

We are IntechOpen, the world's leading publisher of Open Access books Built by scientists, for scientists

6,900

Open access books available

186,000

International authors and editors

200M

Downloads

Our authors are among the

154

Countries delivered to

TOP 1%

most cited scientists

12.2%

Contributors from top 500 universities



WEB OF SCIENCE™

Selection of our books indexed in the Book Citation Index
in Web of Science™ Core Collection (BKCI)

Interested in publishing with us?
Contact book.department@intechopen.com

Numbers displayed above are based on latest data collected.
For more information visit www.intechopen.com



Advances in Craniofacial Surgery

Hamid Mahmoud Hashemi,
Farnoosh Mohammadi and
Shiva Solahaye Kahnemouii

Additional information is available at the end of the chapter

<http://dx.doi.org/10.5772/63739>

Abstract

Calvaria development initiates by growth from primary ossification centers meeting each other to form suture sites. The term craniosynostosis describes premature fusion of one or more of the calvarial sutures. Deformities are usually observable during the first few months of the newborn's life. The premature fusion of sutures could produce intracranial pressure elevation and consequently lead to abnormal neurocognitive = neurologic development. Patients with craniosynostosis require surgical plans containing multiple surgical staging. In the following chapter, we present our experience in surgical treatment of children with various craniosynostosis syndromes.

Keywords: craniosynostosis, craniofacial surgery, syndrome, surgical operation, frontal advancement

1. Congenital craniofacial abnormalities

The term craniofacial anomaly describes all congenital deformities of the cranium and the face [1]. A deformity is an alteration in shape due to unusual pressure or positioning in the uterus during late pregnancy. Some deformities resolve spontaneously without any interference within a few days after the newborn's birth, but others persist and require surgical treatment (**Figure 1**) [1, 2].



Figure 1. Congenital craniofacial abnormalities.

A malformation describes as an error in normal development of the organs or tissues. Malformations may arise from chromosomal abnormalities, teratogenic agents, single-gene defects, or a combination of genetic and environmental factors; albeit few cases are idiopathic [1, 2].

2. Craniofacial abnormalities (CFA)

Various craniofacial abnormalities result from mal-development of the first and the second visceral arches, which form the facial bones and ears during the second month of gestation. Causes include several thousand genetic syndromes as well as prenatal environmental factors [3]. Each specific congenital anomaly may be associated with several different genetic syndromes. The patients should be evaluated for other probable associated physical anomalies and delayed development that may require treatment and involvement.

3. Craniosynostosis

Craniosynostosis describes as early fusion of one or more of the cranial sutures of infant's head. **Figures 2 and 3** show the different sutures, which could be involved in craniosynostosis syndromes [3].

Figure 2. Different sutures which could be involved in craniosynostosis syndromes.

IntechOpen

Figure 3. Sutures which could be involved in craniosynostosis.

During brain growth, open sutures allow the skull to expand and develop a normal head shape. If one or more of the sutures closes early, it causes the skull to expand in the direction of the open sutures lead to an abnormal head shape and may cause increased pressure on the growing brain in severe cases (**Figure 4**) [3, 4].

Figure 4. Abnormal head shape from early suture closure.

3.1. Types of craniosynostoses

In sagittal synostosis also named scaphocephaly, the sagittal suture is prematurely closed. So the head does not expand in width and grows long and narrow to accommodate the growing brain. [In Greek, the word skaphe means “light boat or skiff” and kephale means “Head” which resembles an inverted boat]. The sagittal suture is statistically the most common single suture involved in craniosynostosis.

Metopism is known as a condition of having a persistent metopic suture or persistence of the frontal metopic suture in the adult human calvarium. The main factor of metopic synostosis is to increase the volume of the anterior cranial fossa (**Figure 5**) [5, 6].

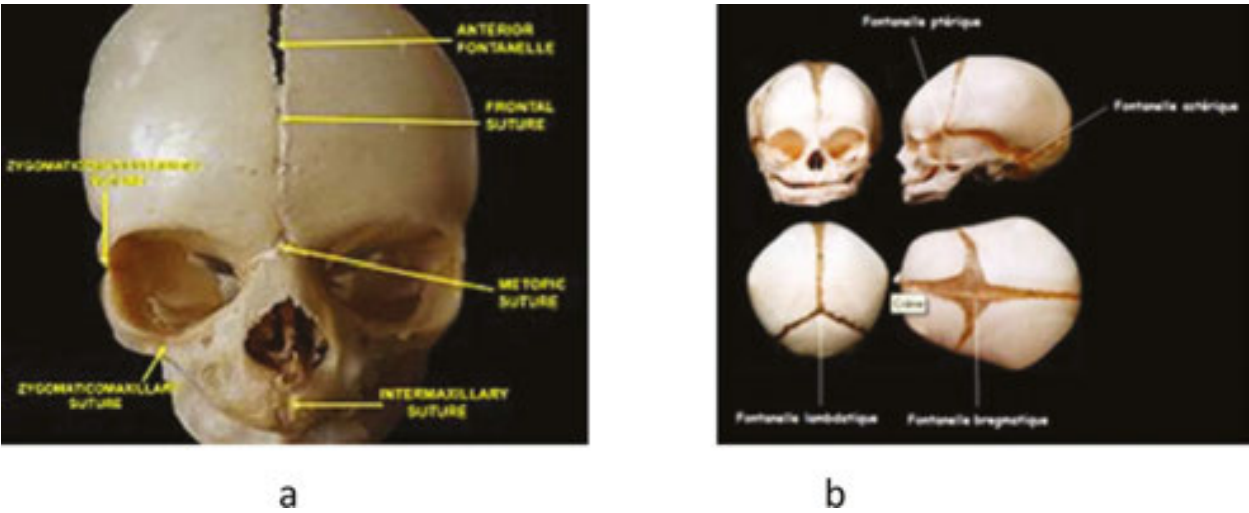


Figure 5. (a, b) Baby skull and the metopic (frontal) suture.

During the embryonic period at the frontal region, there is a membranous tissue between the right and the left halves. On each half, a primary ossification center manifests about the end of the second month of the fetus. At birth, the frontal bone contains two portions, separated by the metopic or frontal suture. Normally, the frontal suture is obliterated, except at its lower part until the 8th year, but rarely persists during the entire lifetime (**Figure 6**) [5, 6].

