

Rehabilitation of A Patient with Ectodermal Dysplasia: An Unconventional, Systematic Approach

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ABSTRACT

Background The prosthetic rehabilitation of an adolescent patient with ectodermal dysplasia constitutes an amalgamation of thorough treatment planning, behavior management and restoration of the compromised intra-oral status, while preserving the health of the dentition and associated structures. The unconventional approach utilized in this treatment protocol included a trial wax-up splint guided by digital smile designing and an esthetic removable clip-on prosthesis as a transitional-continuous prosthesis until the patient reaches her skeletal maturity for the fabrication of a definitive prosthesis. This removable prosthesis has several advantages such as increased comfort, esthetics, maintenance of proprioception, minimal tissue coverage, conservation of tooth and supporting oral structures and cost effectiveness.

Keywords: Ectodermal dysplasia, removable prosthesis, esthetic clip-on prosthesis, oligodontia, adolescent.

INTRODUCTION

Rehabilitation of a young patient presenting with partial or complete anodontia is one of the greatest challenges encountered by a restorative prosthodontist. The practitioner faces several hurdles beyond the limitations of dental treatment. These include several factors related to the psychological status of the patient such as low self esteem, lack of confidence, social withdrawal and emotional instability in the most vulnerable stages of life. Therefore, it is the duty of a clinician to not only reestablish function and esthetics, but to also empower children who are crippled by their existing abnormality.

Ectodermal dysplasia is one such rare, congenital disorder characterized by abnormal development of tissues and structures of ectodermal origin.^[1] The ectodermal germ layer gives rise to organs including the nose, eyes, hair, nails, sweat glands and enamel.^[2] Individuals with this disease

generally exhibit a triad of symptoms: hypodontia, hypotrichosis, and hypohidrosis.^[3]

Oral manifestations range from anodontia to hypodontia; affecting both, the deciduous and permanent dentitions.^[4] The existing teeth are predominantly small, tapered, conical or peg-shaped.^[5] The alveolar process is often poorly developed and knife edged, decreasing the occlusal vertical dimension (OVD); hence giving an illusion of an aged appearance, similar to edentulism. However, the overall growth and development of the child, is normal.^[6]

CASE REPORT

A 14 year old girl presented to the department of Prosthodontics with a chief complaint of difficulty with mastication and poor esthetics. The patient displayed thin, scanty hair with stunted hair growth and slightly protuberant lips due to a loss of vertical dimension (Fig. 1). Additionally, she had a low smile line with less than 50% visibility of the maxillary

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Fig 1: Pre:treatment extra:oral view.



Fig 2: Pretreatment smile.



Fig 3a: Pre:treatment intraoral view, maxillary arch showing widely spaced conical teeth.



Fig 3b: Pre:treatment intra:oral view : mandibular arch showing partial anodontia, widely spaced conical teeth and a knife:edged ridge.



Fig 4: Mounted maxillo:mandibular relation at the restored vertical dimension of 3mm.



Fig 5: Esthetic wax:up on the trial thermoplastic splint (according to the smile design proportions).



Fig 6: Intraoral trial of waxedup splint.



Fig 7: Shade selection using a classic Vita shade tab.



Fig 8: Autopolymerising tooth-mold acrylic resin adapted to definitive thermoplastic splint.



Fig 9: Autopolymerising pink acrylic adapted to gingival areas.



Fig 10: Clip-on removable prosthesis – frontal view.



Fig 11: Clip-on removable prosthesis – intaglio surface.



Fig 12: Post-treatment smile.



Fig 13: Post-treatment extra-oral view.

central incisors on maximal smiling (Fig. 2). On intra-oral examination, the patient exhibited classic features associated with ectodermal dysplasia, including hypodontia, microdontia, widely spaced conical teeth and underdeveloped maxilla-mandibular alveolar ridges (Fig. 3a, 3b). The patient gave a history of occurrence of periodic dermal eruptions on the extremities and corneal erosions once in 3 to 5 years.

Both intra-oral and extra-oral photographs were taken according to the digital smile designing protocol and transferred to the software for the same. This digital approach allowed an esthetic analysis and determination of the final prosthesis.

Diagnostic casts were made, duplicated and mounted on a semi-adjustable articulator using a face-bow transfer at the restored vertical dimension of occlusion (Fig. 4).

A splint made from thermoforming material (Erkodur), 2mm in thickness was adapted to the existing models using vacuum suction (Erkopress). A diagnostic wax-up (Fig. 5) was performed on the adapted splint according to the proportions decided by the digital smile designing software (Exocad). The posterior occlusion was created using a mutually protected, group function occlusal scheme.

The trial of the waxed up splint was done in the patient's mouth (Fig. 6) to assess the restored vertical dimension using esthetics and phonetic parameters. The centric occlusion was verified along with elimination of interferences during excursive movements, followed by shade selection of the teeth (Fig. 7) and gingiva using a classic Vita shade tab and Ivoclar gingival tab respectively.

