Case Report - Congenital Deformity

Crouzon syndrome: Virtual planning of surgical treatment by application of internal distractors

Access this article online
Website:
www.amsjournal.com
DOI:
10.4103/2231-0746.186144
Quick Response Code:

Chingiz Rahimov, Ruslan Asadov[1], Gunel Hajiyeva, Nazim Verdiyev[2], Zaur Novruzov[3], Ismayil Farzaliyev

Departments of Oral and Maxillofacial Surgery and [3]Children's Dentistry, Azerbaijan Medical University, [2]Republic Neurosurgical Hospital, Baku, Azerbaijan, [1]Scientific Center of Medical Care for Children with Developmental Disabilities of Craniofacial Region and Congenital Diseases of Nervous System, Moscow, Russian Federation

Address for correspondence:
Prof. Chingiz Rahimov, Bakichanov Street 23, Baku 1022, Azerbaijan.
E-mail: chinrahim@hotmail.com

ABSTRACT

Crouzon syndrome is one of the frequent pathologies within craniosynostosis syndrome. Current progress in computers and biotechnologies allows improving surgical approach and forecasting final result of reconstruction as well. We present a case of successful surgical treatment of Crouzon syndrome, done by application of virtual planning allowing determining "monobloc" features, type of reconstruction and distraction protocol as well. A 20-year-old female had presented with craniofacial deformity. Clinical and radiological investigation revealed Crouzon syndrome. The "monobloc" creation, cranioplasty and internal distractors positioning, direction and schedule of advancement were done according to preoperative virtual planning data achieved by Materialise Mimics Research software. Nine months postoperative functional and esthetic result and radiological findings showed to be reasonable. That application of virtual simulation significantly allows to determine best direction of distraction and improves postoperative outcomes of surgical treatment of Crouzon syndrome.

Keywords: Crouzon syndrome, distraction osteogenesis, "Monobloc" osteotomy, virtual planning

INTRODUCTION

Crouzon syndrome is one of the most frequent pathology within syndrome craniosynostosis. It is characterized by synostosis of coronal, lambdoid and sagittal sutures.[1,2] Typical symptoms are proptosis, midface hypoplasia, hypertelorism and Class III malocclusion.

A gold standard of reconstruction is simultaneous bifrontal craniotomy and Le-Fort IV osteotomy, application of distractors with further advancement of upper and midface as one unit.[3-9] Current progress in computers and biotechnologies allows improving surgical approach and forecasting final result of distraction as well. We present a case of surgical treatment of Crouzon syndrome, by application of virtual planning allowing determining "monobloc" features, type of cranioplasty and direction of internal distractors as well as advancement.

CASE REPORT

A 20-year-old female has been admitted to the hospital with symptoms of facial and head dysmorphism, hypertelorism malocclusion, impaired speech and eating difficulties. Upon anamnesis certain pathology is congenital and first signs were noted immediately after birth. No specific treatment or diagnosis

Cite this article as: Rahimov C, Asadov R, Hajiyeva G, Verdiyev N, Novruzov Z, Farzaliyev I. Crouzon syndrome: Virtual planning of surgical treatment by application of internal distractors. Ann Maxillofac Surg 2016;6:135-40.

was done to the date. Clinical investigation revealed slight disproportion in head and trunk proportions; wide, flat and oblong forehead; expressed supraorbital ridge, short and wide face, beaked nose deformity. Intraoral investigation revealed Class III malocclusion, maxillary hypoplasia, dome-like deformity of the palate, abnormal anatomy of teeth, narrowing of the upper and lower dental arches [Figure 1]. Ophthalmological investigation detected strabismus, exophthalmos, proptosis [Figure 2].

