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Ellis-Van Creveld Syndrome, neonatal teeth and breastfeeding impairment: a case report

Síndrome de Ellis-Van Creveld, dentes neonatais e dificuldade de aleitamento materno: um relato de caso

Renata Zoraida Rizental **DELGADO**[1] https://orcid.org/0000-0002-0885-0056

Ana Carolina Fernandes **COUTO**[1] https://orcid.org/0000-0002-3025-3493

Romulo Andrade **MARCATO**[1] https://orcid.org/0000-0001-9159-8527

Danielle **PORTINHO**[2] https://orcid.org/0000-0001-5477-0666

Wanda Terezinha Garbelini **FROSSARD**[1] https://orcid.org/0000-0002-8222-3771

Cássia Cilene Dezan **GARBELINI**[1] https://orcid.org/0000-0002-8660-0677

ABSTRACT

Because of multisystemic impairment in patients with Ellis-van Creveld syndrome, multidisciplinary care may be demanded since birth to assure breastfeeding. This report presents a case of an Ellis-van Creveld infant that was facing breastfeeding difficulties because of maxillary neonatal teeth. A 3 months old male infant with Ellis-van Creveld syndrome was referred to Pediatric Dentistry Department because of two upper neonatal teeth causing breastfeeding difficulties. Clinical examination revealed that teeth position was compatible to 51 and 61, and both presented uncommon ectopic soft tissue placement, conical crown and hypoplastic enamel covered by a large amount of dental biofilm. Radiography indicated they were of normal series and had 2/3 of crown completion. Due to teeth mobility that impaired breastfeeding, treatment option was exodontia. Early tooth eruption, such as in natal and neonatal teeth, by itself can't be considered a reason for exodontia. But exodontia must be considered when an early erupted tooth(s) impairs breastfeeding, especially in systemically compromised infants. In this present case report, after teeth extraction, the infant was able to breastfeed and gain weight properly.

Indexing terms: Breast feeding. Ellis-Van Creveld syndrome. Infant. Natal teeth.

RESUMO

Devido ao comprometimento multissistêmico em pacientes com Síndrome de Ellis-Van Creveld, cuidados multidisciplinares podem ser necessários desde o nascimento a fim de assegurar o aleitamento materno. O presente relato apresenta um caso de um bebê portador de Síndrome de Ellis-Van Creveld, o qual apresentava dificuldades durante a amamentação devido a dentes neonatais superiores.

[1] Universidade Estadual de Londrina, Faculdade de Odontologia, Departamento de Medicina Oral e Odontologia Infantil. Rod. Celso Garcia Cid, km 380, Campus Universitário, Londrina, PR, Brasil. Correspondence to: RZR Delgado. E-mail: <renata.rizental@gmail.com>.

[2] Universidade Estadual do Oeste do Paraná, Faculdade de Odontologia, Departamento de Odontologia. Cascavel, PR, Brasil.

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Um bebê de 3 meses de vida, portador da Síndrome de Ellis-Van Creveld foi encaminhado ao Departamento de Odontopediatria, por apresentar dois dentes neonatais superiores, os quais estavam causando dificuldades durante o aleitamento materno. O exame clínico demonstrou que a posição dos dentes era compatível com os dentes decíduos #51 e #61, ambos apresentavam implantação ectópica incomum em tecido mole, apresentavam coroa cônica e esmalte hipoplásico coberto por grande quantidade de biofilme dentário. O exame radiográfico indicou que os dentes eram compatíveis com a série normal e apresentavam desenvolvimento completo de 2/3 da coroa. Devido a mobilidade dentária, a qual prejudicava o aleitamento materno, a opção de tratamento foi a exodontia. A erupção dentária precoce, como ocorre com dentes natais ou neonatais, por si só não pode ser considerada motivo para a realização de exodontia, porém esta deve ser considerada, quando prejudica o aleitamento materno, especialmente em bebês sistemicamente comprometidos. No presente relato de caso, após a exodontia, o bebê conseguiu realizar o aleitamento materno e apresentou ganho de peso correto.

