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Craniofacial Anomalies : A Study Of Deformities Of Craniofacial Structures.

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Abstract :-
Craniofacial anomalies are deformities occurring in the growth of head and facial bones, including the structures of nose, mouth, jaws and ears. Craniosynostosis is a developmental anomaly that Occurs as a result of abnormal and non typical sutural fusion. Cleft lip and Cleft palate are Openings or space or splits in the roof of the mouth and lip. And spacing in the lip while forming cleft palate. Hemifacial microsomia is congenital malformation in which one side of the face is underdeveloped. Ankyloglossia, also known as tongue-tie is a condition where a band of tissue connects the underside of the tongue to the floor of the oral cavity that prevent the tongue from moving freely. Plagiocephaly is an deformation characterized by asymmetrical distortion or flattening of skull.
Keywords :- *Ankyloglossia, cleft lip and palate, craniosynostosis, Hemifacial microsomia, plagiocephaly.*

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I. INTRODUCTION

Head is considered as the most complex structure of our body. The skull and facial bones encloses the brain and supports the functions of breathing, feeding, hearing, vision and speech [1] . And these structures are susceptible to formational error that results in craniofacial anomalies.

Craniofacial anomalies are malformations occurring in the growth of head and facial bones, including the structures of nose, mouth, jaws and ears. They can be congenital and usually occurs during embryological development. Congenital craniofacial anomalies are those including cleft lip and cleft palate commonly and craniosynostosis, Hemifacial microsomia. Studies shows that each year 35% of babies are born with such dysregulations that affects the individual's appearance And aesthetics , vision, hearing, breathing and many such

functions. The following anomalies discussed below are part of such pathologies portraying the craniofacial manifestations [2].

CRANIOSYNOSTOSIS

Craniosynostosis is a developmental abnormality which Occurs as a result of improper or abnormal and nonphysiological sutural fusion. In a newborn, When one or more Sutures of the head are prematurely closed, The Result is an abnormally shaped skull as well as sensory, Respiratory disturbances and neurological dysfunctions [3,4,5,6,7]. The Prevalence of craniosynostosis is determined to be 1 in 2100-2500 births [3,8,9]. predisposing factors for this condition includes Both environmental factors like abnormal position, prenatal exposures to teratogens, maternal smoking, and drugs mainly antiepileptics such as valproic acid and phenytoin and also genetic factors like single gene mutations, chromosome abnormalities account for approximately 20% of all craniosynostosis. Most of the genes that are linked to craniosynostosis are inherited in an autosomal dominant manner [3,6 ,10-13].

TYPES OF CRANIOSYNOSTOSIS

Scaphocephaly

It is mainly due to early closure of the sagittal suture which runs front to back, down the middle of top of the head and is seen commonly in premature infants. In this, The head is elongated in anterior-posterior direction and is shortened in bilateral direction. In some, frontal bossing is clearly visible and the ridging of sagittal suture is palpable. There is male predominance, with a ratio of 3.5:1 [3,14,15,16,17].

Anterior plagiocephaly

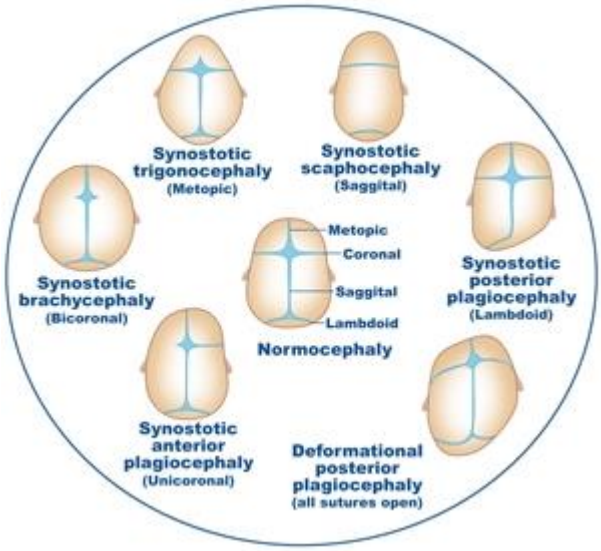
This results from premature closure of the coronal suture. due to arrested growth, forehead is flattened On the affected side and higher supraorbital margins forms a characteristic sign known as the Harlequin sign is seen on the radiographs. The forehead is protruded forward On the opposite side. nasal septum deviation towards the normal side is observed. It is more common in girls than in boys with the ratio of 2:1 [3,14,15,16,1718].

