ORIGINAL RESEARCH

Cleft lip and palate in a Brazilian subpopulation

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ABSTRACT

Background: This work aimed to access the profile of cleft lip and palate patients of a sub-population in Mid-West Brazil.

Materials & Methods: Research was carried out through a cross-sectional study at the Craniofacial Rehabilitation Center of the University General Hospital of the University of Cuiabá, Mato Grosso, Brazil. Variables related to oral cleft type, gender, race, age and presence or absence of associated congenital anomalies or syndromes were analyzed.

Results: 313 patients treated at the institution from 2004 to 2007 were recruited. There were 54% male and 46% female patients with the mean age of 11.4 years. Cleft lip and palate was the most prevalent alteration in 49.6% of cases. Caucasians were the most affected in 54.6% of cases. 6.4% of patients had other anomalies or syndromes associated with cleft.

Conclusion: More comprehensive surveys should be conducted in order to supply the lack of data on the occurrence and determinants of oral clefts in this region.

Key Words: Cleft lip, Cleft palate, Patients with special needs, Dentistry.

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Introduction:

Congenital anomalies affect about 5% of all newborns around the world[1]. Among the facial congenital anomalies, cleft lip and cleft palate are considered the most frequent[2-4]. These are the result of failure in the embrionary development or maturation processes, which usually take place between the fourth and the eight week of pregnancy[5-6].

Even though it was first described almost two centuries ago; its etiology remains not completely understood. Most of the times, its occurrence is attributed to the Multi-factorial Theory, which can be summarized as the interaction of genetic and environmental factors[3-5, 7-8].

The prevalence of the cleft lip and cleft palate varies according to the geographical location and ethnic group being studied. It is more commonly found in Asian descents as opposed to other groups. However, it is more common among Caucasian when compared to afro-descent[9-11]. In regards to gender, cleft lip, with or without the occurrence of cleft palate, is more frequent

in males, while the occurrence of isolated cleft palate without the presence of cleft lip is more common in females[7, 12].

Throughout the world, the incidence of cleft lip and cleft palate vary from 1.0:1000 to 2.29:1000. A greatest incidence takes place in Bolivia (2.29:1000), followed by Japan (1.60:1000), Paraguay (1.49:1000), Germany (1.39:1000), China (1.36:1000), Holland (1.35:1000), Norway (1.33:1000), Denmark (1.30:1000) and an average of (1.0:1000) for South America[9,13]. Even tough there are no accurate statistics to determine the occurrence of cleft lip and palate in Brazil, it is estimated that the frequency of cleft lip, associated or not with cleft palate, reaches 1:1000 newly born babies, varying from 0.7 to 1.3[14], .

In Brazil, the geographic distribution of the health centers which are part of the network of Craniofacial Deformity Treatment Reference Centers is mostly concentrated in the Southeastern region, specially in the state of São Paulo, and an insufficient number of health centers in the North, Northeast and Mid-West regions, generating a flow of patients in search of treatment in locations far from their homes, which, for this reason, many times discontinue treatment[15].

In November of 2004, through an agreement between the Health Department of the State of Mato Grosso, the Craniofacial Rehabilitation Center in the University General Hospital of the University of Cuiabá (HGU-UNIC), it became a state reference in the treatment of patients with cleft lip and/or cleft palate. The service is pioneer in the State and counts with a team of several professionals in different specialties offering rehabilitating treatment to the patients.

Since there is almost no data on patients with cleft lip and/or cleft palate in isolated states of Brazil, the work at hand presents the profile of patients treated at the Craniofacial Rehabilitation Center in the University General Hospital of the University of Cuiabá (HGU-UNIC).

Material and Methods

This is a study of transverse cutting involving the patients received at the HGU-UNIC Craniofacial Rehabilitation Center in the period between November 1st, 2004 and February 28th, 2007.

The archives, files and records of all the patients received at the Cleft lip and palate Rehabilitation Center in the University General Hospital of the University of Cuiabá (HGU-UNIC) were analyzed in the given period, according to the following variables: gender, age, color of skin/race, type of cleft and the concomitant occurrence of other congenital defects. Patients with incomplete data were excluded from the study.

The defects were classified as cleft lip, cleft palate and, when they occur simultaneously, as cleft lip and palate. As for the color of skin/race classification, the criteria used by the Center is the one in which the doctor indicates whether the patient is are white, mixed, black or Native American.

After collecting the data, descriptive statistical analysis was performed to establish the relative and average frequencies. Possible association between the variables were evaluated using the chi-square test and considered statistically meaningful when $p < 0.05$.

