

## Early Dental Rehabilitation of a Young Patient with Ectodermal Dysplasia-Case Report

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### Case Report

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**Abstract:** Ectodermal dysplasia (ED) is a rare hereditary disease characterized by a congenital dysplasia of several structures of ectodermal origin. Common manifestations include defective hair follicles and eyebrows and deficiency of sweat glands. Intraorally, anodontia, hypodontia or oligodontia are the most common findings. We report the case of a four-year-old girl who was referred to the pediatric dentistry department of hospital la Rabta (Tunisia) and exhibited many of the dental manifestations of ED as well as general abnormalities. Our early oral rehabilitation was launched to improve oral functions and esthetics by including the fabrication of two removable prostheses and acid-etched composite resin restorations. The patient reported a good adaptation of the conventional dental removable prosthesis and parents were satisfied with the treatment.

**Keywords:** Ectodermal dysplasia, hereditary disease, syndrome, specific oral findings, dental management, early oral rehabilitation

### INTRODUCTION

Ectodermal dysplasia (ED) is a hereditary disorder linked to a recessive gene from X chromosome that can affect several developing ectodermal origin structures which may include: skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear, and parts of other organs [1]. The incidence of the disease is about 1:100,000 births and has a 30% mortality rate in childhood due to intermittent hyperthermia [2]. Several types of ED are known but the severity of the disorder varies from patient to patient. In general, ED syndromes display trichondysplasia, onchondysplasia, hypohydrosis and abnormal dentition: anodontia or hypodontia and conical teeth.

The most reported ED syndrome is hypohidrotic (anhidrotic) ED known also as Christ-Siemens-Touraine syndrome. This affects one to seven individuals per 10,000. [3] Yavuz *et al.* reported that the most frequent abnormality in ectodermal dysplasia is skin disorders (93%), followed by hair and nail disorders (86%) [5]. The phenotype characteristics of ectodermal dysplasia are those of aged face. Specific facial features include little hair, lack of eyelashes, prominent brow, labial protuberance, flattened bridge of the nose, small vertical facial height, small palatal base, bulging eyes, ferveescene, depressed nasal bridge, jaw deformity [4, 5]. Besides, dental abnormalities will be found too. We can observe lack of teeth on both deciduous and permanent dentitions which means hypodontia, oligodontia and even anodontia. Some studies showed that premolars were the most commonly missing teeth. [5] in addition, enamel dysplasia, microdontia, malformed and conical teeth, diastema; underdeveloped alveolar ridges are observed too.

Furthermore, hyposalivation and missing teeth lead to swallowing difficulties and affected maxillofacial growth leading to atrophy and underdevelopment of the stomatognathic system. Children with ED usually have a normal mentality and life expectancy, and their facial appearance warrants professional concern for their emotional well-being and social progress [6].

### CASE PRESENTATION

The affected propositus is a female patient aged 4 years old. She had the classic triad of hypotrichosis, hypohydrosis and hypodontia, and was thus diagnosed with Ectodermal dysplasia. She was referred to the Department of Oral Medicine, hospital La Rabta, Tunis, Tunisia, with the chief parent's complaint of missing teeth and difficulty of chewing and speaking.

Family history revealed that the mother was healthy but the father and the sister were affected. A history of the genetic disorder was reported in relatives too (figure 1A, 1B).



(A)



(B)

**Fig-1: the family's child: father and sister are affected**

At the age of 4 years, the time of our first extra oral observation, the facial appearance revealed dysmorphic facial features. These are characterised by a senile facial appearance. The patient presented dry and

rough skin, sparse scalp hair, missing eyelashes and eyebrows, severe hypohidrosis and hypotonicity of the perioral musculature (figure 2A, 2B).



(A)



(B)

**Fig-2: specific facial features: senile facial appearance on frontal and profile view**

Intra oral examination showed the absence of several primary teeth, thin bone bridge and conical shape of incisors 51 and 61 (figure3).



**Fig-3: Intra oral examination: absence of several primary teeth and conical shape of incisors 51 and 61**

The orthopantomogram confirmed hypodontia. Seven primary teeth were missing: 52, 61, 62, 64, 72, 71, and 81. Agenesis of permanent germs was noticed

too. At this early age we can confirm the absence of germs 12, 22, 23, 24, 34, 31, 41 (figure4).



**Fig-4: The orthopantomogram**

Agenesis of Second premolars 25, 35, 45 could not be confirmed at this age because the physiological date of crypt appearance is not reached yet. Thus, as we have the congenital absence of more than six permanent teeth aside from the genetic syndrome, we can confirm oligodontia.

