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RESEARCH ARTICLE

ANHIDROTIC ECTODERMAL DYSPLASIA: PROSTHETIC MANAGEMENT

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ABSTRACT

The anhidrotic ectodermal dysplasia or Christ-Siemens-Touraine syndrome is a scarce pathology, characterized by abnormalities occuring during the morphogenesis of tissues and organs of ectodermal origin, induced by genetic disorders. This syndrome is characterized by the classic symptomatic triad: anhidrosis / hypohidrosis, anodontia / hypodontia, hypotrichosis (4). Dental abnormalities lead dentist to make an early diagnostic of this syndrome. These dental abnormalities can cause physiological and psychological disorders. Oral management includes oral hygiene education, prevention, conservative dental management and prosthetic rehabilitations. This paper describes the different prosthetic stages of oral rehabilitation in a 7-year-old child with Christ-Siemens-Touraine ectodermal dysplasia.

INTRODUCTION

Ectodermal dysplasias constitute an heterogeneous group of rare diseases characterized by abnormalities in the morphogenesis of tissues and organs of ectodermal origin, induced by genetic disorders (11). These abnormalities affect the skin, the nails, the hair, the teeth, the cutaneous glands, the anterior ocular part, the central nervous system. The anhidrotic ectodermal dysplasia or Christ-Siemens-Touraine syndrome is the most common and the most frequently reported form (4). It is a hereditary syndrome which transmission is linked to the X chromosome in most cases.

Clinical features

The clinical expression of anhidrotic ectodermal dysplasia is characterized by the classic symptomatic triad (1):

- **Hypohidrosis:** It is a decrease or even an abolition of sweat secretion due to the aplasia of the sudiparous glands Anhidrosis can cause thermoregulation disorders resulting in hyperthermia causing dehydration, convulsions and sometimes even death.
- **Hypotrichosis:** lack of hair development, localized or generalized to all the hairy regions. Hair is sparse, thin, rough and grows slowly, the hair is lighter than normal. Eyebrows may be abundant or absent.
- **Hypodontia:** absence of a number of teeth or anodontia: absence of all or almost all the teeth.

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Clinical consequences

- **The facies:** the development of the upper third of the face is accentuated, the developpement of the two lower parts middle are reduced. The forehead is high, bulging and the frontal bumps are marked, the eyes are often widely separated, and seem sunken in their orbits.
- **The skin:** during the childhood the skin is thin, very dry and finely wrinkled, premature skin aging is possible (10).

Complications

- **Hyperthermicexess:** the deficit in sweat glands is responsible for a lack of regulation of thermolysis. Hyperthermal attacks are therefore frequent and unexplained and can occur following exercise or during periods of extreme heat. It is often treated spontaneously or with symptomatic treatments (3).
- **Respiratory Complications:** the absence of mucus in the respiratory tracks can cause sinusitis, respiratory infections, feeding problems during childhood, xerostomia, dysphonia, recurrent bronchitis, dysphagia, gastric reflux. oesophageal, laryngitis and nasopharyngitis (15).
- **Ocular Complications:** they are related to lacrimal glands aplasia or the agenesis of the lacrimal ducts. the dryness of the ocular mucosa causes a weakening of the cornea, responsible for photophobia, which requires a permanent wear of sunglasses (11).
- **Intellectual Retardation:** This deficiency is one of the consequences of phonation and hearing disorders and a decrease of the rate of linear growth (12).

- **Weight Delay:** Due to multiple dental agenesis, chewing is disrupted. The diet is semi-liquid and not varied and therefore this leads to a delay in the development of growth (1).

Oral manifestations of anhidrotic ectodermal dysplasia

Dysplasia manifests by dental anomalies such as: number abnormalities (hypodontia or anodontia) and abnormalities of morphology:

The clinical examination shows the following signs:

- the Absence of permanent teeth.
- Persistence of temporary teeth on the arch a very late age
- Dental anomalies such as: conical teeth, microdontics, teeth with short roots. These anomalies are frequently associated with one or multiple dental agenesis.

The positive diagnosis of agenesis is reveled after a panoramic X-ray accompanied by retro-alveolar imaging

Dental abnormalities have serious consequences (13):

Due to hypodontia or anodontia, the absence of effective functions (chewing, swallowing and breathing) does not allow optimal stimulation of bone structures. insufficiently in the stimulation of the maxillary suture leads to maxillary hypoplasia. According to Desplats J and coll (6): Multiple agenesis has an important impact on facial growth and is accompanied by functional and aesthetic disorders that worsen in adulthood. Dentition process that govern the placement of temporary and permanent dentures accompany the stages of growth and development of the maxillae and the face. In addition, the mandibular condyle, and general mandibular growth are disrupted by the absence of the occlusion and unfunctional masticatory muscles.

