Life is what happens when you're busy making other plans.

- John Lennon

The body's most intricate structure is found in the cranium. Along with connective tissue, muscle, vasculature, and accompanying innervation, the skull, which includes bones that surround and protect the brain and sensory organs, serves as a structure for the face to support the activities of eating and breathing. Endoderm, mesoderm, ectoderm, and cranial neural crest cells (CNCCs) and their offspring together give rise to these tissues.[1] Signalling between these cellular components and to the craniofacial mesenchyme (formed primarily by CNCCs with a mesodermal contribution) provides positional cues and regulates growth and differentiation.[1] Fig 8.1

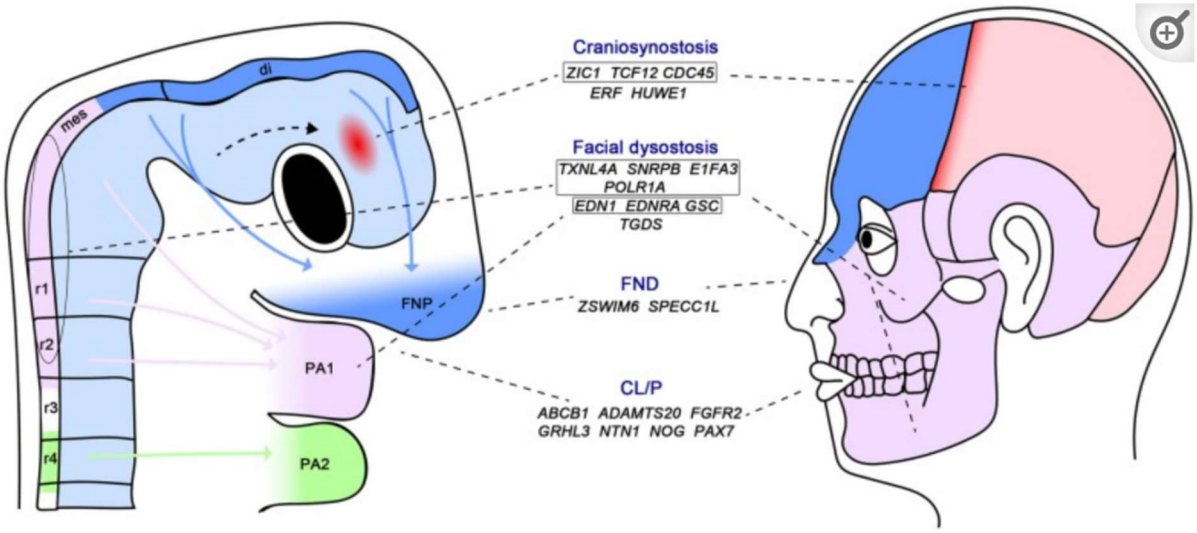

Fig 8. 1 Growth Of The Head And Face, As Well As Freshly Discovered Craniofacial Genes

TREACHER COLLINS SYNDROME (TCS)

Mandibulofacial dysostosis, or Treacher Collins syndrome (TCS), is an autosomal dominant disorder with varying degrees of expressivity. In individuals with fully developed TCS, the facial profile may display a retrusive lower jaw and chin, a prominent nasal dorsum, and a horizontally deficient convex appearance. The palpebral fissure in the area around the eyes has an antimongoloid slope because of colobomata, hypoplasia of the lower lids and the lateral canthi, partial lack of the eyelid cilia, and dystopia and hypoplasia of the inferolateral orbit. Hair growth with tongue-like structures is common in the preauricular area. [2] Hearing is compromised due to varying degrees of external auditory canal hypoplasia and middle ear

ossicles that are hypoplastic. The external ears are missing, deformed, or malposed. Hypoplasia of the malar bones, frequently accompanied with clefting across the arches and restricted creation of the remaining zygomas, including the glenoid fossa component, is a distinctive feature. [3] The temporomandibular joints (TMJs), the muscles of mastication, and the muscles of facial expression are all impacted differently by the hypoplastic characteristics of the maxilla and mandible bones. It's interesting to note that TCS does not reveal hypoplasia of the soft tissues in the middle of the face.[4] In general, the maxillomandibular complex is steeply rotated clockwise and there is an Angle Class II anterior open-bite malocclusion. The lower face height in the front is longer than the lower facial height in the back. The jaws' clockwise rotation is consistent with the A-to-B-point connection in profile. The occurrence of cleft palate (with or without cleft lip and choanal atresia of the nasal cavity) is unpredictable.

The categorization of Mandibular Malformation and Temporomandibular Joint falls into several classifications [5,6]

Type I TMJ–mandibular malformation involves. minimal hypoplasia of the glenoid fossa and the condyle– ascending ramus on each side. All of the skeletal components are present.

Type IIA TMJ little degree of glenoid fossa and condyle-ascending ramus hypoplasia are present in TMJ-mandible malformation.

Type IIB Condyles suffer from significant hypoplasia in mandibular deformity.

Type III A mandibular defect known as glenoid fossa-mandibular malformation prevents the lower jaw from stopping against the cranial base in the posterior position.

Staging of Skeletal Reconstruction: Timing and Techniques

- Zygomatic and Orbital Reconstruction
- Maxillomandibular Reconstruction
- Nasal Reconstruction
- Facial Soft-Tissue Reconstruction
- External Ear Reconstruction

To address the unique and specific components of the Treacher Collins malformation, the reconstruction and rehabilitation process must include the following aspects: 1) the zygomatic

and orbital region; 2) the maxillomandibular region; 3) the nasal region; 4) the soft-tissue envelope; 5) the external ears; 6) the external auditory canals; and 7) the middle-ear structures.

The orbit and zygoma Reconstruction

Studies have suggested that sculpted full-thickness calvarial bone grafts used in a three-dimensional approach can be a viable strategy for repairing malar defects and orbit defects in people with varying degrees of Treacher Collins syndrome. The neo-zygoma is subsequently fixed with a microplate and screws to support it. Without the necessity for further periorbital incisions, a coronal scalp incision provides exposure for the reconstruction. (7) Fixed split-thickness autogenous cranial bone can be utilized to reconstruct the deficient areas of the orbital floor. After removing cranial vault as a donor, the affected sites can be reconstructed through the use of either autogenous split cranial grafts or synthetic materials. When non-autogenous restoration is required for small cranial donor sites, a titanium mesh base attached to the adjacent skull is a preferred option. A replacement for bone is then used to fill in the mesh. To attach to the new lateral orbital rim, lateral canthopexies are completed via the coronal scalp incision and secured in place. CT scanning pre and post-surgery in our investigation showed that individuals with TCS had significantly longer lateral orbital walls (depth), wider lateral orbital rims, wider interzygomatic arches, and longer zygomatic arches. The changes made to address the zygomatic and orbital deformities in TCS have been shown to persist in postoperative scans taken after a year. Different surgeons have suggested various techniques and schedules for treating these anomalies. According to some experts, the use of full-thickness autogenous cranial bone to construct the entire zygomatic complex is deemed more efficient in preserving volume and contour, as opposed to onlay grafts, which have been linked to less favorable outcomes. Despite the use of onlay autogenous bone grafts from different donor sites (such as skull, hip, or rib) to reconstruct various regions in the craniofacial area (such as supraorbital ridge, zygoma, anterior maxilla, angle of mandible, or chin), they have displayed significant and unanticipated resorption over time. Also, it shouldn't be anticipated that the graft would develop or expand in volume at a rate that corresponds to that of the underlying bone, such as the zygoma.

