

Distraction osteogenesis in Goldenhar Syndrome: Case report and 8-year follow-up

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ABSTRACT

Goldenhar syndrome is a well-known condition featuring the following triad of anomalies: ocular abnormalities, microtia and vertebral anomalies. This syndrome involves structures arising from the first and second branchial arches. Craniofacial anomalies, including mandibular, zygomatic and/or maxillary hypoplasias are found in 50% of patients with Goldenhar syndrome. Patients with this syndrome may present unilateral or bilateral underdevelopment of the mandible. Several treatments for the correction of the dento-facial deformity have been described, among them distraction osteogenesis is one that shows promising results. Distraction osteogenesis is the process of bone formation that occurs during slow separation of the segments of bone after an osteotomy and it has been used to alleviate facial asymmetry. Mandibular distraction osteogenesis has been applied for many years, but long-term reports present controversial results. The purpose of the case report is to describe the immediate and long-term effects of distraction osteogenesis used to treat mandible asymmetry in a 5-year-old boy with Goldenhar syndrome.

Key words: Goldenhar syndrome, asymmetry, distraction osteogenesis.

INTRODUCTION

Goldenhar syndrome is a well-known developmental anomaly consisting of the triad of craniofacial microsomia, ocular dermoid cysts, and spinal anomalies. It was first described in 1952 by Goldenhar and was later included to a broader classification called oculo-auriculo-vertebral spectrum. This syndrome involves structures arising from the first and second branchial arches (1).

One of the most common craniofacial anomalies observed in this syndrome is unilateral underdevelopment of the mandible. The preferred method for treatment of hemifacial microsomia in children is distraction osteogenesis that consists on bone development through osteotomy and

sequential stretching of the healing callus (2-5). Mandibular distraction osteogenesis has been applied for many years, but long-term reports present controversial results (5,6). The aim of this paper is to describe a case of a child with Goldenhar syndrome in which distraction osteogenesis was used to treat the mandibular asymmetry and to discuss its follow-up.

CASE REPORT

A 3-year-old boy was examined at the Special Care Dentistry Center of the School of Dentistry, University of São Paulo. Extra-oral physical examination showed left microtia, strabism, left facial asymmetry, hypoplasia of the mandible,



Fig. 1. A. Lateral photograph showing the scars from periauricular appendices. B. Irregular skin depigmentation limited to the patient's left side.



Fig. 2. Postero-anterior radiography showing distraction device in place.



Fig. 3. A. Frontal photograph of the patient with Goldenhar syndrome before distraction. B. One year post-distraction. C. Eight-year post-distraction. Note the improvement on the facial contour after distraction and the relapse after 8 years of postdistraction. D. Underdevelopment of the patient's left hand.

mouth corner dropping on the left side, scars from previously removed periauricular appendices and irregular skin depigmentation limited to the left side of the upper body by the midsagittal plane (Figs. 1A,B). These facial features were consistent with a diagnosis of Goldenhar syndrome, further examinations revealed vertebral anomalies, scoliosis and right-side inguinal hernia.

The intra-oral aspects revealed asymmetry of the tongue, with the left side of the tongue smaller than the right side, besides the patient presented poor oral hygiene with caries in almost every teeth. Therefore it was important to treat the carious teeth and to improve oral health conditions before submitting the patient to a surgery procedure to treat the asymmetry. For this reason, specific oral hygiene preventive methods including motivation, and treatment procedures were used to obtain appropriate oral hygiene levels.

The patient was submitted to a distraction osteogenesis when he was 5 years old. The procedure included a small incision just above the angle of the mandible and an osteotomy at the ramus of the mandible, superior to the entrance of the inferior alveolar nerve into the mandibular canal, the surgical steel distraction device was secured onto both segments of the ramus with 2,4mm diameter screws (Fig. 2). The distraction device was activated once a day for 18 days, and then it was left in place for an additional 2 months in order to stabilize the area, the initial appearance and the final outcome of the distraction technique are shown in Figures 3A and 3B. Improved symmetry was obtained in the mandible and the occlusal plane was almost leveled, although post surgery was satisfactory, the 8-year follow-up revealed a skeletal relapse if compared to short time follow up as we can see in Fig. 3C. In Figure 3D, we can see that the patient's left side development was much smaller than the right side.

DISCUSSION

Goldenhar syndrome is a rare condition with an incidence rate ranging from approximately 1:3000 to 1:5000 live births, it is more common in males, with a male-female ratio of 3:2 and the patients right side of the face, body or both is generally more commonly and severely affected than the left (6).

