

Case report

Hemifacial Microsomia: Case series and overview

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Abstract

A congenital facial abnormality called hemifacial microsomia (HFM) that causes hypoplasia of the first and second pharyngeal arches derivatives like the temporomandibular joint, mandibular ramus and body, masticatory muscles, the ear, and occasionally the facial nerve. In terms of developmental craniofacial anomalies, cleft lip and palate, which typically occurs unilaterally but can occasionally occur bilaterally, is the most prevalent, followed by HFM. There can be a number of defects, such as conductive hearing loss brought on by middle and external ear malformations. Diagnostic imaging and face structure categorization using the OMENS system are crucial for the pre-surgical evaluation of this abnormality. This developmental abnormality is managed in a multidisciplinary manner. We are showcasing a trio of examples with a variety of clinical and radiological characteristics, ranging from moderate ear deformity and facial asymmetry.

Case Series

Case 1

A 15 years old female was referred to department of Radio-diagnosis for cross-sectional Non-contrast CT scan of Face with 3D reconstruction by department of plastic surgery. Patient has complaints of facial asymmetry with skin tags in right pre-auricular region since birth (Figure 1 and 2). NCCT with 3D reconstruction reveals small sized right mandibular ramus, and both right condylar and coronoid process (Figure 3) however condylar process articulating with glenoid fossa with reduced right temporomandibular joint as compared to left side (Figure 6). Also, we can see the right parotid gland is lying anterior as compared to left side with a skin tag adjacent to right parotid gland. (Figure 4)

Case 2

A 17 years old female was referred by department of oral-maxillo-facial surgery for cross-sectional Non-contrast CT scan of Face with 3D reconstruction to department of Radio-diagnosis for complaints of facial asymmetry, malocclusion of teeth, right sided hearing loss. She also gave the history of right sided facial muscle paresis. NCCT was performed reveals facial asymmetry (Figure 7 and 8) & malocclusion

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Figure 1 and 2: Right sided facial asymmetry with skin tag on pre-auricular region.

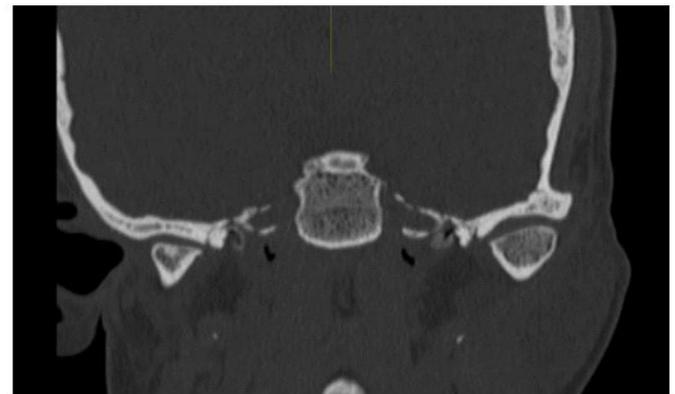


Figure 3: Demonstrating hypoplastic right condylar process and glenoid fossa as compared to left side.



Figure 4: Abnormal position of right parotid gland (Red arrow) as compared to left. Skin tag can also be seen (Yellow arrow)

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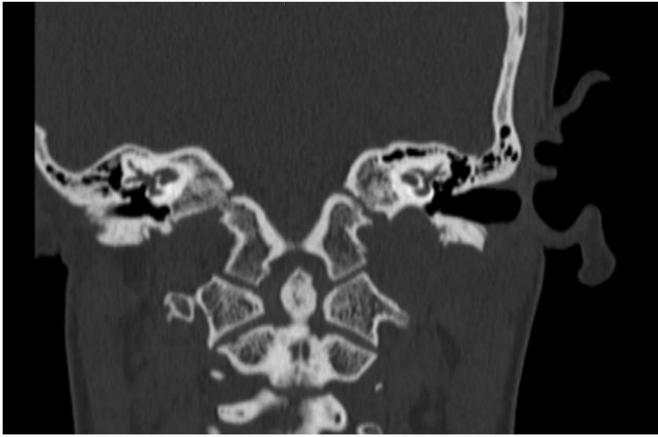


Figure 5: Normal B/L internal ears and external auditory canal



Figure 9: 3D reconstruction image showing malocclusion of teeth.

