Case Report

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Prosthodontic Rehabilitation of a Patient Suffering from Ectodermal Dysplasia: A Case Report

Abstract:

Ectodermal dysplasia is a rare congenital disease that affects several ectodermal structures. Children with ectodermal dysplasia may have various manifestation of the disease that differ in severity and may involve teeth, skin, hair, nails, sweat glands and sebaceous glands. A 16 year Old Girl reported in the department of Prosthodontics and crown bridge, with chief complain of of inability to masticate and her unesthetic appearance. She desired replacement of her missing teeth. Family history revealed similar findings in her 18-year old brother. Clinical findings and family history suggested a diagnosis of hypohydrotic ectodermal dysplasia. The present clinical report demonstrated that maxillary flexible RPDs associated with direct composite restorations, Zirconia crowns and mandibular complete denture with CEKA supported Stud attachment are a reversible, relatively inexpensive method of treatment for ectodermal dysplasia patients especially during their growth years. The esthetic, light weight, and good retentive and fracture properties of the flexible denture base materials make a good alternative to the conventional and cast partial dentures.

Key-words: Ectodermal dysplasia; Denture, Overlay Denture Precision attachment

Introduction:

Ectodermal dysplasia (ED) is a collection of genetic diseases characterized by abnormalities in the development of at least two tissues originating from the embryonic ectoderm. However, due to overlapping characteristics, it is challenging to categorize them clearly. The two primary classifications of ED are Hidrotic (Clouston's syndrome) and Hypohidrotic (Christ-Siemens-Touraine syndrome)[1]. The symptoms of the sweat glands differ between these two types; sweat glands are normal in Hidrotic Ectodermal Dysplasia, while they are absent in Hypohidrotic Ectodermal Dysplasia (HED), which is inherited through an autosomal dominant gene. HED, which is inherited by an X-linked recessive gene, is the most prevalent form of ED syndrome, affecting males more severely and frequently[1]. Approximately 1 in 100,000 people have HED[2].

Patients with HED do not sweat at all or very little sweat, which results in excessively high fevers since the skin cannot regulate temperature as it should. The skin is light-coloured and thin. There may be little or no hair. Some patients' hair growth after puberty improves. There may be little or no hair on the eyebrows, eyelashes, or other parts of the body. Fingernails and toenails may be tiny, thick or thin, brittle, discoloured, broken, and/or ridged due to improper development.

Extraoral symptoms of ectodermal dysplasia (ED) include frontal bossing, a depressed nasal bridge, protruding lips, and hypotrichosis. Dental features of ED include hypoplastic

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conical teeth, underdeveloped alveolar ridges, and missing primary and/or permanent teeth (anodontia or hypodontia)[3,4.] The alveolar bone, where missing teeth are typically replaced, does not develop correctly, resulting in a reduced vertical dimension and a distinctive aged appearance in the face[5,6]. Treatment with functional and prosthetic appliances can help to reduce the deviation from normal facial development with age in individuals with HED[7].

When planning a treatment for ectodermal dysplasia (ED) patients, several factors should be considered, including the patient's age, the stage of development in relation to the missing teeth, any anomalies in the soft tissue, the presence of a malformed dentition, and the psychological condition of the patient. All of these factors are important in determining the appropriate course of therapy for the patient[8]. As a result of the orofacial features manifesting at such a young age in patients with ED, restoring look and function is more difficult than normal. There are several possible treatments for this problem, however removable prosthodontics is the most popular prosthetic therapy for ED in young patients[9]. There is adequate research to show that full dentures are frequently made for anodontia patients while a more permanent prosthesis is made after the completion of growth[10].

In this case presentation, the effectiveness of using a flexible denture base material is examined for creating a maxillary removable denture and tooth-supported stud attachment for a child with HED. The denture is fabricated using conventional acrylic resin overdenture denture methods.

Case Report:

Diagnosis:

A 16-year-old female patient visited the Prosthodontics and Crown & Bridge department of Teerthanker Mahaveer Dental College and Research Centre in U.P. with a complaint of difficulty in chewing and unappealing appearance. She expressed her desire to have better aesthetics and to replace her missing teeth. Upon further inquiry, the patient disclosed that she had a history of not sweating, dry skin, and increased body temperature. The extraoral examination revealed several physical characteristics such as thinning hair, protruding forehead, prominent eyebrows, swollen lips, and a decreased lower facial height. These factors contributed to her inability to chew properly and affected her overall appearance. (Fig-1A)

