

CLASSIFICATION OF CLEFT LIP AND CLEFT PALATE-A REVIEW

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ABSTRACT

Classification for cleft lip and palate is important for both clinical research and epidemiological investigation. Classification of the cleft lip and cleft palate plays an important role in diagnosis and planning the treatment. It involves the embryological processes, the frontonasal and the right and left processes of the maxilla. The incisive foramen is a basic anatomic landmark for classification of cleft lip and palate. Davis and Ritchie's classification was a fundamental classification, which is followed by symbolic representation of Kernahan and their modifications. Newer approaches have also used mathematical expressions to provide a complete description of the deformity including those which can be used for computerized data analysis. This article is a review of the past and the most recent classifications, a bird's eye view on how improvements/advancements in the field have led to a better understanding and representation of the various types of cleft deformities.

KEY WORDS: : Taxonomy, classification, cleft lip, cleft palate

INTRODUCTION

Taxonomies of cleft lip and palate deformities have seen a sea of changes, each with a different basis for classification, ranging from anatomic and embryologic considerations to the complexity of the deformity. Cleft deformities exhibit variations that may bear on surgical procedures and dental management. A fundamental classification was put forth by Davis and Ritchie¹ in 1922 which was used for years, despite its short comings. The Kernahan and Stark's² classification and diagram is one of the most used around the world. Newer approaches have also used mathematical expressions to provide a complete description of the deformity including those which can be used for computerized data analysis. This article is a review of the past and the most recent classifications, a bird's eye view on how improvements/advancements in the field have led to a better understanding and representation of the various types of cleft deformities.

First person to classify malformations of the face was **Forster³ in 1861**, a Pathologist from Wurzburg (Germany) as shown in **Fig.1**. **Davis and Ritchie¹** in 1922 classified the congenital clefts into three groups according to the position of the cleft in relation to the alveolar process. Group I: Pre-alveolar clefts, unilateral, median, or bilateral; Group II: Post-alveolar clefts involving the soft palate only, the soft and hard palates, or a submucous cleft; Group III: Alveolar clefts, unilateral, bilateral, or median. Their classification had many shortcomings such as, insufficient descriptions of cleft lip, cleft of the primary palate with intact secondary palate and presence or absence of alveolar involvement, and the incisive foramen.

Veau⁴ in 1931 put forth the classification of Cleft lip and cleft palate which was divided into four types as shown in the **Fig.2**

Type-1: Cleft of the soft palate only; **Type-2:** Cleft of the hard and soft palate extending no further than the incisive foramen, thus involving the secondary palate alone; **Type-3:** Complete unilateral cleft, extending from the uvula to the incisive foramen in the midline, then deviating to one side and usually extending through the alveolus at the position of the future lateral incisor tooth; **Type-4:** Complete bilateral cleft, resembling type 3 with two clefts extending forward from the incisive foramen through the alveolus. When both clefts involve the alveolus, the small anterior element of the palate, commonly referred to as the premaxilla, remains suspended from the nasal septum.

In 1942 **Fogh Anderson⁵** gave a very similar classification based on embryological development, which is as follows: **Group 1** – clefts of the lip- unilateral or bilateral; **Group 2** – clefts of the lip and cleft palate (single or double); **Group 3** – clefts of the lip and palate upto the incisive foramina.

In 1958 **Kernahan and Stark²** recognized the need for a classification based on embryology rather than morphology. Primary palate comprised of premaxilla, anterior septum, and lip. The roof of the mouth - from the incisive foramen or its vestige, the incisive papilla, to the uvula - is termed the secondary palate. The incisive foramen is the dividing line between the primary and secondary palates. Their classification was as follows:

Clefts of primary palate: Unilateral, Bilateral and median
Clefts of Secondary palate: Unilateral, Bilateral and median

Clefts of primary and secondary palate: Unilateral, Bilateral and median

To this classification must be added the cleft of the mesoderm of the palate, or submucous cleft, which may be camouflaged unless the uvula is cleft.

Based on embryological principles used by Kernahan and Stark² **Harkins and associates**⁶ (1962), presented a classification of facial clefts. A modified version is as follows:

I. Cleft of Primary Palate

A. Cleft Lip

- (1) Unilateral: right, left (a) Extent: one-third, two-thirds, complete
- (2) Bilateral: (a) Extent: one-third, two-thirds, complete
- (3) Median (a) Extent: one-third, two-thirds, complete
- (4) Prolabium: small, medium, large
- (5) Congenital scar: right, left, median (a) Extent: one-third, two-thirds, complete

B. Cleft of Alveolar Process

- (1) Unilateral: right, left (a) Extent: one-third, two-thirds, complete
- (2) Bilateral: (a) Extent: one-third, two-thirds, complete
- (3) Median (a) Extent: one-third, two-thirds, complete
- (4) Submucous: right, left, median
- (5) Absent incisor tooth

2. Cleft of Palate

A. Soft Palate

- (1) Posteroanterior: one-third, two-thirds, complete
- (2) Width - maximum (mm)
- (3) Palatal shortness: none, slight, moderate, marked
- (4) Submucous cleft (a) Extent: one-third, two-thirds, complete

B. Hard Palate

- (1) Posteroanterior (a) Extent: one-third, two-thirds, complete
- (2) Width - maximum (mm)
- (3) Vomer attachment: right, left, absent
- (4) Submucous cleft (a) Extent: one-third, two-thirds, complete

3. Mandibular Process Clefts

- A. Lip (a) Extent: one-third, two-thirds, complete
- B. Mandible (a) Extent: one-third, two-thirds, complete
- C. Lip Pits: Congenital lip sinuses

4. **Naso-ocular:** Extending from the narial region toward the medial canthal region.