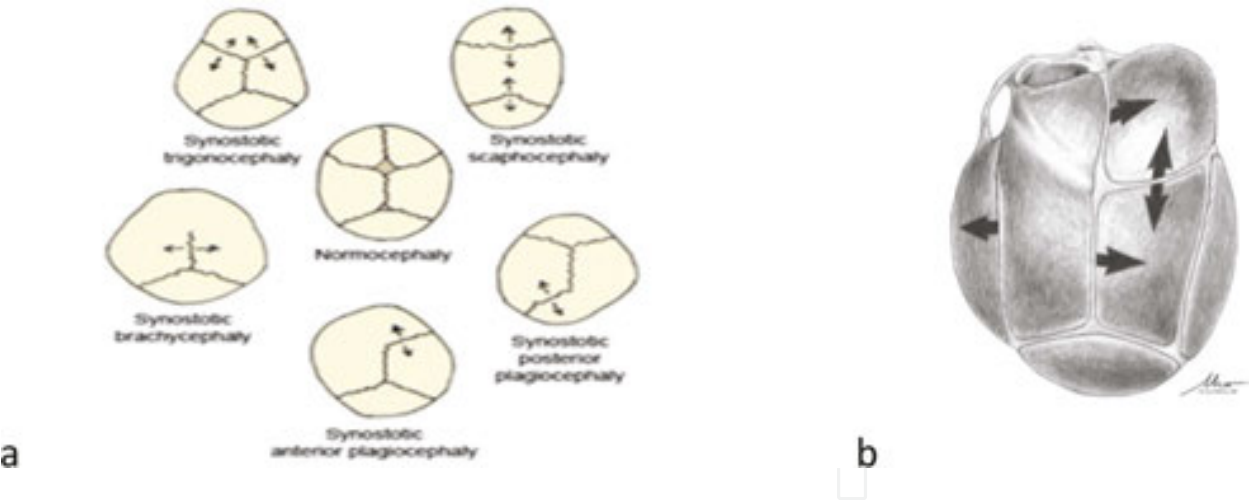


Figure 6. Various craniosynostoses. (a) Various types of craniosynostoses. (b) Possible directions of craniosynostosis in calvarium sutures

Plagiocephaly, also known as flat head syndrome, is a condition with asymmetrical distortion and flattening of one side of the skull. The coronal suture goes from ear to ear on the top of the head. Early closure of one side, unilateral coronal synostosis creates a flattened appearance on that side at forehead and orbital rim areas and causes the “winking” effect. It divides into two groups, namely synostotic plagiocephaly, with one or more fused cranial sutures, and nonsynostotic (deformational) plagiocephaly [7, 8].

Brachycephaly [7, 8] describes a very wide head shape with a flattening across the whole back of the head. In brachycephaly, both sides are fused. The patient may have a very flat and recessed forehead. Flat spot on the back or one side of the head may be caused by remaining in a supine position for a long time [5–8].

3.2. Confirmation of diagnosis of craniosynostosis

The first clue is a misshaped head. The anterior fontanel or soft spot may be open or fused. Diagnosis is confirmed by x-rays like CT scan evaluation to make sure there are no underlying brain abnormalities [5, 8].

Eye anomalies may include the following:

- Hypertelorism [9]
- Hypotelorism [9]
- Coloboma [10, 11]
- Microphthalmia [9–12]
- Anophthalmia [9–12]

3.3. Classification of craniofacial anomalies

Craniofacial anomalies are divided into three main subgroups [5]:

- Craniosynostoses [13–15]
- Craniofacial clefts
- Miscellaneous craniofacial anomalies

Types of craniosynostosis include the following:

(1) Primary craniosynostosis

This is an idiopathic developmental error occurring in otherwise normal conditions with no previous familial incidence.

(2) Secondary craniosynostosis

Premature fusion of the cranial sutures could result from failure of brain growth as in microcephaly or an encephaloclastic process which occurs during the first years of the newborn's life. When severe hydrocephalus has been treated with a low-pressure shunt, a similar process occurs.

(3) Metabolic craniosynostosis

Premature fusion of the sutures may arise due to the obvious biochemical disorders (mucopolysaccharidoses, rickets, hypophosphatasia, or hypercalcemia) (**Table 1**).

Affected suture	Sagittal	Metopic	Unicoronal	Bicoronal	Multiple sutures	Oxycephaly	Kleeblattschädel
Traditional name	Scaphocephaly	Trigonocephaly	Plagiocephaly	Brachycephaly	Turricephaly	Sharp skull	Clover-leaf skull
Literal translation	Boat skull	Triangle skull	Oblique skull	Short skull	Acrocephaly	Decreased	Decreased
Skull length	Increased	Normal	Decrease	Decreased	Tower skull	Increased	Increased
Skull width	Decreased	Increased	normal	Increased	Decreased	Increased	Increased
Skull height	Normal or increased	Increased	Increased		Increased		

Table 1. The various types of calvarial deformity.

Side problems due to the calvarial synostosis could be as follows: [16–18]

- Raised intracranial pressure
- Exorbitism and orbitostenosis
- Orbital hypertelorism
- Orbital dystopia
- Airway restriction
- Speech problems
- Mastication problems

Syndromic craniofacial synostosis usually has autosomal dominant genetic causes; these syndromes affect growth of the midface and the skull base. These conditions lead to insufficient space for the growing brain so intracranial pressure rises. Also they cause airway restriction and various ophthalmologic problems.

3.3.1. Apert syndrome

Apert syndrome is an autosomal dominant disorder and is a congenital disorder characterized. Acrocephalosyndactyly is an autosomal dominant disorder. Males and females are affected equally [19]. In Greek, “acro” means “peak,” referring to the “peaked” head and “Cephalo” means “head.” Almost certainly, all cases are sporadic and have de novo mutations or environmental insult to the genome [20–25]. It is caused by a defect on the fibroblast growth factor receptor 2 gene on chromosome number 10; usually due to a C to G mutation at the 755 position on the FGFR2 gene that causes a Ser to Trp change in the protein [15].

3.3.1.1. Apert syndrome characteristics

- Unusual headshape
- Wide-set eyes with shallow sockets and poorly closing eyelids
- Recessed midface
- Beak-shaped nose
- Underdeveloped jaws with under bite and crowded dentition
- Cleft palate
- Impaired hearing capabilities
- Atypical spine development
- Fused fingers and toes
- Limited intellectual development
- Gastrointestinal malfunctions
- Cardiac malformations
- Hyperhydrosis and heavy sweetening
- Severe acne
- Patches of lost hair (**Figure 7**)





Figure 7. Photograph of a 9-year-old patient with history of cranioplastic surgery at the age of 1 year. Preoperative photograph of the patient, above. Photograph of the patient during the corrective surgery: the frontofacial monobloc advancement surgery, middle. Note the globs in both sides of the calvarium vault are exposed in the superior view. Postoperative photograph of the patient: The same patient 8 months after the frontofacial monobloc advancement surgery, above.

3.3.2. Carpenter syndrome

For the first time, George Carpenter (1859–1910) described this condition and its related features [24–28].

Its features contain as follows:

- Tower-shaped skull because of craniosynostosis
- Additional or fused fingers and toes
- Obesity
- Reduced height

Patients may have average intellectual capacity and intellectual disability [29–32].

Malformation of the skull is the primary diagnostic factor. The two most common types of craniosynostosis in this syndrome are the sagittal and the bicoronal sutures. Mutations in the RAB23 gene located on chromosome 6 are responsible for these features. Also three key SNPs in the MEGF8 gene, located on chromosome number 19 at 19q13.2, have been identified as primary causes [33].

3.3.2.1. Carpenter syndrome surgical treatment

Surgical operations to correct the malformations of the skull should be done during the first year of infancy because modifying the bones is so much easier when the skull is in the growing stage; therefore, performing surgery at a young age increases the chance of obtaining good results. The fused sutures should be broken to allow for brain growth. The cranial plates are removed, reshaped, and replaced back on the skull. Despite the broken sutures, they will quickly refuse, and sometimes holes form in the plates allowing cerebral spinal fluid to escape into cyst-like structures on the external surface of the head (**Figure 8**).



Figure 8. A 3-year-old boy with carpenter syndrome: Preoperative photographs of the patient, above. Photographs during the monobloc frontofacial advancement surgery, middle. Postoperative photographs of the same patient: 6 month after the monobloc frontofacial advancement surgery, above.

