Oral Considerations for a 5-year old Patient with Trisomy 18: A Case Report

De Leon-Balitaan, Marivic DMD; Olson, Tanner DDS; Stewart, Ray DMD, MS; Tanbonliong, Thomas DDS

University of California, San Francisco, Division of Pediatric Dentistry

UCSF School of Dentistry

Introduction

Trisomy-18, commonly known as Edward's Syndrome, is a severe genetic condition involving low birth weight, multiple birth defects, and several defining physical characteristics. Affected individuals often present with overlapping fingers, low-set ears, hypotonia, club feet, and heart and lung abnormalities among others[1]. For babies who survive pregnancy and labor, only 10% survive their first year[2]. Children born with trisomy-18 require specialized, interprofessional care to address their unique symptoms. Secondary to hypotonia and feeding difficulties, patients require foods to be high in sugar and in puree form. Along with an inability to independently perform oral hygiene, patients face a high risk for developing dental caries.

ETIOLOGY AND EPIDEMIOLOGY

- 1 in every 6,000 to 8,000 live births[1]
- 1 in every 2,500 to 2,600 pregnancies[1]
- Prevalence rises with higher maternal age[3]
- De novo genetic mutation at the time of fertilization[3]
- "Mosaic trisomy 18" occurs when some cells in the body do not have three copies of chromosome 18[4]

Case Report

This presentation details a case report involving a 5-year old boy with Trisomy-18 who presented for treatment of severe early childhood caries at the UCSF Pediatric Dental Clinic. Legal guardian gave written and verbal permission for use of radiographs and photos.

MEDICAL HISTORY

Due to extensive medical history and behavioral components, treatment was rendered under general anesthesia in a hospital setting. Patient required oral intubation with UCSF's cardiac anesthesia team.

- Unrepaired ventricular septal defect
- Patent ductus arteriosus (small)
- Pulmonary hypertension
- Seizures
- Cleft lip and palate (right)
- Crossed renal ectopia with fusion anomaly
- Obstructive sleep apnea
- Slow weight gain
- Feeding problems – G-tube fed and pediatric standard formula
- Inferior orbital lesion (left)
- Restrictive strabismus

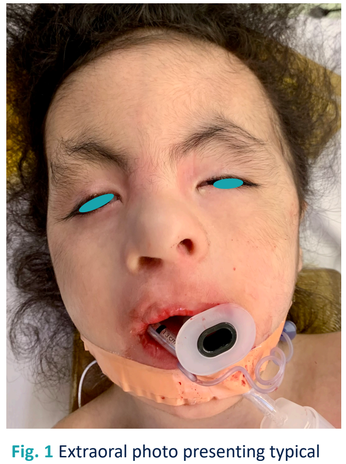


Fig. 1 Extraoral photo presenting typical features of Trisomy 18: dolichocephaly, ptosis, short palpebral fissures, cleft lip and palate, and micrognathia.

PHYSICAL CHARACTERISTICS

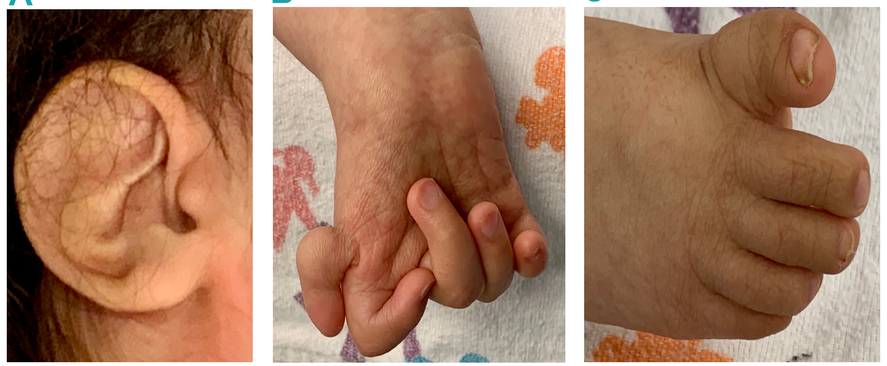


Fig. 2 Extraoral photographs of physical characteristics unique to Trisomy 18. A: microtia of right ear, B: clenched right fist with overlapping fingers and polydactyly, C: clubbing of right foot with abnormal hallux.

