Life Beyond Surgery

ADDRESSING THE NEGLECTED ISSUE OF PROVIDING LONG-TERM CONGENITAL HEART CARE IN MIDDLE-INCOME COUNTRIES

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EXECUTIVE SUMMARY

Congenital heart disease [CHD] is among the top four causes of infant mortality in all middle-income countries, yet minimal attention has been given to this issue in global health efforts. As childhood survival increases due to improved detection and surgical access, life-long care planning will be needed for patients suffering from chronic congenital heart disease. The goal of this project is to inform these efforts by addressing three areas: adult congenital heart disease [ACHD] burden, ACHD resource needs, and in-country perspectives on barriers to long-term care.

ACHD patients in high-income countries have high rates of morbidity and mortality, resulting in rising costs to healthcare systems, and initial data suggest that disease burden may be higher in middle-income settings. Middle-income countries do not have needed resources for long-term CHD care for children or adults, and in-country experts report major gaps in services, low levels of CHD awareness, and societal barriers to care. Existing ACHD guidelines were seen as unrealistic or inappropriate for many middle-income settings.

The consideration of ACHD disease burden will lead to better-informed surgical decision making and can help guide the structure of developing congenital cardiac care systems. Using a proposed life-long care model, ACHD needs can be incorporated into current efforts to expand pediatric cardiac services. Many actions can be initiated now, such as establishing follow-up plans at the primary care level, initiating targeted interventions in high-risk populations, incorporating ACHD in existing training efforts, and improving education regarding follow-up needs for both parents and providers.

The provision of life-long CHD care will require a significant expansion of awareness, resources, and government investment, and an advocacy road map is proposed to guide efforts by healthcare providers and patient groups. If these efforts are successful, today's children will benefit from the improved health that comes from high-quality CHD care across the life-span.

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BACKGROUND AND JUSTIFICATION

CONGENITAL HEART DISEASE AND INFANT MORTALITY

Congenital heart defects are in-born malformations of the heart's structure such as valve deformities, underdeveloped ventricles, or holes in the heart's inner walls. The International Classification of Diseases recognizes 59 different congenital heart defect diagnoses, representing an array of conditions varying in impact and severity.[1] It is the most common birth defect with reported global birth prevalence between 6.5 and 9.3 per 1,000 births, and accounts for just under one third of all birth defects[2,3] Each year approximately 1.35 million children worldwide are born with heart defects[2], 90% in low resource settings.[4] The majority of these children require childhood heart surgery to survive, and over one quarter will die without surgical repair within the first weeks or months of life[5]. Prompt access to congenital heart surgery allows ~90% of children with congenital heart disease [CHD*] in high-income countries to survive to age 18[6,7] , and the majority of CHD patients are now post-repair adults[8].

CHD rank in < 1 Mortality		
	2002	2017
Low SDI	8	5
Low-middle SDI	6	4
Middle SDI	3	2
High-middle SDI	2	1
High SDI	2	2
Global	5	4

Table 1
Source: Institute for Health Metrics and Evaluation (IHME). http://vizhub.healthdata.org/gbd-compare. (Accessed May 19, 2019)

In contrast, most children born with CHD in low- and middle-income countries have no access to the care needed for survival. A major barrier to progress in CHD care access is the continuing "invisibility" of CHD from the global health agenda and lack of awareness of its prevalence and impact.[9] In countries still struggling to fight communicable disease, children typically die undiagnosed, masking CHD's role in overall infant mortality. However, due to the decrease of communicable disease, CHD has begun to emerge as a major cause of infant mortality in low and middle-income countries, now ranking first in high-middle, second in middle, fourth in low-middle, and fifth in low-income countries [Table 1].[10]

DEVELOPMENT OF CHD SURGERY IN LOW AND MIDDLE-INCOME COUNTRIES

Mirroring overall tertiary care development, CHD surgery in most low-income countries is available only by transporting children to charitable care at foreign medical centers, or through medical missions able to treat a small number of children each year. In these countries, the development of

***Congenital Heart Defect vs. Congenital Heart Disease:** Congenital heart defects are in-born structural anomalies in the heart, whereas congenital heart disease is the negative health impact that results from these defects. The terms "congenital heart disease" and "congenital heart defect" are commonly used interchangeably. However, many congenital heart patients object to the use of "disease" in describing their underlying condition as it implies they were born "diseased"[11]. In this paper, "ACHD" and "CHD" will be used to refer to adult congenital heart disease and congenital heart disease. "Congenital heart defects" will not be abbreviated.*

healthcare basics is needed before sustainable in-country pediatric cardiology surgery can begin. The focus of this project is middle-income countries, in which progress in CHD surgical availability is increasing, via independent pediatric heart centers, adult cardiology facilities, or both. Two organizations, the World Society of Pediatric and Congenital Heart Surgery and the International Quality Improvement Collaborative for Congenital Heart Disease, offer participation in surgical registries to promote research and quality improvement. More than 70 CHD surgical centers in middle-income countries currently participate in these databases, which collectively enrolled over 100,000 cases since 2007. Although this represents a tiny portion of children in need of surgery in that time period, it demonstrates growing access to CHD surgical care.

In addition to availability of diagnosis and medical facilities, cost continues to be a major barrier to care. The majority of health systems currently require partial payment for CHD surgery. This can lead to catastrophic health care costs and delays in access that have major impacts on outcomes.[9] There is growing recognition that creating sustainable pediatric cardiac care requires government engagement, with surgical funding available for those unable to cover the costs.[12]

Efforts to include congenital heart disease in global noncommunicable disease efforts have begun, led by organizations such as Children's HeartLink and the Global Alliance for Rheumatic and Congenital Hearts. Thus far, fundraising, advocacy and awareness efforts have primarily focused on securing cardiac surgery for the tens of thousands of children who die each year due to untreated CHD. Reaching this goal will allow middle-income countries to make the transition high-income countries achieved in the 1970's - the evolution of CHD from a fatal to chronic childhood-onset disease.

ACHD CARE IN HIGH-INCOME COUNTRIES: MISSED OPPORTUNITIES, CONTINUED CHALLENGES

Despite expectations that most CHD surgeries would be curative, as substantial numbers of CHD patients started to reach adulthood, unexpectedly high rates of late-onset complications began to be reported. By the early 2000's a substantial body of research documented high rates of cardiac and non-cardiac complications, new hemodynamic problems requiring additional surgery[13–15] , and significant pregnancy-related morbidity and mortality. [16]In 1996 the Canadian Adult Congenital Heart Network was formed and, due to the highly complex needs of the patients and the lack of adult congenital heart disease [ACHD] expertise offered by community cardiologists, created a national network of specialized ACHD programs led by cardiologists with training in adult congenital heart disease.[17] Over the next fifteen years, multiple management guidelines were published reiterating the need for ACHD-specific tertiary care centers.[18–21] These recommendations were strengthened with publication of studies reporting significantly lower morbidity and mortality among patients receiving care at ACHD programs.[22–25]

Two major barriers were identified to implementing guideline-compliant care: the severe shortage of medical staff with training in ACHD and the fact that large numbers of adult congenital heart patients had been lost to follow up during childhood or adolescence.[26] Two major drivers of these gaps in care were found to be a lack of awareness of need for follow-up and minimal knowledge of the long-term risks of their disease.[11,27,28] In 2011, transition guidelines were published emphasizing the need for targeted education and structured transfer to adult congenital heart disease programs during

adolescence or young adulthood.[29] To address the workforce gap, the American Board of Internal Medicine began formal subspecialty certification in adult congenital heart disease in 2014.[30]

Yet despite over twenty years of efforts, major workforce gaps persist, access to recommended care remains limited, and significant numbers of congenital heart patients continue to be lost to care.[31] A recent study found that only 25% of pediatric patients in Germany successfully transferred to ACHD care[32]; in the United States reported rates fall to 12%,[33] with one 2019 study reporting that 47% of families were lost to cardiac care before age 5.[34] In Japan, fewer than 5% of ACHD patients are estimated to be receiving guideline-compliant care in 2016 ,[35] and a 2019 survey of 96 European ACHD programs found that only 22% of the ACHD patient population were receiving care from ACHD specialists and 96% of European ACHD centers were not fully compliant with European ACHD guidelines.[36]

The late adoption of CHD surgeries allows middle-income countries to benefit from fifty years of growing congenital heart knowledge, including the large body of research regarding the long-term health risks of CHD. Countries now expanding congenital heart care systems can avoid the mistakes made in high-income countries by planning ahead for the long-term needs of those living with chronic congenital heart disease. Surgical decision-making can be based not only on short-term outcomes, but also on the on-going burden of disease and level of life-long care patients will need from their families and the medical system. Middle-income countries also have the opportunity to develop strategies appropriate for their settings and identify novel approaches to care for this new and challenging patient population.

OBJECTIVES

The objective of this project is to make the case for the need for ACHD planning in middle-income countries and provide information to inform efforts to initiate such care. The report provides an overview of ACHD burden of disease in high- and middle-income countries, provides a summary of ACHD care guidelines to identify needed services, and provides expert in-country perspectives on current barriers to care and priorities for action. In addition, novel strategies to promote ACHD care planning in middle-income countries are proposed.