Treatment of anhidrotic ectodermal dysplasia

A multidisciplinary approach is strongly recommended for these patients. there is no specific treatment for this syndrome management is essentially preventive and symptomatic.

• Preventive treatment (10):

- Prevention of hyperthermia consists of avoiding the exposure to the sun, avoid heat and intense physical activity, to cool the body with wet clothes, to drink cold drinks.
- Use of artificial tears to prevent eyes dryness
- The use of artificial saliva reduces dry mouth and difficulty in swallowing.

• Symptomatic treatment (10):

Nasal crusts and spills are treated with nasal saline drops and a humidifier. Early oral care is needed to rehabilitate aesthetics, chewing and phonation functions. Oral care must be done by a multidisciplinary team composed of a pedodontist, a prosthodontist, an orthodontist. Dental treatment aims restoring the various functions of phonation, chewing, swallowing and restoring an acceptable facial esthetics and thus allow a normal psycho-emotional development of the child.

Clinical case

The patient K kh, aged 7 years, presents a ectodermal dysplasia anhidrotic and came into consultation for the absence of all the teeth on the maxillary and the mandibular arch and the rehabilitation of the lower part, the motive is as well aesthetic as functional. Exobuccal examination (Fig. 1) shows a high forehead, marked frontal bumps. eyes are apart, a flattened nose, thick, bent, dry lips, slightly detached ears, a decrease in the vertical dimension of occlusion, the forward position of the chin giving the child an "old" woman's physique. The endobuccal examination (Fig. 2) reveals edentulous arches (maxillary and mandibular): the maxillary tuberosities are reduced and the alveolar processes are underdeveloped, the crests are narrow, thin and underdeveloped ".

