Content available at: https://www.ipinnovative.com/open-access-journals

IP Annals of Prosthodontics and Restorative Dentistry

Journal homepage: https://www.aprd.in/

Case Report

Smiles reimagined: The transformative power of overlay complete dentures in prosthodontic rehabilitation of ectodermal dysplasia patient

Tapan Kumar Giri[1], Bidhan Chandra Mondal[1], Palak Khanna[1]*, Tridib Banerjee[1], Debanti Giri[2]

[1]*Dept. of Prosthodontics, Dr. R Ahmed Dental College and Hospital, Kolkata, West Bengal, India*
[2]*Dept. of Oral and Maxillofacial Medicine and Radiology, Dr. R. Ahmed Dental College and Hospital, Kolkata, West Bengal, India*

ARTICLE INFO

Article history:
Received 02-10-2023
Accepted 16-11-2023
Available online 04-12-2023

Keywords:
Ectodermal dysplasia
Anodontia
Overlay dentures

ABSTRACT

Ectodermal dysplasias (EDs) encompass a diverse set of conditions characterized by developmental abnormalities affecting ectodermal tissues. Hypohidrotic ectodermal dysplasia is a congenital syndrome marked by features such as sparse, delicate hair, skin and nail anomalies, reduced sweating capacity (attributed to a deficiency in sweat glands, resulting in sweat-related issues), and underdeveloped or missing teeth (partial or complete absence of primary and/or permanent dentition). In most instances, individuals with EDA can expect a typical lifespan and exhibit normal cognitive abilities. Nonetheless, the absence of sweat glands poses a risk of overheating, potentially leading to brain damage or even fatality in infancy if not promptly recognized. Therefore, early diagnosis holds significant importance. This case report presents a documented instance of ectodermal dysplasia, complemented by a comprehensive literature review.

1. Introduction

Ectodermal dysplasia (ED) is a genetic disorder characterized by congenital abnormalities affecting two or more ectodermal structures, which can include skin, hair, nails, teeth, nerve cells, sweat glands, and parts of the eye and ear, among others. There are 20 common types of ED, each with varying degrees of severity. The diagnosis of ED can be challenging due to the diversity of types and the range of abnormalities present in affected individuals. Identifying the specific components of the disorder is crucial for providing appropriate treatment and improving the quality of life for ED patients. Understanding the genetic inheritance patterns is also important for counseling parents of affected children and predicting the risk of future offspring being affected.[1]

ED is caused by defective genes, which can be inherited from one or both parents or result from gene mutations. Any ectoderm-derived structure can be affected in ED, and the severity of the disorder varies from person to person. Common features in affected children include lightly pigmented and thin skin with visible blood vessels, especially around the eyes and on the elbows, palms, and soles. The skin is typically dry, scaly, and prone to irritation due to poorly developed or absent oil glands. Treatment for dry skin involves daily bathing with specialized soap.[2]

Sweat glands may be absent or nonfunctioning, leading to an increased risk of elevated body temperature, especially during illness, physical activity, or hot weather. Preventive measures include limiting physical activity in warm conditions, increasing fluid intake, and appropriate clothing choices. Overheated children can benefit from a lukewarm or sponge bath to lower body temperature.[3]

* *Corresponding author*.
E-mail address: pkhannaji@gmail.com (P. Khanna).

https://doi.org/10.18231/j.aprd.2023.046

Hair on the scalp, eyebrows, eyelashes, and body may be absent, sparse, fine, or abnormal in texture. Fragile and unruly hair is common due to the lack of oil glands. Treatment options include gentle shampoos and wigs for severe cases of hair loss. Fingernails and toenails may also exhibit abnormalities, and treatment involves keeping them lubricated and seeking medical attention for persistent fungal or yeast infections.[4]

One of the most common issues in ED is the absence or abnormal development of teeth, often associated with cleft lip and palate. Treatment options include removable or fixed partial dentures, complete denture prostheses, and implant-retained prostheses as needed. In cases involving cleft lip and palate, a combination of plastic surgery, oral and maxillofacial surgery, and maxillofacial prosthetics may be necessary.

