LIVE-BORN INFANTS WITH CLEFT LIP AND/OR CLEFT PALATE: CONTRIBUTION OF SPEECH PATHOLOGY SCIENCES TO SINASC

Live-born infants with cleft lip and/or cleft palate: contribution of speech pathology sciences to Sinasc

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ABSTRACT

Purpose: to characterize all cleft lip and/or cleft palate patients from a state reference center in Pernambuco and all live-born infants notified to Live Birth Information System. The aim was to measure the effectiveness of cleft lip and/or cleft palate patient notification in the System. **Methods:** it was developed an epidemiological study of children born in 2009 treated at the craniofacial malformation reference center, and cleft lip and/or cleft palate live-born babies (born in the same year) notified to SINASC. A deterministic linkage between both data sources matched the names, and those who were on the center's list, but did not appear in the system were considered sub-notifications. **Results:** a total of 138 patients were listed at the reference center. Of these, 37.70% were born with transforamen cleft and 66.40% were male. Seventy-eight live-born children with craniofacial malformation were notified to SINASC. Most were male, and an even distribution among cleft types was observed. Eighty-six of the 138 cases listed as patients at the centerwere not notified to SINASC. Therefore, 110.3% of the cases were sub-notified. **Conclusion:** the sub notification rate of cleft lip and/or palate cleft at SINASC is high. Although only data from Pernambuco were analyzed here, the national scenario is likely comparable.

KEYWORDS: Cleft Lip; Cleft Palate; Information Systems; Live Birth

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Conflito de interesses: inexistente

INTRODUCTION

Cleft lip and/or cleft palate are among the most common craniofacial malformations in the human species, and they occur due to lack of fusion in embryonic processes responsible for the formation of the face and the palate, still in intrauterine life[1].

Such malformation demands a treatment protocol with a complexity level that varies according to the extension of the cleft[2] and that has aesthetical and functional implications, interfering in the individual's communication[3-8] and its social relations, including the psychological impact suffered by the individual itself and its parents and relatives[9-11].

That way, a surgical-therapeutic intervention is necessary on the first months of life, as well as

follow-up assistance throughout the years[2]. The therapeutic interventions influence the facial development and must be done in specialized centers[3] so that the treatment is not performed wrongly, resulting in injuries more damaging than the malformation itself.

In Brazil, there are centers recognized as of reference in the treatment of this malformation[12]. However, being a country with large geographic dimensions, populations of some regions may suffer due to lack of resources for specific treatments, demanding them to look for care in other regions. The main problem is the population's lack of information concerning causes, consequences and possible treatments for cleft lip and/or cleft palate.

Besides, it is important to have detailed information about the live-born infants with such malformation, through precise registries, in order to establish the offer of the most adequate treatment, aiming to recover an appropriate communicative and functional pattern.

According to data from the Ministry of Health (MS), the occurrence of this malformation in Brazil, in 2009, was of five cases for every 10.000 live-born infants. Such occurrence has remained stable on the period from 2000 to 2011[13], despite the expansion of the Family Health Strategy, which distributes folic acid at no cost, the deficiency of which during pregnancy represents one of the main causes for the occurrence of cleft lip and/or cleft palate[14,15].

Brazil has a wide network of Health Information Systems (HIS), in national range, with most of its information available online[16]. Among them, there is the Information System on Live-born Infants (*Sistema de Informações sobre Nascidos Vivos* – Sinasc), implanted nationally on the 1990s, with the goal of identifying the profile of live-born babies and contributing to produce health indices specific to that population. Nowadays, the data produced by Sinasc allow diagnoses, management and vigilance of priority areas, especially because it makes possible to explore aspects related to life and health conditions and to the geographical distribution of births. From data of this system, it is possible to evaluate actions specifically aimed to mother and child healthcare, serving as source to scientific production as well as to planning and evaluating interventions based on the needs of that population[17].

The main document of Sinasc, the Declaration of Live-born Infants (*Declaração de Nascidos Vivos* – DNV) is composed by 52 variables, among them two that refer to the existence of congenital malformations. However, it is still a challenge to develop a pattern on the registration of these two variables, resulting in inaccuracies in the data generated by Sinasc[18]. A study in Rio de Janeiro registered 11,7% of the DNV with ignored information about cleft lip and/or cleft palate malformation, in the period of 1999 to 2001[19]. In another study performed in services of reference to people born with cleft lip and/or cleft palate, it was shown that only 53,3% of the DNV presented the malformation registered; concerning the description, the cleft palate presented the highest number of mistakes, being described accurately only in 25% of the cases[20].

