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Surgical Treatment of Dystrophic Spinal Curves Caused by Neurofibromatosis Type 1

A Retrospective Study of 26 Patients

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Abstract: Dystrophic scoliosis in neurofibromatosis type 1 (NF-1) is difficult to treat. The purpose of this study was to review the clinical and radiological outcome of surgical treatment of dystrophic spinal curves in NF-1, for analyzing its efficacy, safety, and possible complications.

This retrospective study consisted of 26 NF-1 patients with spinal deformities treated between 2003 and 2012 in our department. Preoperative X-ray, 3D-CT, and MRI were performed to evaluate the deformities of dystrophic scoliosis accurately. All patients were treated with posterior instrumented fusion alone using screws and hooks. According to the anatomical development situation of each patient's pedicles and the transverse processes, we chose different fixations and different fixed segments. The clinical and radiological outcomes of surgical correction were evaluated postoperatively.

The average preoperative kyphosis was 43° (range 15–86°). The postoperative kyphosis had an average of 20° (range 10–39°) yielding 53% correction. At final follow-up, there was an average of 4.6% correction loss. The preoperative scoliosis Cobb angle had an average of 47° (range 35–96°). The postoperative scoliosis Cobb angle had an average of 21° (range 10–37°) yielding 55% correction. At final follow-up, there was an average of 6.6% correction loss. The apical vertebral body rotation was corrected by an average of 48%. At final follow-up, the score of the SRS-30 questionnaire ranged from 97 to 135 with an average of 109.

In conclusion, the deformities of dystrophic scoliosis can be accurately determine through preoperative radiographic evaluation, which plays an important role in guiding the correction of scoliosis program development. The results of this study demonstrate that satisfactory therapeutic effects can be achieved in the dystrophic scoliosis patients by preoperative meticulous surgical plans, intraoperative careful manipulation, and hybrid instrumentation.

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Abbreviations: CT = computerized tomography, MEP = motor-evoked potential, MRI = magnetic resonance imaging, NF-1 = neurofibromatosis type 1, SEP = somatosensory-evoked potential, SRS = Scoliosis Research Society.

INTRODUCTION

Neurofibromatosis type 1 (NF-1) is a multisystemic disease that mainly affects cellular growth of neural tissue.[1] Most patients undergo some type of bony dysplasia. They include spinal deformities, such as scoliosis or kyphosis, pseudarthrosis of the tibia, and soft tissue tumors.[2,3]

Scoliosis is the most common skeletal manifestation of NF-1, with an occurrence ranging between 10% and 64%.[1,4,5] There are 2 main types of scoliosis in persons who have neurofibromatosis: nondystrophic and dystrophic forms.[6] Dystrophic scoliosis is illustrated by progressive, sharply angulated short segment curvature with severe wedging, rotation and scalloping of the apical vertebral bodies.[7]

The choice of treatment for dystrophic scoliosis in NF-1 is still under discussion. Generally speaking, dystrophic curvatures of less than 20° should be observed in case of progression at 6 months intervals.[4] As for adolescent patients with dystrophic curvature greater than 20° to 40° of angulation, a recommendation is to apply a posterior spinal fusion with segmental spinal instrumentation.[4,8] Thus, in case of patients with more severe dystrophic scoliosis, it is often to perform anterior fusion in addition to posterior spinal fusion, to improve the fusion rate and to decrease the risk for deterioration.

Paraplegia is an uncommon finding in patients who have dystrophic curves.[7,9] In neurofibromatosis patients with scoliosis, rib penetration into the spinal canal is uncommon but may be more commonly identified with modern imaging techniques. This protrusion has the potential to cause paraplegia with or without a traumatic episode during spinal surgery or even postoperatively.[10] It is the surgeon's responsibility to stabilize the spine with the most suitable, safe, and permanent method without causing neurologic injury.[2]

The purpose of this study was to review the clinical and radiological outcome of surgical treatment of dystrophic spinal curves in NF-1, for obtaining a good efficient result with safety and without complication.

METHODS

Clinical Data

This retrospective study consisted of 26 NF-1 patients with spinal deformities treated between 2003 and 2012 in our department. The study was approved by the Ethical Committee

of the Fourth Military Medical University and conducted according to the principles expressed in the Declaration of Helsinki. All participants or their guardians signed an informed consent to participate in the research.

All patients presented with 2 or more criteria (14) to diagnose NF-1. The series included 16 males and 10 females with an average age of 9 years (range 6–15 years) and a positive family history in 7 patients (27%). In this series, 4 cases suffered from lower extremity neurological symptoms. Before being referred to us, 10 patients were ineffectively braced for an average period of 4 months (range 3–8 months).

