

Original Article

## Evaluation surgical treatment results of scoliosis in patients with neurofibromatosis

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### Abstract

**Introduction:** Surgical treatment of spinal deformity resulting from neurofibromatosis (NF) is a major challenge for orthopedic spine surgeons. There are several problems and complications including pseudoarthrosis and cure progress despite treatment. Progressive kyphoscoliosis is the most important spinal deformity. The present study aims to evaluate surgical treatment results in severe spinal involvement cases.

**Methods:** This analytical a-descriptive study evaluated 20 patients with NF, severe scoliosis and kyphosis (up to 50°) hospitalized and treated at our center during the past 10 years. The treatment failure rate and complications were studied.

**Results:** In this study, 20 cases with NF and kyphoscoliosis with the mean age of  $13.00 \pm 7.18$  years were studied. These case series were consisted of 13 (65%) males and 7 (35%) females. Overall treatment failure was 45%. However, it was 55% of failure happened in posterior fusion alone. Failure rate was reported 36% in the combined anterior and posterior fusion and mainly seen in younger than 8-years children. Surgical complications were found in 20% with pseudoarthrosis as the most common one. There were no infections and neurological complications. Statistically, there was a significant negative relation between age and curve progression in scoliosis and kyphosis during the 2 years follow-up period. There was not any significant difference between genders considering curvature progress.

**Conclusion:** The combined anterior and posterior fusion is probably more effective treatment, especially at early ages when more aggressive treatment is required since it reduces the treatment failure possibility.

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### Introduction

Neurofibromatosis (NF) is a genetic congenital disease inherited as dominant autosomal and does not differ in two genders. Incidence rate of the disease is 1:3000 birth and genetic mutations are often seen in 50% of the patients.<sup>1,2</sup>

NF is associated with classic clinical symptoms including stained skin damages (milk coffee stains), IQ and mental changes, and different skeletal deformities. According to recent studies, there is a meaningful relation between spinal involvement and neurological problems, which is often

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progressive.<sup>3,4</sup> There are two types of NF. NF type I (NF-1) is a multi-systemic disease and manifested with disorders in neurological and skeletal systems, soft tissue, and skin.<sup>1</sup> There is a wide range of disorders in NF with skeletal ones as the most prevalent manifestation of NF-1 which is seen as bone dysplasia in most patients.<sup>1,2</sup> According to the studies, skeletal and orthopedic disorders are usually manifested earlier. The orthopedic disorders include scoliosis, kyphosis, and tibia congenital dysplasia in the form of pseudoarthrosis and bowing of the tibia, forearm and other bones.<sup>2,3</sup> The most common of the osseous complications of NF-1 is spinal deformity, occurring in 10-30% of individuals with NF-1. Many of these patients will eventually require surgery for curve progression.<sup>4</sup>

More growth of upper limbs, as well as soft tissue tumors, is reported in this disease. Considering the high prevalence of spinal involvement and progressiveness of the skeletal disorders and their effect on patients' life, the present study aims at evaluating treatment results in patients suffering from NF.

## Methods

In a descriptive-analytical study, the study sample was consisted of 20 patients suffering from NF with spinal scoliosis surgically treated at Shohada Hospital of Tabriz, Iran, within the last 10 years (2003-2013). The inclusion criteria were patients' satisfaction to participate in the study, lack of any records of spinal tumor and fracture, suffering from NF with clinical criteria of this disease, severe spinal involvement in the form of kyphosis (up to 50°) and severe scoliosis, and patients treated with spinal fusion surgery method. Patients were excluded for other deformities in limbs exception of spinal deformity.

Figure 1 demonstrates clinical scheme of a patient with coffee milk stains and severe spinal deformity. Clinical examinations and new radiographs in standing the position were used to evaluate the corrected angels, and previous radiographs were used to determine spinal deformity variations. The

