Cri-du-Chat syndrome: conservative dental treatment in an 8-year old child

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ABSTRACT

The aim of this article was to report the case of an 8-year old patient with Cri-du-Chat syndrome (CdCS) referred to the pediatric dental clinic for urgent dental treatment. The chief complaints were a traumatic injury to both permanent maxillary central incisors and difficulty performing oral hygiene. The patient was extremely cooperative during clinical evaluation, demonstrating ability to withstand conservative treatment options. Dental examination revealed accentuated overjet, carious lesions, and very poor oral hygiene. Clinical and radiographic evaluations were necessary to diagnose and determine treatment strategies for the traumatic injuries, carious lesions, and gingivitis, which were implemented over a total of five dental visits. Outcomes of the treatment strategies adopted are described. This case report illustrates the variety of clinical findings that pediatric dentists may encounter in the oral cavity and face of patients with CdCS and the problems that these alterations may cause. When faced with a CdCS patient, dentists should try to provide the best treatment possible and prioritize the use of conservative techniques.

Keywords: Cri-du-Chat Syndrome, Maxillofacial Injuries, Local Anesthesia, Atraumatic Restorative Treatment.

Síndrome de Cri-du-Chat: tratamento odontológico conservador em uma criança de 8 anos de idade

RESUMO

O objetivo deste artigo é relatar o caso de uma paciente de 8 anos de idade com a síndrome do miado do gato (CdCS) que foi encaminhada à clínica de odontopediatria para tratamento odontológico de urgência. As principais queixas eram uma lesão traumática nos incisivos centrais superiores permanentes e dificuldade em realizar a higiene oral. A paciente foi extremamente cooperativa durante a avaliação clínica, demonstrando capacidade de colaborar com opções de tratamentos conservadores sob anestesia local. O exame clínico mostrou overjet acentuado, lesões de cárie e uma higiene oral muito pobre. Avaliações clínicas e radiográficas foram necessárias para diagnosticar e determinar estratégias de tratamento das lesões traumáticas, lesões de cárie e

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gengivite, as quais foram realizadas ao longo de cinco consultas odontológicas. Os desfechos do tratamento são descritos. Este caso ilustra a variedade de achados clínicos que os odontopediatras podem encontrar na cavidade oral e face de pacientes com CdCS e os problemas que essas alterações podem causar. No atendimento desses pacientes, os dentistas devem tentar fornecer o melhor tratamento possível, priorizando o uso de técnicas conservadoras.

Palavras-chave: Síndrome do Miado do Gato, Traumatismos Maxilofaciais, Anestesia Local, Tratamento Dentário Restaurador sem Trauma.

INTRODUCTION

Cri-du-Chat syndrome (CdCS) (MIM ID # 123450) is a genetic disease resulting from the total or partial deletion of the short arm of chromosome number 5 (1,2). The condition was first described by Lejeune et al. in 1963 (3). *Cri du Chat* in French means "cat-like cry" and refers to the characteristic cry of affected children at birth. Diagnosis can be made immediately after birth, based on the peculiar cry, probably caused by abnormal development of the larynx or a variety of neurological disorders (4). Studies have reported incidence rates ranging from 1:15,000 to 1:94,000 live births (1,5), but the prevalence found by Niebuhr[4] in 6,000 individuals with mental retardation was approximately 1:350. Furthermore, data reported by Higurashi's et al. (5) suggested a significantly higher prevalence of the disorder among females. Mainardi et al. (6) analyzed 80 patients and 148 parents from the Italian CdCS Register and found a paternal origin of the deleted chromosome 5 in 90.2% of the cases. Clinical evaluation of those patients revealed higher degree severity for microcephaly, dysmorphism, and psychomotor retardation related to the size of deletion of chromosome 5, i.e., the more severe cases were those with larger deletions.

