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Case Report
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# Early Prosthetic Treatment of Young Children with Ectodermal Dysplasia: Two Case Reports

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## Abstract

Ectodermal dysplasia (ED) is an inherited disorder characterized by the abnormal development of ectodermally derived anatomic structures, with features such as onchodysplasia, trichodysplasia, dyshidrosis, and oligodontia. Hypoplasia of the alveolar ridges is also associated and removable prosthodontics is the most common treatment modality for dental management. Oral rehabilitation is recommended for the positive development of self-esteem as well as for improved speech, masticatory function and facial esthetics. The purpose of this article is to present early prosthetic rehabilitation of two toddlers or very young patients with ED, aiming to contribute to enhance the current literature, considering that it does not provide much information on this topic.

## Introduction

Ectodermal dysplasia (ED) is an inherited disorder characterized by the abnormal development of ectodermally derived anatomic structures [1] and was defined by Freire-Maia [2,3] as any syndrome with at least two of the following features: onchodysplasia, trichodysplasia, dyshidrosis, anodontia or oligodontia. Midface hypoplasia is frequently observed along with other orofacial characteristics including hypoplastic conical teeth, underdevelopment of the alveolus, a pronounced supraorbital ridge and frontal bossing, depressed nasal bridge, and protuberant everted lips due to a decreased facial vertical dimension [4-7]. Patients with ED have also demonstrated hypoplasia and aplasia of major salivary glands therefore may exhibit varying degrees of xerostomia [8,9].

With over 190 different types of ED syndromes reported [10], clinical presentation depends on the specific type afflicting the patient. The two major types are: hypohidrotic, where the sweat glands are absent or significantly decreased, and hidrotic, in which the sweat glands are normal. The most frequently reported ED syndrome is the X-linked hypohidrotic ED [11], also known as Christ-Siemens-Touraine syndrome [12] or Clouston [13], with a reported incidence of 1/10,000 births [14]. Hypohidrotic ED can also be associated with sensitivity to heat, frequent high fevers, and is associated with more dental defects [15].

Hypodontia of the primary and permanent dentition and associated hypoplasia of the alveolar bone with poorly formed alveolar ridges are both commonly occurring [4,16]. Removable prosthodontics is the most common treatment modality for the dental management of ED [17] often providing an acceptable esthetic and functional result; however, hypoplasia of the edentulous alveolar ridges can significantly compromise retention and stability of a removable prosthesis. When teeth are present, they are often conical in shape, making them less than ideal as removable partial denture abutments. These teeth can however be used as an option for abutments with conventional complete overdentures [18-20], whose advantage of preserving bone makes it the desired treatment for the young ED patient[21-28]. Several studies have reported the successful use of endosteal implants for the treatment of ED [29-40]. Implants could preserve alveolar bone and improve the psychological well-being of the patient, however caution should be used in placing implants in children since they could become malpositioned or displaced if placed during active growth. Several clinical reports have demonstrated the use of dental implants to rehabilitated patients with ED, however few prospective clinical trials of implants placed in the young ED patient have been published.

The appearance of patients with ED can affect their social and psychological development, therefore oral rehabilitation is recommended for the positive self-esteem development. Additionally, improved speech, masticatory function and facial esthetics contribute to normal dietary habits and

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**Figure 1:** A and B) Full facial and profile views of the patient, illustrating the deficient proportion of the lower third of the face.

**Figure 2:** Intraoral examination revealed severe hypodontia with only the primary mandibular second molars present.

social development. While individuals with ED report more issues with chewing, swallowing, dry mouth and speech, orofacial function has not been studied systematically [41]. Oral-motor exercises (OME) can be incorporated for patients who have feeding and swallowing problems [42] with the goal of aiding in adjusting to prostheses. OMEs include stretching, massage, vibration, and passive range of motion exercises with the assistance of a caregiver. These procedures are applied to provide sensory input and it has been theorized these techniques normalize feeding patterns by reducing abnormal oral reflexes, facilitating normal muscle tone, or desensitizing the oral region [43]. Although clinical evidence is not yet available for these principles, reasonable rationale for their use has been provided [44-46]. The addendum at the end of the article provides desensitization instructions and steps to follow regarding OMEs.

## Aim

The purpose of this article is to present early prosthetic rehabilitation of two young patients with ED. Informed consent was obtained from the parents of the participating subjects.

## Case Presentation

### Patient 1

A 3-year, 8-month-old female (Caucasian was referred to the Department of Pediatric Dentistry, University of Florida in Gainesville, FL for evaluation and treatment. She presented with classical features of hypohidrotic ED including diffusely sparse hair, hypohidrosis, onchodysplasia, oligodontia, and impaired hearing. The facial profile showed a depressed nasal bridge, prominent forehead, and the lower third of the face was deficient, consistent with decreased vertical dimension of occlusion (Figure 1A and 1B). Intraoral examination revealed macroglossia, minimal keratinized tissue, adequate salivary flow, oligodontia, underdeveloped alveolar ridge, shallow vestibular space and flat palatal vault (Figure 2). Radiographic examination confirmed clinical findings and revealed only primary mandibular second molars with conical contours and permanent mandibular first molars were present bilaterally. Severe hypotonicity of the peri-oral and masticatory muscles was apparent with reported difficulty in speech and mastication. The mother also presented with similar oligodontia, reporting she only had her mandibular first molars present and had been wearing a complete maxillary denture and mandibular removable partial denture since the age of eight.