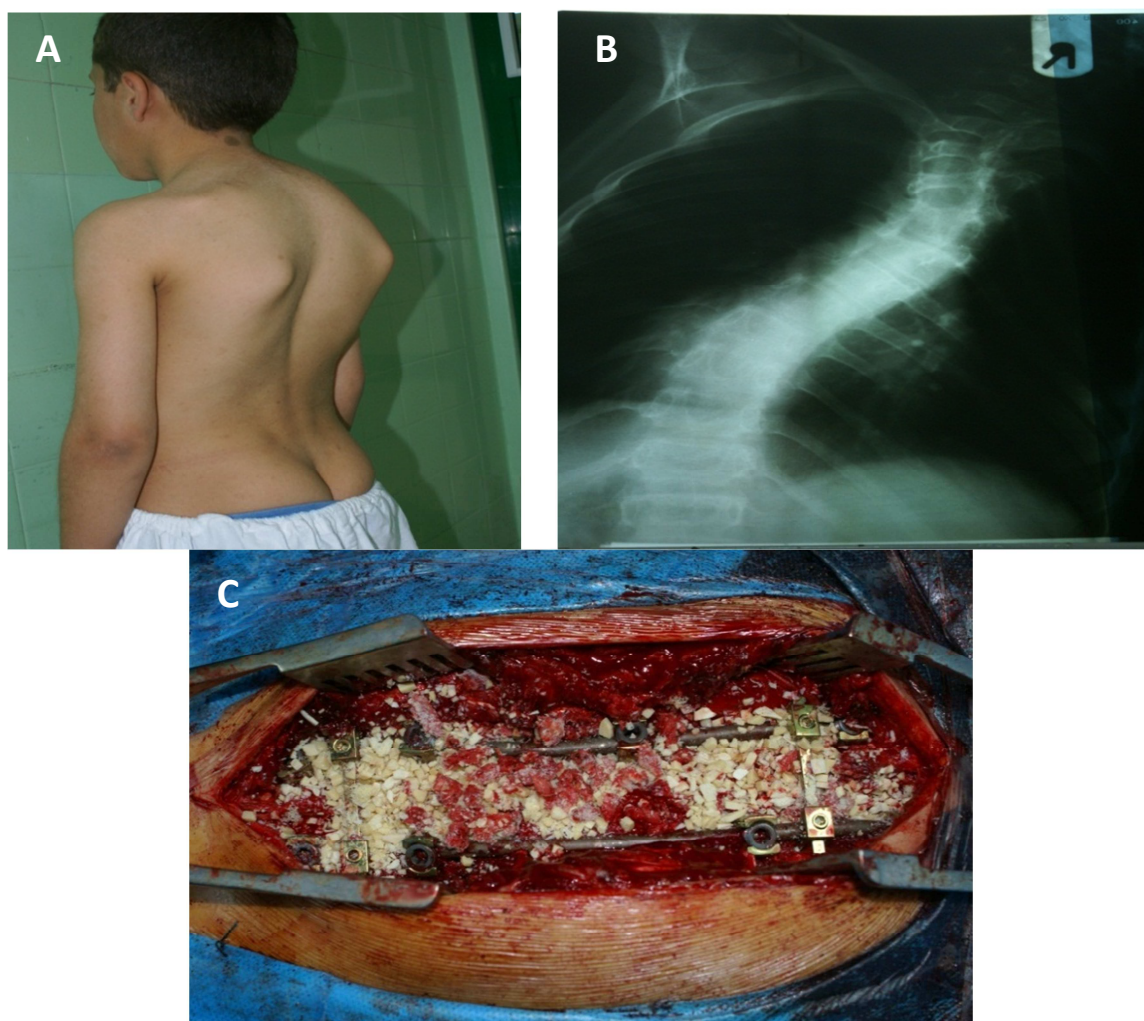
Scoliosis Research Society/MODEM's questionnaire was used for the final outcome after 2 years follow-up. The study was approved by Research Committee of Tabriz University of Medical Sciences.

Quantitative data is presented as mean and standard deviation and qualitative one as frequency and percentage. t-test and independent samples are used to compare quantitative data and chi-square test, or Fischer's exact test is used to compare the qualitative ones. Correlation between deformity degree and age was performed by Pearson statistical test. In all cases,  $P < 0.05$  was considered to be significant. The measurements were analyzed with SPSS for Windows (version 17, SPSS Inc., Chicago, IL, USA).