CHAPTER 1: HOW DOES ACHD BURDEN DIFFER IN HIGH- AND MIDDLE-INCOME COUNTRIES?

METHODOLOGY

A review of ACHD research was executed to provide an overview of current evidence regarding the status and impact of ACHD in high- and middle-income countries. Priority was given to studies referenced in the most recent published ACHD care guidelines, systematic reviews, studies using large data sets such as single- or multi-country registries or administrative data sources, and those published within the last three years. The history and current status of ACHD care in high-income countries, ACHD prevalence, mortality, morbidity, mental health, quality of life, health care utilization, and economic impact in high- and middle-income countries was summarized, and similarities and differences between settings were highlighted.

LIMITATIONS

Neither a structured literature review nor a systematic review was executed, and although systematic reviews were cited when possible, few are available. The review articles that served as primary sources of research citations in middle-income countries were published before 2017 and, given the rapid recent increase in ACHD research publications in these regions, it is likely that relevant articles were missed. ACHD research from middle-income countries is almost exclusively single-center studies and illustrates the need for research development.

RESULTS

ACHD BURDEN OF DISEASE IN HIGH-INCOME COUNTRIES

In high-income countries, the burden of morbidity and mortality in all forms of congenital heart disease is significantly higher than in the general population, with severity of impact rising with disease complexity.[13] Congenital heart disease is typically classified using complexity categories first proposed by Webb et al at the 32nd Bethesda Conference on the Care for Adults with Congenital Heart Disease.[19] Diagnoses are defined as simple, moderately complex, and highly complex, with moderately and highly complex defects representing an estimated 50% of all defects. Table 2 lists the most common diagnoses in each category.[19]

Mortality and morbidity risk in ACHD patients

Simple Defects
Small unrepaired atrial septal defect
Mild valve disease
Repaired Ventricular Septal Defect
Repaired Atrial Septal Defect
Repaired Patent Ductus Arteriosus
Moderately Complex Defects
Simple defects w/residual or new cardiac issues
Coarctation of the Aorta
Tetralogy of Fallot
Moderate to severe valve disease
Highly Complex Defects
Single Ventricle – all forms
Transposition disorders– all forms
Eisenmenger Syndrome

Table 2: ACHD Complexity Categories

Although simple repaired defects were previously found to be relatively low risk compared to the general population,[13] more recent studies have reported that those with repaired simple defects have between a 72% and two-fold increase in mortality risk[37,38] ; in one study, this increased risk was only seen in women, and correlated to mortality during labor and delivery.[39] Higher mortality in simple defects is also significantly associated with later repair.[40] CHD patients with moderately complex defects show mortality rates two to three times higher than their age-matched peers, and those with highly complex defects are between five and 23 times higher risk of mortality.[38] Heart failure is a leading cause of mortality and is responsible for between 26% and 43% of all deaths.[38,41] Arrythmia is over three times more common in this group than in the general population and rises with age, with 29% of those with simple defects, and 49% of those with more complex conditions, developing arrythmia by age 50.[39]

ACHD impact on pregnancy

Although successful pregnancy and delivery is now possible for the majority of women with CHD, it continues to be associated with higher morbidity and mortality. A recent study using registry data from 28 countries reported an overall maternal mortality rate of 1%, with significantly higher rates reported for those with more complex defects.[42] Rates of recurrence of heart defects among the children of ACHD patients appears to vary widely based on defect, with a 2019 study reporting that although the overall recurrence rate was 1.8%, among certain defects there was a 50% rate of recurrence.[42]

Acquired heart disease risk in ACHD patients

In addition to the direct effects of their underlying condition, CHD patients are at increased risk from adult-onset cardiovascular conditions, with a recent UK study finding that ACHD patients with simple heart defects and two cardiovascular disease risk factors were twice as likely to experience acute coronary syndrome or stroke than non-CHD patients with five risk factors.[43] A 2018 review of research on acquired heart disease risks concluded that 80% of ACHD patients have one or more risk factor, with high reported rates of physical inactivity, hypertension, metabolic syndrome, and obesity rates as high or higher than the general population.[44–46] ACHD patients have also been reported to have a 70% increased risk of cancer[47] and to develop premature dementia more than twice as often as the general population.[48]

Impact of ACHD care on healthcare systems

The rapid growth of the ACHD population has resulted in significant burden to the healthcare system, with US ACHD hospitalizations doubling between 1998 and 2011,[49] outpacing pediatric CHD admissions.[50,51] A recent systematic review of 21 studies on ACHD healthcare utilization in Europe, Canada, and the US reported that compared to the general population, ACHD patients had a 4- to 8-fold

increased risk of hospital visits, with 60% experiencing multiple hospitalizations and 69% reporting emergency room visits within a five year period.[51] ACHD hospital stays, whether for cardiac or non-cardiac causes, are longer and more expensive [49,52] than those for non-CHD patients with the same conditions.

Psychosocial impact of ACHD

In addition to mortality and physical morbidities, ACHD patients report higher rates of depression and anxiety than their peers.[53–55] Data on quality of life are mixed, but lower quality of life has been consistently associated with more physical limitations, inability to perform desired activities, lower levels of education, unemployment, and not having adequate income.[56–64] A recent systematic review by Seckeler et al concluded that ACHD patients have significant barriers to education, employment, and earning power, as well as additional financial burdens due to direct and indirect healthcare costs and lack of access to life, disability, and health insurance.[65]

ACHD BURDEN OF DISEASE IN MIDDLE-INCOME COUNTRIES

In contrast to the several-fold increase in ACHD research in high-income countries, research in middle-income countries has lagged.[66] However, a significant increase has occurred in the last five years, including four publications which summarize current ACHD research findings, workforce gaps, and care needs.[66–69]

ACHD prevalence and clinical profile in middle-income countries

Due to limited access to childhood surgery, the age structure of CHD in middle-income countries shows a large predominance of children under five, with an estimated 30% survival to five, and 10% to 20% survival to age 18.[67] This population is comprised of the small subset of CHD patients able to survive without surgery, as was seen in high-income countries until the mid-20th century. Research on prevalence in ACHD has not been done in the community, but of those presenting for surgery, 52% to 75% have simple defects; tetralogy of Fallot, a moderately complex defect, is present in 10%-26% of cases. Reported rates of previous surgeries range from 14% in New Delhi to 34% in Singapore and South Africa[67,69] Between 6% and 20% are reported to have developed Eisenmenger Syndrome, a complex multi-system disorder that results from long-term cyanosis and precludes surgical repair.[69] When possible, surgical or catheter-based repair is attempted with higher rates of morbidity and mortality.[67,69] Outcomes data are largely limited to surgical outcomes, and minimal data are available on long-term morbidity and mortality.

ACHD impact on pregnancy

A recent registry-based study by Roos-Hesselink et al. found that women in middle-income countries experienced significantly higher mortality and morbidity than those in high-income settings, although the findings were limited due to small sample size.[70] Studies on pregnancy management in middle-income countries consistently report significant numbers of women with Eisenmenger syndrome,[71,72]which is in the highest risk category (class 4) in the World Health Organization classifications of cardiovascular disease in pregnancy.[73] Although research is lacking, multiple pregnancies among women with CHD in middle-income countries appear to be more common, and

experiencing two or more pregnancies has been associated with poorer maternal outcomes.[74] In areas in which marriage to relatives is common, significantly higher rates of CHD incidence have been reported, and studies from Pakistan and Saudi Arabia report up to a 20% prevalence of CHD in areas where consanguity rates are high.[75–77] No data on rates of CHD recurrence in ACHD patients in middle-income countries is available, but it can be hypothesized that consanguity will increase the risk of recurrence.

Psychosocial Impact of ACHD

Little research has been done on mental health, quality of life, or impact on income or employment in ACHD patients in middle-income countries, but initial data reports similar or worse outcomes than high-income countries.[78–80] One recent study by Moons et al. exploring variation in ACHD quality of life in fifteen countries found that ACHD patients from countries with higher standard of living and health care expenditure had higher quality of life, but only a single center from one middle-income country (India) was included in the analysis[81].

DISCUSSION

Anticipating ACHD long-term care needs in middle-income countries

As greater numbers of children in middle-income countries gain access to CHD surgery, one can anticipate that the ACHD case mix will more closely resemble that in high-income countries. Repair of moderately complex defects is increasingly routine, and repairs on highly complex conditions are steadily increasing. However, even if the rate of CHD surgical development increases dramatically over the next ten years, changes in the ACHD case mix can be expected to occur slowly over the next decades.

Because of the predominance of CHD patients with simple defects, which have lower rates of long-term morbidity and mortality than more complex defects, it has been argued that ACHD patients in middle-income settings will have less need for on-going ACHD care. However, simple repaired defects recently have been associated with substantial increases in morbidity and mortality.[37,39] Tetralogy of Fallot, a moderately complex defect, is common, and Eisenmenger syndrome, a highly-complex condition with exceptionally high morbidity and mortality, makes up a significant proportion of the case mix .[82,83] In addition, the majority of CHD survivors in middle-income countries will have undergone late repair, which increases risks of long-term complications in all forms of CHD.[84–86] However, it is important to note that this means many will be “natural survivors”, whose individual constellation of heart and lung anatomy enables survival to an older age. The phenomenon of “natural survivorhood” is not well-understood, but it has been hypothesized that as a group they not only have more favorable anatomy but also greater overall health resilience.