Therefore, this study aimed to characterize cases of cleft lip and/or cleft palate presented in a center of reference in the State of Pernambuco and the live-born infants registered in Sinasc with this malformation, measuring the under-notification of such cases in the system.

METHODS

This work was approved by the Committee of Ethics in Research of the Institute of Integral Health Professor Fernandes Figueira (Imip) and it respects the standards for researches with human beings by the Brazilian National Council of Health (466/2012).

This work consists in an epidemiological study, exploratory and population-based, developed in Pernambuco, state located in the Northeast Region of Brazil, with a current population of 8.931.028 inhabitants[13]. The universe in this study is formed by individuals with cleft lip and/or cleft palate registered in the Center of Attention to Facial Malformations (Cadefi/Imip), born in 2009, and by the live-born infants notified in the Sinasc with either or both types of cleft. The Cadefi/Imip is a regional reference in the treatment of cleft lip and/or cleft palate and other craniofacial defects, considered the third biggest in the country, and it offers high complexity specialized services, involving the main therapeutic practices necessary to the full rehabilitation of individuals with craniofacial anomalies.

This work used as data source the Sinasc bank of the State of Pernambuco and the registration files of Cadefi/Imip, which is filled in every first consultation and is available as part of the documentation, on the hospital records. Based on the Sinasc data bank, it were used for the analysis the live-born infants, from 2009, whose DNV had a cleft lip and/or cleft palate notification, codified in Q35 to Q37 of the CID-10.

The variables *live-born infant gender* and *type of cleft* were collected from the Sinasc data bank. From the Cadefi/Imip, it was observed: gender; type; extension and laterality of the cleft; use of folic acid during pregnancy; prenatal visits; diagnosis of the cleft during prenatal; and family history of the cleft.

A deterministic linkage was performed relating the two sources, taking the mother's name as principal variable of search and, as variables of confirmation

of the real pair, the child's date of birth, the mother's date of birth and the child's gender, respectively. The under-noticed cases were the ones present on the lists of Cadefi/Imip which were not notified on Sinasc as presenting the congenital malformation.

The data were processed and analyzed through the software EpiInfo for Windows, version 3.5.4, and the Bioestat, version 5.0. Absolute and relative frequencies were applied, using Yates' Chi-squared test to evaluate the difference between proportions, adopting α = 5%. The incidence of live-born infants with cleft lip and/or cleft palate was measured from cases notified to the Sinasc; the corrected incidence was calculated by adding the under-notified cases to the numerator.

RESULTS

138 children born in 2009 registered in the Cadefi/Imip with cleft lip and/or cleft palate were found. Of these, 66,4% were males; 37,7% were born with transforamen cleft, 57,3% had full damage and 38,2% had partial damage of the lip and/or palate structures isolated. When related to the laterality, 42,6% were on the left side, and 38,3%, bilateral (Table 1).

The ingestion of folic acid by mothers during pregnancy was notified in 49,3% of the cases, and 97% had access to prenatal visits. Of those who performed the prenatal procedures, 6,9% received the diagnosis of the congenital malformation during pregnancy. When related to family history, 71,2% of the cases had no other occurrences in the family (Table 1).

Of all the variable analyzed on the Cadefi/Imip, only the type of cleft had no statistical relevance ($x^2 = 0,844$; $p = 0,656$). The variables *extension of cleft* and *laterality of cleft*, although statistically significant, presented high rates of ignorability, respectively 20,3% and 31,9%.

On Sinasc, 78 live-born infants were registered, residing in the state of Pernambuco, born in 2009, with some kind of cleft lip and/or cleft palate. Of these, 65,40% (51) are males. There was homogeneous distribution concerning the type of cleft. Each of the three categories (cleft lip; cleft palate; cleft lip and cleft palate) summed 33,33% (Tabela 2).

Of the 138 cases registered in Cadefi/Imip, 86 were not notified to Sinasc, resulting in a under-notification of 110,3% in this system. In other words, for that year, the incidence of live-born infants with the malformation is of 11,55 for every 10.000, and not 5,50 for every 10.000, as measured by direct data provided by Sinasc.

