Burden of Surgical Congenital Anomalies in Kenya: A Population-Based Study

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Summary

Background: Congenital malformations are a significant component of the global burden of disease among children, accounting for 25 million disability-adjusted life years (DALYs) worldwide. Unfortunately, efforts to estimate the burden of pediatric surgical disease in Africa are limited by the absence of population-based data. The objective of this study was to estimate both the prevalence and the disease burden of several common congenital surgical malformations among children in Kenya.

Methods: Community volunteers randomly surveyed households at sites across Kenya. Caretakers were asked to identify on a photographic portfolio, several congenital malformations present among the children in their household, including club foot, hypospadias, hydrocephalus, spina bifida/encephalocele, cleft lip, bladder extrophy and imperforate anus. DALYs were then calculated based on life expectancy tables and published and estimated disability weights for the conditions encountered.

Results: The caregivers of 5559 children (54% female) were surveyed in 1909 households, 56% of which were rural, 31% suburban and 12% urban. The overall prevalence of congenital malformations was 6.3 per 1000 children, amounting to 54–120 DALYs per 1000 children, depending on the life tables used. The most prevalent condition in the survey was club foot, whereas spina bifida had the highest burden of disease.

Discussion: This study is the first to document the prevalence of selected surgical congenital malformations among children in Kenya and the burden of disease associated with them. The results will serve to inform strategies aimed at reducing the unmet burden of surgical disease in resource-limited regions.

Key words: burden of disease, congenital malformations, Africa, pediatric surgery, population survey.

Congenital malformations are a significant cause of morbidity and mortality, especially in low- and middle-income countries (LMICs). Debas et al. [1] estimate that congenital malformations account for 9% of the surgical burden of disease (BoD), contributing to the disability experienced by 150 million children around the world, with disabilities being more common among children in LMICs [2]. These disabilities represent a burden placed on child development and family responsibilities, and are both the cause and the result of poor socioeconomic status [2, 3].

The burden of childhood surgical disease from congenital malformations is significant in sub-Saharan African settings. In Gambia, such conditions represented the second highest proportion of children presenting for surgical care, preceded only by injuries [4, 5]. Surgical procedures to address congenital malformations comprised 40% of all operations performed at a major teaching hospital in northern Nigeria [6], and nearly one in four children presenting at a major sub-Saharan hospital suffered from congenital malformations [4, 5]. In studies that...
focused on specific malformations and/or geographic regions, the incidence of congenital malformations in LMICs was calculated to be between 3.9 and 11.8 per 1000 live births [7, 8]. However, in such studies, congenital malformations are likely to be underreported due to stigmatization or fear associated with being rejected by the community [9, 10].

Based on the premise that about half of all congenital malformations are surgical [1], an obvious method of decreasing the burden of childhood disease is the prevention and early treatment of neonatal surgical conditions, many of which result in disability or death when left untreated [11]. Surgical care can have a significant impact on the health burden of congenital malformations, which account for 3% of pediatric deaths and 24 million disability-adjusted life years (DALYs) worldwide [12]. DALYs are a widely accepted metric of disease burden, as they take into account both the years of life lost and the reduced quality of life resulting from disability.

There are limited data on the extent and impact of pediatric disabilities in LMICs. In response to the increased recognition of disabilities as a major global health concern, a comprehensive review of three large databases was undertaken to identify publications focused on pediatric intellectual, hearing, speech, vision, motor and neurological disabilities in LMICs [9]. The reviewers noted a paucity of information on most pediatric disabilities, with the exception of intellectual and hearing impairments, and few LMICs have epidemiological information on pediatric disabilities in their region. The Kenya National Survey for Persons with Disabilities recently estimated a 2.4% overall prevalence of disability among Kenyan children aged 0–14 years [13]. There is a need, however, for more detailed data on specific disabilities to identify the most effective health care interventions for alleviating the morbidity and mortality of pediatric congenital disease. Moreover, quantification of the disease burden of congenital malformations potentially avertable by surgery allows for comparisons with other health interventions competing for limited resources. We are not aware of any population-based study quantifying the burden of surgically treatable congenital malformations in an African country. The objective of this study therefore was to estimate the prevalence and burden of selected surgical congenital malformations among children in Kenya.

Methods

Design

The study is a population-based prevalence survey. It entailed field workers surveying households and looking for family members 16 years of age or younger who have any of a set of eight visible congenital malformations. The field workers were recruited by liaising with the local community health office at each designated site. The workers were already volunteering at, or employed by, the health office and were known in the community and familiar with visible disabilities. They were trained on-site by one of three study coordinators, who would then follow them on their initial home visits to ensure adequate performance. The study sample consisted of caretakers—parents and other guardians—from a sample of 10 urban and rural locations in Kenya. These locations were selected to match the population distribution in Kenya by province.