2. Case Presentation

A 15 -year-old female reported to the department of prosthodontics and crown and bridge (Dr. R. Ahmed dental college and hospital, Kolkata, west bengal, India) with a chief complaint of missing upper and lower teeth since birth. Patient gives no history of exfoliation of teeth but gives a history of delayed eruption of teeth (at the age of 2yrs only). Patient also complains of dry skin and absence of sweat in his skin. she is intolerable to withstand hot water and hot environment. Patient gives history of dry mouth and difficulty in swallowing. On general examination, the patient was well oriented with time, place and was cooperative. On examination extra orally patient had scanty hair in eyebrows and eyelashes (Figures 1 and 2). There was frontal bossing and depressed ala of nose (Figure 1). Her skin appeared soft, dry in face, upper and lower limbs. Nails appear normal. Scanty eyebrows were seen. Areas of hyperpigmentation were seen around ala of nose(Figure 2). Intra orally patient was partially edentulous teeth present were 13,16,23,26 appears conical (Figure 3). Upper and lower alveolar ridges appear normal, lower ridge is knife edged (Figure 3). Her panoramic radiograph was relevant to the above findings (Figure 4). So with the above clinical findings we came to an diagnosis of Hypohidrotic ectodermal dysplasia.

2.1. Prosthodontic rehabilitation

Canines were labially placed and required modification for accommodation of artificial teeth over it. They were endodontically treated before modification of form (labial reduction and undercut removal)(Figure 4). Primary impressions were made with irreversible hydrocolloid impression material(Figure 5) as it is comfortable and can be easily removed from undercut areas. Casts were prepared with type III dental stone. Custom trays were prepared, and border moulding was done with low fusing green stick impression compound. Tray adhesive was applied after spacer removal and scraping of borders. The final impressions of the maxillary and mandibular arches were made with addition silicone light body impression material(Figure 6). Maxillo-mandibular relation was recorded, and the master casts were mounted on a semiadjustable articulator (mean value articulator) (Figure 7). The teeth were arranged according to a balanced occlusal scheme [Figure 8]. Try-in was done (Figure 9) and after careful evaluation prosthesis was fabricated in the conventional heat cure acrylic resin (Figure 10). The dentures were then inserted in the patient's mouth and adjustments were done using pressure indicating paste (Figures 11 and 12).

Table 1: Differences between hydrotic and hypohydrotic type of ectodermal dysplasia

	Hypohydrotic/ anhidrotic ectoderamal dysplasia	**Hydrotic ectodermal dysplasia**
Also called as	Christ-Siemens-Touraine syndrome	Clouston syndrome
Mode of inheritance	X-linked recessive	Autosomal dominant
Prevalence	more	less
Scalp hair	Fine, stiff and short	Soft, dawny and dark coloured
Dental anomaly	Hypodontia / anodontia	Hypodontia/ anodontia
Sweat glands	Reduced / absent	Not much affected
Nails	No abnormality	Dystrophic nails
Nasal bridge	underdeveloped	No flattening
Lips	protruding	normal
Eyelashes / auxiliary hairs	Variably affected	Scanty / absent

3. Discussion

Hereditary ectodermal dysplasia refers to the abnormal development of one or more structures originating from the ectoderm layer. This condition was first described by Thurnam in 1848, and the term "ectodermal dysplasia" was coined by Weech in 1929. Typically, it follows a recessive inheritance pattern, which means that it is more prevalent and severe in males compared to females. Freire-Maia characterized it as a developmental defect that affects the ectoderm during embryonic development, subsequently impacting tissues and structures derived from it. This condition influences the development of keratinocytes, resulting in abnormalities in various body parts such as hair, sebaceous glands, sweat glands, nails, teeth, lenses and conjunctiva of the eyes, anterior pituitary gland, nipples, and ears.[5,6]

