CASE REPORT

A simple prosthetic restorative solution of a single peg-shaped upper central primary incisor in a case of ectodermal dysplasia

ANCA MARIA RĂDUCANU[1)], MIHAELA PĂUNA[2)], I. V. FERARU[1)]

[1)]*Department of Pediatric Dentistry*
[2)]*Department of Removable Prosthodontics*
Faculty of Dental Medicine,
"Carol Davila" University of Medicine and Pharmacy, Bucharest

Abstract

Introduction: This clinical report describes the oral rehabilitation of a 7-year-old male diagnosed with hypohidrotic ectodermal dysplasia and anodontia. *Background*: Ectodermal dysplasia (ED) comprises a large, heterogeneous group of inherited disorders, which are defined by primary defects in the development of two or more tissues derived from the embryonic ectoderm (skin, hair follicles, nails, teeth, sebaceous and sweat glands). Our patient exhibited the typical general and cranio-facial features of ED as well as emotional and behavioral problems. The patient presents only one conical primary central incisor. *Clinical management*: The treatment was focused to improve his esthetic needs and oral functions and included the fabrication of removable prostheses and of an acrylic crown on the single tooth existing in his mouth, manufactured by an original simple method. Six months follow-up points out that restorative treatment provided psychosocial and functional comfort to the young boy. *Conclusions*: This clinical report demonstrates the importance of the prosthodontic treatment for oral rehabilitation in children with ED. The partial and complete removable dentures associated with the acrylic crown can be a reversible and inexpensive method of treatment for ED patients.

***Keywords*:** hypohidrotic ectodermal dysplasia, ED, anodontia, prosthodontic treatment, acrylic crown.

Introduction

Ectodermal dysplasia (ED) comprises a large, heterogeneous group of inherited disorders (there may be over 150 different types) [1–3], that are defined by primary defects in the development of two or more tissues derived from embryonic ectoderm [1, 2, 4–7]. ED is present worldwide affecting all races, most often in whites [2].

The most common EDs are X-linked recessive hypohidrotic/anhidrotic ED [2, 5, 7, 8]. It has full expression only in males. Female carriers show little or no signs of the condition [2].

Most of the EDs manifest variable defects in the morphogenesis of ectodermal structures such as the hair, skin, nails, teeth and sweat glands witch produce numerous *general findings* and symptoms, thus the clinical features associated with ED may vary from case to case [7–10].

Characteristic *deformities of the skull are*: square aspect of the forehead, frontal bossing, prominent supra-orbital ridges, saddle nose, the alae nasi appears hypoplastic, the midface is depressed and hypoplastic, the cheekbones are high and broad (malar hypoplasia) and appear flat and depressed as well (dished-in appearance), the chin may be pointed and the lips are thick and everted, cleft lip or palate [3, 11]. The patient may resemble an edentulous old person [3]. The cranio-facial deviations increase with advancing age, in non-treated patients, with a tendency towards a Class III pattern, decreased lower facial height, the maxilla is more retruded as the mandible, the mouth width is smaller than normal [11].

The *dental anomalies* are: number anomalies (anodontia or severe hypodontia) [1, 5, 6, 8], shape anomalies (peg-shaped conical incisors and canines both in deciduous and permanent dentitions, talon cusp, taurodontia) [1, 4, 5, 8], size anomalies (microdontia in both affected males and carrier females) [5], structural anomalies (hypoplasia). Partial or total anodontia results in some loss of function, such as chewing and esthetics [12].

Other oral manifestations are: small or atrophic gums, inflammatory and/or infectious stomatitis, dysphonia, dysphagia, feeding difficulties, hearing impairment [8], xerostomia causes problems with chewing, swallowing food, lack of sense of taste or smell [6, 9, 12].

Patient, Material and Methods

The oral rehabilitation of a 7-year-old male with X-linked recessive anhidrotic ectodermal dysplasia is presented.

The Romanian mother displays some milder clinical manifestations of ED such as: absence of the maxillary lateral incisors, some facial distortions (Figure 1). The Chinese father and his family are completely healthy.

The boy exhibited the typical general, cranio-facial and oral features of ED.