5. **Oro-ocular:** Extending from the angle of the mouth toward the palpebral fissure.

6. **Oro-aural:** Extending from the angle of the mouth toward the auricle.

In 1964 **Pfeiffer** introduced **symbolic representation of cleft lip and cleft palate**^{7, 8}, at the 2nd International Symposia on Cleft Lip and Palate in Hamburg as shown in the **Fig.3**. It is a pentagon that consists of a

vertical block of three pairs of rectangles representing lip, alveolus, and hard palate standing on top of a triangle representing the soft palate. In 1987, Pfeifer introduced another diagram that enables one not only to represent the cleft but also the surroundings of the cleft malformation. Both diagrams are easy to use, but they did not consider the malformed nose and Vomer.

In 1971 **Kernahan**⁹ further modified this classification into a striped Y symbolic classification (**Fig. 4**). He has represented the most severe and extensive form of cleft lip with cleft palate deformity as a 'Y'. The incisive foramen can be represented symbolically by a small circle with the dividing pointing between the primary and secondary palates. Each right and left limb is divided into three portions representing respectively the lip, alveolus and area between alveolus and incisive foramen. The stem of the Y is similarly divided into three portions representing hard palate (7, 8) and soft palate (9). Each individual can be diagrammatically represented by stippling appropriate areas of clefting. In submucous cleft of palate the appropriate section is cross hatched, Simonart's band can be represented by cross hatching the anterior portion of the limb of the Y. By assigning numbers to the striped Y segments, classification and retrieval of information can be achieved with ease. Shortcomings of the Kernahan Striped Y system are as follows:

1. The degree of cleft is ambiguous
2. Premaxillary protrusion and alveolar arch collapse cannot be depicted.
3. The palate is not divided into its hard and soft portions for differential description in partial and complete clefts.
4. Function is not illustrated along with structure, so there is no indication of velopharyngeal incompetence.
5. The diagram lacks labeling for patient name, date and stage in the course of the treatment.
6. Inadequate detail for recording cleft lips, especially asymmetric deformities in bilateral cleft lip;
7. Inadequate detail for assessment of palatal deformities associated with speech results and rates of fistula formation
8. Potentially misread data that was hard to analyze by computer.

The classification was modified later by other investigators, **Elsahy**¹⁰, **Millard**^{11, 12}, **Friedman et al**^{13, 14} and **Smith et al**¹⁵ in 1998. The description of the cleft deformities became more detailed. To overcome the shortcomings of Kernahan and to permit the recording of further details **Elsahy**¹⁰ (1973) modified **Kernahan Striped Y classification** in the following ways:

1. New triangles 1 and 5 atop the arms of the Y represent the right and left nostrils floors respectively
2. Circle 13 between the arms of the Y represents the premaxilla.

3.

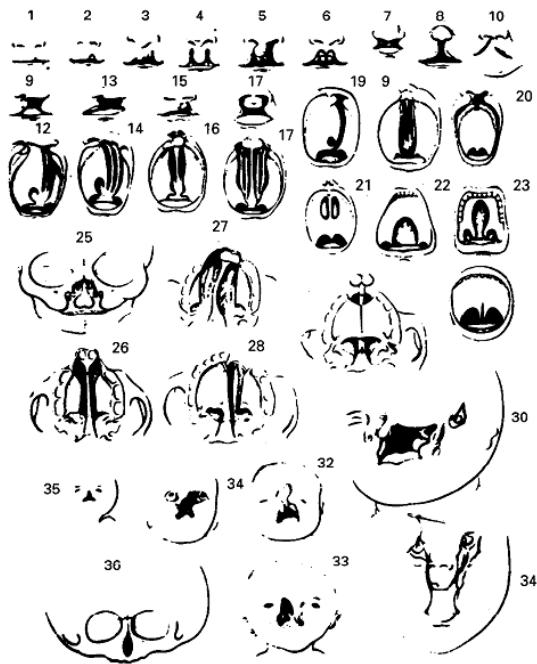


Fig. 1 .Table XXV of Forster's publication
Koch et al., British Journal of Oral and Maxillofacial Surgery (1995) 33, 51-58.

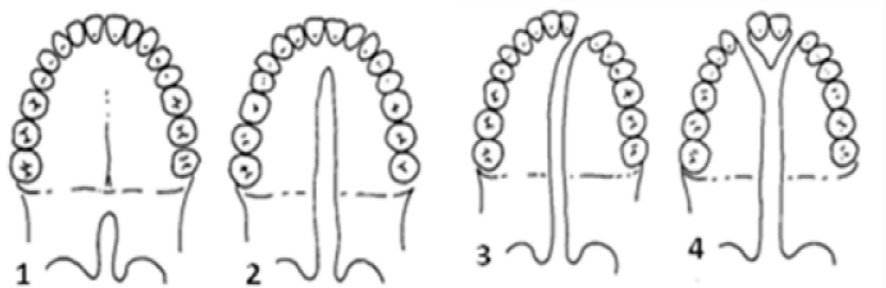


Fig.2. Veau's Classification of cleft lip and palate
Whitaker et al., Cleft Palate Journal, July 1981, Vol.18 No.3, 161-76.

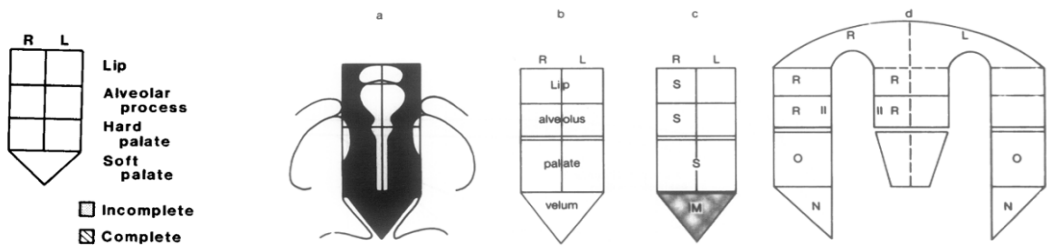


Figure: 3 Symbolic representation of Pfeiffer 1966 and 1987
British Journal of Oral and Maxillofacial Surgery (1995) 33, 51-58.

