Prosthetic rehabilitation of ectodermal dysplasia: A review article.

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Abstract
Ectodermal dysplasia (ED) is a congenital syndrome characterized by developmental failure of two or more ectodermal structures and their accessory appendages. ED is commonly a difficult condition to manage with prosthodontics because of the typical oral deficiencies and because the afflicted individuals are quite young when they are evaluated for treatment. It is important that these individuals receive dental treatment at an early age for physiologic and psychosocial reasons. Patient with this disease often need a multidisciplinary approach to treatment planning and dental treatment to regain appropriate function, esthetics, and comfort. The definitive treatment plan may include removable, fixed, or implant-supported prosthesis or a combination of these options. This article reviews the literature that pertains to the prosthodontic treatment of the ED and the review includes considerations in patient management and timing of treatment.

Keyword: Ectodermal dysplasia, implants, hypodontia, removable prosthesis, rehabilitation.

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Introduction

Ectodermal dysplasia (ED) is a large group of heterogeneous heritable conditions characterized by congenital defects of two or more ectodermal structures and their appendages: hair (hypotrichosis, partial or total alopecia), nails (dystrophic, hypertrophic, abnormally keratinized), teeth (enamel defect or absent) and sweat glands (hypoplastic or aplastic).[1,2] This syndrome was first reported in 1792 by Danz and Wedderburn found in inbred Indian group in 1838.Thurnam published the report of a patient with ectodermal dysplasia in 1848.[3] The term ectodermal dysplasia was not coined until 1929.[4] In 1971 only eight forms of ED were known.[1,5] Now approximately 200 different ED have been delineated; about 30 have been identified at the molecular level with identification of the causative gene.[6] These disorders are relatively rare and occur in 1:10,000 to 1:1,00,000 birth.

Classification

There are several classifications given by different authors. Some are based on clinical features and others on genetic component of the disorder.[7,8] Hidrotic and anhidrotic (hypohidrotic form) are two major types of ED. The hidrotic form, which is autosomal trait, affects teeth, hair and nails but rarely affects the sweat glands which was first described by Clouston in 1929.[9] Hypohidrotic form (Christ-Siemens-Touraine Syndrome) is most common type, X-linked recessive trait occurs with an incidence 1 to 7: 10,000 live births.[10,11] Hypodontia, hypotrichosis and hypohidrosis which form a triad are the characteristic feature of the hypohidrotic form.[12,13] Clinical signs include trichodysplasia (abnormal hair) in 91% of cases, tooth agenesis in 80%, onychondysplasia

(abnormal nails) in 75%, and dyshidrosis (abnormal sweat glands) in 42%.[2]

Genetic aspects

Hypohidrotic (or anhidrotic) ED (HED) is can be inherited in an X-linked (XL), autosomal recessive or autosomal dominant manner.[15] X-linked HED was the first in which the defective gene was cloned as a novel signaling molecule of the tumor necrosis factor (TNF) superfamily named as ectodysplasin (EDA).[16] This EDA gene was located to Xq 12-13 by Zonana et al. [17] Autosomal forms of HED are due to mutation in the EDA receptor (EDAR). EDAR binds the A1 isoform of EDA (EDA-A1) but not binds not the EDA-A2 isoform. Autosomal HED may also be caused by mutation in a cytosolic, EDAR-specific adapter molecule named EDAR-associated death domain (EDARADD).[14,15,18,19] The EDA, EDAR and EDARADD genes helps to give instruction in making proteins during development of embryo. These proteins form part of a signaling pathway that is critical for the interaction between two cell layers, the ectoderm and the mesoderm. It forms many structures that arises from ectoderm, the skin, hair, nails teeth and sweat glands. Mutation of those genes prevents normal interaction between mesoderm the ectoderm which impairs the development of hair, sweat glands and the teeth.[7,8] The inappropriate formation of these ectodermal structures helps to form the characteristic features of HED.

Dental Abnormalities

The most striking oral feature of ED may range from hypodontia to anodontia of the primary or permanent teeth (with or without cleft lip and cleft palate), associated with hypoplasia of the alveolar bone structure. Consequently, the reduction of the vertical dimension of the lower face; the vermilion border disappears which results in in protuberance of lips.[21,22] Vierucci and co-workers have shown hypohidrotic type ectodermal dysplasia children have maxillary retrusion due to sagitally under developed maxilla, forward and upward displacement of the mandible and collapsed lower anterior facial height.[23] It is not uncommon for the face of an affected child taking appearance characteristics of old age.[24] The maxillary central incisors, maxillary first molars, and maxillary canines are teeth most often present which teeth are present are often conical, malformed and widely spaced.[25] True anodontia cases were extremely rare conditions. In such cases, multidisciplinary team is generally advocated to be the most appropriate approach.[20,26] Treatment decisions of the dental team depend on the patient's remaining teeth condition, age, needs, wishes, their willingness to undergo minor or major treatment with different impact, and also on the economic possibilities. This review article has written about the prosthodontic treatment of ED, including the patient management and timing of treatment.

