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Article

PATIENTS' PROFILE OF CLEFT LIP AND PALATE : 3 YEARS EVALUATION AT SURABAYA CLP CENTER FOUNDATION

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ABSTRACT

Introduction: Cleft lip and cleft palate are the most common birth defects affecting newborns, yet their variability across each geographic origin is not clearly understood. This study aims to investigate the CL/P prevalence, profile, and corrective surgery timing in Surabaya, Indonesia.
Method: This retrospective research used a descriptive method by collecting medical data records from the Surabaya CLP Center Foundation from January 2021 to December 2023, which related to sociodemographic problems, clinical features, and management timing were then statistically analyzed.
Results: A total of 495 patients underwent cleft surgery, of which 59.4% were male and 40.6% were female. The most common type of Craniofacial Cleft observed is Combination Cleft Lip and Palate "CLP" (71.1%). CLP is more common in male patients than in female patients (43.4% vs. 27.7%); while CL is more common among males (14.1% vs. 9.7%) and CP is more common in females (3.2% vs. 1.8%). Significantly, complete clefts (72.9%) were more common than the incomplete clefts (27.1%). Unilateral clefts were more common than bilateral clefts (76.2% vs 18.8%). It was observed that only 22% of patients had a family history of CL/P. Overall, 74,9% of patients underwent corrective surgery within the right period.
Conclusions: The data shows that most patients tended to seek medical advice and treatment at appropriate times, suggesting a high awareness of CLP as the result of proactive education and social outreach by the charity institution. Further investigations using data from other institutions are suggested to conclude the CLP management in Surabaya entirely.

Key words: *Orofacial cleft; Congenital; Cheiloplasty; Palatoplasty*

Pendahuluan: Celah bibir dan celah langit-langit adalah cacat lahir yang paling umum terjadi pada bayi baru lahir. Namun, variasi kejadiannya berdasarkan asal geografis belum sepenuhnya dipahami. Penelitian ini bertujuan untuk menyelidiki prevalensi, profil, dan waktu pelaksanaan operasi koreksi CL/P di Surabaya, Indonesia.
Metode: Penelitian retrospektif ini menggunakan metode deskriptif dengan mengumpulkan data rekam medis dari Yayasan CLP Center Surabaya selama periode Januari 2021 hingga Desember 2023. Data yang berkaitan dengan masalah sosiodemografis, karakteristik klinis, dan waktu pelaksanaan penanganan dianalisis secara statistik.
Hasil: Sebanyak 495 pasien menjalani operasi sumbing, di mana 59,4% adalah laki-laki dan 40,6% adalah perempuan. Jenis celah kraniofasial yang paling umum adalah kombinasi celah bibir dan langit-langit "CLP" (71,1%). CLP lebih sering terjadi pada pasien laki-laki dibandingkan perempuan (43,4% vs. 27,7%); sementara CL lebih sering terjadi pada laki-laki (14,1% vs. 9,7%) dan CP lebih umum pada perempuan (3,2% vs. 1,8%). Secara signifikan, celah lengkap (72,9%) lebih sering terjadi dibandingkan celah tidak lengkap (27,1%). Celah unilateral lebih umum dibandingkan celah bilateral (76,2% vs. 18,8%). Tercatat hanya 22% pasien yang memiliki riwayat keluarga dengan CL/P. Secara keseluruhan, 74,9% pasien menjalani operasi koreksi dalam periode waktu yang tepat.
Kesimpulan: Data menunjukkan bahwa sebagian besar pasien cenderung mencari perawatan medis pada waktu yang tepat, mengindikasikan kesadaran yang tinggi terhadap CLP sebagai hasil dari edukasi proaktif dan kegiatan sosial yang dilakukan oleh Yayasan CLP Center Surabaya. Penelitian lebih lanjut dengan menggunakan data dari institusi lain disarankan untuk mendapatkan gambaran menyeluruh tentang penanganan CLP di Surabaya.

Kata Kunci: *Celah orofasial; Kongenital; Cheiloplasty; Palatoplasty*

Conflicts of Interest Statement:
The author(s) listed in this manuscript declare the absence of any conflict of interest on the subject matter or materials discussed.

