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Development of an international standard set of clinical and patient-reported outcomes for children and adults with congenital heart disease: a report from the International Consortium for Health Outcomes Measurement Congenital Heart Disease Working Group

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Aims Congenital heart disease (CHD) is the most common congenital malformation. Despite the worldwide burden to patient wellbeing and health system resource utilization, tracking of long-term outcomes is lacking, limiting the delivery and measurement of high-value care. To begin transitioning to value-based healthcare in CHD, the International Consortium for Health Outcomes Measurement aligned an international collaborative of CHD experts, patient representatives, and other stakeholders to construct a standard set of outcomes and risk-adjustment variables that are meaningful to patients.

Methods and results The primary aim was to identify a minimum standard set of outcomes to be used by health systems worldwide. The methodological process included four key steps: (i) develop a working group representative of all CHD stakeholders; (ii) conduct extensive literature reviews to identify scope, outcomes of interest, tools used to measure outcomes, and case-mix adjustment variables; (iii) create the outcome set using a series of multi-round Delphi processes; and (iv) disseminate set worldwide. The Working Group established a 15-item outcome set, incorporating physical, mental, social, and overall health outcomes accompanied by tools for measurement and case-mix adjustment variables. Patients with any CHD diagnoses of all ages are included. Following an open review process, over 80% of patients and providers surveyed agreed with the set in its final form.

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Conclusion This is the first international development of a stakeholder-informed standard set of outcomes for CHD. It can serve as a first step for a lifespan outcomes measurement approach to guide benchmarking and improvement among health systems.

Graphical Abstract

Keywords Outcomes • Congenital heart disease • Patient-reported outcomes

Introduction

Congenital heart disease (CHD) is the most common category of congenital malformations, with a prevalence of nine per 1000 live births worldwide.[1–3] As a condition, CHD is a heterogeneous set of hundreds of unique diagnoses, ranging from relatively minor defects not requiring intervention, to complex CHD requiring several surgical interventions in the first days to year of life to insure survival.[4] With rapid advancements in diagnosis and surgical and catheter-based interventions, there has been significant increased early and late survival of patients with CHD.[5,6]

Despite improvement in survival rates, the worldwide burden of CHD morbidities remains high, with significant variation in care before and after surgical repair, particularly in low-middle income countries (LMICs).[7–11] Once patients become adults, the care varies even more widely, depending in part on the geographic availability of adult healthcare providers.[8] The global burden of resource utilization from acute care for CHD is estimated to be 6 billion dollars annually, notwithstanding the costs of significant chronic care and lost employment from patients and family members.[9,12–15]

As centres may lack sufficient volume to effectively compare outcomes, CHD registries have increased in availability over the past decade, primarily tracking mortality, surgical, and immediate post-surgical complications.[13,16–18] Despite these advances, long-term outcomes and patient-reported outcomes remain tracked in only a minority of cases and measurement of success remain in siloes, resulting in inconsistent definitions for equivalent outcomes.[19]

This has resulted in unclear value of the care delivery of CHD longitudinally. A condition such as CHD therefore will benefit from a standard set of core outcomes that are meaningful to patients and care systems to better provide high-value care. As an effort to streamline global outcome measures, the International Consortium for Health Outcomes Measurement (ICHOM) formed a CHD working group. ICHOM was founded as a not-for-profit organization in 2012 to promote comprehensive standardized outcome measurement and align outcome measurement efforts globally.[20] Over 600 organizations have implemented ICHOM sets, which are reviewed annually by ICHOM. The CHD Working Group (WG) aimed to develop a multi-stakeholder informed standard set of outcomes for CHD, focusing on patient long-term perspectives. The process to develop the standard set is the first step to fulfil a key tenant of value-based healthcare: to deliver patient-centred comprehensive health outcomes throughout a lifetime.

The primary aim was to identify a stakeholder-informed minimum standard set of outcomes for patients with CHD to be used by health systems worldwide. The goals of the standard set were to enable measurement in routine clinical practice, strive to improve decision-making between providers and patients, and to facilitate quality improvement (QI). The outcomes specifically embodied metrics familiar to current registries (i.e. mortality), as well as patient-reported health status [i.e. burden of disease, health-related quality of life (hrQOL)]. Importantly, collecting data for the standard set will not exclude any other registry data collection. The secondary aim of this initiative was to develop benchmarking opportunities across hospitals and countries (creating the ability to compare outcomes between systems).

Methods

The methodological process included four key steps, which have previously been outlined and utilized to create effective standard sets of diverse conditions: (i) develop a working group representative of all CHD stakeholders, with key focus on patient representation from LMICs; (ii) conduct extensive scoping reviews of international CHD classifications to identify populations for inclusions in the standard set, outcomes currently sought and tracked, and current tools used to measure outcomes; (iii) create a minimum set of standard outcomes for CHD using a worldwide stakeholder multi-round Delphi process; and (iv) disseminate the CHD standard set worldwide. Importantly, each of the four steps above integrated patient representation and involvement and operated to conclude the set in a timely manner of less than 18 months from start to conclusion, beginning in July of 2018.

