CASE STUDY

PROSTHETIC REHABILITATION WITH DENTAL IMPLANTS AFTER ILIAC CREST BONE GRAFTING ON A PATIENT OF ECTODERMAL DYSPLASIA WITH PARTIAL ANODONTIA: A CASE REPORT

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ARTICLE INFO	ABSTRACT
	Ectodermal dysplasia (ED) is a hereditary disorder associated with developmental disorders of two or more structures of ectodermal embryonic origin. The triad of nail dystrophy, alopecia or hypotrichosis and palmoplantar hyperkeratosis is usually accompanied by a lack of sweat glands. Varying degrees of Hypodontia or anodontia of the primary and permanent dentition poorly developed alveolar ridges and improper maxillo - mandibular relations, are the most common oral manifestations. It usually affects males and females are the carriers. **Case presentation:** A 28 year-old young man with hypohidrotic ectodermal dysplasia (HED) presented in this article, had typical features of HED: hypohidrosis, hypotrichosis, severe hypodontia, atrophic alveolar ridges, old-looking facial expression. According to the patient's age and clinical findings, implants supported fixed prosthesis in both arches was the treatment of choice.

INTRODUCTION

Ectodermal dysplasia represents a group of inherited conditions in which two or more ectodermally derived anatomic structures fail to develop. Thus depending on the type of ectodermal dysplasia, hypoplasia or aplasia of tissues (e.g., skin, hair, nails, teeth, and sweat glands) may be seen. The various types of this disorder may be inherited in any one of several genetic patterns, including autosomal dominant, autosomal recessive, and X-linked patterns. Thurman published the first report of a patient with ED in 1848, but the term was not coined until 1929 by Weech. Freire-Maia and Pinheiro described numerous varieties of ectodermal dysplasia involving all possible Mendelian modes of inheritance. Freire-Maia and Pinheiro proposed the first classification system of the ectodermal dysplasias in 1982, with additional updates in 1994 and 2001. Their original classification system stratified the ectodermal dysplasias into different subgroups according to the presence or absence of (Nevill *et al*., 2008) hair anomalies or trichodysplasias, (Buyse, 1990) dental abnormalities, (Freire-MaiaandPinheiro,1998) nail abnormalities or onychodysplasias, and (Pinheiro and Freire-Maia, 1994) eccrine gland dysfunction or dyshidrosis.

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With the recent identification of the causative genetic defect for a number of the ectodermal dysplasias, newer classification systems have been devised. In 2003, Lamartine reclassified the ectodermal dysplasias into the following 4 functional groups based on the underlying pathophysiologic defect:

- Cell-to-cell communication and signaling
- Adhesion
- Development
- Other

Similarly, in 2001, Priolo and Laganà reclassified the ectodermal dysplasias into 2 main functional groups:

- Defects in developmental regulation/epithelial mesenchymal interaction
- Defects in cytoskeleton maintenance and cell stability.

Other classification systems categorize the ectodermal dysplasias based on defects in cell-cell communication and signalling, adhesion, transcription regulation, or development. Several ectodermal dysplasia syndromes may manifest in association with midfacial defects, mainly cleft lip, cleft palate, or both. The three most commonly recognized entities are

- Ectodermal dysplasia, ectrodactyly, and clefting (EEC) syndrome;

- Hay-Wells syndrome or ankyloblepharon, ectodermal dysplasia, and cleft lip/palate (AEC) syndrome;
- Rapp-Hodgkin syndrome. Ectodermal dysplasias is usually transmitted as an X linked recessive trait in which gene is carried by female and manifest in male. The prevelence of population has been assessed as between 1:10000 and 1:100000 live male birth.

