

Ali Rıza ALPÖZ

Hydrotic Ectodermal Dysplasia (Associated With Numerous Enclused Permanent Teeth Including a Primary Molar)

Department of Pedodontics,
Faculty of Dentistry, Ege University,
Bornova 35100 Izmir-Turkey

Received: March 13, 1998

Key Words: Hydrotic ectodermal dysplasia.

Ectodermal Dysplasia (ED) is a hereditary congenital disease that effects several ectodermal structures. Although the existence of an autosomal recessive inheritance is reported , the disease is usually transmitted as an X - linked recessive trait (1-4). ED patients require dental treatment primarily because of other dental needs and not always for good appearance. Restoration of good appearance is merely one of the outcomes of a succesful treatment. Ectodermal dysplasia is characterized by the clinical manifestations of hypodontia, hypohidrosis and hypotrichosis (1). The pathognomy of the face in ED individuals of both sexes is similar and characteristic (1,3).

The purpose of this article is to present a female patient with ED associated with numerous enclused permanent teeth including a primary molar.

A female, 12 year - old patient was attended to the clinic of the department of Pedodontics, Ege University with her parents because of the eruption delay and dental caries problems. On extraoral physical examination wet face appearance was found as the main finding (fig.1). Oral and radiographic examination showed mixed dentition with unerupted numerous teeth. (fig.2). She had no systemic disorders and also no medicine intake regularly. No contributory family history and similar dental manifestations were found from her parents,



Figure 1. View of the female patient with hydrotic ectodermal dysplasia.



Figure 2. Ortopantomograph film shows the unerupted permanent teeth and the enclosed primary molar.

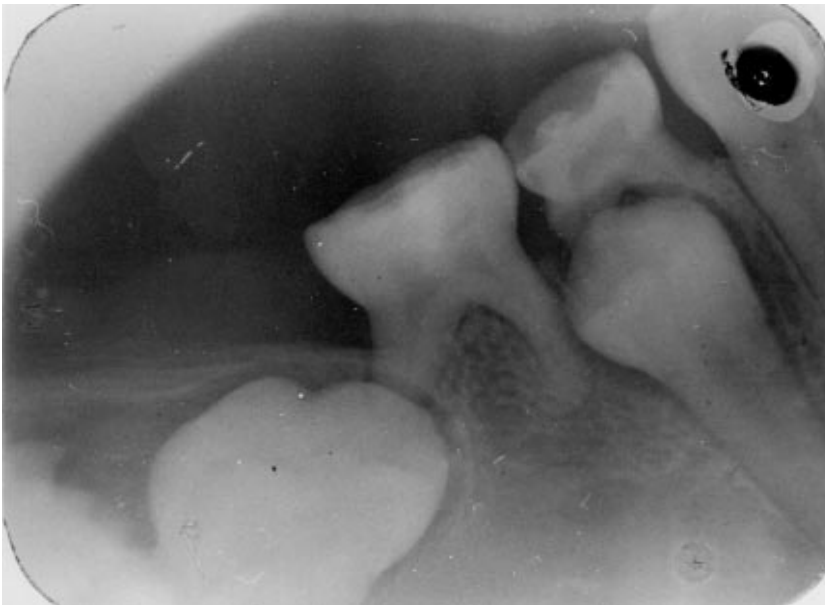


Figure 3. Periapical film shows the congenitally lost lower right permanent second premolar (45).

older and younger sisters. Radiographic examination showed that only lower right permanent second premolar was congenitally lost (fig 3) , while the other teeth were present as erupted or unerupted. Lower left second primary molar (75) was found unerupted and to be closely depressed together with second permanent premolar (35) and first permanent molar (36) (fig.2). Consultation was established with the department of Pediatric Genetics of the Medicine Faculty, Ege University for the diagnosis of the case. The case was described as Autosomal Recessive, Hydrotic Ectodermal Dysplasia by the Medicine faculty, Ege University.

