

Early orthodontic management of Crouzon Syndrome: a case report

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Received: 02 February 2009 / Accepted: 02 March 2009
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Abstract Crouzon Syndrome is an autosomal dominant disorder with complete penetrance and variable expressivity. Described by a French neurosurgeon in 1912, it is a rare genetic disorder. Crouzon syndrome is caused by mutation in the fibroblast growth factor receptor 2 (FGFR2) gene. The disease is characterized by premature synostosis of coronal and sagittal sutures which begins in the first year of life. Once the sutures become closed, growth potential to those sutures is restricted. However, multiple sutural synostoses frequently extend to premature fusion of skull base causing midfacial hypoplasia, shallow orbit, maxillary hypoplasia and occasional upper airway obstruction.

The case of a 7-year-old South African black boy with Crouzon Syndrome is presented. He presented with characteristic triad of cranial deformity, maxillary hypoplasia and exophthalmos. The clinical, cephalometric features and initial orthodontic management of this patient are discussed as part of multidisciplinary management.

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Keywords Craniosynostosis • Anterior crossbite • Fibroblast growth factor

Introduction

Crouzon Syndrome is an autosomal dominant disorder with complete penetrance and variable expressivity or can appear as a mutation [1]. Described by a French neurosurgeon in 1912 [2], it is a rare genetic disorder. It may be transmitted as an autosomal dominant genetic condition

Crouzon syndrome is caused by mutation in the fibroblast growth factor receptor 2 (FGFR2) genes [3]. The disease is characterized by premature synostosis of coronal and sagittal sutures which begins in the first year of life. Once the sutures become closed, growth potential to those sutures is restricted. However, multiple sutural synostoses frequently extend to premature fusion of skull base causing midfacial hypoplasia, shallow orbit, maxillary hypoplasia and occasional upper airway obstruction.

Crouzon syndrome occurs in approximately 1 in 25,000 births worldwide [4]. Prevalence in the United States is 1 per 60,000 [5]. Crouzon syndrome makes up approximately 4.8% of all cases of craniosynostoses, making it the most

common syndrome of the more than 100 within the craniosynostosis group [6]. No known race or sex predilection exists.

While many cases have been documented worldwide, none have been reported in the South African black race group; therefore a documented case of a 7-year-old South African black boy with Crouzon syndrome is presented.

Case report

A 7-year-old black boy presented for orthodontic treatment complaining of upper front teeth biting inside his lower teeth. The child's mother stated that the boy constantly snores. Review of medical history was unremarkable, specifically, the mother reported normal labor and delivery. There were no anomalies in any siblings or near relatives reported. The child was not on any medications and denied any medical allergies. He has never been to a dentist before and this was the first dental consultation.

Further medical history revealed that the condition was noted by the mother at 6

months when the fontanels were rising. The child was operated at the ages of 3, 4 and 5 years where a craniotomy was performed, (Fig 1), with subsequent repeated operations due to complications, and none have caused mental complications and he is progressing well in a normal school. The final operation is scheduled at 18 years for the reshaping of the head as well as ocular adjustment.

Extra-oral examination revealed elliptical shaped head, with dolichofacial growth pattern, and convex facial profile. The presence of prominent eyeballs, which is the characteristics of the Crouzon disease triad, can be observed (Fig. 2).

Intra-oral examination showed that the patient is in early mixed dentition, with all primary teeth present in the maxillary arch, and in the mandibular arch only central permanent incisors having erupted in the midst of primary teeth. He displayed dental malocclusion characterized by anterior crossbite (Fig. 3). The maxilla was narrow with high arched palate.

Orthopantomograph evaluation (Fig. 4), demonstrated the presence of all permanent succeeding teeth, indicating

severe space shortage and potential crowding of some of these permanent teeth. The right mandibular permanent canine showed signs of impaction and is horizontally placed. Cephalometric analysis (Fig. 5), indicated skeletal discrepancy complicated by maxillary hypoplasia and vertical growth pattern.

Early orthodontic management included maxillary anterior expansion to correct anterior crossbite. The patient was treated with a removable expansion appliance (Fig. 6), which was activated twice a week. A posterior bite plane was incorporated to open the bite and allow crossbite correction. The patient wore this initial appliance for 12 months, thereafter a new appliance was constructed after the exfoliation of maxillary primary central incisors.

The initial anterior expansion was obtained after 18 months of treatment with a removable appliance resulting in almost edge-to-edge bite (Fig. 7). The patient is still continuing with anterior expansion appliance with the objective of obtaining a positive overbite. In addition the patient is under the management of a multidisciplinary team consisting of neurosurgeon, ophthalmologist, and ENT surgeon.

