Overdenture prosthesis for oral rehabilitation of hypohidrotic ectodermal dysplasia: A case report

Esteban D. Bonilla, DDS*/Luis Guerra, DDS**/Oscar Luna, DDS***

Abstract

The dentition of a patient with ectodermal dysplasia was restored with a modified hollowed maxillary overdenture opposing a conventional mandibular overdenture. Lingualized occlusion was used because it was the ideal occlusal scheme for this patient to achieve denture stability. The lingual cusps of the maxillary posterior teeth contacted the fossae of the mandibular teeth to create freedom of movement and to prevent lateral interference. (Quintessence Int 1997;28:657-665.)

Clinical relevance

An overdenture with lingualized occlusion preserved the existing dentition and created freedom of movement for the patient with complete or partial absence of teeth due to hypohidrotic ectodermal dysplasia.

Introduction

Ectodermal dysplasia (ED) is a hereditary disorder associated with defective ectodermal structures and their accessory appendages. There are two forms of ED, the hidrotic and the hypohidrotic. The teeth and the hair are affected similarly in both forms, but the manifestations in nails and sweat glands and the hereditary patterns tend to differ (Table 1).

The most frequently reported ED syndrome is x-linked hypohidrotic ectodermal dysplasia (HED), which affects 1 to 7 individuals per 10,000; males are afflicted more frequently than females. The hidrotic form is largely confined to families of French or French-Canadian origin. The disorder probably appears during the first trimester of pregnancy. If it is severe, it appears before the sixth week of embryonic life, and, consequently, the dentition will be affected. After the eighth week, the other ectodermal structures may be affected.

The most remarkable characteristics of HED is hypohidrosis, because physical features are not as apparent in the first year of life. It may be diagnosed clinically before the second year of life only after repeated episodes of unexplained fever.

According to Perabo et al, ectodermal dysplasia may have been recorded as early as 1792 by Danz. In 1838, Wedderburn documented ectodermal dysplasia in a letter to Charles Darwin, describing a case of 10 Hindu male family members. The letter was quoted by Darwin in Variations in Plants and Animals Under Domestication. Thurnam, in 1848, reported two cases of the hypohidrotic form. He found that two cousins and their maternal grandmother had very light and very fine hair, anodontia, and an inability to sweat. Williams described one of his young female patients as having fine, scanty white hair, as well as hypodontia. In 1883, Guilford described a 48-year-old man, edentulous from birth, who had never perspired and had soft, scant, downy hair on his head. Hutchinson,
Table I  Differential diagnosis of hypohidrotic ectodermal dysplasia

|                      | Hypohidrotic (anhidrotic) ectodermal dysplasia | Hidrotic ectodermal dysplasia | Rothmund-Thomson syndrome | Chondrodysplasia 

<table>
<thead>
<tr>
<th>Generic transmission</th>
<th>X-linked recessive</th>
<th>Autosomal-dominant</th>
<th>Autosomal-recessive</th>
<th>Autosomal-recessive</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset of symptoms</td>
<td>First year</td>
<td>First year</td>
<td>1-5 years</td>
<td>1-2 years</td>
<td>First year</td>
</tr>
<tr>
<td>Skin</td>
<td>Dry, thin, soft, smooth</td>
<td>Dry, rough, with crusty texture. Hyperpigmentation in some areas</td>
<td>Atrophy, brownish pigmentation. Telangiectasias beginning during 3 to 6 months</td>
<td>Occasional hypotrophy, eczema, petechiae</td>
<td>Dermatoglyphic changes</td>
</tr>
<tr>
<td>Palms and soles</td>
<td>Small hyperkeratosis</td>
<td>Hyperkeratosis</td>
<td>Hyperkeratosis</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Sebaceous glands</td>
<td>Markedly decreased</td>
<td>Slightly decreased</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Throat glands</td>
<td>Severe, decreased, or absent</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Mammary glands</td>
<td>Aplastic/hypoplastic</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Hair</td>
<td>Fine, scanty and light-colored. Lanugo hair</td>
<td>Dry, fine; usually hypotrichosis</td>
<td>Hypotrichosis of scalp and body. Occasional alopecia</td>
<td>Thin, brittle, sparse, and hypochronic</td>
<td>Sparse scalp hair</td>
</tr>
<tr>
<td>Nails</td>
<td>Normal or spoonshaped</td>
<td>Short, thick; elevated tip</td>
<td>Frequently dystrophic (rough, ridge, heaped up)</td>
<td>Dysplastic (brittle, furrowed, and underdeveloped)</td>
<td>Absent to hypoplastic fifth fingernail and toenail. Other nails occasionally hypoplastic or absent</td>
</tr>
</tbody>
</table>

