Review Article

A REVIEW OF CLASSIFICATION SYSTEMS FOR CLEFTLIP AND PALATE PATIENTS II. EMBRYOLOGICAL CLASSIFICATIONS

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ABSTRACT

The second and final article of this series describes the embryological classifications of cleft lip and palate patients, their usage and deficiencies.

INTRODUCTION

This article has described classification systems based on the embryological features. The structures contributing to the formation of face are the frontonasal prominence, maxillary prominence and mandibular prominence. The frontonasal prominence forms forehead, bridge of nose, medial and lateral nasal prominence. The maxillary prominence forms cheeks and lateral portion of upper lip and the mandibular prominence forms the lower lip. The medial nasal prominence forms the philtrum of the upper lip, crest and tip of nose whereas the lateral nasal prominence forms the alae of nose[1].

During the fifth week of intrauterine life, nasal placodes invaginate to form nasal pits. The prominence on the outer edge of pits is the lateral nasal prominences and those on the inner edge are the medial nasal prominences. During following two weeks, maxillary prominences grow medially, compressing medial nasal prominence towards midline. Hence, upper lip is formed by two medial nasal prominences and two maxillary prominences. Cleft deformities of lips is due to partial or complete lack of fusion of the maxillary prominence with the medial nasal prominence on one or both sides[1].

The two medial nasal prominences merge not only at the surface but also at a deeper level and form a structure known as intermaxillary segment. Intermaxillary segment is responsible for the formation of philtrum of upper lip, jaw component which carries four incisors and palatal component which forms triangular primary palate[1].

During the sixth week of development, palatine shelves appear as two shelves like outgrowths from the maxillary prominence directing obliquely downward on each side of tongue. In the seventh week, palatine shelves ascend to attain horizontal position above the tongue and fuse forming secondary palate. Anteriorly, the shelves fuse with triangular primary palate. The incisive foramen is the midline landmark between primary and secondary palates[2].

The incisive foramen is the dividing landmark between the anterior and posterior cleft deformities in all embryological classifications.

The previous article of this series described the morphological classifications of cleft lip and palate. This article has described some of the more well-known embryological classifications.

1. KERNAHAN STRIPED Y CLASSIFICATION:

Kernahan proposed this classification in 1971[3]. This system provides graphic classification scheme using Y-configuration as shown in figure 1.

- Areas 1 and 4 - Lip (It represents the fusion line between the maxillary prominence and medial nasal prominence at lip level).
- Areas 2 and 5 – Alveolus (It represents the fusion line between the maxillary prominence and intermaxillary segment).

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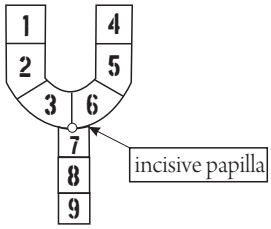


Fig. 1: Kernahan striped Y classification

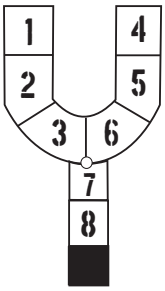
- Areas 3 and 6 – Primary Palate (It represents the fusion line between the primary and secondary palate and lies anterior to the incisive foramen).
- Areas 7 and 8 - Hard palate (It represents the fusion line between palatine shelves of the hard palate posterior to the incisive foramen).
- Area 9 - Soft palate

The small circle represents incisive papilla.

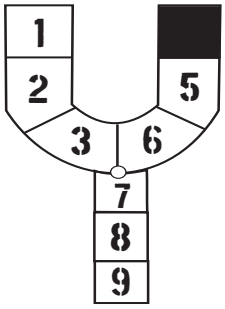
In order to show the cleft, stippling of the appropriate areas is done on the above diagram.