3.3.3. Crouzon syndrome's surgical treatment

3.3.3.1. Crouzon syndrome

Crouzon syndrome also known as a branchial arch syndrome is an autosomal dominant genetic disorder that affects the first branchial (pharyngeal) arch. Mutation in the FGFR2 and FGFR3, located on chromosome 10, is responsible for it. For the first time, Octave Crouzon, a French physician, described the mentioned condition [34–39]. He noted that the affected patients were a mother and her daughter, implying a genetic basis. Low-set ears are a typical characteristics in all branchial arch syndromes. It usually presents as brachycephaly resulting in an appearance of a short and broad head. Exophthalmos and psittichorhina could be accompanied. External strabismus is a common feature. Hypoplastic maxilla and insufficient growth of the midface result in relative mandibular prognathism appearance so give the patient having a concave face. Typically, surgery is used to prevent the closure of skull sutures around the developing brain. Otherwise, blindness and mental retardation are typical outcomes of premature suture fusion (**Figure 9**).

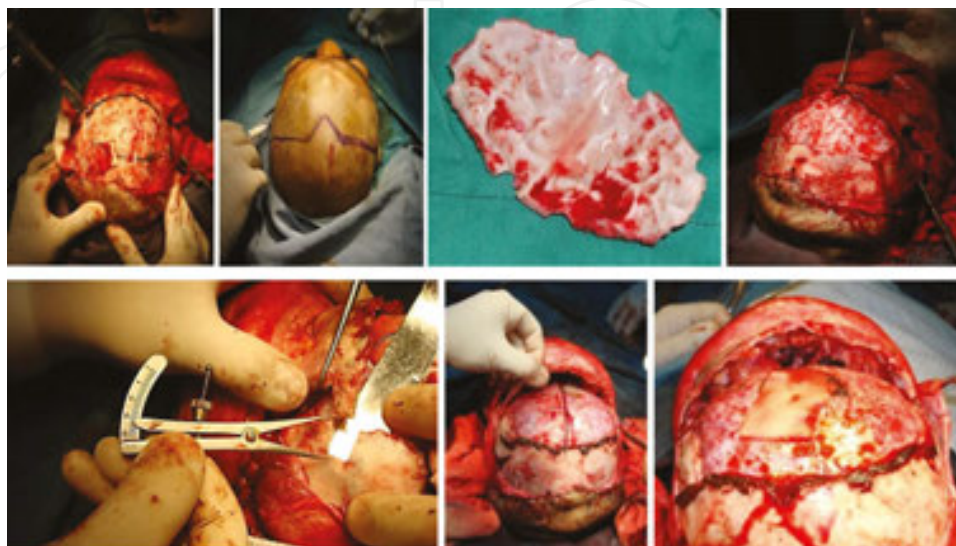




Figure 9. Photograph of the patient an 8-year-old girl with the Crouzon syndrome: Preoperative photographs of the patient, above. Exposure the calvaria by the Bicoronal Flap, middle. Note: pressure effect of brain on the inner table of the frontal bone segment is obvious. Monobloc advancement: 17 mm advancement is obvious in zygomatic arch region. Note: Creating frontal intracranial volume by the surgery, below. The same patient 8 years later before and after orthognathic surgery, last.

3.3.4. Goldenhar syndrome

Goldenhar syndrome is defined as congenital defects due to involvement of the first arches during intrauterine development and includes incomplete development of the ear, lip, nose, mandible, and soft palate [40–45]. It is also known as oculo-auriculo-vertebral (OAV) syn-

drome. In 1952, this condition was documented by the Belgian ophthalmologist Maurice Goldenhar for the first time.

3.3.4.1. Signs and symptoms

Incomplete development of the ear, nose, soft palate, lip, and mandible on usually one side of the body is the main hallmarks. Some patients will show growing problems with internal organs, especially the lungs, heart, and kidneys [47, 48].

3.3.4.2. Cause

The cause is unknown but involves the branchial arch development in the first trimester and probably is multifactorial with genetic components which would account for certain familial patterns [45, 46].

3.3.5. Treatment

Surgical intervention is essential to help the patient continue natural development, for example, jaw distraction; bone grafts; ocular dermoid debulking; repairing palatal or lipclefts; repairing heart malformations or spinal surgery. Patients may require assistance of hearing aids or eye glasses as they grow.

3.3.6. Muenke syndrome

Muenke syndrome also known as FGFR3-related craniosynostosis is distinguished by the premature closure of certain bones of the skull during development. It was first described by Maximilian Muenke [50–57].

3.3.6.1. Signs and symptoms

Affected patients have premature fusion of skull bones especially along the coronal suture leading to abnormally shaped head, wide-set eyes, low-set ears, and flattened cheekbones and sometimes have macrocephaly and hearing loss.

Most affected patients have normal intellect with no mental retardation, but developmental delay and learning disabilities are possible problems. The condition is caused by a FGFR3 gene mutation with an autosomal dominant pattern. This mutation causes the overexpression and overactive FGFR3 protein, so this protein interferes with normal bone growth leading to premature fusion of the skull bones. Strabismus is the most common ocular finding [49].

3.3.6.2. Treatment

Surgical correction of the abnormal skull shape and coronal craniosynostosis is planned although abnormal growth patterns continue during the growing years.

3.3.7. Parry-Romberg syndrome

Parry–Romberg syndrome is a neurocutaneous syndrome distinguished by progressive shrinkage and degeneration of the tissues beneath the superficial skin, usually on only one side of the face as hemifacial atrophy but occasionally extending to other parts of the body known as progressive hemifacial atrophy. It probably has an autoimmune background; the syndrome could be considered a variant of localized scleroderma and a type of connective tissue disease. The condition is usually accompanied by significant neurological, ocular, and oral signs and symptoms [58–64].

During the first or early second decade of life, symptoms and physical findings usually become apparent. The average age of onset is 9 years of age, and the majority of individuals experience symptoms before 20 years of age. The disease may progress for several years before eventually going into remission.

Neurological abnormalities might be afflicted with trigeminal neuralgia. Some patients develop seizures. The type of the seizure is typically Jacksonian and occurs on the side contralateral to the affected side of the face. Enophthalmos is the most common eye abnormality.

Oral tissues are commonly involved in Parry-Romberg syndrome. Affected individuals develop dental abnormalities on the affected side include resorption of the dental roots, dental root exposure, or delayed tooth eruption. Patients may have difficulty or inability in normal mouth opening or other jaw symptoms, including temporomandibular joint disorder and spasm of the muscles of mastication on the affected side. Patients may experience atrophy of one side of the upper lip and tongue.

Diagnosis could be made by history and physical examination according to the facial asymmetry. In patients with neurological symptoms like migraine or seizures, brain MRI scan is the choice imaging modality. Detection of autoantibodies by diagnostic lumbar puncture and sera tests may be indicated for those whom present seizures of recent onset. Immunosuppressive drugs might be indicated to control the course of the disease. Autologous fat transfer or fat grafts could be useful for of a more acceptable facial contour. Larger defects require microsurgical reconstructive surgeries.

3.3.8. Pfeiffer syndrome

For the first time, in 1964, Rudolf Arthur Pfeiffer described a list of features included a coronal synostosis, turribrachycephaly, and maxillary hypoplasia (**Figure 10**). Pfeiffer syndrome, a genetic disorder, distinguished by premature craniosynostosis also affects the hands and feet bones. The patients usually have hearing loss and dental problems. Extracranial features include broad thumbs and toes of the hands and feet. The thumbs and first big toes are wide and bend away from the other digits. Short fingers and toes (brachydactyly) and webbing or syndactyly may be seen [65–72].