The main characteristics of CdCS are a high-pitched cat-like cry, distinct facial dysmorphism, microcephaly, severe psychomotor and mental retardation, as well as abnormal facial features (1), e.g., a round face, hypertelorism, micrognathia, epicanthal folds, and low-set ears (3). Cardiac malformations, neurological and renal abnormalities, preauricular tags, syndactyly, hypospadias, and cryptorchidism can also be found (1,2,4).

Reported orofacial manifestations include mandibular microretrognathia, high palate, variable malocclusion, enamel hypoplasia, poor oral hygiene, generalized chronic periodontitis, and delayed tooth eruption (2,4,7). Yáñez-Vico et al. (8) evaluated the craniofacial morphology of 10 CdCS patients using standard cephalometric methods. The main characteristics found were skeletal class II malocclusion caused by mandibular retrognathism, dental biprotrusion, and a small upper airway. Additionally, 70% of the patients had a steep palatal plane angle and a flattened skull base angle.

Cornish and Pigram (9) assessed 27 children with CdCS and demonstrated that, with early special education and an atmosphere of family support, some affected children reached psychomotor and social levels compatible with those of normal children. That study demonstrated the potential of children with this syndrome to develop and maintain important skills, including the ability to communicate needs, to socially interact with

others, and to develop some degree of mobility. These findings underscore the need for these patients to receive a multidisciplinary treatment.

Whenever general anesthesia is needed, it should be carefully accomplished using all necessary measures, as this procedure increases the risk of complications. In addition, intubation may be hindered by retrognathia and deep palate. When possible, dental treatment should be carried out under local anesthesia, and sedatives should be used if necessary to improve cooperation and avoid injuries as much as possible (10). In view of the scarce literature describing dental conditions and dental treatments in CdCS patients, the aim of this article was to report the case of a CdCS female patient who received conservative dental treatment under local anesthesia, as well as describe the peculiarities related to this syndrome.

CASE DESCRIPTION

An 8-year old female child with CdCS was referred to the pediatric dental clinic at the School of Dentistry of Federal University of Rio Grande do Sul for urgent dental treatment. The chief complaints were a traumatic injury to both permanent upper central incisors and difficulty performing oral hygiene. The patient's mother was 32 years old when she became pregnant. She received medical follow-up throughout pregnancy, and syphilis was diagnosed at 39 weeks' gestation, after which delivery was immediately undertaken. The mother did not report the presence of congenital anomalies in other family members. During the clinical interview, the mother informed that the girl had CdCS and that she had been under medical supervision since birth. It was not possible to obtain the father's medical history.

At the moment of the interview, the child was not under any systemic drug therapy and did not present any involvement of vital organs. Notwithstanding, she showed some degree of mental retardation, chronic anemia, worms, and flu. The mother also informed that the patient consumed mainly pasty foods due to mastication problems. The patient did not present congenital heart alterations. She was extremely cooperative during the interview and clinical evaluation, demonstrating ability to withstand conservative treatment options under local anesthesia if necessary.

During the first clinical evaluation, some characteristics were recorded: facial asymmetry, low-set ears, preauricular tags, eyes slightly separated, epicanthal folds, down-turned corners of mouth, convex facial profile with mandibular microretrognathia, difficulty opening the mouth, especially on the left side, hypofunction of perioral muscles, cheeks and tongue, and absence of lip seal (Figure 1a-b). Dental examination revealed vestibular projection of upper central incisors, which suffered lateral and extrusive luxation, carious lesions affecting all posterior teeth on the left side, due to the difficulty opening the mouth, and very poor oral hygiene. Other structures showed normal characteristics for an 8-year old child. Clinical and radiographic evaluations were necessary to diagnose and determine treatment strategies for the traumatic injuries, carious lesions, and gingivitis (Figure 1c-d). At the first periapical radiograph, no sign of resorption or apical lesion

was diagnosed, indicating the monitoring of trauma. Incomplete root formation was also observed in both teeth.

