

Alternative rehabilitation treatment for a patient with ectodermal dysplasia

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The ectodermal dysplasia constitutes a group of hereditary disorders whose clinical manifestations can be defects in ectodermal structures. The hypohidrotic and anhidrotic are commonly types of ectodermal dysplasia. The main characteristics are dental anomalies, hypotrichosis and hypohidrosis. The oral rehabilitation of this patients is important for better social living, self esteem and oral function. This paper had as objective to relate and discuss a case of anhidrotic ectodermal dysplasia, describing the positive influence of an alternative rehabilitation treatment.

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INTRODUCTION

Ectodermal dysplasia is part of a group of hereditary diseases that affects the development of two or more ectodermal derived structures, for instance, hair, fingernails, teeth and skin.¹

Ectodermal dysplasia is a relatively rare disorder having a related cases variation in frequency of 1:10,000 up to 1:100,000 of those born alive.² It is more frequent in boys and in most cases it is of recessive autosomal character,^{3,8} but it could also be dominant autosomal or linked to chromosome X.² It is usually diagnosed in childhood, subsequent to a feverish episode of unknown origin.^{2,4,5,8}

The types of dysplasia commonly observed are the hypohidrotic and anhidrotic. The first has as principal characteristic scarce hair, dystrophic fingernails and dental anomalies, not presenting involvement of the sudoriparous and sebaceous glands.^{1,5,9,10} The anhidrotic

type shows itself fundamentally through hypohidrotic, hypotrichosis, hypodontia^{1,5} and cranial anomalies.²

The patients with anhidrotic ectodermal dysplasia present with: face often smaller due to the prominent front swellings, in cell nasal bridge, soft skin, flat and dry, partial or total absence of the sudoriparous glands, not supporting temperatures discharges. The hair and eyebrow are fine, scarce and blond.⁵

Regarding the oral findings, in the deciduous or permanent teeth, the presence of anodontia or hypodontia is common as are bad dental formations, such as conoid teeth. The bone growth usually continues except for the alveolar process, due to the multiple dental absences, so reducing the vertical dimension and, as consequence, resulting in protuberant lips, dry and split, due to the hypohypohidrotic,^{6,10} besides the facial aspect of senility, because of the decrease of the vertical dimension.⁵ The buccal mucous membrane presents itself dry and there is a decrease of saliva due to the reduction of the number of salivary glands.¹

Ectodermal dysplasia causes a physical and emotional problem for the patient. Oral rehabilitation is one of the possible means to improve the quality of life.² In this way, the objective of the present article was to describe a case of an anhidrotic type of ectodermal dysplasia, with the approach of the influence of an esthetic rehabilitation and functional alternative in the improvement of the quality of life.

CASE REPORT

A female patient, 5 years old, was sent to the pediatric dental clinic of a public university of Rio de Janeiro (RJ), Brazil, accompanied by her mother, with her main complaint dental absences and presence of "small" teeth.

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Figure 1. Patient, 5 years old, with anhidrotic ectodermal dysplasia (prominent front swellings; in cell nasal bridge; hair and brow slender and brittle; salient eyebrow area; soft, flat and dry skin; protuberant lips).

During the anamnesis, no alteration in the general health of the patient was identified, however, in the clinical exam ectodermal dysplasia was suspected. The mother had a problem-free pregnancy and childbirth was cesarean. At the age of 2 months the child presented repeated feverish episodes due to throat problems. The child possessed normal mental clinical development and did not have any deleterious buccal habit.

In the clinical extra-buccal exam, voluminous and protuberant lips can be observed, associated with delicate fissures on the mouth and eyes. The nose and the nasal bridge were in unison, and there was frontal swelling. The ears were well formed and without oblique implantation. The hair and the brow, though not scarce, were fine and brittle; the eyebrow area was salient (Figure 1). The patient presented dry skin and the fingernails, which possess any dystrophic alteration.

In the clinical intra-oral exam (Figures 2a and 2b), the presence of normal gums was verified. Agenesis of teeth 53, 63, 74, 81 and 84 was observed. The agenesis was confirmed through the radiographic panoramic (Figure 3), where the absence of teeth 11, 12, 13, 21, 22, 25, 35, 41, 44, could also be identified, causing loss of vertical dimension, as well as the presence of conoid teeth (51, 61 and 71). The patient still presented partial ankyloglossis (Figure 2b). The oral hygiene was satisfactory, the patient did not have active carious lesions, gums and periodontic tissues were healthy (Figures 2a and 2b). A diet diary was requested and it was verified that the patient only consumed liquids and soft foods.

