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Median cleft of the upper lip: A new classification to guide treatment decisions

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ABSTRACT

Median cleft of the upper lip (MCL) is a specific and rare entity on the spectrum of facial clefts. MCL have different clinical expressions and can be either isolated or part of multiple malformations. Confusion still exists regarding the explanation and classification of MCL; some cases have been reported in the literature, but no studies carried out a complete review of the literature.

This study reviewed cases of MCL in 2 French units and conducted a systematic review of the literature, in order to derive a new classification.

Fourteen patients with MCL in the 2 units and 195 cases in the literature were reviewed. They involved complete (42%), incomplete (49%), and minor forms (9%). Epidemiological and clinical data were collected, from which a classification was derived, based on the type of cleft and its belonging to other syndrome(s). Three main groups were distinguished, namely, isolated MCL, MCL within craniofacial malformations, and MCL with extrafacial malformations. Each group and subgroup was associated with a prognosis and led to specific management.

This study reviewed all of the various forms of MCL and their associated anomalies, in order to have a global view of MCL and to derive a useful classification scheme to guide management of care.

1. Introduction

Median cleft lips (MCL) are part of an extremely rare group of facial clefts defined by a cleft involving the upper lip, situated on the median line. MCL have different clinical expressions, from a small notch of the vermilion to a cleft of the entire length of the philtrum with nasal and maxillary extension (Elias et al., 1992; Lee, 1985; Urata and Kawamoto, 2003; Jian et al., 2014). MCL can be isolated or associated with other craniofacial malformations such as hypotelorism, hypertelorism, holoprosencephaly, or lipoma of corpus callosum and other malformations of the body (Allam et al., 2011). The main craniofacial malformations associated with MCL are the holoprosencephaly spectrum and frontonasal dysplasia (DeMyer et al., 1963, 1964). Holoprosencephaly includes a spectrum of defects, from cyclopy with an alobar holoprosencephaly to mild bilateral cleft lip with lobar holoprosencephaly. Frontonasal dysplasia, with hypertelorism, is specially associated with the 0–14 cleft of the Tessier classification (Tessier, 1976).

Confusion still exists regarding the explanation and classification of MCL, despite some embryological theories having been advanced (DeMyer, 1975; Stark, 1954). Some authors suggest a theory of failure of the 2 medial nasal processes to fuse in the midline (DeMyer, 1975). Others support the theory of the failure of mesodermal migration at the primary palate (Veau, 1938; Stark, 1954). This failure of mesodermal penetration results in a breakdown in the median element of the prolabial element and then a median cleft of the upper lip. Based on the embryological approach, MCL could be secondary to the agenesis or hypoplasia of the median element of the prolabium, or secondary to a cleft of this element (Braithwaite and Watson, 1949; Millard and Williams,

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1968). On the other hand, the anatomical criteria for the median cleft lip and extent to adjacent structures are essential for surgical treatment (Veau, 1938).

Some cases of this type of cleft have been reported in the literature, but no studies have carried out a complete review of the literature. Here, we describe cases of MCL registered in 2 French tertiary centers over 25 years, and we systematically review of the literature. This study aimed to derive a comprehensive and useful classification of MCL from a large group of patients, in order to guide the management of these patients. The second objective was to calculate the correlation between clinical expression and associated malformations.

2. Material and methods

2.1. Chart review

A retrospective chart review of MCL was carried out from 1989 to 2015 (25-year period) in 2 French tertiary centers: the craniomaxillofacial surgery department of Lyon and the pediatric plastic surgery department of Montpellier. We collected epidemiological data (diagnostic age, ethnicity, and gender) and clinical data of the cleft and associated anomalies. Clinical data are reported in Fig. 1.

The size of the cleft lip was classified according to the following 3 categories: (1) minor forms, grouping normal lip and simple notch in the vermilion (i.e., cleft at the vermilion, with a height of no more than 6 mm above the edge of the lip); (2) incomplete forms, grouping clefts limited to the vermilion (clefts more than 6 mm deep, not exceeding the limit between red and white lip) and clefts reaching the white lip but not the full height of it; and (3) complete forms, in which the lip is clefting over its entire length.