4. Squares 2 and 6 represent the right and left aspects of the lip, respectively.
5. Squares 3 and 7 represent the right and left alveoli, respectively
6. Squares 4 and 8 represent the prepalate (i.e. that portion of the premaxilla immediately anterior to the incisive foramen) on the right and left sides, respectively.
7. Squares 9 and 10 represent the hard palate proper (i.e. posterior to the incisive foramen) with both right and left sides respectively.
8. Square 11 represents the Velum, both right and left sides.
9. Circle 12 below the stem of the Y represents the posterior pharyngeal wall.

The numbering of the segments in the striped Y and addition of the triangles and circles as described above are shown in **Fig. 5**. Elsayhy gave further instructions for elaboration of his modified striped Y as follows: Protrusion of maxilla can be shown by extending a line from circle 13, by which the length represents its degree. Notching of the vermillion border or alveolar ridge can be indicated by a narrow band of stippling in the lower portion of segments 2/6 or the upper portion of 3/7 respectively. Maxillary segment collapse can be depicted by shading or stippling segments $\frac{3}{4}$ or $\frac{7}{8}$ for right and left sides respectively. Displacement of palatal segments in complete cleft palate can be shown either by drawing double vertical lines on the sides of segments 9 and 10 with right and left arrows to indicate the direction of deflection or by drawing an X over the appropriate right and /or left arrow on the diagram. Submucous clefting of the palate can be depicted by cross hatching. The competence of velopharyngeal closure can be denoted by drawing a line between square 11 and circle 12, the length of which represents closure adequacy from no line (= no closure) to full length connection(=complete closure)

This classification has the following advantages over the original Striped Y:

1. It gives information about the degree of cleft lip.
2. It indicates the presence or absence of collapse of the alveolar arch
3. It describes the state of the hard and soft palate as a separate identity.
4. It describes the position of the palatal segments in complete cleft palate.
5. It indicates the presence or absence of velopharyngeal closure, thus giving some idea of the patient's speech.
6. It indicates the absence or presence of protruding maxilla and the degree of protrusion.
7. It facilitates comparison between different patients and different stages in the same patient (preoperative and post operative)
- 8.

Millard^{11,12} (1977) endorsed Elsayhy's revision of Kernahan's striped Y classification. He further modified it by adding inverted triangles atop the upright triangular segments 1 and 5 to stand for the right and left aspects of the nasal labial respectively. In his symbolic representation

(**Fig.6**), horizontal lines in these nose segments, of density proportionate to the degree of nasal deformity, can be used to mark it. Horizontal lines can also be employed to show submucosal clefts. Stippling depicts over clefts.

In 1979, the embryological classification was integrated into the International Classification of Diseases (ICD) by the **World Health Organization**¹⁶ in 1979. The sequence though, was not absolutely correct, it is as follows: 749.0 cleft palate; 749.1 cleft lip; 749.2 cleft lip and palate. In Chapter XVII of **WHO ICD Version 2007**¹⁷, discusses about the congenital malformations, deformations and chromosomal abnormalities (Q00-Q99) and Cleft lip and cleft palate (Q35-Q37). Q 35 Cleft palate includes fissure of palate, Palatoschisis and excludes cleft palate with cleft lip. Q35.1- cleft palate, Q35.3- cleft soft palate, Q35.5- cleft hard palate with soft palate, Q35.7- cleft Uvula, Q35.9- cleft palate unspecified. Q 36 Cleft lip includes Cheiloschisis, congenital fissure of lip, hare lip, labium leporinum and excludes cleft lip with cleft palate. Q 36.0 Cleft lip, bilateral, Q 36.0 Cleft lip, bilateral, Q 36.1 Cleft lip, median, Q 36.0 Cleft lip, unilateral. Q37 includes cleft palate with cleft lip. Q 37.0 Cleft hard palate with bilateral cleft lip, Q 37.1 Cleft hard palate with unilateral cleft lip, Q 37.2 Cleft soft palate with bilateral cleft lip, Q 37.3 Cleft soft palate with unilateral cleft lip, Q 37.4 Cleft hard and soft palate with bilateral cleft lip, Q 37.5 Cleft hard and soft palate with unilateral cleft lip, Q 37.8 Unspecified cleft palate with bilateral cleft lip and Q 37.9 Unspecified cleft palate with unilateral cleft lip.

In 1991 **Friedman et al**^{13, 14} proposed the modification which combines the graphic and striped schemes of Elsayhy and Millard; further it incorporates various cleft microforms and assigns severity scores to the anatomic and functional deformities. Instead of shading the blocks in the diagram to indicate the severity of the deformity a number is placed in each diagrammatic segment to represent, as shown in the **Fig.7**

Spina¹⁸ in 1974 proposed a modification of classification presented by the nomenclature committee of the American Cleft Palate association. The reference point for the proposed classification is the incisive foramen.

Group I: Pre-incisive foramen clefts (clefts lying anterior to the incisive foramen), Clefts of the lip with or without an alveolar cleft: A. Unilateral B. Bilateral C. Median

Group II: Trans-incisive foramen clefts (clefts of the lip, alveolus, and palate).A. Unilateral. B. Bilateral.

Group III: Post-incisive foramen clefts and Group I: Rare facial clefts.