Still during the first visit, the child and her mother received instructions on how to perform effective oral hygiene as a strategy to reduce gingivitis. In addition, chlorhexidine gluconate 2% was prescribed once a day, for 10 days, to improve periodontal status at the site of dental trauma. After 4 days, the patient showed better gingival condition, and a flexible splint was made using nylon floss and composite resin and anchored to the primary molar to allow periodontal healing of both traumatized incisors (Figure 2a-b).

Three weeks later, in the first periapical control radiograph, an initial process of external root resorption in both traumatized teeth was observed. In this case, as the patient seemed able to undergo local anesthesia, pulpectomy was the treatment proposed. The mother and patient readily agreed with the suggestion, and pulpectomy was initiated using the tell-show-do technique, under local anesthesia.

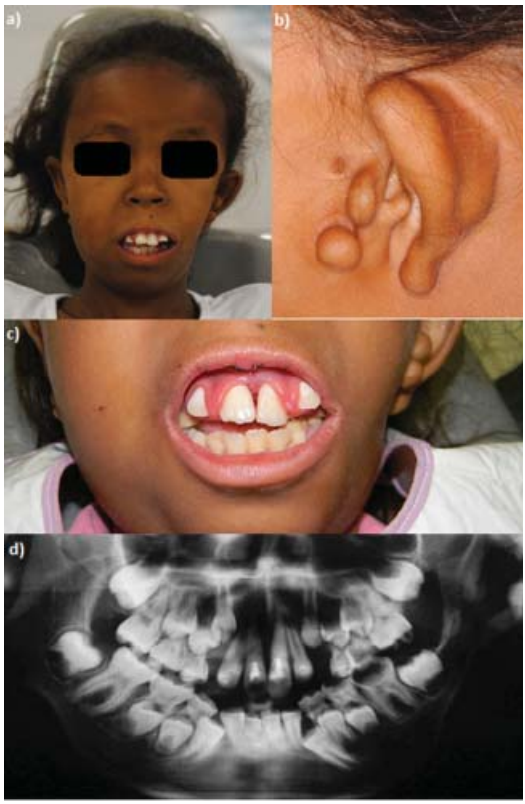

FIGURE 1 – Clinical and radiographic findings: a) face asymmetry, low-set ears, preauricular tags, eyes slightly separated, epicanthal folds, down-turned corners of mouth, convex facial profile with mandibular microretrognathia, hypofunction of perioral musculature, cheeks, and tongue, and absence of lip seal; b) preauricular tags; c) vestibular projection of upper central incisors, which suffered lateral and extrusive luxation; d) panoramic radiograph obtained after the introduction of calcium hydroxide paste to stimulate apexification of immature teeth.