Rehabilitation protocol for ED patients

Nowak[28] stated that "treating the children with ED requires the clinician's knowledge in growth and development, management of behaviour, techniques in prosthesis fabrication, the ability to motivate the patient and their parents for use of the prosthesis, and the long-term follow-up for the any type of modification and/or replacement of prosthesis." There are Various types of prostheses have been used, including overdenture, complete or partial removable prostheses, or fixed prostheses, implants.[27] After taking proper history, clinical & radiographical examination of the individuals we have to consider the treatment plan accordingly.

The use of conventional prostheses before placing implants is encouraged; since, it

helps to condition the growing patient and provides esthetic and functional information for the subsequent implant treatment plan.[29] According to Nowak229] a series of introductory visits may be needed before treatment commences, to attain the required patient trust.

Available treatment strategies

Removable prosthodontics

For the dental management of ED (Fig 1).[35] Removable prosthesis is the most frequently reported treatment modality. In childhood a complete denture, removable partial denture (RPD) or overdenture is often the treatment of choice because it is easily modify the intraoral prosthesis during rapid growth periods. These treatment options affordable for the ED patient and his or her family and it is also a reversible method of oral rehabilitation. Cooperation of the patient as well as the support of the family is necessary if removable prosthesis is to be successful in young patients.[30] Other problems related to removable prosthesis are speech difficulties, dietary limitations, and loss of the prosthesis.[28]

Overdentures

Overdentures are amenable to long- term maintenance and allow progressive changes to be made to the prosthesis. Implant - supported overdentures are also the best treatment option (Fig 2,5d,5e).The advantages of preserving the natural teeth are (i) Preservation of alveolar bone, (ii) Preservation of proprioception, (iii) Improved retention (iv) Improved support, and above all (v) Less psychological trauma of loosing natural teeth for patients.[31].Patients treated with complete maxillary overdenture and mandibular overdenture demonstrates less vertical alveolar bone reduction than patients with conventional complete maxillary and mandibular dentures.[32] Periodic recall of young ED patients is also important because prosthesis modification or replacement will needed as a result of continuing growth and development.[33,34] In addition to adjustments related to fit, the occlusion of prosthesis must be monitored for changes because of jaw growth.[34]

Fixed prosthodontics

Fixed prosthodontic treatment is seldom used exclusively in the treatment of ED, primarily because many afflicted individuals have a minimal number of teeth (Fig 4b). In additions, ED patients are quite young when they are first treated, and fixed partial dentures (FPDs) with rigid connectors should not give in young and growing patients. This is because rigid FPDs could interfere with jaw growth especially if the prosthesis crosses the midlines.[35] Hogeboom[35] presented a case that dramatically demonstrated the occurrence of a detachable fixed prosthesis separated at midline in two segments because of transverse jaw growth in an ED individual which was treated by fixed prosthesis. Individual crown restorations have no age restrictions related to jaw growth, but larger pulp sizes and shorter crown height may cause concern.[36] Crowns and direct composite restorations are often used in combinations with RPDs in the prosthodontic management of these patients.[37-40]

Implant prosthodontics

The literature indicates that endosteal implants were being used more widely in the prosthodontic management of ED (Fig 6). [29,41] Ekstrand and Thomsson [42] , Bergendal et al.[41] , Smith et al.[43] and Cronin et al.[44] have also reported situations in which endosteal implants were successfully used in ED patients. A number of studies indicate an improvement in the psychologic and physiologic function of adult patients with an implant supported denture when compared

with their condition before implants were placed in an edentulous control group with complete dentures.[45-48] Tallgren[49] reported that the annual mean crestal bone loss, in denture wearers, in the anterior mandible is 0.4 mm. If the prosthetic rehabilitation consists of an implant-retained overdenture, then the annual bone loss is 0.1 mm or 0.5 mm over 5 years.[50]

It has been recommended that treatment with implants must be delayed until the age of 13; since an implant which was placed at age 7 or 8 may not be in a favorable position at age 16.[29] The survival of dental implants after 10 years varies between 82% and 94%.[51] Meta-analysis estimate survival rates of 86.7% for implant-borne fixed dental prostheses (FDPs), 77.8% for tooth-implant-borne FDPs, and 89.4% for implant-borne single crowns.[52] Ledermann et al.,[53] In their 7-year follow-up with a mean length of 35.5 months reported a 90% success rate in 9-18 years age group with 42 endosseous dental implants placed in 34 patients. Brugnolo et al.[54] noted the infraocclusion of implants placed in patients aged 13-14. It is recommended to wait before planning to place an implant for the completion of skeletal and dental growth; though, many physiologic and psychological factors create pressure to commence for earlier treatment.[55] Odman et al. [56] concluded that implant should not be placed in children "until the permanent dentition is fully erupted.'' A very few study favor implant placement in children because from a physiologic stand point, the conservation of bone may be the most important reason for the use of dental implants in growing patients.[57] Other factors that favor implant placement in children are their excellent local blood supply, positive immunobiologic resistance, and uncomplicated osseous healing.[53] Placement of endosteal implants is not suggested during the time of maxillary growth, determined as being up to 15 years of age. Isolated treatment of young HED patients using a combination of conventional endosseous implants with specially designed zygomatic implants has also been presented with satisfactory results. Thilander et al.[58] longitudinally followed 27 single-crown restored implants placed in 15 adolescents whose ages ranged from 13 to 19 years. Even though in most restorations minor infraocclusion did occur, they concluded that implants are acceptable for that age group when growth and development is complete, with all fully erupted teeth. Cronin et al. [44] provide a description of mandibular jaw growth and the possible consequences of early implant placement in the mandible. They concluded that implants should placed girls after age 15 years for and boys after 18 years provide most predictable prognosis.