The idea that "even if the graft partially resorbs, at least something has been achieved" should be disregarded since it frequently leads to surface abnormalities that worsen the underlying abnormality.

Maxillomandibular Reconstruction

Clarifying any unique airway requirements is a key factor in deciding when and how to do a jaw restoration. If first stage early lower jaw reconstruction is required before growth

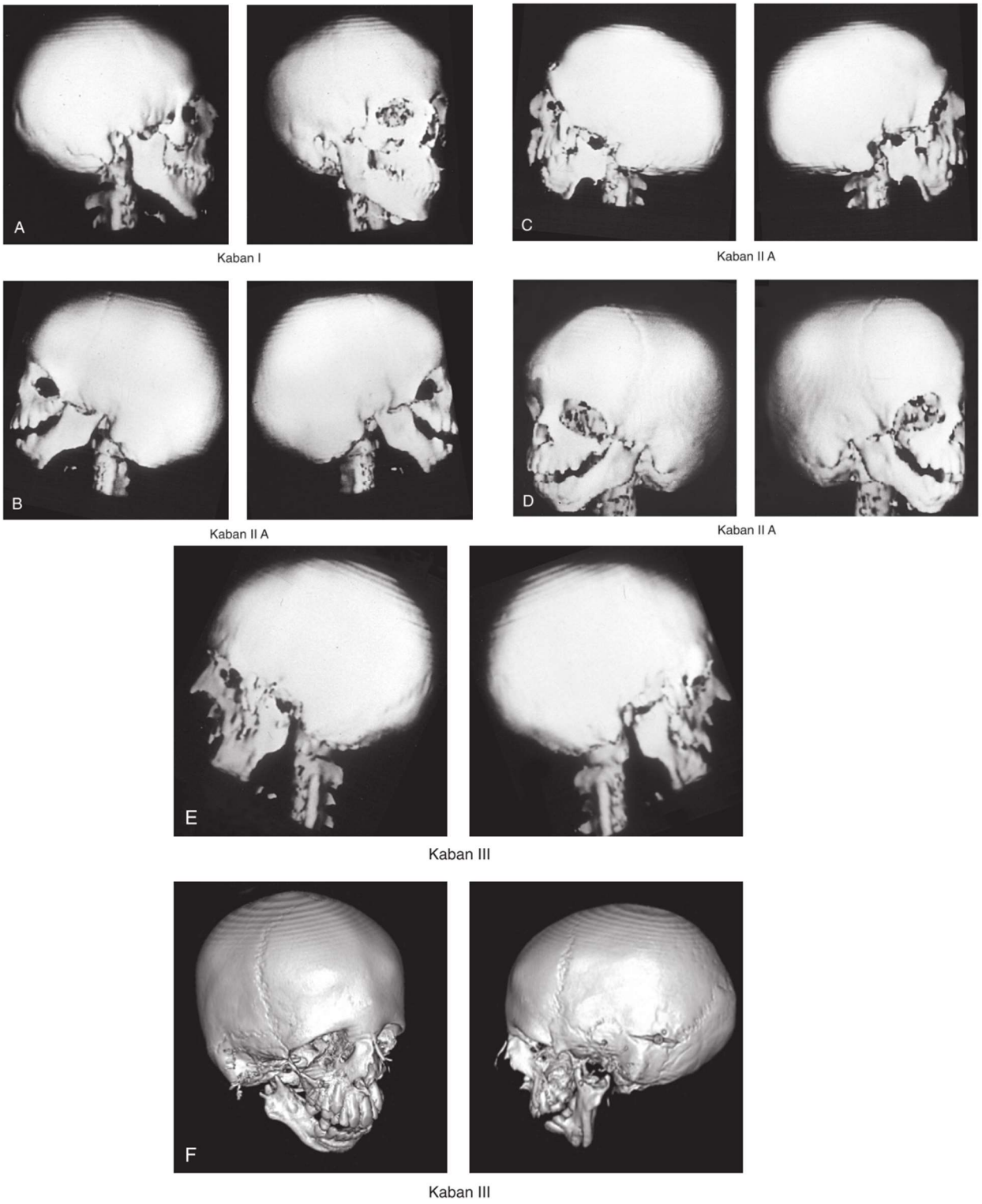

Fig 8.2: The Level Of Temporomandibular Joint-Mandibular Deformity Seen In Individuals With Hemifacial Microsomia, Kaban And Colleagues Developed A Categorization System.

complition, this will determine if it is essential. The amount of TMJ-mandibular deformity is the second factor to be taken into account. Abnormalities of Type I and IIA do not necessitate the construction of glenoid fossa or condyle. Patients with these diseases have sufficient TMJ architecture, allowing for mandibular reconstruction using ramus osteotomies while yet maintaining a functional TMJ. [8]

Individuals diagnosed with TCS may require reconstruction of the maxillomandibular complex due to three primary dysmorphologies: changes in facial proportions resulting in increased anterior lower face height and decreased posterior facial height; absence of a horizontal jaw,

