

Ectodermal Dysplasia in a 6 Years Old: A Clinical Challenge

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ABSTRACT

Ectodermal dysplasia depicts a group of inherited disorder in which two or more than two ectodermally derived structures fail to develop perfectly. Intervention at an early stage not only improves patient's appearance but also minimize the associated psychological and emotional problem in such children. This paper presents a case of a 6 years old girl who was referred to our department with complain of absence of multiple teeth for which oral rehabilitation was done.

Keywords: Ectodermal dysplasia, Hypodontia, Oral rehabilitation.

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INTRODUCTION

Ectodermal dysplasia (ED) has been defined by National Foundation for ectodermal dysplasia as a genetic disruption in which there are congenital birth defects of two or more ectodermal structures.¹ First case of ectodermal dysplasia was documented by Thurman in 1848, but the term was coined in 1929 by Weech.² Female carriers outnumber affected men and Pigno in 1996 stated that females show little or no signs of the condition.³

Patients with ectodermal dysplasia are characterized by hypoplasia or aplasia of ectodermal structures, such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye, ear and other organs.⁴ Ectodermal dysplasia might be inherited in any form of several genetic patterns, including autosomal-dominant, autosomal-recessive, and X-linked modes.¹ Although, more than 170 different subtypes of ectodermal dysplasia have been identified, this genetic disorder is relatively rare with an estimated incidence of one case per 100,000.⁵

Ectodermal dysplasia can be of two types clinically as follows:

1. Hypohidrotic form/Christ-Siemens-Touraine syndrome
2. Hidrotic/Clouston form

The hypohidrotic form exhibits classical triad of hypohidrosis, hypotrichosis and hypodontia with usual X-linked recessive pattern. Males are affected severely while females show minor defects. In this form of ED, sweat glands are usually spared.⁶

In the oral cavity, the most prominent feature in oligodontia. Teeth in anterior maxilla and mandible are conical in shape.⁷

In hydrotic form of ED, chief clinical features include nail dystrophy, palmoplantar keratosis and hair defects. The patients have normal sweating and facies. Ectodermal dysplasia patients require both functional and esthetic facial rehabilitation. It is a challenge to manage in children which requires teamwork from medical and dental professionals.

CASE REPORT

A 6 years old female child reported to department of pedodontics and preventive dentistry, with the chief complain of lack of esthetics and missing teeth because of which patient developed tongue thrust habit. Extraoral examination revealed sparse hair on scalp, thin eyebrows, sparse eyelashes, frontal bossing, protuberant lips and decreased lower facial height. Dryness of skin was absent, sweating and nails were also noted to be normal. Speech defects in the form of difficulty in pronouncing labiodental consonants (f, v) and lingua-alveolar consonants (s, z, t, f) was observed.

Intraoral examination revealed missing primary maxillary lateral incisor as well as primary mandibular central and lateral incisors, canines and right left first molar. However, in right mandibular region a protrusive ridge was observed which was suspected to be erupting primary canine. Alveolar ridge in edentulous areas appeared thin and vestibule shallow. The primary canines had characteristic conical form and first primary molars had the bud crown form (Figs 1 and 2). Orthopantomograph confirmed not only the presence on right mandibular primary canine but also revealed congenitally missing tooth buds of right permanent mandibular first and second premolars, canine and incisors as well as left permanent mandibular incisors (Fig. 3).

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Fig. 1: Intraoral photograph of teeth in maxillary arch



Fig. 2: Intraoral photograph of teeth missing in mandibular arch



Fig. 3: Orthopantomograph depicting the status of primary and permanent teeth

Family history regarding similar features was negative suggesting autosomal recessive mode of inheritance.

Upper and lower impression were made and poured in dental stone, and acrylic plate was made using cold cure pink acrylic on the mandibular cast (Fig. 4) after which bite registration was done using Futar bite registration material. The articulated casts were then mounted on a mean value articulator for building up of occlusal rim.

Teeth setting were done on the fabricated rims and patient was recalled for trial fitting of denture (Fig. 5). The unacrylized denture was then sent to laboratory for heat cure acrylization using pink acrylic. Patient was recalled again, and the denture was inserted and postinsertion instructions were given (Fig. 6).

DISCUSSION

The case presented above had defective hair and partial anodontia with no family history. So, a diagnosis of ED of autosomal recessive inheritance was concluded. However, further studies on this condition to define phenotypic spectra are required to point out the precise pathogenesis.

Oral rehabilitation of patients with ED is necessary to improve esthetics, speech, masticatory efficiency and sagittal and vertical skeletal relationships during crani-ofacial growth.⁸

Guckes et al have stated that placement of implants in young children (<13 years of age) may result in displacement of the implant, but he suggested that implant may be used in anterior mandible of young patients routinely.⁹



Fig. 4: Maxillary and mandibular casts after impression making



Fig. 5: Articulated casts on mean value articulator



Fig. 6: Postoperative photograph depicting mandibular arch after delivery of the denture

It has been reported that child's self image is completed by 4 to 5 years of age; therefore, cosmetic measures must be initiated as soon as possible to have the child resemble his peer group. Relining or rebasing should be done in every 1 to 2 years because growth of jaw is independent of presence of teeth.^{10,11}

Fabrication of denture for a young child may not be complicated but requires thorough knowledge of growth and development, behavior management and skills for fabricating dentures.¹² A thorough follow-up is required until patient reaches adult dentition and then a fixed treatment like fixed partial denture, overdentures or an implant-retained prosthesis may be opted for.

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