

Complete denture prostheses in an 8-year-old child with hypohidrotic ectodermal dysplasia

Vidhyasagar Mopagar¹, Shradha Naik^{1*}, Rutuj G. Jadhav¹, Ashish Raurale²

¹Departments of Pedodontics and Preventive Dentistry, Rural Dental College, ²YCMM and Rural Dental Development College, Ahmednagar, Maharashtra, India

ABSTRACT

Ectodermal dysplasia (ED) is a large, heterogeneous group of inherited disorders that are defined by primary defects in the development of two or more tissues derived from embryonic ectoderm. The intimate origin of the diverse ectodermal structures account for the wide spectrum of dysplasia. Clinically, the hair (hypotrichosis, partial, or total alopecia), nails (dystrophic, hypertrophic, or abnormally keratinized), teeth (enamel defects or absence), and sweat glands (hypoplastic or aplastic) are most commonly affected in this group of disorders. Dental treatment is often necessary in patients with some forms of ED and some children may need dentures as early as 3 years of age. It is important to seek dental advice early, as maintenance of the alveolar ridge is important for a more comprehensive treatment in adulthood. The peculiar clinical features and oral rehabilitation of an 8-year-old patient with ED have been described in detail in the following sections.

Key words: Complete dentures, Ectodermal Dysplasia, Syndrome

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INTRODUCTION

Ectodermal dysplasia (ED) is a rare group of disorder with an estimated incidence of seven in 100,000 births for all ED.^[1] More than 170 clinically distinct hereditary syndromes are present in which ED is present.^[2] Out of the numerous different classifications of EDs, a simple attempt was made by Nelson, which included five categories: Hypohidrotic (anhidrotic); hidrotic (Clouston's syndrome); ectrodactyly ectodermal dysplasia syndrome; Rapp-Hodgkin syndrome; and Robinson's disease.^[3]

Clinical features

The EDs may be defined as conditions with at least one of the following four features: Trichodysplasia, dental defects, onychodysplasia, or dyshidrosis as well as at least one sign showing involvement of another structure of ectodermal origin.^[4] Partial or complete absence of sweat glands in these patients makes them unable to tolerate warmer temperatures; hypohidrosis being the most characteristic feature. Dry palmer plantar surfaces

with white keratotic patches, smooth and dry skin, sparse, and thinned out hair all over the body are a few important features. In females usually, the mammary glands are aplastic or hypoplastic. Other features include abnormal development of nails, malfunction of lacrimal glands, and increased susceptibility to skin infections and allergies.^[5]

Dysmorphic facial features such as prominent supraorbital ridges, frontal bossing and a depressed nasal bridge ('saddle' nose formation) are usually observed. Other facial symptoms include maxillary hypoplasia, prominent lips and linear wrinkles around the eyes.^[2,5] Intraorally, beside mucous symptoms, patients with hypohidrotic ED suffer from both an altered shape and a reduced number of teeth, most of them having conical shaped teeth.

Other manifestations are seen in the general growth and development of the child, with below average height and weight, chronic respiratory tract infections, speech defects, and the overall quality of life.^[6-8]

*Address for correspondence:

Dr. Shradha Naik, Department of Pedodontics and Preventive Dentistry, Rural Dental College, Loni - 413 736, Maharashtra, India.
E-mail: shrads88@gmail.com

CASE REPORT

An 10-years-old male patient reported with the chief complaint of missing teeth, inability to chew hard food and low body weight. The patient's parents wanted a prosthesis, which could help the child with mastication and improve his social acceptance. History revealed that the patient had missing teeth since birth. The patient had got dentures made elsewhere two years ago but did not wear them due to the ill fitting nature and soft tissue trauma. Also, he did not bring with him the previously made dentures. Based on the detailed medical history and general evaluation, we suspected a case of hypohidrotic ED, which was later confirmed on referral to a pediatrician and dermatologist. Health histories and oral examination of both the parents and three siblings ruled out any form of ectodermal dysplastic condition in them.

