

Prosthetic rehabilitation of an adolescent with ectodermal dysplasia utilizing multidisciplinary approach: A case report

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ABSTRACT

Ectodermal dysplasia (ED) is a hereditary disorder characterized by abnormal development of two or more embryonic ectodermal structures such as skin, nails, teeth, hair, eyes, and sweat glands. The main oral manifestations of ED include oligodontia, malformed teeth and, deficient alveolar ridges. Inheritance of ED is X-linked and expressed typically in males. This case report explains a multidisciplinary treatment of a 16-year-old adolescent suffering from ED with underdeveloped premaxilla, underdeveloped alveolar ridges, retained deciduous teeth, and thick fibrous frenal attachment. Prosthetics rehabilitation was done using removable cast partial dentures with lip support. It also enumerates the important role of a multidisciplinary approach in dental treatment, helping the patient's healthy transition from childhood to adulthood. Dental professionals should be able to identify the oral features of ED and provide proper oral care (as early as 3–4 years) to restore the oral functional demands and esthetics.


Key words: Cast partial denture, Corneal opacity, Ectodermal dysplasia, Ectropion, Multidisciplinary approach, Underdeveloped premaxilla

Ectodermal dysplasia (ED) is a rare genetic disorder characterized by aplastic or dysplastic development of two or more tissues having an embryonic ectodermal origin [1]. Thurnam (1848) firstly described and reported two typical patients with ED in the medical literature [2]. Abnormal gland, tooth bud, hair follicle, eye, nail development, and skin are commonly encountered findings in individuals suffering from ED. The estimated incidence of ED is about 7 in 10,000 births. All possible inheritance modes of Mendelian trait (autosomal dominant or recessive, X-linked dominant or recessive) are followed in ED [3]. Usually, inheritance of ED is X-linked, which is typically expressed in males and less commonly in females who carry it as carriers [4]. Hypohidrotic recessive and autosomal hidrotic dominant are the two most frequent types of X-linked ED [5]. Dental characteristics are also present sometimes, characterized by conical or peg-shaped teeth, hypodontia, or anodontia (with both deciduous and permanent dentition), malformed teeth, underdeveloped alveolar ridges, and delayed eruption (permanent teeth) [2].

CASE REPORT

A 16-year-old male, a student by occupation, had reported to the Department of Prosthodontics complaining of dental pain, mobile teeth, inability to chew food for the past 6–7 months, improper speech, and poor facial esthetics. The patient's medical history revealed that he was not on any medication and was prone to recurrent throat infection and pyrexia. There was no previous history of ED in the family and all his siblings were healthy. The patient had no previous dental history.

On general examination, the patient presented with hypohidrotic ED with corneal opacity, pale skin with no body hair on arms and legs, deformity with the left foot, dry eyes, ectropion with the right eye, and short upper lip (Fig. 1). Corneal opacity was seen with the entire right eyeball and had lost 80% of its vision. He had an ovoid facial form and straight profile with reduced vertical dimension giving a typical senile appearance. Oral examination revealed underdeveloped premaxilla, underdeveloped alveolar ridges, retained deciduous teeth, thick fibrous frenal attachment extending till the incisive papilla (Fig. 2a), and oligodontia (25 permanent teeth were absent). Teeth which were present were 14, 24, 26, 34,

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37, 44, and 47 (permanent dentition) and 55, 65, 71, 73, 75, 81, and 85 (deciduous dentition). All four present premolars were rotated. 26, 37, and 47 had pit and fissure caries, 65 was grossly decayed, 71 and 81 were Grade III mobile, and 73 was Grade II mobile (Fig. 2a and 2b). The tongue was normal in size. Due to the lack of teeth in occlusion, horizontal and vertical relations could not be determined.

Panoramic radiograph examination revealed physiologic root resorption with 65, 71, 73, 75, 81, and 85, and deficit bone formation (Fig. 3a). An intraoral periapical radiograph of 55 was taken to determine the length of available root length.

