Cardiology in the Young

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Patient-reported outcome measures in congenital heart surgery: a systematic review

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Review

Cite this article: Francis J, Prothasis S, George J, and Stoica S (2023) Patient-reported outcome measures in congenital heart surgery: a systematic review. *Cardiology in the Young* **33**: 337–341. doi: 10.1017/S1047951123000057

Received: 15 November 2022
Revised: 2 January 2023
Accepted: 2 January 2023
First published online: 18 January 2023

Keywords:
Patient-reported outcome measures; patient-reported outcome measures; CHD; heart surgery

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Abstract

Background: Patient-reported outcome measures are commonly used to evaluate the effectiveness of treatments. CHD remains the most common congenital malformation. There has been a gradual shift in evaluating the outcome of surgery for CHD from mortality to morbidity and now to self-reported outcomes. *Aims:* We aimed to review studies assessing patient-reported outcome measures as a useful marker of outcome for patients, both children and adults, who underwent surgery for CHD. *Methods:* A systematic database search was conducted of original articles that explored the application of patient-reported outcome measures in the CHD surgical setting in PubMed and SCOPUS from inception to February 2022. *Results:* Our search yielded 1511 papers, of which six studies were included in this review after screening abstract and full-text, with a total sample size of 5734 patients. The main areas of discussion were the utility of patient-reported outcome measures, determinants of patient-reported outcome measures, and the need for a congenital cardiac surgery-specific patient-reported outcome measure for paediatric patients and their parents/guardians and adult patients. *Conclusion:* This systematic review reports the use of patient-reported outcome measures to be a useful indicator to gain insight into the patients' perspective to provide holistic and patient-centred management. However, further studies are required to assess the utility of patient-reported outcome measures in a congenital cardiac surgical setting.

Over 1.3 million babies are born each year with CHDs.[1] Due to the nature of the disease, immediate surgery is often required. Follow-up surgery during adolescence and adulthood is not uncommon. With the change in medical practice to patient-centred care, utilising patient-reported outcome measures allows for patients to further take part in clinical decision-making. This also allows the clinicians to identify and quantify the impact of CHD surgery on patients and their families, including quality of life.

Perceived health, from both physical and mental perspectives, ought to be evaluated to obtain a comprehensive view of patient status. Furthermore, in different cardiac populations, patient-reported outcomes are found to be independent predictors of mortality.

The objective of this systematic review was to consolidate evidence on the use of patient-reported outcome measures for patients undergoing surgery for CHD. We investigated the types of patient-reported outcome measures used in both paediatric and adult patients, the variation and determinants of patient-reported outcome measures, and their utility in the clinical setting.

Methodology

Search strategy

A search for all relevant literature was performed using the PubMed and SCOPUS databases in February 2022. Search terms included: ("PROMS" OR "PREMS" OR "PROM" OR "PREM" OR "Patient Reported Outcome Measures" OR "Patient Report Outcome Measure" OR "Patient Reported Experience Measure" OR "Patient Reported Experience Measures" OR "Qualitative Methods" OR "Qualitative Methodology" OR "Quality of Life Measurement" OR "Capability Well-being Measure" OR "PRO" OR "PROS" OR "Quality of Life" OR "HRQL" OR "HRQoL" OR "QoL") AND ("Congenital Cardiac" OR "Paediatric Cardiac" OR "Congenital Cardiac Surgery" OR "Paediatric Cardiac Surgery" OR "Congenital Heart Disease" OR "Adult Congenital" OR "Adult Congenital Surgery" OR "Congenital Surgery").

Search criteria included articles being available in the English language, full-text articles, and primary research papers. The population was inclusive of both adult and paediatric patients with CHD who had undergone cardiac surgery. References of identified papers were also reviewed to ascertain if any further papers could be included for screening.

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Study selection

Duplicates were removed prior to screening. All articles yielded in our search were screened using the title and abstract in Rayyan[2] by two independent reviewers (JF and JG) before a decision was made. A third independent reviewer (SS) resolved any disagreements between the reviewers. Subsequently, screened articles were analysed through a full-text review. Figure 1 outlines the preferred reporting items for systematic reviews and meta-analysis flow diagram.