Table 1 continues on next page.
<table>
<thead>
<tr>
<th>Hypohidrotic (anhidrotic) ectodermal dysplasia</th>
<th>Hidrotic ectodermal dysplasia</th>
<th>Rothmund-Thomson syndrome</th>
<th>Chondro-ectodermal dysplasia</th>
<th>Coffin-Siris syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eyelashes/ eyebrows</td>
<td>Missing</td>
<td>Absent or scanty</td>
<td>Usually fall out during the first year of life and remain sparse or absent thereafter</td>
<td>Absent or scanty</td>
</tr>
<tr>
<td>Lacrimal gland function</td>
<td>Normal or reduced</td>
<td>Normal or reduced</td>
<td>None</td>
<td>Normal</td>
</tr>
<tr>
<td>Psychomotor</td>
<td>Occasional mental deficiency</td>
<td>Occasional mental deficiency</td>
<td>None</td>
<td>Occasional mild mental retardation</td>
</tr>
<tr>
<td>Systemic abnormalities</td>
<td>Distinct intolerance to heat</td>
<td>None</td>
<td>None</td>
<td>Congenital heart disease</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>None</td>
<td>Occasional short stature</td>
<td>Short stature. Occasional small hands and feet.</td>
<td>Short limbs. Dwarfism. Bilateral polydactyly (affects hands and feet)</td>
</tr>
<tr>
<td>Skeletal</td>
<td>None</td>
<td>Occasional small stature</td>
<td>Occasional hypogonadism</td>
<td>Hypoplastic genitalia</td>
</tr>
<tr>
<td>Endocrine</td>
<td>Usually none</td>
<td>Occasional pituitary and suprarenal deficiency</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>Allergic disorders</td>
<td>Asthma and eczema6</td>
<td>Occasional asthma</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>Prognosis for life</td>
<td>Above average</td>
<td>Average</td>
<td>Above average</td>
<td>Death before 55 years of age</td>
</tr>
</tbody>
</table>

In 1886, described similar hereditary ectodermal anomalies. The term congenital ectodermal defect was not used until a case was reported by Christ in 1913.\(^\text{28}\)

Weech,\(^\text{29}\) in 1929, introduced the term hereditary ectodermal dysplasia and suggested the term anhidrotic for those with the inability to perspire. In the same year, Clouston\(^\text{30}\) analyzed 119 cases of the hidrotic form in a group of people of French-Canadian origin for six generations, and in 1939 he coined the term hypohidrotic ectodermal dysplasia.\(^\text{31}\)

Felsher,\(^\text{32}\) in 1944, changed the adjective anhidrotic to hypohidrotic because the author argued that persons with the hypohidrotic form are not truly devoid of all sweat glands. Since then, cases of ectodermal dysplasia of both types have been reported in the dental literature over a period of many years.\(^\text{17-19,23,33-39}\)

Only a few illustrated clinical presentations have described the management of the patient with ectodermal dysplasia.\(^\text{40-43}\)

Patients afflicted with a hereditary deformity such as ectodermal dysplasia suffer from poor psychological and physiologic development as a result of unacceptable esthetics and abnormal function of orofacial structures. Dentists have a responsibility to rehabilitate these patients to improve appearance, mastication, and speech. Dental treatment depends on the severity of the disorder; therefore, treatment varies according to the age, growth, and development of the stomatognathic system of the patient.

