Clinical management of hypohidrotic ectodermal dysplasia with anodontia: case report

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Ectodermal dysplasia is a rare hereditary disorder with a characteristic physiognomy. The case of a 5-year-old child with hypohidrotic ectodermal dysplasia and complete anodontia is presented. Because of the anodontia and the need for treatment at an early age, the prosthetic management of such a young child can be difficult. Complete dentures were provided to encourage a normal psychological development and to improve the function of the stomatognathic system.
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INTRODUCTION

Ectodermal dysplasia (ED) is a hereditary disorder with abnormal development of tissues and structures of ectodermal origin.[1,2] Over 100 different forms of ED have been described.[1,3] Two different forms are clinically distinguished, an autosomal inherited hydrotic form (Clouston´s syndrome) and a hypohidrotic form (Christ-Siemens-Touraine syndrome).[4,5] The latter form is an X-linked recessive trait,[2,6,7] with an incidence of 1 to 7 per 100,000 live births.[8] Characteristic of the hypohidrotic form include the triad of hypodontia, hypotrichosis and hypohidrosis[2] from the decreased number and distribution of sweat pores.[9]

The oral manifestation of this disorder create treatment challenges for the dentist.[10] Oligodontia or complete anodontia leads to an atrophy of the alveolar bone.[11,12] Teeth that do erupt are delayed in eruption, and vary in morphology with large pulps and cone shaped crowns.[2,5] The permanent dentition may also present with impactions and transpositions.[4] For children of pre-school age, proper prosthetic treatment is necessary for the function of the stomatognatic system and for the psychological development.[4,5, 13-18]

The prosthetic treatment of a young child with hypohidrotic ectodermal dysplasia with complete anodontia is described.

CASE REPORT

A 5-year-old Negroid male presented to the Department of Pediatric Dentistry due to lack of teeth and problems with speech and mastication.

The general medical history was non-contributory, but with a weight and height both within the 10% percentile.[19] An evaluation by a pediatrician resulted in the diagnosis of hypohidrotic ectodermal dysplasia with hypotrichosis, hypohidrosis and asthma bronchiale. In addition an atrophic rhinitis (ozena) with a fetid green secretion has been reported. There were no signs of mental retardation.

The family history revealed no other cases of ectodermal dysplasia.

The extraoral examination (Figure 1) revealed the typical facial physiognomy of hypohidrotic ectodermal dyplasia with prominent forehead and ears, protuberant lips and a saddle nose. The patient also presented with sparse scalp hair, missing eyelashes and eyebrows. The skin appeared dry with a hyperpigmentation around the eyes and the mouth. A diminished lower facial height contributed to a senile facial expression.

The intraoral examination revealed complete absence of primary teeth (Figure 2). Evaluation of the diagnostic casts showed underdeveloped alveolar ridges with minimal height (3mm/4mm in the lower/upper incisal region, 2mm/0.5mm in the lower/upper region of the supposed 2nd primary molar) and width (2mm/2mm in the lower/upper front and 1mm/0.5mm at the side, respectively). The palate was shallow and the oral mucosa was healthy with a slight dry appearance. The tongue was relatively large, but no signs of macroglossia could be detected.

Radiographic examination (Figure 3) revealed the complete absence of permanent tooth germs. Only a

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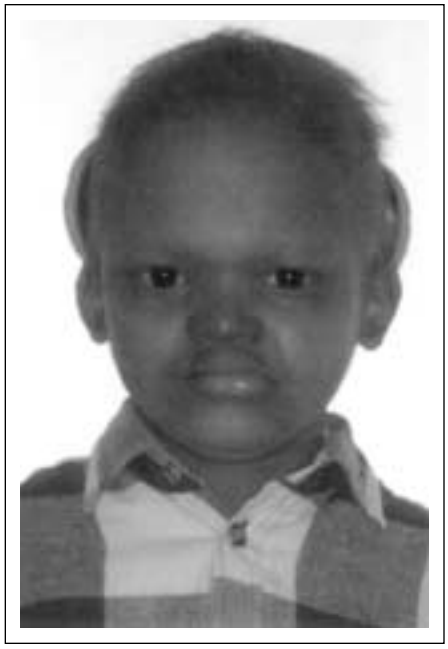

Figure 1. Facial view of the patient showing the characteristic physiognomy.

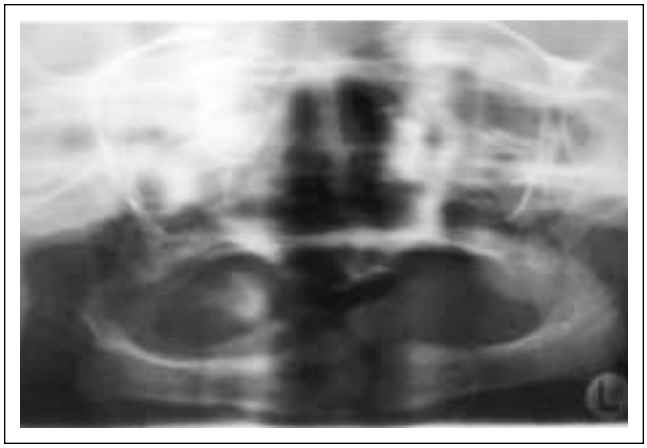

Figure 3. Panoramic radiograph showing complete absence of teeth.