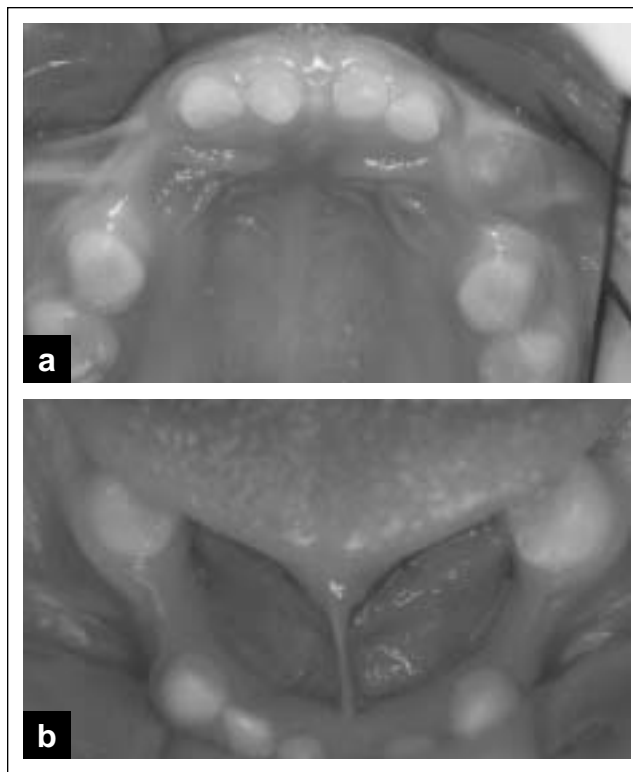


Figure 2. A clinical view of the upper arch (2a) and lower arch (2b) notice dental absences and conoid teeth.

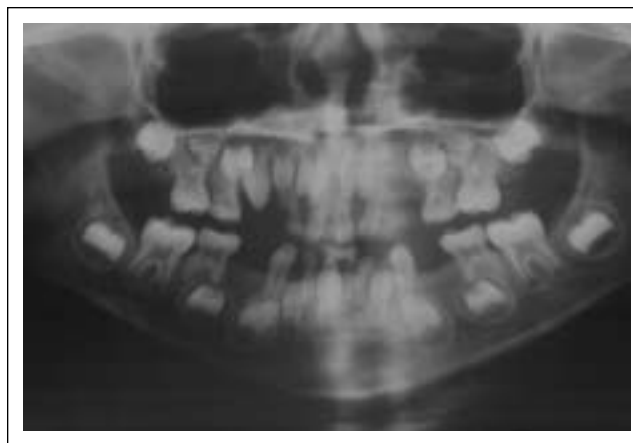


Figure 3. Panoramic radiograph showing the dental agenesis.

In the phonetic evaluation, it was observed that the child presented difficulty in speech and voice alteration (hoarseness), hypertrophy of the pharyngeus membrane being identified.

Until the age of 5 the condition had not been diagnosed. The hypothesis of the occurrence of ectodermal dysplasia was raised by the pediatrician in the first consult. In order to verify the suggested hypothesis, the patient was directed to the Stomatology Department of the same institution for confirm the anomaly, where the diagnosis of anhidrotic ectodermal dysplasia was established.

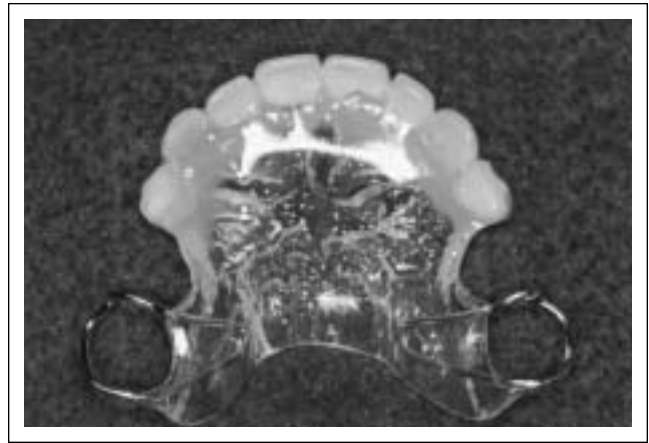
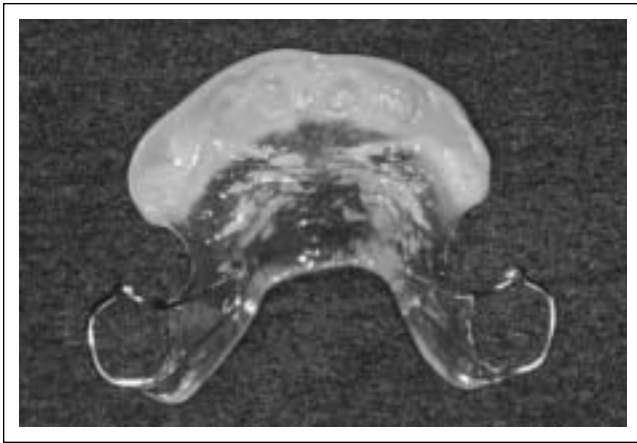


Figure 4. A view of the retention area (4a) where conoid teeth have arisen (encapsulated prosthesis). The occlusal side of the prosthesis (4b).



Figure 5. The lower arch with the removable prosthesis.