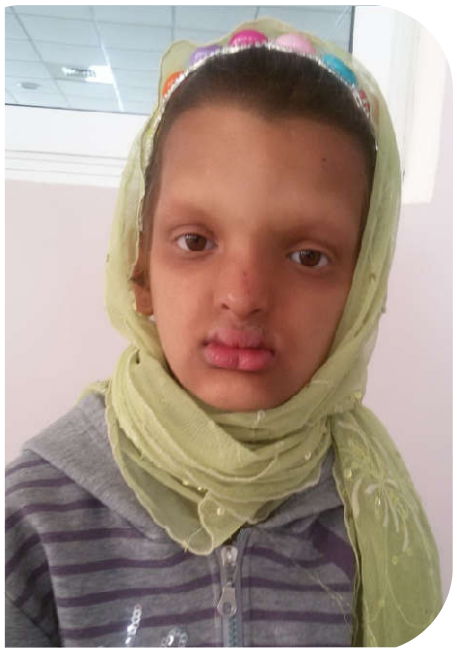

Figure 1. Front view

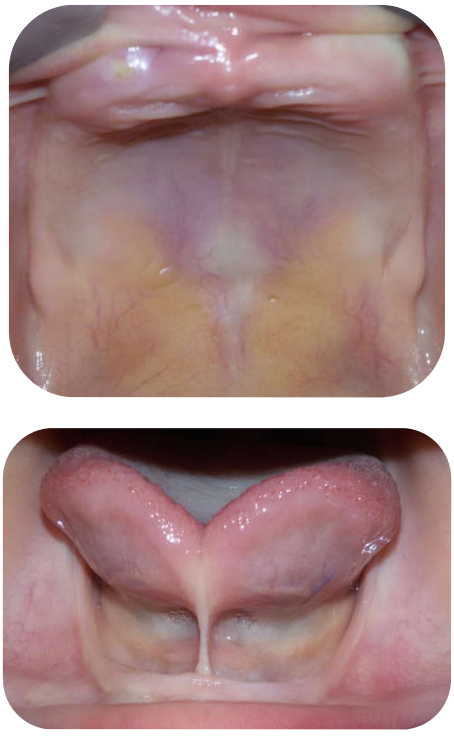

Figure 2. Intra-oral view

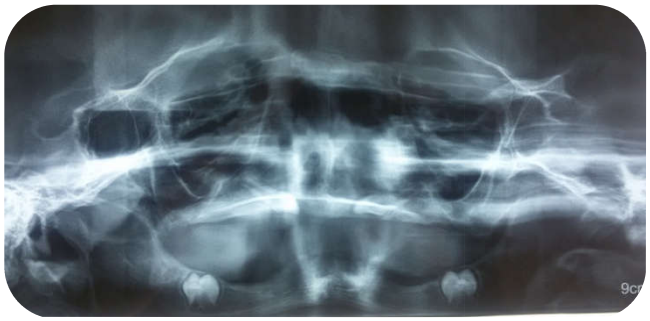

Figure 3. Panoramic X-ray

The radiological assessment (Fig 3): shows the presence of: 2 germs of maxillary canines and 2 germs of mandibular molars. These elements allowed us to make the diagnosis of Anhidrotic Ectodermal Dysplasia (AED) confirmed by our fellow specialists in internal medicine and dermatology.

3The treatment aimed:

- the functional restoration of dental arches, allow phonation, improve facial esthetics and stimulate growth through functions
- The design of the prosthesis should take into account phenomena of dentition. The removable prosthesis remains the best choice in these clinical situations (14).

The therapeutic decision made for our young patient was a total removable prosthesis.

To win the child's trust:

the first session aims to reassure the patient and explain the hole management process. Our young patient was cooperating and she expressed the need for this prosthesis and a great desire to make up for her absence of teeth.

- The presence of parents and their cooperation are highly desirable (8). The prosthetic steps:

The techniques for making complete dentures for children are identical to those for adults, the materials, the "manners". .. are sometimes modified and adapted to a child's mouth (9).

The primary impression with alginate (Fig 4)

The primary impressions are made with alginate, and are intended for the creation of an individual impression tray.
The use of warm water will reduce the time taken in the mouth to sit.

- A small plastic tray adapted to the patient's mouth was used.

The registration of the peripheral seal with a thermoplastic paste (Fig5)

The registration of the peripheral seal with a thermoplastic paste such as green kerr paste is not necessary in a very young age (2 years and 3 years for example), it can be done in the case of our young patient (Fig. 5). This recording of the peripheral seal follows the setting in the mouth of the individualized tray, the practitioner asks the child to move his face.

The anatomo-functional impressions (Fig. 6)

The maxillary and mandibular impressions are taken with a sufficiently precise material, that has a relatively short setting time ; and pleasant taste. We chose to take the impression with eugenol zinc oxide paste (Fig. 6).

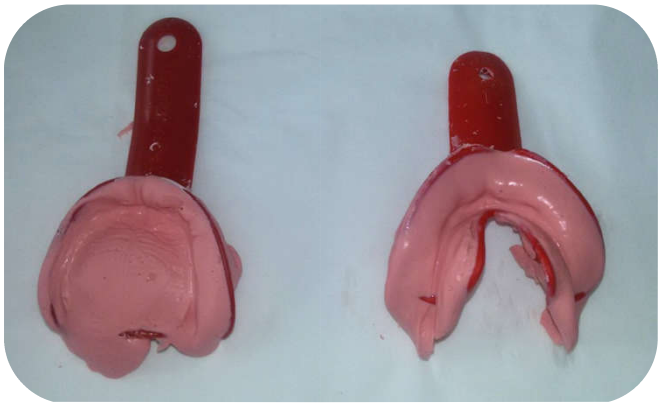

Figure 4. Primary impressions

Fig. 5. The registration of the peripheral seal with a thermoplastic paste

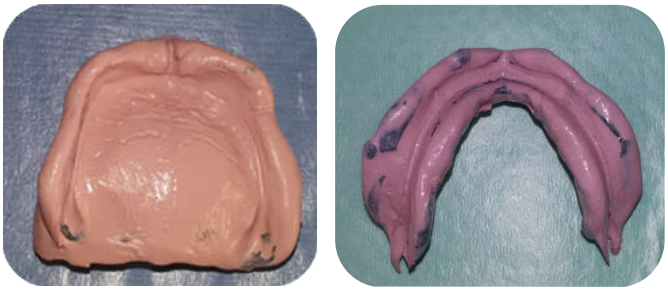

Fig. 6. The anatomo-functional impressions

Occlusion registration

Registration of maxillomandibular relations is difficult in children (7).

- Adjustment of the occlusalplane: the occlusion rim is set to have a parallelism with the bipolar line anteriorly and the posterior plan using the occlusion fox plane adapted to the size of the patient's mouth (Fig. 7).
- The volume of the occlusion rim should support the lips and cheeks and restore a harmonious appearance to the face.
- The vertical dimension of occlusion: is determined by the swallowing test.
- The centered relationship position is difficult to record in the child. The latter must be relaxed: the practitioner must seek the first contact of the occlusion rim during

the first swallowing, it is necessary to exert a pressure during the bite. The child bites with the head in hyper-extension (7) (Fig. 8)

The choice of the teeth

The assembly is done with prosthetic teeth which present an anatomy of temporary teeth: type "bambino tooth"

The shade of these teeth should be close to the natural temporary teeth. theprosthetist must leave proximal diastema between the temporary teeth (14) (Fig. 8).