Group IV: Rare Facial clefts

A completely new recording-system for the diagnosis of cleft lip and palate malformations is the **LAHSHAL** system that **Kriens**¹⁹ introduced in Bremen in 1985. He projects the first letter of the English terms for Lip, Alveolus, Hard, and Soft Palate in one

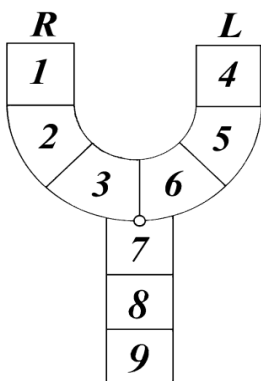


Fig. 4. The Kernahan striped Y classification of cleft lip and palate.
1 - Right lip; 2 - right alveolus; 3 - right premaxilla; 4- left lip;
5 -left alveolus; 6 -left premaxilla; 7 -hard palate; 8 - soft palate; 9 -submucous cleft.
Elsahy NI. Cleft Palate J. 1973;10:247–250

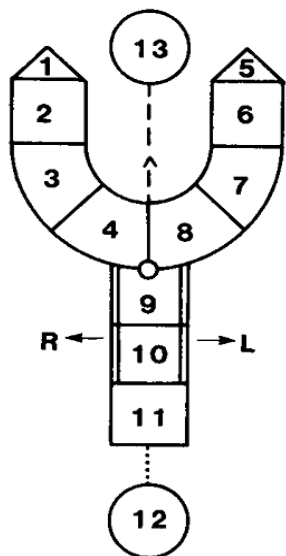


Fig.5. Symbolic representation of Elsahy
Elsahy NI. Cleft Palate J. 1973; 10:247–250

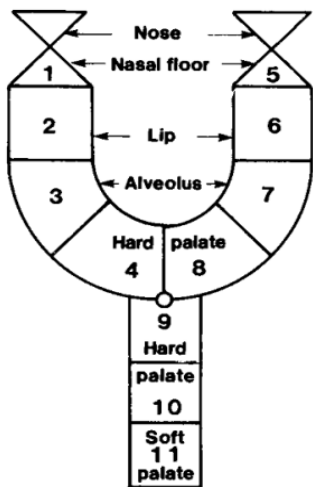


Fig.6. Symbolic representation of Millard
Cleft Palate J 1991; 28: 252.

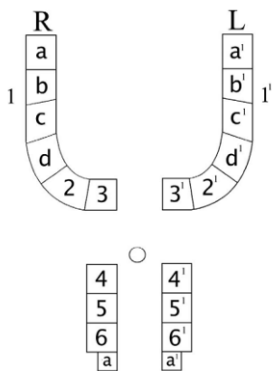


Fig. 8. The Smith et al. (1998) modification of Kernahan's striped Y classification.
1 complete cleft; a through d incomplete cleft from minor to lips with Simonart's band;
2 alveolus; 3 primary palate; 4 cleft up to the palatine process of the maxillary bone;
5 cleft up to the palatine process of the palatine bone; 6 cleft soft palate; a submucous cleft.
Smith AWPlast Reconstr Surg. 1998;102: 1842–1847.

Table 1. A Concise Description of the LAPAL

Side anatomic component	Right		Middle	Left	
	Lip	Alveolus and Primary palate	Palate	Alveolus and Primary palate	Lip
Complete	4	4	4	4	4
Larger than half	3	3	3	3	3
Smaller than half	2	2	2	2	2
Subcutaneous or Submucous	1	1	1	1	1
Intact	0	0	0	0	0

System for Classification of Cleft Lip and Palate
Qiang Liu et al., Craniofacial Journal, September 2007, Vol.44 No.5, 465-68.

Table-2 Modification of Tessier’s cleft classification system

- A. Basic Considerations
1. The point of reference is the orbit with the clefts found in two different hemispheres.