Several prosthodontic treatments, such as complete dentures, removable partial dentures, overdentures, and implants, are available to rehabilitate patients with HED. These treatment approaches can be used individually or in combination to provide an optimal
The literature shows that endosteal implants have been used widely in rehabilitating patients with HED. A consensus conference on implantology suggested that implants not be placed until maximum maxillary growth has occurred, which was reported as being up to 15 years of age. This recommendation is well supported by a preliminary study with a 90% dental implant success rate in HED patients to 69 years old.

The purpose of this article is to illustrate the oral rehabilitation, with an overdenture prosthesis, of a patient with hypohidrotic ectodermal dysplasia.

Ectodermal dysplasia is characterized mainly by (1) a severe reduction in the numbers of eccrine sweat glands (hypohidrotic); (2) scanty, fine, light hair on the scalp and eyebrows (hypotrichosis); (3) complete or partial absence of teeth (anodontia or oligodontia); and (4) abnormal nails (onychodysplasia). Other associated signs are a pronounced supraorbital ridge and frontal bossing, a depressed nasal ridge, and proterant everted lips due to a decreased facial vertical dimension. However, cephalometric analysis has shown normal jaw and facial development.

Case report

The patient, a 5-year 1-month-old Latin American boy, was referred to the Dental Clinic for examination, evaluation, and treatment of his disorder. The child exhibited the classic characteristics of ectodermal dysplasia: hypodontia, hypohidrosis, hypotrichosis, prominent forehead, saddle nose, and everted lips (Fig 1).

His parents' main concern was their son's lack of teeth and inability to eat properly, although he was under the regular care of a pediatrician. His disorder had been diagnosed in November 1985. He had also had asthma since birth, but there was no other history of abnormal skeletal development, cardiovascular disease, cerebral, mental, or neuromuscular disorders, allergies to any medication, or bleeding disorders. All his vital signs and blood laboratory tests were normal. He was the sixth affected male in four generations.

An oral examination revealed a dry and sticky oral mucosa with localized mild gingivitis. Six maxillary teeth, four peg-shaped primary anterior teeth and two deformed small primary molars were present, and there was only one mandibular primary canine-like tooth. He exhibited aplasia of alveolar bone in the edentulous area (Figs 2a to 2c). A complete radiographic examination was performed. Cephalometric analysis revealed a skeletal Class I profile (Fig 3).

In November 1989, when the child was aged 5 years 4 months, a panograph revealed three unerupted mandibular teeth. One of them was surgically exposed while the patient was under intravenous sedation. The maxillary arch had six erupted primary teeth and six unerupted teeth (Fig 4). Orthodontic consultation was required. A tooth positioner was made to upright both mandibular canines. Then a fixed appliance was used to complete the uprighting of the canines.

A conventional overdenture was the treatment of choice for this patient, because the objectives were to preserve the remaining dentition, to restore function and esthetics, and to allow certain modifications to be made to meet the needs of the developing stomatognathic system (Figs 5a and 5b).

One year 10 months after fabrication of the first set of complete overdentures, the patient started to feel some discomfort as a result of changes in the dento-osseous structures of the maxillary and mandibular arches. Later, the maxillary overdenture did not fit because of the eruption of the maxillary permanent first molar on both sides. Therefore the authors decided to make a new modified complete overdenture set with a different occlusal scheme to achieve proper balanced occlusion.

Fabrication of the modified overdenture

Accurate irreversible hydrocolloid impressions were taken with modified stock metal trays. Custom acrylic resin trays were fabricated for final impressions. The maxillary and mandibular trays were border molded.
Fig 2a  Maxillary occlusal view.

