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ORIGINAL ARTICLE

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Prevalence of cleft lip and palate in Bauru, SP – concordance among registries of HRAC/USP, DNV and SINASC

Prevalência da fissura labiopalatina em Bauru, SP – concordância entre os registros do HRAC/USP, DNV e SINASC

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ABSTRACT

Objective: Congenital defects, including cleft lip and palate, increase the morbidity and mortality in the affected population. This study aimed to determine the prevalence of cleft lip and palate in the city of Bauru, Brazil, by evaluation of registry in the Brazilian Livebirth Certificate (DNV) and the Information System on Livebirths (SINASC), and analyzed the concordance of diagnosis compared with registries of the Hospital for Rehabilitation of Craniofacial Anomalies (HRAC/USP), located in the same city. **Material and Methods:** This retrospective observational study comprised analysis of all DNVs and identification of individuals with clefts born and living in Bauru, comparing with data from HRAC/USP. The prevalence was calculated by dividing the number of children born with clefts in the study period by the total number of livebirths registered. The reporting of different types of clefts was compared by the chi-square test. **Results:** Overall, 50,898 DNV were evaluated, among which there were 25 reported cases of cleft lip and/or palate. In the same period, HRAC/USP registered 77 cases born in Bauru, representing 67.5% of underreporting of the occurrence of clefts. Cleft palate was the most prevalent (34.9%), followed by cleft lip and palate (31.7%) and cleft lip (30.2%), mostly affecting males (58.5%). The reporting of cleft palate (16.12%) was lower compared to cleft lip (43.75%) and cleft lip and palate (54.54%). **Conclusion:** The study revealed predominance of cleft palate, with significant underreporting of clefts in the public health system, especially for cleft palate as compared to cleft lip and cleft lip and palate.

KEYWORDS

Cleft lip; Cleft palate; Epidemiology; Prevalence; Reporting.

RESUMO

Objetivo: Os defeitos congênitos, incluindo fenda labial e palatina, aumentam a morbidade e mortalidade na população afetada. Este estudo teve como objetivo determinar a prevalência de fissura labiopalatina na cidade de Bauru, Brasil, por meio da avaliação do registro na Certidão de Nascimento de Vivos (DNV) e no Sistema de Informação de Nascidos Vivos (SINASC), e analisar a concordância do diagnóstico em comparação com registros do Hospital de Reabilitação de Anomalias Craniofaciais (HRAC / USP), localizado na mesma cidade. **Material e Métodos:** Este estudo observacional retrospectivo compreendeu a análise de todas as DNVs e a identificação dos indivíduos com fissuras nascidos e residentes em Bauru, comparando com os dados do HRAC / USP. A prevalência foi calculada dividindo-se o número de filhos nascidos com fissura no período do estudo pelo total de nascidos vivos registrados. O relato de diferentes tipos de fissuras foi comparado pelo teste do qui-quadrado. **Resultados:** No geral, foram avaliadas 50.898 DNV, entre as quais houve 25 casos notificados de fenda labial e / ou palatina. No mesmo período, o HRAC / USP registrou 77 casos nascidos em Bauru, representando 67,5% de subnotificação da ocorrência de fissuras. A fenda palatina foi a mais prevalente (34,9%), seguida da fenda labiopalatina (31,7%) e fenda labial (30,2%), ocorrendo principalmente no sexo masculino (58,5%). O relato de fissura palatina (16,12%) foi menor em comparação com fissura labial (43,75%) e fissura labiopalatina (54,54%). **Conclusão:** O estudo revelou predomínio de fissura palatina, com subnotificação significativa de fissuras no sistema público de saúde, principalmente para fenda palatina em relação à fenda labial e labiopalatina.

PALAVRAS-CHAVE

Fenda labial; Fissura Palatina; Epidemiologia; Prevalência; Notificação.

INTRODUCTION

Depending on the severity, congenital defects account for a large number of intrauterine deaths and for infant morbidity and mortality. It is estimated that 3 to 6% of newborns are affected by some type of congenital anomaly [1] and, according to Monlleó and Gil-da-Silva-Lopes (2006) [2], these are also a frequent cause of death during the first year of life. The epidemiological approach to congenital anomalies is fundamental for the allocation of resources for the appropriate treatment of each disorder. Among the congenital anomalies, cleft lip and palate are the most common congenital craniofacial defects and can occur as isolated defects or associated with syndromes, caused by lack of union between the embryonic facial processes due to a multifactorial etiology. Depending on the development stage in which the etiological factors act, cleft lip and palate can cause esthetic, functional, psychological and social disorders, of variable severity depending on the extent and type of the defect, which can affect the lip and oral cavity, even causing oronasal communication, besides psychological changes in the newborn's family, which is also concerned with the future social integration. The treatment is performed by a highly specialized interprofessional team with professionals from the areas of plastic surgery, speech therapy, dentistry, nursing, psychology, social work, and others. [3]