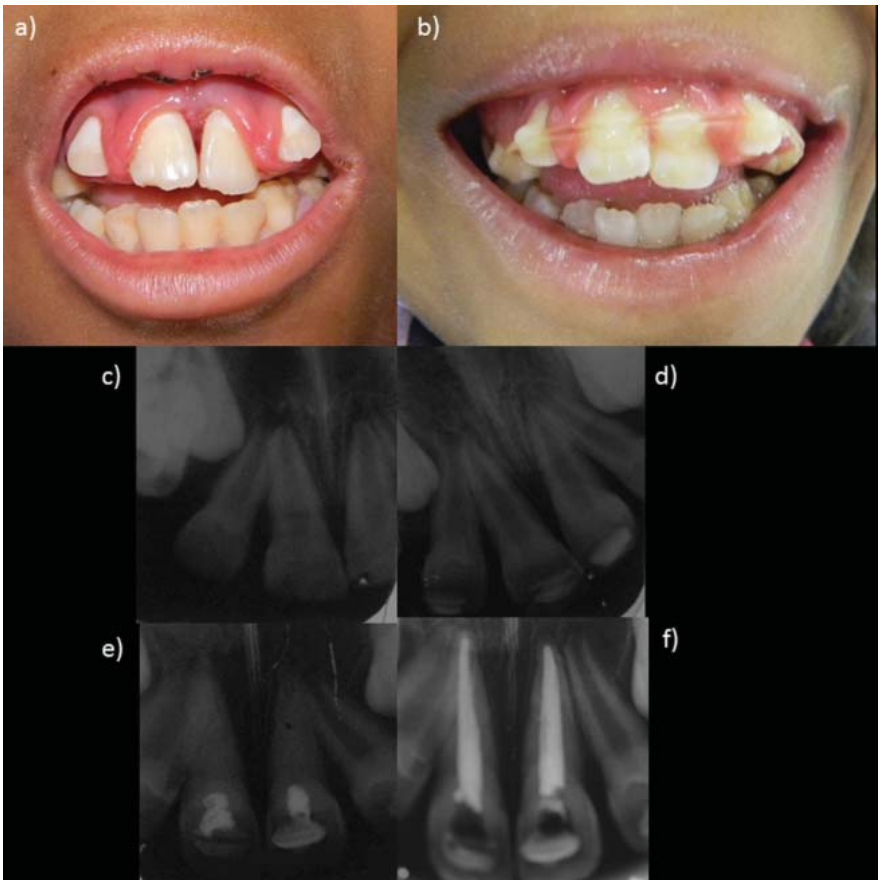

FIGURE 2 – a) Traumatized upper central incisors at first visit; b) 4 days later, with better gingival condition, a flexible splint was anchored to the primary molar to allow periodontal healing of both traumatized incisors; c) periapical radiograph obtained at first visit; d) 3 weeks later, an initial process of external root resorption in both teeth was observed; e) introduction of calcium hydroxide paste to stimulate apexification of immature teeth; f) 5 months later, successful apical barrier formation was observed, with no signs of external resorption progression and final obturation of both incisors.

At first, a calcium hydroxide-based intracanal medicament (Calen, S. S. White, Rio de Janeiro, RJ, Brazil) was introduced into the root canals to stimulate apexification of the immature teeth (11). The patient was extremely collaborative throughout the procedure, demonstrating ability to undergo more conservative treatments without the need for general anesthesia. After 5 months and two exchanges of the calcium hydroxide intracanal dressing, successful apical barrier formation, and no signs of progression of the external resorption process, final obturation of both incisors was performed, followed by restoration with composite resin (Z350XT-3M ESPE, Sumaré, SP, Brazil) (Figure 2c-f). None of the endodontic procedures was performed under rubber dam isolation, due to patient's difficulty in opening her mouth but extreme collaboration.

All carious lesions affected posterior teeth located on the left side, probably due to difficulties cleaning that side as a result of limited mouth opening on that side. They all involved up to the inner half of dentin, with no pulp involvement. The patient and her mother did not report spontaneous pain. As a conservative treatment option, the partial caries removal technique (12,13) was performed, without local anesthesia or rubber dam, and teeth were restored using a modified glass-ionomer cement (Vitremer-3M ESPE, Sumaré, SP, Brazil) after indirect pulp capping with calcium hydroxide cement (Dycal, Dentsply, Petrópolis, RJ, Brazil).

After 3 months, the patient presented good oral hygiene and absence of periodontal disease. The flexible splint was removed, and the first primary molars were extracted as part of a serial extraction plan to allow the correct eruption of permanent teeth. The girl is now 10 years old and has lost all her primary teeth. She returns for regular maintenance visits to monitor the traumatized teeth and oral hygiene conditions (Figure 3).

