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Case Report

Facial Asymmetry in Goldenhar Syndrome: A Case Series with Review of Management Protocols

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Abstract

Background:

Goldenhar syndrome, also known as oculo-auriculo-vertebral spectrum or hemifacial microsomia, is a rare congenital condition characterized by craniofacial, ocular, auricular, and vertebral anomalies. Its incidence ranges from 1 in 3,500 to 1 in 5,600 live births, with underdiagnosis contributing to its low reported prevalence in India.

Objective:

To evaluate the clinical outcomes of distraction osteogenesis in patients with Goldenhar syndrome and highlight the importance of a multidisciplinary approach in achieving functional and aesthetic rehabilitation.

Methods:

A retrospective analysis was conducted on patients diagnosed with Goldenhar syndrome who underwent distraction osteogenesis for mandibular deformities. Treatment planning was individualized based on skeletal maturity and severity of asymmetry. Orthodontic therapy was integrated to correct occlusal discrepancies and enhance postoperative stability. Clinical and radiographic assessments were used to evaluate outcomes.

Results:

All patients demonstrated significant improvement in facial symmetry and underlying bone growth. Distraction osteogenesis facilitated early intervention, avoided donor site morbidity, and minimized the need for bone grafting. No major complications such as distractor malfunction or vector loss were observed. Orthodontic support contributed to long-term occlusal stability. Kaban's treatment objectives were successfully achieved in all cases.

Conclusion:

Distraction osteogenesis is a reliable and minimally invasive technique for correcting mandibular asymmetry in Goldenhar syndrome. When combined with orthodontic and multidisciplinary care—including ophthalmology, ENT, neurosurgery, and orthopedics—it offers optimal functional and psychosocial outcomes. Early diagnosis and individualized treatment planning are key to successful rehabilitation.

Introduction

Goldenhar syndrome, also known as hemifacial microsomia, is a rare congenital condition first identified by Dr. Maurice Goldenhar. It arises from disrupted development of the first and second branchial arches during embryogenesis, leading to a distinct set of craniofacial and systemic anomalies. The hallmark features include mandibular hypoplasia, which causes noticeable facial asymmetry, along with oculo-auricular malformations and vertebral anomalies [1].Ocular manifestations may range from microphthalmia and anophthalmia to cleft eyelids and exophthalmia, while ear-related abnormalities often include microtia, auricular fistulas, and external auditory canal atresia. Skeletal involvement can present as vertebral defects, cleft spine, or microcephaly, and in some cases, there may be associated internal organ anomalies, adding to the complexity of clinical management [2]. Craniofacial anomalies commonly associated with Goldenhar syndrome may include cleft lip, cleft palate, macrostomia, and bifid tongue. Patients often exhibit facial asymmetry, along with malocclusion and various dental irregularities, such as misaligned or missing teeth, which can complicate both function and aesthetics [3]. Poswillo's experimental studies in mice demonstrated that exposure to certain teratogenic agents can lead to the formation of hematomas in the stapedial artery, a vessel crucial for supplying blood to the first and second branchial arches during embryonic development. This vascular disruption may contribute to the pathogenesis of craniofacial anomalies [4].

In cases where facial bones are affected unilaterally, there is often associated involvement of the trigeminal and facial nerves. The facial nerve dysfunction is typically linked to abnormal development of the temporal bone on the affected side. The extent of these neuroanatomical and skeletal anomalies can range from mild asymmetry to severe deformities, depending on the severity of the underlying condition [5]. This paper presents the surgical strategies employed to correct facial asymmetry in three individuals diagnosed with Goldenhar syndrome. The cases include two patients—an adolescent and an adult—classified as Type 2A according to the Pruzansky-Kaban system, and one pediatric patient with complete mandibular agenesis, corresponding to Type 3.

