CASE REPORT

Hypohidrotic Ectodermal Dysplasia with Anodontia: A Rare Case—Rehabilitation by Prosthetic Management

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ABSTRACT

Ectodermal dysplasia is a hereditary disorder characterized by developmental dystrophies of ectodermal derivatives. It is characterized by triad of signs comprising sparse hair, abnormal or missing teeth and inability to sweat. This case of 12-year-old boy with hypohidrotic ectodermal dysplasia and complete anodontia of both primary and permanent dentition is presented. Owing to the need for treatment at an early age for anodontia and due to the ill-formed maxillary and mandibular residual ridges, the prosthetic management can be difficult. Complete dentures with soft liners and hollow maxillary denture were provided to encourage normal psychological development and to improve the function of the stomatognathic system.

Keywords: Ectodermal dysplasia, Anodontia, Soft liner.

INTRODUCTION

Ectodermal dysplasia (ED) is the term used to describe a group of rare inherited disorders characterized by dysplasia of tissues of ectodermal origin—primarily nails, teeth and skin and occasionally dysplasia of mesodernally derived tissues.¹ The condition is thought to occur in approximately 1 of 100,000 live births.¹² Clinically ED is divided into two broad categories, the hypohidrotic form (X-linked recessive) and the hidrotic form (autosomal inherited). Hypohidrotic ED also termed as Christ Siemens Touraine syndrome,³⁴ is more common and is characterized by a triad of signs comprising sparse hair (hypotrichosis), abnormal or missing teeth (hypodontia or anodontia) and an inability to sweat because of lack of sweat glands (hypohidrosis). Hypodontia in turn causes reduced alveolar bone growth and lack of development of alveolar ridges which often appear clinically to be extremely narrow and concave linguually. Teeth, if present, are often conical in shape, malformed and widely spaced. Other symptoms include saddle nose, prominent lips, linear wrinkles and hyperpigmentation around eyes, atopy, mild facial dysmorphic features and increased susceptibility to respiratory infections.⁵⁶

This case report essentially emphasizes the prosthodontic management of appearance and functionality of treatment in the form of a complete denture provided to the patient.

CASE REPORT

A 12-year-old boy reported to the Department of Prosthodontics with the complaint of absence of teeth since birth, inability to eat and difficulty in speech. The general medical history and family history was noncontributing. The boy was diagnosed to have hypohidrotic ED. The boy exhibited the classical features of ED: Complete anodontia, hypohidrosis, hypotrichosis, prominent forehead, saddle nose, diminished lower facial height, sparse scalp hair, missing eyelashes and eyebrows and protuberant lips (Fig. 1).

The intraoral examination revealed complete absence of deciduous and permanent teeth as a result of which the edentulous alveolar ridges were deficient in both height and width. The mandibular ridge was of the knife edge variety and the maxillary ridge was depressed posteriorly with a flat and shallow palate. The oral mucosa had a slight dry appearance and the tongue seemed relatively large (Figs 2A and B).

Radiographic examination revealed complete absence of permanent tooth buds and underdeveloped maxillary and mandibular alveolar ridges (Fig. 3). Later the clinical diagnosis was confirmed by both oral medicine and oral pathology departments after taking the patients case history.

Fig. 1: Preoperative photograph of ectodermal dysplasia patient
Implants retained prosthesis was not considered to the patient because of highly resorbed residual ridges and even economic constrains of the family.

A removable maxillary and mandibular complete denture was planned for the patient, considering the patients need for mastication, esthetics, speech, comfort and overall psychological development. Although routine procedures for construction of complete denture were performed, case-specific modifications were required as hollow maxillary denture due to large interridge space and permanent soft liner to both maxillary and mandibular complete dentures.

Clinical Procedures

The maxillary and mandibular primary impressions were recorded using stock trays and polyvinyl siloxane putty impression material and primary casts were made. Acrylic custom trays with spacer were fabricated on the primary casts and border molding was done using low fusing compound and final impression were made with monophase elastomeric impression material and master casts were obtained.

Temporary record bases and wax occlusal rims were made and jaw relation was recorded conventionally, after assessing the phonetics and esthetics, face bow transfer was done and mounted on to the Hanau wide view semiadjustable articulator. Teeth arrangement was customized in accordance to patient’s esthetics and age as mild crowding, mamelons. Following the try in appointment, the waxed up denture was planned for acrylization.

