

Introduction:

Cardiofaciocutaneous syndrome (CFC), is a genetically heterogeneous syndrome and results from mutations in the Ras/mitogen-activated protein kinase (MAPK) signaling pathway, which affects the receptor tyrosine kinase (RTK), plays a role in the craniofacial and dental development. CFC syndrome is a rare disorder characterized by craniofacial dysmorphology, intellectual disability, congenital heart disease, growth retardation, dermatological and neurologic anomalies. Children with CFC require multidisciplinary care from specialists and comprehensive management to achieve a better life prognosis.

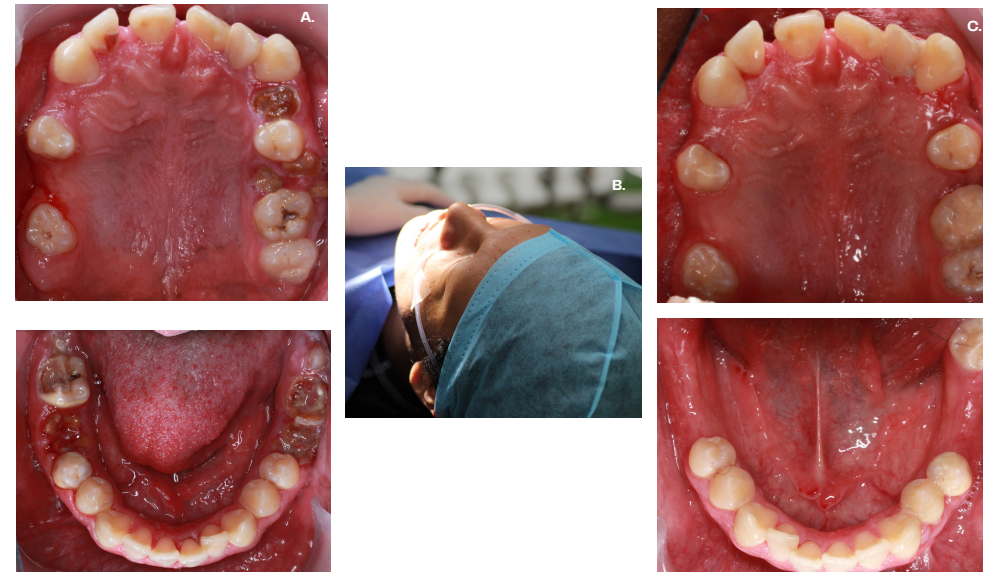


Figure. 2 Initial intraoral photographs and control after 1 year of the rehabilitation. In figure C we can observe a radical change in habits that promote oral health.

Cone Beam Features:

General crestal bone loss, retained #5, multiple supernumerary teeth, periapical lesion in #7, 19, 30. (Fig. 3)

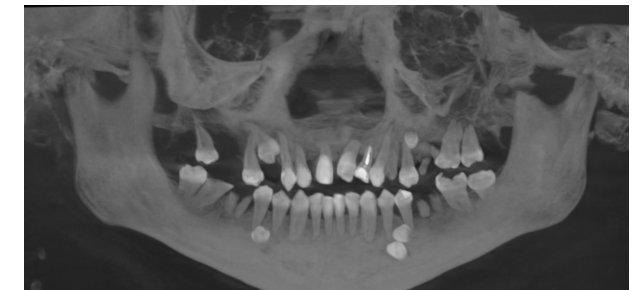


Figure 3: Initial Cone Beam of the year 2022.

Treatment:

From previous medical evaluation, prevention habits were instructed to the patient's caregivers, and oral rehabilitation was performed under 2 first interventions with conscious sedation. In the first intervention, sedation was required, during which the treatments were an indirect pulp therapy and restoration with a hybrid glass ionomer in #15, and endodontic treatment in #7. In the next intervention, the extraction of radicular remains and supernumerary teeth was performed under conscious sedation. In further interventions, conscious sedation was not required for the composite restoration in #1, 7, 15, 20, 26 and incisal reduction of #8 and 25, to reduce dental trauma (Fig. 2)

Conclusion:

Management of patients with CFC is difficult since the intervention of health specialists is vital to assure their safety. Also is necessary to achieve motivation of the caregivers and establish special oral care techniques to promote the maintenance of oral health and encourage regular dental visits.

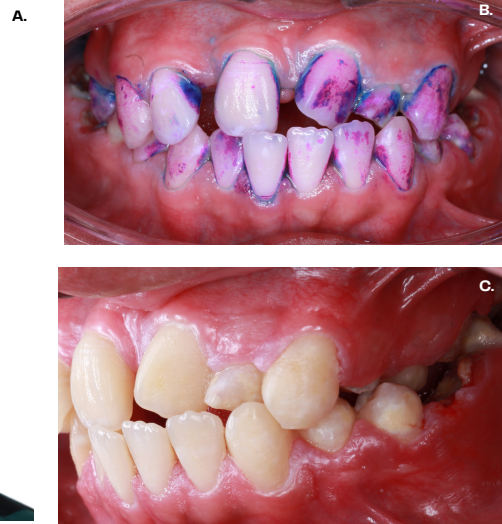


Figure. 1 Extraoral photograph where we can observe clinical features of CFC syndrome, in Figure B the initial plaque control with mature biofilm, and Figure A dental anterior and posterior crossbite.

Case Report:

Seventeen-year-old male presented to the pediatric clinic, accompanied by his parents, who noticed that their son had dental pain for several months. His medical history includes a diagnosis of CFC, intellectual disability, and congenital hypothyroidism which is currently managed with daily levothyroxine of 150 mg. Clinical examination revealed poor oral hygiene, anterior and posterior crossbite, periodontitis, existing restorations, MIH, active carious lesions in multiple teeth, radicular remains of #12, 14, 19, 30, 31, semi erupted lower third molars and missing #1, 3 and 5. (Fig. 1, 2)