

Ectodermal dysplasia: case report of aesthetic and functional rehabilitation

Displasia ectodérmica: relato de caso de reabilitação estético-funcional

Nattália DI LANARO¹, Khawana FAKER¹, Viviane CANCIO¹⁻², Mônica Almeida TOSTES¹

1 - School of Dentistry, Federal Fluminense University (UFF), Niterói / RJ, Brazil.

2 - Salgado de Oliveira University (UNIVERSO) - Niterói - Rio de Janeiro - RJ - Brazil.

ABSTRACT

Objective: The aim of this study is to report a case of esthetic and prosthetic rehabilitation of a male 6-year-old patient diagnosed with Hypohidrotic Ectodermal Dysplasia, assisted at the clinics for individual with special needs held in the extension project of the Federal Fluminense University. **Methods:** This is a clinical, observational and descriptive study where the restorative and rehabilitative treatment was conducted and reported corresponding to the functional and esthetic needs of the patient. **Results:** The anatomical shape of teeth 51 and 61 was reconstructed and a partial removable prosthesis was made to cover the absence of teeth 71, 72, 74, 81 and 82, offering function, esthetics and satisfaction, as reported by the patient. **Conclusion:** This case contributed to increase the knowledge about the types of Ectodermal Dysplasia, the importance of differential diagnosis, as well as teaching of undergraduate students in the extension project.

KEYWORDS

Dental Care for Disabled; Permanent Dental Restorations; Ectodermal Dysplasia; Removable Partial Denture; Children with Disabilities.

RESUMO

Objetivo: Relatar o caso de reabilitação estético-protética de um paciente do gênero masculino com 6 anos de idade diagnosticado com Displasia Ectodérmica Hipohidrotica, atendido na clínica de pacientes especiais realizada no projeto de extensão da Universidade Federal Fluminense. **Métodos:** Trata-se de um estudo observacional, descritivo onde o tratamento restaurador e reabilitador foi realizado e retratado correspondendo às necessidades estéticas e funcionais do paciente. **Resultados:** Realizou-se reanatomização dos elementos 51 e 61 e confecção de prótese removível para suprir as ausências dos elementos 71, 72, 74, 81 e 82, propiciando função, estética e satisfação relatada pelo paciente. **Conclusão:** Este relato de caso contribuiu para o aumento de conhecimento acerca das formas de Displasia Ectodérmica, sobre a importância do diagnóstico diferencial das mesmas, bem como para o ensino dos alunos de graduação do projeto de extensão.

PALAVRAS-CHAVE

Assistência Odontológica para Pessoas com Deficiências; Restaurações Dentais Permanentes; Displasia Ectodérmica; Prótese Parcial Removível; Crianças com Deficiência.

LITERATURE REVIEW

Ectodermal Dysplasia is considered a rare genetic disorder (1: 100,000) of environmental origin, with recessive trait linked to the X chromosome when present in males and dominant when in females, and may be described as Hidrotic, Hypohidrotic or Anhidrotic [1]. This disorder is characterized by developmental defects in ectoderm-derived structures such as hair, sweat glands and teeth. According to Murdoch-Kinch et al [2], patients often have hair and pelage in smaller quantities or absent, and sweat glands may be absent or malformed. Guedes-Pinto [3] described that, in the oral cavity, teeth may be absent or present in lower number (Hypodontia), and may exhibit morphological variations. The patient with Ectodermal Dysplasia can present more intense characteristics or not, depending on the dominance/expression of the disorder.

The central purpose of this report is to present the esthetic and prosthetic rehabilitation of a male patient, aged six years old, with low dominance of the disorder, whose parents sought treatment at the clinic of special patients held in the extension project of the UFF.

CASE REPORT

The caretakers of LCL, a male child aged six years old, diagnosed with Hidrotic Ectodermal Dysplasia of low dominance, sought treatment for him in the clinics for individuals with special needs conducted at the extension project, with complaint about the “lack of some teeth”. The patient had physical characteristics of the disorder to a lesser degree, with the presence of hair and pelage, slightly dry skin, everted lips, saddle-shaped nose, and large ears. (Figure 1) Anamnesis did not reveal any reports of problems in supporting high temperatures or hypohidrosis, and there were no reports of neurological or behavioral disorders. The mother reported no other cases in the family.