Termos de indexação: *Aleitamento materno; Síndrome de Ellis-Van Creveld. Lactente. Dentes natais.*

INTRODUCTION

Ellis Van-Creveld Syndrome (EVCS), also known as chondroectodermal dysplasia, is a complex genetic disorder [1]. It presents multisystemic features, signs and symptoms ranging from bone and cardiac abnormalities until stomatologic ones [2]. The general occurrence is low, with a prevalence of 7:10 births [3, 4] affecting both genders and without race predilection [1]. Congenital heart disease may support the diagnosis and appears to be the main determinant of longevity [1], 50% of childhood deaths are associated with cardiac and respiratory changes that patients may present [3].

Oral aspects include labiogingival adherences, accessory labiogingival frenula, labiogingival frenulum hypertrophy, conical teeth, enamel hypoplasia and hypodontia, natal and neonatal teeth, changes in eruption and exfoliation dental patterns, malocclusion and some others abnormalities [1,5,6].

A multidisciplinary approach is necessary for the management of the various clinical manifestations of EVCS. Pediatric dentistry follow-up is important for the prompt diagnosis and treatment of oral disorders, especially in newborns and infants who present conditions that may compromise breastfeeding and may increase the risk of bronchoaspiration [7]. It is fundamental to provide parents information about measures to oral health promotion, aiming at improving patients' quality of life. The aim of this study is to report a case of an infant with EVCS that was facing breastfeeding difficulties due to rare maxillary neonatal teeth. The care plan developed to ensure continued breastfeeding is presented.

CASE REPORT

The pediatric cardiology team of the University of Londrina referred to the Pediatric Dentistry Department a 3-month and 21-day-old male EVCS infant. He was the first child of non-consanguineous parents who reported no history of relatives with EVCS or EVCS related characteristics. Infant's medical history was included:

a) Gestational and childbirth history: previous diagnosis of suspected syndrome on ultrasonography at the 34th gestational week, in which short bones characteristic of dwarfism and pulmonary hypoplasia were detected; cesarean delivery at 40 weeks, Apgar 7 and 8, weighing and measuring, respectively, 3.155 kg and 45 cm.

b) Neonatal Period: intensive care unit monitoring for 55 days due to pulmonary hypertension, cardiac atrial septum defect, persistent left superior vena cava, bicuspid aortic valve, tricuspid insufficiency, and coarctation of the aorta. He was submitted to corrective heart surgery at 56 days of life, with good evolution.

c) Secondary Clinical Findings: syndromic face, retrognathia, low-set ears, hypertelorism and hypoplastic thorax due to short ribs, pulmonary hypertension secondary to thoracic deformity, single transverse palmar crease, polydactyly affecting hands, rocker-bottom feet, nail hypoplasia, flat hemangioma in left eyelid and in occipital, and convergent strabismus [figure 1].

Oral clinical examination revealed fusion of upper lip and labial gingiva, multiple frenulum's, segmentation of upper and lower anterior alveolar ridge, upper labiogingival hypertrophy and eruption of 51 and 61. Both teeth was composed by hypoplastic enamel, conical-shaped crowns; and no alveolar bone was identified surrounding the early stage dental structure highlighting an uncommon ectopic soft tissue placement. Radiographic examination indicated that both teeth were of normal series [figure 2]. According to mother's report, both teeth erupted some days after birth, because of it they were denominated as neonatal teeth (NNT). Until then, they had not been interfering with

