



Dental Management of Persons with Ectodermal Dysplasia

Ectodermal dysplasias (ED) are a group of inherited disorders, initially classified by Freire-Maia in 1971 into 117 varieties.¹ This classification was further defined in 2014, identifying at least 186 types of ED.² While the different types vary in presentation, each is characterized by abnormal development in at least two ectodermally derived structures (hair, teeth, nails, and sweat glands). The incidence of ED worldwide is roughly 7 in 10,000 births and typically appears in the first trimester of pregnancy.

There are many modes of inheritance, the most common being X-linked recessive. As a result, the gene is more often carried by females and manifested in males.³ In-depth information concerning genetics and research related to the ectodermal dysplasias is available through the websites of the National Foundation for Ectodermal Dysplasias, the National Organization For Rare Disorders, and Geneskin.⁴⁻⁶

The defects associated with ED are not usually life threatening when managed appropriately; however, all can significantly affect the quality of life. This is particularly relevant to dentistry, as there are many oral manifestations of ED. Eruption is often affected, including tooth agenesis of primary and/or permanent dentition and incorrect eruption order or timing. In addition to missing teeth, the morphology may be affected including undersized, conical, and/or fused teeth, and taurodontism. Teeth may also be malpositioned and have abnormal spacing.⁷ These presentations can impact the development of the jaws and alveolar processes. Considering the extensive needs, genetic testing and counseling should be considered; this can help secure insurance coverage and other forms of support.

As a result, these patients often require a wide scope of dental procedures. All aspects of restorative dentistry are often required for dental rehabilitation of patients affected by ED. A team of dentists with training in pediatric dentistry, orthodontics, prosthodontics, and oral and maxillofacial surgery are often involved in establishing and carrying out a treatment plan.⁸⁻¹¹ The goals of dental treatment for individuals affected by ED syndromes are to provide an age-appropriate dentition that optimizes chewing function (and thus nutrition), oral/facial development, speech, swallowing, and esthetics.

Comprehensive treatment also aims to enhance physical, emotional, and psychosocial development for affected individuals. Items that need to be addressed in planning dental treatment include age, psychosocial environment, teeth present, oral hygiene, occlusal vertical dimension, bone volume, jaw growth and development, orthodontics and/or orthognathic surgery, implants, time required for treatment, maintenance, and cost of treatment. Since the dental manifestations of the ED syndromes persist throughout life, dentists must anticipate working closely with children, adolescents, and adults.¹²⁻¹⁴

As ED is typically diagnosed early in life, a pediatric dentist is often the first dental team member. Their role in maintaining primary dentition is often critical, as implant therapy occurs much later in life. Preservation of deciduous and permanent teeth is desirable to preserve investing bone, help support and retain removable prostheses, and possibly serve as orthodontic anchorage. Deciduous teeth may be removed as skeletal growth occurs; however, they should be preserved for as long as possible. These considerations are particularly crucial when a primary tooth is missing its successor, malposition makes maintenance difficult, or malformed enamel heightens caries risk. If orthodontics is part of a patient's



treatment plan, consideration should be given to forces, as hypodontia may put these patients at higher risk for root resorption.¹⁵ Tooth agenesis and malformation often results in diminished function and esthetic concerns, from both the patient and their families. Consequently, treatment is often initiated early. Composite restorations can improve the contour of abnormally shaped anterior teeth, further improving function and esthetics.

Interim acrylic removable partial/complete dentures are often utilized in this early phase as a minimally invasive option to improve appearance and function. While they can be effective, patient compliance with home care is critical and often challenging. Patients and providers alike must understand that these removable prostheses will require frequent relining or remaking to accommodate growth, fractured prostheses, and evolving esthetic needs. This is particularly evident as ED patients transition from primary to permanent dentition. Resultingly, a systematic review found that dentists were not satisfied with the results in as many as 30% of these cases, and prostheses had to be remade an average of 3.5-4 years.¹⁶

As growth occurs, definitive treatment can be rendered, which often involves dental implants. Multidisciplinary planning is often necessary in treatment of the mature ED patient, including consideration of orthodontics to position the permanent teeth in the best position in combination with dental implants and orthognathic surgery to correct skeletal jaw relationships.

In ED patients with anodontia, dental implants have been placed in children and pre-adolescents in the anterior mandible to support a partial or complete overdenture. Skeletal growth requires close attention to maintenance of the implant-supported prostheses, which will require repairs, relines, or remakes. In many cases, mini-implants or small diameter implants are an option, especially in children and adolescents with narrow ridges. This treatment option helps the patient with improved retention of the prosthesis, increased masticatory efficiency, and improved self-image, providing a better quality of life.¹⁷

There have been few clinical reports on the use of implants and mini-implants in children and adolescents with ectodermal dysplasia.¹⁸⁻²⁴ While there are some concerns related to growth patterns, and early implant placement should not be done routinely, there are some indications to use them in young patients with anodontia or severe oligodontia. Currently, it appears that implant placement in these cases does not affect the craniofacial growth significantly, but there might be a slight increase in implant failure.²⁵⁻²⁷

Definitive dental treatment that includes implant placement in the maxilla and posterior mandible is best carried out following completion of skeletal growth. A systematic review found success rates from 88.5-97.6% for implants placed in ED patients.²⁶ Clinical reports indicate that implants can be successful in patients with ectodermal dysplasia if bone volume is adequate for implant placement.²⁸ With that in mind, grafting procedures are often required in areas of tooth agenesis. Some evidence indicates lower implant success rates in younger patients and those that require extensive grafting.¹⁶

While many dentists and dental specialists may contribute to the treatment of ED patients with anodontia and severe hypodontia, it is the position of the American College of Prosthodontists that a

prosthodontist is often the best choice to be the lead provider in planning, organizing, and managing the definitive care.

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