Composition of working group

ICHOM incorporated an international assembly of clinical and healthcare leaders of CHD to work with patients to create the set, including expertise in clinical registries, hospital administration, patient-centred outcomes research, psychology, QI, and patient advocacy (*Figure 1*). Specifically, over half of the providers care for adults with CHD and multiple nurses in the working group have specialization in CHD quality. Patient advocates were essential to decision-making at every step, and included patients and parents of patients with paediatric and adult CHD. Of the working group members present for every decision-making session, 20% of the working group were patients with CHD or parents of patients with CHD, and represented the largest advocacy groups worldwide for CHD. Patient advocacy groups of many of the largest CHD advocacy groups worldwide were included, incorporating representatives of patient focus groups specifically providing input to patient outcomes of interest. This process included a patient focus group with all of the patient representatives based on open questions to elicit outcomes that may not have come from the literature. Any additional outcomes from the patient focus group were added to be discussed and any outcomes that came out of the focus group were highlighted specifically to the working group as being important to patients and families.

Search strategy and outcome and case-mix variable selection criteria

There were four separate literature reviews stemming from the initial scoping review, performed in accordance with Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA): (i) Scoping review, (ii) Outcomes review, (iii) Tool review, and (iv) Case-mix adjusted variable review (variables that allow different groups to more easily be compared). Each review was conducted via Google Scholar, PubMed, and Medline with the following medical subject headings (MeSH): CHD, adult CHD outcomes, outcomes, patient-reported outcomes, and psychometric analysis. Additional articles were supplemented to the lists based on recommendations from experts in respective fields. Registries and databases for inclusion were selected based on results of the review and with aid of expert opinion from the WG (Supplementary material online, *Table S1*).

Review process of outcome measures and measurement tools

The WG completed three Delphi rounds to build majority consensus of population scope, measurement tools, risk factors for case-mix adjustment, and 15 outcomes. These variables were discussed over a combination of nine teleconferences and surveys to forge consensus. The

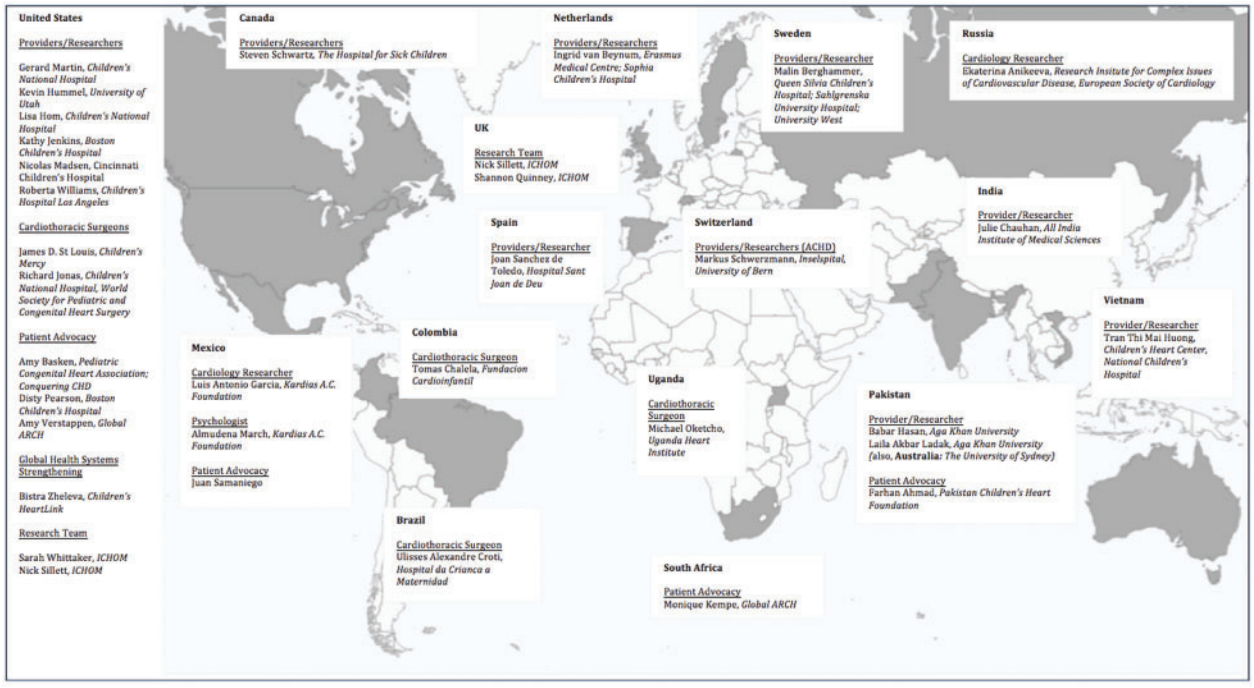

Figure 1 Working group composition. The world map displays working group members by (i) country of work, (ii) type of contributor (cardiology provider or researcher, cardiothoracic surgeon, patient advocacy/representation, or research team member), and organization or institution that the member is affiliated.

modified Delphi process used involved three rounds in which items were rated on a 9-point Likert scale according to importance. If outcomes were not voted with >80% agreement as essential or non-essential, items were re-discussed and revised if necessary prior to a second round of voting, and eventually a final round of 'Yes/No' voting requiring >70% agreement for inclusion. At completion of the final set, all members of the WG voted 'Yes/No' whether they approved the final standard set, voting unanimously in approval.

