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Cleft Lip and Palate Complicated by Hypoxic Ischemic Encephalopathy A Need for Psychosocial Counseling

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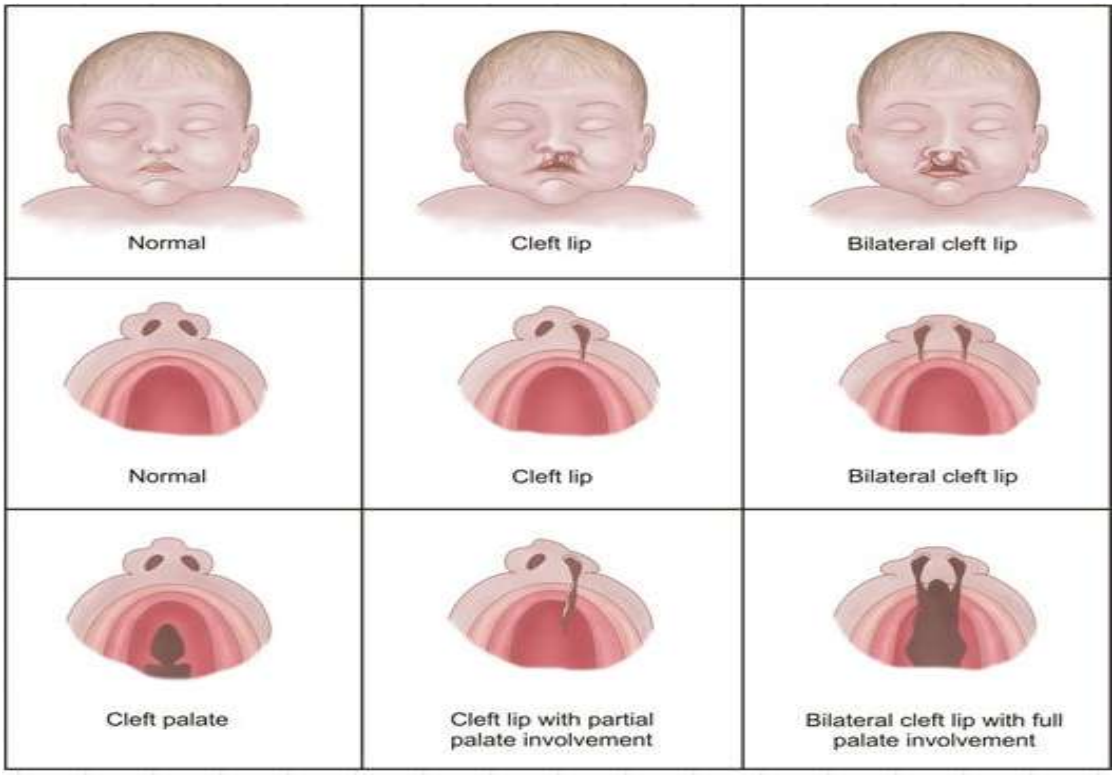
Abstract
Cleft lip and cleft palate are openings or splits in the upper lip, the roof of the mouth (palate) or both. Cleft lip and cleft palate result when facial structures that are developing in an unborn baby don't close completely. Cleft lip and cleft palate are among the most common birth defects. They most commonly occur as isolated birth defects. The main reasons of cleft in infants may be either environmental (such as smoking, alcohol, poor nutrition) or but are also associated with many inherited genetic conditions or syndromes, genetic factors. Having a baby born with a cleft can be upsetting, but cleft lip and cleft palate can be corrected. In most babies, a series of surgeries can restore normal function and achieve a more normal appearance with minimal scarring. A number of specialists involves in treatment of cleft and decides the best treatment plan depending on the site of defect and age of the infant. In this case of cleft lip and cleft palate is rare due to complications that occurred as a result of milk aspiration, which led to a lack of oxygen that resulted in hypoxic ischemic encephalopathy, convulsions, disseminated intravascular coagulopathy, and multisystem organ failures.