Figure 1: Side profile of the patient before rehabilitation

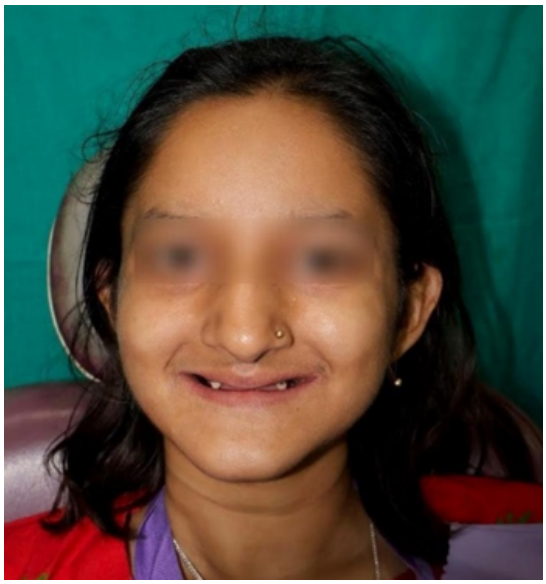

Figure 2: Facial profile of the patient before rehabilitation

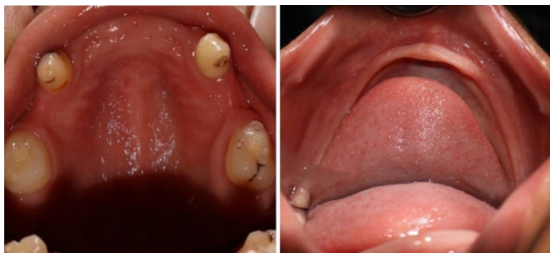

Figure 3: Maxillary and mandibular arch

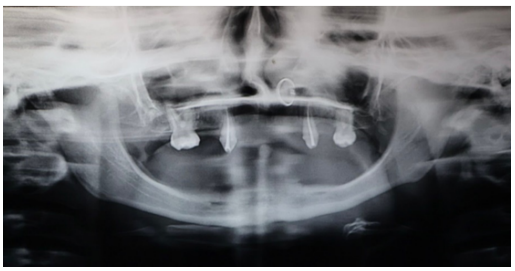

Figure 4: OPG- X-ray

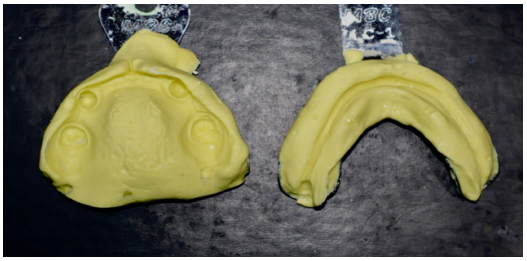

Figure 5: Impressions taken with irreversible hydrocolloid impression material

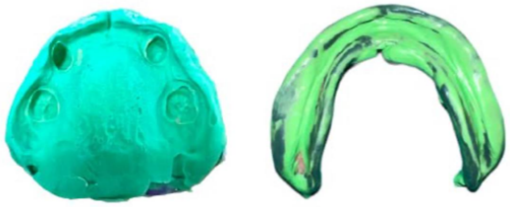

Figure 6: Final impressions

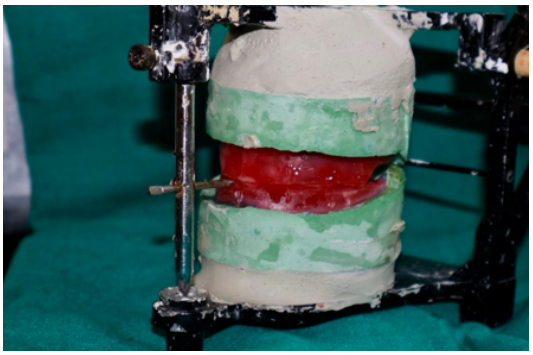

Figure 7: Jaw relation record taken with maxillary and mandibular record bases

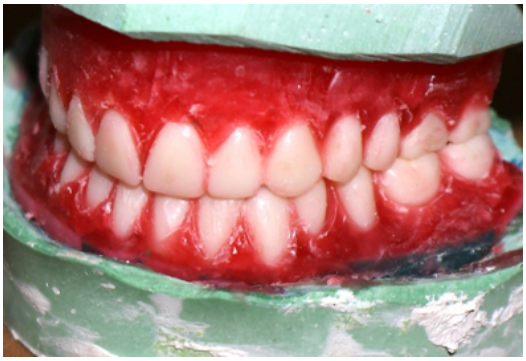

Figure 8: Teeth setting

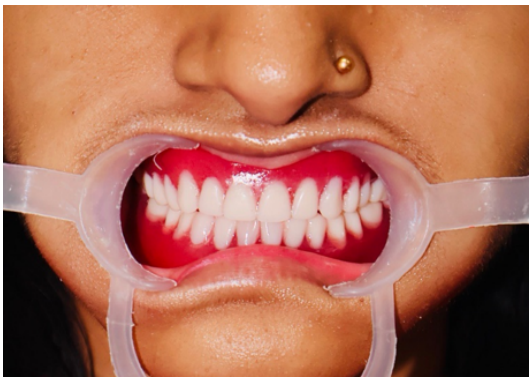

Figure 9: Try-in

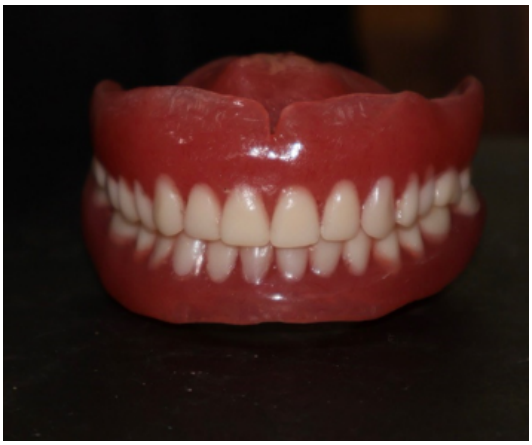

Figure 10: Processed denture

