



# Classification Systems for Orofacial Clefts

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## ABSTRACT

Classification systems proposed are based on the cleft lip and palate morphology and a few are based on the embryonic principles, the main aim to propose these classification systems was to ease the treatment planning. Not only this it helps the patient to name, grade, remember, plan and discuss the clinical scenario. Some of the classifications are Tessier type, Veau, and Davis and Ritchie classifications. The American Cleft Palate Association and Kernahan and Stark classifications probably represent the best variable systems today, and they are presented in some detail for comparison. But a more standard classification is still awaited.

**Keywords:** Orofacial clefts, Cleft lip, Cleft lip and palate (CLP), Cleft palate (CP).

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## INTRODUCTION

The group of orofacial cleft anomalies is heterogeneous. It comprises typical orofacial clefts, such as cleft lip, cleft lip and palate, and cleft palate, and atypical clefts, including median, transverse, oblique, and other Tessier types of facial clefts (Tolarova, 1998; Tessier, 1976).<sup>1,2</sup> Typical and atypical clefts can both occur as an isolated anomaly, as part of a sequence of a primary defect, or as a multiple congenital anomaly (MCA). In an MCA, the cleft anomaly could be part of a known monogenic syndrome, part of a chromosomal aberration, part of an association, or part of a complex of multiple congenital anomalies of unknown etiology.

Cleft lip can occur as a unilateral (on the left or right side) or as a bilateral anomaly. The line of cleft always

starts on the lateral part of the upper lip and continues through the philtrum to the alveolus between the lateral incisor and the canine tooth, following the line of sutura incisiva up to the foramen incisivum. The clefting anterior to the incisive foramen (i.e. lip and alveolus) is also defined as a cleft of primary palate. Cleft lip may occur with a wide range of severity, from a notch located on the left or right side of the lip to the most severe form, bilateral cleft lip and alveolus that separates the philtrum of the upper lip and premaxilla from the rest of the maxillary arch when cleft lip continues from the foramen incisivum further through the sutura palatina in the middle of the palate, cleft and palate (either unilateral or bilateral) is present. A wide range of severity may be observed. The cleft line may be interrupted by soft (skin or mucosa) bridges, hard (bone) bridges, or both, corresponding to a diagnosis of an incomplete cleft. This occurs in unilateral and bilateral cleft lip and palate. Cleft palate is etiologically and embryologically different from cleft lip and palate. Several subtypes of cleft palate can be diagnosed based on severity. The uvula is the place where the minimal form of clefting of the palate is observed. (However, a relatively high prevalence of this anomaly in the general population suggests that a certain proportion may represent the very far end of a normal variability). A more severe form is a cleft of the soft palate. A complete cleft palate constitutes a cleft of the hard palate, soft palate and cleft uvula. The clefting posterior to the incisive foramen is defined as a cleft of secondary palate.

In a significant proportion of patients, the cleft of the hard palate is covered by mucosa and continues through the soft palate, forming a so called submucous cleft palate. A submucous cleft palate may occur in the hard palate only and continue to the open cleft of the soft palate, or it may occur as a submucous cleft of the soft palate with or without a notch into the hard palate. Careful clinical examination may reveal a blue triangle in continuation of the cleft of the soft palate, which represents a cleft of the bone palate underneath mucosa. The cleft palate may take 2 distinguishable forms—a V shape, which is most common in isolated clefts, or a U shape, which is most common in Robin sequence and in syndromic clefts.<sup>2</sup>

## Various Classifications of Cleft Lip and Palate

The palate has always been included in the various classifications, sometimes with separate specification of clefts of the hard plate, soft plate and even uvula.

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Lip cleft have had a far more varied treatment. Some early classifications permitted the observation of isolated clefts of the lip, while others recognized lip clefts only as extensions of palatal clefts, or ignored them entirely. The alveolar process has had even more erratic consideration. It is completely ignored in some systems, or added as a degree of prominence with its own listing. The submucous or occult cleft has also had treatment ranging from complete omission to a separate categorical entry.

### Drawings of the Clefts

The simplest system for recording morphological details with a minimum of errors due to unclear definition of terms is to present a picture of the condition. The worksheet may include a basic outline of the palate and face, on which the observer draws the appearance of the cleft. A serious failing of this system is that it is no classification. With a drawing each individual is unique, and there is no means of grouping or comparing patients until patterns of defects are identified and given names.<sup>2</sup>

### Veau and Davis and Ritchie Classifications

The most popular classification systems were those of Veau and Davis and Ritchie. It is disheartening to note that a number of books on cleft palate have been published in this decade which completely ignore more recent contributions. These two classifications can be used as examples of some of the problems represented by older systems.

#### Veau described Four Types of Cleft

*Group I:* Cleft of the soft palate only.

*Group II:* Cleft of the hard and soft palate to the incisive foramen.

*Group III:* Complete unilateral cleft of the soft and hard palate, and the lip and alveolar ridge on one side.

*Group IV:* Complete bilateral cleft of the soft and hard palate, and the lip and alveolar ridge on both sides.