Goldenhar syndrome is well known for its classical triad of ocular, auricular and vertebral disturbances (7), this syndrome may also present heart diseases (5-58% of the patients) (8), hypoplasia of the zygomatic, mandibular and maxillary bones, muscle hypoplasia, anatomical and morphological abnormalities of the tongue, vertebral anomalies, cleft palate, disturbance of the central nervous system and other visceral anomalies (9,10). Craniofacial anomalies, including mandibular, zygomatic and/or maxillary hypoplasias are found in 50% of patients with Goldenhar syndrome (11). An interesting finding in this case was the irregular skin depigmentation limited to the patient's left side of the back and chest, which is not a documented characteristic of this syndrome.

When the patient has hypoplasia of the mandible, orthog-

natic surgery or distraction osteogenesis can be used to correct the asymmetry (2), according to Wiens et al. (2) three advantages are associated to the use of distraction osteogenesis in the mandible. First, this procedure requires only 1 surgical site. Conventional ramus graft surgeries, which are associated with increased morbidity, use iliac crestal bone as an autogenous graft and thus require 2 surgical sites. Second, newly formed bone can be distracted more than once, enabling additional distractions as the patient grows. In fact, the distraction device components may be left in place by simply removing the transcutaneous pin through the incision and reinserting it at a later date. Third, soft tissues in the area accommodate and stretch with the distraction device and newly formed bone is of the same diameter and strength as the surrounding bone.

There is no consensus in the literature on when this procedure should be started. The authors agree that the facial development on the unaffected side proceeds normally, growing more than the distracted side, and propose over-correction and/or second-stage distraction in cases when the distraction is performed in early stages, before skeletal maturation. Therefore, early mandibular reconstruction would allow maxillary and dento-alveolar development to take place, reducing the need for or the amount of later surgery, besides it improves body image and socialization of the child (12), for this reason we decided to treat the patient when he was only five years old. Short time follow-up presented good results with asymmetry improvement, but 8 years after the distraction it could be noted a substantial return of the phenotype. The genetic input on neuro-muscular and skeletal architecture tends to slowly mould the distracted skeleton back to the original configuration (5), besides the patient's left side development was much smaller than the right side as we can see by comparing his hands (Fig 3D). Another way of explaining the relapse is based on patient's initial skeletal characteristics. The pressure from the distraction might have caused the proximal segment of the mandible to be rotated clockwise instead of lengthening the ramus height and positioning the mandible forward, and ramus inclination of both sides to be swung to the nonaffected side. These could be related to the lower improvement of sagittal, vertical, and asymmetric relationships between the maxilla and mandible. Therefore, in cases with initial skeletal characteristics such as more retrognathic mandible, smaller ramus height and articular angle, and a more vertical growth pattern, a delay in early distraction osteogenesis treatment may sometimes be the best option to prevent second surgical intervention at a later stage (13). The relapse shown in the present case may be due to the criteria mentioned above, but in 1998 when the surgery was performed the data were not available yet and the surgery was performed based on the patient's severe asymmetry.

Besides all these facts, some authors have proposed that the treatment of mandible asymmetry requires an interdisciplinary approach including maxillofacial surgery and orthodontics (14,15). According to Tehranchi & Behnia (14) hybrid functional appliances can be used to continue the

process of gradual lengthening of the mandible in order to improve neuromuscular function. Gradual distraction plus functional orthodontic therapy enhances facial symmetry and minimizes relapse. In this case, the patient did not have a correct orthodontic treatment, because of his social and economic conditions. After the surgery he moved to another city and could not continue the treatment so the follow-up was discontinued. Co-operation not only within the team, but also with the patients and their families is essential in order to achieve the best results (15). By putting these facts together we can conclude that the patient's skeletal characteristics and absence of the orthodontic treatment, which could have helped a redirection of the mandible growth, contributed to the treatment relapse. For this reason a new distraction osteogenesis will be scheduled when the patient turns 14 years old, when bone maturation is almost complete.

The decision about when the distraction osteogenesis can be performed should be based on the severity of the deformity. The ideal is to postpone the surgery until the complete development of the mandible is complete, but if there is a great asymmetry, the surgery must be carried through in early stages to avoid psychologic problems. In these cases, the patient's family has to know that relapse might occur and another surgery might be necessary. Long-term evaluation of a large number of patients will be necessary to evaluate the efficacy of this treatment protocol.

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