Radiolographic Evaluation

Radiographic studies included preoperative standing Anteroposterior (AP), lateral, and supine side-bending radiographs. The following parameters were measured pre- and postoperatively for correction assessment: the frontal and sagittal curve measurements were made by Cobb technique; apical axial rotation was determined by the method of Perdriolle and Vidal.[11] The intracanal rib head and the development of pedicle and the transverse processes were assessed by 3-dimensional computed tomography (CT) scan. Magnetic resonance imaging (MRI) of the whole spine was performed for each patient to reveal any intraspinal lesions and assess the relationship between the rib head and the dural sac (Figure 1). Typical dystrophic curves had occurred for all patients, and at least 3/5 of the following criteria including vertebral scalloping, pencil-ing of the ribs, severe apical vertebral rotation, spindled transverse processes, and foraminal enlargement. The apex of the deformity was thoracic (n = 14), thoracolumbar (n = 9), and lumbar (n = 3).

Operation Plan

A posterior vertebral column resection was performed according to each patient's spinal deformity. If the intracanal rib head is obviously visible, the spinal cord injury may occur during the process of correction, then we removed the intracanal rib heads completely (Figure 2). According to the anatomical development situation of each patient's pedicles and the transverse processes, we chose different fixations and different fixed segments. If the pedicles were large enough, we preferred to select the pedicle screw fixation to ensure sufficient fixing strength. If the pedicles were too thin, we considered to choose the transverse process hook fixation technique.

Surgical Procedure

Posteriorly, the patient was in the prone position, and a midline incision was made. Extreme care was taken during exposure due to occasional thinning of the laminae. All patients, after well-performed exposure, were treated with posterior instrumented fusion by using screws, hooks, and/or wires; in addition, hybrid instrumentation was also used. For the posterior instrumented fusion, hooks were used for proximal fixation, pedicle screws were used for the distal fixation, and hooks were also placed selectively on the convexity of the curve.

Gradual correction was done with a combination of translation/derotation maneuvers. All patients were closely monitored intraoperatively using both transcranial electric motor-evoked potential (MEP) and somatosensory-evoked potential (SEP). Meticulous decortication was performed and grafted with generous autograft and artificial bones. Nine levels on average (6–14 levels) are included for the fusion, extending the fusion area to vertebrae that were neutral and stable before surgery in both the coronal and sagittal planes. Then, approximately 5 days after surgery, all the patients resumed walking, while were required to wear a thoraciclumbar vest for 3 months on average in purpose of protecting the fusion site.

Clinical Examination

Clinical examination included a thorough neurologic examination and assessment of curve flexibility. In this series, 3 cases suffered from lower extremity neurological symptoms. The Scoliosis Research Society (SRS)-30 questionnaire was

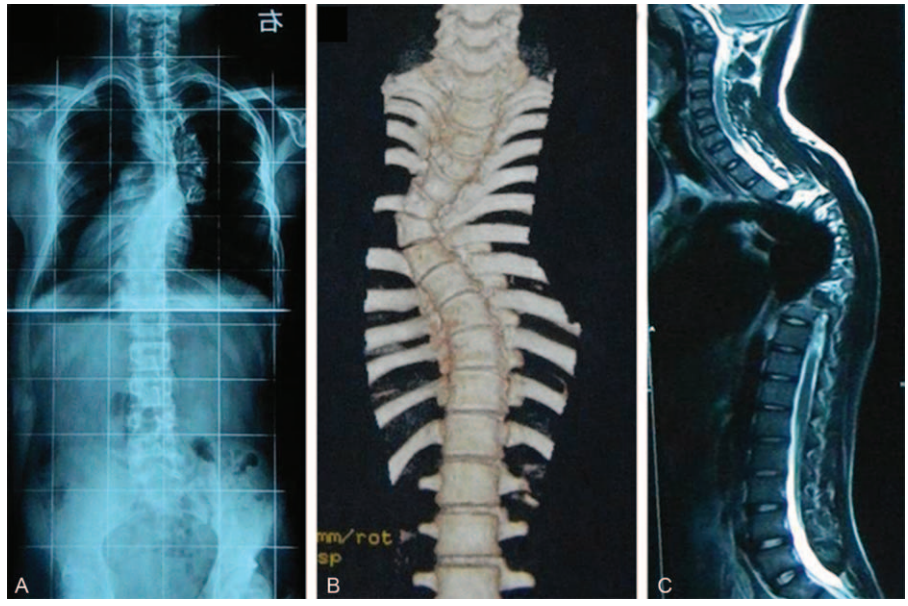

FIGURE 1. Accurately determine the deformities of dystrophic scoliosis preoperatively through X ray (A), 3D-CT (B), and MRI (C).