The study was approved by the Ethics in Research Committee of the Julio Müller Hospital, at the Federal University of Mato Grosso.

Results

From a total of 324 patients assisted by the Craniofacial Rehabilitation Center (HGU-UNIC) during the research, 11 were excluded from the study for incomplete data; leaving a total 313 patients to be evaluated.

The average age was 11.4 years, varying from 4 days to 72 year-old patients. Table 1 shows the patients participating in the study according to the gender and age group.

From the 313 patients, 169 (54.0%) were males and 144 (46.0%) were females (M/F ratio of 1.2:1), without a meaningful statistic difference between genders ($p = 0.158$).

The distribution of patients according to the color of skin/race can be observed on Table 2. The lip and palate clefts were more frequent on white individuals than other color of skin/races (54.6%), with meaningful statistic difference ($p < 0.001$).

The lip and palate clefts were the most frequent (49.5%), followed by the cleft lip and cleft palates each with 25.2% of the cases. The prevalence of the isolated lip and palate clefts showed a small difference between

Table 1: Distribution of patients by gender and age group.						
Age Group (years)	**Gender**				**Total**	
	Male		**Female**			
	n	**(%)**	**n**	**(%)**	**n**	**(%)**
00 to 10	112	(66.30)	85	(59.00)	197	(62.90)
11 to 20	28	(16.60)	23	(16.00)	51	(16.30)
21 to 30	19	(11.20)	19	(13.20)	38	(12.10)
31 to 40	3	(1.80)	10	(6.90)	13	(4.20)
41 to 50	4	(2.40)	4	(2.80)	8	(2.60)
51 to 60	2	(1.20)	1	(0.70)	3	(1.00)
Above 60	1	(0.60)	2	(1.40)	3	(1.00)
Total	**169**	**(100.00)**	**144**	**(100.00)**	**313**	**(100.00)**

Table 2. Distribution of patients according to race.			
Race	**n**	**(%)**	**Chi-Square**
White	171	(54.60)	χ^2= 182.425 p < 0.001
Mixed	81	(25.90)	
Black	54	(17.30)	
Native American	7	(2.20)	
Total	**313**	**(100.00)**	

Table 3. Occurrence of different types of cleft in relation to gender.							
Kind of Cleft	**Male**		**Female**		**Total**		**Chi-Square**
	n	**(%)**	**n**	**(%)**	**n**	**(%)**	
Lip	45	(57.00)	34	(43.00)	79	(25.20)	χ^2= 1.532 p=0.216*
Palate	32	(40.50)	47	(59.50)	79	(25.20)	χ^2= 2.848 p=0.091*
Lip & Palate	92	(59.40)	63	(40.60)	155	(49.60)	χ^2= 5.426 p=0.020**
Total	**169**	**(54.00)**	**144**	**(46.00)**	**313**	**(100.00)**	
*Not Meaningful Difference. **Meaningful Difference							

Table 4. Occurrence of different types of oral cleft in relation to race.											
Type of Cleft	**White**		**Mixed**		**Black**		**Native American**		**Total**		**Chi-Square**
	n	(%)	n	(%)	n	(%)	n	(%)	n	(%)	
Lip	47	(59.50)	23	(29.10)	08	(10.10)	01	(1.30)	79	(25.20)	χ^2= 77.439 p<0.001**
Palate	43	(54.40)	19	(24.10)	16	(20.30)	01	(1.30)	79	(25.20)	
Lip and Palate	81	(52.30)	39	(25.20)	30	(19.40)	05	(3.20)	155	(49.60)	
Total	171	(54.60)	81	(25.90)	54	(17.30)	07	(2.20)	313	(100.00)	
*No significant difference. ** Significant difference											

genders, but not enough to be considered statistically significant (p = 0.158). However, the prevalence of cleft lip and palate together was 1.46 times greater on males when compared to females (Table 3), presenting a statistically meaningful difference (p= 0.020).

