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THE IMPORTANCE OF ADEQUATE DENTAL MANAGEMENT TO THE PATIENT WITH CLEIDOCRANIAL DYSOSTOSIS: CASE REPORT

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ABSTRACT

Cleidocranial Dysostosis (CCD) is characterized as a rare autosomal dominant genetic syndrome with no gender preference, characterized by a defect in the gene CBFA1 of chromosome 6p21, which modulate the formation of osteoblasts. Its clinical characteristics are clavicle aplasia or hypoplasia, changes in skeletal level with influence on the anatomical structure of the wearer and polydontia. There is no specific treatment, and the main conduct is to offer the best possible quality of life to the bearer, and this involves oral health. In this context, dentistry has an important role, mainly in what concerns the maintenance of the developed dental organs besides the physiological quantity (polydontia) due to the dysfunction. Thus, the present article aims to present the main characteristics of the DCC and report a case in which dental care was important for the improvement of the quality of life of the bearer.

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INTRODUCTION

Cleidocranial Dysostosis (CCD) was described by Pierre Marie and Paul Sainton in 1898 (Marie, Sainton, 1898; Esquerro, Lopez, 2011) and is characterized as a rare congenital syndrome, autosomal dominant, with no predilection for race or gender, in which a defect in the gene CBFA1 of the chromosome 6p21, also described as RUNX2 (Kuczyk *et al.,* 2018), is observed, that modulate the formation of osteoblasts from mesenchymal stem cells and, consequently, promote changes in bone, in addition to influencing the development of dental organs.

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The diagnosis is based on physical and radiographic characteristics based on the identification of supernumerary teeth, aplasia or hypoplasia of clavicle and sagittal suture or open fontanelles (Araya *et al.,* 2011; Almeida Júnior *et al.,* 2012). There is a hereditary bias, characterized by the passage of the gene from generation to generation (Roberts *et al.,* 2013), being common, more members of the same family present the syndrome or characteristics of this, which can be evidenced by the construction of a heredogram (Porciuncula *et al.,* 2013). It is observed in the holder of the DCC, clavicle aplasia or hypoplasia, changes in bone level and dental anomalies (Bir *et al.,* 2017). These manifestations together influence the configuration of limbs and the height of the wearer.

