

MANAGEMENT OF HEREDITARY ECTODERMAL DYSPLASIA OF A CHILD : A CASE REPORT

Dr. Dimpleja J*, Dr. Mishal Muhammed Haris*, Dr. Zarrin Rahnuma*
Dr. Dulal Das**, Dr. Mehendi Tirkey***

ABSTRACT

Ectodermal dysplasia is an inherited disorder which affects the ectodermal derivative such as hair nail teeth skin, sweat gland, sebaceous gland. Clinically, based on the presence of the sweat glands involvement it is divided into two major types are: (1) Hypohidrotic or anhidrotic (Christ-Siemens-Touraine syndrome) in which sweat glands are either absent or significantly reduced in number; (2) Hidrotic (Clouston syndrome) in which sweat glands are normal. Hypodontia or complete anodontia is the most prominent dental feature present in ED patient. National Foundation for Ectodermal Dysplasia goal of dental treatment for individuals affected by the ectodermal dysplasia is to provide an age-appropriate dentition that optimizes chewing function (and thus nutrition), oral/facial development, speech, swallowing, and aesthetics. Prosthetic rehabilitation is the major dental need of ectodermal patient to face the society with confidence. In this case report patient rehabilitated with removable partial denture.

KEY WORDS

**Ectodermal Dysplasia,
Anodontia, Prosthetic Rehabilitation**

ABOUT THE AUTHORS

* Post Graduate Trainee

**Assistant Professor

***Dental Surgeon and Clinical Tutor

Department of Pedodontics & Preventive Dentistry

Dr. R Ahmed Dental College and Hospital, Kolkata

CORRESPONDING AUTHOR

Dr. Dimpleja J

Post Graduate Trainee

Department of Pedodontics & Preventive Dentistry

Dr. R Ahmed dental college and Hospital

Email Address : dimpletweez20@gmail.com

Phone Number : 8056812110

INTRODUCTION

Ectodermal dysplasia is an inherited disorder, it affects the ectodermal derivative such as skin, hair, nail, teeth, sweat glands and sebaceous gland¹. So, for 170 different subtypes of ectodermal dysplasia identified². Mode of inheritance of ectodermal dysplasia are autosomal-dominant, autosomal-recessive, and X-linked mode. The international prevalence of ectodermal dysplasia (ED) is approximately 7 per 10,000 births³. Clinically, based on the presence of the sweat glands involvement it is divided into two major types are: (1) Hypohidrotic or anhidrotic (Christ-Siemens-Touraine syndrome) in which sweat glands are either absent or significantly reduced in number; (2) Hidrotic (Clouston syndrome) in which sweat glands are normal². Defects of dental and hair involvement is quite similar in both the types but involvement of sweat glands are different in both the types. Hypohidrotic ectodermal dysplasia as the most common type seems to show an X-linked inheritance pattern with the gene mapping to Xq12-q134; therefore, males are more susceptible than females¹. Hidrotic type is inherited in an autosomal dominant pattern. No clinical findings can detect in new born. Dental, hair and nail anomalies evident during infancy or childhood. Number of hair follicles, sweat glands and sebaceous glands varies. Hair follicles are reduced and hair is very sparse, short, fine, dry all over the body not only scalp. In hypohidrotic ectodermal dysplasia, Eccrine sweat glands are absent or sparse and rudimentary. Some cases mucous glands are absent in upper respiratory tract and in bronchi, oesophagus and duodenum. Dryness of mouth and eye are appreciable features due to the hypoplasia of salivary glands and lacrimal glands^{1,5}. Facial features are frontal bossing, sunken cheeks, saddle nose, thick and everted lips, wrinkled hyperpigmentation are seen around the eyes and low set ears are seen. Dental manifestation are conical or peg shape teeth, hypodontia or complete anodontia and delayed eruption of permanent teeth present. Other features are dry skin, brittle thin nails, lack of breath development, hearing or vision defect, missing toe or finger. Cleft lip and palate also reported in some cases of ectodermal dysplasia. These characteristics features are not obvious at the birth but extensive scaling of skin and unexplained pyrexia is present¹.



