



## Prosthetic Rehabilitation of a Child with Anhidrotic Ectodermal Dysplasia: A Case Report and a Review of Literature

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### Abstract

**Patients:** A 13-year-old girl was referred to the paediatric dentistry and prevention department, hospital Rabta of Tunis, Tunisia with the chief complaint of unstable old prostheses.

According to clinical and radiographic manifestations, the patient was diagnosed with anhidrotic ectodermal dysplasia. The treatment began with composite resin restorations on the maxillary incisors followed by placing maxillary and mandibular removable partial dentures.

**Discussion:** Ectodermal dysplasia is a large group of rare congenital diseases affecting the ectodermal origin tissues including sweat glands, skin, nails, hair and teeth. The frequent type is anhidrotic-hypohidrotic ectodermal dysplasia known also as Christ Siemens Touraine syndrome. It is characterized by missing teeth, hypohidrosis or anhidrosis, hypotrichosis, conic shape of teeth, everted lips, flat nose and frontal prominence. Removable dentures are the most common treatment modality reported for dental management in children with ectodermal dysplasia in order to enhance their aesthetic appearance and to overcome their masticatory, deglutition and phonation problems.

**Conclusions:** Early dental management should be provided to children with anhidrotic ectodermal dysplasia. It should include preventive procedures, composite resin restorations and prosthodontic rehabilitation to improve their quality of life and increases their self-esteem.

**Keywords:** Ectodermal Dysplasia; Oligodontia; Paediatric Dentistry; Prosthetic Rehabilitation

### Abbreviations

DEA: Anhidrotic Ectodermal Dysplasia

### Introduction

Ectodermal dysplasia is a large group of rare congenital diseases affecting the ectodermal origin tissues, including sweat glands, skin, nails, hair, and teeth [1]. The estimated prevalence rate of ectodermal dysplasia is 1 in 100000 births and it is more frequent among boys than girls [2]. This condition exists in more than 170 forms. The most frequent forms are hidrotic and anhidrotic hypohidrosis which differ in sweat gland performance and hereditary pattern [1,2].

The anhidrotic hypohidrotic ectodermal dysplasia, known also as the Christ Siemens Touraine syndrome, is characterized by hypohidrosis or anhidrosis due to reduction or absence of sweat glands, hypotrichosis, with other features like everted lips, flat nose, and frontal prominence [3]. The oral manifestations consist of hyposalivation, missing teeth, conic shape of teeth, enamel hypoplasia, taurodontic primary second molars, and thin, reduced alveolar ridge height. Tooth abnormalities can affect the primary and permanent dentitions [4]. The affected children usually have a facial appearance of an old man. To enhance their aesthetic appearance and to overcome their masticatory and phonation problems, prosthetic rehabilitation is usually indicated [2].

The aim of this paper was to describe the clinical manifestations and dental management of a case of anhidrotic ectodermal dysplasia with a literature review of reported cases.

**Outline of the case**

A 13-year-old girl was referred to the paediatric dentistry and prevention department (hospital Rabta of Tunis, Tunisia), with the chief complaint of unstable old prostheses. Her past medical history revealed a heat intolerance. The extra-oral examination revealed a senile facial appearance, dry and light-colored hair with some alopecia areas, periorbital hyperpigmentation and wrinkles, sparse eyebrow and eyelash hair, frontal bossing, everted lips, concave facial profile, mandibular prognathism. Her skin was thin, dry, and slightly wrinkled, especially on the hands. The nails were normal but brittle (Figure 1). Intraoral examination revealed the presence of 6 teeth: 2 conic incisors, 3 upper molars, and 1 lower molar. The panoramic radiograph showed the presence of 16, 11, 21, 26, 27, and 37 with reduced alveolar ridge height (Figure 2). On the basis of clinical and radiographic features, the girl was diagnosed with anhidrotic ectodermal dysplasia with oligodontia. An examination of the primary casts alone then mounted on a semi-adjustable articulator was done to finalize the treatment. It revealed an asymmetric maxillary arch, a thin knife-edge alveolar ridge, and a cl III intermaxillary situation with mandibular prognathism. The vertical dimension of occlusion was determined by the 27 and antagonistic teeth 37 (Figure 3). The treatment began with an aesthetic reconstitution of the conic incisors 11 and 21 using composite resin followed by placing removable partial dentures. The occlusion record was done using denture bases and occlusal rims made on the working casts. After selecting the shade and teeth arrangement, the dentures were checked in the mouth of the patient. The dentures were then fabricated with heat-cured acrylic resin (Figure 4a). After the insertion of the removable partial dentures, an occlusal equilibrium was preceded and an improvement of the facial appearance and other oral functions were noted (Figure 4b,c). Oral hygiene and maintenance of dentures instructions were given to the patient and her mother.

