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

M Montanari¹, F Battelli¹, M Callea²*, G Corinaldesi³, L Sapigni³, C Marchetti³, G Tadini⁴,⁵, EG Mancini⁶, F Grecchi⁶, G Clarich², CF Salinas⁷, G Fedele⁸, G Piana¹

Abstract
Introduction
Ectodermal dysplasia syndromes are a heterogeneous group of inherited diseases characterised by abnormal development of tissues of ectodermal origin. The most common form of ectodermal dysplasia syndromes is X-linked hypohidrotic ectodermal dysplasia characterised 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, aesthetics 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. This study reports a 9-year follow-up case of a child affected with X-linked hypohidrotic ectodermal dysplasia accompanied by anodontia.

Case report
At the age of 2 years, conventional upper and lower removable prostheses were fabricated. Subsequently, at the age of 11 years and 11 months; the patient was treated with a lower implant supported overdenture placed on two tapered implants (3.8 × 10 mm) in the anterior mandible. Cone beam computer tomography of the mandible was done and Dicom data used to obtain a rapid stereolithographic model.

Conclusion
Implants can be successfully placed, restored and loaded in growing patients with ectodermal dysplasia.

Introduction
Ectodermal dysplasias are a heterogeneous group of inherited disorders characterised 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 characterised by the triad oligo/anodontia, hypotrichosis, 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 is responsible for X-linked and autosomal HED². Even rarer is a mutation in EDARADD, and recently WNT10A has been reported to be causative of HED³. 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 temperature⁴. 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 taurodontism⁵. 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 age⁶. 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 paediatric 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, aesthetic and psychological indications⁷. Conventional prosthetic rehabilitation in young patients is challenging because of the anatomical abnormalities of existing teeth.
and alveolar ridges. 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. This study reports a case of oral rehabilitation with implant-supported overdenture in a child with HED.

**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 denture was indicated. The implants were two endosseus implants (position #33 and #43) in the anterior aspect of the mandibular jaw. The preprosthetic diagnostic steps included obtaining an orthopantomogram and cone beam computer tomography (MyRay®, Cefla, Italy) 3D-images of the patient. Raw 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 (Figure 1), therefore, two tapered implants measuring (3.8 × 10 mm) was the option of choice. A virtual model of mandibular bone used to obtain a resin model of the mandibular jaw of the patient. Implants under local anaesthesia with a novel biomimetic calcium-phosphate enriched titanium treatment (Anodic Spark Deposition, BioSpark) was possible and resulted in safe primary stability (Figure 3). A cephalometric radiograph was taken after implant placement to evaluate correct implant positioning. After a submerged healing period of 2 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 mandibular and mandibular impressions were obtained using stock trays with an irreversible hydrocolloid material. Final impressions were made with light-body polysulfide rubber base impression material. On the final casts, a base of auto polymerising 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 aesthetic criteria. The mandibular cast was mounted on the articulator. Acrylic resin teeth specific for children dentures were selected and mounted. Denture try-in was performed and, after adjustments, was inserted on ball-attachment. The patient was monitored clinically every month for the following 3 years.

**Discussion**

Early oral rehabilitation improves oral function, phonetics and aesthetics, 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 3 years of follow-up, the mandibular implant-supported overdenture was well accepted from the patient who reported excellent masticatory and aesthetic improvements (Table 1).

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. Others reported a great number of implant failure in these patients 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 years to avoid displacement of implants or exposition of implants because of craniofacial growth. On the other hand, Gukes et al. placed implants in a 3-year-old patient.

In this case report, implants were placed when he was 7 years old because the most important centre of growth had already performed its function and after this age the growth occurred where the prosthesis could not interfere. 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 aesthetics and substantial masticatory improvement maintained the patient’s acceptance of the prosthesis.

### Conclusion

Implants can be successfully placed, restored and loaded in growing patients with ectodermal dysplasia. The replacement of teeth by implants is usually restricted to patients with completed craniofacial growth; however, implants can be used as abutments for overdentures. This study re-ports a 9-year follow-up case of a child affected with X-linked hypohidrotic ectodermal dysplasia-sia accompanied by anodontia where a complete oral rehabilitation with an outstanding outcome has been carried out.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

### References


### Table 1 Evaluation of prosthesis acceptance, masticatory improvement, aesthetic improvement and phonetic improvement

<table>
<thead>
<tr>
<th>Patient</th>
<th>Number of mandibular teeth</th>
<th>Prosthetic acceptance</th>
<th>Masticatory improvement</th>
<th>Aesthetic improvement</th>
<th>Phonetic improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>K.I.</td>
<td>0</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
</tr>
</tbody>
</table>

+, fairly good; ++, good; +++ very good.