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Case Report

Prosthetic Management of a Patient with Anhidrotic Ectodermal Dysplasia: Clinical Case Report

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Abstract

Background: The anhidrotic ectodermal dysplasia gathers a set of diversified pathologies characterized by the triad anhidrosis, hypotrichosis, anodontia (hypodontia); the management of young patients with this condition often requires a multidisciplinary approach within the scope of prevention, pediatric and prosthodontic dentistry.

Case Presentation: The following case describes the clinical steps of a prosthodontic rehabilitation of a 9 years-old patient affected by anhidrotic ectodermal dysplasia, complete dentures were made using a 3D conceived and printed Fox plan.

Conclusion: The prosthetic rehabilitation of these young patients will not only restore all the manducatory functions but also their psychomotor apparatus affected by the aesthetic repercussions of the ectodermal dysplasia.

Keywords: Ectodermic Anhidrotic Dysplasia; Removable Prosthesis; Oligodontia

Introduction

Ectodermal dysplasias (ED) were described for the first time in 1848 by Thurman. They represent a large and heterogeneous group of rare diseases whose common denominator is a genetic disorder inducing abnormalities in the morphogenesis of ectodermal organs and which are often associated with dental abnormalities [1].

According to Perabo., *et al*. the first case of ectodermal dysplasia could have been recorded as early as 1792 by Danz. In 1838, Wedderburn documented ectodermal dysplasia in a letter to Charles Darwin, describing the case of 10 Hindu family members. Thurman in 1848 reported 2 cases of hypo-hidrotic form. Similar cases were reported by Guilford and Hutchinson in 1883 and 1886 respectively. Weech, in 1929, introduced the term hereditary ectodermal dysplasia and suggested the term anhidrotic for people unable to sweat. Felsher, in 1944, changed the adjective to anhidrotic hypo-hidrotic, because the author agreed that people with a hypohidrotic form are not entirely devoid of sweat glands [2].

They include more than 200 clinical syndromes involving abnormalities of the skin, nails, hair, sweat glands, eyes, lungs, digestive

system, and teeth. The most common form (80% of ED) is X-linked anhidrotic ectodermal dysplasia, or Christ-Siemens-Touraine syndrome, whose incidence is estimated at 1/100,000 births. It finds its etiology in a mutation of the gene coding for ectodysplasin A (EDA) and is characterized by the triad hypodontia or anodontia, hypotrichosis, and hypohidrosis. The latter is responsible for disorders of thermoregulation that may compromise the prognosis of the young child during hyperthermic attacks. Once passed the first years, the vital prognosis improves then joins the general population's one [3].

The suggestive clinical presentation of children with ED includes several symptoms revolving around the triad nail dystrophy (onycho-dysplasia), alopecia or hypotrichosis (thinning hair, light and fine hair on the scalp and eyebrows), and palmoplantar hyperkeratosis is usually accompanied by a lack of sweat glands (hypohidrosis) and a partial or total absence of primary and/or permanent dentition; giving these young patients a typical "Elderly" facies. Mucosal involvement results in chronic rhinitis and conjunctivitis-like recurrent eye infections as well as epistaxis, chewing, swallowing, and phonation problems [4].

Concerning the stomatognathic system, the characteristics are:

- Anomalies in the number (default), shape, and structure of the temporary and permanent teeth;
- Diastemas;
- Disorders of maxillomandibular growth;
- An increased carious risk (oligoptyalism and weakened enamel).

Number abnormalities can be classified according to 3 categories: hypodontia (absence of fewer than 6 teeth), oligodontia (absence of 6 teeth or more), anodontia (total absence of teeth). Patients with ectodermal dysplasia have hypodontia in 80% of cases, usually more severe in the mandible than in the maxilla. The second temporary molar, if present, is frequently affected by taurodontism. Permanent incisors and canines are small, spaced, very conical or conoidal, sharp curved pointed, and very unaesthetic; the molars show sharp cusps. the alveolar edentulous ridge's form is a knife blade, the lower face height's growth is altered, the vermilion edge disappears and the lips are everted and prominent, the oral mucosa is dried and the child presents a pseudo-class III appearance. Children with ectodermal dysplasia should be given early and multidisciplinary management involving not only pediatric and prosthetic odontology but also genetics, otolaryngology, ophthalmology, dermatology, psychology, speech therapy, oral and maxillofacial surgery and dentofacial orthopedics [3,4].