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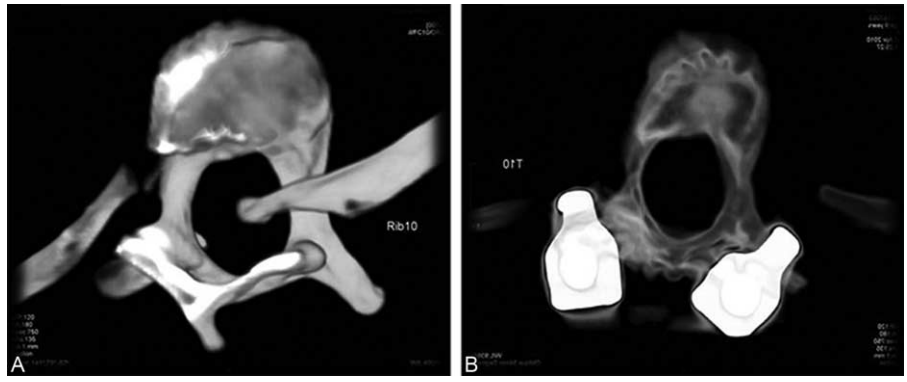

FIGURE 2. (A) Preoperative 3D-CT showed the intracanal portion of dislocated rib head; (B) postoperative 3D-CT showed that the intracanal rib head has removed completely; the spinal canal was very broad and smooth.

used to evaluate the satisfaction rate with surgery. Follow-up was performed 3, 6, 12, and 24 or more months after operation.

RESULTS

The total operative time had an average of 340 min (range 260–490 min). The total blood loss had an average of 475 ml (range 330–740 ml). Radiographic measurements were made on AP and lateral radiographs of the spine by a single independent blinded reviewer. The radiographs analyzed were those obtained preoperatively, immediate postoperatively, and at the final follow-up (Figure 3). The average preoperative kyphosis was 43° (range 15–86°). The postoperative kyphosis had an average of 20° (range 10–39°) yielding 53% correction. At final follow-up, there was an average of 4.6% correction loss. The preoperative scoliosis Cobb angle had an average of 47° (range 35–96°). The postoperative scoliosis Cobb angle had an average of 21° (range 10–37°) yielding 55% correction. At final follow-up, there was an average of 6.6% correction loss. The apical vertebral body rotation was corrected by an average of 48%. Three-dimensional CT results showed that the intracanal rib head was completely removed and the spinal canal was very broad and smooth. At final follow-up, the score of the SRS-30 questionnaire ranged from 97 to 135 with an average of 109. Results are summarized in Table 1.

In 4 cases with incomplete paralysis, 3 cases completely recovered neurologic function after surgery. One patient still

