South African Journal of Surgery. 2021;59(2):57-61
https://doi.org/10.17159/2078-5151/2021/v59n2a3220

Open Access article distributed under the terms of the
Creative Commons License [CC BY-NC-ND 4.0]
http://creativecommons.org/licenses/by-nc-nd/4.0

South African
Journal of Surgery
ISSN 038-2361
© 2021 The Author(s)
PAEDIATRIC SURGERY

The spectrum of orofacial clefts and treatment logistics at Universitas Academic Hospital, Bloemfontein, South Africa

N Blake,[1] H Edeling,[2] D Bekker,[2] PJ Kotzé,[2] R du Plessis,[2] JJF Steyn,[2] G Joubert,[3] JF Jooste[1,2]

[1] Department of Plastic and Reconstructive Surgery, Universitas Hospital, University of the Free State, South Africa
[2] School of Clinical Medicine, Faculty of Health Sciences, University of the Free State, South Africa
[3] Department of Biostatistics, School of Biomedical Sciences, University of the Free State, South Africa

Corresponding author, email: *drnikitablake@gmail.com*

Background: Cleft lips and/or palates should be identified early and be operated on at specific ages according to international recommendations. In African countries, however, cleft lip and palate surgeries are often delayed.
Methods: A retrospective, descriptive study was done to determine the distribution, specific time delays, demographics and loss to follow-up of patients with cleft lip and/or palates treated at Universitas Academic Hospital over a 10-year period. Data was obtained from outpatient files from the Plastic and Reconstructive Surgery Department at Universitas Academic Hospital. Two hundred and three of 318 records (63.8%) had the defined variables documented.
Results: The median time from first presentation to specialist consultation was 1.9 months. The median ages for first presentation was 2.2 months and for specialist consultation 5.2 months. Patients mainly had isolated cleft palates (42.4%), followed by both cleft lip and palate (31%) and isolated cleft lips (24.6%). A quarter of patients (25.6%) were lost to follow-up. More than a third (36.5%) of patients were referred from the local Motheo district and 12.8% were referred from Lesotho.
Conclusion: In our setting, patients with cleft lip and/or palate are generally diagnosed and referred late. These patients also have delayed access to specialist consultation. Often patients are only evaluated by specialists at ages whereby they should have already undergone their first surgeries. Too many patients are lost to follow-up.
Keywords: orofacial cleft, delay, presentation, consultation, surgery

Introduction

Orofacial clefts can be described as incomplete development of anatomical structures that are meant to separate the nasal and the cavities. These structures include the lip, alveolus, and both the hard and soft palates. Orofacial clefts are common congenital abnormalities of the head and neck with an overall prevalence of up to 1 in 700 live births worldwide.[1,2] Orofacial clefts include isolated cleft lip (CL), isolated cleft palate (CP) as well as cleft lip and palate (CLP). CLs may present unilaterally but also bilaterally.[3]

In African countries, cleft patients often have delayed presentation, and untreated cleft lips and/or palates are found with increasing incidence.[4,5] This leads to numerous problems, including feeding, hearing, dental and speech impairment.[3,6] All of these complications can have a major impact on the life and development of the child affected.[7] Factors that contribute to the late diagnosis of the CP include lack of medical aid and funds, transportation, availability of a tertiary institution as well as a lack of basic health care in these countries.[8,9] In Ghana, the most common reason for the delay in cleft repair was lack of finance (47.5%). Other reasons included a lack of realisation of treatment availability, superstition, fear of death from surgery and long distance to health facilities.[7] Olasoji et al. found that mothers in Nigeria commonly believe cleft deformities are due to supernatural forces and seek help from traditional healers, further delaying their presentation to specialised care.[10] In Madagascar, a study found that shame in the community was the main contributing factor in the delay of cleft care.[11] Cultural beliefs and perceptions regarding CLP deformities prevent optimal treatment in African countries.