a. Those of the lower lid are classified as facial clefts

b. Those of the upper lid are classified as cranial clefts

c. Combined or craniofacial clefts may occur

2. This system describes both the surface and underlying bony anatomy.

3. The extent of involvement of soft and bony tissue is variable.
- B. The Classification
- CENTRIC

Facial Clefts

Corresponding Cranial Extension of Facial Clefts

No. 0

No. 1

No. 2

No. 3

No. 14

No. 13

No. 12

No. 11

ACENTRIC

Facial Clefts

Cranial Clefts

No. 4

No. 5

No. 6

No. 7

No. 8

No. 10

No. 9

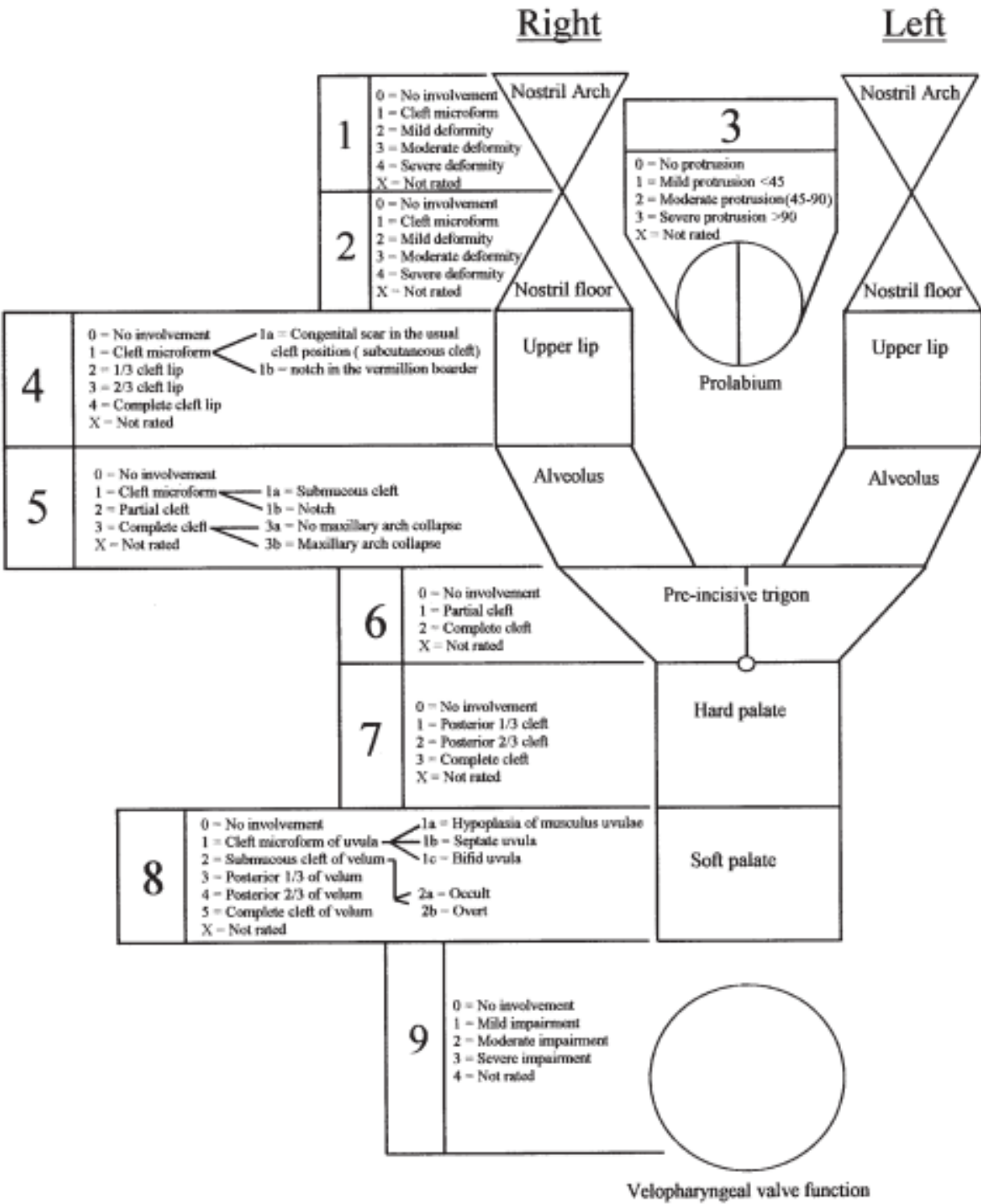
David, J. David; Moore, M.H.; Cooter, R.D.; Cleft Palate Journal, July 1989, Vol. 26, No. 3, (163-185)

Table-3 Transverse view of Koch’s prearranged graphic.

Extent		Shape	
Grade 1	microform	submucous	1
Grade 2	subtotal	partly open/partly submucous	2
Grade 3	total	open	3
I	not affected region		

Koch et al British Journal of Oral and Maxillofacial Surgery (1995) 33, 51-58.

Fig.7 . Data collection sheet and Symbolic representation of Friedman



Davison et al., British Journal of Plastic Surgery, 1998, 51, 281-284

line. A bilateral total cleft of Lip, Alveolus, Hard and Soft Palate is recorded like this: 'LAHSHAL' and a left cleft of lip and alveolus is recorded as '...AL' consequently reading like a roentgenograph. Total clefts are documented in capitals while for subtotal ones small letters were used.

The main disadvantage of the LAHSHAL system is the inflexibility to describe a complex cleft malformation. So, it cannot tell a submucous cleft from a microform. And it is impossible to record a cleft region that is partly submucous and partly open.

Smith et al.¹⁵ (1998) modified the Kernahan Y classification further, in an attempt to make up for the shortcomings. The description of the cleft deformities became more detailed (Fig. 8). Incomplete cleft lip was denoted as letters "a" to "d" for minor defects to lips with Simonart's band. A similarly detailed description also was used to describe a secondary palatal deformity by subdividing it into three segments: palatine process of the maxillary bone, the palatine process of the palatine bone, and the soft palate. The letter "a" denotes a sub-mucous cleft. In addition, there is an indication of the cleft side of the secondary palate based on its relationship to the Vomer. The Smith et al. (1998) modification is more comprehensive than the Kernahan Y classification.

However, due to simultaneous input of numbers and the lettering system used for sub grouping, it is cumbersome to gather data with the systems currently in use. On the one hand, if the numerical values of the Kernahan classification were introduced into a computerized system, as many as nine digits would be required to identify a complete bilateral cleft. The Smith et al. (1998) modification adds details to the Y classification and can describe any kind of cleft deformity. At the same time, this modification adds complexity; recording symbols are mixed with numbers, alphabets, primes, virgules, and even commas. The recording symbols are difficult to use for computerized data analysis.

Using the Kernahan concept with modification, Schwartz et al.²⁰(1993) developed a three-digit numerical system **RPL system** to record the location and number of anatomic components involved in cleft deformities. The right limb of the Kernahan Y classification (1, 2, and 3) is represented by the first digit of this recording system (R). The base of the Y (7, 8, and 9) is represented by the second digit (P), and the left limb (4, 5, and 6) is identified by the third digit (L). Each digit is represented by the numerals 1 to 3, consistent with the anatomic components involved in an anteroposterior direction. Any of the 63 cleft possibilities in the Kernahan classification can be represented by three digits only, allowing immediate identification and computerized data analysis. However, the RPL system is too simple to describe the incomplete and asymmetry of cleft deformities.

