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Case Report

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Implant-supported Oral Rehabilitation in Child with Ectodermal Dysplasia — 4-year Follow-up

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Abstract

Ectodermal dysplasia (ED) is an anomaly determined by genetic factors that alter ectodermal structures such as skin, hair, nails, glands, and teeth. Children affected by this condition require extensive, comprehensive, and multidisciplinary treatment. An 8-year-old female patient visited the Dentistry Clinic of the Federal University of Santa Catarina with the chief complaint of multiple missing teeth. The mother reported that the patient had ED. Clinical and radiographic examination revealed the congenital absence of several primary and permanent teeth and tooth germs. Subsequent oral rehabilitation comprised the application of a maxillary denture and mandibular implant-supported fixed prosthesis. The child was also supplied with a wig for further enhancement of esthetics aimed at improving her emotional wellbeing. Psychological follow-up and speech therapy were also provided. After 4 years of follow-up, implant-supported oral rehabilitation has proved to be a satisfactory treatment option, allowing restoration of masticatory, phonetic, and esthetic function, as well as an improvement in the patient's self-esteem and social wellbeing.

Key words: Child — Ectodermal dysplasia — Oral rehabilitation — Dental implant

Introduction

Ectodermal dysplasia (ED) is defined as a hereditary condition in which two or more anatomic structures derived from the ectoderm either undergo alterations during development or do not develop at all. It comprises a group of congenital disorders that

affect a range of tissues, including the glands (sudoriferous, sebaceous, lacrimal, mucosal, and salivary), skin, hair, nails, and/or teeth[17,18]. This rare syndrome affects approximately one in 100,000 live births, with the proportion of male to female at five to one. Generally, it is manifested in males, but transmitted by females[5].

The most common oral manifestations are hypo- or anodontia of both the primary and permanent dentition; teeth with morphological (conoid, sharp incisors, molars with a small crown diameter) or developmental anomalies (enamel hypoplasia); and compromised development of alveolar bone, resulting in a reduction in the vertical dimension, which gives the patient a senile appearance[17,18]. The mucosa is usually dry and salivary secretion is reduced due to hypoplasia of the salivary glands, which may be a predisposing factor for caries[10].

The diagnosis is essentially clinical and sometimes determined in infancy if the patient presents with a fever of obscure origin[10]. An early diagnosis is crucial in allowing steps to be taken to ensure the normal physical, emotional, and social development of the child, as such patients are generally withdrawn, timid, and bothered by their abnormal appearance and missing teeth[1]. The oral conditions imposed by this syndrome necessitate multidisciplinary dental treatment aimed at achieving balanced development of both the functional and esthetic aspects of the dentition as far as is possible[12].

The purpose of this report is to describe oral rehabilitation comprising complete implant-supported dentures for the mandible and a regular denture for the maxilla in a girl with hypodontia due to ectodermal dysplasia.

Case Presentation

An 8-year-old girl was referred to the Department of Pediatric Dentistry at our institution with the main complaint of multiple missing teeth. Her mother reported that the patient was currently under medical care elsewhere and that her condition had been diagnosed as ED; she also reported the same syndrome in other family members (cousins). The patient showed characteristic signs of the condition: light-colored hair, which was thin and dry; thin and sparse eyebrows and lashes; dry skin; prominent lips; and a senile appearance (Fig. 1). Informed consent was obtained from the parents for scientific publication of this case.

Fig. 1 Patient at age of 8 years with senile appearance

Clinically, the dentition was characterized by microdontia, a high degree (3) of mobility, and some evidence of pulp necrosis (Fig. 2A–C). Measurements with a Willis compass revealed that the lower third of the face was proportionally smaller than the middle third. A phonetic test was used to confirm this finding. The upper labial frenulum was hypertrophic and had low insertion. The child also exhibited phonetic alteration and chewing difficulties.