Improved CHD outcomes, continued chronic disease

Today’s surgical practices reflect the evolution of pediatric cardiology over the last six decades, and the high morbidity and mortality seen in long-term survivors may result in part from more primitive surgical techniques used in an earlier era. New technologies such as catheter-based valve replacements continue to improve outcomes[87], and a growing body of high-quality evidence, including findings from

the first large prospective cohort studies and randomized controlled trials, promises to improve clinical care[66]. However, structural defects are only one aspect of the complex cardiovascular abnormalities that affect CHD patients throughout the lifespan, and all heart surgeries and interventions continue to be associated with higher rates of long-term morbidity and mortality. Fundamentally, CHD patients will never have normal hearts, and the combination of underlying anatomy and surgical insult will result in a growing cohort of patients suffering from chronic CHD.

RECOMMENDATIONS

Anticipating ACHD risks in middle-income countries

The data on ACHD disease burden can offer anticipatory guidance on actions that can be taken now to prevent future problems likely to be heightened in middle-income settings. For example, the rapid increase in behavioral risk factors for adult-onset heart disease in many middle-income countries, combined with the heightened risk from acquired heart disease reported among ACHD patients, suggests that interventions to promote physical activity be prioritized. The combination of risk of pregnancy, higher fertility, and lower access to family planning could be addressed by heightened attention to reproductive issues.

Using long-term CHD outcomes to inform surgical decision-making

Reviewing the literature on ACHD burden of disease raises difficult questions about surgical decision-making in low resource settings. In high-income countries, progress in pediatric cardiology has been defined as the expansion of surgical options to allow survival for those born with even the most severe heart defects. This has resulted in rapidly growing patient populations with high levels of mortality and complex morbidities requiring high levels of on-going tertiary care.

Although morbidity and mortality are high in all those with complex defects, outcomes for those born with one working ventricle are strikingly worse. These infants require a series of open-heart surgeries that result in a heart that allows survival but has profoundly abnormal anatomy. Although post-surgical childhood mortality is low in other forms of complex CHD, those with univentricular hearts experience high childhood mortality, with one study reporting 24% mortality within ten years of surgery.[88] In adulthood, multi-system disorders, such as liver failure and protein-losing enteropathy, often develop, further worsening outcomes. Whereas the overall risk of death in highly complex CHD is 14 times higher than the general population, it is 23 times higher for those living with single ventricles, and at age 20 their risk of death is equivalent to non-CHD patients at age 64.[38]

The surgical risk rating system used in congenital heart surgery defines reparative surgeries for many complex defects, including several forms of univentricular hearts, as mid-level operations [89]. As facilities and expertise improve, it is likely that increasing numbers of surgical centers will be able to execute these procedures in the near future. The decision to operate should consider not only the immediate surgical risk, but also the on-going burden of disease and high level of long-term care these patients will need from their families and the medical system.

CHAPTER 2: WHAT RESOURCES ARE NEEDED TO CARE FOR ACHD PATIENTS?

METHODOLOGY

The goal of the analysis was to summarize the minimum core requirements for a guideline-compliant ACHD program. All three published ACHD-specific care guidelines, as identified in a systematic review executed during the development process of the 2018 American College of Cardiology/American Heart Association ACHD Care Guidelines [90], were reviewed. These Guidelines were developed by the American College of Cardiology and the American Heart Association[90] , the European Society of Cardiology[86] and the Canadian Cardiovascular Society.[91] The description of the required elements of an ACHD program were reviewed to identify common elements. Overall recommendations regarding which diagnoses required care at an ACHD program, how often, and in which situations were also reviewed and summarized. Recommendations for clinical management were consulted to identify commonly-needed cardiac testing technology, medications and devices, but diagnosis-specific treatment guidelines and clinical content were not reviewed. Recommendations regarding exercise, psychosocial and mental health needs, and patient education were also reviewed to identify both commonly-identified needs and recommended practices. In addition to the clinical care guidelines reviewed above, the guidelines on CHD care transition and physical exercise were consulted.[29,92] A brief summary of commonly identified needs and recommended best practices was then created.

LIMITATIONS

Although the ACHD program elements and follow-up guidelines clearly outlined essential elements, key priorities among the psychosocial and educational recommendations were not as clearly defined, increasing the potential for author bias.

RESULTS

All guidelines reviewed based their ACHD program elements and follow-up guidelines on those originally proposed by Webb et al in 2002, and referenced this source as their model.[19] Table 3 summarizes the essential elements of an ACHD program The list of technology, devices, and medications were extrapolated from the list of needed services, as well as the clinical recommendations.

Table 3: Essential ACHD Program Elements

Essential Staffing	
Minimum 2 congenital heart surgeons Minimum two formally trained ACHD cardiologists ACHD-experienced Electrophysiologist Interventionalist Advanced cardiac imaging specialist Anesthesiologist Nurse Practitioners or Physicians assistants	
Essential Services	
ACHD Outpatient Clinic Adult Inpatient ward Intensive Care Unit Pulmonary hypertension Management	High-risk Obstetrics Genetic Counselling Social workers Psychological services Access to Heart Transplant
Essential Technology and Devices	
Electrocardiogram Echocardiography Cardiac magnetic/nuclear testing	Pacemakers/implantable defibrillators Tissue/mechanical valves Closure devices
Essential Medications	
Diuretics Betablockers ACE inhibitors Pulmonary hypertension medications	Anti-arrhythmia medications Warfarin Heparin

Staffing: All ACHD programs are required to have access to two congenital heart surgeons, defined as pediatric heart surgeons with expertise in ACHD, and an ACHD-experienced surgical team and anesthesiologist. A minimum of two cardiologists with two years of formal ACHD fellowship training are required, and at least one interventional cardiologist, electrophysiologist, cardiac imaging specialist, and mid-level provider (i.e. nurse practitioner, physician assistant, nurse educator) with ACHD expertise are

recommended. Guidelines differed as to how formal training was defined, and the United States guidelines mandates formal ACHD subspecialty certification, which requires completing a specified course of study and passing an exam administered by the American Board of Internal Medicine.

Services: All ACHD programs are required to offer an ACHD-specific outpatient clinic and heart transplant services. High-risk pregnancy services, pulmonary hypertension management and genetic counselling should also be available. The more recent guidelines also mandate access to palliative care and a structured transition program.

Table 4: ACHD Follow-up Recommendations	
Category	**ACHD Follow-up needed**
Simple Defects	Single Visit to confirm No symptoms No hemodynamic issues Follow up plan with local cardiologist/PC
Moderately Complex Defects	ACHD follow-up visit every 2 years or less All cardiac procedures done at ACHD center ACHD check before pregnancy ACHD Consultation during pregnancy ACHD Consultation before anesthesia ACHD-specific family planning provided
Highly Complex Defects	ACHD follow-up visit every 1 year or less All cardiac procedures done at ACHD center ACHD consultation before pregnancy All pregnancy and delivery at ACHD program All surgery and anesthesia at ACHD program ACHD-specific family planning provided

Devices, Technology, and Medication Needs: All guidelines assume access to pacemakers and implantable cardiac defibrillators, tissue and mechanical heart valves, and catheter-implanted devices such as stents, closure devices, and valve replacements. The full range of electrophysiologic testing, such as electrocardiograms, echocardiography, and magnetic resonance and nuclear testing, is also needed. There are no CHD-specific medications, but access to standard cardiac medications such as blood pressure medications, diuretics, and antiarrhythmics, as well as pulmonary hypertension medications, is required. Anticoagulation medication such as warfarin and heparin, and access to regular anticoagulation management, is also mandated.

Need for Follow-up: A summary of follow-up recommendations is available in Table 4. All three guidelines advise minimum follow-up using the complexity-based system originally defined by Webb et al[19]. The recent US Guidelines combine these complexity categories with four defined "physiologic" stages of disease severity which incorporate metrics such as exercise function and severity of complications.[90] In this system, patient's need for follow-up is determined both by their diagnosis and by their current symptoms. The European and Canadian guidelines do not formally define physiologic stages of disease but emphasize that those experiencing more severe symptoms should be seen more frequently.

For those with simple defects, an ACHD program visit is recommended to evaluate the presence of cardiac symptoms or abnormal hemodynamics which can develop from the underlying defect or leakage in the repair.[90] If no problems are found, a follow up plan is established with a local cardiologist and/or primary care provider, which typically recommends cardiac checks at three- to five-year intervals and referral back to the ACHD program if new problems occur.

Regular checks by the ACHD program are recommended for all other ACHD patients, with the minimum follow up interval every two years for moderately complex CHD and every year for complex CHD. A pre-pregnancy visit is mandated in both moderately and highly complex defects in order to provide genetic counselling and assess overall pregnancy risk. If considered safe, those with moderate defects can

deliver in a standard setting in consultation with the ACHD team. Those with highly complex defects should have their pregnancy directly managed at the ACHD program. Similarly, those with moderate CHD should consult with their ACHD team before undergoing any procedure requiring anesthesia, and those with highly complex CHD should have all anesthesia executed under the direct supervision of the ACHD program.

