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Ectodermal dysplasia treated with one step surgical rehabilitation: a case report

ABSTRACT

Ectodermal dysplasia (ED) comprises a large, heterogeneous group of inherited disorders that are defined by primary defects in the skin, hair, nails, eccrine glands, and teeth. The most characteristic findings are the reduced number of teeth. Any rehabilitative program involves the correct evaluation of skeletal relationships. Implant-prosthetic treatment can be performed at the end of bone growth. In this paper a case of ED treated with Le Fort I for maxillary advancement, femur homografts, implants' insertion and immediate loading is reported. In December 2007, a 38 year-old female was referred to the Maxillofacial Department of Galeazzi Hospital (Milan, Italy) who had a diagnosis of ED. Twelve implants were inserted in one step surgical procedure. To evaluate the clinical outcome several variables (related to anatomy, implant, and prosthesis) were investigated. Implant failure and peri-implant bone resorption were considered as predictors of clinical outcome. Kaplan-Meier algorithm and Cox regression analyses were then performed to detect those variables statistically associated with the clinical outcome. The occlusion was stable after 15 months follow-up. No implants were lost. Only one implant has a crestal bone resorption higher than the cut-off value (i.e. one clinical failure over 12 implants, success rate = 91.6%). None of the studied variable has impact on the clinical outcome. In the present case, one step oral rehabilitation was stable in term of occlusion and implant outcome after 15 months. This procedure could be performed in adults with ED, significantly reducing the time of oral and facial rehabilitation.

KEYWORDS Alveolus; Bone; Fixture; Remodelling; Resorption; Ridge.

INTRODUCTION

Ectodermal dysplasia (ED) comprises a large, heterogeneous group of inherited disorders that are defined by primary defects in the development of 2 or more tissues derived from embryonic ectoderm. The tissues primarily involved are the skin, hair, nails, eccrine glands, and teeth (1).

This condition is classified as 2 major types: hypohidrotic, in which the sweat glands are absent or significantly decreased and hidrotic, in which the sweat glands are normal. Hypohidrotic ED is the more severe form and it affects males more than females. It is associated with sensitivity to heat and frequent high fevers (2). The time of clinical detection varies from birth to childhood, depending on the severity of symptoms and associated complications, but the diagnostic tool is the typical clinical physiognomy (3). Men have an easily recognizable *facies*, also referred to as an 'old man' faces. The forehead appears square, with frontal bossing, and there is a prominent supra-orbital ridge. The nose has a depressed nasal bridge and is called saddle nose. The midface is depressed and hypoplastic, giving it a "dished-in" appearance. The cheekbones are high and broad, although they appear flat and depressed as well. The chin may be pointed and the lips exerted and protuberant (4). Some infants have a premature look with scaling of the skin. This can also form a clue to the diagnosis: the number of sweat glands is reduced and both scalp and body hair are scarce, with lack of eyebrows and eyelashes (3, 4). This remarkable variability in facial dimensions and harmony found in patients with ED probably corresponds to the different kinds of dysplasia, with different expression of the interested genes (5).

The most characteristic findings in man are the reduced number and abnormal shape of teeth. The dental findings of ED range from complete anodontia

to hypodontia of the primary or permanent teeth with or without cleft lip and palate. The delay in teething is often the first step in the diagnosis. However, if teeth are present, they may be tapered, malformed, and/or widely spaced. Congenital absence of teeth is more frequent in the lower jaw and affects the growth of the jawbones, leading to a lack of alveolar bone in both height and width (3, 6). Early dental treatment of patients with ED is necessary, because it gives the child the opportunity to develop normal functions of speech, chewing, and swallowing as well as normal facial support. It improved also temporo-mandibular joint function and self-esteem (7). The course of the treatment is to restore the function and the aesthetics of the teeth, normalize the vertical dimension and support the facial soft tissues (8). Any rehabilitative program involves the correct evaluation of skeletal relationships, which will eventually be corrected using the appropriate orthodontic techniques. The use of a total or partial removable prosthesis or overdentures is often the initial treatment of choice (1). However, prosthetic-implantological treatment at the end of bone growth must be implemented, with the possibility of restoring *ad integrum* the patient's masticatory function and aesthetics (9). In this article a case of ED treated with Le Fort I for maxillary advancement, femur homografts, implants' insertion and immediate loading is reported.