It is imperative that the treatment of patients with CLs and/or CPs should be multidisciplinary.[3] This includes various role-players such as plastic surgeons, dentists, orthodontists, otolaryngologists, paediatricians, geneticists, speech therapists, dieticians, psychologists and nurse practitioners.[12,13] The optimal timing of CL and CP repairs remains disputed, but most craniofacial centres recommend CL repair at between three to four months of age. A commonly used rule for the timing of CL repair is at least 10 weeks or 10 pounds (approximately 4.5 kg). CP repair is done when the child is older, typically 6 to 12 months of age, although we prefer to do this repair after 9 months of age.[13,14] A delay in the management of these abnormalities leads to difficulty suckling and feeding, speech disabilities, abnormal dental and midface development and hearing impairments due to secondary middle ear infections.[3,12,13] CP surgeries are prioritised highest at our centre as this deformity has the most devastating repercussions if left untreated. Additional

surgeries may need to be performed later in life, such as alveolar bone grafts, orthodontics and rhinoplasty.[3,12-14]

The age at first presentation entails the first time any doctor, clinician or nurse realises the cleft condition and subsequently seeks specialist help. In countries with under-resourced healthcare services, there is often a delay in diagnosis as well as delay in specialist consultation.[5] This study aimed to describe the spectrum of cleft abnormalities and treatment logistics related to time delays and the loss to follow-up of these patients for comparison to international and African reports.

Methods

This was a retrospective descriptive study conducted at Universitas Academic Hospital (UAH), a tertiary institution which hosts specialist services including a Plastic and Reconstructive Surgery Department. Patients requiring cleft surgeries are identified at the Plastic and Reconstructive Surgery outpatient department (PSOPD) at the monthly cleft clinic, commonly known as the Smile clinic. The majority of the CL and CP surgeries (provided they fit into the patient's timeframe for ideal surgical age) take place in designated weeks, referred to as Smile Weeks, which occur twice a year. Smile Weeks consist of mass cleft surgery lists that are funded by the Smile Foundation of South Africa, a non-profit organisation that raises funds for surgeries at government hospitals for children with congenital abnormalities. Smile Weeks provide dedicated theatre time to perform cleft surgeries, thus reducing usual waiting times. This also creates an opportunity for intensive multidisciplinary management as well as the encouragement of parental support groups.

The target population of this study was all patients with CL and/or CP seen and treated at the Department of Plastic and Reconstructive Surgery at UAH, Bloemfontein, 2008 to 2017. This included patients referred from Lesotho as well as from other regions in South Africa.

Data was extracted to data collection forms from the cleft clinic files of the 318 patients identified with cleft abnormalities. Patients with insufficient data in their files were excluded. No patient identifiers were recorded to preserve anonymity of the subjects. Information on the data collection forms was transcribed into a Microsoft Excel spreadsheet for analysis.

Analysis was conducted by the Department of Biostatistics, University of the Free State (UFS). Results were summarised by frequencies and percentages (categorical variables) and medians and interquartile ranges (numerical variables due to skew distributions).

Results

Two hundred and three of 318 records (63.8%) had the defined variables documented. Forty-eight per cent of patients were male and 52.2% were female patients. The highest percentage of patients had isolated CP (42.4%), while 31.0% had both CL and CP, and 24.3% had isolated CL.

Patients first presented at a median age of 2.2 months and their first specialist consultation was at a median of 5.2 months. The median time between first presentation and first consultation was 1.9 months. Table I indicates the time delays from first presentation to first consultation for each specific cleft deformity, as well as cleft type per gender. For all three deformities, the median number of days was between 50 and 60 days. Twenty-five per cent of cleft patients presented later than 9.7 months, still untreated. Once diagnosed, only 50% of patients were seen at UAH Plastic and Reconstructive Surgery Department congenital

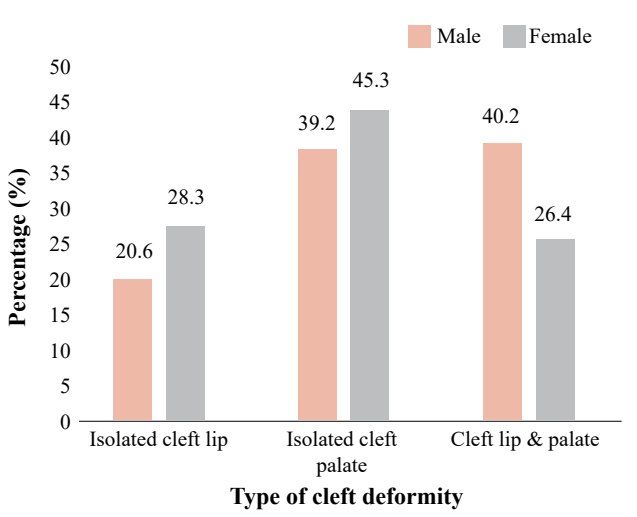

Figure 1: Cleft types per gender

Table I: Time delays per cleft type and gender (*n* = 203)