Table 1 – Distribution of live-born infants with cleft lip and/or cleft palate, born in 2009, registered in the Center of Attention to Facial Malformations (Cadefi/Imip)

Variables	*N*	*%*	Yates' x^2 test
	Gender		
Masculine	91	65,9	
Feminine	46	33,3	
Ignored	1	0,7	
Total	*138*	*100,0*	$\chi^2 = 63,83$; $p < 0,0001$*
	Type of cleft		
Pre-foramen	43	31,2	
Post-foramen	43	31,2	
Transforamen	52	37,7	
Total	*138*	*100,0*	$\chi^2 = 0,844$; $p = 0,656$
	Extension of cleft		
Complete	63	45,7	
Incomplete	42	30,4	
Mixed	3	2,2	
Submucous	2	1,4	
Ignored	28	20,3	
Total	*138*	*100,0*	$\chi^2 = 71,45$; $p < 0,0001$*
	Laterality of cleft		
Right	18	13,0	
Left	40	29,0	
Bilateral	36	26,1	
Ignored	44	31,9	
Total	*138*	*100,0*	$\chi^2 = 8,35$; $p = 0,0393$
	Folic acid during pregnancy		
Yes	68	49,3	
No	70	50,7	
Total	138	100,0	$\chi^2 = 0,02$; $p = 0,968$
	Prenatal monitoring		
Yes	129	93,5	
No	4	2,9	
Ignored	5	3,6	
Total	*138*	*100,0*	$\chi^2 = 162,91$; $p < 0,0001$*
	Received the cleft diagnosis during prenatal		
Yes	9	6,5	
No	122	88,4	
Ignored	7	5,1	
Total	*138*	*100,0*	$\chi^2 = 136,49$; $p < 0,0001$*
	Family history		
Yes	38	27,5	
No	94	68,1	
Ignored	6	4,3	
Total	*138*	*100,0*	$\chi^2 = 62,63$; $p < 0,0001$*

* Statistically significant

Table 2 – Distribution of live-born infants in 2009 with cleft lip and/or cleft palate notified to the Information System on Live-born Infants (Sinasc)

Variables	*N*	*%*	Yates' x^2 test
	Gender		
Masculine	51	65,4	
Feminine	27	34,6	
Total	*78*	*100,0*	χ^2 = 9,49; p = 0,0029*
	Type of cleft		
Lip	26	33,3	
Lip and palate	26	33,3	
Palate	26	33,3	
Total	*78*	*100*	*χ^2 = 0,000; p = 1*

* Statistically significant

DISCUSSION

The results of the present study reassure the epidemiological characterization of cleft lip and/or cleft palate found in other parts of the world.

A study undertaken with 541.540 live-born infants in China also found a higher occurrence of the cleft in male children, at the rate of 1,4:1[21]; in another similar study, the rate was of 2,02:1, in a population of 360.990 inhabitants. The higher occurrence in males was also found in Brazil[22,23] and in other parts of the world[24,25].

Related to the type of cleft, the findings in this study confirm the predominance of the transforamen cleft, as observed by other researchers[2,15,21,22]. In the Coope *et al.* study (2000)[21] for instance, the occurrence of cleft lip and palate (65%) was approximately twice as high as cleft lip only (35%). Marques (2000)[26] stated that transforamen cleft was 2,20 times more frequent than pre-foramen type, and 1,96 times more frequent than post-foramen kind.

The preponderance of left laterality corroborates the results from other studies[2,4], as well as the lack of diagnosis of the cleft during prenatal attendance[9,11].

Concerning the omission of registry in records about the extension and laterality of the cleft in some cases, it is relevant to warn the hospital staff about the importance of the registration routine of such data.

It is also important to highlight that the use of folic acid during pregnancy only happened in half of the cases. The low usage of this vitamin, despite its free distribution, is something to be considered, being the lack of both information and family planning some possible causes for such practice.

Comparing the results of registries by Cadefi/Imip and Sinasc, it was verified that the latter presented high under-notification of cleft lip and/or cleft palate among live-born infants, showing that the amount of cases closer to reality would be of 169, born in 2009 in the state of Pernambuco. Therefore, for that year, the incidence of live-born infants with the malformation was of 11,55 for every 10.000, and not 5,50 for every 10.000, as measured by direct data provided by Sinasc.

Concerning the corrected incidence of cleft lip and/or cleft palate in Pernambuco, it was observed that the levels are similar to the ones found in South America (1:1.000)[27]. Generally, there is high variability in the cleft incidence in different regions of the world (from 1/1.000 to 1,81/1.000). The highest rate was found in Czechoslovakia (1,81/1.000), followed by France (1,75/1.000), Finland (1,74/1.000), Denmark (1,69/1.000), Belgium and Netherlands (1,47/1.000), Italy (1,33/1.000), California (1,12/1.000) and South America (1/1.000).

In general terms, it is stated that the Sinasc is a potential source for the study of live-born infants with some kind of congenital malformation and that it keeps important information for the analysis of the occurrence of the cleft nationwide.