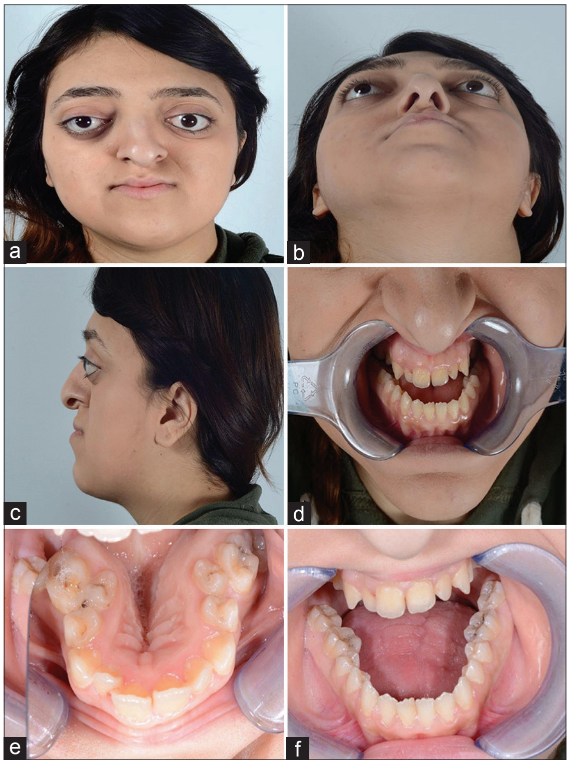

Figure 1: Clinical examination of the patient revealed: (a) facial disharmony; (b) exorbitism; (c) hypoplasia of midface; (d) Class III occlusion; (e) narrowed upper dental arch; (f) impaired lower dental arch

The computed tomography (CT) scan showed no coronal and sagittal sutures on cranial vault, increase in the distance of C5–C6, increase in the volume of ephippium, tower-shaped deformity of the cranial vault as well as increased diameter of the head and cephalometric index up to 82.35 (the normal range is 78.3),

Figure 2: Ophthalmological investigation of the patient revealed: (a) strabismus; (b) proptosis; (c) and (d) exophthalmos

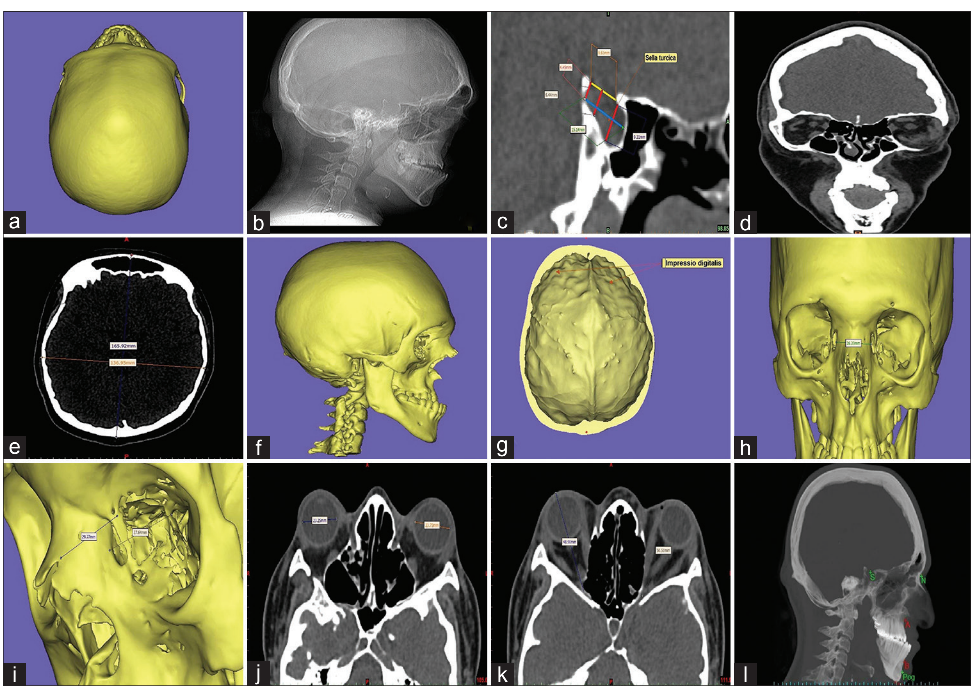

Figure 3: Radiological investigation of the patient detected: (a) no coronal and sagittal sutures; (b) increased distance between C5–C6; (c) increased volume of ephippium; (d) tower-like deformity of cranium; (e) increased cranial index; (f) midfacial hypoplasia; (g) digital impressions on inner surface of the cranial vault; (h) normal intercanthal distance; (i) decreased length of the orbits; (j) diameter of the eyeballs are normal; (k) the length of the optic nerves are normal; (l) SNA is compromised

