Surgical treatment of scoliosis in rare diseases: Arthrogryposis

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Background: The reported incidence of scoliosis in arthrogryposis varies from 30% to 67% and, in most cases, the curves progress rapidly and become stiff from early age. The authors report six cases of scoliosis in arthrogryposis to assess the role of surgical treatment.

Methods: Six cases (3 males, 3 females; mean age at surgery 13.2 years) with arthrogryposis multiplex congenital associated with the characteristic amyoplasia were reviewed: they were operated on for scoliosis at the authors' Spine Surgery Department between 1987 and 2008. Surgery was performed using the Harrington-Luque instrumentation (2 cases), the Luque system (1), a hybrid segmental technique with hooks and screws (1) and spinal anchoring with pedicle screws (2).

Results: The patients were clinically and radiologically reviewed at a mean follow-up of 4.2 years, ± 2.7 (range, 1 to 9 years). Three minor postoperative complications were encountered; a long-term pulmonary complication was seen in one case after reintervention and was successfully resolved after 10 days. Surgery was successful in the other 5 cases, where solid arthrodesis was achieved and no significant curve progression was observed at followup.

Conclusions: The experience acquired with the present case series leads the authors to assert that prompt action should be taken when treating such aggressive forms of scoliosis. In case of mild spinal deformities in arthrogryposis, brace treatment should be attempted, the evolution of the curves being unpredictable; however, when the curve exceeds 40° and presents with marked hyperkyphosis, hyperlordosis or pelvic obliquity, surgery should not be delayed.

Biography
Gianluca Colella is specialized works in Orthopaedics and Traumatology at University Federico II – Napoli, Fellow at Spine Surgery Division - Istituto Ortopedico Rizzoli. He has done his Master Degree in Medicine and Surgery at University Federico II Naples, with a discussion “Trattamento chirurgico e tecniche ricostruttive dei tumori del femore prossimale: limb salvage surgery”, and also Abilitation to Medical and Surgical Profession. . He is also responsible for a specialized clinic dedicated to the study and treatment of severe spinal deformities associated with rare diseases at the Rizzoli Orthopedic Institute.

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