Figure 10. Photograph of a Pfeiffer syndrome patient before and after the corrective surgery.

Pfeiffer syndrome has three subtypes:

- **Type 1:** Classic type has symptoms as described above. Usually, they have normal intelligence and a normal life span.
- **Types 2 and 3** are more severe forms that often accompany nervous system involvements. The premature fusion of skull bones provides limitation for brain growth leading to delayed development and more neurological problems.

Type 2 is differentiated from type 3 by a cloverleaf-shaped head, caused by more extensive fusion of bones of the skull.

3.3.9. Saethre-Chotzen syndrome

Saethre-Chotzen syndrome (SCS) or Acrocephalosyndactyly type III is a congenital disorder accompanying cranio synostosis and affects the craniofacial shape resulting in a cone-shaped head and an asymmetrical face [73–77]. Patients have ptosis, hypertelorism, and syndactyly. In severe cases, mild-to-moderate mental retardation or learning disabilities can be noted. Depending on the level of severity of the condition, medical or surgical intervention may be needed (**Figure 11**).

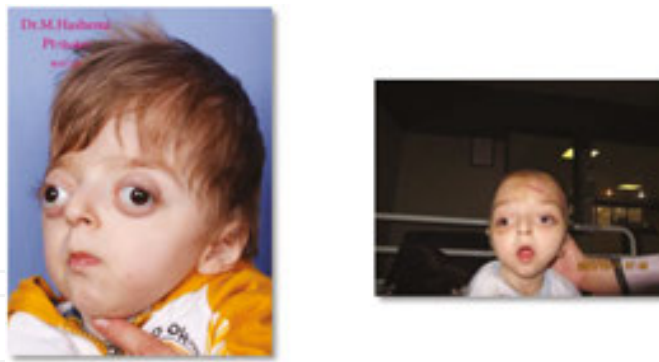


Figure 11. Photograph of the patient with the Saethre-Chotzen syndrome before and after the corrective surgery.

Even within the family, affected individuals have different features. The majority of individuals with SCS are moderately affected, with uneven facial features and a relatively flat face due to underdeveloped eye sockets, malar bones, and the mandible. Growth delays like relatively short stature also are noted. Albeit most individuals with SCS are of normal intelligence, some individuals may have mild-to-moderate mental retardation (IQ levels 50–70).

3.3.9.1. Cranial defects:

- Flat and asymmetric head and face
- Acrocephaly and/or brachycephaly and/or dolichocephaly
- Short head from front to the back
- Lopsided face
- Low-set hairline causing forehead to appear tall and wide

3.3.9.2. Defects of the hands and feet

- Syndactyly between the second and third fingers and between the second and third toes
- Short fingers and toes (brachydactyly)
- Broad thumb and/or a broad hallux (big toe) with a valgus deformity (outward angulation of the distal segment of a bone/joint)
- Hands have a single palmer flexion crease

3.3.9.3. Ocular defects

- Strabismus
- Hypertelorism
- Tear duct stenosis

- Ptosis
- Nearsightedness
- Epicanthal folds
- Blepharophimosis
- Optic atrophy
- Refractory errors

3.3.9.4. *Ear, nose, and mouth defects*

- Small, low-set ears may be rotated somewhat backward and has a bulging pinna.
- Beaked nose; slightly bent downward at tip, that is, slightly off center and contains a deviated septum.
- Malocclusion associated with dental abnormalities, including enamel hypoplasia and hyperdontia and peg teeth.
- Cleft palate with high arch.

3.3.9.5. *Less common defects*

- Short stature
- Vertebral fusions
- Congenital heart problems
- Speech problems
- Malformed rectum
- Cryptorchidism
- Renal abnormalities
- Personality disorders

3.3.10. *Treacher Collins syndrome (TCS)*

Treacher Collins-Franceschetti syndrome [78–84] or mandibulofacial dysostosis is distinguished by craniofacial deformities such as absence of malar bones and has an autosomal dominant pattern. The typical accompanying features include downward-slanting eyes, micrognathia, conductive hearing loss, underdeveloped zygoma, drooping part of the lateral lower eyelids, and malformed or absent ears (**Figure 12**).



Figure 12. Photograph of a patient with Treacher Collins syndrome before and after corrective surgical treatment.

The presentation of symptoms varies. Some individuals may be so mildly affected and remain undiagnosed; others may show severe facial involvement and life-threatening airway compromises. Most of the features are bilateral and are already recognizable at birth.

3.3.10.1. Accompanying abnormalities:

- Facial bones hypoplasia
- Ear anomalies
- Eye problems
- Cleft palate
- Airway problems

Dental anomalies include tooth agenesis, enamel deformities, and misplacement of the maxillary first molars could be noted and usually in combination with mandible hypoplasia results in a malocclusion with problems in food intake and the ability of chewing.

3.3.10.2. Less frequent features:

- Nasal deformity
- High-arched palate
- Coloboma of the upper lid
- Ocular hypertelorism

- Choanal atresia
- Macrostomia
- Preauricular hair displacement

Intelligence of TCS patients is generally normal. The psychological and social problems associated with facial deformity may affect the quality of life in a number of patients.

3.3.10.3. *Mandible:*

- M0: Normal mandible.
- M1: Small mandible and glenoid fossa with short ramus.
- M2: Short and abnormally shaped ramus.
 1. 2A: Glenoid fossa in anatomical acceptable position.
 2. 2B: Temporomandibular joint inferiorly, medially, anteriorly displaced with severely hypoplastic condyle.
- M3: Complete absence of ramus, glenoid fossa, and TMJ.

3.3.10.4. *Ears*

- E0: Normal ear.
- E1: Minor hypoplasia and cupping with all structures present.
- E2: Absence of external auditory canal with variable hypoplasia of the auricle.
- E3: Malposition of the lobule with absent auricle, lobular remnant usually inferior anteriorly displaced.

The treatment requires a multidisciplinary approach. Imaging evaluation consists of X-rays, CT scans, MRI, and ultrasound.

The primary concerns are breathing and feeding problems. Some when even a tracheostomy is requisite to adequate airway preservation. Sometimes during the protection of the airway a gastrostomy can be done to provide an adequate caloric intake. Depending on the development state, surgical treatment is done for restitution of a normal facial contour and the structure of the face at certain times. Hearing loss is caused by deformed structures in the outer and middle ear. The hearing loss is generally bilateral with a conductive loss of about 50–70 dB. The ossicular chain is often malformed even in cases with normal auricles and open external auditory canals.

Surgical treatment can be done early during the first year of life or later. For psychological and social support, every effort is made as soon as possible [80–84].

3.4. Surgical treatment of craniosynostosis

Early: (Before 1 year of age)

1. Strip craniectomies
 - Limited
 - Extended
2. Frontal bone advancement with or without strip craniectomies
3. Cranial vault remodeling
4. Monobloc or craniofacial advancement
5. Shunt surgery for hydrocephalus

Late (After 1 year of age)

1. Frontal bone advancement
2. Le fort III and frontal bone advancement
4. Monobloc or craniofacial advancement
5. Le fort II advancement
6. Maxillary/mandibular/zygomatic surgery

Early surgical treatment of craniosynostosis

3.4.1. Before 1 year of age

The goals of surgery for newborn with a craniosynostosis are twofold:

- (1) Decompression of the intracranial space to reduce intracranial pressure, prevent visual problems, and permit normal mental development.
- (2) Achievement of satisfactory craniofacial form.