Material and Methods

The surgical management of facial asymmetry in Goldenhar syndrome was approached through individualized treatment planning. Preoperative assessment included detailed clinical examination, radiographs and occlusal analysis to evaluate mandibular hypoplasia and associated craniofacial anomalies. All procedures were performed under general anaesthesia with multidisciplinary collaboration involving maxillofacial surgeons, orthodontists, and anaesthesiologists. Postoperative care included antibiotic prophylaxis, physiotherapy, and long-term follow-up to monitor growth and occlusal stability.

Cases

This paper discusses in detail the surgical management of 3 patients with Goldenhar Syndrome.

Case 1:

A 13-year-old adolescent presented to the outpatient department with classical features of Goldenhar Syndrome, including auricular deformities, coloboma, preauricular tags, and ocular anomalies (Figure 1). Clinical and radiographic evaluation revealed mandibular hypoplasia on the left side, characterized by reduced length of the ramus and body (Figure 2), consistent with Type 2A classification under the Pruzansky-Kaban system. Distraction osteogenesis was selected as the preferred modality for skeletal correction. A unilateral distraction procedure was performed to lengthen the ramus and body of the left mandible (Figure 3 & 4). An oblique osteotomy cut was strategically placed at the mandibular angle to facilitate controlled elongation. To support occlusal rehabilitation, an orthodontic appliance incorporating an anterior bite plane was fabricated, intentionally creating a posterior open bite (Figure 5). This allowed for passive eruption of the maxillary molars, aiding in occlusal plane levelling and contributing to the stabilization of the corrected mandibular contour (Figure 6 & 7).

Figure 1: An adolescent with typical features of Goldenhar syndrome.

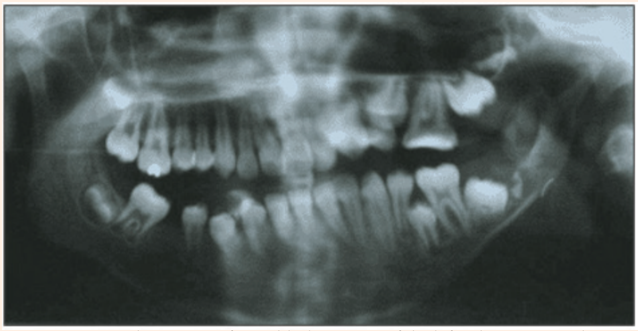

Figure 2: Shortening of mandibular ramus of the left side appreciated.

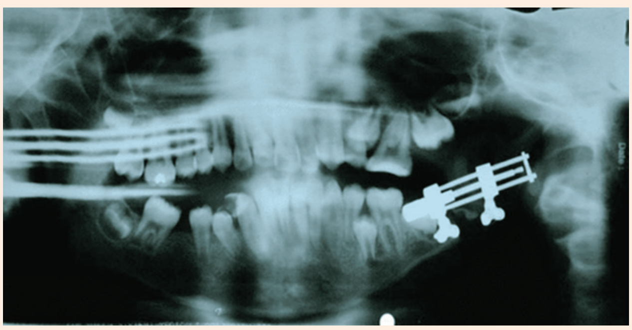

Figure 3: Day 10 of Distraction osteogenesis to increase the underlying bone.

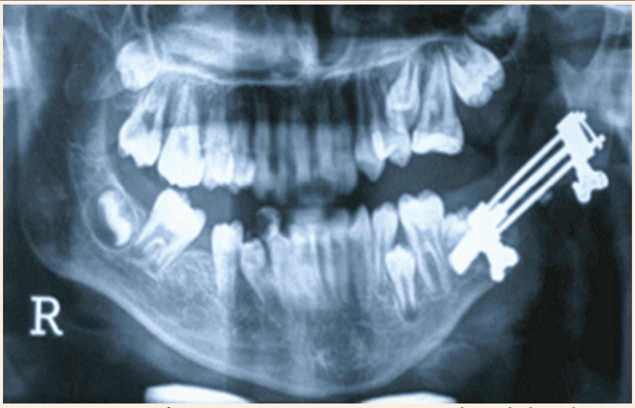

Figure 4: Day 25 of Distraction osteogenesis to increase the underlying bone.

