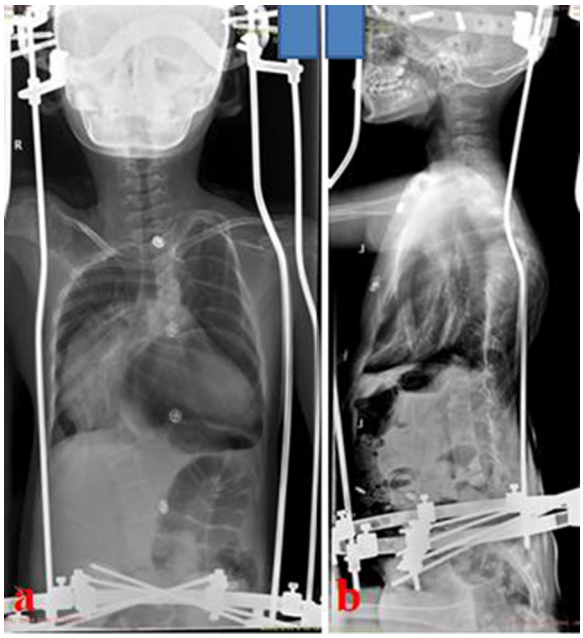

Fig. 6 Anteroposterior (**a**) and lateral (**b**) radiographs demonstrated that the scoliosis and kyphosis curve was improved through the distraction

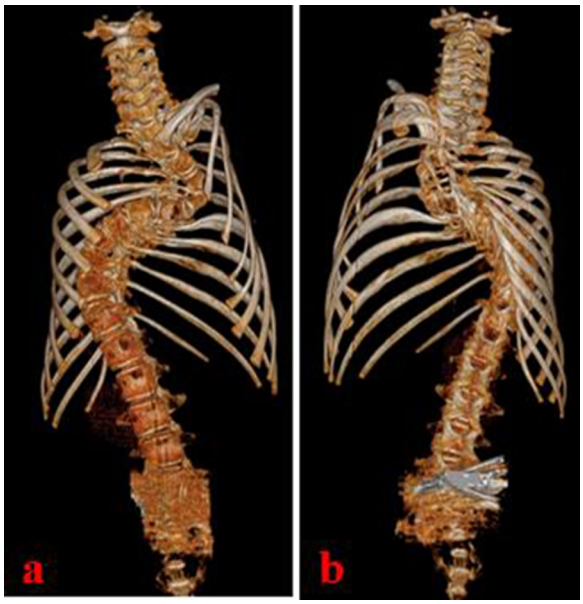

Fig. 7 Three-dimensional reconstruction using computed tomography of spine after the distraction

subsequent spinal fusion, with the objective of avoiding neurological deficit during the preoperative period [12]. A short and sharply angular thoracic deformity, which is a common dystrophic scoliosis associated with NF1, may directly compress the spinal cord, potentially resulting

