Dental Management of Patient with Cardio-Facio-Cutaneous Syndrome Under General Anesthesia: A Case Report

Fatemeh Golzadeh, Melba Esquilin

University of Puerto Rico, Medical Science Campus-School of Dental Medicine
San Juan, PR

Introduction

Cardio-Facio-Cutaneous Syndrome (CFC Syndrome) is an uncommon genetic condition, affecting approximately 1 in 810,000 people. It is characterized by autosomal dominant inheritance, arising from mutations in various genes that play crucial roles in cell signaling pathways crucial for development and growth.[2] Those with CFC Syndrome often exhibit congenital heart anomalies, such as pulmonary valve stenosis, hypertrophic cardiomyopathy, and atrial septal defects, alongside distinctive facial features including a high forehead, wide-set eyes (hypertelorism), crossed eyes (strabismus), rapid involuntary eye movements (nystagmus), drooping eyelids (ptosis), a short nose, low-set ears, a prominent chin, and hair that may be sparse, curly, fine, or thick. Eyelashes and eyebrows might also be absent or sparse. Frequently, individuals experience developmental delays in physical, motor, and cognitive areas. Severe feeding and swallowing difficulties are common, potentially leading to inadequate weight gain and growth failure in infancy. Seizures are also a possibility and may be difficult to manage with treatment.[3] Recognizing the physical and health challenges faced by these individuals is critical for providing comprehensive dental care, especially for those with significant dental decay and a high risk of caries.

Case Report

A 9-years-old male patient, weighing 17.6 kg, actively receiving care at a pediatric dental clinic in a hospital setting, presents with a past medical history of Cardio-Facio-Cutaneous syndrome. This history includes clinical manifestations such as seizures, hearing loss, and intellectual disabilities. Currently, the patient is on a medication regimen that includes 7ml of Keppra and 5ml of Trileptal. There are no reported food or drug allergies. Due to generalized muscle weakness, he is wheelchair-bound and incapable of walking. Clinical examination has identified multiple dental caries, gingivitis, and enamel defects.

Physicals Characteristics :[1]

Facial features: Large forehead, head microcephaly and right occipital plagiocephaly, ears with overfolded helix and prominent anti-helix. Maxillary, ethmoid, and sphenoid sinus mucosal thickening.

Chest: Protuberant, **abdomen**: scaphoid and **Joint:** contractures.

Skin and hair: Skin thickened (hyperkeratotic), eczema (extreme dryness of skin and Wrinkled skin, hair sparse, brittle and eyebrow sparse.

Eye: wide-spaced (hypertelorism), strabismus, nystagmus, down-slanting and ptosis (droopy eyelid).

Heart: Pulmonarias stenosis, atrial septal defects, ventricular septal defects, with regular rate and rhythm. EEG presented abnormal.

Lungs: Clear and no clinical evidence of visceromegaly.

Feeding/ Gastrointestinal (GI): Difficulty feeding, failure to thrive, reflux, vomiting, intestine malrotation, hernia and constipation.

Neurologic: Low muscle tone and strength with gross motor delay and no speech.

Brain MRI: Callosal agenesis with bilateral colpocephaly and temporal horn dilatation, hippocampal atrophy, lissencephaly and midbrain hypoplasia.

Craniofacial Features: Convex facial profile, posterior crossbite.

Preoperative Management

Preoperative:

- A multidisciplinary team consisting of Pediatrician, Genetics, Cardiologist, Gastroenterologist, Neurologist were consulted and gave recommendations and clearance to do dental rehabilitation under general anesthesia.
- Cardiologist: IV dextrose 5% recommended to avoid hypoglycemia and avoidance of medications that may cause QTc prolongation.
- Laboratory result presented without complications.

General Anesthesia Management

Premedication:

- 1.5ml Versed (0.5mg/1ml) used 20 min before starting the anesthesia to help that patient be relax, fall sleep and blook memory before a surgical procedure.

General anesthesia:

- Intubation could not be done through intranasal and was achieved through oral route.
- General anesthesia was done with 80mg Propofol (10mg/ml). It used to control QT interval to avoid affecting heart rhythms and sudden cardiac arrest.
- Dextrose 0.5% used to treat hypoglycemia.
- Fentanyl 25 mcg used to relieve severe pain during and after surgery.
- Decadron 2mcg used to reduce the risk of nausea, vomiting and swelling of the oral tissue.

Clinical Examination

Fig 1: Patient physical Picture[1]

Fig 2: pretreatment intraoral Picture, A)Maxillary view, B)Mandibular view

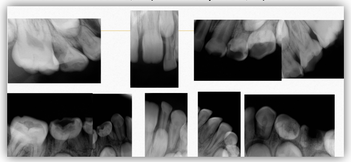

Fig 3: Preoperative Radiographies

Dental Treatment Result

Operative:

- A Full mouth X-Ray was taken. Oral exam achieved and patient presented multiples dental caries.
- Oral prophylaxis and ultrasonics scaling was performed on the patient. Dental caries was removed on teeth #B(O), #H(F), #19(O), #30(O) and restoration was done with resin composite#A2.
- Local anesthesia was not use for the extractions due to the patient's medical conditions. Extraction was done on teeth #A, #I, #J, #K, #L, #S, #T. Gelform was placed, and hemostasis achieved. Sutures were made with Polysorb braided absorbable size (4-0).
- Sealant was placed on teeth #3 and #14. Finally, Sodium fluoride varnish 5% was applied.

Post operative:

- The team recommended the patient to be admitted to the ICU for one day for postoperative monitoring.

Conclusion

- Clinicians are urged to carefully consider and swiftly address potential medical and surgical complications, aiming to minimize adverse outcomes through tailored strategies such as thorough preoperative evaluations, diligent intraoperative monitoring, and immediate interventions when necessary. This proactive approach is essential for risk reduction and the enhancement of patient outcomes.
- Furthermore, the early adoption of a comprehensive, personalized preventive care strategy can significantly lower the risk of dental caries development and, consequently, reduce the necessity for surgical interventions, such as undergoing dental procedures in an operating room.
- This preventive approach underscores the importance of early intervention and personalized care in maintaining oral health.

References

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