The WG selected outcomes based on 4 criteria: (i) the frequency of the outcome; (ii) its impact on the patient and/or their family; (iii) the potential to modify the outcome; and (iv) the feasibility of 'capturing' the outcome in clinical practice. Additional criteria for patient-reported outcome measures (PROMs) included (i) the domain coverage; (ii) the psychometric properties; (iii) the feasibility to implement; and (iv) the clinical interpretability. Next, time points for data collection were selected for each outcome. Risk-adjustment variables were selected based on three criteria: (i) the relevance (strength of the causal linkage between the risk factor and the outcome), (ii) the risk factor independence, and (iii) feasibility of measurement.

Open review

Following WG consensus, there was a period of endorsement and open review by patients and professionals. This process spanned >2 months with a priori recognition that >70% agreement for aspects of the survey is consistent with support for the standard set. The validation and open review aimed to reach feedback from all six continents involved in the standard set, and include LMIC and high-income countries.

Results

Scoping review

The scoping review was informed by a literature review from PubMed, Medline, and Google Scholar, resulting in 2340 articles identified and 216 articles fully analysed for population inclusion and exclusion in accordance with PRISMA (*Figure 2*). The WG defined the scope of the population for the standard set as any patient with CHD, regardless of severity, with all ages included, including children and adults with CHD. Diagnoses were based on International Nomenclature Working Group definitions.[21] The scope of population spanned the lifetime, with many long-term outcomes appreciated into adulthood despite childhood surgical repair. Additionally, it was recognized that in LMICs, patients often present late for diagnosis, at times into adulthood, which precludes the opportunity for repair in childhood.[22]

Outcome domains

The CHD working group reviewed the outcome domains and definitions from 42 registries (Supplementary material online, *Table S1*), three expert-panel selected guidelines (American Heart Association/American College of Cardiology Adult CHD Guideline, American College of Cardiology/American Academy of Pediatrics Policy Statement for Care of Children with CHD, and European Society of Cardiology Guideline for management of Grown Up CHD), and a literature review and analysis of 571 articles.[23–25]

Outcome classifications were defined as physical functioning, social functioning, mental functioning, and overall health

