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Case Report

Dental aspects of Crouzon syndrome: a case report

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ABSTRACT

Crouzon syndrome is a rare genetic disorder characterized by the premature fusion of certain sutures of the cranial vault, base, orbital, and maxillary region, leading to craniofacial dysmorphology. The severity of craniosynostosis varies among individuals. The clinical presentation of this congenital deformity depends on the pattern and timing of sutural fusion. Crouzon syndrome is caused by a mutation in genes that control bone development, particularly the fibroblast growth factor receptor 2 (FGFR2) gene. This report highlights the varied clinical presentation of a 13-year-old patient with Crouzon Syndrome, featuring the craniofacial manifestations, associated dental characteristics, and the benefit of a multidisciplinary rehabilitation approach focusing on management of the condition in the dental context.

Keywords: Crouzon Syndrome, Craniosynostosis, FGFR2

INTRODUCTION

Craniofacial development is a complex and highly coordinated process occurring exclusively under genetic control and environmental influence. The blueprint for building the human head and face is established early during embryonic development.[1] This process is crucial for proper development, and any Gene perturbations can lead to cranio-skeletal malformations.[2] Craniosynostosis is a disorder that involves the premature fusion of one or more cranial vault sutures before the brain is fully formed. Brain growth continues, giving the head a misshapen appearance. Overall, all forms of craniosynostosis affect approximately 1 in 2,000 to 2,500 live births.[3]

Crouzon syndrome, an autosomal dominant disorder, was first described in 1912 by a French physician Octave Crouzon when he identified both a mother and daughter with what was initially called "craniofacial dysostosis." He described a triad of skull deformities, facial anomalies, and proptosis. This triad of findings was then re-labelled "Crouzon syndrome." This affects both males and females and is the most common craniosynostosis syndrome. It occurs in approximately 1.6 per 100,000 people.[4]

Etiopathogenesis includes mutations in the FGFR2 gene, which likely overstimulate signalling by the FGFR2 protein, causing the skull bones to fuse prematurely. Once there is premature synostosis of coronal and sagittal sutures, the growth potential of those sutures is restricted.[5] When premature fusion of sutures occurs, maxillofacial deformities follow, requiring special attention. A defining characteristic of Crouzon syndrome is craniosynostosis, which results in an abnormal head.[4]

Crouzon syndrome affects coronal, sagittal, or lambdoid sutures. Key features include midface hypoplasia, a beaked nose, exophthalmos, hypertelorism, and cervical vertebral fusion. Additional features include thin vermilion of the upper lip and mandibular prognathism. Some patients can present with cleft lip or palate. These patients typically have normal intelligence.[4] CS is distinguishable from other craniosynostosis syndromes by the lack of hand and/or foot abnormalities.[6] Exclusive

eye abnormalities include Exophthalmos (bulging eyes), Proptosis, Hypertelorism (widely spaced eyes), and External strabismus. Sometimes leads to a loss of vision.[7]

Intraoral manifestations include dental crowding and a V-shaped maxillary dental arch. Narrow, high, or cleft palate and bifid uvula can also be seen. Least common findings like oligodontia, macrodontia, peg-shaped, and widely spaced teeth have also been reported.[8]

The diagnosis of Crouzon syndrome is predominantly clinical, with genetic testing and radiological imaging providing additional support. It is usually diagnosed clinically based on the appearance of the child's face and skull. A craniofacial surgeon and geneticist can confirm the diagnosis. Imaging studies such as CT scans or MRI are necessary to assess craniofacial dysmorphology and molecular genetic testing to detect FGFR2 gene mutations.[9]

Management varies based on the patient's age and the severity of the disorder. Optimal treatment planning involves early diagnosis and a multidisciplinary approach. Ideally, the release of prematurely fused sutures should be performed by a neurosurgeon within the first year of life (between 3 to 6 months) to ensure adequate cranial volume for brain growth and expansion. Ophthalmologists play a crucial role in evaluating and monitoring blindness caused by optic atrophy.[7]

Dental professionals address malocclusion and other dental issues. Maxillary hypoplasia and anterior crossbite can be corrected using rapid maxillary expansion, while maxillary protraction can be achieved with a facemask or distraction osteogenesis. Early orthodontic management is essential to prevent severe skeletal discrepancies and may be supplemented with rhinoplasty, genioplasty, and bone grafting.[10] In this context, the dentist plays a fundamental role in the oral rehabilitation of these patients, promoting improvements in aesthetics and quality of life.