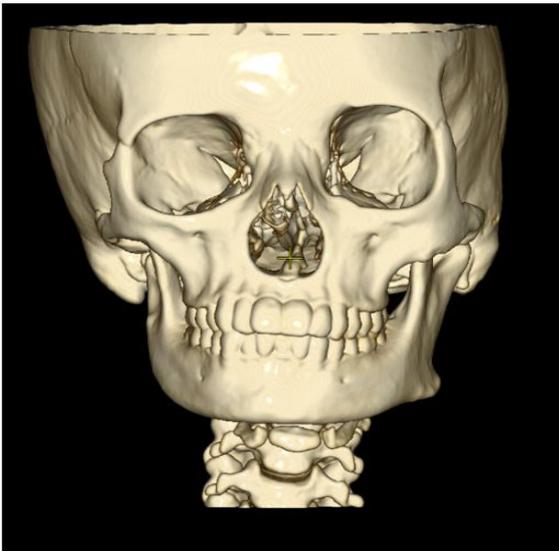


Figure 6: 3D reconstructed image reveals hypoplastic right ramus of mandible.

of teeth (Figure 9). The left mandibular ramus, left mandibular head appears small sized and hypoplastic glenoid fossa and it is not articulating with glenoid fossa (Figure 10,14 and 15) with aplastic external & middle ear and aplastic external auditory canal with non-visualization of ear ossicles. Also noted complete lack of pneumatization of left hypoplastic mastoid air cells (Figure 13). However, internal ear appears normal, skin tag noted in left pre-auricular region. Also, there is hypoplastic left maxillary sinus reduced height of left maxilla with mucoperiosteal thickening of left maxillary sinus with hypodense collection with right sided deviated nasal septum with hypoplastic left zygomatic

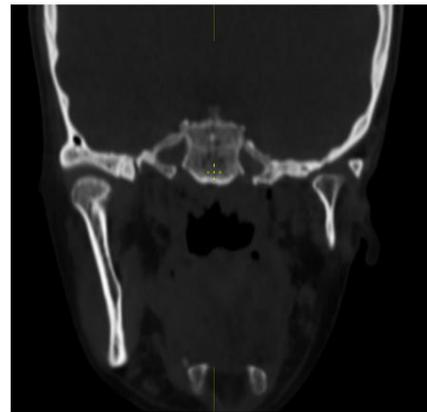


Figure 10: Hypoplastic left ramus of mandible, left condylar process and glenoid process.



Figure 11: Reduced height of left maxillary sinus with hypodense collection within the maxillary sinus.



Figure 7 and 8: 3D reconstruction image showing facial asymmetry with rudimentary left ear pinna with skin tags.

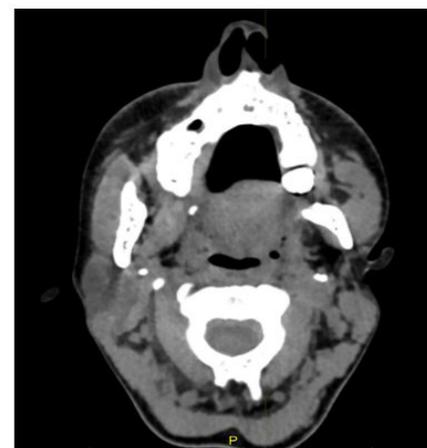


Figure 12: Absent left parotid gland

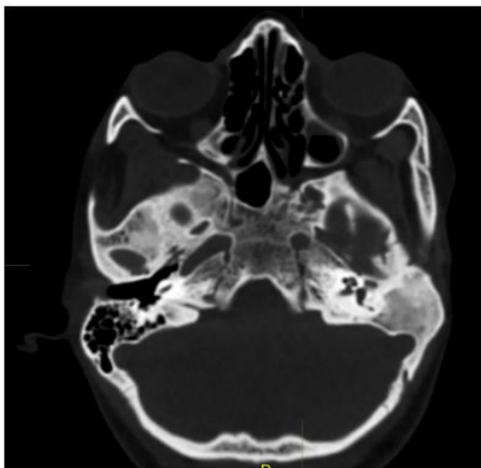


Figure 13: Aplastic external & middle ear and aplastic external auditory canal with non-visualization of ear ossicles with lack of pneumatization of left mastoid.