There are two primary clinical variants of ectodermal dysplasia: the hypohidrotic form, also known as Christ-Siemens-Tourian Syndrome, and the hidrotic form, referred to as Clouston syndrome. The hypohidrotic form typically presents with a triad of symptoms: reduced sweating (hypohidrosis), sparse hair (hypotrichosis), and missing teeth (hypodontia). This variant is often inherited as an X-linked recessive trait, resulting in more severe effects in males and milder ones in females. In contrast, the hidrotic

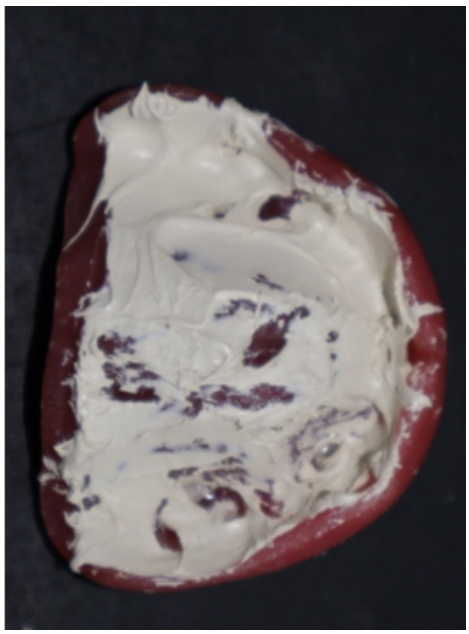

Figure 11: Denture adjustment with the help of pressure indicating paste

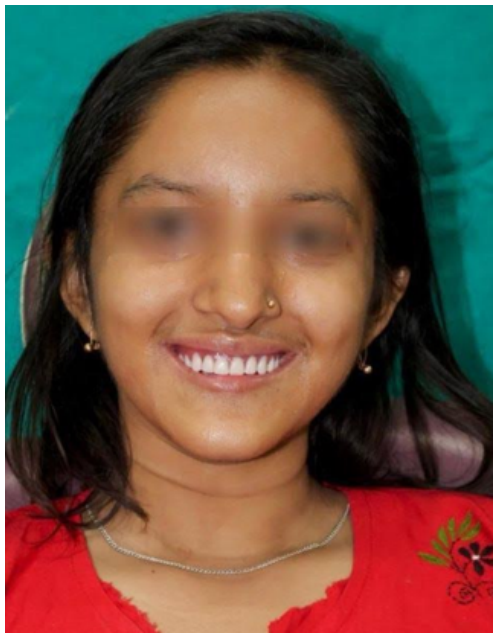

Figure 12: Facial profile of the patient after prosthodontic rehabilitation

form affects teeth, hair, and nails, while the sweat glands are usually unaffected. It is usually inherited as an autosomal dominant trait, although autosomal recessive inheritance patterns have also been reported.[7]