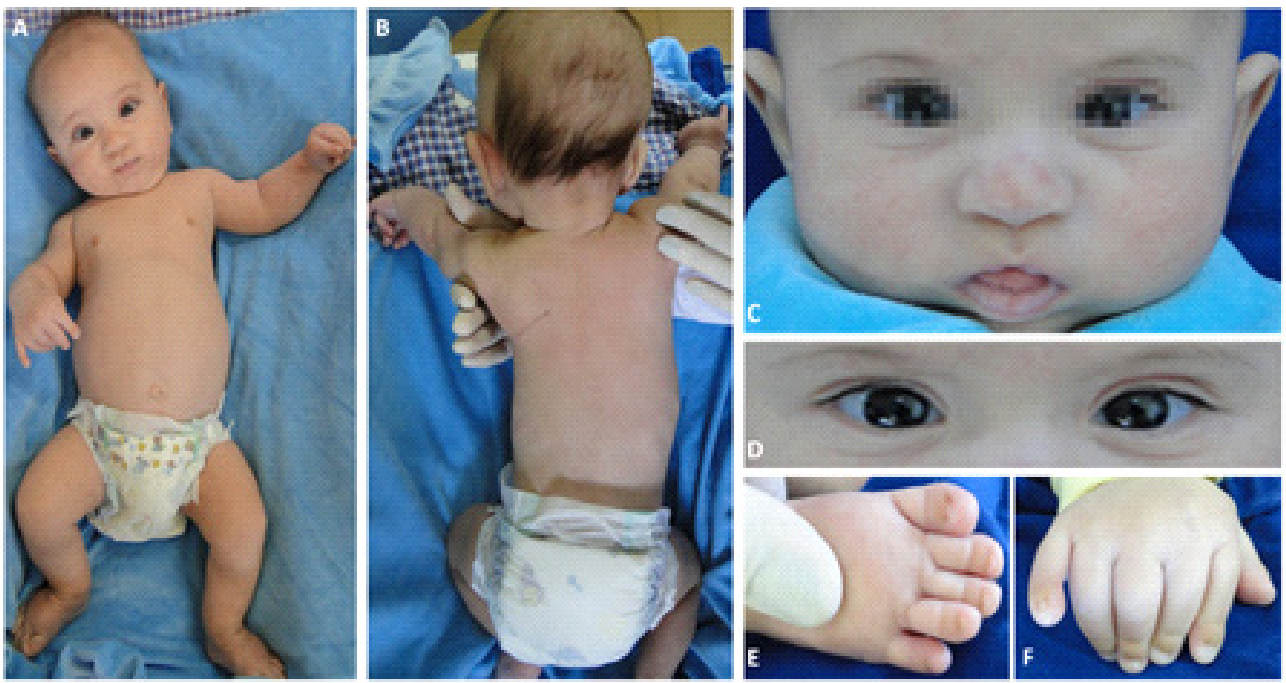

Figure 1. General physical features (A-F) and buccal aspects (G-I). (A) Front view showed mammary hypertelorism and hypoplastic thorax due to short ribs. (B) Back view – fine sparse hair and cicatricial tissue from corrective cardiac surgery at left subscapular region. (C) Front view showing syndromical face, low set ears and upper lip hypertrophy. (D) Convergent strabismus. (E) Image of foot presenting hypoplastic fingernails. (F) Image of hand showing polydactyly and hypoplastic fingernails.

breastfeeding. Since the third month, the mother noticed that the infant started to experience difficulties because of the teeth.

NNT implantation characteristics represented a risk factor for bronchoaspiration. This finding, together with the mother's report of not being able to carry out breastfeeding satisfactorily, led us to opt for extraction of both teeth.

Medical team was contacted and authorized to carry on exodontia in outpatient clinic and antibiotic prophylaxis was done with amoxicillin oral suspension

