

Initial approach in cleidocranial dysplasia: the role of Pediatric Dentistry

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ABSTRACT

Objective: to emphasize the importance of Pediatric Dentistry in the dental treatment planning as a first step of rehabilitation in a pediatric patient diagnosed with Cleidocranial Dysplasia (CCD). **Case Report:** this study presents the case of a ten years old girl, with CCD diagnosis, which attended the Sector of Pediatric Dentistry of São Leopoldo Mandic School of Dentistry - Campinas / São Paulo sent by orthopedist, your main complaint was the delay on superior anterior teeth eruption. Needs preventive, curative, rehabilitative, orthodontic and surgical dental treatment. After an interdisciplinary planning, a treatment proposal was nominated and accepted by responsables' patient.

Keywords: Cleidocranial dysplasia; Supernumerary tooth; Osteochondrodysplasias.

Introduction

The cleidocranial dysplasia (CCD) is an autosomal dominant inheritance pattern of disease and can also happen spontaneously, without gender predilection or ethnic group. It is also known as Marie & Sainton Disease, Scheuthauer-Marie-Sainton Syndrome, Mutational Dysostosis or Cleidocranial Dysostosis.¹ The disease was described on 1898,² but the current nomenclature Cleidocranial Dysplasia is defined to describe the wide spectrum of signs and symptoms.³

Its occurrence is rare, estimated at 1: 1.000,000 born.⁴ A defect of the gene CBFA1, present in 6p21 chromosome, also known as RUNX2 (Runt-Related Transcription Factor 2) is reported as the origin of the disease.⁵

Patients with CCD usually have a life without interferences and major medical complications,⁶ so the diagnosis of the disease may be late.⁷

However, the clinical phenotype of CCD is quite characteristic: short stature, due to the occasional stop of long bones growth, and clavicles with hypoplasia and malformation, to varying degrees, and may be absent, unilaterally or bilaterally, in 10% of cases. Clavicular hypoplasia gives a long neck appearance and allows for extensive shoulder movements to the point of touching one another in the medium body line, and its function is rarely affected.⁵

In the dental changes, there is atresic maxilla with a deep palate resulting relative mandibular prognathism, prolonged retention of deciduous teeth, impacted permanent teeth or with late eruption, multiple supernumerary teeth, crown and root abnormalities, crypt formation around impacted teeth.¹

Among the craniofacial characteristics, frontal and pa-

rietal prominence, delayed closure of the anterior fontanelle, hypoplasia of maxilla and zygoma, underdeveloped and narrow paranasal sinuses, wide nose with depressed nasal bridge and ocular hypertelorism.⁵

In view of all these characteristics, it is evidenced that CCD requires great efforts due to the complexity of the treatment, because in addition to the patient's aesthetic-functional rehabilitation there is also psychological support and adequate motivation for the patient and family.¹

The importance of a multidisciplinary therapeutic planning involving several specialties is highlighted, in which the success of the treatment depends on the moment of the intervention.⁶

Based on this principle, the importance of Pediatric Dentistry is emphasized in the success of dental treatment planning, since it is the specialty referring to the initial approach of the patient. The Pediatric Dentistry approach is always preventive, to prepare the patient for the following phases of multidisciplinary treatment. It may also be therapeutic, when the patient has some prior need to continue treatment of oral changes of Cleidocranial Dysplasia.

Therefore, the aim of this study is to emphasize the importance of Pediatric Dentistry in the dental treatment planning as a first step of rehabilitation reporting a case of pediatric patient diagnosed with CCD and needs of clinical, surgical and orthodontics origin.

Case Report

A 10-years-old female patient was presented with your parents to the Pediatric Dentistry Clinic of the São Leopoldo Mandic School of Dentistry - campus Campinas / São Paulo, sent by the orthopedist with a diagnosis of Cleidocranial

Dysplasia. The patient's main complaint was the delay on superior anterior teeth irruption. The treatment and scientific divulgation were accepted and signed by responsible's patient on a consent term.

