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Basics of Craniofacial Surgery

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1. Introduction

1.1. History

Craniofacial surgery includes a wide range of procedures in the face and cranium from congenital malformations such as orofacial clefts to traumatic deformities. Many scientists contributed to develop this field of surgery. Rene Le Fort was a French surgeon who discovered the lines of weakness in the facial skeleton. He classified the pattern of fractures through these lines into three categories known as LeFort 1, 2 and 3.[1] Gillies was the first surgeon who performed Le Fort 3 osteotomy for a patient with Crouzon or Pfeiffer syndrome, but the result was not satisfactory.[2] Tessier accomplished LeFort 3 osteotomy in a different and of course a more accurate way for a child with Crouzon's syndrome. [3] The presentation of this surgery had a great influence on this remarkable field and become a turning point in craniofacial surgery. Tessier introduced different approaches to the craniofacial skeleton such as transcranial approach to the orbital hypertelorism, transconjunctival approach to the orbital floor or development of subperiosteal facelift technique.[1]-[3] He is named the father of craniofacial surgery. During the years much progress has been made in this field. Refined surgical techniques and instruments, new imaging techniques like 3D computed tomography(CT) scan has had a great impact on craniofacial surgery, not only in diagnosis of craniofacial anomalies but also in treatment planning of surgery. Stereolithographic models are 3D printing models which can replicate the actual shape of the defect. These models facilitate reconstruction of prosthesis; they can help in determining the site of insertion of prosthesis or the correct position of the plates or devices like distraction osteogenesis.

2. Craniofacial anomalies

Craniofacial anomalies (CFA) include different dysmorphogenic conditions in this region of the body. People with these anomalies generally have problems not only in function but also with their ordinary life in the society. These people, especially children with CFA have less social competence and lack of self-esteem. The aim of craniofacial surgery is to give a normal appearance to the patients. Specialized centers would be a great help in treating patients with CFA. Accurate data of epidemiology of CFA is of utmost importance for managing and treatment of patients.

Cleft lip and palate are the most common congenital anomalies which affect the orofacial region. The incidence of oral clefts in United States is 1 in 700 births.[4] Orofacial clefts are more common in boys but cleft palate without cleft lip have a slight tendency to involve girls. One fourth of oral clefts are bilateral and the rest are unilateral. In unilateral cases the left side is affected more frequently.

Craniosynostosis means premature fusion of cranial vault sutures. There are six major cranial vault sutures. Any of these sutures can be affected in craniosynostosis, alone or in combination with other sutures. In this section we discuss the prevalence of nonsyndromic single suture synostosis.

Sagittal suture synostosis or scaphocephaly is the most common single suture synostosis with the prevalence of 1 in 5000 live births. [5] Boys are affected three times more frequently than girls.

Coronal suture synostosis or anterior plagiocephaly is the second most common single suture synostosis. The prevalence is approximately 1 in 10000 births. [6]

Metopic suture craniosynostosis or trigoncephaly is an unusual type of synostosis with an approximate prevalence of 1 in 15000. [7]

Lambdoid suture synostosis or posterior plagiocephaly is a rare entity with a prevalence of 1 in 150000 live births. [8]

Bilateral coronal suture craniosynostosis or brachycephaly is also rare.

3. Craniofacial pathology

Pathology includes any deviations from normal function and structure. The pathologic conditions show themselves as aplastic, hypoplastic, hyperplastic, neoplastic, traumatic or developmental entities. In craniofacial pathology a good access to the lesion and preservation of vital structures are important factors in a successful operation. Most operations in this field should be carried out in a team work manner engaging both the maxillofacial surgeon and the neurosurgeon.

3.1. Fibrous dysplasia

Fibrous dysplasia is a benign fibro-osseous lesion. Fibro-osseous lesions are a category of entities in which normal bone tissue is replaced with fibrous and mineralized tissue. In fibrous dysplasia normal bone is substituted with cellular fibrous tissue and immature bone.[9]

In most of cases it is monostotic involving a single bone.[10] The rate of growth is very slow. The maxilla and frontal bone are the most affected sites.[11] The most common feature is painless swelling. Radiographic feature of ground glass is of significant importance for diagnosis. This pattern is due to superimposition of disorganized poorly calcified bone trabecular.

