Epidemiological profile of children and adolescents with oral cleft treated at a referral center in Curitiba, PR, Brazil

Perfil epidemiológico da criança e do adolescente com fissura oral atendidos em um centro de referência em Curitiba, PR, Brasil

Introduction: Oral cleft is the second major cause of congenital anomalies and represents a major craniofacial alteration in live births. The objective of this study was to analyze the epidemiological data collected from the Center for Comprehensive Care to Individuals with Cleft Lip and Palate in the period from January 2011 to December 2014.

Methods: This retrospective study evaluated 1,262 medical records of patients with an oral cleft. After applying the inclusion and exclusion criteria, 52.7% of the medical records were included in the study.

Results: Among the 666 medical records, 57.4% were of male patients and 42.6% were of female patients. Of these, 34.8% of the patients had a trans-foramen cleft, 27.2% had a pre-foramen cleft, 25.8% had a post-foramen cleft, and 12.2% had another type of cleft. Patients from Curitiba and the metropolitan region constituted 36.6% of the cases, and patients from rural areas of Paraná represented 61% of the visits to the care center. The median age at the first visit of the patients from Curitiba/metropolitan region and rural areas of Paraná was 1 and 2 months, respectively. The first surgery was performed at the care center at the age of 6 months in patients from Curitiba and metropolitan region and 7 months in patients from rural areas of Paraná.

Conclusion: There was a predominance of boys and a higher prevalence of incomplete post-foramen clefts in the total population. Despite the long distance to the care center, children from rural areas of Paraná underwent the correction surgery and were treated at the referral center with an age difference of only 1 month compared with patients who lived in Curitiba, where the care center is located.

Keywords: Epidemiology; Cleft lip; Reconstructive surgery.
INTRODUCTION

An oral cleft (OC) develops unevenly on the face during the embryonic and fetal periods and is the second major cause of congenital anomalies in live births. OC is clinically characterized by nonclosure of the lip, palate, or both and affects approximately 1 in 700 live births. In Brazil, the exact number of subjects with OCs is unknown, but it is estimated that 180,000 individuals are affected by this anomaly, and mortality in the first year of life is approximately 35%. Approximately 10% of the cases have other associated anomalies, characterizing different syndromes.

The Center for Comprehensive Care to Individuals with Cleft Lip and Palate (Centro de Atenção Integral ao Fissurado Labiopalatal-CAIF) is a care center of the State Health Secretariat of Paraná (Secretaria de Estado da Saúde do Paraná-SESA) and a member of the Reference Network for the Treatment of Craniofacial Deformities, an institution created in 1998 by the federal government for accreditation of care services specializing in the treatment of craniofacial deformities. All treatment modalities are provided by the Unified Health System (SUS), social contributions, and contributions from nongovernmental organizations (NGOs), including Operation Smile, which is an international NGO that recruits volunteer physicians to operate on individuals with cleft lip and palate at no charge. Therefore, comprehensive and multidisciplinary treatment is available to patients with OCs.

OC has a negative impact on speech, hearing, physical appearance, and cognition and affects the health and social integration of individuals with OCs. OC causes high morbidity, emotional distress, and can lead to social exclusion. This anomaly is caused by genetic and environmental factors, which can act in isolation or together. The treatment of OCs is multiprofessional and should be integrated, comprehensive, and continuous, starting at the age of 3 months.

Knowledge of the epidemiological profile of patients with OC treated at a specialized center is essential for the community, allowing evaluation of effectiveness of care services, enabling better structuring of these services, and subsidizing new research.
OBJECTIVE

The objective of this study was to analyze the epidemiological data obtained from the CAIF - a referral center in Curitiba, Paraná, Brazil - which treats patients from the state of Paraná and 17 other states, and evaluate the epidemiological profile of patients with cleft lip and/or cleft palate admitted to this center.

METHODS

This descriptive, cross-sectional, and retrospective study was approved by the Research Ethics Committee of our institution under Certificate for Ethics Assessment (Certificado de Apresentação para Apreciação Ética-CAAE) No. 44484015.0.0000.0093.

In total, 1,262 medical records of patients with OC were analyzed, and the records were subdivided into four age groups-infants (aged 0 to 2 years), preschool children (aged 2 to 4 years), schoolchildren (aged 4 to 10 years), and adolescents (aged 11 to 19 years)\(^{11}\), treated at the CAIF, located at the Worker’s Hospital in Curitiba, Paraná.

