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Growth-Friendly Surgery Is Effective at Treating Early-Onset Scoliosis Associated With Goldenhar Syndrome

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Abstract

Objective: To evaluate the radiographic results and complications of growth-friendly (GF) surgery in the treatment of early-onset scoliosis (EOS) associated with Goldenhar syndrome.

Background: Goldenhar syndrome has been associated with spinal deformity, which may be progressive. Efficacy and complication rate of GF treatment has not been reported for this population of patients with EOS.

Methods: Patients with Goldenhar syndrome and EOS with two years' follow-up were identified from two international multicenter EOS databases. Scoliosis, kyphosis, spine height, and hemithoracic height/width were determined preimplant, immediately postoperative, and at the two-year follow-up. Severity of complications (SV) was recorded (Smith et al. *JPO* 2015).

Results: Ten patients met inclusion criteria and had a mean age of 4.6 ± 2.5 years at GF implantation (one spine and nine rib-based). Mean preoperative scoliosis was 64°, postimplant 52°, and at mean follow up of 2.4 ± 0.5 years was 50° ($p = .09$). Preoperative kyphosis was 36°, postimplant 38°, and final 42° ($p = .08$). Preoperative T1–S1 height was 23.5 cm, postimplant 23.6 cm, and final 27.3 cm ($p = .06$). Preoperative convex hemithoracic height was 10.4 cm, postimplant 7.9 cm, and final 12.8 cm ($p < .05$). Preoperative concave hemithoracic height was 8.4 cm, postimplant 8.8 cm, and final 9.9 cm ($p = .30$). Preoperative right hemithoracic width was 8.02 cm, postimplant 7.22 cm, and final 7.86 cm ($p = .07$). Preoperative left hemithoracic width was 7.18 cm, postimplant 7.86 cm, and final 8.60 cm ($p = .43$). Eight patients had ⩾1 complication with SV I ($n = 7$), SV II ($n = 2$), and SV IIA ($n = 7$). These included infection ($n = 4$), migration ($n = 3$), pneumonia ($n = 2$), and instrumentation failure ($n = 2$).

Conclusion: At minimum two-year follow-up, GF surgical intervention for the treatment of EOS associated with Goldenhar syndrome trended toward improvements in scoliosis and spine height, but had a significant improvement in convex hemithoracic height; however, the majority of patients experienced severity grade I or II complications.

Level of Evidence: Level IV.

Keywords: Goldenhar syndrome; Early-onset scoliosis; Growth friendly; Oculoauriculovertebral dysplasia spectrum

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Introduction

In 1952, Goldenhar described three patients with phenotypic characteristics including epibulbardermaids, preauricular skin tags, mandibular asymmetry, and cervical vertebral abnormalities (eg, butterfly vertebrae, hemivertebrae, supplemental vertebrae, and rib anomalies) [1]. This constellation of anomalies has come to be known as Goldenhar syndrome or oculo-auriculo-vertebral spectrum (OAVS) [2], recognizing cardiac, renal, skeletal, and other anomalies that occur in addition to structural facial abnormalities, representing a spectrum of developmental abnormalities [3]. The incidence of Goldenhar syndrome has been estimated by prospective newborn studies to be between 1/3500 and 1/5600. This large range is due in part to the variable clinical expression of OAVS [4]. The spinal anomalies associated with Goldenhar syndrome have been poorly defined, which is likely due to the rarity of the syndrome and that the care of these patients is often directed by specialists whose principal interest is in facial reconstruction [5]. The prevalence of vertebral abnormalities in Goldenhar syndrome is estimated to be 16% to 60% of patients with the syndrome [3]. These congenital scoliosis curves may have variable severity that depends on age, size of deformity, location of the deformity, and type of vertebral anomaly [6,7].