During intraoral examination, the patient demonstrated a good oral hygiene condition, with no biofilm and carious lesions. Clinical and radiographic examination revealed absence of deciduous and permanent teeth, 52, 62, 71, 72, 74, 81, 82, and 12, 22, 31, 32, 41, 42, respectively. Teeth 13 and 23 were impacted and teeth 51 and 61 erupted with unusual axial tilt. The upper and lower deciduous canines exhibited conoid anatomy (Figure 2).

The first treatment was reconstruction of the anatomical shape of teeth 51 and 61, which erupted with unusual axial tilt, and light rotation. For making more accurate restoration of a previous molding, it was performed with a putty condensation silicone to obtain a palatal tab (Figure 3). The restoration was carried out with 3M microparticulate photopolymerizable resin in colors A2 and E1 (Figure 4). Also, in the lower arch, a partial removable prosthesis was made to cover the absence of teeth 71, 72, 74, 81 and 82, for better masticatory function.



Figure 1 - Facial aspect of LCL patient, 6 years old, with Ectodermal Dysplasia of low expressivity.



Figure 2 - Anterior open bite due to absence of deciduous teeth 52, 62, 71, 72, 81 and 82.



Figure 3 - Restoration with previous molding, were performed with a putty condensation silicone.



Figure 4 - Reconstruction of the anatomical shape of teeth 51 and 61 with light-curing resin and partial denture insertion containing the teeth 71, 72, 75, 81 and 82.

DISCUSSION

According to the author Murdoch-Kinch et al [2] and Guedes Pinto [3], the main features found in patients with Hidrotic Ectodermal Dysplasia of low expressivity are: presence of pelage and hair, slightly dry skin, saddle-shaped nose, large ears, hypodontia and atypical tooth morphology, which corroborate the features found in the present patient.

Oral rehabilitation of the young patients with missing teeth depends on the age, number, condition of present teeth, and the state of growth of the patient [3]. It includes prosthesis fabrication, maintaining the remaining dentition, accommodation of growth, and development and behavior management for long-term follow-up [2]. Hypodontia adversely affects physiologically as the absence of teeth is accompanied by vestigial ridge; therefore, restoring function and appearance are more challenging than usual [4]. It affects psychologically in the childhood as young patients realize themselves different from other children, hence psychological boost up is

one of the primary objectives of the treatment plan [4]. The need for the prosthesis is important during the preschool years and continues into the adulthood. Early rehabilitation is critical considering that the establishment of lifelong dietary patterns occurs during childhood and absence of several primary teeth may result in speech abnormalities as learning and reinforcement of articulation keeps on until 8 years of age.

Souza et al [4] cites that the main treatment option for hypodontia in these patients is the use of removable partial dentures in pediatric patients, being one of the treatments of choice. The patient's caretakers were informed about the need to continue the treatment in adulthood and may require the use of prostheses or implants due to congenitally missing teeth in the permanent dentition.

CONCLUSION

This case report contributed to increase the knowledge about Ectodermal Dysplasia

and contributed to teaching of undergraduate students in the extension project. The treatment was carried out according to the patient's needs and cooperation. Early prosthetic treatment is of great value to the patients with this systemic condition missing teeth from functional and psychosocial standpoint. The physiological and psychological conditions of both the parent and the patient improved notably with simple and conservative prosthetic rehabilitation.

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Viviane Andrade Cancio de Paula
(Corresponding address)

Professional Address: Universidade Federal Fluminense,
Centro de Ciências Médicas, Faculdade de Odontologia.
Rua Mário Santos Braga, n.28. Centro- Niterói, RJ, Brazil.
Postal Code: 24020-140.
Email: vicancio@ig.com.br

Date submitted: 2016 Jul 29

Accept submission: 2016 Oct 31