DOI: 10.21276/aimdr.2021.7.1.AT2

Original Article ISSN (O):2395-2822; ISSN (P):2395-2814

Different Types of Cleft Palate with an Embryological Review in Infants of Eastern Odisha- An Observational Study

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Received: October 2020
Accepted: December 2020

ABSTRACT

Background: Developmentally the palatine processes of the maxillae and that of the palatine bones arise as shelves of tissue which grow medially and meet the elements of the opposite side in the midline. But the part of the alveolar process which bears the central incisor teeth is formed by a median down-growth (the medial nasal process). The alveolar process may show either a unilateral cleft or a bilateral cleft due to defective fusion of the medial nasal process with the lateral nasal and palatine processes. Rarely a large midline defect is also seen due to failure of development of the medial nasal process. **Objectives:** To assess the socio demographic determinants of cleft palate patients. To identify the factors that contribute to the etiology of cleft palate. **Methods:** Study design: A cross sectional study was conducted at Dept of Paediatrics in a tertiary care hospital, in Odisha during the year 2018-20. A pre-designed questionnaire, personal interview with mothers was used as study tool. Study Materials: Inclusive Criteria: Patients having Congenital Anomalies of face i.e Cleft palate admitted in the Department of Paediatrics. Exclusive Criteria: Multiple Congenital anomalies of face and other associated system or other Craniofacial abnormalities. **Results:** In type III and type IV variety 16(66.7%) and 6(60%)were females respectively. Out of 86 infants majority of the patients 64(74.4%) did not have any positive family history of cleft palate. 51(59.3%) had no medical history of mother during the 1st trimester of pregnancy. Only 35(40.7%) had history of fever or other associated illness during 1st trimester. 38(44.2%)had history of Drug intake in mother during the 1st trimester of pregnancy. Rest 48(55.8%)had no history of drugs intake during 1sttrimester. **Conclusion**: The risk for recurrence of cleft lip or palate in future pregnancies of parents with one affected child is 2-5 %, and 1-2 % for cleft palate alone. But the risk is 5-6 % for bilateral cleft lip or palate which is very high as compared to 2-3 % for cleft lip without cleft palate.

Keywords: Cleft palate, palatine processes, medial nasal process, lateral nasal and palatine processes.

INTRODUCTION

The palate which intervenes between the nasal fossae and the mouth cavity is extremely important for proper complete swallowing and articulation of the sounds used while speaking.[3] Its functional efficiency is very much dependant upon the ability of the palate to close off completely from the mouth cavity as well as from the oropharynx. This is actually possible when the musculature of the soft palate acting in conjunction with that of the pharynx brings about apposition between the soft palate and the posterior pharyngeal wall According to Wardill closure must take place and must be complete between certain stages of phonation and that even a gap of about 1 mm between the soft palate and the posterior pharyngeal wall will be responsible for defective speech.[4,5]

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Embryological explanation

According to the neural crest developmental process the specialized embryonic cells or neural crest cells migrate to different places at a different pace. When there is failure of migration of the cells or there is a defect in the rate of migration then facial abnormalities or clefts are seen.[6] Developmentally it is seen that the frontonasal process along with the right and left nasomedial processes of the maxilla gives rise to the formation of the upper lip and the premaxilla. For the development of the palate a plate like shelf called as palatal process grows medially from each maxillary process. Then the definitive palate is formed by the fusion of certain processes which are as follows

a) Each palatal process fuses with the posterior margin of the primitive palate.
b) The two palatal processes fuse with each other in the midline and their fusion starts anteriorly and then proceeds backwards.
c) The medial edges of the palatal processes then fuse with the free lower edge of the nasal septum. Thus the two nasal cavities are separated from each other and from the mouth.

At a later stage the mesoderm in the palate undergoes intramembranous ossification to form the hard palate. However the most posterior portion remains as the soft palate because ossification does not extend into that region. The part of the palate which is derived from the frontonasal process forms the premaxilla and this carries the incisor teeth. Closure of the lip occurs during the 5th or 6th week of embryonic life while during the 8th or 9th week of gestation closure of the palate occurs.[7]

When there is failure of fusion of the median down growth with the alveolar process of the maxilla a cleft develops at this line and the lateral incisor lies defectively on either side of the cleft.[8] Thus it is seen that a cleft palate proper is almost always seen as a defect in the midline, due to failure of approximation of the two palatine processes. The alveolar process may show either a unilateral cleft or a bilateral cleft due to defective fusion of the medial nasal process with the lateral nasal and palatine processes.[9] Rarely a large midline defect is also seen due to failure of development of the medial nasal process.

