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**Oral rehabilitation with implant-supported overdenture in a child with hypohidrotic ectodermal dysplasia**

Montanari M[1], Battelli F[1], Callea M[2♦], Corinaldesi G[3], Sapigni L[3], Marchetti C[3], Tadini G[4,5], Mancini EG[6], Grecchi F[6], Clarich G[2], Salinas CF[7], Fedele G[8], Piana G[1]

[1]*Unit of Odontostomatological Sciences - Department of Biomedical and NeuroMotor Sciences, University of Bologna, Bologna, Italy;*
[2] *Institute for Maternal and Child Health - IRCCS "BurloGarofolo"- Trieste, Italy,*
[3] *Unit of Oral Surgery, University of Bologna, Bologna, Italy*
[4,5] *Fondazione IRCCS Cà Granda – Ospedale Maggiore, Policlinico di Milano, University of Milan, Department of PediatricDermatology; Fondazione IRCCS Ca' Granda Ospedale Maggiore, Policlinico di Milano, University of Milan, Department of Pediatric Clinic 1*
[6]*U.O.C. Chirurgia Maxillo Facciale, IRCCS, Istituto Ortopedico Galeazzi, Milan, Italy,*
[7] *Medical University of South Carolina, College of Dental Medicine, Department of Pediatric Dentistry and Orthodontics,Division of Craniofacial Genetics, Charleston SC, USA,*
[8] *President ANDE ( Associazione Nazionale Displasia Ectodermica), Via Cascina Fidelina, Carugate, Milano, Italy,*

**Corresponding author:**
♦Dr. Michele Callea
Department of Pediatric Stomatoly,
Institute for Maternal and Child Health,
IRCCS BurloGarofolo, Trieste, Italy
+39 040 3785675
+39 333 2081878
mcallea@gmail.com

**Abstract**

Ectodermal Dysplasia syndromes (EDs) are a heterogeneous group of inherited diseases characterized by abnormal development of tissues of ectodermal origin. The most common form of EDs is X-linked Hypohidrotic Ectodermal Dysplasia (XL-HED) characterized by abnormalities of the skin, teeth, hair and sweat glands. The intraoral abnormalities include hypodontia, malformed teeth (conically shaped) and reduced alveolar ridge height. It causes severe impairment of chewing, swallowing, speech, esthetics and affects social relation. Early dental treatment at 2-3 years is essential to improve oral function and reduce the social impairment. This may include resin bonded restorations to conventional prosthetic treatment. In some cases suffering from severe hypodontia, however, conventional prostheses are inadequate due to lack of retention and instability. The replacement of teeth by implants is usually restricted to patients with completed craniofacial growth; however implants can be used as abutments for overdentures. The present study reports a 9-year follow up case in a child affected with XL-HED accompanied by anodontia. At 2 yrs of age, conventional upper and lower removable prostheses were fabricated. Subsequently, at age 11 years and 11 months; the patient was treated with a lower implant supported overdenture placed on two tapered implants (3.8 x 10 mm) in the anterior mandible. CBCT (Cone Beam Computer Tomography) of the mandible was done and dicom data used to obtain a rapid stereolithographic model.

## Introduction

Ectodermal Dysplasias are a heterogeneous group of inherited disorders characterized by dysplasia of tissues of ectodermal origin (hair, nails, teeth, skins and glands).[1] Clinically, it may be divided into two broad categories: the X-linked hypohidrotic form and the hidrotic form. Hypohidrotic Ectodermal Dysplasia (HED) is characterized by the triad oligo/anodontia, hypotricosis, hypo/anhydrosis (Christ-Siemens-Tourane syndrome). The incidence of HED is about 1/100,000. Mutation in the ectodysplasin-A (EDA) and ectodysplasin-A receptor (EDAR) genes are responsible for X-linked and autosomal HED.[2] More rare occurs a mutation in EDARADD, and recently WNT10A has been reported to be causative of HED.[3] The clinical features include sparse, fine hair, missing or conical teeth, decreased sweat and mucous glands, hypoplastic skin, and heat intolerance with exercise or increased ambient temperature.[4] Complete or partial anodontia and malformation of teeth are the most frequent dental findings. Incisors and canines are often conical in shape, while primarily second molars, if present, are mostly affected by taurodontism.[5] The diagnosis of HED in the neonatal and early infancy period may be difficult since sparse hair and absent teeth are a normal finding at this age.[6] During childhood the diagnosis is more easily made on the basis of history and clinical examination. Dental abnormalities are the most common complaint. Treatment is supportive and includes protection from heat exposure, skin, hair ear, nose and nail care, genetic counseling for family planning and early oral prosthetic rehabilitation. A dental multidisciplinary team that includes a pediatric dentist, an orthodontist, a prosthodontist and an oral and maxillofacial surgeon is necessary for a successful outcome. Prosthetic rehabilitation has been recommended as an essential part in HED management due to functional, esthetic, and psychological indications.[7] Conventional prosthodontic rehabilitation in young patient is challenging because of the anatomical abnormalities of existing teeth and alveolar ridges.[8] The