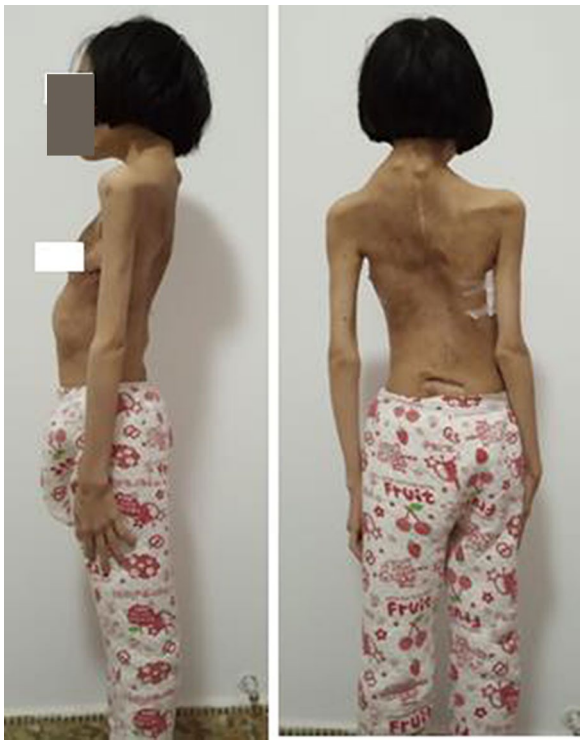

Fig. 8 Appearance of the spine after the entire treatment

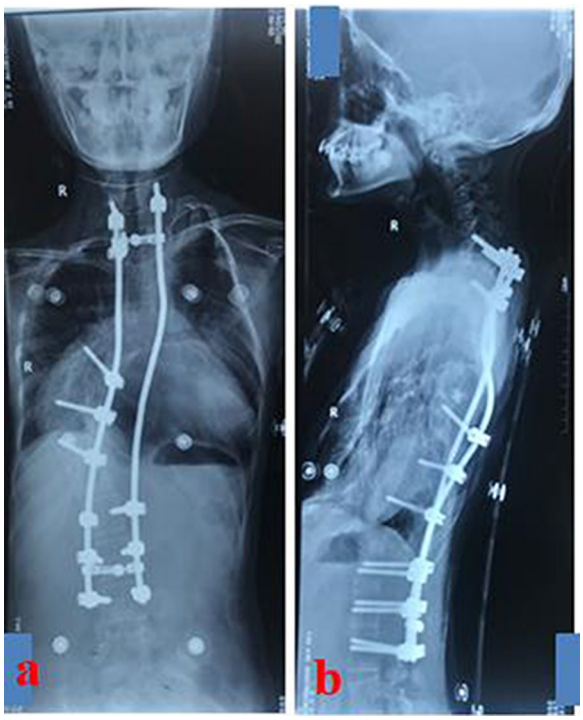

Fig. 9 Postoperative anteroposterior (**a**) and lateral (**b**) radiographs

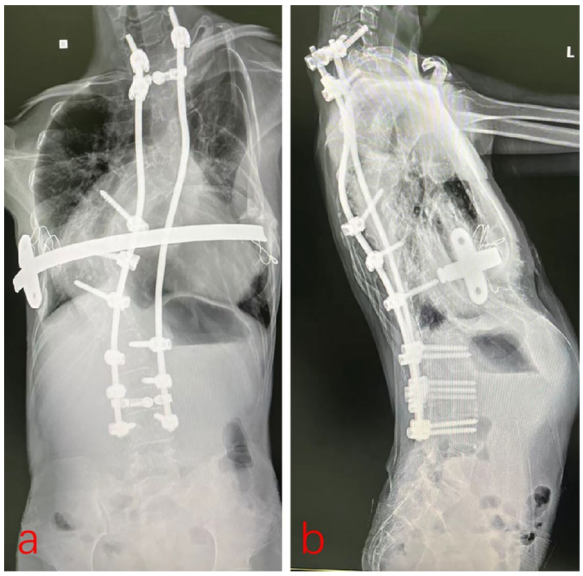

Fig. 10 Anteroposterior (**a**) and lateral (**b**) radiographs at 3-year follow-up

in neurological deficit. Stoker *et al.* reported one case in which the patient had been diagnosed with neurological deficit on admission and underwent halo-gravity traction and posterior vertebral column resection (pVCR) thereafter. Ultimately, satisfactory curve correction and neurological function recovery were achieved [13].

Severe dystrophic scoliosis due to NF1 presents a significant challenge to surgical correction. Furthermore, additional risks may be present, including the potential for bleeding due to hypervascularity associated with the neurofibroma, respiratory compromise secondary to their deformity, progression, and hyperkyphosis uredespite fusion [9, 13, 14]. A circumferential fusion via a combined anteroposterior approach has been demonstrated to be an effective procedure [4]. Sing *et al.* reported a simultaneous anterior–posterior approach for vertebrectomy and fusion for patients with severe dystrophic kyphoscoliosis in NF1. This approach allows for proper visualization of the spinal cord, achievement of solid arthrodesis, and minimization and prevention of deformity progression [15]. Posterior vertebral column resection (pVCR) has been demonstrated to be an effective treatment for severe deformities in recent years [16]. However, the high incidence of complications may prove disadvantageous to pVCR, particularly in the absence of extensive experience among the surgical team [16, 17].

In this case, numerous challenges were encountered, including a low body mass index (BMI), markedly severe kyphoscoliosis, significant pulmonary dysfunction, and paraplegia. We were dedicated to identifying effective treatment options for this patient. A two-stage strategy, including halo-pelvic traction and spinal fusion with pedicle screws, was employed to correct the curve and improve the patient's pulmonary dysfunction. Paraplegia completely recovered during the distraction process. Following the distraction, the correction of the scoliosis and kyphosis curves was satisfactory, and there was no requirement of osteotomy to reconstruct the coronal and sagittal balance. It can therefore be concluded that this two-stage surgical strategy represents a safe and effective treatment option for severe kyphoscoliosis associated with NF1.

