

Case Report**Prosthetic Reconstruction of a 4 Year Old Child with Hypohidrotic Ectodermal Dysplasia Born to Parents with Consanguineous Marriages; Case Report**

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Ectodermal dysplasia is a disease that affects components of body with ectodermal origin, so it is manifested by thin hair, malformed or missing teeth and lack of sweating. In this case, I present a 4 year old boy with hypohidrotic ectodermal dysplasia. He had psychological issues and difficulty in eating. In this young child, with prosthetic treatment that included partial removable denture, his problems was dissolved.

INTRODUCTION

Ectodermal dysplasia (ED) encompasses a set of genetic diseases that affects components of body with ectodermal origin. These structures include skin, hair, teeth, nails, and sweat glands (1). Ectodermal dysplasia's inheritance may be via X-linked, autosomal-dominant, and autosomal-recessive patterns (2). Ectodermal dysplasia has the prevalence from 1:10 000 to 1:100 000 (3).

Clinical forms of ectodermal dysplasia may be Hypohidrotic or Hydrotic. Hypohidrotic form also described as Christ-Siemens Touraine syndrome (4,5) is characterized by signs contain thin hair, malformed or missing teeth (hypodontia or anodontia), and lack of sweat glands (6).

Ectodermal dysplasia in oral area has features like hypodontia or anodontia, deficiency in alveolar ridge, lower face's reduction in vertical dimension, disappearance of the vermilion border, malformation of teeth, dryness of the oral mucosa. The child usually has the senile appearance (7).

Other characteristics include; a depressed nasal bridge, frontal bossing, protuberant lips (8).

This case underlines prosthetic treatment included partial denture at an early age.

CASE PRESENTATION

A 4 year old boy was referred to the department of pediatric dentistry, Tabriz Azad University, Tabriz, Iran, with

the complaint of missing teeth, inability to eat, difficulty in speech and poor esthetics.

Family history revealed that he was the only child of parents with consanguineous marriage and none of the other relatives had similar condition.

The medical history confirmed existence of ectodermal dysplasia. The parent were aware of this condition. They mentioned that child was intolerant to heat. His skin was dry with sparse body hair.

Extra-oral examination revealed classic features of ectodermal dysplasia like frontal bossing, absent eyebrows, loss of eyelashes, saddle nose, protuberant lips and a diminished lower facial height (Figure 1).

The intra-oral examination showed presence of second primary molars in maxilla and mandible and a conical shaped lateral primary incisor in maxilla. Deficiency in alveolar ridges was obvious in height and width. The oral mucosa was dry in appearance. There was a caries on upper left second primary molar (Figure 2).

Radiographic examination showed presence of four first permanent molars tooth germs (Figure 3).

The upper left second primary molar's caries was filled with amalgam (SDI, Australia). Removable partial dentures was planned for maxilla and mandible. Initial impressions using irreversible hydrocolloid were made (TECO, Holland). Border moulding was done after preparation of custom trays.

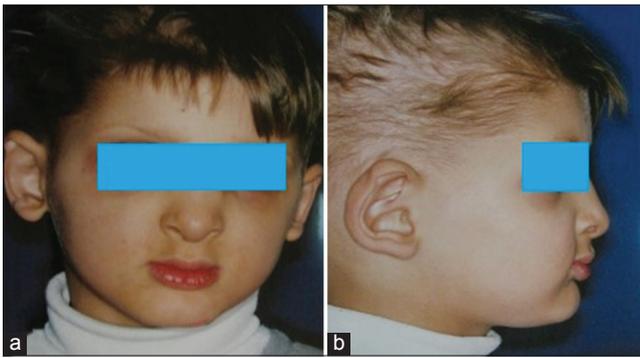


Figure 1. a) Frontal view of the patient, b) Lateral view of the patient



Figure 2. Intra-oral views before treatment. a) frontal view, b) maxillary occlusal view, c) mandibular occlusal view

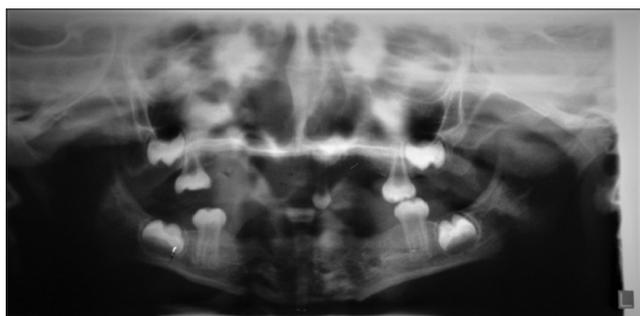


Figure 3. Panoramic radiography of the patient before treatment

For arches of maxilla and mandible final impressions were made with irreversible hydrocolloid (TECO, Holland). Jaw relationship was recorded, followed by trial dentures and finally finished partial dentures were given to complete oral and psychological rehabilitation (Figure 4).

Instructions for oral hygiene and not eating sticky foods were provided. Follow up visit was scheduled 2 weeks later. The parents noted that the child was pleasant and his self-esteem improved. Other appointments for follow up visits were 3 and 6 months later.

Adjustment of partial dentures by relining or replacement will be needed due to growth phase of the child.

DISCUSSION

Early oral rehabilitation has an important effect in life of patients with ectodermal dysplasia because of aesthetic, functional and psychological reasons.



Figure 4. Extra and Intra-oral views after treatment. a) Full Frontal view, b) intra-oral 3/4 view, c) maxillary occlusal view, d) mandibular occlusal view

Dentist must be well informed in diagnosing the patient with ectodermal dysplasia, his growth and development, restoring existing teeth using various materials and methods of prosthetic rehabilitation. The clinician must have the ability to motivate the child for using the prosthesis and follow up for a long period (9).

Schnabl et al. perform a systematic review and found that the mean age for reconstruction with prosthesis in patients with ectodermal dysplasia is 4 years (10). Treatment of patients with ectodermal dysplasia in early age has many benefits like normalizing masticatory muscles' operation, improvement of the senile profile that caused by reduction in vertical dimension of face and absence of teeth. It also helps to increase the child's self-esteem.

Using dental implants in children with ectodermal dysplasia is becoming popular and have benefits, but their placement in growing phase may also have concerns. In pediatric patients, Implants can be used in anterior mandible before skeletal maturation (11).

An appropriate choice for treatment of children with ectodermal dysplasia, is a removable denture or an over denture due to easy modification of denture that may be needed during growth phase of child (12). For this patient, treatment goals were to provide a functional occlusion, esthetic appearance, improvement in patient's nutrition and self-esteem increase.

In this case, parents of the child had consanguineous marriage. According to More et al., 68% of the cases with ectodermal dysplasia borned to parents with consanguineous marriage, but he noted that it is necessary to founding the prevalence of this occurrence. He also noted that it is important to take family history (13).

SUMMARY

This case addressed the oral construction of a patient with ectodermal dysplasia. Early treatment has a positive effect on aesthetics, masticatory function, speech and patient's psychological development, thus enhancing the quality of life of the patient.

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