Age at Primary Cleft Lip Repair

Vanderburg, Richard; Alonso, Nivaldo; Desai, Priya; Donkor, Peter; Mossey, Peter; Stieber, Erin

Published in:
Plastic and Reconstructive Surgery - Global Open

DOI:
10.1097/GOX.0000000000003657

Publication date:
2021

OPEN

PRS GLOBAL OPEN

ORIGINAL ARTICLE

Craniofacial/Pediatric

Age at Primary Cleft Lip Repair: A Potential Bellwether Indicator for Pediatric Surgery

Richard Vanderburg, RN, BSN*
Nivaldo Alonso, MD, PhD†
Priya Desai, MPH‡
Peter Donkor, BDS, MDSc§
Peter Mossey, PhD¶
Erin Stieber, JD‡
Felicity V. Mehendale, MD||

Background: The bellwether procedures described by the Lancet Commission on Global Surgery represent the ability to deliver adult surgical services after there is a clear and easily made diagnosis. There is a need for pediatric surgery bellwether indicators. A pediatric bellwether indicator would ideally be a routinely performed procedure, for a relatively common condition that, in itself, is rarely lethal at birth, but that should ideally be treated with surgery by a standard age. Additionally, the condition should be easy to diagnose, to minimize the confounding effects of delays or failures in diagnosis. In this study, we propose the age at primary cleft lip (CL) repair as a bellwether indicator for pediatric surgery.
Method: We reviewed the surgical records of 71,346 primary cleft surgery patients and ultimately studied age at CL repair in 40,179 patients from 73 countries, treated by Smile Train partners for 2019. Data from Smile Train's database were correlated with World Bank and WHO indicators.
Results: Countries with a higher average age at CL repair (delayed access to surgery) had higher maternal, infant, and child mortality rates as well as a greater risk of catastrophic health expenditure for surgery. There was also a negative correlation between delayed CL repair and specialist surgical workforce numbers, life expectancy, percentage of deliveries by C-section, total health expenditure per capita, and Lancet Commission on Global Surgery procedure rates.
Conclusion: These findings suggest that age at CL repair has potential to serve as a bellwether indicator for pediatric surgical capacity in Lower- and Middle-income Countries. *(Plast Reconstr Surg Glob Open 2021;9:e3657; doi: 10.1097/GOX.0000000000003657; Published online 24 June 2021.)*

INTRODUCTION

With the passing of the World Health Assembly resolution 68/15 for "Strengthening Emergency and Essential Surgical Care and Anesthesia as a Component of Universal Health Coverage"[1] and the ratification of the Sustainable Development Goals, member states have committed to scale up the delivery of essential surgery to achieve universal health coverage within their countries.[2,3] Childhood mortality and life-long disability can be substantially reduced by improving the accessibility of pediatric surgical care in low- and middle-income countries (LMICs). However, as noted by Ozgediz et al, "If surgery is the neglected step-child of global health, then pediatric surgery is the child not yet born."[4]

Although there exists no universally accepted methodology for designating bellwether procedures[5] it has been suggested that a "bellwether" satisfies the expectation that it describes a surgical procedure that, when recorded and studied, could facilitate the assessment of a hospital's ability to perform essential surgical care. The Lancet Commission on Global Surgery (LCoGS) used the concept of "bellwether procedures" to describe the availability of essential surgery at district hospitals, and more recently, there have been calls to expand the thinking around bellwether procedures to be more inclusive of the functioning of the broader surgical system.[6] However, the three adult bellwether procedures did not correlate with common and essential pediatric procedures for club foot, cleft lip (CL) and neonatal surgery.[7]

*From *NGO Strategic Consulting, Norfolk, Va.; † Hospital Das Clinicas, Craniofacial Unit, Hospital for Craniofacial Anomalies, University of Sao Paulo (USP), Sao Paulo, Brazil; ‡ Smile Train, New York, N.Y.; §Department of Surgery, School of Medical Sciences, Department of Maxillofacial Sciences, Dental School, College of Health Sciences, Kwame Nkrumah University of Science and Technology (KNUST), Kumasi, Ghana; ¶Dundee University Dental School, Dundee, Scotland, United Kingdom; and ||Global Cleft Lip and Palate Research Programme, Global Health Research Centre, Usher Institute, University of Edinburgh, Edinburgh, Scotland, United Kingdom.*

Received for publication January 19, 2021; accepted May 5, 2021.

