

Dental Management of Ectodermal Dysplasia in a Pediatric Patient: A Case Report

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Background

The ectoderm is one of the three germinal layers from the early embryo which eventually forms the epidermis, hair, nails, tooth enamel, sweat glands, nerves, and other structures. The term ectodermal dysplasia (ED) is used to describe a disorder when two or more of the structures of the ectoderm have not properly formed. More than 180 types of this condition have been identified each with their own specific gene mutations. ED is transmitted as x-linked recessive, autosomal-dominant, or autosomal recessive modes. Males are most often affected, and incidence is estimated to be 1 case per 100,000 individuals. Hypohidrotic ectodermal dysplasia is the most common form.

Clinical Presentation

Clinical presentation may vary depending on the subtype of ED and variations within that specific subtype. Individuals also present with varying degrees of abnormalities.

Skin/Hair/Nails

Skin appears to be thin and is lightly pigmented in those with ED. ED patients may also have thin, sparse hair and reduced density of hair in their eyebrows and eyelashes. Hair on the scalp often appears dry and fragile. Nails may also appear thin, thick, brittle, ridged, or small.

Sweat Glands

According to sweat gland involvement, two groups are distinguished: (1) Hypohidrotic/Anhydrotic where sweat glands are completely absent or reduced in number or (2) Hydrotic where sweat glands are normal. Without sweat glands, ED patients have a much more difficult time cooling down when overheated. ED patients often experience increased frequency of fevers.

Teeth

Hypodontia is the most common oral abnormality in those with ED. Hypodontia may be mild, moderate, severe, or complete (anodontia). When teeth are absent, alveolar ridges are often under-developed due to lack of tooth bud formation. Enamel dysplasias may also occur. Tooth enamel may present as hypoplastic, hypocalcified, or hypomature. Abnormalities of the enamel may cause higher risk for dental caries. In some forms of ED, saliva production may also be reduced leading to xerostomia. Clefts may also be present in specific types of ED.

Dental Considerations

The overall goal of dental treatment for those with ED is to provide an age-appropriate dentition that facilitates proper oral/facial development, optimizes chewing function, swallowing, speech, and esthetics. Dental treatment may also elevate psychosocial, physical, and emotional development for affected patients. Comprehensive care should be obtained for these individuals, working closely with a team of specialists (pediatric dentists, orthodontists, general dentists, prosthodontists, and oral surgeons) to provide optimal care throughout the patient's life.

