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ORIGINAL ARTICLE

Frequency of various craniofacial clefts observed in a single center during a period of 34 years

Authors' Contribution:
A–Study Design
B–Data Collection
C–Statistical Analysis
D–Data Interpretation
E–Manuscript Preparation
F–Literature Search
G–Funds Collection

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ABSTRACT:

Background: Orofacial clefts are the most common congenital anomalies of the head and neck.

Aim: The aim of this paper is to present own group of patients with different types of isolated orofacial clefts.

Material and method: A retrospective study presenting patients with different facial clefts was carried out in the group of 473 patients born in the city of Lodz.

Results: In the whole group there were: 434 patients with cleft lip and/or palate, 28 with nose clefts and 11 with rare facial clefts classified according to Tessier categorization.

Conclusion: In our group, the majority of cases comprised of typical cleft lip and/or palate. Nose clefts are not always a part of craniofacial clefts, but isolated nose clefts can occur and can be observed as midline defects or ala nasi anomaly. A simple system may be adequate for surgical repair, with the more detailed classification schemes important for further understanding of the disease.

KEYWORDS: classification, cleft, types

INTRODUCTION

Orofacial clefts are the most common congenital anomalies of the head and neck [1, 2]. The prevalence is estimated to be 1 per 500 to 2 500 births depending on the type of cleft, ancestry, geographic residential location, maternal age, prenatal exposures and socioeconomic status [1–6]. These malformations constitute a serious problem of both medical and social nature, mainly due to lifelong negative implications on health and well-being and the numbers of people affected [1].

There are various types of oro- and/or craniofacial cleft, that is why efforts put into their categorization are much needed. However, a universally accepted classification that fully encompasses, accurately describes and integrates all the various types of clefts, does not exist [5]. When composing a classification, different aspects of clefts should be taken into consideration. The Van der Meulen's categorization has embryological basis and the Tessier's classification has anatomical one [7, 8]. Tessier described a numeric classification for rare craniofacial clefts. He first emphasized that a fissure of the soft tissue corresponds, as a general rule, with a cleft of the bony structure. He introduced a cleft-numbering system around the orbit from 0 to 14 depending on its relationship to the zero line – the vertical midline cleft of the face. He also added number 30 to describe lower midline facial cleft (median mandibular cleft). His proposition is easy to understand and has become widely accepted because the recording of the malformations was simple and communication between observers was facilitated [9]. However, due to its descriptive character, even experienced surgeons have to consult a facial diagram to describe the location of a cleft properly [10].

The aim of this paper is to present our patient experiences with different types of isolated orofacial clefts and to comment upon existing classifications with some insights worth taking into consideration while planning the healthcare for children with facial anomalies.

MATERIAL AND METHODS

A retrospective study aiming to present our experience with patients with different types of isolated facial clefts was planned. Medical data of all patients with isolated facial clefts, who were born in Lodz city and were treated in our Clinic between years 1981–2015, were carefully studied. Children with cleft in the facial area with co-existing anomalies or other genetic syndromes or chromosomal abnormalities were excluded from this research. Each patient underwent a clinical examination and as a rule photographic documentation was carried out. Thorough medical history of each patient was recorded. A patient database was created on the basis of the medical data, ambulatory cards, hospital histories and photos. The collected data included: age, sex, type of cleft, affected side of the body and introduced or planned treatment. We divided our group of children with isolated facial clefts into three groups: isolated typical cleft lip and/or palate, isolated nose cleft and isolated rare cleft (rare clefts were numbered according to the Tessier's classification).

Lodz is the third-largest city in Poland. It is located in the central part of the country. The area of the city is estimated for 293.25 square kilometers and per November 2018 its population was 687 702 people. Our out-patient Clinic and hospital Department are the only plastic centers in the whole district so consequently our Department is the only taking care of kids with orofacial malformations. The study design was approved by the local Ethics Committee of the Medical University of Lodz (agreement No. RNN/234/19/KE) and was conducted in accordance with the principles outlined in the Declaration of Helsinki.

