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CLASSIFICATION OF CLEFT LIP AND PALATE - PAST TO PRESENT

Orthodontics

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ABSTRACT

Cleft lip and palate is the most common congenital anomaly affecting 1.6 per 1000 live births. Cleft lip and palate may involves the lip, alveolus, hard palate and soft palate in different combinations. As cleft lip and palate involves diverse combinations and requires an interdisciplinary approach towards treatment, a common language of communication is required. The classification of cleft lip and palate has come a long way since the first classification system by Davis and Ritchie in 1922 till now. This article explores the different classification systems and the evolution of classification systems of cleft lip and palate till now.

KEYWORDS

INTRODUCTION

Cleft lip and palate is the most common congenital deformity of the craniofacial region with a global incidence of 1 in 700[1]. The cleft of the lip and palate may involve several combinations of the lip, alveolus, hard palate and soft palate including complete and incomplete. To facilitate communication among clinicians, patients and hospital administrators a common understandable language is required. A good classification system aids in better communication, organization of data into a detailed and simplified system, aids in treatment planning as well as record keeping. Classification of cleft lip and palate requires a universal, simple, and practical classification system. This article explores the diverse classification systems of cleft lip and palate from past till the present.

Classification systems:

Davis and Ritchie (1922)

John Staige Davis and Harry P. Richie were among the first advocates who believed in discarding the age-old term "hair lip", renaming it as the "congenital cleft of the lip" at the American Medical Association in St. Louis in 1922[2].

They classified cleft lip and palate on the basis of surgical consideration taking the alveolar process as the basis of classification[2]. They classified cleft lip and palate into three groups:

Group I: Pre-alveolar process cleft
1. Unilateral (right/left, complete/incomplete)
2. Bilateral (right-complete/incomplete, left-complete/incomplete)
3. Median (complete/incomplete)

Group II: Postalveolar process cleft
1. Soft palate
2. Hard palate

Group III: Alveolar process cleft
4. Unilateral (right/left, complete/incomplete)
5. Bilateral (right-complete/incomplete, left-complete/incomplete)
6. Median (complete/incomplete)

This classification system has historical significance being the first classification system, but still it doesn't hold universal acceptance. One of the attributable reasons could be the consideration of surgical perspective which is subject to change as opposed to anatomy. Another drawback of this classification system includes the ambiguity and overlapping among the groups. According to Davis and Ritchie any cleft of the alveolus irrespective of the lip or palate should be categorized under Group III making it unclear.

Brophy (1921 to 1923)

Tuman W. Brophy gave an extensive classification system based on 5076 operations of cleft palate repair and 2676 operations of cleft lip repair[3]. The exhaustive classification system consisted of 16 types with his consideration to include every muscle and bone involved in the deformity[4]. Brophy was applauded for his classification system but according to many surgeons it was a complex classification system which had many categories that could have been grouped together[5,6].

Veau (1931)

Victor Veau introduced his classification system in his book "Division Palatine"[6]. Veau gave a simple classification system consisting of four types:

I. Clefts of the soft palate
II. Clefts of the soft and hard palate, up to the incisive foramen
III. Clefts of the soft and hard palate extending unilaterally through the alveolus
IV. Clefts of the soft and hard palate extending bilaterally through the alveolus

In1938,Veaue classified cleft lip separately according to laterality into unilateral, bilateral and median and according to extent into simple and total[7]. This classification system is simple with practical applications but it does not classify rare cleft deformities such a median cleft of the lip.

Fogh Andersen (1942)

Poul Fogh Andersen was a surgeon who considered the incisive foramen, instead of the alveolar process, as a better basis of division for classification of cleft lip and palate[8]. He observed that labial clefting was very commonly associated with the cleft of the alveolar process, suggesting an embryological association among the two, hence introducing an embryological perspective on the cleft classification.

He classified cleft lip and palate into four categories:
1. Harelip (single or double) - signifying the cleft of the lip
2. Harelip with cleft palate - signifying the cleft of the lip along with cleft palate
3. Isolated cleft palate
4. Rare atypical clefts, e.g., median cleft lip

Kernahan and Stark (1958)

Desmond A. Kernahan and Richard B. Stark like Fogh Anderson suggested the embryological basis of cleft classification, taking into account the incisive foramen as the key point[9].