Figure 1 – *The patient's mother – facial and oral aspects.*

General features

The general examination showed typical features: the body skin is abnormally thin, dry and soft, hyperpigmented and wrinkled, appearing prematurely aged, the patient presents unusual scaling or peeling of the skin, hyperkeratosis and chronic eczema on both palms and soles (Figure 2), the body hair is absent, perspiration is absent with frequent episodes of hyperpyrexia, the nails are thin and fragile, chronic respiratory infections, frequent middle otitis, epistaxis, normal intelligence.

Figure 2 – *Hyperkeratosis and chronic eczema on palms and soles.*

Facial and oral features

The patient presented characteristic *facial distortions*: prominent frontal ridges and chin, brittle and fine hair, a saddle shaped nose, an extended philtrum, a thin upper lip, an everted lower lip, a horizontal groove on the chin, thick cheeks and large ears, reduced lower face, dark hair and scanty eyebrows and eyelashes (Figure 3).

Figure 3 – *7-year-old boy – facial and oral aspects. Peg-shaped upper central incisor.*

The oral examination showed the presence of one maxillary cone-shaped tooth (Figure 3). The patient exhibited the loss of facial vertical dimension, the absence of alveolar processes and an abnormal development of alveolar ridges, dysphagia and dysphonia. The absence of teeth had allowed the tongue to spread. The oral mucosa, palate and the floor of the mouth are normal.

Oral treatment

The objectives of the oral treatment were to improve esthetics and correct oral functions, especially mastication, in order to finally allow the normal psychological development.

To accomplish these objectives we decided for a simple conventional prosthetic treatment. Due to the age of the patient and the poor amount of alveolar bone, a partial maxillary removable and a complete mandibular denture were provided.

Alginate (Tropicalgin – Zhermach®) was used for primary impressions due to its rapid-setting qualities and pleasant smell and taste. The impressions were considered acceptable despite their imperfections and the preliminary casts (Figure 4) were poured. The casts were subsequently corrected with blue modeling wax.

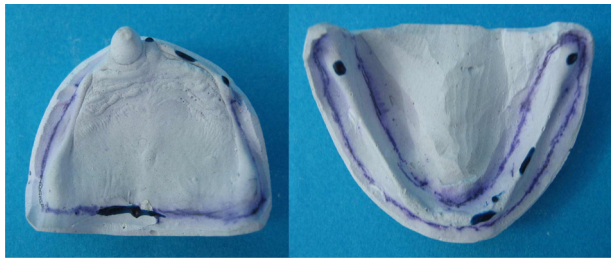

Figure 4 – *Preliminary casts.*

For the secondary impression a polysiloxane (Oranwash – Zhermach®) was used. The impressions were boxed to preserve the periphery created by the muscles.

Occlusion rims constructed on the working casts were transferred in the mouth to establish the occlusal vertical dimension and centric relation. The vertical dimension was first determined by swallowing, checked by the rest position and associated free way space, and finally verified by means of phonetics. The vertical occlusal dimension was increased in order to establish better muscle tonus and lip support.

Acrylic teeth (LuminaAcryl V, Vita Zahnfabrik) with reduced cusps inclines were used and the trial dentures were tested.

The dentures (Figure 5), processed with a conventionnal type resin, were placed into the mouth and the necessary adjustments were carried out (Figure 6). The patient was examined the next day.

Figure 5 – *The maxillary and mandibular dentures.*

Retention and stabilization of the dentures were clinically acceptable. Discomfort areas were relieved. The mother was instructed about the home care of the prosthodontic appliances and the need for periodic

recall for re-evaluation and remanufacture of the dentures as growth occurs.

The patient had no discomfort and seemed to be adapting well on the following weekly visits.

The conical maxillary tooth (1.1) was reconstructed with an acrylic crown made for an optimum esthetic reason, using the homologous acrylic tooth (the remainning acrylic tooth after the anterior teeth arrangement on the wax base), carving it in order to obtain a veneer which was completed on the palatal side with baro-polymerized acrylic resin (Figures 7 and 8). The partial upper denture was configured to accommodate with this crown. The result was not perfect due to the higher location of the gingival margin of the 1.1 crown compared to 2.1, but this was obvious only in a large smile.

