Comprehensive and reliable classification system for primary diagnosis of cleft lip and palate

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ABSTRACT

Objective: Due to the lack of a universally accepted classification system, we are aiming to introduce a modified comprehensive, precise and relatively simple classification system for primary diagnosis of cleft lip and palate.
Methods: The proposed classification is based on the Kernahan's striped Y diagram with more details in cleft extent and with the addition of severity scores to each cleft component. Clear definitions of cleft extents and severity degrees were described based on 400 consecutive primary cases. Two medical students were taught the classification then diagnosed photographs of 100 cases twice to test its reliability.
Results: The students' results were 11% and 13% wrong diagnoses for student 1 and 2 in the first time, 8% and 10% in the second time, respectively. The inter-rater reliability for the two students in the first and second time was 0.716 and 0.878, respectively. The intra-rater reliability for student 1 and 2 were 0.826 and 0.755 respectively. The average duration to diagnose a case was less than a minute.
Conclusion: This classification is comprehensive and records many diagnostic variables with high reliability and precision.

1. Introduction

Cleft lip and palate is one of the most common congenital anomalies, however, a universally accepted classification system is still unavailable. Such classification is important for standardization of patients' registration, caregivers' communication, in addition to research and epidemiological purposes.

The main reason for lack of such classification is the wide spectrum of presentation of cleft patients. To reach a complete diagnosis, it is important to describe not only the anatomical 'extent' of the cleft (how much is the extension of clefting? such as cleft uvula, cleft soft palate, cleft soft and hard palate, ...), but also to determine the different degrees of 'severity' for each cleft 'extent' described (how much is the width of the cleft?, are the alveolar segments wide or collapsed?, how severe is the nasal deformity?...). So, each cleft 'extent' can be presented with different 'severity' scores. For example, a diagnosis of unilateral complete cleft lip and palate (the same cleft 'extent') could be presented with different 'severity' degrees as

regards nasal deformity, width of cleft lip, collapse of the alveolar segments, width of the cleft palate...etc.

The primary 'severity' degree could impact the difficulty of repair, technique selection and the management protocol (Rossell-Perry, 2009). Furthermore, it could influence the final outcome such as the increased incidence of maxillary growth restriction associated with both wider cleft palate (Liao et al., 2010) and wider alveolar cleft (Tomita et al., 2012). Recently, studies have shown that wider clefts have not only increased risk for palatal fistula (Yuan et al., 2016), but interestingly also for velopharyngeal insufficiency (Lam et al., 2012; Yuan et al., 2016). Moreover, a correlation between the initial 'severity' of unilateral cleft lip and the final nasolabial appearance was reported (Mortier et al., 1997). Therefore, recording the 'severity' for each cleft 'extent' is essential for the complete diagnosis. Nevertheless, such 'severity' degrees were ignored by most of the classifications (Kernahan, 1971; Elsahy, 1973; Kriens, 1990; Schwartz et al., 1993; Koch et al., 1995; Smith et al., 1998; Liu et al., 2007).

The simplest system for recording cleft lip and palate is to present a diagram with the visual summary of the anatomical defect (Friedman et al., 1991). This might be the reason behind the gained reputation of the Kernahan striped Y symbolic classification.

Despite its simplicity, it lacks many cleft 'extents' and totally ignores the 'severity' (Kernahan, 1971). Kernahan divided the clefting into the primary and secondary palate with the incisive foramen as a landmark between them (Kernahan and Stark, 1958). Later on, they designed their Y symbolic classification system based on that principle (Kernahan, 1971). In fact, this sound embryological background was first introduced by Fogh-Anderson (Fogh-Anderson, 1942).

Over the years, Kernahan's classification has undergone many modifications to extend it and few trials to add 'severity' scores to some components. Still these modifications were incomplete and sometimes complicated (Elsahy, 1973; Millard, 1990; Friedman et al., 1991; Smith et al., 1998). In parallel, newer systems and diagrams were described (Kriens, 1990; Schwartz et al., 1993; Koch et al., 1995; Liu et al., 2007; Rossell-Perry, 2009), but they have not gained wide recognition due to either insufficiency or complexity. We are proposing a comprehensive and relatively easy classification system for primary diagnosis of cleft lip and palate based on the Kernahan striped Y classification.

2. Patients and methods

The classification was applied on more than 400 consecutive primary cases. It is an extension of the striped Y symbolic classification of Kernahan (1971). The classification is based on 4 main components of deformities; the nose (N), the lip (L), the premaxilla or alveolus (A) and the secondary palate (P). Both the cleft 'extent' and its 'severity' were recorded for each component. The 'extents' of cleft are represented in the main Y diagram, while the 'severity' scores are located beside each correspondent component. The nose has only a 'severity' score above the Y diagram as it is not clefted. Some components have right and left limbs to indicate the sidedness when appropriate. When using the system, the boxes of the cleft 'extents' for each component are to be marked in sequences, while the needed degree only is marked to record the 'severity'. Certain views are needed when examining the patient for 'severity' scores.