Review articles and secondary articles were excluded from our study. The inclusion criteria were as follows:

1. Articles focused on patient-reported outcome measures or self-reported quality of life assessment.
2. The patient population include parents of children who underwent surgery for CHD, paediatric patients who underwent surgery for CHD, or adult patients who underwent surgery for CHD.

Data extraction and analysis

Using a pre-established protocol, information was extracted from the final studies by two authors (JF and JG). Tabulated data include the type of study, sample size, population demographics, number of patient-reported outcome measures, type of patient-reported outcome measures used, what patient-reported outcome measures evaluated, and main outcomes. The correctness of the tabulated data was validated by a third author (SS). Due to the heterogeneity in the "quality of life" data, it was not possible to conduct a meta-analysis.

Results

Study selection

A total of 1511 articles were identified in the literature search, of which 46 were screened following duplication and were read in full and assessed in accordance with the inclusion and exclusion criteria. A total of six studies were included in this review following critical appraisal, featuring 5734 patients. A summary of the studies collected and their respective designs, type of outcomes measured, and their main reported outcomes are found in Table 1.

Variations in patient-reported outcome measures and their characteristics

Sixteen different types of patient-reported outcome measures were used in the six studies. Of the six selected studies, the distribution per age was as follows: three utilised patient-reported outcome measures in the adult CHD population,[4,7,8] one was conducted in adolescents and young adults with CHD,[5] one was conducted by parents who were told to report from the child's perspective,[3] one was conducted by children between the ages of 10 and 14, in addition to their parents answering the questionnaire.[6]

Discussion

Survival rates of patients with CHD have significantly increased over recent years steering research into the assessment of long-term functioning and well-being by use of patient-reported outcome measures. The publication of studies that apply patient-reported outcome measures in the setting of CHD showed an important increase over time.[9] It is increasingly evident that understanding children's views about living with CHDs is vital to bolstering their successful participation in daily life, school, and peer relationships.

There was significant variability in patient-reported outcome measures used, all of which evaluated a variety of constructs through different instruments. Patient-reported outcome measures instruments were not surgery specific and generally assessed health-related quality of life, functional status, reports of adverse effects, perceptions of well-being, and satisfaction with treatment. We found evidence in support of patient-reported outcome measures increasing disease awareness, treatment response, and allowing physicians to provide multi-faceted care to ensure that they carry out a more holistic management of patients.

There was no agreed consensus on which instrument to use with each study using a variety of self-completed questionnaires. Most of these questionnaires were previously validated patient-reported outcome measure tools, but none of them were specific to congenital heart surgery.

The patient-reported outcome measures studies we reviewed gave an insight that otherwise could not be gained. This specifically involved the determinants of patient-reported outcome measures. For example, Guerra et al. (2013)[3] found that children who underwent CHD surgery at an older age had significantly worse health-related quality of life than those who had surgery in early infancy. These findings favour the notion of early complete repair where possible and preferably before school age when self-awareness and experiences consolidate. Interestingly, children with biventricular repairs had significantly lower total PedsQL 4.0 scores than the control population, reflecting overall worse health-related quality of life. These children had a high score in physical functions, but a lower score in all other dimensions including emotional, social, school, and psychosocial functioning. These findings reflect that clinical condition or disease severity do not necessarily correlate with perception of quality of life.

Silva et al (2011)[5] found that patients with better social support showed better quality of life in all dimensions than those with poorer social support. Crucially, CHD patients who had not undergone surgery had a better quality of life in the physical dimension compared to surgical patients. It is important to delineate whether the functional consequence of surgery appears as a limitation to our patients, or whether being surgery-free is more important to not feel limited. Patients stated that frequent hospitilsation and check-ups restricted their day-to-day life and placed limitations on their physical activities. However, the authors found that CHD patients had a higher overall quality of life than the control Portuguese population. This may be due to disease identity, where patients, especially those with congenital defects, become more resilient and adaptative early on in life.

Several authors also investigated the impacts of cyanosis and the number of surgical procedures on quality of life. They concluded that cyanosis itself did not have a significant effect; however, factors such as number of surgical procedures and the severity of residual injury did negatively affect their quality of life, physical and psychological health.