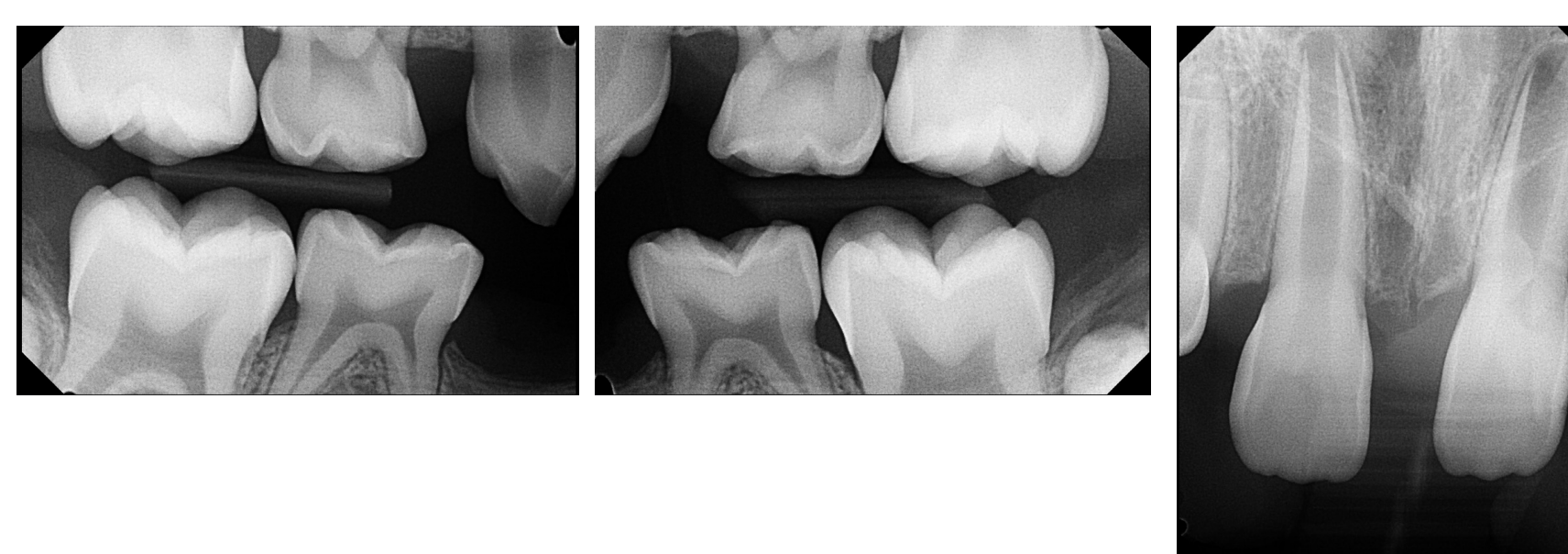
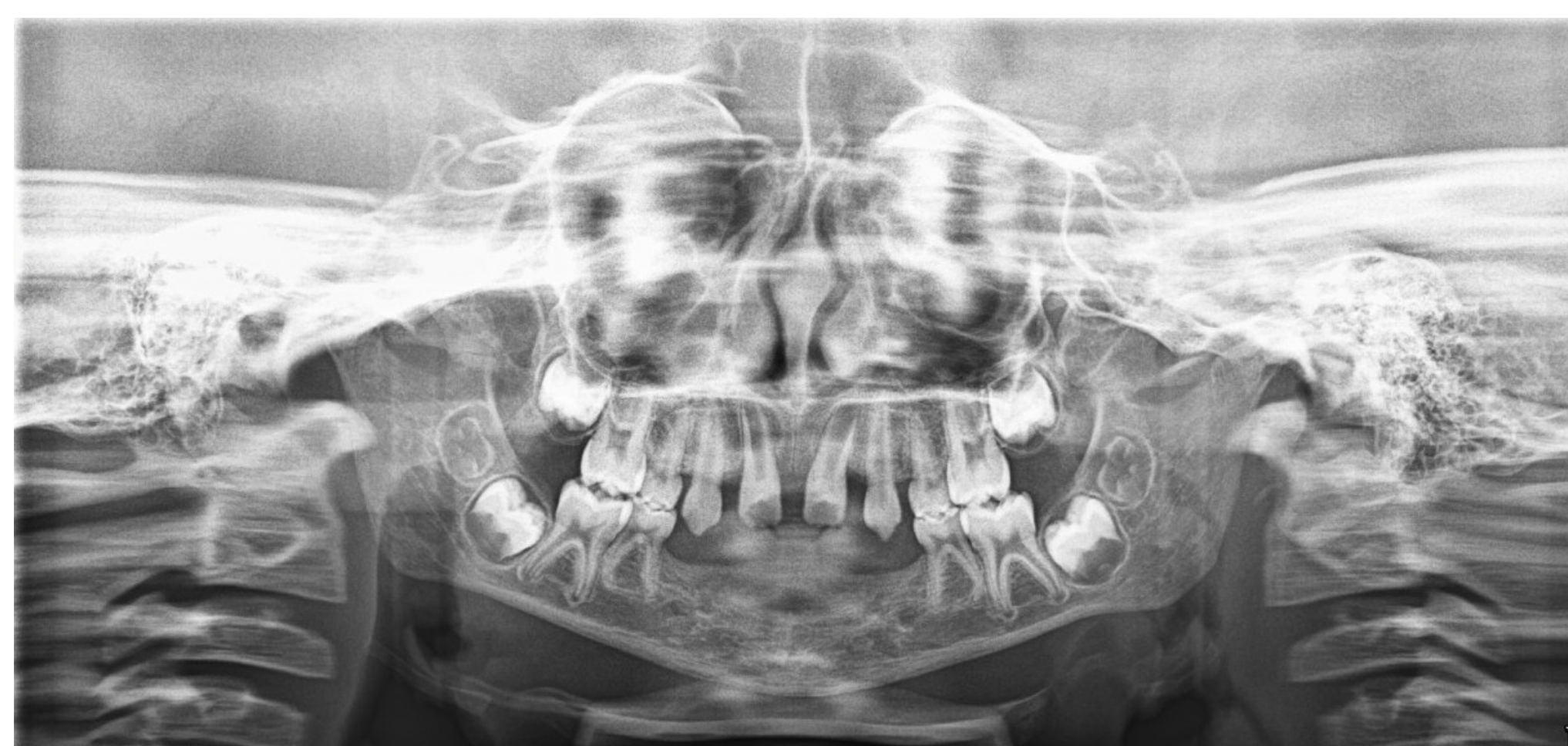
Case Description