Kay words*: Cleft lip, Cleft palate, Encephalopathy, Psychosocial Counseling*

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I. Introduction

Clefts of the lip and/or palate (CLP) are currently the most common craniofacial birth defects that arise as a result of failure of facial embryonic processes to fuse .These congenital growth diseases can be isolated or associated with other anomalies or part of recognized syndromes. The majority of these orofacial clefts are non-

syndromic (1). In general, the types of orofacial clefts are classified into a cleft lip either with or without a cleft palate or a cleft palate only. CLP etiology, which involves both genetic and environmental factors, is highly complex; its molecular basis remains largely unknown. Epidemiological studies on different populations have revealed marked geographical and ethnic differences with the prevalence of disease which is ranging from 1:500 to1:2500 live births Disease has a major impact especially on the affected individual, their families and society that needs long rehabilitation between birth and adulthood. The effects on an individual's speech, hearing, appearance, social integration and psychology can lead to long-lasting adverse outcomes for health and wellbeing. Early referral to a craniofacial clinic is needed to ensure the best outcomes for these children. Most individuals with CLP require the coordinated care of providers (multidisciplinary care) in many fields of medicine and dentistry. Depending on the location and severity of the cleft, subsequent cosmetic operations may be necessary. Treatment of CLP anomalies is still challenging, costly, lengthy and requires years of specialized care (2).

Case presentation

A.W 13 day old male neonate full term born in a hospital to a primary gravida 24y old healthy mother and 26 y old healthy father with positive consanguinity. Admitted to hospital due to milk aspiration. He is a known case with history of admission to our nursery soon after birth due to Clift lip and palate where screening was done for other anomalies birth and abdomen U/S was normal. The family was reassured about the good prognosis and was discharged to home after the family learned how to deal with their baby feeding with special nipple and the need for surgical repair later.

According to the family he was relatively well until few hours before presented after his father fed him with a special nipple , he suddenly develop to blush discoloration of the skin for which the family rushed to KO hospital , where ABC was done , intubated immediately received shock therapy ,D10 , NaHco3, and put under strict management care.

On arrival to our nursery he was in extremely critical general condition, intubated on ambo-bag, completely flaccid, unconscious .He was immediately connected to M.V. and received another shock therapy, D10 and NaHco3.

During examination he developed abnormal movement myoclonic in rather in lower limp and upper limp improve after he was given phenobarbital loading dose.

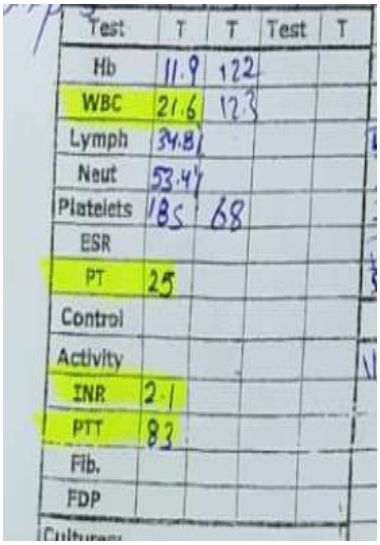
After stabilization. V/S HR 150 bpm, RR 45, temp. 37c, O2 SAT 100%, BP 78/48. Anthropometric Wt. 2.8 Kg, L.t 48 cm, HC 32 cm.

Examination

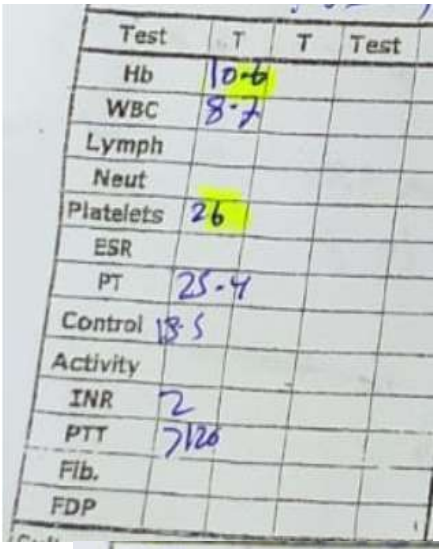
- Chest/ on M.V equal air entry bilateral, no add sound. , no signs of air leak accepted expansion.
- Heart/ normal S1, S2, no murmur hypo profusion, CRT > 35 cc and palpable femoral bilateral but week.
- Abdomen/ soft, lax, no organomegaly, normal genitalia, bloody stool.
- CNS/ un conscious, encephalopathy , flaccid, not responding to painful stimulation , hypotonia , convulsion controlled with phenobarbital . AF open ,flat not pulging
- Pupil/ miosis reactive to light.