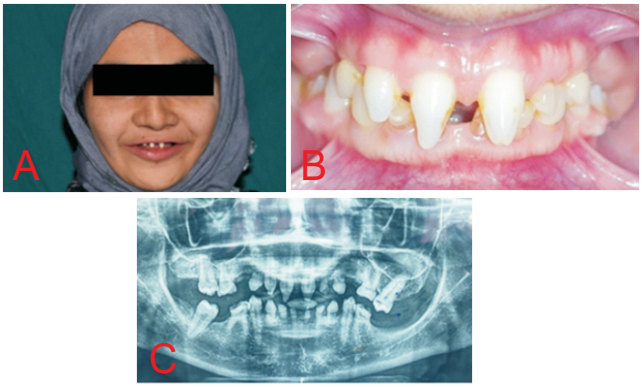

Figure 1: Pre-Operative Presentation of patient
A-Extra oral B- Intra Oral C- OPG

Intraoral examination revealed very less amount of saliva. Permanent teeth present with respect to (w.r.t) 11,12,16,21,26,34,44,47, and deciduous teeth w.r.t 54, 55, 64, 65,71,72,73,75,81,82,83,85 (World Dental Federation notation (FDI)). (Fig-1B) Also, showed the presence of cone-shaped teeth in positions 11, 12, and 21. In addition, the mandibular edentulous ridge in the third quadrant was found to be resorbed, and the maxillary tuberosity in the second quadrant was enlarged.

The patient's family history indicated that her 18-year-old brother had similar dental findings. Based on these clinical and family history findings, the patient was diagnosed with HED.[11,12]

Treatment Planning:

Panoramic radiograph revealed complete root formation w.r.t 11,12,21,34,44,47 and there was no evidence of any impacted or developing permanent tooth (fig-1C). Deciduous teeth w.r.t 54,55,64,65 along with all the permanent teeth were planned to save and remaining undeveloped and resorbed deciduous teeth were extracted. (fig-2)

Figure 2 – Post Extraction Intra-oral pictures
A- Maxillary, B- Mandibular, C & D – Occlusal

Primary impression was made using irreversible hydrocolloid (Algitex,DPI)and poured in type 2 gypsum (Doctors brand, Jai durga sales) and Diagnostic jaw relation and mounting was done in a mean value articulator to access the inter-arch distance which revealed that there is minimal space available in the 3rdquardrant to replace teeth due to the enlargement of the maxillary tuberosity in the 2nd quadrant, hence 65 was extracted along with 5mm of tuberosity reduction. (fig-3)

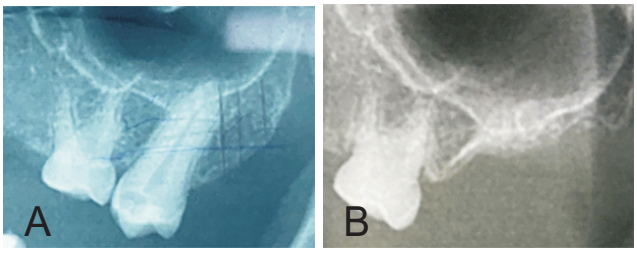

Figure 3: Pre and Post Operative radiograph after maxillary tuberosity reduction.

Patient was not economically sound enough, so implant supported prosthesis was not an option. To achieve both functional and aesthetic improvements, a maxillary flexible removable partial denture (RPD) with zirconia crowns on the anterior teeth and a mandibular overdenture with the CEKA Preci Line-PreciClix Radicular RC attachment System (CEKA-attachment) were proposed. [12] (fig-6A)

Considering the patient's age and the condition of the remaining natural teeth, a Valplast flexible partial denture was proposed as it offers several benefits such as increased retention, durability, biocompatibility, patient comfort, and requires no tooth or tissue preparation. The Retento Grip tissue-bearing mechanism employed by the Valplast flexible denture base material was deemed necessary for retention in this case. [13,14]

CEKA attachment (fig-6) system was planned due to residual ridge resorption in the 3rd quadrant. CEKA attachment is tooth supported and can be placed in a single appointment and it is easy to use and more economical when compared with the ball attachment system, the patient receives secure retention and stability for their overdenture. This attachment system comes with different retentive rubber female cups, yellow females have normal retention, White females have reduced retention, Red females have increased retention. In this case yellow female component was used (fig-6B)

Treatment Phase:

The treatment plan was comprised of three phases: oral prophylaxis, endodontic treatment, and prosthetic treatment. The patient was informed about the procedures and gave consent. During the endodontic phase, intentional root canal therapy was performed on teeth 34 and 44.

