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Dental Management of Ectodermal Dysplasia: A Case Report

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Abstract
Aim: *To present a case report of an interdisciplinary treatment of young patient with ectodermal dysplasia associated with severe oligodontia*
Material and method: *This report presents a case of a 9 year old child affected by ectodermal dysplasia with hypodontio in the upper jaw, atypically located maxillary canines and edentulism in the lower jaw. Treatment of thos patient was interdisciplinary including an orthodontics, prothodontics and pedodontics.*
Results: *Conservative and prosthodontic treatment made significant imporevement of the extraoral profile as well as intraoral aesthetics. Beside that improvement in masticatory function and speech was also noticed after the treatment. The results had major impacts on self-esteem, masticatory function, speech and facial esthetics.*
Conclusions: *Interdisciplinary management and communication of different specialties has once again proved to be essential in treatment of dental problems in complex cases such as ectodermal dysplasia.*
Keywords:*hypodontia, anodontia, oral rehabilitation, removable prosthesis*

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I. Introduction

Ectodermal dysplasia is a hereditary disorder associated with abnormal development of embryonic ectodermally-derivated organs including teethe,nails and sweet glands. It can be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recesive, and X-linked modes. These gene abnormalities can be inherited from one or both parents, or they can arise as a new genetic event.

Althought more than 170 different subtypes of ectodermal dysplasia have been identified, these disorders are considered to relatively rare with an estimated incidence of 1 case per 100 000. The most common form of ectodermal dysplasia ussually affects men. Other forms od the disease affects men and women equallly.

According to the state of sweat glands involvment, two major groups are distinguished: hypohidrotic or anhydrotic (Christ-Siemens –Touriane syndrome) in which sweat glands are absent or significantly reduced in number, or hydrotic (Cloustone Syndrome) in which sweat glands are normal. The hypohidrotic as the most common type seems to show X-linked inheritance pattern, therefore males are more susceptible than females. On the other hand hydrotic type is inherited in an autosomal dominant pattern. Prenatal diagnosis is available for some families with X-linked hypohidrotic dysplasia through the use of DNA probes. This is not possible for all families.

The teeth are markedly reduced in number (oligodontio or hypodontio). And often manifest abnormal development in shape which may appear tapered, conical or pointed in incisors/. Molars might be observed in reduced size. The lack of tooth bud formation causes hypoplastic alveolar bone, leading to reduced vertical dimension of occlusion. Therefore an old appearance is common in affected individuals.

There is no specific treatment for this disorder. But here are some usefull recomandations in such cases: wearing a wig or dentures to improve appearance, using artificial tears to replace normal tearing and prevent drying of the eyes, spraying the nostrils with saline nose spray often to remove debris and prevent infection., living in cooler climate and taking cooling water baths or using water sprays to keep a normal body temperature .

A new treatment that could help children born with a rare genetic condition has been developed at the Washington University School of Medicine in Misuri, The new treatment which is in clinical trail now, is a recombinant protein that initiates a cascade of molecular and cellular events that leads to the proper

development of hair, teeth and sweat glands. Dr. Anand Srivastava, associated director od JC Research Institute's Centar for Molecular Studies at GGC. Prior to arriving at GGC in 1995, Srivastava identified the gene that causes EDA while he was a researcher at Washington. In the intervening years he and a team of international colleagues continued to work on the condition.

The drug, which was developed by Edimer Pharmaceuticals, has been shown to correct some of the symptoms of the condition. Clinical trials began 2012 that call for five doses of the drug administered intravenously during the first month of life.

We will represent interdisciplinary rehabilitation of a boy with hereditary ectodermal dysplasia associated with severe oligodontia .

