Ectodermal Dysplasia: A Family Tree

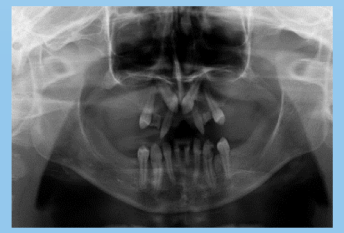
Elizabeth Potee DMD; Gerald Ferretti DDS, MS, MPH; Margaret Ferretti DMD, MPH; Ying An DDS, PhD
Department of Pediatric Dentistry, University Hospital's Rainbow Babies and Children's Hospital
Case Western Reserve University School of Dental Medicine
Cleveland, OH

Abstract

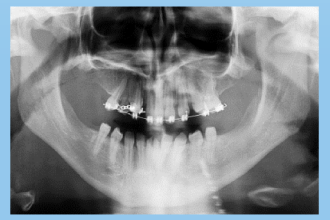
Ectodermal dysplasia (ED) consists of a large group of inherited disorders in which two or more of the ectodermally-derived structures such as the skin, sweat glands, hair, nails, and teeth develop abnormally. Especially important in pediatric dentistry are the distinguishing oral characteristics including hypodontia, hypoplastic conical teeth, and underdeveloped alveolar ridges. Due to children afflicted with ED often having low self-esteem, treatment goals should be aimed at creating a normal dental appearance. This presentation discusses three siblings with an autosomal dominant form of ectodermal dysplasia. All siblings are currently followed by the craniofacial clinic at Case Western Reserve University. This poster will evaluate and compare the panoramic films of each sibling, with a focus on the youngest sibling who has undergone dental treatment in the OR under general anesthesia for treatment of decay using esthetic restorations.

Patient and Family History

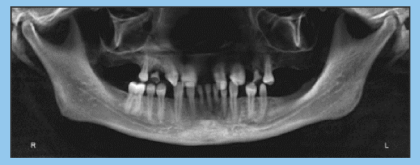
Patient is an 11 year old Caucasian male with a past medical history of familial ectodermal dysplasia, OCD, ADHD, asthma, and epitaxis. His medications include trazodone, vyvanse, seroquel, and adderall. Patient is the youngest of 3 children, all with an autosomal dominant form of ED from paternal side. All siblings are followed by the craniofacial clinic at Case Western Reserve University. The oldest sister currently wears a full maxillary overdenture. The middle sister is currently in orthodontic braces prior to partial prosthesis fabrication. The youngest brother was referred to our clinic by the craniofacial team due to extensive decay and a request for esthetic concerns to be managed with composite restorations. They included a request to not extract primary teeth C and H in order to preserve bone for eventual exposure of impacted canines.



11yo; youngest brother
22 congenitally missing permanent teeth



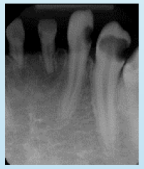
15yo; middle sister
16 congenitally missing permanent teeth

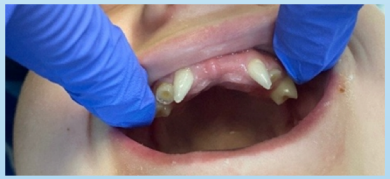


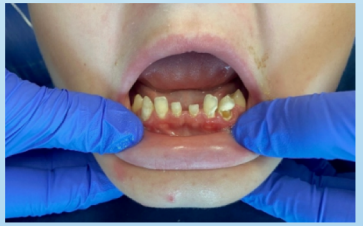
19yo; oldest sister
17 congenitally missing permanent teeth

Pre-op Presentation

Initial appointment at the resident dental clinic on June 30, 2021.
Present teeth: 5, C, 7, 10, H, 12, 21-23, O, P, 26-28
Impacted teeth: 6 and 11
Missing teeth: 1-4, 8-9, 13-20, 24-25, 29-32
Conical laterals: 7, 10
Decay: 22-MDF, 23-DLF, 26-F, 27-F, 28-B
(teeth 22 and 23 vital and asymptomatic)
Occlusion: retrognathic maxilla with extreme underbite (-3mm)





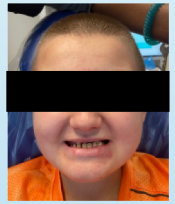


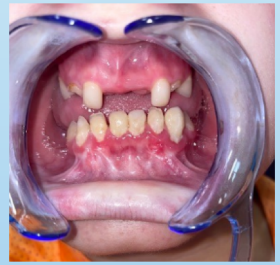
Treatment

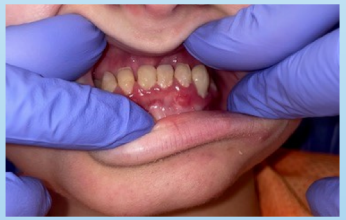
Patient was seen at Rainbow Babies and Children's Hospital in Cleveland for treatment in the OR under General Anesthesia on October 8th, 2021.
For esthetics, resin crowns were placed on teeth 7, 10, O, and P.
Due to extensive decay, direct pulp caps with Biodentine were placed on teeth 22 and 23 after disinfecting with cotton pellets soaked in chlorhexidine. A full coverage resin crown restoration placed on tooth 22. And resin composite restorations were placed on teeth: 23-DFL, 26-F, 27-F, and 28-B.

Post-op Clinical Presentation

Patient was seen at our clinic for a follow-up on December 8th, 2021; approximately 2 months after OR procedure. Patient presented with significant plaque build-up and gingivitis, signs of continued poor oral hygiene. However, patient was very pleased with the restorative esthetics and denies any sensitivity from teeth 22 and 23 that received direct pulp caps. Both teeth tested asymptomatic, vital.







Continued Care

Patient is now on a routine recall schedule at our clinic and continues to be seen regularly with the craniofacial team at CWRU. Per the craniofacial fellows, their plan is to have patient referred to OMFS for exposure of 6 and 11, extraction of C and H, and then bond lower arch in the future. As of now, mom does not think he will tolerate any type of appliances or wires due to his severe OCD.

References

About Ectodermal Dysplasias.*NationalFoundation for Ectodermal Dysplasias*.http://nfed.org/index.php/about_ed/about-ectodermal-dysplasias. PinheiroM, Freire-Maia N (November 1994). "Ectodermal dysplasias: a clinical classification and a causal review". *Am. J. Med. Genet.*53 (2): 153–62

McDonald, Ralph E., Avery, David R., Dean, Jeffery A., "Dentistry for the Child and Adolescent, 8thEdition." Philadelphia,PA, USA:2004.