

## SAINT OR SINNER ??? -A COMPLETE REHABILITATION OF A FULL BLOWN CONDITION OF ECTODERMAL DYSPLASIA: CASE REPORT

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### ABSTRACT

Ectodermal dysplasia consists of clinically and genetically heterogeneous groups of disorders characterized by absence of or incomplete or delayed development of one or more of the appendages derived from epidermal tissue (hair, sweat gland, teeth, skin, and nails) or of oral ectodermal origin during embryogenesis. A case of a seven year-old child with full blown condition of ectodermal dysplasia is presented. Common dental, oral, and physical conditions were taken into consideration. Clinical management consisted of an overdenture to improve psychological development and to promote better functioning of the stomatognathic system.

**KEYWORDS:** Child, ectodermal dysplasia, prosthetic treatment.

### INTRODUCTION

*“Every child comes with a message that God is not yet discouraged of man” – Tagore.*

The most beautiful creation is the innocent smile blossoming on the face of a young child. Eric Erickson has coined a wonderful term in child psychology to illustrate the child's behavior “d'terrible two's” – Little devil & little angel. Although one side of coin of God's creation can be written with exclamatory mark, the other side, we have to look for, which make us to raise queries??. One such query is a condition thrown with poignant smile is ectodermal dysplasia( ED).

ED is a congenital, diffuse and non-progressive disease. It is a large group of inherited disorders characterized by the primary defect in hair, teeth, nail or sweat gland function in addition to another abnormality in the tissue of ectodermal origin. E.g.: ears, eyes, lips, mucous membrane, mouth or nose<sup>1</sup> Their incidence is estimated to be 1/100000 births<sup>2</sup>. In children with ED, the appearance of the teeth is extremely important because it can affect the patients self esteem<sup>3</sup> which creates challenges for the pediatric dentist<sup>4</sup>. A complete oral rehabilitation of these patients is of great value from the functional stand point as well as for physiologic and psychosocial reasons. The present report describes

the complete rehabilitation of a young child with a full blown condition of ED.

### Case Report:

A 7 year old girl reported to the Department of Pedodontia of J.K.K.Nataraja Dental College, Komarapalayam with missing teeth, speech problems, mastication difficulties which resulted in a restricted diet and esthetic concerns. Her mother gave a history of frequent bout of fever and intolerance to heat and take dips in summer to stay cool. She was moderately built and poorly nourished. Her skin was dry and body hair was scanty, prominent forehead and ears, protuberant and averted lips. The nails were normal (Fig.1 to Fig.3). Intra oral examination revealed severe hypodontia in both deciduous and permanent dentition. In the maxillary arch 1st and 2nd right deciduous molar and 1st left deciduous molar were seen. In the mandibular arch only single deciduous peg lateral was found (Fig.4 and Fig.5). Radiographic investigation (OPG) showed tooth germ of two maxillary central incisors (Fig 6). The family pedigree (Fig.7) was studied and an autosomal recessive character of the 5th generation was identified in the young girl and her young brother. Her brother revealed similar features (Fig 8) of ED. Since hypotrichosis, hypodontia and hypohidrosis were evident, she was diagnosed with ED.



Fig 1. Frontal view



Fig 2 . Lateral view



Fig 3. Normal nails



Fig 4. Intra oral view (Maxillary arch)

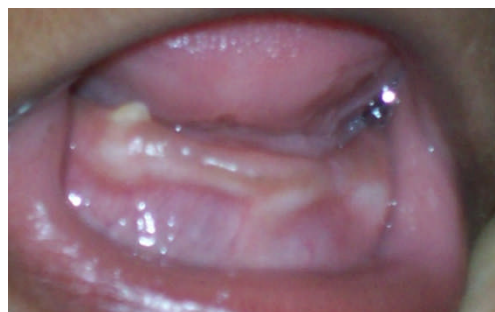


Fig 5. Intra oral view (Mandibular arch)

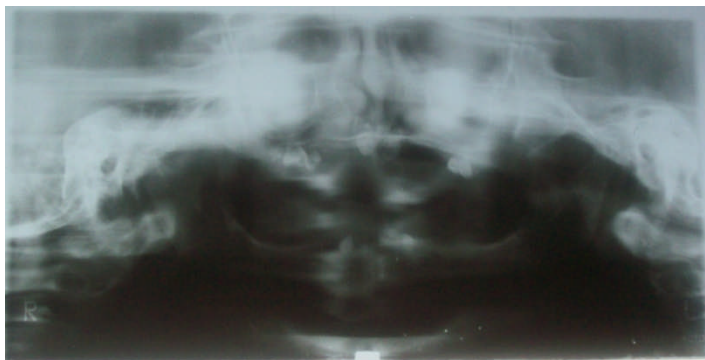


Fig 6. Panoramic Radiograph

**Prosthetic therapy:**

To avoid extraction and to maintain the alveolar bone height, we planned to retain the deciduous teeth till the normal exfoliation occurs and thus she was provided with an overdenture. The basic principles of prosthodontic treatment for children have been reported<sup>(5,6)</sup> and were followed in the case presented here. Preliminary impression was made using alginate. Special trays were fabricated using auto polymerizing resins after applying spacer of 2 mm in the region of retained

teeth to allow for the physiologic eruption of teeth and the undercuts were blocked. Border moulding was done using green stick compound. Secondary impressions were made using zinc oxide eugenol paste. Record bases (Fig 9) were made and jaw relations registered. Considering the age, inadequacy of arch length and to preserve the remaining teeth, teeth setting were meticulously made.