The treatment was planned and performed in the following manner: Extraction of all deciduous teeth except 55 followed by a cast partial denture (CPD) with the enhancement of the lip support followed by fixed oral rehabilitation by bone grafting, and implant placement after growth completion. The patient was reluctant to wait and undergo intentional root canal treatment; hence, a CPD was planned instead of an overdenture. The dental team comprised a prosthodontist, an oral surgeon, an endodontist, a periodontist, and an oral radiologist, who divided treatment into the restorative phase, surgical phase, prosthetic phase, and maintenance phase.

The patient was explained about the course of treatment and informed consent was obtained from the patient to use his data for educational purpose. Preliminary impressions were made using irreversible hydrocolloid and poured in Type III dental stone. Diagnostic facebow transfer was taken and transferred to a semi-adjustable articulator and was mounted in an existing bite of the patient. After evaluating the panoramic and intraoral periapical

radiographs, a decision was made to extract all the deciduous teeth except 55 as root appeared to be long enough on the intraoral periapical radiograph to act as an abutment.

Restorative Phase

Endodontist performed the restorative treatment with 26, 37, and 47 using composite by occlusal stamp technique before the planned extraction was executed. Before the cavity preparation, the occlusal stamp of the existing occlusal surface was made using the flowable composite and applicator tip. Caries were excavated and packable composite was used to restore the teeth. After placing the packable composite in the prepared cavity preparation, a Teflon tape was placed on the occlusal surface and the occlusal stamp was pressed on it, thereby recreating the existing anatomy.

Surgical Phase

Once the restorative phase of the treatment plan was complete, extractions were done. Oral surgeon extracted the teeth according to the formulated treatment plan in two visits under local anesthesia. The extraction of teeth in the second and third quadrants was done in the first visit and extraction of the fourth quadrant in the second visit.

After the wound healing of the extraction site, a frenectomy was performed by a periodontist. Frenectomy was accomplished



Figure 1: Corneal opacity and ectropion with the right eye

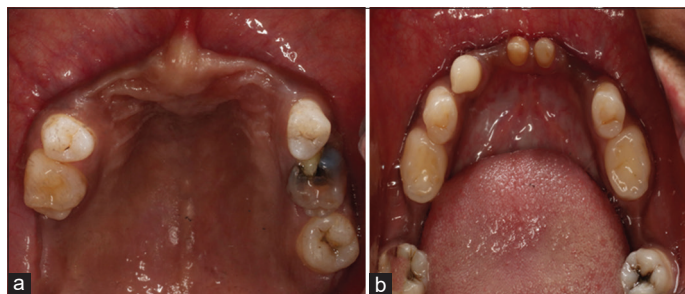


Figure 2: (a) Intraoral occlusal view of maxillary arch depicting underdeveloped premaxilla and thick fibrous frenal attachment; (b) intraoral occlusal view of mandibular arch with over-retained deciduous teeth



Figure 3: (a) Pre-operative panoramic radiograph showing physiologic root resorption of deciduous teeth and deficit bone in premaxillary region; (b) addition of modeling wax and teeth arrangement beyond the metallic framework to make up for the underdeveloped premaxilla for sufficient lip support; (c) mandibular trial denture: Placement of four anterior teeth and molar to verify occlusion and esthetics

by routine scalpel technique under local anesthesia. A hemostat was inserted into the depth of vestibule engaging the fibrotic frenal attachment. Until the hemostat was free, incisions were placed on the upper and undersurface of the hemostat. After the removal of thick fibrotic attachment, blunt dissection was performed to relieve the fibrous attachment. Interrupted sutures were placed using 4-0 black silk suture material. A periodontal pack was used to cover the operated site. One week post-surgery, the pack, and sutures were removed.

Prosthetic Phase

Preliminary impressions were made using irreversible hydrocolloid after completion of the surgical phase. Impressions were poured in Type III dental stone and surveying was done. Survey lines were too low, close to the gingival margin since the teeth were lingually tilted. Designing of CPD was finalized and required preparation on teeth was done. Surfaces of teeth were altered to raise the survey line and rest seat preparation was done. Dimpling was done with 37 and 47 since they were lingually tilted.