## Results

In this study, 20 patients with kyphoscoliosis resulting from NF with the mean age of  $13.30 \pm 7.18$  years were studied. The case series was consisted of 13 (65%) males and 7 (35%) females. The patients underwent posterior fusion in 9 cases (45%) and combined posterior and anterior fusion in 11 cases (55%). The patients were averagely followed-up for  $4.5 \pm 2.1$  years. Table 1 refers to pre-operation measurements of scoliosis and kyphosis and post-operation re-measurements and correction rate during the last follow-up. According to this table, a significant correction is seen in scoliosis and kyphosis rate of the patients ( $P < 0.05$ ). Clinical photos of a younger than 8-year-old patient are demonstrated in figure 2, that progressive curvature is seen in spite of treatment.

According to the last follow-up, treatment failure was 45% in the patients. It was 55% in the patients underwent posterior fusion alone. All patients were younger than 8 years, and more surgical interventions were required to treat these patients. Failure rate was reported 36% in the combined anterior and posterior fusion. In other words, deformity progressed in spite of surgical treatment. Three patients were younger than 3 years, and one patient was older than 8 years. Evaluating the relation between age and curve size, the study



**Figure 1. A 12-year-old boy with severe kyphosis and scoliosis treated with spinal fusion**  
 A: Clinical photo with milk coffee stains in neck and trunk, B: Preoperative radiography, C: Posterior spinal fusion

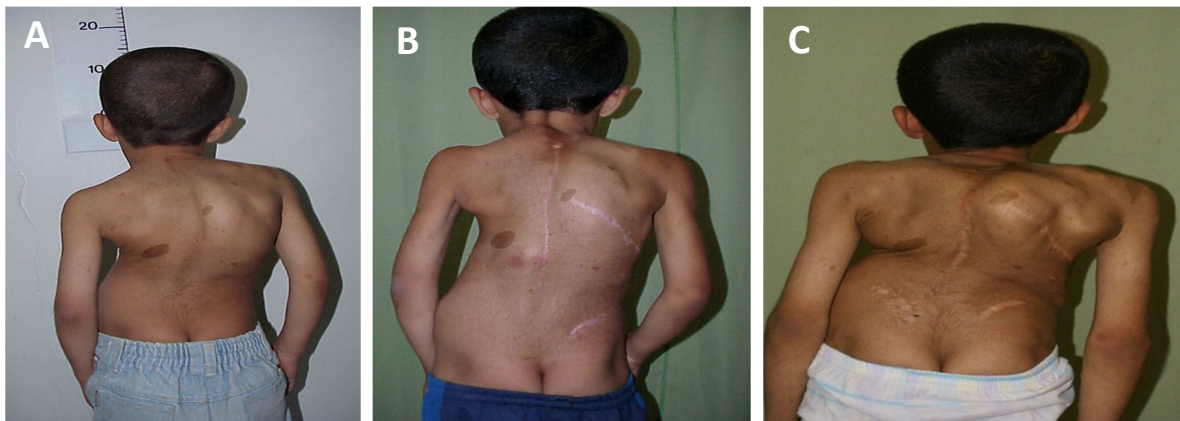
**Table 1. Scoliosis and kyphosis rate in patients with neurofibromatosis before, after and during follow-up period**

Variable	Mean ± Standard deviation
Pre-operation scoliosis (°)	82.7 ± 20.5
Post-operation scoliosis (°)	54.1 ± 13.6
Scoliosis during the last follow-up period	65.6 ± 21.3
Pre-operation kyphosis	61.8 ± 11.9
Post-operation kyphosis	42.5 ± 13.7
Kyphosis during the last follow-up period	51.4 ± 7.7

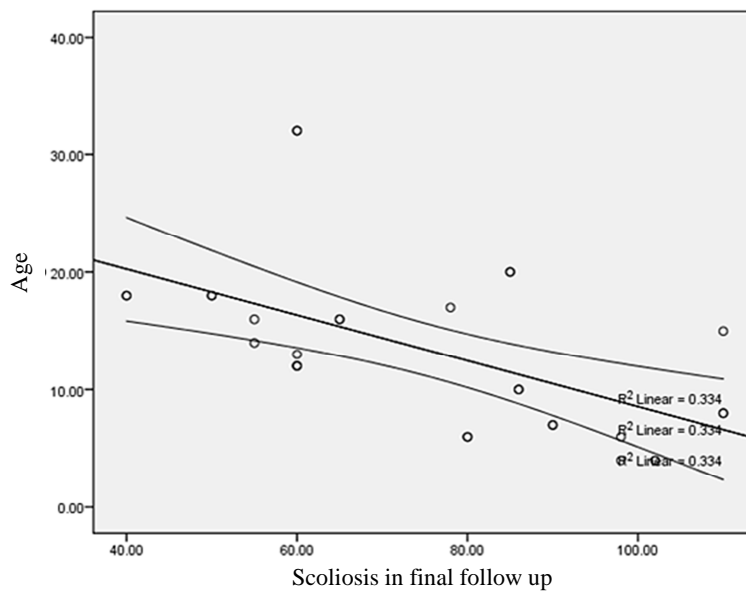
indicates negative linear relation between age and progressive rate of scoliosis and kyphosis in the NF patients. There was a significantly negative relation between age and scoliosis size ( $R = -0.5, P = 0.001$ ). Statistically, a meaningful negative relation was found for kyphosis, too

