Neurofibromatosis and spinal deformities: The severity in pediatric age and treatment

**Background:** Scoliosis in neurofibromatosis type I (NF-I) ranges from 10% to 60%; can be divided into three groups 1) dystrophic scoliosis, 2) non-dystrophic scoliosis and 3) early onset scoliosis. Surgical treatment is very demanding in early onset, severely progressive spinal column deformities.

**Materials & Methods:** Twenty-three patients, aged between 4 and 11 years, were surgically treated at the Authors’ Spine Surgery Division in the past 15 years. Mean follow-up is 5 years (range, 18 months to 15 years). Mean age at the time of surgical procedure was 9.1 years (range, 4 years to 11 years). Average scoliosis was 48° (range, 38° to 82°) and skeletal maturity according to Risser sign was 0 in all of the patients. Patients were divided into 2 groups according to the surgical procedure adopted. Posterior only instrumentation was performed in 16 patients those presented with a thoracic kyphosis lower than 50° (Group A), in the remaining 7 patients showing thoracic kyphosis exceeding 50°, combined anterior and posterior instrumented arthrodesis was performed (Group B). One patient, belonging to Group A, was instrumented with growing rod without fusion.

**Results:** Average correction of scoliosis was 60%, overall complication rate 24% and major 7%. Crankshaft phenomenon was observed in 21% (Group A). In these cases, anterior arthrodesis was performed after a mean 15-month from first surgical procedure. Fusion failure was observed in 1 (Group B) patient who underwent revision of posterior instrumentation. Clinical and radiographic evaluation at F-up showed good outcome in terms of deformity progression and quality of life.

**Conclusion:** Early and aggressive surgery is the most effective management for dystrophic curves in neurofibromatosis which was proven. Our experience confirms the need for spinal stabilization even in pediatric age in rapidly progressive spinal deformities. The growing rod technique should be carefully evaluated in each single case.

**Biography**

Stefano Giacomini is a Medical Director and performs orthopedic examinations in Free Job regime at the Outpatient Clinic of Rizzoli. He has done his specialization in Orthopedics and Traumatology in 2001 at the University of Bologna. He is the author of over 50 scientific publications. He has participated, even as a speaker at dozens of national and international conferences and *socio GIS* (Italian Scoliosis Group).

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