Radiographic examination revealed the congenital absence of several primary and permanent teeth and tooth buds, deficient root development, and a large pulp chamber (Fig. 3). Complementary examination including carpal, antero-posterior, and lateral cephalometric radiographs, and cone beam computed tomography (CBCT) was used to determine the stage of bone development in order

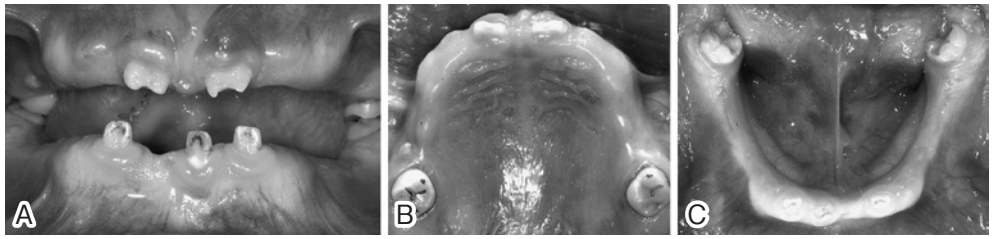

Fig. 2 Intraoral photograph showing severe hypodontia in both maxillary and mandibular arch

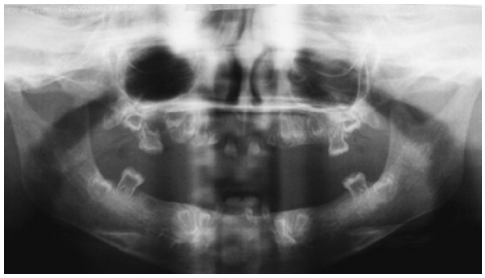

Fig. 3 Panoramic X-ray
Note absence of several primary and permanent teeth.

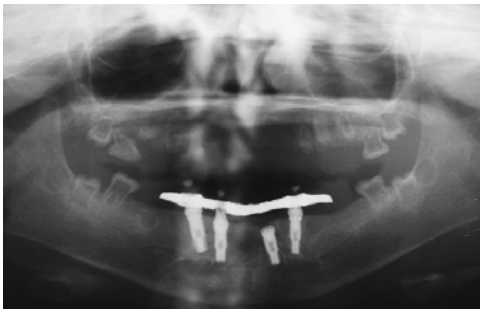

Fig. 4 Panoramic X-ray after implant placement procedure
Insertion torque of approximately 40 N/cm was not achieved in only one implant.

to plan the placement of dental implants.

Clinical Procedures

Prior to implant placement and fabrication of maxillary and mandibular dentures, the low insertion of the upper labial frenulum was surgically removed and maxillary infected teeth extracted.

Functional impressions of the maxilla and mandible were obtained using polyether impression material (Impregum Soft, 3M ESPE, USA), custom open trays, and impression abutments. Base plates and wax rims were used to register the occlusal relations. Rehabilitation with complete fixed implant-supported dentures was not indicated as the patient had a reduced vertical dimension. The dimension was therefore scheduled to be increased to eliminate phonetic difficulties. To achieve this, the artificial teeth were proofed in wax to allow verification of occlusion and the relationship between the upper and lower lips; phonetic and swallowing tests were given and esthetics confirmed. All this was carried out in a single session.

The patient was called in again 1 week later for delivery of the complete maxillary denture and for a test of a lower multifunctional surgical stent. Surgical placement of the implants was carried out under local anesthesia (Articaíne 100, Articaína HCl 4% + Epinefrina 1: 100.000, 72 mg + 18 μg/carpule, 1.8 ml, DFL, Brazil). This comprised a horizontal incision on the crest of the lower ridge with a number-15C scalpel (Swann-Morton, England), followed by complete detachment of the vestibular and lingual flaps. All the mandibular anterior teeth and were extracted together with the tooth buds. The implant sites were then prepared with sequential drill bits up to 2.0 mm in diameter, deepening the

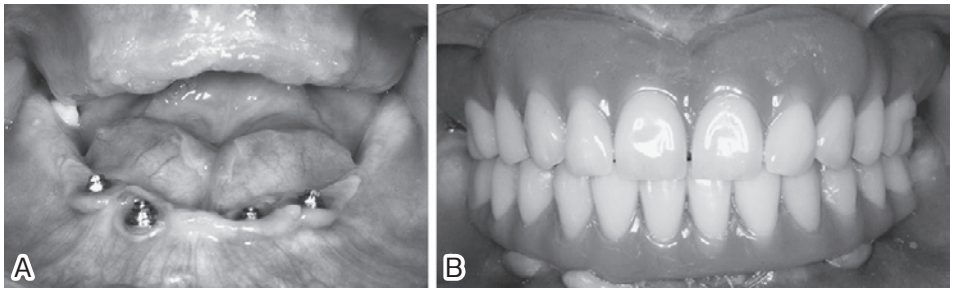

Fig. 5 A: Intraoral view of 4 implants placed, B: Extraoral view of prosthesis.

