

## Ectodermal Dysplasia: Dental Management and Complete Denture Therapy

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**Abstract:** Ectodermal dysplasia is a genetic disorder in which there are congenital birth abnormalities of 2 or more ectodermal structures. The management of ectodermal dysplasia is complicated due to the craniofacial dysmorphology, wide array of dento-facial defects and because the individuals are quite young when they are evaluated for treatment. This article reviews the various dental treatment options for ectodermal dysplasia patients and presents a detailed report describing the prosthodontic management with complete denture therapy, suitably modified for the young patient.

**Key words:** Hypohidrotic ectodermal dysplasia • Anodontia • Complete denture • Prosthodontics

### INTRODUCTION

Ectodermal dysplasia (ED) is a congenital syndrome characterized mainly by tissue abnormalities of ectodermal origin, namely skin, nails, hair and teeth [1]. They constitute a wide spectrum of congenital diseases that was first described by Thurman [2]. The etiology of ectodermal dysplasia lies in genetic anomalies, which can be inherited through either parents or manifested via gene mutations [3]. It displays an autosomal dominant, autosomal recessive or X-linked pattern of inheritance [4, 5].

ED can be classified into hypohidrotic, in which the sweat glands are absent or significantly decreased; and hidrotic, in which the sweat glands are normal [6]. Hypohidrotic ectodermal dysplasia (HED, also called as Christ-Siemens-Touraine syndrome), [2, 7] is the more severe form of the disease and is associated with sensitivity to heat, frequent high fevers and dento-facial defects. It is the most frequently reported ED variant with an X-linked recessive inheritance gene mapped to Xq12-q13. [4, 5] HED affects men more severely and frequently, whereas female heterozygotes present minor defects [2].

Most patients exhibit marked facial asymmetry which include frontal bossing, depressed nasal bridge, reduced

vertical facial height and depth, small palatal and cranial base widths, small malar processes and high-set orbits [1]. Severe maxillary hypodontia in the permanent dentition may cause cranio-facial asymmetry [2]. Additionally, atrophy of the alveolar process in dentulous and edentulous region may cause reduction in the lower facial height [1, 8]. These features give the child a distinctly aged facial appearance similar to an edentulous patient [1]. HED patients usually exhibit slight mental impairment, have a senile appearance and are usually socially isolated [9].

As hypohidrotic ectodermal dysplasia presents the most dental anomalies, it is of considerable interest to the dental community [1]. Its diagnosis is based on characteristic symptoms, that include hypodontia (80%), [6, 10] anodontia or oligodontia (reduced number of teeth) with conically shaped teeth, hypohidrosis (decreased perspiration) with concomitant hyperthermia caused by a lack of sweat glands and hypotrichosis (sparse hair) [9]. Most of the teeth that develop are often conical shaped, complicating its use as abutments for removable partial dentures; however, they may serve as abutments for overdentures in some situations [2, 11, 12]. Reduced salivary secretion, dry oral mucosa and hoarse voice quality have also been reported in these patients [1].

This article reviews the various dental treatment options available for the HED patient and describes the prosthodontic treatment with complete denture therapy, suitably modified for the young patient.

**Dental Management of Ectodermal Dysplasia:** The dental management of the ED patient requires clinical knowledge of growth and development, behavior management, pedodontics, prosthodontics, orthodontics and oral surgery [7, 13]. Since ED patients usually present themselves at a very young age with a multitude of anomalies, a multidisciplinary team approach is required. The initiation of dental treatment should be decided by the dentist along with the parents and patient [7]. Clinicians have suggested that the child patient must have an initial prosthesis before he/she begins school, so as to adapt and get used to the prosthesis [7, 14].

Behavioral management techniques like “tell-show-do” play an important role in conditioning young patients to conquer their fear, anxiety and establish a trustworthy relationship with them [7, 15, 16]. Uncooperative young patients may need sedation for extensive prosthodontic procedures; however, Nowak stated that sedation compromises the patient’s understanding and compliance, which are essential for the success of a prosthesis based treatment [7, 13]. Parents play an important role in the rehabilitation of ED patients by understanding the treatment process and teaching the child how to adapt and use the prosthesis effectively [7, 16].

The oral rehabilitation of ED patients usually consists of complete or removable prosthesis in the development years, followed by a definitive prosthesis based on fixed partial dentures and/or endosseous implants, after the complete development of the alveolar process [7].

