# Case Of Anodontia With Ectodermal Dysplasia: A Multidisciplinary Management

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**Abstracts:** Ectodermal dysplasia is a heterogeneous group of disorders characterized by a constellation of findings involving defects of two or more of the following: teeth, skin, and appendageal structures including hair, nails, and eccrine and sebaceous glands. Ectodermal dysplasia might be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes. Oral traits may express themselves as anodontia, hypodontia, and conical teeth. Anodontia also manifests itself by a lack of alveolar ridge development. In this case report; it is aimed to describe the prosthetic rehabilitation of the Hypohidrotic ectodermal dysplasia.[Patil R NJIRM 2015; 6(4):107-110]

**Key Words**: Anodontia, Ectodermal dysplasia, Prosthetic oral rehabilitation.

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**Introduction:** Ectodermal dysplasia is a heterogeneous group of disorders characterized by a constellation of findings involving defects of two or more of the following; teeth, skin, and appendageal structures including hair, nails, and eccrine and sebaceous glands[1]. Ectodermal dysplasia might be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes[2.] The earliest recorded cases of ectodermal dysplasia were described in 1792[3].Thurman published the first report of a patient with ED in 1848[4]. The term was not coined until 1929 by Weech[5]. Since then 100 types of ectodemial dysplasia syndromes have been identified. The clinical expression of ectodermal dysplasia varies, depending on the specific syndrome. The group was defined by Friere-Maia as expressing at least two of the following traits: trichodysplasia, abnormal dentition, onchodysplasia, and dyshidrosis[6]. The incidence of ectodermal dysplasia is estimated to be 0.7 to 1 per 100000 births [7]. Ectodermal dysplasia (ED) is a large and complex group of disorders defined by the abnormal development of two or more structures derived from the ectodermal layer.

The most frequently reported manifestation of ED is Hypohidrotic Ectodermal Dysplasia (HED), also termed Christ-Siemens-Touraine syndrome. The ectoderm, one of three germ layers present in the developing embryo, gives rise to the central nervous system, peripheral nervous system, sweat glands, hair, nails, and tooth enamel[8, 9]. As a result, HED patients exhibit the following clinical signs: hypotrichosis, hypohidrosis, and cranial abnormalities.

Patients often exhibit a smaller than normal face because of frontal bossing, a depressed nasal bridge, the absence of sweat glands resulting in very smooth, dry skin and/or hyperkeratosis of hands and feet. Oral traits may express themselves as anodontia, hypodontia, and conical teeth. Anodontia also manifests itself by a lack of alveolar ridge development[8, 10, 11]. Hypohydrotic type is inherited in an autosomal dominant pattern[12]. In general, the skin of affected children is lightly pigmented and appears thin and almost transparent; surface blood vessels are easily visible. Other manifestations include fine sparse hair, reduced density of eyebrow and eyelash hair. When hair is present, it may be fragile, dry, and generally with unruly appearance as a result of poorly developed or absent sebaceous glands. Fingernails and toe nails may also show faulty development and be small, thick or thin, brittle, discolored, cracked, and/or ridged. In this case report; it is aimed to describe the prosthetic rehabilitation of the Hypohidrotic Ectodermal Dysplasia.

**Case Report:** A 17 years old female patient visited to our unit with the complaint of missing teeth, inability to eat and difficulty in speech. The patient had history of dry or scaly skin since birth. Clinical examination of the patient revealed a fine, sparse

scalp hair, dry lips, microstomia everted lower lip, small face and smooth dry skin (Figure.1 and 2).

**Figure 1: Facial frontal view**

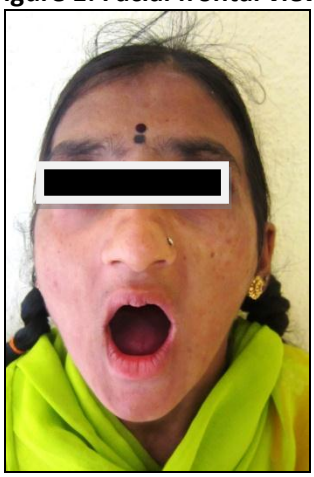
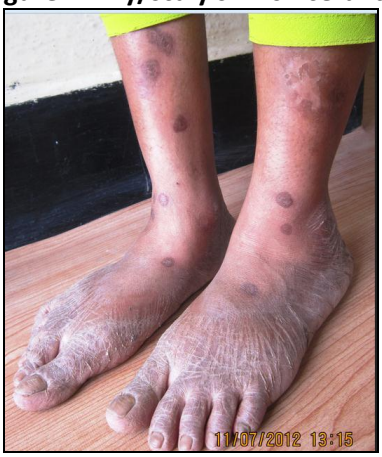
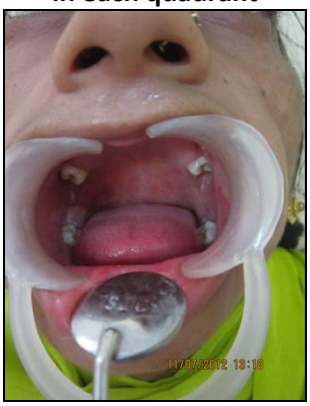


**Figure 2: Dry/scaly skin since birth**



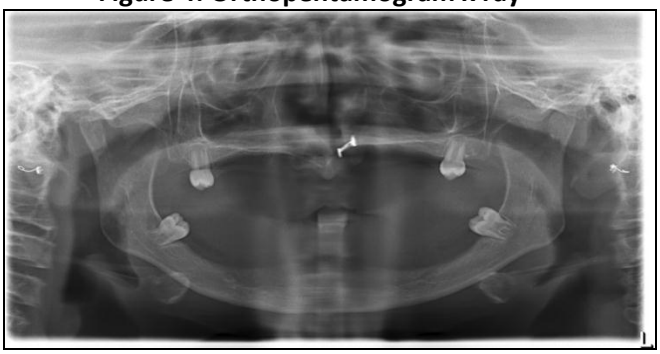

On oral examination patient presented with hypodontia, presence of only second molars in each quadrants, and resorbed ridges (Figure.3).