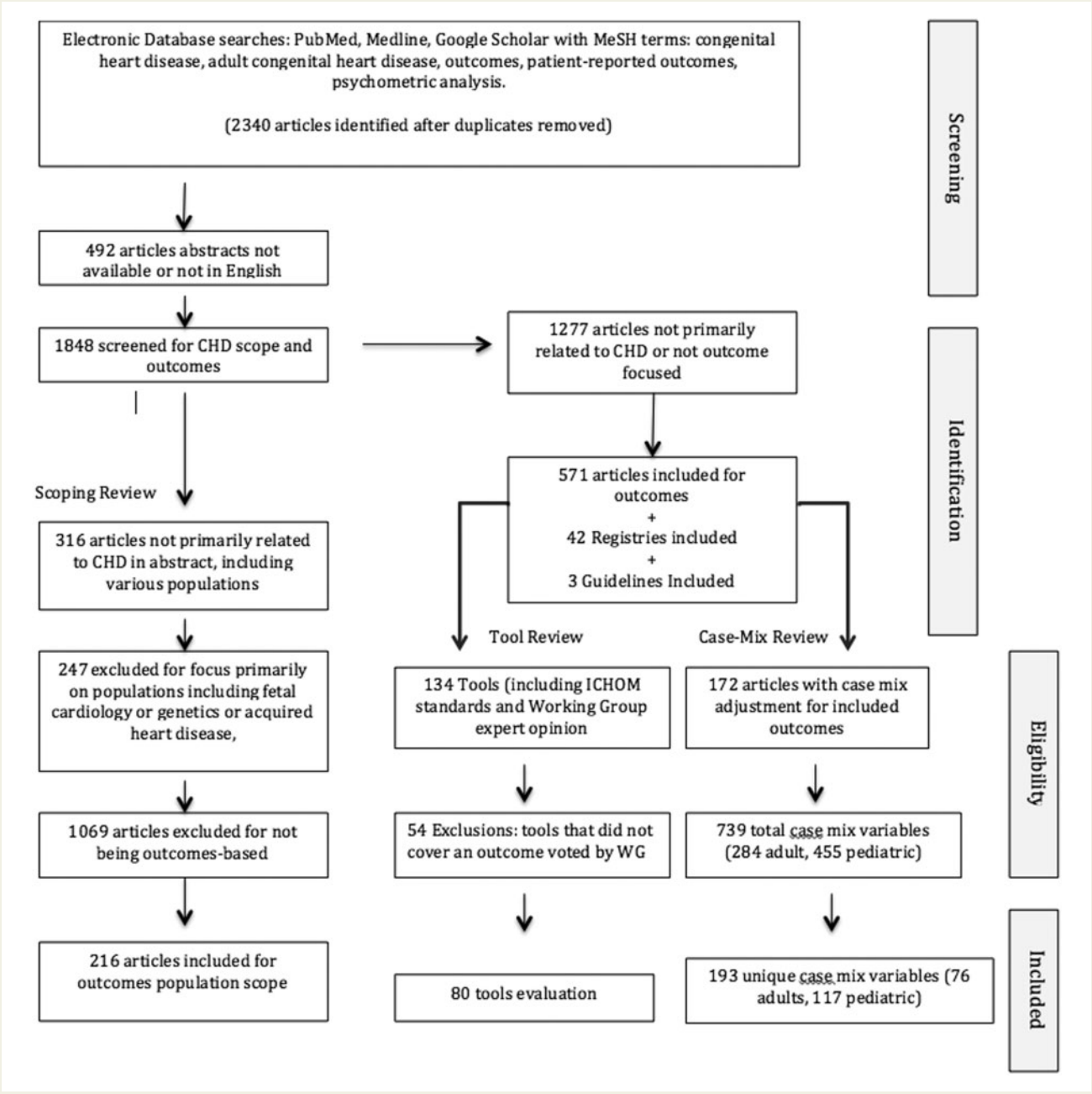

Figure 2 Literature review. There were four separate literature reviews stemming from the initial scoping review, completed in accordance to PRISMA guidelines: (i) Scoping review to identify the population included in the set, (ii) Outcomes review to determine available outcomes in CHD literature, (iii) Tool review to identify all available tools to measure the determined outcomes, and (iv) Case-Mix review to identify all risk factors for variable outcomes of CHD. Each review was conducted via Google Scholar, PubMed, and Medline and supplemented with article recommendations from experts in respective fields.

outcomes; they were stratified by current health state, effect modifiers, and future health state, and by clinical or patient-reported outcomes and the majority included both adult and paediatric ages (*Table 1*).

Measurement tool evaluation

Each outcome was assessed based on availability of validated tools to measure respective outcomes. Both clinical metrics and PROMs were assessed. In total, 80 tools were evaluated from the initial literature review, as well as from key international health surveys, and expert suggestions. Recommendations were made for the implementation and timeline for measuring each outcome based on psychometric properties of tools (*Table 2*). The timeline for collecting the data is recommended to follow the generic recommendations for clinic visits for patients with CHD or adult CHD.[26]

Table 1 Standard set of congenital heart disease outcomes and measures

Category	Outcome	Data source	Measurement Tool	Age Range	Timing
Physical functioning	Survival	Administrative/clinical	Indicate if the patient has died	All ages	Initial, annually
	Heart failure	Administrative/clinical	Indicate the date of death	All ages	Initial, annually
	Activity level and exercise	Administrative/clinical	Indicate if the death is related to CHD	All ages	Initial, annually
	Arrhythmias	Administrative/clinical	Indicate the cause of death	All ages	Initial, annually
	Pregnancy	PROM	Indicate if the death was within 30 days of a procedure for CHD or in hospital following procedure for CHD	<18 years	Initial, annually
	Motor developmental delay	PROM		≥18 years	Initial, annually
	Growth	Clinical		≥6 years	Initial, annually
		Administrative/clinical		All ages	Initial, annually
		PROM	Ross classification for heart failure in children	≥18 years	Initial, annually
		PROM		<18 years	Initial, annually
		Administrative/clinical	New York Heart Association functional classification	<18 years	At 6 months, annually 1–5 years, 10 years, 18 years
			6-min walk test		
			Presence of arrhythmias		
			Have you ever been pregnant? Did you receive clinical counselling or care from a cardiologist during pregnancy? How many pregnancies have you had (and live births)? If you have had a termination, were you medically advised to?		
			National Survey of Children's Health		
			WHO Growth Charts		
Social functioning	Productivity	PROM	Number of school days missed in the past year due to CHD	<18 years	Initial, annually
	Financial Burden	PROM		≥18 years	Initial, annually
		PROM	Work Productivity and Impairment Questionnaire	All ages	Initial, annually
			Is your CHD causing a financial burden to you? Are you having difficulty accessing the care you need for your CHD because of financial burden?		
Mental functioning	Cognition	PROM	No tool included due to lack of a free, feasible measure	≥18 years	Initial, annually
	Behavioural/emotional development	PROM		<18 years	Initial, annually
		PROM	National Survey of Children's Health	≥18 years	Initial, annually
	Depression			≥18 years	
	Anxiety		Patient Health Questionnaire-9		
			Generalized Anxiety Disorder-7		
Overall health status	Quality of Life and Perceived Health Status	PROM	Satisfaction with Life Scale—Children	9–14 years	Initial, annually
		PROM		≥15 years	Initial, annually
		PROM	Satisfaction with Life Scale	≥18 years	Initial, annually
	Family quality of life	PROM	PROMIS Scale v1.2—Global Health	8–18 years	Initial, annually
			Pediatric Cardiac Quality of Life Inventory		

CHD: congenital heart disease; QHO: World Health Organization.