Psychosocial and Educational Needs: All the guidelines offered extensive information regarding the psychosocial challenges which have been synthesized down to a core list of psychosocial challenges and best practices to address these issues. [Table 5].

Table 5: ACHD Psychosocial Recommendations

ACHD Psychosocial Issues	ACHD Guideline Recommendations
• Heightened risk of anxiety, depression, and/or PTSD • Social isolation and/or feeling "different" • Social stigma and workplace discrimination • Barriers to education and employment • Barriers to exercise due to misperception of risk/limitations • Concerns about dating, marriage and becoming a parent • Financial burden of health care and associated costs • Social and emotional challenges of managing illness, disability, and premature mortality	• ACHD patients should be screened for mental health problems. • Appropriate treatment and counselling should be available. • Opportunities to meet other ACHD patients should be provided. • Appropriate career/educational planning should be encouraged. • Exercise should be encouraged when possible, and specific safe activities identified. • Education should be provided on healthy eating and lifestyle choices.

All guidelines stressed the need to offer mental health screening and services and recommended providing opportunities to meet other patients and families. The issue of career planning was highlighted to ensure that patients defined long-term career goals that maximized their abilities and were within their physical capabilities. Given the widespread inactivity among ACHD patients, often due to concerns about risk to health, the importance of not only promoting exercise but also defining specific safe activities was emphasized. Guidance on healthy eating and weight control was also recommended since ACHD patients may not perceive themselves at risk for acquired heart disease.

Education and Transition Recommendations:

Table 6: ACHD Education and Transition Recommendations

• Families and the medical team should provide children on-going developmentally-appropriate education about their heart condition beginning at age 3 – 5. • The information that CHD is a chronic condition and life-long follow-up is needed should be directly communicated to families and patients and reiterated regularly. • Opportunities should be offered to meet with the cardiology team independently beginning in late childhood/early teens. • Discussions about the need for family planning should begin in the teen years.	• By age 18 - 21, patients who are cognitively/developmentally ready should be able to independently take care of their heart health needs, with families providing support • Patients leaving pediatric care should be provided a written care plan and understand how and when to seek care. • Families and patients should keep on-going records of their medical procedures and issues. • In addition to diagnosis and procedures, education should include specific risks, and how to recognize and respond to cardiac symptoms.

Table 6 lists those recommendations that deal with the patient's understanding the disease and need for follow-up and ongoing care; most include a specific action that should be done by a specified time. The transition guidelines were consulted to identify the age range in which certain milestones should be reached. All documents consulted agreed on the need for on-going education to support the ability of the patient to manage his/her own condition, regardless of whether care transfers to an ACHD program

or continues with the pediatric cardiologist. This education should start in childhood and be specific, understandable, and tailored to the developmental stage of the patient. The importance of ensuring understanding that the patient had not been "cured" was emphasized.

DISCUSSION

Basic cardiac services and staff are lacking in middle-income settings

The ACHD program specifications require access to secondary and tertiary level cardiac services, such as pacemaker and ICD implantation, advanced cardiac imaging, interventional cardiology, cardiac surgery, and access to heart transplant, rarely available in low resource settings. A recent review of cardiovascular bioimaging in middle-income countries concluded that although the availability of echocardiography had significantly increased, access to cardiac magnetic and nuclear testing is minimal.[93] Electrophysiology access is also minimal, and low-resource countries implant fewer than 10 pacemaker per one million people, with some programs now turning to pacemaker reuse to address this critical need[94,95] New initiatives have been proposed to expand global access to treatment for rhythm disorders, but as yet overall availability is not significantly increasing.[94] Cardiac surgery is not among 44 proposed essential surgical procedures in the 3rd Edition of Disease Control Priorities,[96]and even in middle-income countries such as India and China, where availability is rapidly increasing, access remains largely limited to those in urban areas with access to private healthcare services.[97] Because the cardiac medications used in ACHD care are the same as those used in other cardiovascular disease, they are included on the essential medicine list and typically available in middle-income countries, but access challenges due to supply and cost are widespread.[98] All of these challenges must be taken into account when designing models of ACHD care appropriate for low-resource settings.

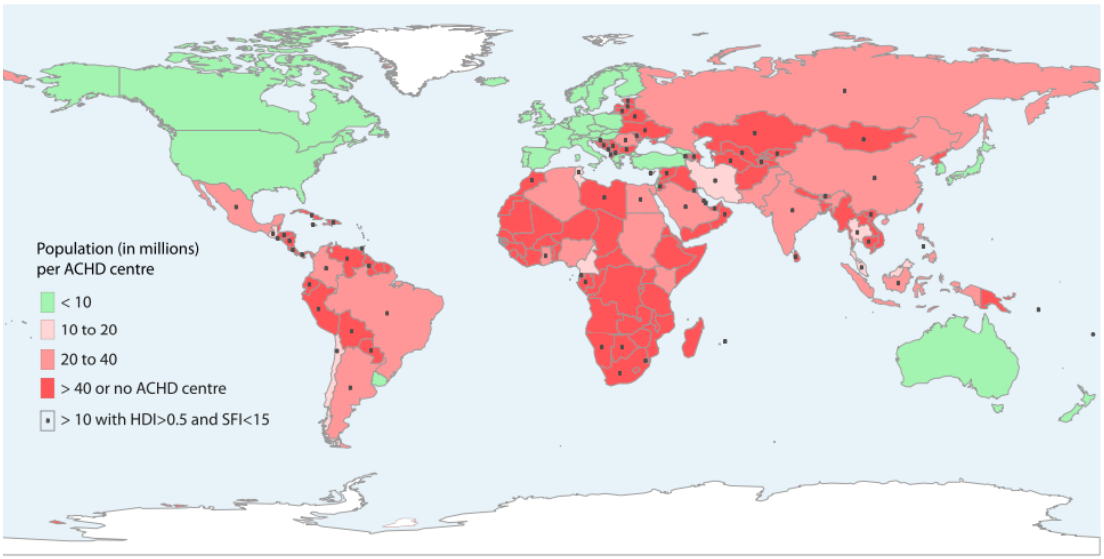

Figure 1: Worldwide distribution of ACHD programs

Source: Kempny et al; Meeting the challenge: The evolving global landscape of adult congenital heart disease. Int J cardiol. 2013 [66]; used with permission

Absence of ACHD-trained staff is a fundamental barrier to care

Foundational to the ACHD program is the provision of care from cardiologists with formal training in ACHD. Although specific research on the level of ACHD training among cardiologists in middle-income countries is lacking, four recent analyses of ACHD care in low-resource settings have concluded that the absence of any ACHD-trained staff is the fundamental barrier to the provision of ACHD care.[66–69] In *"Meeting the challenge: The evolving global landscape of adult congenital heart disease",* Kempny et al analyzed the worldwide distribution and volume of ACHD research published between 1995 and 2011, and identified any center publishing one or more ACHD research publication as an ACHD center. Using this broad definition, a profound gap was found between recommended ACHD centers per person of 1 per 10 million population and the global distribution of ACHD care.[66] (Figure 1). With the exception of Lebanon, no middle-income countries were found to meet the 1 per 10 million threshold, with numbers of ACHD programs calculated to be between 10% and 50% of this level.[66] It is important to note that, given that the criteria for being an "ACHD Center" was publication of a single ACHD-related study, it is likely that the actual availability of trained ACHD providers is substantially lower.

Improving ACHD mortality demands high-quality care

In high-income countries, ACHD programs were not created because ACHD patients lacked access the cardiac services, but to prevent the excess mortality and morbidity seen when patients receive low quality care from cardiologists with minimal training in their complex needs. These quality problems become acute when ACHD patients are operated on by non-CHD surgeons. Low volumes of congenital heart surgery have been consistently linked to high levels of excess mortality in both children and adults**,** leading many European health systems to establish minimum volumes of both pediatric and adult CHD surgery necessary for all CHD surgical centers. [99]

All CHD care begins with sustainable, high-quality pediatric heart surgery

At its foundation, ACHD care is predicated on the development of congenital heart surgery, and this development is still in its infancy in most low-resource settings. The typical CHD patient undergoing surgery in low-resource settings will be operated on by a surgical team lacking in both CHD training and experience. But even the best-trained team cannot optimize outcomes without all the components essential to minimize CHD surgical mortality. Although these challenges face all surgical development in low-resource settings, CHD surgery involves performing complex open-heart procedures on tiny hearts, within cardiac centers able to keep infant patients alive.

Children's HeartLink, a U.S non-profit organization that provides capacity building to pediatric heart centers in middle-income countries, has created a health system model defining the essential elements needed for high-quality pediatric heart care. [Table 7]. A useful comparison can be made between the highly-complex and comprehensive services needed for an ACHD program, and the foundational elements needed for pediatric cardiac care. Each of these elements are essential to adults as well as children, and key ACHD program elements, such as echocardiography and catherization, are included. Others, such as interventionalists and electrophysiology, are not, reflecting Children's HeartLink current focus on needed services between initial diagnosis to operative repair.