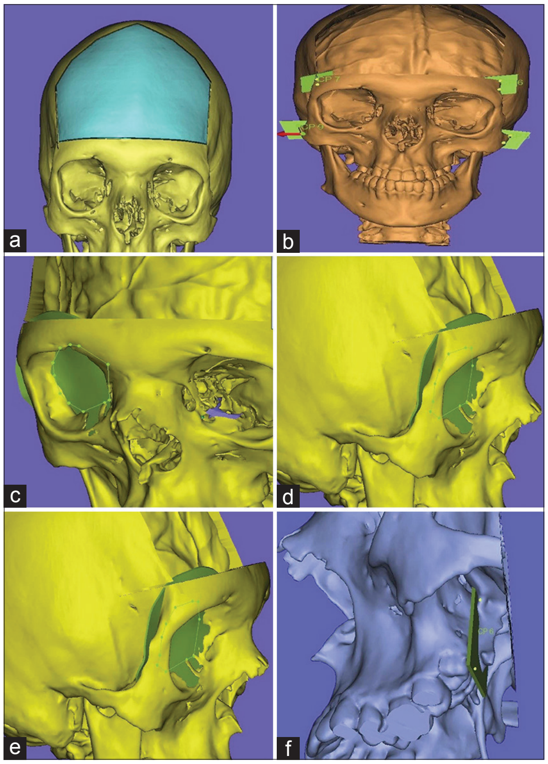

Figure 4: Preoperative virtual planning: (a) anterior craniotomy; (b) facial bones osteotomy; (c) and (d) circular osteotomy; (e) vomer osteotomy; (f) pterygo-maxillary disjunction

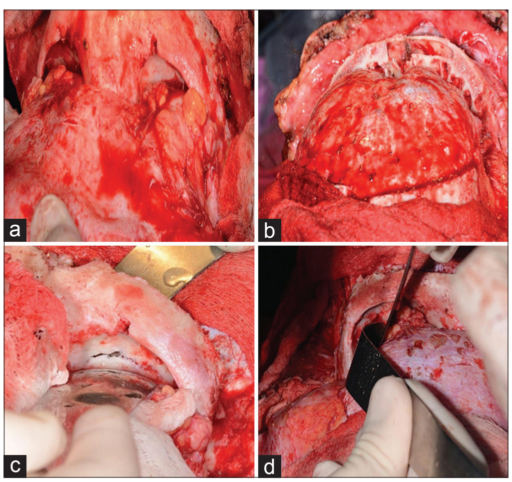

Figure 6: Surgical procedure: (a) Exposure of cranial skeleton; (b) access to anterior cranial fossa; (c) orbital roof osteotomy; (d) lateral orbit osteotomy

hypoplasia of the midface, digital impressions on the inner surface of the cranial vault, normal intercanthal distance, decreased depth of both orbits (29.77 mm on the right side and 27.64 mm on the left side) and normal anatomy of both eyeballs [Figure 3]. Three-dimensional cephalometric investigation revealed impaired SNA and SNB [Figure 4].

As result of clinical and radiological investigation diagnosis of Crouzon syndrome was established with 75% of accuracy.

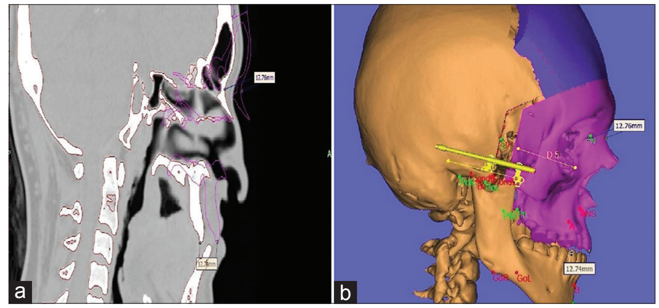

Figure 5: Preoperative virtual planning: (a) monobloc advancement; (b) distractors positioning