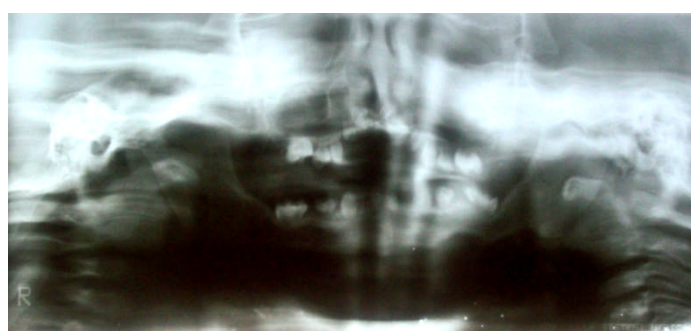
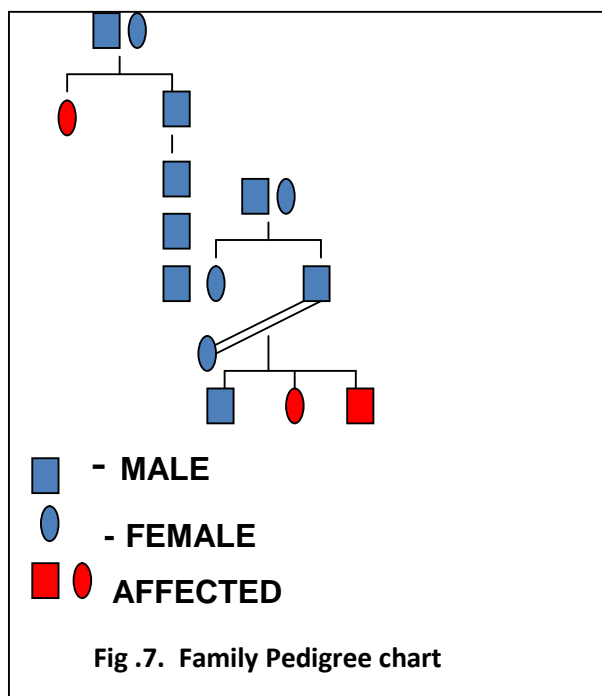


Fig 8 . Sibling of the patient with features of ED

Since the girl is 7 years old two upper and lower permanent central incisors were placed and due to inadequacy of the arch length the 1st permanent molar was not placed in both the arches. Acrylic dentures were processed(Fig 10) through compression moulding and polished dentures were inserted (Fig.11). After the final insertion routine oral hygiene instructions for the

overdenture were given to the patient and her parents.

**Complete rehabilitation – The treatment journey.....**During the initial visit, the child was reluctant to accept the treatment. First, we counseled the parents about the importance of the treatment not only to enhance her appearance but the overall wellbeing of the child. Her first oral examination was done in an isolated special need set up because of her timid, reluctant behavior. Then during the first week we trained her slowly in our dental setting. Now she was placed in the dental chair and treated as usual with the other children.