Investigation

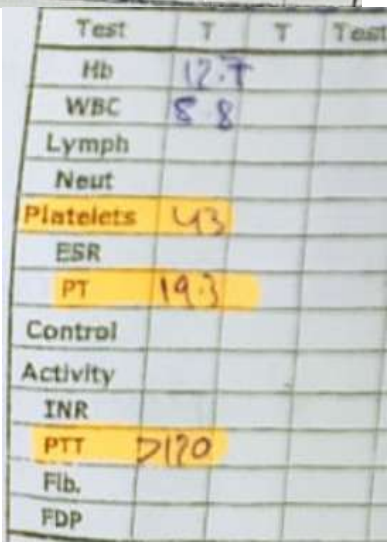
- **CBC, PT, and PTT**



Test	T	T	Test	T
Hb	11.9	122		
WBC	21.6	123		
Lymph	34.8			
Neut	53.4			
Platelets	185	68		
ESR				
PT	25			
Control				
Activity				
INR	2.1			
PTT	83			
Fib.				
FDP				

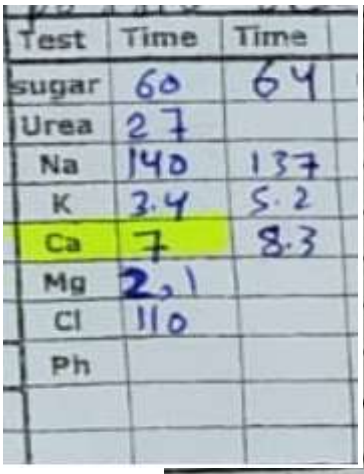


Test	T	T	Test
Hb	10.6		
WBC	8.7		
Lymph			
Neut			
Platelets	26		
ESR			
PT	25.4		
Control	18.5		
Activity			
INR	2		
PTT	>120		
Fib.			
FDP			

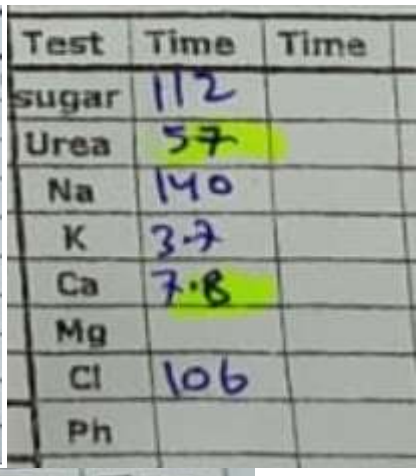


Test	T	T	Test
Hb	12.7		
WBC	5.8		
Lymph			
Neut			
Platelets	43		
ESR			
PT	19.3		
Control			
Activity			
INR			
PTT	>120		
Fib.			
FDP			

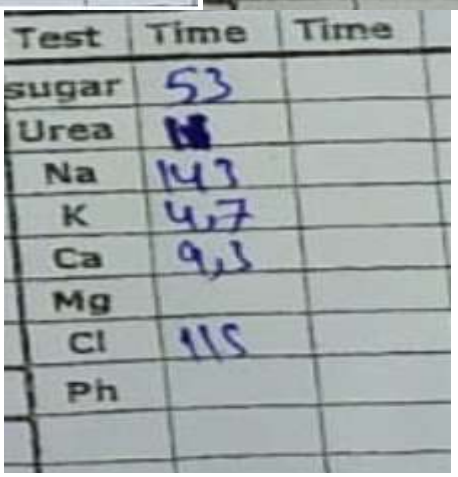
- **Chemistry**



Test	Time	Time
sugar	60	64
Urea	27	
Na	140	137
K	3.4	5.2
Ca	7	8.3
Mg	2,1	
Cl	110	
Ph		

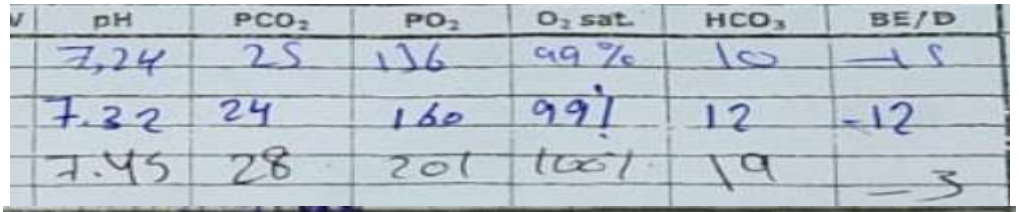


Test	Time	Time
sugar	112	
Urea	57	
Na	140	
K	3.7	
Ca	7.8	
Mg		
Cl	106	
Ph		

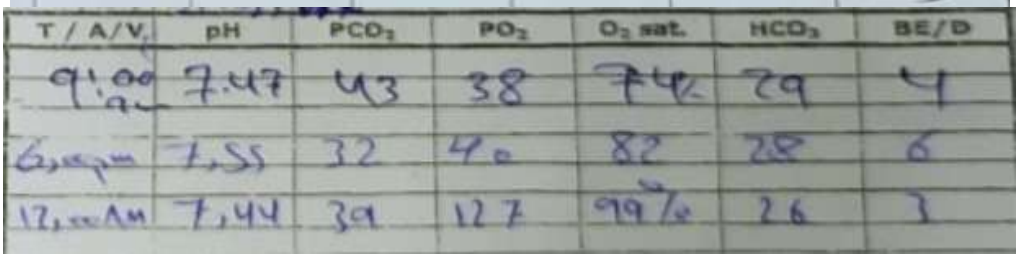


Test	Time	Time
sugar	53	
Urea	[illegible]	
Na	143	
K	4,7	
Ca	9,3	
Mg		
Cl	115	
Ph		