Within each location, interviews were conducted by the field worker in a random selection of households, using a method of the World Health Organization Expanded Programme on Immunization, as described by Durkin et al. [14]. According to this method, in each study community, the field worker began in a central location (such as the community’s local government health center or chief’s office), and walked in one of four possible directions (north, south, east or west). The field worker, through a random process, then selected a household to interview that was:

(i) along the left or right side of the road (by throwing a dice, with 1–3 = left, 4–6 = right); and
(ii) between one and six houses away from his/her starting location (e.g. for a four, go to the fourth house on the side of the road that was decided in step 1).

A household was defined as a family, including its caretakers/servants, who shared a single front door within a residential property that borders the side of the road. The field worker continued in the direction of travel using the aforementioned process of sampling until the community boundary was reached. This process was then repeated in the three other remaining directions. In the event that the direction of travel was no longer possible (such as at a T-junction), the field worker used a random process to decide a new direction to travel (left or right) until the earliest opportunity to resume the original direction. In the event of no children living in the selected household, or if nobody was at home, the next closest household was approached.

A standard portfolio of photographs of congenital malformations was provided to each community worker as a screening tool to help caretakers identify the conditions of interest (cleft lip, club foot, spina bifida/encephalocele, hydrocephalus, hypospadias, bladder extrophy and imperforate anus). These conditions were chosen because they are easily identifiable, visible deformities for which surgical treatment is available in the country. Caretakers from each interviewed household were presented with the screening tool and shown additional pictures if there was confusion in the identification of the condition.
Caretakers were also asked to provide additional information, including education level, household income and their child(ren)'s place of birth. The survey and the photographic portfolio are available for download from http://dl.dropbox.com/u/9254603/KIDS%20study/KIDS%20survey.docx and http://dl.dropbox.com/u/9254603/KIDS%20study/KIDS%20photo%20portfolio.pdf, respectively. Similar door-to-door interview techniques, along with the use of photographic albums, have been previously used to determine the community-based prevalence rate of pediatric disabilities [7, 15].

**Sample size**

The overall prevalence of pediatric disabilities in community-based studies in LMICs varies between 0.4 and 12.7% [3, 15–20], and the prevalence of neurological and motor disabilities among children in LMICs varies between 0.2 and 6.1% and 0.2 and 2.8%, respectively [17, 18, 21, 22]. Using a point estimate of 4% as the prevalence of all the disabilities that we seek to identify in our study, and applying standard statistical formulae for calculating 95% confidence intervals from the binomial distribution, we estimated that a survey of 5000 children would provide 95% confidence intervals of 3.5 and 4.6%. We therefore intended to survey an average of 500 children within each of the 10 communities sampled. Assuming an average household size of 5.3 [23], we anticipated, on average, interviewing the caretakers of 3 children per household, 167 households per community and 1670 households in total. The sample size calculation was performed using a standard online statistical calculator [24].

**DALY calculation**

BoD estimates were calculated using previously established methods [25] and universal and country-specific life tables provided by the Global Burden of Disease (GBD) study [26] and the World Health Organization [27]. DALYs are calculated as the sum of life years lost due to premature mortality (YLL) and years lived with disability (YLD), where YLD is the product of the life expectancy at disease onset and the disability weight (DW). In light of the fact that the current study only attempted to identify prevalent conditions among living children and that the study assumed that these conditions do not affect mortality rates in these children, the DALY calculation included only the YLD. As the GBD study provides DWs for only a few selected pediatric surgical conditions, DWs for the remainder were estimated using the scale endorsed by the World Health Organization [28] and shown in Table 1.

**Statistical analysis**

Descriptive statistics were applied to the demographic variables and DALYs, using Microsoft Excel® software.

**Ethics**

This study was approved by the Research Ethics Committee of AIC Kijabe Hospital in Kijabe, Kenya. Before entering each locale, the community chiefs were first approached for written consent to undertake this study in their jurisdiction. Verbal consent was also obtained from the household caretakers before proceeding with the interviews.