Pediatric Cardiac Care in an Ideal Health System

		Level of Care			
Capacity	Continuum of Care	Village Health Center	First-Level Hospital	Second-Level Hospital	Third-Level Hospital
Prenatal care and attended birth	Early Detection	X	X		
Universal screening and referral	Early Detection	X	X	X	X
National congenital birth anomaly reporting mechanism	Surveillance	X	X	X	X
Antibiotic availability and delivery	Prevention (RHD)	X	X	X	X
Pulse oximeter	Diagnosis	X	X	X	X
Echocardiogram, electrocardiogram, and chest x-ray	Diagnosis		X	X	X
Fetal echocardiogram	Diagnosis			X	X
Cardiac catheterization	Diagnosis & Treatment				X
Cardiac medications availability and delivery	Treatment	X	X	X	X
Spinal and general anesthesia	Treatment		X	X	X
Open heart surgery	Treatment				X
Intensive care unit	Treatment				X
Infection control	Quality Assurance	X	X	X	X
Continuing medical education	Quality Assurance	X	X	X	X

Table 7: Pediatric Cardiac Care in an Ideal Health System *Source: Children's HeartLink; used with permission*

RECOMMENDATIONS

Expanding long-term investment in sustainable high-quality CHD surgery

As communicable disease decreases, further improvements in infant mortality will demand the availability of surgical centers able to execute CHD surgery without the high levels of mortality often evident in low-resource settings.[100] But this ability is dependent on sustained capacity building, and typically takes from five to ten years. Most high-quality surgical centers in middle-income countries currently receive significant support from international charitable institutions. The expansion of high-quality CHD care needed to impact CHD mortality will require significant long-term government investment in infrastructure and training.

Including ACHD training in CHD quality improvement efforts

The International Quality Improvement Collaborative has demonstrated that CHD centers in low-resource settings can achieve outcomes comparable to high-income settings.[100]The International Quality Improvement Collaborative is a network of 64 CHD surgical centers in low-resource settings which execute coordinated quality improvement interventions. As part of these efforts, they provide regular remote and in-person training to IQIC hospital staff. By including ACHD training in their efforts, they could improve the quality of care for the ACHD patients currently seen in these centers.

Planning for life-long care

By combining the elements included in the Children's HeartLink Model with the required ACHD program elements, a model identifying all suggested services for on-going CHD care can be created [Table 8]. With the exception of screening, family planning, and genetic counselling, all services listed are needed by children and adults with CHD. Using this life-span model could assist pediatric programs in identifying the components they need as more of their patients not only stay in care but begin reaching their teens.

Table 8: Life-Long Congenital Heart Care in an Ideal System

Capacity	Infants/children (Pre/Post surgery)	Children (On-Going Care)	Teens	Adults
Universal screening and referral*	X			
Pulse Oximetry*	X	X	X	X
Fetal/Child/Adult Echocardiogram*	X	X	X	X
MRI/Cardiac CT*	X	X	X	X
Cardiac Catherization*	X	X	X	X
Open Heart Surgery*	X	X	X	X
Intensive Care Unit*	X	X	X	X
Spinal/General Anesthesia*	X	X	X	X
Access to Heart Transplant *	X	X	X	X
Interventional cardiology*	X	X	X	X
Electrophysiology*	X	X	X	X
Antibiotic Access*	X	X	X	X
Cardiac Medications*	X	X	X	X
Pacemaker/ICD Access*		X	X	X
Psychosocial Support/Mental Health Services*		X	X	X
Family Planning/Birth Control*			X	X
Genetic counselling				X
Prenatal care and attended birth/obstetrics*	X			X
National CHD Reporting/Tracking Mechanism*	X	X	X	X
Infection Control*	X	X	X	X
Continuing Medical Education*	X	X	X	X

Partnering with Primary Care Systems for Long-term Care

ACHD care guidelines state that simple repaired defects without residual issues can be followed up at the primary care level, with cardiac testing performed every three to five years. As more children in middle-income countries undergo successful repair for simple defects, many can be given cardiac care plans and transferred back to local follow-up. This will require establishing partnerships with local healthcare providers, as well as educating parents, primary care providers, and local cardiologists on long-term risks and needs in repaired CHD.

CHD and RHD services: collaboration opportunities

In many middle-income countries, significant numbers of children and young adults in need of cardiac care are rheumatic heart disease [RHD] patients, who are largely treated at the same centers that provide CHD surgery. A recent systematic review of RHD clinical management defined the essential services needed for on-going care of RHD patients.[101] Based on this review, all CHD services also required by RHD patients have been indicated with an asterisk. It is noteworthy that, with the exception of genetic counselling for prevention, all listed services for life-long CHD care are also required by survivors of RHD. Greater recognition of the mutual long-term needs of RHD and CHD patients, resulting in more investment in the above-described services, would benefit both communities.

CHAPTER 3: WHAT DO PROVIDERS AND PATIENTS IDENTIFY AS NEEDS IN ACHD CARE?

METHODOLOGY

CHD professionals and patient and family group leaders from middle-income countries with substantial expertise in ACHD issues in their country were recruited via the Global Alliance for Rheumatic and Congenital Hearts and Children's HeartLink. When possible, group leaders and professionals from the same country were included to allow for better comparison of perspectives.

Seven CHD professionals and nine CHD patient group leaders from seven countries (South Africa, Pakistan, India, Mexico, Brazil, Lebanon, and Malaysia) were interviewed. All of the health care providers interviewed take care of children and teens with CHD. Four direct formal ACHD clinics or programs, one heads a CHD teen transition program, and one serves as a pediatric nurse educator and program coordinator. Four of the patient groups represented provide major funding for CHD surgeries and provide on-going capacity building to a partnering hospital. Five groups work with government health officials, and two provide direct clinical care. Five groups provide support and education services to patients and families. Four of the group leaders are ACHD patients and five are CHD family members (one father, one grandmother, three mothers). One group leader also serves a public health official and one ACHD patient is a surgical fellow. A listing of all interviewees and their affiliations is available in Appendix 1.

All interviewees received the summary ACHD program requirements and psychosocial recommendations presented above. One to two-hour interviews via phone or video were executed using an unstructured format. Each interview began with an invitation to identify the main barriers to ACHD care and key challenges facing ACHD patients in their settings. All interviewees were asked to comment directly on the relevance and feasibility of the ACHD guideline recommendations provided, which were reviewed together at the time of the call. In addition, interviewees were asked to identify priority actions needed to address the unmet needs of ACHD patients in their settings. The information and opinions shared were then synthesized in the summary below. Common and divergent themes were identified, including areas in which patient, family, and provider perspectives significantly differed.

LIMITATIONS

The goal of the interviews was to identify expert opinions, and no formal qualitative research was conducted. The author has had previous contact with four of the healthcare providers and eight of the nine group leaders interviewed, thus potentially biasing the opinions expressed to the author's own. However, all four healthcare providers are authors of multiple papers addressing CHD in developing countries, and two authored papers specifically addressing ACHD in middle-income countries. Similarly, the group leaders were selected not based on familiarity, but because they represent organizations that have had exceptional impact in their country. As with the professionals, their participation ensured that top leaders in CHD advocacy offered their expertise. Their views are not intended to represent those of the average person or CHD patient group leader, but rather reflect their extensive experience working with patients, families, healthcare providers, and government officials in their country.

The absence of Chinese and Vietnamese interviewees is a major limitation given the burden of CHD and the dramatic expansion of CHD services in these areas. Congenital heart surgical centers in China and Vietnam were contacted but did not respond to invitations to participate. No patient support groups from middle-income countries in these areas were identified.

RESULTS

ACHD CLINICAL CARE GUIDELINES

Overall Feasibility and Relevance

All the cardiologists interviewed were familiar with the existing ACHD care guidelines and used them to help guide clinical decision-making, but none felt that implementing the recommended care structure was feasible at this time. Opinions differed about the extent to which meeting the proposed standards should be the long-term goal, and when this might be achievable. Both Brazilian health care providers stated that although trained ACHD staff were not available, their programs often were able to provide recommended CHD testing and follow up. The Indian and Pakistani ACHD program directors, and the South African CHD program director, felt strongly that their countries should aspire to meet the standard but recognized that given the overall health system challenges in their country other health priorities would need to come first. They described the guidelines as a long-term aspirational goal, with several commenting that they were a useful tool to document ACHD care needs to health systems officials.

Two of the cardiologists interviewed agreed that having an aspirational standard was useful but expressed reservations about whether the ACHD care structure proposed was fully needed in their country. One felt that the existing guidelines did not account for important health system differences, such as the fact that many cardiologists in low resource settings see both children and adults and therefore have more expertise in CHD than those in high-income countries. He suggested that ACHD specialty care be reserved for those with more complex defects, and those with valve problems (acquired, congenital, or rheumatic heart disease-related) be managed by structural cardiologists who focus their practice on valve disorders. Another thought that the overall structure was good but that the recommendations on frequency of checkups and testing were excessive and driven by the practice style in high-income countries. The Indian group leader, who is also a public health official, talked extensively about the ways in which basic level ACHD services might be included in current expansions of the community outreach system and the training of community health workers.