DOI: 10.1097/GOX.0000000000003657

Disclosure: *Mr. Vanderburg is a compensated Consultant for Smile Train. Ms. Desai and Ms. Stieber are employees of Smile Train. All the other authors have no financial interest to declare in relation to the content of this article.*

A pediatric bellwether indicator would ideally be a routinely performed procedure, for a relatively common condition that, in itself, is rarely lethal at birth, but that should ideally be treated with surgery by a standard age. Conditions should be easy to diagnose, to minimize the confounding effects of delays or failures in diagnosis.

Given the differences between the types of burden of disease between adult and pediatric surgical patients (ie, high proportion of congenital birth defects requiring surgery in pediatric patients), there is a need for bellwether indicator(s) for pediatric surgery. However, many pediatric surgical conditions are unsuitable as bellwether indicators for a variety of reasons. These include a low prevalence of individual conditions, as well as early and rapid mortality (which may occur before diagnosis and referral for surgery). Lastly, conditions that are not easily diagnosed (eg, congenital inguinal hernia, cleft palate) based upon only a simple physical examination may be referred late (or not at all) for surgery. Thus, numbers of operations carried out for such procedures may be excessively influenced more by diagnostic failures than by limited surgical capacity. A pediatric surgery bellwether procedure needs to be minimally affected by such confounding variables to represent surgical capacity as accurately as possible.

In this article, we propose using age at CL surgery as a bellwether indicator for pediatric surgery in LMICs for the following reasons. CL and/or palate is the most common craniofacial birth condition, affecting one in 700 live births.[8] CL with or without an associated cleft palate (CP) requires no specialist skills or investigations to make a diagnosis. Thus, its value as a bellwether indicator for pediatric surgery is less likely to be confounded by diagnostic limitations and delays. This is in contrast to isolated CP, which is often missed unless a thorough intra-oral examination is performed.[9] Mortality associated with CL is poorly understood, but it is assumed that most infants survive while waiting for surgery, thus allowing analysis of age at surgery as an indicator of access to surgical care. Most surgical protocols aim to perform CL repair between the age of 3–6 months.[7] In contrast, there is less consensus on the optimum age for palate repair.[10] Facilities present within second-/third-level hospitals as described by the Global Initiative for Children's Surgery[11] are required for CL repair under a general anesthetic. Deidentified surgical data was used in this study. Therefore, no ethical approval was required.

LITERATURE REVIEW

The Disease Control Priority Project (DCP3)[4] described a package of essential surgery that if implemented, would address the majority of surgical burden of disease. This package of essential surgery included delivery of CL surgery to be performed at second- and third-level hospitals.

The LCoGS[12] found that essential surgery,[13] if addressed would avert 80% of the burden of surgical disease at district hospitals. The LCoGS identified three procedures that correlated with the majority of the other essential surgery procedures: emergency caesarian, emergency laparotomy, and open long bone fracture. These three procedures were described as "bellwether procedures." LCoGS authors proposed using the bellwether procedures as a proxy for understanding district hospital surgical system performance. The bellwether procedures were used to model the proportion of the population that had access to essential surgery within 2 hours. The 2-hour availability threshold was included as an indicator of the availability of surgery and as a recommendation from the Commission's report and emphasizes the importance of timely intervention.

Research has been done to understand other potential bellwether procedures (eg, neurosurgical care[5] at the district hospital, neonatal surgery[14]). There have also been calls to establish bellwether procedure(s) for pediatric surgery[15,16] as children represent a population with unique surgical needs, including anesthetic, preoperative, and postoperative capacity requirements.[17] Despite pediatric conditions accounting for a large proportion of unmet surgical needs,[18] pediatric surgery has been largely absent from discussions around surgical coverage.

