

A First Look: Determinants of Dental Care for Ectodermal Dysplasia Patients.

¹Dr. Aruna Kanaparthi, BDS, MDS, ²Dr. Rosaiah Kanaparthi, BDS, MDS

¹Assistant Professor Department of Operative Dentistry, College of Dentistry, Jizan University, Jizan, Saudi Arabia

²Assistant Professor Dept of periodontics, College of Dentistry, Jizan University, Jizan, Saudi Arabia

Abstract: Ectodermal Dysplasia (ED) is a hereditary disorder characterized by abnormal development of certain tissues and structures of ectodermal origin. The most frequently reported ED syndrome is X-linked hypohidrotic dysplasia, also known as Christ-Siemens-Touraine syndrome, which affects one to seven individuals per 10,000 live births. Orofacial characteristics of this syndrome include anodontia or hypodontia, hypoplastic conical teeth, underdevelopment of the alveolar ridges, frontal bossing, depressed nasal bridge, protuberant lips, and hypotrichosis. The characteristics associated with ED will often result in afflicted individuals having an abnormal appearance. Normal social and psychologic development of young ED patients dictates that they look and feel as normal as possible. Also, a “tell-show-do” approach to treatment is recommended for the young ED patient. The multi-disciplinary treatment can involve composite restorations, fixed, removable, or implant prosthodontics, singly or in combination.

Key-words: Ectodermal dysplasia, hypodontia, hypotrichosis, anodontia, hypohidrotic

I. Introduction

Freire-Maia defined the nosologic group of ED as any syndrome that exhibits at least two of the following features:

- (1) trichodysplasia (abnormal hair),
- (2) abnormal dentition,
- (3) onchodysplasia (abnormal nails), and
- (4) dyshidrosis (abnormal or missing sweat glands).

Certain oral and facial characteristics may also be associated with the syndrome. Because there are more than 120 different ED syndromes, clinical manifestations depend on the specific syndrome afflicting an individual.

Therefore, dental appearance in these patients is extremely important because it can affect their self-esteem. The national foundation for ectodermal dysplasia (nfed) defines ectodermal dysplasia (ed) as a genetic disorder in which there are congenital birth defects abnormalities of 2 or more ectodermal structures. these structures may include skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear, and parts of other organs. The nfed lists 20 common types of the disorders. severity differs, even among people affected with the same type of ed.

The diagnosis of ED can be difficult because of the variety of types, range of abnormalities, and severity of defects singularly and collectively. It is important to identify the diagnostic components of the disorder so that appropriate treatment can be rendered to ensure the best quality of life for ED patients. It is also important to understand the genetic hereditary patterns so that the parents of the affected child can be counseled and better predict the chances that future offspring will be affected. Defective genes cause Ectodermal Dysplasia; these genes can be inherited from one or both parents or manifested through gene mutation.

Any structure derived from the ectoderm can be defective in ED. Each type of ED involves different structures and the severity of the disorder varies from patient to patient. In general, the skin of affected children is lightly pigmented and appears thin and almost transparent; surface blood vessels are easily visible. Pigmentation is heaviest around the eyes (usually wrinkled) and on the elbows, palms, and soles, with the latter 2 areas hyperkeratotic in nature. The skin is usually dry, scaly, and easily irritated as a result of poorly developed or absent oil (sebaceous) glands.^[1,2,3]

Prosthodontic treatment of ED

- Fixed,
- Removable,
- Implant-supported prostheses.

These treatment approaches can be used individually or in combination to provide an optimal result.

II. Patient Management Considerations

There is little information in the literature other than clinical reports regarding dental management of the young ED patient. An article by Nowak provides the most complete review to date on the dental management of these young patients. Till and Marques also provide some insights into the subject and are cited in this review. Nowak stated that “treating the pediatric patient with ED requires the clinician to be knowledgeable in growth and development, behavioral management, techniques in the fabrication of a prosthesis, the modification of existing teeth utilizing composite resins, the ability to motivate the patient and parent in the use of the prosthesis, and the long-term follow-up for the modification and/or replacement of the prosthesis. If the treating dentist is not knowledgeable in one or more of these areas he should obtain consultations or refer treatment when needed. A multidisciplinary team composed of a pediatric dentist, a prosthodontist, an orthodontist, and an oral and maxillofacial surgeon has been advocated in some reports and recommended to ensure proper treatment of young ED patients.

