

Radiographic study of patients with ectodermal dysplasia and partial anodontia

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ABSTRACT

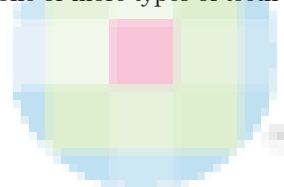
Aim: To investigate the association between gender and type of cleft with hypodontia and to verify if the presence of the cleft interferes with hypodontia of one or more type of teeth in patients with ectodermal dysplasia attending the Hospital for Rehabilitation of Craniofacial Anomalies (HRCA), University of São Paulo (USP).

Materials and Methods: Panoramic radiographs of 54 patients of both genders (29 males, 25 females), from the files of the oral radiology sector of HRCA/USP, presenting with ectodermal dysplasia and cleft lip and/or palate were evaluated by radiographic observation of hypodontia of one or more types of teeth.

Statistical Analysis: We performed descriptive statistics and statistical analysis by Fisher test.

Results: Hypodontia was observed in 50% of females and 50.88% of males; 50% for cleft palate and 50.59% for complete cleft lip and palate. Only 22.22% of patients with cleft palate and 6.67% with complete cleft lip and palate presented with hypodontia of one type of teeth, whereas 77.78% of cases with cleft palate and 93.33% with complete cleft lip and palate displayed hypodontia of more than one type of teeth.

Conclusion: Based on the present methodology, there were no statistically significant differences in hypodontia of one or more types of teeth between genders or types of cleft.



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Ectodermal dysplasia is a hereditary condition in which most frequent genetic inheritance is sex-linked recessive. It consists of genetic and environmental alterations characterized by absence, delayed or incomplete development of structures of ectodermal origin as hair, nails, teeth, and skin.^[1-5] It is more frequently observed in white individuals and male sex.^[2,6,7]

The frequency of ectodermal dysplasia has been estimated at 1:100,000 births.^[5,8-11] The two classic forms of this syndrome are anhidrotic/hypohidrotic and hidrotic. The characteristics of hypohidrotic ectodermal dysplasia (HED) include the triad hypodontia, hypotrichosis, and hypohidrosis.^[1,4,9,11,12] Patients with the hidrotic form present dystrophic nails, scarce hair, and tooth abnormalities, without involvement of sweat and sebaceous glands.^[12,13]

Cleft lip and/or palate are a congenital malformation caused by deficiency or lack of fusion of maxillary, mandibular, or frontonasal facial processes. It is highlighted by the prevalence of 1:600 to 1:800 births, depending on the population, with esthetic and functional implications. According to Larson *et al.*,^[14] there is a strong association between occurrence of cleft and partial anodontia (especially of maxillary incisors and premolars), as confirmed by recent histological and genetic examinations.^[15]

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Partial anodontia may be divided into hypodontia and oligodontia. In the former, there is anodontia of one or few teeth, commonly affecting the maxillary lateral incisors, maxillary and mandibular second premolars.^[16] Hypodontia is a frequent signal of ectodermal dysplasia in 80% of cases.^[6,17] Oligodontia is a rare alteration, with prevalence of 0.3% in permanent teeth, and refers to anodontia of six or more teeth.^[18]

The correct choice of the best form of treatment for patients with HED and partial anodontia (hypodontia or oligodontia) is very important in order to stand out the significance of early diagnosis and intervention. During the examination, the patient receive education and preventive procedure, such as instruction of oral hygiene and feed diet, control of bacterial plaque, removal of oral or deleterious habits, and procedures of oral rehabilitation.^[4,13]

Hypodontia of primary and permanent dentition is one of the most frequently occurring oral symptoms in patients with HED. These features of poor esthetic affect the social and the psychological well-being of the patient. A prosthetic rehabilitation of a HED patient is usually the treatment chosen (prosthodontic and restorative), which improved esthetic and oral functions, so as to improve psychological and social comfort to the child, who will remain under dentistry treatment until it is completely grown-up and orally developed.^[3,11,17,19]

The types of teeth most frequently affected by hypodontia in patients with ectodermal dysplasia and cleft are the maxillary second premolars and lateral incisors.^[14] According to Becktor *et al.*,^[8] the types of teeth usually absent are the maxillary lateral incisors, second molars and premolars, mandibular incisors, second molars and premolars. Balshi and Wolfinger^[19] reported that mandibular anterior teeth are usually absent in patients with ectodermal dysplasia.