Even though by some accounts more than 170 different subtypes of ectodermal dysplasia can be defined, these disorders are considered to be relatively rare, with an estimated frequency of seven cases occurring in every 10,000 births. For fewer than 20% of these conditions, the specific genetic mutations and their chromosomal locations have been identified. Systematically classifying these conditions can be challenging because of their wide-ranging clinical features; however, some investigators have suggested that a classification scheme based on the molecular genetics alteration associated with each type might be appropriate. Thus groups of ectodermal dysplasia syndromes could be categorized as being caused by mutations in genes encoding cell-cell signals, genes encoding adhesion molecules, or genes regulating transcription. Perhaps the best known of the ectodermal dysplasia syndromes is hypohidrotic ectodermal dysplasia. In most instances, this disorder seems to show an X-linked inheritance pattern, with the gene mapping to Xq12-q13.1; therefore, a male predominance is usually seen.

- X-linked hypohidrotic ED has been mapped in the proximal area of the long arm of band Xq-12-q13.1. Decreased expression of the epidermal growth factor receptor has been proposed as playing a causal role in this condition's phenotype. The gene ED1 responsible for the disorder has been identified.
- Autosomal recessive disorders, phenotypically indistinguishable from the X-linked forms, exist in humans. A candidate gene has recently been identified at the dl locus (downless) that is mutated in mice.
- The gene that causes hidrotic ED (Clouston syndrome) has been identified to be GJB6, which encodes for connexin-30. GJB6 has been mapped to the pericentromeric region of chromosome 13q.
- Mutations of the gene PVRL1, encoding a cell-to-cell adhesion molecule/herpesvirus receptor, have been reported in those with cleft lip/palate ED.

However, a few families have been identified that show autosomal recessive or autosomal dominant patterns of inheritance. Affected individuals typically display heat intolerance because of a reduced number of eccrine sweat glands. Sometimes the diagnosis is made during infancy because the baby appears to have a fever of undetermined origin; however, the infant simply cannot regulate body temperature appropriately because of the decreased number of sweat glands. Uncommonly, death results from the markedly elevated body temperature, although this generally happens only when the condition is not identified. Sometimes, as a diagnostic aid, a special impression can be made of the patient's fingertips and then examined microscopically to count the density of the sweat glands. Such findings should be interpreted in conjunction with appropriate age-matched controls. Other signs of this disorder include fine, sparse hair, including a reduced density of eyebrow and eyelash hair. The periocular skin may show a fine wrinkling with hyperpigmentation, and midface hypoplasia is frequently observed, often resulting in protuberant lips. Because the salivary glands are ectodermally derived, these glands may be hypoplastic or absent, and patients may exhibit varying degrees of xerostomia. The nails may also appear dystrophic and brittle. The number of hair follicles, sweat glands, and sebaceous glands varies. Symptoms of a reduction in hair follicles vary from sparse scalp hair (usually short, fine and dry) to a complete absence of hair. Hair bulbs may be distorted, bifid, and small. Eccrine sweat glands may be absent or sparse and rudimentary, particularly in those with hypohidrotic EDS. In some cases, mucous glands are absent in the upper respiratory tract and in the bronchi, esophagus, and duodenum. The mouth may be dry from hypoplasia of the salivary glands; lacrimal glands also may be deficient. Teeth Show abnormal morphogenesis or are absent. Nails are often brittle and thin or show abnormal ridging, but they may be grossly deformed. Other signs and symptoms like lack of breast development, deficient hearing or vision, cleft lip and/or palate and missing fingers or toes are also seen. The presence or absence of these abnormalities defines the different syndromes.

The teeth are usually markedly reduced in number (oligodontia or hypodontia), and their crown shapes are characteristically abnormal. The incisor crowns usually appear tapered, conical, or pointed, and the molar crowns are reduced in diameter. Complete lack of tooth development (anodontia) has also been reported, but this appears to be uncommon. Female patients may show partial expression of the abnormal gene; that is, their teeth may be reduced in number or may have mild structural changes. This incomplete presentation can be explained by the Lyon hypothesis, with half of the female patient's X chromosomes expressing the normal gene, and the other half expressing the defective gene.