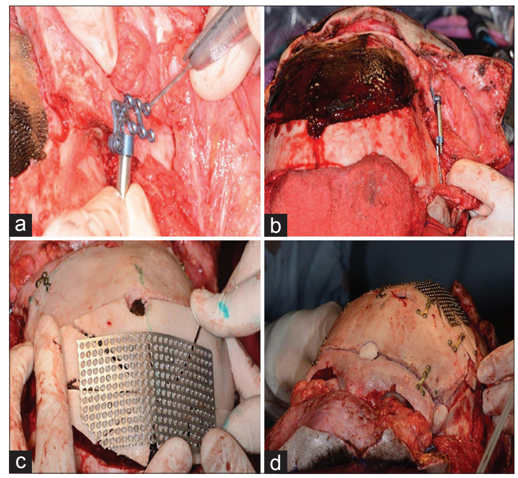

Figure 7: Surgical procedure: (a and b) internal distractors placement; (c and d) cranioplasty

By taking into account the age of the patient and clinical symptoms, monobloc advancement by means of internal distractors was planned. Therefore, a virtual planning protocol was used for procedure preplanning.

Virtual planning step

Materialise Mimics Research version (Materialise, Belgium) was used. Virtual data achieved as DICOM format burned on CD and imported to software. All virtual procedures were done in systemic fashion and well protocoled. As the first step virtual frontal craniotomy area was designed based on monobloc advancement principles. The area was determined and the squama of the frontal bone was removed virtually to access anterior cranial fossa.

As the standpoint for advancement the virtual cephalometry data was chosen with two conditions: (1) advancement should strictly be parallel to occlusion plane; (2) during advancement original A point should reach normal ranges of SNA index. Thus, the value of advancement was 12.76 mm. In next step facial bones osteotomy, including zygoma arches, frontozygomatic suture and Sphenozygomatic suture was done. Virtual circular orbitotomy was done both sides, thus segmentation of mid-third of monobloc was done. In the last step of virtual osteotomy separation of vomer and both pterygo-maxillary sutures was performed. As a result complete segmentation of virtual monobloc was achieved and its virtual advancement was visualized [Figure 4].

Because of primary idea to perform monobloc advancement by the means of internal distractors, the virtual analogues were used to determine their best position and distraction direction [Figure 5]. Appropriate virtual measurements were done and all data were well documented to be used during surgical procedure.

Surgical step

Surgery was done under general anesthesia delivered by orotracheal intubaton. Bilateral tarsoraphy and hair styling was done as a part of preoperative preparation. Then a standard coronal approach was created and extended to the nasal base, medial and lateral wall of both orbits. A great attention was paid to temporal muscle dissection, which was completely separated from its origin. As result of this dissection the frontal bone squama, both temporal fossa, and medial, upper and lateral walls of the orbit as well as the nasal base were free from soft tissue.

In the next step, frontal craniotomy was done according to preplanned lines of cutting. The squama was removed and the frontal lobe of the brain was approached. Then the frontal lobes of the brain were carefully dissected from the anterior cranial fossa including transection of anterior 2/3 of olfaction fibers and the

Figure 8: Postoperative clinical view of the patient: (a) facial harmony restored; (b) no exorbitism occurs; (c) maxillary hypoplasia eliminated; (d) Class I oclussion achieved

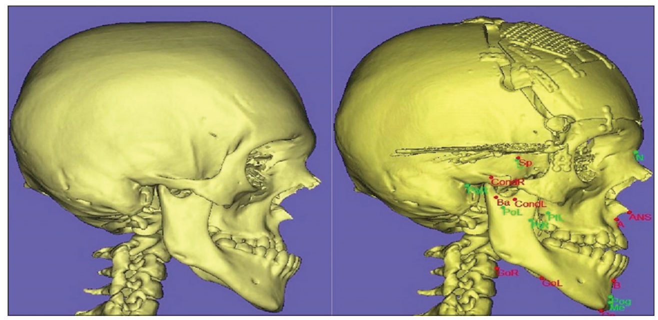

Figure 9: Postoperative computed tomography scan: Comparative computed tomography scan proves proper monobloc advancement