Residual lesions were also seen to be a factor that contributes to difference in outcomes between patient groups and their perspective on quality of life. Patients with severe or moderate lesions scored lower on physical as well as psychological functions and reported worse quality of life than individuals with mild lesions.[5] This could be explained by the differences in individuals and their general functionality. Individuals with severe lesions are more likely to have increased limitations in their daily habits and have

https://doi.org/10.1017/S1047951123000057 Published online by Cambridge University Press

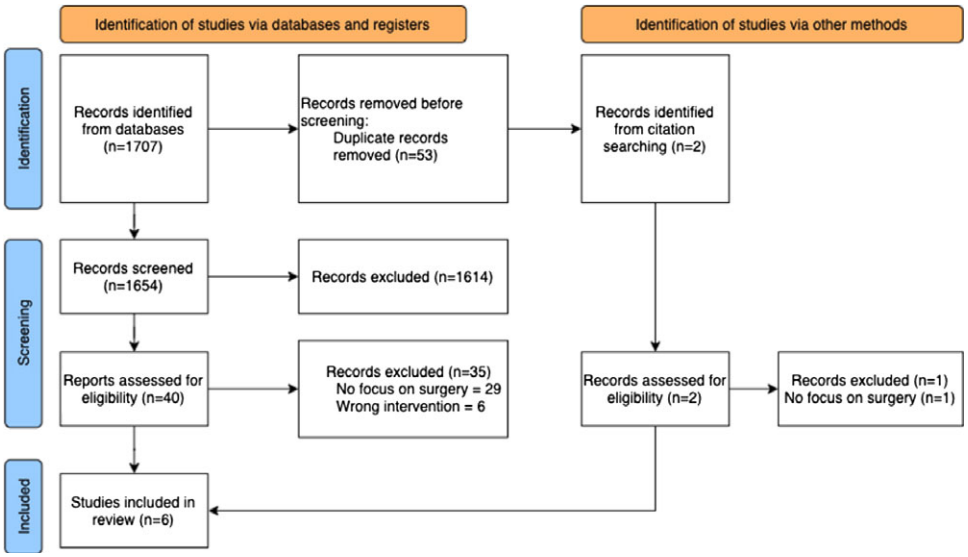


Figure 1. PRISMA diagram of study identification and selection.

resulting self-esteem issues, mental health limitations, and freedom in functionality than those with mild lesions. However, the authors found that CHD patients had a better psychological profile than the healthy population. This was believed to result from the differing social and family relationships between the two groups. Individuals with CHD are more likely to have protective and united family and support group because they required extra attention and care from a young age. This increased support and thereby likely improved mental health will likely have a positive effects on patients' physical domains. Within CHD patients, older patients are more likely to report better psychological scores as they have managed their illness for a long period of time and have therefore adjusted to their lifestyle well and are functioning well with their routine. Furthermore, we can deduce that the patients' age when reporting for patient-reported outcome measures could also influence their response as older individuals with CHD are more likely to report a better quality of life than younger individuals.

Children with increased number of cardiac operations as well as presence of post-surgical residual defects reported to have worse quality of life. It is agreed within the literature reviewed that social support is a clear factor in compensating for the health problems for CHD patients, including severity of disease and residual disease. Social support can likely improve perceived quality of life in all domains.

Knowles et al. (2016) used an open question providing an opportunity to capture novel data in an unstructured format from children. One 11-year-old girl reported that "it's quite hard to make friends because. .. you're different to them.. . it took almost a year for me to make friends in my high school". This type of insight can help identify and promote strategies to successfully negotiate the challenges of living with a long-term condition. It is also essential that a CHD-specific patient-reported outcome measures is standardised to account for parents of neonates and infants, children, and parents of children undergoing treatment, teenagers, and adults with CHD.

