Original Article

Epidemiological profile of patients with orofacial cleft treated by a reference surgical team in the State of Amazonas, Brazil

Perfil epidemiológico dos pacientes portadores de fissuras labiopalatinas atendidos por equipe cirúrgica de referência no Estado do Amazonas

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■ **ABSTRACT**

Introduction: Orofacial clefts are congenital malformations with an estimated occurrence of 1:650 births in Brazil. The most widely adopted classification system in that country is the method developed by Spina, and cheiloplasty and palatoplasty are the main surgeries performed. **Methods:** This was a retrospective descriptive study using data collected from the Smile Train Express organization regarding patients with orofacial clefts treated by a reference surgical team between March 1, 2014 and December 1, 2016. **Results:** A total of 477 patients were identified, predominantly male and in the first two years of life at admission. The most prevalent type of malformation was left unilateral transforamen cleft. The most frequent surgical treatment was cheiloplasty. **Conclusions:** The epidemiological pattern is consistent with the findings described in the national literature.

Keywords: Orofacial cleft; Cleft lip; Maxillofacial abnormalities; Epidemiology; Classification.

■ **RESUMO**

Introdução: As fissuras labiopalatinas são malformações congênitas e, no Brasil, estima-se a ocorrência de 1:650 nascimentos. A classificação adotada é a de Spina. A queiloplastia e a palatoplastia são as principais cirurgias executadas. **Métodos:** Estudo retrospectivo descritivo com obtenção de dados a partir do sistema *Smile Train* Express referente a pacientes com fissura labiopalatina atendidos por equipe cirúrgica de referência entre 1 de março de 2014 e 1 de dezembro de 2016. **Resultados:** Foram identificados 477 pacientes, predominando o sexo masculino e os dois primeiros anos de vida na admissão. A fissura mais prevalente foi transforame e unilateral esquerda. O tratamento cirúrgico mais frequente foi a queiloplastia. **Conclusões:** O padrão epidemiológico está em consonância com a literatura nacional.

Descritores: Fissura palatina; Fenda labial; Anormalidades maxilofaciais; Epidemiologia; Classificação.

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INTRODUCTION

Orofacial clefts (OFCs) are congenital malformations that occur between the 4th and 12th weeks of the embryonic period owing to lack of fusion of the maxillary and mid-nasal processes[1]. These malformations are attributed to genetic and environmental factors, which can act alone or in combination. More than half of individuals with a cleft have relatives who also have the malformation. The most frequently associated environmental factors are: maternal stress; medication use; ionizing radiation; nutritional, toxic, and infectious agents; and smoking during fetal development[2].

OFCs are the most frequent facial malformations, with an estimated occurrence on the order of 1 in 650 births in Brazil[2,3]. Most individuals with OFC do not present with any other abnormalities (non-syndromic OFC), but a significant proportion (30 to 50%) manifest other malformations and may exhibit a particular syndrome (syndromic OFC)[4].

Several proposed classifications have been developed that are based on morphological and/or embryonic aspects. The most widely used in Brazil is the Spina Classification, divided into four groups: incisional pre-foramen cleft or, simply, cleft lip (CL), incisional post-foramen cleft or palatine fissures (PF), transforamen cleft or orofacial cleft (OFC), and rare clefts of the face (Figure 1).

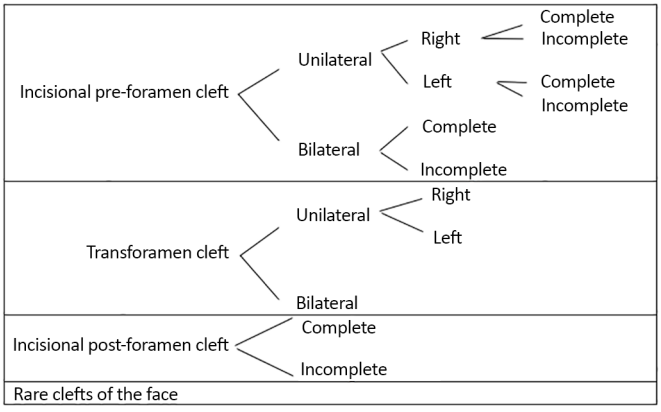

Figure 1. Classification of the types of orofacial cleft, according to Spina.

The Spina Classification is based on the embryological theory that recognizes the independent developmental mechanisms of the anterior structures (originating from the primary palate) and those posterior to the incisive foramen (originating from the secondary palate), with this anatomical reference point chosen for this particular classification. The current limited knowledge in the field regarding the etiology of OFC hinders even the descriptions and distinctions of the various forms of presentation of these malformations[5].

OFCs have a significant impact on speech, hearing, and appearance, as well as a major influence on the health and social integration of a patient, owing not only to morbidity, but mainly to emotional disturbances, stigmatization, and social exclusion, as these malformations interfere with the development of self-esteem, interpersonal relations, and incorporation into the socioeconomic and cultural environment[6].

It is important for the individual with a cleft to undergo early follow-up, performed by a multiprofessional team, with an interdisciplinary approach, as well as integral treatment, continuing from birth to adulthood, providing the individual with the necessary tools for adjustment to society.

