Ectodermal dysplasia: Dental management and benefits, a case report

**ABSTRACT**

**Aim** This case report describes a method of restoring function and aesthetics in a 9-year-old girl with ectodermal dysplasia with 15 years follow-up. Ectodermal dysplasia is both physically and emotionally devastating to patients. It is important that they are treated at an early age to help their social interaction. With proper dental intervention, the quality of life can be improved for patients with ectodermal dysplasia.

**Keywords:** Ectodermal dysplasia; Hypodontia; Hypohidrotic dysplasia; Partial denture; Fixed prosthesis.

**Introduction**

The earliest recorded cases of ectodermal dysplasia were described in 1792. Since then, 100 types of ectodermal dysplasia syndromes have been identified. Ectodermal dysplasia is a group of disorders defined by the abnormal development of two or more structures derived from the ectodermal layer. 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 enamel of the teeth [Larson, 1993; Neville, 1995; Perabo et al., 1956].

The most frequently reported manifestation of ectodermal dysplasia is hypohidrotic dysplasia, also termed Christ-Siemens-Touraine syndrome and anhydrotic dysplasia. Patients with this form of ectodermal dysplasia exhibit the following clinical traits: hypotrichosis, hypohidrosis, and cranial abnormalities. The face of these patients is often smaller because of frontal bossing and a depressed nasal bridge. Oral traits such as anodontia, hypodontia, and conical teeth can be detected; anodontia also manifests itself by a lack of alveolar ridge development.

Hypohidrotic dysplasia is usually diagnosed during infancy, following a bout of fever of unknown origin. These infants have an inability to regulate body temperature adequately because they have fewer sweat glands. The absence of these sweat glands results in very smooth, dry skin. Linear wrinkles are usually present around the eyes and mouth. The skin of the hands and feet often exhibits hyperkeratosis.

The clinical expression of ectodermal dysplasia varies, depending on the specific syndrome. The group was defined by Friere-Maia [1977] as expressing at least two of the following traits: trichodysplasia, abnormal dentition, onchodysplasia, and dyshidrosis. The disease may be inherited by autosomal-dominant, autosomal-recessive, or X-linked genetic transmission. These disorders, relatively rare, occur in 1 in 10,000 to 1 in 100,000 births [Neville, 1995; Megarbane et al., 1998].

The clinical manifestations of ectodermal dysplasia cause considerable social problems. Prosthodontic treatment of the clinical traits of ectodermal dysplasia can have a deep impact on these patients: the possibility to look and feel like their peers is paramount for their psychological development. The literature has demonstrated the benefits that corrective prosthodontics has for the self-esteem and social well-being of these patients [Roff and Wirt, 1984].

**Case report**

A 9-year-old girl was referred for management of the oral manifestations of her ectodermal dysplasia (Fig. 1). After her medical history was discussed, a complete examination was performed, including panoramic radiograph (Fig. 2), diagnostic casts, and clinical examination. A treatment plan consisting of periodontal therapy, caries management, extraction of non-restorable primary teeth, restoration with preformed paediatric crowns (Fig. 3) and fabrication of a removable denture was discussed with the patient's parents, who showed some hesitation concerning the removable denture. We modified the treatment plan to fit the patient demand in order to give her a better quality of life, convinced that a fixed solution will be a big plus, helping the patient psychologically and socially.

In the mandible the treatment consisted in a transitional first phase that combined a fixed restoration in the anterior region consisting of a temporary six elements bridge from cuspid to cuspid fixed on teeth 32, 33, 42 and 43 with two pontics in the edentulous region of 31 and 41, with a removable denture in the posterior edentulous region waiting for the eruption of the molars to be completed in order to be able to proceed with the second phase that consisted of a fixed full-arch bridge of 12 elements (Fig. 4). In the maxilla the axes of the teeth needed to be adjusted in order to be more aligned to receive a fixed full arch upper bridge. To preserve the dental substance and avoid removing too many teeth, the alignment was made using slight orthodontic movements (Fig. 4).

After eruption of the molars was complete, the lower partial denture and the provisional resin bridge were replaced by a fixed metal ceramic bridge of twelve...
elements fixed on teeth 36, 33, 32, 42, 43 and 46 with six pontics filling the edentulous region of teeth 35, 34, 31, 41, 44 and 45.

After teeth alignment in the maxilla, a conservative preparation of the upper teeth was made in order to receive the fixed ceramic to metal bridges fixed on teeth 17, 16, 14, 13, 11, 21, 23, 24 and 27 with five pontics filling the edentulous region of teeth 15, 12, 22, 25 and 26 (Fig. 5).

Since a common oral manifestation of ectodermal dysplasia is the loss of occlusial vertical dimension, a correct vertical dimension is paramount to make enough space for the teeth arrangement. The patient’s vertical dimension of occlusion was established by assessing phonetic criteria and sibilant sounds. Preliminary occlusal relations were recorded, and then the bridges were fabricated.

After the controls and the occlusal adjustments, the ceramic to metal bridges were fixed on the teeth with a polycarboxylate cement (Fig. 6, 7, 8).
Discussion

In this case the patient was considered mentally retarded as she did not communicate properly with her family and friends. However, during and after the dental treatment, clear psychological and social progress were noticed. The positive changes in her behavior were profoundly appreciated by her family and friends. Note the tattoo at the eyebrows region (Fig. 4 and 7), reflecting her gain of self-esteem. It seems that gaining self-confidence after dental rehabilitation contributed tremendously to the development of this patient.

This case report describes a method for fixed prosthetic treatment of patients with ectodermal dysplasia. Excellent oral hygiene is crucial to the successful treatment of these patients.

The patient should use topical fluoride daily for prophylaxis against caries during the treatment.

Other treatment options were described in the literature, consisting of a removable prosthesis, has shown also excellent results when the patient maintains proper oral hygiene [Brewer and Morrow, 1975; Abadi and Herren, 2001]. Nevertheless a fixed solution is closer to the normality and allows the young patient affected by ectodermal dysplasia to integrate more easily into the society.

Conclusion

Dental management using complete fixed prosthesis for a patient with ectodermal dysplasia has been outlined. Dental restoration aids the patient in developing proper speech, deglutition, and mastication, and may have dramatic social and psychological benefits for these patients. It seems that gaining self-confidence after dental rehabilitation at an early age highly affects the development of these patients.

References