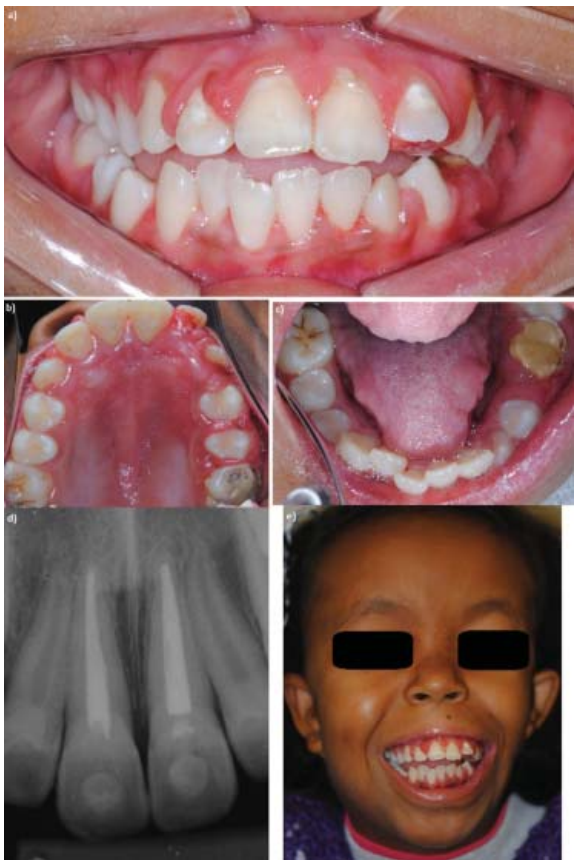

FIGURE 3 – a-c) Clinical features 1 year and 10 months after the first visit, demonstrating adequate hygiene and the correct eruption of permanent teeth; d) control periapical radiograph of both traumatized incisors obtained 17 months after final obturation; e) final facial aspect of patient.

DISCUSSION

Even being considered a rare genetic autosomal disorder, not common in dental practice, CdCS presents orofacial manifestations that require specific and specialized dental treatment. Consequently, it is important that health professionals have some understanding of the developmental and behavioral characteristics of children with this condition. The present case report describes orofacial characteristics that are in accordance with previous descriptions found in the literature (1-6).

The increased risk for caries and periodontal disease in these patients can be associated with the orofacial manifestations of the syndrome and poor oral hygiene resulting from difficulties opening the mouth (2). Anterior open-bite and accentuated overjet, common craniofacial characteristics of the syndrome, may have contributed to the traumatic injuries observed in our patient (8, 14).

Carious lesions were treated conservatively by partial caries removal (12, 13) and restored using a modified glass-ionomer cement after pulp capping with calcium hydroxide. This technique involves minimal intervention, reducing the risk of pulp exposure and technical complications (13), advantages that are extremely important for this special patient.

Flexible splinting and apexification were used to treat the traumatic injuries of permanent incisors. This procedure enabled periodontal healing and apexification of the roots, enhancing endodontic treatment conditions. The calcium hydroxide apexification technique, which was part of the conservative treatment proposed for this child, is well established in the literature, with proven biocompatibility and high success rates stimulating apexification of immature permanent teeth (11).

Due to the patient's difficulty opening the mouth, the whole treatment was adapted to the patient's oral condition, and thus performed without rubber dam isolation. During partial caries removal (12, 13), no local anesthesia was used, and the patient did not report any discomfort. Another treatment option in this situation (syndromic patients) would be the extraction of teeth with difficult access to treatment pulpectomy in one session for traumatized teeth, under general anesthesia. According to Ohtawa et al. (15), this choice should be considered in cases of poor patient cooperation, extremely difficult type or location of treatment, when a lot of time is required, or when the patient is required to attend the clinic several times.

Considering that other dental clinics had previously refused to provide dental treatment to the patient because of the diagnosis of CdCS, the fact that the patient presented the syndrome was not an impediment for clinical dental treatment. In this case, after full explanation of the treatment plan to the mother and child using the tell-show-do technique, the patient fully collaborated with all procedures, allowing the conservative treatments proposed.

CONCLUSION

This paper illustrates the variety of clinical findings that pediatric dentists may encounter in the oral cavity and face of patients with CdCS and the problems that these alterations may cause. It also shows that, even though patients with CdCS may present some difficulties because of their condition, a well-prepared team of dentists should be able to treat them. In sum, when faced with a CdCS patient, dentists should try to provide the best treatment possible according to patient cooperation and prioritize the use of conservative techniques with local anesthesia when necessary.

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