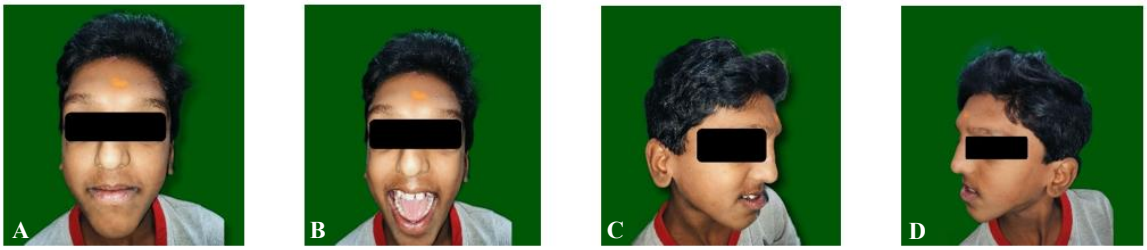
CASE REPORT

A 13-year-old boy reported to the Department of Pediatric and Preventive Dentistry with a chief complaint of crowding in his upper teeth. Due to the boy's unusual facial appearance, a detailed family and medical history was obtained. His prenatal, natal, and postnatal history were unremarkable, with no reported family abnormalities. However, his parents, though a non-consanguineous couple, presented with an advanced paternal age. The child had attained developmental milestones in the normal range and had normal intelligence but had minor speech difficulties. No anomalies were reported among his siblings or close relatives. The patient did not present with any digit abnormalities or hearing defects. Behaviourally, the patient was slightly anxious, had an interactive nature, and was cooperative during all the visits.

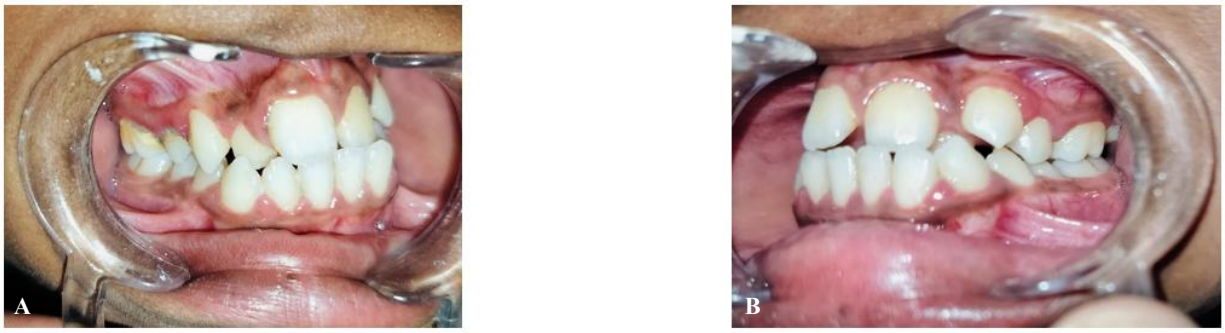
On extraoral examination, the patient presented with proptosis of the eyes, down slanting palpebral fissures, strabismus, hypertelorism, maxillary hypoplasia associated with pseudoprognathism of the mandible, incompetent lips, and a thin vermilion of upper lip (Figure 1). Intraoral examination revealed a V-shaped maxillary arch with a high-arched palate, all characteristic features of this syndrome. Occlusal examination revealed Angle's Class II Division I malocclusion, anterior crowding, anterior and posterior crossbite, mesiodens, and ectopically erupted maxillary left and right second premolars (Figure 3). The chronology and eruption status of the teeth were typical for the child's age.