Fig :1. A Sparse scalp hair



Fig :1. B Extra Oral Examination



Fig 2 : Intraoral Examination



Fig :3. Panoramic Radiograph

CASE REPORT

A 8 yrs.old boy was referred from Chittaranjan Dental Hospital to the department of paediatric dentistry, Dr. R. Ahmed Dental College and Hospital, with the complaints of absence of many teeth. Family history reveals that a maternal grandfather and uncle also suffered from oligodontia and heat intolerance. Extra oral examination reveals sparse scalp hair, thin eye brows, depressed nasal bridge, retarded malar prominence and protruded lower lip (fig1A,1B). Intra oral examination revealed conical shaped canine, primary second molar, permanent first molar in both right and left side of upper jaw. Primary second molar, permanent first molars in both right and left side of lower jaw. Other oral finding is thin alveolar ridge, reduced vertical bone height, loss of vestibular depth in the lower jaw and also reduced salivation noted (fig2).The radiographic findings revealed two unerupted premolar germ, one unerupted molar germ in right upper jaw and two unerupted premolar germ &two unerupted molar germ in right and left lower

jaw (fig3).Other noticeable features are heat intolerance, reduced sweating, Dry skin, thin nails. Patient was diagnosed with Hypohidrotic ectodermal dysplasia based on the features.

Treatment plan was based on age and Economic background, mainly to preserve the alveolar bone integrity present at the time, so decided to fabricate removable maxillary and mandibular partial denture. Impression taken with irreversible hydrocolloid (Alginate) and cast made with dental stone (fig4A). Bite registration done in the patient mouth and on the same day shade selection of the tooth done according to the adjacent tooth shades (fig4B). Registered bite transferred to articulator and teeth settings done (fig4C). Trail denture adjustments done by checking phonetics (fig4D). Heat cure acrylic resin was used for fabrication of denture. Denture insertion done (fig4E), patient was satisfied both functionally and aesthetically.



Fig :4. A Alginate Impression



Fig :4.B Bite Registration



Fig :4.C Teeth Setting



Fig4. D : Trial denture Try-in



Fig 4. E : Final Insertion of Acrylic Denture

DISCUSSION

Early Dental rehabilitation of patients with ectodermal dysplasia is needed to lead normal social life and also it will preserve the alveolar bone integrity and improve the skeletal relationship of jaw both sagittal and vertical. According to the National Foundation for Ectodermal Dysplasia goal of dental treatment for individuals affected by the ectodermal dysplasia is to provide an age-appropriate dentition that optimizes chewing function (and thus nutrition), oral/facial development, speech, swallowing, and aesthetics. Parents or guardians of young persons with ED often desire to have treatment of children begin as soon as possible. Items that need to be addressed in planning dental treatment include age, psychosocial environment, teeth present, oral hygiene, occlusal vertical dimension, bone volume, jaw growth and development⁸.

The first dental visit of patients with ED should occur as soon as the first tooth erupts, in order to establish the dental home and to explain to the parents the treatment stages required as the child grows¹⁰. Pigno et al¹². recommended use of a dental prosthesis

before the child goes to school at around 3-4 years of age. Schnabl et al., who found that the median age of prosthetic rehabilitation in patients with ED is 4 years. FPDs in children are gaining popularity because of their superior aesthetics, and improved retention and stability¹⁴. The National Foundation for Ectodermal Dysplasia recommends the placement of dental implants in the anterior mandible of children older than school age (7 years and older)⁸. Fixed prosthesis and implant placement as a definitive, comfortable and aesthetic solution for patients with ectodermal dysplasia, could be taken into account once the skeletal growth is completed. Periodic dental recall of patients with ED should be done at regular intervals to be able to monitor the patient's growth and development and consequently adjust or replace the prosthesis accordingly. Vergo recommended 15 relining/rebasing an intraoral prosthesis in a growing patient every 2-4 years and remaking a new prosthesis every 4-6 years. Treatment should be effective in both functionally and aesthetically.

CONCLUSION

The child was successfully rehabilitated in terms of function and esthetics. He was able to eat the food with soft and medium consistency and was happy with his esthetics. Patients with ectodermal dysplasia require a multidisciplinary strategy for treatment planning and dental treatment to restore social well-being, esthetics and function. Provision of removable dentures is a rational, feasible and economic option at this age of the patient.

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