Fig. 6 New implant-supported mandibular denture segmented in midline showing almost no bone alteration between mandibular foramens

sites until they reached the length of the implants. Two implants measuring 3.75 mm in diameter and 11 mm in length and 2 more implants measuring 4.0 mm in diameter and 11 mm in length (Conexão, Brazil) were installed between the mental foramens, with an approximately 40 N/cm insertion torque, which enabled immediate loading (Fig. 4). Implant size was determined after examining CT scans and bone density intraoperatively. An insertion torque of approximately 40 N/cm was not obtained in only one implant, making it unsuitable for initial use as a support for the denture.

Conical mini-abutments 1 mm in height were installed (Conexão) and transfer impression procedures performed. Square copings (Conexão) were installed and bonded with Duralay resin (Reliance Dental Mfg., USA). The template copings were then bonded and polyether (Impregum Soft, 3M ESPE) impressions of the mandible obtained for construction of the plaster casts. The casts were mounted on a semi-adjustable articulator following inter-occlusal registration by means of a multifunctional template. Next, a metal structure was fabricated on the plaster cast of the jaw. The marginal adaptation of this temporary metal bar was evaluated and acrylic resin then applied. The definitive mandibular fixed implant-supported denture was installed and occlusal adjustments made (Fig. 5A, B). Next, the mother and child received instructions on oral hygiene and prosthesis placement, which emphasized flossing and the maintenance of full maxillary prosthesis hygiene. The patient reported good adaptation to the dental prosthesis and satisfaction with the treatment; the resultant phonetics, esthetics, and masticatory function were all good. The concomitant increase in self-esteem also resulted in an improvement in her social skills.

Fig. 7 Clinical state after 2 years
Note anterior open bite of superior prosthesis with right mandibular molar.

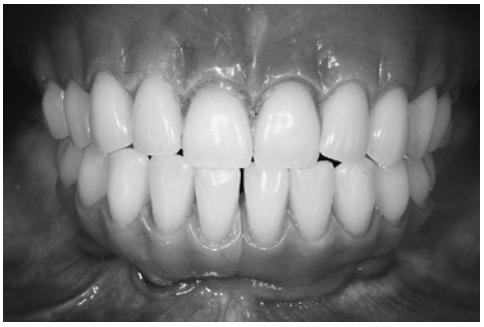

Fig. 8 Prosthesis *in situ* after occlusal adjustment

Fig. 9 Extraoral view of patient at 4-year follow up

Fig. 10 Panoramic X-ray at 4-year follow up

Outcomes

After 1 year, both the maxillary and mandibular dentures were replaced at the request of the patient who reported that the old ones had become uncomfortable. These were fabricated using the same techniques as described above. Even though there is usually almost no alteration in bone between the mandibular foramens at this stage, the new implant-supported mandibular denture was segmented at the midline to allow evaluation of potential transversal growth over the next few years (Fig. 6A–C).

The patient makes daily use of the dentures, which have provided improvements in esthetics and speech. In the present case, implant therapy was central to achieving predictable functional and aesthetic rehabilitation. The mother of the child subsequently reported that the dietary intake and psychological wellbeing of the child had improved greatly after insertion of the prosthesis. The girl is currently undergoing periodic clinical and radiographic follow-up examinations to monitor the implants and dentures together with other aspects of her development, including bone growth, as the dentures will eventually have to be replaced as she continues to age. In addition, the girl was also provided with a wig to further improve esthetics and enhance her emotional wellbeing. Psychological follow-up and speech therapy is also being given, which will all help her adapt to her new physical and oral conditions and social environment.

At a 2-year follow-up examination, change was observed in occlusion due to vertical growth in the posterior region of both the maxilla and mandible (Fig. 7). Therefore, the dentures were repositioned and occlusal adjustment performed (Fig. 8).

The patient is currently coming in for follow-up examinations at 6-month intervals. The girl has rehabilitated easily and maintains excellent oral hygiene. The mother has reported great changes in her dietary intake. At a 4-year follow-up, excellent results were observed in terms of esthetics, speech, and function (Figs. 9 and 10).