RESULTS

The study was carried out in the group of 473 patients with isolated orofacial clefts. In the analyzed group, there were 237 males

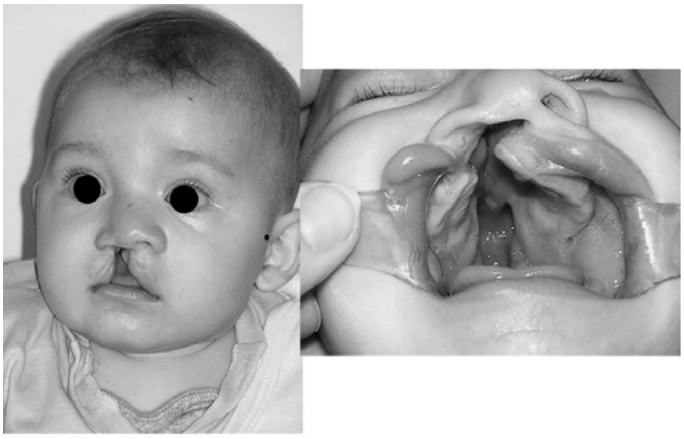

Fig. 1. A child with a typical right-sided clef of the lip and palate.

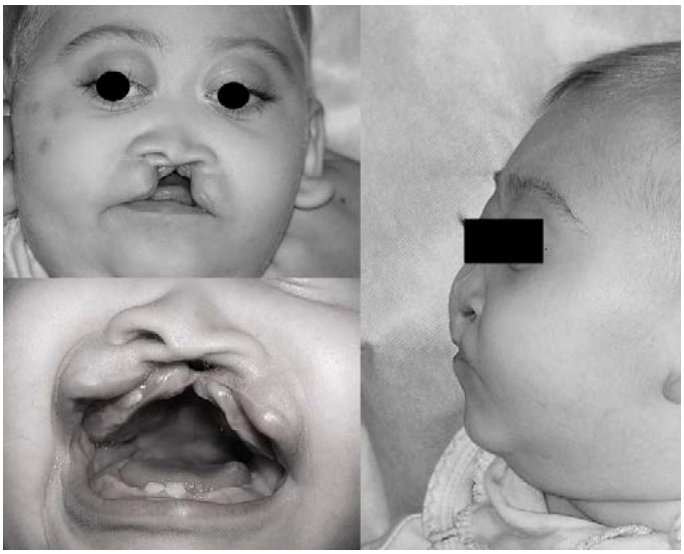

Fig. 2. A male with No 0 cleft according to Tessier. This anomaly known as midline cleft includes upper lip and nose in soft tissues; osseous characteristics include alveolar bone and/or nasal bones.

and 236 females. The patients' age, at the time of the first consultation, was from 3 weeks of age to 26 years (the mean age was 1 year and 8 months) (Tab. I.–III.). The majority of patients were children (<18 years of age), only one male and three females were above 18 (at the age of 22, 19, 20 and 26, respectively).

In the whole group, there were: 434 patients with typical isolated cleft lip and/or palate (91.8%), 28 with isolated nose clefts (5.9%) and 11 with isolated rare facial clefts (2.3%) (these rare clefts were classified according to the Tessier's categorization).

Among the patients with cleft lip and/or palate the most frequent condition was isolated cleft palate (181 children), followed by cleft lip and palate (156 children). The less frequent one was isolated cleft lip (97 children) (Fig. 1.). Unilateral cleft lip and palate was two times more common than bilateral malformation (105 vs. 51). When analyzing sex in the whole group of typical clefts it could be stated that the frequency of cleft lip and/or palate is equal between females and males (216 vs. 218). However, while examining the particular types of clefts it should be emphasized that isolated cleft palate is definitely more common in females than in males (112 vs. 69), while cleft lip and palate are more common in males than in females (96 vs. 60). The frequency of isolated cleft lip was comparable between male and female patients (53 vs. 44). In the group of 97 children with isolated cleft lip, left-sided cleft was two times more common than right-sided cleft (60 vs. 37). The same relation was observed in the group of patients with unilateral cleft lip and palate – the left side was affected in 70 children, while the right side in 35 children (Tab. I.).

