Ectodermal dysplasia is an hereditary condition associated with the defective development of tissues of ectodermal origin. This syndrome is classified as hidrotic, and anhidrotic. It manifests at least two of the following diagnostic features (1).

1. Hypotrichosis is characterised with fine, scanty, blond hair, eyelashes and eyebrows.
2. Dental anomalies (Hypodontia, oligodontia, and anodontia, “Both permanent and deciduous dentitions are affected”).
3. Hypohydrosis and anhydrosis are due to hypoplasia or absence of the sebaceous structures or decrease in the number or complete absence of sweat glands.
4. Onychodysplasia appearing as spoon-shaped, abnormal nails.
5. Depressed nasal bridge, frontal bossing, prominent ears, pouting everted lips (1-5).

Case Report

A 5-year-old Turkish male was seen at the Gülhane Military Medical Academy, Dental Sciences Center, on May 7, 1998 who exhibited the typical features of ectodermal dysplasia. He had fine, thin, blond hair, and scanty eyebrows and eyelashes. The skin of the entire body was dry. Although he did not have frontal bossing a saddle nose in profile, the vertical dimension was considerably lost. The lips were everted and the vermilion border was indistinct (Fig. 1).

Oral examination showed total anodontia in both maxillary and mandibular arches. The absence of teeth had allowed the tongue to spread. The buccal mucosa, palate and the floor of the mouth were normal. However, the alveolar ridges appeared to be considerably shallow (Fig. 2).
Radiographic evaluation revealed that only the germs of the upper right canine and lower left first molar were present (Fig. 3).

His parents seemed to be normal and they denied any history of this condition in their respective families.

The patient was treated with upper and lower complete dentures.

Primary and secondary impressions were taken with alginate because of its rapid-setting qualities, and pleasant smell and taste.

The impressions were boxed with strips of wax to preserve the periphery created by the muscles.

Occlusion rims constructed on the working casts were transferred to the mouth to establish the vertical dimension and centric relation. The vertical dimension was first determined by swallowing, checked by the rest position and associated free way space, and finally verified by means of phonetics.

The centric relation was registered utilizing occlusion rims with a check bite procedure. Face-bow mounting was used to relate the maxillary cast on the articulator relatively in the same position as the maxilla is to the natural condyles.

Plastic teeth with cuspal occlusal forms were used. Their inclines were reduced and central fossae enlarged both anteriorly and laterally to allow freedom in centric position. The teeth were set on the articulator to achieve balance in protrusive and lateral excursions (Fig. 4). The dentures, processed with a conventional type resin, were inserted and the necessary adjustments were carried out (Fig. 5). The patient was seen the next day.

Figure 2. Photographs show intraoral structures.
   a. Photograph of total anodontia of the patient.
   b. Prosthetic rehabilitation.

Figure 3. Radiograph shows germs of upper right canine and lower left first molar.
Figure 4. Photographs show waxing completed and ready for flasking.

Figure 5. Upper and lower total prosthesis.
   a. Occlusal view of the finished denture.
   b. Impression view of the finished denture.
Retention and stabilization of the dentures were clinically acceptable. Discomfort areas were relieved. He had no discomfort and seemed to be adapting well on the following weekly visits. The increased vertical dimension had established better muscle tonus and lip support. The patient was called for periodic check on a monthly basis.

Anodontia has many adverse effects on the psychological and physiological conditions of patients during childhood. Therefore, proper complete dentures must be applied. As a rule, the younger the child, the easier will be the adaptation to the denture. However treatment is completely dependent on patient-parent cooperation. As is known, there are many difficulties when treating young patients (1-3). One of these problems is the difficulty experienced during impression making. Patient cooperation can be achieved by the technique that Franks termed “operant conditioning”. During the first visit, the patient is given an upper stock tray to take home. The parent is instructed to supervise at home while the child inserts the tray into the mouth. This approach will minimize difficulty with impression making. Another problem can be frightening the child when recording centric relation, as something similar to a flame is introduced into the patient’s mouth. To avoid this problem, any adjustment of the occlusion rims should be done out of the child’s sight (4).

Mathewson and Primosch (1), reported that children with genetic diseases in which missing teeth are part of the syndrome often need complete dentures to restore primary and permanent dentitions.

The patient and parent should be instructed in the home care of prosthodontic appliances. They need to be informed of the need for periodic recall and refabrication of the dentures as growth occurs.

The instructions given to parents are reported by Ettinger and Pinkham (5). Parents should check the child’s mouth for supporting tissue irritation. The dentures should be removed for sporting activities, because children are unpredictable and a child with a denture may often abuse or break it. The child should be evaluated every three months for changes.

Bergendal et al. (6) reported that the development of techniques for osseointegrated implants offer new possibilities for the oral rehabilitation of children with ectodermal dysplasia. Kearns et al. (7) suggested that endoosseous implants can be successfully placed and can provide support for prosthetic restoration in patients with hereditary ectodermal dysplasia. However, vertical dentoalveolar growth results in the submergence of the implant relative to the adjacent natural dentition when implants are placed adjacent to erupting permanent teeth. Yuan et al. (8) also reported that these patients are provided with improved esthetics, restored chewing function, and space maintenance by the construction of a prosthetic restoration. However, with limited tooth support and an unusual occlusal pattern, it is difficult to obtain satisfactory retention and esthetics with traditional prosthetic techniques. Children suffering from ectodermal dysplasia often need extensive and complicated prosthetic treatment.

In our case, the patient who had ectodermal displasia with anodontia was treated with complete dentures. The psychological conditions of both the patient and the parents improved. Also, the facial profile as well as the function and esthetics of the patient improved. Notable improvements in the speech and nutritional status of the patient were obtained.

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References