**Discussion**

Anhidrotic ectodermal dysplasia (AED) is a genetic disease, diagnosed essentially with the identification of clinical characteristics such as hypotrichosis, common facial features, and dental abnormalities. A skin biopsy could be performed to investigate hypoplastic or absent sweat glands, and a genetic analysis could be used to confirm the diagnosis [2,4].

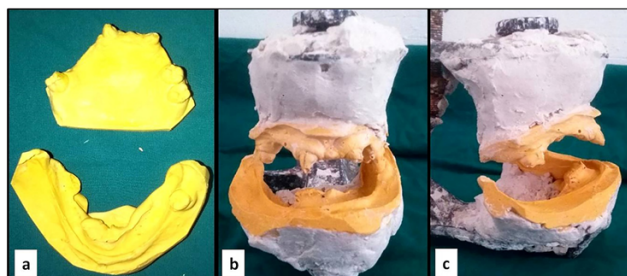
The abnormal appearance of children with AED could affect their physiologic development, self-confidence, and social integra-



**Figure 1:** Extraoral views showing characteristic features of ectodermal dysplasia. (a) Frontal view of the patient. (b) Profile view of the patient. (c) Areas of alopecia. (d) Dry wrinkled hand.



**Figure 2:** Panoramic radiograph.



**Figure 3:** (a) Examination of the primary casts. (b,c) Primary casts mounted on a semi-adjustable articulator.



**Figure 4:** (a) Removable partial dentures. (b,c) post-operative appearance.

tion [15,17]. Thus, the oral management of these patients should be provided at an early age. It should include preventive procedures, composite resin restorations, and prosthodontic rehabilitation [3,4].

This paper highlights the extraoral features and oral manifestations of AED in children as well as the importance of oral management and follow-ups in restoring function and enhancing aesthetics.

A review of the literature was conducted for the period from 2012 to 2023. Ten case reports and two case series were reported. The first case was published in 2012 and the latest was in 2021. Of the 12 patients, 10 were males. The age was between 2-10 years old. Removable partial dentures were used in 6 cases, overdentures and Implant-supported prostheses were used in 3 cases, and removable complete dentures were used in 2 cases (Table 1).

Authors	Year	Gender	Age (Years old)	Extraoral features	Intraoral manifestations	Prosthetic rehabilitation
Shigli and Sarkar [10]	2012	Male	10	hypohidrosis, hypotrichosis, prominent forehead, saddle nose, sparse scalp hair, missing eyelashes and eyebrows, protuberant lips	Oligodontia, knife-edge mandibular ridge	Maxillary and mandibular removable partial dentures
		Male	9	Sparse scalp and eyebrows hair, dry skin, normal nails, frontal and temporal prominence, saddle nose, large ears, protuberant lips, nose and periorbital hyperpigmentation	Oligodontia, thin maxillary and mandibular alveolar ridges, conic primary canines and first molars	Maxillary removable partial dentures Mandibular complete dentures
		Male	9	hypohidrosis, hypotrichosis, saddle nose and everted lips	Oligodontia, dry mucosa, conic central and lateral incisors, mesio dens	Maxillary and mandibular Overdentures
Paulus and Martin [13]	2013	Male	2	Thin and dry skin, periorbital and peri-buccal wrinkles, hyperpigmentation periorbital, thin and sparse hair, commissural perleche, labial vertical defect	Oligodontia, conic canines	At the age of 2 years old: removable dentures  At the age of 6 years old: Implant-supported prostheses
Aydinbelge, et al. [12]	2013	Female	7	Hypohidrosis, hypotrichosis, prominent forehead, saddle nose, an extended philtrum, large ears, diminished lower facial height, sparse scalp hair, missing eyelashes and eyebrows, fragile and thin nails, protuberant lips	Maxillary oligodontia, Mandibular anodontia, conic central incisors, dry oral mucosa, reduced height and width of alveolar ridges	Maxillary overdenture, Implant-supported mandibular prostheses
Fraiz, et al. [3]	2014	Male	3	facial characteristics of ectodermal dysplasia	Oligodontia, abnormal shape of the primary maxillary incisors	Maxillary and mandibular removable partial dentures
Mello, et al. [14]	2015	Female	9	Reduced vertical dimension, senile facial appearance, dry skin, sparse scalp hair, missing eyelashes and eyebrows, hypohidrosis	Oligodontia, xerostomia; maxillary anterior diastema, conic incisors	Mandibular implant-supported prosthesis
Maroulakos, et al. [15]	2015	Male	2.5	Fine sparse hair, scant eyelashes and eyebrows, dry skin, flattened facial appearance, pseudo-class III jaw relationship	Hypodontia Conic maxillary central incisors, slightly hypoplastic right maxillary primary canine underdeveloped alveolar ridges	Maxillary and mandibular removable partial dentures
		Male	4	Aged profile, thin and dry skin, sparse fine hair, slight eyebrows and eyelashes, reduced lower face height	Oligodontia, conic canines, underdeveloped alveolar ridges	Maxillary and mandibular overdentures
Pinto, et al. [16]	2016	Male	6	Dry skin, sparse hair, normal nails, saddle nose, frontal bossing, prominent lips	Hypodontia, conic teeth, ogival palate	mandibular removable partial dentures