Treatment Planning:

After the permission was received from the parents for the presentation of the child, full history with

clinical and radiological examination were obtained and primary carious molar teeth were treated with glass ionomer restorations. It was decided to operate the enclosed primary molar (75) in the department of oral surgery , faculty of dentistry and follow the eruption status with three months regularly. It was also decided to prepare upper and lower partial dentures for stimulation of the permanent teeth and to restore the anterior esthetic appearance . The patient was operated approximately 20 days after the initial visit. Eruption direction of 35 and 36 was seen transversally 40 days after the operation (fig.4). Healthy dental follicular structure was found in the soft tissue around extracted primary molar in the laboratory of oral pathology , faculty of dentistry. Alginate impressions were received

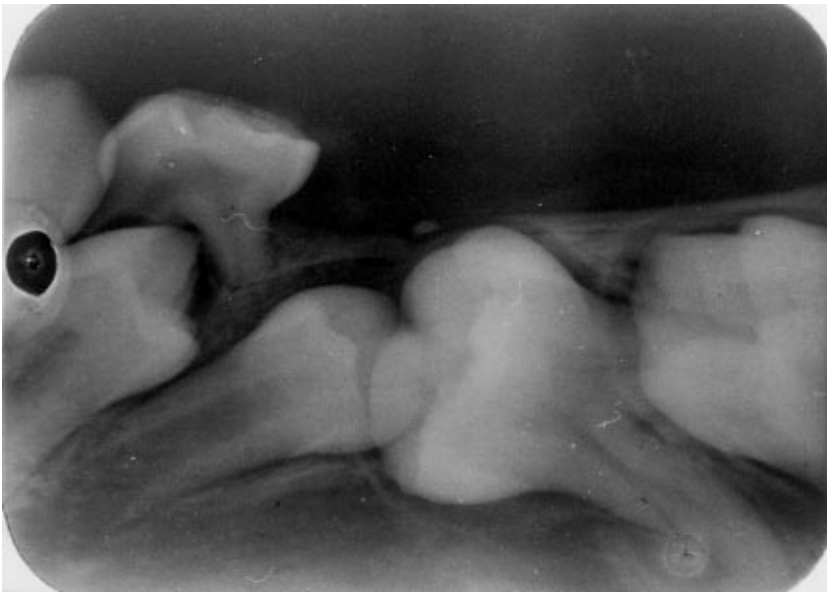


Figure 4. Eruption direction of the lower enclosed left permanent premolar (35) and the first molar (36) after the operation.

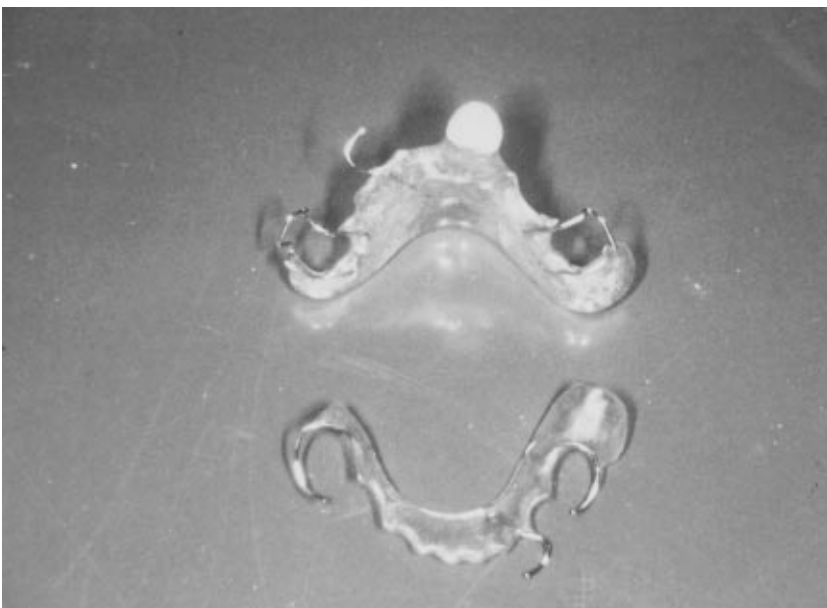


Figure 5. Upper and lower partial dentures prepared for the correction of the esthetic appearance and the stimulation of the eruption.