conically shaped teeth and “knife-edge” alveolar ridges result in poor retention and instability of dentures. Moreover, dentures must permit a correct pattern of growth in addition to jaw expansion.[9]

## CASE REPORT

A 2-year-old patient affected with Ectodermal Dysplasia and anodontia was rehabilitated with removable upper and lower prostheses. The prosthetic rehabilitation was provided to allow a correct masticatory function and normal physiological development. A monthly follow up of the patient was performed and, with time, conventional prostheses showed reduced retention especially in the mandibular jaw; therefore a different prosthetic treatment approach was necessary. At the age of 11 years and 11 months, the fabrication of an upper conventional and a lower implant-supported overdenture was indicated. The implants were two endosseus implants (position #33 and #43) in the anterior aspect of the mandibular jaw. The pre-prosthetic diagnostic steps included obtaining an Orthopantomogram (OPT) and CBCT (MyRay®, Cefla, Italy) 3D-images of the patient. Row DICOM data were elaborated using a 3D imaging software (OsiriX®, Pixmeo, Switzerland). The radiographic images showed a remarkable multi-dimensional atrophy of the mandibular alveolar process (Fig. 1), therefore two tapered implants measuring (3.8 x 10 mm) was the option of choice. A Virtual model of mandibular bone was used to obtain a resin model of the mandibular jaw of the patient (Fig. 2). The surgical procedure of implant placement was simulated in the resin model of the mandible and a surgical template was fabricated to guide implant placement on the anterior aspect of the mandible. The insertion of two tapered screw implants under local anesthesia with a novel biomimetic calcium-phosphate enriched titanium treatment (Anodic Spark Deposition, BioSpark) was possible and resulted in safe primary stabil-

ity (Fig. 3). A Cephalometric radiograph was taken after implant placement to evaluate correct implant positioning. After a submerged healing period of two months, the implants were exposed and two ball-attachments (Rhein 83, Bologna, Italy) were connected to the implants in order to increase lower prosthesis retention. In order to fabricate custom impression trays, initial maxillary and mandibular impressions were obtained using stock trays with an irreversible hydrocolloid material. Final impressions ware made with light-body polysulfide rubber base impression material. On the final casts, a base of auto polymerizing resin was constructed and a wax rim was added to the base. Preliminary occlusal relations were recorded and the patient's vertical dimension of occlusion was established by assessing phonetic and esthetic criteria. The mandibular castwas mounted on the articulator. Acrylic resin teeth specific for children dentures were selected and mounted. Denture try-in was performed and, after adjustments, were inserted on ball-attachment. The patient was monitored clinically every month for the following three years.

**DISCUSSION**

Early oral rehabilitation improves oral function, phonetics and esthetics, reducing social impairment. Mandibular growth in a sagittal and transverse direction showed no adverse effects on implant position. The fixtures advanced with the mandible, maintaining their original relationship with the bone. After three years of follow-up, the mandibular implant-supported overdenture was well accepted from the patient who reported excellent masticatory and esthetic improvements (Tab. I).

Implants can be successfully placed, restored and loaded in growing patients with Ectodermal Dysplasia. Several Authors in the literature reported good results with implant-supported overdentures in patients with Ectodermal Dysplasia.[10] Others reported a great number of implant failure in these patients[11] that can be due to the rigid connection of implants and the large diameter

of implants compared with the width of bone crest. The majority of authors placed implants after 13 yrs to avoid displacement of implants or exposition of implants because of craniofacial growth.[12] On the other hand, Gukes et al placed implants in 3-year-old patient.[13]

In the present study case report implants were placed when he was 7 years old because the most important center of growth had already performed its function and after this age the growth occurred where the prosthesis could not interfere.[14, 15] The prosthesis was connected with implants using two ball-attachments in order to avoid a rigid connection to allow mandibular growth and to reduce interference with the patient's growth. Factors such as good stability and retention of the implants-supported overdenture, reduction of micro-movement typical of conventional prostheses, excellent esthetics and substantial masticatory improvement maintained the patient's acceptance of the prosthesis.[16]