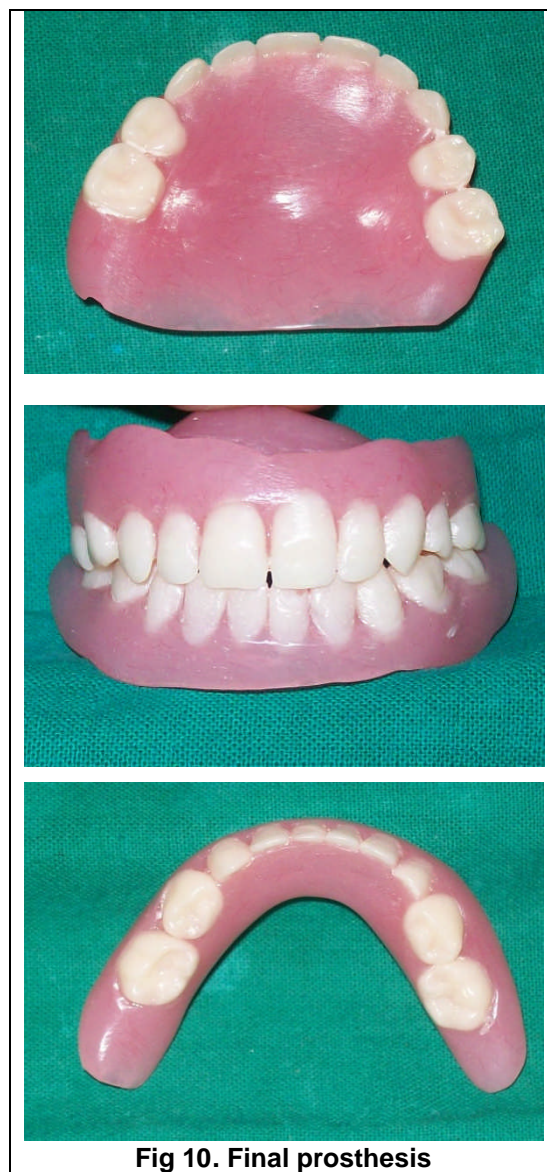


**Fig. 9. Bite Rim**

Later she became the model for the other children with potentially cooperative behaviour and she herself seemed to be anxious to improve her appearance and her positive attitude enhanced her compliance with the treatment. Immediately after the denture she was reluctant to use it, so to put her into ease we made her to wear the denture two to three hours /day. Later her frequency of using the denture had been slowly increased. Then she got used to the denture, now she can't stay without the denture. The food items such as meat, apples and biscuits which she avoided prior because of her chewing inefficiency had become her favorite item now. Initially she was kept under weekly recalls, then on monthly recalls. The fluoride application was done on the retained teeth to prevent plaque accumulation and caries. After three months her body weight was compared with the body weight ( 12 Kgs) which was taken on the day of her first visit to us. It was significantly increased by two Kgs. (15 kgs) Since tooth germs of 11 & 21 are seen in the OPG, constant monitoring is needed for which we are planning later to create a window to allow the path of eruption of 11 & 21. The child has come out of the social stigma and she is able to move around with the peer group comfortably. Her parents were happy with the significant improvement not only in her oral health but her general health as well. Now they are eagerly involved in the next journey of her younger brothers well being. This treatment journey as a whole resulted in the overall improvement in the growth and development And a good psychological self image in the child as well as her parents.

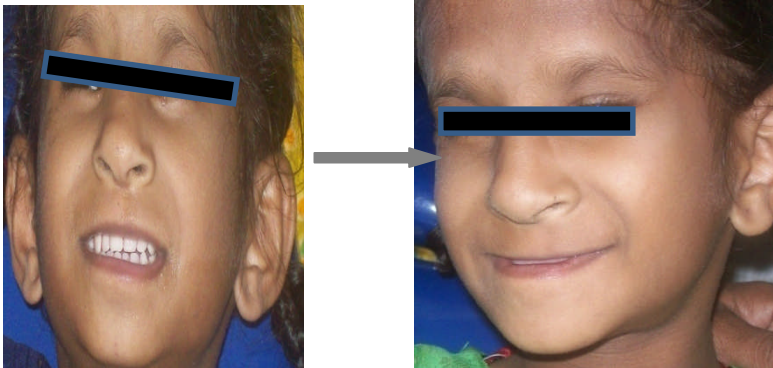
**Discussion:**

The treatment plan of a child with ED not only includes complete oral rehabilitation but also psychological boost up<sup>7</sup>. The common dental treatments in cases with ED are conventional dentures, overdenture and removable prosthesis. Patient with ED presents a thin underdeveloped ridge and decreased flow rate are the main problems encountered in treating such cases.<sup>8</sup> New alternatives for rehabilitation with ED, such as the use of implants, must be carefully considered, taking into account the presence of underdeveloped thin alveolar bone and age.



**Fig 10. Final prosthesis**

Fig 11. A poignant smile to confident smile



There was no definite time to begin the treatment but *Till and Marques*<sup>9</sup> recommended that the initial prosthesis could be delivered when the child starts school, so that the child would have a better appearance and will also have the time to adapt to the prosthesis. Usually by the time the child reaches the school age, he or she is old enough to recognize the esthetic handicap and is willing to cooperate. In this case initial molding was the key in determining the patient's cooperation as she wanted to have a better appearance. Periodic recall of young patients with ED is also important, because prosthetic modification or replacement will be need as a result of continuing growth and development<sup>10</sup>

#### CONCLUSION:

Dental treatment as a phase in the management of ED is essential. The first goal of the treatment therapy is to meet the immediate needs of the young patient, which includes mastication, esthetics, speech development and improvement of psychological factors. To obtain successful outcome, there needs to be good communication between the professionals in an interdisciplinary team and the parents.

.....the smile of a child can bring light even to the darkest corners and crack open even the sturdiest of all, yet when the special smile is marred by something despairing as a poignant smile, one cannot but wonder whether God has forsaken us after all.

We as pediatric dentist cannot blame ourselves for not preventing this but provide them with better facility and do what men/women of science would do believing that these children are neither a Saint nor a Sinner but a Human being after all.

Being in the hands of us, these children are given life to enlighten their smile.....

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