Conclusion

The halo-pelvic traction technique is an effective method for producing high corrective forces to correct the severe spinal deformity in a slow and safe manner. In this case, paraplegia due to severe kyphoscoliosis resolved gradually following the correction of kyphoscoliosis via halo-pelvic traction. The two-stage treatment strategy for patients with severe kyphoscoliosis in NF1 may offer an alternative approach to correcting the severe curve while avoiding the potential complications associated with a rapid, one-stage correction.

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Author contributions
Conceptualization—DZ and YJL; methodology—YXL, FYY, and YLY; formal analysis—YXL; investigation—YXL, FYY, and YLY; writing—original draft—YXL and DZ; writing—review and editing—DZ.

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Availability of data and materials
The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
This study was approved by the Institutional Review Board of the Third People's Hospital of Chengdu. All authors critically revised and reviewed the article and approved the final version of the article before submission. All authors read and approved the final article.

Consent for publication
Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
There are no competing interests to declare.

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References

1. Gutmann DH, Ferner RE, Listernick RH, Korf BR, Wolters PL, Johnson KJ. Neurofibromatosis type 1. Nat Rev Dis Primers. 2017. https://doi.org/10.1038/nrdp.2017.4.
2. Uusitalo E, Leppävirta J, Koffert A, Suominen S, *et al*. Incidence and mortality of neurofibromatosis: a total population study in Finland. J Invest Dermatol. 2015;135(3):904–6.
3. Akbarnia BA, Gabriel KR, Beckman E, Chalk D. Prevalence of scoliosis in neurofibromatosis. Spine. 1992;17(8 Suppl):S244-248.
4. Tsirikos AI, Saifuddin A, Noordeen MH. Spinal deformity in neurofibromatosis type-1: diagnosis and treatment. Eur Spine J. 2005;14(5):427–39.
5. Korf BR. Diagnosis and management of neurofibromatosis type 1. Curr Neurol Neurosci Rep. 2001;1(2):162–7.
6. Crawford AH, Herrera-Soto J. Scoliosis associated with neurofibromatosis. Orthop Clin North Am. 2007;38(4):553–62.
7. Winter RB, Moe JH, Bradford DS, Lonstein JE, Pedras CV, Weber AH. Spine deformity in neurofibromatosis. A review of one hundred and two patients. J Bone Joint Surg Am. 1979;61(5):677–94.
8. Tauchi R, Kawakami N, Castro MA, Ohara T, *et al*. (2017) Long-term Surgical Outcomes After Early Definitive Spinal Fusion for Early-onset Scoliosis With Neurofibromatosis Type 1 at Mean Follow-up of 14 Years. J Pediatr Orthop
9. Parisini P, Di Silvestre M, Greggi T, Paderni S, Cervellati S, Savini R. Surgical correction of dystrophic spinal curves in neurofibromatosis. A review of 56 patients. Spine. 1999;24(21):2247–53.
10. Khoshhal KI, Ellis RD. Paraparesis after posterior spinal fusion in neurofibromatosis secondary to rib displacement: case report and literature review. J Pediatr Orthop. 2000;20(6):799–801.
11. Zhao X, Li J, Shi L, Yang L, *et al*. Surgical treatment of dystrophic spinal curves caused by neurofibromatosis type 1: a retrospective study of 26 patients. Medicine. 2016;95(14): e3292.
12. Mukhtar IA, Letts M, Kontio K. Spinal cord impingement by a displaced rib in scoliosis due to neurofibromatosis. Can J Surg. 2005;48(5):414–5.
13. Stoker GE, Lenke LG, Dorward IG. Posterior vertebral column resection for the treatment of dystrophic kyphosis associated with type-1 neurofibromatosis: a case report and review of the literature. Spine. 2012;37(26):E1659-1664.
14. Murray PM, Weinstein SL, Spratt KF. The natural history and long-term follow-up of Scheuermann kyphosis. J Bone Joint Surg Am. 1993;75(2):236–48.
15. Singh K, Samartzis D, An HS. Neurofibromatosis type I with severe dystrophic kyphoscoliosis and its operative management via a simultaneous anterior-posterior approach: a case report and review of the literature. Spine J. 2005;5(4):461–6.
16. Lenke LG, Sides BA, Koester LA, Hensley M, Blanke KM. Vertebral column resection for the treatment of severe spinal deformity. Clin Orthop Relat Res. 2010;468(3):687–99.
17. Yang C, Zheng Z, Liu H, Wang J, Kim YJ, Cho S. Posterior vertebral column resection in spinal deformity: a systematic review. Eur Spine J. 2016;25(8):2368–75.

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