

Prosthodontic Rehabilitation of Ectodermal Dysplasia with Oligodontia : A Case Report

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Introduction

Ectodermal Dysplasia (EDs) is a rare group of inherited disorders characterized by developmental defects in the tissue formed from the outer embryonic layer. These include anomalies in the skin, the mucous membrane and their adnexes and are of two types: X-linked anhidrotic or hypohidrotic ED (Christ-Siemens-Touraine syndrome) and Hidrotic ED (Clouston's syndrome)^(1,2) Genetic studies show that mutations in the Ectodysplasin-A and Ectodysplasin-A receptor genes are responsible for X-linked and Autosomal Hypohidrotic Ectodermal dysplasia^(2,3) Dental abnormalities of size, shape and number are commonly seen. Incisors and canines are often conical-shaped while primary second molars, if present, are mostly affected by taurodontism.^(1,2)

This clinical report aims to present the prosthetic rehabilitation of a patient with Hypohidrotic Ectodermal Dysplasia.

Case report

A 3 year old boy reported to the Department of Paediatric and Preventive Dentistry with a complaint of multiple missing teeth, inability to eat properly and defective speech. The child was diagnosed with Ectodermal Dysplasia since birth with no relevant family history.

Extraoral examination revealed classical features of ED. Sparse hair on scalp, saddle nose, frontal bossing, sunken cheeks, hyperpigmented skin around the eyes, protuberant lips and decreased lower facial height were noted.

Intraoral examination showed presence of primary maxillary second molars and mandibular canines and second molars. Presence of thin, atrophic maxillary and mandibular ridges and multiple missing teeth were noted.

OPG showed only the developing tooth buds of maxillary and mandibular permanent first molars and oligodontia. The parents desired a prosthesis which could facilitate eating as well as improve his societal acceptance so a treatment plan was formulated accordingly. The patient was planned for removable prosthetic rehabilitation of the maxillary and mandibular jaws and informed consent was taken.

Specialized custom-made primary acrylic trays fabricated on

an ideal primary cast were used for this purpose. Impressions were taken with light-body vinyl siloxane impression material. The removable dentures were fabricated with conventional denture making procedure. The patient was recalled after 24 hours. Fluoride mouthwash was prescribed. The patient seemed well adjusted to the dentures after 3 weeks. The parents noticed a drastic improvement in the child's eating habits, speech and aesthetics as well as a positive attitude and acceptance of the child towards the dentures. He was scheduled for recall for every 3 months to evaluate his oral hygiene status and maintenance of dentures. (Figure 1)

Discussion

Patient's age, tooth agenesis and its degree of malformation, growth and development of stomatognathic system of patient and patient's motivation are factors which influence treatment plan of a patient with ED.⁽⁴⁾ A multidisciplinary approach is required for the treatment of such cases. The desire to be like others with teeth can be a motivator for cooperation in even the young child. (Pipa Vallejo et al 2008).^(5,6) Our patient in this case report presented with Hypohidrotic ED based on his clinical manifestations.

Nowak stated that 'treating the paediatric patient with ectodermal dysplasia(ED) requires the clinician to be knowledgeable in growth and development, behavioural management, techniques in the fabrication of a prosthesis, modifying the existing teeth using various techniques for restoration as well as the ability to motivate not only the patient but also the parent in the usage of the prosthesis, and the regular follow-up for the modification and/or replacement of the prostheses.'⁽⁷⁾ Many authors have proposed various types of rehabilitation possibilities for these patients, of which the use of removable prosthesis during the early stages of growth (3-5 years) has been recommended to control the vertical dimension and to avoid ridge resorption (Tarjan et al 2005).⁽⁶⁾ Removable prostheses provide the patient an effective, affordable, easy, and reversible method of oral rehabilitation.⁽⁸⁾ Retention and stability for the prostheses are difficult to obtain due to dryness of oral mucosa and the under-developed alveolar ridges. (Pigno MA