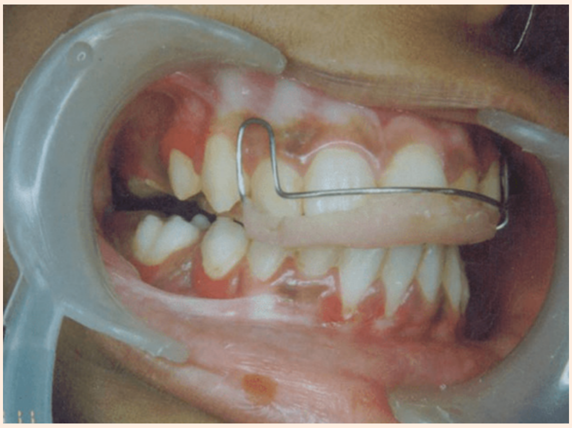

Figure 5: Anterior bite plane fabricated.

Figure 6: Post-op photos shows improvement in asymmetry

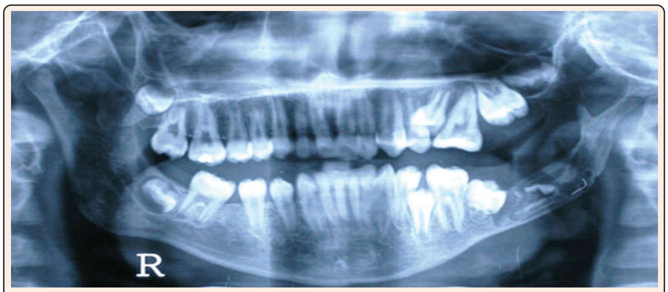

Figure 7: Post-op OPG after distractor removal showing new bone formation

Case 2:

An adult patient with a syndromic presentation reported with concerns of facial asymmetry. Clinical evaluation revealed auricular dysplasia, ocular anomalies, microphthalmia, and visual impairment, along with hypoplasia of the left mandibular ramus and body (Figure 8). Based on the anatomical and radiographic (Figure 9) findings, the condition was categorized as Type 2A according to the Pruzansky-Kaban classification. Notably, despite the skeletal deficiency, the patient exhibited adequate occlusal intercuspation, although an occlusal cant was evident (Figure 10), accompanied by soft tissue deficiency on the affected side. Given the patient's skeletal maturity and stable occlusion, Molina's Technique of simultaneous mandibular and maxillary distraction was considered appropriate for correction of occlusal cant. A simultaneous distraction osteogenesis of both the mandible and maxilla was performed using distractor placement in left mandible and LeFort 1 osteotomy in maxilla (Figure 11 & 12). Inter-maxillary ligation was initiated on the fifth day post-latency and continued through the six-week consolidation phase. The procedure resulted in a successful correction of the occlusal cant and facial asymmetry, with satisfactory aesthetic and functional outcomes (Figures 13-15).

Figure 8: A female syndromic patient with hypoplasia of left mandible.

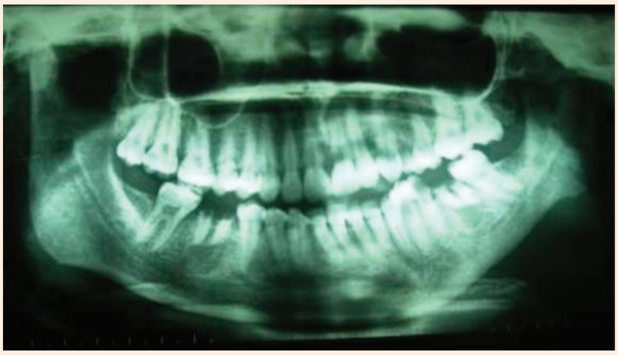

Figure 9: Orthopantomogram confirming hypoplasia of left ramus and body of mandible.

Figure 10: Good occlusal intercuspation but occlusal cant noted.