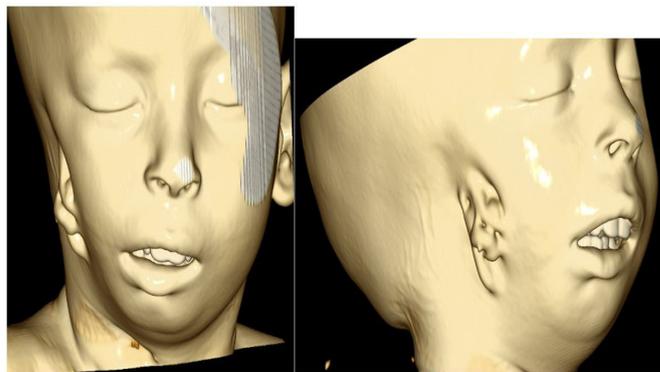


Figure 16 & 17: Facial asymmetry and rudimentary right ear lobe with skin tags. Also noted asymmetrically large right side of the head.

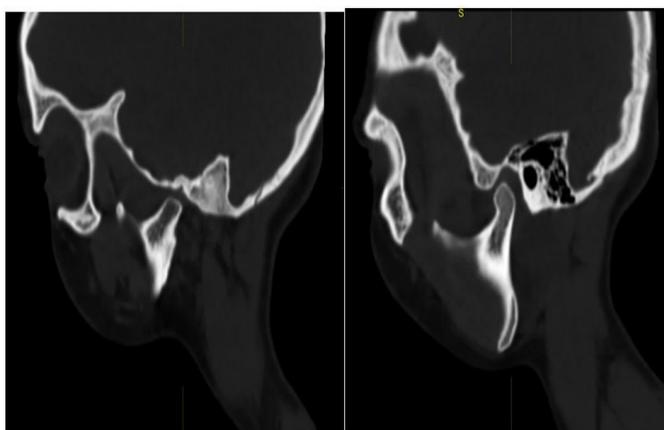


Figure 14 & 15: Comparing left and right ramus of mandible- The left mandibular ramus, left mandibular head appears small sized and hypoplastic glenoid fossa and it is not articulating with glenoid fossa arch. There is also absent left parotid gland (Figure 12)

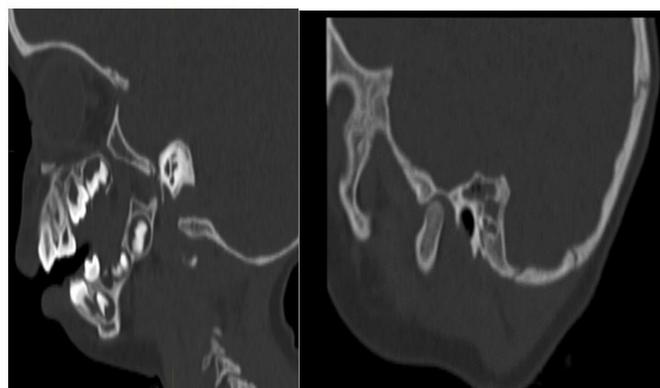


Figure 18 & 19: Small sized right mandibular ramus, not articulating with aplastic glenoid fossa and absent TMJ however left sided ramus appears normal.

Case 3

A 1 years old female child was referred by department of oral-maxillo-facial surgery for cross-sectional Non-contrast CT scan of Face with 3D reconstruction to department of Radio-diagnosis for complaints of facial asymmetry, malocclusion of teeth, right sided rudimentary ear lobule and absent external auditory canal. NCCT reveals of small sized right mandibular ramus, not articulating with aplastic glenoid fossa and absent right TMJ (Figure 18,19) with aplastic external & middle ear and aplastic external auditory canal with non-visualization of ear ossicles with asymmetrically large right side of the head. Also noted complete lack of pneumatization of right hypoplastic mastoid air cells . However, internal ear appears normal with anteriorly displaced dysmorphic ear auricle(pinna) (Figure 20,21 and 22).