Following are the best-defined syndromes within this group

Hypohidrotic (anhidrotic) ED (Christ-Siemens-Touraine syndrome) is the most common phenotype in this group and is usually inherited as an X-linked recessive trait; autosomal recessive and autosomal dominant forms have been reported but are rare. It is characterized by several defects (e.g. hypohidrosis, anomalous dentition, onychodysplasia, hypotrichosis). Typical facies are characterized by frontal bossing; sunken cheeks; saddle nose; thick, everted lips; wrinkled, hyperpigmented skin around the eyes; and large low-set ears. Because such characteristics are not obvious at birth, clinical clues for diagnosis in the neonatal period are extensive scaling of the skin and unexplained pyrexia. Dental manifestations include conical or pegged teeth, hypodontia or complete anodontia, and delayed eruption of permanent teeth. The prevalence of atopic eczema is high. Other common signs are short stature, eye abnormalities, decreased flow of tears and photophobia. Intelligence is normal. According to Bessermann-Nielsen, the salivary glands, including the intraoral accessory glands, are sometimes hypoplastic in this disease. This results in xerostomia, and the protuberant lips may be dry and cracked with pseudorhagades formation. As a related phenomenon, there may be hypoplasia of the nasal and pharyngeal mucous glands which leads to chronic rhinitis and/or pharyngitis, sometimes with associated dysphagia and hoarseness. Hidrotic ED (Clouston syndrome) is inherited in an autosomal dominant manner; the homozygous state may be lethal. Clinical features include nail dystrophy associated with hair defects and palmoplantar dyskeratosis. Nails are thickened

and discolored; persistent paronychial infections are frequent. Scalp hair is very sparse, fine, and brittle. Eyebrows are thinned or absent. Patients have normal facies, normal sweating and no specific dental defect is seen.

Case Report

A 28 year old young man with Ectodermal Dysplasia came to our private practice with chief complaint of not able to chew food with current prosthesis which was a removable denture for both maxillary and mandibular arches. He got this removable prosthesis made at the age of 23 years and on history taking told us that he wears them on; on and off basis as they were not satisfactory according to him. So he desired a fixed prosthesis.

CASE HISTORY

After the family and medical history were taken, a complete examination was performed, including clinical examination, panoramic radiography and Lateral Cephalogram. Patient had retained deciduous teeth A, E, J, K and T. But when he showed up at the clinic only teeth E and T were present.

Intraoral Findings

Congenitally absent maxillary and mandibular anteriors. Deciduous retained tooth E and T. Present permanent teeth 2,3, 14,15, 18,19, 30,31. Maxillary alveolar bone was very thin due to congenitally absent teeth. Intraoral examination revealed undeveloped maxilla with poorly expressed tubers, flat palatal vault, hypertrophic gingivobuccal plicas. Alveolar ridges were rather atrophic (knife - ridge) except in the areas where teeth were present. The color of alveolar mucosa and gingiva was normal. Severe hypodontia was present with missing most of the permanent teeth. Only two primary teeth existed in the jaws: E in the maxilla and T in the mandible. Salivary secretion was a little bit reduced, but he did not experience any problems during the mastication or wearing the dentures. Underdevelopment of alveolar ridges was also confirmed by orthopantomogram that revealed one deciduous teeth in the maxilla and one in mandible, as well as eight permanent teeth in the posterior regions of the maxilla and mandible in both the quadrants. The retained deciduous mandibular second molars are rectangular in shape with wide pulp chamber and the bifurcation seen only few millimeters above the apex of the root which gives the appearance of a taurodont. The radiograph also reveals congenital absence of permanent maxillary central incisors, lateral incisors, canines, all premolars, third molars and mandibular anteriors and third molars. A diagnosis of partial anodontia was made from clinical and radiographic features. Complete rehabilitation of dentition was planned with fixed partial denture and the patient was informed about the treatment. The treatment objective is to provide an ordered pattern of occlusal contact and articulation to optimize oral function, health of TMJ, occlusal stability, esthetics and comfort.

