

Introduction

Goldenhar syndrome, also known as: oculoauriculovertebral spectrum is a congenital condition that is caused by developmental defects in the first and second brachial arches. It is characterized by a classic triad: unilateral facial asymmetry, ear and/or eye malformations and vertebral anomalies but can also present with renal, cardiac, GI, and central nervous system defects. Dental manifestations include malocclusion, congenitally missing teeth, supernumerary teeth, cleft lip and/or palate, micrognathia, enamel hypoplasia and gingival hypertrophy. More severe dental presentations include complete absence of the mandibular ramus and TMJ.

Case Report

This presentation discusses a 5-year-old female who presented to Children's Wisconsin Dental clinic by referral from Plastics. Patient reported inconsistent pain on lower left side of her face. Patient's medical history is significant for Goldenhar syndrome with left hemifacial microsomia/microtia/agenesis of left mandible and left TMJ. Patient has history of left mandibular graft necrosis which was partially removed. No known drug allergies or daily medications. Clinical and radiographic examination revealed multiple carious lesions of varying extent, left side tongue fibroma, enamel hypoplasia, congenitally missing teeth, and remaining hardware present in left side soft tissue. Treatment was completed under general anesthesia in coordination with oral surgery. This report will include clinical, radiographic, histological findings and treatment rendered.

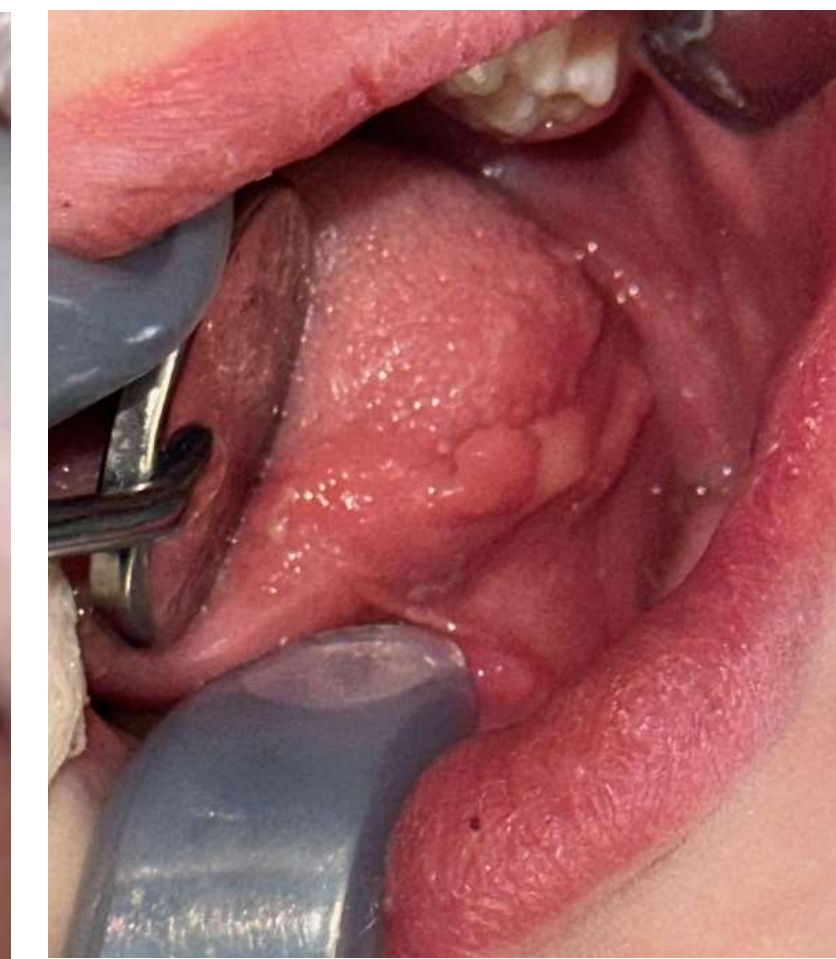
Clinical & Radiographic Findings

Clinical Findings:

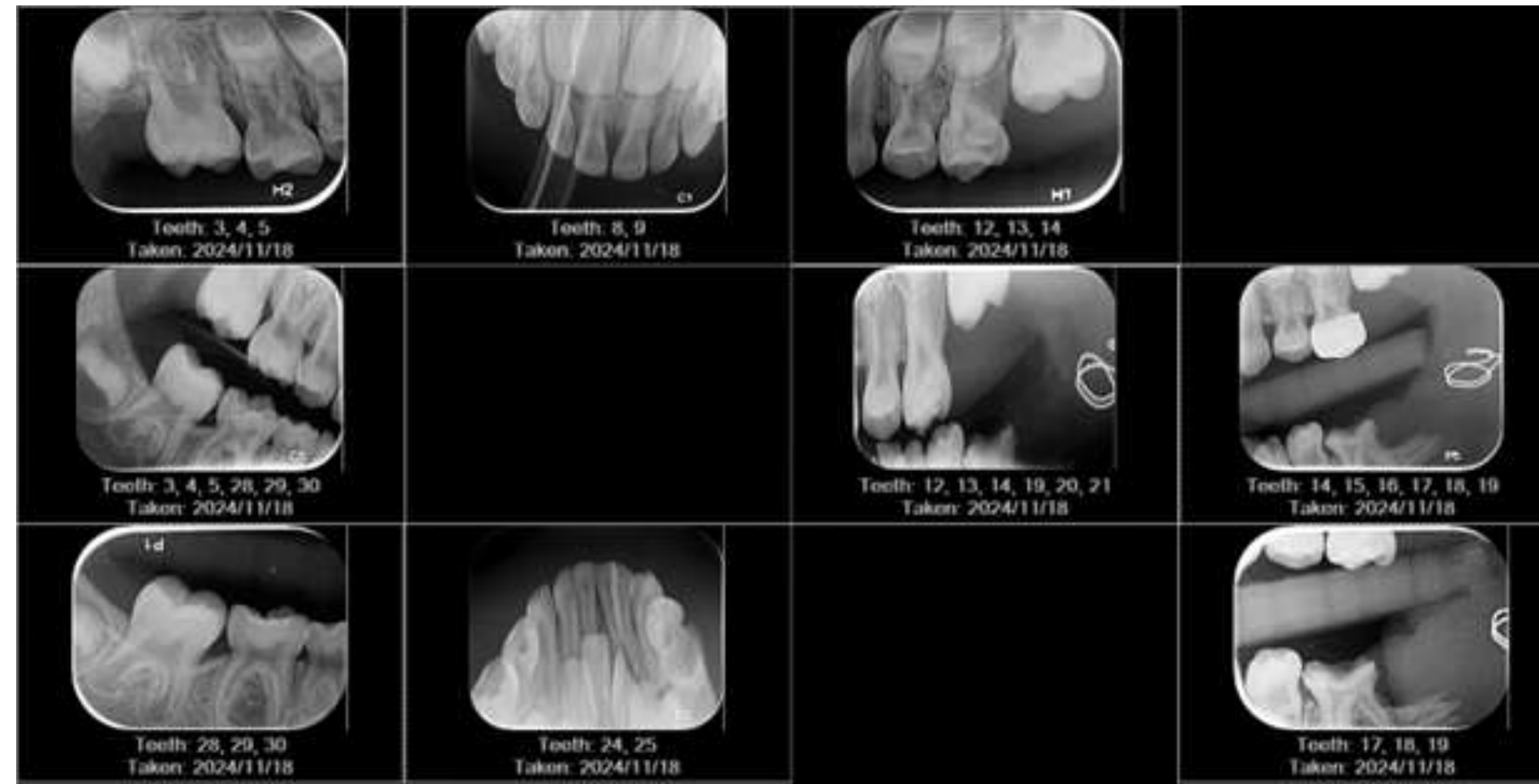
- Congenital deformity of the left ear
- Gingivitis
- Enamel hypoplasia
- Attrition
- Severe lower anterior crowding
- Dental caries
- Non-restorable carious tooth, ankylosis and enamel hypoplasia: #K
- ~4mmx4mm round pink fibroma located to the left lateral border of the tongue

Radiographic findings:

- Dental caries
- Agenesis of the left mandible and left TMJ
- Congenitally missing teeth
- ~8-12mm retained wire located in left cheek soft tissue mucosa