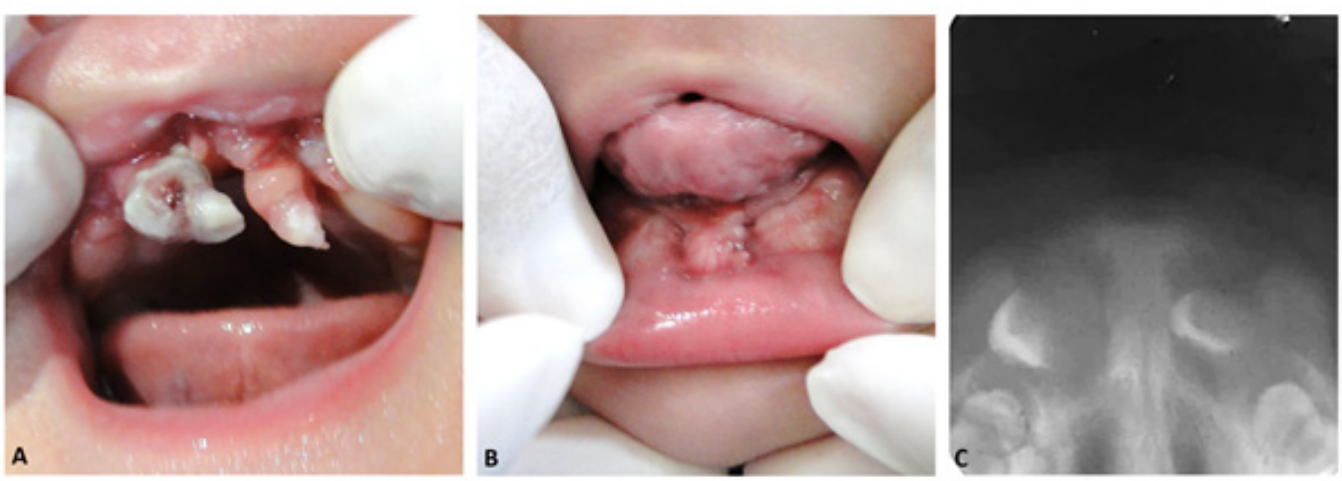

Figure 2. (A) Superior arch - note uncommon ectopic soft tissue placement of teeth #51 and #61, labiogingival hypertrophy, absence of mucobuccal fold and multiple labiogingival frenula. Serrated anterior/superior ridge. (B) Lower arch. Showing absence of mucobuccal fold and multiple labiogingival frenula. Segmental alveolar ridge. (C) Occlusal radiograph. Note that the teeth are part of the deciduous dentition.

(250 mg/ml - 3.33 ml). Patient protective stabilization was then performed, antisepsis of the operative field (0.12% chlorhexidine digluconate), topical anesthesia (tetracaine hydrochloride 1% and phenylephrine hydrochloride 0.1%), excision by cutting, irrigation with 0.9% sodium chloride saline, and sterile gauze compression to blood clot stabilization. After 1 week the region presented evolution compatible with normal healing [figure 3].

The biological material was sent for anatomopathological examination. Histopathology revealed fibrous connective tissue with mixed inflammatory process and granulation tissue fragments, ulcerated surface covered by purulent fibrous membrane, bacterial colonies and fragments of hard tissue compatible with enamel, dentin and pulp tissue [figure 4].

Although apprehensive about the surgical procedure, the parents accepted the proposed treatment because the infant had been presenting weight loss due to breastfeeding difficulty. A few hours after the procedure, breastfeeding was performed without any intercurrences.

The patient is under continuous medical and dental follow-up, presenting health and cognitive development compatible with his condition. Furthermore, after healing process, the infant presented weight gain.

DISCUSSION

Some infant's oral cavity anomalies may impair breastfeeding [8] being ankyloglossia the most studied one. EVCS infants present a diversity of oral problems that can affect breastfeeding - i.e. labiogingival adherences, accessory labiogingival frenula, labiogingival frenulum hypertrophy, natal and neonatal teeth, change in dental eruption timing [1,5,6].

In view of this, pediatric dentists' inclusion in multidisciplinary care team to ensure breastfeeding in