Early-onset scoliosis (EOS) is defined as scoliosis with onset at an age less than 10 years, regardless of etiology [8,9]. EOS is complex to manage, as the growing spine is a variable and dynamic structure. It has been classified into four main etiologies—congenital, neuromuscular, syndromic, and idiopathic—as this group of patients is quite heterogeneous [10]. The important objective when managing this heterogeneous population is to prevent progression of spinal deformity while maintaining spine, chest wall, and lung development. To accomplish these goals, surgical intervention with growth-friendly treatment is often required. Currently, posterior distraction–based implants, such as spinal growing rods and rib-based devices including vertical expandable prosthetic titanium rib (VEPTR) (DePuy-Synthes Spine, Raynham, MA), are some of the most commonly used surgical treatments for EOS (Fig. 1). Although growth-friendly treatment is the gold standard for the treatment of EOS, it is known to have a high complication rate [11,12]. Smith et al. have described a classification system to grade the severity of complications that can be device-related or disease-related: device-related severity grade (SV) I is a complication that does not require unplanned surgery as it can be treated at the next scheduled surgery, SVII requires an unplanned surgery

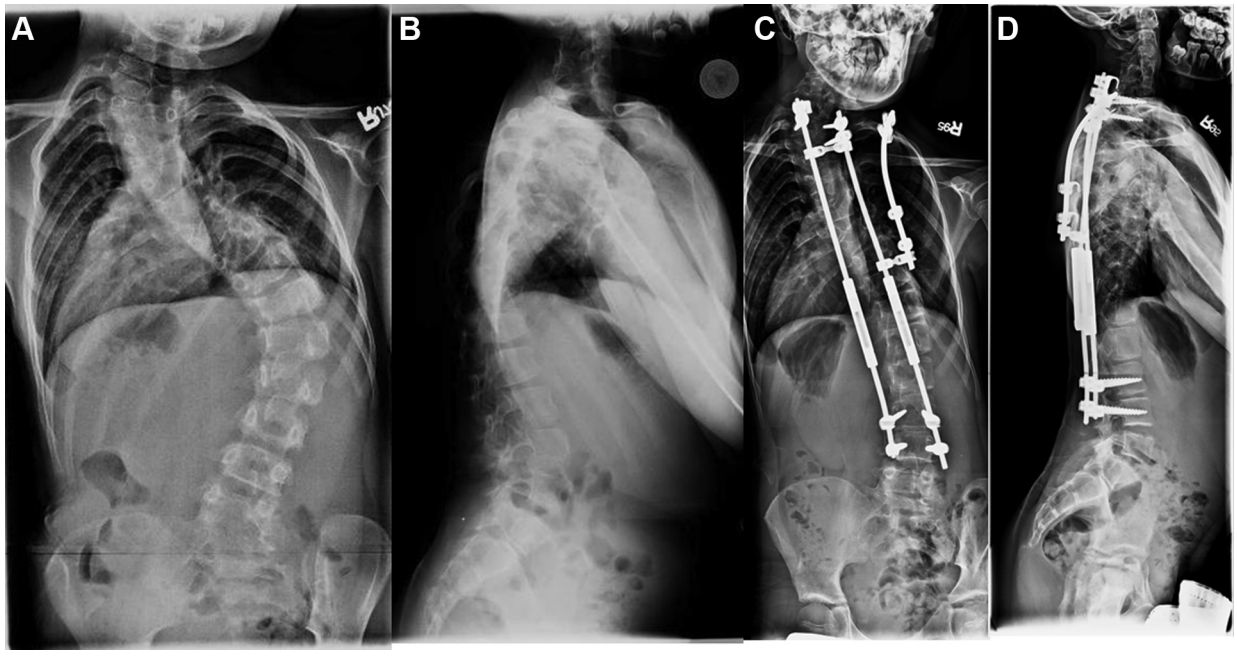

Fig. 1. Seven-year-old girl with Goldenhar syndrome. (A) Preoperative anteroposterior radiograph demonstrates 87° scoliosis. (B) Preoperative lateral radiograph. (C) Two-year postimplantation of rib-based distraction device demonstrates that T1–S1 height has increased from 28.3 cm postimplantation to 30.6 cm at two-year follow-up. (D) Two-year postimplantation lateral radiograph.