It would obviously be difficult to describe clefts of the lip and alveolar process without palatal clefts using this system.

Davis and Ritchie, who based their system on the location of the cleft relative to the alveolar process, used three major headings with some subdivisions under each.

*Group I:* Prealveolar cleft—Lip clefts only with subdivisions for unilateral, median and bilateral.

*Group II:* Postalveolar cleft—Degrees of involvement of the soft and hard palates could be specified, up to the alveolar ridge, submucous clefts could also be included.

*Group III:* Alveolar cleft—Complete clefts of the palate, alveolar ridge and lip, with subdivisions for unilateral, median and bilateral.

Clefts from groups I and II could be reported in the same subject, if there were no involvement of the alveolar ridge. There is no opportunity to report a cleft of the lip and alveolar process under these strict definitions, but Davis and Ritchie recommended adding them to group III. They also suggested this is the best place for a cleft of the palate and alveolar process without lip involvement.

The differences between these two popular systems can be noted immediately, as well as the inadequacies in both for recording all observable cleft types.<sup>4</sup> Another problem inherent in this form of numbered system is the tendency of users to revert to reporting just numbers.

### Classifications based on Embryological Patterns

Probably the most significant rallying point for the reformation of the classification system was the concept of a morphological system based on embryological patterns.

#### Cleft of the Lip, Alveolus and Palate

(Classification based on embryological principles).<sup>2,6</sup>

##### *Group I: Clefts of Anterior (Primary) Palate*

- a. Lip: right and/or left
- b. Alveolus: right and/or left.

##### *Group II: Clefts of Anterior and Posterior*

(Primary and secondary) palate:

- a. Lip: right and/or left
- b. Alveolus: right and/or left
- c. Hard palate: right and/or left.

##### *Group III: Clefts of Posterior (Secondary) Palate*

- a. Hard palate: right and/or left
- b. Soft palate: medium.

For further subdivision the terms 'total' and 'partial' should be used.

### Rare Facial Clefts

(Classification based on topographical findings)

- a. Median clefts of upper lip with or without hypoplasia or aplasia of premaxilla
- b. Oblique clefts (oro-orbita)
- c. Transverse clefts (oroauricular)
- d. Clefts of lower lip, nose, and other very rare clefts.

### Some of the Major Objections to this Classification

1. Numbering the groups is an invitation to refer to clefts by the identifying numerical types. This not only interferes with communication by requiring the reader to know the meaning of the numbers in this classification but also leads to further confusion with previous Veau and Davis and Ritchie numbers.<sup>5</sup>

2. Introduction of the term anterior palate in place of Kernahan and Stark's 'primary palate' and the American Cleft Palate Association's 'prepalate' does not resolve the conflict between terms, because it could be more easily misinterpreted than either of the others.
3. The term alveolus is presented as synonymous with 'alveolar ridge' or 'alveolar process.' This has been a common surgical colloquialism, but has never received approval by anatomists.
4. Median clefts are listed under facial clefts rather than as clefts of the lip. This represents a conceptual difference with both the Kernahan and Stark and the ACPA classifications.
5. Alveolar ridge clefts are given the appearance, in group,<sup>2,7,8</sup> of having the same embryological independence as the lip and palatal clefts.

It is hoped that further revisions are planned for this classification.

## Two Best Classifications

The American Cleft Palate Association and Kernahan and Stark classifications probably represent the best variable systems today, and they are presented in some detail for comparison.<sup>2</sup>

It should be noted first that the two classifications are identical in basic concept. They are both predicated on current embryological theory about the development of the face and recognize the independent mechanisms of development anterior and posterior to the incisive foramen. They both have three major headings.<sup>2,7</sup>

1. Cleft anterior to the incisive foramen.
2. Clefts posterior to the incisive foramen.
3. Combinations of both types of clefts.

It is immediately obvious that the Kernahan and Stark (K&S) classification is much briefer than that the American Cleft Palate Association (ACPA), but this need not be related to accuracy.

Under the major heading of cleft of primary palate (K&S) or prepalate (ACPA) both lip and premaxillary or alveolar process clefts are included; however, only in the ACPA classification can clefts of the lip and alveolar process be identified separately. The primary palate heading is essentially the same as that of the prepalate without subdivision. Some authorities (e.g. Pruzansky would consider the possible specification of lip and alveolar process separately at best unnecessary, and at worst misleading. Fogh-Andersen and others believe that the clefts of the alveolar process and lip are so closely related that the extent of one can usually be predicted from observation of the other. In fact Pruzansky believes at birth there is always at least a dimple in the alveolar process when there is a cleft lip, even though this may grow out and

disappear completely in a short time. Clinically, however, children with cleft lips present varying degrees of clefts of alveolar process, which may have some relationship to dental and speech problems. Since, there may be some significance in the consideration of the alveolar ridge independently of the lip, it seems wiser to retain this opportunity for specificity until it can be clearly proved that this is a difference that makes no difference.