Due to the nature of CHD, different patient-reported outcome measure instruments are required to assess patient outcomes of different ages. One of the primary aims of patient-reported outcome measures is to help children successfully negotiate the challenges of living with a long-term condition. Therefore, assessing outcomes from a young age all the way up to adulthood is imperative to achieve this goal. However, each stage requires a different approach based on the age, literacy, and communication of the patient. Consequently, assessing parents/guardians to try report from the child's perspective is a useful tool. To provide a holistic understanding of the patients' status, it will require a consensus panel of various stakeholders including patients, parents, surgeons, physicians, mental health professionals, etc.

Knowles et al. (2016) states that the coping strategies patients utilise are similar in different severity groups, including accepting their condition as a part of their identity and normalising their experience by emphasising that their experiences are similar to others. This self-reassurance allows patients to positively "reframe" their journey in this lifelong condition. Interestingly, patients with a lower disease severity often denoted their condition to be "in the past". Perhaps it would be useful to discern the ways in which how the degree of acceptance and self-reassurance improves patients' self-reported quality of life.

However, to contrast and compare patient-reported outcome measures in CHD, consensus in which outcome measures and instruments to use is required. For example, in rheumatology, the Outcome Measures in Rheumatology initiative has defined core domain sets (what outcomes to measure) and core outcome measurement sets (how to measure the outcome, i.e., which patient-reported outcome measure to use).[10]

Interestingly, Moons et al (2005) found that disease severity only had a detrimental impact on patients' lives only when it was measured in terms of poor functional status. Contrary to Knowles et al. (2016), a diagnosis of CHD is not a significant determinant of patient-reported outcome measures, rather only the functional outcome of the disease or treatment determines one's own quality of life.

Variations will occur between patients from different socio-economic classes, genders, functional class, and degree of support provided. Moons et al. (2018) found that of the countries in the APPROACH-IS, patients from Switzerland, Sweden, and the Netherlands showed the most favourable patient-reported outcome measures. Whereas patients from Japan, France, and India reported lower patient-reported outcome measures. However, the analysis used showed great variation in assessing the outcome of patient-reported outcome measures instruments. For example, the authors previously noted that Australian patients had the best

https://doi.org/10.1017/S1047951123000057 Published online by Cambridge University Press

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Table 1. Resultant studies exploring the use of PROMs in congenital cardiac surgery patients.

Study	Year	Type of study	Author location	Sample size	PROMs instrument	Primary aim	Main outcome
Guerra et al.	2013	NM, P	Canada	130	1. PedsQL 4.0	Determine health related QOL at 4 years after children underwent CHD surgery	(1) Health-related QOL was significantly lower in children who underwent surgery for CHD in early infancy. (2) An association was found between age at surgery and postoperative low cardiac output, socioeconomic status, and QOL
Moons et al.	2018	M, P	Belgium	4028	(1) SF-12, (2) EQ-5D, (3) HADS, (4) HBS-CHD, (5) LAS, (6) SWLS	(1) Investigate inter-country variation in PROMs in adults with CHD. (2) Identify patient-related predictors of PROMs. (3) Explore standard of living and healthcare system characteristics as predictors of PROMs	(1) Substantial inter-country variation was observed, with Switzerland having the highest composite PROMs score and India the lowest. (2) Functional class, age, and employment status were patient-related factors that independently and consistently predicted PROMs. (3) Standard of living and healthcare system characteristics predicted PROMs above and beyond patient characteristics
Silva et al.	2011	NM, P	Portugal	40	(1) WHOQOL-BREF	(1) Assess perception of quality of life of adolescents and young adults with CHD. (2) Examine variables that have a negative impact and add resilience to self-perception of quality of life	(1) Compared to a healthy Portuguese population from a previous study, the patients in this study showed a better perception of QOL. (2) The number of surgical procedures and the persistence of moderate-to-severe residual injuries had considerable detrimental effect
Knowles et al.	2016	M, P	United Kingdom	436	PedsQL 4.0	Assess self-reported health experiences and coping with CHD	(1) Children's reported experiences were not dependent on their cardiac diagnosis, although there were clear qualitative differences by clinical severity group. (2) Children's concerns emphasised social participation and the findings imply a need to shift the focus from monitoring cardiac function to optimising participation
Bay et al.	2017	M, P	Sweden	471	1. SF-12, (2) HBS-CHD, (3) LAS, (4) SWLS), (5) SOC-13	Analyse factors associated with physical activity in adults with CHD using PROMs	(1) Physical Component Summary Score (PCS) and Mental Component Summary Score (MCS) are more strongly associated with physical activity level than age and medical factors. (2) The use of PROMs could provide valuable information on the benefit of personalised advice regarding physical activity to patients with CHD
Moons et al.	2005	NM, P	Belgium	629	(1) LAS, (2) SWLS, (3) SEIQoL-DW	(1) Explore whether the severity of CHD is associated with quality of life and perceived health status of adult patients	(1) Severity of CHD is marginally associated with patients QOL and perceived health. (2) Functional status was more related to patients' assessment of QOL than was the initial diagnosis or illness course