The most common prosthetic treatments for dental management of ectodermal dysplasia are removable prostheses.

Since the development of alveolar bones depends on the presence of teeth, and the latter are present in small numbers in the case of ED, residual bone inheritance in children with this disease remains very limited; therefore, restoring function and appearance is more difficult than usual. Follow-up by a multidisciplinary team is considered the most appropriate approach in such cases [5].

This work aims to describe the prosthetic management of a patient with ectodermic anhidrotic dysplasia.

Case Report

Identification

The 9-year-old male patient, presenting anhidrotic ectodermal dysplasia, referred by the department of pedodontics and prevention in the Dental Consultation and Treatment Center (D.C.T.C) of Casablanca for aesthetic and functional rehabilitation, at first the patient seems calm, distant, and familiar with the university hospital's environment. The geological examination shows that the patient is the only member of the family affected by anhydrous ectodermal dysplasia the patient has only one brother whose general condition is normal.

The examination reveals that the patient is followed in the pedodontics and preventive service in which he has performed several dental care and extractions before benefiting two years ago from a partial denture in maxillary resin and a complete prosthesis to the mandible, the latter are no longer adapted to the new intraoral dimensions of the patient, modified by growth and oro-facial development.

Extra-oral examination (Figure 1)

The extra-oral examination shows an expansion of the cranial perimeter, increased by an excessive width of the frontal bone, contrasted by the reduction of the lower face height's; the external pavilion of the ears appears broad and falling.

The dryness of the ocular, nasal, and perioral mucous membranes is reflected by the presence of several zones of desquamation and the accentuation of the expression of the perioral grooves,

this chronic dermatological dryness is marked by areas of hyperpigmentation. Given the systematic involvement of the dander in the case of DE, the patient's hair and eyebrows are sparse and thin, the nails also appear streaked and fragile.

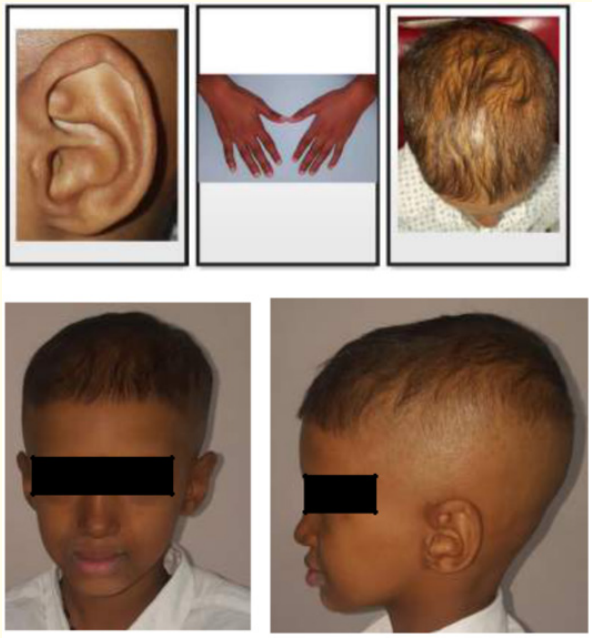

Figure 1: Extra-oral pictures face and profile of the patient.

Intraoral examination (Figure 2)

Intraoral examination shows xerostomia, the only residual teeth are 54,64 and 65. The maxillary arch is hyperbolic with residual ridges sufficient in height and width, while the parabolic mandible presents more severe bone resorption.

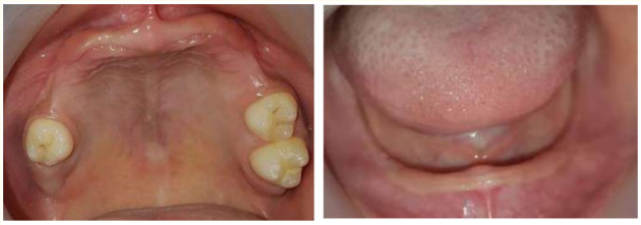

Figure 2: Intraoral photographs of the patient.

