Gianluca Colella, J Genet Syndr Gene Ther 2018, Volume 9
DOI: 10.4172/2157-7412-C1-017

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Surgical treatment of early onset scoliosis associated with rare disease

Background: Kyphoscoliosis is the most common and rapidly evolving spinal deformity in rare syndromes. Early Onset and Congenital deformities, often associated with a lot of malformation, are difficult to manage. The aim of the study is to describe an approach to spinal deformities in rare syndromes and assess the effectiveness of growing systems.

Methods: We report 11 cases of pediatric patients with scoliosis (7 males, 4 females) : 2 NF1, 2 arthrogryposis, 1 Spondylo-rib dysplasia, 1 trisomy 8, 1 syringomyelia, 1 Arnold Chiari type I, 1 Prader Willi, 1 Kabuki syndrome, 1 Escobar. This is a retrospective study from 2006 to 2011. The inclusion criteria were 1) EOS or congenital scoliosis in rare disease, 2) Growing rod system, 3) follow-up 24 months (12-36) . Surgery was performed using VEPTR in 4 patients (Mean age 1st visit: 3 yy (1-7) at surgery: 5 yy (3-9), GROWING ROD in 7 patient (Mean age 1st visit: 6 yy (3-10) at surgery: 9 yy (5-12).

Results: The patients were clinically and radiologically reviewed at a mean follow-up of 11 months. We made 11 first surgery and 15 lengthening procedures. The thoracic curve correction was of 50% (from 65° to 39°). Children are grown on average of 2,5 cm/ each follow up year, results so similar to the phisiological growth. We had 8 postoperative mechanical complications on 26 surgeries: 4 screws loosening, 3 broken rods, 1 rib-hook loosening; 6 cases were treated during the planned lenghthening and 2 with revision surgery not originally planned.

Conclusions: Our strategy is for each patient working in team, with other colleagues more specialist and a particular route starts from a dedicate ambulatory (Rare disease ambulatory). The first step is to be aware of the specific diagnosis of a suspected syndrome; without a confirmed genetic diagnosis of a suspected syndrome, it is impossible to plan preoperative strategy for major spine surgery to minimize the risk of increased morbidity and mortality due to the syndrome.The preoperative programs need a particular flow chart with all exams to study the case, such as X-Rays, MRI, Cardio and abdominal US, CT scan, anaesthesiologist, neurological, pneumological visits. The growing systems are good tools and effective in the treatment of early-onset scoliosis in rare syndromes. Early surgical treatment is needful for these deformities that don't respond at the conservative treatment. The correction rate depends on the age at first surgery and earlier we start better will be the results.

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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