At the general physical examination (Figure 1A) was observed a short stature with a thin trunk, short hands and toes, and drooping shoulders, which are found in the patient's medium body line when requested. In the facial analysis, an enlarged cranial dimension was observed, with maxillary deficiency and absence of zygomatic prominence, giving the characteristic of a III facial pattern; facial profile of an adult, not compatible with the age of the patient.



Figure 1. General physical examination, observing the meeting of the shoulders in the medium body line (A); intrabuccal examination: occlusion (B), upper arch (C) and lower arch (D)

In the oral examination, maxillary atresia was observed, resulting in an arched and deep palate with posterior crossbite. Besides of anterior open bite due to deleterious habit (finger) - as observed in Figure 1B. In Figures 1C and 1D, we can observe the prolonged permanence of deciduous incisors (51, 52, 61, 62, 72) and delay in the irruption of the permanent successors; unsatisfactory restorations with presence of mechanical failure on teeth 55 and 84; caries lesion score 3 active ICDAS on teeth 16 and 26; provisional restorations on teeth 74 and 75.

Radiographic investigations included: panoramic radiography (Figure 2), in which it was possible to observe the presence of eight supernumerary teeth and permanent teeth in process of eruption with radicular formation without alterations. It was still detected that the teeth 74 and 75 had an endodontic provisional treatment without the end of definitive treatment. Teleradiography in the lateral norm demonstrated maxillary atresia, obtuse gonial angle with vertical

tendency of mandibular growth, lower incisors with lingual inclination at the mandibular symphysis. Periapical radiographs were also performed in the incisor region for the numerical identification of supernumerary teeth.



Figure 2. Panoramic radiography

After these complementary exams and associated with the clinical examination, it was possible to bring together an interdisciplinary team to carry out an integrated planning that would allow a holistic approach.

The interdisciplinary team was composed of specialists in pediatric dentistry, dentistry for special patients, orthodontists and bucco-maxillo-facial surgeon. The following treatment sequence was defined: clinical, orthodontic and surgical.

The clinical treatment stages developed at the pediatric dentistry clinic were curative and preventive, consisting: orientation of oral hygiene habits, ionomeric sealant on teeth 16 and 26; composite resin repair on teeth 55 and 84; endodontic treatment and aesthetic-functional rehabilitation on teeth 74 and 75.

After the clinical treatment was completed, the patient began orthodontic treatment for maxillary disjunction, in order to correct the posterior crossbite and improve the shape of the upper arch.

The patient is undergoing orthodontic treatment and follow-up, waiting the opportune moment for the surgical treatment with extractions of supernumerary teeth.

Discussion

Cleidocranial Dysplasia represents pathology of extreme importance for the dentist, especially for the specialist in Pediatric Dentistry. Knowledge of developmental disorders involving cranio-dento-facial structures is very important, because are the main reason for dental consultations, evidencing the relevance of the dentist to the diagnosis of this condition.⁵ In the present case, it was possible to observe pathognomonic features, like the absence of clavicles, broad skull sutures and many supernumerary teeth, localized in

maxilla and mandible.

Cleidocranial dysplasia should be considered during all phases of diagnosis, planning and treatment of the patient, it is still a debatable subject among professionals.¹ The treatment of the stomatognathic system changes seeks the elaboration of an interdisciplinary and complete therapeutic planning, with functional and aesthetic oral rehabilitation. The treatment objective in our case was an initial approach in dentistry (Figure 3), considering both the physical and psychological aspects of our patient.