Shows reduced height of right maxilla with hypoplastic right zygomatic arch with hypoplastic right sided pterygoid plates with absent right medial and lateral pterygoid muscle, right temporalis muscle. (Figure 23,24 and 26)

Non- visualization of right parotid gland (? Absent). Also noted partial fusion of lamina and spinous process of C2 & C3 on right side and lamina of C5-C6 on left side. (Figure 27 and 28)

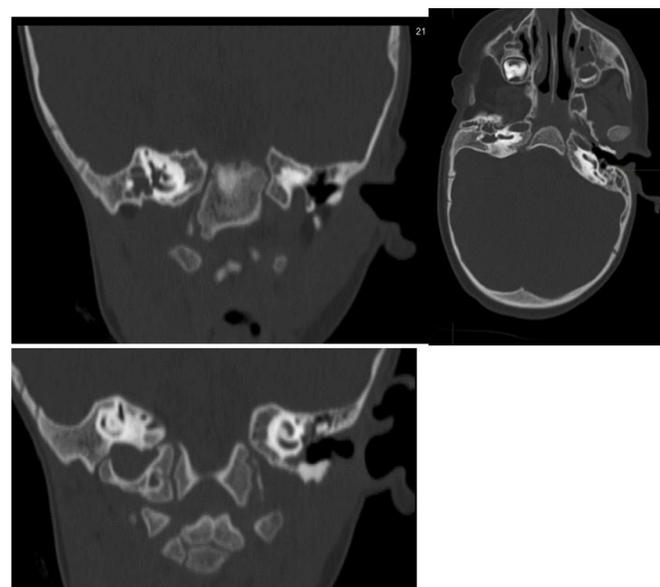


Figure 12: Aplastic external & middle ear and aplastic external auditory canal with non-visualization of ear ossicles with asymmetrically large right side of the head with complete lack of pneumatization of right hypoplastic mastoid air cells. However, internal ear appears normal with anteriorly displaced dysmorphic ear auricle(pinna).

Discussion and review of literature

Carl Ferdinand Von Arlt published the first description of hemifacial microsomia in 1881. First and second branchial arch derivatives with a wide range of phenotypes are involved in HFM. Other names for it in-

clude otomandibular-facial dysmorphogenesis, lateral facial dysplasia, and first and second branchial arch syndrome [1]. The first branchial arch plays a major role in HFM, which also affects the temporomandibular joint, mandibular ramus, masticatory muscles, and ear. Microtia or pinna atresia is caused by abnormal auricular hillock development, and its severity is inversely correlated with that of faulty external auditory canal development [2]. HFM affecting about one in 5,000 newborns , More than 15 terms, each representing the viewpoint of a different physician, have been used to describe this condition, includ-