Discussion

In the present case, the clinical characteristics of a patient with ED were evident: thin, light-colored, sparse hair; smooth, dry, thin skin; and missing teeth. The genetic pattern of the present case was also characteristic, as the mother reported that the patient had two female cousins with the same syndrome, although with less severe characteristics. The deficient root development and large pulp chamber in the present case are not commonly described in the literature[10] and may be associated with the fact that the syndrome compromises structures of ectodermal origin.

Treatment complexity is directly related to patient age and degree of severity[5,14]. In children, the most conservative treatment approach is to wait for completion of skeletal and dental growth before formulating an implant-based prosthetic treatment plan[2]. However, physiological and psychological factors favor implant placement before completion of skeletal growth. Psychological well-being and good nutrition are the most important reasons for placing an implant in a growing individual. In addition, excellent blood supply and good bone healing lead to increased stability and retention of prosthetic rehabilitation in patients during the growth phase[13,19].

Maxillary development occurs through apposition and surface remodeling[6], with the latter taking place as growth proceeds downward and forward in relation to the cranium and base of the skull. It should be borne in mind that the largest portion of the anterior surface of the maxilla is an area of resorption rather than apposition[6,16].

Mandibular growth, on the other hand, proceeds nearly exclusively through backward and upward growth of the condyle and backward growth of the ramus in order to allow eruption of the molars through resorption of the anterior portion of the ramus and apposition in the posterior portion of the ramus. As a result, anterior mandibular width stabilizes relatively early and only increases weakly through appositional growth[6,16]. As the symphysis begins to close a few months after childbirth, there is no risk of implant surgery traumatizing the growth site, and there is little possibility that dentures crossing the median line will limit transverse growth[15,19].

In the present case, undeveloped alveolar bone and muscle activity compromised retention of the conventional removable denture, so an endosseous implant-retained removable overdenture was planned. The implants were subsequently positioned in the anterior region of the mandible, between the mental foramens, an approach which has proved highly successful in previous studies[10,19].

According to the literature[11], there is no ideal chronological age for the placement of endosseous implants. The most important factor determining the ideal time to place implants may be level of skeletal maturity. However, in children, early implant placement may be considered in severe cases of ED in order to minimize poor functioning of removable dentures[20]. Implant placement in patients 5–6 years of age has been reported in the literature[3,7]. Implants placed in 3-dimensional growing bone have been described as behaving like ankylosed primary teeth, and eventually become submerged due to growth associated with the continued eruption of neighboring natural teeth[8,9].

Due to disease, the present patient had no teeth adjacent to the implants, so the risk of

the implants becoming submerged was lower. So far, at 4 years postoperatively, all the implants continue to show favorable positioning and osseointegration. Regular postoperative follow-up visits are essential in young patients with ED, as the prostheses must be changed in line with new growth and development[5]. The present patient came in for such examinations every 6 months over a 4-year period, during which time the maxillary complete dentures were changed twice and the implant supported prosthesis given occlusal adjustments. Besides improvements in esthetic and functional factors, an increase is also soon observed in self-esteem in patients with ED following oral rehabilitation. Here, both the patient and her mother were very satisfied with the outcome of the oral rehabilitation and wig. The patient's previously senile appearance improved, which contributed to her physical, psychological, and social wellbeing.

A number of case reports and cohort studies have described the use of implants to support a mandibular prosthesis in children with ED. Studies with different follow-up periods (ranging from 9 months to 12 years) reported low rates of failures[4]. One retrospective study examined 61 implants in 14 adolescents and young adults at 1–5 years postoperatively and reported a success rate of 67%[4]. A prospective study of 51 patients followed for up to 78 months reported 91% survival rates in the mandible and 76% in the maxilla[4]. Taken together with these previous studies, the present results suggest that continued use of implants in young children is feasible as long as it does not interfere with mandibular and/or maxillary growth.

As there is no standard protocol or even a general consensus among researchers regarding the procedures and indications for placement of conventional or temporary dental implants, all risks and benefits should be assessed on a case-by-case basis and clinicians should select the technique to be employed based on common sense, knowledge, and professional experience.

The placement of dental implants and complete dentures in children with ectodermal dysplasia is a viable rehabilitation alternative that allows restoration of masticatory, phonetic, and esthetic function, as well as improvement in the patient's self-esteem and social wellbeing.

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