Some studies point to advances in the coverage and quality of information from the system in Pernambuco and Brazil[28,29]. Nonetheless, the data brought by Sinasc are only the tip of the iceberg, exposing the challenge of improving the attainment of malformations data in a more precise and realistic way.

CONCLUSION

The present study brought as contributions the acknowledgment of the characteristics of children born with cleft lip and/or cleft palate, as well as the under-notified cases in the health system, providing the possibility of updating the information generated by Sinasc.

Although the data presented is restricted to the state of Pernambuco, it is believed that it reflects a national scenario. Therefore, it is essential to produce more studies in order to expand this discussion and stimulate strategies to improve the attainment of cleft lip and/or cleft palate information by Sinasc.

RESUMO

Objetivo: caracterizar os casos de fissura de lábio e/ou palato atendidos num centro de referência do estado de Pernambuco e os nascidos vivos notificados ao Sistema de Informação sobre Nascidos Vivos com essa malformação, mensurando a subnotificação dos casos de fissura nesse sistema. **Métodos:** desenvolveu-se um estudo epidemiológico, cuja população foram crianças nascidas em 2009 e atendidas no centro de referencia para deformidades craniofaciais em Pernambuco, e pelos nascidos vivos no mesmo ano, notificados ao Sinasc com a fissura. Realizou-se um linkage determinístico entre ambas as fontes de dados e foram considerados subnotificados os casos que constaram na lista de atendimentos do centro de referência, mas que não foram notificados no Sinasc. **Resultados:** dos 138 casos com fissura encontrados no Centro de Referência, 37,70% nasceram com fissura transforame e 66,40% do sexo masculino. No Sinasc, foram notificados 78 nascidos vivos com essa malformação, sendo a maioria do sexo masculino e com distribuição homogênea em relação ao tipo da fissura. Dos 138 casos localizados no Centro de Referência, 86 não foram notificados ao Sinasc, perfazendo uma subnotificação de 110,3%. **Conclusão:** existe uma alta subnotificação da fissura de lábio e/ou palato no Sinasc. Apesar dos dados fazerem referência ao estado de Pernambuco, acredita-se que reflete uma realidade nacional.

DESCRITORES: Fenda Labial; Fissura Palatina; Sistemas de Informação; Nascimento Vivo

REFERENCES

1. Genaro KF, Yamashita RP, Trindade IEK. Avaliação clínica e instrumental da fala na fissura labiopalatina. In: Fernandes FDM, Mendes BCA, Navas ALPGP. Tratado de Fonoaudiologia. São Paulo: Editora Roca; 2009. p. 488-503.

2. Silva Filho OG, Freitas JAS. Caracterização morfológica e origem embriológica. In: Trindade IEK, Silva Filho OG. (org.) Fissuras labiopalatinas: uma abordagem interdisciplinar. São Paulo: Santos; 2007. p. 17-49.

3. Silva DP, Dornelles S, Paniagua LM, Costa SS, Collares MVM. Aspectos Patofisiológicos do Esfíncter Velofaríngeo nas Fissuras Palatinas. Arq. Int. Otorrinolaringol. / Intl. Arch. Otorhinolaryngol. 2008;12(3):426-35.

4. Hsieh CH, Ko EW, Chen PK, Huang C. The effect of gingivoperiosteoplasty on facial growth in patients with complete unilateral cleft lip and palate. Cleft Palate Craniofac J. 2010;47(5):439-46.

5. Yamashita RP, Oliva TRT, Fukushiro AP, Brustello CMB, Trindade IEK. Efeito da veloplastia intravelar sobre o fechamento velofaríngeo avaliado por meio da técnica fluxo-pressão. Rev Soc Bras Fonoaudiol. 2010;15(3):362-8.

6. Brustello CMB, Fukushiro AP, Yamashita RP. Resistência laríngea em indivíduos com fechamento velofaríngeo marginal. Rev Soc Bras Fonoaudiol. velofaríngea: revisão de literatura. Rev CEFAC. 2012;14(3):528-43.

8. Yamashita RP, Carvalho ELL, Fukushiro AP, Zorzetto NL,Trindade IEK. Efeito da veloplastia intravelar sobre a nasalidade em indivíduos com insuficiência velofaríngea. Rev CEFAC. 2012;14(4):603-9.

9. Robbins JM, Damiano P, Druschel CM, Hobbs CA, Romitti PA, Austin AA MS, et al. Prenatal diagnosis of orofacial clefts: association with maternal satisfaction, team care, and treatment outcomes. Cleft Palate Craniofac J. 2010;47(5):476-81.

10. Lei R, Wang S, Cheng C, Chen PK, Chin C. Psychometric evaluation of the stress scale for parents with cleft lip and/or palate children—a preliminary study. Cleft Palate Craniofac J. 2010;47(5):482-90.