MATERIALS AND METHODS

A 38 year-old female was referred to the Maxillofacial Department of Galeazzi Hospital (Milan, Italy) who had a diagnosis of ED in December 2007. Informed written consent, approved by the local Ethics Committee, was obtained from the patient to use her data for research purpose.

The patient is neither a smoker nor drinker and she is a nurse. She had no previous major operation in the head or neck region. Examination of the oral cavity showed the presence of two lateral incisors, two canines, one right premolar and one molar in the mandible (Fig. 1). Faces and skin annex were typical for ED and the clinical history was suggestive for a hidrotic ED (no high fever episode). Orthopantomography and lateral telerradiography (Fig. 2 and 3) were performed and impressions and custom model splint for maxilla reposition were taken.

An operation was planned to insert implants in the upper and lower jaws (Fig. 4): after placement of the surgical guide, a mucotomy was performed, the bone drilled and implants inserted (Neoss S.r.l., Milan, Italy). The implant platform was positioned at alveolar crest level. Then, a maxillary



Fig. 1 Frontal view of the occlusion.



Fig. 2 Presurgical panoramic radiograph.

advancement by means of a Le Fort I osteotomy, temporary inter-maxillary fixation (Fig. 5), femur homografts (banked from living donors, Bone Bank of Orthopaedic Institute Gaetano Pini, Milan, Italy) insertion in the osteotomic gaps, and internal rigid fixation were performed. The provisional restoration was immediately delivered and after 8 weeks the final restoration was applied.

The immediate postoperative clinical (Fig. 6) and radiological controls (Fig. 7 and 8) demonstrated a successful outcome. The 15 months follow-up demonstrated the stability of the result (Fig. 9-11).

Variables

Several variables were investigated: anatomic (i.e. maxilla and mandible, tooth site), implant (i.e. length and diameter), surgical (i.e. grafted and non-grafted site) and prosthetic (i.e. loaded and unloaded fixtures) variables.

Primary and secondary predictors of clinical



Fig. 3 Presurgical telerradiography.

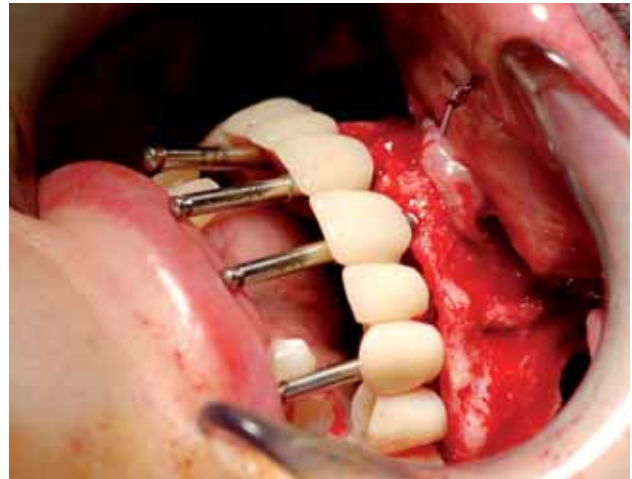


Fig. 4 Implant inserted by means a customized provisional prosthesis.



Fig. 5 The temporary intermaxillary rigid fixation.



Fig. 6 The occlusion at the end of the operation.