The LAPAL system²¹ 2007 consists of **only five Arabic numerals** that describe accurate anatomic components and the extent of any cleft. Numerals are ordered from the right side to the left side, corresponding to what one sees when facing a patient. One numeral is used for the palate posterior to the incisive foramen for the following reasons: (1) Clefts in the posterior hard palate and soft palate are almost in the midline; (2) A bilateral cleft palate is not attached to the nasal septum; and (3) The soft palate has no relationship to the Vomer, although a unilateral cleft palate is fused with the nasal septum on one side. The extent of cleft deformities (i.e., intact to complete cleft) is represented by Arabic numerals 0 to 4 in order to provide more detailed information, even though some minor clefts such as a minor degree of cleft alveolus, do not have a great bearing on management. This procedure is consistent with clinical appearances and helps explain the system. The simplicity and precision of the LAPAL system means it is understood easily and can be used for computerized data analysis. The LAPAL system has universal application for clinical research and epidemiological investigation. Labelling according to LAPAL system is presented in **Table-1**.

Following are some examples of LAPAL system:

Example-1: A complete cleft lip and palate on the left side would be recorded as 00444

Example-2: A bilateral complete cleft lip with complete cleft alveolus and palate on the left side and cleft alveolus on the right side would be recorded as 43444;

Example-3: A cleft soft palate and submucous cleft would be recorded as 00200;

Koch and Koch²² in 1995 proposed a new extended classification, **LAHSN** of cleft deformities. In addition to the lip, alveolus, hard palate, soft palate, they also considered the Vomer and the micro forms in three dimensions. The anatomical regions-lip, alveolus, hard and soft palate, and nose (LAHSN) can be affected single, or they can be affected in all combinations with each other. The severity of all single and combined malformations of LAHSN depends on its extent in sagittal, transverse and vertical directions, (**Fig.9, Fig.10 and Fig.11**) and it depends on whether they are submucous or open forms. For a better estimation of the severity, and for a description of the real extent of a cleft, we think, it is necessary to have a gradation for each cleft region. A classification considering this should have the same gradation for each region and be applicable to all the various types of clefts. It must satisfy the clinical demands, be reproducible, and be simple. Since the severity of a cleft malformation depends on its extent in transverse, vertical and sagittal direction and its shape-whether it is an open or submucous form, this has to be considered when a cleft diagnosis is going to be recorded (**Table-2**).

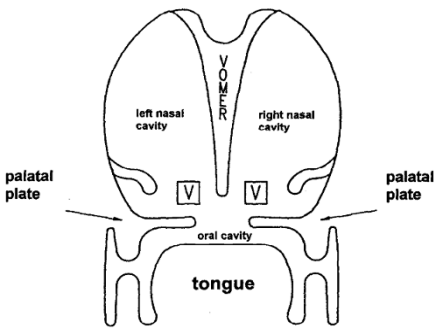


Fig.9 : Frontal view of Koch's prearranged graphic.

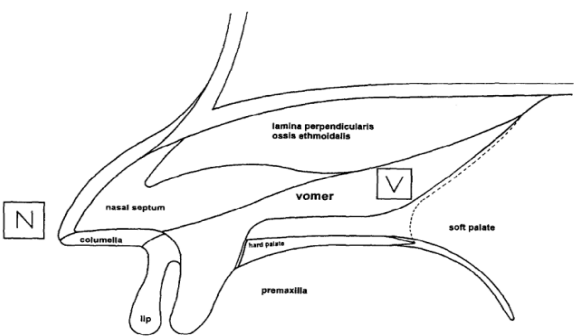


Fig.10. Sagittal view of Koch's prearranged graphic

British Journal of Oral and Maxillofacial Surgery (1995) 33, 51-58

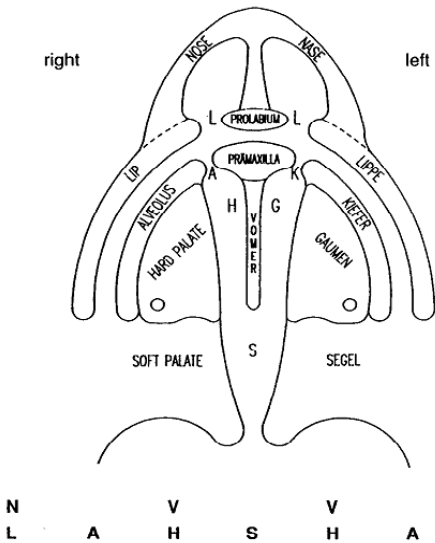


Fig.11 Transverse view of Koch's prearranged graphic
British Journal of Oral and Maxillofacial Surgery (1995) 33, 51-58.

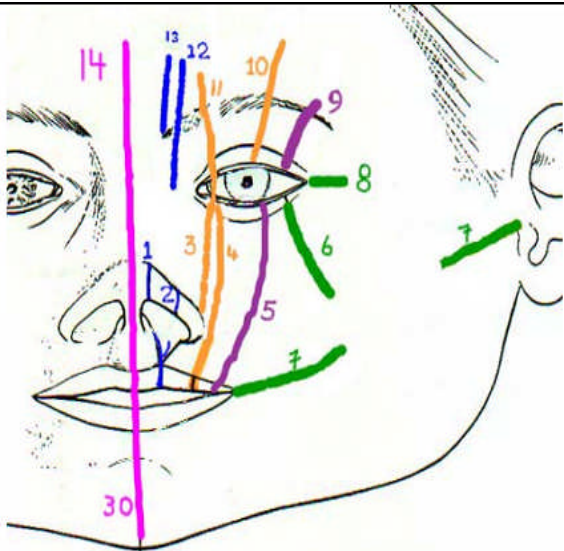


Fig.12. Tessier's cleft classification system Cleft Palate Journal, July 1989, Vol. 26, No. 3, (163-185)