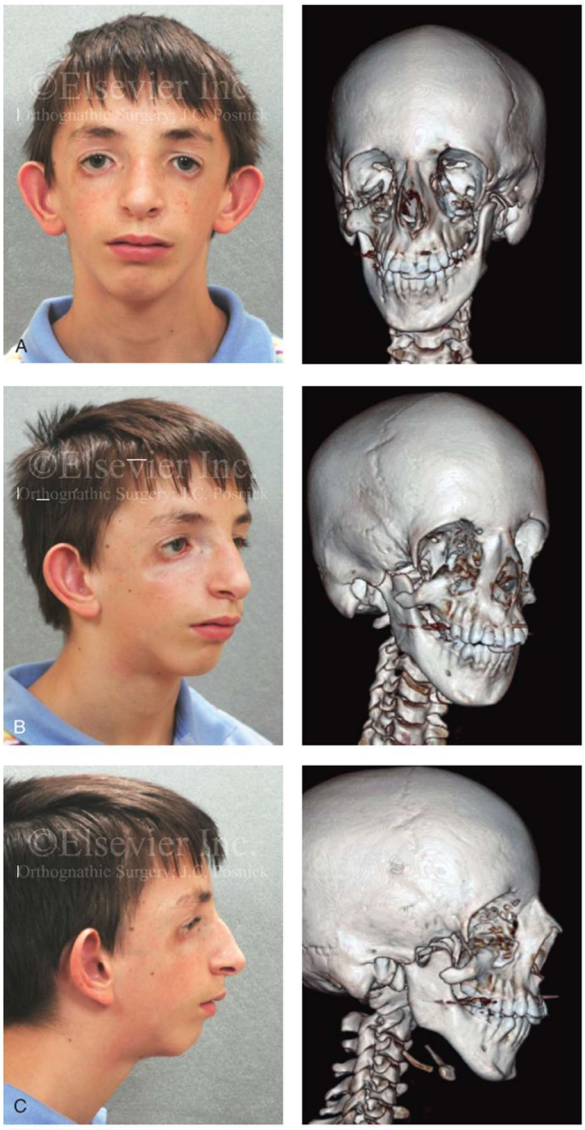

Fig8.3: Treacher Collins Syndrome Is Completely Exhibited In An 11-Year-Old Kid.

which is particularly prominent in the mandible but may also occur in the maxilla; and chin dysplasia, characterized by increased vertical length and sagittal retrusion.

When seen in profile, these abnormalities cause the maxillomandibular complex to rotate too much clockwise and to have an undesirable A-point-to-B-point connection. An Angle class II anterior open-bite malocclusion is the most common kind. It's interesting that some people will exhibit a Class III or Class I connection. For the airway room to be increased, the deformity of occlusion to be fixed, and the facial equilibrium to be restored, the maxillomandibular complex must be rotated anticlockwise.[9]

The most favorable cosmetic outcomes are typically observed in patients who receive definitive reconstructive jaw surgery after attaining early maxillofacial skeletal maturity (usually after the age of 13-15 years), and when this surgery is combined with effective orthodontic treatment during the permanent dentition. To correct the inclination of anterior teeth, decompress the occlusion, and alleviate dental root crowding caused by orthodontic treatment, mandibular and, in some cases, maxillary premolar extractions are often required to properly reposition the jaws for aesthetic and airway purposes.

The condyle that is congenitally absent must be surgically constructed in cases of type IIB mandibular abnormalities. Osteotomies by themselves—with the skeletal segments' immediate or progressive DO—will not be sufficient. Despite its inherent drawbacks, mandibular reconstruction with a costochondral graft is still favoured for treating the Type IIB abnormality. In order to attain facial equilibrium and a sufficient airway, the entire maxillomandibular complex must be rotated anticlockwise. To achieve reliable and effective repair, new condylar construction, followed by maxillary and mandibular osteotomies near or at skeletal maturity, represents the only dependable approach. The comprehensive approach to jaw reconstruction encompasses intranasal interventions aimed at improving airway patency, such as septoplasty, inferior turbinate reduction, and reshaping of the nasal floor and walls.

To address the Type III TMJ-mandibular deformity, the non-existent glenoid fossa and condyle present since birth must be reconstructed surgically. Incorporating the new glenoid fossa into the zygomatic and orbit reconstruction is desirable since it is an essential component.[10]

Indications for First-Stage Mandibular Reconstruction in the Newborn

Treacher Collins syndrome (TCS) is a congenital disorder that affects the first and second branchial arches on both sides of the face, resulting in a range of upper airway anomalies that can be recurrent and variable. One of the syndrome's consistent yet variable elements is hypoplasia of the jaw. Studies have revealed that sleep apnea affects over 25% of individuals with TCS and is a risk factor for the disorder. In cases where OSA patients are dependent on tracheostomy, sagittal advancement of the mandible may be used as an alternative therapy approach to create additional space in the oral cavity, push the tongue forward, and open the retromandibular airwayFor the past 20 years, DO methods have been used to improve the airway by carefully relocating and then maintaining the mandibular segments until they are consolidated. Many writers described the use of this procedure as an efficient way to decannulate people who would otherwise be tracheostomy-dependent using TCS. Over the last two decades, DO techniques have been utilized to enhance the airway by gently repositioning and retaining the mandibular segments until consolidation takes place.

Anderson and colleagues reported long-term outcomes of lower jaw lengthening with a DO method that was performed during the youth of the patient to treat airway obstruction in individuals with TCS. During the distraction period, the patient's minimum cross-sectional airway increased in size, but it remained below the normal levels for their age, according to their findings. Number of scans performed afterward for a period of a decade of progressive growth showed minimum size of the airway did not increase. Furthermore, they illustrated through the analysis of three-dimensional CT images that the mandibular development followed the dysmorphic TCS pattern similar to the pre-distraction intervention.

As a result, the mandibular plane angle was excessively steep and turned clockwise. The authors come to the conclusion that opening the retromandibular airway in a kid with TCS who is having OSA may be helped by a mandibular DO surgery. Yet, throughout chronologic development, the upper airway's early improvement was not sustained. Despite more than 20 years of international experience, further studies on long-term outcomes are required to determine the role of mandibular distraction osteogenesis in the treatment protocol for infants and children with treacher collins syndrome. In the absence of trustworthy data, it is recommended to exercise caution and discretion while using mandibular osteotomies and lengthening techniques through any method during infancy and adolescence. [11]

Indications for First-Stage Mandibular Reconstruction during the Mixed Dentition.

If a Type IIB condylar deformity requires a first-stage mandibular reconstruction, it is advisable to wait until the child is between 7 to 10 years old and undergo the procedure only after the permanent first molars in the mandible have emerged. Sadly, the mandibular and other maxillofacial abnormalities cannot be permanently corrected with a first-stage early lower jaw reconstruction. Reconstructive surgeons are still plagued by issues like infection and TMJ ankylosis, among others, costochondral graft overgrowth, undergrowth, and asymmetrical growth. During early skeletal maturity, the maxillary jaw, mandibular jaw, and chin area will all need their final orthognathic correction. The argument holds true for patients with typical dentofacial deformities, as well as those with associated syndromes or cleft lip and palate jaw deformity. It is suggested to delay the correction of maxillomandibular deformity until early skeletal maturity, which typically occurs between the ages of 13 to 15 years. It is not advisable to undergo mandibular surgery during mixed dentition due to various reasons. Firstly, it may cause damage to the developing permanent dentition and inferior alveolar nerves, leading to long-term harm. Secondly, delaying the surgery until a later age may prevent cutaneous scarring. Thirdly, postponing the surgery could limit perioperative airway and infection complications. Fourthly, delaying the surgery may also help in preventing negative psychosocial memories in the patient. Lastly, performing mandibular surgery during mixed dentition could lead to iatrogenic mandibular and TMJ deformation, making it challenging to achieve a successful long-term restoration.

