Prosthetic rehabilitation of a Crouzon patient: A case report

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Abstract

Crouzon syndrome is a rare genetic disorder, which can be defined as a variation of craniofacial dysostosis caused by the premature obliteration and ossification of two or more sutures. The growth pattern results in pseudoprognathism and malocclusions including an overcrowded or a widely spaced dentition. Specifically maxillary arch is narrow, high, and V-shaped. Cleft palate and bifid uvula are other possible features in the oral cavity. This report describes a non-surgical treatment model to overcome the remaining significant Class III intermaxillary relation and excessive tooth loss to recover function and aesthetics for a 25-year-old Crouzon patient.

Keywords: Crouzon syndrome, overdenture, prosthetic treatment

Introduction

Crouzon Syndrome is a genetic disorder showing autosomal dominant trait. Originating from the mutation of fibroblast growth factor receptor-2 (FGFR2), this syndrome affects the first branchial arch, which is the precursor of the maxilla and mandible.[1]

The most notable characteristic of Crouzon Syndrome is cranial synostosis (a union between adjacent bones or parts of a single bone), but it is usually presented as brachycephaly (fusion of the coronal suture), which results in the appearance of a short and broad head. The craniofacial skeletal findings are believed to result from the cranial base, possibly resulting in premature suture fusion of both the cranial base and cranial sutures.[2] Fusion of different sutures leads to different patterns of growth of the skull. Premature synostosis of the coronal, sagittal, and lambdoid sutures begins in intrauterine life and growth restriction in the neighboring bones lead to abnormal bone growth and produce facial deformities.[3] Multiple sutural synostosis initiate the premature fusion of the skull base sutures causing midfacial hypoplasia, shallow orbits, a foreshortened nasal dorsum, maxillary hypoplasia, and, in severe cases, upper airway obstruction. Although usually presented at birth, craniofacial deformities are prominently presented with time, leading to mental retardation in 12% of the cases.[4]

Physical features include:[5-7]

- Craniosynostosis, which mostly affects the coronal and sagittal sutures, starting from intrauterine life up to 2–3 years of age.
- Due to the lateral and anteroposterior flattening of the acrocranium, anteroposterior diameter of the head is smaller than transverse diameter.
- Vertical growth pattern of the cranium leads to high and widened forehead.
- Hypoplastic maxilla and wide face produce pseudoprognathism and malocclusions.
- Oral peculiarities include narrowed and high V-shaped maxillary dental arch, overcrowding or widely spacing of the upper teeth, cleft palate and bifid uvula.
- Deviation of the nasal septum, narrowed or obliterated anterior nares, nasopharyngeal narrowing and wide beaked nose are present that cause upper airway obstruction. The nose is curved in shape resembling a parrot nose.
- Ocular proptosis is presented, that is caused by very shallow orbits.
- Hypertelorism, divergent squint, and downward slanting of the upper eyelids cause “frog face” appearance.
- The upper lip is shortened and sometimes cleaved.

This case report presents a choice of prosthetic treatment for a partially edentulous patient with Crouzon syndrome.

Case Report

A 25-year-old male patient was referred to İstanbul University Faculty of Dentistry by his dentist as a complicated partial edentulous case. The patient was free of systemic diseases and was not receiving any medication. He also did not have a story of allergy. The patient apparently represented physical means of Crouzon syndrome like craniosynostosis, involving high and widened forehead, parrot like nose, deep V shaped maxillary arch, proptosis of the eyes, abundant, and thick dark hair, and as the most distinct feature anteroposterior flattening of the cranium [Figures 1 and 2].

Dental history revealed that both primary and permanent
dentitions were complete initially but a removable partial denture was fabricated at the age of 10 due to early tooth loss caused by severe periodontal destruction. Unfortunately, the patient had not used the dentures as he could not have adopted himself to it.