11. Berberian AP,Tonocchi R, Souza D, Moleta F, Correia-Lagos HN, Zanata IL. Fissuras orofaciais: aspectos relacionados ao diagnóstico. Distúrb Comum. 2012;24(1):11-20.

12. Monlleó IL, Gil-Da-Silva-Lopes VL. Anomalias craniofaciais: descrição e avaliação das características gerais da atenção no Sistema Único de Saúde. Cad. Saúde Pública. 2006;22(5):913-22.

13. BRASIL. Departamento de Informática do SUS. Informações de Saúde. [citado 2013 Dez 17]. Disponível em: http://www.datasus.gov.br

14. Vieira AR. Unraveling human cleft lip and palate research. J Dent Res. 2008;87(2):119-25.

16. Lima CRA, Schramm JMA, Coeli CM, Silva MEM. Revisão das dimensões de qualidade dos dados e métodos aplicados na avaliação dos sistemas de informação em saúde. Cad. Saúde Pública. 2009;25(10):2095- 109.

17. Paiva NS, Coeli, CM, Moreno AB; Guimarães RM, Camargo Júnior, KR. Sistema de informações sobre nascidos vivos: um estudo de revisão. Ciênc. saúde coletiva. 2011;16 (supl. 1):1211-20.

18. Nunes LMN. Prevalência de fissuras labiopalatais e sua notificação no sistema de informação. [dissertação] Piracicaba (SP): Universidade Estadual de Campinas; 2005.

19. Theme Filha MM, Gama SGN, Cunha CB, Leal MC. Confiabilidade do Sistema de Informações sobre Nascidos Vivos Hospitalares no município do Rio de Janeiro, 1999-2001. Cad Saúde Pública. 2004;20(supl.1):83-91.

20. Pereira AC, Nunes LMN, Queluz DP Fissuras orais e sua notificação no sistema de informação: análise da Declaração de Nascido Vivo (DNV) em Campos dos Goytacazes - RJ, 1999-2004. Ciênc. saúde coletiva. 2010;15(2):345-52.

21. Cooper ME, Stone RA, Liu Y, Hu DN, Melnick M, Marazita ML. Descriptive epidemiology of nonsyndromic cleft lip with or without cleft palate in Shangai, China from 1980 to 1989. Cleft Palate Craniofac. J. 2000;37(3):274-80.

22. Martelli DRB, Cruz KW, Barros LM, Silveira MF, Swerts MSO, Martelli Júnior H. Avaliação da idade materna, paterna, ordem de paridade e intervalo interpartal para fissura lábio-palatina. Braz J Otorhinolaryngol. 2010;76(1):107-12.

23. Di Ninno CQMS, Fonseca LFN, Pimenta MVE, Vieira ZG, Fonseca JA, Miranda ICC, et al. Levantamento epidemiológico dos pacientes portadores de fissura de lábio e/ou palato de um centro especializado de Belo Horizonte. Rev CEFAC. 2011;13(6):1002-8.

24. Natsume N. Kawai T, Kohama G, Teshima T, Kochi S, Ohashi Y, et al. Incidence of cleft lip or palate in 303738 japanese babies born between 1994 and 1995. Br. J. Oral. Maxillofac. Surg. 2000;38(6):605-7.

25. Nazer J, Hubner ME, Catalán J, Cifuentes L. Incidence of orofacial cleft in the University of Chile Maternity Hospital and other hospitals participating in the Latin American Collaborative Study of Congenital Malformations (ECLAMC). Rev. Med. Chil. 2001;129(3): 285-93.

26. Marques D. Estudo genético-clínico de pacientes com fissuras labiopalatais do Núcleo de pesquisa e reabilitação de lesões lábio-palatais, de Joinville/SC. [Dissertação]. Curitiba (PR): Universidade Federal do Paraná; 2000.

27. Derijcke A, Eerens A, Carels C. The incidence of oral clefts: a review. Br. J. Oral Maxillofac. 1996;34(6):488-94.

28. Frias PG, Pereira PMH, Andrade CLT, Lira PIC, Szwarcwald CL. Avaliação da adequação das informações de mortalidade e nascidos vivos no Estado de Pernambuco, Brasil. Cad Saúde Pública. 2010;26(4):671-81.

29. Rodrigues M, Bonfim C, Frias PG, Braga C, Gurgel IGD, Medeiros Z. Diferenciais na adequação das informações de eventos vitais nos municípios de Pernambuco, 2006-2008. Rev Bras Epidemiol. 2012;15(2): 275-84.

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