There is not a definitive time to begin treatment, but Till and Marques recommend that an initial prosthesis be delivered before the child begins school so that the child has a normal appearance and time to adapt to the prosthesis. Ultimately, the decision to begin treatment should be made by the treating dentist along with the parents and patient. Because individuals with ED are quite young when they are evaluated for treatment, the treating dentist should have some knowledge and ability in the behavioral management of pediatric patients. Nussbaum and Carrel advocate sedation for managing problem children who need extensive prosthodontic treatment. Nowak does not recommend sedation, stating that it will result in prosthesis failure because ultimate success depends on patient understanding and compliance.

Instead of sedation, Nowak advocates conditioning to the pending dental procedures by a “tell-show-do” approach. Other authors have also successfully used this conditioning approach. According to Nowak, a series of introductory visits may be needed before treatment commences, to attain the required patient trust. Parents may facilitate or impede the success of this process, and their involvement must be assessed on an individual basis. When treatment commences, it is important that the patient understands each procedure and its outcome. In addition, the child and parents should have a reasonable idea of what the final prosthesis will look like, how it will benefit the patient, how to care for it, and any limitations it will have. Clinical case studies by Sarnat et al and Nomura et al have indicated that jaw growth in individuals with ED is within normal limits. Although these studies are convincing, each study involved only one subject. Studies with larger sample sizes are needed before any definitive conclusions can be made.^[4,5]

III. Treatment Modalities

Fixed partial dentures (FPD)

Fixed prosthodontic treatment is seldom used exclusively in the treatment of ED, primarily because many afflicted individuals have a minimal number of teeth. In addition, ED patients are often quite young when they are first treated, and fixed partial dentures with rigid connectors should be avoided in actively growing patients. This is because rigid fixed partial dentures could interfere with jaw growth, especially if it crosses the midline. Hogeboom presented a case that dramatically demonstrated the occurrence of jaw growth in an individual treated for ED in which two segments of a detachable fixed prosthesis separated at the midline because of transverse jaw growth.

Individual crowns

Individual crown restorations have no age restrictions related to jaw growth, but larger pulp sizes and shorter crown heights may cause concern. Most cases require intentional root canal therapy prior to crown placement. In spite of these concerns, crowns are often used in the treatment of young ED patients.

Direct composite restorations, veneers

Recently direct composite restorations have become the more desirable method of restoring normal form, function and esthetics to hypoplastic teeth commonly found in ED patients.

Removable Prosthodontics

Removable prosthodontics is the most frequently reported treatment modality for the dental management of ED. Because anodontia or hypodontia is typical in individuals with this condition, complete dentures, partial dentures, or overdentures are often part of the treatment provided.

Removable partial dentures (RPD)

Crowns and direct composite restorations are often used in combination with removable partial dentures (RPDs) in the prosthodontic management of these patients. Composite restorations are usually necessary to provide proper contours on the hypoplastic teeth that will be used as abutments for RPDs.

Complete dentures

Although complete dentures can provide an acceptable esthetic and functional result, underdevelopment of the edentulous alveolar ridges in individuals with ED can compromise denture retention and stability.^[6]

Overdentures

When there are teeth present for support, overdentures are a desirable treatment option for these patients. Crum provided an excellent overview of the advantages of conventional overdentures as opposed to complete dentures. One important advantage is that overdentures preserve alveolar bone. As for accommodation by young ED patients to complete dentures, published cases usually report good adaptation to the prostheses. To facilitate accommodation in young children, some clinicians have reported the delivery of one denture at a time. An important factor in patient acceptance is the appearance of the denture teeth in relation to the patient's age. The dentition stage depicted in the denture should be appropriate. This is accomplished by using primary prosthetic teeth for the very young patients and making modifications to existing or new dentures as the patient grows older. Periodic recalls of young ED patients are also important because prosthesis modification or replacement will be needed as a result of continuing growth and development. In addition to adjustments related to fit, the occlusion of the prosthesis must be monitored for changes because of jaw growth.