Cheiloplasty and palatoplasty are the initial reparative plastic surgeries performed during the long and complex treatment of OFC. For the child who is born with an OFC, reconstructive surgery is a challenge not only aesthetically, but mainly functionally. Cheiloplasty consists of reconstructive surgery of the cleft lip, and palatoplasty is the reconstruction of the cleft palate[7].

The treatment protocol most often used, including by the team currently working in Manaus, is closure of the lip at 3 months of age, and closure of the palate, in a single procedure, at 1 year of age. Alveolar bone graft is performed in patients between 7 years and 9 years of age, when the canine teeth are close to eruption. Orthognathic surgery is performed in patients older than 15 years of age. The final surgery of the patient with a cleft is a secondary rhinoplasty, to correct any residual nasal deformity[8].

In 2014, the State Health Secretariat of Amazonas (SUSAM) signed an agreement with *Smile Train Express*[9], an international non-governmental organization, founded in 1999, which aims to provide comprehensive treatment to patients with OFCs around the world. With this partnership, the surgical team became a reference for patients diagnosed with OFCs in the State of Amazonas. Surgeries for OFC are performed in the Dr. Fajardo Children's Hospital; in addition, there are corrective surgeries to treat this malformation performed as part of a yearly communal work effort in the city of Manaus.

OBJECTIVE

The purpose of this study was to present the main epidemiological features of OFC in the population of patients treated by a reference surgical team for OFC in the State of Amazonas.

METHODS

This was a retrospective descriptive study performed at the Dr. Fajardo Children's Hospital, in Manaus, AM, with data acquired from the *Smile Train Express*[9] organization regarding patients with a diagnosed

OFC treated and operated on by a reference surgical team in Amazonas between March 1, 2014 and December 1, 2016. The Research Ethics Committee of the Adriano Jorge Hospital Foundation approved this research under opinion number 1,899,716/2017.

Data from a total of 477 patients were collected, with the following analysis variables: cleft types according to the Spina Classification[5], sex, origin, age, and surgical condition at hospital admission, as well as the surgical treatment performed. Regarding surgical condition at admission, any history of previous surgical treatment was analyzed and, in case of previous surgery, an attempt was made to identify the type of surgery performed.

RESULTS

During the study period, 477 patients with OFC who underwent surgery performed by the reference surgical team were identified. There was a slight male predominance of 52%.

Table 1 shows the distribution of these patients regarding age at admission to the service. The minimum age was 17 days and the maximum age was 47 years.

Table 1. Distribution of patients with cleft according to age group at admission to referral service in the State of Amazonas.

Age (years)	n	%
0 to 1	167	35
2 to 4	82	17
5 to 9	95	20
10 to 14	67	14
15 to 24	48	10
25 to 40	16	3.5
Over 40	2	0.5
Total	477	100

Almost half of the patients studied came from the interior of Amazonas, as shown in Table 2.

Table 2. Distribution of patients according to region of origin.

Origin	n	%
Capital	241	51
Countryside	236	49
Total	477	100

Regarding the type of cleft, the most prevalent was incisive transforamen (60%), followed by incisive pre-foramen fissure (24%), and, finally, incisive foramen cleft (15%). There were five cases in which patients had pre- and post-foramen (1%) fissures, concomitantly. There was a predominance of left side clefts. Table 3 shows that the types of OFC were regrouped into: pre-foramen cleft (complete and incomplete/right, left, and bilateral), 113 cases; post-foramen cleft (complete and incomplete), 68 cases; pre- and post-foramen cleft (right and left), 5 cases; and transforamen cleft (right, left, and bilateral), 291 cases. All transforamen clefts affect the incisive foramen and all pre- and post-foramen clefts do not.

As shown in Figure 2, half of the patients with a cleft had undergone previous surgery, with cheiloplasty (65%) the most frequent, followed by cheiloplasty and palatoplasty (25%) and, finally, palatoplasty (10%).

The surgical treatments performed in these patients are detailed in Figure 3, with cheiloplasty being the most frequent (35%), followed by palatoplasty (27.5%). Procedures for correction of previous surgeries (secondary surgeries) were also performed, and 11.5% of patients underwent simultaneous surgeries ("combo").

DISCUSSION

During the study period, 477 patients were attended to by a reference surgical team in the State, and a total of 495 procedures were performed.

The present study shows that, among the analyzed patients, OFCs were more frequent among the male sex (52%); and although the difference was not significant, this finding is consistent with most national statistics[10-14].

The most frequent age at admission of patients in the referral service was between one month and two years (35%), which was considered adequate; this time frame is compatible with the chronology of primary lip and palate surgeries and allows early treatment, especially in cases of bilateral transforamen cleft, since the structures involved and the extent of the lesion result in greater complexity, requiring a longer duration of treatment[15].

Another study finding was that an approximately equal number of individuals with OFC came from the countryside and the capital of the State of Amazonas. It is necessary to take into account that at various times, communal chirurgical efforts were undertaken in some municipalities, such as Tefé, Parintins, Itacoatiara, Tabatinga, Eirunepé, and Benjamin Constant, and that most municipalities have geographical peculiarities that result in difficulties in access to specialized services for the population. Therefore, it is not possible to state that the numbers obtained in this study represent the reality for the entire state.