Kernahan and Stark classified cleft lip and palate into three categories:
1. Clefts of structures anterior to the incisive foramen
2. Clefts of structures posterior to the incisive foramen
3. Clefts of the structures both anterior and posterior to the incisive foramen.

Modifiers were used to signify laterality (unilateral,bilateral and median) and severity (total and subtotal). Kernahan (1971) depicted their classification system in the form of a pictorial representation known as the "striped Y" diagram"[10] (Figure 1(a)). The Y diagram consists of nine boxes with nasopalatine foramen in the center. The right and left limb of the Y represents the lip (1-right aspect, 4- left aspect), alveolus (2- right side, 5-left side) and area between the alveolus and incisive foramen (3-right side, 6- left side) respectively. The stem of the Y is divided into areas representing the hard palate (7, 8) and soft palate (9).

Stippling in the segments represents the extent of the cleft, and a straight line vertically between the arms of the Y represents a median cleft, and blocking of the area indicates complete absence of the primitive palate. A lazy S at the junction of the top two limbs of the Y depicts an isolated cleft lip nose[11].

The drawbacks of this pictorial representation includes[12]:
1. The degree of cleft was not represented.
2. Premaxillary protrusion and alveolar arch collapse cannot not be depicted.
3. Velopharyngeal incompetence cannot be illustrated as it lacks representation of functional aspects.
4. The pictorial representation does not have provision for recording patient details.
5. There is inadequate information for recording asymmetric cleft lip deformities in bilateral cleft lip patients.
6. Incorporation of data into the computer is difficult.

The Kernahan classification system has been modified by Elsahy[13], Millard[11,14], and Freidman et al[15].

Elsahy (1973)[13] modified the Kernahan striped Y classification by adding triangle number 1 and 5 representing the nostrils of the right and left side respectively. Circle 13 represents the premaxilla, squares 2 and 6 represent the lip, and squares 3 and 7 represent the alveoli. While squares 4 and 8 represent the pre-palate and squares 9 and 10 represent the hard palate proper of the right and left sides respectively. Square 11 represents the Velum, and circle 12 represents the posterior pharyngeal wall (Figure 1(b)). With this modification even the functional aspect including the velopharyngeal incompetence could be depicted.

Millard(1977)[11,14] further modified Elsahy's revision of Kernahan's striped Y diagram by adding inverted triangles at the top of triangle 1 and 5 to represent the right and left aspect of the nasal arch (Figure 1©).

Friedman et al (1991)[15] gave the modification combining the schemes of Elsahy and Millard, by incorporating cleft microforms and assigning severity scores to the anatomic and functional deformities. The data collection sheet by Friedman et al. was further improvised by Davison et al. (Figure 1(d))[16].

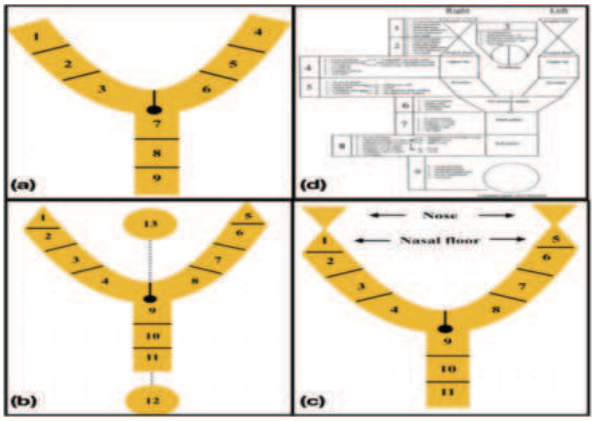

Figure 1(a). Kernahan striped Y pictorial representation, (b) Elsahy's modification of the Y diagram, (c) Millard modification of the Y diagram and (d) Friedman modification.
*- derived from Davison et al.[16]

American Cleft Palate Craniofacial Association Classification (Harkins et al., 1962)
The American Association for Cleft Palate Rehabilitation chaired by Cloyd S. Harkins appointed a committe for the formulation of a classification system[17].