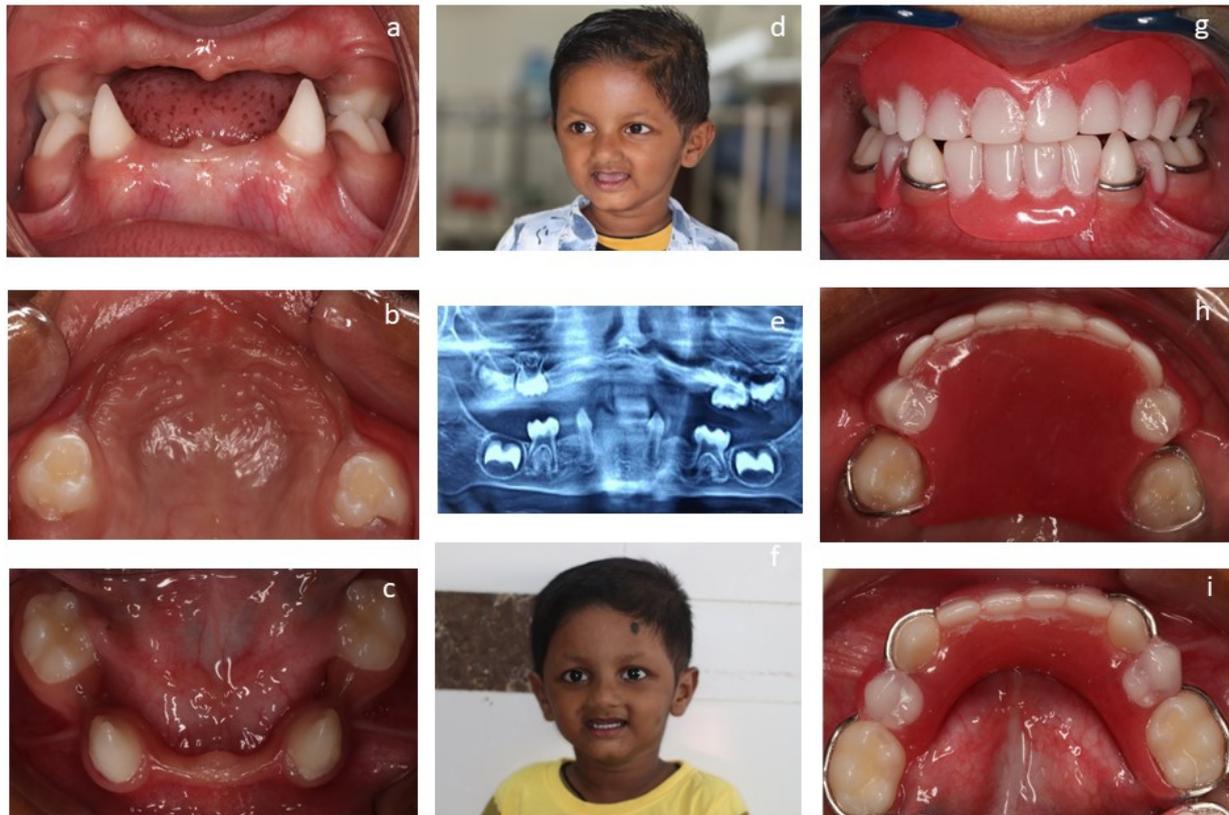


Figure 1: a-Preoperative occlusion, b- Preoperative maxillary arch, c- Preoperative mandibular arch. d- Preoperative smile of the patient, e-OPG showing oligodontia, f-Postoperative smile of the patient. g- Postoperative occlusion. h- Postoperative maxillary arch. i-Postoperative mandibular arch

et al 1996). When teeth are present in the mouth, overdentures are the preferred treatment option. Fixed Prosthodontic treatment is rarely used because of decreased number of abutments, and moreover the patient is too young. For adult patients with ED, dental implants are the preferred treatment of choice because growth has stabilized and implants can be used to support, retain and stabilize the prosthesis.⁽⁹⁾

Conclusion

The meticulous follow-up of these patients after removable prosthetic rehabilitation is essential to ensure success of the treatment and to avoid any complications. Education of the patient and the parents regarding possible problems and maintenance of any prosthesis is mandatory. Cooperation of the patient and family is a crucial factor for the removable prosthesis to have a successful prognosis.

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