Also, due to the location of the growing teeth and anticipated induced development limits, bone surgeries in the deformed maxillary jaw and chin are rarely performed in childhood. The literature has now established that Le Fort I osteotomy does not results continuing horizontal maxillary development. Moreover, it is not practical to surgically position the jaws in a way that would guarantee long-term functional occlusion and improved aesthetics during the mixed dentition. A Le Fort I osteotomy is necessary in TCS patients to treat the vertical, horizontal, and transverse maxillary abnormalities. When pitch direction needs to be corrected, the maxillomandibular complex invariably benefits from anticlockwise rotation. In most cases, anterior maxilla intrusion is required to create a more typical contact between the top lip and the teeth while smiling and when the mouth is relaxed. The posterior maxillary height may need to be lengthened, but more often than not, it can stay in a position that is quite near to

neutral. In order to achieve the appropriate anticlockwise rotation of the mandible and chin, a maxillary plane change is performed, which is also referred to as pitch orientation. The primary objectives of orthognathic surgery are to achieve the following three goals. Firstly, to restore the normal anterior facial height. Secondly, to improve the upper airway by increasing the space for better airflow. Lastly, to improve the cosmetic appearance by enhancing the connection between the A and B points when the patient is viewed from the side. By achieving these goals, orthognathic surgery can help patients achieve better oral function, improved breathing, and enhanced facial aesthetics. In 1993, Rosen published research on the long-term stability of the lower jaw's anticlockwise rotation when it is necessary to do so in order to improve face aesthetics and function in people with mandibular micrognathia. Similar clinical trials conducted by others have supported the mandibular anticlockwise rotation's long-term skeletal stability when used in an orthognathic repair. In most cases, bilateral intraoral sagittal split osteotomies are considered the preferred ramus procedures for Type I and IIA mandibular malformations. These procedures can effectively achieve horizontal advancement, anticlockwise rotation of the mandible, and sufficient bone contact across each osteotomy site without the need for lengthening the posterior facial height. Titanium plates and screws are used as necessary to stabilize each mandibular osteotomy site. When necessary, an interposed corticocancellous bone graft (such as an autogenous iliac graft) is employed. Splitting in the ramus area at the time of final reconstruction is desirable when possible for Type IIB and Type III rib or fibula graftconstructed mandibles that were developed earlier throughout the patient's life. With the advancement of the distal mandible, there is often just a little amount of bone contact across the osteotomy site. It is best to prepare for the potential necessity for an interpositioned corticocancellous bone transplant (such as an autogenous iliac). As compared to traditional osteotomies, DO osteogenesis approaches have often failed to show lower morbidity or better soft-tissue response in patients with TCS. Results show that when the treatment is done during childhood, there is no continuous jaw development and a considerable amount of relapse occurs during the consolidation phase of DO methods. The use of curvilinear DO for the repair of complicated mandibular abnormalities in severe types of TCS is still being improved by Kaban, colleagues, and others. Despite best efforts when applying DO procedures, achieving the desired proportions and symmetry of osteotomized skeletal segments through reliable three-dimensional vector control remains a work in progress. For these reasons, we choose the traditional orthognathic correction of the maxillomandibular lack of balance should

be considered when practical, while improvements in DO technology and its clinical applications are pending.

Nasal Reconstruction

Adults with treacher collins syndrome frequently have a mild to moderately wider nasal bridge, they almost always have a mid-dorsal hump. The nose's length is often normal, but it seems longer because the upper and bottom skeleton thirds of the face are out of harmony. Moreover, the tip frequently droops and lacks the desired projection. It is preferable to hold off on the rhinoplasty until after successful orthognathic surgeries have been completed in order to attain the optimum cosmetic effects. Exposure is given via an open approach (a columella-splitting incision).

Facial Soft-Tissue Reconstruction

Although surgeons have made well-intentioned attempts for decades to address soft-tissue deficits of the eyelid-adnexal areas in individuals with TCS, there are only a few recorded cases of cosmetically appealing soft-tissue eyelid reconstructive outcomes. From a technical perspective, transferring pedicled upper eyelid skin and muscle flaps to underdeveloped lower eyelid areas is not a complicated procedure. However, the resulting adnexal scarring often produces an artificial or "operated" appearance that cancels out the potential benefits.

Another commonly cited alternative is to apply full-thickness skin grafts to the lower eyelids. It leaves a predictable "patchy" appearance and should only be used on people with stubborn corneal exposure issues. To address the inferior displacement of the lateral canthi caused by both orbital dystopia and hypoplasia of eyelid structural components, correcting orbital dystopia and performing direct lateral canthopexies is often necessary. However, these procedures may not fully correct the multilayer soft-tissue hypoplasia of specialised adnexal structures such as skin, cartilage, eyelashes, ligament, fascia, and tendon. Hence, even relatively straightforward eyelid surgeries for individuals with TCS should always be discussed with a qualified oculoplastic surgeon.

To summarize, the temporal fossa's hollowing is a common feature seen in individuals with TCS, which can be attributed to the hypoplasia of various structures such as the epidermis, temporoparietal fascia, temporalis muscle, and reduced bitemporal breadth of the anterior cranial vault. The treatment for subcutaneous deficit in the temporal or adnexal region is limited

to using transposed peri cranial or temporoparietal flaps, due to the congenital hypoplasia of these areas.

When the treatment is carefully planned and executed, using microvascular reanastomosis to transfer soft-tissue flaps from other parts of the body is a viable option for repairing TCS. Since Dos Santos originally described parascapular free flaps, they have been the go-to procedure when this amount of face soft tissue restoration is necessary. According to Siebert and colleagues, the parascapular flap is effective in correcting contour defects in the lateral areas of the face by providing vascularized soft tissue in the subcutaneous plane and minimizing scarring. This improves cosmetic outcomes in certain patients with TCS. It is fortunate that patients with TCS do not have deficiencies in the centrally located soft tissues of the face, such as the forehead, nose, upper and lower lips, chin, and submental area. Techniques such as adipose tissue grafts, autologous fat injections, and other methods for augmenting soft tissue are now frequently employed. Soft-tissue augmentation techniques are becoming more common. The outcomes of autogenous fat injection are unpredictable and thought to be technique-dependent, yet they are typically positive and obviously promising. Tanna et al. investigated the effectiveness of serial autologous fat grafting for soft-tissue contour restoration in individuals with hemifacial microsomia. The study involved two groups of patients with moderate to severe hemifacial microsomia. Group I received microvascular free-flap repair using inframammary extended circumflex scapular flaps, while Group II underwent multiple stages of autogenous fat grafting. Both groups received microvascular free-flap reconstruction. The pre-reconstruction face symmetry ratings for the two patient groups were similar (74% and 75%, respectively) as were their OMENS values (2.4 and 2.3, respectively). The microvascular free-flap group had a face symmetry score of 121% at the final evaluation, while the fat-grafting group received a score of 99%. Likewise, neither the patient nor the doctor's assessment of overall satisfaction between the fatgraft and microvascular groups was significantly different. It is reasonable to extrapolate these study results to the person with Treacher Collins Syndrome.