Variable	*n*	Time delays					
		Age at 1st presentation		Age at specialist consultation		Time from 1st presentation - specialist consultation	
		Months		Months		Days	
		Median	IQR	Median	IQR	Median	IQR
Isolated CL	50	1.4	0.1; 5.2	3.9	2.3; 9.4	54.5	15; 93
Male	20	3.0	0.2; 5.1	6.1	3.4; 9.6	85	7; 128
Female	30	0.6	0.03; 5.2	2.7	1.7; 7.4	44	21; 78
Isolated CP	86	4.7	0.8; 37.8	10.7	3.0; 53.1	58.5	7; 154
Male	38	5.6	0.9; 43.5	11.1	4.0; 46.8	38.5	6; 104
Female	48	4.3	0.5; 32.6	10.7	2.7; 58.3	65	14; 183
CLP	67	0.3	0.03; 4.2	3.6	1.6; 9.2	55	13; 120
Male	39	0.3	0.03; 4.9	3.6	1.5; 10.2	64	26; 120
Female	28	0.2	0; 3.9	3.1	1.7; 9.1	52.5	8.5; 159.5

IQR – interquartile range

Table II: Number of patients and loss to follow-up per referral area

Variable	Cleft patients and loss to follow-up per area			
	Cleft patients per area		Loss to follow-up per area	
	n	%	*n*	%
Central Free State	74	36.5	21	39.6
Nothern Free State	19	9.4	6	11.3
Eastern Free State	34	16.7	10	18.9
Western Free State	34	16.7	5	9.4
Southern Free State	7	3.5	3	5.7
Northern Cape	3	1.4	2	3.8
Lesotho	26	12.8	6	11.3
Other	6	3	0	0
Total	**203**		**53**	

defect multidisciplinary clinic under 5.2 months, and 25% of patients were older than 15.8 months.

The majority of female patients had isolated CP deformities (45.3%) and isolated CL deformities (28.3%). Male patients had the highest percentage of CLP deformities (40.2%). There were 117 CL patients in total; 50 with isolated CL and 67 with CLP deformities. For most of these patients (75.2% – 72.9% of males and 77.6% of females), the cleft was unilateral.

Table II shows the number of patients per referral area with their respective loss to follow-up rates. The local Motheo region (Central Free State) ranks as the area from which the largest number of referrals were received (36.5%, n = 74). Of the main geographical regions represented, Motheo district had the lowest time to first presentation (median 12.5 days) and Lesotho the longest (median 135 days). Lesotho patients, however, had the shortest median time from first presentation to specialist intervention (8.5 days).

Patients were classified as loss to follow-up when they defaulted on their follow-up appointment date and did not return again within 6 months. A quarter (26.6%) of the 203 patients were lost to follow-up. The most patients lost to follow-up per year was 9 patients (17.3%) in 2013, however, there has been a steady decline since then with only one patient (1.9%) lost to follow-up in 2017. Of the 53 patients lost to follow-up, 24 (42.4%) were isolated CP patients, followed by 14 (33%) CLP patients and 15 (24.6%) CL patients. This correlates with the total distribution in number of clefts per type.

Discussion

Our study concluded a median age at first presentation of cleft deformities of 2.2 months. This is a significant concern as these deformities should ideally be diagnosed at birth. Patients are delayed to specialist consultation for a variety of infrastructure reasons; an overloaded system with overbooked clinics, shortage of medical personnel, limited access to public and hospital transport, severe financial constraints, and ignorance regarding the importance of referral from the peripheral hospitals and clinics. There are, however, situations in which the delay is planned. Cleft patients are often born prematurely or have concurrent syndromes and spend prolonged periods in neonatal ICU units; in this case, the consultation as outpatient takes place later in life when the patient is in a more stable condition. This would be an acceptable reason for delay to consultation as well as delay to surgery, but was not specified in this study.

A study of cleft patients in Nigeria revealed that the average age of presentation was 2.47 years of age.[8] Similar studies in Ghana showed age at treatment of respectively 3.9 years and 6.7 years.[4,7] Conway et al. found that of the 27 880 patients receiving cleft surgery in Africa, more than half presented after four years of age. The average age of patients at surgery was 9.34 years. Interestingly, they report a clear association between poor economic development and delay in cleft care. African countries with very low gross domestic product (GDP) like Sudan, Guinea and Ethiopia all have average cleft presentations at older than 10 years of age. South Africa has a GDP in the top 3 in Africa and this is reflected in a much younger average age for surgery at 2.22 years as reported in their research.[2]