II. Aim Of Study

- *To present a case report of young patient with ectodermal dysplasia accompanied with severe oligodontia*
- *To determine the benefits of an complete interdisciplinary (orthodontic-prothodontic-pedodontic) treatment ofsubject with ectodermal dysplasia*

III. Materials And Methods

A 9 year old boy was refered to our Clinic at the Department of orthodontic, with complains of missing teeth. The extra oral examinations revealed: dry skin, hypoplastic midface, prominent lips and sparse fine hair. These findings matched typical features of anhidrotic ectodermal dysplasia.

The intraoral examination revealed: complete edentulism in the lower jaw, atypically located canines and only 3 molars present in the upper jaw, thin alveolar ridge and reduced vertical bone height and loss of vestibular bone in the lower jaw.

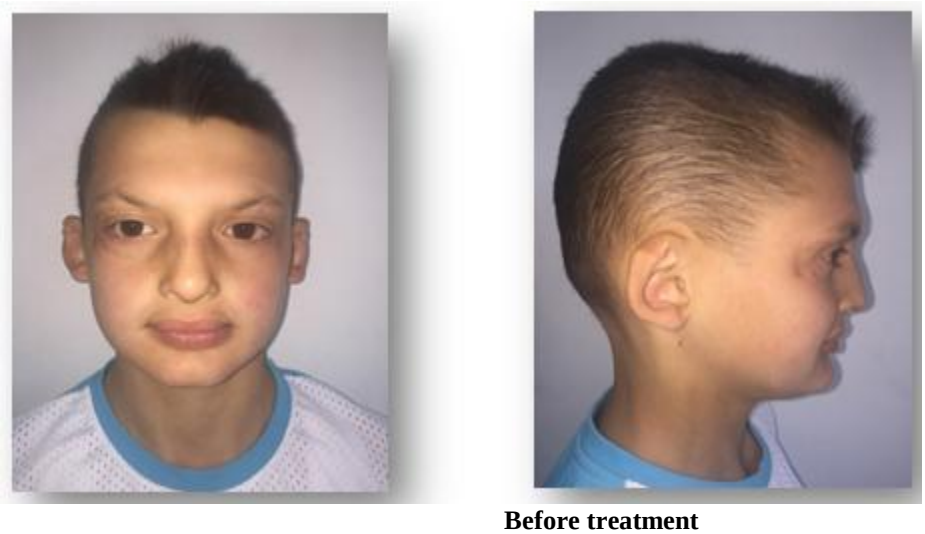
The radiographic findings (Picture 4) also confirmed the clinical diagnosis. Oral rehabilitation was accomplished with composite esthetic restaurations and removable acrylic prostheses.

IV. Results

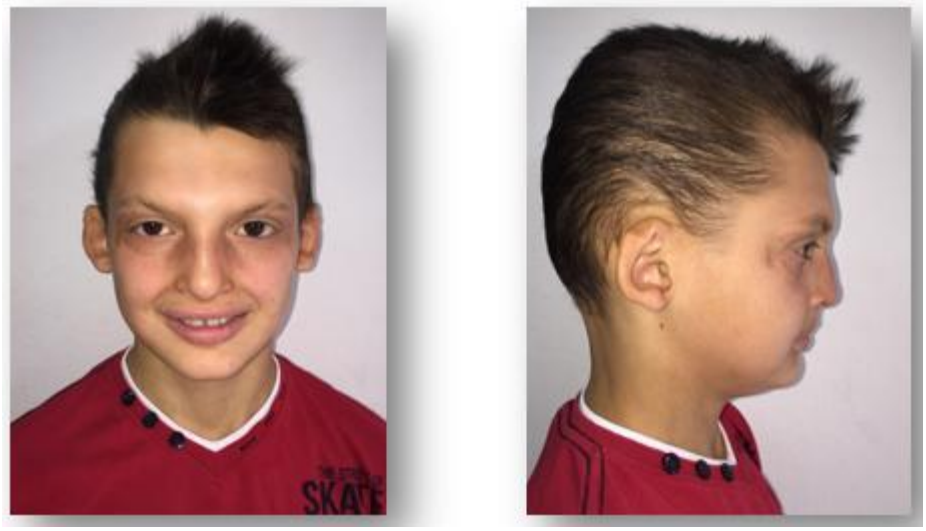
The interdisciplinary treatment of this subject with ectodermal dysplasia included: conservative treatment of the upper central incisors and prosthodontic treatment of the oligodontia with removable dentures.

