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Severe scoliosis treatment in patient with Type III Osteogenesis Imperfecta Case Report

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Osteogenesis imperfecta (OI) is a genetic disorder of increased bone fragility, low bone mass and connective-tissue manifestations. Overall prevalence is estimated between 1/10,000-20,000. Children affected with type III tend to become wheelchair-bound and non-ambulatory. Axial skeletal is involved, with progressive platyspondyly and kyphoscoliosis. The treatment of these patients remains a challenge. There are several tips and tricks to attempt to decrease the risk of complications. The authors report the treatment of a severe scoliosis in type III OI, to illustrate a possible strategy and pitfalls.

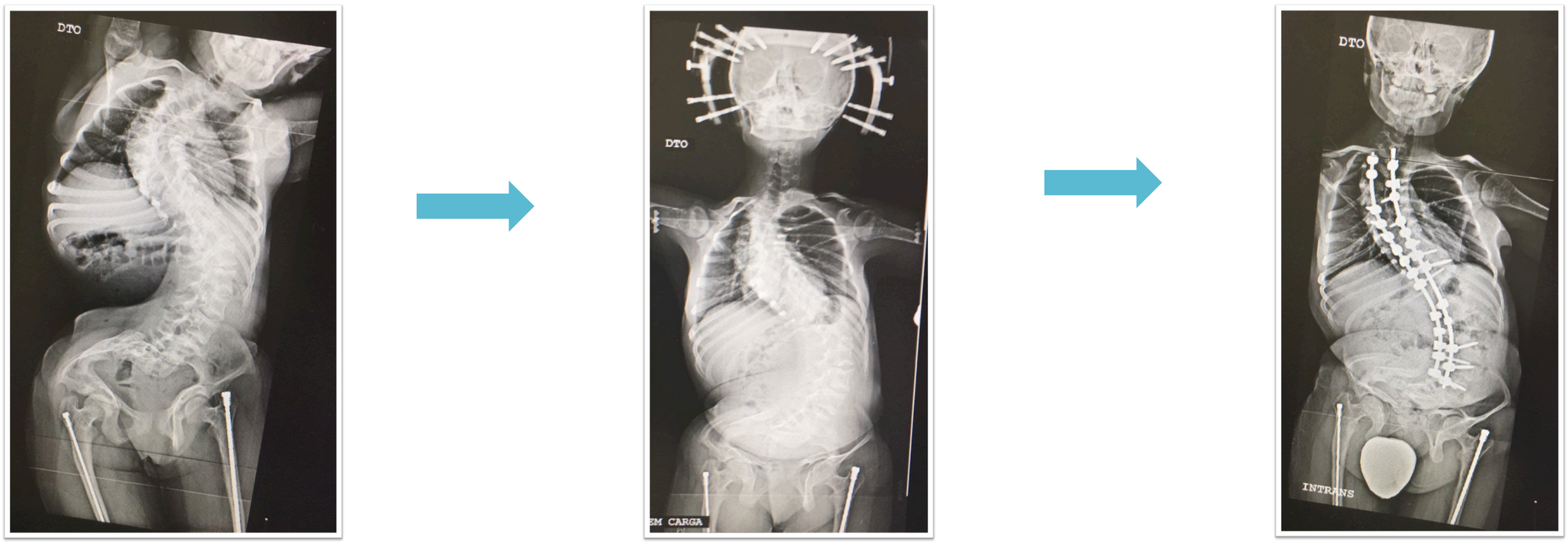
Female, 15 yo - Type III OI, non-ambulatory

Severe major curve scoliosis (thoracic curve 110º; lumbar curve 113º), complicated with pulmonary insufficiency.

Previous treatment with pamidronate

Treatment strategy: pre-operative halo-gravity traction with following posterior correction and arthrodesis.
The patient tolerated gradual traction increases for 30 days, to a maximum of 37% of her body weight. Improvement of the curve was evident (30% thoracic curve and 43% lumbar curve); no traction related complications reported. A T2 to L4 posterior arthrodesis was then performed using pedicle screws and hooks.

Pre-operative | **After halo-gravity traction** | **Post-operative**



A 40,9% reduction of the thoracic curve and 59% of the lumbar curve was achieved, as well as an improvement of the patient's spinal height and balance. The patient needed new surgery to repositioning one pedicle screw responsible for bronchial perforation. Transient neurological deficits included fecal incontinence and palsy of left lower limb.
3 months after surgery the patient has completely recovered her previous neurological status. 2 years down the line the patient maintains the correction achieved, with and improved trunk control and no respiratory complaints.

Spinal surgery in OI patients is challenging due to bone fragility and dysplasia. Pre-operative treatment with pamidronate can be helpful to modulate the disease and improve bone quality. Halo-gravity traction allows a gradual curve reduction with continuously neurological monitoring and low rate of complications. It can also decrease the risk of implant failure, even with poor quality bone. Intra-operatively, neurological monitoring is imperative, however neurological complications can still go unnoticed. Navigation decrease screw malpositioning in very dysplastic spines, however this technology is not widely available. A multidisciplinary team and an intense rehabilitation program are crucial for these patients recovery. The purpose of surgical treatment can be just to improve mobility, respiratory function and sitting ability. Difficult cases can be solved in spite of clinical risks, but patient and family's expectations should be deal with extreme care.

Bogunovic L, Lenke LG, Bridwell KH, Luhmann SJ. Preoperative Halo-Gravity Traction for Severe Pediatric Spinal Deformity : Complications , Radiographic Correction and Changes in Pulmonary Function. Spine Deform. 2013; Palomo T, et al. Intravenous Bisphosphonate Therapy of Young Children With Osteogenesis Imperfecta: Skeletal Findings During Follow Up Throughout the Growing Years. Journal of Bone and Mineral Research, Vol. 30, No. 12, December 2015; Sato A, et al. Scoliosis in osteogenesis imperfecta caused by *COL1A1*/*COL1A2* mutations — genotype–phenotype correlations and effect of bisphosphonate treatment. The Bone Journal, Vol 86, 2016.