Investigations included radiographic, hematological, and systemic evaluation. An orthopantomogram (OPG) revealed normal dental age and development; condyles appeared normal, with no signs of ankylosis (Figure 4B). The lateral cephalogram revealed maxillary hypoplasia and a shallow orbit (Figure 4C). Routine hematological (CBP, CT, BT) and biochemical tests were within normal limits. Even though the patient didn't have signs and symptoms related to vision and neurological issues, the expert opinion of an ophthalmologist and a neurosurgeon was sought to evaluate for the same.

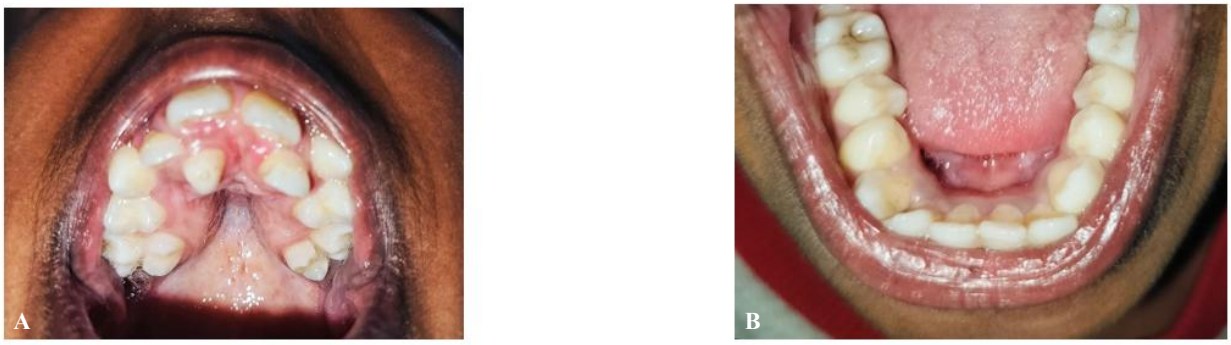
It was identified that the patient had Crouzon Syndrome (CS), which affects the craniofacial structure and impacts the teeth. The patient displayed the characteristic triad of Crouzon Syndrome, including skull deformities, facial anomalies, and proptosis. Additional features like brachycephaly, wide-set (hypertelorism) bulging eyes, a flattened forehead, a beaked nose, and an underdeveloped upper jaw (maxillary/midface hypoplasia) that defines the Crouzonoid face were also evident in this patient.



Figures 1: (A and B) Frontal view of the face; (C and D) lateral view showing hypoplasia of the maxilla associated with pseudoprognathism of mandible, beaked nose, incompetent lips and thin vermilion of upper lip.



Figures 2 (A and B): Angles class II div I malocclusion with anterior and posterior crossbite.



Figures 3: (A) V-shaped Maxillary arch with high arched palate, crowding in anterior teeth, mesiodens, palatally erupted maxillary left and right second premolars; (B) occlusal view of mandibular arch.

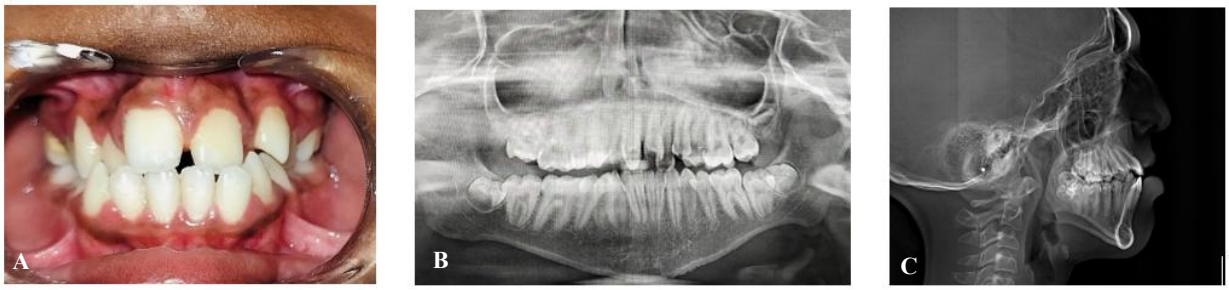


Figure 4: (A) Occlusion showing anterior crossbite, (B) orthopantomogram showing maxillary teeth crowding and meOodens; (C) lateral cephalogram showing hypoplastic maxilla.