Positional plagiocephaly

Positional plagiocephaly is characterized by displacement of the ipsilateral ear and forehead anteriorly, giving a parallelogram shaped head. The ipsilateral occipital flattening and contralateral occipital bossing is commonly seen. The effects are primarily cosmetic and it does not require surgical intervention [3,7,19].

Trigonocephaly

Trigonocephaly is due to premature closure of the interfrontal suture. In this, The head is broad at back and the forehead is pointed and narrow. The forehead has a triangular shape When viewed from above,. The orbits are abnormally close together (hypotelorism) [3,7,14,15].



Brachycephaly

Brachycephaly is fusion of right or left side of the coronal suture that runs across the top of the head from ear to ear. It is also known as coronal synostosis . The skull appears short as a result of the fused coronal

suture. The forehead and occipital part of the head are usually flattened and the frontal bone is more prominent and elongated in vertical direction. Harlequin sign and hypertelorism of the orbits is seen [3,20].

Diagnosis

Congenital craniosynostosis can be observed at birth or during the first year of life of the affected infant.

The most common and accurate method is the CT scan with 3D reconstruction, but since it has a high possibility radiation risk, this option requires consideration. MRI is considered as a less accurate option when compared to CT and is usually reserved for children in which the CT revealed any anomalies of the brain [3,21].

Genetic testing is done for FGF receptor genes, which have been studied as the genes that are most commonly associated with these syndromes. The genes FGFR2 and FGFR3 also undergo testing, as well as transcription factors (TWIST, MSX2) as it has been identified that 57 genes have a relationship and are the possible underlying cause of the craniosynostosis. [3,21,22,23].

Treatment and management

The management depends on what kind of craniosynostosis it is. The uncomplicated and nonsyndromic types can be managed surgically while some syndromic forms requires urgent surgical intervention to prevent the involvement of other systems.

Endoscopic suturectomy: it is done in a patient less than six months of age as the bone is observed to be more flexible and manageable by an endoscope. The postoperative recovery is faster, there is minimal blood loss, and the duration of the surgery is much shorter compared to an open craniotomy. The only disadvantage of this method is that there is a need to use of remodeling helmet for 4 to 6 months postoperatively [3,6,21].

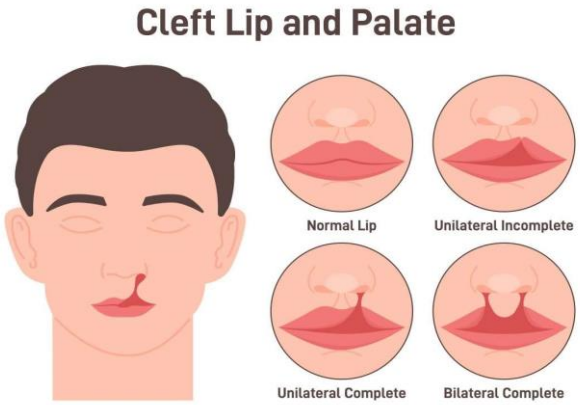
Open craniotomy: this is done in patients who are of more than six months of age because the bones have now become more rigid and cannot be easily manipulated with an endoscope. This allows for a better remodeling of the skull and decreases the postoperative need for helmet use [3,6,21].

The main goal in the management of conditions like this is to create enough space in the cranium for the brain to grow and to undergo proper development to provide the child with decent appearance.

The best time for correction of craniosynostosis is observed to be between 6 to 12 months of age where there are no signs of increased Intracranial pressure or airway complication [3,21].

CLEFT LIP AND PALATE

A separation in the lip, palate, or both is known as cleft lip and palate. Among craniofacial abnormalities, cleft lip, and cleft palate are the most often seen at birth. Clefts arise when the fusion process is harmed during the fifth and seventh intrauterine weeks. This irritates the nasal septum, which leads to recurrent infections of the middle ear and respiratory system. Including the possibility that, when swallowing or masticating, the tongue may be positioned in the cleft gap, thus widening it even further [24, 27].