The measurement of the prevalence of surgical burden of disease generally, and particularly related to pediatric surgery, is done at an early stage.[19] A larger burden of disease is accrued within a pediatric population due to the potentially life-long effect of the condition accruing disability[20] and of the impact of premature mortality across an expected lifetime.[21] These factors, combined with the greater proportion of children in LMICs, lead to estimates of a large pediatric surgical burden of disease in LMICs.

There have been urgent calls from the pediatric surgical community to collect and analyze data[22] and provide information for policy-makers in the allocation of resources for health system scale up.[23] The aim of this article is to propose the age at cleft lip (CL) repair as a bellwether indicator for pediatric surgery.

METHODS

Cleft Surgical Data

Surgeons in 73 countries, across 1110 hospital sites perform cleft surgical procedures supported by Smile Train and upload details of all cleft surgical procedures to Smile Train's online database, Smile Train Express. We selected the most recent complete calendar year (2019) for analysis. Surgical procedures include primary surgery (repair of the original CL and/or CP) and secondary (or revision) surgery. Variables such as patient choice and quality of original surgery influence whether, and at what age, secondary lip surgery is performed. Therefore, secondary surgery was not included in this analysis.

Data on all CL repairs for 2019 were exported from Smile Train Express. Number of procedures per country/center, age at surgery, and type of anesthesia were studied. Patients recorded as treated in "Palestinian territories" were excluded from the analysis due to the lack of available economic and health statistics.

Patients who present at an older age with an unrepaired CL and CP require unique consideration. Surgeons may opt to do primary CL repair and primary CP repair simultaneously (CLP) due to concerns about the patient and family's capacity to return for further surgery. These patients were included in the analysis of age at CL repair but are reported separately for clarity. Additionally, some older patients with isolated CL (no palatal involvement) may have surgery under a local anesthetic. These patients are reported for clarity, but only patients who had a general anesthetic are included in the analysis of age at CL repair.

Health Indicators

Country-level indicators were extracted from the World Bank and WHO[24] databases (Table 1). The authors chose to use LCoGS procedure rates versus World Bank procedure rates, which provided a more complete data set for comparison.

Statistical Analysis

Data were imported to Microsoft Excel, version 16.43.1 (Microsoft Corporation). Bivariate correlations were conducted between age at surgery for CL repair and the national patient data and the health and economic indicators accessed from the World Bank and WHO.

RESULTS

The total number of cleft surgical procedures reported in 2019, from 73 countries, was 104,349. Of these, 33,003 procedures were excluded for the following reasons: procedures recorded as secondary surgeries, surgeries that were performed on adults (age > 18 years), patients operated on in Gaza/West Bank (for whom no referenceable population-level data were available) and patients operated on under local anesthesia (Table 2).

DISCUSSION

A large proportion of the unmet pediatric surgical burden of disease is related to congenital anomalies[17] for which LMICs incur 94% of the burden[4]. When attempting to identify a bellwether indicator for safe pediatric surgery under general anesthesia, it is important to separate indicators of surgical capacity and access from delays in referral to surgical services as a consequence of late or missed diagnosis. The latter are, of course, important and relevant

Table 1. Indicators (and Sources) Used for Comparison to Smile Train Data

Indicator	Source
Total health care expenditure per capita	World Bank
Risk of catastrophic health expenditure for surgery	World Bank
Life expectancy at birth	World Bank
Maternal mortality rate	World Bank
Infant mortality rate	World Bank
Child mortality rate	World Bank
Specialist surgical workforce	World Bank
Hospital bed density	World Bank
GINI index	World Bank
Gross domestic product per capita	World Bank
Proportion of deliveries via cesarean section	WHO
Surgical procedure rates	LCoGS

Table 2. List of Exclusions*

Reason for Exclusion	No. Procedures
Secondary (revision) surgery only	29,534
Adult (over 18 y)	8874
Surgery performed in Gaza/West Bank	126
Surgery performed under local anesthesia	3404

*A single patient may have had more than 1 reason for exclusion.

Of the remaining 71,346 patients, 39,053 underwent CL repair, and 1126 underwent combined CL and CP repair (CLP). In total, 58.6% patients were men (Table 3), which would be expected as there is a male predilection in CL(P).

