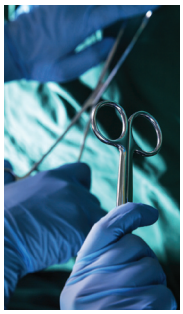

Chapter 8

Surgical Interventions for Congenital Anomalies

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INTRODUCTION

Great strides have been made during the past 50 years in the diagnosis and management of congenital anomalies, once referred to as birth defects. Formally fatal conditions can now be treated with success rates exceeding 90 percent. Yet improvements in care have been largely limited to high-income countries (HICs), even though many anomalies can be cured with simple operations (Chirdan, Ngiloi, and Elhalaby 2012). If surgery is the neglected stepchild of global health (Farmer and Kim 2008), then pediatric surgery is the child not yet born.

Improving the accessibility and quality of pediatric surgical care in low-income (LICs) and lower-middle-income countries (LMICs) has the potential to substantially reduce childhood mortality and lifelong disability. Data on congenital anomalies in these countries are sparse, including on the incidence (conservatively estimated at 3 percent to 6 percent [CDC 2012; Christianson, Howson, and Modell 2006]), country-specific differences in disease burden, and cost-effective interventions. These areas of knowledge must be developed to identify implementation and surveillance priorities, and to advocate for resources.

This chapter briefly summarizes the growing body of knowledge on surgical congenital anomalies in LICs and LMICs, highlights prevalent anomalies that exemplify the unrealized promise of pediatric surgery, and concludes with crucial future steps to reduce the burden of disease.

Chapter 21 addresses economic evaluation of cleft lip and palate repair.

MORBIDITY, MORTALITY, AND ADDITIONAL ADVERSE CONSEQUENCES

Congenital anomalies are one of the leading causes of global disease, responsible for a staggering 57.7 million disability-adjusted life years (DALYs) lost worldwide (WHO 2013b). DALYs—a measure of the number of healthy life years lost to premature death or disability—are an established metric for the burden of disease.

Current estimates of the surgical burden of disease are acknowledged to be a "best educated guess," given the "near total lack of pertinent data" (Jamison and others 2006, 1246). Even less is known about pediatric surgical disease (Bickler and Rode 2002). The studies that have begun to fill this knowledge gap paint a brutal picture. The burden of congenital anomalies falls most heavily on LICs and LMICs, where 94 percent of anomalies occur (WHO 2012). Higher fertility rates translate to higher birth rates and more children born with anomalies. Disease incidence (or frequency of disease occurring in the population) is also higher, a phenomenon attributed to higher micronutrient and macronutrient deficiencies,

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exposure to teratogens, prevalence of intrauterine infection, and self-medication with unsupervised drugs or traditional remedies (Christianson, Howson, and Modell 2006; Penchaszadeh 2002). Though decreased fertility may reduce incidence of anomalies, most are not otherwise preventable and are treated through surgical interventions.

Some anomalies are "quick fixes" that can be easily repaired; others require staged, or multioperation, surgical interventions, and delays in treatment may result in lifelong illness, disability, and poor quality of life. The paucity of surgical resources in LICs and LMICs means that anomalies attributed to the former category in HICs often fall into the latter in LICs and LMICs. Anomalies resulting in visible deformity—such as clubfoot and cleft lip—cause stigma, which can trigger abandonment or infanticide. Invisible deformities that result in chronic disability can lead to similar outcomes. A long-term, "incurable" anomaly may also endanger families' well-being because key resources are allocated to care for the afflicted child. Families may fracture, with one or both parents leaving the child with other family members.

Improving the pediatric surgical capabilities of LICs and LMICs will dramatically reduce this burden. Because children are the future economic engine powering the development of these countries, the value of investing in surgical care for children extends beyond DALYs averted to encompass the future socioeconomic well-being of LICs and LMICs themselves. It is critical to address the gaps in knowledge that impede the development of effective care systems.

Data Collection Challenges

Many LICs and LMICs lack rigorous congenital anomaly surveillance programs, making calculations of incidence and prevalence (the total number of individuals in the population with a given disease) difficult (Penchaszadeh 2002). Estimates, which range from 4 to 12 cases per 1,000 births, likely undervalue the problem because of stigma and exclusion (Bickler and others 2010; Goksan and others 2006; Wu and Poenaru 2013). LICs and LMICs often report incidence in cases over time, as opposed to using standard metrics, such as incidence per 10,000 live births. Incidence and prevalence data are also skewed by the survivability of the anomaly under consideration. Because children with anomalies that are not immediately life threatening are more likely to reach medical centers, the relative incidence and prevalence of immediately life-threatening impairments appears comparatively lower in hospital-based data (Nandi, Mungongo, and Lakhoo 2008). Population-based surveys—which directly collect data from noncentralized sites—are one approach to addressing this challenge (Wu, Poenaru, and Poley 2013).

Mortality Rate Estimation Challenges

The burden of disease associated with congenital anomalies in LICs and LMICs is most often calculated as the mortality rate, including neither measures of morbidity nor the cost of ongoing illness (table 8.1). Analysis of mortality data in these countries can be challenging; for example, autopsies were performed in only 0.8 percent of nearly 1,100 neonatal deaths in Benin. In all examined cases, autopsies provided additional information on the cause of death (Ugiagbe and Osifo 2012). Furthermore, a high proportion of children with surgical diseases do not reach a health facility and die at home or in transit, suggesting a sizable hidden mortality (Mo Suwan and others 2009; Ozgediz and others 2008). Nonfatal anomalies can result in extensive, ongoing morbidity. The burden of disease is grossly underestimated if this measure of impairment is not included. Extant calculations do, however, highlight marked disparities in outcomes between HICs, on the one hand, and LICs and LMICs, on the other hand.

CHALLENGES TO PROVIDING CARE FOR CONGENITAL ANOMALIES

Despite the higher incidence of congenital anomalies in LICs and LMICs, lower population prevalence is seen compared with HICS, most likely indicating a high infant and child mortality rate (Christianson, Howson, and Modell 2006). *In LICs and LMICs, up to 10 percent of infants die during the neonatal period* (Zupan 2005); a considerable portion of this mortality can reasonably be attributed to congenital anomalies.