Shade selection and Composite build was done followed by tooth preparation w.r.t. 11,12,21, gingival retraction using 000 non-impregnated, knitted retraction cord (sure-cord,superendo sure-dent corp) was done prior to impression making (Fig-4A), impression were further made by Poly vinyl siloxane (PVS) putty and light body impression material(photosil, dental product of India DPI) and poured in type 4 gypsum (kalrock, kalabhai Karson pvt. Ltd.) later Zirconia crown were fabricated and were cemented w.r.t 11,12,21 using resin cement(multilink-speed, IvoclarVivadent). (Fig-4B) After cementation of the crowns PVS impression was made and cast was poured in type -3 gypsum (kalrock, kalabhai Karson pvt. Ltd.), surveying was done using Jelenko surveyor to determine favourable and unfavourable undercuts later all the unfavourable undercuts was blocked out using type 2 gypsum from the cast and a path of insertion was determined for the maxillary RPD.

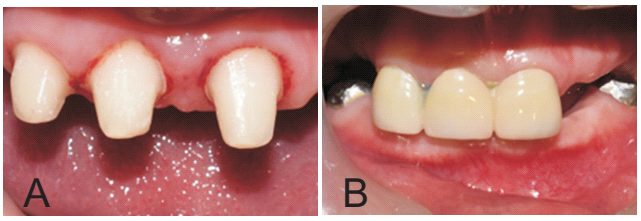

Figure 4: A-Composite Build up and Crown preparation, B-Cementation of Zirconia Crown

For the mandibular overdenture, teeth were reduced upto the level where attachment would come w.r.t 34,44 and dome shaped tooth preparation was done w.r.t 47 and then impression was made using PVS impression material and cast was poured in a type 4 gypsum later temporary copings were made using a bis-acrylic composite (Protemp-4,3M) up to a level where attachment would come, metal coping was fabricated using the same cast w.r.t 47 as no attachment was to be given over it, later temporary crowns were cemented using temporary cement w.r.t 34,44 and metal crown was cemented using glass ionomer luting cement(GC Gold label glass inomer, GC corporation, Tokyo, Japan) w.r.t 47.

An irreversible hydrocolloid-alginate was used to make a primary impression of the mandibular arch. Custom trays were then made using cold-cure acrylic resin, and border molding was performed. A final impression of the mandibular arch was made using light body PVS impression material, which was poured with type 3 gypsum. Occlusal rims were prepared for both arches using a facebow transfer (HANAU

Spring Bow, Whip Mix Corporation, USA) (Fig5A), and the jaw relation was articulated in a semi-adjustable articulator (HANAU Wide-Vue Articulator, Whip Mix Corporation, USA). After teeth arrangement and try-in (FIG-5B), minor adjustments were made before the fabrication of final overdenture and RPD.

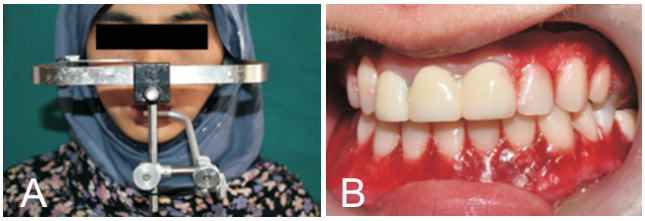

Figure- 5: A-Facebow transfer , B- Try in

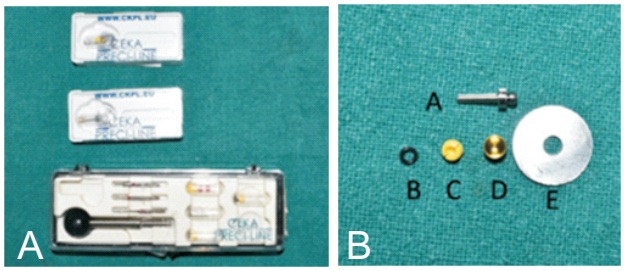

Figure 6: A-CEKA Attachment and tools

B- Attachment

a- ceka metal post with male attachment

b- rubber spacer

c- yellow female ruber cap

d- metal female housing

e- attachment holder

To place the attachments, the temporary coping was removed from teeth 34 and 44. The gingiva level of these teeth was reduced, and any sharp edges were rounded off. For post placement, pre-drilling burs from the CEKA kit were used to prepare the post space, while diamond burs were employed to prepare the base of the Preclix post. The post diameter was then expanded using a reamer(Fig-7A). The post was cemented with resin cement (Fig-7B), after which yellow female retentive rings were inserted into the CEKA female housing using a CEKA insertion tool. Female housing along with yellow retentive ring was placed over the Male ball attachment which was attached with the metal post, (Fig-7C) denture was seated over the female housing to verify occlusion and pre-determined vertical height the seating of the denture over the post was easy as denture already had space for female housing because of the temporary crown space which was made at the level of attachment. After a satisfactory verification, female housing was picked up in the denture using autopolymerising acrylic resin excess flush were trimmed using a round bur. The denture underwent adjustments and was checked again to ensure appropriate border extensions, vertical and horizontal dimensions, and aesthetics.(Fig-8).