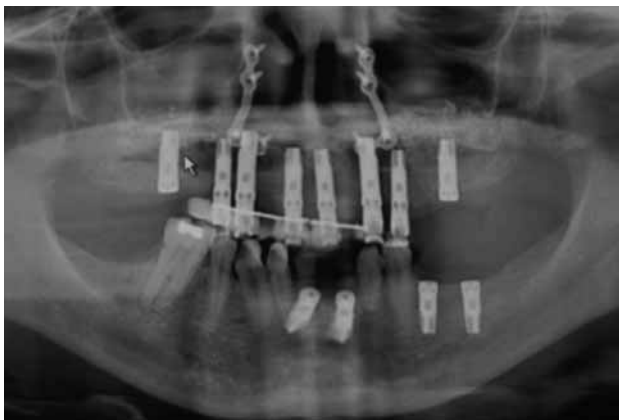


Fig. 7 The immediate postsurgical panoramic radiograph.

Fig. 8 The immediate postsurgical telerradiography.



outcome were used. The primary predictor was the presence/absence of the implant at the end of the observation period. The defined survival rate (i.e. SVR) was the total number of implants still in place

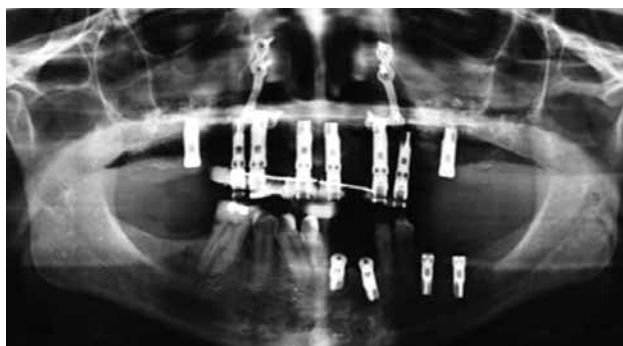


Fig. 9 The 15 months follow-up panoramic radiograph.



Fig. 11 The 15 months follow-up frontal view of the occlusion.

at the end of the follow-up period.

The second predictor of outcome was peri-implant bone resorption. It was defined as the implant success rate (SCR) and was evaluated according to the absence of persisting peri-implant bone resorption at a rate greater than 1.5 mm during the first year of loading and 0.2 mm/year during the following years (10).

Data collection methods and summary of operative methods

Before, after surgery and at the end of the follow-up radiographic examinations were performed. Peri-implant crestal bone levels were evaluated by the calibrated examination. Measurements were recorded after surgery and at the end of the follow-up period. The measurements were carried out medially and distally to each implant, calculating the distance between the implant platform and the most coronal point of contact between the bone and the implant. The bone level recorded just after the surgical insertion of the implant was the reference point for the following measurements. The measurement was rounded off to the nearest 0.1 mm. The radiographs were taken using a long x-ray tube at 70 Kw of power, transferred to a computer system (Gendex, KaVo ITALIA srl, Genova) and saved in uncompressed TIFF format for



Fig. 10 The 15 months follow-up teleradiography.

classification. Each file was processed with the Windows XP Professional operating system using Photoshop 7.0 (Adobe, San Jose, CA), and shown on a 17" SXGA TFT LCD display with a NVIDIA GÈ Force FX GO 5600, 64 MB video card (Acer Aspire 1703 SM-2.6). Starting from the implant dimensions, it was possible to establish the distance from the medial and distal edges of the implant platform to the point of bone-implant contact (expressed in tenths of a millimetre).

The difference between the implant-abutment junction and the bone crestal level was defined as the Implant Abutment Junction (IAJ) and calculated at the time of operation and at the end of follow-up. The delta IAJ is the difference between the IAJ at the last check-up and the IAJ recorded just after the operation. Delta IAJ medians were stratified according to the variables of interest.

Peri-implant probing was not performed because controversy still exists regarding the correlation between probing depth and implant success rates (11, 12).

Data analysis

Disease-specific survival curves were calculated according to the product-limit method (Kaplan-Meier algorithm) (13). Log rank testing was used to compare survival/success curves, generated by stratifications for a variable of interest.

Cox regression analysis was then applied to determine the single contribution of covariates on the survival/success rate (14). Stepwise Cox analysis allowed us to detect the variables most associated with implant survival and/or clinical success.