Table 3. Distribution of Primary CL, CLP, and CP Repairs by Gender and Age

Surgical Procedure	Girls n	Girls %	Boys n	Boys %	Total	Average Age (mo)
CL	15,145	38.8%	23,908	61.2%	39,053	19.5
CLP	366	32.5%	760	67.5%	1126	33.14
CP	14,053	45.1%	17,114	54.9%	31,167	37.76
Total	29,564	41.4%	41,782	58.6%	71,346	27.4

The average age at surgery for all primary surgery was 27.4 months. Primary CL surgery averaged 19.5 months, primary CP surgery averaged 37.76 months, and simultaneous primary CLP surgery average of 33.14 months (Table 3).

The results of the bivariate correlation demonstrate a moderate positive correlation between maternal mortality rate, infant mortality rate, and child mortality rate. A moderate negative correlation was also noted between age of primary CL (± CLP) surgery and life expectancy, proportion of deliveries via cesarean section, specialist surgical workforce, and LCoGS procedure rates. (Table 4).

Table 4. Correlation between Average Age at Surgery in Months versus National Health and Economic Data (r*)

	CL (n = 39,053)	CL ± CLP (n = 40,179)
Population	−−0.04	−0.04
Total health expenditure per capita (USD)	−0.31	−0.32
Risk of catastrophic health expenditure for surgery (%)	0.29	0.30
Life expectancy at birth (y)	−0.49	−0.50
Maternal mortality rate (per 100,000 live births)	0.43	0.43
Infant mortality rate (per 1000 live births)	0.55	0.55
Child mortality rate (per 1000 live births)	0.56	0.57
Deliveries via C-section (%)	−0.44	−0.46
Deliveries via C-section—poorest wealth quintile (%)	−0.39	−0.41
Specialist surgical workforce (per 100,000 population)	−0.37	−0.38
Hospital beds (per 1,000 population)	−0.29	−0.29
LCoGS procedure rates	−0.31	−0.32
GINI	0.13	0.13
GDP / capita (current USD)	−0.32	−0.33

*r = Pearson Correlation Coefficient.

Average age of patients was organized by procedure types (CL, CLP) by country, and World Bank groups. (Table 5).

Table 5. Number of Procedures, % Distribution of Procedures, and Average Age by World Bank Group

No. Procedures, % Distribution within Procedure Group, and Average Age by World Bank Income Group			
Income Group	No. Countries	CL Avg. Age (mo)	CLP Avg. Age (mo)
Low income	22	23.2	41.5
Low middle-income	31	18.3	43.7
Upper middle-income	17	15.6	26.6
High income	2	7.6	34

to neonatal care, new-born screening, and broader pediatric medical care. It may be argued that an ideal pediatric surgical service would contribute to raising awareness of surgically treatable conditions by training neonatal clinicians and pediatricians in early diagnosis of such conditions. However, a sensitive pediatric surgery bellwether indicator would ideally minimize the confounding impact of missed diagnoses and late referral for surgery.

CL addresses these criteria because, unlike many congenital conditions, it is easy to diagnose, even by non-healthcare professionals. Additionally, CL and CP are highly prevalent conditions (birth incidence estimated at one of every 700 live births).[25] Based on the prevalence of CL and CP as a congenital birth condition that comprise a large proportion of the unmet pediatric surgical burden of disease and the availability of multinational treatment data from a large INGO, the consideration of CL as a bellwether for pediatric surgical system performance is warranted.

Recommendations for optimum age for CL repair vary between countries. High-income countries such as the UK have standards that recommend CL repair between the ages of 3–6 months (unless there are specific clinical contraindications).[26] A study in Egypt, a lower middle-income country, also suggests the majority of CL cases are repaired between 3 and 6 months.[27] From the patient data reviewed, the average age at surgery for CL surgery was found to be 19.5 months and for CLP surgery, it was 33.14 months. The delayed age at surgery indicates a large backlog of unmet burden of disease for CL or CLP surgery and for pediatric surgery more generally. The average age of presentation was highest in low-income countries, progressively decreasing in age to low middle-income, upper middle-income, and high-income countries (Table 5). This progression points to the relationship between level of economic development and issues of accessibility and availability of a specific elective but essential pediatric surgical procedure.