2.1. Description of the new classification system (Fig. 1)

- **Nasal deformity severity**:

    Unilateral (basal view) (Fig. 2):
    S1: The cleft-side nostril is traversed by horizontal line bisecting the non-cleft side nostril.
    S2: The cleft-side nostril is below that line.
    S3: S2 + depressed curvature of the ala.
    Bilateral (basal view) (Fig. 3):
    S1: Columellar length ≥1/4 of the nasal tip height from the columellar base.
    S2: Columellar length <1/4 of the nasal tip height from the columellar base.
    S3: No columella.

- **Cleft lip extent:**

    E1: Microform.
    E2: Cleft ≤1/2 lip height.
    E3: Cleft >1/2 lip height.
    E4: Complete cleft lip with Simonart's band.
    E5: Complete cleft lip.

- **Cleft lip severity:**

    Unilateral (frontal view) (Fig. 4):
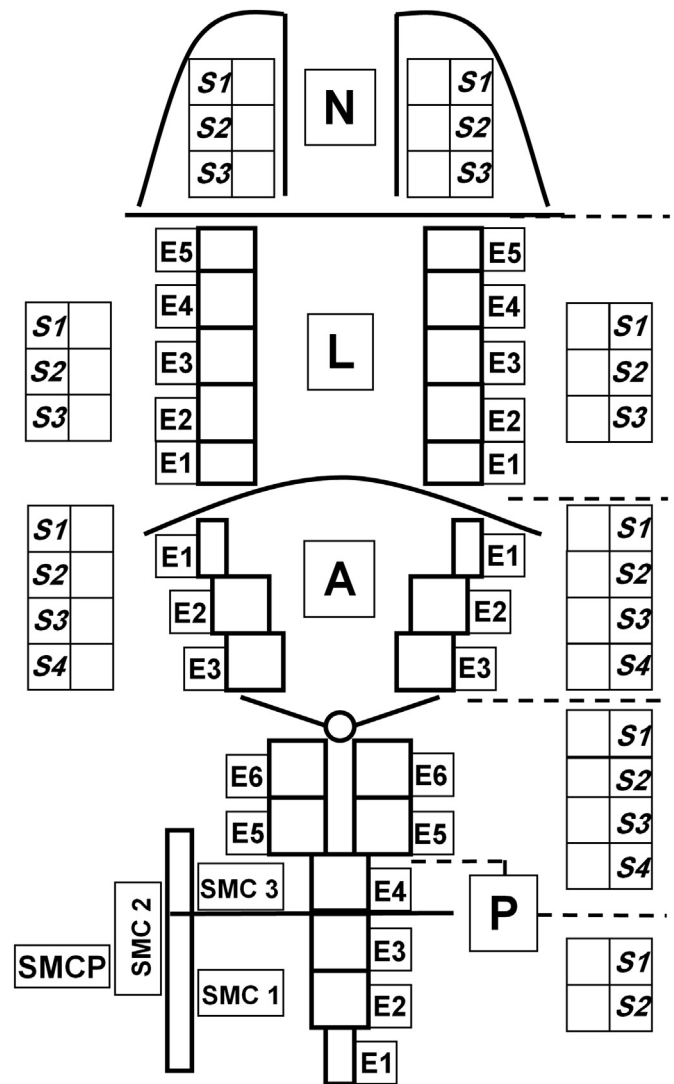   The medial and lateral lip elements at the vermilion are:

Fig. 1. The new classification system.

S1: about to touch each other.
S2: separated by ≤ width of the non-cleft side nostril.
S3: separated by > width of the non-cleft side nostril.
Bilateral (basal view) (Fig. 5):
Prolabium forward protrusion by:
S1: ≤1/2 of the vertical nasal height from the alar base to the tip.
S2: >1/2 to <3/4 of the vertical nasal height from the alar base to the tip.
S3: ≥3/4 of the vertical nasal height from the alar base to the tip.

- **Cleft alveolus extent:**

    E1: Notch.
    E2: Cleft of the alveolar margin not reaching the incisive foramen.
    E3: Cleft of the alveolar margin reaching the incisive foramen.

- **Cleft alveolus severity (intraoral basal view)** (Fig. 6):

    S1: Narrow, no collapse.
    S2: Narrow, collapse.
    S3: Wide (≥4 mm), no collapse.
    S4: Wide (≥4 mm), collapse.