**Figure 3: Hypodontia showing only second molars in each quadrant**



Blood investigations revealed low hemoglobin level (Hb-07mg/dl) with Niacin deficiency for which she was consulted the physician and under medication. On radiographic examination orthopentamogram (OPG) showed that generalised bone loss of maxillary and mandibular arch and missing of all the teeth except second molars in each quadrant (Figure.4).

**Figure 4: Orthopentamogram x ray**



Case was planned for vestibuloplasty for resorbed ridges, root canal treatment for all remaining second molars and restoration with metal copings to fabricate the over dentures.

Procedure: Mouth preparations for the maxillary and mandibular arches were started with vestibuloplasty for the resorbed arches and intentional root canal treatment for the remaining second molars in all the quadrants. Root canal treated canals were prepared with peeso reamer, hot condenser was used to remove the guttapercha, the diameter of the canal was prepared till one third of the width of the tooth and the length two third was prepared, after that the tooth preparation was done, post core fabrication was done by direct technique using orthodontic wire points and applying the resin, resin pattern with post and core was casted, finished and polished. Cast metal copings were cemented to all the second molars with glass ionomer cement (Figure.5 and 6).

**Figure 5: Cast metal copings cementation in maxilla**

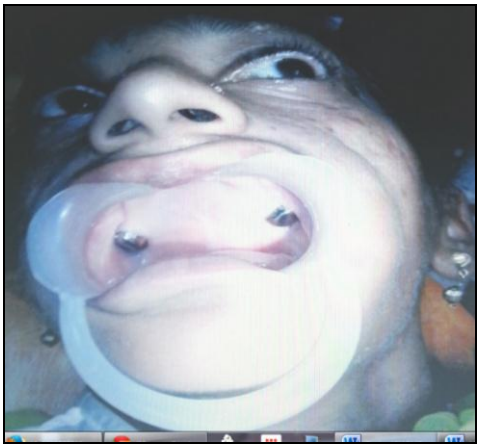


**Figure 6: Cast metal copings cementation in mandible**

Primary impression of maxillary and mandibular arch was done with impression compound, poured with plaster to obtain the primary cast; custom made tray was fabricated , border molding & secondary impression was made and poured with dental stone to obtain the secondary cast, denture base & occlusal rims were constructed and jaw relation was recorded, casts mounted on an articulator. Teeth arrangement was done. Try-in done, after final try-in the waxed dentures were processed in a heat polymerized denture base resin (Figure.7). The complete dentures were placed in oral cavity (Figure.8)

**Figure 7: Denture fabrication**

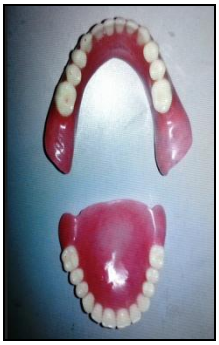
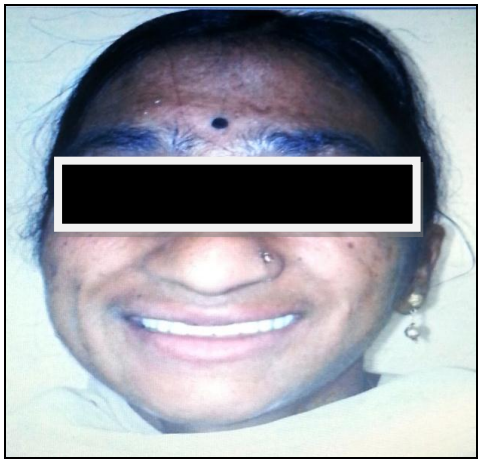


**Figure 8: Denture placement with complete oral rehabilitation**



**Discussion:**

The ectodermal dysplasias comprise a large, heterogenous group of inherited disorders that are defined by primary defects in the development of two or more tissues derived from the embryonic ectoderm. The condition is thought to occur in approximately one in every 100,000 birth[14].

Oral rehabilitation of the ectodermal dysplasia patient is necessary to improve both the sagittal and vertical skeletal relationship during craniofacial growth and development as well as to provide improvements in esthetics, speech, and masticatory efficiency[12].

Treatment generally includes a removable and or fixed partial denture, an overdenture, complete denture prosthesis or an implant retained prosthesis. In cases where there is associated cleft lip and palate the treatment may consist of intervention by a plastic surgeon and an oral and maxillofacial surgeon. In such cases a maxillofacial prosthesis may be indicated. In the present case prosthodontic management was done by over denture. Options which could be considered in our patient were of overdenture after intentional root canal treatment of the existing teeth. Overdenture indicated as it involves retaining the tooth or tooth root after intentional root canal treatment.

**Conclusion:** Dentists are often the first who diagnose these patients. Therefore, they should be aware of the clinical manifestations of this syndrome which helps in proper diagnosis, early interventions and appropriate therapies.

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