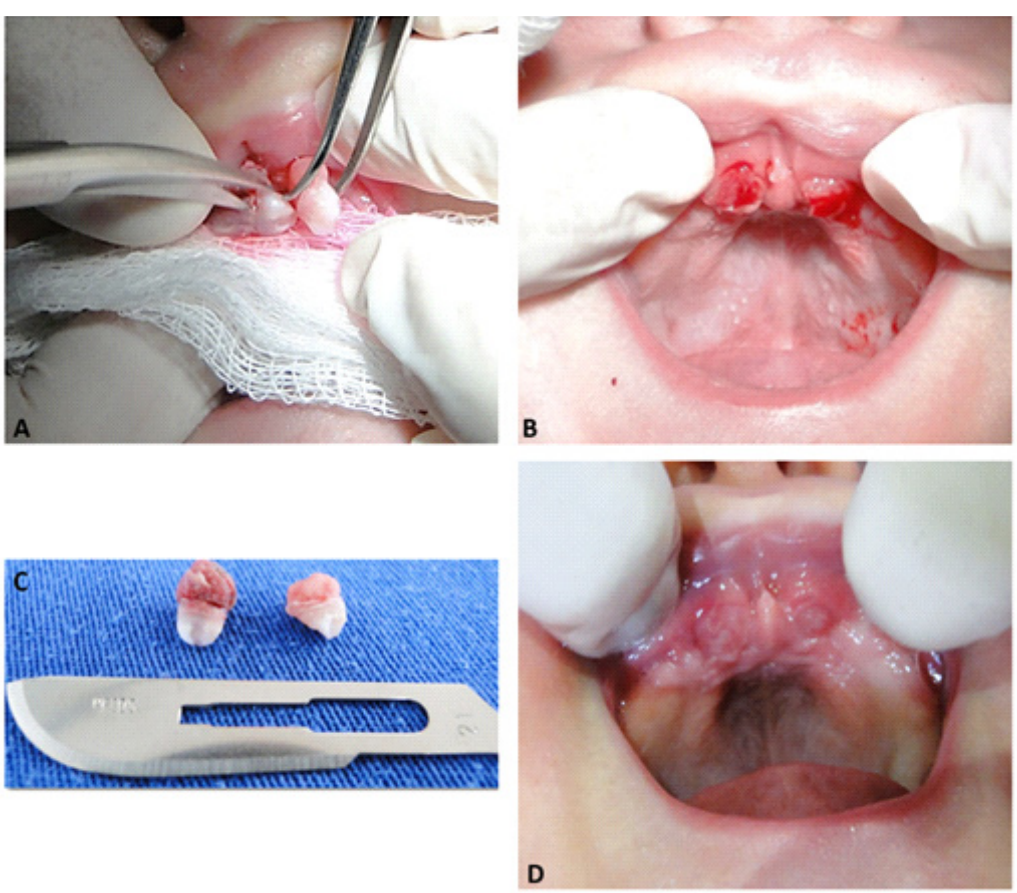

Figure 3. Surgical procedure under local anesthesia. (A) Exertion with Goldman-Fox scissors. (B) Photographic image of the region operated immediately after the surgical procedure. (C) Anatomical parts. (D) Intraoral image showing the wound healing after 7 days.

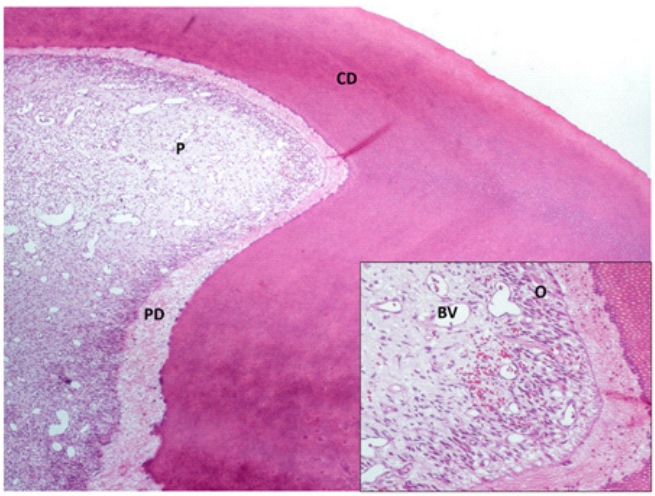

Figure 4. Histological photomicrography of H&E stained. The image shows mineralized tissue with tubular morphology compatible with coronary dentin (CD), odontoblast layer (O), organized predentin (PD), pulp tissue (P) composed of loose connective tissue showing numerous blood vessels (BV). (10x).