External Ear Reconstruction [12]

One can complete the surgical reconstruction of the auricle using a stepwise approach in the hands of skilled professionals. The techniques Brent presented serve as the industry standard for external ear reconstruction. A key aspect of successful auricular reconstruction is the implantation of a well-crafted cartilage framework. According to Brent, most children have

sufficient rib cartilage for reconstruction by the age of 6. The substantial cartilage block needed to create the ear's structure will thereafter be provided by the synchondrotic area of ribs 6 and 7, which is taken from the side opposite the ear being built. A tiny preauricular incision is created after determining the location of the ear using the preoperatively obtained measurements and a premade template. Vestigial cartilage that is useless is removed. In order to recruit enough tension-free skin covering, the dissection is prolonged beyond the specified auricular outline, resulting in the development of a thin-skinned pocket. Brent prefers to harvest the cartilage grafts for each ear several months apart in patients who have bilateral microtia. The simultaneous repair of both ears would require bilateral chest incisions, significant splinting, and the possibility of respiratory difficulties. The lobule transposition, skin graft-assisted auricular detachment, hairline management, and tragus building are further phases of the auricular construction. To correct bilateral microtia, Brent recommends a series of five procedures spaced three months apart.

Future middle ear and external auditory canal surgery in the patient with TCS must be taken into account while finishing microtia repair. Any middle-ear surgery that involves the formation of the auricle should come first because, once an effort is made to open the canal, the skin envelope's scarring drastically reduces the likelihood of a successful auricular repair. Brent recommends sculpting the cartilaginous framework with a bigger concha to enable a future surgically constructed canal when middle-ear surgery is an option.

Alloplastic and homologous frameworks are further choices for external ear restoration. Silicone and Medpor are two currently utilised alloplastic materials. The implanted materials are more susceptible to infection, soft tissue wound dehiscence, and vulnerability to slight trauma even after many years of repair. One method to create an artificial ear involves generating bovine cartilage cells in a laboratory, placing them on a synthetic ear structure that is biodegradable, and then inserting the structure under the skin of a mouse with an intact immune system. The use of tissue engineering techniques has shown potential for the future, however, until the immunogenic issues are resolved, sculpted autogenous rib cartilage will remain the material of choice for surgical ear repair. Nevertheless, the likelihood of obtaining an aesthetically pleasing outcome is low unless the option of sculpting autogenous rib cartilage is carried out by a skilled auricular reconstructive surgeon. Given the unique and varying severity of TCS, it is important to note that not all patients require or may even benefit from reconstructive surgery. In some cases, the use of a prosthetic external ear may be a viable

option. The decision to pursue reconstruction or a prosthetic should be made on an individual basis, taking into consideration the patient's specific circumstances and desires.

External Auditory Canal And Middle-Ear Reconstruction.

The hearing loss in TCS patients is attributed to not only external auditory canal stenosis or atresia but also hypoplastic middle-ear cavities and ankylotic or absent ossicles, which account for an average of 44 dB hearing loss. Ossicles that are ankylosed or non-functional seem to have the same impact on hearing conduction as if they were absent. Gains in hearing can be anticipated in people with TCS who attempt middle ear reconstruction. The middle ear dysmorphology, which persists even after the ossicles are mobilised and cause a residual hearing loss, makes the patient dependent on some kind of amplification. Jahrsdoerfer reported on a cohort of patients with TCS who underwent surgery aimed at improving their hearing. He reaffirmed the efforts to "non-aided" long-term hearing that have been mostly ineffective. For adequate amplification in the majority of patients, including those who have had reconstruction as well as those who have not, a type of bone-conduction hearing aid is necessary. A bone-conduction hearing aid can come in different forms, including a traditional device with amplification adjustments, a modified amplification system, or a bone-anchored device. According to Brent, only selected patients with bilateral and unilateral microtia, high levels of personal motivation, the presence of favourable radiologic evidence of middle ear development, and an experienced otologist's expectation of a favourable outcome should undergo any middle ear surgery because the benefits must outweigh the risks and complications of the procedure. Moreover, ear-canal atresia, middle-ear deformities, and microtia correction require the expertise of an auricular/reconstructive surgeon and an otologist who must work together to plan the procedure. The auricular/reconstructive surgeon begins middle ear surgery by carefully conserving both the connective tissue on the underside of the framework and the general circulation of the flap before pulling the rebuilt ear from its bed. The otologist then performs ossiculoplasty, drills a bony external auditory canal, and repairs the tympanum with a temporal fascia graft. After removing the soft tissues to externalise the meatus through the chondral area, the auricular/reconstructive surgeon harvests a skin graft that the otologist uses to line the new external auditory canal.

The current method for treating deformities brought on by TCS involves staging the reconstruction to match face growth patterns, visceral function (such as breathing, eating, chewing, and swallowing), and psychosocial requirements. Each patient's precise morphologic

study and the realisation that a tiered reconstructive strategy is required assist to make the goals at each interval clear to the doctors and the family. The outlook for people with TCS will be much improved by continuing to establish a justification for the time, procedures, and degree of surgical, medical, and dental treatments and then objectively evaluating head and neck function, face aesthetics, and psychosocial results.

HEMIFACIAL MICROSOMIA

Dysmorphology Associated with Hemifacial Microsomia

Fig 8.4: 9-Year-Old Kid Who Was Born With Unilateral Cleft Lip And Palate And Left-Sided Hemifacial Microsomia.

Fig 8.5: A 14-Year-Old Female With External Auditory Canal Absence And Right-Sided Hemifacial Microsomia, Including Microtia.