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INTRODUCTION

Cleft lip and palate (CLP), despite it being one of the most common birth anomalies, has not been attributed to an exact root cause. It may occur due to various genetic, environmental factors, and socioeconomic conditions. Its variability across geographic origin, race, and ethnic groups is also unclear. The global average incidence of CLP is estimated to be 1.08%. Previous studies show that Asia has the highest incidence of CLP.[1] In broad terms, cleft lip and palate cases are typically categorized into two main groups: cleft lip with or without cleft palate (CLP) and isolated cleft palate (CP). Additionally, depending on the presence of associated anomalies, these cases are further classified as either syndromic or non-syndromic (nsCLP) clefts.[2] The generally accepted average incidence of nsCLP across the global population is 1 in 1000 births. However, there is a clear and significant disparity between different races and ethnic groups. The incidence of nsCLP is lowest in African populations (0.3 in 1000 births). In general, based on the presence of associated anomalies, these cases are further categorized as either syndromic or non-syndromic (nsCLP) clefts. This classification system allows for a better understanding of the diverse presentations of cleft lip and palate conditions, distinguishing between those occurring independently and those associated with other conditions.[2] For additional information, the two most prevalent syndromes associated with orofacial clefts are Van der Woude syndrome and Pierre Robin sequence. Van der Woude defects typically manifest as bilateral isolated cleft lip (CL), whereas the Pierre Robin sequence often presents as cleft lip and palate (CLP). These syndromes are important to recognize in the context of orofacial clefts, as they may have implications for treatment planning and outcomes.[3] In contrast, it is highest in American populations (3.6 in 1000 births) and Asian populations (1.4-2.1 in 1000 births). European populations report a number closer to the global average, i.e. 0.7-1.3 in 1000 births.[2] Specifically in Indonesia, based on the Basic Health Research (Riset Kesehatan Dasar/RISKESDAS) studies, the recorded national prevalence rate of cleft lip and palate increased from 0.08% in 2013 to 0.12% in 2018. The National Guidelines for Medical Services, which addresses the management of cleft lip and palate, indicates that the national prevalence of cleft lip in Indonesia is 0.2%. The annual incidence of cleft lip and palate in Indonesia is estimated to be around 7,500 cases.[4] It is common for CLP patients to suffer impaired facial growth, dental defects, speech disorders, poor aural faculty, and difficulties in social and psychological well-being.[5] In addition to these health problems, affected individuals face potentially long-term social difficulties such as social rejection and even perjury from their peers and communities. Even if reconstructive surgery is performed in the early stage of development, scarring, and other abnormal facial developments may cause lifelong functional and psychosocial problems.[2] Moreover, psychological, nutritional, and speech difficulties experienced by patients are made worse by postponing surgeries, as well as the increase in overall social, psychological, and financial burden to the patients and their families. Delays in surgery are linked to a worse long-term prognosis, and/or in some cases may result in heightened morbidity and mortality.[4]

Therefore, it is necessary to study the profile and prevalence specifically for Cleft Lip and Palate patients in Surabaya. The classification used is LAHSHAL annotation which involves a palindromic diagram to characterize cleft pattern. LAHSHAL system uses various symbols: a strip line (-) as representation for normal anatomy, an asterisk (*) for microform or submucous cleft, lowercase letter (l, a, h, s) for incomplete clefts, and an uppercase letter (L, A, H, S) for complete cleft at the respective region (Figure 9),[6] it is to ensure that the most appropriate corrective surgery is performed at the correct age. Additionally, proactive education and socialization outreach can be carried out to patient's parents and families to seek medical advice and assistance as early as possible, so that the most optimal reconstructive can be planned and timed according to standard time schedule made by CLP Center to achieve the best possible outcome. Completing and analyzing this study will provide a better understanding of CLP prevalence and surgical outcomes in Surabaya.

METHOD

This retrospective research used the descriptive method by collecting medical data records from the Surabaya CLP Center

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Foundation. Surabaya CLP Center Foundations is one of the charity institutions in Surabaya, Indonesia, providing free cleft repair and follow-up treatments. Most of the cleft patients cared for by this foundation belong to the East Java Region. The study was conducted with patients' data from January 2021 to December 2023. The data related to sociodemographic problems and clinical features were collected employing electronic questionnaires completed by surgeons and healthcare providers and recorded in an online medical record database. The data is then compiled and statistically analyzed based on the distribution of gender, classification of cleft location (CL/P), completeness of clefts (complete or incomplete), side involvements (unilateral, bilateral, or palate only), sites/locations of cleft (right or left), and age of patients at the time of surgery. These data sets were then tabulated and analyzed using an Excel spreadsheet.