Cleft lip

Splits or openings in the lip and roof of the mouth. As well as lip spacing during cleft palate formation. Occurs when there is a hole in the roof of the mouth that can extend into the nasal cavity and does not completely seal. Any portion or side of the palate may be affected by the cleft [26]. It may be in the throat or at the front of the mouth. From a hard to a soft palate, the lip may also be included in the cleft.

Operation

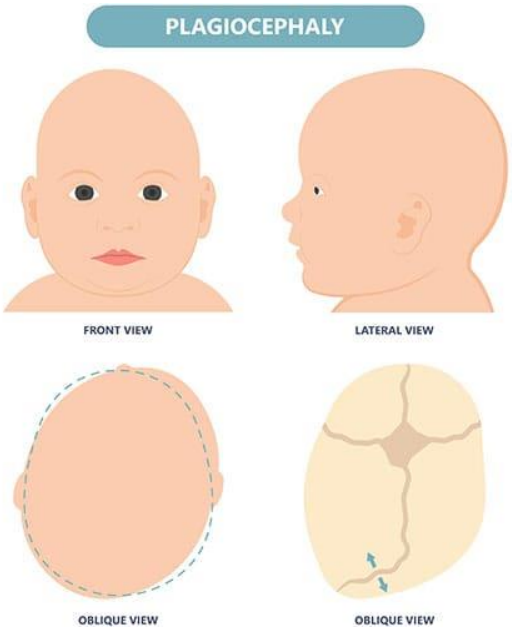
The age range for cleft lip therapy is three to four months. While some people have their cleft palate surgery later, it is usually advised to finish the procedure between the ages of 7 and 12 months. Restoring palatal function and proper speech development, as well as returning the lip and nose to their anatomical structures, are the primary surgical aims of cleft therapy. Reconstructive surgery is tailored to the specific cleft type of each patient, taking into account various surgical approaches [29, 30].

Avoidance

To help prevent neural tube abnormalities, the Institute of Medicine and the US Preventive Services Task Force advise childbearing women to take 0.4 mg of folic acid daily [30].

PLAGIOCEPHALY

Plagiocephaly is increasing in the infants. Plagiocephaly is an asymmetric deformation of the skull due to a lot of reasons; first birth, assisted labor, multiple pregnancy, prematurity, congenital muscular torticollis, and position of the head and neck asymmetries are common in typical healthy new-borns Within these asymmetries, positional plagiocephaly is a general in cranial distortion positional plagiocephaly features are asymmetrically seen in occipital flattening, accompanied by anterior displacement of the ear on the same side, parietal protuberance and often ipsilateral frontal protuberance, with fellow frontal flattening[31,33]. These characteristics make the head look like a parallelogram viewed from above Facial findings can be associated with the condition, but positional plagiocephaly is not implied. First births, aided labor, numerous pregnancies, birth injuries, congenital abnormalities, preterm newborns, males, and particularly congenital muscle torticollis are risk factors for positional plagiocephaly. The condition is commonly caused by birth deformities of the head and neck, which are not repaired since the baby sleeps in the same posture. The effects of gravity, the fluidity of the cranial sutures, and the plasticity of the brain cause the skull to change form during birth. This makes it possible for the relatively big head to pass through the small birth canal [33].



Presentation of Clinical Data

When caregivers notice an irregular head shape during a baby's movements or when guardians voice worries about their infants' aberrant head shape, DP is suspected. Right-sided anomalies are more prevalent than left-sided ones. Fetuses often lie with their heads toward the right side of the mother's pelvis and the left side of their forehead against her lumbosacral spine when still in the womb. Consequently, a lot of babies still spend more time on the left Differential Diagnoses Differential diagnoses include central nervous system (CNS) diseases and abnormalities, vertebral deformities, craniosynostosis, DP, and Brachycephaly for newborns presenting with cranial deformity, with or without evident CMT or positional preference [29, 32]. Urgent diagnosis and treatment are necessary for infants who have suspected CNS tumors, vertebral abnormalities, or craniosynostosis. Healthcare professionals must rule out these serious illnesses before examining alternative possibilities.