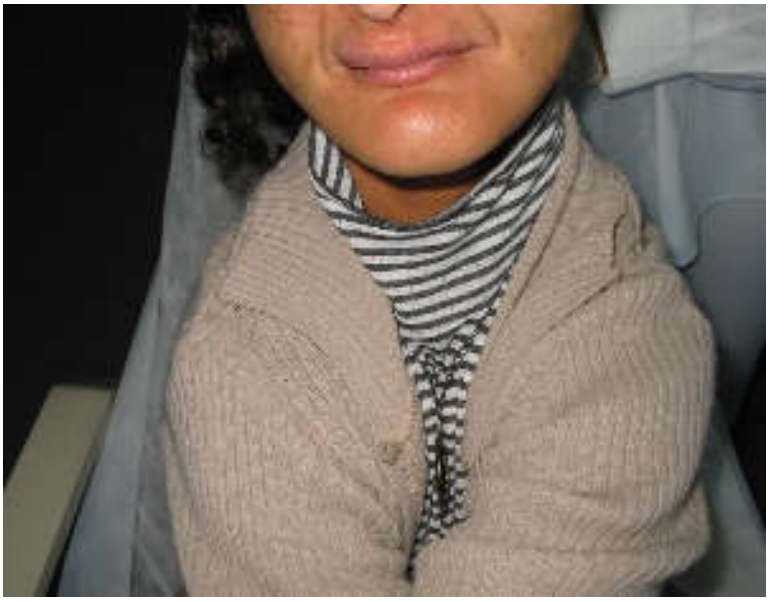

Fig. 1. Aspect of shoulder hyperlaxity in clavicle aplasia factor

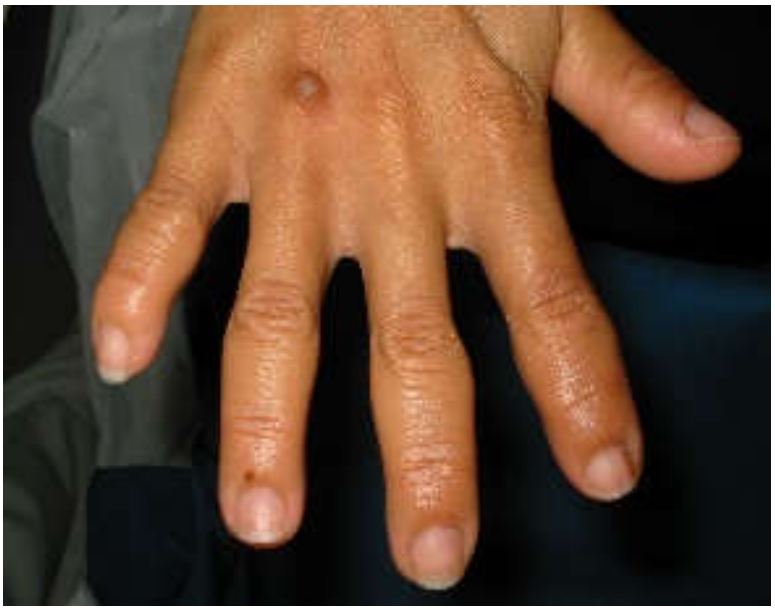

Fig. 2. Aspect of the patient's hand evidencing alteration in the bone constitution

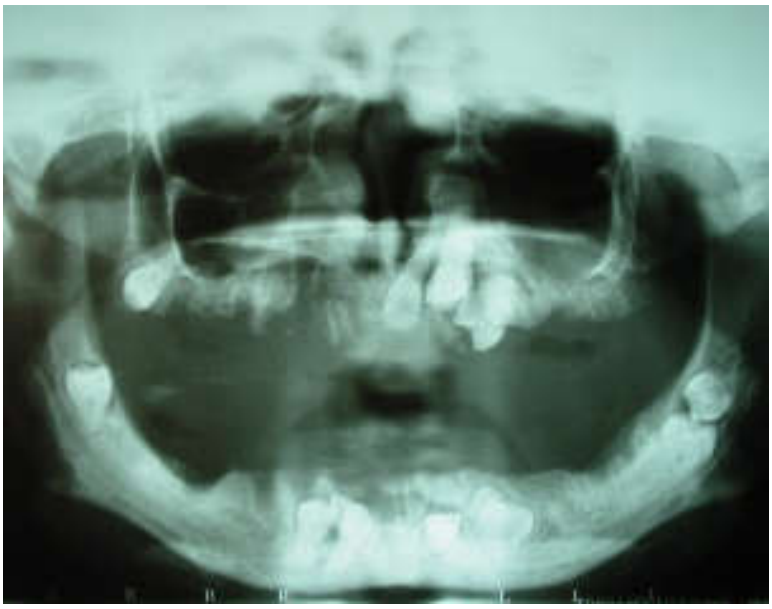

Fig. 3. Panoramic X-Ray. Note the supernumerary teeth in the maxilla and mandible

