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Research paper

Crouzon's syndrome: a new surgical approach

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Abstract

Crouzon's syndrome is a rare genetic disorder characterized by distinctive malformations of the skull and facial region, premature cranial suture closure is the most common skull abnormality, optic disc edema and proptosis are among the most common ocular findings. It is a genetic disorder of gene FGFR-2 (Fibroblast Growth Factor Receptor-2) in 95% of cases, and in 5% of cases, FGFR-3 (Fibroblast Growth Factor Receptor-3) mutation occurs.
Once a suture becomes fused, growth perpendicular to that suture becomes restricted and the fused bones act as a single body structure. Compensatory growth occurs at the remaining open sutures to allow continued brain growth, resulting in abnormal bone growth and producing facial deformities.
In the new born child, some potential problems that may need to be addressed include respiratory difficulties, feeding problems, neuro-logic complications such as hydrocephalus and the potential risk of developmental delay.
We represent a literature review and a rare case of Crouzon's Syndrome, who wanted facial correction to be done at the age of 24years. We planned two stage surgical procedure, for correction of facial deformity.

Keywords: *Crouzon's Syndrome; Facial Deformity; Exophthalmos; Craniosynostoses; Fibroblast Growth Factor; Surgically Assisted Rapid Maxillary Expansion (SAME).*

1. Introduction

Crouzon's syndrome is a rare autosomal dominant genetic disorder characterized by distinctive malformation of the skull and the facial skeleton [1]. It is characterized by premature closure of cranial sutures, exophthalmos and midfacial hypoplasia. It occurs in approximately 1 in 25000 live births [1], [6]. Majority of patients have mutations in the extracellular immunoglobulin III domain of the Fibroblast Growth Factor Receptor 2(FGFR 2) gene [1], [16], [17]. Approximately 4.8% of all cases of craniosynostosis are Crouzon's syndrome [5], [18]. No known sex or race predilection [20]. The syndrome is characterized by premature synostosis of coronal and sagittal sutures, which usually begins in the first year of life. Once the sutures become fused, growth potential to those sutures is restricted [10], [15]. However, multiple sutural synostoses frequently extend to premature fusion of the skull base causing midfacial hypoplasia, shallow orbit, exophthalmos, maxillary hypoplasia, and occasional upper airway obstruction. Intraoral manifestations include mandibular prognathism, overcrowding of upper teeth, and V-shaped maxillary dental arch. Narrow, high, or cleft palate and bifid uvula can also be seen. Occasional oligodontia, macrodontia, peg-shaped and widely spaced teeth have been reported [1], [3], [21]. Multiple staged surgeries are the general treatment plan for patients. We present a case of Crouzon syndrome in a 24year old patient.

2. Case report

A 24 year old male patient reported to the department of oral and maxillofacial surgery with a chief complaint of, difficulty in closing eyelids and concerned about aesthetic appearance [Fig. 1].



Fig. 1: Pre-Op Photographs Showing Facial Asymmetry with the Chin Deviated to the Right Side, Retrognathic Maxilla and Exophthalmos.

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The patient was treated for hydrocephalus at the age of one year. There was no history of consanguinity in parents. No other family member or sibling was affected with the same complaints. On examination, the patient was oriented in time, place, and person. The examination of the face revealed enlarged cranial vault with frontal bossing, maxillary hypoplasia, and relative mandibular prognathism. Flattening of the malar prominence and facial asymmetry was also noted. The ocular manifestations included shallow orbits and bilateral proptosis. Multiple impacted teeth, peg shaped and widely spaced teeth were present [Fig. 2, 3]. His cornea had no signs of exposure keratitis.



Fig. 2: Intra Oral Photograph, Showing Peg Shaped and Widely Spaced Teeth.

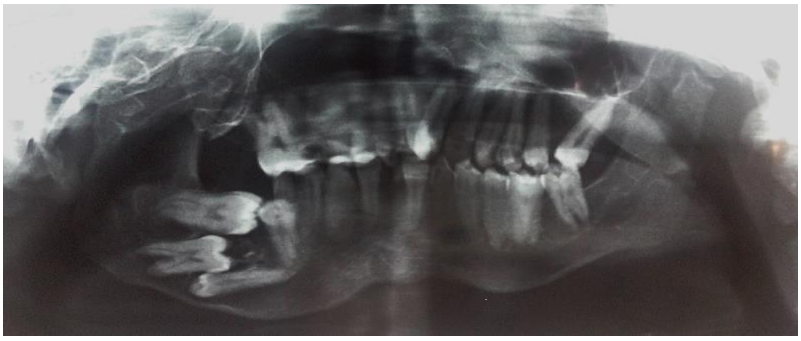


Fig. 3: OPG, Showing Multiple Impacted Teeth.

