Five-Year Follow-Up Treatment of an Ectodermal Dysplasia Patient with Maxillary Anterior Composites and Mandibular Denture: A Clinical Report

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Abstract

This clinical report describes the 5-year follow-up treatment of an 11-year-old boy with ectodermal dysplasia. The dentition was initially restored with a combination of maxillary anterior-bonded composite restorations and a mandibular complete denture. This individualized approach allowed successful management of the patient.

Ectodermal dysplasia is a disorder affecting the ectodermal structures including hair, skin, nails, and teeth.[1] According to Pinheiro and Freire-Maia, the ectodermal dysplasias form a group of over 150 conditions, classified into 11 clinical subgroups, and include both pure dysplasia and syndromes; 101 (66%) of them have a genetic cause (41 autosomal dominant, 52 autosomal recessive, 8 X-linked), and the remaining 53 (34%) conditions have an unknown cause.[2]

The most common type of ectodermal dysplasia is hypohidrotic ectodermal dysplasia (HED).[3] Its frequency is between 1/10,000 and 1/100,000 births, and it has X-linked inheritance, therefore, affecting mostly males.[1]

Patients with HED manifest hypohidrosis, hypotrichosis, and hypodontia.[3] They can present with sparse hair and thin, dry skin and have an inability to sweat.[4]

One of the most common oral abnormalities is the absence of most deciduous and permanent teeth. The existing anterior teeth can have conical-shaped clinical crowns.[3] Due to the number of congenitally missing teeth, most patients present with reduced vertical dimension and esthetic problems.[5]

Eczematous skin lesions, decreased vertical dimension, and oligodontia give children suffering HED "an old man appearance," affecting their self image.[6] From around age 9, children affected with various disabilities become aware of their differences, often causing depression.[7]

Many prosthodontic treatments have been described in the literature to rehabilitate patients with HED, depending on the age and development of the stomatognathic system.[8] Complete dentures,[4] removable partial dentures (RPDs),[9] and overdentures[10] individually or in combination are alternatives for young children. These interim prostheses may ease the transition of a child into adolescence; then as young adults, endosteal implants[11-14] may be considered as more definitive restorations.

Children and adolescents with this disorder suffer social isolation and emotional problems. Many families turn to support organizations to face these challenges. Dentistry forms a part of the interdisciplinary team that treats these young people, providing successful clinical outcomes and proper emotional development.

The aim of this clinical report is to describe the dental irregularities and oral rehabilitation and long-term follow-up of a young patient with HED.

Figure 1 Frontal bossing, depressed nasal bridge, and periorbital pigmentation observed.

Clinical report

A 6-year-old Caucasian male was referred to the Advanced Prosthodontics Department at the University of Southern California in May 2001. The parents' chief complaint was "a disorder causing missing teeth." Their main concern was their son's esthetic appearance and difficulty chewing food.

General and dental features

The patient presented with fine, sparse blonde hair and scant eyebrows. His small face was characterized by frontal bossing and a depressed nasal bridge. His skin appeared thin and dry with periorbital increased pigmentation (Fig 1).

An intraoral exam revealed normal mucosa and salivary flow. The patient had a total of six caries-free primary teeth. The maxillary central incisors and canines had cone-shaped clinical crowns, and the two maxillary primary molars appeared malformed with reduced occlusal tables (Fig 2). The mandible was completely edentulous, which is a rare finding.[3] According to the Prosthodontic Diagnostic Index, both maxillary and mandibular arches can be diagnosed as Class IV, describing multiple missing teeth, deformed teeth, and altering vertical dimension with the denture. Vertical dimension was reduced due to a lack of occlusion and any vertical stop causing protuberant lips. It was noticed during the intraoral exam that the patient had a strong gag reflex. A panoramic radiograph revealed an unerupted permanent tooth in the anterior maxillary region of 8 and confirmed the absence of any primary or permanent teeth in the mandible (Fig 3).