Facial Soft Tissues

Clinically speaking, the external ear, eyelid-adnexal structures, preauricular-cheek-lip soft tissues, and temporal fossa are the four anatomical areas of the head and neck that are thought to be affected by the soft-tissue abnormalities linked to HFM. The majority of the cutaneous and subcutaneous tissue, the volume of fat, the muscles of mastication and facial expression, the cranial nerves, and the parotid and submandibular glands are among the soft tissues within

each area that may be lacking or dysmorphic. According to Kane and colleagues' research, the degree of hypoplasia in particular mastication muscles in individuals with HFM typically predicts the degree of dysplasia in the osseous origin and insertion of those muscles.[13]

The coronoid process will be lacking if the temporalis muscle is hypoplastic. The gonial area of the jaw will be lacking if the masseter muscle is hypoplastic. The condylar head is missing or nonexistent when the lateral pterygoid muscle is gone. Soft tissue thickness and skin surface area in the preauricular-cheek region are typically correlated with skeletal deficits. Little remnants of epithelial tissue known as skin tags are often located near the breach between the first and second arches. Little cartilaginous residues that are present in the subcutaneous tissue are usually linked to skin tags. If the sinus passages are blocked, inclusion cysts may develop or an infection may develop. [14]

Macrostomia: Clefting or failure of the first branchial arch and the maxillary and mandibular processes to fuse are referred to as macrostomia. This causes a cleft to form directly through the oral commissure, separating the orbicularis oris muscle from the surrounding skin and mucosa. [15]

Also possible are anophthalmia and microphthalmia. Colobomata of the iris or eyelids are frequently found without eyelashes. Ptosis of the upper eyelid, caused by dysfunction of the levator palpebral muscle, is frequently observed together with constriction of the vertical palpebral fissure. It is typical to have lateral canthi deficiencies with a narrowed horizontal fissure. The majority of patients have epibulbar dermoid cysts, which are solid, yellowish or pinkish-white, ovoid masses that can range in size from the size of a pinhead to 8 to 10 mm in diameter. The inferotemporal region of the limbus is where cysts are most frequently seen. It often has a smooth surface and many fine hairs. [16]

These cysts may be mobile or attached to the dermis, and they can develop anywhere on the planet or in the orbit. Several lesions may develop in each eye, and most individuals experience unilateral epibulbar dermoid cysts. On rare occasions, encroachment on the pupillary axis or lipid infiltration of the cornea might impair vision. The external ear frequently exhibits abnormalities, which can vary from anotia to a moderately dysmorphic ear. Inability to show a connection between the degree of microtia and the severity of skeletal deformity was a problem for Farkas and James.

The External Auditory Canal, the Middle- and Inner-Ear Structures, and Audiologic Findings

Patients with modest external ear deformities often have a small external auditory canal, although atretic canals are anticipated in more severe instances. On occasion, a little external ear with a typical middle-ear structure is visible. The kind of hearing loss will be determined by audiometry; 15% of patients will likely have conductive loss and, less commonly, sensorineural loss. It's possible for the ossicles to hypoplasia or agenize. In a thorough investigation, 57 individuals with hemifacial microsomia were assessed using air and bone conduction audiometry and temporal bone tomography. The authors were unable to link hearing function to the severity of auricular (external ear) malformation. The best method for documenting the anatomy of the middle ear is focused temporal bone CT scanning. [17]

Maxillomandibular Region

The first and second branchial arches' skeletal structures will experience varying degrees of hypoplasia. As a result, the damaged side's anteroposterior, transverse, and vertical dimensions are reduced, and the opposite side develops secondary abnormalities. Particularly in the maxillomandibular area.

Cranio–Orbito–Zygomatic Region

It is common to discover the zygomatic complex to have varying degrees of hypoplasia. Clinical manifestations of zygomatic hypoplasia include orbital dystopia, maxillary hypoplasia, glenoid fossa deficiency, and squamous aspect of the temporal bone deficit. A three-dimensional reconstruction of a craniofacial CT scan is the most effective way to see this. The asymmetry of the malformation makes it challenging to pinpoint constant, repeatable markers in the upper face. As a result, quantifying the skeletal deformity is challenging and inaccurate.

STAGING OF SKELETAL RECONSTRUCTION: TIMING AND TECHNIQUES

The patient's face rehabilitation should focus on the specific and distinctive aspects of the HFM, which may include the following: the external ear, the auditory canal, the zygomatic and orbital areas, the maxillomandibular regions, the facial soft tissues, and the middle ear structures.

Zygomatic and Orbital Reconstruction

According to this source, unless a functional handicap calls for it, repair of the cranial vault, malar, and orbital defects before the age of seven is not recommended. The cranio-orbito-zygomatic skeletal development is nearly complete by the time the patient reaches this age. When necessary, an adult-sized anterior cranial vault, orbit, and zygomatic complex may be built and matched with the opposite normal side with minimal consideration for how further growing may change the findings obtained. Because bicortical cranial bone makes it easier to divide the inner and outer tables for effective reconstruction after the age of 7, donor site skull reconstruction is likewise simpler after that age than it is in younger children. The posterior skull's graft donor site may also be rebuilt using artificial bone material. Several techniques and timeframes for reconstructing the upper face skeletal abnormalities caused by HFM have

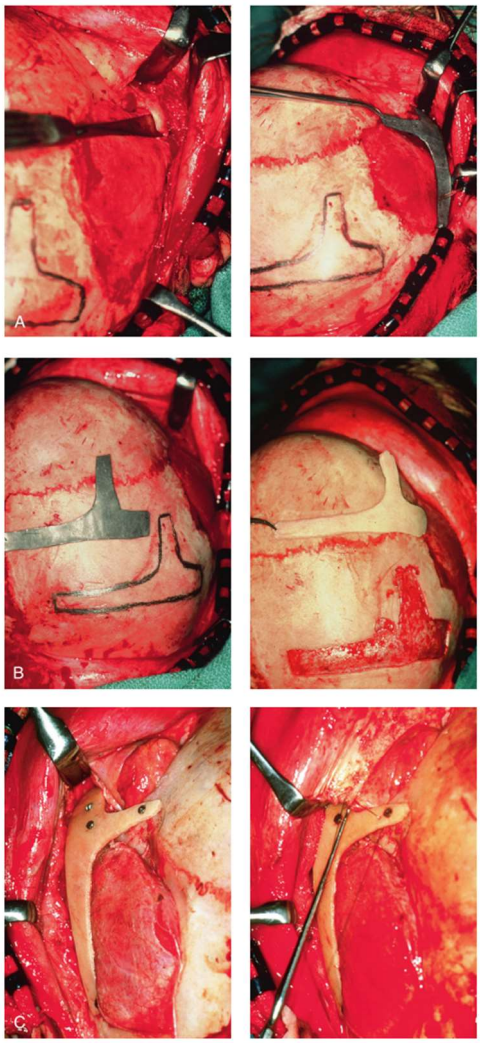

Fig 8.6: A 7-year-old child's intraoperative images illustrate the zygomatic-orbital reconstructive method covered in this chapter.

been suggested by other surgeons. Posnick and colleagues verified that full-thickness autogenous cranial bone creation of the entire zygomatic complex preserves volume and form better than onlay grafts, which had proven universally unsatisfactory. Onlay autogenous bone grafts from all attempted donor sites (e.g., skull, hip, rib) that have been implanted in the craniofacial area (e.g., supraorbital ridge, zygoma, anterior maxilla, angle of mandible, chin) over time have shown considerable and unexpected resorption. Also, it shouldn't be anticipated that the graft would develop or expand in volume at a rate that corresponds to that of the underlying bone, such as the zygoma. Others argue that even if the onlay graft partially resorbs, at least something has been accomplished. This viewpoint, however, has to be rejected since surface imperfections generate secondary deformities that worsen the initial deformity.