In view of the patient's age, good behavior, and with the mother's understanding of the syndrome, a maxillary complete denture and a mandibular complete overdenture were determined to be the treatment of choice. Preliminary impressions were made with modeling plastic impression compound (Red Cake, Kerr Corporation; Orange, CA). Custom trays were fabricated with light-polymerized acrylic resin (Triad TruTray, Dentsply; York, PA) and definitive border molding and impressions were made using polyvinylsiloxane impression material (Aquasil, Dentsply Caulk; Milford, DE). Maxillomandibular relation records were made using wax occlusion rims and the casts mounted on a semi-adjustable articulator. Prosthetic deciduous denture teeth (Bambino, Pearson Lab; Sylmar, CA) were adjusted and arranged for an age appropriate appearance. The vertical dimension of occlusion, lingualized-balanced occlusal wax trial denture and the positions of the prosthetic teeth were evaluated intraorally, and

**Figure 3:** A and B) Wax trial denture with verified tooth position and corrected vertical dimension of occlusion.

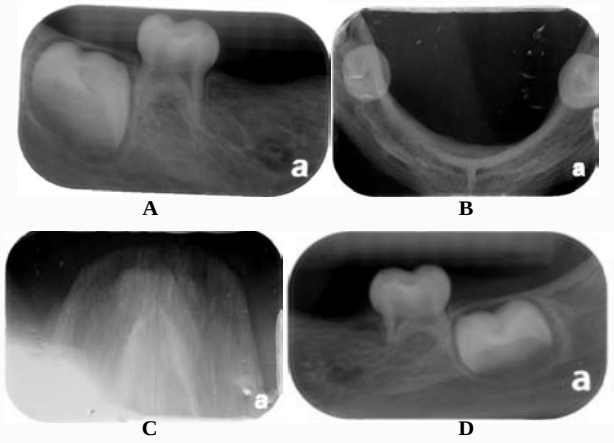



**Figure 4:** (A) Mandibular left periapical (B) Mandibular right periapical (C) Mandible (D) Maxilla.

adjustments were made at this time (Figure 3). The dentures were then processed with heat-activated acrylic resin (Lucitone 199 Original, Dentsply; York, PA).

Upon insertion of the dentures, the vertical dimension of occlusion and facial profile proportions were restored to normal for the patient's age. Specific instructions were given to the parent to wear the dentures only a few hours a day initially to facilitate accommodation to the prosthesis, as well to maintain a soft diet for the first month. Instructions were given the parents were instructed on proper oral hygiene to prevent decay of abutment teeth. Teeth were to be brushed twice daily as well as routine removal of the prosthesis after each meal and at night. The patient was asked to return to clinic the next day, after one week and after 2 months for inspection and adjustments of the prosthesis (Figure 4). Oral motor function therapy instructions were given and asked to be performed twice daily. Upon recalls, the dentures appeared clean and well maintained with excellent oral hygiene. The accommodation to both the maxillary complete denture and mandibular complete overdenture were favorable and rapid as the patient was wearing the dentures daily with reported improved masticatory efficiency. Initially the patient was placed on three-month recalls and was encouraged to return if any discomfort was reported. The three month recalls were maintained as a standard for better follow up to evaluate growth and development and confirm ideal functioning of the dentures in re-establishing the oral motor function.

### Patient 2

A 3-year, 5-month-old male patient African American was referred to the Department of Pediatric Dentistry, University of Florida in Gainesville, FL for evaluation and treatment. This patient, as compared to the first case, presented with an increased expression of hypohidotic ED including onchodysplasia, hypotrichosis, dyshidrosis, and hypodontia. He had a pronounced supraorbital ridge and frontal bossing, a depressed nasal ridge, large protuberant lips (Figure 5) as well as very dry and scaly skin. Intraoral examination revealed macroglossia, minimal keratinized tissue, slightly dry oral mucosa, minimal to no vestibular depth, underdeveloped alveolar ridges, flat palatal vault, and severe hypodontia (only primary maxillary central incisors present). Radiographic examination confirmed the clinical findings with only primary central incisors and no permanent teeth present (Figure 6).

**Figure 5:** Facial view of patient with depressed nasal ridge, dry scaly skin, large protuberant lips, and evidence of a decreased occlusal vertical dimension.