Classification consisting of four categories was devised:
1. Clefts of the prepalate
a. Cleft lip
b. Cleft alveolus
c. Cleft lip, alveolus, and primary palate

2. Clefts of the palate
- Cleft of the hard palate
- Cleft of the soft palate
- Cleft of the hard and soft palate

3. Clefts of the pre-palate and palate
4. Facial clefts other than prepalatal and palatal
- Cleft of the mandibular process
- Naso-ocular clefts
- Oro-ocular clefts
- Oroaural clefts

Each cleft was further divided according to laterality into left, right, bilateral, or median. In terms of extent, the cleft was divided into 1/3, 2/3, or 3/3 the length of the deformity. In 1962, a Nomenclature Committee which was agreed by the American Association for Cleft Palate Rehabilitation which formulated the criteria for an ideal classification system[17] (Figure 2).

Criteria for ideal classifiction system:

I. **Concise, clear definitions of terms,**
A. Rejection of the meaningless, the ambiguous, and the irrelevant
B. Preference for simple, descriptive English terms
C. Retention of established customary terms, where possible, in order to avoid duplication and confusion
D. Formation of new terms only where necessary
E. Indication of synonomous terms, especially those of wide usage such as ones based on Latin or Greek, to facilitate comprehension and use of the system proposed here

II.Convenience of use through:

A. Economy of expression
B. Logical arrangement of classification conformable with
C.Standardized methods of measurement

III. Stimulation of scholarly and clinical research by:
A. Standardized procedures for observation and reporting B. Provision for rare conditions

Figure 2- Criteria for an ideal classification system

Sancho 1962
Vilar Sancho classified clefts based on Greek nomenclature. Lip was designated by "K" (keilos), alveolus by "G" (gnato), hard palate by "U" (urano) and soft palate by "S" (stafilos). Capital letters were used to represent complete clefts and partial clefts were represented by small letters. "2" was used to indicate bilateral, "d" to represent right, "l" indicated left, an "I" to represent incomplete and "o" to indicate operated. Due to the incompleteness and language barrier this classification system could not be universally adopted[18].

International Classification (Broadbent et al., 1969)
The International Confederation for Plastic and Reconstructive Surgery in devised a classification system of cleft lip, alveolus, and palate based on embryology and facial clefts based on topography[19].

Classification of the lip, alveolus, and palate:
1. Clefts of the anterior palate
2. Clefts of the anterior and posterior palates
3. Clefts of the posterior palate

Classification of rare facial cleft (based on topographical findings):
A. Median clefts of the upper lip, with/without hypoplasia or aplasia of the premaxilla
B. Oblique clefts (oro-orbital)
C. Transverse clefts (oroauricular)
D. Clefts of the lower lip, nose, and other very rare clefts

Dahl 1970

Dahl gave a simple classification scheme dividing clefts into three main categories as CL-cleft lip, CP-cleft palate, and CLP- clefts of the lip and palate. Cleft of the lip and palate was further subdivided into UCLP-unilateral clefts of the lip and palate, and BCLP-bilateral cleft lip and palate[20].

A modification of Dahl's classification system was introduced by Sandham who added a fifth group describing the rare types of facial clefts including median clefts of the lip and oblique facial clefts[21].

Spina 1973

Victor Spina[22] did a minor modification of the International Classification's first tier, giving an updated version of the previous system:
Group I: Pre-incisive foramen clefts (Unilateral/Bilateral/Median)
Group II: Trans-incisive foramen clefts (Unilateral/Bilateral)
Group III: Post-incisive foramen clefts
Group IV: Rare Facial clefts

Indian classification 1975

The Indian classification system given by Balakrishnan described cleft deformities into three different groups[23]:
Group I : Cleft of Lip
Group I(A) : Cleft of Lip with Cleft of Alveolus
Group II : Cleft of Palate
Group II(S) : Submucous Cleft of Palate
Group III : Cleft of Lip and Palate

The side of the cleft is depicted by: (R) for right, (L) for Left and (RL) for bilateral, and each component of combinations are separated by '/'.