Table 2 Tool criteria for inclusion and psychometric properties assessed

Tool evaluation criteria		Strongly supported	Weakly supported	Not supported
Validity	Sensitivity to change	Clear evidence	Weak evidence	No evidence found
	Content validity	Clear evidence	Weak evidence	No evidence found
	Construct validity	Clear evidence	Weak evidence	No evidence found
Reliability	Test–retest reliability	$r \geq 0.7$	$r < 0.7$	No evidence found
	Internal consistency	$\alpha \geq 0.7$	$\alpha < 0.7$	No evidence found
Translation	Number of translations	>10	2 to 10	1
Interpretability	Scoring: non-proprietary scoring available	Yes	No	No evidence found
	Recall period			
	Availability of clinical cut-offs, normative data	Clear evidence	Weak evidence	No evidence found
Cost	Fee for routine clinical use	No	Yes	No evidence found
Burden	Patient burden (question number)	<11	11 to 29	>29
Reporter	Self, parent, clinician			
Age group coverage	Age range of tool			
Tool scores	Scores produced by the tool			

Overall health outcomes

Patient quality of life (QOL) is a broad multidimensional concept assessing both positive and negative aspects of life, whereas perceived health status indicates individual view of their own health. Tools to measure overall QOL are the Satisfaction with Life Scale-Child for paediatrics, and the Satisfaction with Life Scale plus the PROMIS Scale Global Health (v1.2) for adults. For hrQOL, we recommend the Pediatric Cardiac Quality of Life Inventory (PCQLI), a disease-specific hrQOL instrument, which was designed for clinical and research at Lurie Children's Hospital and is feasible to implement.[27] The PCQLI is free-for-use and short in length, contrasting the cost and length of the PedsQL-Cardiac Module, a widely used research assessment.[28] For adult hrQOL, the WG recommends the generic PROMIS Scale v1.2-Global Health. There is no recommendation for a cardiac-specific measure because there are no tools created for clinical use, with only one adult-specific hrQOL identified, which is used exclusively in research settings.[29]

Physical health outcomes

Survival

The WG agreed unanimously to assess survival of all patients at every encounter with patients with CHD in the cohort. Categorization of cause of death, both attributable to CHD and not related to CHD, was based on an extensive review of the late causes of death after paediatric cardiac surgery over the past 50 years.[5]

Growth

Consistent with other ICHOM standard sets, the World Health Organization (WHO) Growth Charts were selected as the most appropriate metric to assess paediatric growth with CHD. There were not condition-specific growth metrics identified. The WG suggests a timeline with frequent measurement over the first 3 years of life, since significant early child development occurs in the first 1000 days of life (*Table 1*).[30]

Activity level

Both the expert panel of providers and patient representatives agreed on the importance of clinical and patient-reported activity level as an outcome. The 6-min walk test, valid in both paediatric and adult CHD, can freely and effectively be measured in the spectrum of healthcare settings and was voted for inclusion by the WG. Additional measurements will overlap with heart failure measures and through cardiac and generic HRQoL measures; specifically the PCQLI is not activity-specific, but incorporates assessment of capacity within the overall measure.

Heart failure

Patients with CHD of all ages are to be included for measurement of heart failure both with clinical measures and PROMs At all encounters, a clinical assessment will indicate the presence of heart failure based on clinical exam or findings in ancillary studies.[31] Adult patients, on an annual basis, will complete the New York Heart Association (NYHA) Classification for heart failure, with levels I-IV indicating varying severity of symptoms the patient experiences for heart failure.[32] Similarly, the Ross Classification for Heart Failure was selected as a PROM measurement for paediatric patients with CHD, mirroring the NYHA classifications for heart failure.[33]

Arrhythmias

Of complications that were considered life altering and significant enough to include in the standard set, arrhythmias was selected from the review as an outcome for patients of all ages. Assessment includes clinical review of whether the patient had the occurrence of an arrhythmia, including those who had undergone successful treatment. Additionally, sudden cardiac arrest, as an adverse event to assess the burden of arrhythmias, was recommended.[34,35]

Motor development (paediatric)

Twenty-one tools were analysed for assessing developmental delay in paediatrics within the CHD population. The National Survey of Children's Health (NSCH), while not used specifically in CHD, directs families to indicate if they are aware of their children having a co-morbid developmental delay that has been ever diagnosed or discussed with them by a care provider.[36] A subset of questions from the NSCH was included in the standard set for assessing developmental delay. Many of the other tools assessed require a high cost or the length of completion (60–90 min) were not considered feasible.

Pregnancy

The ability for a patient with CHD to become pregnant and have a child was acknowledged by the WG's patient representatives as one of the first questions from parents of children with CHD. While the review identified tools to assess the ability of a pregnant woman to deliver a child, there is no clear mechanism for measuring the number of patients with a desire to become pregnant and have a child that are able to successfully do so. Because of the weight the WG attributed to pregnancy as an outcome in ACHD, the group selected a series of questions that address the outcome (*Table 1*).

