



University of Colorado Anschutz Medical Campus

Introduction

Ectodermal dysplasia affects the development of organs derived from the ectoderm including nails, skin, salivary glands, and teeth.¹

The common form, hypohidrotic ectodermal dysplasia, is caused by EDA and EDAR gene mutations.² The inheritance pattern can be X-linked, autosomal recessive, or autosomal dominant.³ Ectodermal dysplasia affects about 1 in every 100,000 live births.³

Clinical Features:

- Fine hair
- Little to no eyebrows
- Periorbital pigmentation
- Frequent sinus infections
- Low set and pointed ears
- Depressed nasal ridges
- Increased risk of bronchial asthma and allergic rhinitis³

Oral Findings:

- Hypodontia/oligodontia and tooth development⁴
- Compromise esthetics, phonetics, chewing, and swallowing⁴
- High palatal arch, cleft lip/palate
- Cone-shaped lateral teeth
- Enamel hypoplasia
- Asymmetric alveolar ridge and reduced lower facial height¹

Case report and Objectives

A 35-month-old male initially presented to the University of Texas Health Houston Pediatric Dental Clinic with discomfort in the lower anterior region. His medical history was significant for ectodermal dysplasia, eczema and seizures prior to ectodermal dysplasia diagnosis. Medications included hydroxyzine, fexofenadine, topical steroids, cetirizine, and epi pen for allergies to eggs, peanuts, fish, mold, dust, and seasonal allergens.

Radiographic findings revealed oligodontia. The cause of discomfort at initial presentation was determined to be secondary to the eruption of primary canines. The patient returned for regular exams for 1.5 years but was subsequently lost to follow-up and was next seen at age 7 years and 11 months, when he presented with concerns about esthetics.

At that visit, the extraoral exam was significant for the presence of sparse, fine hair and dry patchy, pigmented skin. Intraoral exam revealed conically shaped teeth in abnormal positions, clinically absent primary teeth, a stable occlusion, and deficient maxillary and mandibular alveolar ridges.

Objective:

• Fabricate a removable jackscrew-activated maxillary prosthesis to restore esthetics, phonetics and function

• Allow continued growth of the orofacial complex

Adjustable Removable Prosthesis for Primary Dentition in Ectodermal Dysplasia: a Case Report

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Results





Figure 1. Maxillary (a) and mandibular (b) occlusal radiographs taken at chronologic age 2 years and 10 months demonstrate the development of #C, #H, #M, and #R,





Figure 2. (a) Panoramic radiograph taken at chronologic age 4 years and 3 months demonstrates movement distortion, deficient maxillary and mandibular alveolar ridges and numerous missing primary and permanent teeth. (b) Panoramic radiograph taken at chronologic age 7 years and 11 months demonstrates movement distortion, deficient maxillary and mandibular alveolar ridges, oligodontia, and the presence of erupted #A, #B, # *C*, #*H*, #*I*, #*J*, #*K*, #*M*. #*R*. and #*T*.



Figures 3. Intraoral photographs taken at age 7 years and 11 months demonstrate (a) irregular spacing between erupted teeth, conical shaped # C and # R, and right sided class III canine occlusion. (b) maxillary and mandibular occlusal relationships from an anterior view, deficient maxillary and mandibular alveolar ridges, and clinically absent incisors. (c)irregular spacing between erupted teeth, conical shaped # H and # M, and left sided class I canine occlusion.



Figure 4. Extraoral photos taken at age 7 years and 11 months demonstrate the presence of fine, sparse hair, thin eyebrows, glasses, dry patchy and pigmented skin, and reduced lower facial height. (a) and (c) lateral views demonstrate straight facial profile. (b) Anterior posterior view demonstrates frontal bossing, a depressed nasal bridge, low smile line and dark buccal corridors.



Figure 5. (a) Extraoral image of the patient with prosthesis at try-in. (b) Intraoral image of prosthesis at delivery. A midline jackscrew was incorporated into the acrylic appliance to allow for transverse growth of the maxilla creating a midline diastema between #8 and #9 when activated. Adams claps were placed on # B and # I for retention. (c) Illustration depicting the design of the prosthesis.



Discussion

Fabricating a removable prosthesis for children with ectodermal dysplasia is challenging due to the ongoing growth of the craniofacial complex, the aberrant position of existing teeth, and the reduced lower facial height.

Typically, three treatment options may be considered:

- Removable partial dentures
- Overdentures
- Fixed partial dentures⁵

Overdentures and fixed prostheses provide enhanced retention and are preferred when natural teeth are available for support. Compared to complete dentures, overdentures contribute to alveolar bone preservation. However, a significant drawback of overdentures is the need for extensive tooth preparation and elective endodontic treatment of otherwise healthy teeth. Additionally, one concern with fixed prostheses is that they may restrict craniofacial growth⁶.

Removable partial dentures are the most common and least invasive option but require frequent adjustments and replacement. An adjustable removable dental prosthesis enhances esthetics, phonetics, and function while accommodating growth during the transition from primary to permanent dentition. Regular dental visits are essential for monitoring growth and development, ensuring timely adjustments or replacements of the prosthesis.⁶

In the present case, a removable maxillary partial denture was selected because the erupted maxillary teeth were present in a favorable position allowing retention of the prosthesis. Additionally, a jackscrew-activated RPD would permit unrestricted growth of the maxilla. A key limitation of this case is the patient's failure to return for follow-up after appliance delivery, preventing necessary adjustments and replacement, monitoring of growth and evaluation of patient satisfaction.

Conclusions

- Early prosthetic intervention in the management of ectodermal dysplasia is important to improve esthetics and facilitate emotional development of the child.
- An adjustable appliance facilitates maxillary growth in all directions, and with regular follow-up, can allow for continued growth and development of the orofacial complex.
- Multidisciplinary care between pediatric dentistry, orthodontics, and prosthodontics is important to ensure comprehensive and timely interventions.

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References