Maxillomandibular Reconstruction

Understanding the presenting TMJ-mandibular anatomy is crucial when deciding on the timing and methods for maxillomandibular reconstruction in a patient with HFM. The TMJ-mandibular malformation categorization, which was discussed previously in this chapter, is clinically applicable and improves communication. The fundamental maxillomandibular skeletal asymmetry and dysmorphology that needs repair in the patient with HFM and either a Type I or Type IIA malformation include the following: Reduced posterior facial height on the ipsilateral side is indicative of 1) degrees of changed facial height; 2) lessened horizontal projection; and 3) decreased facial breadth (i.e. deficiency on the ipsilateral side). These malformations frequently lead to canting (or roll orientation) of the pyriform apertures, the maxilla, and the gonial angles; shifting (or yaw orientation) of the maxillary, mandibular, and chin midlines away from the facial midline; clockwise rotation (or pitch orientation) of the occlusal plane; and an asymmetric class II malocclusion frequently with an anterior open-bite malocclusion. (18) A patient who has a Category IIB deformity needs to have a neocondyle built, in accordance with the categorization system given. Moreover, the creation of a neo-glenoid fossa is required for the person who additionally has a Type III deformity. Patients who have definitive reconstructive surgery of the maxilla, mandible, and chin regions at any time after jaw maturity (i.e., at 13 to 15 years of age for the HFM patient) typically achieve the best long-term function (i.e., speech, swallowing, chewing, breathing) and improved facial aesthetics. This should be done in conjunction with efficient orthodontic treatment after the permanent teeth have erupted and before the patient graduates. Similar with other dentofacial

abnormalities, extractions may be necessary to correct the inclination of the anterior teeth and to reduce orthodontic dental root crowding in order to prepare the jaws for surgical relocation.

The optimal time to restore type I and type IIA mandibular abnormalities is when all permanent teeth have erupted and orthodontic objectives have been met. Sagittal split ramus osteotomies of the mandible, together with Le Fort I osteotomies (typically performed in segments), and osseous genioplasty, can be used to achieve surgical goals. This combination uses conventional methods and does not call for bone transplants. By orthognathic operations, mirrorimage symmetry and Euclidian proportions of the skeleton must be roughly approximated for the reconstruction to be successful. Notwithstanding its drawbacks, this surgeon continues to favour costochondral graft restoration of the defective condyle ascending ramus at the time of skeletal maturity for Type IIB mandibular deformities (see the section to follow concerning condylar reconstruction with the use of costochondral graft). On the opposite side, a sagittal split ramus osteotomy is finished in order to derotate the distal mandible. This is paired with an osseous genioplasty and a Le Fort I osteotomy (typically performed in parts). To prevent relocation of the distal mandible, the contralateral side of the jaw must first have a ramus osteotomy. To achieve the desired lower jaw reorientation, the distal mandible is then fixed to the maxilla using intermaxillary fixation with a specially made acrylic splint. Before moving the distal mandible, the ipsilateral coronoid process may need to be removed. After that, bicortical screws are used to tightly anchor the contralateral ramus osteotomy. The autogenous costochondral transplant is used to repair the ipsilateral proximal mandible. The finest shape for mandibular repair comes from harvesting the rib graft from the opposing chest wall. A titanium miniplate and 2.0-mm or 2.3-mm screws are used to secure the rib transplant to the natural distal mandible.

The fixation plate runs along the inferior border of the mandibular body from the graft onward (fig. 7). Before attaching the graft, it is typically essential to recontour (with a bur on a rotary drill) the distal mandible's outer cortex. A Risdon neck incision made outside of the mouth is frequently used for graft implantation and fixation. With this treatment, avoiding intraoral incisions in the ipsilateral mandibular ramus area may reduce the risk of infection. Successful reconstruction depends on the neocondyle being properly seated in the glenoid fossa. Depending on the severity of the underlying soft-tissue and skeletal deficiencies, some patients may benefit more from a vascularized fibula composite flap than from a costochondral transplant.

The surgical reconstruction of the congenitally absent portions is necessary for the Type III glenoid fossa-mandibular deformity. With the Type IIB deformity, the mandibular repair is often performed as previously described. The zygomatic complex will need to be constructed whenever the glenoid fossa does (i.e., when there is a Type III deformity). Before mandibular reconstruction and when the patient is at least 7 years old (see the preceding section regarding orbito-zygomatic reconstruction), the glenoid fossa-zygoma and orbital reconstruction are best performed as separate operations. For the mandibular reconstruction, a vascularized fibula composite flap rather than a costochondral graft may be advantageous for the insufficiency of both the condyle-ascending ramus and the surrounding soft tissues.

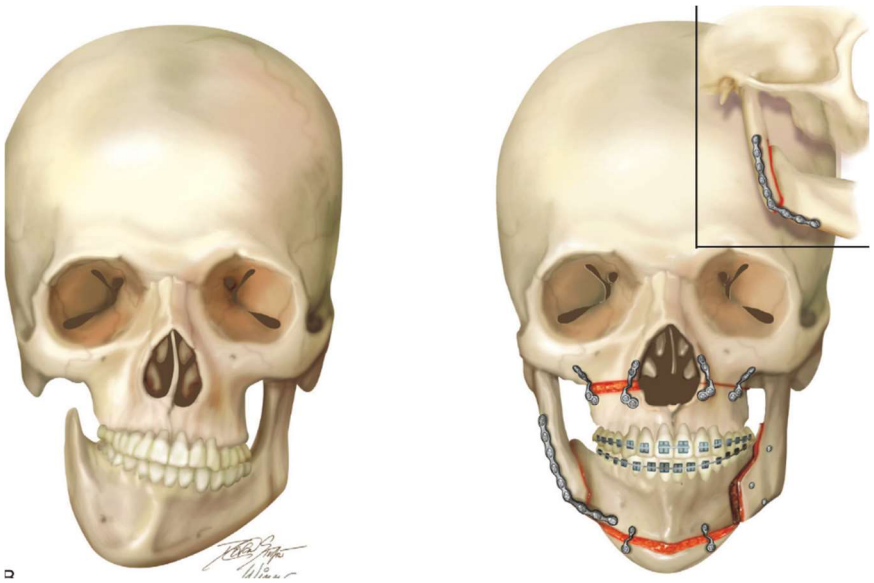

Fig 8.7, Illustrations of the presenting skeletal deformities and of the reconstruction that was carried out. Part B modified from an original illustration by Bill Winn