Treatment Delays

Heightened mortality rates stem from delays in treatment caused by the paucity of health professionals trained to identify and treat anomalies and by pejorative cultural beliefs surrounding anomalies. In LICs and LMICs, many births occur at home, either with no assistance or with traditional birth attendants (TBAs), and patients must often travel great distances to reach medical facilities. Hypothermia—a dangerous drop in body temperature—is common following medically unsupervised transport over long distances, with severe repercussions on patient outcomes (Agarwala and others 1996;

Table 8.1 Prevalence and Mortality of Selected Congenital Anomalies in Selected Countries

	High-income countries		Selected countries		
Congenital anomalies	**Reference prevalence**	**Reference mortality (percent)**	**Country of reporting institution**	**Prevalence at reporting institution**	**Mortality (percent)**
Anorectal malformations	1 per 5,000 live births[a]	<5[a]	Iran, Islamic Rep.[b]	22 (1993–96)	27.3 (1993–96)
				106 (2002–05)	13.2 (2002–05)
			Nigeria[c]	88 over 17 years	30.2
			India[d]	125 over 2 years	22.0
			Nigeria[e]	81 over 8 years (38 percent of neonatal obstructions)	32.0
			Nigeria[f]	54 over 10 years	20.4
			Ethiopia[g]	27 over 5 years	33.0
			India[h]	948 over 14 years	15.0–20.0 (staged repair)
					4.5 (primary repair)
			Nigeria[i]	55 over 10 years	20.0
Hirschsprung's disease	1 per 5,000 live births[a]	Less than 5–10[j, k]	Iran, Islamic Rep.[b]	8 (1993–96)	25.0 (1993–96)
				50 (2002–05)	4.0 (2002–05)
			Nigeria[e]	30 over 8 years (14 percent of neonatal obstructions)	20.0
			Nigeria[i]	24 over 10 years (18.7 percent of neonatal obstructions)	20.8
			Bangladesh[l]	1,273 over 5.5 years	14.3
			Burkina Faso[m]	52 over 7 years	16.0
			Eritrea[n]	11 over 5 years	9.1
			Nigeria[o]	78 over 10 years	22.6
Congenital heart disease	8.2 per 1,000 live births (Europe)[p]	3–7[q]	Guatemala[r]	1,215 over 8 years	10.7 (overall) 32.1 (highest-risk patients)
	6.9 per 1,000 live births (North America)[p]		India[s]	330 over 8 years	21.4 (1999–2001)
					4.3 (2002–06)
			Nepal[t]	5.8 per 1,000 children	20.2
			Sri Lanka[u]	102 over 1 year	18.6
Esophageal atresia	One per 3,500 live births[a]	5–15[a]	India[v]	50 over 2 years	30.0
			Barbados[w]	2.06 per 10,000 live births	30.8
			Saudi Arabia[x]	48 over 20 years	25.0
			Bangladesh[y]	21 over 2 years	47.6
			China[z]	15 over 10 years	46.7 following surgery
			Malaysia[aa]	52 over 10 years	23.0

table continues next page

Table 8.1 Prevalence and Mortality of Selected Congenital Anomalies in Selected Countries (continued)

	High-income countries		Selected countries		
Congenital anomalies	**Reference prevalence**	**Reference mortality (percent)**	**Country of reporting institution**	**Prevalence at reporting institution**	**Mortality (percent)**
			Pakistan[ab]	80 over 1 year, excluding isolated esophageal atresia patients	58.0
			Saudi Arabia[ac]	94 over 15 years	30.8
			Iran, Islamic Rep.[b]	22 (1993–96)	75.0 (1993–96)
				106 (2002–05)	58.8 (2002–05)
			Ethiopia[g]	12 over 5 years	91.7
Gastroschisis	3.3 to 5 per 10,000 live births[a, ad, ae]	1 (30-day mortality)[af]	Ghana; Nigeria; South Africa[af]	Over four years:	23.0 (30-day mortality)
				2 (Ghana)	
				5 (Nigeria)	
				19 (South Africa)	
			Iran, Islamic Rep.[b]	2 (1993–96)	100 (1993–96)
				7 (2002–05)	85.7 (2002–05)
			Iran, Islamic Rep.[ag]	0.65 per 1,000 births	80.0 (2005–07)
				10 per 15,321 live births (2005–07)	
			Nigeria[ah]	14 over 10 years	71.4
			Nigeria[ai]	12 over 11 years	33.0
			Colombia[aj]	32 over 9 years	18.8
			South Africa[ak]	106 over 6 years;	43.0 (total)
				6 neonatal surgical admissions (2003);	68.0 patients with staged, silo-assisted closure
				15 neonatal surgical admissions (2007)	
			Thailand[al]	49 over 3 years	14.0
			Nigeria[am]	7 of 2,381 patients treated at the pediatric surgery unit over eight years	57.1
Omphalocele	2.18–5 per 10,000 live births[a, ad, ae]	5[a]	Iran, Islamic Rep.[b]	12 (1993–96)	75.0 (1993–96)
				68 (2002–05)	58.8 (2002–05)
			Iran, Islamic Rep.[ag]	2.1 per 1,000 births	20.0 (2005–07)
				42 of 15,321 live newborn births (2005–07)	
			Nigeria[ai]	42 over 11 years	43.0
			Colombia[aj]	23 over 9 years	43.5
			Nigeria[am]	49 over 8 years	32.4 (patients with omphalocele major)

table continues next page

Table 8.1 Prevalence and Mortality of Selected Congenital Anomalies in Selected Countries (continued)

	High-income countries		Selected countries		
Congenital anomalies	**Reference prevalence**	**Reference mortality (percent)**	**Country of reporting institution**	**Prevalence at reporting institution**	**Mortality (percent)**
Congenital diaphragmatic hernia	1 per 2,000 to 1 per 5,000 live births[a]	≤ 10[an]	Turkey[ao]	10 over 4 years	50.0
			Tunisia[ap]	28 over 13 years	39.0
			Nigeria[aq]	64 over 24 years	35.5 (overall)
					60.0 in patients born in hospital
					28.5 in patients born outside hospital who survive until presentation
			Nigeria[ar]	7 over 6 years	43.0
			Malaysia[as]	21 over 6 years	47.6

Sources: Anorectal malformations, Ethiopia: Anorectal malformations, India: Anorectal malformations, Iran, Islamic Rep.: Anorectal malformations, Nigeria:
a. Coran and others 2012. b. Peyvasteh and others 2011. c. Adejuyigbe and others 2004. d. Chalapathi and others 2004. e. Adeyemi 1989. f. Archibong and Idika 2004. g. Tefera, Teka, and Derbew 2007. h. Gangopadhyay and others 2005. i. Ekenze, Ibeziako, and Ezomike 2007. j. Rescorla and others 1992. k. Swenson 2002. l. Banu and others 2009. m. Bandre and others 2010. n. Calisti and others 2011. o. Chirdan and Uba 2006. p. van der Linde and others 2011.q. Bernier and others 2010. r. Larrazabal and others 2007. s. Bakshi and others 2007. t. Shah and others 2008. u. Wickramasinghe, Lambabadusuriya, and Narenthiran 2001. v. Upadhyaya and others 2007. w. Singh and others 2012. x. Rayes 2010, y. Islam and Aziz 2011. z. Yang and others 2006. aa. Narasimman, Nallusamy, and Hassan 2013. ab. Anwar-ul-Haq and others 2009. ac. Al-Salem and others 2006. ad. Sadler 2010. ae. Canfield and others 2006. af. Manson and others 2012. ag. Askarpour and others 2012. ah. Ameh and Chirdan 2000b. ai. Uba and Chirdan 2003. aj. Toro, Rave, and Gomez 2010. ak. Sekabira and Hadley 2009. al. Saranrittichai 2008. am. Abdur-Rahman, Abdulrasheed, and Adeniran 2011. an. Chiu and Hedrick 2008. ao. Ozdogan and others 2010. ap. Khemakhem and others 2012. aq. Adegboye and others 2002. ar. Abubakar and others 2011. as. Rohana, Boo, and Thambidorai 2008.

