

Prosthetic Rehabilitation Of A 7-Year-Old Child With Ectodermal Dysplasia: A Case Report

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Abstract

Ectodermal Dysplasia is a developmental defect which affects one or more ectodermal structures including teeth, hair, nails, and sweat glands. There are clinical and oral manifestations to it. The clinical manifestations include loss of hair, sparse hair or thin and dry hair. The most common oral manifestation is hypodontia, which demands the patients for prosthetic need. The term 'oligodontia' is the condition whereby ≥ 6 permanent teeth are agenetic excluding third molars. The present case report describes the prosthetic rehabilitation of a 7-year-old child with ectodermal dysplasia thereby restoring function, improving esthetics, and imparting psychological assurance to the child.

Keywords: Ectodermal Dysplasia, Hypodontia, Oligodontia, Complete Denture, Prosthetic Rehabilitation.

Date of Submission: 06-07-2023

Date of Acceptance: 16-07-2023

I. INTRODUCTION

Ectodermal Dysplasia was defined by the Ectodermal Dysplasias Classification Working Group as a group of genetic conditions that affect the development and/or homeostasis of two or more ectodermal derivatives, including hair, teeth, nails, and some glands; their genetic causes and clinical phenotypes are heterogeneous. (1) The prevalence of ED ranges from approximately 1:10,000 to 1:100,000 worldwide, and it mostly affects males. (2)

Oral manifestations include partial or complete anodontia, abnormal shape of the teeth, enamel hypoplasia, reduced asymmetric alveolar ridge height, maxillary retrusion, and high palatal arch. Absence of teeth may cause masticatory difficulties, nutritional deficiencies, speech problems, and compromised facial appearance. (2)

The functional and aesthetic rehabilitation of these patients is a challenge for dentists. Early treatment during childhood is essential to solve the problem of multiple missing teeth and enhance the growth of the jaws to achieve better oral function and facial aesthetics. (1) Removable (partial) dentures are usually a well-accepted treatment offering good aesthetic outcomes and functional mastication even though they have to be replaced every 3.5–4 years. (3)

II. CASE PRESENTATION

A 7-year-old male patient was brought to the department of Pediatric and Preventive Dentistry by his mother with the chief complaint of missing teeth in the mouth. Only a few teeth were present in the upper jaw.

Medical history revealed dryness of skin and absence of sweating since birth. There was a h/o multiple raised lesions over cubital fossa, popliteal fossa, dorsum of hand and feet. There was reduced hair growth since birth. The parent gave a history of child showing intolerance to heat. There was a h/o crusted nasal septum and multiple episodes of epistaxis in the past 5 years.

Family history revealed similar condition in maternal uncle.

On taking dental history, parent reported absence of multiple primary teeth.

On general examination the child was presented with sparse hair growth on the head and eyebrows. Hair showed less pigmentation and reduced thickness. There was a generalized dryness of skin. There were multiple discrete and confluent skin-colored papules over cubital fossa, popliteal fossa and posterior aspect of neck.

On extraoral examination of head and neck, features included frontal bossing, pursed lip, peaked nose and depression of nasal bridge. Nails were normal. (Figure 1)



Figure 1

On intraoral examination, the teeth present in the maxillary arch included 16, erupting 26, and grossly decayed 55 and 65. The mandibular arch was edentulous. (Figure 2)



Figure 2

OPG was advised to find out whether any other permanent teeth were developing. But the radiographic findings were in accordance with the clinical findings. Only 55,16,65 and 26 were present in the oral cavity. (Figure 3)



Figure 3

The case was provisionally diagnosed as Ectodermal Dysplasia. The treatment plan was extraction of grossly decayed primary teeth followed by prosthetic rehabilitation.

Extraction of grossly decayed primary teeth were done. Removable partial denture for maxillary arch and complete denture for mandibular arch were fabricated in the following steps.

Impression was made using elastomeric impression material and the cast was poured in dental stone.

Jaw relation records were taken. The upper and lower occlusal rims were sealed and mounted on the articulator. Adult teeth of lightest and smallest size were selected.(Figures 4,5,6)



Figure 4

Figure 5

Figure 6

Try in was done. Following that, upper RPD and lower complete denture were inserted and instructions were given. (Figure 7,8) The patient was recalled after 24 hours for any necessary corrections. Regular followup was advised as the denture might require refabrication and customization in the future to suit the esthetic needs of the patient.



Figure 7

Figure 8

III. DISCUSSION

Ectodermal dysplasia is a developmental defect which affects one or more ectodermal structures including teeth, hair, nails, and sweat glands. There are clinical and oral manifestations to ED. The clinical manifestations include loss of hair, sparse hair or thin and dry hair. The child become prone to rash skin. In addition, thick or thin, abnormally shaped or ridged nails were noted.

The most common oral manifestation is hypodontia, which demand the patients for prosthetic need.(4) Tooth agenesis or hypodontia refers to cases where one or more teeth fail to develop. In its most severe presentation, no teeth are present at all (anodontia). The term 'oligodontia' is the condition whereby ≥ 6 permanent teeth are agenetic excluding third molars.(5)

The present case is a case of oligodontia. The main goal of treatment for this child was to extract the grossly decayed primary teeth to eliminate the source of infection and prosthetic rehabilitation of missing teeth to restore function, improve esthetics, and imparting psychological assurance. In the initial visits, the child was classified as Frankl's negative behavior. Non pharmacologic behavior management techniques including communication, tell-show- do, modelling and voice control were used during the treatment. In the subsequent visits, the child started cooperating with the treatment.

The treatment was accomplished within six visits. By the time the child came for denture insertion, he showed an improvement to a definitely positive behaviour.(Figure 9) During the recall visit, the child became well adapted to the denture. The parent reported improvement in mastication, speech as well as social behavior of the child.



IV. CONCLUSION

The most common oral finding in patients with Ectodermal dysplasia is Hypodontia/anodontia. The treatment of concern from a pediatric dentists' point of view is definitely prosthetic rehabilitation and thereby improving their function, esthetics and social living.

REFERENCES

- [1]. Cerezo-Cayuelas M, Pérez-Silva A, Serna-Muñoz C, Vicente A, Martínez-Beneyto Y, Cabello-Malagón I, Et Al. Orthodontic And Dentofacial Orthopedic Treatments In Patients With Ectodermal Dysplasia: A Systematic Review. *Orphanet J Rare Dis* [Internet]. 2022;17(1):1–18. Available From: <https://doi.org/10.1186/s13023-022-02533-0>
- [2]. Alnuaimi R, Mansoor M. Prosthetic Rehabilitation With Fixed Prosthesis Of A 5-Year-Old Child With Hypohidrotic Ectodermal Dysplasia And Oligodontia: A Case Report. *J Med Case Rep*. 2019;13(1):1–6.
- [3]. Kratochvilova L, Dostalova T, Schwarz M, Macek M, Marek I, Malíková M, Et Al. Ectodermal Dysplasia: Important Role Of Complex Dental Care In Its Interdisciplinary Management. *Eur J Paediatr Dent*. 2022;23(2):140–6.
- [4]. Kassar WM. Dental Rehabilitation Of A Child With Ectodermal Dysplasia: A Case Report. *Int J Clin Pediatr Dent*. 2019;12(4):362–5.
- [5]. Filius MA, Cune MS, Raghoobar GM, Vissink A, Visser A. Prosthetic Treatment Outcome In Patients With Severe Hypodontia: A Systematic Review. *J Oral Rehabil*. 2016;43(5):373–87.