EXAMPLES:

1. Clefts of the soft palate will be represented by the following diagram.



2. Left Cleft lip will be represented by the following diagram.



4. MODIFIED STRIPED-Y ELSAHY CLASSIFICATION:

Elsahy modified Kernahan Striped –Y classification in 1973 by recording more details[4] as shown in Figure 2

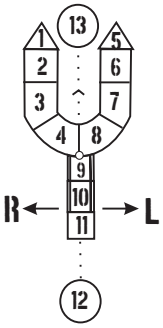


Fig. 2: Modified striped-Y Elshay classification

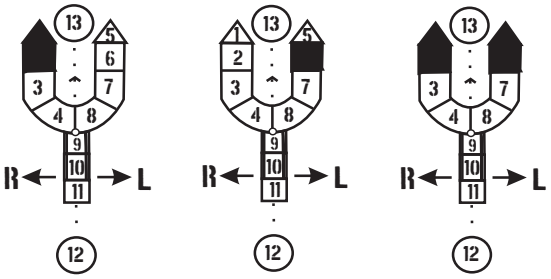
- Triangle 1 represents the fusion line between the maxillary prominence and medial nasal prominence at nostril floor level on right side while triangle 5 represents the fusion line between the maxillary prominence and medial nasal prominence on the left side at the nostril floor level.
- Square 2 represents the fusion line between the maxillary prominence and medial nasal prominence at upper lip level on the right side while square 6 represents the fusion line between the maxillary prominence and medial nasal prominence at upper lip level on the left side.
- Square 3 represents the right alveolus while square 7 represents the left alveolus (fusion line between the maxillary prominence and intermaxillary segment).
- Square 4 and 8 represents the fusion line between primary and secondary hard palate (which lies anterior to the incisive foramen) on the right and left side respectively.
- Squares 9 and 10 represents the fusion line between palatine shelves of hard palate and lies posterior to the incisive foramen.
- Square 11 represents the soft palate.
- Circle 12 represents the posterior pharyngeal wall.
- Circle 13 represents the pre-maxilla.

This classification has following advantages over original striped-Y.

1. It gives information about the degree of cleft lip. For example, if there is unilateral complete cleft lip(involving lip and nostril floor) stippling will include the triangle and square on that side (incase of bilateral cleft lip, the respective areas

will be stippled on both sides). If the cleft lip is incomplete i.e. it does not include nostril floor, then the stippling will include square 2 or 6 only(if bilateral, then squares on both sides).

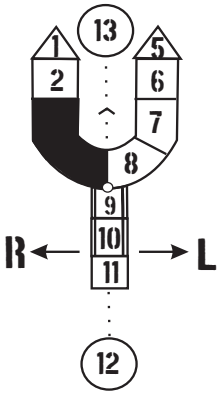
Examples:



Right Complete cleft lip | Left Incomplete Cleft lip | Bilateral complete cleft lip

2. It can indicate the collapse of the alveolar arch. For example, if there is collapse of the lateral maxillary segment, squares 3 and 4 or 7 and 8 (depending upon the side) will be shaded black instead of stippled.

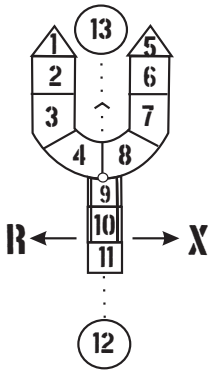
   Example: Collapse of Right maxillary segment will be shown as follows:



3. It represents hard and soft palate separately. Squares 9 and 10 represents hard palate and are bordered by 2 lines (to distinguish it from soft palate) whereas square 11 represents soft palate and is bordered by a single line.

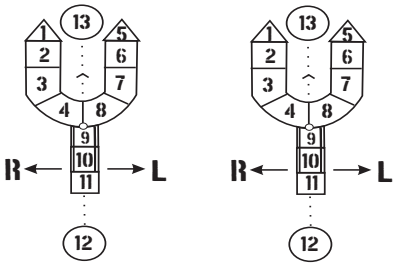
4. It indicates displacement of palatal segments. If there is displacement of the palatal segment towards the right, an **X** can be drawn over the arrow on the right side, if displacement is towards left, then **X** will be drawn on the left.

   Example: Displacement of palatal segments towards the left will be represented by following diagram.



5. It gives information about the state of velopharygeal closure. As, circle 12 represents the posterior pharyngeal wall, if there is a good velopharyngeal closure, a line is drawn on the dots to join circle 12 with square 11. If there is no velopharyngeal closure, no line is drawn. Thus, the length of the line drawn between circle 12 and square11 indicates the degree of velopharyngeal closure.