Treatment

Primary impressions

muco-static primary impressions were performed using an irreversible hydrocolloid {Alginate} (Figure 3), pedodontic plastic impression trays were tested and validated, the set time was reduced using warm water during mixing. After disinfection, these impressions were cast, and individual impression trays (IIT) in chemo-polymerizable resin were made in the laboratory (Figure 4).

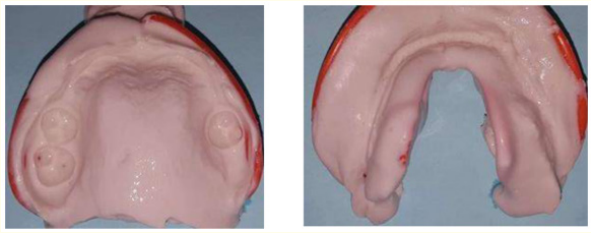

Figure 3: Primary impressions in alginate.

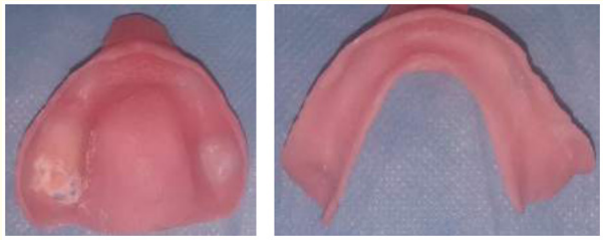

Figure 4: Individual resin trays.

Functional secondary impressions (Figure 5)

After the fitting and adaptation of the individual trays, functional secondary impressions were made in two stages, first Functional margins were formed with a thermoplastic material "Kerr® thermoplastic compound" under the patient's active movements; The Second functional impression was made by paste based on eugenol's zinc oxide "Impression Past®" in the mandible, while In the maxilla, given the persistence of the temporary molars, Thiocol (Permlastic Regular®) was chosen (Figure 5).

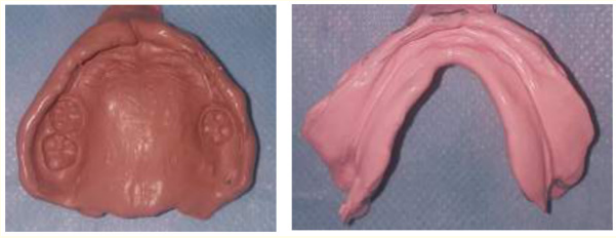

Figure 5: Functional secondary impressions.

The disinfected secondary impressions were sent to the laboratory to be boxed (Figure 6) and cast to make occlusion models (Figure 7).

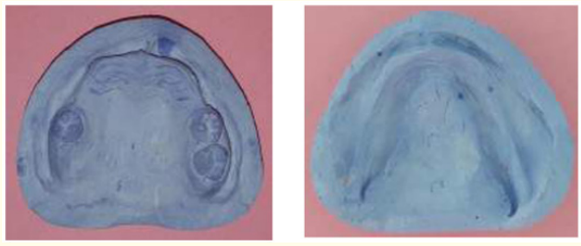

Figure 6: Secondary models.

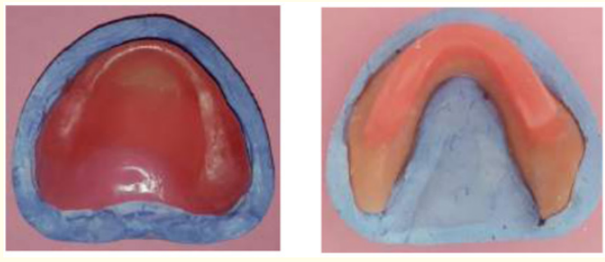

Figure 7: Wax rims.

The record of intermaxillary relations (Figure 9)

The adjustment of the occlusal plane began with the validation of adequate lip support, followed by verification of the parallelism of the different parts of the occlusion bead (the anterior part parallel to the bi-pupillary line and the posterior segment parallel to Camper's plan).

