

**Early Management of Hypohidrotic Ectodermal Dysplasia: Conventional Removable Prosthesis.**

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**Abstract**

**Introduction**

Hypohidrotic ectodermal dysplasia (HED) is a genetic disease, the development of ectoderm characterized by malformations of ectodermal structures such as skin, hair, teeth and sweat glands. (2, 17 , 23)

It is composed of three clinically indistinguishable subtypes associated with impaired sweating as a major symptom: X-linked DEH, autosomal recessive HED (AR-HED) and autosomal dominant HED( AD-HED), as well as a rare fourth subtype with immunodeficiency as the main manifestation (HEDwith immunodeficiency). Its incidence is estimated at 7/10,000 births. (2, 7,19, 23)

HED is clinically characterized by oligodontia (absence of at least six permanent teeth), dental morphology abnormalities, craniofacial dysmorphism and alveolar bone hypoplasia, associated with other ectodermal derivative defects. (1, 5, 18)

Children with ectodermal dysplasia have characteristic facial abnormalities that can look like old people.

The ectodermal anhydrous dysplasia (EAD) or X-linked ectodermal dysplasia (XED), is the most frequent of the ED because it represents 80% of the ED (incidence estimated at 1 / 100,000 births).

In this work, we recall the characteristics of HED and specify, through a clinical case of prosthetic rehabilitation, the interest of treatment with conventional prostheses

**Clinical Case**

A 5 years old male child went to the Department of Pediatric Dentistry in Casablanca Dental Consultation and Treatment Center (CCTD) with his mother. The latter suffers from an aesthetic and functional gene related to the delay of eruption of the teeth.

On the exobuccal examination, we notice a particular facies. The nose is flat and broad, the lips are thick and everted, the chin prominent, the ears large and loose, the eyelashes and eyebrows are scanty and the hair sparse.



Figure 1 : Front view



Figure 2 : Profile view

At the endobuccal examination, we notice the absence of several teeth with only the presence of the two posterior maxillary teeth.

At the mandible, the presence of an alveolar ridge in a shape of a knife blade.



Figure 3 : Severe oligodontia with the presence of 2 knife-edge bone crest posterior teeth.

The panoramic X-ray of the child, revealed the presence of 2 teeth and a germ, in the upper maxilla. While in the mandible, presence of anodontia.



Figure 4 : Panoramic X-ray: alveolar ridge in a shape of a knife-edge following absence of teeth

The diagnosis of hypohidrotic ectodermal dysplasia was discussed in relation to the clinical and radiological signs presented by the child.

A prosthetic rehabilitation is then proposed, to restore the function and aesthetics.

Primary dental impressions with alginate are performed. The plaster models from these dental impressions permit to create the individual impression gates, necessary for the perfect reproduction of the dental arches.



Figure 5 : Secondary dental impressions

Elastomeric secondary impressions, reproducing the static and dynamic movements of the patient.

Realization of wax occlusion models; it will help to evaluate the correct vertical dimension, using the phonetic and aesthetic criteria and to record the relationship between the two arches.

It is necessary to record on the occlusal wax the interincisal medial, the verticality and the smile line.



Figure 6 : Occlusion models & occlusal wax



Figure 7 : The prothesis



Figure 8 :Front photograph with prosthesis

### Discussion

The oligodontia is generally severe when associated with the various forms of ectodermal dysplasias, both in temporary and permanent dentition, which has multiple morphological and functional consequences.

Thus, diastema, alveolar bone hypoplasia, subsidence of the lower facial height with incisal supraclulsion are some examples of clinical manifestations directly related to the dental phenotype. An average of 8 dental agenesis is described in temporary dentition, an average of between 11 and 16 agenesis being found in permanent dentition, which corresponds to severe oligodontia phenotypes.

A specific topographic distribution of dental agenesis is found in ectodermal dysplasias, which guide the clinical diagnosis. crown abnormalities in size and morphology are generally observed in ectodermal dysplasias, consisting of conoid anterior teeth and microdontia ( 19, 22)

The craniofacial features described in the various forms of ectodermal dysplasia associates a large number of dysmorphic elements such as hypoplasia of the middle third of the face, maxillary retrognathia-brachygnathia, mandibular prognathism or pseudo-prognathism, facial concavity and frontal prominence, for example.

Depressed nasal bridge is noticed, which accentuates the effect of a small face (21,22)

The mento-labial crease is pronounced, the lips are everted and thick. Their prominence is due to the decrease of the vertical dimension (22)

As a consequence of dental agenesis, the formation of the alveolar ridge is inhibited. In general, the eruption of the teeth can be considerably delayed.

Multiple dental agenesis, as well as the maintenance of temporary teeth, which are not replaced beyond physiological age by permanent teeth, cause functional disturbances in chewing, swallowing, breathing and phonation. . (1,3,4,7)

A poorly diversified semi-liquid or liquid diet is often necessary because of the weakness of the dental organ.

These foods do not stimulate the action of the masticatory muscles, so we observe a hypertrophy and spreading of the tongue, accompanied by cheeks and lips interposition ( 8, 9, 10)

To allow social integration if the young patients despite the missing teeth, early prosthetic rehabilitation is indicated. It can improve aesthetics in a decisive way, but

also masticatory function and phonetics and therefore self-confidence. There is no definite age to start dental care. This must be done early, as soon as the cooperation of the child allows it. (11).

Early prosthetic rehabilitation is necessary, usually in preschooler age. From the age of 2 to 3 years, children can already benefit from dental replacements, made with simple and non-invasive procedures.

The pathology of these children is sometimes detected quite late, sometimes after the age of 3, when parents begin to notice certain dental abnormalities in their child or simply the absence of lacteal teeth. (article prosthesis dysplasia) indeed, our patient consulted only at the age of 5 years, referred by his pediatrician, because of the delay of teeth eruption.

For the oral management of these children, a multidisciplinary approach should be conducted with different prosthetic options, depending on the degree and site of dental and alveolar impairment, in combination with controlled local hygiene and conservative restorations (9, 11, 12, 13).

Regarding our patient, the first step in his treatment plan was oral hygiene motivation, so child can adopt daily oral hygiene behaviors once the prosthesis placed Then prophylactic care was performed on all existing teeth.

The dental manifestations represented by severe hypodontia are the origin of the significant alveolar bone deficit. The absence of permanent teeth and the lack of stimulation of the jaws will lead to an irregular alveolar volume and an alteration of its structure. In fact, the lingual spread will cause a deviation of the alveolar development. However the absence of teeth will not prevent the growth of the basal bone, maxillofacial development is almost normal. (14,15,16) which will allow us to realize our removable prosthesis.

Commercial dental impression trays are most often unsuitable, hence the interest of creating an individual impression tray to reproduce the dental arches of the patient. This will enable us to obtain stable and adapted occlusion models.

In order to achieve a good secondary impression or functional anatomy, the child must be accompanied for the different movements.

To record the inter-maxillary report, the models must be well adjusted to ensure good labial support, with a slight increase in the vertical dimension of occlusion.

The esthetic validation of the prosthesis by the patient is essential, before the polymerization.

Removable prosthesis must be considered as a temporary solution. It must facilitate the normal completion of dental and maxillary development, thereby playing a role of orthopedic or orthodontic device.

The follow-up consists of a constant adaptation of the prosthetic devices to the growth, with rebasings and renewals according to the individual growth rate, a tri-annual consultation being systematically recommended (6)

### **Conclusion**

Early prosthetic rehabilitation with removable resin-based prosthesis best meets the therapeutic objectives. These prostheses stimulate bone growth and allow progressive muscular, articular and phonetic conditioning.

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