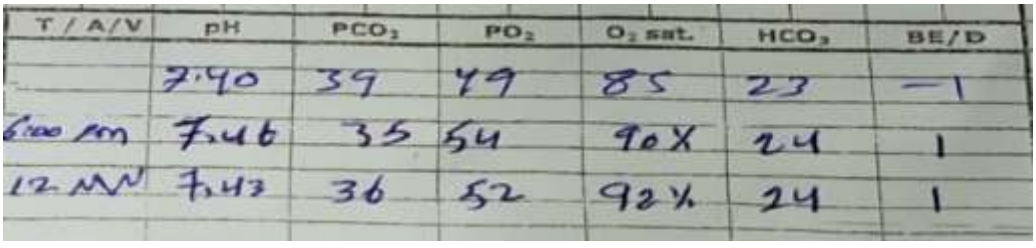
- **ABG**



V	pH	PCO_2	PO_2	O_2 sat.	HCO_3	BE/D
	7,24	25	136	99 %	10	−15
	7.32	24	160	99!	12	-12
	7.45	28	201	100!	19	−3

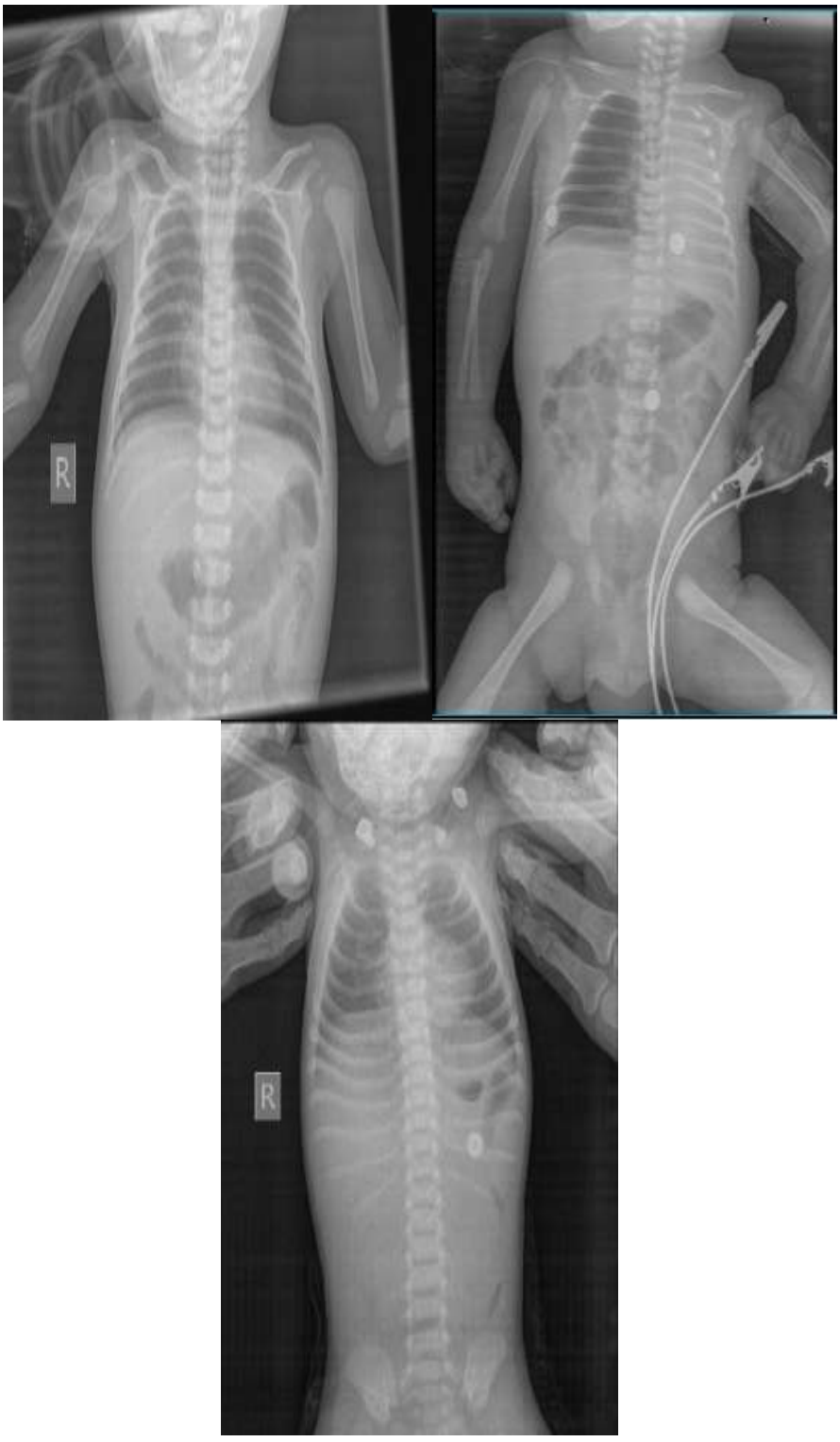


T / A/V	pH	PCO_2	PO_2	O_2 sat.	HCO_3	BE/D
9:00	7.47	43	38	74%	29	4
6,[illegible]	7,55	32	40	82	28	6
12,[illegible]AM	7,44	39	127	99%	26	1



T / A/V	pH	PCO_2	PO_2	O_2 sat.	HCO_3	BE/D
	7.40	39	49	85	23	−1
6[illegible]	7.46	35	54	90X	24	1
12 MN	7.43	36	52	92%	24	1

- **X-Ray**



Neonate was admitted to our nursery for 10 day during which his course was remarkable for MSOF, DIC.
Respiratory frailer for which he was connected to M.V. for 5 days, extubated successfully and was kept on nasal oxygen. Circulatory failure for which he received dopamine and hydrocortisone.
Encephalopathy and convulsion controlled with phenobarbital.
Anemia, Coagulopathy and thrombocytopenia "DIC" received FFP, Vit K, PLT and PRBCs.
Sepsis received Ampicillin, Cefotaxime, Amikacin and Metronidazole for the first 3 days but as there was no proper response to Ampicillin.