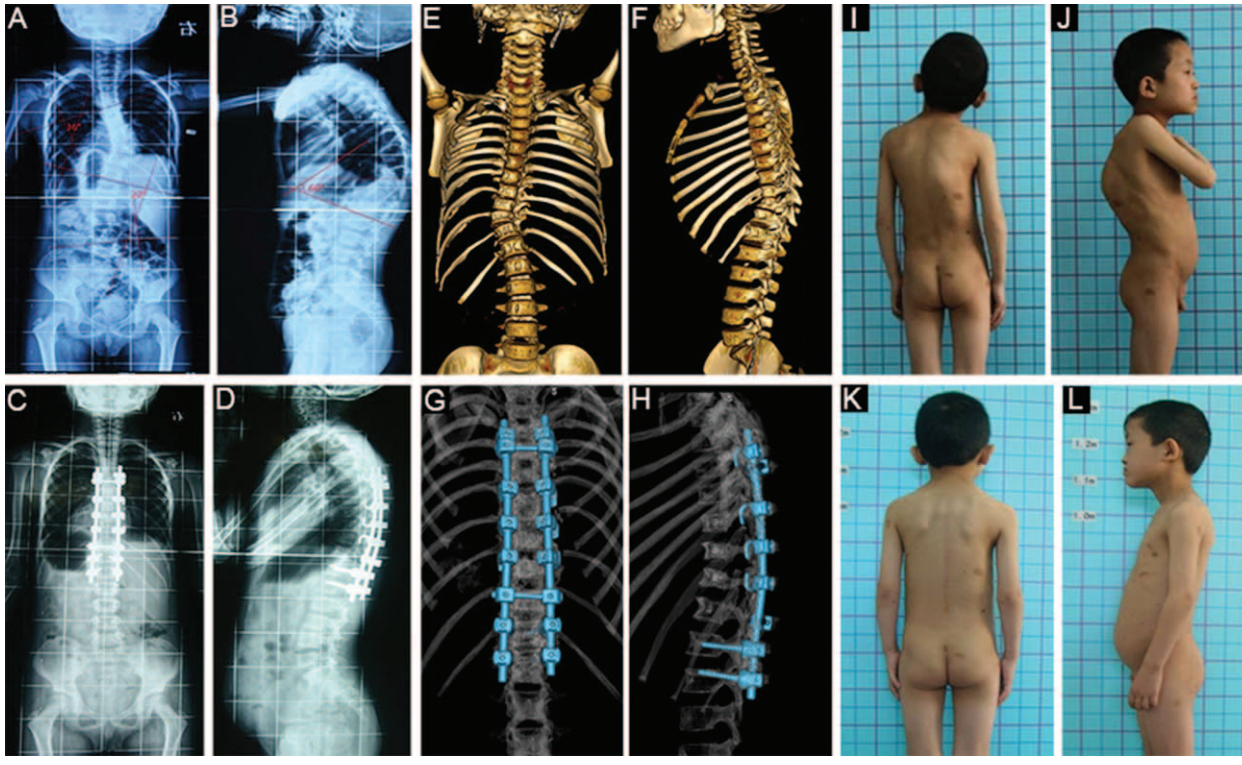

FIGURE 3. A 9-year-old male with a dystrophic curve preoperative and follow-up AP X-rays (A) and 3D-CT (E) revealing a 36° scoliosis corrected down to 11° (C and G). Preoperative and follow-up lateral X-rays (B) and 3D-CT (F) revealing a 60° thoracic hypokyphosis corrected to 18° (D and H). Preoperative (I and J) and 2-year follow-up (K and L) photographs.

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TABLE 1. Summary of Operative Data

Total number of patients	26
Female	10
Male	16
Mean age (range)	9 (6–15)
Operative time (min; range)	340 (260–490)
Blood loss (ml; range)	475 (330–740)
Average preoperative kyphosis (°; range)	43 (15–86)
Average postoperative kyphosis (°; range)	20 (10–39)
% of correction	53%
Average preoperative Cobb angle (°; range)	47 (35–96)
Average postoperative Cobb angle (°; range)	21 (10–37)
% of correction	55%
The score of the SRS-30 questionnaire	109 (97–135)

SRS = Scoliosis Research Society.

suffered from lower extremity hypermyotonia, but significantly improved compared with preoperatively. Within these patients, 1 of them had a dural tear that was successfully repaired, 1 patient suffered superficial infection, had been responded well to antibiotics. Two patients, after 10 months follow-up, had hook dislodgement and pseudarthrosis presenting. All the procedures including exploration, replacing the hook and regrafting had been performed, together with the rest of the follow-up were all smooth.

DISCUSSION

A short-segment and sharply angulated curve normally used for describe dystrophic scoliosis, and the vertebral bodies is associated wedging and scalloping. And dystrophic scoliosis could be accompanied by the phenomena including vertebral body rotation, widening of the intervertebral foramina and penciling of rib head; and it could tend to develop a severe deformity.[1]

In young patients with progressive dystrophic deformities, it is believed that only applying posterior spinal fusion is contraindicated. In accordance with the literature report, after applying posterior spinal fusion alone, pseudarthrosis is occurred to be up to 60%.[12] In spite that some surgeons may perform posterior fusion with abundant autologous bone graft and pedicle screw instrumentation, which is of perfect long-term results, anterior and posterior spinal fusion with segmental instrumentation and bone grafting are the more predictable and successful procedure.[13–15] What's more, based on the acknowledged natural history of certain progression for this type, the young child with dystrophic scoliosis greater than 40° is applicable to be fused.[4,8] And because of poor growth potential of the involved segments, minimal stunting of growth is caused by an early fusion.[2]

There is a prominent question raised in severe scoliosis, that is, will posterior surgery alone is adequate or both anterior and posterior surgeries are a necessity. As the anterior/posterior approach is concerned, the approach combines anterior distraction with posterior compression of the vertebrae; as a result, it causing a more comprehensive correction of the deformity possible. The more extensive operation is necessary, but the operation involves complications with higher risk, including reduced respiratory function and excessive bleeding. In the contrast, the posterior approach only is of fewer invasions, while it has been associated with high rates of instrumentation failure, pseudarthrosis, and postoperative progression of scoliosis.[16,17] As it has been reported by Li et al[17] that 19 patients with NF-1 treated surgically with posterior instrumented spinal fusion alone. In accordance with dystrophic thoracic curves, the Cobb angles were 68°, 27°, and 33° respectively at preoperation, postoperation, and at the final follow-up in the coronal planes; were 31°, 28°, and 30° respectively at preoperation, postoperation, and at the final follow-up in the sagittal planes. There were 8 (42.1%) complications, 3 intraoperative, and 5 postoperative. And 1 of the patients (5.2%) of pseudarthrosis with instrumentation failure requires revision surgery. In our study, the mean corrections achieved in these patients are comparable to those reported by authors using anterior–posterior fusions.[18,19] There were no cases of decompensation among the patients in this series, and the loss of correction was limited at follow-up.