Sekabira and Hadley 2009; Uba and Chirdan 2003). The misdiagnosis of anomalies as better-known infectious diseases, and added delays for invisible anomalies, may further hinder the provision of timely, appropriate services. These multifactorial delays are a crucial hurdle in treating both immediately and non-immediately life-threatening anomalies. While non-immediately life-threatening anomalies often require emergency interventions, the period before these conditions become emergencies can be better used to dramatically improve outcomes.

Scarcity of Skilled Surgeons

The scarcity of trained surgeons in LICs and LMICs also significantly contributes to the burden: one pediatric general surgeon may serve millions of children (Chirdan and others 2010), and physicians performing pediatric surgery may have little or no pediatric surgery training (Ekenze, Ibeziako, and Ezomike 2007; Mhando, Young, and Lakhoo 2008). Whereas North America has an estimated one pediatric cardiac surgeon per 3 million people, Sub-Saharan Africa has one per 38 million people (Bernier and others 2010); 75 percent of the world's population is estimated to have no access to cardiac surgery (Hoffman 2013). Similarly, 33 percent of the world's population is covered by 5 percent of its neurosurgeons (Warf 2013). Expanding the pool of specially trained surgeons and surgery teams must be a fundamental goal of ongoing and future programs to address the pediatric surgical burden of disease.

ESTIMATING THE IMPACT OF PEDIATRIC SURGERY ON THE GLOBAL BURDEN OF DISEASE

In the World Health Organization's (WHO's) most recent Global Health Estimates, congenital anomalies constitute 2.1 percent of the total disease burden and rank eleventh in the causes of disease burden (WHO 2013b). Although impressive, these figures are likely to be underestimates because of the limited number of anomalies included in the analysis and the difficulties in evaluating incidence, morbidity, and mortality. Only six congenital surgical conditions had disability weights in the 2004 estimates, and congenital anomalies were not among the new disability weights estimated in 2012 (Saloman and others 2012; WHO 2008).

Some researchers have tried to fill the gap with evidence-based estimates of selected disability weights (Poenaru and others 2013). Of the conditions measured in the Global Health Estimates, cardiac anomalies represent the greatest overall burden (table 8.2), and, along with neural tube anomalies and cleft lip and palate, cause

32 million DALYs. Some 57 percent, or 18 million, of these DALYs are estimated to be surgically preventable if outcomes in HICs could be achieved in LICs and LMICs (Higashi, Barendregt, and Vos 2013). These anomalies typify the reservoir of unmet need that congenital anomalies in LICs and LMICs create (table 8.3).

Only a small body of literature evaluates the potential of surgery to reduce this burden in terms of DALYs averted or cost-effectiveness. Yet, these foundational studies have provided compelling evidence that pediatric surgery is a cost-effective intervention with the potential to avert more than 67 percent of the DALYs associated with congenital anomalies (Corlew 2010; Higashi, Barendregt, and Vos 2013; Ozgediz and Poenaru 2012; Poenaru 2013; Wu and Poenaru 2013; Wu, Poenaru, and Poley 2013). Favorable outcomes have been reported in HICs for such conditions as anorectal malformations (ARMs) and congenital diaphragmatic hernia (Poley and others 2008). In LICs and LMICs, the human capital approach to cleft lip and palate repair (see chapter 21) has provided very favorable cost-effectiveness analysis estimates. An extension of this methodology to treatment for congenital swelling of the brain in Uganda has also yielded favorable results, at a cost of US$59 to US$126 per DALY averted (Warf and others 2011). Surgical repair of congenital inguinal hernias in Uganda has been estimated to have an incremental cost-effectiveness of US$12 per DALY averted (Eason and others 2012). Another report from Cambodia estimates a cost-effectiveness of US$99 per DALY averted over three months for reconstructive surgery for an array of anomalies (Rattray and others 2013).

Table 8.2 Burden of Disease due to Congenital Anomalies

Anomaly	DALYs (thousand)	YLDs
Cardiac	20,760	565
Neural tube	10,075	759
Down syndrome	2,939	1,225
Cleft lip	709	254
Other chromosomal	2,941	694
Other congenital	20,272	1,835
Total	57,696	5,332

Source: WHO 2013b.
Note: DALYs = disability-adjusted life years; YLDs = years living with disability.

Table 8.3 Prevalent Congenital Anomalies and Avertable Disease Burden

Congenital anomaly	Brief description	Treatment opportunities
Cardiac anomalies	Most prevalent anomalies *Incidence*: Approximately 8 per 1,000 births *Most common*: Ventricular septal defect (hole between the lower chambers of the heart)	Backlog of 1 million to 2 million children need congenital cardiac surgery in India. Requires relatively resource-intensive treatment compared with other anomalies; estimated US$2,500 per operation in some programs (Hoffman 2013). Cost containment and capacity-building strategies have been described (Rao 2007).
Neural tube defects	*Incidence*: Nearly 1 per 1,000 births *Most common*: Spina bifida	Preventable through folate supplementation, a major public health strategy in many LICs and LMICs. ETV is an innovative, sustainable strategy to treat associated hydrocephalus, or swelling of the brain, with favorable results compared with traditional treatment. Longer-term follow-up confirms the feasibility and effectiveness of community-based strategies for ETV (Warf and others 2011; Warf 2011).
Cleft lip and palate	*Incidence*: 1 per 700 live births; slightly higher in some regions, like Sub-Saharan Africa (Poenaru 2013) Approximately 25 percent of cases associated with other anomalies	Global backlog of unrepaired cleft cases is between 400,000 and 2 million cases. Guidelines suggest that cleft lip should be repaired in the first six months of life; cleft palate, in the first year to 18 months. Average age at time of repair is nearly age 10 years in Sub-Saharan Africa (Poenaru 2013). Cleft lip may require a single corrective operation. Approximately 20 percent of palate repair cases may require subsequent surgery; postoperative speech therapy is essential (Semer 2001).

Sources: Hoffman 2013; Poenaru 2013; Rao 2007; Semer 2001; Warf and others 2011; Warf 2011.
Note: ETV = endoscopic third ventriculostomy; LICs = low-income countries; LMICs = lower-middle-income countries.

The benefits of improved pediatric surgical services in averted morbidity and cost extend across the lifespan. Treating congenital disease at its inception may result in a significantly greater reduction in the burden of disease. The following sections present examples of anomalies that are prime targets for intervention. Because of the lack of cost-effectiveness data, these anomalies are presented as case studies that highlight the preventable burden of disease and the potential of low-cost measures adapted to low-resource settings (i.e., low-income countries [LICs] and LMICs) to substantially improve outcomes. Further research is urgently needed to develop and evaluate cost-effective treatment programs to take advantage of the substantial DALY upside of treating congenital anomalies. Congenital heart conditions are discussed in more detail in volume 5, *Cardiovascular, Respiratory, Renal, and Endocrine Disorders*.