Patient: 7y6m male presents with CC "I am missing a lot of teeth and am starting to get made fun of at school for how I chew my food."

Medical History: Patient is currently undergoing genetic testing for Ectodermal Dysplasia

Medications/ Allergies: No current medications or detectable allergies

Family/ Social History: Patient's younger sister also presents with similar clinical features but has not yet been diagnosed.



Extraoral Exam:

Sparse/thin hair on scalp, eyebrows, and eyelashes. Nails appear to be brittle. Guardian reports that patient gets hot easily. Skin lightly pigmented compared to parents.

Intraoral Exam:

Present teeth: #s 3, A, C, 8, 9, H, J, 14, 19, K, T, and 30
#s 2, 15, 17, 18, 31, and 32 are not yet developed/erupted
High lingual, maxillary labial, and mandibular labial frenum attachments

Radiographic Exam:

No radiographic decay noted
Decreased height of mandibular anterior alveolar ridge
Incomplete root development of teeth # 8 and # 9

Dental Treatment

Lingual and mandibular labial frenectomies were first completed using a CO2 diode laser with the intent to create better retention of future mandibular prostheses. The patient returned 4 weeks following treatment (once tissue healing had occurred). During the follow up visit, tissue healing had occurred, but both lingual and labial frenum attachments were still not optimal. It was determined that additional frenectomies would be necessary for optimal retention for future prostheses, but the retention gained would be sufficient for the patient's first prosthesis. Next, maxillary and mandibular impressions were taken using alginate. The partial was designed to be made in acrylic with Adam's clasps on K and T. During the delivery of the partial, it was determined that the Adam's clasps did not adapt to the teeth and provide adequate retention needed for function of the prosthesis.

The partial was returned to the lab, with a new design of C clasps as the major connectors. During the delivery of the second design, the clasps were adjusted and fully adapted to teeth without tissue impingement. Both the patient and guardian were happy with the esthetics and function of the partial.



Discussion

Lifelong dental management is necessary for those with Ectodermal Dysplasia. As the individual grows, new prostheses will need to be fabricated to fit changing oral structures and provide chewing function. Eventually, orthodontic treatment and prosthetic placement of implant-supported dentures will yield the most functional and esthetic results. Patients with ED present unique dental challenges and treatment is best approached through a multi-disciplinary methodology. Treatment may be a delayed process and patient compliance is necessary for success.

References

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Treatment Plan

This patient had no previous history of dental treatment. The treatment plan was as follows:

- Soft tissue lingual and mandibular labial frenectomies and mandibular anterior alveolar ridge gingivoplasty
- Mandibular acrylic partial denture designed with Adam's clasps as direct retainers on K & T
- Future orthodontic treatment to close maxillary diastema and create space for prosthetic lateral incisors once root development is complete of #s 8 and 9