M = multicentre, NM = non-multicentre, P = prospective, NP = non-prospective.
PedsQL 4.0 = Paediatric quality of life version 4.0.
SF-12 = 12-item short-form survey.
EQ-5D = Euro-Qol-5D.
HADS = Hospital anxiety and depression scale.
PROMIS Anxiety = Patient-reported outcomes measurement information system anxiety.
PROMS Depression = Patient-reported outcomes measurement information system depression.
HBS-CHD = Health behaviour scale-congenital heart disease.
LAS = Linear analogue scale.
SWLS = Satisfaction with life scale.
WHOQOL-BREF = World Health Organization quality-of-life scale.
CD-RISC 10 = Connor–Davidson resilience scale.
SEIQoL-DW = Schedule for the evaluation of the individual quality of life-direct weighting.

quality of life when measured with a linear analogue scale; however, the more recent study suggested that the Australian patients' scores were closer to the median when quality of life was assessed with other patient-reported outcome measures. It is not known whether geographical variations actually represent genuine differences in perceived quality of life, or whether a specific type of patient-reported outcome measures is unable to capture the cultural, language, and social differences across borders.

It is also crucial to consider that patients with CHD are differently impacted by the economics of the country they reside in, with factors such as standard of living and differences within healthcare systems all influencing their quality of life varyingly. This further signifies the importance of having an evidence-based congenital heart surgery specific patient-reported outcome measures, whilst having variations which account for age and background.

Overall, none of the patient-reported outcome measures instruments used in the studies were specific to congenital heart surgery or transcatheter intervention. Furthermore, no absolute differences were stated in defining patient-reported outcome measures and self-reported quality of life. It is important to note that patient-reported outcome measures are a multi-dimensional tool that can assess self-reported quality of life amongst other things including functional status, physical well-being, mental well-being, impact on social life and academia, etc. Although a few patient-reported outcome measures aimed to assess the quality of life holistically, no consensus was made about which patient-reported outcome measures is most effective for congenital heart surgery patients. Importantly, as surgical technique treatments continue to evolve, it may be worthwhile to assess how the use of patient-reported outcome measures pre-operatively can guide treatment plans by tailoring support patients require or want. Such findings warrant the need for a novel patient-reported outcome measures that can be used in the context of congenital heart surgery.

Patient-reported outcome measures instruments are a useful indicator of outcome, but the instruments we analysed were not surgery specific and assessed general quality of life, physical-, mental-, and emotional status postoperatively. Surprisingly, in addition to the severity of CHD, the complexity of the surgery, and the number of operations predicting patient-reported outcome measures, socio-economic status, age at the first operation, employment status, and support available were also strong determinants of patient-reported outcome measures. When patient-reported outcome measures did not match their respective CHD severity, qualitative differences were observed between different clinical severity groups.

Major concerns expressed by children and adults include the lack of inclusion within society, fear of further operations and treatments, and the constant worry about their health deteriorating. Using patient-reported outcome measures can help clinicians offer various types of support to facilitate better coping strategies, whether it be group-based discussions, academic support, or family support. Compared to other specialties, few studies and patient-reported outcome measures instruments exist in congenital heart surgery. Further research can look into developing a congenital heart surgery specific patient-reported outcome measures with the aim of highlighting current limitations in assessing outcomes postoperatively.

Acknowledgements. None.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Conflicts of interest. None.

Ethical standards. NA.

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