Case Study 1: Congenital Colorectal Disease—Anorectal Malformations and Hirschprung's Disease

Description. ARMs and Hirschsprung's disease (HD) are two of the most prevalent congenital anomalies.

- ARMs are physical anomalies that prevent the passage of fecal matter through a distinct anus. Examples include absence of an anus or fusion of the anus to other openings in the body (for example, the urethra).
- HD is a functional obstruction of the bowel caused by the absence of the nerve cells needed to stimulate normal contractile movement of the bowel. If food matter cannot move through the bowel, material collects in the preceding bowel and dilates it, causing megacolon. The intestinal tract may perforate, causing widespread infection and death.

HD and some ARMs are not immediately life threatening when partial passage of fecal material is possible. For example, female children in LICs and LMICs with the most common female ARM—vestibular fistula, whereby the rectum opens into the vagina—often remain undiagnosed until much later in life. Untreated, however, non-immediately life-threatening conditions can lead to substantial morbidity and eventual mortality due to intestinal rupture.

The etiology of ARMs is unclear, but both genetic and environmental factors have been implicated (Davies, Creighton, and Wilcox 2004). The incidence of ARMs is cited as one per 3,000 to 5,000 live births (Chalapathi and others 2004; Chowdhary and others 2004; Eltayeb 2010), but this incidence varies with ethnicity and geography (Moore and others 2008; van Heurn and others 2002). ARMs are reportedly more common in Sub-Saharan Africa and constitute a significant clinical load (Calisti and others 2011; Moore and others 2008). HD has been associated with a number of congenital syndromes and anomalies, and may have various genetic causes (Amiel and others 2008). The incidence of HD is comparable to that of ARMs (Coran and others 2012) and is one of the leading causes of pediatric intestinal obstruction in LICs and LMICs (Adeyemi 1989; Ameh and Chirdan 2000a; Saha and others 2012). Hidden mortality and traditional health practices (for example, enemas) mask prevalence (Bandre and others 2010), suggesting that the burden of disease may be significantly underestimated.

Diagnosis and Treatment in LICs and LMICs. ARMs are usually diagnosed on physical examination. HD is not visually identifiable, so must be diagnosed based on the symptoms—feeding intolerance, vomiting, abdominal distension, delayed passage of the meconium (the first stool passed by a newborn) and severe neonatal intestinal infection, or enterocolitis (Amiel and others 2008). Meconium passage may serve as a valuable screening tool for HD in HICs, LICs, and LMICs; 95 percent of children with HD do not pass the meconium within the first 24 hours of life, while only 1 percent of children without HD experience a comparable delay.

HD is definitively diagnosed via rectal biopsy[1] (Amiel and others 2008), but diagnostic ability may be limited in LICs and LMICs because of the cost of biopsy analysis and the scarcity of pathology services (Bandre and others 2010). Many practitioners are forced to provide definitive treatment without confirmation of the HD diagnosis. Analytic protocols for biopsy specimens can, however, be adapted to the resources of medical laboratories in LICs and LMICs (Babu and others 2003; Poenaru and others 2010), and inexpensive radiography can replace costly endoscopic technologies in preoperative planning (Pratap and others 2007).

In LICs and LMICs, late presentation for nonemergency congenital colorectal disease is the norm. More than 60 percent of patients with HD present late, as children, adolescents, and even adults (Ameh and Chirdan 2000a; Poenaru and others 2010; Sharma and Gupta 2012; Vincent and Jackman 2009). Delayed diagnosis results from a web of interacting societal, cultural, and socioeconomic factors that delay the diagnosis of other anomalies detailed in this chapter. Presentation and diagnosis are delayed by geographical and financial barriers to care, social taboo, cultural norms (for example, routine traditional enemas), lack of awareness among medical personnel in first- and second-level facilities, inaccurate medical advice, and failed or unwarranted procedures at other medical facilities (Al-Jazaeri and others 2012; Bandre and others 2010; Ekenze, Ngaikedi, and

Obasi 2011; Sharma and Gupta 2012; Sinha and others 2008; Vincent and Jackman 2009). Delayed diagnosis of ARMs leads to severe morbidities and elevated mortality, which are further heightened for infants in rural areas where delayed diagnosis is coupled with subsequent transport to distant medical facilities (Adejuyigbe and others 2004; Chalapathi and others 2004; Eltayeb 2010; Turowski, Dingemann, and Giilick 2010).

Delayed presentation of HD is characterized by chronic constipation, abdominal distension, bowel loops visible through the abdominal wall, failure to thrive, anemia, malnutrition, and fecal impaction (Al-Jazaeri and others 2012; Coran and Teitelbaum 2000; Ekenze, Ngaikedi, and Obasi 2011; Frykman and Short 2012; Prato and others 2011; Sharma and Gupta 2012). Some patients present with bloody diarrhea due to HD-associated enterocolitis, the leading cause of HD-related morbidity.

In HICs, mortality for ARMs is negligible. In LICs and LMICs, mortality reaches 20 percent to 30 percent (Adejuyigbe and others 2004; Adeyemi 1989; Archibong and Idika 2004; Chalapathi and others 2004; Ekenze, Ibeziako, and Ezomike 2007; Peyvasteh and others 2011; Tefera, Teka, and Derbew 2007). The mortality rate for HD is less than 5 percent to 10 percent in HICs (Rescorla and others 1992; Swenson 2002) but jumps to 20 percent to 43 percent in LICs and LMICs (Adeyemi 1989; Ameh and Chirdan 2000a; Bandre and others 2010; Ekenze, Ibeziako, and Ezomike 2007). These disparities are linked to delayed diagnosis and treatment, sepsis,[2] and the absence of critical care when patients present with advanced complications (Adeniran and Abdur-Rahman 2005; Chalapathi and others 2004; Chowdhary and others 2004).

Both ARMs and HD can be treated with either primary (one-step) or staged (multistep) surgical repair (Coran and others 2012). These operations generally do not require intensive postoperative care, but they do require general anesthesia. Delays in diagnosis often preclude primary repair because these repairs cannot be performed when the bowel is grossly distended. In such cases, a colostomy[3] is a life-saving first step in staged repair. Colostomy complications, however, are common in HICs, LICs, and LMICs (Chalya and others 2011; Chandramouli and others 2004; Patwardhan and others 2001). The risks are compounded in LICs and LMICs by the prohibitive cost of colostomy bags, cultural prejudice, and limited parental understanding (Adeniran and Abdur-Rahman 2005; Chandramouli and others 2004; Olivieri and others 2012). Although simple, inexpensive treatments may ameliorate some of these challenges (Chalya and others 2011), a significant burden remains. Many children live for years with colostomies without receiving definitive repair. The burden on families of caring for children with long-term colostomies has not yet been well captured.