In the clefts of the primary palate (K&S), it is not clear whether the subtotal and total median clefts are matters of degree of involvement, as is the specification of median cleft under 'prepalate' (ACPA). For the total absence of the premaxilla, which appears qualitatively different from the minor midline notches in the vermilion border of the lip, the ACPA classification suggests 'prepalate arrest'. This is one of the opportunities to search for the theoretical condition, and eventually confirm or reject the condition of the basis of research stimulated by the classification.<sup>2</sup>

In the clefts of the secondary palate, Kernahan and Stark disregard the usual divisions into hard and soft palates. Embryologically this may be justified, but in terms of habilitative procedures there are important theoretical differences. The attachment of the vomer to the cleft palatal shelves, provided for in the ACPA classification, has been a matter of some interest to surgeons, so that a classification that does not provide for this observation would probably be considered inadequate in practice. (Surgeons often record this observation as unilateral or bilateral clefts, as under Kernahan and Stark's clefts of primary and secondary palates). Palatal length, a matter of some concern in evaluating the structures for procedures which will lead to good speech, is included in the ACPA outline. It may be that this dimension does not have the predictive potential that has been theorized for it, and the methods of judging may be inadequate, but this can best be determined by permitting it to be included in the classification, with later evaluation of the contribution it has made to understanding the problem.<sup>9</sup>

In combined clefts of the primary and secondary palate, Kernahan and Stark have used essentially the same subheadings as in clefts of the primary palate. This suggests that a fairly simple description could cover all types of complete clefts. Of course this is not true in combinations such as a cleft of the left side of the lip and alveolar ridge, plus a cleft of the velum. The authors are aware of this, because they provide an illustration of this type of mixed condition, but their classification does not call for the kind of words (or perceptual set) that encourages such observations. The ACPA classification takes refuge in the statement that combined clefts are to be described separately as clefts of the prepalate and palate.

Both classifications recognize submucous clefts, but Kernahan and Stark do so only for clefts of the secondary



palate, thus omitting specification for conditions such as congenital scars of the lip or incomplete lip musculature. Also both classifications are weak in descriptions of palatopharyngeal incompetence and displacement of palatal segments in complete clefts. As noted above, the ACPA classification may include some conditions that do exist. Neither classification provides for a description of the shape of palatal cleft or height of the palatal shelves. These details, however, if they should ever be included, would fit comfortably as further subdivisions within the three general headings provided by both classifications.

The ACPA classification is obviously more complex than the Kernahan and Stark classification. This has probably worked to its discredit, by discouraging its use among practitioners who do not see its basic form and assume it requires more information than they are interested in obtaining. However, a good example of the need for recording more detail than is provided in the Kernahan and Stark classification can be found in the 1968 article by Conway et al,<sup>2</sup> who reviewed and categorized 850 cases of cleft lip and palate. They used the Kernahan and Stark classification, but found that they had to add additional subcategories.

### Mortier's Classification

Mortier et al (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. While these approaches attempt to characterize many features of primary and secondary palate clefts, a methodology still does not exist to adequately characterize other important features that relate to complete clefts, such as magnitude of segment separation. That is, the repair of a complete primary palate cleft with segment separation of 15 mm undoubtedly involves greater surgical complexity than one with segment separation of only 3 mm. None of the representational forms proposed to date for primary palate clefts considers this important parameter. In addition, elements associated with the patient's esthetics and functionality is considered only in a limited fashion. Therefore, a new approach to the description of primary and secondary cleft palates was proposed incorporating an element that are related to the palate, lip, and nose and that reflects their complexity from a surgical perspective. This work was developed jointly with the cleft lip and palate team at pediatric Hospital of Tacubaya, which belongs to the Health Institute of the Federal District Department in Mexico City, Mexico.<sup>3</sup>

### Recommendations for a Standardized Classification

The time for agreement on some standardized morphological classification of long overdue. The need for a universal consistent method of reporting findings has been cogently argued in print for over 15 years, and several individuals and groups have attempted to produce acceptable patterns. Among the classifications that have been published at least for the last 10 years there appears to be more similarity in basic organization than diversity.<sup>13,15</sup>

In at least the first levels of description, the terms of one classification can be transferred to the other.<sup>1</sup> There should be specification of prepalatal (or primary palatal) clefts, including degree of cleft of both the lip and the alveolar process.<sup>10-12</sup> There should be specification of palatal (or secondary palatal) clefts, including involvement of both the hard and soft palates. Clefts involving both the prepalate and palate (or primary and secondary palates) will still need individual specification relative to these two developmental systems.<sup>14,15,17</sup>

At least on this basic level, scientists and clinicians used some rigor and consistency in reporting observations according to this pattern, and actively resisted using traditional or strictly local forms of recording data.<sup>16,17</sup>

There are standard methods of recording observations in an orderly fashion, with level of generalization following each other. If the patient has, for example, a cleft of the lip, this should be reported first as a cleft of the prepalate, then of the lip, then of the particular side and extent, and possibly other subdivisions as considered appropriate or necessary.<sup>18</sup>

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