The median age at first presentation was 2.2 months and specialist consultation was 5.2 months in this study, much earlier than the reports from other African countries. This could be due to higher levels of patient and healthcare worker education or simpler access to resources and is a topic for further research. Timely attendance at the specialist unit is also imperative for patients requiring multidisciplinary services not available at their referral centres. Despite this, these time frames still represent significant delays that prevent presurgical moulding, typically applied to CL, which is only effective up to 3 months of age.[14]

Our study population consisted of well-represented gender groups (47.8% male, 52.2% female) which makes it unlikely that gender discrimination in seeking cleft care is as prevalent in our setting as it in some other African countries.[2]

When reviewing the cleft type distribution, it is interesting that isolated CP (42.4%) was the most common variant in our sample. This differs from most international statistics where combined CLP is predominant, representing around 50% of clefts, followed by 30% isolated CP and 20% isolated CL.[3,13] The findings in other African studies differ. Agbenorku et al. conducted a study in Ghana that recorded a 52.5% rate of CL, with male predominance, in their sample of 61 patients. This is an interesting finding in an African country setting where the mean age at surgery was 6.7 ± 2.7(SD) years. This study also emphasised that lack of finances was the biggest barrier to earlier repair.[7] The findings could be biased and relate to more patients seeking help for a visible deformity, or higher mortality rates amongst CP patients. Another study in Ghana by Donkor et al. also found high rates of CL surgeries

(78%) being performed compared to CP surgeries. They proposed that CP is underdiagnosed in their community and that patients with CLP deformities would often not return for palatal surgery after the lip has been repaired.[4] This has also been an observation at our facility when patients default on their follow-up.

Conway et al. conducted a large study of orofacial clefts in Africa in which they state there were far fewer CP and syndromic patients treated than they had expected. They also propose that these vulnerable groups are likely underrepresented in their results.[2] Isolated CP has a strong association with genetic abnormalities resulting in syndromes.[13] This correlation was not recorded in our study and could form part of future research.

Our results also showed that females have a higher prevalence of isolated cleft deformities, while males have a higher prevalence of CLP. International data from Robin et al. and Neligan et al. found the same cleft gender ratio as this study.[3,14] It is generally accepted that unilateral CL are nine times as common as bilateral clefts lips.[1,13] Ibrahim et al. found that bilateral CL represented 15% of their cases in Nigeria.[8] Our findings of 24.8%, therefore, agree with these two sources that unilateral lips are more common than bilateral.

Out of 203 files included in this study, 25.6% of the patients were lost to follow-up. This is a high number, but it must be considered that most of these patients have long-term follow-up until 16 to 18 years of age.[12] During data collection, we did not specify when in their treatment protocol these 52 patients defaulted. The loss to follow-up per cleft type corresponded to the overall distribution and no type defaulted significantly more than another. Often CLP patients do not return after lip surgery as they do not see a visible problem anymore and are uneducated to the extent of the deformity.[2] Migrant parents, transport issues and financial constraints could also contribute to loss to follow-up.[2,4] CL patients generally require rhinoplasty, which is only performed in the later teenage years, and in our experience, patients often decline this surgery or do not return for this, especially if the deformity is not severe. Another observation is that CP patients may default appointments if they are satisfied with their result despite the multidisciplinary speech therapy and dental management that may be required for the best possible result. Taking this all into account, we should still not be "losing" so many patients that do not return for proper follow-up visits. Every effort is made to contact the family telephonically, and if this is unsuccessful, the social worker becomes involved. If there is no return of the patient after 6 months, the patient is classified as a defaulter.

Lesotho patients utilise Free State Department of Health specialist services that are unavailable in their country. This arrangement does add to an already overloaded healthcare system, but the biggest frustration is delay in referral from the Lesotho centres. Lesotho also lacks speech therapy services and patients have to travel to South Africa for these follow-ups, which can be a costly exercise. Lesotho patients would benefit greatly from patient education, prompt referral policies and investment in paramedical services such as speech therapists. Most patients were referred from the Motheo area, which is expected as it is local and densely populated. The loss to follow-up is also predominantly from this area despite it being the closest to UAH. It is important to evaluate referral areas for appropriate allocation of intervention strategies such as community awareness and education regarding cleft deformities. Similar programmes have been implemented in various countries around the world.[11,15] Foreign patients (Lesotho citizens) contribute a substantial number of cleft patients that rely on the Free State Department of Health for their management and pose fiscal and logistical constraints in providing them with optimal care.