( $R = -0.6, P = 0.001$ ) (Figures 3 and 4). The complications were observed in 20% (four patients) with pseudoarthrosis as the most important one. The average blood loss was  $1132 \pm 76$  ml, and the average surgery time was  $248 \pm 63$  min. Fifteen of the patients required an intra-operative transfusion, which averaged  $787 \pm 87$  ml. There was no neurological impairment, intra-canal hematoma, dural tear or other form of complication after surgery.

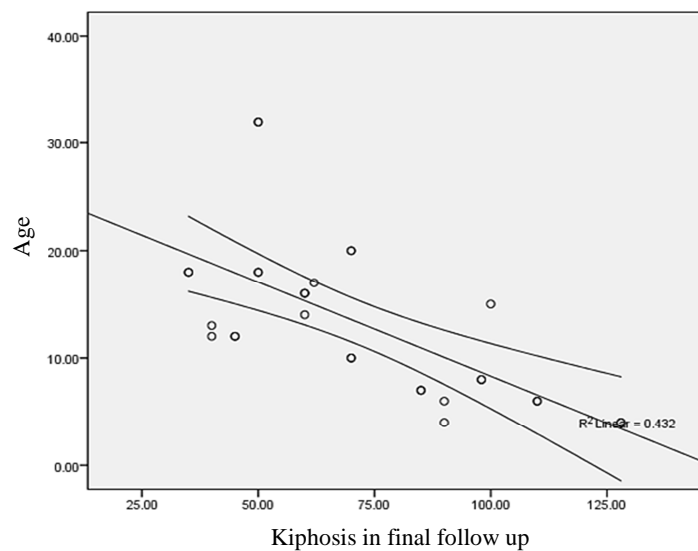
Clinical and radiographic scheme of a younger than 8-year-old patient are demonstrated in figure 1A-C where progressive curvature is seen in spite of treatment. Furthermore, figure 2A-C refer to an older patient experienced relatively fixed condition in spite of being treated using spinal fusion.



**Figure 2. A: A 6-year-old boy before spinal fusion (combined anterior and posterior fusion), B: Six months after surgery and C: One year after surgery with sever progressive kyphosis and scoliosis**



**Figure 3. Statistically negative relation between early age and curvature progress of scoliosis in neurofibromatosis patients**



**Figure 4. Statistically negative relation between early age and curvature progress of kyphosis in neurofibromatosis patients**

## Discussion

NF is a genetic disease with neuroectodermal involvement and mesenchymal dysplasia<sup>1</sup> and its clinical symptoms were initially explained by Von Recklinghausen in 1882.<sup>1,2</sup> Progressive kyphoscoliosis is an important spinal deformity pattern. NF is classified in two types.<sup>2</sup> In type I, there is more peripheral involvements and is associated with skeletal and orthopedic disorders. Central involvement is seen in type II and there are less orthopedic involvements.<sup>1</sup> In addition to spinal involvement directly resulted from vertebra disorder, indirect involvement occurs as progressive functional scoliosis due to the disorder in other limbs including lower limb disorder resulting from hypertrophy or dysplasia.<sup>5</sup>

Surgical treatment of spinal deformity due to NF is a major challenge for orthopedic surgeons. There are several problems and complications such as pseudoarthrosis and progressive curvature despite of treatment.<sup>6,7</sup> Spinal skeletal disorders are the most prevalent clinical symptoms of NF. Posterior correction and fusion were performed with segmental spinal instrumentation consisting of lateral mass screws, lamina screws, pedicle screws, and polyethylene tape for sub-laminar wiring. Anterior spinal fusion was performed using a fibula strut to induce solid bone fusion. All patients used a halo vest for postoperative external fixation.<sup>8</sup>

