

Esthetic, Functional and Psychological Management of Ectodermal Dysplasia using a Full Mouth Rehabilitation Approach

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ABSTRACT

Congenital and developmental disorders often pose a challenge to the rehabilitation of patients with not only severe dental destruction but also significant psychological trauma. Patient care in such cases often forms a different genre of therapy as it requires special attention and meticulous care by the rehabilitating dentist. The remarkable impact of successful treatment in these cases not only far exceeds the dentofacial benefits but also provides profound psychosocial recuperation to these young patients. The literature has demonstrated the benefits that corrective prosthodontics has for the self-esteem and social well-being of these patients. This case report highlights the intricacies of one such full mouth rehabilitation of a young patient with ectodermal dysplasia using an interdisciplinary approach.

Keywords: Ectodermal dysplasia, Hypohidrotic, Anhidrotic, Full mouth rehabilitation.

How to cite this article: Chablani AA, Ram SM, Shetty O. Esthetic, Functional and Psychological Management of Ectodermal Dysplasia using a Full Mouth Rehabilitation Approach. J Contemp Dent 2013;3(2):92-97.

Source of support: Nil

Conflict of interest: None declared

INTRODUCTION

Ectodermal dysplasia (ED) is a hereditary disorder characterized by abnormal development of certain tissues and structures of ectodermal origin and their accessory appendages. It occasionally involves dysplasia of the mesoderm-derived tissues.¹

The National Foundation of Ectodermal Dysplasia (NFED) defines this condition as a genetic disorder in which there are congenital birth defects (abnormalities) of two or more ectodermal structures.² These structures may include skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye and ear and parts of the other organs.³ Diagnosis of ED can be complicated due to the variation in types, the range of abnormalities and the severity of the defect.

ED has two forms: The hidrotic and hypohidrotic (anhidrotic) type. In both these types, the manifestations of teeth and hair are similar but those of nails, sweat glands and hereditary patterns tend to differ. The hidrotic form is inherited as an autosomal dominant trait that affects teeth, hair and nails but usually spares the sweat glands.³ Hypohidrotic ectodermal dysplasia is an X-linked recessive mendelian character, males being affected much more frequently. Also known as Christ-Siemens-Touraine

syndrome, it is characterized by a clinical triad of hypohidrosis, hypotrichosis and hypodontia.⁴ Individuals affected usually exhibit a soft, smooth, thin, dry skin with partial or complete absence of sweat glands. Such persons present with significantly reduced perspiration and consequently suffer from hyperpyrexia with an inability to endure warm temperatures.⁵ The sebaceous glands and hair follicles are often defective or absent. The bridge of the nose is depressed. The supraorbital ridges and frontal bosses are pronounced.

Intraoral findings invariably manifest complete or partial anodontia with frequent malformations (cone shaped) of any teeth present complicating their use as removable or fixed partial denture abutments, but sometimes offering the possibility for use as abutments for overdentures.^{6,7} It is seen that even when complete anodontia exists, the growth of the jaw is not impaired thus implying that the development of the jaws, except for the alveolar process, is not dependent upon the presence of teeth. However, as the alveolar process does not develop in the absence of teeth, there is a reduction from normal vertical dimension resulting in the protuberant lips. In addition, the palatal arch is frequently high and a cleft palate may be occasionally present.¹ According to Bessermann Nielsen, the salivary glands, including the intraoral accessory glands are sometimes hypoplastic in this disease which may result in xerostomia and dry, cracked lips.¹ 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.

The remarkable impact of successful treatment in these cases not only far exceeds the dentofacial benefits but also provides profound psychosocial recuperation to these young patients. The literature has demonstrated the benefits that corrective prosthodontics has for the self-esteem and social well-being of these patients. This clinical report highlights the intricacies of a full mouth rehabilitation procedure for a patient with ED using an interdisciplinary approach.

CASE REPORT

A 25-year-old female reported to the Department of Prosthodontics with a chief complaint of multiple missing teeth and the remaining teeth being malformed, thus giving

an unpleasant appearance and a lack of chewing efficiency. A detailed medical and social history was obtained followed by an extraoral and intraoral examination.