RESULTS

A total of 495 patients underwent cleft surgery at Surabaya CLP Center over the three years (January 2021 –December 2023), of which 294 (59.4%) were male and 201 (40.6%) were female.

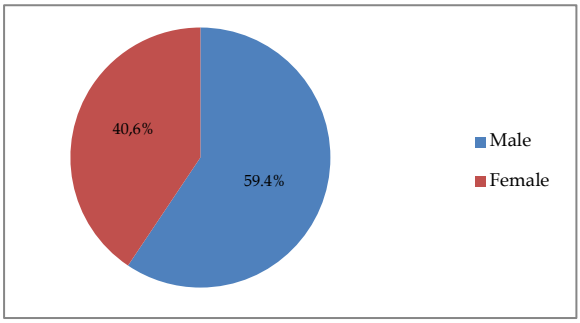

Figure 1. Gender distribution of Cleft lip and/or palate patients

The most common location of the Cleft observed is the Combination of Cleft Lip and Palate "CLP" including LAHSHAL, LAHS---, ---SHAL, lahSHAL, ---Shal, lahs---, ---SHAl, --hsh-l, ---SHal, laHSHAL, laHS---, laHS---, la-S---, lahShal, LAhSh-l, LAHSHal, LAHShal, lAHS---, laHSHal, ---Shal, --HSHAL, LAHShal, --hShal, ---S-al (352; 71.1%) followed by Cleft Lip only "CL" involving la-----, la---al, -----al, LA-----, LA---AL, La---al, l------, ------l, L------, ------L, l-----l, L-----L, LA----l, l----al, l-----aL , (l)------, ------(l) (118; 23.8%) and Cleft Palate only "CP" featuring ---S---, --HSH--, --hSh-- (25; 5.1%).

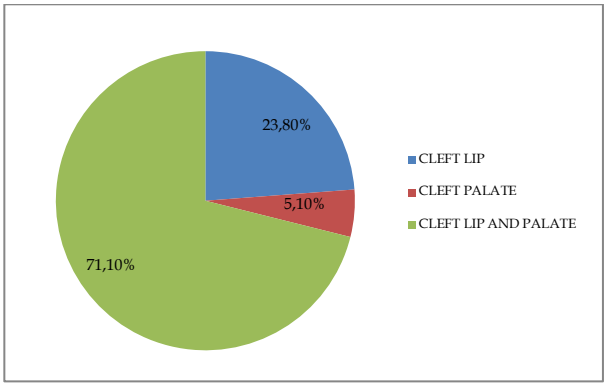

Figure 2. Classification of Cleft lip and/or palate patients

CLP is more common in male patients than in female patients (43.4% vs. 27.7%), CL is more common among males (14.1% vs. 9.7%), and CP including hard and/or soft palate is more common in females (3.2% vs. 1.8%).

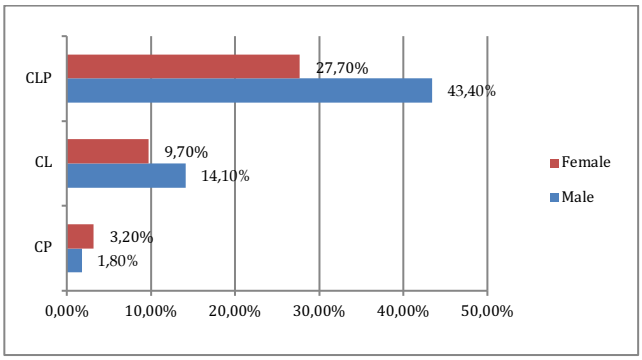

Figure 3. Classification of the cleft with gender distribution of the cleft lip and/or palate patients

We observe that significantly the more common involvement of clefts is complete cleft (361;72.9%) and incomplete cleft is less common (134;27.1%).