The following variables were searched in the medical records: age, sex, place of origin, type of cleft (according to the Victor Spina classification\(^{12}\)), age at the first visit, age at the first surgical intervention, and history of previous surgery. The inclusion criteria were cleft lip, cleft palate, or cleft lip and palate, with a follow-up at the CAIF from January 2011 to December 2014. The exclusion criteria were age over 19 years, incomplete and/or illegible medical records, and any other craniofacial anomaly.

The classification of clefts most commonly used in CAIF is that of Victor Spina. This classification takes into account the incisive foramen, the border between the primary and secondary palates, and allows the separation of clefts into three types: pre-foramen (cleft lips, which may be categorized into unilateral, bilateral, or median), post-foramen (cleft palates, which may be complete, involving the entire hard palate, or incomplete, involving only the uvula or a portion of the palate), and trans-foramen (unilateral or bilateral, involving the lip, dental arch, and whole palate)\(^{12,13}\).

After applying these criteria, 408 medical records were excluded because they involved patients with craniofacial anomalies, 20 medical records because they were incomplete and/or illegible, and 168 records because the patients were older than 19 years. In total, 666 medical records were included in the present study, corresponding to 52.7% of all records analyzed.

Statistical analysis

The data were tabulated and expressed as medians, means, and standard deviations or as frequencies and percentages. Statistical analysis was conducted using the statistical package Prism version 5.0 (GraphPad, San Diego, CA, USA) using the Kolmogorov-Smirnov test to assess data normality. Categorical variables were expressed as percentages and compared using the \(\chi^2\) test or Fisher’s exact test, as appropriate. A \(p\)-value of less than 5% was considered statistically significant.

RESULTS

The demographic characteristics and classification of cleft lips and palates in children treated at the CAIF during the study period are shown in Table 1. It was not possible to include a group from other regions in the statistical analysis because of the low number of patients (\(n = 16\)).

In the present study, the number of male patients with OCs was significantly higher than that of female patients (\(p < 0.0001\)). In both sexes, incomplete post-foramen cleft was the most common anomaly (25.8\%), including 74 cases in male patients and 72 cases in female patients, followed by left unilateral trans-foramen cleft, which occurred in 59 boys and 39 girls. Among the participants, 137 patients (20.6\%) had a history of OC in the family, 471 (70.7\%) had a history of no OC in the family, and 58 (8.7\%) had no history.

The medical records of 106 patients were not considered for the statistical analysis in the interval between the first consultation and the first surgery performed at the CAIF because these records were incomplete or the patients had not yet undergone corrective surgery by the date of the survey. Among these, 49 patients were from Curitiba or the metropolitan region, 50 patients were from rural areas of Paraná, and seven patients were from other geographical regions.

The median age at the first visit to CAIF was higher in patients from rural areas (\(p = 0.0001\)) than that of patients living in Curitiba or the metropolitan region (Table 2 and Figure 1).

There was no significant difference in the interval between the first consultation and the surgical procedure performed at the CAIF (\(p = 0.91\)) in the patients from the capital and those from rural areas of Paraná (Table 2 and Figure 2).

DISCUSSION

An OC limits many aspects of the patient’s life, including speech, hearing, and cognition, and hinders the interaction of the patient with social groups. However, this effect can often be reversed when interventions are performed early\(^{14}\). OC needs to be diagnosed early to ensure that the treatment follows established guidelines and achieves the best possible
Table 1. Demographic profile and classification of cleft lip and palate in children treated at CAIF from January 2011 to December 2014.