There was no syndactyly seen which is a differentiating feature between Crouzon's and Apert syndrome. No cardiac abnormality were detected. Computed tomography (CT) facial skeleton reveals bilateral proptosis with shallow orbits and narrow bilateral bony optic canals, craniosynostosis deviated mandible, deviated nasal septum [Fig. 4 A-D].

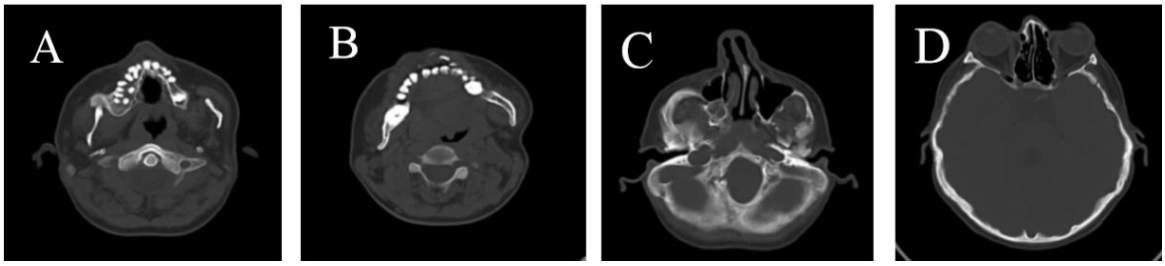


Fig. 4: CT scan Showing A. Multiple Impacted Teeth, B. Deviated Mandible, C. Deviated Nasal Septum and D. Bilateral Proptosis.

The final diagnosis of craniosynostosis, most likely Crouzon syndrome with ocular complications was made, on the basis of clinical and radiological findings.
Stereolithographic model was fabricated based on the CT scan and the model surgery was performed for the advancement of the orbitozygomatic complex [Fig. 5 A-C].

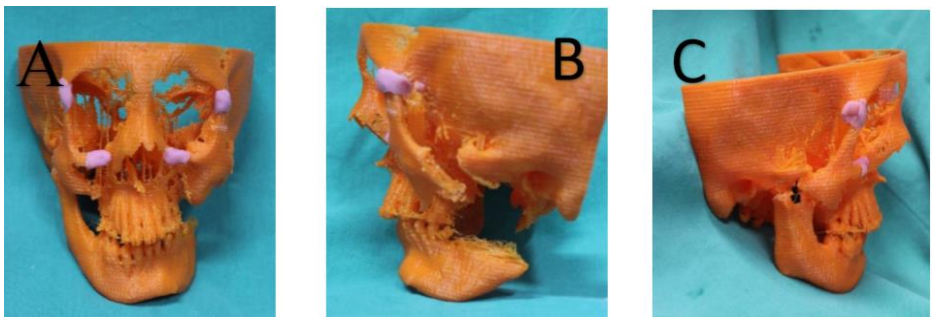


Fig. 5: (A-C): Stereolithographic Model For Surgical Planning and Model Surgery.

3. Stage I surgery

Two stage surgery was planned, In the Stage I surgery, the patient was surgically treated by advancement of orbitozygomatic complex in order to give a malar prominence and an advancement of 7mm was achieved on the right side and 5mm on the left side[Fig. 6 A, B].

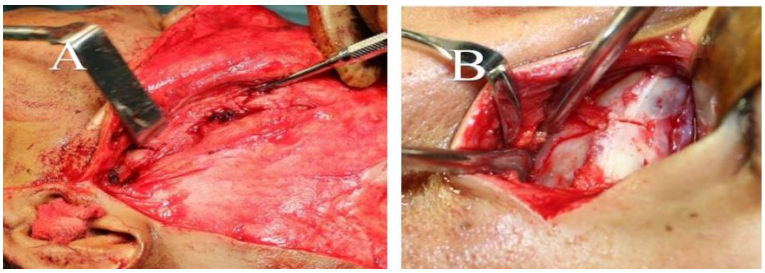


Fig. 6: A. Bicoronal Incision, B. Infraorbital Incisions and Bony Cut.