The bellwether procedures described by the LCoGS used a 2-hour threshold to estimate timely access to essential surgical care. In contrast, timeliness of surgery for non-life-threatening congenital birth defects, including primary CL repair, are determined largely by the age of the child and are often considered in the timeframe of months and sometimes years. This time frame should conceivably include the time required for diagnosis, referral to an appropriate treatment center, and delivery of surgical care.

Our study showed that adults continue to present with unrepaired CLs indicating that there is a residual backlog that will take time to address. It is possible that improving access to, and awareness of, surgical treatment may increase the number of adults seeking treatment, potentially increasing the average age of primary lip repair if adult patients are included in any analysis. We therefore focused our analysis on the age of surgery in pediatric patients.

Comparing data from 73 countries, we have demonstrated that countries that achieve a younger average age at CL repair are those that have better indicators of overall health system performance as well as surgical system performance. Additionally, reductions in age at surgery could serve as a sensitive indicator of increasing and improving pediatric surgical capacity. Thus, supporting the case for age at CL repair as a pediatric surgery bellwether indicator.

Limitations

Through the granting process of Smile Train, the organization has an impact on the economy and capacity of the surgical environment. This impact has the primary effect of enabling more cleft surgery, but the broader impact on overall surgical activity is unknown. The data analyzed are from one organization and represent only operative data. A more meaningful metric of coverage may be to look at total national capacity for cleft surgery.

CONCLUSIONS

Untreated CL and palate are highly prevalent in LMICs. Patients who do not receive surgery are forced to live with life-long disability. These children are a part of the estimated 1.7 billion children who do not have access to surgery,[21] many of them requiring treatment for congenital birth defects.

Age at surgery for CL+CLP for Smile Train patients are correlated with broader indicators of health system (life expectancy, maternal mortality rate, infant mortality rate, etc) and surgical system performance (C-section rate, surgical specialist density, procedure rates etc). The correlation of actual patient treatment data and national level statistics would suggest that age at surgery for CL (± CLP) is a meaningful bellwether for pediatric surgery in LMICs. Increasing surgical capacity is essential for achieving the Sustainable Development Goals, and the utilization of this bellwether indicator for pediatric surgical care has the potential to inform policies for scale up and for understanding and addressing barriers to accessing pediatric surgical care.[25]

Dr. Felicity V. Mehendale, MD
Global Cleft Lip and Palate Research Programme
Global Health Research Centre
Usher Institute
University of Edinburgh
Doorway 1, Old Medical School, Teviot Place
Edinburgh EH8 9AG
E-mail: Felicity.Mehendale@ed.ac.uk

ACKNOWLEDGEMENT

Dr. Mehendale's research at the University of Edinburgh is supported by the Caledonian Heritable Foundation.

REFERENCES

1. Price R, Makasa E, Hollands M. Strengthening emergency and essential surgical care and anesthesia as a component of universal health coverage—addressing the public health gaps arising from lack of safe, affordable and accessible surgical and anesthetic services. *World J Surg.* 2015;39:2115–2125
2. Roa L, Jumbam DT, Makasa E, et al. Global surgery and the sustainable development goals. *Br J Surg.* 2019;106:e44–e52.
3. LeBrun DG, Chackungal S, Chao TE, et al. Prioritizing essential surgery and safe anesthesia for the post-2015 development