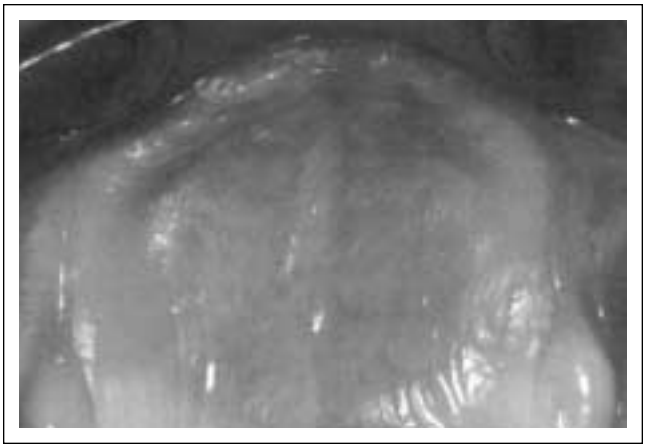

Figure 2. Edentulous upper jaw.

Figure 4. Facial view after treatment.

very thin layer of bone separated the oral cavity from the maxillary sinus.

In order to improve the appearance, mastication, and speech, the child was provided with removable complete upper and lower dentures.

Behaviour management techniques, such as tell-show-do, were used throughout the procedures. Although routine procedures for construction of the dentures were used (Table 1), case specific modifications were required. The glabella bearing was adapted to the depressed nasal bridge with a silicone material. Permanent tooth forms instead of primary tooth forms were used for better static and dynamic occlusion, which improved the balance of the dentures.

Before providing the patient with the dentures the tone of the perioral muscles were trained by a vestibular shield. The vestibular shield is a resin made orthodontic appliance, which fills the space between the dental arch and the lips expanding distally at the most to the region of the first primary molars.[20] The patient would activate the appliance by pulling the extraoral ring of the appliance several times a day.

After the final insertion (Figure 4) routine oral hygiene instructions for dentures were given to the patient and the parents. At the same time a logopaedic therapy has been started. The patient was advised to remove the dentures at night to allow for healing of the oral tissue. At recall appointments, no pressure spots were noticed and the static and dynamic occlusion

Table 1. Routine steps for manufacturing of dentures.

Treatment steps	Materials
diagnostic casts	alginate
custom trays	resin plates
functional modelling of the tray margins	thermoplastic impression compound
modified mucostatic impression	polysiloxane and zinc-oxide eugenol impression material
master casts	plaster
wax rims	
alignment of the upper wax rim to Camper's plane	
vertical intermaxillary relationship	phonetic exercises
facebow transfer	
horizontal and sagittal interocclusal record	aluminum wax, zinc-oxide eugenol
mounting of the casts	semiadjustable articulator
trial fitting	artificial teeth
wax try-in	
heat-curing process	resin
remounting	
final insertion	

showed no interferences. Retention was excellent and the parents reported a significant improvement of his speech and mastication.

The increased self-esteem (Figure 5) improved the socialization skills of the boy.

Further recalls have taken place every three months. After 12 months the dentures still fit well with an excellent occlusion. Future treatment will include modification or replacement of the dentures according to the observed skeletal growth.

DISCUSSION

Concerning the dental symptoms of hypohidrotic ectodermal dysplasia, the Bloch Sulzberger syndrome (incontinentia pigmenti) and the Ellis van Creveld syndrome must be differentiated.[2,4] The child showed no signs of the characteristic hyperpigmentation of the trunk. The observed increased pigmentation in the present case was limited to the area around the eyes and the mouth. Polydactyly and exostosis as described for the Ellis van Creveld syndrome were also not apparent.[2,4]

The decreased vertical facial height as part of the specific appearance[2,5,21-23] of these patients resulted from the congenital absence of teeth.[24] Sarnat and coworkers[25] and Nomura and coworkers,[26] showed that the development of the facial skeleton lies in the lower range of normal.[27] In order to enable a further unrestricted development the dentures have to be adapted to the changing intraoral situation due to growth.[18,26,28]

Figure 5. Extraoral view of the patient with incorporated dentures.

Since alveolar bone development was reported to be dependent on the presence of teeth[29,30] the alveolar ridges on which dentures had to be constructed, were very plain. Franchi and coworkers could not find any significant resorption of the alveolar ridges caused by dentures.[31]

Although a decrease of salivary glands of the intraoral mucosa has been described in the literature,[32,33] in the present case the amount of saliva was sufficient for the adhesion of the dentures.

For favorable development of the stomatognathic system and also for psychologically reasons an early treatment strategy in these cases has been suggested by several authors.[4,5,13-18]

The behaviour management (tell-show-do)[34,35] is important for the success of the treatment concerning the compliance and the prosthetic outcome.[13,16,36]

The most common treatment in cases of ectodermal dysplasia with anodontia is complete dentures[4,5,10,11,13,36-41] as used in the present case. Recently, implant-borne total telescopic dentures have been described as a possible treatment.[15,28] But high costs, difficulties in placement and a high failure rate make use questionable.[18] Analogous to an ankylosed tooth implants cannot erupt and are likely to impede or even stop the normal growth.[42-44] Therefore, this option is not recommended in children before skeletal maturity.[45]

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