<table>
<thead>
<tr>
<th>Variables</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>382</td>
<td>57.4</td>
</tr>
<tr>
<td>Female</td>
<td>284</td>
<td>42.6</td>
</tr>
<tr>
<td>Place of origin</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Curitiba and metropolitan area</td>
<td>244</td>
<td>36.6</td>
</tr>
<tr>
<td>Rural areas of Paraná</td>
<td>406</td>
<td>61</td>
</tr>
<tr>
<td>Other regions</td>
<td>16</td>
<td>2.4</td>
</tr>
<tr>
<td>Classification of cleft</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-foramen* (27.2%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complete unilateral</td>
<td>45</td>
<td>6.7</td>
</tr>
<tr>
<td>Complete bilateral</td>
<td>6</td>
<td>0.9</td>
</tr>
<tr>
<td>Incomplete unilateral</td>
<td>114</td>
<td>17.1</td>
</tr>
<tr>
<td>Incomplete bilateral</td>
<td>16</td>
<td>2.4</td>
</tr>
<tr>
<td>Trans-foramen* (34.8%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left unilateral</td>
<td>98</td>
<td>14.7</td>
</tr>
<tr>
<td>Right unilateral</td>
<td>58</td>
<td>8.7</td>
</tr>
<tr>
<td>Bilateral</td>
<td>76</td>
<td>11.4</td>
</tr>
<tr>
<td>Post-foramen* (25.8%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complete</td>
<td>26</td>
<td>14.7</td>
</tr>
<tr>
<td>Incomplete</td>
<td>146</td>
<td>22</td>
</tr>
<tr>
<td>Others (12.2%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other combinations</td>
<td>7</td>
<td>1</td>
</tr>
<tr>
<td>Submucosal</td>
<td>49</td>
<td>7.4</td>
</tr>
<tr>
<td>Mixed</td>
<td>19</td>
<td>2.9</td>
</tr>
<tr>
<td>Rare*</td>
<td>6</td>
<td>0.9</td>
</tr>
</tbody>
</table>

* according to the classification of Victor Spina (http://ctmc.lusiada.br/malformacoes-craniofaciais/). * p < 0.0001 using the χ² test. CAIF: Centro de Atenção Integral ao Fissurado Labiopalatal (Center for Comprehensive Care to Individuals with Cleft Lip and Palate).

Table 2. Correlation between the place of origin, age (in months) at the first consultation at CAIF, age at the first surgery performed at CAIF, interval between the first consultation and first surgery performed at CAIF, and difference between patients previously operated or not.

<table>
<thead>
<tr>
<th>Origin</th>
<th>Curitiba and metropolitan area, n = 244 (36.6%)</th>
<th>Rural areas of Paraná, n = 406 (61%)</th>
<th>Other regions, n = 16 (2.4%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at first consultation at CAIF (months), median (range)</td>
<td>1 (1-219)</td>
<td>2 (1-218)</td>
<td>6 (1-98)</td>
</tr>
<tr>
<td>Age at the first surgical intervention at CAIF (months), median (range)</td>
<td>6 (1-222)</td>
<td>7 (2-222)</td>
<td>12 (4-81)</td>
</tr>
<tr>
<td>Interval between the first consultation and the first surgery performed at CAIF (months), median (range)</td>
<td>4 (1-41)</td>
<td>4 (1-24)</td>
<td>8 (3-15)</td>
</tr>
<tr>
<td>Previously operated (n = 54, 8.1%)</td>
<td>18</td>
<td>34</td>
<td>2</td>
</tr>
<tr>
<td>Not previously operated (n = 612, 91.9%)</td>
<td>226</td>
<td>372</td>
<td>14</td>
</tr>
</tbody>
</table>

CAIF: Centro de Atenção Integral ao Fissurado Labiopalatal (Center for Comprehensive Care to Individuals with Cleft Lip and Palate).

quality of life for the affected individuals15. Therefore, this study presented epidemiological data to evaluate the contribution of CAIF in the state of Paraná, to collect and disseminate data from this population, and indicate the importance of these data for the identification and effective treatment of children with cleft lip and palate.

The interval between the first consultation and the first surgery needs to be determined to ensure adequate planning of the scheduled surgical procedures. In fact, the earlier the patient is admitted to the CAIF, the higher the possibility of planning the surgery considering the recommended ages. This strategy aims at reducing the likelihood of hearing impairment and limitations to the child’s maxillofacial.
the CAIF was observed, which can be attributed to the geographical distance to the referral center and the consequent displacement of patients coming from other regions to the center, which is located in the capital. However, even in cases in which treatment was provided at a median age of 2 months, the patients from rural areas were treated within the appropriate timeframe for the surgical correction, and the CAIF was effective even for these patients because of the promptness in performing the surgical procedure.

The recommended sequence of surgeries involves closure of the soft palate at the age of 3 to 9 months, depending on the type and extent of the cleft, and secondary closure of the residual hard palate at the age of 15 to 18 months. However, there is no consensus on these age intervals. In the present study, the age at which the CAIF received the patients with OCs was adequate considering the main objectives, which were ensuring speech development and maintaining normal maxillofacial growth in addition to reducing the incidence of otitis media and subsequent long-term hearing loss.