**Results**

Between July 2009 and March 2010, the caregivers of 5559 children were surveyed in 1909 households. All eight provinces were sampled at or above their

<table>
<thead>
<tr>
<th>Description</th>
<th>Weight</th>
</tr>
</thead>
<tbody>
<tr>
<td>Limited ability to perform at least one activity in one of the following areas: recreation, education, procreation or occupation</td>
<td>0.1</td>
</tr>
<tr>
<td>Limited ability to perform most activities in one of the following areas: recreation, education, procreation or occupation</td>
<td>0.2</td>
</tr>
<tr>
<td>Limited ability to perform activities in two or more of the following areas: recreation, education, procreation or occupation</td>
<td>0.4</td>
</tr>
<tr>
<td>Limited ability to perform most activities in all of the following areas: recreation, education, procreation or occupation</td>
<td>0.6</td>
</tr>
<tr>
<td>Needs assistance with instrumental activities of daily living such as meal preparation, shopping or housework</td>
<td>0.8</td>
</tr>
<tr>
<td>Needs assistance with activities of daily living such as eating, personal hygiene or toilet use</td>
<td>0.9</td>
</tr>
</tbody>
</table>
intended target levels. There were no non-responders in the study: of all caretakers who were invited to participate in the study, none refused to participate. The distribution of respondents by location is shown in Table 2. Figure 1 depicts the location of the homesteads surveyed—most children resided in the population-rich Rift Valley and Eastern provinces. Fifty-five percent of the households were in rural areas, and there were 2.9 eligible children, on average, per interviewed household.

Additional demographic characteristics of the caretakers are presented in Table 3. The highest education level among the majority of caretakers was primary school or no formal education at all, with fathers more likely to attain higher education. Almost half of the households (45%) included children who were born in the home rather than at a clinic or hospital. The median household income was 5000 KSh/month (range, 0–6,000,000) or approximately US$65/month. These demographics are fairly consonant with published socio-economic studies for Kenya [32].

There were 35 malformations encountered in the eight selected conditions sought by the current study, 19 males and 16 females (Table 4). Forty-nine percent of the children identified were <5 years of age, 31% were between 5 and 10 years, and 20% were >10 years old.

The population prevalence of these malformations and associated DWs is shown in Table 4. The overall rate of congenital malformations was 6.3 per 1000 children. Club foot was the most prevalent malformation encountered (2.9 per 1000 children) both in males and females.

Figure 2 depicts the spread of estimated DALYs from each malformation by three different calculation methods. Spina bifida carried the highest BoD by all methods (106–234 DALYs in the sample), whereas cleft lip the lowest (3.5–7.7 DALYs). The lowest estimate of total burden of surgical disease in the sample was 303 DALYs (54 DALYs/1000 children) when age-weighted discounted Kenya YLL values were used, and 669 DALYs (120 DALYs/1000 children) when using universal, unweighted and undiscounted YLL values. The standard deviation of the DALYs per 1000 estimated by all methods was 29.

Figure 3 compares the prevalence of the selected malformations with the mean BoD (as calculated by the aforementioned three methods), by gender, associated with them—both per 1000 children. While the prevalence is plotted in increasing order, the corresponding BoD varies widely in order.

Spina bifida resulted in the greatest BoD, with rates of 19–42 DALYs per 1000 children. When stratified for gender, hydrocephalus represented the greatest BoD among males (10–20.7 DALYs per 1000 children) and spina bifida for females (15.1–32.2 DALYs per 1000 children).

**Discussion**

To our knowledge, this is the first population-based study to estimate the prevalence and burden of congenital surgical malformations in Kenya. Population-based estimates of congenital malformations and pediatric disabilities in LMICs are scarce.

One reason for the scarcity of data is the challenge of reliably selecting a random non-biased sample in LMIC populations. Many similar surveys are based
on hospital-based birth data [33], which naturally miss the significant percentage of births in LMICs that occur outside health care facilities. Moreover, surveys are often done in urban areas, because of the challenges of rural surveying. Address registries, village maps and telephone directories were not available for any of the communities targeted, which led us to the sampling method described previously. This simple method allowed us to quickly and inexpensively train community workers to administer the survey. Moreover, the use of photographic portfolios in interviews has been shown to be an effective way of helping respondents identify various congenital malformations [7, 15]—especially important in resource-poor settings where illiteracy may be an issue.

The significance of the study lies in its ability to derive population-based data for common visible congenital anomalies. BoD metrics hold advantages over traditional ones such as prevalence, as demonstrated by Figure 2. Depending on the DW of each condition and the age of each child, some conditions (e.g. imperforate anus, spina bifida and hydrocephalus) carry a disproportionately higher BoD than implied by their prevalence whereas the opposite is true for less disabling conditions such as club foot, cleft lip and hypospadias.

The nature of the survey questions resulted in prevalence rates for these conditions, which may differ from their corresponding incidence rates. While incidence rates are more generally accepted as the metric in epidemiological studies of congenital conditions, prevalence estimates are needed for BoD calculations [14–16]. Prevalence directly allows the estimation of surgical backlog in any area, as well as unmet need or avertable DALYs [25].