Polyostotic Fibrous dysplasia is uncommon. In syndromic conditions like Jaffe-Lichtenstein, McCune-Albright and Mazabraud syndromes, polyostotic fibrous dysplasia forms an important part of these syndromes.

Treatment of fibrous dysplasia depends on the degree of functional or cosmetic impairment from shaving of involved area to resection. The aim is not to remove the entire lesion but to have an acceptable appearance. Regrowth after surgical reduction is unpredictable.

3.2. Sarcoma

One of the most important issues in morbidity and mortality of children is malignant neoplasms.[12] One third of malignant solid tissue tumors during infancy and childhood are caused by sarcomas.[13] The most common sarcoma in children is rhabdomyosarcoma and after that fibrosarcoma.[14] For management of rhabdomyosarcoma, radiotherapy and chemotherapy in combination with surgery are recommended in accessible tumors by many authors.

3.3. Lymphoma

Malignant lymphoid tissue tumors are common in head and neck region. There are different types of classification for lymphoma but separation to Hodgkin and non- Hodgkin types is the most common. Both have manifestations in the head and neck area. In Africa, another type of this condition known as Burkitt's lymphoma is common in children. Surgery is usually not indicated and chemotherapy should be done by an oncologist.

3.4. Melanoma

Approximately 20% of melanomas occur in head and neck regions.[15] An important etiologic factor is excessive exposure to ultraviolet light but many risk factors have been proposed in development of this lethal entity. Most of them arise from pigmented lesions. Melanoma can be categorized to superficial spreading, nodular, lentigo maligna, acral lentiginous and desmoplastic type according to clinical and histological evaluations. Excision of the lesion is the treatment of choice. Elective lymph node dissection is a controversial matter.

4. Nonsyndromic craniosynostosis

The pathophysiology of malformed skulls was described in the 18th century. It was reported that “bony expansion ceases in a direction perpendicular to the synostosed suture with compensatory expansion in the opposite direction.” This is called the Virchow’s law. Early closure of cranial sutures is termed as craniosynostosis which can be categorized to syndromic and non-syndromic synostosis. The latter is the main point of concern of this chapter.

4.1. Anterior plagiocephaly

Plagiocephaly is simply the Greek synonym for slanted head. It is reported in 5% to 25 % of deliveries. It can be associated with external forces or synostosis. Those associated with external forces are deformational and are commonly the result of compressive forces applied by the maternal pelvis to the head of the fetus which is more common on the left side. Anterior plagiocephaly can be due to unilateral coronal synostosis (UCS). This is almost nonsyndromic but can be seen in a familial feature relating to a mutation in fibroblast growth factor receptor gene. A surgical approach to treat this condition is basically to decompress the intracranial pressure which can itself cause brain damage and ophthalmic consequences due to optic atrophy. The superior forehead and superior orbital region is the main affected site. The ipsilateral superior orbital rim is displaced superiorly and nasal root is displaced to the problematic side. Presurgical radiological evaluation of the patient is best carried out by 3D CT scans which are available now and a multidisciplinary approach involving pediatric craniofacial surgeon, pediatric neurosurgeon, ophthalmologist and radiologist is necessary.

The surgical technique presented by Posnick is based on removing, reshaping and reassembling the cranial vault and bilateral three-quarter orbital osteotomies. The procedure is started by a postauricular coronal incision with wide subperiosteal dissection anteriorly to bilateral infraorbital rim regions and superior of the zygomatic bone and maxilla and posteriorly half way from the coronal to lambdoid suture. Bilateral lateral canthotomy is carried out. Frontal bone is removed from the preferred marked lines then the osteotomy is carried out including orbital roof, superior aspect of orbital medial wall, lateral orbital wall and lateral side of inferior orbital floor to the inferior orbital fissure.

4.2. Trigonocephaly

The prevalence of this type of craniosynostosis is 1 in every 15000 newborns. This results from early closure of the metopic suture which makes a triangular shape deformity of anterior cranial vault and anterior cranial base and orbits presents with orbital dystopia. Correction of the position of the superior and lateral orbital rims is the main concern in trigonocephalic patients. Beside the esthetic considerations, preventing the increase in intracranial pressure in growth is an important indication for surgical intervention.