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**Figure legends**

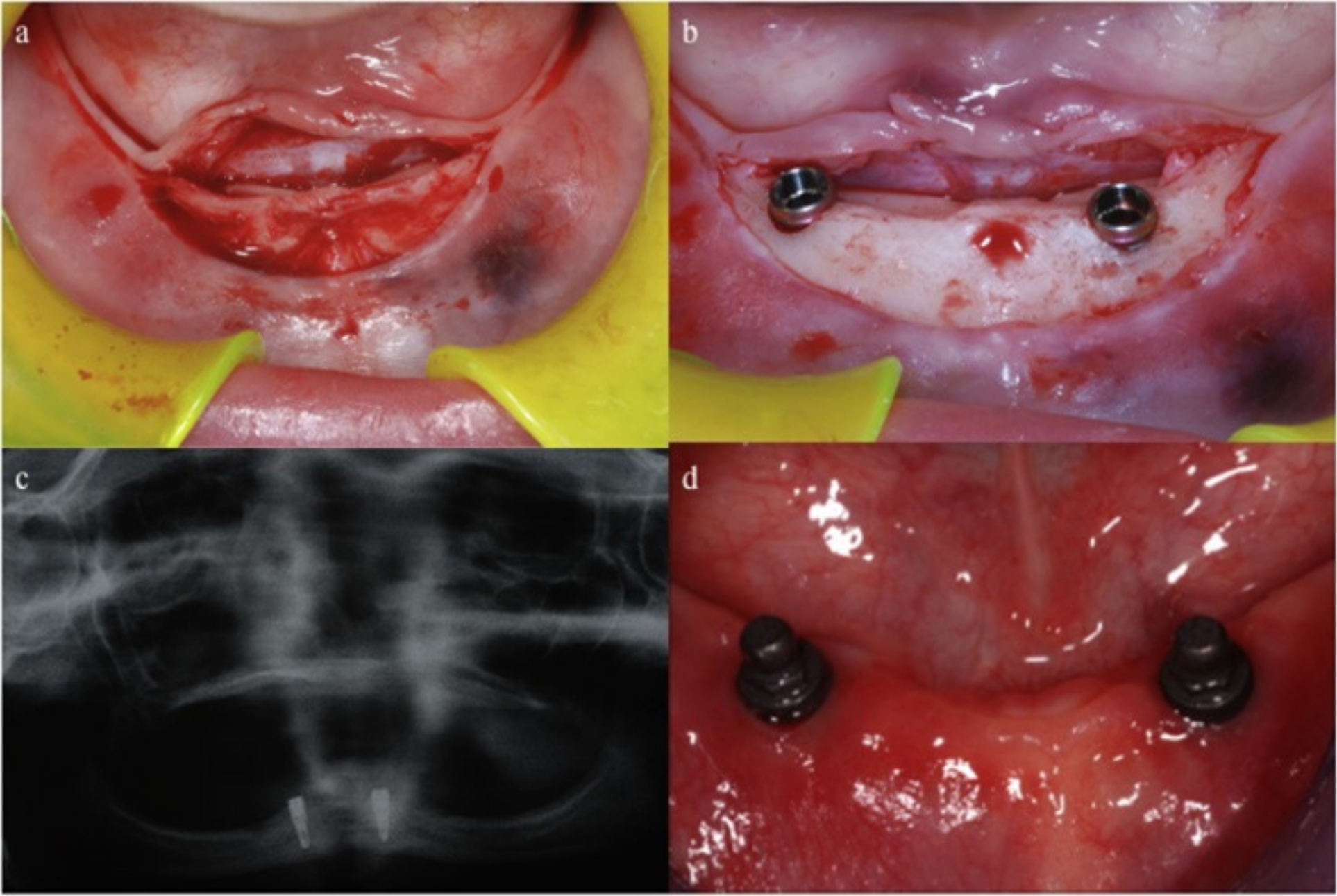
**Fig. 1:** Radiographic images of the mandibular alveolar process

**Fig. 2:** Virtual model of mandibular bone used to obtain a resin model of the mandibular jaw of the patient

**Fig. 3:** Patient after the treatment

**Table I**: Evaluation of prosthesis acceptance, masticatory improvement, esthetic improvement and phonetic improvement. + fairly good, ++ good, +++ very good

| Patient | Number of mandibular teeth | Prosthetic acceptance | Masticatory improvement | Esthetic improvement | Phonetic improvement |
|---|---|---|---|---|---|
| K.I. | 0 | +++ | +++ | +++ | +++ |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