In this study, surgical correction was performed at a median of 6 and 7 months in the patients from the capital and rural areas, respectively. Patients from rural areas had more reasons for nonadherence or might need to wait longer for surgery. However, our results indicated that the longer distance from the CAIF did not represent a significant barrier for this group to attend the visits scheduled for surgical procedures (Figure 2), indicating adequate performance of this referral center in the treatment of patients with OCs.

The number of cases of OC in male patients was higher than that in female patients. This result is similar to that found in other studies conducted in Brazil in São José dos Campos, São Paulo, Recife, Pernambuco, and Minas Gerais and in Africa. Moreover, pre-foramen clefts were more common in boys (11.1%), but this result disagrees with those in the literature. The incidence of trans-foramen clefts was similar to that of other studies, with a lower incidence in girls.

Many studies reported a higher incidence of left unilateral clefts, which contradicts the results of a study conducted in Iran between 1982 and 2011, wherein the most common anomaly was bilateral followed by right unilateral clefts. However, the results of the remaining studies agree with ours, with a higher prevalence of left unilateral clefts (30.7%) followed by right unilateral clefts (16.5%) and bilateral clefts (14.7%). These results indicate the influence of environmental and genetic factors on the incidence and type of OC in different populations.

Epidemiological studies on OC use two common classifications of clefts. One was used in the present

**Figure 1.** Age (in months) of children at the first consultation at the Center for Comprehensive Care to Individuals with Cleft Lip and Palate (Centro de Atenção Integral ao Fissurado Labiopalatal-CAIF). The median age at the first visit to the CAIF was higher in patients from rural areas than that of patients living in Curitiba or the metropolitan region.

**Figure 2.** Period (in months) between the first visit and surgery considering the place of residence of patients treated at the Center for Comprehensive Care to Individuals with Cleft Lip and Palate (Centro de Atenção Integral ao Fissurado Labiopalatal-CAIF). There was no significant difference between the groups.
study, and the other classification simplifies the malformations into cleft lip, cleft palate, and cleft lip and palate. We chose the Victor Spina classification because it was more commonly used in the referral center used for data collection and because it was more comprehensive in the details of the clefts. In our study as well as in other Brazilian studies, there was a predominance of trans-foramen clefts and cleft lips and palates. In a study conducted in Africa and another in Iran, the most common type of malformation was combined cleft lip and palate, occurring in 58% and 50% of patients, respectively.

It was not possible to evaluate the ethnicity of the individuals with OCs treated at the CAIF. This information was not included in several medical records, thus limiting the analysis. Croen et al. evaluated a population in California in the United States and observed that Black people had a lower prevalence of OC than Caucasians and that Asians had the highest prevalence, which agrees with the results of a study conducted in Europe.

The CAIF is an institution founded in 1993 and maintained by the SESA of Paraná with the assistance of the Association for Rehabilitation and Social Promotion of Individuals with Cleft Lips and Palates (Associação de Reabilitação e Promoção Social ao Fissurado Labiopalatal-AFISSUR). With a large, competent, and committed multidisciplinary team, this care center treats children and adults from 18 Brazilian states and partners with national and international organizations and universities. CAIF is recognized worldwide as a referral center for the treatment and rehabilitation of individuals with OCs and craniofacial deformities in general.

CONCLUSION

In the present study, there was a predominance of male subjects with OCs. According to the classification of clefts, the most common deformity was incomplete post-foramen cleft. There was a significant difference in the median age at the first consultation between children living in the capital and those living in rural areas of Paraná. However, there was no difference in age at the time of surgery considering the place of origin of the patients. In addition, the interval between the first consultation and surgery was adequate with respect to what is expected from a referral center.

COLLABORATIONS

AVV Analysis and/or interpretation of data; 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.

CLVS Final approval of the manuscript; conception and design of the study; completion of surgeries and/or experiments.

GTC Final approval of the manuscript; conception and design of the study; completion of surgeries and/or experiments.

MGRMR Final approval of the manuscript; conception and design of the study; completion of surgeries and/or experiments.

WST Final approval of the manuscript; conception and design of the study; completion of surgeries and/or experiments.

MAG Analysis and/or interpretation of data; 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.

RN Analysis and/or interpretation of data; 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.

REFERENCES