The patient’s main expectation was to improve the reduced chewing ability, which was caused by excessive number of tooth loss. There were a total of 14 remaining teeth, 8 in the maxilla and 6 in the mandible, most of which were malpositioned and periodontally involved [Figures 3 and 4]. The probing depths of most teeth were 2 to 3 mm, which was 7 mm in the mesial sight of the maxillary right first molar, 5 mm in the mesial sight of the second left incisor, 4 mm in the maxillary left canine and the mandibular left first molar. The overall periodontal situation was associated with poor oral hygiene. Right maxillary first molar, left maxillary first incisor, both mandibular first incisors, and right mandibular first molar teeth exhibited a mobility of mod 1 according to Miller’s index. The intermaxillary relation of the jaws was Class III resulting nearly in a circular non-occlusion [Figure 5].

Diagnostic casts were prepared via alginate impressions. Casts were mounted on a paralleleometer and were analyzed on the basis of present undercuts, potential guiding surfaces and retentive areas of the remaining teeth for a possible removable partial denture.

A telescopic overdenture for the maxilla and a single piece fixed prosthesis for the mandible were planned for restoring the dentition.
All teeth were prepared with chamfer type margins. The maxillary right first molar, which had a severe soft tissue recession on the buccal side, was prepared at the enamel-cement border to ease the oral care. Interim fixed restorations were fabricated from silicon impressions that were made before tooth preparations. Final impressions for ceramic veneers and telescopic copings were made with an addition type silicone (Panasil; Kettenbach, Eschenburg, Germany) three days after tooth preparations. The copings of the telescopic crowns were cast with retention pearls on the sides and mandibular framework for ceramic veneers was cast as one piece. The try in for the copings and mandibular single piece cast framework were done as in routine. Before making a secondary impression over the copings from the maxillary arch, the retention pearls on the copings were combined with self curing acrylic polymer (Pattern Resin LS, G.C., Alsip, U.S.A.) to make sure that the primers do not change position during impression making. The final impression for the telescopic crowns was also made with the same addition type silicone.

Among the malpositioned teeth, both mandibular first molars were severely tipped to the mesial sides but only left molar had to receive endodontic treatment for proper tooth preparation.
removable partial denture to fixed partial dentures over
in our case. However, in adult patients surgical management
orthodontist, within a developmental context is needed at
interdisciplinary approach, including the pediatric dentist
patterns, visceral function, and psychosocial development.
To maximize surgical outcomes and patient satisfaction, an
prosthodontic treatment seems to be a good choice to
consents of the patient and his family were taken on the
basis of aesthetics and comfort.

The maxillary removable prosthesis was finished with
conventional procedures and the mandibular fixed
prosthesis was glazed [Figure 6]. Mandibular prosthesis
was cemented with resin-type cement (Panavia F, Kuraray
Dental, Okayama, Japan) and the copings of the telescopic
overdenture were cemented with zinc polycarboxylate
cement (Adhesor Carbofine, Spofa Dental, Jicin, Czech
Republic) via closed mouth technique. Verbal and written
instructions about the care of the prosthesis and the
abutment teeth were given to the patient and his family.
Early recalls were made on the first day, first week, and
two weeks after the prostheses were delivered to the
patient. As the cooperation of the patient was poor about
the care of prostheses and the abutment teeth, monthly
recalls were made during the following year to consolidate
the oral care. Later recalls revealed positive feedback, the
remaining abutment teeth and the periodontal structures
were healthy and the prostheses were in good condition,
still serving satisfactory function and aesthetics [Figures
7-10]. After prosthetic rehabilitation, as stated by his family,
the patient was further socialized and at the age of 25 he
was employed for the first time in his life.

Discussion

Treatment planning for Crouzon patients varies according
to the age the syndrome is diagnosed. Multiple staged
surgery is the general treatment plan in infancy, in order
to stage reconstruction to coincide with facial growth
patterns, visceral function, and psychosocial development.
To maximize surgical outcomes and patient satisfaction, an
interdisciplinary approach, including the pediatric dentist
orthodontist, within a developmental context is needed at
eyeal ages.[8] However, in adult patients surgical management
may not be preferred for various reasons by the patient, as
in our case.