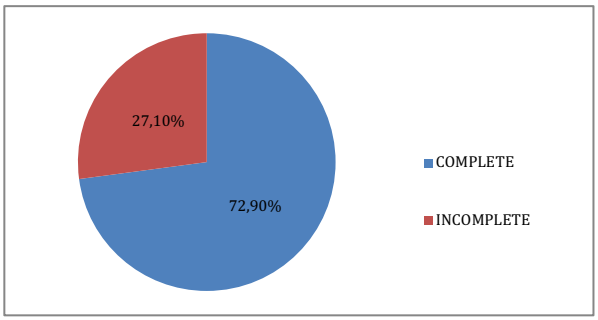

Figure 4. Completeness of clefts

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Unilateral clefts were more common than bilateral clefts (377; 76.2% vs 93; 18.8%), while palate-only clefts were observed only in 25 patients or 5.1% of the total.

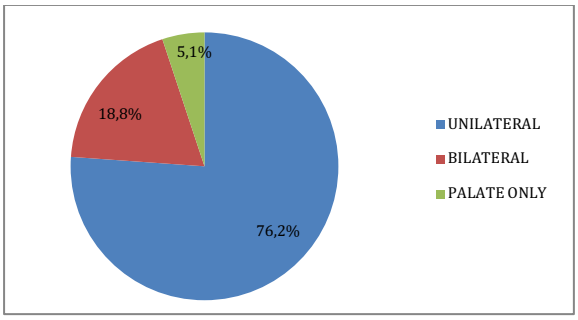

Figure 5. Side involvements of the cleft lip and/or palate patients

This was followed in frequency by left-sided CL with palate (38.2%), right-sided CL with palate (16.4%), left-sided CL only (14.9%), and right-sided CL only (6.7%).

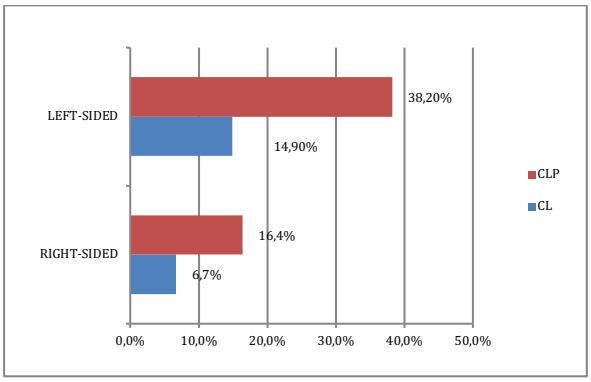

Figure 6. Locations of the cleft lip and/or palate

Overall, 260 (86.1%) patients underwent primary cheilorrhapy within the optimal period (3 -12 months) and 121 (62.7%) underwent primary palatorraphy within the optimal period (10 – 18 months).

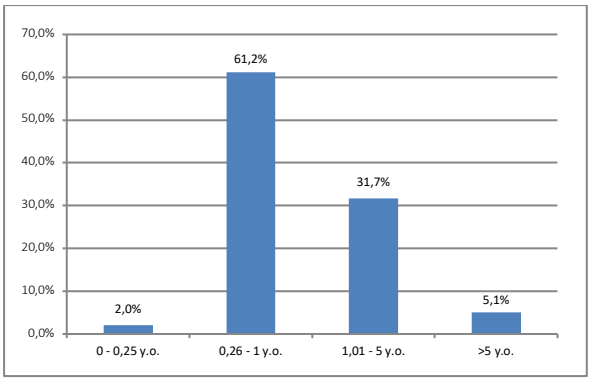

Figure 7. Age distribution of Cleft lip and/or palate patients at the time of surgery

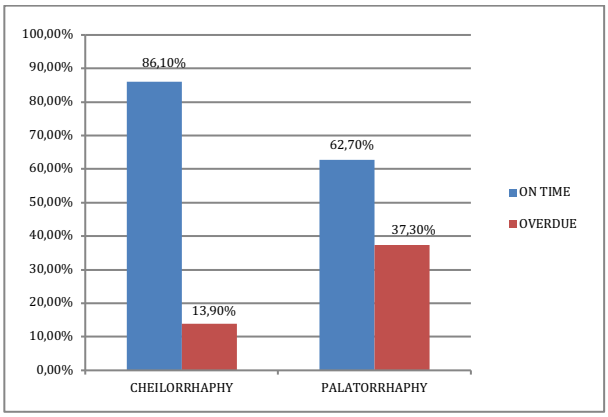

Figure 8. Time compliance of cleft lip and/or palate patients undergoing surgery.