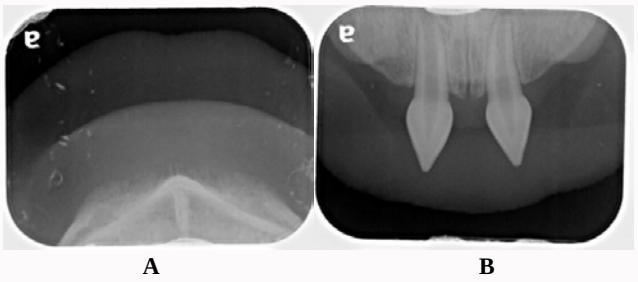



**Figure 6:** (A) Mandible (B) Maxilla Radiograph reveals only the primary maxillary central incisors were present, note the sharp incisal edges and abnormal "spade-like" contours.

The patient did have a more developed orofacial musculature and was able to speak intelligible words compared to the first case, possibly due to the absence of a hearing impairment. The mother was also afflicted with ED but to a lesser extent, as well as the patient's sister and two cousins on his mother's side, all of who had different degrees of hypodontia. Genetic counseling was recommended to determine a more definitive inheritance pattern. The patient was very young for removable dentures, but since the cousins were successfully restored with removable partial dentures, the patient and mother were extremely motivated to have a maxillary complete overdenture and a mandibular complete denture fabricated. Prior to starting the impression procedures, enameloplasty of the primary maxillary incisors was completed to create a more dome-like contour. A maxillary complete overdenture and mandibular complete denture were fabricated as previously described and delivered. Due to his large protuberant lips, the buccal flange and borders of both dentures were thinned to a thickness of 1.5 mm to lessen the lip support. The patient did have a minor gag reflex during insertion initially, but the mother reported this subsided after a few days of wearing the prosthesis (Figure 7). The mother reported only minor problems and the patient adapted quickly, with improvements in speech, esthetics, and mastication, as well as his social behavior recalls were established every three months to follow up on dentures as a rehabilitator of the oral motor function as well to evaluate the need for replacement of dentures due to growth and development of the arches.

## Clinical Update

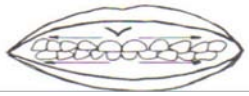
Implant supported prosthesis is an excellent option for patients who do not have any teeth in the jaw, but does have enough bone to support implants. Particularly in patients who do not have mandibular teeth, implant-supported dentures provide a more stable

**Figure 7:** (A) Full facial view immediately after oral rehabilitation, illustrating restored vertical dimension of occlusion and harmonious facial height. (B) Four months post-insertion of prosthesis, patient's self-esteem and confidence is much improved.

**Table 1:** Instructions for desensitization.

| |
|---|
| Always use a gentle but firm stroking movement. |
| Discontinue use with any aversive response to stimuli and regress to previous step. |
| Talk about what you are doing in calm manner and give praise for tolerating a step. |
| Do not rush through steps; desensitization can take time, varying individually. |
| Always start with step 1 at each session, even if early steps are well tolerated. |
| Exercises should be done 2-3 times every day. |

**Table 2:** Instructions for desensitization.

| | |
|---|---|
| Using both hands, start in the middle of the forehead and stroke out to the top of the ear. Starting at the cheekbone, stroke firmly down to the center of the chin. |     |
| Stroke from one side of the lower lips to the other. Repeat on the upper lip, starting from the center to corners of the mouth, repeat 4-5 times. |     |
| Divide inside of the mouth into 4 parts. Place index finger on the gingiva and work from front to back one way, not rubbing continuously back and forth, be careful not to touch frenum. Repeat 10 times one way, and then continue onto the other areas of the mouth in the same way. |  |

option since regular dentures tend to be less stable there (Table 1). Implants in children are typically not recommended due to the continuing development of the mandible and maxilla up to pubescent age. However, there are published studies to support the idea that implant supported dentures are a stable option for children with ectodermal dysplasia.

A prospective study conducted is Smith et al. [31] and Mittal et al. [45] assessed the feasibility of placing endosseous implants in children and adolescents with ectodermal dysplasia and to assess the position and stability of such implants during growth. A total of 41 implants were placed (patient age range 6-11) with an average follow up of 7.8 years. Of the 41 implants, 40 were successfully integrated and were restored, with only one becoming submerged because of adjacent alveolar development (Table 2). These results suggest that endosseous implants can be successfully placed and can provide support for prosthetic restoration in children with ectodermal dysplasia.

## Summary

Toddlers or young patients afflicted with ED have a marked challenge with the development of speech, poor esthetics, underdeveloped orofacial musculature, poor masticatory function and affected social and psychological development. Prosthetic oral rehabilitation seems highly recommended for these patients as early as 3 years of age to stimulate a normal oral functional development. However the fit needs to be evaluated frequently and prostheses should be prefabricated annually to account for growth. Oral rehabilitation of ED patients with removable prostheses at such a young age can prepare them for more definitive prosthetic treatment once growth is completed. The ability of the young patient to adapt to removable denture prostheses is rapid and the benefits of improved speech, esthetics, self-esteem, psychosocial development and overall function are evident.

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