In order to improve the statistical control of vital events and enable the development of more reliable demographic and health indicators, the Information System on Livebirths (SINASC) was established in 1990, which uses the Livebirth Certificate (DNV) as data source, an official document issued by maternity wards which is mandatory for civil registration. Since 1999, the Ministry of Health modified the DNV by the inclusion of field 34 - "Congenital malformation and/or chromosomal anomaly", which includes three filling options: yes (presence of a congenital defect), no (absence of a congenital defect) and ignored. In the presence of a congenital defect, it is subsequently classified by the International Code of Diseases, tenth edition (ICD-10), thus creating the basic conditions for the implementation of a national system for monitoring congenital defects. In 2012, the National Congress published the Law 12662/12, which ensures the national validity of the DNV [4], allowing notaries to provide information to all interested public organizations, which allows integrating the information with SINASC and reducing the cases of underreporting. The DNV is issued in three copies – to the Municipal Health Department, to parents or guardians, and to the files of the Health Unit where the child received the first care. Due to the thorough information it contains, the SINASC represents an important instrument for analyzing and recording the characteristics of mothers and babies at birth. [5,6]

The occurrence of cleft lip and palate in Brazil is estimated at one for every 650 births, data classically known for the pioneer study in this type of birth defect conducted by Nagem Filho et al (1968). [7] However, other studies indicate that the available data on craniofacial anomalies in the Brazilian population are scarce and dispersed, depending on the aspects studied in relation to their occurrence, such as heredity and various environmental factors, highlighting the lack of a standardized protocol in this research field. [8]

The Basic Care Handout for Patients with Cleft Lip and Palate of the Municipal Health Secretariat of São Paulo (2012) [9] reports, based on SINASC, that between 2008 and 2011 there was a mean of 167,863 livebirths per year, with a mean of 88 cases of cleft lip and/or palate per year in the city of São Paulo, but also recognizes the occurrence of errors in hospital records and that studies are necessary to check this information.

Thus, there are some questions about the efficiency in the diagnosis and reporting of congenital anomalies by professionals working in maternity wards, responsible for the physical examination and filling the DNVs and also for

providing guidance and referral to the parents of the newborn. In addition, clinical assistance and prevention strategies are necessary to control the occurrence of congenital defects, and reliable data on the disorder is also essential, since such studies are still scarce not only in Brazil, but also in America in general. [8]

After the pioneer study on the prevalence of cleft lip and palate performed in school population in Bauru in 1968 by Nagem Filho et al., [7] no other population-based study was conducted in the city.

The city of Bauru has a tertiary craniofacial center dedicated to the care of individuals with cleft lip and palate, the Hospital for Rehabilitation of Craniofacial Anomalies of the University of São Paulo (HRAC/USP). This hospital was established in 1967, thus the city has a long experience in the management of children with cleft lip and palate. This hospital belongs to the public health system and assists children with clefts from different Brazilian regions, regardless of the socioeconomic background. Thus, all children born with clefts in the city of Bauru are usually referred to this hospital, which is the only center assisting children with clefts in the entire region.

Considering that SINASC is a system for surveillance of congenital defects that uses DNVs as a source of information and represents an important instrument for analyzing and recording the characteristics of mothers and babies at birth, the objective of this study was to investigate the efficiency of reporting of cleft lip and palate in the Livebirth Certificate (DNV) and their concordance in the Information System on Livebirths (SINASC), as well as to determine the prevalence of different types of cleft lip and palate in the population of the city of Bauru. Secondarily, the number of individuals born and living in the city of Bauru and registered in HRAC/USP in the same period was also assessed, for comparison of findings.

MATERIALS AND METHODS

This retrospective observational study was conducted by evaluating field 34, which includes the reporting and description of existing malformations in all DNVs and identification of individuals with cleft lip and palate born and living in the city of Bauru – SP, Brazil, in a period of 11 years, between 01/01/2000 and 12/31/2010. The analysis of field 34 of the DNV was performed for all individuals born in this period. The reporting was also analyzed by searching the SINASC, from the Municipal Health Secretariat. The study was approved by the local Institutional Review Board under n. 321/2012 and was conducted according to the principles of the Declaration of Helsinki.

The study also surveyed the individuals with cleft lip and palate registered at the Hospital for Rehabilitation of Craniofacial Anomalies (HRAC/USP), born in the same period and also living in the city of Bauru, by accessing the institution database. Individuals not living in the city of Bauru at birth were excluded from the sample.