We studied the family history of CL/P for all patients, 22% of cases had a family history of CL/P, while the remaining 78% had no family history of CL/P.

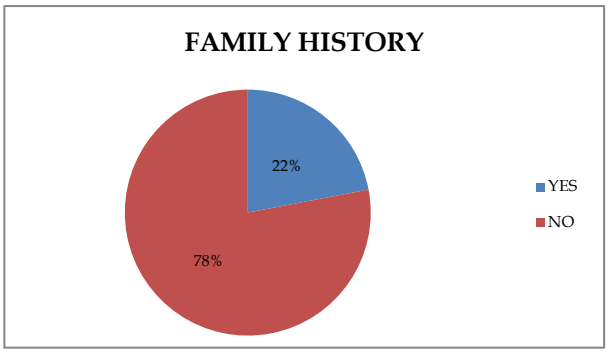

Figure 9. Patients who had a family history of cleft lip and/or palate

DISCUSSION

Cleft lip and cleft palate are the most common birth defects affecting newborn infants. These congenital malformations affect the head and neck and their occurrences, whether at both lip and palate (CLP) or Cleft lip only (CL) or Cleft palate only (CP), significantly impact the quality of life, healthcare utilization, and expenditure of patients and their families as well as public welfare. Additionally, it increases the risk of perinatal deaths.[1] CL/P is most prevalent in the Asian and American populations and occurs the least in African populations. The gender distribution of CL/P is not equal in general.[2] As shown in this study, we observed that CL/P was found more frequently among males (59.4%)

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compared to females (40.6%). This data is relevant to other studies showing similar gender distribution; one study in Damascus, Syria showed a higher percentage of CL/P cases found in males (51.9%),[3] another study in the northern region of Saudi Arabia showed male preponderance (56.6%),[7] and 10 years study in Northern India concluded yet another higher number in males (54.8%).[8] As for the types of clefts found in each gender, the data showed that combined cleft lip and palate (CLP) were more commonly found in males (43.4%) compared to females (27.7%), isolated cleft lip (CL) were higher in males (14.10%) than females (9.7%). On the contrary, isolated cleft palate (CP) was more predominant in females (3.2%). The data showed consistency with other studies in Syria, India, Saudi Arabia, and China, stating a higher incidence of CLP and CL among males while CP had a higher incidence among females. Male predominance in CLP and CL types is coherent with previous studies disclosing higher sensitivity of male fetuses to environmental stress, causing a higher incidence of congenital birth defects.[1] While the female preponderance in CP cases is likely attributed to a longer period of time for palatal closure in female embryos, increasing their probability of exposure to the pathogenic teratogens.[8] The most common type of CL/P based on anatomical involvement in our sample was the CLP (71.1%), followed by CL (23.8%) and CP (5.1%). A study in Saudi Arabia showed a similar distribution of the clefts,[7] other studies in multiple Asian countries also showed the highest prevalence of CLP, with CP as the second most common cleft, rather than CL.[1,3,8,9]

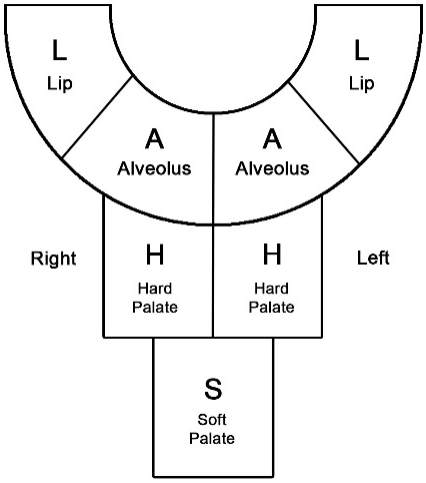

Figure 10. Anatomic location of the cleft LAHSHAL annotation system: Strip line (-) shows normal anatomy, asterisk (*) signifies microform or submucous cleft, lowercase letter (l, a, h, s) indicates incomplete, and uppercase letter (L, A, H, S) represents complete cleft.