In this chapter, we discuss the late surgical treatment of craniosynostosis.

3.4.2. Late surgery

Late surgery is defined as surgical procedures performed on the patient with craniofacial synostosis after 1 year of age.

3.5. Le fort III advancement osteotomy

Gillies and Harrison reported the first high maxillary (modified Le fort III) osteotomy for craniofacial dysostosis. Through external incision, they performed transverse osteotomies that separate the nasal bones from the frontal bones. The osteotomy of the orbital floor was placed immediately within the infraorbital margin and extended across the floor of the orbit to the

medial orbital wall anterior to the lacrimal groove. Intermaxillary fixation was placed. Although no bone grafting was done, a satisfactory result was reported. Seven and one half years after this operation, the patient underwent further surgery to correct persistent exorbitism by removal of the medial portion of the orbital floor, and ox cartilage was also placed over the zygoma for contour improvement. The fate of the ox cartilage was not reported. Techniques of the Le fort III advancement osteotomy through a subcranial (Extracranial) route. Anesthesia is achieved through a transnasal endotracheal tube if the Le fort III advancement is to be performed without a tracheostomy [85–90].

The exposure of the facial skeleton is obtained through three incisions:

- (1) Scalp (Bicoronal)
- (2) Conjunctival or subciliary cutaneous
- (3) Buccal vestibular

The eyelid incisions can be avoided, but the operation is technically more difficult. The bicoronal incision and the raised scalp flap provide access in a subperiosteal plane to the lateral wall and floor of the orbit, to the root of the nose, and to the medial orbital wall. The subperiosteally raised area can then communicate with the area, which will be exposed through the conjunctival or eyelid incision (optional). After the scalp flap is raised, the periorbita is elevated from the roof, the floor, and the lateral and medial orbital walls are exposed; the medial canthal tendon is left undisturbed, and the lacrimal sac is elevated from the lacrimal groove. A transverse cut is made across the orbital floor and joins the inferior orbital fissure to the lower end of the medial wall osteotomy.

The lateral wall of the orbit is sectioned transversely in the region of the frontozygomatic suture line or above it. After retraction of orbital contents medially and the temporal muscle laterally, the lateral orbital wall is divided in a full thickness fashion at its junction with the cranium. The zygomatic arch is likewise sectioned. After all lines of osteotomy are verified, the midfacial skeleton may be loosened with the Rowe disimpaction forceps. Autogenous bone grafts are placed in the defects of the nasofrontal junction, lateral orbital wall, and pterygomaxillary fissure.

Le fort III osteotomy performed through a combined intracranial approach with advancement of the frontal bone.

After the neurosurgeon has removed the frontal bone segment, the supraorbital osteotomy is extended horizontally to the region of the temporal fossa and continued in a stepwise fashion inferiorly toward the base of the skull. A posterior extension is thus outlined, which guides the advancement and maintains bony contact. In a horizontal direction, osteotomy then transgresses the lateral orbital wall and follows a line through the orbital roof at about the junction of middle and posterior thirds of orbit. The procedure is then completed by performing the Le fort III osteotomy. In this way, a horizontal component is also advanced approximately 2 cm in height, containing the frontal bone, part of the roof, and the lateral wall of the orbit (**Figures 13 and 14**).

3.5.1. Monobloc advancement of orbits and midface [90–95]

To increase the orbital volume for the correction of the exorbitism, the subtotal orbits and midface could be advanced as a single skeletal segment. The lines of osteotomy are similar to the previously described for the combined Le fort III–frontal bone advancement except that the nasofrontal junction and frontozygomatic sutures are spared of osteotomies. The technique also has the advantage that a concomitant hypertelorism correction can be done; it suffers the disadvantages of an increased infection rate and limited orbital volume expansion.

3.5.2. Le fort III advancement

The most common indication for Le fort III advancement is the patient with midface hypoplasia and adequate zygomatic projection.



Figure 13. Photograph of a 24-year-old patient with midface deficiency and mandibular prognathism. Before the surgery, top. One week after the Le Fort III osteotomy and mandibular set back surgery, below.

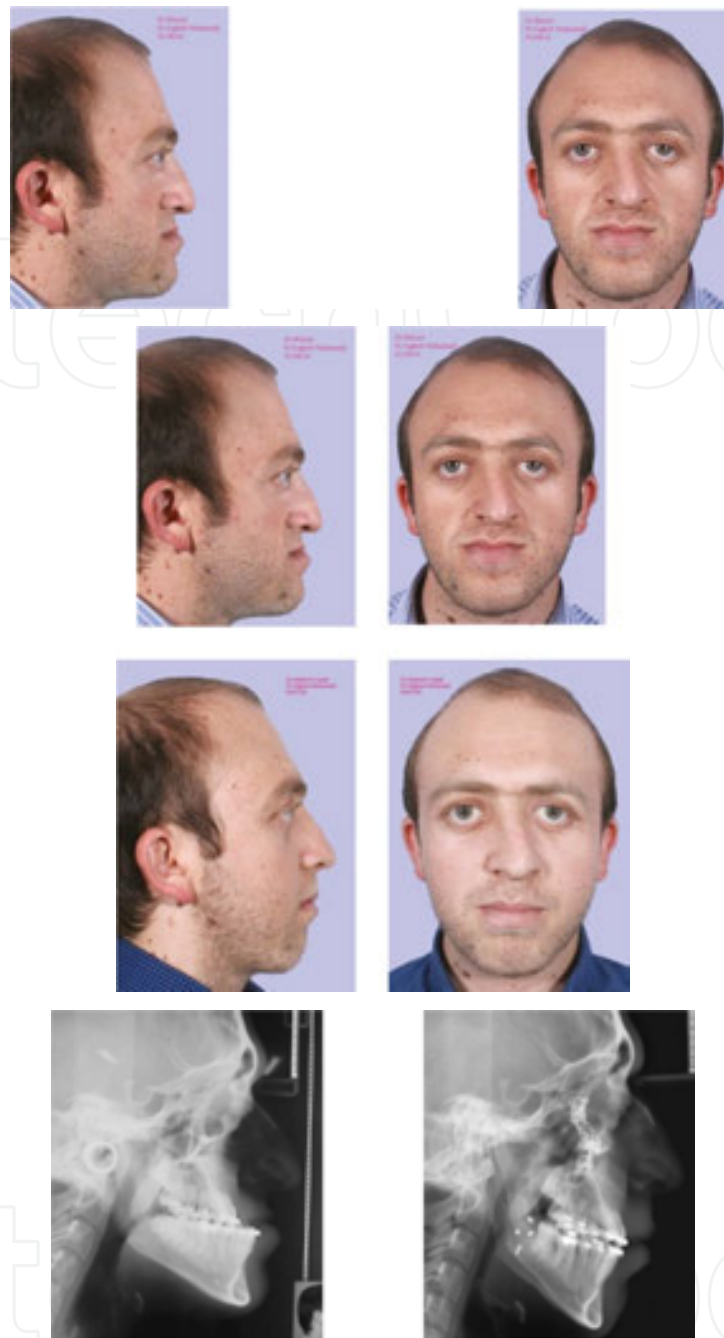


Figure 14. Photographs of a 32-year-old patient with midface deficiency and mandibular prognathism. Photographs of the patient before the orthognathic surgery, top. Photographs of the patient 1 year after the Le Fort III osteotomy and mandibular set back surgery, below.