Other problems related to removable prostheses are speech difficulties, dietary limitations, and loss of the prosthesis.^[7,8,9,10]

Implant Prosthodontics

The literature indicates that endosteal implants are being used more widely in the prosthodontic management of ED. Guckes et al. have reported preliminarily a 90% dental implant success rate at second-stage surgery in ED patients 13 to 69 years old, as part of an ongoing clinical trial at the National Institutes of Health.

Ekstrand and Thomsson, Bergendal et al, Smith et al. and Cronin et al. have also reported situations in which endosteal implants were successfully used in the prosthodontic management of ED.

A number of studies indicate an improvement in the physiologic and psychosocial function of adult patients with an implant-supported denture when compared to their condition before implants were placed or to an edentulous control group with complete dentures.

Kent provides a review of the effects of dental implants on psychologic and social well-being. He concludes that "there is a consistent and clinically significant pattern of results" to report the claim that dental implants can positively affect the well-being and quality of life. As already noted, prosthodontic treatment is extremely important in ED patients for physiologic and psychosocial reasons. As a result, implant-supported prostheses seem to be a desirable treatment option for these patients

Another reason to consider dental implants in the treatment of ED is the beneficial impact they could possibly have on the preservation of alveolar bone. As previously mentioned, the alveolar ridges of individuals with ED are underdeveloped because of the lack of tooth development. These alveolar ridges must support a dental prosthesis over the course of a lifetime.

Thus, treatment measures that will maintain alveolar bone and enhance the prognosis for future prosthodontic treatment are extremely important, especially in individuals with total anodontia. A 1989 Consensus Conference on Implantology concluded that implants should not be placed until maximum jaw growth has occurred, which was reported as up to 15 years of age. In an editorial, Lekholm concurs with criteria concerning maximum jaw growth, giving age guidelines of 14 to 15 years of age for girls and a year later for boys. He also recommends that an individual's growth curve be studied before any implant placement procedure is started

Even though minor infraocclusion did occur in most of the restorations, implants are acceptable for that age group provided that growth and development is complete, with all teeth fully erupted. The authors stress that the dental and skeletal maturity, and not the chronological age of the patient is important to avoid infraocclusion of an implant restoration. It is clear from the literature that the timing of treatment is important if implants are placed in young ED patients because of possible complications resulting from jaw growth.^[11,12,13,14]

IV. Summary

The prosthodontic management of ED requires a broad knowledge base to handle the special problems associated with treatment. For this reason, a multidisciplinary team approach is recommended for optimal dental management of the condition. Although it is important to provide early treatment, it must be remembered that any prosthesis made for a young patient must be closely monitored for needed adjustments or for a replacement prosthesis made necessary by growth and development. A 6- to 12-month recall schedule until skeletal growth is complete is advised.

Regarding fixed prosthodontics for dental management of ED, FPDs should be avoided in young, actively growing patients because they could interfere with jaw growth. This is especially true if the prostheses crosses the midline.

As for the hypoplastic teeth are common with ED, direct composites or crowns are often needed to restore proper contours to the teeth.

Removable prosthodontics is the most frequent treatment modality used for dental management of ED. Though complete dentures are an acceptable form of treatment, overdentures or RPDs supported by natural teeth are desirable for preservation of alveolar bone.

When removable prostheses are fabricated for ED patients, the dentition stage depicted should be appropriate.

In addition, it may be necessary to fabricate and deliver one denture at a time in young patients with no denture experience, to facilitate accommodation.

