Dental Management of Persons with Ectodermal Dysplasia

Ectodermal dysplasias are inherited conditions characterized by defects of the hair, teeth, nails, and sweat glands. The modes of inheritance are X-linked recessive, autosomal recessive, and autosomal dominant. The most common mode is X-linked recessive; therefore, more males are affected. "The ED syndromes are a remarkably diverse group of human disorders in which other parts of the body may be affected. 186 types of ectodermal dysplasia have been identified using the Freire-Maria classification. The current estimate of the incidence of ED is 7 in 10,000 newborns."[1] The defects associated with ED are not life threatening when managed appropriately; however, all can significantly affect the quality of life.[2]

The most prevalent form of ectodermal dysplasia is hypohidrotic ectodermal dysplasia (HED), also known as Christ-Siemens-Touraine syndrome (CST). Eruption of the teeth may be delayed, or only a few teeth may erupt in those affected by HED.[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.[1,2,4]

Due to missing, abnormally shaped, and mal-positioned teeth, a wide scope of dental procedures, including all aspects of restorative dentistry are 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.[5-8] The goals of dental treatment for individuals affected by the 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. Since the dental manifestations of the ED syndromes persist throughout life, dentists must anticipate working closely with children, adolescents, and adults.[9-11]

The literature regarding dental management of persons affected with ectodermal dysplasia includes case reports of patients treated at various ages with removable, fixed, and implant-supported prostheses.[12-15] Follow-up periods vary. Yap and Klineberg published a systematic review of the literature regarding the use of dental implants in treatment of persons with ED in 2009.[16]

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.

Parents or guardians of young persons with ED often desire to have treatment of children begin as soon as possible. Dental defects and esthetics concern families, especially when the affected child is approaching school age.[2] When possible, 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 are removed as skeletal growth occurs. Composite build-ups can improve the contour of abnormally shaped anterior teeth. With the encouragement of parents, young children can successfully wear conventional or tooth-supported complete dentures. All-acrylic interim removable partial/complete dentures can improve appearance and function. Removable prostheses require relining or remaking to accommodate growth. As growth occurs, orthodontics can place the permanent teeth in the best positions to be combined with implants in treatment of the mature patient. Orthognathic surgery may be necessary to correct skeletal jaw mal-relationships prior to initiating a definitive treatment plan.

In ED patients with anodontia, implants have been placed in children and pre-adolescents in the anterior mandible to support an overdenture or fixed hybrid prosthesis. Skeletal growth requires close attention to maintenance of the implant-supported prostheses, which will require repairs, relines, or remakes.

Definitive dental treatment that includes implant placement in the maxilla and posterior mandible is best carried out following completion of skeletal growth. Clinical reports indicate that implants can be successful in patients with ectodermal dysplasia if bone volume is adequate for implant placement.[17] However, the use of dental implants in patients with ED requires consideration of several factors, including age, motivation for seeking treatment, pattern of missing teeth, bone quantity and quality, and patient and guardian expectations. The interaction of these factors is complex and requires careful consideration for each child, adolescent, or adult presenting for treatment.

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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