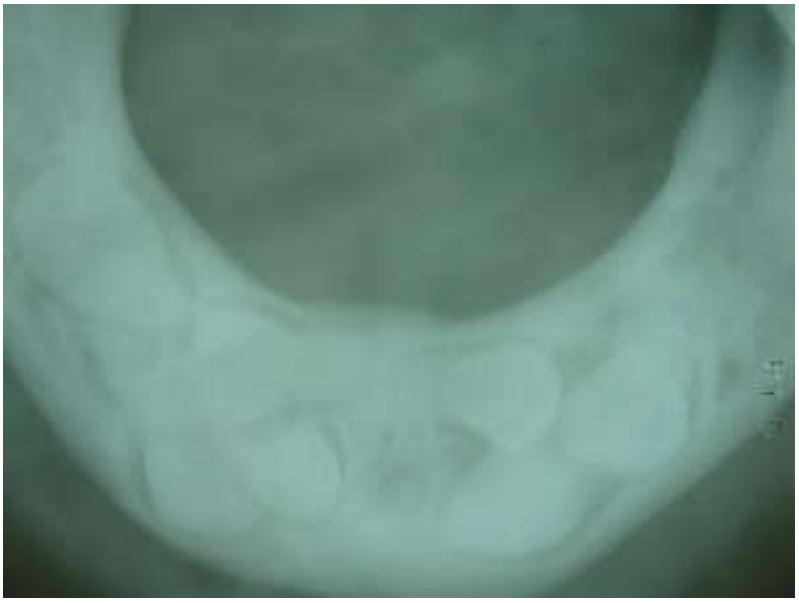

Fig. 4. Occlusal X-Ray of mandible. Note the distribution of supernumerary teeth

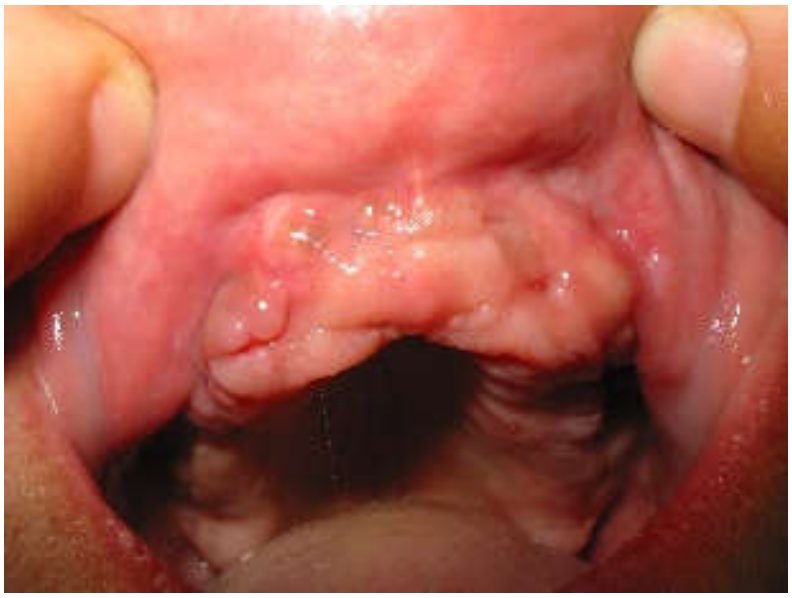

Fig. 5. Intraoral buccal aspect of the maxilla. Area that was initially made the exodontia of erupted teeth

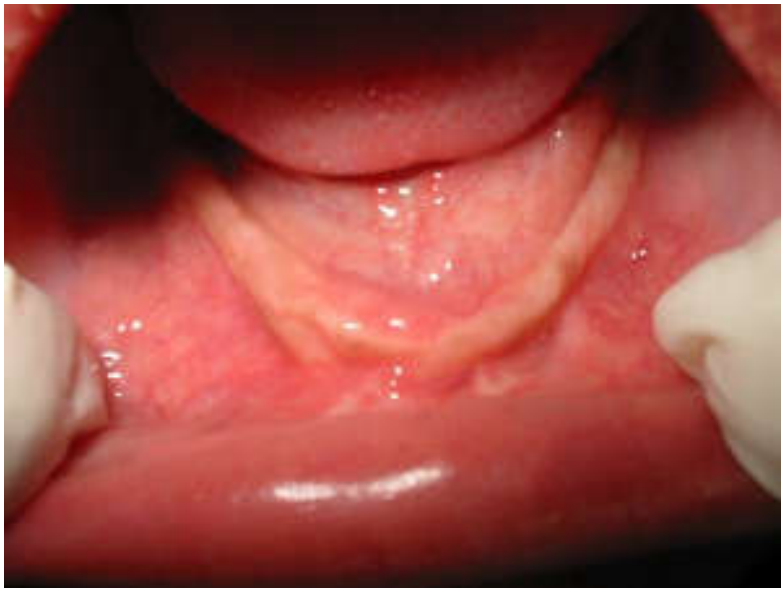

Fig. 6. Intra-buccal clinical aspect of the mandible suggesting edentulism, however, with supernumerary teeth included