In most published national studies[12-16], the percentage of individuals with transforamen cleft has been higher when compared to cleft lip or cleft palate

Table 3. Distribution of patients with orofacial cleft, according to type, extent, and lateralit.

Types of Clefts	n	%	Cleft Extension	n	%	Cleft Laterality	n	%
Pre-foramen	113	24	Complete	27	25	Right	44	38
			Incomplete	86	75	Left	53	47
						Bilateral	16	15
			Total	113	100	Total	113	100
Post-foramen	68	15	Complete	11	15	N/A		
			Incomplete	57	85			
			Total	68	100			
Transforamen	291	60	N/A			Right	69	24
						Left	125	43
						Bilateral	97	33
						Total	291	100
Pre- and post-foramen	5	1	N/A			Right	3	60
						Left	2	40
						Total	5	100
Total	477	100						

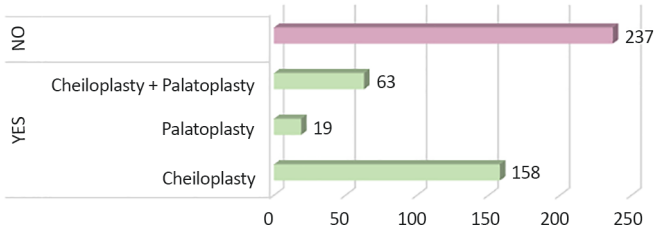

Figure 2. Distribution of clefts according to surgical condition at admission to the referral service in the State of Amazonas.

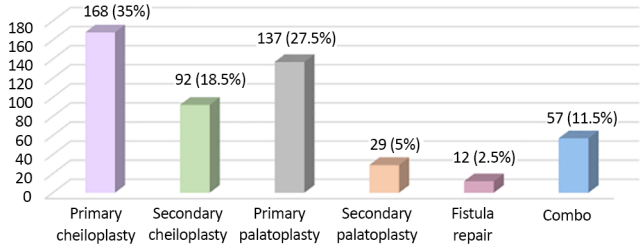

Figure 3. Distribution of surgeries performed in patients with orofacial cleft.

alone, as in our case series (60% *vs.* 24% pre-foramen *vs.* 15% post-foramen). Spina's rank[5] allows the identification of mixed forms (pre-foramen and post-foramen clefts in the same carrier), which occur during different periods of embryological development. We found this type of fissure in 1% of cases in our study.

In the present study, the OFCs were unilateral in most cases, with the left side more commonly affected, as described in the national literature[11,13,14]. The reason for this predilection is not understood. Patterns of laterality defects are known to be observed in various types of anomalies. Groups of genes expressed asymmetrically during the early stages of embryonic development may contribute to this preference, but this hypothesis has not yet been demonstrated by any study[17].

According to the anatomical extension, clefts can be subclassified as complete or incomplete, with the rupture or non-rupture of the incisive foramen as a reference. Such subclassification is only used for pre- or post-foramen clefts[5]. This understanding is necessary for the elaboration of a therapeutic plan, because the greater the extent of the fissure, the greater the functional impairment and, therefore, the greater the resources required for the total recovery of the patient.

In the present study, incomplete clefts (80%) predominated, among those classified as pre- or post-foramen, as in the findings of Gardenal *et al.*[13]. On the other hand, if we add the transforamen clefts to the set, we can observe a predominance of complete fissures (70%), since all the transforamen clefts affect the incisive foramen.

Cheiloplasty and palatoplasty are the first restorative surgeries performed during the long process of treatment of OFCs. In the studies of Alonso *et al.*[10], Gardenal *et al.*[13], and Moura[18], the majority of patients with OFCs had not undergone a previous surgical procedure

to correct this malformation at the time of admission to the service (70%, 80%, and 90%, respectively).

In our study, we did not obtain the same results, since 49.5% of the patients had already undergone some type of primary corrective surgery. According to Gardenal *et al.*[13], this condition is considered unfavorable since multidisciplinary care is usually not delivered after the surgical steps, particularly following cheiloplasty.

Finally, the main surgical modalities (cheiloplasty and palatoplasty) for the treatment of OFC were subdivided into primary and secondary. We observed that, in both cases, cheiloplasty was the procedure with the highest prevalence.

CONCLUSION

Based on the results of this study, we infer that OFCs are more frequent in the male sex, with a predominance in the first two years of life at the time of admission to the surgical service and an equal distribution in origin in these patients. Transforamen and left unilateral cleft were the most prevalent types. Among those classified as pre- or post-foramen, incomplete clefts predominated. Half of the study patients had undergone previous surgery, and cheiloplasty was the most commonly performed surgery, both as a primary and secondary procedure.

COLLABORATIONS

KMGA Analysis and/or interpretation of data; statistical analyses; final approval of the manuscript; conception and design of the study; completion of surgeries and/or experiments; writing the manuscript or critical review of its contents.

AJAS Analysis and/or interpretation of data; statistical analyses; final approval of the manuscript.

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