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A CASE AND REVIEW OF CPO AND CLASSIFICATION OF CLP

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ABSTRACT

Cleft lip and palate is one of the most common congenital defects. CLEFT PALATE with or without cleft lip is seen in about 1/2500 births and mayinvolve only the uvula or extend through the soft and hard palates. Cleft palate only (CPO) is one of the most common malformations presenting at birth. Etiology of CPO is unknown with several genetic and environmental risk factors. Infants with cleft palate may suffer with other complications such as feeding difficulties, eustachian tube dysfunction, middle ear effusions, middle ear infections, hearing loss, speech disorders, dental and orthodontic problems. Feeding plate provides a prosthetic aid for swallowing.

KEYWORDS: cleft palate only, feeding plate, genetic predisposition.

INTRODUCTION

The incidence of isolated cleft palate is low as compared to other orafacial defects, ranging from 1.3 to 25.3 per 10,000 live births. Approximately fifty percent of cases born with cleft palate occur as part of a known genetic syndrome or with another malformation (e.g., congenital heart defects) and the other half occur as solitary defects, referred to often as non-syndromic clefts. It has been found that almost 50% of CPO cases are not associated with any syndrome. (Burg et al. 2016) In non-syndromic patients, subclinical phenotypes such as bifid uvula and submucous cleft palate have been found in family members suggestive of a genetic predilection. (Reiter et al. 2012) In the case mentioned here, bifid uvula was present in the mother. The reported sex ratio of affected males to females by the World Health Organization is 0.93 for non-syndromic CPO. (Mossey and Catilla, 2003) Maternal factors such as smoking, alcohol consumption, intake of drugs during pregnancy, use of corticosteroids, diabetes mellitus, advanced paternal age, insufficient intake of folic acid and obesity seem to influence the occurrence of cleft palate in the offspring. (Burg et al., 2016)

Treatment of orofacial clefts requires multidisciplinary approach and is finally treated by surgical intervention. In the meantime, prosthodontist can play her role by fabricating a feeding plate or obturator. Adequate nutrition is also important for the child to be able to

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undergo the cleft repair surgery, i.e., stable weight gain with no health alterations and the capability to safely receive anesthetics. (Wyszynski, 2002) CPO does not present with compromised esthetics initially but is followed by dental abnormalities, speech disorder, impaired swallowing and growth retardation. (Fisher and Sommerlad, 2011) Feeding difficulties in children with cleft lip and palate (CLP) are frequent and appear at birth due to impairment of sucking and swallowing functions. The use of appropriate feeding methods for the different types of cleft and the period of the child's life is of utmost importance for their full development. (Duarte et al. 2016)

CASE REPORT

A 22 days old male infant was referred to the department of prosthodontics, Dr. Ziauddin Ahmad Dental college, A. M. U., Aligarh by J.N.M.C., A.M.U. with the chief complaint of difficulty in feeding. The mother reported that the infant was not able to suckle milk properly. There was no history of previous treatment or surgery for the defect. The patient's mother had a nasal twang in voice and on examination it was revealed that she also had soft palate cleft (Veau's classification: Type I). [Fig. 1] Intraoral examination of the infant revealed a cleft in the uvula, soft palate and secondary hard palate (Veau's classification: Type II). [Fig. 2] After a detailed examination of the infant and patient's consent, feeding plate was fabricated. [Fig.3]







Fig. 1 – Intra-oral examination of mother.

Fig. 2 – Intra-oral examination of infant.



Fig. 3 – Feeding appliance.

DISCUSSION

The primary challenge in the fabrication of feeding plate for infants is impression making. Many reports have detected intranasal impression material in patients with cleft. (Jones and Drake., 2011), (Reichert et al., 2017) The defect should be filled with gauze on which Vaseline is applied so that it does not stick to mucosa in the defect and easy and complete removal of impression material occurs.

Also this case report proves the genetic predisposition for cleft lip and palate. Maternal history of an incomplete palatal cleft manifested with a complete palatal cleft in the child. Several study designs have been implemented to examine the possible role of interactions of environmental teratogens with genetic mutations on cleft palate formation. CPO has been shown to have a strong genetic component based on its high recurrence rate in families of affected individuals. In population studies, the relative risk of recurrence of CPO among first-degree relatives has been reported to be 56 times greater in Norway and 15 times greater in Denmark than the risk for the general population. (**Sivertsen et al., 2008**)

The varying clinical presentation of cleft lip and palate needs classification system for communication and treatment planning. There are different classifications depending on the condition of cleft, its extension and severity. It helps in proper identification, discussion and treatment planning for the condition. There are mainly two types of classification based on morphological and embryological aspects. Following are some commonly used classification systems.

1. DAVIS AND RITCHIE CLASSIFICATION

The following classification was proposed by Davis and Ritchie in 1922. (**Davis and Ritchie., 1922**) This system broadly categorized the clefts into three groups according to position of cleft in relationto alveolar process.

Group I – Pre alveolar clefts:

- Unilateral cleft lip
- Bilateral cleft lip
- Median cleft lip

Group II - Post alveolar clefts:

- Cleft hard palate alone
- Cleft soft palate alone
- Cleft soft palate and hard palate
- Sub mucous cleft

Group III-Alveolar clefts:

• Unilateral alveolar cleft

2. VEAU CLASSIFICATION



The following classification as demonstrated in Figure 4 was given by Veau in 1931.