Owing to increased interridge space, there would be increase in weight of maxillary denture resulting in regular dislodgment and inconvenience to the patient. So we had planned to make maxillary denture hollow. As the patients residual ridges were severely resorbed and even dry due to reduced salivation we had planned to use permanent soft liner on tissue surfaces of both the dentures.

Laboratory procedures for hollow maxillary denture and soft liner for maxillary and mandibular complete denture:

1. After the trial procedure the land area of the cast was indexed using a conical bur and the trial denture was sealed to master cast. The master cast along with trial denture was duplicated with reversible hydrocolloid and dental stone was poured. A 0.5 mm thermoplastic sheet was adopted to duplicated cast and a clear template was made.

2. The trial denture was processed in a standard manner though the wax elimination stage.

3. Two layers of modeling wax was adopted to the definitive cast, confirming to the border extension. Second flask was used to invest the wax and wax elimination was completed.

Permanent soft liner adaptation:

1. Dewaxed record base was used to create space for the permanent soft line adaptation, record base was trimmed to 1 mm thick and was used to have space for the soft liner, cellulose sheet was adopted over the record base and heat cure acrylic resin was packed and trial closure was done, the flasks were opened and permanent soft lining material was mixed according to manufacturers’ recommendations and packed after removing the record base. The flask was closed and the dentures were processed conventionally.

2. After processing of the permanent record base with the soft liner, the cope was separated without detaching the
permanent record base from the cast. Clear matrix was placed on the definitive cast using the indexes on the land area as seating guides. A calibrated probe was used to measure the space between the matrix and the processed resin.

3. Vinyl polysiloxane putty was adopted to the bur roughened processed resin and was shaped to approximate contours of the matrix. The putty was shaped with a bur to leave 2 to 3 mm space between putty and matrix all over and was fixed to the processed resin using cyanoacrylate.

4. Original cope was reseated on the drag to verify complete closure of the flask. Acrylic resin was mixed, packed and polymerized.

5. After completion of acrylization the processed dentures were recovered, remounted on the semiadjustable articulator and necessary occlusal corrections were made.

6. Two openings were made with a bur into the denture base distal to most posterior teeth and the silicon putty was removed by scraping with a sharp instrument. The opening was widened laterally as necessary to facilitate the access. The putty was removed completely and the openings were sealed with self-cure resin.

7. Dentures were finished and polished, hollowness was verified by immersing the denture in water as it floats (Fig. 4).

After the final insertion, postinsertion instructions for maintenance were given to the patient and parents (Figs 5 and 6). At recall appointments pressure spots were identified and relieved.

Patient was very happy and satisfied with the dentures. Retention of the maxillary denture was good owing to its reduced weight and soft liner. Patient gradually adapted to the lower denture. Patient self-esteem improved satisfyingly as did his socialization skills.

Further recall has been scheduled every 3 months. Further treatment will include modification of the dentures by relining or replacement of the dentures according to observed skeletal growth.

**DISCUSSION**

The typical decreased vertical facial height of this patient resulted from the congenital absence of teeth leading to underdevelopment of the alveolar ridges, causing increased interridge space. The dentures fabricated in such conditions would result in heavy weight as a result there are more chances of looseness of dentures making patients uncomfortable and annoying. Making the maxillary denture hollow decreases the denture weight considerably giving comfort to the patient. Furthermore, studies have proved that increasing the weight of the denture causes accelerated rate of alveolar resorption, so the hollow denture decreases the denture weight and in turn preserves the residual alveolar ridge.

Classically, ED patients have decreased salivation so their oral mucosa becomes dry and are more prone to denture related trauma, soft liner on the tissue surface cushions and protects the oral tissue from trauma. Furthermore, any available undercuts can be used to increase the retention level in such highly resorbed conditions.

Historically prosthetic treatment for ectodermal dysplasia patients involved removable partial dentures, removable partial or complete overlay dentures and fixed dental prosthesis. The advent of dental implants and implant borne total telescopic dentures has provided an additional treatment modality for restoration of dentition in this group of patients. There are esthetic, functional and psychological reasons that make it important to start oral rehabilitation early in life. However, this is usually a difficult condition to manage prosthodontically because of the typical oral...
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Serial sets of dentures may be required as the child matures. Shaw describes the need for these patients to restore vertical dimension and prevent the undesirable protruding lips secondary to overclosure, thereby improving the profile.4

Ideally, a restored dentition should be in place before the child attends primary school as oral rehabilitation has a major psychological impact on the patients and facilitating social acceptance.

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REFERENCES


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