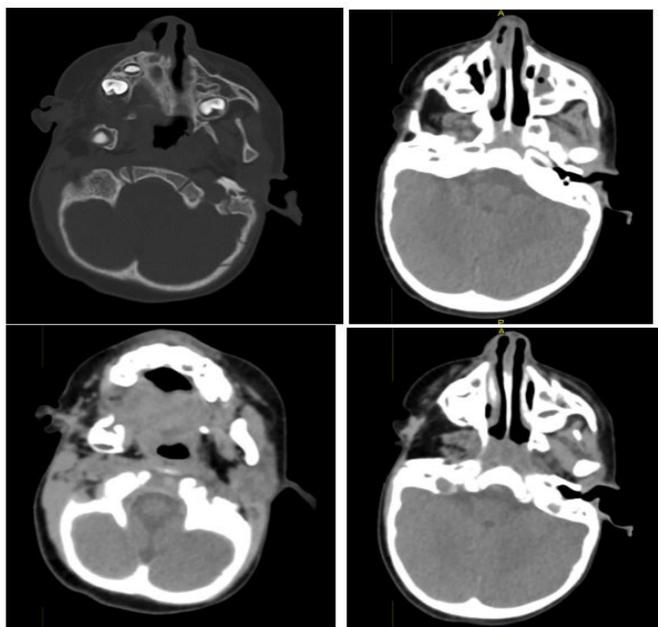


Figure 23,24,25 & 26: Hypoplastic right sided pterygoid plates with absent right medial and lateral pterygoid muscle, right temporalis muscle (Absence of muscles of mastication on right side and they are replaced by fatty tissue). Non- visualization of right parotid gland (? Absent).

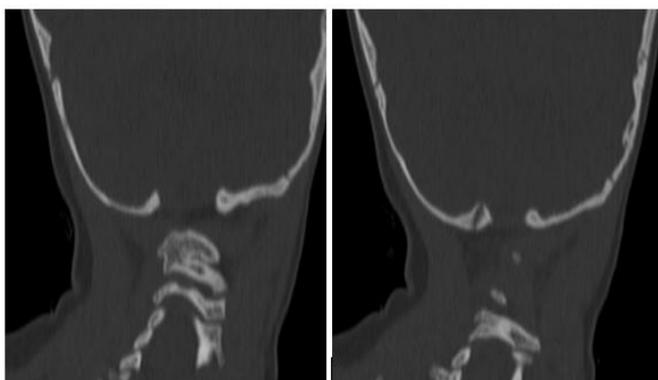


Figure 27 & 28: Partial fusion of lamina and spinous process of C2 & C3 on right side and lamina of C5-C6 on left side.

ing Goldenhar's syndrome (oculoauriculovertebral dysplasia) [3,4,5]

Facial Musculoskeletal Structures and Ossicles

Development of Branchial Arches

Mesenchyme generated from lateral mesoderm makes up the branchial arches at first. The branchial arches begin to grow into distinct structures starting in the fourth week of pregnancy as neural crest cells move into the arches of the future head and neck [6]. In the lateral lips of the neural folds, neural crest cells develop and separate during neurulation. The cardiac conotruncal septum, the adrenal medulla, and melanocytes are only a few examples of the varied structures formed by neural crest cells after they move across the head, neck, and body. An arch artery, a cranial nerve, neural crest-derived muscle anlage, and cartilaginous precursors are all found within each branchial arch.

First Branchial Arch

Indirect ossification of the first-arch dermal mesenchyme results in the direct ossification of the jaw, maxilla, zygomatic temporal bone, and squamous temporal bone [6]. Accurate evaluation of the mandible is crucial for presurgical planning because hemifacial microsomia is characterised by asymmetric development of the mandible [7]. A popular classification scheme for the mandible employs the numbers 1 for

a small but normally shaped mandible, 2 for a small and abnormally shaped mandible, where 2A denotes that the condyle is in the normal position and 2B that it is displaced, and 3 for ramus and condyle aplasia. [8]

The malleus and incus are said to be pre-formed in arch cartilage from the first branchial arch, according to the majority of embryology textbooks [6]. According to a more complex origin theory put forth by other authors, the malleus manubrium and the incus long process are derived from the cartilage of the second branchial arch, and the first arch contributes to the head and neck of the malleus as well as the body and short process of the incus [9]. Malleus and incus malformation, fusion, and lateral displacement against the lateral wall of the tympanic cavity are common anomalies seen in patients with hemifacial microsomia [8].

The muscle anlage of the first branchial arch gives rise to the masticatory muscles. The soft tissues of the face become asymmetrical in appearance and function due to hypoplasia of these muscles. Although the osseous muscle attachment sites and the development of the masticatory muscles are closely related, the muscle mass cannot be predicted by the bone morphology, necessitating an independent assessment of the muscles [10]. Imaging analysis provides substantially more data regarding the condition of these muscles than a clinical examination does. The anterior belly of the digastric muscle, the mylohyoid muscle, the tensor muscle of the velum palatinum, and the tensor tympani muscle are also derived from the first arch. Imaging investigations can show asymmetric development of these structures.