Group I (A) - Defects of the soft palate only

Group II (B) - Defects involving the hard palate and soft palate extending not further than the incisive foramen, thus involving the secondary palate alone.

Group III (C) – Complete unilateral cleft, extending from the soft palate to the alveolus, usually involving the lip Group IV (D) - Complete bilateral clefts, resembles Group III but is bilateral. When cleft is bilateral, premaxilla is suspended from the nasal septum.

3. ARTURO SANTIAGO CLASSIFICATION

Santiago A proposed a classification in 1969, in which he used four digits to indicate presence of cleft and its location. (**Santiago. 1969**) Each digit is followed by letter to indicate condition of cleft(complete, incomplete or sub mucous).

Four digits represent the following four structures affected by cleft.

- The first digit refers to the lip.
- The second digit refers to the alveolus.
- The third digit refers to the hard palate.
- The fourth digit refers to the soft palate.

The numbers used as digits represents the condition of cleft.

- 0= No cleft
- 1= Midline cleft
- 2= Cleft on right side
- 3= Cleft on left side
- 4= Bilateral cleft

The letters indicate more specifically the type of cleft.

- A = An incomplete midline cleft
- B = An incomplete cleft of right side
- C = An incomplete cleft of left side
- D = Bilateral incomplete cleft
- E = Sub mucous cleft

Points to consider when using the Arturo Santiago Classification System:

• When a cleft is not described that it is complete or incomplete, it is always assumed as completecleft.

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- When clefts of lip, hard and soft palate are described without giving any information about alveolus, it is assumed that it is completely affected by cleft.
- All cases will be considered midline cleft unless otherwise specified.

4. LAHSAL CLASSIFICATION OF CLEFT LIP AND PALATE (Figure 5)

Kreins O (Hodgkinson et al. 2005) proposed LAHSHAL system for classification of cleft lip and palate patients which was modified on the recommendation of Royal College of Surgeons Britain in 2005 by omitting one "H" from the acronym "LAHSHAL".

LAHSAL system is a diagrammatic classification of cleft lip and palate. According to this classification, mouth is divided into six parts.



Fig. 5 – LAHSAL classification.

- Right lip
- Right alveolus
- Hard palate
- Soft palate (LAHSAL)
- Left alveolus
- Left lip
- The first character is for patient's right lip and last character for patient's left lip.
- LAHSAL code indicates complete cleft with capital letter and an incomplete cleft with small letter.
- No cleft is represented with a dot.

EXAMPLES

1. Bilateral complete cleft lip and palate

The condition is bilateral cleft lip and palate, so there will be no dot and all letters of LAHSAL code will be written. As, cleft of lip and palate is complete, all the letters will be capital, so the patient with bilateral complete cleft lip and palate will be represented as LAHSAL.

2. Left complete cleft lip

A complete cleft lip will be represented with letter "L", as it is left, so, this "L" will be written at theend. Patient with left cleft lip will be represented as L

3. Right incomplete cleft lip and alveolus

Here, the cleft of lip and alveolus is incomplete, so they are represented with small "l" and "a". To represent a cleft on right side, "l" and "a" will be written in start followed by four dots. Thus, patientwith right incomplete cleft lip and alveolus will be represented as la....

4. Incomplete hard palate, complete soft palate defect Cleft of hard palate is incomplete so it will be represented with "h" and cleft of soft palate is complete so it will be represented with "S", this patient will be represented as. hS.

5. ELNASSRY CLASSIFICATION

Elnassry proposed following classification in 2007. He divided cleft lip and palate patients into seven classes. Class I: Unilateral cleft lip

Class II: Unilateral cleft lip and alveolus Class III:

Bilateral cleft lip and alveolus

Class IV: Unilateral complete cleft lip and palateClass V: Bilateral complete cleft lip and palate Class VI: Cleft hard palate

Class VII: Bifid uvula

6. KERNAHAN AND STARK CLASSIFICATION (Kernahan and Stark, 1958)

This classification has an anatomical basis that it is the incisive foramen and not the alveolus that becomes the dividing point between the different groups of deformities. (Figure 6)



First group: clefts lying anterior to the incisive foramen. It includes minor cleft lip to wholepremaxilla.

Second group: clefts lying posterior to the incisive foramen. It includes clefts of the secondarypalate.

Third group: clefts of both the primary and secondary palate.

7. THE STRIPED Y: A SYMBOLIC CLASSIFICATION OF CLEFT LIP AND PALATE (Kernahan. 1971)

Small circle at the junction of limbs of Y - incisive foramen (Figure 7)

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Fig. 7 - The Striped Y: A symbolic classification of Cleft lip and palate.

1 and 4 - Lip

2 and 5 – Alveolus

3 and 6 – Area of hard palate anterior to incisive foramen 7, 8, 9 – Hard and Soft Palate

CONCLUSION

CPO should not be considered as a simple and easy to repair cleft. Even though surgical correction requires a single operation, the resulting dental and speech problems, along with the associated psychological implications are complex and need long term multidisciplinary effort until the late teenage years. Formation of primary and secondary palate involves different mechanisms as proved by many genetic studies. Different genes may be involved in movement of mesenchymal cells. (Salvia and Stanier. 2014) So CPO should be considered, studied and treated as a separate anomaly.

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