Extraorally, the patient presented with features, like prominent forehead (frontal bossing) and supraorbital ridges, receding hairline, scanty eyebrows, a depressed nasal bridge and protuberant lips (Fig. 1). The patient had a slightly concave profile due to overclosure of the jaws and a reduced vertical dimension of occlusion (Fig. 1). The symptoms given by the patient included xerostomia and cracking of lips, reduced perspiration and an inability to endure warm temperatures. A thorough clinical and



Fig. 1: Preoperative extraoral frontal and lateral views

radiographic examination revealed a case of mixed dentition with partial anodontia (oligodontia) with the conical-shaped maxillary anterior teeth (Figs 2 and 3). The 'freeway' space available was 7 mm bilaterally at the premolar region with a collapsed vertical dimension of occlusion causing the lower anterior teeth to impinge onto the rugae area of the palate leading to a traumatic bite.⁸

The lack of adequate number of abutment teeth, underdeveloped alveolar ridges, inadequate bone height and width and the patient's hesitancy toward intense hard tissue reconstructive procedures made rehabilitation of the maxillary arch using any tooth or implant supported fixed prostheses a difficult option to consider. The mandibular arch, however, had an adequate number of healthy abutment teeth to allow for tooth-supported fixed prostheses. The proposed treatment plan thus comprised of a full mouth rehabilitation procedure restoring the lost vertical dimension using a combination of removable and fixed prostheses.

However, due to the patient's strong desire for fixed replacements as a final treatment solution, it was decided that the maxillary arch would receive full coverage single piece long span splinted ceramometal restorations, whereas a 6 unit ceramometal bridge anteriorly with individual full coverage ceramometal crowns posteriorly would be fabricated for the mandibular arch. As per the concept of a



Fig. 2: Preoperative intraoral views

shortened dental arch, it was planned that the maxillary arch would be restricted to the second premolars in order to minimize undue stresses to the abutment teeth.⁹ The final treatment plan offered to the patient comprised of restoration of the lost vertical dimension of occlusion with full coverage ceramometal restorations for both the arches.

After a thorough oral prophylactic treatment, maxillary and mandibular full arch impressions were made using alginate, an irreversible hydrocolloid impression material [Tropicalgin, Zhermack, Badia Polesine (RO), Italy]. The maxillary diagnostic cast was mounted on a semiadjustable articulator [Hanau H2 series, Whip-mix Corporation, Fort Collins (CO), USA] using a fascia face-bow provided with the articulator (Fig. 4).¹⁰ The patient was deprogrammed with a tightly rolled cotton placed between the incisors for a span of 5 minutes.^{11,12} An interocclusal record, used to mount the mandibular diagnostic cast, was made with a free-flowing recording material [Aluwax, Aluwax Dental Products Ltd, Allendale (MI), USA] at the centric relation (CR) position at a restored vertical dimension.

The articulator was programmed using a mean of the protrusive records obtained from the patient and the lateral

condylar guidance values calculated using the Hanau's formula ($L = H/8 + 12$).¹³ The incisal table was kept 5° steeper than the horizontal condylar guidance values to facilitate posterior disclusion on protrusion. An occlusal plane was developed using a customized Broadrick's occlusal plane analyser (BOPA) (Fig. 5).¹⁴ Diagnostic wax-up incorporated a mutually protected occlusion with a unilateral balance in lateral excursions as the canines were malformed.¹⁵⁻¹⁷

Taking the preoperative OPG, IOPAs and the diagnostic wax-up into consideration, preprosthetic procedures were planned and executed. The over-retained deciduous mandibular central incisors along with a badly broken down maxillary anterior tooth were extracted due to inadequate bone support. Severely maligned teeth underwent root canal treatment and received a prefabricated post [ParaPost X, Coltene Whaledent AG, Cuyahoga falls (OH), USA] and core build-up [LuxaCore Z Dual, DMG Chemisch Pharmazeutische Fabrik GmbH, Hamburg, Germany] for reinforcement. A clear acrylic autopolymerizing resin stent was fabricated as a guide to mark the extent of crown lengthening procedure required. The crown lengthening procedure was performed by the Department of Periodontics and a healing period of 2 weeks was thereby prescribed.

Vacuform trays fabricated on the duplicated stone models of the wax-up of both the arches were used as indices for the temporization. Intraoral tooth preparations and simultaneous temporizations were carried out using the direct method of temporization with an autopolymerizing composite resin [Luxatemp Star, DMG, Chemisch Pharmazeutische Fabrik GmbH, Hamburg, Germany] (Fig. 6). The temporary restorations were finished, polished and cemented with a noneugenol-based temporary cement [TempoSIL, Coltene Whaledent AG, Cuyahoga falls (OH), USA] (Fig. 7). Minor occlusal adjustments, if any, were done intraorally and a mutually protected occlusion with unilateral



Fig. 3: Radiographic examination—OPG



Fig. 4: Orientation relation record



Fig. 5: Broadrick's occlusal plane analyzer



Fig. 6: Abutment teeth preparations

balance was established. These restorations were maintained for an evaluation period of 6 weeks during which no signs of gingival inflammation or any other discomfort related to the temporomandibular joint (TMJ), muscles or teeth were evident.