3.6. Possible surgical complications

In the best of circumstances, surgical complications are inevitable but using theoretical knowledge and sufficient practical knowledge and requisite skills; complications can be reduced or be prevented. Despite all efforts in surgery in any surgery, including craniofacial surgeries, there are possible side effects and even a risk of death.

3.6.1. Some examples of problems during the surgery

- Anesthesia related
- Difficult airway
- Blood loss
- Electrolyte imbalance and hyponatremia
- Venous air or blood or fat embolism
- Intraoperative events
- Incision-related events

3.6.2. Miscellaneous events related by the surgery

- Periorbital or eye related events
- Bone wax granuloma formation
- Infection
- Neurological deficit
- Death

Author details

Hamid Mahmoud Hashemi, Farnoosh Mohammadi and Shiva Solahaye Kahnamouii*

*Address all correspondence to: Shiva.kahnamouii@gmail.com

Department of Oral and Maxillofacial Surgery, Tehran University of Medical Sciences, Tehran, Iran

References

- [1] Cohen, MM Jr. An etiologic and nosologic overview of craniosynostosis syndromes. *Birth Defects*. 1975;11:137–184.
- [2] Cohen, MM Jr. Craniosynostosis and syndromes with craniosynostosis: incidence, genetics, penetrance, variability and new syndrome updating. *Birth Defects*. 1979;15:13.
- [3] Posnick, Jeffrey C. *Orthognatic surgery: principles and practice*. Amsterdam: Elsevier. 2013, pp. 61–68.

- [4] Bertelson, TI. The premature synostosis of the cranial sutures. *Arch Ophthalmol* (Copenhagen). 1958;5:47–66 (suppl).
- [5] Zhou YQ, Ji J, Mu XZ, Zhang RH, Wei M, Yu ZY. Diagnosis and classification of congenital craniofacial cleft deformities. *J Craniofac Surg*. 2006;17(1):198–201.
- [6] Forbes BJ. Congenital craniofacial anomalies. *Curr Opin Ophthalmol*. 2010;21(5):367–374
- [7] In: EP Caronni (Ed.) *Craniofacial surgery*. Little Brown, Boston. 1985.
- [8] Crouzon, O. Dystosto secranio faciale herediture. *Bull Mem Soc Med Hosp Paris*. 1912;33:545.
- [9] Converse, JM, McCarthy, JG. Orbital hypertelorism. *Scand J Plast Reconstr Surg*. 1981;15:265–276.
- [10] Doxanas, MT, Anderson, RL. Eyebrows, eyelids and anterior orbit. In: MT Doxanas, RL Anderson (Eds.) *Clinical orbital anatomy*. Williams & Wilkins, Baltimore. 1984, p. 82.
- [11] Doxanas, MT, Anderson, RL. Eyebrows, eyelids and anterior orbit. In: MT Doxanas, RL Anderson (Eds.) *Clinical orbital anatomy*. Williams & Wilkins, Baltimore. 1984, p. 72.
- [12] Diamond, GR, Whitaker, LA. Ocular motility in craniofacial reconstruction. *Plast Reconstr Surg*. 1984;73:31–35.
- [13] Ortiz-Monasterio, F. Notes on the history of craniofacial surgery. In: Jackson IT, Munro IR, Salyer KE, editors. *Atlas craniomaxillofacial surgery*. St. Louis, Mosby. 1982, pp. vii–xii.
- [14] Matthews D. Craniofacial surgery: indications, assessment and complications. *Br J Plast Surg* 1979;32:96–105
- [15] Tessier P. The definitive plastic surgical treatment of the severe facial deformities of craniofacial dysostosis. Crouzon's and Apert's diseases. *Plast Reconstr Surg* 1971;48:419–442.
- [16] Whitaker LA, Munro IR, Salyer KE, Jackson IT, Ortiz-Monasterio F, Marchac D. Combined report of problems and complications in 793 craniofacial operations. *Plast Reconstr Surg* 1979;64:198–203.
- [17] Poole MD. Complications in craniofacial surgery. *Br J Plast Surg* 1988;41:608–613.
- [18] Converse JM, Wood-smith D, McCarthy JG. Report on a series of 50 craniofacial operations. *Plast Reconstr Surg* 1975;55:283–293.
- [19] Allam KA, Wan DC, Khwanngern K, Kawamoto HK, Tanna N, Perry A, Bradley JP. Treatment of apert syndrome: a long-term follow-up study. *Plast Reconstr Surg*. 2011;127(4):1601–1611.

- [20] Buchanan RC. Acrocephalosyndactyly or Apert's syndrome. *Br J Plast Surg.* 1968;21:406–418.
- [21] Cohen MM Jr. Cardiovascular anomalies in Apert-type acrocephalosyndactyly. *Birth Defects.* 1982; 8:239–241.
- [22] Cuttone JM, Brazis PT, Miller MT et al. Absence of the superior rectus muscle in Apert's syndrome. *J Pediatr Ophthalmol Strabis.* 1979;16:349–354.
- [23] Abe Bert Baker, Lowell H. Baker. Apert's syndrome. *Clin Neurol (Medical Dept., Harper & Row).* 1979;3: 47.
- [24] Carpenter G. Case of acrocephaly with other congenital malformations. *Proc R Soc Med.* 1909;2:45–53, 199–201.
- [25] Jenkins D, Seelow D, Jehee FS, Perlyn CA, Alonso LG, Bueno DF, Donnai D, Josifova D, Mathijssen IM, Morton JE, Orstavik KH, Sweeney E, Wall SA, Marsh JL, Nürnberg P, Passos-Bueno MR, Wilkie AO. RAB23 mutations in Carpenter syndrome imply an unexpected role for hedgehog signaling in cranial-suture development and obesity. *Am J Hum Genet.* 2007;80(6): 1162–1170.
- [26] Twigg SR, Lloyd D, Jenkins D, Elçioglu NE, Cooper CD, Al-Sannaa N, Annagür A, Gillesen-Kaesbach G, Hüning I, Knight SJ, Goodship JA, Keavney BD, Beales PL, Gileadi O, McGowan SJ, Wilkie AO. Mutations in multidomain protein MEGF8 identify a Carpenter syndrome subtype associated with defective lateralization. *Am J Hum Genet.* 2012;91(5):897–905.
- [27] Carpenter G. Two sisters showing malformations of the skull and other congenital abnormalities. *Rep Soc Study Dis Child Lond.* 1901;1:110–118.
- [28] Carpenter G. Case of acrocephaly, with other congenital malformations. *Proc R Soc Med.* 1909;2:45–53.
- [29] Temtamy SA. Carpenter's syndrome: acrocephalopolysyndactyly. *J Pediatr.* 1966;69:111–120.
- [30] Robinson LK, James HE, Mubarak SJ, Allen EJ, Jones KL. Carpenter syndrome: natural history and clinical spectrum. *Am J Med Genet.* 1985;20:461–469.
- [31] Cohen DM, Green JG, Miller J, Gorlin RJ, Reed JA. Acrocephalopolysyndactyly type II –Carpenter syndrome: clinical spectrum and an attempt at unification with Goodman and Summit syndromes. *Am J Med Genet.* 1987;28:311–324.
- [32] Dagan Jenkins, Dominik Seelow, Fernanda S. Jehee, Chad A. Perlyn, Luís G. Alonso, Daniela F. Bueno, Dian Donnai, Dragana Josifiova, Irene M. J. Mathijssen, Jenny E. V. Morton, Karen Helene Ørstavik, Elizabeth Sweeney, Steven A. Wall, Jeffrey L. Marsh, Peter Nürnberg, Maria Rita Passos-Bueno, Andrew O. M. Wilkie RAB23 mutations in carpenter syndrome imply an unexpected role for hedgehog signaling in cranial-suture development and obesity. *Am J Hum Genet.* 2007;80(6):1162–1170.