Supervisory The management guidelines are subject to change and are not supported by sufficient, high-quality research.

Research has suggested that DP can resolve with time without treatment. Once the infant can sit upright independently. However, some researchers believe DP is more likely to worsen over time and will not resolve without intervention [31].

HEMIFACIAL MICROSOMIA

This is mainly known as the unilateral otomandibular dysostosis or lateral facial dysplasia, is mostly asymmetrical, congenital malformation of the 1st and 2nd branchial arches present and the second common craniofacial anomaly after the cleft lip and palate of the human body[34,42,45]**.**

Etiology

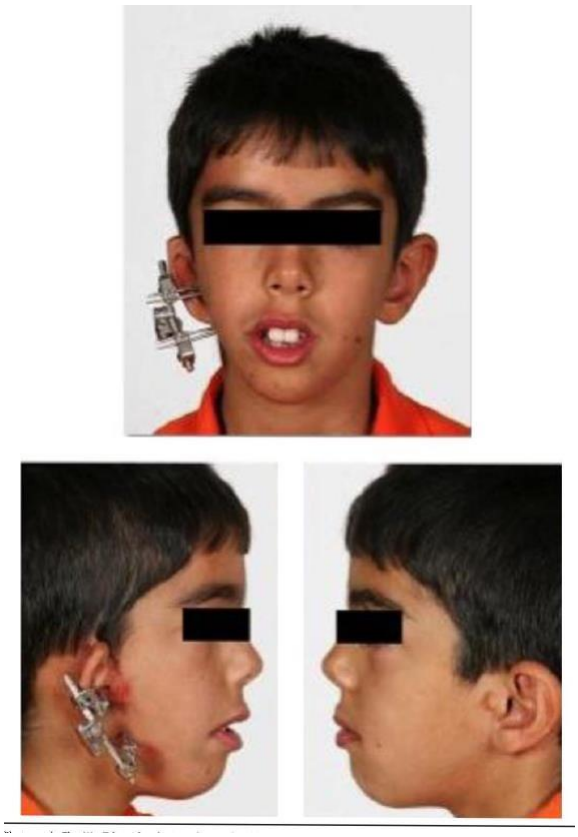
The main reason of HFM is not clear, but two leading theories are of vascular injury of the stapedial artery and anomalous migration of the neural crest cells.[35,37]. The heterogeneous phenotypical appearance of hemifacial microsomia has been done to be caused by the combination of genetic and environmental factors that will disrupt the vascularization and the development of the first and most important second pharyngeal arches in the process.[36]

Pathophysiology

Hemifacial microsomia can present with many deformities which is involving the eyes, ears, and the first two pharyngeal arches most importantly. Ocular deformities will mostly include strabismus, anophthalmia, microphthalmia, eye asymmetry, cleft eyelid, and also exophthalmia. Auricular abnormalities will include preauricular appendage, preauricular fistula, microtia, ear asymmetry, and external auditory canal atresia [34].