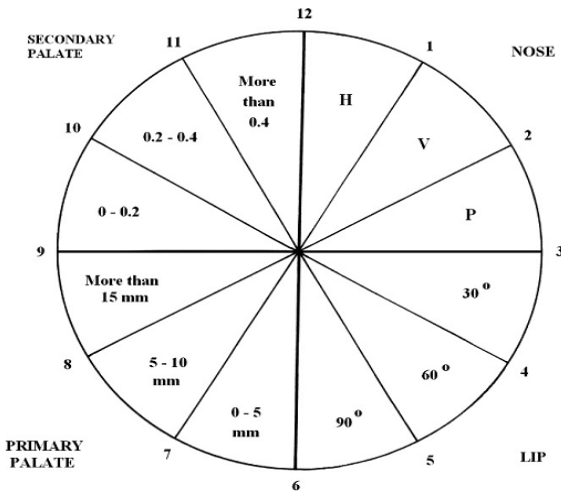


Fig-13. The Clock Diagram
Percy Rossell-Perry (2009) Cleft Palate-Craniofacial Journal: May 2009, Vol.46 No.3, pp. 305-313

Table-4 Score assigned to the clefts in Primary Palate

Primary palate	score
Normal	0
Microform	1
Incomplete1/3	3
Incomplete 2/3	6
Complete with contact of segments	12

M.R. Ortiz-Posadas et al., Cleft Palate–Craniofacial Journal, November 2001, Vol.38 No. 6, 545-50.

Table.5. Factor Corresponding to the Millimeters of Separation of the Segments

Separation in mms	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20
Factor	1. 1	1. 2	1. 3	1. 4	1. 5	1. 6	1. 7	1. 8	1. 9	2. 0	2. 1	2. 2	2. 3	2. 4	2. 5	2. 6	2. 7	2. 8	2. 9	3. 0

M.R. Ortiz-Posadas et al., Cleft Palate–Craniofacial Journal, November 2001, Vol.38 No. 6, 545-50.

Table. 6 . Score Assigned to Secondary Palate Clefts

Secondary Palate	Score
Normal	0
Submucous without bifid uvula (soft palate)	1
Submucous with bifid uvula (soft palate)	4
Incomplete 1/3 central (soft palate only)	8
Incomplete 2/3 unilateral (soft palate 1 one palatal shelf)	13
Incomplete 2/3 bilateral (soft palate 1 both palatal shelves)	14
Complete grade I* unilateral	25
Incomplete 2/3 1 complete grade II†	27
Complete grade I bilateral	28
Complete grade II unilateral	34
Incomplete 2/3 1 complete grade II	36
Complete grade II bilateral	37
Complete grade III‡ unilateral	50
Incomplete 2/3 1 complete grade III	53
Complete grade III bilateral	55

M.R. Ortiz-Posadas et al., Cleft Palate–Craniofacial Journal, November 2001, Vol.38 No. 6, 545-50.

Table 7.Examples of the Score Assigned to Some Clefts in the Primary Palate

Description of Cleft		
Left Side	Right Side	Score
Complete wcs (2 mm)	—	14
Complete wcs (17 mm)	—	32
Incomplete 2/3	Complete wcs (18 mm)	59
Complete wcs (19 mm)	Complete wcs (4 mm)	77
Complete wcs (18 mm)	Complete (18 mm)	100

M.R. Ortiz-Posadas et al., Cleft Palate–Craniofacial Journal, November

In transverse direction the cleft malformation is very easy to localize and to record: left, or right sided, or a bilateral malformation of the lip, alveolus, hard palate and nose, and the medially located cleft malformation of the soft palate.

In the vertical direction the two levels of the malformation-nose and Vomer on the one hand, lip, alveolus, hard and soft palate on the other hand have to be considered.

The sagittal direction extent (microform, subtotal or total) of the malformation of lip, alveolus, hard and soft palate is defined by adding the degree to the symbol representing the affected region (without regard to the shape)

Thus the classification is read as follows:

- bilateral total cleft of LAHS L3 A3 H3 S3 H3 A3 L3
- Right side total LAHS L3 A3 H3 S3 -
- Left side total cleft of lip -L3
- bilateral total cleft of lip and alveolus L3 A3 A3 L3
- Bilateral total cleft of hard and soft palate -H3 S3 H3-
- Uvula bifida -SI-
- The formula is read like a roentgenograph: The right side of the patient is written on the left side of the paper.
- The malformation of the outer nose and Vomer is documented in a second line above the recorded malformation of LAHS (without regard to the shape), for example: bilateral total cleft malformation of the outer nose and Vomer N3 v3 v3 N3

A submucous cleft shows the same pathological findings, except that it is covered with soft tissue. That means that the functional tissue layer (bone, muscle or cartilage) is affected as well as in an open cleft form. These findings should be diagnosed and documented in the same manner. To be able to record a submucous, open or a partly submucous/partly open form of a cleft malformation, we use a second numeral following the degree of the sagittal extent: Submucous 1 ;Partly open/partly submucous 2; Open

Mortier et al²³ (1997) developed a dual scale, which included two indicators: one corresponding to the severity of the cleft (ISS, or initial severity score) and another related to the surgical result (PRS, or postoperative results score). This indicator considered seven features to describe the patient. A comparison of the ISS and PRS allows for more objective judgment of the surgical result. However, it has been applied only to unilateral incomplete clefts of the primary palate.