CLINICAL AND RADIOGRAPHIC FINDINGS

Upon clinical and radiographic examination, findings included the following:

- Right clefting
- Narrow palatal vault
- Generalized mild hypomineralization
- Generalized plaque and calculus consistent with children who are G-tube fed
- Congenitally missing #D
- Taurodontism of all primary molars
- Multi-surface decay of #E, F, G
- Deep pits and fissures of #A, B, ,I, J, K, L, S, T with no clinical or radiographic caries

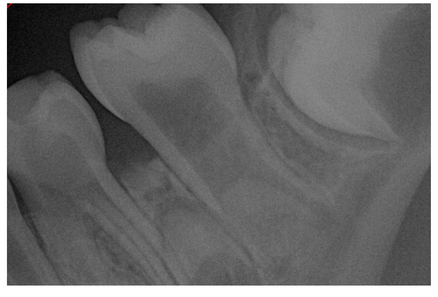
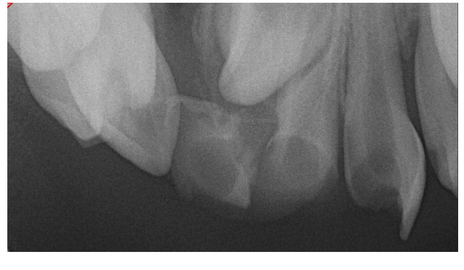


Fig. 3 Radiographic evidence of #L and #K taurodontism, also seen on all other primary molars.



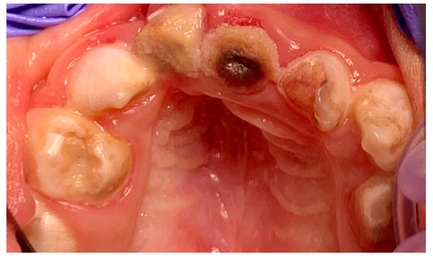


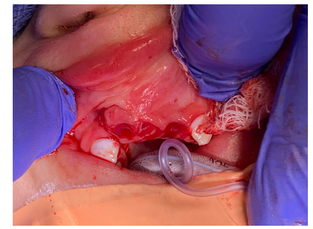
Fig. 4 Maxillary anterior intraoral photo and periapical radiograph showing gross decay of teeth #E-G with narrow high arched palatal vault, congenitally missing #D, and right cleft palate.

Diagnoses

- Caries Risk Assessment: High
- Gingivitis secondary to plaque, calculus, poor oral hygiene
- Severe early childhood caries
- Congenitally missing #D
- Gross caries: #E, F, G
- Deep pits and fissures: A, B, I, J, K, L, S, T

Treatment Considerations

- Due to the challenges in managing these patients, emphasis should be placed on prevention and early intervention focusing on frequent recall periodicity and fluoride exposure.
- For patients with caries, SDF should be utilized to prevent the progression of lesions.
- Glass ionomer can be utilized for protective restorations due to the fluoride release and hydrophilicity of material.
- SSCs and full coverage restorations should be considered for definitive treatment.
- Completed: EO/IO exam, radiographs, prophy with scaling, extractions of E, F, G, restorations of A, B, I, J, L, S, T with Equia Forte glass ionomer, fluoride varnish.



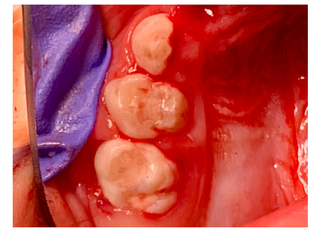


Fig. 5 Maxillary anterior region post-extractions of #E-G, Equia forte occlusal restorations of #A and B.

Conclusions

Children with Trisomy 18 present unique challenges to the pediatric dentist. Secondary to hypotonia and feeding difficulties, patients are often fed a diet high in sugar and require foods to be in puree form. Along with a cariogenic diet, oral hygiene falls on the caretaker as patients are unable to independently perform tasks of daily living. Additionally, general and pediatric dental offices are often unequipped to provide the necessary care for patients with Trisomy 18 resulting in treatment at a hospital-based dental programs. This case report highlights the complexity of dental and medical needs, and treatment rendered for a 5-year-old living with Trisomy 18 at the UCSF Benioff Children's Hospital. These patient often require muti-disciplinary patient centered care between medicine and dentistry.

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