And over the last few days of admission his condition improved gradually, he was weaned from oxygen, inotropes stopped and his condition level improved and tolerated feeding by nasogastric tube. He was discharge to home with family instruction and follow up in outpatient clinic (3,4).

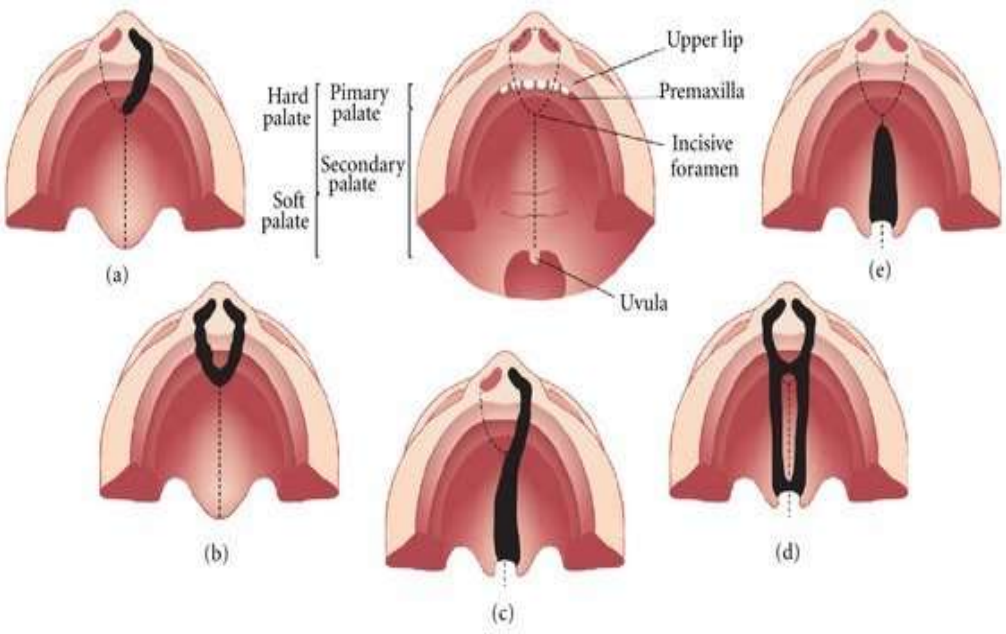
Pathophysiology

Cleft lip:

• Caused by incomplete fusion of the nasomedial or inter maxillary process during the 2nd month of embryonic development.
• Cleft causes structures of mouth and face to develop without the normal restraints of encircling lip muscles.
• May affect external nose, nasal cartilages, nasal septum, and alveolar processes also.
• Usually just beneath the center of one nostril.
• Can occur bilaterally, symmetrically or asymmetrically.
• More complete the cleft lip, the greater the chance that teeth in the line of the cleft will be missing or malformed.
• Complete cleft = entire thickness of the lip.
• Incomplete cleft = only a portion of the lip is involved.