Tessier²⁴ (1976) formulated a classification system based upon his extensive personal experience. This

system uses the orbit as the frame of reference and the clefts are based around this axis. A broad classification is one proposed by Tessier (1976) utilizing a clockface analogy from 0 to 14, Table-2. The point of reference for these clefts is the orbit with the clefts found in two different hemispheres. Those of the lower lid region are facial, while those of the upper lid are cranial. Clefts 0 through 4 have extensions downward to involve the maxilla and fit into the usual cleft lip and palate classifications. Their superior extensions are the more severe major cranial anomalies (Fig.12)

M.R. Ortiz-Posadas, L. Vega-Alvarado, J. Maya-Behar²⁵, proposed a new method, which allows for a complete description of primary and secondary cleft palates, incorporating elements that are related to the palate, lip, and nose that will also reflect the complexity of this problem. They developed a mathematical expression to characterize clefts of the primary palate, including the magnitude of palatal segment separation and the added complexity of bilateral clefts, yielding a numerical score that reflects overall complexity of the cleft. Clefts of the secondary palate are also considered in a separate score. Using this method, it is possible to incorporate elements that are not considered in other approaches and to describe all possible clefts that may exist. In the case of cleft primary palate, along with the surgeon, they determined the necessary elements that to be considered are:

1. The complexity of unilateral complete clefts with contact between the primary palate segments (cbs).
2. The separation, in millimeters, in the case of unilateral complete clefts without contact between the primary palate segments (wcbs).
3. The additional complexity associated with bilateral clefts.

Scores associated with the complexity of unilateral complete clefts with cbs are shown in **Table 4**. Scores range from 0 (normal primary palate) to 12 (complete cleft of the primary palate with contact between the segments). The degree of separation between the segments in unilateral complete clefts with no cbs was used to establish level of complexity. The relationship between the magnitude of segment separation and complexity was considered to be directly proportional (the greater the separation, the greater the surgical complexity). As such, a separation factor was assigned to each millimeter of separation (**Table 5**). From a surgical and aesthetic-functional perspective, the complexity of a bilateral cleft and its repair exceeds the simple summed complexity of the unilateral clefts that form the bilateral cleft. For that reason, bilateral clefts were scored as 1.5 times the sum of the unilateral cleft components. Therefore, in the case of unilateral clefts wcbs, the value 12 (see Table 1), corresponding to a complete cleft with contact between the segments (cbs) is multiplied by the factor corresponding to the millimeters of separation between the segments. For example, a complete cleft wcbs (12

mm) has a score of $12 \times 2.2 = 26$ (rounded off to whole numbers).

As an example of complexity score determination in the case of a bilateral cleft–primary palate with this methodology, consider a bilateral cleft–primary palate with the following characteristics: a left incomplete cleft (one-third) and a right complete cleft, with a 3-mm separation between the segments.

To obtain the overall complexity score:

Calculate the relevance of each unilateral cleft: Left incomplete one-third = 3. Right complete wcbs (3 mm) = $12 \times 1.3 = 15.6$. Sum unilateral cleft complexities: $3 + 15.6 = 18.6$.

Multiply the result by the bilateral cleft complexity factor (1.5) i.e. $18.6 \times 1.5 = 27.9$.

A method that fully describes clefts of the primary and secondary palate, taking aesthetic and functional elements such as the features of the cleft itself and the deformity of the lip and nose into account, (see Table 6 and 7), provides a very valuable tool for the evaluation of progress in the patients' rehabilitation. The advantages of this utility may be seen in the work of Mortier et al²³ (1997), even though their approach is limited to incomplete cleft of the primary palate. Using the method proposed here, all possible cleft forms and their severity can be characterized.

Percy Rossell-Perry²⁶ gave the **Lima clock diagram 2009**, is the design of a new diagram for cleft lip and palate, based on the degree of severity of the four basic cleft components: nose, lip, primary palate, and secondary palate. **The clock diagram, Figure-13** is a circle divided into four areas, one for each cleft component. Each area is subdivided into three segments, which represent the three degrees of severity- mild, moderate, and severe. He assigns the clock numbers (1 to 12) to each degree of severity of the four components as follows:

- Right superior quadrant (nasal deformity). Degrees: Mild (1), Moderate (2), Severe (3).
- Right inferior quadrant (medial segment lip and Prolabium deformity).Degrees: Mild (4), Moderate (5), Severe (6).
- Left inferior quadrant (primary palate). Degrees: Mild (7), Moderate (8), Severe (9).
- Left superior quadrant (secondary palate).Degrees: Mild (10), Moderate (11), Severe (12).

Merits of this Lima Clock Diagram method:

1. Characterize clefts according to their severity.
2. It is possible to incorporate elements that are not considered in other approaches and to describe all possible clefts.
3. Clock diagram describes unilateral and bilateral cleft lips and / or palates, by assessing the severity of each of the four cleft components.
4. This method provides a very valuable tool for the evaluation of progress in patient rehabilitation.

5. This severity-based classification and clock diagram are directly related to the management protocol used in our clinic

Limitation of their system is the absence of lateral segment description on the clock diagram and of other components such as the nasal septum and maxilla.

CONCLUSION

The upper lip, premaxilla, and primary palate are formed by the merging of three structures: the frontonasal process and the right and left processes of the maxilla. Any disturbance in the merging of the above processes results in the formation of the clefts. The incisive foramen is a basic anatomic landmark for classification of cleft lip and palate. There are about one hundred combinations of the cleft lip and cleft palate. Proper diagnosis of this cleft formation and its severity assessment helps in planning and execution of the appropriate treatment. An attempt is made to review the various classifications of cleft lip and cleft palate. An ideal system must be easy to understand, to document, to locate and to quantify the cleft lesion, transcend language barriers, easily applicable to computerized data analysis, should be applicable for both research and clinical applications.

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