Although comparisons with other studies should be made with caution, the prevalence rates obtained

### Table 3
Demographic characteristics of caretakers interviewed

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Number</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Household location</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Urban</td>
<td>232</td>
<td>12.2</td>
</tr>
<tr>
<td>Suburban</td>
<td>634</td>
<td>33.2</td>
</tr>
<tr>
<td>Rural</td>
<td>1043</td>
<td>54.6</td>
</tr>
<tr>
<td><strong>Child(ren)'s place of birth</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Home</td>
<td>454</td>
<td>23.8</td>
</tr>
<tr>
<td>Local clinic</td>
<td>963</td>
<td>50.4</td>
</tr>
<tr>
<td>Both</td>
<td>411</td>
<td>21.5</td>
</tr>
<tr>
<td>No children/declined to answer</td>
<td>81</td>
<td>4.2</td>
</tr>
<tr>
<td><strong>Income distribution</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Average monthly income per household (KES)</td>
<td>1101</td>
<td>2.9</td>
</tr>
<tr>
<td>Percent share of wealth</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Poorest 20%</td>
<td>3036</td>
<td>8.0</td>
</tr>
<tr>
<td>Second 20%</td>
<td>4770</td>
<td>12.5</td>
</tr>
<tr>
<td>Middle 20%</td>
<td>7449</td>
<td>19.5</td>
</tr>
<tr>
<td>Fourth 20%</td>
<td>21,775</td>
<td>57.1</td>
</tr>
<tr>
<td>Highest 20%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Declined to answer: 562 households</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Education level</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Did not attend school</td>
<td>233 (12.2)</td>
<td>183 (9.6)</td>
</tr>
<tr>
<td>Primary</td>
<td>869 (45.5)</td>
<td>631 (33.1)</td>
</tr>
<tr>
<td>Secondary</td>
<td>546 (28.6)</td>
<td>608 (31.8)</td>
</tr>
<tr>
<td>College</td>
<td>166 (8.7)</td>
<td>201 (10.5)</td>
</tr>
<tr>
<td>University</td>
<td>39 (2.0)</td>
<td>69 (3.6)</td>
</tr>
<tr>
<td>Not applicable/declined to answer</td>
<td>56 (2.9)</td>
<td>217 (11.4)</td>
</tr>
</tbody>
</table>

### Table 4
Disability weight, number and prevalence of congenital malformations

<table>
<thead>
<tr>
<th>Condition</th>
<th>Disability weight</th>
<th>Number</th>
<th>Prevalence (per 1000)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bladder extrophy</td>
<td>0.6</td>
<td>1</td>
<td>0.2</td>
</tr>
<tr>
<td>Cleft lip</td>
<td>0.05</td>
<td>2</td>
<td>0.4</td>
</tr>
<tr>
<td>Club foot</td>
<td>0.1</td>
<td>16</td>
<td>2.9</td>
</tr>
<tr>
<td>Encephalocele</td>
<td>0.6</td>
<td>2</td>
<td>0.4</td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>0.4</td>
<td>5</td>
<td>0.9</td>
</tr>
<tr>
<td>Hypospadias</td>
<td>0.1</td>
<td>5</td>
<td>0.9</td>
</tr>
<tr>
<td>Imperforate anus</td>
<td>0.85</td>
<td>1</td>
<td>0.2</td>
</tr>
<tr>
<td>Spina bifida</td>
<td>0.6</td>
<td>3</td>
<td>0.5</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>35</strong></td>
<td><strong>6.3</strong></td>
<td><strong>4.4</strong></td>
</tr>
</tbody>
</table>

The nature of the survey questions resulted in prevalence rates for these conditions, which may differ from their corresponding incidence rates. While incidence rates are more generally accepted as the metric in epidemiological studies of congenital conditions, prevalence estimates are needed for BoD calculations [14–16]. Prevalence directly allows the estimation of surgical backlog in any area, as well as unmet need or avertable DALYs [25].
in this study seem to be lower than in other published reports [7, 8, 34]. This may be in part due to the survey method, as detailed later in the text. Moreover, the study generated prevalence, rather than incidence, rates. In situations where the congenital malformation carries a low mortality (e.g. club foot, hypospadias or cleft lip), the prevalence rate should be similar to the yearly incidence rate. For instance, club foot was estimated in our study at 2.9 per 1000, twice the rate recently found in the Uganda Sustainable Clubfoot Care Project [33]. Given, however, that most malformations still carry with them some risk of death, our results would therefore tend to underestimate the true incidence of the congenital malformations of interest. Therefore, this approach offers minimum prevalence and BoD estimates for the population in question.