The analysis was performed for the universe and year of birth, and the prevalence was obtained by dividing the number of children born with cleft lip and palate during the study period by the total number of livebirths registered in the city in the period. The study also analyzed the variation in the reporting percentage according to the type of cleft. Finally, the agreement between the type of cleft registered in SINASC was compared with the DNV of each individual, to assess the reliability of digitization of each DNV.

Data obtained from HRAC/USP, DNV and SINASC were grouped in a Microsoft Excel spreadsheet and then analyzed for the epidemiological survey. The reporting of the different types of cleft lip and palate was compared using the chi-square test.

RESULTS

Occurrence and registry of cleft lip and palate

Overall, 50,898 DNVs were evaluated for the 11-year period between 2000 and 2010, with the highest number of DNVs of 5,294 in 2000 and the lowest of 4,373 in 2007, respectively, with a mean of 4627.09 DNVs per year of study or 385.5 DNVs per month. Among the 50,898 DNVs evaluated, 25 reported different types of cleft lip and palate, which would reveal a calculated prevalence of clefts of 1:2036.

Conversely, the database of HRAC/USP retrieved records of 77 cases of individuals with cleft lip and palate for the same period, including the cases identified on the DNVs as described above, revealing a prevalence of 1:661, which represents 67.5% underreporting of occurrences of cleft lip and palate.

Table I shows the details on the number of DNVs evaluated per year and the occurrence of cleft lip and palate (CLP).

Table I - Detail of the number of DNVs evaluated per year and occurrence of cleft lip and palate

YEAR	DNV	CLP
2000	5,294	1
2001	4,902	2
2002	4,729	
2003	4,606	2
2004	4,814	2
2005	4,614	
2006	4,458	
2007	4,373	5
2008	4,432	2
2009	4,290	5
2010	4,386	6
TOTAL	50,898	25

CLP= cleft lip and palate; DNV= livebirth certificate.

Table II presents the number of cases reported on the livebirth certificates (DNV) and the cases registered in the Hospital for Rehabilitation of Craniofacial Anomalies (HRAC/USP) during the entire study period.

Table II - Cases reported in the DNV, HRAC and respective calculated prevalences

Types of cleft	Health Secretariat	HRAC
Lip	7	16
Lip and palate	12	22
Palate	5	31
Combinations	1	6
Lower lip		1
Median		1
TOTAL	25 (7F, 18M)	77 (35F, 42M)
Prevalence	1/2,036	1/661
Prevalence per 1000 births	0.49/1,000	1.51/1,000

F = female; M = male.

The reporting of cleft lip and palate in the DNVs varied according to the type of cleft. There was statistically significant difference between cleft palate (16.12%), cleft lip (43.75%) and cleft lip and palate (54.54%) ($x2 = 8.629$, $p = 0.013$). The reporting of cleft palate was significantly lower compared to cleft lip ($x2 = 4.234$, $p = 0.04$) and cleft lip and palate ($x2 = 3.083$, $p = 0.079$). The reporting of cleft lip and cleft lip and palate were similar ($x2 = 0.33$, $p = 0.566$). There were six cases of combinations of clefts (lip + palate), of which only one was reported. There was also one case of lower cleft lip and one case of incomplete median cleft, which were not reported in the DNV.

Notification of cleft lip and palate in SINASC

Regarding the digitization of the DNV of individuals with cleft lip and palate in SINASC, four cases (16%), between 2000 and 2003, did not describe the information corresponding to the occurrence of the cleft, and one case (from 2004)

stated that there was no malformation. Only in nine cases (36%) there was agreement of reporting of cleft lip and palate between DNV and SINASC and eleven DNVs of cases with cleft lip and palate (44%) had incomplete digitization of field 34 in the SINASC. This demonstrates a low reporting efficiency (36%), revealing a high percentage of error.

DISCUSSION

This investigation was a population-based study comprising investigation of DNVs, which means that the sample included only livebirths, excluding stillbirths and cases of pregnancy interruption.

To obtain reliable data, a second data source was used, specifically by consulting the registry of individuals with cleft lip and palate from HRAC/USP, where children born with clefts in the city of Bauru are assisted. These data were compared with information from the Municipal Health Secretariat of Bauru, which is based on the reports on the DNVs and SINASC.

