Dental Treatment and Medical Considerations for Patient with Goldenhar Syndrome

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Abstract

Goldenhar syndrome (GS), or oculo-auriculo-vertebral spectrum (OAV), is a congenital condition that affects one in every 3,000-5,000 births. This condition impacts the development of eyes, ears, and spine. A defining characteristic of OAV is hemifacial microsomia; the underdevelopment of the jaw and facial structures can cause significant challenges for the dental management of patients with OAV. Comprehensive dental treatment for patients with Goldenhar syndrome should include appropriate medical management, often involving a craniofacial team. This case report details the dental and medical coordinated care of a 13-year-old patient with Goldenhar syndrome. Due to the severity of hemifacial microsomia, this patient was treated in the operating room and received extractions, restorative treatment, and non-operative treatment as part of comprehensive dental care. Coordination with the craniofacial team is paramount in guiding dental and orthodontic treatment, particularly at appropriate developmental milestones. Lifelong team coordination can significantly improve patient outcomes and esthetic results.

Etiology and Diagnosis

First described by Maruice Goldenhar in 1952, this condition presents on a spectrum. [2]

Two theories of etiology:

- Abnormal development of the first and second branchial arches [6]
- Changes occur at 30-45 days gestational age based on myriad of anomalies [4]

Diagnosis:

- No universal diagnostic test or set of criteria
- Facial phenotype is often used for diagnostic purposes when there are sufficient anomalies
- Prenatal diagnosis possible with ultrasound imaging in presence of severe mandibular hypoplasia, cleft lip and/or cleft palate, and severe auricle abnormality [4]

Clinical Presentation

Apart from facial structures, anomalies in the renal, cardiac, skeletal, and nervous systems are seen.[4]

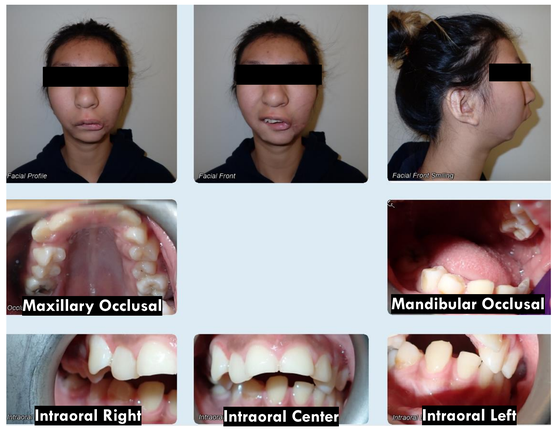
- Systemic features: language and speech delays, cranial nerve involvement, pulmonary anomalies, ventricular septal defects, tetralogy of Fallot, cervical spine instability, radial limb anomalies, imperforate anus, absent kidney, anomalous blood supply to kidney, double ureter, hydronephrosis[4]
- Common facial and dental features: microtia, preauricular skin tags, facial asymmetry, midface deficiency, micrognathia, malocclusion of teeth with poor oral hygiene, caries[3]

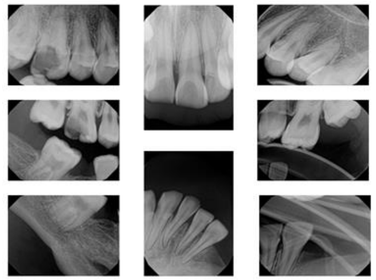
Dental Considerations

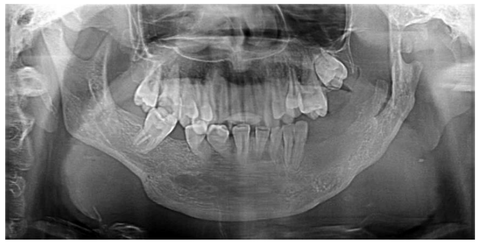
Severity of facial and dental anomalies can guide treatment recommendations/decisions.

- Limited opening
  - Accessibility of dentition/Space for instruments
- Airway concerns
- Caries risk and progress
  - Nutrition and oral hygiene modifications
- Malocclusion/Orthodontic concerns
- Location for dental treatment
- Coordination with other service lines

Case Report







Patient: 13-year-old female presents for routine examination with no chief concern
Medical History: Goldenhar syndrome, Cri-du-chat syndrome, microsomia, mandibular retrognathism, anemia, upper airway resistance syndrome, ear abnormality/hearing loss, speech problems, innocent heart murmur, first-degree AV block and left ventricular hypertrophy
Medications/Allergies: none
Social History: lives at home with mother and father
Oral Hygiene: fair- generalized moderate plaque, no calculus
Diet: sugar-sweetened beverages and snacks throughout the day
Caries Risk Assessment: high
Extraoral Exam: left-sided micrognathia, thick scar tissue along left mandible, limited opening
Intraoral Exam: caries #3, 14, 28, 30; deep grooves and pits #2, 4, 5, 12, 29; class II occlusal tendency

Treatment

Initial treatment plan in clinic: Recommended comprehensive dental rehabilitation in the operating room under general anesthesia. Discussed stainless steel crown vs extraction of #3, composites on #14 and #28, sealant on #2, and stainless steel crown on #31.

While under general anesthesia a full-mouth series of radiographs and exam were completed. Treatment plan was updated and all dental treatment needs completed. Patient received a dental prophylaxis and fluoride varnish application.

Updated treatment plan in operating room: extraction #3 and 13, composites #14-O, 28-B, 30-OB, silver diamine fluoride (SDF) #23-D, and sealants #2, 4, 5, 12, 29

- #3 non-restorable due to extent of caries
- #13 extracted per orthodontist
- #23 unable to access with handpiece; placed SDF until further surgical intervention with craniofacial surgeon allowing for sufficient access to restore tooth

Discussion

Patients with Goldenhar syndrome can present with varying degrees of severity which dictate the type, timing, and extent of treatment needed. As the craniofacial and dental anomalies of this syndrome are the most prominent, patients benefit greatly from interdisciplinary craniofacial teams. Comprehensive craniofacial teams may include the following service lines:

- Plastic surgery
- Otolaryngology
- Oral surgery
- Dentistry
- Orthodontics
- Genetics
- Psychology
- Speech
- Nutrition
- Social work[5]

Minimally invasive dentistry is particularly helpful in this patient population. [1] The principles of prevention that pediatric dentists recommend for all patients apply to patients with GS. Establishing a dental home in accordance with the American Academy of Pediatric Dentistry guidelines, receiving consistent dental exams, and achieving good oral hygiene practices can reduce caries risk and increase dental health for these patients.

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