The philtrum's involvement was classified into five categories: (1) normal philtrum; (2) fistula in philtrum; (3) enlargement of philtrum's ridges; (4) cleft in philtrum; and (5) absence of philtrum (in this case, unlike the cleft, the median part of the philtrum is missing).

The appearance of the columella was classified into 6 categories: normal, enlargement, bifidity, cleft, agenesis, and other columella malformation.

The upper labial frenulum was divided into 4 groups: normal frenulum, bifid frenulum, shortened frenulum, and agenesis.

The premaxilla was classified into 4 categories: normal premaxilla, notch (cleft in the premaxilla less than 6 mm high), cleft, and agenesis.

The associated malformations were cleft palate, hypo- or hypertelorism (intercanthale distance was measured on patients' pictures: it was considered normal when it was equal to the length of the palpebral fissure), holoprosencephaly, skin tags, abnormalities of corpus callosum, digital anomalies (poly or syndactyly), and other head or body malformations.

2.2. Review of the literature

This review included all PubMed publicly available articles describing patients with MCL.

2.2.1. Literature search strategy

The literature search was conducted using PubMed database in English and French languages using MESH terms “median cleft lip” and key words “median cleft lip” and “fente labiale médiane”. All papers found in the PubMed search were reviewed according to a standardized protocol, described below, up to December 2013 (Fig. 2).

2.2.2. Selection of papers

The same reviewer evaluated the abstracts of all of the articles for appropriateness to the study aim. All relevant articles on the MCL in English or French were selected for full-text review to determine whether they met eligibility criteria.

2.2.3. Eligibility criteria

Inclusion criteria were at least 1 patient with MCL described with adequate clinical description of the cleft, and article in English or French language.

Exclusion criteria were as follows: animal studies, nasofrontal dysplasia without median cleft lip, lateral cleft lip, median cleft of the inferior lip, other complex facial cleft, other malformation, holoprosencephaly without cases, dysmorphism without cleft lip, lack of data, no cases described, and same patient described in another article (duplicates).

2.2.4. Data extraction

Information concerning MCL was elicited. We collected the same data as for patients of our chart review: epidemiologic data,

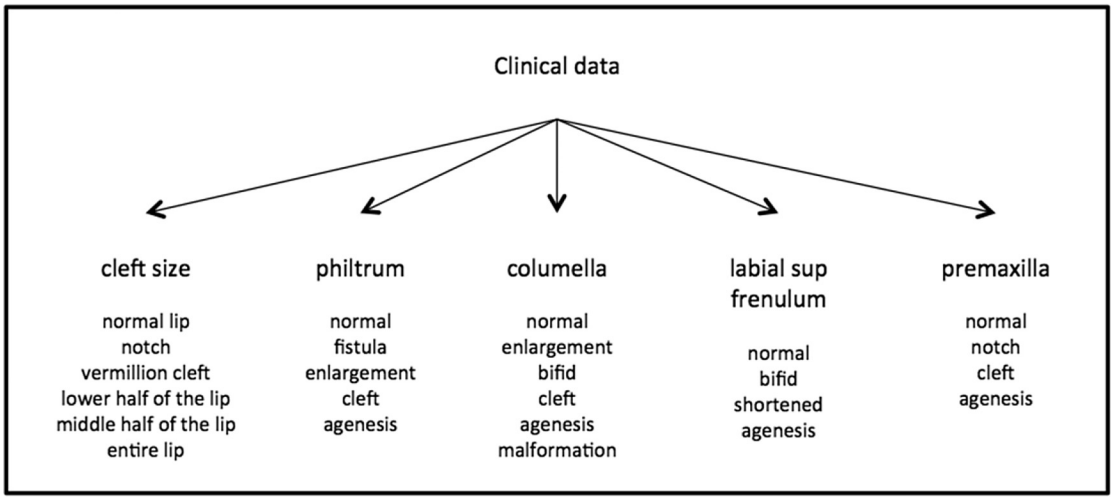

Fig. 1. Clinical characteristics of the median cleft lips recorded in our population of patients.