agenda: operative capacities of 78 district hospitals in 7 low- and middle-income countries. *Surgery*. 2014;155:365–373.
4. Jamison DT, Nugent R, Gelband H, et Al. Surgical interventions for congenital anomalies. In: *Disease Control Priorities*. 3rd ed. Washington, D.C.: International Bank for Reconstruction and Development/The World Bank; 2015:129–149.
5. Dewan MC, Baticulon RE, Ravindran K, et al. Pediatric neurosurgical bellwether procedures for infrastructure capacity building in hospitals and healthcare systems worldwide. *Childs Nerv Syst*. 2018;34:1837–1846.
6. Weiser TG. Bellwethers versus baskets: operative capacity and the metrics of global surgery. *World J Surg*. 2020;44:3310–3311.
7. Shi B, Losee JE. The impact of cleft lip and palate repair on maxillofacial growth. *Int J Oral Sci*. 2015;7:14–17.
8. Warkany J. *Congenital Malformation: Notes and Comments*. Chicago, Ill.: Year Book Medical Publishers; 1971.
9. Habel A, Elhadi N, Sommerlad B, et al. Delayed detection of cleft palate: an audit of newborn examination. *Arch Dis Child*. 2006;91:238–240.
10. Shaw W, Semb G, Lohmander A, et al. Timing of primary surgery for cleft palate (TOPS): protocol for a randomised trial of palate surgery at 6 months versus 12 months of age. *BMJ Open*. 2019;9:e029780.
11. Goodman LF, St-Louis E, Yousef Y, et al; GICS Collaborators. The global initiative for children's surgery: optimal resources for improving care. *Eur J Pediatr Surg*. 2018;28:51–59.
12. Meara JG, Leather AJ, Hagander L, et al. Global Surgery 2030: evidence and solutions for achieving health, welfare, and economic development. *Lancet*. 2015;386:569–624.
13. O'Neill KM, Greenberg SL, Cherian M, et al. Bellwether procedures for monitoring and planning essential surgical care in low- and middle-income countries: caesarean delivery, laparotomy, and treatment of open fractures. *World J Surg*. 2016;40:2611–2619.
14. Ford K, Poenaru D, Moulot O, et al. Gastroschisis: bellwether for neonatal surgery capacity in low resource settings? *J Pediatr Surg*. 2016;51:1262–1267.
15. Qazi SH, Dogar SA, Dogar SA, et al. Global perspective of paediatric surgery in low and middle income countries. *J Pak Med Assoc*. 2019;69(suppl 1):S108–S111.
16. Ullrich S, Kisa P, Ozgediz D. Global children's surgery: recent advances and future directions. *Curr Opin Pediatr*. 2019;31:399–408.
17. Butler EK, Tran TM, Nagarajan N, et al; SOSAS 4 Country Research Group. Epidemiology of pediatric surgical needs in low-income countries. *PLoS One*. 2017;12:e0170968.
18. Grabski DF, Kakembo N, Situma M, et al. Burden of emergency pediatric surgical procedures on surgical capacity in Uganda: a new metric for health system performance. *Surgery*. 2020;167:668–674.
19. Sitkin NA, Farmer DL. Congenital Abnormalities in the context of global surgery. *Semin Pediatric Surg*. 2016;25(1):15–18.
20. Yousef Y, Lee A, Ayele F, et al. Delayed access to care and unmet burden of pediatric surgical disease in resource-constrained African countries. *J Pediatr Surg*. 2019;54:845–853.
21. Wu VK, Poenaru D, Poley MJ. Burden of surgical congenital anomalies in Kenya: a population-based study. *J Trop Pediatr*. 2013;59:195–202.
22. Mock C. Surgery's role in addressing population health needs. *J Pak Med Assoc*. 2019;69(suppl 1):S95–S97.
23. Smith ER, Concepcion T, Lim S, et al; Global Initiative for Children's Surgery. Disability peights for pediatric surgical procedures: a systematic review and analysis. *World J Surg*. 2018;42:3021–3034.
24. Global Health Observatory data repository. Available at https://apps.who.int/gho/data/node.main.BIRTHSBYCAESAREAN?lang=en%20on. Accessed October 8, 2020
25. Poenaru D, Pemberton J, Frankfurter C, et al. Establishing disability weights for congenital pediatric surgical conditions: a multi-modal approach. *Popul Health Metr*. 2017;15:8COMP: doi:10.1186/s12963-017-0125-5
26. NHS Standard Contract for Cleft Lip and/or Palate Services Including Non-Cleft Velopharyngeal Dysfunction (Vpd) (All Ages). Available at https://www.england.nhs.uk/wp-content/uploads/2013/06/d07-cleft-lip.pdf. Accessed October 8, 2020.
27. Abulezz TA, Elsherbiny AK, Mazeed AS. Management of cleft lip and palate in Egypt: a national survey. *Indian J Plast Surg*. 2018;51:290–295.