In the group of 28 patients with isolated nose clefts there were 21 with isolated midline nose clefts and 7 with isolated ala nasi clefts. Among the patients with midline clefts, 15 had total nose malformations while 6 had a defect involving only the tip of the nose. In all patients with ala nasi cleft, the defect was unilateral, in 5 it was left-sided and in 2 right-sided. In both groups of patients with midline clefts and ala nasi clefts, the malformation was more frequent in females (12 with midline cleft and 5 with ala nasi cleft) than in males (9 with midline cleft and 2 with ala nasi cleft) (Tab. II.).

In the group of rare isolated facial clefts there were 11 patients and all of them were classified according to the most common Tessier's categorization describing rare clefts. Two patients presented defects consistent with type 0 cleft, two with type 4 and three with type 6 (one was bilateral), also there were single patients with cleft type 2, 3 and 7 (Fig. 2.). One male had bilateral anomaly but on the right side he presented an anomaly compatible to cleft type 4 and on the left side, cleft type 6. In this group there were 8 males and 3 females. Four patients had a right-sided cleft, 3 left-sided, 2 clefts were medial and, as mentioned above, 2 were bilateral (Tab. III.).

DISCUSSION

Face clefts comprise a large heterogeneous fraction of all human birth defects and are notable for their significant lifelong morbidity and multifactorial etiology [11]. The group of facial anomalies commonly is divided into two groups: typical orofacial clefts (isolated cleft lip, cleft lip and palate and isolated cleft palate) and atypical clefts (median, transversal, oblique as classified by Tessier). Both typical and atypical clefts can occur as an isolated anomaly, as part of a sequence of the primary defect or as a multiple congenital anomaly [12]. In this paper we focused on isolated anomalies in the face area and collected data for the group of 473 patients who were born in Lodz with isolated primary facial clefts.

Eppley et al. and Fearon stated that complete classification of facial clefts with worldwide acceptance does not exist [5, 10]. We agree with the above-mentioned authors and, furthermore, we assume that creating such a categorization is not possible. Different medical specialists focus on different aspects of birth anomalies, which explains why there are several different facial cleft classifications. There are mainly morphological classifications, embryological systems, genetic categorizations and anatomical ones [12]. There also exists a recent review of classification systems carried out in 1981 by the American Cleft Palate Association [12].

Fearon in 2008 proposed a regional craniofacial cleft classification, which is based on Tessier's categorization and its numbers. However, Fearon divided all of Tessier clefts into only four groups according to surgical planning [10]. This surgical classification, very importantly, includes only true clefts and subdivides them into four types: Type I: midline facial clefts (formerly 0–14 and 30); Type II: paramedian facial clefts (formerly 1–13 and 2–12); Type III: orbital facial clefts (formerly 3–11, 4–10, and 5–9); and

Tab. I. The distribution of cleft lip and/or palate in our patients.

TYPE OF CLEFT	NO OF PATIENTS	MALES	FEMALES	UNILATERAL ANOMALY	BILATERAL ANOMALY
CP	181	69	112	-	-
CLP	156	96	60	R – 35 L – 70	51
CL	97	53	44	R – 37 L – 60	-
Sum	434	218	216	202	51
Mean age of males: 2.45 months; SD = 1.23; median = 2.5 months					
Mean age of females: 3.5 months; SD = 1.61; median = 3 months					

CP – cleft palate; CLP – cleft lip and palate; CL – cleft lip; R – right side; L – left side

Tab. II. The distribution of isolated nose clefts in our patients.