Cleft Palate:

• Often associated with cleft lip, but may occur without it.
• Fissure may affect only the uvula and soft palate (secondary palate, formed ~ 9 weeks), or may extend forward to the nostril and involve the hard palate and the maxillary alveolar ridge (primary palate, formed ~ 4-5 weeks).
• Complete= involves the primary and secondary palate.
• Incomplete= involves the secondary palate only.
• Unilateral = on one side the palatal process of the maxilla is fused with the nasal septum.
• Bilateral= not attached to the nasal septum, and the septum is visible through the cleft.
• Cleft occupies the midline posteriorly and can go as far forward as the alveolar process. Clefts involving the palate only are usually midline.
• Some cases, the vomer and nasal septum are partly or completely undeveloped.
• When these facial bones are involved, the nasal cavity and the oral cavity are open to each other (5,6).



Etiology

The etiology of CLP is complex but it's thought to be involving interactions of numerous genetic, environmental and prenatal factors. Although the exact cause of CLP is largely unknown, current published evidence shows that there are numerous risk factors that are involved in etiology (7).

Researches indicate that the genes a child inherits from their parents occasionally make them more susceptible to developing a CLP Compelling evidences suggest that genetic variants play a substantial role in the development of CLP.

Evidence also shows that the risk of oral clefts among first degree relatives of cases is much higher than that in the general population.

Risk factors such as maternal smoking, maternal age alcohol consumption, gender, medicinal drugs, viral infection and exposure to teratogens during early pregnancy have previously been investigated (8).

Prevalence

The overall prevalence of OFC is estimated to be approximately 1 in 700 live births, accounting for nearly one half of all craniofacial anomalies. As reported by the World Health Organization (WHO), the prevalence at birth of OFC varies worldwide, ranging 3.4–22.9 per 10,000 births for CL/P, and 1.3–25.3 per 10,000 births for CPO. The incidence of CL/P and CPO can vary greatly between studies. The inclusion criteria, case definition, data sources, and selection bias contribute to the varying incidence estimates. Even though there are many different variables regarding the inclusion or exclusion criteria of in studies, the majority report a higher incidence of CL/P compared to CPO(9) .

Prevalence has been found to vary based on ancestry, with the highest incidence rates observed amongst Asian populations (0.82–4.04 per 1000 live births), intermediate rates amongst Caucasians (0.9–2.69 per 1000 live births), and the lowest rates amongst African populations (0.18–1.67 per 1000 live births) . Prevalence has also been found to vary further by subgroup, for example, with one study reporting lower rates of OFC amongst Far East Asians compared to Filipinos (10).

Gender ratio

Prevalence of OFC additionally varies according to gender and cleft pattern. Male predominance has been consistently identified in CLP, with a male/female sex ratio of 1.81 (CI 95%: 1.75–1.86). For CP, the opposite has been shown, with a reported sex ratio of 0.93 (CI 95%: 0.89–0.96) ; however, this may be due in part to sampling bias, as one Danish study could not find a significant predominance of females in individuals with CP after combining both surgically treated and no surgically treated cases(11).

Complications

Children with cleft lip with or without cleft palate face a variety of challenges, depending on the type and severity of the cleft.

- Difficulty feeding: One of the most immediate concerns after birth is feeding. While most babies with cleft lip can breast-feed, a cleft palate may make sucking difficult.
- Ear infections and hearing loss: Babies with cleft palate are especially at risk of developing middle ear fluid and hearing loss.
- Dental problems: If the cleft extends through the upper gum, tooth development may be affected.
- Speech difficulties: Because the palate is used in forming sounds, the development of normal speech can be affected by a cleft palate, Speech may sound too nasal challenges of coping with a medical condition.
- Children with clefts may face social, emotional and behavioral problems due to differences in appearance and the stress of intensive medical care (12).

Treatment and management

A cleft lip may require 1 or 2 surgeries, depending on the extent (complete or incomplete) and width (narrow or wide) of the cleft. The first surgery is usually performed by the time a baby is 3 months old.