Border molding was done on the fabricated custom tray, an impression was made in polyether (Impregum Monophase 3M ESPE Pentamix) and master casts were poured using Type 4 die stone. CPD framework was made according to this master cast. While recording the jaw relation, the underdeveloped premaxilla was constructed using modeling wax until proper lip support, facial esthetics, and phonetics were achieved. Facebow transfer was taken and transferred on a semi-adjustable articulator; mounting was done in recorded centric relation. Teeth arrangement was done followed by try-in (Fig. 3b and 3c). Phonetics, facial esthetics, lip support, and teeth visibility were evaluated in the trial dentures.

The final prosthesis was fabricated after the patient's approval regarding esthetics and phonetics. There was a considerable enhancement in the pre-operative and post-operative facial profile



Figure 4: (a) Intraoral pre-operative view in maximum intercuspation; (b) intraoral post-operative view in maximum intercuspation; intraoral post-operative (c) maxillary and (d) mandibular occlusal view

and esthetic outcome until definitive fixed prosthetic treatment is done (Figs. 4 and 5).

DISCUSSION

Craniofacial signs which are typical to ED are mandibular prognathism, maxillary hypoplasia, a reduced lower facial height, facial concavity, lip eversion, prominent forehead, and a saddle nose [6]. Anodontia or hypodontia leads to failure in alveolar process formation, ensuring reduced vertical dimension and protuberant lips [2,7]. Improving sagittal and vertical relationship and developing efficient masticatory function, esthetics and phonetics are necessary for oral rehabilitation of the patient with ED [1]. Despite extensive research and dental treatments, no specific treatment protocol has been formulated till now for dental rehabilitation of ED. The literature is limited to clinical reports stating the use of orthodontic rehabilitation based on removable and/or fixed prosthesis. The fixed dental prosthetic treatment has its own disadvantages and cannot be rendered until growth is completed (especially when planning for implant placement) [5].

Orofacial impairment not only strikes vital functions such as mastication and breathing but also hampers speech and socialization [7]. Evaluation of psychosocial status of ED patient may be a key to prosthodontic treatment. Disturbances in the social life of the affected individual can be seen due to poor esthetics, aged facial appearance, and clinical features of ED. Improving the oral function and facial esthetics with the help of removable prosthodontics to manage orofacial disfigurement can enhance patient's self-esteem and confidence until complete growth has occurred to receive fixed prosthodontic treatment [8].

The treatment can be initiated at an early age to restore functional dental needs. Removable partial dentures and complete overdentures are usually used in growing patients. In general, implant placement is considered when the patient has attained complete growth (not given below 18 years) [9,10].

Hypodontia or anodontia is the most common clinical feature of ED [2]. In this case, preserving the single deciduous tooth and remaining permanent teeth was advantageous with regard to proprioception, retention, stability, and function. Circumferential



Figure 5: (a) Extraoral pre-operative; (b) extraoral post-operative view showing increased lip support and enhanced esthetics

clasps were preferred in this case as gingivally approaching clasps were not possible due to the lingual inclination of the teeth. The patient was expecting a fixed restorative treatment, but he willingly accepted the treatment after explaining him the treatment plan of the post-growth period. The patient was advised not to bite on his anterior teeth and use the posterior teeth for mastication.

Yenisey *et al.* treated a 10-year-old male patient by a multidisciplinary approach by planning an overdenture and an implant-supported prosthesis when the patient had completed his growth [2]. Al-Ibrahim *et al.* used a combination of surgical and prosthetic intervention, a maxillary tooth-supported fixed detachable telescopic prosthesis, and an implant-supported mandibular fixed partial denture was fabricated for the patient [8].

Early prosthetics intervention helps the patient to have a normal life with social, emotional, and psychological stability [8]. Treatment can be started as early as 3–4 years of age of the patient, this instills a positive effect in gaining self-confidence [11]. Limited prosthetic rehabilitation options are available if early intervention is avoided, a similar thing happened with this particular patient. In the era of rapidly advancing implant dentistry, CPDs still are an advantageous treatment option from the perspective of psychological development and esthetics appearance until full growth is achieved and extensive surgical procedures are done.

CONCLUSION

A multidisciplinary approach is an important part of treating a patient with ED. This will ensure that all the patient needs are fulfilled and addressed properly for a smooth transition from “childhood-adolescence-adulthood.”

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