EVCS infants must be considered. Early weaning due to natal/neonatal teeth impairs infants' nutrition, growth and development. Moreover, this event should be avoided [9-11], especially in systemically compromised patients.

Although oral characteristics of EVCS are widely documented, there is a lack of reports about the oral condition of EVCS infants. Knowledge about EVCS infants presenting natal/neonatal tooth(s) is mostly based on case reports about EVCS children, adolescents or adults that presented them, according to parents report (memories) [4,12]. A bibliographic search revealed only three reports of NNT in EVCS infants [12,13]. Of these, only the study of Farah et al. [5] presents NNT confirmation. In the case reported by Shawky, Sadik and Seifeldin [13] there is mention of NNT, however, it is not provided information about teeth eruption period and radiographic examination is absent. In turn, the case reported by Serotkin, Stamberg and Waber [12] did not present main characteristics related to EVCS - cardiac and pulmonary impairment. On the other hand, unusual features of the syndrome were reported by the authors, such as inguinal hernias, supernumerary ribs, and delayed motor and cognitive development. In addition, the genetic involvement described is different (mosaicism of chromosome 17q) from those commonly reported for EVCS [14,15].

Natal tooth and NNT eruption may occur because of superficial position of dental germ, infection, accelerated eruption due to peak fever, heredity and nutritional deficiencies, as well as by genetic conditions related to some syndromes [16]. The frequency of NNT in the mandibular anterior region is 85%, whereas only 11% erupt in maxilla, similar to his present case [7,11]. Although both teeth were of normal series, absence of bone insertion (because they had no root development) caused teeth mobility, what impaired breastfeeding and represented a risk of bronchoaspiration and pulmonary infection. Such situations indicate the need to remove these teeth [5, 7]. As there is no consensus about antibiotic prophylaxis in patients with congenital heart defects [17-19] and considering the infant's recent cardiac surgery, we contacted cardiologic team and follow their recommendation on antibiotic prescription.

The classic tetrad characteristic of EVCS, composed of chondrodysplasia, polydactyly, congenital heart alterations and ectodermal alterations is widely reported by the scientific community [1,2]. The case here reported presented all the above-mentioned findings and others less frequent as: low-set ears, strabismus and flat hemangioma in the occipital region and left eyelid. Ectodermal disorders such as nail hypoplasia, fine hair, change in chronology of dental eruption, and presence of multiple braces and buccal flanges are commonly reported in patients with EVCS [5,20,21].

Consanguinity is associated in approximately 30% of the reported EVCS cases, while other reports do not present this relation [5, 20, 21], as in our study. Chondrodysplasia is the most commonly reported clinical sign and is often diagnosed in the gestational period by ultrasonography [20,22]. In this report, the diagnostic hypothesis of dwarfism was raised at the 34th week of gestation by ultrasound, reinforcing the importance of prenatal follow-up.

CONCLUSION

Although rare, EVCS requires a multidisciplinary approach. Since birth, pediatric dentistry plays an important role in the management of syndrome's oral findings, as described in this paper. In this present case report, after teeth extraction, the infant was able to breastfeed and gain weight properly. At first, the action should ensure breastfeeding by prompt diagnostic and treatment of oral conditions that difficult breastfeeding. Later, it would

focus on in emphasizing preventive treatments such as dietary counseling, oral hygiene and reinforcement about the importance of regular dental care aiming subsequent rehabilitation of aesthetic and masticatory functions.

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Collaborators

RZR DELGADO, ACF COUTO, RA MARCATO, contribute to conception/ design, contributed to acquisition, analysis/ interpretation, drafted the manuscript., contribute to conception/ design, contributed to acquisition, analysis/ interpretation, drafted the manuscript. D PORTINHO, contribute to conception/ design, drafted the manuscript, and critically revised the manuscript. WTG Frossard, contribute to conception/ design, supervised all stages of clinical treatment, drafted the manuscript, and critically revised the manuscript. CCD GARBELINI, Contribute to conception/ design, supervised all stages of clinical treatment, drafted the manuscript, and critically revised the manuscript. All authors reviewed the text and approved its final version.

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