A variety of treatment plans ranging from a conventional
removable partial denture to fixed partial dentures over
remaining teeth or with additional support from implants
were evaluated for reconstructing the mandibular arch.
As the patients’ former experience of a conventional clasp
retained removable partial denture was a failure, this choice
of treatment was ignored. The cost of implant therapy was
the reason to eliminate this choice.

For restoring the maxillary dentition fixed partial dentures or
an overdenture type removable prosthesis were the choices
of treatment. Extreme Class III relation of the jaws restrained
the possibility of constructing the maxillary arch with fixed
partial dentures, even in a cross bite occlusion. Besides these
factors, to achieve maximum support, retention, stability and
to benefit from the splinting effect, a telescopic overdenture
was the final choice.

There were certainly more complicated treatment options,
which mostly included quiet invasive orthognatic surgical
procedures. As the patient refused surgical treatment, these
options were initially eliminated.

Usually, multiple surgeries to prevent maxillary hypoplasia
and cranial synostosis are planned starting from the early
ages. But, for the adolescent and adult patients, little is
achieved with surgery. Most available papers on Crouzon
syndrome present either information about the syndrome
itself or multidisciplinary treatment procedures to overcome
the craniofacial deformities disturbing the growth pattern
and the physical appearance. The dental treatment of a
patient is usually neglected due to the invasive procedures
during early ages when permanent dental treatment is not
possible. Therefore, such patients receive dental care mostly
after the treatment for the vital features of the syndrome.
This situation has led to a lack of literature about the dental
care and treatment of adult patients with Crouzon syndrome.

Prosthodontic treatment seems to be a good choice to
restore not only the dental arches but also facial contours as
well for selected patients with Crouzon Syndrome. The type
of prostheses, the supporting structures or implants should
be evaluated for each patient.

Conclusion

Prosthetic reconstruction of function and aesthetics for
Crouzon patients avoiding further surgical applications
appears out to be a powerful alternative to improve the
quality of life and patients’ satisfaction. This situation has led to a lack of literature about the dental
care and treatment of adult patients with Crouzon syndrome.

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References

1. Bowling EL, Burstein FD. Crouzon syndrome. Optometry 2006;77:217-22.
2. Kreiborg S. Craniofacial growth in plagiocephaly and Crouzon syndrome. Scand J Plast Reconstr Surg 1981;15:187-97.
3. Ulgen O, Gencosmanoglu R, Cankayali R, Tasdemir G, Mutluer S, Songur E. The effects of fronto-parieto-squamosal suture fusion on cranial growth: An experimental study. J Craniofac Surg
4. Richtsmeier JT, Lele S. Analysis of craniofacial growth in Crouzon syndrome using landmark data. J Craniofac Genet Dev Biol 1990;10:39-62.

5. Tay T, Martin F, Rowe N, Johnson K, Poole M, Tan K, et al. Prevalence and causes of visual impairment in craniosynostotic syndromes. Clin Experiment Ophthalmol 2006;34:434-40.

6. Arnaud-López L, Fragoso R, Mantilla-Capacho J, Barros-Núñez P. Crouzon with acanthosis nigricans. Further delineation of the syndrome. Clin Genet 2007;72:405-10.

7. Boutros S, Shetye PR, Ghali S, Carter CR, McCarthy JG, Grayson BH. Morphology and growth of the mandible in Crouzon, Apert, and Pfeiffer syndromes. J Craniofac Surg 2007;18:146-50.

8. Nurko C, Quinones R. Dental and orthodontic management of patients with Apert and Crouzon syndromes. Oral Maxillofac Surg Clin North Am 2004;16:541-53.

9. Bayraktar G, Kaynar A, Duran O, Oner B, Oztürk S, Palandüz S. Case report: A surgical and prosthetic approach to combination syndrome presenting in a patient with craniofacial dysostosis (Crouzon syndrome). Eur J Prosthodont Restor Dent 1998;6:9-12.

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