Removable prosthodontics is a common mode of treatment in ED as such patients are often associated with anodontia or hypodontia [7]. Congenital missing teeth, coupled with tooth loss due to caries or trauma are other situations that demand the need for removable prosthodontic treatment in young ED children [2]. The treatment of such patients would comprise of a complete denture, removable partial denture, overdenture or a combination of the above [7]. Complete denture treatment provides satisfactory functional and esthetic rehabilitation of the patient, but may not be adequately retentive in severely hypoplastic ridges [7]. In such situations, vestibuloplasty and ridge augmentation may enhance the hard and soft tissue support [7, 13, 17].

The advantages of overdentures are well documented and may be used successfully in cases of hypodontia and enamel hypoplasia [7, 18, 19]. Overdentures make use of natural undercuts and precision attachments for improved retention and stability [7, 19]. They also help preserving the residual alveolar ridge for future anchorage of dental implants [7, 19].

Fixed prosthodontic treatment may be initiated for older patients but as most of the ED patients are quiet young, it should be avoided [7]. A rigidly connected fixed partial denture, crossing the midline, may interfere and restrict the normal growth of the alveolar process [7]. Single crown restorations should also be avoided for the younger patient as they could cause exposure of immature and incomplete pulp during tooth preparation [7]. Alternatively, direct composite can be used to develop proper occlusal contacts, contours and normal morphology of hypoplastic teeth, which could later serve as abutments for removable partial dentures [7, 19].

Misaligned teeth may require orthodontic treatment to be positioned favorable prior to removable or fixed partial denture treatment [7]. Functional appliances may be advised for patients with increased overjet, overbite and posterior teeth infraocclusion [1, 20]. Occlusal plane therapy can be initiated when posterior tooth eruption is needed prior to prosthodontic treatment [1].

Dental implants are increasingly being used in the management of ED, [6, 21]. However, concern has arisen regarding the placement of osseointegrated implants in developing alveolar bone [22]. Implants placed in young ED patients may get submerged due to the continuously growing alveolar process and investing tissues [6]. The growth of the adjacent natural teeth relative to the implant supported prosthesis may cause infra-occlusion over a period of time, which would require frequent remakes to correct the plane of occlusion or distraction osteogenesis for repositioning the submerged implant [6, 23]. The submerged implants may be predisposed to peri-implantitis and the inadequate crown/implant length ratios may contribute to increased horizontal forces [23]. In case of severe bone atrophy, implant placement may not be possible without bone grafting [3]. Consequently, clinicians have contraindicated dental implants for children upto 6 yrs of age [9]. Dental implant treatment may be considered in situations of anodontia as a method of preserving the residual alveolar ridge [7, 24-27]. and in patients above 13 yrs of age as most of the alveolar bone growth would have been completed by then [7, 28].

### **Complete Denture Therapy in Ectodermal Dysplasia:**

The 7-year old male patient exhibited the classical features of HED, characterized by hypohidrosis, hypotrichosis and hypodontia [13]. In addition to these signs and symptoms, the patient presented with dry skin, sparse eyebrows and hair. The chin and supraorbital ridges were prominent. Frontal bossing, saddle nose, hyperkeratosis of the palms and soles of the feet were present. Hypertonicity of the peri-oral and masticatory muscles caused speech and masticatory difficulties. The vertical dimension of the lower face was reduced and the lips were protuberant, leading to the characteristic senile appearance.

The patient's history, clinical and radiographic examination revealed the complete absence of the primary and permanent dentition. Intraoral examination revealed a shallow palate, macroglossia and slightly dry oral mucosa. The maxillary ridge was depressed posteriorly (Order VI) and the mandibular ridge was of the knife edge variety (Order IV) [29].

Oral rehabilitation with complete dentures was decided for the patient to improve both the maxilla-mandibular relationship, as well as to provide improvements in esthetics, speech and masticatory efficiency. Although routine procedures for construction of complete dentures were used, case specific modifications were made and described below.

### **Treatment Procedure:**

- The maxillary primary impression was made using polyvinyl siloxane putty impression material (STD Putty, 3M ESPE) supported on an adhesive (Tray adhesive, 3M ESPE) coated stock metal tray.
- Due to the lack of adequate sized stock metal tray, the mandibular primary impression was made by supporting polyvinyl siloxane putty on an 18 gauge orthodontic wire, which was pre-adapted to the shape of the mandibular arch.
- Acrylic custom trays (MP Sai, Mumbai) with wax spacers were fabricated in accordance to the selective pressure impression technique.
- Single step border molding was done using polyvinyl siloxane putty impression material (Express XT putty soft, 3M ESPE). Conventional border molding using low fusing compound was avoided for better patient compliance, avoidance of discomfort, irritation and injury to the mucosa.