Patient was taken up for surgery under strict aseptic conditions, tracheostomy was performed and General Anaesthesia was administered. Bicoronal incision and infraorbital incisions were placed. Bony cuts were performed in the frontozygomatic region, infraorbital region and the zygomatic arch region.

Advancements were achieved and the plating was done. Titanium meshes were placed in the Infraorbital region to give a complete closure of the eyelids [Fig. 7 A, B].

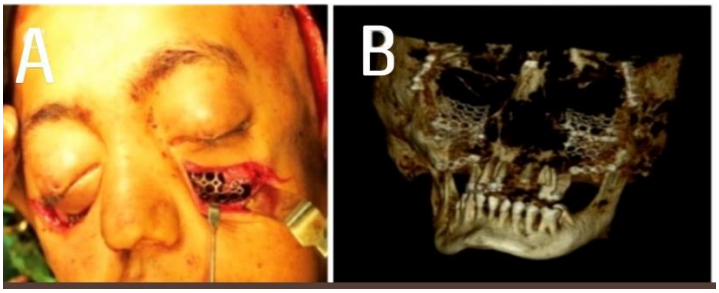


Fig. 7: A. Titanium Mesh Placed in the Infraorbital Region. B. Post-Operative 3D CT scan.

Post operatively, the patient showed improvement in the facial profile and improvement in the closure of the eyes and aesthetics, which further improved during the follow up period [Fig. 8 A-C].



Fig. 8: A) Immediate Post-Operative Profile View. B) Postoperative Closure of the Eyelids. C) Six Months Post-Operative Picture Showing Improved Facial Profile and Aesthetics.

4. Stage II surgery

In the Stage II surgery, surgically assisted rapid maxillary expansion (SAME) using HYRAX (Hygienic Rapid Expander) appliance and mandibular distraction osteogenesis to correct the skeletal midline deviation of lower face was planned. The second stage surgery also performed under general anaesthesia after tracheostomy procedure. Incisions were placed from the midline till the first molar region and the bony cuts were performed and the hyrax appliance placed in the maxilla. Vestibular degloving incision was placed in the right side of mandible and the osteotomy was performed in the premolar region, and corticotomy was done on the left side. Distraction device was fixed and was activated at a rate of 1mm/day after a latency period of 7days [Figure 9 A, B]. And at the end of treatment, in the maxilla 10mm of expansion was achieved and an advancement of 15mm was achieved in the mandible [Fig. 10].

Post operatively the patient showed improved facial appearance and the correction of the chin deformity and the patient was happy with the appearance [Fig. 11].

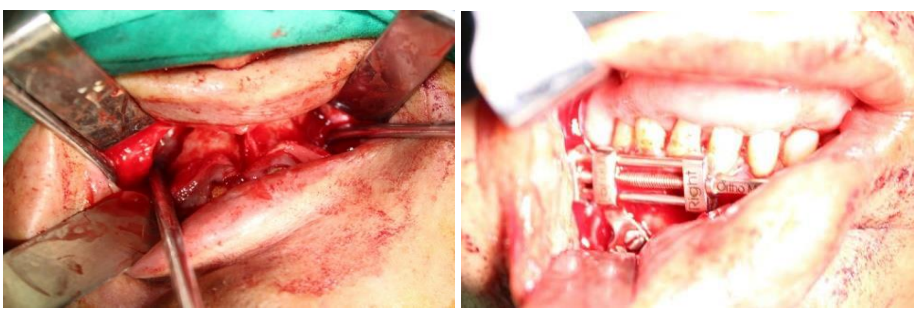


Fig. 9: A) Osteotomy for SAME in Maxilla B) Mandibular Distraction Device in Place.