   Examples:



Complete velopharyngeal closure | Incomplete velopharyngeal closure

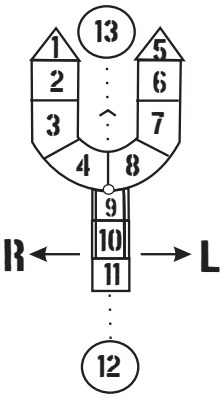
6. It indicates the presence or absence of protruding pre-maxilla and the degree of protrusion. If there is protrusion of pre-maxilla, a line is drawn along the dots in the direction of arrow towards circle number 13. The length of line drawn will indicate the degree of pre-maxillary protrusion.

   Example: The following diagram shows presence and degree of pre-maxillary protrusion.

3. FRIEDMAN'S CLASSIFICATION:

In 1991, Friedman[5] combined Elsahy and millard's classification by using numbers in each segment of the diagram to represent severity of the condition as shown in figure 3.

- Triangles 1 and 6- Nostril arch.
- Triangles 2 and 7- Nasal floor (fusion line be-



tween the maxillary prominence and medial nasal prominence at nostril floor level).

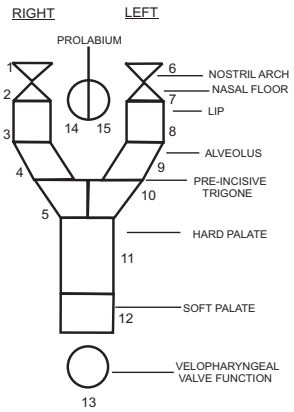


Fig. 3: Friedman's classification

- Square 3 and 8- Lip (fusion line between the maxillary prominence and medial nasal prominence at the upper lip level).
- Squares 4 and 9- Alveolus (fusion line between the maxillary prominence and intermaxillary segment).
- Squares 5 and 10-Primary palate (fusion line between the primary palate and secondary palate and is present anterior to the incisive foramen).
- Square 11- Hard palate (fusion line between the two palatine shelves of hard palate).
- Square 12- Soft palate.
- Circle 13- Velopharyngeal valve.
- Circle 14 and 15- Prolabium.

There are eight steps to record clinical findings and to represent them in this diagram.

STEP I:

This step includes the depiction of the deformity of the nasal arches (triangles 1 and 6)and nasal floors (triangles 2 and 7). In each triangular space, the defect is recorded in the following manner.

0 = No involvement

1 = Cleft microform (e.g; notching)

2 = Mild deformity (i.e; some actual tissue deficiency)

3 = Moderate deformity

4 = Severe deformity

X = Not rated

STEP II:

It includes the depiction of the amount of prolabium or pre-maxillary protrusion on one or both sides (semi circles 14 and 15).

0 = No protrusion

1 = Mild protrusion ($\leq 45^{p}$)

2 = Moderate protrusion ($>45^{p}$,$<90^{p}$)

3 = Severe protrusion($\geq$90p)

X = Not rated

STEP III:

This step depicts the upper lip deformity.

0 = No involvement

1 = Cleft microform

1a = Congenital scar in usual cleft position (subcutaneous cleft)

1b = Notch in the vermillion border

2 = 1/3 cleft lip (cleft in vertical dimension)

3 = 2/3cleft lip (cleft in vertical dimension)

4 = Complete cleft lip

X = Not rated

STEP IV:

It shows the defect in the alveolus.

0 = No involvement

1 = Cleft microform

1a = Submucous cleft

1b = Notch

2 = Partial cleft

3 = Complete cleft

3a = Absence of maxillary arch collapse

3b = Presence of maxillary arch collapse

X = Not rated

STEP V:

It represents the pre-incisive foramena ltrigones.

0 = No involvement

1= Partial cleft

2= Complete cleft

X= Not rated.

STEP VI:

It describes the anatomy of the hard palate

0 = No involvement

1 = Posterior 1/3 cleft

2 = Posterior 2/3 cleft

3 = Complete cleft

X = Not rated.

STEP VII:

It indicates a defect in the soft palate or velum including uvula.

0 = No involvement

1= Cleft microform of uvula

1a = Hypoplasia of musculus uvulae

1b = Septate uvula

1c = Bifid uvula

2 = Submucous cleft of velum

2a = Occult

2b = Overt

3= Posterior 1/3of velum

4= Posterior 2/3 of velum

5 = Complete cleft of velum

X = Not rated.