Ruhin *et al.*^[20] described, in a study of cases with ectodermal dysplasia and cleft, several tooth abnormalities as impaction, microdontia, peg-shaped teeth, taurodontism, hypoplastic enamel, besides absence of maxillary and mandibular premolars.^[12,13] A case of a female patient with maxillary oligodontia was reported, who presented with only peg-shaped incisors and canines in the maxillary arch^[7,21] and total anodontia in the mandibular arch.^[22]

Knowledge on the pattern of hypodontia in individuals with ectodermal dysplasia and cleft lip and palate may be helpful for the establishment of adequate treatment protocols for these patients. Thus, the present descriptive study radiographically investigated the possible association of hypodontia with sex or type of cleft in patients with ectodermal dysplasia and cleft attending the Hospital for Rehabilitation of Craniofacial Anomalies of University of São Paulo (HRCA/USP).

MATERIALS AND METHODS

This project was developed in accordance with the Research Ethics Code (approved under n. 221/2004-UEP/CEP).

The sample consisted of panoramic radiographs of 54 Caucasian patients with ectodermal dysplasia, aged 8 to 37 years, from the files of the Oral Radiology sector of Hospital for Rehabilitation of Craniofacial Anomalies of University of São Paulo (HRCA/USP). The type of cleft presented by the patients was assessed on the patients' records at treatment onset; among the 54 patients, 43 presented with complete cleft lip and palate (31 bilateral and 12 unilateral), nine with isolated cleft palate, and two with isolated cleft lip. There were 25 females and 29 males in the sample.

The radiographs were assessed by a single examiner, in a dark room on a film viewer with the aid of a magnifying glass, at two distinct moments with 21-day interval to evaluate the presence of partial anodontia (hypodontia and oligodontia). Radiographic findings and clinical information (sex, age, registry number, absent teeth, and type of cleft) were recorded on standardized forms.

All permanent teeth were observed in radiographic examination, except for the third molars, because these teeth are extremely variable. The records of 90 patients with ectodermal dysplasia were screened at HRCA/USP. However, panoramic radiographs were available only for 54 patients. Sixteen patients are still under 8 years of age, which is the age that the first panoramic radiograph is obtained according to the institution protocol. Radiographs were not available for another 20 patients for other reasons, such as death, drop-out, or lack of previous attendance at the Oral Radiology sector.

Statistical analysis was performed by the Fisher's Chi-square test in order to take a comparison between two or more samples, which in this study are: Gender and dental absence; fissure type and dental absence; fissure type; and absence of one or more dental groups. The frequency of hypodontia was compared between sex and among types of cleft; the absence of one or more groups of teeth was also compared among types of cleft.

RESULTS

There were no discrepancies between evaluations of any patient at the different moments; therefore, no further statistical analysis for calculation of intra-examiner variability was performed.

Fifty-four cases (29 males and 25 females, aged 8-37 years) had tooth agenesis (all cases: From hypodontia to anodontia), associated with cutaneous dyshidrosis, sparse or absent hair,

nail dystrophy, and hypohidrosis. The skin on most of the body was abnormally thin, dry, and soft with an abnormal lack of pigmentation [Table 1].

The prevalence of partial anodontia was 50% for the female sex and 50.88% for the male, with no statistically significant difference between sex ($P > 0.05$) [Figure 1].

Concerning the type of cleft, 55.55% of patients with cleft palate presented partial anodontia compared to 50.59% of patients with complete cleft lip and palate, without statistical difference in hypodontia between the types of cleft ($P > 0.05$) [Figure 2].

Among patients with cleft palate, 22.22% exhibited hypodontia of one type of teeth and 77.78% of more than one type of teeth, compared to 6.67% and 93.33% for patients with complete cleft lip and palate, respectively. Thus, there was higher occurrence of hypodontia of more than one type of teeth, though without statistically significant difference among types of cleft [Figure 3].