The conservative and prosthodontic treatment made significant imporevement of the extraoral profile (Picture 1) as well as intraoral aesthetics (Picture 2 and 3). Beside that, improvement in masticatory function and speech was also noticed after the treatment.

The results had major impacts on self-esteem, masticatory function, speech and facial esthetics.

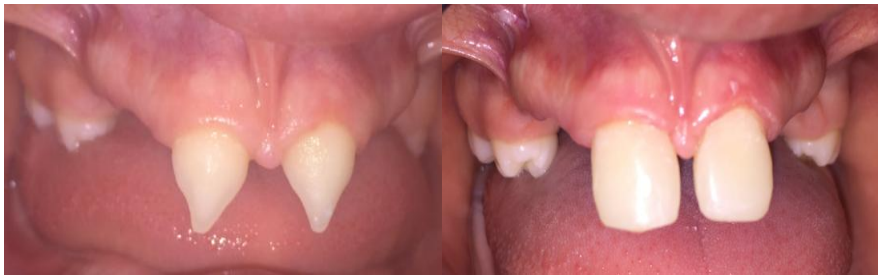


Before treatment



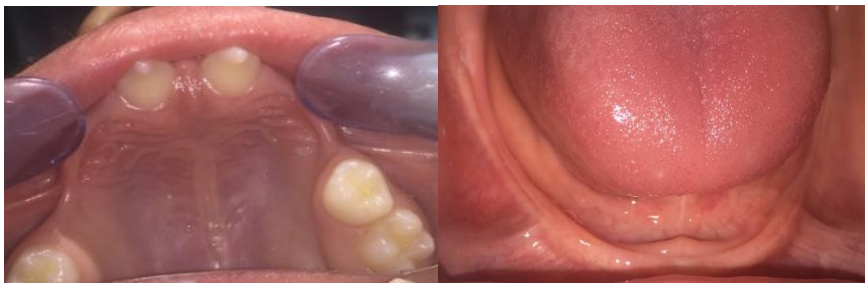
After treatment

Picture 1

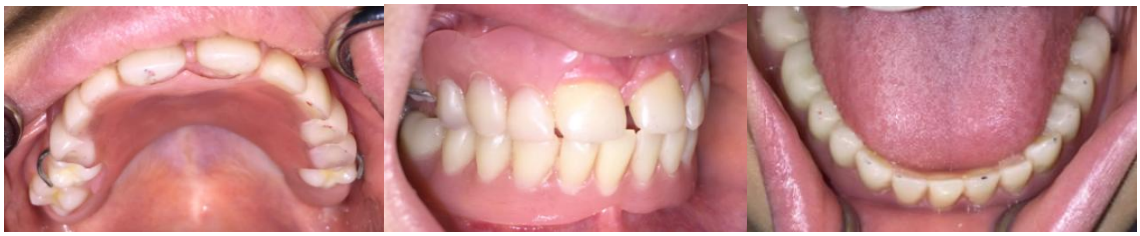


Before treatment After treatment

Picture 2 Conservative treatment of upper central incisors



Before treatment



After treatment

Picture 3 Prosthodontic treatment of oligodontia

Picture 4 OPG

V. Conclusions

Ectodermal dyspasia is commonly associated with dental problems, such as oligodontia. Treatment of these problems has to include several different dental specialties in order to achieve high level of aesthetics and functional improvements.

Interdisciplinary management and communication of different specialties has once again proved to be essential in treatment of dental problems in complex cases such as ectodermal dysplasia.

The results of this interdisciplinary treatment had major impacts on subjects' self-esteem, masticatory function, speech and facial esthetics.

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