A modification of the Indian classification system has been proposed by Agrawal[24] who incorporated newer abbreviations: P(Partial), S(submucous), sb(Simonart's band), micro(microform) and Pmax(Protruding premaxilla), with each component separated by '+' instead of the original '/'.

LAHSHAL system 1985

The LASHAL system was introduced by Kriens (1985) with letters representing the lip, alveolus, soft palate and hard palate. Complete clefts are represented in capitals, incomplete clefts are represented with small case letters[25]. 'LAHSHAL' represents a bilateral total cleft of the Lip, Alveolus, Hard and Soft Palate, while a cleft involving the left side of lip and alveolus would be represented as '…AL'. A small letter marked with an asterisk symbolizes a submucous cleft malformation or a microform.

Even though simple, this classification system lacks the ability in describing complex cleft malformations, also it cannot differentiate between a submucous cleft and a microform.

RPL system 1993

Schwartz et al. (1993) modified the Kernahan classification system by converting the stripped Y into a three digit numerical system known as the RPL system[26]. The right limb of the stripped Y consisting of the right aspect of lip (1), alveolus (2) and area between the alveolus and incisive foramen (3) is represented by R while the base of the Y consisting of the hard palate (7,8) and the soft palate (9) is represented by P, and the left limb consisting of the left aspect of lip (4), alveolus(5) and area between the alveolus and incisive foramen (6) is represented by L. Each is further divided by numerals 1 to 3, consistent with the deformity involved. This system allows quick identification and incorporation in the form of computerized data. However, this is a simple classification system which is incompetent in describing an incomplete cleft deformity.

LAHSN 1995

The classification system described by Koch and Koch[27] known as LAHSN represents the anatomical regions: lip, alveolus, hard palate, soft palate and nose (Table 1). The extent of the malformations of LAHSN depends on its severity in the sagittal, transverse and vertical dimensions, also depending on whether it is submucous or open form. The extent and severity is graded by a tripartition scheme dividing the deformity into microforms (1), subtotal (2) and total (3) form.

In transverse direction the cleft deformity is recorded as left, or right sided, or a bilateral malformation of the lip, alveolus, hard palate and nose, and the medially located cleft deformity of the soft palate. In the vertical direction the two different levels of the deformities i.e the nose and vomer on one hand, and the lip, alveolus, hard and soft palate on the other hand are considered. In the sagittal dimension the extent of the deformity of the lip, alveolus, hard and soft palate is defined by degree. A total bilateral cleft of the lip, alveolus, hard palate and soft palate is represented as L3 A3 H3 S3 H3 A3 L3. Any deformity of the outer nose and vomer is represented by a second line above the recorded deformities of LAHS. A second numeral is used to signify the extent of the submucous, open or a partly submucous/partly open form of a cleft deformity if any.

Table 1- LAHSN classification system

Extent		Shape	
Grade 1	Microform	Submucous	1
Grade 2	Sub total	Partly open/Partly submucous	2
Grade 3	Total	Open	3
Not affected	/		

LAPAL system 2007

The LAPAL system was introduced by Liu et al[28] and is designated by five Arabic numerals representing the extent of the cleft deformities ranging from intact(0) to complete cleft(4) (Table 2). Numerals are designated from the right to the left side, corresponding to how one viewed when seeing the patient. Only one numeral is used to represent the palate posterior to the incisive foramen as the clefts posterior to the hard palate and soft palate mostly lie in the midline.

The LAPAL is a simplified, precise and easy to understand system which is liable to computerized analysis. In the LAPAL system 00444 represents a complete cleft lip and palate on the left side, while 00200 represents a soft palate and submucous cleft.

Table 2- LAPAL system of classification of CLP

Side	Right	Right	Middle	Left	Left
Component involved	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
Submucous	1	1	1	1	1
Intact	0	0	0	0	0

Lima clock diagram 2009

Percy Rossell Perry introduced the Lima clock diagram 2009, designating the degree of severity of the nose, lip, primary palate, and secondary palate[29] (Figure 3). The clock diagram is represented by a circle which is divided into four parts, each one designating a cleft component, each is further subdivided into three components signifying the severity- mild, moderate, and severe. A clock number (1 to 12) is assigned to designate the degree of severity of the four components involved.