- [33] Carinci F, Pezzetti F, Locci P, et al. Apert and Crouzon syndromes: clinical findings, genes and extracellular matrix. *J Craniofac Surg*. 2005;3(16):361–366.
- [34] Hoefkens MF, Vermeij-Keers C, Vaandrager JM. Crouzon syndrome: phenotypic signs and symptoms of the postnatally expressed subtype. *J Craniofac Surg*. 2004 Mar;15(2): 233–40; discussion 241–2. Crouzon syndrome: phenotypic signs and symptoms of the postnatally expressed subtype. Hoefkens MF1, Vermeij-Keers C, Vaandrager JM.
- [35] Cohen MM Jr., Kreiborg S. Birth prevalence studies of the Crouzon syndrome: comparison of direct and indirect methods. *Clin Gene*. 1992;41:12–15.
- [36] Hlongwa P. Early orthodontic management of Crouzon syndrome: a case report. *J Maxillofac Oral Surg*. 2009;8:74–76.
- [37] Cohen MM Jr, Kreiborg S. Birth prevalence studies of the Crouzon syndrome: comparison of direct and indirect methods. *Clin Genet*. 1992;41:12–15.
- [38] Fogh-Andersen P. Craniofacial dysostosis (Crouzon's disease) as a dominant hereditary affection. *Nord Med*. 1943;18:993–996.
- [39] Goldenhar, M. Associations malformatives de l'oeil et de l'oreille. *Journal de Ginitique Humaine*. 1952;1:243.
- [40] Touliatou V, Fryssira H, Mavrou A, Kanavakis E, Kitsiou-Tzeli S. Clinical manifestations in 17 Greek patients with Goldenhar syndrome. *Genet. Couns*. 2006;17(3):359–370.
- [41] Zaka-ur-Rab Z, Mittal S, Optic nerve head Drusen in Goldenhar syndrome (PDF). *JK Science* 2007;9(1):33–34.
- [42] Araneta MR, Moore CA, Olney RS, et al. Goldenhar syndrome among infants born in military hospitals to Gulf War veterans. *Teratology* 1997;56(4):244–251.
- [43] Ng YY, Hu JM, Su PH, Chen JY, Yang MS, Chen SJ. Goldenhar syndrome (oculoauriculovertebral dysplasia): report of one case. *Acta Paediatr Taiwan*. 2006;47(3):142–145.
- [44] Riyaz A, Riyaz N. Goldenhar syndrome with unusual features. *Indian J Dermatol Venereol Leprol* 1999; 65:143–144.
- [45] Kulkarni VV, Shah MD, Parikh AA. Goldenhar syndrome: A case report. *J Postgrad Med* 1985; 31:177–179.
- [46] Reddy MV, Reddy PP, Usha Rani P, HemaBindu L. Facio-auricular vertebral syndrome – a case report. *Indian J Hum Genet* 2005; 11:156–158.
- [47] Nakajima H, Goto G, Tanaka N, Ashiya H, Ibukiyama C. Goldenhar syndrome associated with various cardiovascular malformations. *Jpn Circ J* 1998; 62:617–620.
- [48] Rad EM. Goldenhar syndrome with right circumflex aortic arch, severe coarctation and vascular ring in a twin pregnancy. *Ann Pediatr Cardiol* 2014; 7(3):217–220.

- [49] Barbosa M, Almeida Md R, Reis-Lima M, Pinto-Basto J, dos Santos HG. Muenke syndrome with osteochondroma. *Am J Med Genet Part A*. 2009; 149A:260–261.
- [50] Rannan-Eliya SV, Taylor IB, De Heer IM, Van Den Ouweland AM, Wall SA, Wilkie AO. Paternal origin of FGFR3 mutations in Muenke-type craniosynostosis. *Hum Genet*. 2004; 115(3): 200–207.
- [51] Grosso S, Farnetani MA, Berardi R, Bartalini G, Carpentieri M, Galluzzi P, Mostardini R, Morgese G, Balestri P. Medial temporal lobe dysgenesis in Muenke syndrome and hypochondroplasia. *Am J Med Genet A*. 2003; 120A:88–91.
- [52] Agochukwu NB, Solomon BD, Gropman AL, Muenke M. Epilepsy in Muenke syndrome: FGFR3-related craniosynostosis. *Pediatr Neurol*. 2012a; 47:355–361.
- [53] Agochukwu NB, Solomon BD, Muenke M. Impact of genetics on the diagnosis and clinical management of syndromic craniosynostoses. *Childs Nerv Syst*. 2012b; 28:1447–1463.
- [54] Barbosa M, Almeida Md R, Reis-Lima M, Pinto-Basto J, dos Santos HG. Muenke syndrome with osteochondroma. *Am J Med Genet Part A*. 2009; 149A:260–261.
- [55] Yu JE, Park DH, Yoon SH. A Korean family with the Muenke syndrome. *J Korean Med Sci*. 2010; 25:1086–1089.
- [56] van der Meulen J, van den Ouweland A, Hoogeboom J. Trigonocephaly in Muenke syndrome. *Am J Med Genet A*. 2006; 140:2493–2494.
- [57] Anderson PJ, Molony D, Haan E, David DJ. Familial Parry-Romberg disease. *Int J Ped Otorhinol*. 2005; 69:705–708.
- [58] Mazzeo N, Fisher JG, Mayer MH, Mathieu GP. Progressive hemifacial atrophy (Parry-Romberg syndrome)—case report. *Oral Surg Oral Med Oral Pathol Endod*. 1995;79(1): 30–35.
- [59] Pensler JM, Murphy GF, Muliken JB. Clinical and ultrastructural studies of Romberg's hemifacial atrophy. *Plast Reconstr Surg*. 1990;85:669–676.
- [60] Parry-Romberg syndrome. (2009). <http://www.whonamedit.com/doctor.cfm>.
- [61] El-Kehdy J, Abbas O, Rubeiz N. A review of Parry-Romberg syndrome. *J Am Acad Dermatol*. 2012; 67(4):769–784.
- [62] di Meo N, Stinco G, Nan K, Pinzani C, Trevisan G. Parry-Romberg syndrome: a case with a possible association with Lyme disease. *Acta Dermatovenereol Alp Pannonica Adriat*. 2015; 24(4):77–79.
- [63] Sahin MT, Bari S, Karaman A. Parry-Romberg syndrome: a possible association with borreliosis. *J Eur Acad Dermatol Venereol*. 2004; 18(2):204–207.
- [64] Park MS, Yoo JE, Chung J, Yoon SH. A case of Pfeiffer syndrome. *J Korean Med Sci*. 2006;21(2):374–378.