Patients with HED usually present more cases of hypodontia and other dental abnormalities (such as retention of deciduous teeth) than the patients with hidrotic ectodermal dysplasia. However, in this present study, there was no correlation and significant difference (clinical and radiographic) between the type of ectodermal dysplasia and the occurrence of anodontia and dental abnormalities ($P > 0.05$).

DISCUSSION

Most patients in the present study were males and presented HED, which usually is the most severe type as to the occurrence of dental abnormalities. However, there are some cases of patients with hidrotic ectodermal dysplasia and partial anodontia or other dental abnormalities.^[12,13]

The findings of the present study revealed that patients with ectodermal dysplasia and cleft also presented hypotrichosis and ectrodactyly [Figure 4]. Ectrodactyly is a remarkable feature; at least hypodontia and hypotrichosis should be present for diagnosis of the syndrome.^[4]

Analysis of patients with ectodermal dysplasia and cleft in this sample confirmed the higher occurrence of hypodontia of the maxillary lateral incisors and premolars and mandibular incisors and premolars [Figures 5 and 6], followed by molars and canines. This is in agreement with other studies which also demonstrated that these are

the most affected teeth, yet not necessarily in the same order.^[14,16]

In the present study, retention of deciduous teeth was also observed, including maxillary and mandibular molars and mandibular canines and incisors [Figure 7]. Radiographic analysis of the subjects in this study also confirmed the presence of dental anomalies of shape, including peg-shaped incisors and canines [Figure 8], besides teeth with enlarged pulp. Cases of oligodontia in which only peg-shaped incisors and canines were present were also observed,

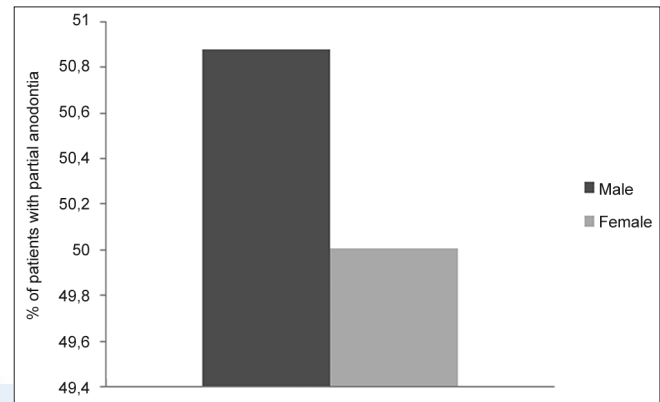


Figure 1: Influence from sex on hypodontia

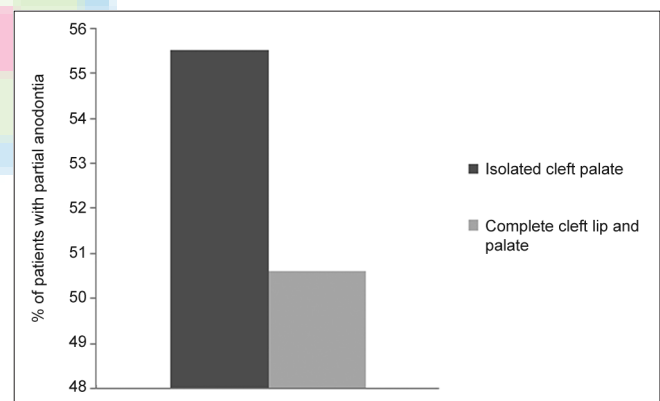


Figure 2: Influence from type of cleft on hypodontia

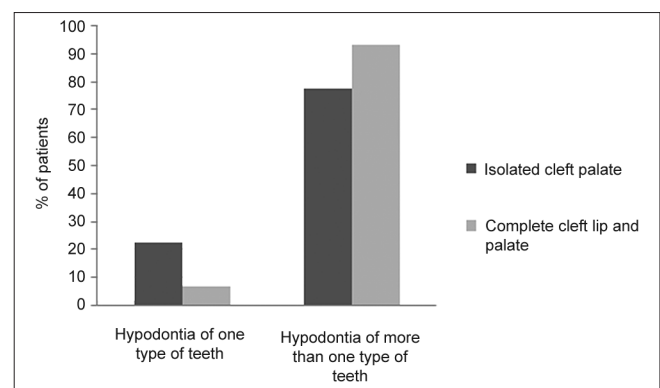


Figure 3: Influence from type of cleft on hypodontia of one or more type of teeth