What should not be overlooked is the harmonization of

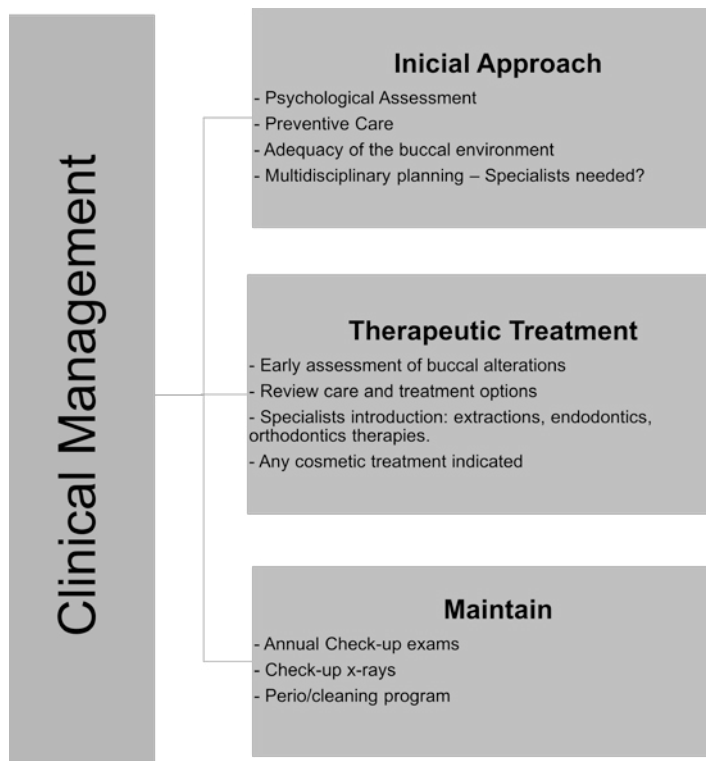


Figure 3. Clinical management of DCC pediatric patient

the patient as a whole, prior to the therapeutic and preventive treatment - be it orthodontic or surgical. Biopsychosocial care should be attend, which involves balancing the patient's health, the adequacy of the buccal environment and changing life habits.¹

The present study, after preventive care in an initial approach, consulted professionals from different specialties to build the patient's treatment plan, considering their expectations and the family, the biological needs and the timing of the treatments.

Regarding the dental treatment of patients with CCD much has been discussed about, but there is a consensus that the intervention is crucial to the success of the treatment, and the sooner the patient with CCD is diagnosed and initially approached, better results are achieved.⁶

However, it is not always easy to determine the most sui-

table aesthetic-functional treatment for each patient with CCD. The timing of diagnosis can also be suggested as a guide for choosing the treatment model.⁸ Thus, the earlier is the intervention, and the greater is the possibility of reduction of surgical and orthodontic extension.

Some studies suggest the extraction of all deciduous teeth and the following osteotomy to allow the spontaneous eruption of permanent teeth,^{7,9,10} which differs from the option followed in this clinical report. In particular, what had considered in this case was the priority of the adequacy of the buccal environment, with aesthetic and functional rehabilitations. As well as the need of previous orthodontic treatment, so that the retained permanent teeth can rupture in a healthy buccal environment. Even that a future ortho-surgical traction will be still necessary.

It is evident that the most favorable period for treatment is dependent on the degree of root development and amount of bone present, which often means performing procedures even during childhood.^{6,8} This fact determines the fundamental role of pediatric dentistry, as it will be the professional trained to deal with the biological characteristics of childhood, in time, as well as psychological, of the patient's behavioral conditioning. Even so, in the scientific literature there are few articles referring to this pediatric approach in patients with CCD.

Conclusion

In view of the complexity of the Cleidocranial Dysplasia dental treatment, it is necessary the interdisciplinary planning among dentistry areas. The Pediatric Dentistry is the first specialty to approach the patient. It is responsible of the clinical success, which should be based on the therapeutic trilogy of preventive, functional and aesthetic goals. Besides of it, the pediatric dentistry is the interface of psychological conditioning to enable dental treatment in the different specialties.

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Mini Curriculum and Author's Contribution

1. Milena Sierra Drumond da Costa – DDS. Contribution: performed the experimental procedures, bibliographical research and wrote the manuscript.
 2. Rafael Celestino Colombo de Souza – DDS and PhD. Contribution: contributed substantially to discussion, bibliographical research, image editing, proofread the manuscript.
 3. Sanny Bozza Silvestri – DDS. Contribution: performed the experimental procedures.
 4. Ana Paula Carvalho – DDS. Contribution: bibliographical research, experimental procedures.
 5. Thaís Gimenez – DDS. Contribution: bibliographical research and manuscript writing.
 6. José Carlos Pettorossi Imparato – DDS and PhD. Contribution: experimental design, work supervisor.
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