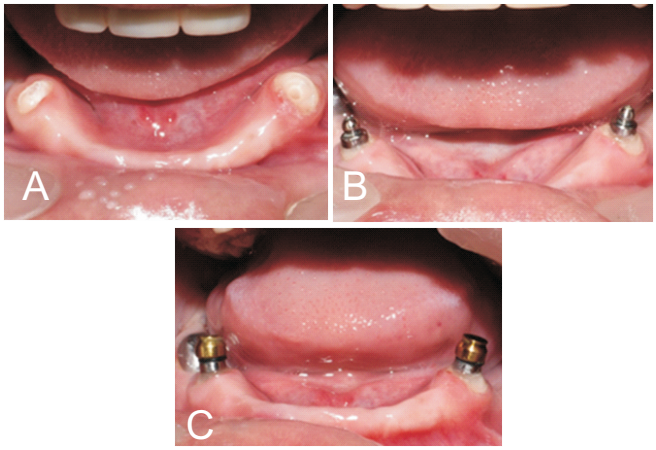

Figure 7: A- Post space preparation

B- Cementation of the CEKA male component

C- Placement female retentive cap over the male component

Figure 8 : Pick up of the Female component in the denture and placement of the retentive rubber sleeves

The patient was provided with post-insertion instructions and was advised to return for a check-up after 24 hours. She received guidance and training on the careful insertion and removal of the denture, and was encouraged to maintain proper oral hygiene to ensure a positive outcome for both the abutments and the prosthesis. Patient and her family were satisfied with the prosthesis and the aesthetic.(Fig-9)

Figure 9: Post Operative Picture

Discussion:

As stated by Sir MM Devan "Perpetual preservation of what remains is more important than the meticulous replacement of what is missing" that is Preventive Prosthodontics should be the rationale of any treatment, specially in childrens with ectodermal dysplasia whenever possible remaining natural tooth structure must be saved for better neuromuscular control thus regulating the biting force over the denture[15,16,17]. Patients with ectodermal dysplasia face numerous challenges when it comes to prosthodontic rehabilitation, such as reduced alveolar bone height, limited remaining tooth structure, and inadequate inter-arch distance, which can impact occlusal stability, aesthetics, and mastication. It is generally advised to postpone implant placement until adolescence in patients with incomplete craniofacial growth to minimize the risk of failure[18]. Implant placement in younger patients may result in adverse outcomes such as damage to tooth germs and constraints on skeletal craniofacial growth. In this instance, implant therapy was not selected due to the patient's ongoing growth and development, inadequate alveolar bone support, and financial constraints.

In young patients, RPDs are a reversible treatment option that can greatly enhance functions and esthetics without compromising compromised dentitions[19]. However, metal clasps on anterior teeth can cause aesthetic issues with the use of cast partial dentures[20]. To address this concern, acetal resins have emerged as a viable alternative tooth-colored denture clasp material to improve aesthetics[21]. Furthermore, the use of thermoplastic materials like Valplast has become increasingly popular in recent years. This innovative procedure involves the softening of a fully polymerized basic material through heat, without any chemical changes, followed by injection molding to create dentures with improved flexibility and comfort.[22]

The Ceka attachment is a type of stud attachment that offers several benefits such as good aesthetics, easy utilization, and assembly in removable partial denture prosthesis. However, it requires high technical skill to be applied effectively. Unlike other attachments, it requires a minimum occluso-gingival abutment height but provides better retention. In this case report, we opted for a Ceka attachment because of limited interocclusal space. However, the attachment has a drawback in that the plastic resilient cap tends to wear out over time and loses its retentive capacity, necessitating replacement. The patient was informed beforehand that they may need to replace the female housing assembly in the future.[23]

In a case done by Sabhlok[23] et al the author used similar attachment system and found a satisfactory result however he stated that CEKA attachment system needs a high technical skill, this is true if attachment is placed in a conventional manner i.e The denture housing was secured onto the laboratory analogue using rubber base impression material, and then the denture was acrylized., this process has high rate of error chances during acrylization process. In this case report to minimise such inconvenience chair side pickup technique was used, this technique not only reduces the technique sensitivity of the conventional manner but also give us the liberty to not use the attachment if retention and stability in the final denture is sufficient.

Conclusion:

The case report discuses the management of a child with Ectodermal Dysplasia with only few teeth present and resorbed edentoulous ridges. The basis philosophy was to restore the esthetic and function of the child at present time, till the complete growth of the jaw has occurred. The final treatment will be implant supported prosthesis with bone augmentation as and when necessary. This is the first part of the longitudinal studies which are to be undertaken for this case. The final outcome from the treatment was satisfactory for patient, her family and us. On the subsequent follow ups she was boasted up with confidence, she was much comfortable now to socializes.

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