Thus clefts in the palate occur when there is defect in the fusion of any of the components of the palate. Clefts of the palate extending upto the anterior end are associated with harelip, as both the upper lip and the palate are formed by fusion of the maxillary processes with the frontonasal process.[10]

The incisive foramen is considered as the dividing landmark between the anterior and posterior cleft deformities. Those anterior to the incisive foramen include lateral cleft lip, cleft upper jaw and cleft between the primary and secondary palates. Anterior clefts vary in severity from a barely visible defect in the vermilion of the lip to extension into the nose. Such defects are due to a partial or complete lack of fusion of the maxillary prominences with the medial nasal prominence on one side or on both the sides or due to lack of fusion of the lateral palatine processes with the primary palate.[11]

In severe cases the cleft extends to a deeper level, forming a cleft of the upper jaw and the maxilla is split between the lateral incisor and the canine teeth. Frequently such a cleft extends to the incisive foramen. Cleft of the soft palate and cleft uvula usually occur when the defect lies posterior to the incisive foramen.[12] Such posterior clefts vary in severity from a cleavage of the entire secondary palate to cleavage of the uvula only. They occur when there is failure of fusion of the lateral palatine processes with each other and so also with the nasal septum and primary palate.[13]

This is how cleft palate occurs due to lack of fusion of the palatal shelves which may be due to smallness of the shelves or failure of the shelves to elevate. It is thus noted that Inhibition of the fusion process itself very well leads to the palatal defects.

Depending on the defective fusion different types of cleft palate were observed.

Type 1 -- Tripartite palate - Complete nonfusion or complete cleft palate, giving rise to a Y shaped cleft, accompanied by bilateral harelip

Type 2 – Bipartite palate -- Cleft palate with unilateral hare lip: Usually the left maxillary process fuses with the premaxilla but it does not fuse with the right maxillary process. This type of cleft palate is accompanied by unilateral cleftlip.

Type 3 -- Midline cleft palate: This cleft extends into the hard palate.

Type 4 -- Cleft of soft palate

Type 5 -- Bifid uvula

MATERIALS AND METHODS

This study is an observational study carried out over a period of two years, where conventional panoramic photographs were taken of 86 newborns and infants with facial defects who were admitted in Sishu Bhavan (SVVPGIP) of SCB Medical College, Cuttack . The sample consisting of 86 infants (within one year of age) were examined thoroughly and the photographs of the face from different angles were taken with the help of a digital camera 40 mega pixel using the standard technique and then were stored in a laptop. All the photographs analysed in the study were taken of the patients of known age and gender. Family history as well as sibling history which was determined and extracted and information about their habits and addiction ascertained by a predesigned questionnaire after taking their informed consent from their parents was documented.

Thus a detailed prenatal history, terratogenic exposure and a three generation family history was enquired for every case study and was very carefully recorded. The family history included queries regarding occurrences of clefting either of the lip or palate, hypodontia, other birth defects, developmental disabilities or any other known genetic syndromes among the other siblings or in the family belonging to both the maternal and paternal side.

A complete physical examination of every case was carried out by the paediatrician on duty and all the cases were also pursued to identify any kind of dysmorphic features and or any kind of associated birth defects or medical concerns.

RESULTS

Among 86 infants with cleftings 18 were found to have the tripartite type of cleft palate and only five were found to have a small cleft in the uvula only. Twenty nine cases were detected to have type 2 variety with cleft of the hard palate along with unilateral cleft lip while twenty four cases were found to have a midline palatal cleft involving both the soft palate and the hard palate. Ten patients whose parents complained of regurgitation of milk

and associated respiratory tract infection when examined were found to have a cleft of the soft palate only.

Out of total 86 cleft palate patients Type II cleft Palate was present in 29(33.7%), Type III in 24 (27.9%), Type I in 18 (20.9%) followed by Type IV in 10 (11.6%) and Type V variety in 5 (5.9%) infants. Considering the sex distribution Out of 18 patients in Type I more than half i.e. 10 (55.6%)were males, similarly in Type II variety 16 (55.6%) were males. In type III and type IV variety 16 (66.7%) and 6 (60%) were females respectively.

Table 1: Sex Distribution of Patients according to the type of cleft palate.