Primary repair reduces the number of surgeries, minimizing treatment costs and averting colostomy-related morbidity and mortality. Good outcomes in LICs and LMICs have been reported (Adeniran and Abdur-Rahman 2005; Elhalaby 2006; Ibrahim 2007; Osifo and Okolo 2009; Pratap and others 2007), with mortality rates rivaling those in HICs (Ibrahim 2007; Osifo and Okolo 2009). However, the prevalence of delayed diagnosis and treatment render routine primary repair risky. Reducing delays is key to relieving the preventable burden of congenital colorectal disease. Suggestions include the following (Adeniran and Abdur-Rahman 2005; Al-Jazaeri and others 2012; Ameh and others 2006; Ekenze, Ngaikedi, and Obasi 2011; Olivieri and others 2012; Peyvestah 2011; Poenaru and others 2010):

- Increase the number of third-level facilities (major hospitals offering a full spectrum of services)
- Increase participation of existing third-level facilities in the training of community health centers in diagnosis and preoperative management
- Improve training at the level of the TBA, primary care provider, and community health worker
- Modify medical education curricula to encompass ARMs and HD
- Target surgeons at first-level hospitals to perform colostomies with available resources
- Institute low-cost modifications to standard repair procedures

Substantial loss to follow-up after colostomy formation in LICs and LMICs remains a challenge, and innovative approaches to ensuring patient return or local follow-up in home regions must be developed to overcome barriers to continuity of care.

Case Study 2: Abdominal Wall Defects—Omphalocele and Gastroschisis

Description. Omphalocele and gastroschisis are abdominal wall defects in which the internal organs, or viscera, protrude through the abdominal wall. In omphalocele, the gut and other abdominal organs, such as the liver, spleen, and gonads, protrude through the abdominal wall into a membranous sac. In gastroschisis, no sac is present and usually only the gut protrudes from the abdomen (Coran and others 2012). Patients with omphalocele can be fed if the sac is intact; those with gastroschisis cannot be fed and quickly perish without treatment.

The incidence of omphalocele is approximately 2.18 to 5 per 10,000 live births; that of gastroschisis is 3.3 to

5 per 10,000 live births (Canfield and others 2006; Coran and others 2012; Sadler 2010; Stoll and others 2001). The incidence of gastroschisis is on the rise and varies geographically (Andrew, Holland, and Badawi 2010; Arnold 2004; Benjamin and others 2010; Castilla, Mastroiacovo, and Orioli 2008; Laughon and others 2003; Loane, Dolk, and Bradbury 2007; Vu and others 2008).

Risk factors for omphalocele include chromosomal anomalies, very young and very advanced maternal age, lack of multivitamin and folic acid supplementation during pregnancy, and maternal history of fevered illness (Botto and others 2002; Botto, Mulinare, and Erickson 2002; Frolov, Alali, and Klein 2010; Mills and others 2012). Gastroschisis has not yet been associated with any particular genes. Risk factors for gastroschisis include young maternal age, low socioeconomic status, poor nutrition, and lack of vitamin supplementation during pregnancy (Coran and others 2012). Mothers in LICs and LMICs are more likely to have children at both younger and more advanced ages, to have limited family planning knowledge and resources, and to suffer nutritional deficiencies. Accordingly, their children are likely to be at greater risk for omphalocele and gastroschisis.

Diagnosis and Treatment in LICs and LMICs. Ultrasonography, a low-cost technology once in place, can detect omphalocele and gastroschisis before birth with high success (Richmond and Atkins 2005). This technology can inform decisions about pregnancy termination and mode of delivery, facilitating improvements in outcomes.

Several effective surgical strategies for omphalocele have been described. Gastroschisis necessitates greater attention to heat loss and moisture preservation because a larger surface area of viscera is exposed. Historically, primary closure has been the treatment of choice in that it limits damage incurred by exposure. Recent studies have shown that using a silo (a moisture-retaining bag that holds the viscera before they are returned to the abdomen) and postponing closure for hours to days can be equally effective (Coran and others 2012).

Treatment is highly effective in HICs; multiple series report survival rates of 70 percent to 95 percent for omphalocele and 90 percent or greater for gastroschisis (Coran and others 2012). In comparison, the mortality associated with omphalocele and gastroschisis in LICs and LMICs is shockingly elevated, with survival falling to less than 20 percent in some studies (Askarpour and others 2012; Richmond and Atkins 2005). Delayed presentation plays a key role because hypothermia and gangrenous bowel may develop in the interim (Ameh and Chirdan 2000b; Sekabira and Hadley 2009; Uba and Chirdan 2003). An additional challenge with gastroschisis is that bowel function can be impaired initially, necessitating the use of intravenous nutrition, often not available in poorer countries. It has been suggested that in-house birth at centers equipped to medically manage patients with abdominal wall defects is essential to improving outcomes, as is improving training for transport personnel, obstetricians, and primary care physicians (Sekabira and Hadley 2009). Training TBAs to recognize omphalocele and gastroschisis, and to place children from the shoulder down in clean polyethylene bags to protect the bowels during transport, is another viable option for reducing complications associated with delayed presentation (Ameh and Chirdan 2000b).

Surgical procedures commonly used in HICs may increase mortality in LICs and LMICs, largely as a result of infection and sepsis (Ameh and Chirdan 2000b; Uba and Chirdan 2003); nonsurgical or altered surgical procedures may be preferable. Uba and Chirdan (2003) report the successful treatment of unruptured omphalocele using daily application of scab-inducing topical ointments, leading to eventual skin growth over the defect. Bedside placement of a silo bag followed by gradual reduction of the viscera into the abdomen and sutureless repair is a potentially cost-effective treatment strategy worthy of additional evaluation in LICs and LMICs. Several studies have investigated low-cost alternatives to silos, including transfusion bags and female condoms (Bustorff-Silva and others 2008; Miranda and others 1999). The female condom is particularly intriguing because its use requires no sutures, surgery, or anesthesia; allows for easy preoperative observation; and allows gravity to gradually move the viscera into the abdomen (Bustorff-Silva and others 2008).

Once infants with omphalocele or gastroschisis survive the neonatal period, there is little to no associated disability or mortality. These anomalies are highly treatable and thus potentially highly DALY averting.

Case Study 3: Clubfoot

Description. Talipes equinovarus, or clubfoot, is a complex congenital anomaly in which the entire foot is rotated inward (van Bosse 2011). Clubfoot may be idiopathic—without an identifiable cause—or associated with other congenital anomalies. The etiology is unknown but likely involves genetic and environmental factors (Dobbs and Gurnett 2009). According to estimates from HICs, the incidence is approximately 1 per 1,000 live births (Coran and others 2012). However, incidence varies widely with ethnicity, and reports from LICs and LMICs have shown that incidence may be about 1 per 500 live births (Dobbs and Gurnett 2009; Mkandawire and Kaunda 2002); 80 percent of children with untreated clubfoot are born in LICs and LMICs (WHO 2013a). Untreated clubfoot

leads to lifelong disability, social stigmatization, and decreased economic self-sufficiency in adulthood (Alavi and others 2012; Lourenço and Morcuende 2007).