Figure 6. A frontal view of the patient occlusion.

Initially, as plan of treatment, it was decided that she should have a lingual frenectomy under local anesthesia and voice control. The alternative prosthetic treatment was planned with a removable prosthesis for the high arch, in which the conoid teeth served as retention for the prosthesis (Figures 4a and 4b), and a removable prosthesis with staples for the low arch (Figures 5 and 6). The teeth were preserved without need for restoration and extraction. These prosthesis need to be changed periodically according to the growth and the bone development. The healthy clinical evaluations of the patient are to be done quarterly until definitive oral rehabilitation is done.

The child was guided properly to perform an efficient hygiene of the prosthetic appliances. She was cooperative and satisfied with her aesthetics.

After the placement of the prosthesis, the patient presented diet alteration starting to consume solid feedings, and, still, a positive feedback regarding psycho-social living, thus improving the quality of life. The phonetic treatment contributed in a significant way at school, positively intervening in her relationship with

other children, and the problem of hoarseness was extinguished.

DISCUSSION

Ectodermal dysplasia is commonly diagnosed in childhood, subsequent to a feverish episode of unknown origin,^{4,5,8} however, in the present report, the condition had not been diagnosed until the age of 5. In the present report, in the clinical exam, ectodermal dysplasia was suspected, due to the appearance of the child and the common characteristics of the syndrome, this being described as the first verified case in the family, although the frequency of ectodermal dysplasia increases when there is a family history.²⁻⁸

It is common for the patients to say that the pediatricians do not diagnose the symptoms of the syndrome in the first years of life, in many cases the diagnosis has been made by a pediatric dentist,¹¹ as in the case here presented. The diagnosis of ectodermal dysplasia and consequent classification is extremely easy when the patient presents with more characteristics and clear clinical forms. In the present case report when these

characteristics are less evident, the diagnosis and the classification of the patient, became a difficult task for the physician.¹²

Absence of mucous glands in the breathing tract can lead to dysphonia due to the atrophied pharynxes or larynxes membrane.⁸ It can result in laryngitis and pharyngitis.^{5,9,10} Neville⁵ also related that the patients came with hoarse voices. In the present report the patient had difficulty when speaking and hoarseness, hypertrophy of the pharyngeus membrane being identified through phonetic evaluation. The phonetic problems were solved after phonetic therapy.

In ectodermal dysplasia there is no specific treatment to minimize the relative consequences for the anomalies of fingernails, skin and hair. Regarding the dental treatment instituted, it was possible to improve the physical condition of the child, as well as the esthetics; the chewing and phonetic functions. These previous conditions harmed the appropriate psycho-social development of the patient. Therefore, the dental treatment was done not only to re-establish the function, but to improve the emotional and social aspect of the child as well.

In patients with ectodermal dysplasia the reduction or absence of the dental sheet justifies the presence of conoid teeth and/or anodontia.¹³ In these cases, it is necessary the use of prosthetic appliances to the reestablishment of aesthetics and function. For the rehabilitation of the oral health of the child, there are several suitable possibilities that also contribute to the increase of vertical dimension. In cases of oligodontia, the election treatment constitutes the placement of partial prosthesis, being proceeded by the clinical and radiographic periodic control due substitutions in function of maxillofacial growth.¹ In the case here presented, to accomplish the alternative rehabilitation treatment, the conoid teeth served as support aided as factors of retention of the prosthesis. Due to the good oral hygiene of the patient, the use of conoid teeth as support and retention was opted.

The child was instructed on how to clean the prosthesis, since her oral hygiene was satisfactory. The patient has been monitored quarterly for the past 15 months since the placement of the prosthesis, in case there is a need adjust these appliances.

The removal of the conoid teeth for the subsequent placement of a prosthetic appliance would further reduce the alveolar edge of the patient.² Therefore, the maintenance of these teeth would promote a larger distribution of the chewing forces and a larger retention of the prosthesis. The teeth were preserved without need for restoration. The child showed great cooperation with the use of braces and was satisfied with the esthetics.

Ekstrand and Thompsom,¹⁴ recommend the placement of implants in the adult phase for the treatment of patient with ectodermal dysplasia, although Guckes, McCarthy and Brahim¹⁵ suggest the placement of

implants in a 3 year-old child. For the patient of this report it was chosen the alternative prosthetic rehabilitation instead of the placement of implants, with fittings in the removable prosthesis according to the growth of the patient. The child adapted herself very well to the prosthesis with the great personal satisfaction.

The largest change observed with the accomplishment of the oral rehabilitation of the patient was from the psychological point of view. Another observed point was the recovery of the chewing function, as seen by the change in the diet. The child demonstrated better school life and an improvement in life quality.

CONCLUSIONS

The alternative rehabilitation treatment, with the use of removable appliances, significantly increased the retention of the removable prosthesis in a patient with ectodermal dysplasia. This alternative treatment was successful, establishing the function and improving the social living and quality of the life of the patient.

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