The patient presented with typical features hypotrichosis, scarce eyebrows and eyelashes, dry anhidrotic skin, depressed nasal bridge, thick lips, dark pigmented skin around periorbital area and a diminished lower facial height contributing to a senile facial expression [Figure 1].

Intraoral examination showed edentulous upper and lower ridge, which were atrophic with decreased height [Figure 2a and b]. The palate was shallow, oral mucosa was normal and dry due to less saliva, the tongue was relatively large. Orthopantomogram did not show presence of any impacted teeth or developing tooth bud [Figure 2c].

Prosthodontic management

To improve the child's appearance, mastication, and speech, he was provided with removable complete maxillary and mandibular dentures. The basic principles of removable prosthodontic treatment for children suffering from ED have been reported^[9] and were followed in the case presented here.

Primary impressions

Initial impression for the purpose of study and record were taken using irreversible hydrocolloid impression material followed by medium bodied rubber based impressions for special tray construction [Figure 3a].

Final impressions

The patient was cooperative and motivated which allowed us to perform border molding with low fusing green stick compound and the final impressions with zinc oxide eugenol irreversible rigid impression paste [Figure 3b].

Jaw relation record

The occlusal vertical dimension (OVD) in these patients is most often collapsed and needs to be restored to a

clinically acceptable position making this step challenging and requiring accuracy. Heat-processed record bases which provide superior stability, confirmation of the final retention, and easier adjustment of occlusion were fabricated. Occlusal rims were fabricated chair side, and



Figure 1: Extraoral features

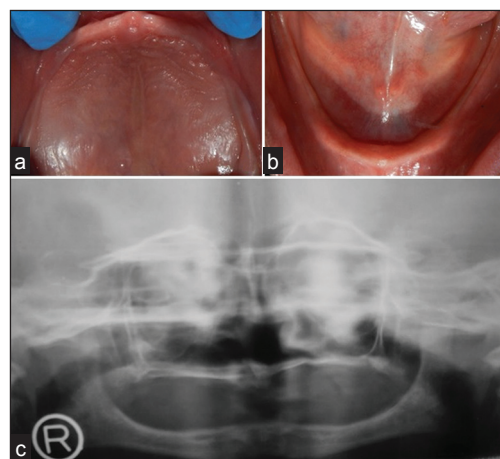


Figure 2: (a) Maxillary alveolar arch. (b) Mandibular alveolar arch. (c) Orthopantomogram showing absence of tooth buds

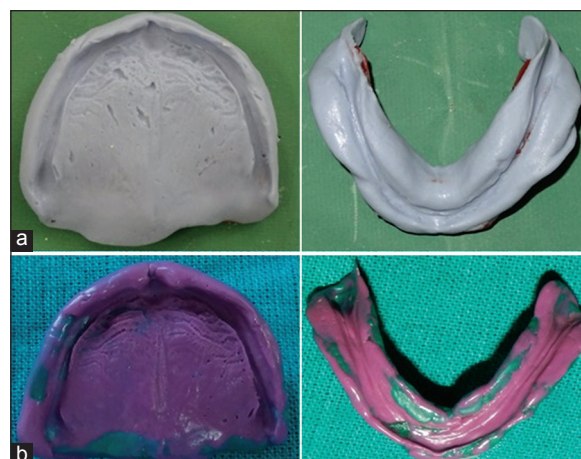


Figure 3: (a) Primary impressions. (b) Final impressions

the appropriate OVD was determined. A fox plane was used to confirm the occlusal plane, and the centric jaw relation was recorded [Figure 4a and b].

Teeth selection and try in

Pediatric mold stock denture teeth were not used since there was a longer arch perimeter in both jaws. Instead, small sized adult stock acrylic denture teeth with appropriate shade were selected to simulate the pediatric dentition (keeping in mind the dentogenic concept of teeth selection for complete dentures). To match the patient's facial skin tone, medium to darker shade of teeth set was used. No gross modifications were needed during try in [Figure 4c].