If there is no sign of deficits due to increased intracranial pressure then it is recommended to postpone the surgical intervention 9 to 11 months of infant age. This supports the fact that at

that age most of the brain growth is done and the outcome is less dependent on brain growth. Also better bony reshaping can be performed and hemodynamic concerns at the time of surgery are more controllable.

It is obvious that every surgical treatment plan should be programmed based on the individual characteristics of the deformity but a general surgical approach involves releasing of the metopic suture and also osteotomies of the anterior cranial vault, temporal and three-quarter orbital osteotomies. A postauricular coronal incision with subperiosteal dissection is carried out. Bilateral lateral canthotomy is performed. Frontal craniotomies are performed by the neurosurgeon considering the retraction of frontal and temporal lobes and remaining anterior to the olfactory bulbs. Orbital osteotomies are done including orbital roof and lateral aspect of orbital floor to the inferior orbital fissure. For better correction of the orbital segment a vertical split osteotomy can be done in the midline to separate sides from each other. Temporal osteotomy can then be performed and reshaping, repositioning and reassembling of frontal bone in strip figures are done.

Bony gaps can be filled with autogenous grafts and segments are fixed by means of screws and microplates.

4.3. Scaphocephaly

Early closure of the sagittal suture which makes the calvarium grow more in the anteroposterior direction is called scaphocephalia. This leads to a more prominent frontal and occipital region. This condition is seen in 1 in every 5000 births. Like other synostotic conditions increased intracranial pressure and hydrocephalus are points of concern. It is accepted that craniotomies can at least release the external pressure on the brain. Papilledema and optic atrophy are other possible situations related to synostotic conditions that can lead to blindness.

There can be 3 types of scaphocephalia depending on the location of synostosis along the sagittal suture. If the anterior portion is involved in synostosis then posterior projection and prominence of the skull occurs. If posterior parts of the suture gets involve then anterior skull projection is seen and there is a condition when the whole suture is synostotic that leads to a more significant growth of the skull in the anteroposterior direction. Again it is recommended that surgical intervention occurs at 9 to 11 months of age supporting the concept that it is the age that most of brain tissue growth has occurred and bony tissue can be reshaped easier and estimated blood loss during the operation is a lesser challenge.

The general concept in the surgical approach which is done again by a post auricular coronal approach is to remove the sagittal suture and portions of temporal bone then reshaping the segments in a manner to decrease the anteroposterior projection while increasing the biparietal length. Dissections are carried out anteriorly to the periorbital regions and posteriorly to the occipital prominence of the skull. Craniotomies are done and brain tissue is retracted gently to allow the surgeon to have access for orbital osteotomies. Strip portions of bone are reshaped in an individualized manner to correct the deformity.

4.4. Brachycephaly

Brachycephaly is the Greek synonym for short headedness, occurs in bilateral coronal synostosis. This condition can be seen in some syndromes like Crouzon, Apert and Pfeiffer. In syndromic conditions other synostoses can be seen like in cranial base and upper face that leads to concave facial profile, proptosis and midface deficiency. In non-syndromic conditions no midface deficiency is present. Forehead height is apparently increased and the orbits are retruded and widened. The general concept in surgical approach is to increase the cranial capacity and therefore decrease the intracranial pressure preventing brain damage and conditions like hydrocephalus. Beside the esthetic concerns these conditions if left untreated may lead to ophthalmic deterioration and even blindness.

Again like other synostotic conditions the forehead and upper parts of the orbit are the center of concern for reconstruction. In these patients the forehead has a retruded position. In a normal condition the eyebrows are slightly more projected anteriorly comparing to the globes in contrast to brachiocephalic patients whose forehead and upper orbital rims have a retruded position. Here an anterior coronal flap is preferred because the malformed bony parts are located in the anterior skull region. Subperiosteal dissection anteriorly to periorbital region and posteriorly halfway between coronal and lambdoid sutures is carried out. As was said before the aim is to remove the synostotic and bad shaped bony parts and reshaping and reassembling them in a new position to correct the deformity. Osteotomies of the frontal and temporal bone are done as well as osteotomies of the orbital roofs. A vertical osteotomy should be carried out in the midline to help reduce the width of the superior orbital parts. In all the dissections in any form of cranial synostosis where lateral canthotomy is performed, at the end of operation lateral canthopexies are done using wire sutures.