As a consequence of this oligodontia, we could notice a spreading tongue into interstices of missing teeth leading notably to deglutition, phonation and proprioception problems.

Besides, at the age of 4, the teeth 63 and 73 are in eruption process which confirm delayed dental eruption aside from number and shape dental abnormalities detected in oral examination.

In our early treatment approach, the maxillary anterior teeth 51 and 61 were restored with composite resin to adjust the incisor shape and improve esthetics and chewing function. Secondly, the confection of two conventional partial removable prosthesis was strongly recommended Figure (6).



Fig-5:

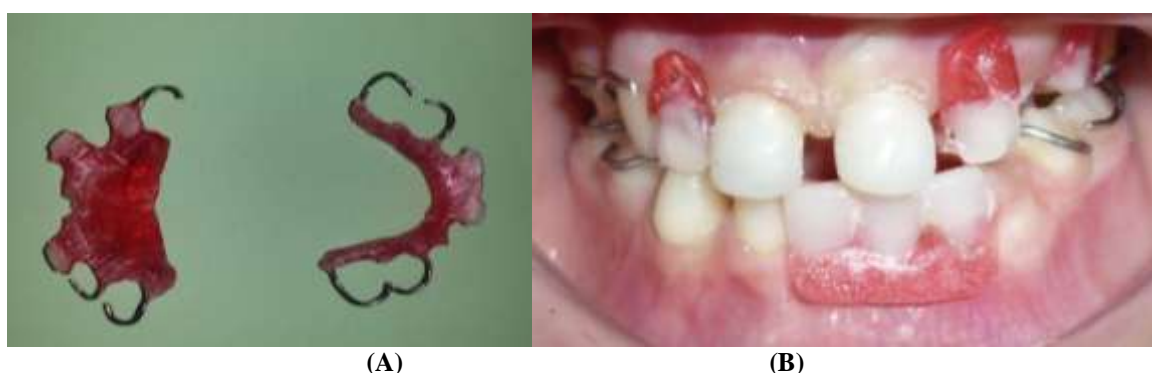


Fig-6: A) confection of two conventional partial removable prosthesis; B): Esthetic reconstruction of anterior teeth and placement of upper and lower removable denture

Generally, children with ectodermal dysplasia must start the prosthetic treatment as soon as possible

for reasons of space maintainer, self-esteem and optimal psychological maturation [7] (Figure 7).



Fig-7: satisfied patient

Authors agree that from the age of 3 to 5 years, the child begins to become sufficiently cooperating to accept such treatment. The partial resin denture is the most commonly used, and is satisfactory esthetically and functionally until a certain age by improving masticatory efficiency, swallowing ability and phonetic capability [8].

#### DISCUSSION

The most common treatment plan for patient with ectodermal dysplasia is the removable prosthesis. Positive effects include more self-confidence, facial esthetics, speech and masticatory function improvement. In addition, studies have shown that

patients affected by oligodontia had higher anxiety and emotional problems than a normative sample even during childhood [9].

As a fact, oral rehabilitation of children with ectodermal dysplasia is necessary to improve sagittal and vertical skeletal relationships during craniofacial growth. However, during this period, the maxilla is still growing. Several options have been proposed to adapt to this growth. Some authors recommend changing or modifying the prosthesis every 2 or 3 months [10]. For others, it is every 6 months [11]. Other therapeutic approach established by Montanari and al is to include a three-way screw in the prosthesis with activation every



2 weeks [12]. Unfortunately, such an approach remains dependent on the periodic visits of control by the patient.

On the other hand, in some cases, if the oligodontia is more than 10 teeth and there is failure of the conventional prosthesis retention, a new treatment approach can be suggested, which includes a mini-implant placement to support mandibular prosthesis both on childhood or adulthood [2, 13]. However, long-term effects of the rehabilitation on the growth and the development of craniofacial structures were not considered. Bergendal *et al.* demonstrated a frequent dental implant loss among patients with ectodermal dysplasia (64.3%), unlike patients with dental trauma and agenesis treated with dental implants. Consequently, Children with ED should be rehabilitated with smaller implants due to the reduced dimension of the maxillary and cortical bones [14]. Many authors suggested this plan treatment over the age of 12 [15]. But in any case, further clinical evaluations must be performed each six months to observe the need of mini-implant replacements and/or substitution of prostheses due to the growth and development processes.

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#### PATIENT CONSENT FORM

Patient consent related case report was taken from concerned parents.

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