The distribution of the involvements in our sample was as follows: the most common were complete cleft (72.9%), rather than incomplete cleft (27.1%). Regarding the side (unilateral) involvements of the CL/P, 18.8% of the cases were two-sided clefts (bilateral), 76.2% were right unilateral cleft and 28.3% were left unilateral cleft. Similarly, the majority of studies in Asian countries showed that unilateral cleft was more frequent than bilateral.[7–9]

One study in Syria showed a higher incidence of right unilateral CL,[3] while other studies in India and Saudi Arabia found a higher occurrence of the left side.[7–9] Previous studies have yet to identify any clear explanation for these differences. One hypothesis is that the blood vessels supplying the right side of the fetal head leave the aortic arch closer to the heart thus providing better blood perfusion to the right side.[8] The causes of non-syndromic cleft lip and/or palate (nsCL/P) are multifaceted, involving a combination of genetic predisposition, environmental influences, and potential gene-environment interactions, with genetic factors estimated to contribute to 90% of nsCL/P cases.[10] This observation is consistent with the findings of Nahas et al., where family histories of CL/P were examined. Their study revealed that 46.6% of cases had a relative with CL/P, while the remaining 53.4% had no family history of CL/P.[3]

Our study similarly found that among all patients, 22% had a family history of CL/P, while 78% did not. This trend aligns with a 2017 study conducted at the CLP Center Foundation in Surabaya, which showed that a higher number of patients undergoing CL/P correction surgery had no family history of CL/P (183 patients), compared to those with a family history (53 patients).[11] Infants and children with these cleft deformities must undergo various stages of treatment and surgery, and various developmental and growth challenges arise from it. In almost all cases, comprehensive and cross-disciplinary management is essential for successful patient care.

It requires a team of specialist doctors from multiple fields, including craniofacial plastic surgery, pediatrics, anesthesiology, otolaryngology, medical rehabilitation, psychiatry, speech therapy, nursing, orthodontics, prosthodontics, and social work.

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Ideally, a long-term treatment protocol for cleft lip and palate patients should be designed, planned, and scheduled to achieve proper occlusion, normal speech function, and a natural appearance, while minimizing the risk of complications.[4]

Schedule intervention and surgery timeline for CLP multidisciplinary care are summarized in Table 1.

Table 1. Timeline of appropriate CL/P interventions

Age	Treatment/Intervention
3 months or more than 10 weeks to 12 months	Cheilonasorraphy can be performed if the 'rule over tens' criteria are fulfilled: - 10 weeks of age - Weights 10 pounds or 5kg - Hemoglobin level > 10gr %
10 – 18 months	Palatorraphy Hearing evaluation
1 – 4 years	Speech evaluation, starting 3 months post-surgery Speech therapy Hearing evaluation
4 - 6 years	In the presence of speech abnormality, consider repalatorraphy or/and pharyngoplasty to minimize nasal speech (nasal escape).
6 years	- Teeth and jaw evaluation - Nasoendoscopy recommended if further hearing evaluation needed
8 – 10 years	Orthodontic treatment to fix alveolar arch
9 – 10 years	Alveolar bone graft
12 – 13 years	Final touch for refining/adjusting previous surgical procedures
Young adults – 17 years (Skeletal maturity)	- Evaluation of facial bones - Advancement osteotomy Le Fort I surgery

Referring to Table 1, we evaluated whether the patients received corrective surgery within the optimal age or came overdue. The study showed that 86.1% of patients underwent primary cheilorrhapy within the optimal age (3 - 12 months) and 62.7% of patients underwent primary palatorraphy within the optimal age (10 – 18 months). The data shows that most patients had the tendency to seek, receive, appropriate medical advice and treatment in appropriate timing, suggesting a high awareness of CLP as the result of proactive education and social outreach by the charity institution. Further investigations using data from other institutions are suggested to entirely concludes the CLP management in Surabaya.

CONCLUSION

The data presented concludes that the most common type of cleft abnormalities operated at Surabaya CLP center were the combination of lip and palate cleft (CLP). Male patients showed higher rate of both cleft lip (CL) and combination cleft lip and palate (CLP), while cleft palate (CP) was more frequent in female patients. Unilateral clefts were more common than bilateral clefts. Complete clefts appeared in higher frequency than incomplete clefts. The data also demonstrates that unilateral left-sided clefts were more common than unilateral right-sided clefts, either of which were more common than bilateral clefts. Majority of patients were shown to seek, and thus receive, appropriate medical advice and treatment in optimal timing.

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N/A

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