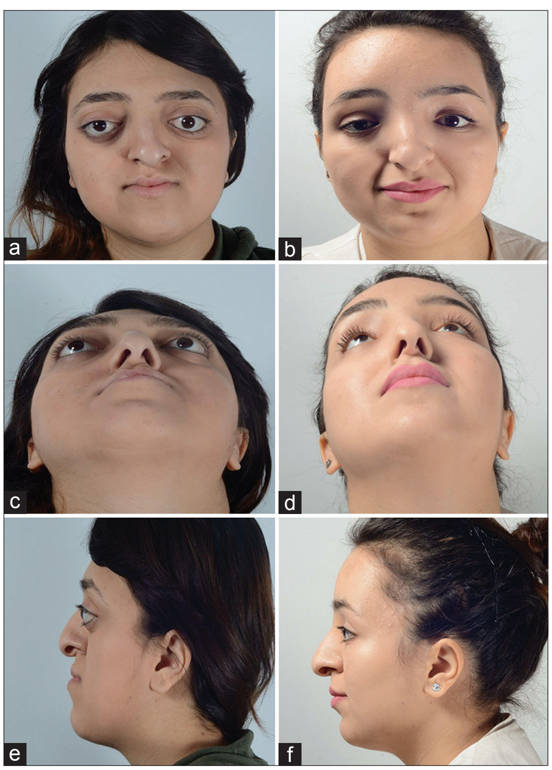

Figure 10: Pre‑(a, c, e) and post‑operative| (b, d, f) clinical view of the patient.

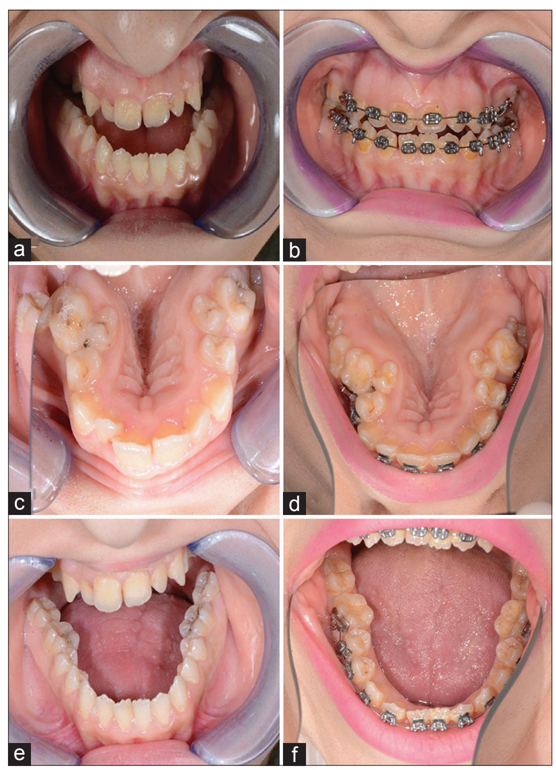

Figure 11: Comparative clinical examination of the patient: (a and b) Class I occlusion achieved; (c and d) upper dental arch and palate is wide; (e and f) lower dental arch is extended

lobes were elevated. This maneuver helps to perform osteotomy of orbital roof. Then circular orbitotomy was performed on both sides starting from lateral and medial orbital walls and finalized on inferior wall, which has been done blunt by the means of chisel [Figure 6].

In the next step pterygo-maxillary disjunction has been done blunt via transoral approach by curved chisel. The last step of segmentation entailed a straight chisel that was placed in the region of crista galli and separation of the vomer bone has been done. Thus, total segmentation of midface was achieved allowing to mobilize this block forwards and downwards.

Next to segmentation placing of distractors was performed according to virtual protocol data. Distractors were activated on the distance planned to be reached and placed to their original position. At the same time second team preformed fontal bone squama to finalize cranioplasty. All segments of monbloc were fixed together and final suturing was done [Figure 7].

Postoperative follow-up

Patient recovered in Intensive Care Unit for 5 days. 7 days postoperative activation of distractors was performed. Distraction schedule included 0, 7-1 mm of distraction per day over 20 days period. Typical symptoms of facial pain and headache have been observed. Following distraction 9 months of stabilization is required. At the same time intraoral palatal expansion device was used for horizontal maxillary expansion. 9 months after complete stabilization of bony structures, internal distractors removal and lateral canthopexy was performed.

On 15 months of follow-up clinical reduction of chief symptoms was noted, including restoring of facial harmony, achieving normal Class I occlusion, treating hypertelorism and bite correction making eating and chewing easier [Figure 8]. Postoperative CT scan showed midface and forehead advancement done properly [Figure 9].