It is difficult to treat dystrophic scoliosis in NF-1 as it has been resulted by a disease process which gets to worsen throughout life. A 38% incidence of pseudarthrosis in 9 out of 23 dystrophic patients after posterior fusion treatment alone were reported by Sirois and Drennan[20]; and it has been shown that 1.7 procedures on average were required to achieve solid fusion in these patients. After making the analysis of the curvatures for all these patients, short and highly dystrophic curves were found with their presence. According to the report by Sirois and Drennan[20] and Winter et al,[21] when dystrophic kyphotic curves of 50° or more, a 64% and a 72% of failure were occurred respectively after the treatment of posterior fusion alone were applied. And in accordance with the report from Hsu et al,[22] regardless of kyphosis, the treatment by combined anterior and posterior fusion for dystrophic curves occur a 7.5% of failure. The authors assumed that the main reason for failure is the inadequate anterior procedure. In a study carried by Parisini et al[23] in 56 patients with spinal deformities, neurofibromatosis evaluating had been applied, and fusion failure was observed in 47% and 33% of scoliotic patients, who underwent posterior instrumented fusion only compared with anterior and posterior fusion. Poor and osteoporotic bone stock makes it difficult to anchor the instrumentation securely.[24] Dural extasia, a pathological widening of the dural sac, can cause vertebral ''scalloping'' and weaken the fixation of posterior instrumentation, making loosening of the hooks and loss of correction common.[25]

In this study, 2 patients had hook dislodgment and pseudarthrosis presenting after 10 months follow-up. Compared with the previous report for patients applied with only posterior instrumented fusion, less smooth exploration, replacing the hook and regrafting, and the rest of the follow-up were performed, which is better compared with some studies on patients performed with combined anterior and posterior fusion.[25] Based on our results, using solid instrumentation and extension of the fusion are suggested to many levels may make successful posterior fusions in these patients.

The diagnosis of NF-1 dystrophic scoliosis can be made based on typical clinical features, such as multiple cafe au lait spots on the skin and a characteristic curve pattern on radiology.[26] The neurological symptoms of NF-1 with intraspinal rib head dislocation are varied, ranging from mild sensory and motor deficits to paraplegia and paraparesis, with 60% of documented cases being asymptomatic.[27] In our study, rib head dislocations were detected on the axial CT slice and 3D reconstruction. All of the dislocated ribs were on the convex side of the curve and were from the levels at the periapical

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regions. Furthermore, MRI scanning demonstrated the relationship between the cord and intracanal rib heads, which was also important to clarify the intracanal deformity and assist with surgical decision making. In this study, there were 6 patients presenting obvious intracanal rib heads. The intracanal rib heads were removed to minimize the risk of spinal cord injury during the process of correction. Three dimensional CT results showed that the intracanal rib head was removed completely, the spinal canal was very broad and smooth. Postoperatively, 1 patient had a transient mild paraparesis which completely recovered at 6 months follow-up.

It is universally agreed that dystrophic curves cannot be corrected by brace treatment.[28] In this study, 10 patients had been ineffectively braced for averagely 4 months (rang 3–8 months) before being referred to us. Usually it is suitable to brace following surgery.[21] In case of severe dysplastic curves that are instrumented into the upper thoracic and cervicothoracic region, bracing may need to be extended to the cervical region. Bracing may also helpful to prevent screw/hook to pull out, which is particularly true for dysplastic curves that have low bone mineral density.[24]

Our study had some major limitations. Firstly, the medical records were reviewed retrospectively; we did not see patients at final follow-up specifically for this study. Secondly, because the follow-up ranged from 2 to 10 years, the long-term clinical outcome of procedures is not known in terms of correction loss and other possible problems.

CONCLUSIONS

In conclusion, the deformities of dystrophic scoliosis can be accurately determine through preoperative radiolographic evaluation, which plays an important role in guiding the correction of scoliosis program development. The results of this study demonstrate that satisfactory therapeutic effects can be achieved in the dystrophic scoliosis patients by preoperative meticulous surgical plans, intraoperative careful manipulation, and hybrid instrumentation.

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