RESULTS

There were a total of 12 fixtures. The mean follow-up after implant insertion is 15 months: six implants

Jaws	Graft	Implant site	Implant length (mm)	Implant diameter (mm)	Prosthetic Type
Mandible 8 (0.3)	Yes 8 (0.3)	Incisors 19 (2.3)	3.5 mm 2 (2.0)	3.5 mm 2 (0.4)	Yes 6 (0.3)
Maxilla 4 (1.2)	No 4 (1.2)	Cuspids 15 (2.2)	13.0 mm 10 (0.3)	4.0 mm 10 (0.6)	No 6 (0.8)
-	-	Premolars 89 (1.8)	-	-	-
-	-	Molars 85 (2.0)	-	-	-

Table 1 Distribution of series the number of cases is out of parenthesis whereas the median delta IAJ is in parenthesis.

were immediately loaded and 6 (2 upper molars and 4 inserted in the mandible) are still not prosthodontized (because of economic problem of patient). The mean post loading follow up is 7.5 months.

A total of 12 (Neoss S.r.l., Milan, Italy) were inserted: 8 into the maxillae and 4 into the mandible. Two measures of fixture length and diameter were used: 11.5 and 13 mm and 3.5 and 4.0 mm, respectively. Implants were inserted to replace 4 incisors, 2 cuspids, 3 premolars and 3 molars. The gap of maxillary osteotomy was filled with femur graft whereas no graft was placed in the mandible. Six over 8 fixtures inserted in the upper jaw were immediate loaded.

No implant was lost (i.e. SVR = 100%) and thus neither univariate nor multivariate analysis showed any differences among the studied variables.

Table 1 reports the median delta IAJ according to the studied variables.

The Kaplan Meier algorithm (i.e. univariate analysis) demonstrated that only implant length is potentially associated with crestal bone resorption (degree of freedom = 1, p value = 0.0253). However, Cox algorithm (i.e. multivariate analysis) did not confirm the result so that none of the studied variables is statistically associated to the clinical outcome.

DISCUSSION

Dental implants have become an accepted treatment modality for aging patients with either completely or partially edentate jaws (15). However, in partially edentulous children who have ED, multiple implant placement is not possible because of the ongoing development of the jaws and insufficient bone. In addition, the bone height and width will not be sufficient for implant insertion without advanced surgical approaches (16). In non-treated patients

with ED, craniofacial deviations from the norm increased with advancing age (17) with a tendency toward a Class III pattern with anterior growth rotation (3). Cephalometric analysis and anthropometry studies show reduced facial dimensions, decreased lower facial height and variable pattern in facial widths. The maxilla has relatively more retruded than the mandible, the nasal alar width and mouth width are significantly smaller (18). In the mandible, sufficient bone may be available only at the mid-symphysial area, where one implant could provide stability for the mandibular denture (19). However, the maxilla of totally edentulous patients frequently requires bone grafting procedures or Le Fort I and grafting (20) before implant insertion, because may affect their retention and stability.

Orthognathic surgical procedures are the treatment of choice in ED patients who exhibit maxillary and/or mandibular skeletal deficiency (21). It has been postulated that the Le Fort I osteotomy that restores normal maxillomandibular relationship, might restimulate maxillary growth (22, 23). First, the jaws must be positioned correctly in relation to each other and the cranium by means of orthognathic surgery; only then can implant placement with autogenous or heterologous bone grafts be attempted (24).

Although these surgical procedures can be performed in most ED patients, removable dentures are usually the only viable treatment alternative when the expenses and morbidity of the surgical procedures are taken into account (21, 25).

In the present report we describe a case of ED treated with simultaneous implant placement, Le Fort I advancement, grafting of maxillary gaps and provisional immediate rehabilitation of upper jaw. Although implants were inserted also in the mandible, these lasts are still not prosthodontized because of economic problem of the patient. The

mandible is bearing removable dentures. The one step oral rehabilitation is stable in term of occlusion and implant outcome after 15 months follow-up. This procedure can be performed in adults. It significantly reduces the time of oral and facial rehabilitation without compromise the medium term clinical outcome also in patients with ED.

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