Clinically, short fingers, narrow hands, altered weight / height relationship, hypertrichosis and hypertelorism (Araya *et al.,* 2011), as well as changes in the genesis of the scapular and sternocleidomastoid musculature can be observed (Rodrígues *et al.,* 2002). In relation to dental aspects, the most expressive feature of this syndrome is the supernumerary formation of teeth, that is, in a permanent physiological dentition, there are 32 teeth, however, in a person with CCD, this number may be higher, characterizing a (Santos *et al.,* 2016), in addition to maxillary hypoplasia (Kuczyk *et al.,* 2018). There is no specific treatment for CCD, and interventions are performed according to the signs and symptoms developed, however, there is a need for continuous monitoring, since changes in bone level may compromise motor functions in the long term. Regarding oral health, supernumerary teeth may interfere with the function of the stomatognathic system, especially with regard to masticatory function. Dental follow-up may include the extraction of all teeth with posterior prosthesis, as well as the management of the deciduous dentition, which may present eruptive retardation (Seow, Hertzberg, 1995) and supernumerary teeth through orthodontic interventions. In relation to implant installation, there are still few research studies in this specific group of patients (Santos *et al.,* 2016). Preventive dental maintenance, as early as possible, is important, since teeth that erupt ectopically or late may compromise occlusal harmony. Another important point refers to the fact that tooth loss due to caries, periodontal disease or other factors may predispose the individual to exposure of the supernumerary teeth due to masticatory efforts, as well as to negatively influence the positioning of dental prostheses. Thus, it is understood that the planning of dental actions during the life of the CCD patient is of great importance (Tripathi *et al.,* 2012). Therefore, the present article aims to present the main characteristics of the DCC and report a case in which dental care was important to improve the quality of life of the patient.

CASE REPORT

Patient N.A.S., female, 38 years old, feoderma, was welcomed with the previous history of having sought dental care for the extraction of all teeth, in order to install total superior and inferior dentures. The exudation of the clinically visible (erupted) teeth was performed without previous X-Ray, and the professional performed anatomical deviations during the execution of the procedure and requested specialized evaluation in the stomatology clinic. In the specialized evaluation, the first course was to request a panoramic X-Ray, which showed the presence of 12 supernumerary teeth included in the mandible and 5 supernumerary teeth included in the maxilla. As a complement, a mandibular occlusal radiograph was taken due to the reduced bone thickness. In view of the condition, the partial extraction of the teeth included in the mandible was performed, due to the risk of bone fragility and consequent fracture, and in the maxilla, the extraction of all dental elements was chosen. After adjustment of the gingival roller, the patient was referred for prosthetic rehabilitation. Regarding the general anatomical aspects of the patient, clavicular aplasia was observed, with hyperlaxity of the shoulders, bone hypo development of the hands, short stature and hypertrichosis. In relation to the intra-buccal aspects, ogival palate was observed, gingival roller with absence of clinically visible teeth and hypodevelopment of the mandibular bone. Due to the neurocognitive aspects, no type of alteration was evidenced. Regarding the patient's self-perception regarding CCD, she reported that she knew about the diagnosis, but had not been guided about odontological care and regarding heredity, could not know if there were more family members with the same syndrome or with characteristics similar to it.

DISCUSSION

The CCD, because it is characterized as a rare congenital syndrome, still presents a great challenge for the health sciences, especially regarding the aspects of maintenance of the bone structures in development and the polydontia framework. The dental management in question is of great importance and the diagnosis of the syndrome should be well defined, especially by the dentist who has elementary knowledge about CCD, especially in functions of the visual clinical aspect of shoulder hyperlaxity as a factor of clavicle aplasia. The identification, as early as possible, of supernumerary teeth (quantities and location) is essential for planning actions that aim at improving the quality of life of patients with CCD, since there is often a need for programmed exodontitis (Santos *et al.,* 2016), which aims to improve the oral environment in relation to masticatory functions, but all based on the radiographic diagnosis by the periapical, occlusal and or panoramic techniques as complementary exams, as a support to determine the therapeutic behaviors for each situation. In addition, there may be delayed eruption of teeth in relation to physiological chronology, as a result of supernumerary ectopic teeth that may prevent the eruption of other teeth in position (Araya *et al.,* 2011). Another important aspect is related to the psychosocial factors, since the physical changes are visible and the inclusion of this individual in the society can be difficult, therefore, the management of the oral health can be characterized as a positive point to minimize prejudices, since the smile, in itself, constitutes a great motivating, aggregating and affective factor. It should be emphasized that oral health is an inseparable factor of the human organism and, therefore, must be inserted in the context of follow-up of patients with CCD from the first years of life and thus, the dental surgeon can work preventive actions and, when necessary and timely, to develop clinical intervention actions within the universe of dentistry specialties.

Conclusion

In view of the above, it can be concluded that DCC deserves multidisciplinary attention as early as possible, since the maintenance of anatomical deviations and the monitoring of potential occurrences during the life cycle constitute important actions for the improvement of the individual's quality of life.
The odontologicsactions should be considered from the first years of life, so that the oral health professional can work preventive actions and clinical intervention, having as principles, the need and the opportunity to act, according to the individual development of the segment and of the systemic conditions inherent in the chronological progression of CCD.

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