Second Branchial Arch

The second branchial arch contributes less to facial development than the first. Cartilage outgrowths of the second arch include the stapes, lesser horn, superior margin of the hyoid bone, styloid process, and stylohyoid ligament [6]. The muscles of the face, the posterior abdomen of the digastric, the stapes, and the stylohyoid [6] arise from the musculature of the second branchial arch.

External, Middle, and Inner Ear

Auricular Hillocks

Six hillocks of the first and second branchial arches that appear on either side of the first branchial cleft give rise to the pinna [6]. The majority of the final pinna comes from the second arch. The severity of the aberrant external auditory canal development is inversely correlated with the severity of the auricular hillock abnormality, which results in microtia or pinna atresia [8]. Preauricular tags that are abnormal accessory hillocks are present in some people with hemifacial microsomia.

First Branchial Cleft (Groove)

The first branchial cleft, which is bordered with ectoderm, deepens to form the external auditory canal [6]. During development, the ectodermal lining cells of the external auditory meatus multiply to produce a meatal plug that later recanalizes. There are different degrees of soft-tissue and bone external auditory canal atresia or stenosis as a result of the failure of cleft deepening or of recanalization of the meatal plug. Although they are caused by buried cell resting or unobliterated sinuses [11], cysts of the first branchial cleft and the branchial sinus are unrelated to hemifacial microsomia.

First Branchial Pouch

The first branchial pouch, which is lined with endoderm, elongates to produce the tympanic cavity. The ossicles, which were independently formed from the cartilage of the first and second arches, are covered in endoderm as a result of the tubotympanic recess gradually cavitating around them. The pharynx and the growing tympanic membrane are both abutted by the increasing tympanic cavity, which is still connected to it through the little auditory tube. An inner layer of endoderm, a middle layer of mesoderm, and an outside layer of ectoderm make up the tympanic membrane. Unfavorable candidates for surgery include patients with aural atresia who have a facial nerve course that crosses the oval window or a tympanic cavity that is less than 3 mm long from the medial promontory of the cochlea to the lateral area of bony plate atresia.

Otic Vesicle

The otic placode develops as a surface thickening of the ectoderm atop the rhombencephalon during the end of the third week of gestation. The placode then pinches off to produce the otic vesicle before invaginating to create the otic pit [6]. The otic vesicle relocates near the growing middle ear and transforms into the intricate membranous labyrinth. At the same time, the otic vesicle causes the mesoderm in the area to condense into the encasing bony labyrinth. Patients with hemifacial microsomia frequently exhibit abnormalities of the membranous labyrinth and bony labyrinth, such as hypoplasia and atresia of the oval and round windows as well as aberrant cochlea and semi-circular canal development [12].

HFM is a complicated malformation syndrome with a wide range of clinical symptoms that affect the facial skeleton and various organ systems, as well as a large number of genetic and teratogenic correlations [13,14]. Auricular defects, preauricular tags and fistulas, microtia-atresia, hypoplasia of the mandible, the maxilla, and the orbit, microphthalmia, epibulbar dermoid, strabismus, conductive or sensorineural hearing loss, and hypoplastic facial muscles are a few examples of deformities [15]. The condition known as Goldenhar syndrome in HFM is characterised by abnormalities of the heart, kidneys, and vertebrae. Gorlin proposed the name "oculoauriculovertebral dysplasia" (OAVD) for this condition [16].

The clinical manifestations of HFM range from mild facial asymmetry to significant underdevelopment of one facial half, with implications for the orbits, a partially developed ear, or even an absence of the ear entirely. The facial midline and chin are misaligned and shift to the affected side. An oblique lip line results when one corner of the mouth is frequently higher than the other. Other asymmetrical symptoms include abnormalities of the external and internal ear, a unilaterally shorter zygomatic arch, and unilaterally hypoplastic maxillary and temporal bones. Middle ear abnormalities frequently result in hearing loss, and the temporal and zygomatic branches of the facial nerve frequently malfunction. [17,18]

Classification Systems

Pruzansky Classification System (1969) [19]- Using x-rays of people with Craniofacial Microsomia(CFM), Pruzansky categorises the

Table 1

Grade I	Smaller than preserved normal side
Grade II	Condyle, ramus, and sigmoid notch identifiable, but grossly distorted in size and shape
Grade III	Grossly distorted ramus with loss of landmarks or agenesis

patterns of mandible deformities and malformation that are discernible in these people.