Social health outcomes

Productivity

Productivity in daily life and work was included for both children and adults, but the review indicated measurement differs between populations. For paediatrics, productivity is recommended to be measured by cardiac and generic hrQOL measures and by the number of school days missed in the past year because of CHD, following the WHO guidelines for school absenteeism.[37] For adults, as a corollary to children school absenteeism, the WG recommends the Work Productivity and Activity Impairment Questionnaire, which scores work absenteeism and presenteeism, work productivity loss, and activity impairment impacting work production.[38]

Family QOL (paediatric)

The burden of a chronic, high resource utilization condition on family members was appreciated and recognized to be an important outcome. In particular, in the families of paediatric patients (primarily indicating parents), there was deemed a need to measure family QOL longitudinally. While some broad QOL tools address family QOL peripherally, these tools were excluded for other reasons such as cost, length, or poor psychometric properties. There were no tools that adequately measure family QOL in a feasible way and thus none included in the standard set.

Financial burden

Financial burden, the distress felt by the patient or family of the patient attributable to financial concerns, either in the form of healthcare costs or loss of wages, was considered a core outcome for adults with CHD. However, there are no tools to assess financial burden that can be applied internationally. With the importance of financial burden recognized, the WG chose to ask families of paediatric and adult patients if CHD is causing a financial burden impacting care access.

Mental health outcomes

Behavioural/emotional development (paediatric)

Behavioural/emotional development was identified as a core outcome of significance in CHD, but lacks a clear tool to track over time, despite widespread literature on the topic. To gauge family awareness of behavioural/emotional developmental delay, the WG selected questions from the NSCH to identify patients with delays and track longitudinally. Twenty-three tools were assessed for psychometrics, many of which are widely used in the research (Peabody Developmental Motor Scales, Bayley Scales of Infant Development) and clinical (Child Behavior Checklist) settings.[39] However, all these above tools were excluded due to limitations to feasibly apply internationally, due to cost, length, burden of tool completion, or availability of tools.

Depression and anxiety (adult)

Mental health disease including depression and anxiety were strongly voted for inclusion in the standard set for measurement in adults. The PHQ-9 and GAD-7 were determined to be adequate for measurement of depression and anxiety, respectively, which reflects the determination by the ICHOM Depression and Anxiety Standard Set.[40,41] As with cognition, these outcomes were not elected in paediatrics, with agreement that longitudinal measurement would be adequately completed with adult measurement.

Cognition

There are limited assessments validated for assessment of cognition in adults with CHD on a regular basis. The most widely used tools for adults, including the Wide range Achievement Test, have not been used in CHD greater than age 12, and other tools with high psychometrics require high cost or technology burden. Cognition measurement in paediatrics did not meet criteria by the WG for inclusion based on the review. However, measures of cognition in paediatrics overlap heavily with development (behavioural/emotional and physical) as well as assessment as adults.

Case-mix adjustment

For both clinical and patient-reported outcomes, the WG defined minimum risk factors as variables for case-mix adjustments. Risk factors were stratified by those affecting all patients with CHD as well as variables specific to populations. Informing the risk factor selections was a review and inclusion of common validated risk models in use for severity of illness and prognosis. Inclusion in the standard set was based on relevance (predictor of included outcomes), practicality of measurement, and internationally comparable (Supplementary material online, *Table S2*).

External endorsement

For publication of the standard set for implementation and benchmarking, a consumer and professional endorsement survey was distributed worldwide to assess for gaps from various stakeholder perspectives. The review included 352 patient reviews with feedback, including >50 responses from patients in North America, Europe, and Asia in countries of varying income statuses; 61% of respondents were adults with CHD, 42% were parents of children with CHD. Additionally, 69 professionals from six continents reviewed the

standard set; the majority were healthcare professionals including primarily nurses and physicians, with responses also from researchers, advocacy professionals, and healthcare administrators. There was >70% agreement with every aspect of the patient survey, except for only a 62% agreement to whether 'all children's outcomes that were important to patients were captured'. The majority of the disagreement for this question stemmed from the LMICs population suggesting financial burden to be included as a paediatric outcome; as a result, the working group agreed with inclusion of financial burden in paediatric and adult groups. For the professional review, there was over 70% in every aspect except that only 68% suggested the set was missing a focus on social health in paediatric patients, with desire for more focus on family QOL.

Discussion

Following extensive reviews, a standard set of outcomes that matter to patients was created. The primary accomplishments included reviewing and outlining the population scope, determining a core set of outcomes, identifying and psychometrically assessing tools to measure each respective outcome, and developing case-mix adjustment variables. Within this process, following conclusions were identified: (i) CHD is a life-long disease and outcomes must be measured longitudinally to include ACHD, not exclusively CHD in paediatrics, (ii) the majority of outcomes that matter are predominantly functional outcomes, and (iii) despite broad recognition of the importance of neurodevelopmental and cognitive outcomes, there is currently a lack of a widely available, low-burden tools to assess such outcomes. With collaboration from patient and family representation, this is the first international effort to recommend a standard set of outcomes that is meaningful to patients and can serve as a framework for measuring CHD success worldwide.

