Clinical Report

Combined denture prosthetic treatment of patients with Crouzen syndrome

Puneet Bhardwaj, Meena Aras
Department of Prosthodontics, Goa Dental College and Hospitals, Bambolim-403 202, Goa, India

For correspondence
Dr. Puneet Bhardwaj, C- 503, Sarita Vihar, New Delhi-110 076, India. E-mail: drpuneetbhardwaj@yahoo.co.in

Crouzon syndrome causes anodontia and hypodontia intraorally, in addition to other symptoms. Partial or total anodontia results in loss of function such as chewing, and affects aesthetics. Prosthodontic rehabilitation can be accomplished with fixed, overdenture, complete, or implant-retained prostheses. For rehabilitation, it is crucial to know the age, number and condition of the present teeth, and the state of growth of the patient. A 21-year-old female patient, who visited our institution, was treated by a multi-disciplinary team of surgeons, endodontist, and prosthodontists. Combined denture (combined fixed-removable tooth replacement) was considered for correcting the angulation and cross bite of maxillary remaining teeth.

Key words: Anodontia, craniosynostosis, Crouzon syndrome, cross bite, exophthalmos, hypertelorism, strabismus

DOI: 10.4103/0972-4052.49025

INTRODUCTION

In 1912, Crouzon described the hereditary syndrome of craniofacial dysostosis in a mother and son. He described the triad of calvarial deformities, facial anomalies, and exophthalmos. Crouzon syndrome is an autosomal dominant disorder, with complete penetrance and variable expressivity. It is characterized by premature closure of calvarial and cranial base sutures, as well as those of the orbit and maxillary complex (craniosynostosis). Other clinical features include hypertelorism, exophthalmos, strabismus, beaked nose, short upper lip, hypoplastic maxilla, and relative mandibular prognathism. Unlike some other forms of autosomal dominant craniosynostosis, no digital abnormalities are present in Crouzon syndrome.[1,2]

Crouzon syndrome is caused by mutations in the fibroblast growth factor receptor-2 (*FGFR2*) gene, but exhibits locus heterogeneity with causal mutations in *FGFR2* and *FGFR3* in different affected individuals. Premature synostosis of the coronal, sagittal, and, occasionally, lambdoidal sutures begins in the first year of life and is completed by the second or the third year. The order and rate of suture fusion determines the degree of deformity and disability. Once a suture becomes fused, growth perpendicular to that suture becomes restricted, and the fused bones act as a single bony structure. Compensatory growth occurs at the remaining open sutures, to allow continued brain growth. However, multiple sutural synostoses frequently extend to premature fusion of the skull base sutures, causing midfacial hypoplasia, shallow orbits, a foreshortened nasal dorsum, maxillary hypoplasia, and occasional upper airway obstruction.[3-5]

CLINICAL FEATURES

Skull

Craniosynostosis: Craniosynostosis commonly begins during the first year of life and usually completes by the second or the third year. Coronal and sagittal sutures are most commonly involved, resulting in acrocephaly, brachycephaly, turricephaly, oxycephaly, flat occiput, and high prominent forehead with or without frontal bossing. Ridging of the skull is usually palpable. Cloverleaf skull is rare and occurs in the most severely affected individuals.

- Flattened sphenoid bone
- Shallow orbits
- Hydrocephalus (progressive in 30%)

Face

- Midface (maxillary) hypoplasia

Eyes

- Exophthalmos (proptosis) secondary to shallow orbits, resulting in frequent exposure conjunctivitis or keratitis
- Ocular hypertelorism
- Divergent strabismus
- Rare occurrence of nystagmus, iris coloboma, aniridia, anisocoria, microcornea, megalocornea,

cataract, ectopia lentis, blue sclera, glaucoma, luxation of the eye globes, and blindness from optic atrophy

Nose
- Beaked appearance
- Compressed nasal passage
- Choanal atresia or stenosis
- Deviated nasal septum

Mouth
- Mandibular prognathism
- Overcrowding of upper teeth, malocclusions, and V-shaped maxillary dental arch
- Narrow, high, or cleft palate and bifid uvula
- Occasional oligodontia, macrodontia, peg-shaped, and widely spaced teeth

Ears
- Narrow or absent ear canals
- Deformed middle ears

Other skeletal features
- Cervical fusion (18%), C2-C3 and C5-C6
- Block fusions involving multiple vertebrae
- Subluxation of the radial heads
- Ankylosis of the elbows

Skin
- Approximately five percent of the patients have acanthosis nigricans, which is detectable after infancy. The hallmark of these lesions is a darkened thickened skin, with accentuated markings and a velvety feel.
- Central nervous system
- Approximately 73% of the patients have chronic tonsillar herniation. Of these, 47% have progressive hydrocephalus.
- Syringomyelia may be present.[1,2,5]

CLINICAL REPORT

A 21-year-old female patient was referred to the Department of Prosthodontics, Goa Dental College, Goa, for examination, evaluation, and treatment. The patient exhibited the classical features of Crouzon syndrome including brachycephaly, flat occiput, and high prominent forehead, and shallow orbits. The facial profile showed midface (maxillary) hypoplasia, ocular hypertelorism, nose with compressed nasal passage and deviated nasal septum [Figure 1]. Intraoral examination revealed oligodontia and overcrowding of the upper teeth, malocclusion and V- shaped maxillary dental arch with narrow and high arch palate [Figure 2A, B].