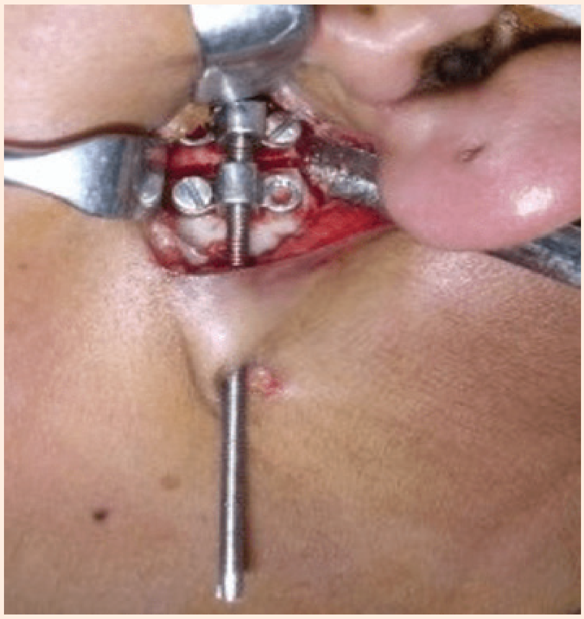

Figure 11: Osteotomy site and distractor placed in the left ramus region.

Figure 12: Simultaneous Le forte 1 osteotomy performed.

Figure 13: PA Skull showing simultaneous maxillary and mandibular distraction by Molina's Technique with distractor device in-situ.

Figure 14: Post op photos showing correction in facial asymmetry.

Figure 15: Occlusal Cant corrected.

Case 3:

A young paediatric patient presented to the outpatient department with complete agenesis of the left mandibular ramus (Figure 16), glenoid fossa, and temporomandibular joint (TMJ). The condition was classified as Type 3 under the Pruzansky-Kaban system, indicating a total absence of the osseous framework necessary for initial distraction osteogenesis (Figure 17). Due to the lack of native bone, a free fibula graft was employed to reconstruct the deficient mandibular segment and associated anatomical structures on the affected side (Figure 18). Once the graft demonstrated successful integration, it was planned to serve as a foundation for future distraction osteogenesis, anticipating asymmetry that could develop from normal growth on the contralateral side. Following the reconstructive procedure, an orthodontic guiding flange was provided to assist in occlusal alignment and stabilization (Figure 19). Improvements in facial symmetry and occlusion were achieved (Figure 20).

Figure 16: Complete agenesis of left side with ear deformity.

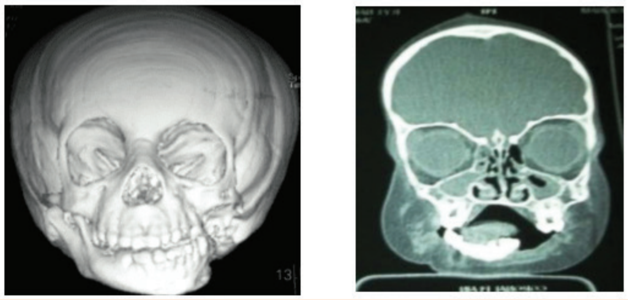

Figure 17: 3D reconstruction CT image showing agenesis of left side of mandible.

Figure 18: Reconstruction using free fibula osteocutaneous flap.

Figure 19: Guide flange prosthesis fabrication.

Figure 20: Follow up photograph and OPG showing Free fibula graft taken up well for reconstruction of left mandible.

Discussion

Goldenhar syndrome presents with a broad spectrum of phenotypic variability, which often complicates its clinical management. Effective treatment planning requires careful consideration of several factors, including the severity of craniofacial deformity, the specific anatomical structures involved, the patient's age at presentation, and the extent of functional impairment. To streamline therapeutic goals, Kaban proposed a structured approach emphasizing key objectives like augmenting mandibular volume and associated soft tissues, establishing a functional temporomandibular joint, encouraging vertical growth of the maxilla, and ultimately achieving a stable and functional occlusion [6].