The procedure of final impression making was undertaken in the fifth week. Knitted retraction cords [000, Ultrapak, Ultradent Products Inc, South Jordan (UT), USA] were used for gingival retraction. Final impressions were made with a double-mix double-step impression technique using additional silicone polyvinyl siloxane impression materials [Aquasil, Dentsply Caulk, Milford (DE), USA] comprising of soft putty and light viscosity wash impression material.¹⁸ An anterior jig using impression compound was fabricated and an interocclusal record was obtained using a bite registration material [Jet Bite, Coltene Whaledent AG, cuyahoga falls (OH), USA] to mount the final casts on the articulator.

The articulator was reprogrammed with the patient's protrusive records at the restored vertical dimension of occlusion. The incisal guidance established in the patient's mouth based on the esthetics and phonetics was transferred

to the articulator with the help of temporary restorations. Metal copings for the full coverage ceramometal restorations were fabricated and evaluated intraorally for the marginal fit, followed by a bisque trial to evaluate the contour and shade of the final restorations.

The final restorations were evaluated for the desired static and excursive occlusal relationships on the programmed articulator. The restorations were cemented with a noneugenol temporary luting cement [TempoSIL, Coltene Whaledent AG, Cuyahoga falls (OH), USA] (Fig. 8). The occlusion planned for the patient during the wax-up and the temporization stages was effectively seen to be replicated in the final restorations. These restorations were permanently cemented with resin-modified glass ionomer luting cement [Fuji CEM, GC Corporation, Tokyo, Japan] (Fig. 9) within a span of 2 weeks once the patient was accustomed with no negative signs or symptoms. A regular follow-up was maintained.

DISCUSSION

The prosthodontic management of ED requires a thorough knowledge of the condition to handle the special problems



Fig. 7: Temporization



Fig. 8: Final restorations in maximal intercuspation position



Fig. 9: Postoperative views

associated with the treatment. A multidisciplinary team approach is often recommended for optimal dental management of the condition.

The management of the patient with ED, using fixed prosthodontics proved to be a highly acceptable option as long as the patient satisfaction was concerned with a strong demand for fixed restorations and a desire to retain as much of her natural dentition as possible. The treatment modalities involving intense surgical reconstructions and implant supported fixed prosthodontics at such an early age, though ideal were considered invasive and unacceptable to the patient.⁵ Another treatment option which would have been more conservative and ideal would be a tooth supported overdenture prostheses for the maxillary arch but was not acceptable to the patient. The one piece long span prosthesis planned for the maxillary arch would be contraindicated if the patient's skeletal growth was incomplete.

The concept of a shortened dental arch wherein the maxillary arch was restricted to the second premolars posteriorly helped minimize undue stresses from being transferred to the abutment teeth.

Group function occlusal scheme provided for a more stable and self-maintaining scheme of occlusion and helped to distribute the force over greater number of teeth on the mediotrusion side.

The rehabilitation procedure followed in this case seemed convenient to plan, execute and helped to obtain the desired outcome without any difficulties. The incisal guidance, the cuspal inclines of posterior teeth, the plane of occlusion, unilateral balance and the esthetic plane obtained during the wax-up stage were all effectively replicated at the temporary stage. Minor discrepancies, during the temporary stage, if any, were adjusted intraorally and indices of the same were used as a guide to contour the final restorations.

CONCLUSION

Congenital and developmental disorders often pose a challenge to the rehabilitation of patients with not only severe dental destruction but also significant psychological trauma. This type of patient care forms a different genre of therapy requiring special attention and meticulous care by the rehabilitating prosthodontist. Treatment, in such cases, is of utmost importance, not only from a functional standpoint but also from a psychosocial health status point. Dental therapy as a phase in the treatment of ED is essential. The method of treatment of a patient with ED has been reported. As a result of dental therapy, the patient has seemed to improve esthetically, functionally, psychologically and physiologically. A well co-ordinated interdisciplinary approach can thus provide the desired dentofacial benefits and profound psychosocial recuperation to the patient.

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