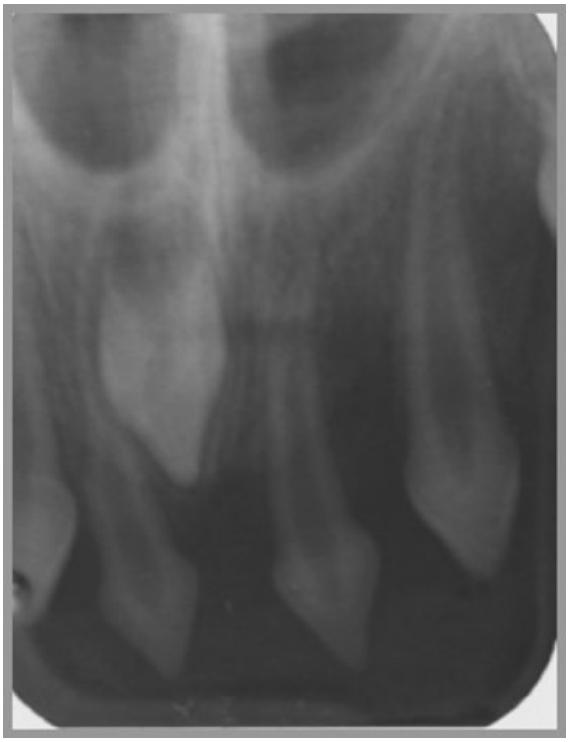

Figure 3 Preoperative intraoral panoramic radiograph.

The patient was very concerned about his appearance. He was very timid and rarely smiled. Restoring his missing teeth would promote self-confidence and psychosocial adaptation.

Dental treatment

Primary impressions were made with irreversible hydrocolloid (Indentic Alginate, Dux Dental, Oxnard, CA), and diagnostic casts were poured and then articulated to evaluate dentition,

Figure 2 Preoperative intraoral photograph.

Figure 4 Initial diagnostic wax-up.

periodontium, and occlusal vertical dimension (OVD). The objectives of the treatment plan were to maintain existing dentition, restore function, and improve esthetics; therefore, a maxillary overdenture and a mandibular complete denture were planned.

Custom maxillary and mandibular acrylic trays were fabricated from diagnostic casts. Both trays were border-molded with modeling impression compound (sds Kerr, Sybron Dental Specialties, Romulus, MI).

A final polysulfide rubber base impression (Heavy-bodied Permlastic, sds Kerr) was obtained with extreme difficulty due to the gag reflex of the young patient. Wax occlusion rims were then fabricated. OVD was duplicated with the old prostheses. A centric relation record was made with PVS (Blue-Bite HP, Henry Schein Inc, Meville, NY), and facebow transfer was recorded. Casts were articulated on a Hanau articulator (Hanau 96H2, Whip Mix Corporation, Louisville, KY), and primary denture teeth (Bambino A-59, Justi Products, Oxnard, CA) were set in wax.

At the wax try-in appointment the mandibular denture was tried intraorally. It was adjusted for proper esthetics and function; however, the patient had difficulty tolerating the maxillary record base, and it became obvious that a complete maxillary overdenture would not be accepted. It was then decided to modify the treatment plan and restore the maxilla with resin composite restorations instead of an overdenture. A space analysis was carried out, taking into consideration the existing distance between the peg-shaped teeth. The wax-up on the initial master cast allowed the determination of more naturally shaped teeth (Fig 4). Lateral incisor pontics were cantilevered off canines.

A silicon matrix was fabricated, permitting a three-dimensional replication intraorally of the restoration as it was created in the mouth. An intraoral chairside composite trial restoration was performed to choose shade prior to any modification of the existing teeth. No tooth reduction was required; however, the enamel surface of the maxillary incisors and canines were roughened with a diamond bur to increase surface roughness and surface area. Thirty-five percent phosphoric acid (Ultra-Etch, Ultradent Product Inc, South Jordan, UT) was placed on the enamel for 30 seconds, rinsed, and dried. Adhesive (optiBond Solo, sds Kerr) was subsequently applied following manufacturer's instructions. A polychromatic layering technique was used to resemble natural dentition.[15-17] A 3-layer concept comprising two sets of dentin and an enamel as described by Dietschi[18,19] was the technique used. The silicone matrix guided creation of the lingual and incisal contours formed using a translucent shade (shade B1) (Kerr XRV Herculite Prodigy, Orange, CA). Subsequently, the second layer consisted of a body shade resembling dentin (shade B1-B2) (Kerr XRV Herculite Prodigy), which was placed in lobes creating the three-dimensional structure of a newly erupted tooth.[18] Finally, a transparent composite (shade B1) was used over the dentin lobes and incisal edge replicating the translucency of enamel. At this appointment, a second mandibular wax try-in was accomplished. Occlusion was adjusted, and both parents and the patient approved the esthetics (Fig 5).

The mandibular denture was festooned and processed. At the insertion visit, the polishing of the maxillary teeth was achieved using a variety of finishing burs, sandpaper disks, rubber wheels, and polishing pastes. (Ultra Denture Kit, Brasseler USA, Savannah, GA) Pressure-indicating paste was used to relieve the pressure areas of the denture. The patient returned for a 24-hour postinsertion visit where minimal adjustments were made (Fig 6).