None of the group leaders interviewed were familiar with the formal ACHD care guidelines, but all understood the need for life-long CHD care, and all felt strongly that meeting the proposed standard was of importance to their communities, with one patient commenting, *"we deserve this."* Four of their organizations (India, Pakistan, Bulgaria, Mexico) have initiated activities to address life-long care issues, including targeted medical education for cardiologists (India), parent education (Pakistan), and direct meetings with government officials regarding the need for life-long care (Pakistan, Bulgaria, India).

Almost all of the cardiologists interviewed noted that creating some kind of ACHD hand-off system was imperative because pediatric cardiology programs are well over capacity, with long surgical waiting lists

and patient loads of over 300 cases a week. Those who had created separate ACHD clinics (Malaysia, Pakistan, South Africa, Brazil) stated that a major driver was the urgent need to open up space for children. In India and Malaysia, both cardiologists and the public health official noted that, due to the rapidly rising rates of acquired heart disease, adult cardiologists are overloaded and have no incentive or desire to take care of ACHD. Interviewees also noted that although the issue of ACHD care was often perceived as a future problem, significant numbers of CHD patients have already reached their teens and are in need of healthcare services that are not currently being provided.

The majority of interviewees reported that, in part due to capacity, many pediatric CHD patients stop receiving care shortly after surgery, and that the barriers facing the provision of life-long care listed below, such as the shortage of trained staff, cost, and resource access, start in childhood. Because of Bulgaria's small size, all CHD surgeries are done at one center, and childhood access was reported as generally good. However, during a recent health system funding crisis, the patient group held public protests in order to prevent government plans to de-fund in-country CHD surgeries.

Identified Barriers to Implementation

Availability of A/CHD-trained staff: A fundamental barrier identified by health professionals interviewed was the absence of trained ACHD providers in their setting. None of the ACHD program directors interviewed had guideline-recommended training, although two had spent time studying at a formal ACHD program abroad, and the third had partnered with ACHD experts from Great Britain to provide on-going training to herself and her staff. As one cardiologist put it, *"In South Africa, there is no such thing as an ACHD specialist"*. In addition, none of the non-MD medical staff at the existing ACHD clinics, such as nurses, echocardiographers, and anesthesiologists, were reported to have ACHD training, although those able to treat patients in pediatric programs generally did so, thus ensuring CHD competence. Many interviewees noted that the shortage of pediatric cardiologists and other medical specialists in their country severely constrained their ability to care for CHD patients of all ages. Although Malaysia and Brazil reported significant challenges with CHD surgical capacity, access to general pediatric cardiology services was felt to be generally good. All ACHD doctors interviewed reported extreme demands on their time and additional clinical duties beyond their ACHD practice.

Divisions Between Pediatric and Adult Care Systems: In Bulgaria, Brazil, South Africa, and Mexico the government health system mandates that only children be cared for in pediatric hospitals, thus preventing ACHD patients access to CHD-trained care. In South Africa, transition to adult care is mandated by age 15 and often happens by age 8. The Bulgarian patient leader reported that some CHD cardiologists were able to provide after-hour unpaid care to ACHD patients, but that re-operations could only be done in adult cardiac centers by surgeons without CHD training and expertise.

Staffing Barriers: All of the cardiologists discussed ways in which staff shortages impacted their ability to deliver care, with several noting that physician "extenders" such as physicians assistants, nurse practitioners, and nurse educators did not exist in their country. Budget constraints on salary were also mentioned, with one cardiologist commenting that her department has no money for administrative help and therefore clinicians were responsible for their own scheduling.

***Availability of Cardiac Testing and Medication*:** All interviewed cardiologists stated that most recommended services, such as electrophysiology, echocardiography, and nuclear testing, were available in cities but not in all regions, and that securing such services often ranged from challenging to impossible. In India, Pakistan, Mexico, and Lebanon, there is a stark difference between availability in the private system, typically well-equipped, and the poorly equipped and staffed public system, with one Indian cardiologist noting that even her poorest patients hate the public hospital and will delay care as long as possible rather than return. Brazil and Malaysia's strong pediatric healthcare systems make services available in most areas, but long wait times could be a challenge, and in Brazil unnecessary hospitalization is sometimes needed to get quick access to cardiac testing. Almost all cardiac medications were reported as available, but scarcity and cost could make access difficult. In Pakistan a major pulmonary hypertension drug (Sildanefil) is banned because it can be used to treat erectile dysfunction under another brand name (Viagra).

Cost: All interviewees identified cost as a major barrier to care, with the extent of the impact varying based on the level of funding provided by the health system. The Indian, Pakistani and Malaysian cardiologists stated that they tailor their advice to match the economic realities of the family, and that both surgical costs and the long-term financial burden of caring for a medically complex child are considerations when deciding whether to recommend reparative surgery. The Pakistani ACHD director explained that decisions to recommend any new testing, medication, or intervention were guided by what the patient identified as his or her own needs, and that *"calm, comfort, and capability",* rather than guideline compliance, was his goal. Several described alternative testing strategies, such as close observation while walking rather than formal exercise testing, that save money and identify patient limitations.

In Brazil and South Africa, which offer free access to medical care, secondary costs such as travel and lost time from work are major barriers. The South African cardiologist noted that even when costs are relatively low, it was unrealistic to expect poor families to spend scarce resources to bring healthy children back for regular checkups. Several group leaders noted that even relatively affluent families struggle with the costs of CHD, and that many private insurance companies deny or only partially reimburse CHD-associated costs. The leaders from Lebanon, Pakistan, and Mexico, all of whom run foundations that fund surgery, reported that their bylaws restrict support to children, making provision of care to adults challenging.

Mismatch between Clinical Recommendations and Low-resource Settings: The planned focus of the interviews was care structure and access rather than clinical guidance, but the issue of the mismatch between the clinical recommendations and their practice needs was raised by the Pakistani, Indian, South African, and Malaysian cardiologists. As noted in the description of burden of disease, many of their patients have not had primary repair, and the existing guidelines were seen as offering minimal advice on decision-making in this context. In addition, guidance regarding Eisenmenger patients did not match their clinical experience of outcomes and impact, and several commented that some of the clinical recommendations were needlessly aggressive and wasteful of resources.

Overall Feasibility and Appropriateness

A number of the recommendations regarding educational and psychosocial issues were identified as culturally inappropriate or challenging by the majority of interviewees. As described below, there were major differences reported between countries, and one instance in which the group leader's perspective differed significantly from the providers. In addition, several health care providers noted that, given their current inability to provide clinical care to their patients, they could not yet address psychosocial issues.

Identified Barriers to Implementation

Education Regarding Need for Life-Long Care: All interviewees thought that explicit and on-going patient and family education regarding the need for life-long care was not only appropriate but essential, and that the absence of such education was a major barrier to follow up for both children and adults. The majority stated that many pediatric cardiologists lacked awareness of ACHD care needs and that some continued to perceive surgeries as *"one and done",* i.e. curative. Both the program manager and cardiologist in Brazil felt that most pediatricians in their area were aware of the need for life-long care, but that some may not be communicating it as clearly and frequently as needed. The Brazilian pediatric practice currently provides each family with written educational materials and a care binder summarizing the patient's medical history; none of the other interviewees reported using formal written materials or care plans.

In India, Pakistan, and South Africa, it was noted that the number of families seen per day severely limits the time available for any family communication, and this issue is made worse the absence of mid-level providers or nurses who provide patient education in high-income settings. Additional barriers mentioned included low levels of education, poor health literacy, language differences, and cultural norms regarding physician/patient interaction. Almost all interviewees stated that even when education was provided, denial on the part of patients and families also impacted decision making. The Indian public health official noted that in general, physicians in India tend not to educate about any health conditions, and those performing interventions do not see follow-up care as their responsibility.

In addition to expectations about physician/patient communication, other cultural norms were identified as impeding education regarding long term risks. Interviewees from Malaysia and Pakistan described a widespread discomfort among both families and physicians with discussing possible bad future outcomes because it is seen as making those events more likely to occur. Certain aspects of religious faith, such as a belief that *"only God can give health",* was also cited as barriers to both understanding and managing CHD health risks. One group leader noted instances where it appeared that the clinical team withheld information about the severity of disease based on the religion of the parent.

Transition to independent health decision making: All interviewees agreed that an effort should be made to educate patients fully about their condition and to create opportunities for individual conversations when possible, and that the patient's own needs and desires should be central in all

decision-making. However, the expectation that parents should have a minor role by early adulthood was identified as reflective of the European and North American settings in which the guidelines were written. In Brazil, Lebanon, Mexico, Malaysia, Pakistan, and India, it was noted that young people are generally expected to live with their parents until they marry, with one commenting that one is not considered an adult until marriage, and another joking, "*in India you only stop being a child when your parents die*". After marriage, many people continue to live in an extended family that actively participates in advising the patient. Several noted that young people are expected to ask for parental guidance in all major decisions, including those regarding health.