Gender	Type I	Type II	Type III	Type IV	Type V
Male	10 (55.6%)	16 (55.2%)	8 (33.3%)	4 (40%)	3 (60%)
Female	8 (44.4%)	13 (44.8%)	16 (66.7%)	6 (60%)	2 (40%)
Total	18 (20.9%)	29 (33.7%)	24 (27.9%)	10 (11.6%)	5 (5.9%)

Table 2: Distribution of patients with familial history of cleft palate.

Family history	No. of Infants(N=86) %
Positive	22 (25.6%)
Negative	64 (74.4%)
Total	86 (100%)

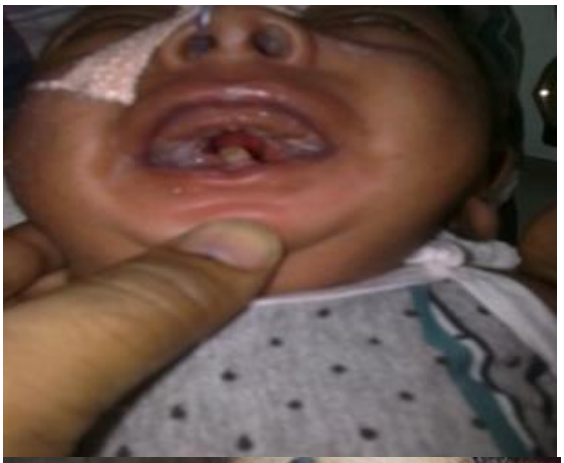
Out of 86 infants majority of the patients 64(74.4%) did not have any positive family history of cleft palate.

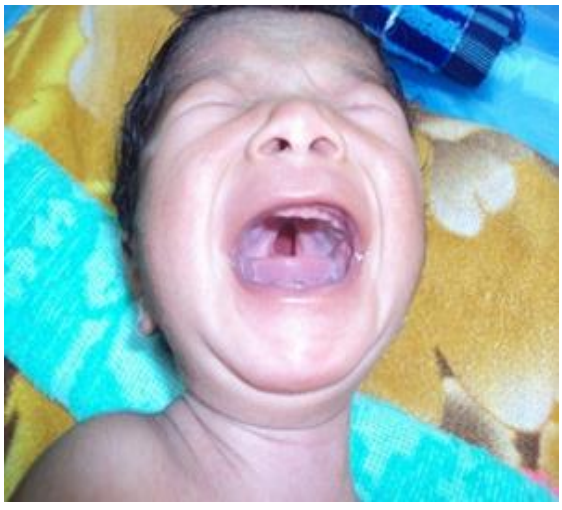
Table 3: Medical history of mother during 1st trimester and type of cleft Palate

Type of cleft Palate	Yes	No	Total
Type I	6 (33.3%)	12 (66.7%)	18 (20.9%)
Type II	11 (37.9%)	18 (62.1%)	29 (33.7%)
Type III	10 (41.7%)	14 (58.3%)	24 (27.9%)
Type IV	6 (60%)	4 (40%)	10 (11.6%)
Type V	2 (40%)	3 (60%)	5 (5.9%)
Total	35 (40.7%)	51 (59.3%)	86 (100%)

Table 4: Intake of Drug by mother during 1st trimester and type of cleft Palate.

Type of cleft Palate	Yes	No	Total
Type I	5 (27.8%)	13 (72.2%)	18 (20.9%)
Type II	12 (48%)	17 (52%)	29 (33.7%)
Type III	13 (54.2%)	7 (45.8%)	24 (27.9%)
Type IV	6 (60%)	4 (40%)	10 (11.6%)
Type V	2 (40%)	3 (60%)	5 (5.9%)
Total	38 (44.2%)	48 (55.8%)	86 (100)





[Table 3] depicts medical history of mother during 1st trimester and type of cleft Palate: Out of 86 patients with cleft palate, 51(59.3%) had no medical history of mother during the 1st trimester of pregnancy. Only 35(40.7%) had history of fever or other associated illness during 1st trimester.

[Table 4] shows Intake of Drug by mother during 1st trimester and type of cleft Palate: Out of 86 patients with cleft palate, 38 (44.2%) had history of Drug intake in mother during the 1st trimester of pregnancy. Rest 48 (55.8%) had no history of drugs intake during 1st trimester.