Several techniques can improve the outcomes of cleft lip and palate repairs when used appropriately before surgery. They are non-invasive and dramatically change the shape of the baby's lip, nose and mouth:

A lip-taping regimen can narrow the gap in the child's cleft lip.

A nasal elevator is used to help form the correct shape of the baby's nose.

A nasal-alveolar molding (NAM) device may be used to help mold the lip tissues into a more favorable position in preparation for the lip repair.

The first surgery, to close the lip, usually occurs when the baby is between 3 and 6 months old. The second surgery, if necessary, is usually done when the child is 6 months old.

Repair of a cleft palate is performed at 12 months and creates a working palate and reduces the chances that fluid will develop in the middle ears. To prevent fluid buildup in the middle ear, children with cleft palate usually need special tubes placed in the eardrums to aid fluid drainage, and their hearing needs to be checked once a year. This is often done at the time of palate repair.

About 30-40 percent of children with a cleft palate need further surgeries to help improve their speech. Speech is usually assessed between ages 4 and 5. Often a nasopharyngeal scope is performed to check the movement of the palate and throat.

A decision is then made, together with the speech pathologist, if surgery is needed to improve the speech. This surgery is usually performed around age 5.

Children with a cleft involving the gum line may also need a bone graft when they are about 6-10 years old to fill in the upper gum line so that it can support permanent teeth and stabilize the upper jaw. Once the permanent teeth grow in, a child will often need braces to straighten the teeth and a palate expander to widen the palate. Additional surgeries may be performed to improve the appearance of the lip and nose, close openings between the mouth and nose, help breathing, and stabilize and straighten the jaw (13.14).

Psychosocial Counselling needed for this case:

Various psychosocial and cultural factors contribute in the development of psychosocial issues among individuals with any form of facial anomaly in general.

This case is considered one of the cases that require psychological and social intervention in addition to the biological therapeutic intervention; because it is related to the acceptance of the shape of the child and the family. The search for psychological support that helps the family to overcome that state, as there is no absolute cause for the condition and therefore psychological support forms an important basis in the treatment phase. So that the surgical procedures are accepted later without rejection and bear the consequences of the matter.

Researches shown that attractive children are seen by others as brighter, having more positive social behavior and receive more positive treatment than their less attractive counterparts. Self-perception plays a pivotal role in influencing an individual's self-esteem and psychological adjustment affected by cleft lip and palate anomaly. Additionally, parental influence also shapes ones psychosocial perception. The attitudes, expectations and degree of support shown by parents can influence a child's perception of their cleft impairment. Parents of children with clefts may be more tolerant of misbehavior in their child and are more likely to spoil their child by being overprotective. Additionally, peer interaction also plays an important role in maintaining psychosocial limitations. Many children with cleft lip and palate may have a less attractive facial appearance or speech than their peers. A high incidence of teasing over facial appearance is reported among those with cleft lip and palate (15).

Social stigma and cleft disorders

A social stigma is created within an individual when he/she is negatively discriminated by labeling him/her different from normal. An individual's thoughts, feelings and behavior related to their physical appearance makes their body image attitudes. A negative response from outsiders, actual or perceived, may adversely affect self-image. Also physical attractiveness plays an important role in the development and maintenance of self-beliefs. Research indicates that preference for attractive individuals subsequently influences self-esteem, social competence, and future ratings of attractiveness. Moreover, being physically attractive appears to be an advantageous trait regardless of age. Physical attractiveness has shown to play a significant role in social set ups like developing relationships during various stages of life, school, courtships, work etc. Social acceptance often depends on one's physical look. These associations between physical beauty and social acceptability indicate the difficulties for cleft lip and palate affected individuals (16).

Prevention

- Primary prevention: Orofacial clefts appear to have substantial environmental causes; the potential for their occurrence thus seems considerable. The pattern of occurrence of orofacial clefts is different from that of neural tube defects so their causes may also be different (17).

Maternal tobacco use has been consistently associated with a modest o elevation in risk of orofacial clefts but the attributable risk may be of public health importance. Moreover tobacco use is rapidly increasing among women, especially in technologically developing countries, and many women are exposed to passive smoking in the home and workplace.

Maternal alcohol use, well known as a cause of the fetal alcohol syndrome, has also been associated with risk of isolated orofacial clefts in some, but not all, studies. The type and context of alcohol consumption differs considerably across populations and more consistent methods are needed for the assessment of maternal alcohol intake. The possible increased risk of orofacial clefts and other CFA related to the common exposures of smoking and alcohol use during pregnancy is a message that should be incorporated into health promotion programs for women of reproductive age. 103 Global strategies to reduce the health-care burden of craniofacial anomalies (18). Maternal nutritional factors have been associated with the risk for s orofacial clefts in human population studies, although strong evidence of a causal relationship is still lacking. The most promising candidate nutrients include folic acid and pyridoxine (vitamin B-6) and some evidence also exists of possible roles for riboflavin (vitamin B-2) and vitamin A (19) .