The emphasis on independence was also identified as not reflective of the economic realities that often face patients in their countries. Many commented that, given the absence of disability pensions and services, on-going family financial support and caretaking were essential to their patient's ability to stay healthy and productive in society, with one commenting that those who do not have such support generally will not live to adulthood. Several noted that extended families often help the patient on the long journey to the clinic, while others stay home with children. Strikingly, all the healthcare providers and many of the group leaders spoke of family support and involvement as a major positive force in their patient's lives which was central to their patient's mental and social health. One commented that she was concerned about her patients who came by themselves as it meant they were not receiving the love and care they needed to thrive.

Although patient group leaders identified many positive aspects to family involvement, several also talked about challenges that can arise. Parent leaders discussed the difficulties many have in balancing their child's independence with the desire to protect their health, while patients reported times in which they felt invisible or unable to talk freely when family attended visits. Some reported conflicts arising at the time of major medical decisions, making difficult situations more stressful. Several noted that, although family expectations in their country were changing, their education and affluence made it likely that their attitudes were not representative of the country as a whole.

Mental Health and Quality of Life: The majority of the interviewees stated that the mental health and quality of life issues listed in the ACHD guidelines were seen in their setting, with one noting that she sees them among the families as well as the patients. Both the Pakistani and Malaysian ACHD directors have published studies documenting these issues in their patients, and several group leaders reported significant struggles with depression or anxiety. The South African cardiologist stated her concern that anxiety and depression were being under-recognized in her patients due in part to the myth that, "*Africans don't get depressed",* and expressed the need to execute more research in this area. One participant noted that although she often saw her patients struggle with sadness and fear in response to difficult health situations, she did not see them as having more diagnosable mental health issues than other people.

Almost all interviewees reported insufficient mental health professionals available in their area, making the provision of recommended screening and treatment recommendations difficult, and the Indian public health official stated, *"In India, we still see mental health problems as madness".* However, she pointed to recent government medical education programs on mental health and felt that some progress was being made in this area. She also noted that due to advocacy by Indian HIV patient and

family groups, mental health needs in HIV were widely recognized, and suggested that the CHD community might build on this model. Several group leaders also reported significant stigma associated with mental health treatment, and one cardiologist noted that her patients did not like to be seen filling prescriptions for psychiatric medications because they come in different packaging than other medication.

***Birth Control, Pregnancy and Pre-Marital Counselling*:** Interviewees from Brazil, Mexico, Bulgaria, and South Africa reported no barriers to implementing ACHD guidelines regarding birth control and the need for planned pregnancy. Birth control and sex education was described as widely available and culturally accepted, and most teens and unmarried women are comfortable discussing their birth control needs. The South African cardiologist noted that, in response to her country's high rates of sexual assault, she initiates conversations as early as possible and encourages even those not sexually active to use birth control *"just in case something bad happens".* Several commented that in many communities, single motherhood is the norm.

The Pakistani, Malaysian, and Indian cardiologists all stated that it was essentially impossible to provide birth control education to unmarried teenagers in their country. Several commented that even mentioning the issue would be seen as an insult to the patient and her family, as it implied that she was engaging in immoral behavior. One noted that some of her colleagues were not comfortable discussing the topic and that conversations regarding this aspect of the ACHD program were quickly terminated. All addressed the need for family planning by advising a follow up visit at the time of marriage and stressed that these conversations require sensitivity and an on-going relationship with the family. The public health leader from India, although acknowledging the existing challenges, thought that new national initiatives promoting adolescent health might offer opportunities to connect teenage ACHD patients with needed information.

Gender expectations and arranged marriages in India and Pakistan were identified as major challenges for girls and women with CHD. In some communities, parents may decide not to opt for reparative surgery in childhood as there is a chance that the daughter would not be able to marry. If a girl has undergone surgery, a scar is often a major stigma. In some communities, having one "defective" child in the family can impact all the children's change of marriage. This leads to situations in which families do not to reveal the prospective bride's heart condition when the marriage is arranged. The cardiologists stressed the need to give clear advice on the risks of pregnancy and point out the inability to keep the scar a secret, and reported that many of their patients do marry and have children. However, one described a situation in which the bride was thrown out of the groom's house when the scar was discovered. One group leader also noted that having a heart condition is a major barrier to marriage for men as well. In India and Pakistan, the need for premarital counselling specifically addressing the risk of consanguity was also identified.

Social Stigma: Almost all interviewees reported social stigma associated with CHD, but the intensity and extent varied significantly between settings. As described above, group leaders and healthcare providers in India, Pakistan, and Malaysia described the ways that having a child with any health problems is seen as "shaming" the family. They noted that fear of other people knowing about one's heart condition was a major impediment to creating CHD patient or parent support groups. In South Africa and Malaysia,

group leaders also commented that adult patients tended to disappear from CHD-related social events, and that stigma and embarrassment likely was a factor.

Education and Employment: Challenges with education, such as the lack of understanding of the needs of CHD children, lack of access to needed educational supports, and discrimination on the part of teachers and fellow students were widely reported. Issues with career planning and employment were also widespread but varied considerably based on setting. In Brazil and in India, almost all employers require the submission of a certificate of health before hiring, making many ACHD patients unemployable in the regular job market. In countries which offer ACHD patients disability pensions and benefits, such as Brazil, South Africa, and Bulgaria, large numbers of ACHD patients opt for government support rather than seeking employment, in part due to high levels of unemployment in their countries. The cardiologists from Brazil and South Africa noted that many healthy patients with simple repaired defects request disability certification, and that the system implicitly discouraged career planning for their patients. The Bulgarian group leader noted that disability benefits in her country were excellent, in part because they provided partial support to people who work part-time. However, she also observed that the system was often used by people with no real disability, and that it sometimes discouraged people from working. In India, Pakistan, and Malaysia, the lack of social welfare benefits for CHD patients was noted, and several cardiologists described how hard even their sickest patients work to find jobs that allow them to support themselves and their families. The Pakistani cardiologist commented that when his ACHD patients die they often leave behind whole families who will now be destitute.

Exercise: All interviewees stated that, as in high-income settings, parents of CHD patients often discouraged exercise, and that many ACHD patients were inactive. None felt that this issue was being adequately addressed by pediatric cardiologists in their country, and several cardiologists mentioned that they have not incorporated advice about exercise into their own standard practice. The Indian public health official noted that this was a national problem in India, and that despite the obesity epidemic doctors generally do not promote exercise.

IDENTIFIED PRIORITIES FOR ACTION

All priorities for action identified by the interviewees are listed below. Identified priorities showed substantial overlap, but the top priorities varied largely based on role. Although acknowledging the need for training, research and funding, group leaders emphasized the need for awareness-building, whereas all cardiologists defined more ACHD training as the top need.

Awareness of the need for life-long care: This issue was mentioned by most interviewees but emphasized by the group leaders. Several incorporate this messaging in their media, but others stated that this issue is not currently addressed by their groups. The need for targeted education of pediatric cardiologists as well as parents was highlighted.

Patient education and empowerment: Group leaders emphasized the need to directly educate patients and families about the need for life long care. Several interviewees stressed the need to empower patients to take charge of their own health, and identified stigma as a major barrier to follow-up, with one patient leader stating, *"we need to be proud of who we are".*

Increase in ACHD training: All providers identified this issue as the foundation of ACHD care development. The creation of ACHD professional associations was seen as one strategy to promote the field as a discipline, as well as training partnerships with ACHD programs in high-income countries. Many group leaders also highlighted the need for more ACHD cardiologists, and one plans to fund an ACHD cardiology position at the hospital his organization supports. ACHD on-line and remote training were also suggested as strategies.

Initiate ACHD clinics within existing pediatric and adult cardiology centers: All the ACHD program directors currently use this strategy, with the location of services determined in part by rules regarding the care of adult patients in pediatric settings. A hub-and-spoke model was promoted, and the Pakistani ACHD program director reported providing remote ACHD care in rural areas.

Increase in ACHD research in middle-income settings: All interviewed cardiologists, and several group leaders, emphasized the need for more ACHD research in their settings. Many suggested that multi-center research networks and registries based on successful models in high-income countries should be initiated and might be promoted by a newly-founded ACHD professional organization.

Better definition of the prevalence of ACHD: Both healthcare providers and group leaders noted the importance of national registries of ACHD patients in order to both plan care and communicate needs to health care officials.

Creation of country/region-specific ACHD care guidelines: Most interviewed cardiologists expressed the need for ACHD clinical and management guidance appropriate for their settings and suggested these be created by ACHD professional organizations for those practicing in low-resource settings.

Government funding for ACHD care: All interviewees stressed the importance of advocating governments to cover CHD care across the life span. The Bulgarian, Pakistani, and Indian group are currently actively working with government on this issue.

Expansion of CHD charitable funding to cover ACHD care: Since many of the current efforts to develop CHD care in middle-income countries rely on charitable support, pushing to expand coverage for ACHD care was identified as a priority by several group leaders.

Better Incorporation of ACHD in existing government initiatives: Several interviewees stressed that existing government health initiatives, such as the development of adolescent and mental health services, could be leveraged to provide services for those with CHD.

Development of Low-cost and remote technology- Given the need for cardiac testing in remote areas, several interviewees stressed the need to develop low-cost echocardiography and other imaging technology. One interviewee is currently developing programs to use artificial intelligence to remotely diagnose CHD.