We designed and printed a pediatric Fox plan according to the patient's oral proportions (Figure 8), the design was carried out by the software "Meshmixer", then the instrument was printed in polylactic acid (PLA) using an FDM (Fused deposition modeling) printer.

The vertical dimension of occlusion was corrected according to the aesthetic standards. The recording of the centric relation (CR) was made by a special wax "Aluwax®" after having validated that the CR is reiterative and reproducible

We choose overdentures covering the residual molars to preserve bone potential surrounding the teeth and to and to allow the patient to maintain periodontal-likely proprioception to offer him a semblance of "dentate state".

Teeth fitting (Figure 10)

After mounting the teeth in the laboratory (Figure 10). The teeth fitting has allowed their aesthetic and functional validation both by us (practitioner) and by the patient himself.

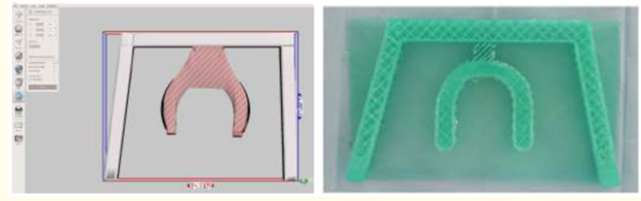

Figure 8: Computer-aided design and manufacturing of PLA pediatric Fox plan.

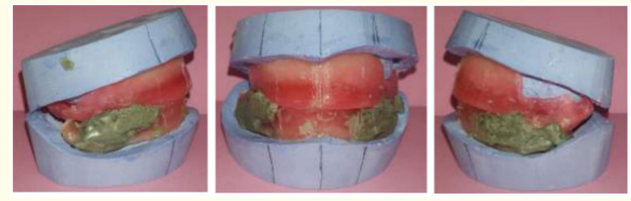

Figure 9: Record of intermaxillary relations.

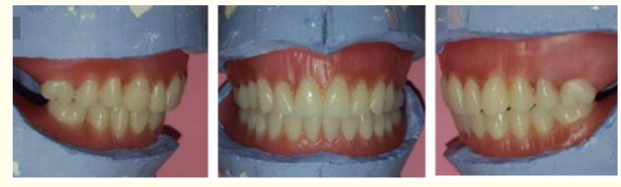

Figure 10: Teeth mounting.

Denture insertion (Figure 11 and 12)

After denture's insertion, an immediate occlusal equilibration eliminated occlusal interferences, reducing the incidence of the grievances, then all the advice relating to oral and denture hygiene as well as to the gradual introduction of hard-boiled foods were given to both the patient and his mother.

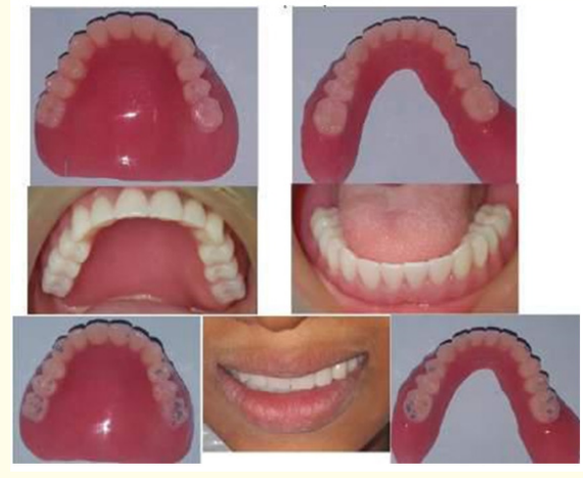

Figure 11: Denture's insertion and immediate occlusal equilibration.

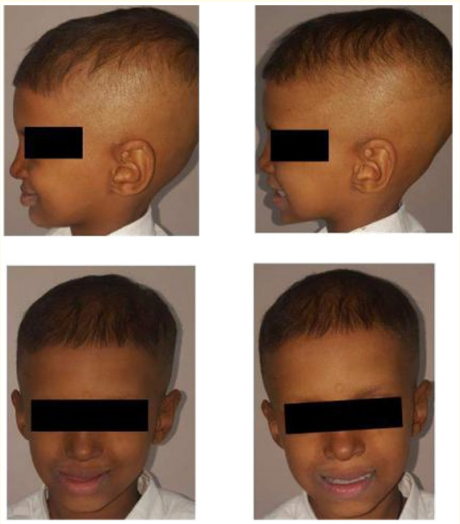

Figure 12: Comparison before and after the insertion of complete dentures.