The dentures were fabricated in heat-processed acrylic resin. Occlusion was refined, and the dentures were finished and polished. The dentures were delivered to the patient who immediately got accustomed. Initially, the patient had some difficulty in keeping the mandibular denture in his mouth. To improve and facilitate its adaptation, he was advised to use a denture adhesive paste for the first few days. A week later, he was completely at ease with his new prostheses [Figure 5a].

On recall appointment after 1 month, some minor occlusal adjustments were made, however the patient did not have any complaints. We did not observe any mucosal changes or reactions. There was significant improvement in patient's appearance, mastication and weight observed during 3 months recall appointment [Figure 5b].

DISCUSSION

There is little information in the literature, other than clinical reports, with regard to the dental management of the young patient with ED. This case was diagnosed only after the patient approached a dentist due to difficulty

in eating and absence of teeth. Due to unawareness and poor socio-economic status of parents, the patient had not visited a general practitioner for his continually deteriorating health, recurrent episodes of fever, inability to tolerate heat, and overall appearance.

This condition can be diagnosed based on clinical appearance, symptoms and certain tests. Radiographs showing skeletal deformities, renal ultrasonography, pilocarpine iontophoresis, iodine starch test and skin biopsy are some of the means which can be used to confirm the presence of hypohidrotic type of ED. Apart from this specific genetic testing can also be done to identify the associated syndrome.^[10]

The presence of complete anodontia is extremely rare and unique to observe and in this case justifies the fabrication of complete dentures. A literature review by Pigno *et al.* stated that patients with ED present a characteristically thin and underdeveloped residual ridge, covered by thin mucosa and topped by movable connective tissue. These features, along with the decreased quantity of saliva, are the main problems encountered in treating such cases.^[11]

There is no fixed age reported for prosthesis; however, it has been recommended to give a temporary as soon as the school starts. This will help the patient to adapt socially, develop proper speech and improve learning through good nutrition and development.^[12] Success with complete dentures at 3 years of age has also been reported.^[13]

In cases, where a few teeth are present various treatment modalities such as endosseous implants,^[1] overdentures,^[14] and removable partial dentures have been tried with considerable success.^[15]

In the present case, the patient was cooperative and well-motivated, because of which it became relatively easier for him to accept and adjust to the appliance. There was

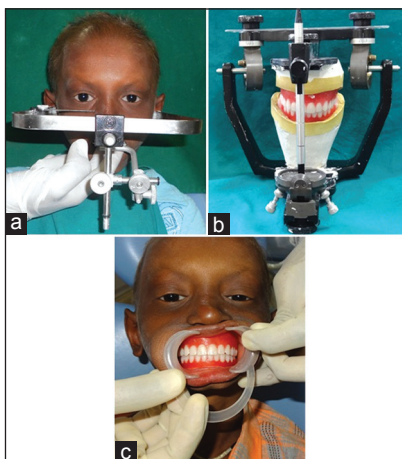


Figure 4: (a) Jaw relation using Hanau's Facebow. (b) Teeth setting. (c) Try in stage

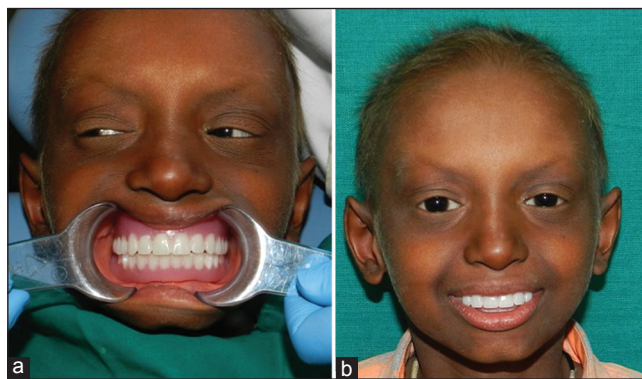


Figure 5: (a) Intraoral view showing maxillary and mandibular complete dentures on denture insertion. (b) Three months recall showing improvement in appearance, check fullness

significant improvement in his weight, speech and overall confidence which was well appreciated by his parents. It is important to keep a periodic recall and modify the dentures accordingly due to the continuously growing arches.

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