- Final impressions were made using light body-injection type polyvinyl siloxane impression material (Express XT light body fast, 3M ESPE) and master casts were poured.
- Temporary acrylic record bases were fabricated on the master casts and wax rims were fabricated. The maxillo-mandibular relationship was recorded conventionally, after assessing the phonetics and esthetics using Silverman's closet speaking space technique [30].
- A deciduous teeth mold (Nissin, Japan) was selected after calculating the inter-canine distance and teeth were arranged using the lingualized occlusion scheme.
- The try-in appointment was carried out conventionally and after the approval by the parents and patient, the waxed-up dentures were processed in heat-polymerizing acrylic resin (Lucitone, Dentsply).
- A laboratory remount was done to correct the changes in the vertical dimension after processing and to ensure the presence of a centric occlusion.
- On the subsequent appointment, the dentures were inserted placed and proper fit was verified. Recall appointments were scheduled for 24 hours, 1 week and 3 weeks.

### **DISCUSSION**

The primary treatment of ED patients with total anodontia is usually by complete dentures and/or dental implants. This poses certain controversial questions, namely, the right age for initiation of prosthetic treatment, residual ridge resorption (RRR) associated with complete dentures [29, 31] and the gradual submerging of dental implants [6] due to continuous skeletal and alveolar bone growth in young patients.

Clinicians have initiated prosthetic treatment for ED patients at the age of 5 yrs stating that it ensures functional, phonational, psychologic and esthetic rehabilitation of the child [6]. Initiating prosthodontic treatment at an early age enhances masticatory muscles tonicity, delays alveolar bone resorption associated with the absence of teeth, compensates for the decrease in vertical dimension and prevents angular cheilitis [22]. Early complete dentures treatment can lead to significant improvements in mastication, appearance, speech and satisfactory diet for the child. It instills self-confidence in the child, which is essential for normal psychological and

social development [9]. Failure to initiate complete denture therapy in young ED patients may cause reduction in the height of the lower third of the face, upward and forward displacement of the chin due to antero-rotation of the mandible and a predilection to Class III malocclusion [2, 32].

The young ED patients differ from geriatric patients in that they are edentulous due to agenesis and are in an active skeletal growth phase [22]. Therefore, RRR may not be as severe in such patients and they may respond positively to the functional and psychological benefits of the prosthesis [22]. Early treatment with a removable appliance can result in a significant growth stimulation of the alveolar processes under the pressure of the interim prosthesis and can provide a better foundation for future implant placement.

Cooperation and compliance to the new prosthesis are some of the main disadvantages of early complete denture therapy in young ED patients [2]. It is therefore necessary that parents take an active part in the treatment phase and ensure proper use of the prosthesis [2]. The dryness of the oral mucosa, immature maxillary tuberosities and alveolar ridges cause difficulty in obtaining adequate retention and stability of the complete denture [32]. This can be counteracted by the impression technique, occlusal scheme, ensuring a broad distribution of occlusal load and complete extension of the denture base [2, 33].

Special attention was paid to the impression technique. Polyvinyl siloxane impression material was used due its biocompatibility, pleasant odor and handling characteristics making it comfortable for the young patient [33]. It is also highly elastic, fast setting and hydrophobic, which aids in making the impression of the dry mucosa [33]. The prosthetic teeth used were modified in width and height to reduce eccentric forces directed to the compromised ridges that can cause alveolar resorption. The cuspal inclines were reduced to a non-anatomic form as it was difficult to obtain an accurate centric relation record. This also ensures further reduction of the detrimental lateral forces during eccentric movement. Lingualized occlusion scheme was used with freedom of movement in the intercusp contact area, to provide centralization of forces and to prevent lateral interferences [2].

Periodic recall and maintenance appointments in young ED patients are important as prosthesis modification or replacement will be needed due to continuing skeletal growth and development [7, 13, 17, 34,

35]. As the ED child matures, the complete denture will have to be relined, rebased, or remade to accommodate change in vertical dimension, loss of occlusion and abnormal mandibular posture to maintain speech, chewing, mastication and swallowing functions [2, 3, 19]. Vergo TJ recommended relining or rebasing prostheses every 2 to 4 years and remaking them every 4 to 6 years [19]. He also stated that failure to do so may result in hyperplasia or epulis formation [19]. The patient was advised to remove the dentures at night to offset the daily stresses placed upon the oral tissues. At recall appointments, pressure areas were identified using pressure indicating paste and relieved accordingly. Retention of the maxillary denture was good and the patient gradually adapted to the lower denture. The parents reported a significant improvement in the child's speech, self-esteem and socialization skills.

Further future treatment will include modification of the dentures by relining or replacement of the dentures according to the alveolar growth and eventually, placement of endosseous implants for additional support of the denture.

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