The modified trial was sent for final fabrication of the removable clip-on prosthesis. Thermoform splints, 2mm in thickness were adapted to the duplicated set of casts and microblasted to facilitate micromechanical retention of the overlying acrylic resin. Autopolymerising tooth-mold acrylic (Ivoclar, Vivadent; A2 shade) was injection molded onto the thermoform splint using the putty index as a guide (Fig. 8). This was followed by the addition of autopolymerising pink acrylic (Ivoclar, Vivadent; G4 shade) in the gingival areas (Fig. 9). The prosthesis was finished and polished (Fig. 10, Fig. 11) with

alpine stones of various grits, silicone polishing points, sandpaper and buffing with a polishing cake.

The occlusion of the final prosthesis was reconfirmed, minor chair-side modifications made and delivered to the patient; hence restoring function and esthetics (Fig. 12, Fig. 13)

DISCUSSION

Patients with ectodermal dysplasia require dental rehabilitation at an early stage in life for the accomplishment of proper function, nutrition, esthetics and emotional stability. Therefore, the treatment modality appropriate for every patient is deciphered and dictated by the skeletal age of the patient.^[7] The skeletal age can be determined using skeletal maturity indicators such as a cephalometric analysis of the cervical vertebrae or a hand-wrist radiograph for the evaluation of the phalanges. An arbitrary indicator would be discomfort due to tightness or the inability to seat the removable prosthesis after a few months because of the underlying growth of the jaw. However, this may be due to other local factors and is not a definitive diagnosis.

A combination of unhampered growth and restoration of the existing dentition is the ultimate objective while planning a prosthesis for an adolescent with such impairment. It must be emphasized that a restriction on the natural growth of the jaws would invariably lead to a disfigurement in the future. Hence, a removable, clip-on smile was the treatment of choice for the growing girl.

In this case the fabrication protocol adopted for the prosthesis was an innovative approach with an amalgamation of digitalization, a waxed-up trial splint and a final esthetic clip-on removable prosthesis. The trial splint was directly transferred intra-orally, giving the patient a true impression of how the final prosthesis will fit. Psychologically this is of great benefit while treating a child, as it acts as a preparatory step before the final prosthesis is fabricated. Further, this trial saves valuable chair-side time and modifications can be made instantly by carving out the wax on the retainer until a satisfactory result is accomplished. Once the trial was approved, it was exactly duplicated for the clip-on prosthesis. Since the final prosthesis was a thermoform splint reinforced with self cure acrylic,

a micromechanical bond was established between the two materials through microblasting. An advantage of this unconventional material combination is that the intaglio surface is a relatively softer thermoform material compared to rigid acrylic, which provides cushioning, superior comfort and reduced possibility of frictional wear between the natural dentition and prosthesis.

The patient was guided to wear the clip-on prosthesis only during wake hours and never while sleeping in order to minimize any kind of restriction on the normal growth. The patient was informed to rinse the mouth and retainer thoroughly after every meal to avoid any food impaction between the natural teeth and the prosthesis; therefore, preventing the development of dental caries and halitosis. The patient was educated on meticulous hygiene maintenance for her dentition and prosthesis. She was instructed to continue brushing her teeth twice a day as usual; while the clip-on prosthesis should be immersed in water overnight, brushed gently with a soft bristled brush and disinfected by soaking it in a denture cleansing agent twice a week overnight. A regular recall every three to six months is of utmost importance for a patient requiring a close assessment of the intra-oral status. This allows the practitioner to examine the oral and periodontal health, conduct a preventive chair-side fluoride treatment at regular intervals and analyze the growth of the adolescent. An additional advantage of this prosthetic design includes the utilization of the prosthesis itself as a custom tray for the biannual fluoride treatment.

Furthermore, the removable clip-on prosthesis is a transitional prosthesis which will require periodic adjustment, modification and replacement depending on the growth of the adolescent. Therefore, one of the criteria for this temporary-continuous treatment was to make it relatively cost effective as the treatment may require multiple fabrications.

Moreover, this treatment modality utilizes an extremely conservative approach, requiring no invasive procedure such as extractions, root canals or tooth preparation; hence maintaining the existing dentition, supporting periodontium and proprioceptive response. Thus improving the prognosis for the definitive prosthesis in the future.

Further, the psychological benefit gained from this prosthesis was unparalleled. There was an immediate change in the patient's confidence, personality, speech and articulation. Hence, this removable appliance will help in her personal growth and development in the most susceptible stage of her life.

In the past, there have been several authors who propagate the use of a removable prosthesis for the management of growing patients with ectodermal dysplasia.^[8] However, the treatment modality was generally confined to the fabrication of overdentures.^[9] This unconventional approach utilizes similar concepts, however it eliminates several disadvantages such as: 1) extensive coverage of soft tissue due to denture extensions, in the process improving patient comfort during speech and mastication; 2) psychologically the patient acceptance is better as this device acts as a clip-on smile rather than a complete denture; 3) it is an extremely conservative and cost effective method, requiring a total of three clinical appointments.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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