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## Hypohidrotic ectodermal dysplasia: prosthodontic treatment in a paediatric patient

### Case report

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### Introduction

Ectodermal dysplasia (ED) comprises a large, heterogeneous group of inherited disorders [1, 2], that are defined by primary defects in the development of 2 or more tissues derived from the embryonic ectoderm. [1, 3, 4, 5, 6, 7, 8]  
The most common EDs are X-linked recessive hypohidrotic/anhidrotic ED [3, 5, 7, 9]. It has full expression only in males; female carriers show little or no signs of the condition. [2] The goals of the dental treatment are: enhancement of functions (aesthetics, mastication, phonetics) and psychosocial activities. [3].

### Objectives

The aim of this paper is the presentation of the clinical aspects and the prosthetic rehabilitation of 7 years old boy with X-linked recessive anhidrotic ectodermal dysplasia.

### Material and Methods

**CASE REPORT:** His Romanian mother and maternal grandmother are healthy carriers of the ED gene and display some milder clinical manifestations of the disease. The Chinese father and his family are fully healthy.

**CLINICAL MANIFESTATIONS:** The patient presents typical general, facial and oral clinical features characteristic to the disease. Defective development of structures of ectodermal origin (skin, hair, nails, sweat glands) produces numerous general findings and symptoms, such as lack of perspiration, frequent episodes of hyperpyrexia, thin and fragile nails, chronic respiratory infections, frequent otitis media, epistaxis.

**TREATMENT:** A simple conventional prosthodontic rehabilitation has been chosen for the enhancement of functions and psychosocial activities. The conical primary maxillary right central incisor was reconstructed with an acrylic crown by an original technique.



Fig. 1a and b: Facial and oral aspects of the mother: some facial distortions and the absence of the maxillary lateral incisors.



Fig. 2: Facial distortions: prominent frontal ridges and chin, brittle and fine hair, a saddle shaped nose, an extended philtrum, an everted lower lip, a horizontal groove on the chin, thick cheeks and large ears, reduced lower face height.



Fig. 3: General features: thin and dry skin, hyperpigmented, appearing prematurely aged, unusual scaling or peeling of the skin, hyperkeratosis and chronic eczema on both palms and soles.

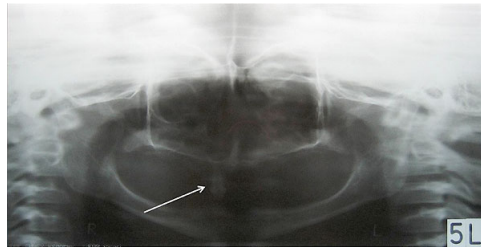


Fig. 4a and b: Oral and radiological features: presence of one primary maxillary cone-shaped tooth, the absence of alveolar processes.



Fig. 5: Partial maxillary removable prosthesis and a total mandibular denture.



Fig. 6: Manufacture of the acrylic crown. For an optimum aesthetic result it was made out of a homologous acrylic tooth, carved in order to obtain a veneer which was completed on the oral side with baropolimerised acrylic resin.



Fig. 7a and b: The patient's facial and oral aspect after the completion of treatment; improved masticatory, phonetic and aesthetic functions.

## Results

The oral rehabilitation of children with ED is necessary in order to improve the patient's functions, aesthetics as well as psychological condition. This frequently involves a complex prosthetic treatment, which presents a number of difficulties given by the patient's continuing growth, the choice of the appropriate prosthetic appliance as well as the uncooperative attitude of the young patient. The behavioural treatment by the "Tell-Show-Do" technique was necessary before and during the prosthodontic treatment. After the completion of the prosthodontic treatment our patient had no discomfort and seemed to be adapting well. However, although both he and his mother were informed about the need for periodic recall in order to observe the integration of the prosthesis and replace the dentures as growth occurs, treatment was abandoned.

## Literature

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*This Poster was submitted by Dr. Mihaela Pauna.*

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## Hypohidrotic ectodermal dysplasia: prosthodontic treatment in a paediatric patient

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### INTRODUCTION

Ectodermal dysplasia (ED) comprises a large, heterogeneous group of inherited disorders [1, 2], that are defined by primary defects in the development of 2 or more tissues derived from the embryonic ectoderm. [1, 3, 4, 5, 6, 7, 8]  
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 The goals of the dental treatment are: enhancement of functions (aesthetics, mastication, phonetics) and psychosocial activities. [3].

### AIM

The aim of this paper is the presentation of the clinical aspects and the prosthetic rehabilitation of 7 years old boy with X-linked recessive anhidrotic ectodermal dysplasia.

### CASE REPORT

His Romanian mother and maternal grandmother are healthy carriers of the ED gene and display some milder clinical manifestations of the disease.  
 The Chinese father and his family are fully healthy.



Fig. 1 Facial and oral aspects of the mother: some facial distortions and the absence of the maxillary lateral incisors.

### CLINICAL MANIFESTATIONS

The patient presents typical general, facial and oral clinical features characteristic to the disease.

Defective development of structures of ectodermal origin (skin, hair, nails, sweat glands) produces numerous general findings and symptoms, such as lack of perspiration, frequent episodes of hyperpyrexia, thin and fragile nails, chronic respiratory infections, frequent otitis media, epistaxis.



Fig. 2 Facial distortions: prominent frontal ridges and chin, brittle and fine hair, a saddle shaped nose, an extended philtrum, an everted lower lip, a horizontal groove on the chin, thick cheeks and large ears, reduced lower face height.



Fig. 3 General features: thin and dry skin, hyperpigmented, appearing prematurely aged, unusual scaling or peeling of the skin, hyperkeratosis and chronic eczema on both palms and soles.



Fig. 4 Oral and radiological features: presence of one primary maxillary cone-shaped tooth, the absence of alveolar processes.

### CONCLUSIONS

The oral rehabilitation of children with ED is necessary in order to improve the patient's functions, aesthetics as well as psychological condition. This frequently involves a complex prosthetic treatment, which presents a number of difficulties given by the patient's continuing growth, the choice of the appropriate prosthetic appliance as well as the uncooperative attitude of the young patient. The behavioural treatment by the "Tell-Show-Do" technique was necessary before and during the prosthodontic treatment.

After the completion of the prosthodontic treatment our patient had no discomfort and seemed to be adapting well.

However, although both he and his mother were informed about the need for periodic recall in order to observe the integration of the prosthesis and replace the dentures as growth occurs, treatment was abandoned.

### REFERENCES

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### TREATMENT

A simple conventional prosthodontic rehabilitation has been chosen for the enhancement of functions and psychosocial activities.



Fig. 5 Partial maxillary removable prosthesis and a total mandibular denture.

The conical primary maxillary right central incisor was reconstructed with an acrylic crown by an original technique.



Fig. 6 Manufacture of the acrylic crown. For an optimum aesthetic result it was made out of a homologous acrylic tooth, carved in order to obtain a veneer which was completed on the oral side with baropolymers acrylic resin.



Fig. 7 The patient's facial and oral aspect after the completion of treatment; improved masticatory, phonetic and aesthetic functions.