DISCUSSION

With reference to [Table 1] it is observed that isolated cleft palate was found in greater percentage in females than in males. This is because of the fact that in females palatal shelves fuse approximately one week later than in males. So if any factor causes a delay in the fusion of the palatal shelves it leads to the cleft which is of higher incidence in females than in males. But it is unlikely in case of cleft lip which was found to be more common in case of males even though it was combined with cleft palate.

Since the palate closes from before backward, there is usually a cleft in the soft palate if the hard palate is also affected.

Goyder et al reported that tripartite palate (bilateral alveolar cleft and complete cleft palate) occurred in 22.8 percent of cases, bipartite palate in 31.9 percent, cleft of the soft palate and part of the hard palate in 28.7 percent, of the soft palate alone in 15.7 percent, and rest of the different varieties were seen in 0.9 percent of cases.[16]

According to Davis et al who had studied around 804 cases of cleft palate, it was seen that 74 cases had unilateral clefts in the lip and alveolar process.[17] In 342 patients unilateral clefts of the entire palate was seen while bilateral cleft was seen in 139 patients. Partial posterior clefts of the palate was seen in 240 cases, and submucous clefts of the hard palate in 9 patients.

Genetics or Dysmorphology

Even though majority of the infants with cleft palate did not have an associated genetic syndrome, but still they have been taken into account in order to guide medical decisions and counseling because these conditions may have variable prognostic implications.

However it should be noted that certain physical features which suggest a genetic syndrome may not develop until the later part of infancy, for which continued vigilance is very much needed. So every child who may or not have clefting should be referred for genetic evaluation because even though he or she may have been thought to have nonsyndromic clefting may be later identified to have developmental delay.[4]

Cleft palate can be induced during different stages of embryonic development. There might be a triggering factor during the blastocyst stage (day 6), during gastrulation (day 14), at the early limb bud stage (fifth week) or when the palatal shelves are being formed (7th week).

Susceptibilty to exposure to different types of terratogens varies with the stage of development at the time of exposure. The most sensitive time period for the induction of the cleft type of birth defect is the 3rd to 8th week of gestation. Knowledge of this time period must be taken into account so that the mother can take every kind of precautionary measure to avoid any kind of terratogenic exposure.[5]

However according to the principles of teratology the maternal genome is also important while dealing with drug metabolism, resistance to infection and other biochemical and molecular processes that affect the conceptus.[5]

The various drugs which are known to be a terratogenic element for cleft lip or palate are amphetamines, trimethadione and antiepileptics 5. But manifestations of abnormal development depends on the dose and duration of exposure to a teratogen.

Hyperthermia may also be a causative factor for cleft lip and cleft palate. So also in addition to febrile illness use of hot tubs and saunas can also produce sufficient temperature elevation to cause the birth defects.

There was also a four fold increase in the incidence of cleft lip with or without cleft palate in off springs whose mothers took diazepam during pregnancy.[5]

The risk for recurrence of cleft lip or palate in future pregnancies of parents with one affected child is 2-5 %, and 1-2 % for cleft palate alone. But the risk is 5-6 % for bilateral cleft lip or palate which is very high as compared to 2-3 % for cleft lip without cleft palate. This risk increases if there are additional family members with clefts and it increases to about 8-10 % for nonsyndromic clefting.

CONCLUSION

Condition specific recurrence risks and prenatal testing options should be provided to families of children with syndrome clefting conditions. A discussion regarding the potential preventative role of preconception, prenatal folate supplementation and avoidance of environmental risk factors (tobacco, alcohol, isotretinion) should be considered.

At around 18-20 weeks of gestation a routine ultrasound has become a standard guideline towards obstetric care of the mother and so also improving technologies has led to increased identification of infants with cleft lip. But identification of isolated cleft palate by ultrasonography is still a difficulty and for this reason they are usually identified at

birth. So ideally a genetic evaluation should be considered at several points of gestation.

If the diagnosis of a cleft palate is made in the newborn period then a prenatal and family history should be taken and the infant should be examined for dysmorphic features and genetic counselling should be offered. The risk of recurrence should be vividly discussed with the parents at the time of infancy only with much relevance to an underlying syndrome, health of the infant and of course parental preference. Finally at adolescence risks of recurrence should be revisited with both the patient and family.

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How to cite this article: Panda M, Padhy N, Jena PK. Different Types of Cleft Palate with an Embryological Review in Infants of Eastern Odisha- An Observational Study. Ann. Int. Med. Den. Res. 2021; 7(1):AT04-AT08.

Source of Support: Nil, **Conflict of Interest:** None declared