Prosthetic Rehabilitation of Ectodermal Dysplasia Patient with Semi-stretched Polyamide Base-material

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Abstract

Ectodermal dysplasia is a hereditary disorder that affects one or more of the ectoderm originated tissues. Sparse hair, dry skin, lack of sweat glands, normal mental status can be observed. As a Dental findings hypodontia, anodontia, conical or malformed teeth are characterized with inadequate alveolar crest. In present case we described the prosthetic rehabilitation with semi-stretched polyamide-base material and follow-up of an ED patient.

Eight year old male patient with ED was referred to the Department of Pediatric Dentistry, Marmara University Faculty of Dentistry. The patient had oligodontia, the conical-shaped canines and incisors according to clinical and radiographic oral examinations. Dental prostheses were planned to restore function and esthetics. Semi-stretched polyamide based denture material (deflex) were applied to the upper and lower removable partial dentures, due to the flexibility and elasticity of material and reduction of the stresses on the erupted teeth. At 4th month follow up, necessary adjustment of the prostheses were done after incisor teeth eruptions. The patient continued to follow up at 2 years. Oral rehabilitation, function and aesthetics as positive psychological effects were observed in the patient.

Keywords: Ectodermal dysplasia; Hypodontia; Prosthetic rehabilitation

Introduction

Ectodermal dysplasia (ED) comprises a large heterogeneous group of come into disorders that are characterized by primary defects during the development of two or more tissues that are derived from embryonic ectoderm. Hair, skin, nails and teeth are primarily involved tissues [1]. These disorders are relatively rare with an incidence of 1 in 10,000 to 1 in 100,000 live births [2]. More than 170 different subtypes of ectodermal dysplasia have been identified [3].

There are 2 major types of this condition depending on the number and functionality of the sweat glands: [4] hidrotic or hypohidrotic. Hypohidrotic types have been defined sweat glands are either absent or significantly reduced in number (Christ-Siemens-Touraine syndrome), and [5] hidrotic types where sweat glands are normal and the condition is come into as autosomal dominant (Clouston’s syndrome) [4-6].

Factors such as patient’s age, stage of growth in conjunction with the missing teeth, soft tissue defects, existence of malformed dentition, and psychological status must be considered in the treatment planning [7]. Since patients with ED have psychosocial issues due to the orofacial manifestations presenting at such young age therefore restoring appearance and function is more challenging than usual. There are multiple treatment options for this condition, but the most frequent prosthetic treatment of ED in young patients is removable prosthetics [5].

Growing ED patients have been traditionally treated with conventional prostheses or conservative restorative modalities [8-14]. Problems associated with anatomic and morphologic abnormalities of existing teeth and alveolar ridges, however, result in poor retention and stability of removable prostheses [15,16]. Progressive alveolar bone resorption (since the edentulous ridge is loaded at an early age), periodontal complications, or increased caries rates secondary to using a removable prosthesis further compromise the prosthetic outcome and lead to prosthetic management challenges as the patient grows [15,16]. In addition, restraints of the removable design of the appliance on the lifestyle of the young patients and the associated lack of compliance with the wear of a conventional prosthesis creates parental concerns, which lead to the request for a more stable prosthesis [9].

Several factors such as the minimal number and the conical shape of the existing teeth, the patient’s young age, and the fact that a fixed prosthesis will interfere with the mandible’s growth if the midline is crossed contraindicate the fabrication of fixed partial dentures in this growing population. Single crown restorations could be made, but the large dental pulps and short clinical crowns typical of the pediatric patient may be a problem [17].

Case Report

An 8 year old boy with chief complaints of inability to masticate and unesthetic appearance was referred to Paediatric Dentistry Department of Marmara University, Dental Faculty, Istanbul, Turkey. He was diagnosed as Ectodermal Dysplasia with history of lack of sweating, dryness of skin, and raised body temperature. Extra oral
examination revealed typical findings as sparse hair, frontal bossing, depressed nasal bridge, prominent supra orbital ridges, sunken cheeks, hyperpigmented skin around the eyes, protuberant lips, and decreased lower facial height (Figure 1).