Transforming health systems to value-based healthcare requires transparent data comparison among systems and countries.[42] To this point, numerous registries, standardized datasets, and QI collaborations aiming to standardize CHD management remain primarily in siloes studying survival. While these efforts have made strides in improving care, these outcomes do not effectively target the care over the lifetime of patients with CHD and are limited in the integration of patient voices. The outcomes identified in this review fill this gap, elucidating specific outcomes that would not have been included in the standard set without significant input from patients with CHD, while adding to the foundation of research established by current strategies. Using outcomes that are driven and reported by patients will allow care to be designed and systems to adapt to the need of patients with CHD worldwide.

The scope includes patients of all ages, specifying measurement of outcomes to paediatrics or adults depending on tools being utilized for the metrics. While initially conceptualized as a paediatric standard set, the patient and family representatives suggested early in the process that CHD is a lifelong disease, and thus outcomes must be assessed longitudinally throughout life. Owing to the primary objective of creating outcomes that matter to patients, the set was restructured to target lifelong outcomes of CHD. Additionally, this perspective is consistent with guidance from the AHA, the AAP, the European Society of Cardiology (ESC), and the ACC.[43–45] When assessing diagnoses by severity levels described by the AHA/ACC and World Bank, the simplest lesions were considered for exclusion because of their relatively low impact (e.g. small left to right shunts with no other lesions).[46] In LMICs, however, there is known to be a higher rate of delayed diagnosis and thus more extensive pathology from relatively simple lesions.[47] It was ultimately concluded that all lesions of severity be included in the standard set.

Physical functioning

Physical functional outcomes are the most commonly measured outcomes today. Survival was unanimously included and is still a benchmark for success in CHD. Growth, measured using the WHO growth curves, is a key driver of successful CHD development. Nutrition was discussed at length by the working group as an outcome of its own, but without clear evidence for effectively measuring nutrition, growth was selected to be a proxy for nutrition.[48] Arrhythmias in CHD are both a complication and outcome of care. This was decided based on patient representatives' strong views that the fear of stress of living with arrhythmias impacts daily functioning, and that arrhythmias serve as a common cause of death in ACHD.[49] While symptom burden is not traditionally viewed as an outcome in itself (but rather its effect on QOL), patients living with the burden or fear of arrhythmias indicated that to truly target outcomes on the minds of patients, arrhythmias must be targeted in the ACHD population to be an inclusive standard set. Immediate post-procedural measures (i.e. complications, length of stay), which make up the bulk of current registries in CHD, are incorporated heavily in the set by case-mix adjusting for these variables (Supplementary material online, *Table S2*).

PROMs and QOL measures

The majority of outcomes draw heavily on PROMs to strategically take an inclusive stance on the patient voice in outcome measurement.[50] The use of PROMs is increasingly recognized as a standard in assessing patient outcomes and in the process of shared decision-making with families, and has been advocated by the AHA among other cardiology societies for both paediatrics and adults.[51,52] In the CHD population, specifically the international ACHD population, PROMs have been evaluated at length in the APPROACH-IS.[53] All tools used in the APPROACH-IS were analysed for inclusion, but only the Satisfaction with Life Scale was included, due to costs or alternative tools that were deemed more directly to align with outcomes of interest to patients and families. In order to reduce barriers to implementation, all PROMs selected for inclusion were under the pretense that they were of no cost to institutions, were in the shortest completion time quartile of all tools assessed, did not depend on independent administrators outside of clinic staff or patients, and are available in many languages.

Currently, many single centre or small scale studies use industry-developed tools such as the Pediatric Quality of Life Inventory (PedsQL), which provides robust, CHD-specific data for tracking outcomes that matter to families.[28] The PedsQL, like many QOL tools, come at high direct and indirect costs. The fee-for-use, length, and need of a proctor of such tools inhibits the use by robust studies, in particular in LMICs. We recognize the cost of developing and validating such tools is high, but to effectively measure meaningful outcomes consistently in CHD, there is a need for leaner tools or

reimbursement strategies encouraging such measurement, which have began to develop in the ACHD population.[54] The low cost and burden of the tools in this standard set both cover the spectrum of CHD outcomes and offer LMICs an opportunity to accelerate their QI infrastructure.