Extraoral Findings

A clinical extraoral examination revealed typical features of ectodermal dysplasia like square and bossing forehead, prominent supraorbital ridges, slightly pigmented and wrinkled eyelids, prominent and pointed ears. The nose was short and wide with anteverted nostrils and depressed nasal bridge, giving the middle face concave appearance. The patient had

large philtrum, protruding lips, small and pointed chin, deep mentolabial and nasolabial folds and lower facial height (vertical dimension) contributed to a senile (old - looking) facial expression. The hair was sparse, very thin, soft, blond and slow - growing (hypotrichosis). Eyebrows and eyelashes appeared one year before. The skin was dry (as result of hypohidrosis), thin with light coloring, rough and shiny, with several wounds caused by scratching. Finger and toe nails appeared short, thick, striated and slow - growing. Because of the hypohidrosis, boy had difficulties with body temperature regulation; boy cannot stand high air temperatures.

Treatment Plan

The patient desired a fixed prosthesis so the treatment plan was planned and followed accordingly. The treatment plan was divided into four phases.

Phase 1

Since the maxillary alveolar bone was not adequate to support and sustain implants, so bone grafting was planned to increase the availability of bone support for the success of implants. A bone graft from Iliac Crest of the patient (Figure 3,4,5,6) was harvested under General Anaesthesia and it was secured into the maxillary bone from cuspid area to the molar region in both right and left upper quadrants with the help of four 2mm screws and sutures were placed. An OPG was taken post operation for confirmation. (Figure 7)

Figure 1. Pre-operative Intraoral view of Maxillary Arch of the patient

Phase 2

Mandibular implants were placed after 2 months of phase 1. Five implants were placed at tooth positions (Everett *et al*., 1952; Kupietzky and Houpt, 1995; Paschos *et al*., 2002; Vieira *et al*., 2007 and Monreal *et al*., 1998), An OPG was taken post operation for confirmation of their desired placement.

Figure 2. Pre-operative Intraoral view of Mandibular Arch of the patient

Figure 3. Iliac Crest Bone is being harvested for the graft

Figure 4. Harvested Iliac Crest Bone Graft

Figure 5. Graft is divided in 2 pieces for implantation in Maxillary Arch

Phase 3 - Maxillary implants were placed after 6 months of phase 1 when Iliac Crest bone graft was taken up by the maxillary bone. Four implants were placed at tooth positions (Hickey and Vergo, 2001; Pipa Vallejo *et al*., 2008; Cronin *et al*., 1994 and Sweeney *et al*., 2005). An OPG was taken post operation for confirmation of their desired placement.
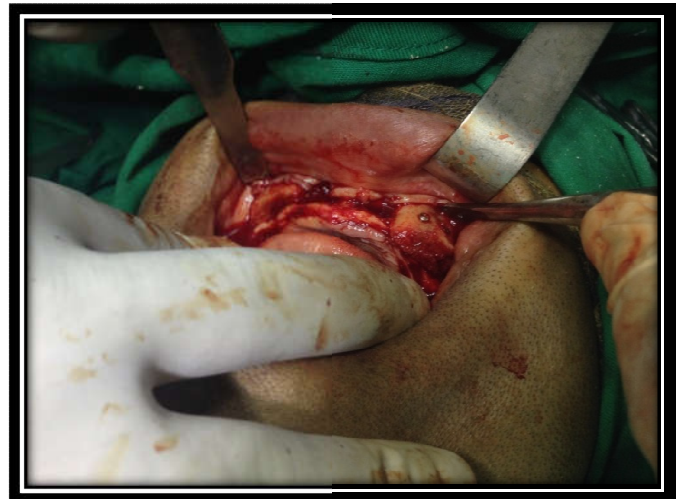

Figure 6. Iliac Crest Bone Graft is implanted with 2mm screws in the Maxillary Arch (Cuspid to Molar Area)

Figure 7. OPG showing graft placement in Maxilla with 2mm screws

Phase 4 - Final prosthesis was given to the patient. All mandibular permanent molars were Root Canal treated to restore functional occlusion. For mandibular arch PFM Crowns were given as fixed prosthesis. In maxillary arch Implant supported screw retained Hybrid Prosthesis was given. (Figure 8, 9, 10) Decisions regarding implant length and width were based on an examination of periapical and panoramic radiographs of the maxillary bone. Mounted diagnostic casts were used to fabricate a guide for implant placement by the surgeon.