Hypohidrotic ectodermal dysplasia is characterized by thin, dry skin with either partial or complete absence of sweat glands, leading to an inability to perspire, heat intolerance, and frequent episodes of hyperthermia. The sebaceous glands are also often defective or missing. Hyperkeratosis (thickening of the skin) is observed on the palms and soles, and pseudorrhagades (wrinkles) may develop around the eyes. Newborns with this condition may have a distinctive "plastic wrap" appearance of the skin. In the hidrotic type, sweat glands develop partially, with the formation of ducts but the absence of secretory coils, resulting in fewer sweat pores. Scalp hair is typically fine, stiff, and short, while eyelashes and eyebrows are often sparse or absent. Facial features associated with ectodermal dysplasia include a prominent forehead, depressed nasal bridge, pronounced supraorbital ridges, prominent and obliquely set ears, a depressed midface, a small lower third of the face due to underdeveloped alveolar bone, and protruding lips. Oligodontia, the congenital absence of teeth, is a prominent feature in the oral cavity, with the present teeth often displaying a conical crown shape. In some cases, individuals may be missing both primary and permanent dentition.[8]

4. Conclusion

The characteristics of ectodermal dysplasias have been outlined, with particular attention to the dental challenges linked to this congenital or developmental disorder. Timely dental treatment can enhance the patient's aesthetic appearance and reduce the emergence of emotional and psychosocial difficulties frequently encountered by individuals with ectodermal dysplasias.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Pigno MA, Blackman RB, Cronin-Jr RJ, Cavazos E. Prosthodontic management of ectodermal dysplasia: a review of the literature. *J Prosthet Dent*. 1996;76(5):541–5.
2. Bhargava A, Sharma A, Popli S, Bhargava R. Prosthodontic management of a child with ectodermal dysplasia: a case report. *J Indian Prosthodont Soc*. 2010;10(2):137–40.
3. Valjakova EB, Misevska CB, Stevkovska VK, Gigovski N, Ivkovska ASS, Bajraktarova B, et al. Prosthodontic management of hypohidrotic ectodermal dysplasia: a case report. *South Eur J Orthod Dentofacial Res*. 2015;2(2):20–6.
4. Gupta S, Tyagi P. Prosthodontic management of anhidrotic ectodermal dysplasia. *Indian J Dent Res*. 2011;22(2):348–51.
5. Ladda R, Gangadhar SA, Kasat VO, Bhandari AJ. Prosthodontic Management of Hypohidrotic Ectodermal Dysplasia with Anodontia: A Case Report in Pediatric Patient and Review of Literature. *Ann Med Health Sci Res*. 2013;3(2):277–81.
6. Nejabi MB, Anwari A, Shadab H, Mtawakel N, Omarzad F, Ahmadi ME, et al. Prosthodontic Management of a Patient with Ectodermal Dysplasia: Case Report. *Clin Cosmet Investig Dent*. 2023;15:133–41. doi:10.2147/CCIDE.S419939.
7. Hickey AJ, Vergo TJ. Prosthetic treatments for patients with ectodermal dysplasia. *J Prosthetic Dent*. 2001;86(4):364–8.
8. Pavarina AC, Machado AL, Vergani CE, Giampaolo ET. Overlay removable partial dentures for a patient with ectodermal dysplasia: a clinical report. *J Prosthet Dent*. 2001;86(6):574–7.

Author biography

Tapan Kumar Giri, Principal and Professor

Bidhan Chandra Mondal, Associate Professor

Palak Khanna, Post Graduate Trainee

Tridib Banerjee, Associate Professor

Debanti Giri, Associate Professor

Cite this article: Giri TK, Mondal BC, Khanna P, Banerjee T, Giri D. Smiles reimagined: The transformative power of overlay complete dentures in prosthodontic rehabilitation of ectodermal dysplasia patient. *IP Ann Prosthodont Restor Dent* 2023;9(4):248-252.