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Surgical treatment of severe progressive scoliosis in neurofibromatosis in pediatric age

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Neurofibromatosis type I is one of the most common genetic disorders of the nervous system. Dystrophic alterations of the spine have been reported in approximately 49% of patients and may develop since early pediatric age. The prevalence of scoliosis ranges from 10% to 60% in the literature. Surgical treatment is a demanding procedure in early onset, severely progressive spinal column deformity. Retrospective evaluation of surgical outcomes in NF-I patients was with severe progressive scoliosis in pediatric age. Twenty three consecutive patients, with an age between 4 and 11 years, have been surgically treated in our Spine Surgery Division in the past 15 years. Mean follow up is 4 years. Mean age at surgical procedure was 9.1 years. Average value of scoliosis before surgical treatment was 48° (min. 38°, max. 82°) Posterior only instrumentation was performed in 14 patients with a thoracic kyphosis inferior to 50° (Group A), while in the remaining 9 patients, with a thoracic kyphosis superior to 50°, combined anterior and posterior instrumentated arthrodesis was performed (Group B). Overall complication rate was 24%. Major complication rate was 7%. Crankshaft phenomenon was observed in 3 Group-A patients (21%); in these cases anterior arthrodesis was performed after a mean 15 months period from first surgical procedure. Clinical and radiographic evaluation at follow up showed good outcome in terms of deformity progression and quality of life. Our experience confirms the need for spinal stabilization even in pediatric age in rapidly progressive spinal deformities. Surgical treatment of spinal neurofibromatosis is a demanding procedure with uncertain outcome and revision surgery may be necessary.

Biography

Greggi T is a surgeon and specialist in Orthopedics and Biomedical Technologies. Since 1991, he has been focusing exclusively on spinal surgery. He has published over 200 scientific papers in reputed journals and has been serving as an Editorial Board Member of repute. Since 2000, he had positions of responsibility at the Department of Surgery of Deformities of the Spine and from 2009 till date, he is the Director of Department of Surgery of Deformities of the Spine.

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