Figure 8 and 9. Post-operative OPG showing Final Placement of Implants and Final Prosthesis

Figure 10. Post-operative Extraoral view of Final Prosthesis

The implant surgery was undertaken under local anesthesia and following the guidelines determined by the manufacturer. The surgical procedure started with an intraoral crestal incision followed by subperiosteal dissection of the mucoperiosteum; flattening of the alveolar crest was performed with a bur and under copious sterile saline irrigation. During bone healing, a temporary prosthesis was not delivered to the patient. At insertion, the implants were placed at a depth according to the guidelines given by the manufacturer. After 2 months from the iliac crest bone grafting 5 Mandibular Implants were placed into the (Everett *et al*., 1952; Kupietzky and Houpt, 1995; Paschos *et al*., 2002; Vieira *et al*., 2007; Monreal *et al*., 1998), regions. Two retained primary teeth were extracted. Then after 6 months of Iliac crest bone grafting into the maxillary arch, Four Implants were placed in Maxilla at (Hickey and Vergo, 2001; Pipa Vallejo *et al*., 2008; Cronin *et al*., 1994) region. Postoperative treatment consisted of the standard analgesics, chlorhexidin 0.2% mouthrinses, antibiotics, and nonsteroidal analgesics for 7 consecutive days. Sutures were removed 1 week after surgery. Standard oral hygiene instructions were given to the patient immediately after surgery. After all of the teeth preparations, the impression copings were placed. Definitive impressions of the maxillary and mandibular teeth and abutments were made with a polyether impression material. The impression copings were fixed onto the abutment analog. Then, cement retained prostheses were completed on abutment level models from a base metal alloy and porcelain and cemented to the abutments in the lower arch. Implant supported screw retained Hybrid Prosthesis was given in the upper arch. Compared with the pre-treatment profile, the post-treatment facial photographs showed a marked improvement in the facial profile.

Follow-up and criteria for success

The patient was followed up 3, 6, and 12 months postoperatively and then annually with visual and radiographic examinations. For the first year after treatment, the patient was followed for routine hygiene and assessment of long-term outcome. The patient acknowledged having improved function and esthetics, and he was pleased with the results. Criteria for success included effective placement and primary stability of the planned implant, stability of the implant (lack of mobility) and absence of pain or any subjective sensation at each visit, lack of peri-implant infection with suppuration, and lack of continuous radiolucency around the implant. Routine radiographs consisted of pano-ramic radiographs taken preoperatively, after placement of implants, at the time of prosthetic loading, and annually thereafter until the end of follow-up.

DISCUSSION

Differential Diagnoses

- Alopecia Areata
- Aplasia Cutis Congenita
- Focal Dermal Hypoplasia Syndrome
- Incontinentia Pigmenti
- Naegeli-Franceschetti-Jadassohn Syndrome
- Pachyonychia Congenita

Investigation

In general, laboratory studies are not useful in the diagnosis or management of the ectodermal dysplasias. Patients with

ectodermal dysplasia associated with immunodeficiency may have hypogammaglobulinemia with impaired lymphocyte proliferation and cell-mediated immunity. An appropriate evaluation, including determination of quantitative immunoglobulin levels and T-cell subset populations, should be performed. Perform orthopantography at an early age if hypodontia or dental abnormalities are present. X-ray films of hands, feet, or both may demonstrate specific skeletal deformities. Sweat pore counts, pilocarpine iontophoresis, and skin biopsy may document hypohidrosis and a reduction in the number of eccrine glands. It is now very well documented that dental findings in ectodermal dysplasia may range from hypodontia to anodontia of the primary or permanent teeth. Comprehensive rehabilitation of patient with ectodermal dysplasia is required to improve both the vertical and sagittal craniofacial relationship in order to provide improved esthetics, speech, and masticatory efficiency. Removable prostheses are the most common treatment method. Although dental literature describes many conventional prosthetic approaches to the clinical management of these patients, osseointegration is also documented as a safe and predictable method for replacement of missing teeth. Dental implants are considered to be a treatment option especially in combination with implant supported dentures for adolescents over 12 years of age are recommended as a treatment choice in literature. In situations where implant therapy is indicated, the main problem is insufficient bone; if bone atrophy progresses in these already alveolar- deficient patients, implant placement may not be possible without bone grafting. The use of osseointegrated implants to aid in restoring missing teeth has become the treatment of first choice, especially for adult patients, in consideration of optimal esthetic characteristics and long-lasting stability. Moreover, an orthodontic appliance or removable denture represents a second-option treatment that should be considered for patients with mixed teeth or limited agenesia. In growing patients with oligodontia, definitive therapies, such as surgical positioning of implants, could determine a risk of developmental deficiencies of local bone ridge. Thus, previous studies suggested postponing surgical positioning of implants to the end of dental growth. In the present case report, the patient was 27 years old and the growth process was completed.