II. Conclusion

The management of cleft lip & palate is necessary at correct time. If delayed in the treatment there may be possibility to developed abnormalities and complications.

So to prevent some problems like speech problem facial asymmetry, feeding problem & infection to nasal cavity & unaesthetic appearance. And in the other side psychosocial intervention for family members especially caring mother considered necessary aspect when treating the case.

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References

[1]. Rahimov F, Jugessur A, Murray JC. Genetics of nonsyndromicorofacial clefts. Cleft Palate Craniofac J. 2012;49:73-9

[2]. Cleft lip and cleft palate: a disease with multiple risk factors in a pregnant woman - Scientific Figure on ResearchGate. Available from: https://www.researchgate.net/figure/A-new-born-with-unilateral-cleft-lip-and-palate_fig1_285639568 {accessed 24 Nov, 2020} .

[3]. D. Shereen N. Abed .

[4]. D. Mohamed A. al jesh .

[5]. Hockenberry, M. J. & Wilson, D. (2007). Wong's Nursing Care of Infants and Children, 8:458-465. St Louis: Elsevier.

[6]. Cute, S. (2009, July). Cultural Considerations in Cleft Lip and Palate: Improving Relationships and Health Care Delivery Between Patients and Professionals. Knol, 25. Retrieved February 1, 2010, from http://knol.google.com/k/cultural-considerations-in-cleft-lip-and-palate#

[7]. Pauws, Erwin, and Philip Stanier. "FGF signalling and SUMO modification: new players in the aetiology of cleft lip and/or palate." *TRENDS in Genetics* 23.12 (2007): 631-640.

[8]. Sivertsen, Å., Wilcox, A. J., Skjærven, R., Vindenes, H. A., Åbyholm, F., Harville, E., & Lie, R. T. (2008). Familial risk of oral clefts by morphological type and severity: population based cohort study of first degree relatives. *Bmj*, *336*(7641), 432-434.

[9]. Ahmed, M. K., Bui, A. H., & Taioli, E. (2017). Epidemiology of cleft lip and palate. In *Designing Strategies for Cleft Lip and Palate Care*. IntechOpen.

[10]. AHMED, Mairaj K.; BUI, Anthony H.; TAIOLI, Emanuela. Epidemiology of cleft lip and palate. In: *Designing Strategies for Cleft Lip and Palate Care*. IntechOpen, 2017.

[11]. Ahmed, Mairaj K., Anthony H. Bui, and Emanuela Taioli. "Epidemiology of cleft lip and palate." *Designing Strategies for Cleft Lip and Palate Care*. IntechOpen, 2017.

[12]. arameters for evaluation and treatment of patients with cleft lip/palate or other craniofacial anomalies. American Cleft Palate-Craniofacial Association. http://acpa-cpf.org/team-care/standardscat/parameters-of-care//. Accessed April 3, 2018.

[13]. Centers for Disease Control and Prevention. Facts about Cleft Lip and Cleft Palate. Accessed 1/2/2019.

[14]. American Cleft Palate-Craniofacial Association. Introduction to Cleft & Craniofacial Conditions. Accessed 1/2/2019.

[15]. De Sousa, Avinash, Shibani Devare, and Jyoti Ghanshani. "Psychological issues in cleft lip and cleft palate." *Journal of Indian Association of Pediatric Surgeons* 14.2 (2009): 55.

[16]. Ahmedani B. K. (2011). Mental Health Stigma: Society, Individuals, and the Profession. Journal of social work values and ethics, 8(2), 41–416.

[17]. Oginni, Fadekemi O, and Anthony T Adenekan. "Prevention of oro-facial clefts in developing world." *Annals of maxillofacial surgery* vol. 2,2 (2012): 163-9. doi:10.4103/2231-0746.101346

[18]. Shaw, William. "Global strategies to reduce the health care burden of craniofacial anomalies: report of WHO meetings on international collaborative research on craniofacial anomalies." *The Cleft palate-craniofacial journal* 41.3 (2004): 238-243. 18

[19]. Shaw GM, Carmichael SL, Laurent C, Rasmussen SA. Maternal nutrient intakes and risk of orofacial clefts. Epidemiology. 2006 May;17(3):285-91. doi: 10.1097/01.ede.0000208348.30012.35. PMID: 16570024.