DISCUSSION

The aim of this article is to describe a case of Crouzon syndrome treated by the means of distraction frontofacial advancement supported by virtual computer planning**.** It becomes the gold standard in Crouzon management because of increase in Class II relationship, this frontofacial monobloc advancement is described by Tessier and published by Ortiz-Monasterio *et al*.[10,11] The general idea of monobloc advancement is to form single piece bone unit which is composed of both from upper dental arch, whole maxillae, zygoma-orbital region and frontal bone. It helps to address three general anatomical problems related with Crouzon syndrome: (1) malocclusion; (2) orbital impairment; (3) tower-like deformity of the forehead [Figures 10 and 11]. However standard approach means that a great displacement could lead to "dead space" formation in retro frontal region communicating with the upper part of the nasal cavity, a cause of meningitis and permanent cerebrospinal rhinorrhea. On other hand application of distractors and their progressive advance, the brain has time to fill this space. In addition, distraction allows greater advances.

At the same time, intraoperative solution for such single piece bone unit formation and distractors positioning could be time-consuming and lead to unfavorable distractors positioning and unpredictable outcomes. The application of virtual planning protocol allows to determine osteotomy lines, monobloc design as well as the type and positioning of the distraction devices. Moreover, a virtual cephalometry supported planning, improvements and predictability of advancements provides surgeon with standpoints for one. It also helps to work out distraction protocol and limits control X-ray investigations during distraction process. At the same time, usage of specific software could also increase the accuracy of preoperative diagnosis by single CT scan investigation.

CONCLUSION

The application of virtual planning seems to be essential for monobloc advancement design both on surgical step and distraction step. It helps perform diagnosis and work out treatment protocol, increasing the accuracy of osteotomy and advancement as well. Moreover, virtual technologies allow to obtain prognosis in cases of application of distraction osteogenesis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Tessier P, Rougier J, Wolfe SA, editors. The craniofaciostenoses (CFS): The crouzon and apert disease, the plagiocephalies. In: Plastic Surgery of the Orbits and Eyelids. New York: Masson; 1981a. p. 200-3.
2. Posnick JC, Ruiz RL. The craniofacial dysostosis syndromes: Current surgical thinking and future directions. Cleft Palate Craniofac J 2000;37:433.
3. Adolphs N, Klein M, Haberl EJ, Menneking H, Hoffmeister B. Frontofacial advancement by internal distraction devices. A technical modification for the management of craniofacial dysostosis in early childhood. Int J Oral Maxillofac Surg 2012;41:777-82.
4. Arnaud E, Di Rocco F. Faciocraniosynostosis: Monobloc frontofacial osteotomy replacing the two-stage strategy? Childs Nerv Syst 2012;28:1557-64.
5. Jackson IT, Munro IR, Salyer KE, Whitaker LA, editors. Craniofacial dysostosis: Late cranial surgery (including forehead and vault). In: Atlas of Craniomax-Illofacial Surgery. Lewiston, NY, U.S.A: Mosby; 1982. p. 499-542.

6. Kamoshima Y, Sawamura Y, Yoshino M, Kawashima K. Frontofacial monobloc advancement using gradual bone distraction method. J Pediatr Surg 2008;43:1944-8.
7. Ko EW, Chen PK, Tai IC, Huang CS. Fronto-facial monobloc distraction in syndromic craniosynostosis. Three-dimensional evaluation of treatment outcome and facial growth. Int J Oral Maxillofac Surg 2012;41:20-7.
8. Nada RM, Sugar AW, Wijdeveld MG, Borstlap WA, Clauser L, Hoffmeister B, *et al.* Current practice of distraction osteogenesis for craniofacial anomalies in Europe: A web based survey. J Craniomaxillofac Surg 2010;38:83-9.
9. Nout E, Cesteleyn LL, van der Wal KG, van Adrichem LN, Mathijssen IM, Wolvius EB. Advancement of the midface, from conventional Le Fort III osteotomy to Le Fort III distraction: Review of the literature. Int J Oral Maxillofac Surg 2008;37:781-9.
10. Guichard B, Davrou J, Neiva C, Devauchelle B. Midface osteotomies lines: Evolution by Paul Tessier, the second Tessier classification. J Craniomaxillofac Surg 2013;41:504-15.
11. Ortiz-Monasterio F, del Campo AF, Carrillo A. Advancement of the orbits and the midface in one piece, combined with frontal repositioning, for the correction of Crouzon's deformities. Plast Reconstr Surg 1978;61:507-16.