Clinical photos during GA: 11/18/2024



Comprehensive film series during GA: 11/18/2024

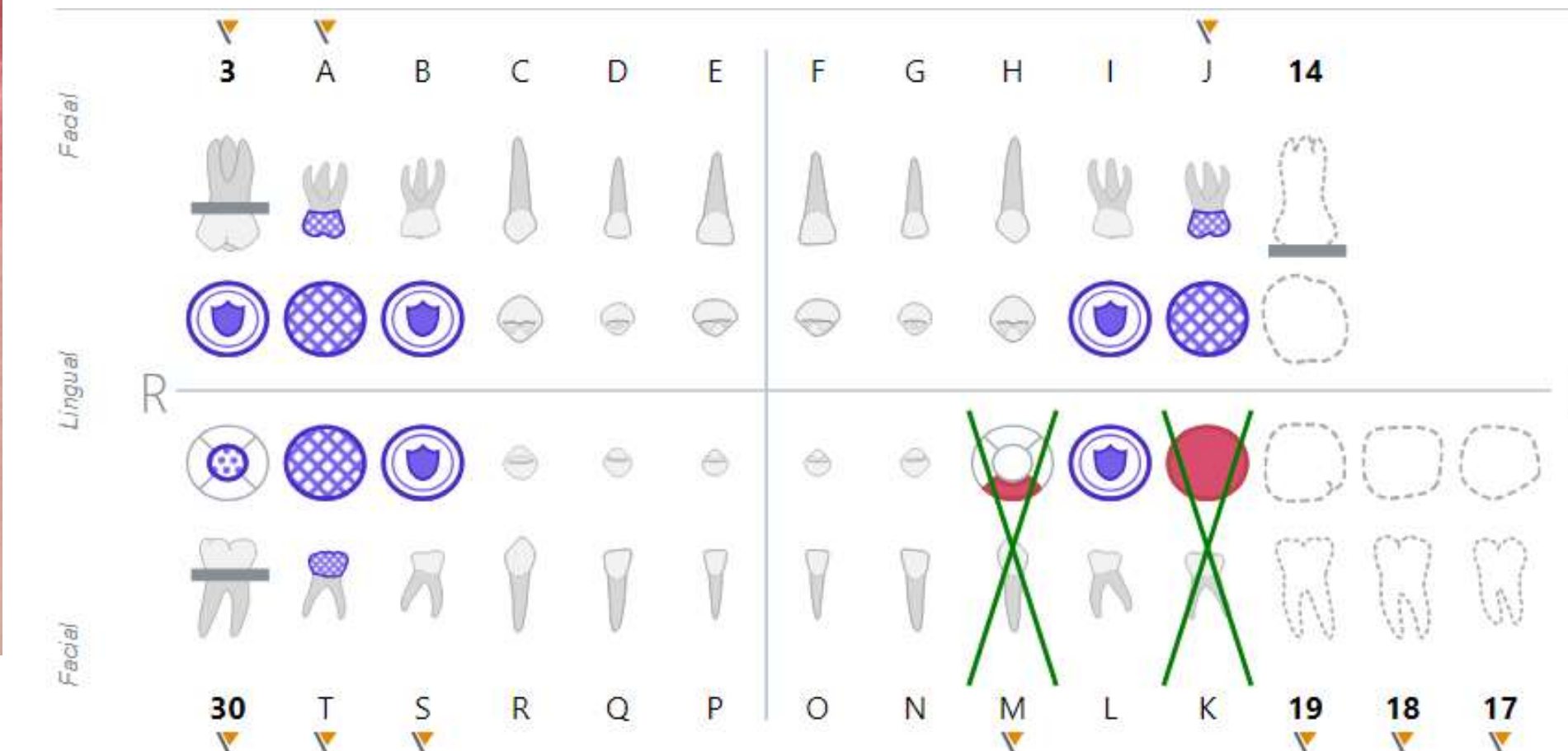


Non-restorable #K & retained wire: 11/18/2024



Panoramic radiograph: 8/14/2024

Treatment Rendered



- Seal #3,B,I,L,S
- Resin #30: Occlusal
- Stainless steel crown: #A,J,T
- Extraction: #K,M
- Removal of retained wire from left soft tissue mucosa
- Excisional biopsy of left tongue fibroma
- Histological Diagnosis:
 - Benign intraoral fibroepithelial polyp with focal degenerative features consistent with traumatic injury

Conclusion

Recommended follow up for this patient is dental recall in May 2025. It is important for patients with Goldenhar syndrome to establish a dental home and follow a 6-month dental recall schedule. They present with unique dental needs that often require specialized and coordinated care. This case report demonstrates the magnitude of medical and dental complexity that encompasses this syndrome and the importance of individualized care.

References

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