Fig 2b  Mandibular occlusal view.

Fig 2c  Buccal views of the right and left sides.

Fig 3  Cephalometric radiograph.

Fig 4  Panograph.
with green and washed with red stick compound (Kerr/Sybron). Final impressions were taken with regular and light-body polysulfide impression material (Perm-elastic, Kerr/Sybron). Maxillary and mandibular occlusion rims were fabricated on the casts obtained from the final impressions.

A facebow transfer was recorded. An occlusal relation record was taken with zinc oxide-eugenol (Superbite, Bosworth), and the casts were mounted on a Hanau articulator. The incisal pin was raised 4 mm to restore the lost vertical dimension of occlusion and to create space for denture teeth.

A trial denture base was made from autopolymerizing acrylic resin, and the selected denture teeth were adjusted in size and set in wax.

An occlusal window was created on the maxillary denture on both sides for the primary second molar and the permanent first molar (Figs 6a and 6b). In overdenture prostheses, an occlusal concept based on balanced occlusion is generally recommended, because it contributes to the stability of the dentures during eccentric movements and minimizes alveolar bone loss. However, for this patient, lingualized occlusion, in which the lingual cusps of the maxillary posterior teeth contact the fossae of the mandibular teeth, was selected to create freedom of movement (long centric) and to prevent lateral interference (Fig 7).

The dentures were processed in heat-polymerizing acrylic resin in the usual manner after a satisfactory try-in.

Both overdentures were placed in the patient's mouth, and adjustments were made. Pressure indicator paste (Mizzy) was used to prevent injury to the oral mucosa.
Fig 6a Modified hollowed maxillary overdenture.

Fig 6b Modified hollowed maxillary overdenture in place, showing the primary second molars and permanent first molars.

Fig 7 Facial view of the overdenture with the teeth in centric occlusion using a lingualized occlusal scheme.

Fig 8a View of the right and left sides showing the permanent first molars 2 years later and a modified overdenture waxup maintaining the lingualized occlusion.

Fig 8b Frontal and profile views of the patient after oral rehabilitation.
The dentures were remounted in the articulator with a new occlusion registration to check and to readjust the centric occlusion and the excursive movements. The maxillary and mandibular overdentures were delivered. Recall appointments were scheduled for 24 hours, 72 hours, 2 weeks, 4 weeks, every 3 months for the first year, and every 4 months for the second year. Written oral hygiene instructions were given and explained to the patient’s parents.

At the end of the 2-year recall, a new set of dentures, with one occlusal window on the maxillary arch for the permanent first molars, was fabricated because of changes in oral hard and soft tissue structures (Figs 8a and 8b).

Summary
A patient with hypohidrotic ectodermal dysplasia was treated with a modified hollowed maxillary overdenture opposing a conventional mandibular overdenture. Lingualized occlusion was the ideal occlusal scheme for this patient to achieve denture stability on function, penetration of the food bolus, centralization of vertical forces on the mandibular teeth, and esthetics. The lingualized concept utilized natural anatomic molars that protruded through the hollowed maxillary denture. The maxillary lingual cusps contacted the mandibular denture teeth in centric occlusion.

Acknowledgement
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References
24. Thurnham J. Two cases in which the skin, hair and teeth were very imperfectly developed. Proc Roy Med Chir Soc 1848;31:71-82.
The University of Manitoba invites applications and nominations for the position of Dean of the Faculty of Dentistry. The appointment is expected to commence on July 1, 1998, and will be for an initial term of five years. The Faculty, which includes the School of Dental Hygiene, is located at the University of Manitoba downtown campus adjacent to the Health Sciences Centre complex. The Faculty offers programs leading to the degrees of Doctor of Dental Medicine, Bachelor of Science in Dentistry, Master of Science and Doctor of Philosophy, and diplomas in Dental Hygiene, Oral and Maxillofacial Surgery and Periodontics.

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