TYPE OF NOSE CLEFT	MALES	AGE	FEMALES	AGE	SUM
Midline cleft	9	From 1 to 22 years	12	From 6 months to 26 years	21
Ala nasi cleft	2	Both 1 year	5	From 5 years to 20 years	7
Mean age of males: 8.13 years; SD = 7.79; median = 8 years					
Mean age of females: 10.7 years; SD = 7.39; median = 7 years					

Tab. III. The distribution of rare facial clefts in our patients described by Tessier's Classification.

NO OF CLEFT	NO OF PATIENTS	SEX	BODY SIDE	AGE
0	2	M	Median	2 months; 11 months
2	1	F	Left	6 months
3	1	M	Left	10 months
4	2	M	Right – 1 Left – 1	11 months; 18 years
6	3	F – 2 M – 1	Right – 2 Bilateral – 1	4 months; 5 months; 4 years
7	1	M	Right	6 months
4 and 6	1	M	Bilateral	2 months
Sum	11	M – 8 F – 3	–	Mean 29 months /2 years and 5 months/
Median age 6 months; SD = 63.29				

Type IV: lateral facial clefts (formerly 6, 7, and 8) [10]. The midline facial clefts may affect the forehead, midface and maxilla, central upper lip or lower lip and mandible. The paramedian facial clefts arise off the midline of the face, but remain medial to the orbit, and may arise anywhere from the frontal bone, down through the nose into the upper lip. The orbital facial clefts may extend anywhere through the orbit and may course into the frontal bone and down through the upper lip. Finally, the lateral facial clefts include clefts running through the oral commissure and also may include any bony clefts occurring lateral to the orbit [10]. Fearon stated that operative approaches to craniofacial clefts are more regionally based and do not necessarily differ from one number to another [10]. For example, the midline facial clefts typically rely on telorbitism corrections, median clefts entail unilateral nasal and lacrimal repair techniques, and orbital clefts require eyelid flap procedures to achieve adequate globe coverage. All clefts extending through the lips, including the lateral clefts, require techniques borrowed from cleft lip and palate corrections, and when clefts involve the underlying bone, they are usually grafted secondarily [10].

As surgeons we agree with the proposition made by Fearon. It is worth to have a short classification of facial anomalies as almost in all cases of such children operative treatment will be necessary to perform at some point of their lives. Such a system facilitates surgeons to describe an observed cleft in such a way that enables others to easily visualize the location and recognize a surgical pattern. We would like to add to Fearon's classification our suggestion to encompass isolated cleft palate as type I and cleft lip as type II cleft. Additionally, specialists taking care of children with clefts should remember that in type I – anomaly such as isolated midline nose cleft can exist and in type II – isolated ala nasi cleft can appear. This was not previously stated by Fearon.

It seems that anatomical classifications are the most logical ones, since anatomy has not changed over the years and doctors of all specialties are familiar with the classic anatomy, which is why communication between different centers is easy and understandable. Anatomical description is precise, and we know exactly which structures are impaired and which need to be reconstructed. However, from the surgical point of view, the most practical classifications are the simple and short ones, like the amended Fearon's classification proposed by us. The key fact is that surgical methods are similar in some groups of anomalies depending on the facial area that they involve. Usually reconstruction of muscle is necessary, also restoration of skin continuity is fundamental, and sometimes bone grafting is needed.

CONCLUSIONS

To sum up, in the group of children with isolated face clefts, the majority of cases constantly comprise of the typical cleft lip and/or palate. The rarest form of defect is the atypical facial cleft cat-

egorized according to the Tessier's classification. It should be pointed out that nose clefts as isolated forms can occur, not being always a part of other craniofacial clefts. Nose anomalies can be divided into midline defect or ala nasi malformation. The classification schemes are often complex, when perhaps a simplified scheme would be presently more adequate for the surgical repair.

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