Diagnosis and Treatment in LICs and LMICs. The most effective treatment for idiopathic clubfoot, the Ponseti method, is largely nonsurgical. We have included this anomaly for analysis because it exemplifies the importance of holistically approaching surgical disease to use both low-cost surgical and nonsurgical interventions to reduce the burden of disease.

The Ponseti method, in which a series of casts are applied between incremental manipulations of the foot by trained practitioners, is successful in up to 98 percent of patients (WHO 2013a). The Achilles tendon is often cut through the skin to correct lingering deformity, and corrective braces are worn for several years to prevent relapse (Dobbs and others 2000). The Ponseti method is also successful in treating neglected idiopathic clubfoot, in which the anomaly is not treated before walking age (Lourenço and Morcuende 2007). If surgery alone is used to correct this aggravated form of the deformity, functionality is low and may degrade into crippling pain and weakness in adolescence and adulthood (Gupta and others 2008).

The Ponseti procedure is well suited to use in LICs and LMICs; it is a low-cost intervention that can be performed by health and allied health professionals[4] (Janicki and others 2009; Lavy and others 2007; Mayo and others, n.d.), an advantageous attribute in contexts with few orthopedic surgeons. It requires no specialized surgical facilities and produces a functional foot (Gupta and others 2008).

Treatment programs using the Ponseti method have been set up in many LICs and LMICs, with success rates that regularly approximate those seen in HICs (Goksan and others 2006; Gupta and others 2008; Jawadi 2010; Makhdoom and others 2011; Mendez-Tompson and others 2012; Panjavi and others 2012; Sarrafan and others 2012). The Ponseti method is also successful in treating neglected clubfoot in LICs and LMICs, yielding superior outcomes and incurring significantly lower costs than purely surgical interventions (Adegbehingbe and others 2010; Hegazy, Nasef, and Abdel-Ghani 2009; Spiegel and others 2009).

The success of national clubfoot programs (box 8.1) provides strong evidence for the utility of task-shifting, that is, training paramedical practitioners to perform select health care tasks. Nonphysician practitioners trained in orthopedics and the Ponseti method can achieve results comparable to those of surgeons in HICs (Tindall and others 2005). Collaborations among diverse partners, including nongovernmental organizations, ministries of health, and academic institutions,

Box 8.1

The Uganda Sustainable Clubfoot Care Project

Clubfoot, the most common cause of locomotor disability in low-income countries leads to profound impairments in activities of daily life, social exclusion, and abandonment. After several years of pilot intervention, the Uganda Sustainable Clubfoot Care Project was implemented in 2000. Essential elements include the following:

- Development of a national strategic plan addressing all levels of the health system
- Endorsement by the Ministry of Health and incorporation into the National Health Policy
- Community awareness campaigns
- Sensitization of maternity units via education
- Training: Ponseti method trainers at the national hospital to train nonphysician orthopedic officers in serial casting in rural hospitals and medical officers in tenotomy
- Development and distribution of locally made orthoses, or mechanical devices (for example, braces) to provide support and correct alignment of the clubfoot.

In 2006 and 2007, 872 children were treated, nearly 800 providers were trained, and services were made available in 21 hospitals. Using this experience in Uganda and a similar program in Malawi as a basis, interventions have been implemented in 10 other countries; early two-year follow-up data suggest similarly successful results.

Source: Pirani and others 2009; Tindall and others 2005.

are instrumental for increasing the number of trained paramedical practitioners, and thereby the availability of treatment (Owen and others 2012). To achieve these goals, it is critical to implement a coordinated, standardized program to decentralize care, and to integrate education, awareness, and service delivery into the public health sector.

IMPLEMENTATION AND SURVEILLANCE PRIORITIES

Addressing the Burden of Disease of Congenital Anomalies

Strategies to increase the accessibility of surgical care for children with congenital anomalies include development of treatment centers for specific conditions (niche hospitals), short-term surgical missions, partnerships to train local providers, and the transporting of patients to other countries for care. Interventions at the policy level may also play a role, as may novel tools such as telemedicine. The strengths and weaknesses of these strategies are summarized in table 8.4. It is critical that local expertise and buy-in be integrated into all efforts to increase the accessibility of pediatric surgery so as to create sustainable systems that increase long-term capacity and take advantage of the substantial potential intellectual, creative, and personnel resources in LICs and LMICs.

Strategy 1. Cultivating Treatment Centers in LICs and LMICs

Recent humanitarian efforts have favored establishing sustainable surgical centers in LICs and LMICs, staffed by either local or foreign personnel. This approach maximizes the number of children treated and enables local trainees to learn to practice in their future professional environments, preventing later struggles to adapt protocols learned abroad to local resources (Larrazabal and others 2007). The entire surgical team can be concurrently trained, prepping both the surgeon and the center to operate independently (Larrazabal and others 2007; Loisance 2012).

Table 8.4 Increasing the Accessibility of Pediatric Surgery

Strategy	Advantages	Disadvantages
Treatment centers in LICs and LMICs	Creates infrastructure and expertise to broadly affect the burden of disease caused by congenital anomalies Streamlines care if center focuses on one pathology or organ system Provides treatment in home countries	Streamlined centers often not integrated with local training programs and public health care systems
Surgical missions	Minimizes costs and culture shock for patients Streamlines care if missions focus on one condition Offers opportunity to train local personnel Are well suited for nonemergency conditions with backlogs	Episodic Limited time window restricts number of patients treated Pressure to deliver care in limited period may restrict training efforts Follow-up limited Integration with existing services may be limited Not designed to treat emergency life-threatening conditions
Academic partnerships	Provides coordinated interface for students and professionals from HICs, LICs, and LMICs to learn from and collaborate with each other Opportunity for training and resource sharing Facilitates scholarly approach to intervention and evaluation	Not as focused on delivery of care Research may not be truly collaborative Potential for medical tourism by HIC practitioners, especially if training not provided to LIC and LMIC partners Financial sustainability may be limited by availability of institutional funding
Human resources and policy changes Telemedicine	Task-shifting increases health care access	Necessitates rigorous regulation and standardization

table continues next page

Table 8.4 Increasing the Accessibility of Pediatric Surgery (continued)

Strategy	Advantages	Disadvantages
	Potentially reduces transport risks and costs	No opportunity for hands-on training
	Builds local capacity through consultation	
	Improves communication between different points of health care access	
Health care delivery outside of LICs and LMICs	Increases level of care compared with local systems	Costly
		Benefits only a few patients
		Does not contribute to developing capacity in LICs and LMICs

Note: HICs = high-income countries; LICs = low-income countries; LMICs = lower-middle-income countries.

Successful centers have developed from international partnerships, foreign humanitarian initiatives, and home-grown efforts in LICs and LMICs (Bode-Thomas 2012), with evidence that continuous support through development programs can build pediatric surgery capacity. Networks of faith-based hospitals also provide specialized surgical services, such as the CURE network, which focuses primarily on neurosurgery and orthopedics; CBM International, which supports an array of surgical services; and the Pan-African Academy of Christian Surgeons network, which has developed training programs in association with BethanyKids, a faith-based pediatric surgical organization. Surgical mission–oriented organizations may also develop treatment centers, as has been done by Operation Smile, a mission program specializing in cleft lip and palate repairs (Patel and others 2013).