Implant-supported restorations can improve physiologic and psychosocial function when compared with complete dentures, but their placement in growing jaws can cause complications. When implant placement in young ED patients is being considered, their dental and skeletal maturity, not their chronological age, should be the determining factor. An individual's growth curve can help in this determination.

For dental implants to become a more desirable and recommended treatment option for young ED patients, more research is needed to determine guidelines for dental implant placement in children and the effectiveness of the concept of preventive implantation with dental implants in maintaining alveolar bone.

V. Conclusion

Young children with anodontia caused by Hypohidrotic Ectodermal Dysplasia (HED) not only have difficulties in eating and speaking but can also sense that their appearance is different than others. Enabling children with HED to look and act more like their peers through the use of well-fitting and functioning dentures with age-appropriate denture teeth will greatly assist in their transitioning into the school years.

Although denture fabrication requires multiple patient appointments and good cooperation, it is shown that even young children can co-operate for the denture-making process. The desire to be like others who have teeth can be a motivator for cooperation in even the young child. Children should be given every opportunity to develop to their fullest potential. A multi-disciplinary team of dentists can make a significant contribution to the overall development and well-being of a child with ED.

References

- [1]. Pinheiro M, Freire-Maia N Ectodermal dysplasias: a clinical classification and a casual review. *Am J Med Genet* 1994;53:153-62
- [2]. Lowry RB, Robinson GC, Miller JR. Hereditary Ectodermal Dysplasia : symptoms, inheritance patterns, differential diagnosis and management. *Clin Pediatr* 1966;5:395-402.
- [3]. Blattner RI. Hereditary Ectodermal Dysplasia. *J Pediatr* 1968;73:444-47.
- [4]. Nowak AJ. Dental treatment for patients with Ectodermal Dysplasia. *Birth Defects* 1988;24:24.3-52.
- [5]. Pigno MA, Blackman RB, Cronin RI Jr, Cavazos E. Prosthodontic management of Ectodermal Dysplasia: a review of the literature. *J Prosthet Dent* 1996;76:541-45.
- [6]. Ramos V, Giebink DL, Fisher JG, Christensen C. Complete dentures for a child with hypohidrotic Ectodermal 4. Dysplasia: a clinical report. *J Prosthet Dent* 1995;74:329- 31.
- [7]. Bonilla ED, Guerra L, Luna O. Overdenture prosthesis for oral rehabilitation of hypohidrotic Ectodermal Dysplasia: a case report. *Quintessence Int* 1997;28:657-65.
- [8]. Deshraj Jain, Sandhya Jain, Alok Kumar, Tripty Rahangdale. Hypohidrotic Endodermal Dysplasia: Prosthetic and Endodontic Management. *International Journal Of Clinical Pediatric Dentistry*, Jan-April 2010;3,(1):63-67
- [9]. Shigli A, Reddy R.P.V, Hugar S.M, Deshpande D. Hypohidrotic Endodermal Dysplasia: A unique approach to esthetic and prosthetic management: A case report, *J Indian Soc Pedo Prev Dent*, Mar 2005, 31-34
- [10]. Akshay Bhargava, Arun Sharma, Sachet Popli, Renu Bhargava, Prosthodontic Management of a Child with Ectodermal Dysplasia: A Case Report *J Indian Prosthodont Soc* 2010 10(2):137-40
- [11]. Guckes AD, Brahim IS, McCarthy GR, Rudy SF, Cooper LI. Using endosseous dental implants for patients with Ectodermal Dysplasia. *J Am Dent Assoc* 1991;122:59-62.
- [12]. Guckes AD, Scurria MS, King TS, McCarthy GR, Brahim IS. Prospective clinical trial of dental implants in persons with Ectodermal Dysplasia. *J Prosthet Dent* 2002;88:21-25
- [13]. Cronin RI, Oesterle LI, Ranly DM. Mandibular implants and the growing patient. *Intl Oral Maxillofacial Implants* 1994;9:55-62.
- [14]. Lekholm U. The use of osseointegrated implants in growing jaws. *Int J Oral Maxillofac Implants* 1993;8:243-44.