The Clock diagram is capable of describing the severity of the deformity, and takes all the four components i.e nose, lip, primary palate, and secondary palate involved in the deformity into consideration. Also in comparison to the Kernahan Y diagram this diagram represents the severity of the deformity involved. According to Perry et al. this classification system is related to the management protocol at their center and can also be used to evaluate the progress. Limitations of this system include the absence of description of other components such as the nasal septum and maxilla.

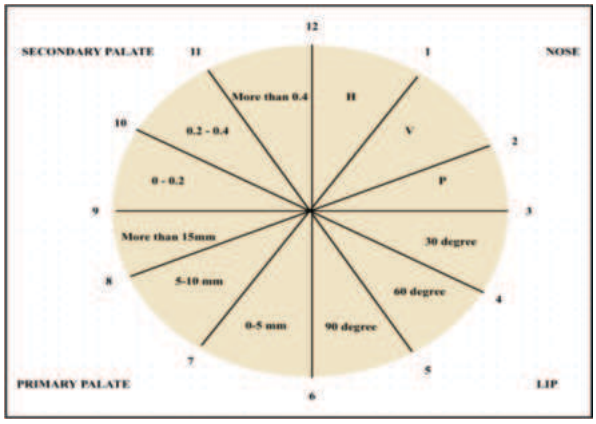

Figure 3- The Clock diagram

Liujsterburg 2014

Liujsterburg et al. examined the records from the Dutch Oral Cleft Registry and developed a classification system based on the embryology of the primary and secondary palate[30]. The records were divided into three categories: cleft lip/ alveolus (CL/A), cleft lip/alveolus and palate (CL/AP), and cleft palate (CP). The records were further subdivided into three groups: fusion defects, differentiation defects, and fusion and differentiation defects based on the events during the embryonic process (Table 3). This classification system allows classification based on embryological events and can be used for research, but has little role in clinical practice.

Table 3- Classification of Cleft deformity according to Fusion and/or Differentiation Defects.

Fusion defects	Primary palate	Complete cleft lip Complete cleft alveolus Incomplete cleft alveolus (only if lip is normal or has a complete defect)
Fusion defects	Secondary palate	Complete cleft hard palate Incomplete cleft hard palate Complete cleft soft palate Incomplete cleft soft palate Complete uvular cleft Incomplete uvular cleft
Differentiation defects	Primary palate	Incomplete cleft lip Submucous cleft lip Hypoplastic lip Incomplete cleft alveolus (only if the lip has an incomplete or submucous cleft) Submucous cleft alveolus Hypoplastic lip/alveolus
Differentiation defects	Secondary palate	Submucous cleft hard palate Hypoplastic hard palate Submucous cleft soft palate (including uvula) Hypoplastic

DISCUSSION

The classification of CLP has come a long way from the first classification system by Davis and Ritchie[1] in 1922 till now. The paradigm has changed from the anatomical perspective to the embryological perspective. The anatomical or surgical considerations are subject to change as opposed to the embryological events. The embryological classification has also been integrated into the International Classification of Diseases by the World Health Organization (WHO)[27]. In the present scenario the Kernahan and Stark classification system still holds popularity among other classification systems due to its versatility, pictorial representation and constant modifications. But still no classification system can be regarded as a universal classification sytem[13-16].

According to Millard the attributable reasons could be the language differences, inaccuracies, omissions and lack of simplicity[11]. Moreover the treatment of CLP patients requires an interdisciplinary approach, which requires consideration of a multitude of other factors as well. A researcher might prefer a classification with an embryological perspective while a clinician might prefer a simple classification system that correlates with treatment goals and principles, on the other hand an epidemiologist might prefer a classification system that is comprehensive and versatile[4]. An ideal system must be easy to understand, simple to document and quantify, should be devoid of language barriers, and liable to computerized data analysis. The classification system of CLP hence, will always be open to debate, criticism and constant modification with time and progress in treatment and understanding.

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