Treatment at these centers can be tailored to available resources. Less-expensive, but effective, diagnostic and therapeutic modalities can be used. Simple, palliative surgeries may predominate in the neonatal period if early definitive repair is too risky. Cheaper surgical materials are produced by countries like Brazil and China; medical equipment companies may be persuaded to make donations; and some disposable surgical materials can be sterilized and reused (Rao 2007; Senga and others 2013).

The few cost-effectiveness analyses that have been published support the feasibility of developing local treatment centers. For example, at the CURE Children's Hospital in Mbale, Uganda, children with hydrocephalus are treated at an estimated cost of US$59 to US$126 per DALY averted, with US$3 million to US$5 million saved to Uganda, and US$5 million to US$188 million saved based on statistical calculations of the value of a life (Warf and others 2011).

Strategy 2. Surgical Missions

The short-term surgical mission, also commonly referred to as humanitarian assistance or a volunteer trip, is an established health care delivery model that is becoming increasingly popular. Individual providers or organized teams travel from HICs to deliver surgical care in LICs and LMICs (Martiniuk and others 2012). Smile Train and Operation Smile, the two largest global cleft lip and palate charities, exemplify two different models of successful, long-running surgical missions. Operation Smile primarily funds teams from HICs to provide short-term health care delivery and training in LICs and LMICs; Smile Train[5] funds local providers to offer outreach and training (Poenaru 2013). In some regions, these teams provide the only surgical services for children (Walker and others 2010). In recent years, Operation Smile has also developed treatment centers as a more sustainable approach (Magee and others 2012).

Sustainability, follow-up for postsurgical complications, and integration with existing health systems are significant challenges for the inherently episodic surgical mission model. Perhaps most important, surgical missions are better suited for "prevalent" rather than "incident" conditions. Prevalent conditions incur increasing morbidity, whereas incident conditions are surgical emergencies. A greater proportion of the burden of disease may be averted by targeting emergency conditions, but emergency treatment cannot be improved without improving capacity at the systems level (Bickler and others 2010).

Strategy 3. Partnerships between Academic Organizations and Development Programs

Academic organizations have increasingly focused on augmenting surgical capacity in LICs and LMICs through partnerships (Calland and others 2013; Qureshi

and others 2011). Some organizations subsidize academic surgeons, who then deliver care, train providers, and conduct research in LICs and LMICs. Partnerships also provide mechanisms by which practitioners from LICs can obtain foreign training in middle-income countries (MICs) (for example, a Sub-Saharan African practitioner training in India) or HICs. Training outside of the home environment has the disadvantage that the trainee may not return for practice. Funding the training of local practitioners by underwriting the presence of visiting surgeons in partnered development programs may be an effective alternative. For example, during the course of a four-year project funded by the Italian Cooperation in Eritrea, local Eritrean surgeons achieved independence and favorable outcomes in the treatment of a wide range of congenital anomalies (Calisti and others 2011). Overall, the scope of activities undertaken by academic organizations in these partnerships varies widely, and outcomes of many permutations of partnership are not yet well characterized.

Strategy 4. Human Resources and Policy Changes

Policy changes, such as the provision of free health care to children, may increase access to pediatric surgery (Groen and others 2013). The expansion of training programs is key to ensuring that children with anomalies are diagnosed in a timely manner and can access appropriate surgical services. Regional professional societies offer a limited number of training spots and scholarships, and training programs need support (Elhalaby and others 2012). More training positions are required to meet the need for trained pediatric surgeons and the interest level of general surgery trainees. To date, only limited analyses of the cost-effectiveness of such programs have been conducted.

Specially trained paramedical practitioners may be a viable solution to the scarcity of providers, as demonstrated by the success of clubfoot programs; however, the scope of treatment may be limited to select conditions (Mayo and others, n.d.; Pirani and others 2009; Tindall and others 2005). The scope of practice for a pediatric general surgeon includes congenital anomalies, acquired surgical diseases, and a high proportion of emergencies. This breadth has made it challenging to design and implement intervention packages in hospitals and health care systems. Other surgical providers may do their best to meet the need, but the shortfall is tremendous. Pediatric anesthesia providers are similarly few, as are trained pediatric neurosurgeons, cardiac surgeons, orthopedic surgeons, and other surgical subspecialists. Although task-shifting has been promoted to meet the need for essential surgery in adults, its applicability to pediatric surgical conditions has not been specifically analyzed.

Strategy 5. Telemedicine

Telemedicine[6] may increase the accessibility of limited surgical specialists to large populations requiring care. Chaotic roads and a lack of medical transport complicate the great distances that patients in LICs and LMICs must travel to reach care. Children seeking treatment may present in critical and sometimes unsalvageable condition following such physiologically stressful transit (Rao 2007). With telemedicine, physicians at local or regional medical centers can interact with experts at centralized third-level facilities to guide patient treatment, potentially circumventing the need for patients to undergo life-threatening journeys. The cost burden of travel and treatment for families is decreased, and unspecialized providers can receive training from specialized peers (Sekar and Vilvanathan 2007). Telemedicine can also link medical centers for educational or research purposes (Hadley and Mars 2008). This developing technology has the potential to beneficially decentralize specialized care and education in LICs and LMICs.

Strategy 6. Health Services Delivery Outside of Local Systems

Families may seek care in foreign treatment centers, funded out of pocket, through community fundraising, or by humanitarian organizations. Increased access to advanced treatment methods saves lives that would otherwise be lost. However, transporting children to foreign centers limits the number of children treated. Because the cost of surgery abroad often surpasses a family's annual income in LICs and LMICs, even the highest-earning families must rely on limited governmental and nongovernmental agency funds (Sadoh, Nwaneri, and Owobu 2011). If patients require multiple surgeries, financial sponsors must decide whether to perform the surgery at all, abstain from follow-up surgeries, or fly the patient back for additional surgeries. If patients die abroad, the families must wrestle with substantial financial and emotional challenges in coordinating burial arrangements (Bode-Thomas 2012). Furthermore, this approach does not build local surgical capacity. The social and economic costs for patients and families who seek care abroad are likely significant but have not been estimated; an estimate may provide incentives for governments and funders to invest in local care.

These strategies are not mutually exclusive; they may coexist or evolve into different models.

Although the development of health care capacity within LICs and LMICs is crucial to the long-term reduction of the burden of disease associated with congenital anomalies, other intervention strategies provide additional and immediate opportunities to improve pediatric surgical care and outcomes.

ACTION PLAN

Clinical Intervention at Every Level

Given that platforms for delivery vary by setting, we propose only general guidelines for congenital anomalies (table 8.5). Robust data from LICs and LMICs are sparse; therefore, the following recommendations—based on the available literature—should be understood as the best available at this time. Future research will undoubtedly lead to more precise, substantiated recommendations.