AlNuaimi., <i>et al.</i> [1]	2019	Male	5	Periorbital discolouration, sparse scalp, eyebrow and eyelash hair, flat nose, dry lips, retrognathic maxilla, concave facial profile, prominent chin, reduced lower facial height	Oligodontia, deep overbite, knife-edge alveolar ridges, enlarged tongue, dry oral mucosa	Nance space maintainer with a saddle in the maxilla 8-unit ceramic bridge in the mandible
De Castro., <i>et al.</i> [17]	2021	Male	3	Hypohidrosis, hypotrichosis, saddle nose, prominent lips	Anodontia, reduced bone height, thin alveolar ridges	Removable complete dentures
Current case	2023	Female	13	Senile facial appearance, sparse dry and light-coloured hair, sparse eyebrow and eyelash hair, thin dry and wrinkled skin, brittle nails, periorbital hyperpigmentation and wrinkles, frontal bossing, everted lips, concave facial profile, mandibular prognathism	Oligodontia, conic incisors, thin and reduced alveolar height ridge	Removable partial dentures

**Table 1:** Reported cases in English literature using prosthetic rehabilitation.

It has been reported that initial prosthodontic treatment should be provided at preschool age [5]. In fact, many authors recommend using prostheses at the age of 3-4 years old in order to improve masticatory deglutition and phonation functions, induce basal bone growth, and adjust the vertical dimension. Thus, we could achieve aesthetic appearance amelioration, and self-esteem improvement in the child [6-8,11]. Removable complete dentures, removable partial dentures or overdentures are the most common treatment modality reported for dental management because they can be easily adjusted and adapted to jaw growth [2]. In a systematic review, it has been reported that in children with oligodontia, removable partial dentures with clasps are the classic restoration [9]. However, some authors reported difficulty to find stability and retention of the complete and partial dentures, due to hyposalivation, oral mucosa dryness, and alveolar ridge resorption [3]. Thus, overdentures could be a good treatment option as it increases the retention of various attachments to the presented teeth [11]. Nowadays, the using of dental implants in ectodermal dysplasia children is the common treatment option [1]. The placement of implants in children aged 3 years old was reported [12]. As the anterior mandible completes its lateral growth at the age of three, and the symphyseal suture transversal growth ends at the age of 3 months, the dental implant can be placed safely [2,13]. However, the placement of implants in the maxilla should be contraindicated because it can affect transverse growth, which is achieved at the same age as mid-palatal suture fusion [13]. The National Foundation for Ectodermal Dysplasia recommends using dental implants only in the anterior mandible of children aged at least 7 years old [2].

Children with AED must be monitored frequently to supervise the growth and development of the patient and consequently modify the prosthesis until skeletal development is attained [1,2]. It's recommended that every 2-4 years, a rebasing of the prosthesis should be done and every 4-6 years a new prosthesis should be fabricated [1].

In the present case, the characteristics of AED were well presented. Composite resin restorations of the maxillary incisors were done to ameliorate the conic shape, then maxillary and mandibular removable partial dentures were provided to replace the missing teeth, restore oral function and enhance the aesthetic appearance of the child. During follow-up, the patient reported an amelioration of her masticatory capabilities, dietary behaviour, and her self-confidence.

**Conclusion**

Early dental management should be provided to children with AED. It improves their quality of life and increases their self-esteem. So when treating those children, it's important to be well-informed about the development and growth process, psychological management, and aesthetic and functional rehabilitation.

**Conflict of Interest**

No conflicts of interest to declare.

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