Outcomes lacking feasible measurement tools

There remain outcomes of utmost importance to patients that do not have feasible measurement tools (*Table 3*). Obstetric care in adults with CHD patients is a growing field, and pregnancy is recognized as a primary meaningful outcome across life.[55] However, there is no consistency in measuring or epidemiologically outlining what successful pregnancy entails, though the Registry of Pregnancy And Cardiac Disease provided direction to the WG.[56] The WG included self-selected questions to assess pregnancy as an outcome measurement, which was based on Delphi voting from the expert panel, without inclusion of a validated tool. Difficulty measuring financial burden is not a CHD-specific problem, but with CHD representing a high resource utilization and lifelong disease, there is a need for improved measurement of this outcome. Additionally, family QOL, despite demand from the professional open review, lacks a clear tool applicable as an international measure. For the questions added for pregnancy and financial burden, significant research was undertaken including a review of the literature search, consultation to adult CHD pregnancy experts, and continued discussion with patient family focus groups and representatives of patient advocacy groups as to what information was important to them to be asked. The authors are in agreement that tools with psychometric validity are ideal gold, but that the limitations in availability of such tools should not inhibit data collection of these important outcomes.

Neurodevelopmental outcomes (NDO), including emotional and behavioural development in paediatrics and cognition in adults, is among the most intensely studied aspects of CHD over the past decade, and effort has been undertaken to describe and create guidelines for the recognition of NDO.[26,27,57] However, the ability to implement feasible, longitudinal measurement is lacking. Selected questions from the NSCH offers a surrogate measure of NDO, recognizing that the tool is not CHD-specific, nor tailored uniquely to measuring outcomes. A key finding from this process is the need for tools to effectively measure NDO that can be widely applied.

Set strengths and implementation challenges

A particular strength of this review includes the unique breadth of insight of the CHD WG. Patient and family representatives involved have extensive experience developing CHD guidelines and advocating on behalf of CHD patients worldwide. International physician leaders added specific expertise in measuring QOL in CHD patients, developing international QI collaboratives, leading networks of registries, and in CHD healthcare delivery within LMICs. Representatives of Children's HeartLink, a non-governmental organization engaged in worldwide advocacy activities, contributed with their extensive experience in LMICs.[58] All standard set decisions were based on peer-reviewed literature, with the majority of recommendations leaning heavily on current widely accepted societal guidelines for CHD and ACHD care.[25,43] To ease the burden of data collection, all data from every metric in the set is acquirable directly from the patient (or parent) or the chart, with no tools requiring a third party. Finally, it is recognized that CHD, as a lifelong chronic disease, will overlap heavily with patient's burden of other medical conditions. The CHD WG aligned its mission and efforts with ICHOM standard sets assessing overall paediatric health and overall adult health, to Co-ordinate measurement timing and assure efficiency in data collection.

Next steps will be assessing the feasibility of implementing the set. Implementation at the individual patient level will allow short term

Table 3 Availability of feasible measurement tools specific to outcomes

Outcome	*Measurement tool widely available (at no cost with many translations and low burden of implementation)	No tool available, alternative questions developed to assess outcome	No tool available, outcome not assessed
Survival	■		
Heart failure	■		
Activity level and exercise	■		
Arrhythmias		■	
Pregnancy		■	
Motor developmental delay		■	
Growth	■		
Productivity	■		
Financial burden		■	
Cognition (adults)			■
Behvaior/emotional development		■	
Depression	■		
Anxiety	■		
QOL and perceived health status	■		
Family QOL	■		

learning towards the broader goal of understanding the impact of CHD to facilitate value-based healthcare. Establishing pilot use at institutions of variable income status and cultures will be paramount in gauging the set's implementation potential. Finally, linking financial reimbursement for clinical services using this set will be a key success measure for its adoption; including it among health priorities for multicultural agencies such as WHO, Worldbank, and UNICEF in LMICs will leverage the set as a framework for measuring and delivering high-value healthcare.

Limitations

Limitations to this standard set are inherent to a fluid process of data collection once implementing the standard set. While the current outcomes are derived comprehensively from extensive reviews, it is reasonable that individual patients will seek other outcomes as higher priority. The standard set is by intention a minimal set of outcomes to encourage feasibility of implementation, with the drawback that many outcomes were excluded. The ability to effectively measure outcomes will be dependent on overcoming the following limitations: (i) cost limitations from participating members, (ii) data inconsistencies and interoperability with health records, (iii) concurrent alignment with registries that are currently in-use that provide benefit to institutions. Importantly, while this aimed to be an inclusive international set, there are limitations in the generalizability in LMICs. The cross-cultural validity and reliability of these tools is unknown, having been established primarily in high-income countries, which is reflected by a lack of data from LMICs in peer-reviewed journals. The set is flexible in its makeup though, so as implementation is undertaken by a system, it is perceivable that outcomes of greatest cultural or geographic relevance could be focused on first, rather than instituting the set in whole.

Conclusion

CHD is a worldwide lifelong condition characterized by chronic morbidities and acute episodes of acute healthcare utilization, with patients facing physical, social, and mental challenges. This set can serve as a model for a lifespan approach of chronic disease to provide a baseline for decision-making, comparisons among health systems beyond the existing paradigms, and the development of comparative effectiveness and QI initiatives. This is the first international development of a standard set of outcomes for CHD and a first step towards value-based healthcare in CHD.

Supplementary material

Supplementary material is available at *European Heart Journal – Quality of Care and Clinical Outcomes* online.

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Data availability statement

The data underlying this article are available in the article and in its online supplementary material.

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