The prevalence data for cleft lip and palate published in the literature are varied due to factors that hinder the standardization of a universal epidemiological survey protocol. These factors include the multifactorial etiology, besides the difficulty in obtaining a sample and describing or even classifying the different types of clefts. These considerations partly explain the discrepancy in records between databases from the basic public health system and the records of specialized institutions such as HRAC/USP. [10]

According to information from the handout "Livebirth certificate: Field 34 – Handout of Congenital Anomalies" (São Paulo: Municipal Health Secretariat, 2008), the organization of the different fields of the DNV aims to outline the epidemiological profile of pregnant women and newborns, considering demographic, socioeconomic and health risks, but it also confirms the occurrence of underreporting of congenital anomalies corresponding to field 34, either due to difficulties in diagnosis or even due to professionals' lack of knowledge on the reporting, a fact evidenced in the present results and which agrees with the literature. [11]

The first investigation on the prevalence of cleft lip and palate in Bauru [7] revealed a result of 1:650, while the present study revealed that the occurrence of cleft lip and palate showed a slight decrease over time with a ratio of 1:661, with the male gender being the most affected by the malformation (54.5%). Regarding the most prevalent type of cleft, cleft palate had the highest occurrence (40.25%), this being the greatest variation in relation to data reported in a previous study on the distribution of cleft lip and palate at HRAC/USP, [12] which revealed the occurrence of complete cleft lip and palate in 37.1% of cases. This inversion may be related to environmental factors such as the time of exposure to teratogens, nutrition and the general health condition of the pregnant woman. Concerning the registries, the literature records similar data, cleft palate being the most underreported. [11]

The existence of hospitals specialized in the rehabilitation of this anomaly favors the achievement of more reliable epidemiological data, as observed in the comparison between data from HRAC/USP (77 cases) and those found in the DNVs (25 cases), a relationship that shows underreporting over 60%. Concerning the underreporting of the different types of cleft lip and palate, cleft palate presents the worst scenario, perhaps due to its anatomical location inside the oral cavity, followed by combinations of cleft palate associated with cleft lip, probably omitted by the difficulty of professionals to identify them within the ICD-10 coding. These data are similar to others presented in the literature. [13]

Considering the 77 cases, the calculated prevalence was 1:661, very distant from that calculated considering only the 25 occurrences found in the DNVs, which would be 1:2,036

livebirths. From an epidemiological point of view, this evidences a lack of knowledge on this anomaly and its etiological factors, precluding strategies for preventive measures and the calculation of financial costs and allocation of resources for a scientifically based and timely rehabilitation treatment, which must be performed by a specialized multidisciplinary team. This process starts soon after birth, continuing throughout growth and development until adulthood. [14]

Nunes et al. (2010) [15] found lack of agreement between DNVs and SINASC with underreporting of 46.7%, with cleft palate being the most underreported (65%), thus highlighting the lack of efficiency in these services. For all types of clefts, Souza and Raskin (2013) [16] found 49.9% of underreporting, and Santana et al. (2015) [11] found 43.47% of underreporting of cleft lip and palate. These data agree with those found in the present study, which revealed 67.5% underreporting, and lack of agreement between DNV and SINASC in 60%.

Assistance and prevention strategies are necessary to control the occurrence of congenital defects; therefore, it is necessary to know reliable data on the occurrence of the problem, which are still scarce for Latin America. [17] Despite the previous efforts of the Ministry of Health to reduce the underreporting of this type of anomalies, the large discrepancy in records suggests the need to reinforce the improvement of professionals involved in completing the DNV and SINASC, important documents designed to record the epidemiological profile of the population for the strategic implementation of centers specialized in the rehabilitative treatment of congenital anomalies in the different Brazilian states. Mandatory filling of all fields of the DNV by qualified professionals could be an alternative to enhance the importance of this document, also enhancing the reliability of information recorded in the digital databases of the public health system. Preventive actions related to environmental etiological factors of the Ministry of Health and genetic counseling in centers specialized in the rehabilitation of these defects have undoubtedly contributed over the years to decrease the occurrence of cleft lip and palate.

The study highlights the importance of the DNV not only for the patient, but mainly for the public health system to outline the occurrence of congenital defects and provide adequate treatment for the entire affected population. The present data demonstrate that efforts must be made to ensure the correct and complete filling of the DNV and digitization of this information into SINASC, which should be in charge of properly trained personnel aware of the significance of their work, in order to make this data system that truly reliable. Comparative studies with registries of rehabilitation institutions specialized in different types of congenital anomalies might be conducted to expand the findings.

CONCLUSIONS

According to the DNV, during the study period, 25 cases of cleft lip and palate were reported in the DNV, revealing an "official" prevalence of 1:2,036. However, in the same period, 77 cases were registered at HRAC/USP, revealing a prevalence of 1:661, which represents 67.5% of underreporting of cleft lip and palate. Cleft palate was the most frequent, followed by cleft lip and palate and finally by cleft lip, predominantly affecting males. The reporting of isolated cleft palate was lower compared to cleft lip and cleft lip and palate.

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Conflict of interest

The authors have no proprietary, financial, or other personal interest of any nature or kind in any product, service, and/or company that is presented in this article.

Regulatory Statement

This study was conducted in accordance with all the provisions of the local human subject's oversight committee guidelines and policies of: Declaration of Helsinki. The approval code for this study is: 321/2012.

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