Anteroposterior and lateral cephalograms of the temporomandibular joint and its deformities were described by Kaban et al. in their addi-

Table 2

Grade I	Small mandible
Grade IIA	Short mandibular ramus of abnormal shape; glenoid fossa in satisfactory position
Grade IIB	TMJ abnormally placed inferiorly, medially and anteriorly
Grade III	Absent TMJ

tion and modified it. [20,21] Depending on where the glenoid fossa was located, the type II mandible was classified into two groups.

This was a crucial differential since they believed that type IIA mandibles did not require surgical TMJ therapy but type IIB mandibles did. Along with more recent categorization schemes that take into account characteristics seen on high-resolution three-dimensional computed tomography (CT) scans, these classification schemes of the mandibular defects linked to CFM are still widely employed. [22] The other craniofacial abnormalities connected to CFM are not included in these classification schemata, though.

Sat classification

The skeletal, auricle, and soft tissue (SAT) classification system of hemifacial microsomia [23]

Skeletal categories

- S1 = Small mandible with normal shape
- S2 = Condyle, ramus, and sigmoid notch identifiable but grossly distorted; mandible strikingly different in size and shape from normal
- S3 = Mandible severely malformed, ranging from poorly identifiable ramal components to complete agenesis of ramus
- S4 = An S3 mandible plus orbital involvement with gross posterior recession of lateral and inferior orbital rims
- S5 = The S4 defects plus orbital dystopia and frequently hypoplasia and asymmetrical neurocranium with a flat temporal fossa.

Auricle categories

Orbit

O0	Normal orbital size and position
O1	Abnormal orbital size
O2	Abnormal orbital position (arrow up or down)
O3	Abnormal orbital size and position

Mandible

M0	Normal mandible
M1	The mandible and glenoid fossa are small.
M2A	Short ramus, glenoid fossa is in anatomically acceptable position
M2B	Short ramus, TMJ is inferiorly, medially and anteriorly displaced with hypoplastic condyle
M3	Complete absence of ramus, glenoid fossa and TMJ

Ear

E0	Normal Ear
E1	Mild hypoplasia & cupping, all structures present
E2	Absence of external auditory canal with hypoplasia of concha
E3	Mal-positioned lobule with absent auricle, lobular remnant inferiorly and anteriorly displaced

Facial Nerve

N0	No facial nerve involvement
N1	Upper facial nerve involvement (temporal zygomatic)
N2	Lower facial nerve involvement (buccal, mandibular, cervical)
N3	All branches of facial nerve affected

Soft Tissue

S0	No obvious soft tissue or muscle deficiency
S1	Minimal subcutaneous/muscle deficiency
S2	Moderate—between the two extremes S1 and S3
S3	Severe soft tissue deficiency due to subcutaneous and muscular hypoplasia

All 3 cases which have been reported here were classified by using various above mentioned classifications [Table3].

S.No	Features	Case 1	Case 2	Case 3
1	Facial Deformity	Mild	Severe	Severe
2	Orbit size / Position	Normal	Normal	Normal
3	Small or Anomalous Pinna	Normal	Rudimentary with absence of EAM	Rudimentary with absence of EAM
4	Pre-auricular skin tags	Present	Present	Present
5	Condylar aplasia/hypoplasia	Mild Hypoplasia	Condylar hypoplasia	Aplasia of ramus, condyle & coronoid process.
6	Malocclusion	Absent	Present	Present
7	Parotid Gland	Anteriorly placed	Absent	Absent
8	Hearing loss	Absent	Present	Present
9	Facial Paresis/ Palsy	Absent	Mild facial paresis	Mild Facial Paresis
10	Pruzansky Classification (Modified)	Grade I	Grade IIA	Grade III
11	SAT Classification	S1A0T1	S2A2T1	S3A3T2
12	OMENS Classification	O0M1E0N0S0	O0M2AE3N2S2	O0M3E3N2S3 with cervical vertebral segmentation anomaly

A0 = Normal

A1 = Small, malformed auricle retaining characteristic features

A2 = Rudimentary auricle with hook at cranial and corresponding to the helix

A3 = Malformed lobule with rest of pinna absent.