Considering the clinical situation and the financial condition of the patient, maxillary fixed restoration and upper and lower cast partial denture were determined to be the treatment of choice.

Ectopically erupted 24 (maxillary left first premolar) was extracted by the surgeon, because of its malaligned position and to help develop a simple and straightforward treatment plan.

Preliminary impressions were made with irreversible hydrocolloid impression material (Neocolloid; Zhermack, Badia Polesine, Italy). Diagnostic cast was made in type III stone (Kalstone, Kalabhai Karson Pvt. Ltd., Mumbai, India) and mounted in Artex semiadjustable articulator, with the help of Rotofix facebow and interocclual records in centric relation. Diagnostic wax up of the upper arch was done to correct the angulation and occlusal scheme of the patient within functional and aesthetic limitations.

Teeth no. 12, 13, 14, 15, 22, 23 and 25 were treated endodontically, to correct the angulation and occlusal scheme by giving fixed restoration. Teeth no. 12 and 22 were reduced to overdenture abutment without coping, because of their severe mesiolingual tilt and to prevent bone loss in the anterior region due to extraction of two [Figure 3].

Long span FPD was not selected as a treatment option because of malaligned posterior teeth and irregularly resorbed alveolar ridge in the maxillary central incisor region. this resorbed portion has caused a deep cleft like defect, which is difficult to clean and maintain under fixed restoration. Four incisors were replaced with fixed provisional restoration, to check for aesthetics, phonetics, function and comfort. Such a long bridge is also difficult to cast, without marginal discrepancies.

Provisional restorations were made according to diagnostic wax-up and placed in the patient's mouth after performing teeth preparation procedure. Provisional restorations were cemented in the patient's mouth for a period of one month for aesthetics, function and comfort [Figure 4]. A final impression was taken with addition silicon impression material (Flexitime, Easy Putty; Heraeus Kulzer, Hanau, Germany). Metal framework of fixed restoration was tried for contact, contour and marginal adaptation. Metal framework of fixed restoration was provided with precision milling on the lingual surface for frictional seating of the cast partial denture on fixed restoration, with elimination of clasps in the anterior region, for aesthetic purpose. Pick up impression of the metal framework was taken in heavy body addition silicon impression material (Flexitime, Easy Putty; Heraeus Kulzer, Hanau, Germany) and cast partial framework was fabricated on this final cast. Both fixed and removable metal framework was tried in the patient's mouth for precise sitting, close

Figure 1: Appearance of the patient's face before treatment

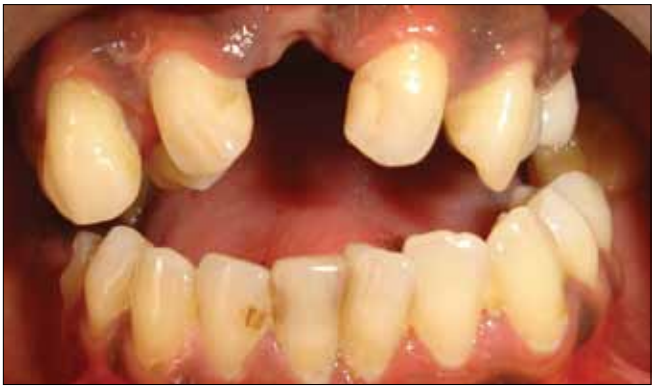

Figure 2 (a): Intraoral view of the patient, showing malocclusion and oligodontia

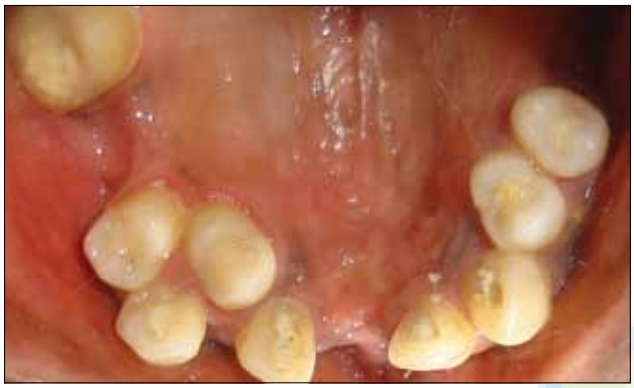

Figure 2 (b): Intraoral view of the patient, showing malocclusion and oligodontia