Follow-up

Several check-appointments were organized at close intervals at the beginning (48h, 1 week then 2 weeks) then progressively further apart (3 weeks then once a month and then once every 3 months). we also programmed the resumption of the prostheses in 2 years as soon as the patient's growth will have modified again its intra-oral dimensions making the current dentures unusable.

Discussion

Ectodermal dysplasia brings together a wide range of rare diseases that affect several aspects: the clinical, pattern of genetic transmission, and the molecular pathways involved.

The first classifications of ectodermal dysplasias were based solely on clinical manifestations as a function of ectodermal derivatives affected, then evolved into clinical and molecular classifications, with the contribution of molecular biology and knowledge. molecular signaling pathways [6,7].

In the context of anhidrotic ectodermal dysplasia, the anomaly of the number concerns the two dentitions, on average for the temporary toothing 3, 5 teeth are absent in the maxillary and 5, 3 in the mandible while for the permanent toothing 5, 4 teeth are absent in the maxilla and 5.8 in the mandible. The teeth present in most cases a dental dysmorphism which is characterized by an atypical aspect. Incisors and canines are conoidal and molars have sharp cusps. In edentulous areas, the alveolar bone does not develop or is absent, hence the need to pair children with this disease. The dental disturbances of interest to both dentitions in these subjects cause a disorder of the physiology of the masticatory muscles and alteration of the facies which is reflected by a decrease in the vertical dimension of occlusion; In the sagittal direction, the absence of dental wedging causes mandibular protrusion, anterior inverted articulate, and Class III lateral dental reports; this lack of wedging is translated in the frontal direction by a lateral deviation of the incisive medians in the position of maximum inter-cuspidation.

The alteration of all the manducatory functions is reflected by a liquid or soft diet, which alters the development of weight and weight. These subjects are therefore leaner and smaller than subjects of the same age. The involvement of the mucous glands responsible for xerostomia can cause dysphagia and ageusia, as well as increased susceptibility to caries and gingivitis. Macroglossia, palatal division and absence of the tongue brake are observed. Primary swallowing resulting in poor lingual posture is favored because there is no previous wedging or posterior wedging. It also promotes the development of maxillary endo-gnathic and mandibular prognathism [8].

The conservation of temporary teeth offers several advantages, firstly it contributes to the comfort of the patient while maintaining a masticatory potential even if it is limited, by maintaining the alveolar bone, it remains possible to avoid the bone resorption for a future implant rehabilitation at the end of growth [9]. This offers greater potential for implant placement without bone grafting; on the other hand, there is a decrease in the width of the mandibular crest of 25% during the first 4 years after the extraction of the second molar and 30% after 7 years, this resorption concerns both the width and the height of the arcade [10].

In addition to restoring all the manducatory functions, the prosthetic rehabilitation of these young patients favors maxillofacial growth by stimulation of the peri-oral musculature; it also restores a good vertical dimension of occlusion and inter-arch space.

The restoration of an adequate aesthetic makes it possible to compensate for the psychic consequences caused by edentulousness [9].

The check-up appointments show that the patient is comfortable with his prostheses, has no complaints, and has been wearing them since the first day of mouth insertion.

The prototype of the FOX plan created in PLA has also been used in other cases, indeed we have individualized its proportions to adapt it in a patient suffering from auto-immune lymphoproliferative ALPS syndrome aged 14 years with a severe microstomia, it has also been used in an 85-year-old patient with squamous cell carcinoma associated with filling of the right posterior vestibule following a post-surgical scar bridle. the speed and ease of both design and manufacture of this single-use device facilitate the recording of inter-maxillary reports in patients whose oral proportions are reduced [10].

Conclusion

The anhidrotic ectodermal dysplasia in children is reflected by an oligodontia/anodontia, the prosthetic rehabilitation in these patients is essential to ensure a better quality of life and an efficient social reintegration.

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