According to a new study, showed that early and aggressive surgery is the most effective management for dystrophic curves in NF.<sup>9</sup> In a study conducted by Hunt and Pugh, its prevalence was estimated 36%.<sup>10</sup> Dystrophic changes rate in sagittal plan plays the most important role in determining surgical approach in treating these patients.<sup>11</sup>

Patients with 20-40° scoliotic deformity and less than 50° kyphosis are treated using only one approach, for example, spinal post-instrument and fusion. Surgical treatment and correction are very difficult in cases with severe involvement, and higher degrees of kyphosis and scoliosis such that clinical outcome of the patients cannot be anticipated.<sup>11,12</sup> According to Sirois and

Drennan, surgical treatment was associated with some complications such as pseudoarthrosis in 38% of patients with severe deformity resulting from NF.<sup>13</sup> Probability of treatment failure and curvature progress is another problem of severe dystrophic spinal deformities such that curvature progress rate and overall failure was reported as 12.7° and 70%, respectively.<sup>13</sup>

In our study, the complication was seen in 20% of NF with severe spinal involvement who underwent surgical treatment. Similar to Sirois and Drennan, pseudoarthrosis was introduced as the most important complication.<sup>13</sup> The overall failure rate is 45% which is less than the previous study. As mentioned, treatment of spinal involvement in NF is a great challenge. According to Winter et al., failure in severe deformity cases with kyphosis (up to 50°) was seen in 64% of posterior fusion.<sup>12</sup>

Hsu et al. reported a 7.5% failure rate in patients underwent combined posterior and anterior surgical treatment and kyphoscoliosis progress was seen during follow-up period of these patients.<sup>14</sup> Parisini et al. studied 56 patients and reported 53% of overall failure rate in cases underwent posterior fusion.<sup>15</sup> Comparing with combined treatment, it was significantly different such that treatment failure was seen in 23% of these patients. According to our study, the overall failure occurred in 45% of scoliokyphosis resulting from NF in spite of surgical treatment. The findings are similar to that of the previous ones, including Parisini et al.'s.<sup>15</sup>

In posterior fusion cases, it was about 50%, and curvature progress was seen in spite of treatment and stabilization. In combined anterior and posterior fusion, treatment failure was reported as 36% that is correspondence with findings of the previous studies. As mentioned, severe spinal deformity in the NF patients is a major treatment problem for the surgeons. In our study, we noticed role of age in treatment results such that the highest rate of treatment failure may be attributed to lower ages. In most cases, the patients were younger than 8 years that was ignored in the previously

conducted studies. Contrary to findings of our study, only in the study conducted by Calvert et al.<sup>1</sup> on 45 patients, there was not found any meaningful relation between age and progressive curvature. In this study, of course, some of the patients did not undergo surgical treatment and progress was evaluated only considering age. In studies conducted by Rezaian<sup>16</sup> in Iran, severe spinal involvement was reported only in 15% of the patients with kyphosis (more than 50°).

Posterior fusion treatment method and a 9-month follow-up period were efficient ways where pseudoarthrosis was introduced as the most important complication, similar to our study. In this study, infection and paraplegia were seen in one case, but there was not any case of infection and neurological involvement in our study. In more recent studies conducted by Tsirikos et al.,<sup>5</sup> there was not any complication including infection and paraplegia after fusion surgery of NF patients and pseudoarthrosis was reported as the most important complication. In Sun et al. studied

were no surgery-related complications any time after the posterior instrumentation and spinal fusion surgery in patients with NF-1 scoliosis.<sup>17</sup> New technique according to Stoker et al. showed posterior-only vertebral column resection represents a safe and efficacious, but technically challenging option for the treatment of angular kyphotic spinal deformity and associated neurological deficit in patients with NF-1.<sup>18</sup>

Spinal deformity treatment in NF is a great challenge. Combined posterior and anterior fusion is more effective treatment, especially at early ages. In addition, more aggressive treatment is required at early ages since it reduces the probability of treatment failure.

#### Conflict of Interests

Authors have no conflict of interest.

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