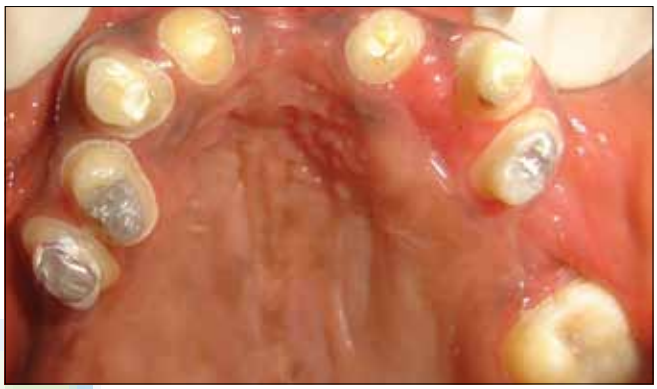

Figure 3: Prepared teeth for fixed restorations

Figure 4: Patient with provisional restoration

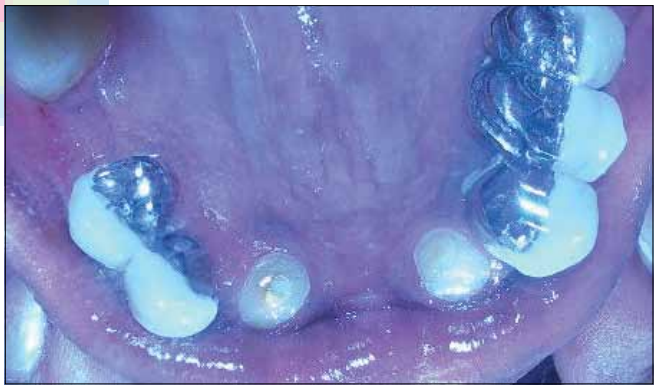

Figure 5: Patient with fixed restoration showing precision milling

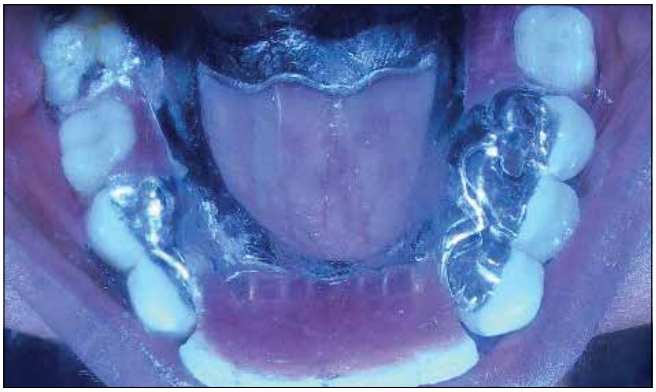

Figure 6: Combined denture (combined fixed-removable tooth replacement) partial denture restoration

Figure 7: Patient after prosthetic treatment

adaptation and frictional resistance of removable framework. Fixed restoration was cemented after doing bisque trial and final glazing of porcelain [Figure 5]. Cast partial denture with arranged teeth was tried in the patient's mouth and finally acrylized and delivered to patient [Figures 6-7].

DISCUSSION[6,7,8]

Prosthetic treatment modes using RPDs or complete dentures and dental implants are the primary treatment alternatives for the clinical management of young patients with severe hypodontia. Oligodontia or anodontia totalis associated with Crouzen syndrome, is often characterized by underdeveloped alveolar bone structures with missing or reduced alveolar ridges. This results in less volume of bone for support of conventional prosthetic dentures, and it can also affect the bone volume available for the placement of dental implants. In developing the optimal surgical and prosthetic approach, the patient's age, dental and skeletal maturity, and the bone volume that is available at the time of intervention must be considered.

With a combined denture (combined fixed-removable tooth replacement) restoration is possible today in countless variations, for nearly every situation in the partially edentulous arch. Properly constructed, these quite complex dentures have proven effective in the restoration of the masticatory function for decades. Also in terms of aesthetic aspects and periodontal hygiene they offer the patient a flawless restoration.

REFERENCES

1. Peterson JS, Pruzansky S. Palatal anomalies in the syndromes of Apert and Crouzon. Cleft Palate J 1974;11:394-403.
2. Cohen MM Jr. Craniosynostosis update 1987. Am J Med Genet Suppl 1988;4:99-148.
3. Wilkie AO, Slaney SF, Oldridge M, Poole MD, Ashworth GJ, Hockley AD, *et al*. Apert syndrome results from localized mutations of FGFR2 and is allelic with Crouzon syndrome. Nat Genet 1995;9:165-72.
4. Reardon W, Winter RM, Rutland P, Pulleyn LJ, Jones BM, Malcolm S. Mutations in the fibroblast growth factor receptor 2 gene cause Crouzon syndrome. Nat Genet 1994;8:98-103.
5. Cohen MM Jr. An etiologic and nosologic overview of craniosynostosis syndromes. Birth Defects Orig Artic Ser 1975;11:137-89.
6. Yalisove IL. Crown and sleeve-coping retainers for removable partial dneutres. J Prosthet Dent 1966;16:1069-85.
7. Gungor MA, Artunc C, Sonugelen M. Parameters affecting retentive force of conus crowns. J Oral Rehabil 2004;31:271-7.
8. Saito M, Miura Y, Notani K, Kawasaki T. Stress distribution of abutments and base displacement with precision attachment and telescopic crown-retained removable partial dentures. J Oral Rehabil 2003;30:482-7.

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

AUTHOR INSTITUTION MAP FOR THIS ISSUE

Please note that not all the institutions may get mapped due to non-availability of requisite information in Google Map. For AIM of other issues, please check Archives/Back Issues page on the journal's website.