- The village health center should have health providers trained to identify anomalies. In Tanzania, educating TBAs and using a birth card to register and record birth data are being evaluated as tools for improving identification of treatable anomalies (Norgrove Penny, personal communication) and may be useful in other practice settings. Public education

Table 8.5 Pediatric Surgery Capacity in an Ideal System

Capacity	Village health center	First-level hospital	Second-level hospital	Third-level hospital
Airway management, fluid replacement, bleeding control, antibiotic therapy	X[a]	X	X	X
Blood transfusion		X[a]	X	X
Tracheal tube		X[a]	X	X
Local anesthesia	X	X	X	X
Spinal and general anesthesia		X	X	X
Pediatric hernia		X	X	X
Pediatric hernia (infant)			X[a]	X
Umbilical hernia		X	X	X
Pyloric stenosis			X[a]	X
Colostomy		X[a]	X	X
Neonatal bowel obstruction (atresia, stenosis, malrotation)				X
Tracheoesophageal fistula repair				X[a]
Clubfoot		X	X	X
Cleft lip		X	X	X
Anorectal malformations or Hirschsprung's disease (first stage; often colostomy)		X	X	X
Anorectal malformations or Hirschsprung's disease (definitive treatment)			X	X
Abdominal wall defects			X[a]	X
Hydrocephalus			X[a]	X
Congenital cardiac anomalies			X[a]	X
Spina bifida				X
Bladder extrophy				X
Undescended testicles			X	X
Hypospadias				X

Note: "Capacity" is defined as trained staff available 24 hours, seven days a week, with adequate equipment and supplies.
a. Capacity should already be there but usually is not.

is important for raising awareness that congenital anomalies are treatable and not a death sentence; this vital information prompts families to seek care.
- The first-level hospital should have the ability to stabilize pediatric patients with surgical emergencies and to definitively treat conditions for which the capacity exists. In-depth guidelines have been suggested for first-level hospitals but have not yet been tested or validated (Bickler and Ameh 2011).
- The second-level hospital should provide life-saving surgical treatments, especially those that are part of staged repair procedures, and should house at least one specialist surgeon with training in pediatric general surgery.
- The third-level hospital must be able to provide treatment for a broader range of neonatal emergencies and more complex urgent conditions. The availability of specialist pediatric surgeons and anesthesia providers with expertise in infants and children is critical.

Research Priorities

Improved data collection and identification of disparities will fuel advocacy and inform targeted intervention programs for congenital anomalies. Research priorities include the following:

Data Expansion for Further Evaluation. Epidemiology, prevalence, and incidence of disease in various health contexts need further evaluation. Epidemiology may vary locally, but additional data are needed (Bickler 2000). Registries for selected anomalies may assist in improving surveillance (for example, via participation in the International Clearinghouse for Birth Defects Surveillance and Research). The evaluation of hidden mortality and morbidity will better approximate the true burden of disease.

Capacity Assessment and Guideline Development. The capacity for pediatric surgical care at various levels of the health system must be assessed, and guidelines for minimum human resources and infrastructure for countries at different stages of development should be created. The WHO Tool for Situational Analysis to Assess Emergency and Essential Surgical Care includes only two items pertaining to pediatric surgical care, and an alternative capacity tool has been proposed (Nacul and others 2013). This tool could be refined and further evaluated as it is piloted in different countries. Although surgical outreach programs can tackle the backlog of nonemergency conditions, emergency conditions require development of the whole health system. More work is needed to define and develop the mechanisms to strengthen systems for pediatric surgery.

Metric Optimization. The quantitative metrics of disease burden should be optimized. Although DALYs are an accepted metric, they are difficult to apply practically. Surgical backlogs can be calculated for congenital anomalies and can be a useful advocacy tool for estimating the resources needed to treat prevalent, nonfatal anomalies. Improved measurements of the burden imposed by delayed access to care have not yet been developed. In middle- and high-income countries, many prevalent congenital anomalies are treated in the first year of life; in LICs, they are never treated or are treated years later, after children have suffered unnecessary complications. Many diseases with 100 percent survivability in HICs result in death or permanent disability in LICs and LMICs. Improved measurement of disparities must be highlighted as an advocacy tool for health equity.

Integration Model Evaluation. Models for the integration of pediatric surgical services within existing child health initiatives should be evaluated. Large-scale child health initiatives exist (such as the WHO's Integrated Management of Childhood Illness but have not historically included surgical care. Providers of children's surgical services share a general concern that if the particular needs of children are not specifically addressed, then they are often neglected. Although congenital anomalies are sometimes considered noncommunicable diseases (as in the Global Burden of Disease study), the agenda of the noncommunicable disease movement has not addressed them. Furthermore, greater planning is needed between networks of specialty organizations and providers treating a broad range of congenital anomalies to collaborate where possible.

Cost-Effectiveness Data Generation. Cost-effectiveness data should be generated to evaluate and select models to improve access to care. Although cost-effectiveness has been estimated for adult general surgery wards in selected hospitals (Gosselin, Thind, and Bellardinelli 2006), only one attempt has been made for pediatric surgical wards (Rattray and others 2013). Low-cost technological innovations and modification of surgical procedures hold great promise to improve perioperative care (Hadley 2008). Cost-effectiveness analysis of training programs could also aid advocacy for greater resources for training.

Marketing and Advocacy Alignment. Marketing and advocacy should become more aligned. Congenital

anomalies vary greatly in scope and severity. Some treatable visible anomalies have received greater emphasis than those invisible anomalies for which it has been more difficult to engage donor programs for targeted support. Improved multidimensional measurements of the burden may help to make children and families suffering from all treatable anomalies more visible to the public health community. Development of innovative strategies for this process is needed.

CONCLUSIONS

This chapter highlights the considerable burden of disease associated with congenital anomalies and outlines key strategies for intervention. The consequences of nonintervention are readily apparent. It is both an economic and a moral imperative that global partners invest in pediatric surgery as a vital component of reducing the burden of disease and improving the public health and economic fortunes of LICs and LMICs.

NOTES

The World Bank classifies countries according to four income groupings. Income is measured using gross national income (GNI) per capita, in U.S. dollars, converted from local currency using the *World Bank Atlas* method. Classifications as of July 2014 are as follows:

- Low-income countries (LICs) = US$1,045 or less in 2013
- Middle-income countries (MICs) are subdivided:
  - Lower-middle-income (LMICs) = US$1,046 to US$4,125
  - Upper-middle-income (UMICs) = US$4,126 to US$12,745
- High-income countries (HICs) = US$12,746 or more

1. Rectal biopsy: A surgeon removes small samples of tissue from the patient's rectum. These samples are subjected to laboratory tests that lead to a diagnosis.
2. Sepsis: Severe widespread infection within the body that can lead to death.
3. Colostomy: Temporary creation of an opening in the abdomen that is connected to the intestine; fecal material exits the opening into a colostomy bag.
4. Allied health or paramedical practitioners: Nonphysician health professionals who provide supplementary or emergency health services.
5. Smile Train. "Smile Train Report Card." http://www.smiletrain.org/our-model/.
6. Telemedicine: Using technology to exchange information between different locations to improve training or clinical care.

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