The parents were counselled regarding the nature of the disorder. As part of dental management, the decision to perform comprehensive dental treatment, including surgical extraction of supernumerary and ectopically erupted teeth followed by functional orthodontics needed was explained to the parents. Additionally, the parent signed the Informed Consent Form (ICF) and granted authorization for the use of data, and images. The child was managed using different non-invasive behavioural techniques, including conditioning, gradual desensitization, and positive reinforcement. Based on the patient's chief complaint and his oral conditions, the following treatment procedures were carried out in sequence:

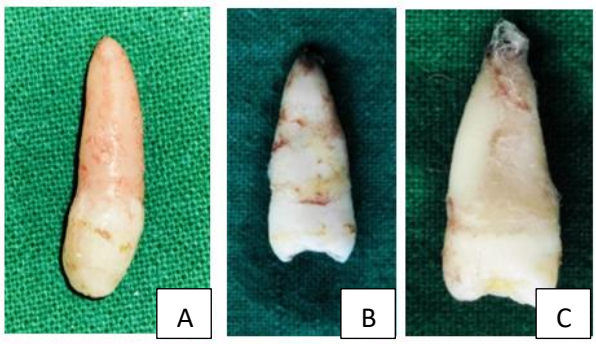


Figure 5 (A-C): Extracted mesiodens tooth, extracted maxillary left and right second premolars

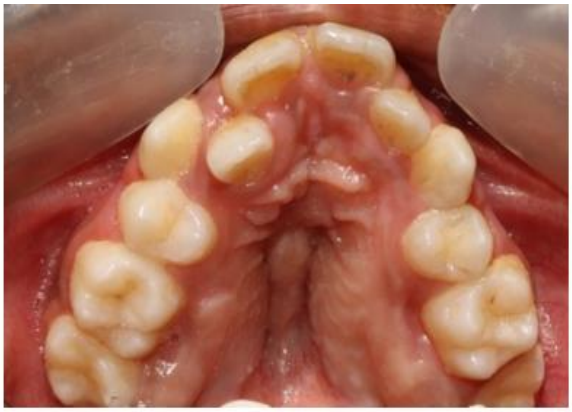


Figure 6: Maxillary arch after 1 month follow-up and before orthodontic treatment.

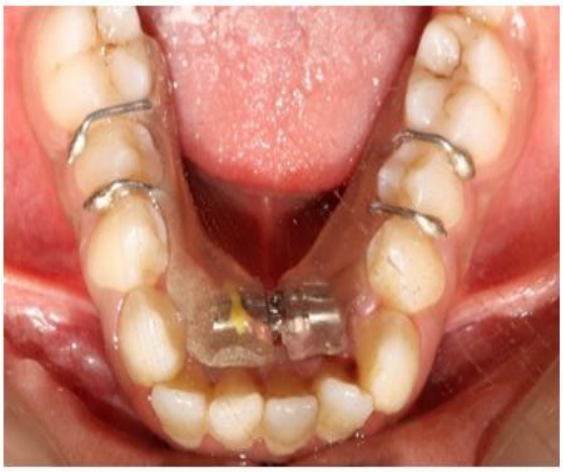


Figure 7: Schwartz expander for the lower arch.