As far as the association to other congenital defects along with the oral clefts, only 20 patients (6.4%) had other birth defects, where 9 of them presented syndromes and 11 showed smaller defects. Among the patients with syndromes or recognizable patterns, the Pierre Robin Sequence was the most prevalent appearing in 3 cases (33.3%); other detected syndromes were a single case of each of the following: the Turner Syndrome, the Moebius Syndrome, the Appert Syndrome, the Oral-Facial-Digital Syndrome and Digital Fibroma. The most common oral cleft among the patients was the cleft palate, with 7 cases (77.8%), the cleft lip and the cleft lip and palate occurred in the patients with the Oral-Facial-Digital Syndrome and the Turner Syndrome respectively. Out of the 11 cases with other malformations, 2 patients presented more than one, revealing 15 alterations, the most frequent, with 3 cases (20%) was cardiac murmur, followed by hypertelorism and hearing problems, each present in 2 cases (13.3%). The other malformations took place once each, they were: mental deficiency, microcephaly, macrocrany, small stature, scoliosis, hemophilia, congenital rubella and strabismus. The cleft lip and palate was the most common among the patients with other abnormalities, with 4 cases (36.4%), the cleft palate took place in 3 cases (27.3%).

While seeking the relation between the cleft lip and/or cleft palate and the different color of skin/races (Table 4), it was observed that the white race presented the largest number of cases of cleft lips (59.5%), cleft palates (54.4%), and cleft lip and palate (52.3%). The mixed race comes second with 29.1% of the cases of cleft lip, 24.1% of cleft palate, and 25.2% of cleft lip and palate cases. The Native American and black races showed the smallest number of occurrences. The chi-square test shows statistically meaningful differences ($p<0.001$) between the races and the occurrence of cleft lip and palate when analyzed separately.

Discussion

Even though there is a large number of published epidemiological studies about oral clefts around the world, still there are very few studies done in Brazil. This is the first research done in the State of Mato Grosso. Moreover, such studies offer an understanding about the occurrence of oral clefts in different population groups, creating subsidies so that the public administration may elaborate prevention and assistance strategies for patients suffering from oral clefts and their families.

The age of the patients assisted during the study averaged 11.4 years (varying from 4 day-old newborns to 72 year-olds), much higher than the average found by Freitas et al.[10,] of 2.8 years (varying from 6 days to 49 years). This age was also much higher than ideal[13, 15-16] to start treatment, which should begin at pregnancy when the cleft is diagnosed and right after birth, as soon as possible. It is important to emphasize that 37.1% of the patients started their rehabilitation at the center after they were 10 years old. Since the cleft lip and palate affects the patients' aesthetics, their capacity to communicate and the ability to feed themselves, the introduction of the rehabilitation center in Mato Grosso allowed treatment to people who would otherwise be left unattended.

The cleft lip and palate was the most prevalent, with 49.6% of the cases, followed by the cleft palate and the cleft lip, each with the same percentage of 25.2% of the cases, matching other studies[10-11,17-20].

While the occurrence of oral clefts was predominant on male patients, when comparing numbers from other studies[7, 10-12, 17-20], the difference was not found to be significantly different in this study.

The male gender predominated on the cleft lip and cleft lip and palate, while the females were predominantly affected by cleft palate, in accordance with existing data[10-11,17], however, the cleft palate did not show statistically meaningful difference between genders, this only occurred with cleft lip and palate, as found by Martelli-Junior et al.[19].

Even though the white color of skin/race represented 54.6% of all the patients in the study, this number was less than what was found by Martelli-Junior et al.[19] (85.7%) and Freitas et al.[10] (80.0%). These results, however, conflict with other studies[9-11] that showed the yellow and native american to be the most affected by cleft lip and palate. One explanation to these differences

could be the large mixture of races that took place amongst the Brazilian population and the concentration of certain races on the different regions of the country.

The occurrence of oral cleft in patients with syndromes or other malformations occurred in 6.4% of the cases, and the cleft palate was present in 77.8% of the cases; the sequence of Pierre Robin represented most of the cases (33.3%). Existing literature makes reference to the existence of an association between clefts and syndromes, at the proportion of 1.4 to 18% of total cases. Al Omari and Al-Omari[17] verified an 18% association rate, McLeod et al.[18] verified 14%, and González et al.[20] verified 1.44%.

Conclusion

It is important to emphasize the importance of treating the patients affected by cleft lip and palate soon after birth and maintaining the treatment throughout their lives, including multiple aspects of their existences, seeking full social integration. The decentralization of rehabilitation centers tends to decrease the government costs in transportation of patients to perform treatment; it is also likely to improve chances that the patients will not abandon treatment and will obviously bring easier access to patients. The high average age of the patients is probably related to the difficulty to reach rehabilitation center at distant locations. Since there are few publications on the occurrence of cleft lip and palate, we would like to emphasize the importance and the need for more studies, seeking to find the causes and determinant factors of these malformations, as well as broader research in the State of Mato Grosso, given the lack of data in the region.

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