STEP VIII:

It represents velopharyngeal valve dysfunction

0 = No impairment

1 = Mild impairment

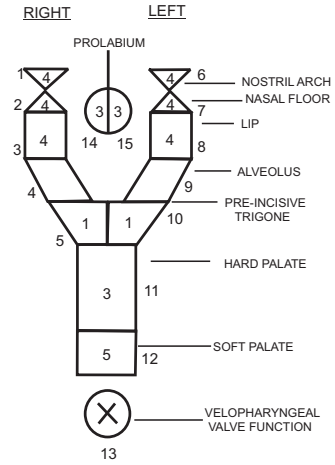
2 = Moderate impairment

3 = Severe impairment

X = Not rated

EXAMPLE:

A 3 months old child has bilateral complete cleft lip and palate. Patient has a severe nasal deformity, marked pre maxillary protrusion, complete cleft lip, alveolus, primary and secondary palate as well as complete cleft in soft palate. Assessment of the velopharyngeal valve dysfunction cannot be made until the child is 2-3 years of age. The child's deformity will be represented by following diagram.



DISCUSSION

Kernahan and Stark[6] in 1958 were for the first time struck by the idea of the importance of classification based on embryology unlike previous classifications based on morphological features. They took the incisive foramen or papilla as the dividing point between the primary and secondary palate. This classification divided clefts into three groups: clefts of primary palate, clefts of secondary palate and clefts of both primary and secondary palate.

Kernahan[3] in 1971 converted his previous embryologically based classification of 1958 from descriptive format into symbolic representation (known as "Striped Y"). It gained more popularity than previous one because of its self explanatory and easy de-

lineation. However, there were some shortcomings, i.e. the degree of cleft lip cannot be properly shown, premaxillary protrusion, alveolar arch collapse and velopharyngeal incompetence cannot be depicted. Elsahy in 1973 modified Kernahan's Striped Y classification by including more details. His classification described degree of cleft lip, collapse of the alveolar arch, displacement of palatal segments, state of velopha-ryngeal closure, absence or presence of protruding premaxilla and degree of pre-maxillary protrusion. Millard[7] in 1977 further modified the Elsahy striped Y by adding inverted triangles atop the upright triangular segments 1 and 5 to stand for the right and left aspects of the nasal arch. This revised classification received considerable fame amongst surgeons.

Until 1991, when attempts were being made all over the world to present a classification system which fully describes the cleft lip and palate condition, no one highlighted the importance of severity of cleft which is a very important factor in predicting prognosis of patient. Friedman et al[5] in 1991 was the first one to describe the severity of the anatomical and functional deformity by further modifying Millard's striped Y and introducing a number in each diagrammatic segment. Though Harkins et al[8] gave importance to severity of cleft but his classification also did not fully explain the condition of cleft. Drawback of Fried-man's classification is that some features need subjective interpretation like deformity of nose, velopharyngeal incompetence so different clinicians may give different severity score to the same patient which may create confusion among clinicians. Schwartz et al[9] in 1993 presented a system which was also based on Kernahan Striped Y and classified clefts using three digits, one for the left side of the Y, another for the right side and a third for the base of Y.

Posadaset al[10] in 2001 proposed an embryological classification in which he incorporated elements related to palate, lip, and nose that described their complexity from a surgical perspective.

Different classifications of cleft lip and palate have been proposed since 1921, but attempt has never been made to gather them under one article. For easy understanding of the reader, this series has classified "Classifications of cleft lip and palate" for the first time and has also described in detail some of the more famous and easily applicable classifications. The first article described morphological classifications in which different cleft conditions were classified on the basis of morphology whereas second article describes classification based on embryology. Most of the embryological classifications use "Striped Y" initially proposed by Kernahan. Striped Y gives true depiction of the fusion lines of the orona-salembryological structures pertaining to cleft lip and palate and makes representation of cleft condition of patient very easy. Upper two limbs of Y represent fusion line of primary palate with secondary palate and lower limb of Y represent fusion line of two palatine shelves of hard palate. These fusion lines are the sites where the cleft can appear.

This article has described embryologic classification systems of cleft lip and palate which are easy to use and at the same time fully explains condition of cleft. Most of the embryologic classifications have graphic representation which makes their use easier.

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