According to Cronin et al. [44] possible consequences of early implant placement include implant submergence because of jaw growth, implant exposure because of bone resorption associated with jaw growth, implant moves due to jaw growth, and jaw growth is limited if the implants are connected by a rigid prosthesis which crosses the midline. For implant placement in young ED patients, their dental and skeletal maturity has to be considered, their chronological age should not be the determining factor. Growth curve of an individual can help in this determination. It is clear from the literature that if implants are placed in young ED patients the timing of treatment is important because of possible complications resulting from jaw growth. Individuals affected by ED face a lifetime of special needs, which may include dentures at a young age with frequent adjustments and replacements; proper special diets to meet dental/nutritional needs; air-conditioned environments; wigs to conceal hair and scalp conditions; carrier-identification testing; protective devices from direct sunlight; osseointegrated dental implants; and respiratory and speech therapy.

Conclusion

This review article provides the literature that is concern to the prosthodontic rehabilitation of the ED and this includes the considerations in patient management and treatment timing. ED patients with different age groups, the prosthodontic approaches may vary. The literature has demonstrated the benefits that corrective prosthodontic therapy at the appropriate time aids the patient in developing proper mastication, deglutition, speech and esthetics which may have dramatic psychological and physical benefits for these patients. These early dental interventions can improve the patient's appearance and psychological problems as long as close follow up and prosthetic modification are maintained due to possible continuing growth.

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Figures

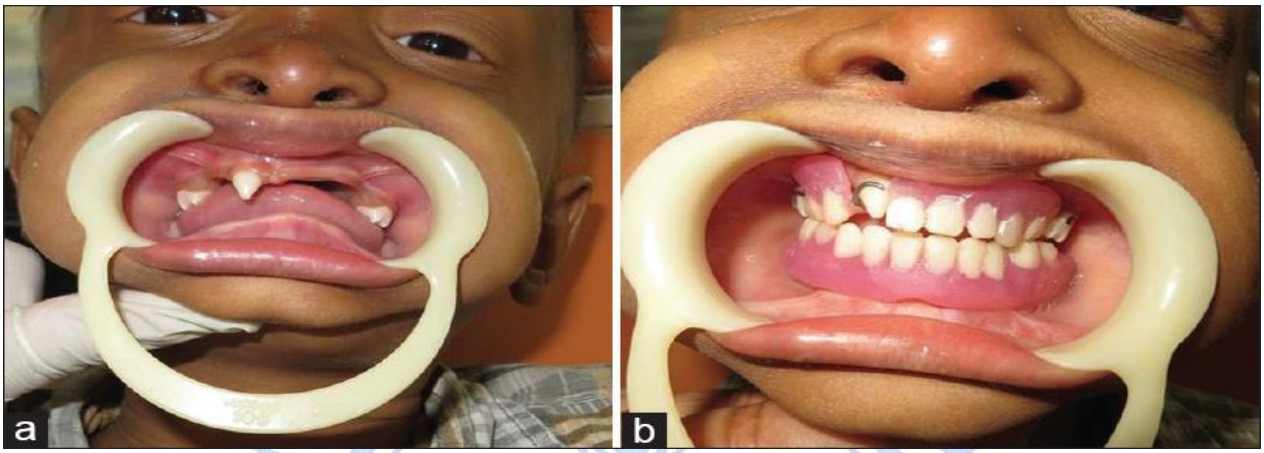

Figure 1-(a) Preoperative, (b) with removable partial dentures

Figure 2-(a)preoperative showing complete anodontia,(b) implants placed in mandible,(c)with maxillary complete denture and mandibular overdenture

Figure 3- (a)Intra-oral view showing conical incisors and underdeveloped mandibular alveolar ridge. (b) Intra-oral mandibular occlusal view (c) intra-oral maxillary occlusal view

Figure 4- (a)Metal try-in for maxillary fixed partial prosthesis. (b) Completed maxillary prosthesis in place, try-in of primary copings in mandibular arch. (c) Metal framework with secondary copings on cast.

Figure 5-(a) Metal framework with occlusal rim for jaw relation recording (b) metal framework with porcelain application on secondary copings (c) anterior try-in (d) final prosthesis occlusal view (e) final prosthesis intaglio view

Figure 6-(a)Patient's occlusion before treatment (b)Definitive cement retained restorations in place(c)preoperative panoramic radiograph (d) postoperative panoramic radiograph of osseointigrated implants.