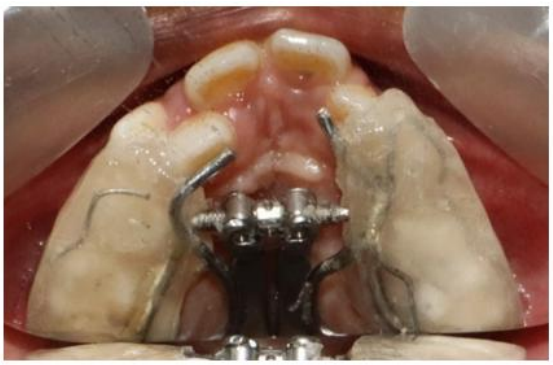


Figure 8: Fan-shaped Hyrax appliance with a posterior bite plane for the upper arch.

Oral Prophylaxis and Oral Hygiene instructions; Pit and fissure sealants for tooth #36 and #46; Extraction of Mesiodens under Local Anaesthesia (Figure 5A); Bilateral extraction of ectopically erupted maxillary second premolars in two visits. (Figure 5B, C); Orthodontic and dentofacial orthopedic treatment was recommended, which included the correction of anterior and posterior crossbite using rapid maxillary expansion and reverse pull headgear. The patient who is currently undergoing orthodontic treatment with a fan-shaped Hyrax appliance with a posterior bite plane for the upper arch (Figure 8) and a Schwartz expander for the lower arch (Figure 7) would be followed by fixed orthodontic appliance therapy to achieve proper levelling and alignment of the dentition.

DISCUSSION

Crouzon syndrome is typically diagnosed in early childhood due to its characteristic craniofacial anomalies. However, there are exceptional circumstances where this syndrome may be diagnosed later, when craniofacial features become apparent during late childhood or due to the onset of secondary complications, such as dental or orthodontic problems, prompting thorough medical evaluation.[11] The current case also reported concerns mainly related to dental abnormalities.

The manifestations of CS may vary in severity from a mild presentation with just midface hypoplasia to severe forms, including early fusion of multiple cranial sutures and varying patterns of suture fusion.[8] The patient presented here exhibited atypical craniofacial anomalies like maxillary hypoplasia, hypertelorism, and maxillofacial dysmorphology. These distinctive features, though visible, were not detected early in their development.

Advanced paternal age may be associated with a higher risk for certain single-gene disorders caused by mutations of the FGFR2, FGFR3 and RET genes.[11] These conditions include Apert syndrome, Crouzon syndrome, and Pfeiffer syndrome among others.[12] The paternal age effect may have been reflected in the present case as the father's advanced age might have contributed to the biological effect on the child. CS is distinguishable from other craniosynostosis syndromes like Apert and Pfeiffer by the lack of hand and/or foot abnormalities. These syndromes share overlapping features but have distinct skeletal and limb anomalies.[6] The present CS case did not present with hand or foot anomalies.

Diagnosis is primarily clinical, supported by imaging techniques such as CT scans, which reveal a "beaten copper" skull appearance, and MRI to assess intracranial abnormalities.[6] Prenatal genetic testing for FGFR2 mutations is an option for at-risk families.[13]

Treatment protocols vary depending on the age, signs, symptoms, and severity of the condition in a particular patient. Orthodontic evaluation plays a crucial role in the early and accurate detection of late-onset forms of Crouzon syndrome, even in cases where the disorder may not have been identified during early childhood by routine medical evaluation.[10]

CS requires multidisciplinary treatment, including dental care, and the clinician must be aware of the causes and

limitations of the condition to recognize and adequately treat the patient. Coordination among pediatric dentists, orthodontists, and psychologists is vital to provide a comprehensive approach to the patient. Continuous coordination among team members is essential to plan and execute appropriate interventions, ensuring the best possible outcome for the patient.[11]

The prognosis largely depends on the severity of malformations and timely intervention. Long-term follow-up is necessary to monitor skull growth, dental development, and vision. With early intervention and coordinated care, children with Crouzon syndrome can achieve improved functionality, facial aesthetics, and quality of life.[4]

CONCLUSION

Genetic syndromes substantially impact the self-image and psychosocial prognosis of affected individuals. Timely intervention can significantly improve the patient's quality of life by addressing the maxillofacial deformities and preventing associated complications. Collaboration among healthcare professionals is essential for optimizing treatment outcomes and providing comprehensive care.

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