DISCUSSION

Major barriers impede care access for both CHD children and adults

Not only are ACHD-specific services and staffing unavailable in middle-income countries, but access to essential pediatric services and staffing continues to be challenging. All of the barriers to CHD care described above, such as insufficient trained staff, infrastructure, and funding, are common to all efforts to expand care access in low-resource settings. However, these challenges are heightened in CHD, which demands a high level of specialized expertise and access to cutting-edge technology. Even countries like Bulgaria, which have achieved universal childhood surgery and excellent national healthcare access, are not currently able to extend resources to ACHD care.

Different health systems create different barriers to care

The range of ways different health systems impact access to ACHD care is noteworthy. For example, in Brazil, Bulgaria, and South Africa, the same government health plans that provide publicly-funded health care create major barriers for ACHD care access, whereas in India and Pakistan, the lack of oversight has allowed more flexibility in where to provide ACHD services. Brazil, Mexico, and Malaysia have made substantial strides in access to childhood surgery, whereas others, like Pakistan, have made minimal progress on improving access nationally. These differences were clearly reflected in opinions on the need for special guidelines, with the cardiologists in Brazil, where most children now access early repair, seeing them as unnecessary, whereas cardiologists in India, Pakistan, Malaysia, and South Africa, defined their lack as a critical gap in care.

Cultural and societal factors impact ACHD care

Even more striking than the differences in health system impact were the ways in which cultural and societal factors affected practice. These factors were identified as barriers to providing education on the life-long care needs of CHD patients, as well as a major barrier to implementing recommendations regarding birth control and family planning.

Shared Priorities for Action

The high level of overlap in priorities demonstrates that the CHD healthcare providers and group leaders interviewed share a common vision of what is needed to improve life-long CHD care. The combination of shared goals and different areas of emphasis means that group leaders and cardiologists can focus on their areas of impact and expertise, while also pursuing collaborative efforts.

RECOMMENDATIONS

Focus on life-long care rather than transition

A major theme in the interviews was the disconnect between the “loss to care” efforts in high-income countries, which focus on adult care access, and the current inability for middle-income countries to provide on-going care for children. In high-income countries, pediatric cardiology was developed before ACHD care was needed, thus resulting in two parallel structures. Current efforts in care maintenance in high income countries largely focus on the point of hand-off and the provision of planned transition and

transfer. But research suggests that this focus does not address the root of the problem: large numbers of pediatric patients lost to follow-up as children. As more pediatric heart centers in middle-income countries begin providing follow-up care for post-operative children, the need to maintain this care can be stressed to families, patients, and local healthcare providers. Initiating a focus on care maintenance from childhood may have more impact than the limited improvements in follow-up seen thus far in many high-income settings.

Creation of ACHD guidelines for middle-income countries

As noted by many interviewees, the clinical needs of ACHD populations in middle-income countries often differ significantly from those in high-income countries. Developing country or region-specific guidelines will help improve outcomes in these patient populations. But in addition to clinical guidance, the interviews made clear that culturally-appropriate guidance on non-clinical aspects of care are also needed. Given the high risk of pregnancy-related mortality in this population, the inability to directly discuss birth control and family planning in some settings is a major gap in needed care. If new guidelines are created, specific guidance on strategies for promoting family planning should be prioritized. Similarly, the extensive comments on the high burden of social, emotional, and economic pressure on these patients, combined with the wide range of ways in which they manifest, demonstrates the need for region-specific psychosocial guidance. Finally, the interviews revealed a fundamental bias towards Western-style concepts of adulthood in current recommendations on transition. This bias is likely to limit their utility not only in middle-income countries, but also in minority communities in high-income settings that hold similar views about the role of extended families in health. In both high and middle-income settings, approaches to transition that acknowledge the many ways adulthood is defined should be developed.

Unified Action for Life-Long Care

As noted by all interviewees, not only do middle-income settings lack ACHD-specific training, research, and registries, but these essential healthcare components are also not available to children. All the ACHD priority actions noted above, including more training, research, patient empowerment, and advocacy, have been identified as essential to improving pediatric congenital care in middle-income countries.[102–105] As with the ACHD and pediatric care models, these priorities can be combined to take a life-long approach, summarized in Table 9.

Table 9: Advocacy Road Map for Life-long CHD Care		
Goal	Audience	Tactics
Awareness of CHD as a Life-Long Issue	General Public Local/National Government Surgical Missions/ Non-profits	Media Lobbying Direct Outreach Summits
Long-term engagement in Cardiac Care	Parents/patients	Patient and Family Education Patient Empowerment Social Events to Reduce Stigma
Awareness of life-long CHD Clinical Issues	Pediatric Cardiologists General Cardiologists Surgical Missions/Non-profits	Publications Conferences Direct outreach
Awareness of A/CHD as a medical specialty	Pediatric Cardiologists General Cardiologists	Creation of local/regional CHD Professional Organizations
More pediatric/adult CHD cardiologists	CHD Funders A/CHD Training Programs	in-country fellowship creation A/CHD training partnerships Fellowships for A/CHD training in low-resource settings
Improved clinical guidance	A/CHD Providers	Country/region specific A/CHD care guidelines
More CHD research	A/CHD Providers	Creation of research networks/registries
Private funding for life-long CHD Care	CHD Charities Surgical Missions/ Non-profits	Direct Outreach
Government funding for life-long CHD Care	Health Funding Authorities	Lobbying Direct Outreach
Inclusion of CHD in existing health programs	Public Health Officials	Lobbying Direct Outreach Partnerships
Inclusion of CHD in global health agenda	World Health Organization Global Health Advocacy Organizations	Global Health Event Engagement Lobbying Partnerships

Many of the ACHD priorities can be addressed by expanding existing efforts to promote pediatric cardiac care. For example, partnerships between high and middle-income surgical programs are common, and have been shown to be effective in supporting capacity building, expanding workforce, and improving quality. [106,107] Many partnering hospitals in high-income countries have established ACHD programs, whose staff could be included in these training efforts. The ACHD professional association, the International Society of Adult Congenital Cardiac Disease, has identified the expansion of ACHD care to low resource countries as an organizational priority,[68] and would be available to facilitate these efforts. Other priorities are currently in the development stage, such as creating a multi-institutional research registry and network of CHD research centers in middle-income countries. Unlike in high-income countries, where most registry efforts have focused on either ACHD or pediatric patients, these registries could enroll all ages of CHD patients from their inception.

Strengthening Partnerships between Providers and Patient Organizations

Finally, what is evident in the interviews is the high level of engagement and collaboration that is already occurring between CHD healthcare providers and CHD patient groups in these settings. Unlike in high-income countries, where groups tend to focus primarily on peer support and education, these groups play a major role in funding surgery, facilitating care access, and advocating for the better provision of services and healthcare coverage for those with CHD. By strengthening these partnerships and expanding their efforts to include a life-course approach, they will create a better future for all those affected by CHD.

CONCLUSIONS

The goal of increasing access to pediatric cardiac services is not only to improve childhood survival; it is to create a healthy future for every child born with heart defects. By recognizing the long-term needs of this population now, the foundation can be laid for life-long care. These efforts do not need to start with separate, adult-specific initiatives, but can be built into current efforts to create sustainable pediatric cardiology services. As outlined above, multiple opportunities exist to include ACHD training and planning in current activities aimed at improving global CHD care. The focus on long-term care maintenance should begin in childhood, thus preventing early loss to follow-up. It can also take advantage of telemedicine and portable testing technology, and be incorporated within not just tertiary care, but also at the primary and secondary care level. These care models can provide new strategies to prevent the widespread care gaps still seen throughout high-income countries. If these efforts are successful, today's children will benefit from the improved health and well-being that comes from high-quality CHD care across the life-span.

In 2016, 193 countries committed to two Sustainable Development Goals directly relevant to CHD: the end to preventable infant mortality, and a 1/3 reduction of deaths from noncommunicable disease. As communicable disease continues to drop, sustained reductions in infant mortality will depend on the prompt diagnosis and treatment of CHD. This will result in a steady increase of those living with an emerging noncommunicable disease: congenital heart disease. The challenges of providing for this population are an extreme version of those found in all chronic disease. Regular access to care must be maintained from birth and continue when patients transition from pediatric to adult care. Continued good health relies not just on the availability and access to cardiac testing and medications, but also to tertiary level cardiac services even more specialized than those needed in acquired heart disease. Most importantly, this care must be of the quality needed to keep even those with complex conditions thriving. This need for high-quality care is essential for improving the health of all people living in low-resource settings. In *"High Quality Health Systems in the Sustainable Development Goals Era: Time for a Revolution",* Kruk argues that only through improving quality will current goals for health improvements be reached, and states,

> "*Quality should not be the purview of the elite or an aspiration for a far distant future; it should be in the DNA of all health systems. Furthermore, the human right to health is meaningless without good quality care because health systems cannot improve health without it."*[108]

Too often, governments and health policy experts identify congenital heart disease as a priority for *"a far distant future".* But by taking on this challenge, they will both improve their chances of meeting their sustainable development goals and honor these patient's basic human rights.

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APPENDIX 2: ACRONYMS

ACHD – Adult Congenital Heart Disease
CHD – Congenital Heart Disease
SDI – Sustainable Development Index