Subsequently, the patient was placed on a 6-month recall schedule where four relines using Viscogel (Densply Detrey GmbH, Konstanz, Germany) as functional impression material were performed during the next 3 years. In May 2006, the patient returned to the clinic with a broken mandibular denture. Periapical X-rays revealed the maxillary right primary central incisor restored with composite was being resorbed by the erupting permanent central incisor. The remaining composites, despite some discoloration, remained intact (Fig 7).

The permanent tooth was allowed to fully erupt, and 2 months later the existing composite restorations were completely removed. No decay was found under the composite restorations. New impressions were made, and casts were articulated to create the second wax-up. At this treatment time the esthetic challenges were greater, as the permanent maxillary right central incisor had erupted labially compared to the remaining primary incisors, creating a gingival marginal discrepancy. Orthodontic extrusion was proposed to address soft tissue esthetics; however, the parents declined any additional dental procedures, especially since the patient did not display the cervical areas of the teeth when smiling. The uneven clinical crown length was accepted, and the resin composite restorations were formed using another silicone matrix developed in the same way as the first. Darker composite shades (A1-A2) (Kerr XRV Herculite Prodigy) were used to characterize the gingival, middle, and incisal areas. Esthetic parameters related to the lower lip, including size, shape, and incisal lengths, were used as far as possible to obtain an acceptable result. A new mandibular denture was delivered. Both patient and parents were happy with the results (Figs 8–10). Oral hygiene instruction was discussed. The patient once again returned for a 24-hour check, where minimal adjustment was made to the denture. Subsequently, the patient was checked at 72 hours and 1 week. Thereafter, the patient was placed on a 6-month recall, which included a periodontal check-up and a mandibular denture reline.

Discussion

The typical treatment of a patient with HED is achieved in several phases, depending on the craniofacial development, thus requiring a comprehensive and multidisciplinary approach. The different treatment modalities are dictated by the manifestation of hypodontia and resulting malocclusions. Operative and prosthodontic restorations will satisfy the principal aims of dental treatment, which are enhancing esthetics and improving masticatory function.

Removable prosthodontics is the most frequent modality used for dental treatment of ectodermal dysplasia.[20] Complete dentures are an alternative for patients with complete anodontia. These prostheses may be carried out as early as preschool years where young children usually adapt to their use.[4,21] Overdentures can be used to restore ideal occlusion and usually allow the preservation of existing dentition;[8] however, for optimal results, patients must follow a rigorous oral-hygiene

Figure 5 Mandibular denture teeth set-up.

Figure 6 First postoperative photograph.

Figure 7 Erupting right maxillary permanent incisor.

Figure 8 Second postoperative intraoral photograph, after 5 years. Right maxillary central erupted labially compared to the remaining primary incisors creating a gingival marginal discrepancy.

Figure 9 Postoperative occlusal photograph.

Figure 10 Second postoperative photograph, after 5 years.

regime to avoid development of caries and periodontal problems.[22] RPDs represent another treatment approach. These prostheses can be fabricated according to the number of remaining teeth and are dependent on how malformed the existing teeth are. Primary and permanent artificial teeth consistent with the appearance of early mixed dentition stages may be used.[7]

Direct bonding in combination with removable appliances as described in this particular clinical report is also an advocated treatment modality.[23] To achieve successful results using resin composite restorations, occlusion must be carefully analyzed. Opposing natural teeth in an edge-to-edge position against full contoured composites may be contraindicated, as resulting forces may cause breakage of the bonded restorations.[7] In this patient, the mandibular denture occluding against the two remaining maxillary molars reduced occlusal forces on the anterior composites.[24] In addition, the excellent bond strength between enamel and composite and the patient's past history with composite restorations indicated a good long-term prognosis for this treatment. Another advantage of this treatment is that it is reversible, since no preparations were required to execute the resin composite restorations.

It is anticipated that as this patient continues to grow and mature, multiple prosthetic replacements and follow-up care will be required throughout his developing years until more definitive implant-assisted prostheses can be delivered. Ideally, implant placement should be considered when the patient stops growing;[9] however, the literature reports the use of endosseous implants in patients as young as 7 years of age in the anterior mandible, since the growth centers are located in the ramus area.[25-27] Some authors report placing implants in the anterior maxillary area before growth is completed. With this approach, orthognatic surgery and prosthesis remake must be anticipated to correct jaw size discrepancies.[27]

Conclusion

This clinical report demonstrates that direct anterior composite restorations in combination with complete dentures can be considered as an alternative when treating patients with HED. These cost-effective, reversible restorations not only improve masticatory function but enhance esthetics, allowing patients to develop socially.

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