Table 1: Report of clinical characteristics for each of the 54 cases

Abnormality	54 cases (100%)
Complete cleft lip and palate bilateral	31 cases (57.4%)
Complete cleft lip and palate unilateral	12 cases (22.2%)
Isolated cleft palate	9 cases (16.6%)



Figure 4: One case of ectrodactyly in patient with ectodermal dysplasia



Figure 5: Panoramic radiograph exhibiting hypodontia of maxillary lateral incisors and premolars and mandibular incisors and premolars



Figure 6: Clinical absence of maxillary lateral incisors and mandibular incisors



Figure 7: Panoramic radiograph exhibiting retention of deciduous teeth and hypodontia of permanent teeth

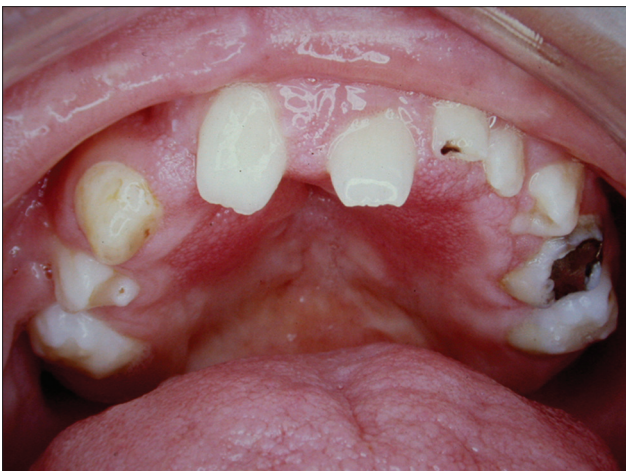


Figure 8: Clinical aspect of absence of maxillary lateral incisors and peg-shaped incisors and canines

as well as complete anodontia of mandibular teeth.^[23] This is in agreement with other studies which stated that patients with ectodermal dysplasia may present oligodontia, delayed tooth eruption, taurodontism, and retention of

deciduous teeth.^[9,12,20] Depending on the severity of the condition, various prosthodontic treatments are available to improve appearance, mastication, and speech.^[2] The prosthetic dental treatment in patients with anodontia or hypodontia is important for physiological and psychosocial reasons. Treatment of the HED patient generally includes a removable and/or fixed partial denture, a complete denture prosthesis (overlying affected teeth when the vertical dimension of occlusion permits), and an implant-retained prosthesis when indicated.^[12,21,23-25] This treatment approaches can be used individually or in combination to provide an optimal result. The proper sequencing of treatment is important to achieve the desired function and esthetic results. Because of early-age intervention and the need to easily modify the intraoral prosthesis during rapid growth periods, a removable partial denture or complete denture prosthesis is indicated initially.^[2] In the present study, only one male patient in the present sample presented ectodermal dysplasia, complete bilateral cleft lip and palate and total anodontia, which assigned a senile facial aspect

to the patient. A total prosthetic rehabilitation was the treatment chosen in order to improve esthetics, function, and physiologic condition of this patient. A multidisciplinary team approach is recommended for successful treatment of affected individuals. The team should include at least a dentist, a prosthodontist, orthodontist, and an oral and maxillofacial surgeon.^[2,26]

CONCLUSION

Based on the present methodology, it could be concluded that there appears to be no statistically significant association between hypodontia and sex or type of cleft ($P > 0.05$). There was higher occurrence of hypodontia of more than one type of teeth, yet without significant difference between types of cleft as to this aspect. The types of teeth most frequently absent were incisors and premolars, followed by molars and canines. There is no statistically significant difference between the type of ectodermal dysplasia and the occurrence of dental abnormalities.

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