To optimize individualized treatment planning, it is essential to accurately characterize the condition, identify the specific anatomical regions affected, and assess the extent of structural involvement. Given the wide variability in clinical presentation, multiple classification systems have been developed to categorize the severity of craniofacial anomalies associated with Goldenhar syndrome, aiding in both diagnosis and therapeutic decision-making. One of the earliest classification systems for mandibular deformities in craniofacial microsomia was proposed by Pruzansky, who categorized the anomalies into three types based on radiographic morphology [7]. This system primarily focused on the structural integrity of the mandibular ramus and condyle. Later, Kaban refined Pruzansky's framework by subdividing Type II into IIA (where the temporomandibular joint is reconstructable) and IIB (where reconstruction is not feasible), thereby integrating both anatomical deformity and functional assessment of the TMJ [8].

In 1987, David et al. introduced the SAT classification, which systematized craniofacial anomalies into three domains: Skeletal (S), Auricular (A), and Tissue (T) components. Each domain was graded independently to reflect the severity of deformity, offering a modular approach to clinical evaluation [9].

Building on this, Vento et al. developed the OMENS system in 1991, which remains one of the most comprehensive and widely adopted classification tools. OMENS evaluates five key anatomical regions: Orbit, Mandible, Ear, Nerve, and soft tissue. Each component is scored based on severity, and the system was later expanded to OMENS-Plus to include extra-craniofacial anomalies such as cardiac and vertebral defects [10].

The surgical approach to mandibular deformities in Goldenhar syndrome is guided by the Pruzansky-Kaban classification and the severity of anatomical disruption [11]. In patients with mild anomalies, such as Type I and Type IIA, active surgical intervention during growth is often unnecessary. Instead, orthopaedic appliances may be employed to guide mandibular positioning and correct occlusal cant. Upon reaching skeletal maturity, orthognathic surgery becomes the preferred modality, with bimaxillary procedures and genioplasty frequently recommended to achieve facial symmetry and functional occlusion.

For adolescent patients with Type IIA deformities, distraction osteogenesis (DO) offers a reliable method for early correction. This technique facilitates simultaneous expansion of bone and soft tissue, allows for strategic overcorrection, and may obviate the need for future grafting or soft tissue augmentation. In adult cases, simultaneous maxillary and mandibular distraction can effectively restore ramal height, correct occlusal cant, and improve soft tissue volume, often minimizing the need for extensive orthodontic preparation.

Management of Type IIB deformities hinges on the integrity of the temporomandibular joint (TMJ). If adequate bone is present, DO may be feasible; however, more severe deficiencies necessitate bone grafting followed by orthognathic surgery. In such cases, the skull base may function as a pseudo-TMJ, with distraction vectors planned accordingly. Consolidation is typically marked by ipsilateral open bite, and further correction may involve Le Fort I osteotomy to address residual asymmetry.

Type III deformities, characterized by complete absence of the mandibular ramus and TMJ, require complex reconstruction. Vascularized free flaps—such as fibula or iliac crest grafts—are preferred due to their capacity to provide both bony structure and soft tissue bulk. These grafts may later support DO or orthognathic procedures. Although total joint replacement (TJR) is a potential option, its application in paediatric patients remains controversial due to concerns regarding growth inhibition and the likelihood of future revision surgeries.

Across all classifications, the overarching goals of surgical management include restoration of vertical ramal height, establishment of a functional neocondyle, and achievement of stable occlusion and facial symmetry. Treatment planning must be individualized, multidisciplinary, and responsive to both anatomical constraints and developmental considerations.

Distraction osteogenesis offers several clinical advantages, including the elimination of intermaxillary fixation, minimal intraoperative blood loss, avoidance of bone grafting procedures, and absence of donor site morbidity. Most notably, it enables early surgical intervention during growth phases, which is particularly beneficial in craniofacial anomalies. While certain drawbacks may be encountered—such as patient discomfort from the distractor device, the need for a secondary procedure for its removal, potential mechanical failure, or unintended vector deviation—these limitations are generally minor and were not observed in our clinical experience.