5. Distraction osteogenesis

This method increases the length of bone by means of gradual distraction. Distraction techniques provide circumstances to achieve large advancements in craniofacial anomalies. Not only lengthening of the skeleton but also distracting the overlying soft tissue occurs with this technique. Because of this fact, some prefer to use the term "distraction histogenesis" than distraction osteogenesis. Ilizarov a Russian orthopedic surgeon published the first case series of DO for limb lengthening.[16] The first reports of mandibular lengthening by DO were reported by Molina and McCarthy.[17],[18] Since that time DO has been used for patients with various craniofacial deformities. Advantages of DO include achieving large advancements, obviating bone grafting and lesser risk of relapse than conventional osteotomies. We may however, encounter some problems. Higher rates of postoperation infection than conventional osteotomy, difficulty in control of vector direction, nonunion or malunion of the surgical site are disadvantages of this method.

The procedure of DO entails a corticotomy at the site where the device would be placed. During the procedure special consideration should be given to protection of periosteum. Minimal disturbance of the periosteum is an important matter in success of DO.[4] The aim of this step

is to provide an environment for remodeling and growth of the bone without significant tissue damage or vascular supply insufficiency. Corticotomy and device placement cause tissue damage like other surgical procedures. A latency period is needed for inflammatory mediators and subsequent inflammation to subside. This period ranges from 0 to 10 days depending on the age of the patient and conditions of the surgical site. After this period, distraction phase begins. Distraction rate is approximately 1mm per day. Soft tissue matrix covering the bone is distracted with this rate of traction. The term distraction histogenesis can describe this phenomenon better than distraction osteogenesis. Distraction at a higher rate may result in malunion or nonunion of involved bone and distraction at a lower rate will cause bone healing without any changes in length of the bone. After distraction, consolidation begins. Time lapse for this phase ranges from 3 months to 6 months depending on the amount of advancement, age of the patient and the type of deformity. Consolidation phase can be confirmed with radiography.[19]

5.1. Mandibular distraction osteogenesis

In severe mandibular retrognathia DO is indicated. This condition can be acquired or congenital. Congenital mandibular retrognathia includes craniofacial anomalies such as Treacher-Collin's syndrome, Pierre Robin sequence and hemifacial microsomia. Acquired anomalies include traumatic events to the mandible such as condylar fractures in young children in which ankylosis of the temporomandibular joint and subsequently, retardation of mandibular growth occur.[19], [20]

5.2. Maxillary distraction osteogenesis

Midface deficiency can occur in a variety of syndromic and nonsyndromic patients. In cleft patients, post-surgical scar tissue causes retardation in maxillary growth. Because of scar tissue formation, Le Fort advancement with conventional osteotomy and rigid fixation are not appropriate for these patients. Difficulty in mobilization of the maxilla and a tendency to relapse are the major concerns in these cases with conventional osteotomy.[21], [22] Craniofacial syndromes like Apert, Crouzon or Pfeiffer's syndromes show different degrees of paranasal hypoplasia. Midface advancement at the Le Fort 1 or 3 levels with DO technique is a good method for large advancements. Lower rate of relapse is another advantage of this technique.

5.3. Cranial vault distraction osteogenesis

Conventional treatment for patients with craniosynostosis is an aggressive dismantling of the cranial vault. A high rate of morbidity and even mortality in some cases is of major concern in these patients. Recently, corticotomy of the cranial vault and slow distraction with DO has shown new horizons in correction of calvarial deformities. Although some advantages like decreasing morbidity or reducing scar formation are mentioned for distraction techniques, its disadvantages cannot be ignored.[23], [24] A 3D deformity cannot be corrected with a unidirectional force. However, accurate selection of position of the appliance and prebending it as much as possible may be helpful.

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