Facial Soft-Tissue Reconstruction

Several well-intentioned surgical attempts have been made to treat the soft-tissue deficits of the eyelid adnexal areas in HFM patients. The translation of a pedicled upper eyelid skin-muscle flap to the atrophic area of the lower eyelid is not technically challenging, but the adnexal scarring that ensues inevitably gives the surgery a surgical appearance that usually overshadows any benefits. For this reason, even modest eyelid surgical treatments for the kid with HFM may benefit from the patient and family consulting a skilled paediatric oculoplastic surgeon. Based on ocular function and eyelid aesthetics, decisions on removing epibulbar or bulbar dermoid cysts from the adnexal area are made. In a patient with HFM, the inferior displacement of the lateral canthus is a reflection of both orbital dystopia and hypoplasia of the

lateral canthi and other eyelid structural components. Osteotomies, bone grafting, and direct lateral canthopexy completion are useful and need to be performed when necessary, although these treatments won't completely reverse hypoplasia of the lateral canthal complex. When the temporoparietal fascia, the temporalis muscle, and the squamous component of the temporal bone all exhibit multilevel hypoplasia, there is a clinically apparent hollowing of the temporal fossa in the HFM patient. Local pericranial or temporoparietal flap attempts to replace the soft-tissue deficit are typically unsuccessful since these tissues constitute a component of the shortfall. The temporoparietal fascia and the temporalis muscle are these flaps, and their inherent hypoplasia explains why they are only marginally useful for adnexal area subcutaneous augmentation.[19]

In order to enhance the bulk of the soft tissues (i.e., the skin, subcutaneous tissue, and muscle), proponents of the DO approach for mandibular reconstruction in patients with HFM had hoped. Unfortunately, clinical trials have not shown evidence of this hoped-for benefit. The fact that good skeletal reconstruction, regardless of the method, will positively affect the soft-tissue envelope's aesthetics while without directly increasing the volume of the soft tissues, is crucial. [20]

Dermal fat grafts, autologous fat injections, and other soft-tissue augmentation techniques are becoming more and more common. Although the outcomes of autogenous fat injection might vary and are thought to be technique-dependent, they are often positive and definitely beneficial. Tanna and associates looked at the use of repeated autologous fat grafts to improve soft-tissue shape in HFM patients. The microvascular free-flap group had a face symmetry score of 121% at the final evaluation, whereas the fatgrafting group received a score of 99%. Additionally, neither the patient nor the doctor reported any statistically significant differences in the overall satisfaction ratings between the microvascular and fat graft groups. [21]

External Ear Reconstruction

A phased approach can be successfully used for surgical reconstruction of the external ear in the hands of a select group of specialists. The techniques Brent outlined remain the gold standard for auricular reconstruction. 10-15 The basis for an effective auricle repair is the installation of a well-sculpted autogenous cartilage structure. As most children's rib cartilage is sufficient for the restoration by the time they are at least 6 years old, Brent likes to wait till that time. The substantial cartilage block needed to create the ear's structure will then be found

in the synchondrotic area of the sixth and seventh ribs. [22,23,24] The position and size of the ear are selected, and a tiny preauricular incision is created using a prefabricated template of the contralateral ear and measurements of the face taken prior to surgery. The detachment of the ear with a skin graft, hairline management, and tragus building are additional phases of auricular construction. Alloplastic and homologous frameworks are further choices for external ear restoration. Currently used alloplastic materials include silicone and medpor (Porex Surgical, Inc, College Park, Ga). Even decades after repair, these alien substances are more prone to infection, soft tissue wound dehiscence, and mild trauma. Bovine cartilage cells can be produced in the lab and seeded on a synthetic, biodegradable ear template that is then implanted beneath the skin of an immunocompetent mouse as part of tissue engineering techniques that are still being evaluated. Regrettably, sculpted autogenous rib cartilage will continue to be the material of choice for the surgical restoration of the ear until tissue engineering advances past the currently encountered immunogenic issues.[25]

A determination must be made regarding the ultimate goal of middle ear surgery prior to concluding microtia repair. Every middle-ear surgery should be followed by auricular construction because, once an atretic ear canal is opened, scarring of the soft-tissue envelope significantly reduces the likelihood of a successful auricular reconstruction. Moreover, the requirement for condyle-ascending ramus restoration should be taken into account (i.e. Type IIB and Type III malformations). The location of each donor site becomes even more crucial if CCJ/rib graft harvesting is also necessary.

External Auditory Canal and Middle-Ear Reconstruction

Neurosensory hearing loss may occasionally be present in patients with HFM, however most hearing loss is caused by external auditory canal stenosis or atresia, middle ear cavity hypoplasia, or ankylotic or absent ossicles. Hearing conduction is restricted to the same degree if ankylotic or non-functioning ossicles were absent. For HFM patients, efforts to restore the middle ear's bones and the external auditory canal are often not made. Clinical issues often solely concern the patient's ability to pinpoint the source of the sound as long as adequate hearing is available in the contralateral ear. The majority of patients experience persistent conductive hearing loss as a result of efforts to restore middle-ear function, making "stereo" hearing uncommon.[26]

Most cases of hemifacial microsomia are random and lack a family trend. Variable deficits of the skeletal and soft-tissue components inside the first and second branchial arches, particularly on one side, are its defining features. Airway, eating, hearing, vision, and the need for psychological family support are all things that are taken into account during infancy and the early years of childhood. The level of deformity in the patient with HFM at birth is thought to be comparatively constant and nonprogressive with age.[27]

Nowadays, the reconstruction is staged to correspond with face growth patterns, visceral functions, and psychosocial development in order to repair the abnormalities linked to HFM. The goals of each stage and manner of therapy are made clear to the physicians and the family by a detailed morphologic study of each patient's abnormalities followed by a comprehensive reconstructive plan. We will enhance the outlook for those who are impacted by HFM as we continue to outline our justification for the timing, strategies, and scope of therapies before objectively assessing the functional, morphologic, and psychosocial results.[28]

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