The fitting of wax casts: the models are tested to check the position of the teeth and the occlusalrelatioship then the prostheses are polymerized.

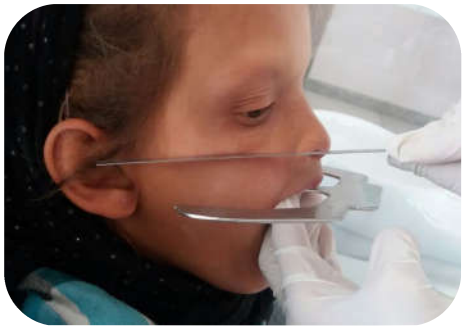

Figure 7. Adjustment of the occlusal plane

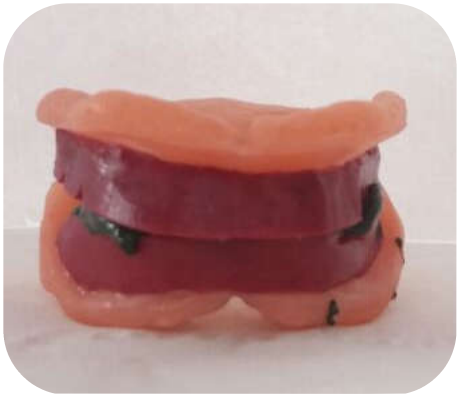

Figure 8. Registration of maxillomandibular relations

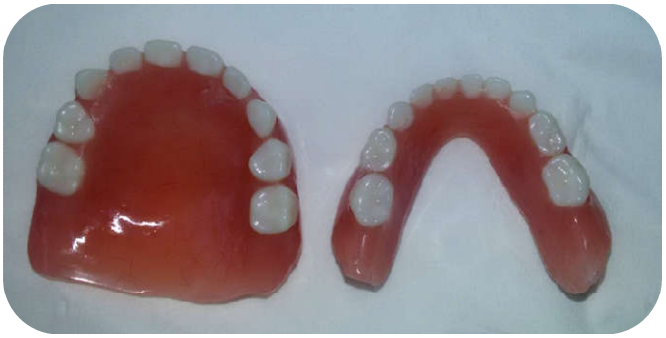

Figure 8. The total removable

The insertion of the prostheses (Fig 9)

An immediate equilibration is very important in order to fit the prosthesis.

Hygiene instructions and maintaining advise is given to children and parents

Parents should be aware of the need to monitor their child's mouth for signs of trauma or tooth eruption and to be consious to the child's complaints (5).

Follow-up and complaints:
Regular checkups are essential.

Check ups are made at three days, one week, two weeks, one month, three months, then every three months (14). It is necessary to adapt the prostheses to the dental eruptions, modifications of the denture repairs of the prostheses are necessary following the changes engendered by the development of the arches.

Growth goes through periods of acceleration and periods of slowdown (2):

- up to 30 months: maximum growth
- From 30 months to 6 years: slower growth
- 6 years to puberty: moderate growth
- at puberty: significant acceleration of growth

Prostheses are remodeled every two to four years on average during the child's growth (14).

The wearing of prostheses allows a harmonization of the oral sphere, with less pronounced cheeks, well-supported lips and a posteriorly positionned chin. This new appearance promotes the psychological well-being of the child (Fig 9).

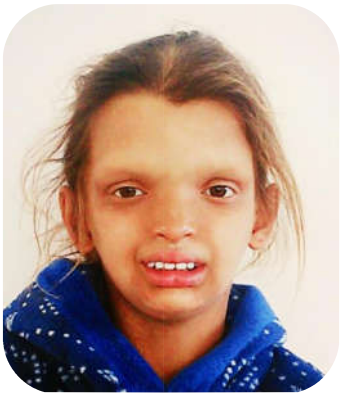

Figure 9. The insertion of the prostheses

Conclusion

The placement of the total removable prosthesis allows an almost normal functioning of the oral system and consequently an acquisition of the language and a muscular balance promoting a harmonious facial growth. The wearing of this prosthesis also allows effective chewing promoting a healthy and balanced diet and consequently a normal growth and weight.

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