and partial dentures were prepared (fig.5) and placed into the mouth (figs.6 and 7). Since no change at the eruption direction was observed three months after the operation (Fig.8), it was decided to apply orthodontic treatment for the eruption of the enclosed teeth numbered 21, 35 and 36 by using orthodontic brackets surgically, combined with removable orthodontic appliances.

Dental findings of Ectodermal Dysplasia are well known and optimal treatment for children with ED

requires the multidisciplinary efforts of pediatric dentists (1,4,5). Usually the initial diagnosis should be made by the pediatrician when the patients complains from unexplained episodes of fever or intolerance to heat and lacks of teeth particularly in the patients with hypohydrotic ectodermal dysplasia (1). It has been found that during the primary dentition phase many abnormalities of the primary teeth could be identified as congenitally missing or misshapen primary teeth, taurodontism and diastema (1,5,6). While no taurodont



Figure 6. Intraoral view of the patient without the partial dentures.



Figure 7. Intraoral view of the patient with partial dentures.

and misshaped teeth were found, only one congenitally missed permanent premolar (45) and numerous enclused permanent teeth were found in our female case (Figs.1 and 2). Typical dental findings for ED such as conical incisors and congenitally lost anterior teeth were also not found. Treatment of a child with ectodermal dysplasia requires knowledge of growth and development, behavioral management, the fabrication of a prosthesis, the modification of existing teeth with resin composites, the motivation of the patients and parent in the use of the prosthesis, and long term follow - up for the modification and /or replacement of the prosthesis. During the mixed dentition stage, the prosthesis will need

to be modified to accommodate the loss of exfoliated primary teeth and the appearance of newly erupted permanent teeth. When the patient is in the permanent - dentition stage, the removable prosthesis may be replaced by a fixed restoration, depending on the number and position of the remaining permanent teeth (1). Children effected with hydrotic ectodermal dysplasia usually show less typical dental findings (3,4). In conclusion , the initial diagnosis should be made as soon as possible and the optimal treatment for children with ectodermal dysplasia should require the multidisciplinary collaborative efforts of pediatric professionals.

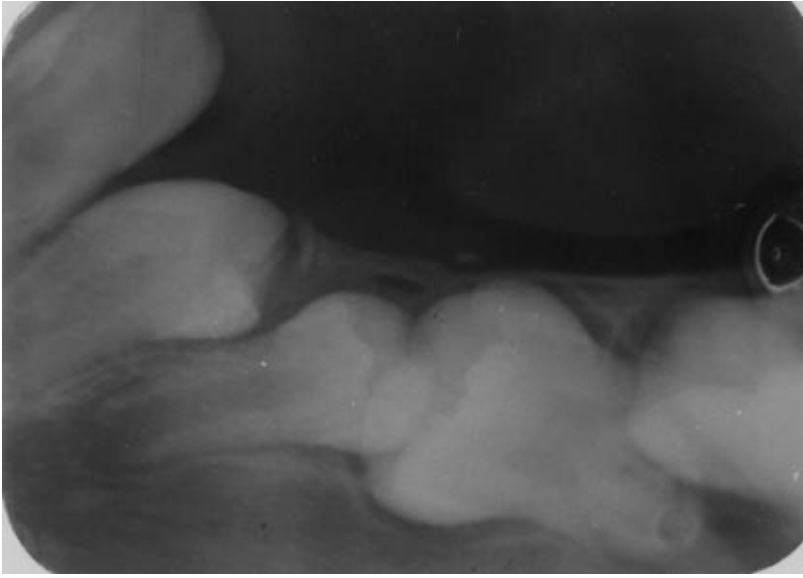


Figure 8. Control film three months after the operation shows no difference on the eruption direction.

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