- [65] Ettinger N, Williams M, Phillips JA. Variable expressivity and clinical heterogeneity can complicate the diagnosis and management of Pfeiffer syndrome [Case Reports, Journal Article, Research Support, Non-U.S. Gov't]. *J Craniofac Surg* 2013; 24(5):1829–1832.
- [66] Badoe EV. Pfeiffer syndrome type 3, a rare and unusual craniosynostosis: case reports from Korle Bu Teaching Hospital, Accra, Ghana. *West Afr J Med* 2013; 32(3):228–230.
- [67] Annick V, FRYNS JP. Pfeiffer syndrome. *Orphanet J Rare Dis*. 2006; 1:19.
- [68] Bueno MP, Sertie AE, Jehee FS, Fanganiello R, Yeh E. Genetics of craniosynostosis: genes, syndromes, mutations and genotype-phenotype correlations. *Front Oral Biol*. 2008; 12:107–143.
- [69] Naveh Y, Friedman A. Pfeiffer syndrome: report of a family and review of the literature. *J Med Genet*. 1976;4: 277–280.
- [70] Stoler JM, Rosen H, Desai U, Mulliken JB, Meara JG. Cleft palate in Pfeiffer syndrome. *J Craniofac Surg*. 2009;5: 1375–1377.
- [71] Fearon JA, Rhodes J. Pfeiffer syndrome: a treatment evaluation. *Plast Reconstr Surg* 2009; 5:1560–1569.
- [72] Peña WA, Slavotinek A, Oberoi S. Saethre-Chotzen syndrome: a case report. *Cleft Palate Craniofac J*. 2010;47(3):318–321.
- [73] Tahiri Y, Bastidas N, McDonald-McGinn DM, Birgfeld C, Zackai EH, Taylor J, Bartlett SP. New pattern of sutural synostosis associated With TWIST gene mutation and Saethre-Chotzen syndrome: peace sign synostosis. *J Craniofac Surg*. 2015; 26(5):1564–1567.
- [74] Di Rocco F, Benoit A, Vigneron J, Segura PB, Klein O, Collet C, Arnaud E. Y-craniosynostosis by premature fusion of the metopic and coronal sutures: a new nosological entity or a variety of Saethre-Chotzen syndrome? *Birth Defects Res A Clin Mol Teratol*. 2015; 103(4):306–310.
- [75] Sharma A, Patel N, Arora S, Ramachandran R. Child with Saethre-Chotzen syndrome: anesthetic management and literature review. *Acta Anaesthesiol Belg*. 2014;65(4):179–182 (Review).
- [76] Thakur AR, Naikmasur VG. A case of Robinow–Sorauf syndrome (Craniosynostosis-Bifid Hallux syndrome): the allelic variant of the Saethre-Chotzen syndrome. *Indian J Dent*. 2014; 5(2):96–99.
- [77] Massalska D, Bijok J, Kucińska-Chahwan A, Jamsheer A, Bogdanowicz J, Jakiel G, Roszkowski T. Prenatal diagnosis of craniosynostosis (compound Saethre-Chotzen syndrome phenotype) caused by a de novo complex chromosomal rearrangement (1; 4; 7) with a microdeletion of 7p21.3–7p15.3, including TWIST1 gene--a case report. *Ginekol Pol*. 2014;85(7):541–544.

- [78] Chang CC, Steinbacher DM. Treachercollins syndrome. *Semin Plast Surg.* 2012; 26(2): 83–90.
- [79] Maeda T, Oyama A, Funayama E, Yamamoto Y. Reconstruction of low hairline microtia of Treacher Collins syndrome with a hinged mastoid fascial flap. *Int J Oral Maxillofac Surg.* 2016 Jun; 45(6):731–4.
- [80] Plomp RG, van Lieshout MJ, Joosten KF, Wolvius EB, van der Schroeff MP, Versnel SL, Poublon RM, Mathijssen IM. Treacher Collins syndrome: a systematic review of evidence-based treatment and recommendations. *Plast Reconstr Surg.* 2016;137(1):191–204.
- [81] Argenta LC, Iacobucci JJ. Treacher Collins syndrome: present concepts of the disorder and their surgical correction. *World J Surg.* 1989; 13:401–9.
- [82] Nardini G, Staffenberg D, Seo L, Shetye P, McCarthy JG, Flores RL. Treacher Collins syndrome and tracheostomy: decannulation utilizing mandibular distraction osteogenesis. *Plast Reconstr Surg.* 2015;136(4 Suppl):47.
- [83] Ligh CA, Swanson J, Yu J, Samra F, Bartlett SP, Taylor JA. A morphological classification scheme for the mandibular hypoplasia in Treacher Collins syndrome. *Plast Reconstr Surg.* 2015; 136(4 Suppl):46.
- [84] Joseph G. McCarthy. Plastic surgery 8–volume set hardcover, 3rd edn, vol 4. W.B. Saunders Company. 1990.
- [85] Bachmayer DI, Ross RB, Munro IR. Maxillary growth following Le Fort III advancement surgery in Crouzon, Apert, and Pfeiffer syndromes. *Am J Orthod Dentofac Orthop.* 1986; 90:420–30.
- [86] Cohen SR, Boydston W, Burstein FD. Monobloc distraction osteogenesis during infancy: Report of a case and presentation of a new device. *Plast Reconstr Surg.* 1998;101:1919.
- [87] Fearon JA. The Le Fort III osteotomy: To distract or not to distract. *Plast Reconstr Surg.* 2001;107:1091–106.
- [88] Fearon JA, Whitaker LA. Complications with facial advancement: A comparison between the Le Fort III and monobloc advancements. *Plast Reconstr Surg.* 1993;91:990.
- [89] Gosain AK, Santoro TD, Havlik RJ, Cohen SR, Holmes RE. Midface distraction following Le Fort III and monobloc osteotomies: Problems and solutions. *Plast Reconstr Surg.* 2002;109:1797–808.
- [90] McCarthy JG, LaTrenta GS, Breitbart AS, Grayson BH, Bookstein FL. The Le Fort III advancement osteotomy in the child under 7 years of age. *Plast Reconstr Surg.* 1990;86:633.
- [91] Molina F, Ortiz-Monasterio F. Mandibular elongation and remodeling by distraction: A farewell to major osteotomies. *Plast Reconstr Surg.* 1995;96:825.

- [92] Ortiz-Monasterio F, Fuente del Campo A, Carillo A. Advancements of the orbits and the mid-face in one piece, combined with frontal repositioning for the correction of Crouzon's deformities. *Plast Reconstr Surg.* 1968;61:507.
- [93] Polley JW, Figueroa AA. Management of severe maxillary deficiency in childhood and adolescence through distraction osteogenesis with an external adjustable, rigid distraction device. *J Craniofac Surg.* 1997;8:181–7.
- [94] Whitaker LA, Bartlett SP, Shut L, Bruce D. Craniosynostosis: An analysis of the timing, treatment, and complications in 164 consecutive patients. *Plast Reconstr Surg.* 1985;80:195–212.
- [95] Whitaker LA, Munro IR, Salyer KE, Jackson IT, Ortiz-Monasterio F, Marchac D. Combined report of problems and complications in 793 craniofacial operations. *Plast Reconstr Surg.* 1979;64:198–203.