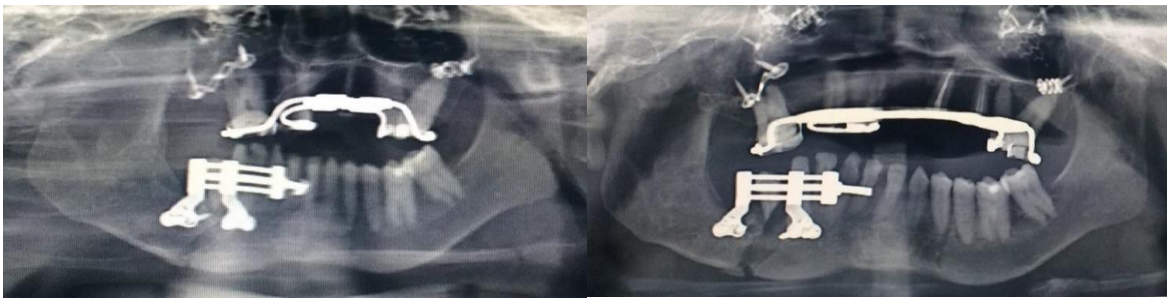


Fig. 10: Post-Operative OPG Showing the HYRAX Appliance and the Mandibular Distractor in Place, and End Treatment OPG Showing Expansion of Maxilla and Advancement of Mandible.



Fig. 11: Six Months Post Op Photograph Showing Improved Facial Appearance and Correction of the Chin Deformity.

5. Discussion and Background

In 1912 Octave Crouzon described the triad of skull deformities, facial anomalies and exophthalmos in a mother and son. Premature craniosynostosis, midfacial hypoplasia and exophthalmos forms a triad now [9], [12].
It is characterized by premature closure of cranial sutures resulting in abnormal growth affecting growth and development of orbits and maxillary complex. Premature sutural fusion most commonly involves the sagittal sutures (56-58%), followed by the coronal sutures (18-28%) and rarely the metopic and lambdoid sutures (5%) [10], [13], [15]. Clinical features usually seen are strabismus, hypertelorism, exophthalmos, beaked nose, maxillary hypoplasia and prognathic mandible [21] [Table 1].

Table 1: Clinical Features of Crouzon's Syndrome

Head	Brachycephaly, dolichocephaly, Trigonocephaly, ridges felt at sutural lines
Brain	Hydrocephalus, raised intracranial pressure, Arnold Chiari malformation
Eyes	Exophthalmos, hypertelorism, reduced monocular visual acuity, reduced binocular visual acuity, amblyopia, optic atrophy, ametropia and hypermetropia, myopia, Manifest strabismus, keratopathy, astigmatism, anisometropia, exotropia and esotropia
Nose	Small beaked nose, Deviated nasal septum
Mid-face	Mid-face retrusion
Jaw	Relative mandibular prognathism
Ears	Low set ears, conductive hearing loss, external, middle and inner ear abnormality
Mouth	High arched palate, overcrowding of teeth, class III malocclusion
Spine	Cervical spine abnormality, fusion of cervical vertebra, scoliosis
Respiratory system	Upper airway obstruction, mouth breathing, sleep apnoea, snoring
Cutaneous	Acanthosis nigricans
Intelligence	Normal in most cases

There are no digital abnormalities present [14]. Craniofacial abnormalities are often present at birth and may progress [12], [19].
Male to female ratio is 3:1, with more than 30 mutations within the gene fibroblast growth factor receptor II (FGFR 2) [24], [25], [30].
The symptoms vary in severity from 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 respiratory problems [7], [29]. Increased intracranial pressure leads to optic atrophy and blindness [8].
Management is multidisciplinary. In the first year, release of synostotic sutures to allow brain growth and expansion [26], [27]. As age advances, reshaping may need to be repeated. Midfacial advancement by distraction osteogenesis or osteotomy can be done to provide adequate orbital volume and reduce exophthalmos [11], [22], [23], [28]. Correction of occlusion and of maxilla and mandible can be done later.
Our patient reported late at the age of 24years with the syndrome.

Treatment options were Lefort II, III distraction osteogenesis [11] and mandibular osteotomy, because of patients compliance we preferred osteotomy of zygomatico orbital region and advancement by 7mm. Patient was able to close his eyes and there was considerable increase in the patient's appearance.
After 6months, rapid maxillary expansion and distraction osteogenesis of the mandible was performed.
The case was reviewed again after 6months and the patient was happy with the aesthetics.

6. Conclusion

Crouzon's syndrome, the treatment is of multidisciplinary approach. Early diagnosis is better. If the patient reports late, then the priority of the treatment will be for the aesthetics i.e., the correction of exophthalmos, advancement of the midface either by osteotomies or Distraction osteogenesis, correction of mandibular prognathism and facial deformities. Correction of occlusal abnormalities by fixed orthodontics or rehabilitation by prosthodontist also needs to be done to ensure specific treatment to prevent late diagnosis effects.

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