Conclusion

Our study highlights poor follow-up and quality of record-keeping, with over a third of our patients not having enough data points to contribute to this analysis. A record that collects a standardised data set detailing all aspects of the patient's encounters and interventions in navigating the road of cleft care is required. Epidemiological findings are similar to those of international studies with the exception of isolated CP being the most common variant in our population group. In our setting, patients with CL and/or CP are generally diagnosed and referred late, however, we compare favourably with other African countries. Further research regarding specific causes of delays is recommended in order to address these issues appropriately.

Acknowledgements

The authors thank Sr Y Molusi for assisting with access to the patient files at Universitas Academic Hospital and Ms T Mulder, medical editor, Faculty of Health Sciences, UFS, for her contribution in technical preparation of the manuscript.

Conflict of interest

The authors declare no conflict of interest.

Funding source

None.

Ethical approval

The protocol was approved by the Health Sciences Research Ethics Committee, UFS (UFS-HSD2018/0399/2808). Permission was also obtained from the Free State Department of Health to use the patient files from the Department of Plastic and Reconstructive Surgery at UAH.

ORCID

N Blake https://orcid.org/0000-0001-9333-2829
H Edeling https://orcid.org/0000-0002-6755-7722
D Bekker https://orcid.org/0000-0001-5210-4010
PJ Kotzé https://orcid.org/0000-0002-1124-804X
R du Plessis https://orcid.org/0000-0002-4082-9601
JJF Steyn https://orcid.org/0000-0003-3554-0149
G Joubert https://orcid.org/0000-0002-3728-6925
JF Jooste https://orcid.org/0000-0002-9793-3805

REFERENCES

1. Mossey PA, Modell B. Epidemiology of oral clefts 2012: An international perspective. Front Oral Biol. 2012;16:1-18.
2. Conway JC, Taub PJ, Kling R, et al. Ten-year experience of more than 35 000 orofacial clefts in Africa. BMC Pediatr. 2015;15:8.
3. Robin NH, Baty H, Franklin J, Guyton FC. The multidisciplinary evaluation and management of cleft lip and palate. South Med J. 2006;99(10):1111-20.

4. Donkor P, Bankas DO, Agbenorku P, Plange-Rhule G, Ansah SK. Cleft lip and palate surgery in Kumasi, Ghana: 2001 –2005. J Craniofac Surg. 2007;18(6):1376-9.
5. Pham AM, Tollefson TT. Cleft deformities in Zimbabwe, Africa. Arch Facial Plast Surg. 2015;9(6):385-91.
6. Bruneel L, Luyten A, Bettens K, et al. Delayed primary palatal closure in resource-poor countries: Speech results in Ugandan older children and young adults with cleft (lip and) palate. J Commun Disord. 2017;69:1-14. https://doi.org/10.1016/j.jcomdis.2017.06.010.
7. Agbenorku P, Thomas D, Agbenorku M, et al. Orofacial clefts: a clinical community study in a developing country. ISRN Plast Surg. 2013;2013:5-7.
8. Ibrahim A, Mshelbwala PM, Ononiwu CN, et al. A descriptive study of clefts of the primary and secondary palate seen in a tertiary health institution in Nigeria. Niger J Surg Res. 2013;15(1):7-12.
9. Adetayo O, Ford R, Martin M. Africa has unique and urgent barriers to cleft care: lessons from practitioners at the Pan-African Congress on Cleft Lip and Palate. PanAfrican Med J. 2012;12(15).
10. Olasoji HO, Ugboko VI, Arotiba GT. Cultural and religious components in Nigerian parents' perceptions of the aetiology of cleft lip and palate: implications for treatment and rehabilitation. Br J Oral Maxillofac Surg. 2007;45:302-5.
11. Mandrano NA, Tosa Y, Kuroki T, et al. Beliefs and perceptions that impair cleft care treatment in Madagascar: a qualitative study during humanitarian mission. J Cleft Lip Palate Craniofacial Anomalies. 2017;4(2):149-53.
12. Nahai FR, Williams JK, Burnstein FD, et al. The management of cleft lip and palate: pathways for treatment and longitudinal assessment. Semin Plast Surg. 2005;19(4):275-85.
13. Thorne CH, Chung KC, Gosain AK, et al. Grabb and Smith's Plastic Surgery. 7th ed. Philedelphia: Wolters Kluwer Health Adis (ESP); 2014. p. 173-99.
14. Neligan P, Warren R. Plastic Surgery. 3rd ed. Vol. 3. Seattle: Elsevier Saunders; 2012. p. 517-83.
15. Mossey P. Global strategies to reduce the healthcare burden of craniofacial. Br Dent J. 2003;195(10):613.