Figure 6 – ***Facial aspect after placing of the prosthesis.***

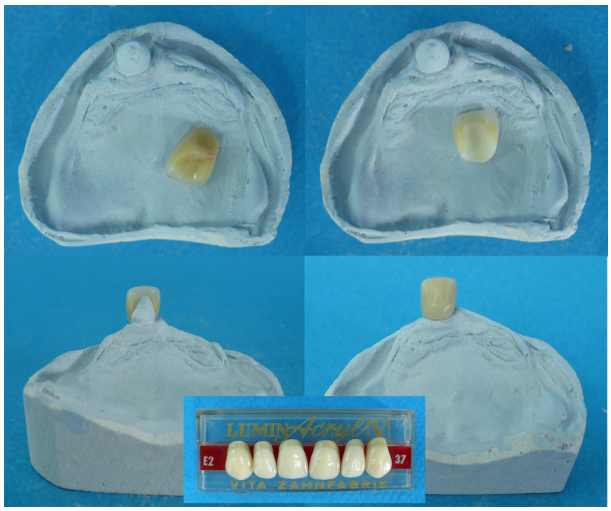

Figure 7 – ***Stages of the reconstruction of the conical maxillary tooth (1.1).***

Figure 8 – ***Final facial and oral aspects.***

The management of our boy with ectodermal dysplasia included instructions for the maintenance of oral hygiene and periodic recall visits.

Discussion

There is no general cure for ED, but *preventive treatment* consisting in genetic counseling and prenatal diagnosis and many treatments addressing the symptoms are available [11, 13, 14].

General treatment may require the coordinated efforts of a multi-disciplinary team of specialists who need to systematically and comprehensively plan an individualized treatment. The general treatment is symptomatic and depends on which ectodermal structures are involved [1, 2, 8, 12]. Medical care also improves esthetics and functions [2, 8].

Dental treatment is usually necessary. Optimal dental treatment requires: early and continuous dental interventions during many years, numerous phases of treatment, the involvement of a multi-disciplinary dental team (oro-maxillo-facial surgeon, orthodontist, prosthodontist, pedodontist) and a marked financial effort of the patients and their families [4, 14].

The main goals of *dental treatment* in patients with ED are enhancement of functions (esthetics, mastication, phonetics) and psychosocial activities [4, 15–17]. Dental treatment is directed towards the specific symptoms that are apparent in each individual [4, 15].

Children may need dentures as early as two years of age. Multiple denture replacements are often needed as the child grows [18], and dental implants may be an option in adolescence. In other cases, teeth can be capped. Orthodontic treatment also may be necessary [14]. In young patients, the main concern of prosthodontic treatment is to enlarge the facial height and depth [19, 20].

Removable prosthesis is the most frequently reported treatment modality for the dental management of ED [19]. However, this is usually a difficult condition to treat with prosthodontic devices [16]. Treatment should be commenced as soon as possible [21] in order to avoid possible resorption and atrophy of the alveolar ridges, and to control the vertical dimension, which can be severely affected by the total or partial lack of teeth [22].

Along the treatment, we confronted with a series of difficulties, caused by the partially uncooperative behavior of the child, the low level of health education in his family, their reduced financial possibilities and the insufficient technical equipment.

The oral rehabilitation being completely dependent on the patient–parent cooperation, behavior management was performed, by the "Tell–Show–Do" method, in all the stages of the prosthodontic treatment.

The difficulty experienced during impression due to the lack of a pair of small impression trays adequate for an edentulous patient was bypassed by overfilling a pair of small tray for dentate patients. The centric relation was registered by simple, non-stressing clinical methods.

The main difficulty consisted in finding a set of very small teeth according to the dimensions of the child edentulous ridges. In the lateral areas, only premolar acrylic teeth were used in order to fit to the reduced anterior-posterior available space. The increase of the vertical occlusal dimension (VOD) was of utmost importance because it radically improved functionality, especially esthetics.

Conclusions

Our pediatric patient with subtotal anodontia was treated by a simple prosthetic method consisting in two conventional dentures and an acrylic crown made by an original, cheap method.

The psychological conditions, the facial profile as well as the functions and esthetics of the patient improved. Notable improvements in the speech and nutritional status of the young patient were obtained.

Continuous evaluation of the patient, in order to monitor the growth, is necessary but our patient abandoned the treatment after a few appointments.

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Corresponding author

Anca Maria Răducanu, Associate Professor, MD, PhD, Department of Pediatric Dentistry, Faculty of Dental Medicine, "Carol Davila" University of Medicine and Pharmacy, 19 Plevnei Street, 010221 Bucharest, Romania; Phone +4021–315 86 49, Mobile +40744–489 595, e-mail: raducanu2000@yahoo.com

Received: August 29th, 2009

Accepted: March 15th, 2010