Discussion

Craniofacial abnormalities are often present at birth and may progress with time. The appearance of an infant with Crouzon syndrome can vary in severity from a mild presentation with subtle midface deficiency to severe forms with multiple cranial sutures fused and marked midface and eye problems. Upper airway obstruction can lead to acute respiratory distress [2,7] Increased intracranial pressure leading to optic atrophy may occur, which can produce blindness if the condition is not treated.

Differential diagnosis of Crouzon syndrome considers Apert syndrome and other problems including Carpenter syndrome, Pfeiffer syndrome, Seatre-Chotzen syndrome, Jackson Weiss syndrome. Patients with associated acanthoses migricans can have FGFR3 mutation.

Management of Crouzon disease is multidisciplinary and early diagnosis is important.

In the first year of life, it is preferred to release the synostotic sutures of the skull to allow adequate cranial volume thus



Fig. 1 Patient's head profile showing the healed scar of craniotomy surgery

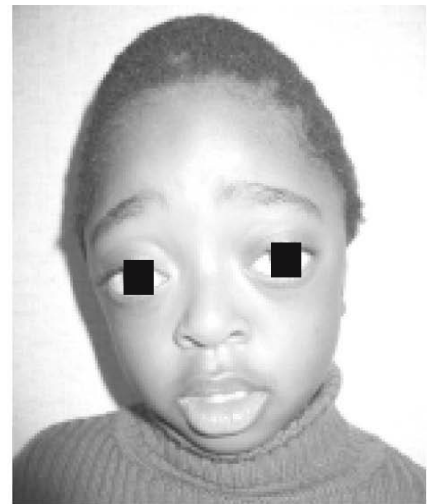


Fig. 2 Patient at initial examination. Note the prominent eyeballs



Fig. 3 Anterior intraoral occlusal showing anterior crossbite



Fig. 4 Orthopantomograph

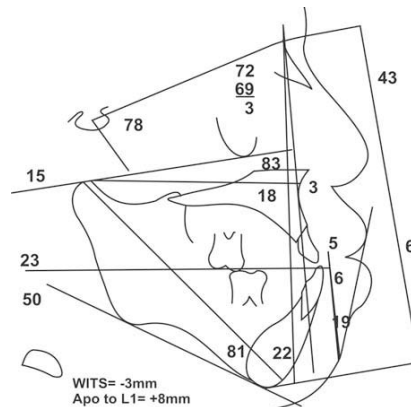


Fig. 5 Cephalometric analysis

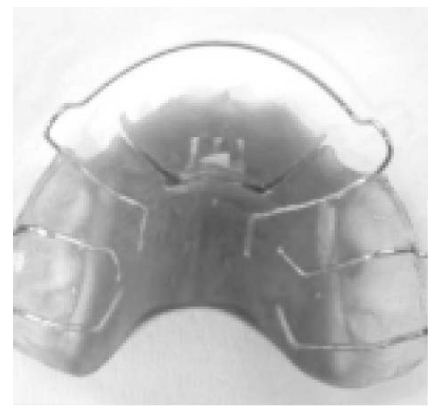


Fig. 6 Anterior expansion appliance



Fig. 7 Intraoral after 18 months of anterior expansion

allowing for brain growth and expansion. Skull reshaping may need to be repeated as the child grows to give the best possible

Fig. 6 Anterior expansion appliance results. If necessary, midfacial advancement and jaw surgery can be done to provide adequate orbital volume and reduce the exophthalmus to correct the occlusion to an appropriate functional position and to provide for a more normal appearance. Prognosis depends on malformation severity.

Orthodontic evaluation should begin at an early age in these children. The upper jaw is usually set back and small in all dimensions. This results in severe crowding of the permanent teeth, as well as a significant anterior crossbite, a condition which can develop into severe skeletal

discrepancy if not treated early as the maxillary teeth are trapped inside of the mandibular teeth. Delayed dental eruption is common, and extra or missing teeth have can be noted on occasion. Treatment might require extraction of some permanent teeth to alleviate the crowding, as well as expansion of the maxilla, either surgically or through the use of orthodontic devices.

Conclusion

Although Crouzon syndrome requires the multidisciplinary team management, dental malformations such as anterior cross-bites, crowding, underdeveloped maxilla, patients may initially present to dental clinic for their primary care and relief. An

understanding of these abnormalities is necessary for the dental team to make the appropriate referrals to insure the patient receives the best available care. The orthodontist can be an integral part of the multidisciplinary team.

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Source of Support: Nil, **Conflict of interest:** None declared.