The DALY calculations were significantly dependent on the life expectancy tables used—although all values were reassuringly well within the same order of magnitude. The age-weighted life tables slightly increased the DALY values, whereas future discounting decreased the values by roughly 50%. As discussed earlier, the choice of age-weighting and discounting is debatable, carrying a fair amount of social value judgment and therefore uncertainty [35]. In the absence of a clear consensus, sensitivity analysis at least presents a spectrum of plausible results [35, 36]. The use of universal, rather than country-specific, life tables is also not standardized. While the majority of surgical studies simply followed the universal tables introduced in the original GBD study [28], the availability of country-specific tables allows us and others to be more specific in the calculations [37].

The BoD associated with congenital malformations is significant by comparison with adult surgical or medical conditions. This is due to a unique mix of features of congenital malformations: young age, typically infant (thus maximum number of years potentially to be lived with a disability); often severe nature (thus associated with either premature death or high DW); stable, chronic condition (unlike trauma or other surgical emergencies, in the absence of treatment, the disability simply persists for decades) and frequently some measure of residual life-long disability even after successful treatment (for instance, impaired mobility in spina bifida, speech problems in cleft palate or incontinence in imperforate anus). The accurate measurement of the BoD associated with congenital malformations can therefore play a significant role in the efforts to document the importance of surgical intervention for congenital malformations. In as much as the GBD study set out ‘to disentangle epidemiology from advocacy in order to produce objective, independent and demographically plausible assessments of the burdens of particular conditions and diseases’ [35], such objective assessments as can be generated must be put to use for the sake of improved health care for those in need [38].

Ultimately, an area-wide survey of congenital malformations achieves much more than help derive some prevalence and BoD values. As Pirani et al. [33] found out in their country-wide Ugandan study, their survey raised awareness of congenital malformations within the population, and provided...
a unique opportunity for mass education on congenital disabilities and their treatment.

Limitations

There are several limitations to this study. In the first place, we based our sample size calculation on a point estimate that is greater than the true prevalence calculated at the end of the study. On data analysis, this produced confidence intervals that were skewed around their point estimates. Such values should be interpreted with some caution, as they do not precisely mirror the binomial distribution that the sample size was based on.

Secondly, the results of this study may underestimate due to the caretakers’ difficulty in disclosing their children’s health status. In our setting, the sampling requirements and literacy considerations resulted in the natural use of community workers for interviewing caretakers. However, the use of interviewer-administered surveys for potentially sensitive topics may lead to less truthful information gathered when compared with other methods such as paper-based surveys [39]. In fact, we noticed caretakers commonly responding that they have seen children in their community who had conditions mentioned in the portfolio. This may indicate parents’ willingness to participate in the survey, but an unwillingness to disclose when the child with the malformation in question was theirs [9, 10].

Thirdly, the photographic portfolio was useful for most, but not all, conditions surveyed. In the case of genitourinary and anorectal conditions, cultural sensitivity required that photographs be only used if requested by the respondent, which may have led to some inaccuracy in the responses.

Finally, the DWs used in this study are simple estimates using the rather coarse WHO tool. The fact is that the GBD study has not generated many DWs within surgery, especially pediatric surgery. Currently there are published DW values for only six pediatric surgical conditions (abdominal wall defects, anorectal atresia, cleft lip and palate, esophageal atresia, congenital heart anomalies and spina bifida) [26]. It is obvious that an accurate measurement of the burden of pediatric surgical disease will require the formal establishment of DWs associated with a large variety of pediatric surgical conditions.

Future Considerations

This study provided an important first step in establishing prevalence rates and disease burden for surgically correctable congenital malformations in Kenya, a topic that has received little previous attention. The project reinforced the feasibility of low-scale population-based BoD studies in resource-limited settings. Future studies may include surveys assessing both the incidence and prevalence of congenital malformations, as well as larger sample sizes. Further studies also need to cover a broader range of both congenital and acquired malformations—this study focused on relatively common and externally visible congenital anomalies—to provide a more comprehensive scope of the burden of congenital disease and disability that can potentially be treated by surgery.

The recognition of surgery as an effective intervention in LMICs has been hindered by the perception that it is a non-cost-effective luxury [40]. In recent years, however, surgery has gained attention as a cost-effective way of addressing the global BoD [11]. Similarly to the existing cost effectiveness analysis (CEA) studies in general and trauma surgery [41, 42] in LMICs, CEA studies in pediatric surgery are essential tools in the global advocacy effort toward appropriate resource allocation for the surgical treatment and long-term rehabilitation of children with congenital malformations.

References