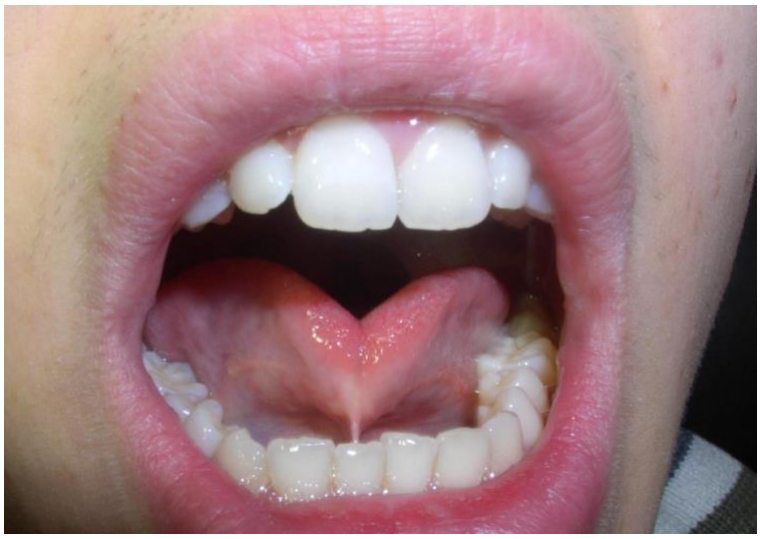
Treatment

Cartilage and bone can be harvested from most important costochondral cartilage, iliac crest, temporal bone, or fibula to augment the hypoplastic mandible [45]. Disadvantages of this technique of grafting include high chances of wound infection, creation of a donor site defect, re-ankylosis of the joint, possible fracture, resorption of graft material, and recurrence of asymmetry. Ramus importantly, the main part as we know is related to the mandible region of the body, which needs to be corrected.[48].



ANKYLOGLOSSIA

This mostly also known as tongue-tie [47,49,50]. This is a highly important topic which can be discussed. There is a lack of consensus regarding all aspects of this disease.[46]



The board of Tongue-Tie Professionals have define this lingual frenulum as a tissue remnant that is located in the midline between the tongue's ventral surface and the mouth's floor. When the lingual frenulum limits the function of the tongue, it is called the symptomatic tongue-tie or symptomatic ankyloglossia tongue tie, this means there will be peculiar analysis with efficient methods[51,54,56].

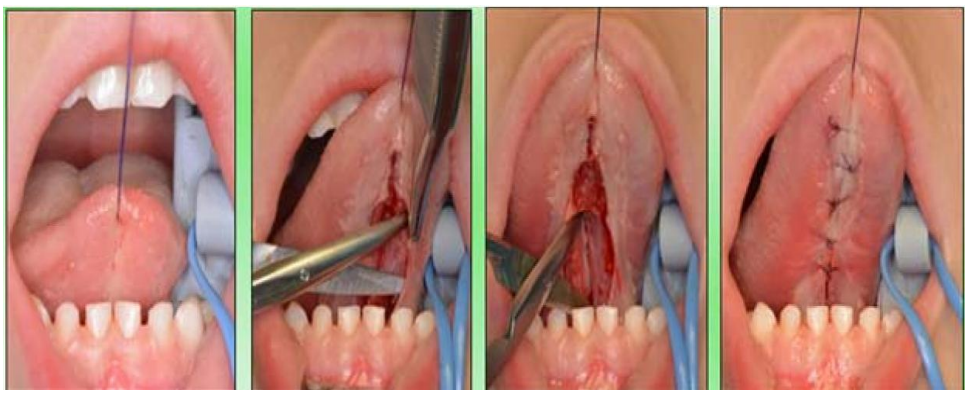
Clinical features

The mother usually reports mild to acute pain while breastfeeding, inadequate milk secretion and incomplete emptying due to an insufficient infant suck [48,50]. Examination findings in the mother of the baby may include nipple infection, ulceration, or bleeding. This may result in early abandonment of breastfeeding due to frustration, anxiety, and feeling of failure.

This hereditary defect is linked mainly to the development of malocclusion, particularly the Class III malocclusion. It is mainly in association with mandibular prognathism and underdevelopment of the maxilla stems [48].

Treatment / Management

Frenotomy is usually and mostly recommended in infants who are diagnosed with ankyloglossia experiencing breastfeeding difficulties after other conservative treatments have failed miserably this can mostly be due to increased uncomfortably. Early release of the lingual frenulum will decrease mainly the possibility that the mother abandons her breastfeeding journey. Notably, there will be limited evidence to support that frenotomy and is associated with positive outcomes in other issues besides breastfeeding [51,53].



The main risks and complications of frenotomy are mostly uncommon but have been described. Rare complications include the bleeding, airway obstruction, damage to the surrounding structures, scarring, and oral aversion. Bleeding is the most common [47].

II. CONCLUSION

The true value of health is felt only when the pathology hits and causes destruction to normal morphology and functions. These are one such pathology that arises due to various aetiologies that results in mild to fatal consequences with respect to morphology and function. Craniofacial abnormalities are a significant part of syndrome and dental professionals have an essential role to play in diagnosis of syndromes[2].

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