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Surgical Treatment With Pedicle Screws of Scoliosis Associated With Osteogenesis Imperfecta in Children

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Abstract

Study Design: Retrospective study.
Objective: To assess results of posterior instrumented fusion using pedicle screws in 12 children with osteogenesis imperfecta (OI) with spinal deformity at a single institution from 2001 to 2012.
Summary of Background Data: This is the first case series of OI patients who underwent non−cement augmented screw-rod instrumented fusion published in the literature.
Methods: Of a total of 54 children with spinal deformity associated with OI, 12 (22.2%) were submitted to posterior spinal fusion with pedicle screws (80% density) because of severe spinal deformity. Here we reported the results in seven females and five males.
Results: Five thoracic (41.7%), five double (thoracic and lumbar 41.7%), and two lumbar (16.7%) curves were considered. The mean number of fused levels was 11.8 (range: 5 to 16). Mean age at surgery was 13 years 8 months. Mean follow-up was 7 years 11 months (range: 3 years 7 months to 16 years 1 month). The mean preoperative scoliosis angle was 75.6°, whereas the postoperative angle was 31.4° (58.5% correction rate). The mean preoperative kyphosis angle was 57.4° and the postoperative angle was 42.3°. We observed one superficial infection, one dural tear, and three cases of proximal junctional kyphosis; two patients required one revision surgery each (2 years and 4 months postoperatively on average).
Conclusions: To our knowledge, this is the first case series published in the literature regarding OI with instrumented fusion with non−cement augmented pedicle screws exclusively in children with spinal deformity. We found that posterior spinal fusion with the screw-rod system in OI deformity in children is feasible and reliable, and has acceptable clinical and imaging results in the long-term follow-up.
Level of Evidence: Level IV

Keywords: Spine; Pediatric surgery; Osteogenesis imperfecta; Pedicle screws instrumentation; Deformity surgery; Scoliosis

Introduction

Osteogenesis imperfecta (OI) is a rare group of hereditary disorders of connective tissue affecting the quality and quantity of type 1 collagen. Around 250 mutations have been described for OI [1]. Depending on the type of OI, it may show an autosomal dominant or recessive inheritance pattern, affecting 1 in every 20,000 births [2-6].

Bone fragility, short stature, triangular facial shape, blue scleradentinogenesis imperfecta, hearing loss, vertebra plana, multiple episodes of bone fractures, and scoliosis/kyphoscoliosis are some of the most common signs that define this disease [6]. The primary diagnosis is made based on clinical findings, whereas skin biopsy and molecular studies remain the gold standard [1,7].

Considering the clinical and radiologic features, Sillence developed a classification that is still in use and divides OI into four different types [2]. Type 1 is the most frequent, with the least clinical impairment. Type 2 is the most severe form

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of OI, most likely lethal in the perinatal period. Type 3 is characterized by severe skeletal involvement and accounts for 20% of all four types. Type 4 is moderate to severe OI and may present with spinal deformity [2,8]. Later on, several modifications were added to the original classification, up to 17 different types are described in published literature.

Data from the literature show that pulmonary function is compromised when the spinal curve is over 60° [9-11], and respiratory insufficiency is the most common cause of death in young adults with OI [10-12]. Bone mass Z score is inversely correlated with the severity of scoliosis/kyphoscoliosis [12]. At the same time, the magnitude of the deformity is strictly correlated with the severity of the disease. The incidence of spine deformity in children with OI is between 39% and 80% [2,8,9,13-17].

Multidisciplinary management is considered vital. Osteogenesis imperfecta patients require clinical and imaging assessment and improvement of bone mass in quality and quantity and muscle strength to obtain full or partial mobility [18].

Orthotic treatment of patients with spine deformity associated with OI has been abandoned worldwide because of the lack of positive results [12,19,20]. Currently, the surgical treatment of choice consists of posterior instrumented fusion with pedicle screws [2,8,13,14]. In the past, when spinal arthrodesis with instrumented fusion was performed in patients with OI, the correction rate of spinal curves was not considered among the goals because of poor bone quality. Today, the goals of surgical treatment with pedicle screws is not only to prevent progression but also to correct the spinal deformity, with very satisfactory results. When a spinal curve shows a progressive pattern or is greater than 40°, posterior instrumented fusion with a screw-rod system should be considered [19]. Optimum age for instrumented fusion remains unknown [7,19].

The aim of this study was to assess postoperative results of 12 patients who underwent posterior instrumented fusion with a pedicle-screw-rod system for severe scoliosis/kyphoscoliosis deformity at a single institution from 2001 through 2012.

Materials and Methods

We evaluated 54 children with spinal deformity associated with OI with complete clinical and imaging exams from our database. We found 12 children who underwent posterior instrumented fusion with pedicle screws. Inclusion criteria were based on severe scoliosis/kyphoscoliosis, trunk shift, deformity progression, and 2 years minimum follow-up. We excluded patients with incomplete data or imaging films, or lost during follow-up. We analyzed sex, age, curve size, curve pattern, bone maturity and the Gross Motor Function Classification System (GMFCS). Data were obtained between 2001 and 2012.

Children were evaluated preoperatively with clinical exams, radiographs (including manual traction/benders films), spine magnetic resonance imaging, and eventually a spine CT scan, by two different spine surgeons. In addition, patients were assessed at the Departments of Skeletal Dysplasia, Genetics, Endocrinology, Pediatric Orthopedics, Physical Therapy, and Psychology when needed. This research project was approved by the IRB committee at Hospital de Pediatria Prof. Dr. Juan P. Garrahan.

Indications of surgery were as follows: scoliosis $>40°$, kyphosis $>75°$, trunk shift, progression of $>10°$ a year, or immature skeleton. At surgery, the patient was placed in prone position (extremely careful concern when positioning and transferring to the operating table considering as many people as possible to this procedure) and were intraoperative neurophysiological monitored during the entire procedure with somatosensory evoked potentials, transcranial motor evoked potentials, and electromyography taking into account a low motor intensity stimulation to prevent potential fractures during the monitoring (proper baselines traces were achieved during every procedure), we also used intraoperative halo cranial traction (8 cases, 50% of the total body weight). Surgical technique was addressed thoroughly concerning pedicle screws instrumentation; once the facets and transverse process were identified, using free hand technique, a gauge was used to prepare the pedicle entry point and we used a ball tip all the way till the anterior vertebra wall (Lenke's probe was basically not needed). Very carefully, Smith Petersen osteotomies were performed in all 12 patients. Deformity correction rate was adjusted to previous bendings and lateral or manual traction radiographs, taking into account not to hypercorrect with benders, preventing potential vertebra fracture and screw pullout. During surgery, we were assisted by a fluoroscopic image intensifier system to eventually check screws positioning when needed. We used irradiated bone allograft and autograft from spine exposure in every patient; non-BMP was used because of economic reasons.

Postoperatively, strict and thorough clinical exams were also conducted by two different surgeons at every visit and standard anteroposterior and lateral radiographs were taken every 3 months for the first year and once a year until the 5th year due to the extremely poor bone mass Z score (Table).

Results

We assessed the results of 12 children who underwent spinal surgery because of severe spinal deformity associated with OI. Seven of the patients were female (58.3%) and five were male (41.7%), with a mean age at surgery of 13 years 8 months (10 years 5 months to 18 years 5 months). The mean follow-up was 7 years 11 months (3 years 7 months to 16 years 1 month).

Three patients had type 1 OI (25%), one had type 2 (8.3%), seven had type 3 (58.3%), and one had type 4 OI (8.3%). Eleven patients (91.6%) had a history of several bone fractures (on average, 14.4 fractures per patient). We