Intraoral examination revealed dry oral mucosa probably due to inadequate salivary secretion. Cone-shaped teeth were present as $52,53,62,63,73$, and $83$ with underdeveloped edentulous mandibular alveolar ridge (Figure 2). Panoramic radiograph showed that $11,13,21,23,33$, and $43$ were present and unerupted (Figure 2).

Maxillary and mandibular partial dentures were planned to provide immediate aesthetic results, re-establish the occlusion by replacing missing teeth. Semi-streched polyamide based denture material (deflex) were applied to the upper and lower removable partial dentures, due to the flexibility and elasticity of material and reduction of the stresses on the erupted teeth (Figure 3). At $4^{th}$ month follow-up, necessary adjustment of the prostheses was done after incisor teeth were erupted.

When patient come $12^{th}$ month later as follow-up, eruptions of tooth $11$ and $21$ were seen as complete. And crown restorations for conical teeth were planned in order to improve the esthetics and to aid for retention of the prostheses (Figure 4).

Heavy silicone (for better handling) was used to take some preliminary impressions and make a personalized tray and later an alginate impression was taken under more favourable conditions. It was considered appropriate to make a lower and upper acrylic partial prosthesis (Figure 5) using steel retainers on some of the teeth present in the arcade.

This design allows modifications to be made as and when necessary, as well as providing a reasonable esthetic result, an acceptable masticatory function on complementing the lack of dentition, at the same time as attempting to maintain the alveolar ridges free of atrophy, and allowing an almost normal social life, which is so important at this stage in a child's personal and psychological development.
Discussion

In a growing child, the rehabilitation of the health issues is a real challenge in paediatrics as well as in pediatric dentistry. Since up to 154 different types and 11 subgroups were cited in the dental literature for ED cases, every child patient must be considered as a unique case in order to overcome the oral discomfort of the ED patient [18,19]. As a main choice for treatment, prosthodontic intervention in an early age may help the child to adjust with the prosthesis and develop normal appearance, speech, mastication, and swallowing as well as temporomandibular joint function. Apart from dental benefits, an early age intervention may also provide psychosocial benefits. But the intraoral prosthesis in childhood needs to be followed and modified cautiously in 3 to 6 months’ time intervals according to the growth spurts or rapid growth periods [20].

In ED child-patients, reduced alveolar bone height with “knife-edge” morphology and limited remaining tooth structure make prosthodontic rehabilitation a challenging task. The replacement of teeth by implants is usually restricted to patients with completed craniofacial growth and is probably best to hold off until adolescence. Implants placed in ED patients younger than 18 years have a higher risk of failure [21].

Disadvantages of this treatment option could be the additional cost of the surgery and the implant parts, the extra time for treatment plan completion, and the patient’s young age, which might potentially complicate the surgical procedure. Children with EDs do not have normal patterns of growth and a risk and benefit analysis must be made to access the value of implant placement. Replacement of teeth by implants is usually restricted to patients with completed craniofacial growth [22].

Flexible dentures have got various advantages over the traditional rigid denture bases [23]. I was reported that the translucency of the material picked up underlying tissue tones, making it almost impossible to detect in the
mouth. As considering the material stronger and flexible better biocompatibility was achieved because the material was free of monomer and metal, these being the principle causes of allergic reactions in conventional denture materials. Also flexible dentures may not cause sore spots due to negative reaction to acrylic resins and absorb small amounts of water to make the denture more soft tissue compatible. And finally there was no requirement of tooth preparation. In our case, implant therapy was not the treatment choice due to ongoing growth and development and insufficient alveolar bone support.

**Conclusion**

In this case, the patient was very comfortable with partial dentures after 2 years of use. Good retention was observed, and the parents reported a significant improvement in terms of speech and mastication. Oral rehabilitation, function and aesthetics as positive psychological effects were observed in the patient. The patient is under our observation to monitor oral hygiene and to make adjustments and modifications to the prosthesis.

**References**


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