Soft tissue categories

T1 = Minimal contour defect with no cranial nerve involvement

T2 = Moderate defect.

The OMENS categorization scheme, later renamed OMENS+ to cover extracranial symptoms, is the most extensively used system[24,25]. Each feature is given a severity score, and the acronym stands for orbit, mandible, ear, nerve, and soft tissue. A visual illustration of the OMENS system was published more recently[26].

TREATMENT

The oral occlusion and the underlying bone condition must be taken into account when planning a treatment plan. Usually, a surgical and orthodontic technique is used.[27] Functional appliances, or growth-directing devices, were originally employed to promote growth and

reduce the extent of orthognathic surgery required once the kid had finished growing. Congenital micrognathia, facial asymmetry, maxillo-mandibular hypoplasia, and HFM are examples of skeletal malformations that are routinely treated in non-growing adult patients with osteotomies, acute orthopaedic mobility, and osseous fixation.[28-32] Unfortunately, the benefits were frequently undermined by the inherent danger of relapse brought on by muscles' incapacity to be stretched severely. [33] Inadequate soft-tissue adaptation may also affect function and appearance. [34] Infection, discomfort, and donor site morbidity were significant postoperative concerns when autogenous costochondral grafting was performed in more severe abnormalities. [35] Distraction osteogenesis is a substitute technique that is now routinely used. [36-40] New bone is created between the surfaces of bone segments that are gradually being pulled apart by incremental traction throughout this process. Gavril A. Ilizarov, an orthopaedic surgeon, initially reported this approach of gradually forming bone after a surgical corticotomy—sectioning of the cortical plates—in 1988[41,42]. It has since found widespread application in the field of craniofacial

abnormalities.[43-46]

Since Pierre Fauchard first documented the use of the expansion arch in the 18th century, dental traction principles have been used to treat skeletal abnormalities. [47] Recently, distraction osteogenesis has been used to advance severe retrognathia in Pierre Robin sequence patients, preventing the necessity for tracheostomy surgery to treat obstructive sleep apnea.[48] In patients with craniofacial abnormalities, Figueroa and Polley[49] employed distraction osteogenesis to treat midface hypoplasia of the maxilla. Ko and colleagues have shown that distraction of the maxilla can improve facial equilibrium and aesthetics. [50] Given the wide range of uses for the oral and maxillofacial region, it is crucial for dental professionals to understand the physiological and physical mechanisms behind distraction osteogenesis. [51] Distraction osteogenesis entails a corticotomy with only minor endosteal and periosteal disruption. The gradual separation of the two surgically separated bony segments then results in the formation of new bone. [52] They are divided and lengthened every day until the appropriate length is reached. This treatment enables the growth of healthy new bone that shares the same properties as the nearby bone. In addition to exerting traction stresses on the bone, soft tissue is

also put under tension, which starts a chain of adaptive alterations known as distraction histogenesis. [53,54] The lower jaw can expand in multiple directions thanks to the concurrent development of the soft-tissue functional matrix. [55] Active histogenesis occurs in skin, fascia, blood vessels, nerves, muscle ligaments, and periosteum under the impact of tension brought on by distraction.[56] The soft tissue can stretch and adapt progressively without losing feeling since the lengthening operation moves slowly over several weeks. Due to the absence of both bone and the surrounding soft tissue in patients with HFM, this secondary gain is considerable. [57-63]

Patient consent declaration

The authors confirm that they have all necessary patient permission paperwork on file. The patient's/patients' form having granted their permission for his/her/their pictures and other clinical data will be reported in the journal. Patients are aware of their names and Initials

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