Dental implants can be used to support, retain and stabilize the prosthesis. Success of the implant therapy depends on the age of the patients. Ectodermal dysplasia doesn't have negative effect on survival rate of implants when inserted on adult patients. Guckes *et al*. Presented 91% survival rate of endosseous dental implants that were placed in anterior mandible and 76% of those placed in anterior maxilla in a group of ED patients aged 8-68. Same authors described successful placement of implants in 3 year - old patient with ED but only in mandible. As the lateral growth of the anterior mandible is usually completed by 3 years of age, implant placement can be considered if anodontia exists in this area. Implants' relative position within the anterior mandible remains unchanged as growth takes place in the rami and condyles. In any case, it is not recommended to place implants in growing children as a routine practice, especially in maxilla. According to the guidelines of National Foundation for ED, implants are recommended for the anterior portion of mandible in children older than preschool age (7 years or older). Kramer *et al*. Successfully inserted and functionally loaded implants into the anterior mandible in 8 year - old children with severe hypodontia. According to Bector *et al*., dental implants

inserted at 8 years of age can be successfully integrated following maxillary and mandibular growth displacement; minor impaction of the maxillary implants and change in mandibular implants' inclination were detected 12 years after insertion.

Conventional prosthodontic rehabilitation is well suited in patients in whom the condition is not too severe and in places where advanced oral and maxillofacial deformity surgery is not available. However, no more implants than necessary should be inserted, or it could be difficult to achieve a satisfactory esthetic result with the prosthetic. Therefore, in the present clinical report, the definitive therapeutic approach was carried out using endosseous implants and a conventional, cement-retained, fixed prosthesis to allow normal masticatory and phonetic function. The patient's esthetics, facial profile, oral function, speech, and psychological status were significantly improved after the treatment. This was a result of coordinated efforts between the surgeon, prosthodontist, and laboratory technician. With an interdisciplinary approach and communication between the specialists during and after treatment, fewer compromises occur and more ideal restorations can be developed in the patients with oligodontia. Cronin *et al*. have concluded that implants placed after age 15 years for girls and 18 years for boys, provided the most predictable prognosis. Yet, Sweeney *et al*. found 20% failure in implants placed in anterior maxilla and only 7.8% failure in anterior mandible. There are different consequences of oral implants, if placed before dental and skeletal maturation: multidimensional restrictions of craniofacial skeletal growth, unpredictable implant dislocations as they do not participate in the maxillary growth process of drift and displacement, implant exposure because of bone resorption, limitation of maxillary growth if implants are connected by a rigid prosthesis that crosses the midline, infraocclusal position of inserted implants compared with adjacent natural dentition, as well as trauma to adjacent teeth buds leading to abnormalities in teeth development.

Conclusion

This case presentation showed how important it is to treat patients with developmental anomalies not only from the functional point of view but also with regard to the patient's appearance and the effect of the treatment on his or her psychology. This case highlights the positive effect of oral rehabilitation on the physical, emotional and social life of the patients with ED. Considering an age when patients should be dentally treated, Maxillo-facial surgery, implant placement and fixed prosthesis with rigid connector crossing the mid line, as a more comfortable and aesthetic solution for patients with ectodermal dysplasia,

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