Goldenhar syndrome encompasses a broad spectrum of craniofacial and systemic anomalies, necessitating a holistic and individualized treatment strategy. The maxillofacial surgeon plays a pivotal role in orchestrating comprehensive care, which must be delivered through a multidisciplinary framework. Collaboration with ophthalmologists, otolaryngologists, neurosurgeons, orthopedic specialists, and orthodontists is essential to address the diverse needs of these patients and ensure optimal functional and aesthetic outcomes.

Results

The patients with Goldenhar syndrome discussed in this article exhibited significant improvement in facial symmetry following surgical intervention. Distraction osteogenesis proved effective in promoting underlying bone regeneration and compensating for skeletal deficiencies. Adjunctive orthodontic therapy played a vital role in addressing occlusal discrepancies and enhancing the long-term stability of the reconstructed bone. Clinical and radiographic evaluations confirmed favorable outcomes, with measurable gains in both hard and soft tissue architecture. The treatment goals outlined by Kaban's protocol were successfully met across all cases, underscoring the efficacy of a structured, multidisciplinary approach in managing craniofacial anomalies associated with Goldenhar syndrome.

Conclusion

Goldenhar syndrome, also referred to as hemifacial microsomia, has a reported incidence ranging from 1 in 3,500 to 1 in 5,600 live births [12], with its prevalence in India likely underestimated due to underdiagnosis or misclassification. The absence of a definitive diagnostic test and the phenotypic overlap with other craniofacial syndromes contribute to diagnostic challenges. Accurate identification requires a thorough evaluation of the patient's craniofacial morphology and skeletal maturity. Once the underlying deformity is understood, a patient-specific treatment plan is developed, taking into account the psychosocial stages of development. Distraction osteogenesis serves as a valuable modality in this context, offering the potential to achieve stable facial symmetry, minimize the need for multiple surgical procedures, and positively influence the patient's self-confidence and psychological well-being.

References

1. Katara P, Gupta R, Jain M (2023) Goldenhar Syndrome In A Paediatric Patient: A Case Report. IOSR Journal of Dental and Medical Sciences 22: 22-23.
2. Schmitzer S, Miruna Burcel, Dăscălescu D (2018) Ioana Claudia Popteanu. Goldenhar Syndrome - ophthalmologist's perspective. Romanian Journal of Ophthalmology 62(2): 96.
3. Villani R, Michelina V, Fabbri E, Zarabini A, Morselli P (2017) Central Bringing Excellence in Open Access Goldenhar Syndrome: Surgical Planning in a Severe Case with Cleft Lip and Palate, Coloboma and Macrostomia.
4. Poswillo D (1988) The aetiology and pathogenesis of craniofacial deformity. Development. 103(Supplement): 207-212.
5. Renkema RW, Caron CJJM, Mathijssen IMJ (2017) Vertebral anomalies in craniofacial microsomia: a systematic review. Int J Oral Maxillofac Sur 46(10): 1319-1329.
6. Mommaerts MY (2015) Hemifacial microsomia: management of the vertical ramus compartment. Plast Aesthet Res 2: 99-106.
7. Pruzansky S (1969) Not all dwarfed mandibles are alike. Birth Defects 4: 120.
8. Kaban LB, Padwa BL, Mulliken JB (1998) Surgical correction of mandibular hypoplasia in hemifacial microsomia: The case for treatment in early childhood. J Oral Maxillofac Surg 56(5): 628-638.
9. David DJ, Mahatumarat C, Cooter RD (1987) Hemifacial micro- somia: A multisystem classification. Plast. Reconstr. Surg 80(4): 525-535.
10. Vento AR, LaBrie RA, Mulliken JB (1991) The O.M.E.N.S. classification of hemifacial microsomia. Cleft Palate Cranio- fac J 28(1): 68-76.
11. Murray JE, Kaban LB, Mulliken JB (1984) Analysis and Treatment of Hemifacial Microsomia. Plastic and Reconstructive Surgery 74(2): 186-199.
12. Kokavec R (2006) Goldenhar Syndrome with Various Clinical Manifestations. The Cleft Palate-Craniofacial Journal 43(5): 628-634.