

Quality of life in adults living with congenital heart disease: beyond morbidity and mortality

Amanda L. Hunter, Lorna Swan

Adult Congenital Heart Disease Unit, Royal Brompton & Harefield NHS Foundation Trust, London, UK

Correspondence to: Dr. Lorna Swan, Adult Congenital Heart Disease Unit, Royal Brompton & Harefield NHS Foundation Trust, London, UK. Email: L.Swan@rbht.nhs.uk.

Provenance: This is an invited Editorial commissioned by the Section Editor Haiyun Yuan (Department of Cardiovascular Surgery, Guangdong Provincial Cardiovascular Institute, Guangdong General Hospital, Guangzhou, China).

Comment on: Apers S, Kovacs AH, Luyckx K, *et al.* Quality of Life of Adults With Congenital Heart Disease in 15 Countries: Evaluating Country-Specific Characteristics. *J Am Coll Cardiol* 2016;67:2237-45.

Submitted Nov 01, 2016. Accepted for publication Nov 04, 2016.

doi: 10.21037/jtd.2016.12.16

View this article at: <http://dx.doi.org/10.21037/jtd.2016.12.16>

“Living well is as important to most people as living longer” is the opening statement of Apers *et al.*'s recently published paper in the *Journal of the American College of Cardiology* addressing quality of life (QOL) amongst adults living with congenital heart disease (CHD) (1). As survival with CHD continues well into adulthood, owing to the dramatic success story of pediatric cardiovascular diagnostics and cardiothoracic surgery, the population of adults with CHD is growing, forming more than 60% of the total CHD population (2), and now exceeding the number of newborns with CHD annually (3). Almost all adults with CHD require periodic evaluation by a congenital cardiologist throughout their lifetime, creating an ever-increasing pressure on healthcare services (4,5). Furthermore, as life expectancy has increased, so too has the prevalence of age-related co-morbidities that add a further layer of complexity to the healthcare needs of this patient group. However it is vital that, as clinicians, we do not lose sight of the fact that reducing the burden of disease by improving overall well-being is as important to our patients as their long-term survival and indeed, the two are likely to be inextricably linked.

QOL is defined as the degree of overall life satisfaction that is positively or negatively influenced by an individual's perception of certain aspects of life important to them including matters both related and unrelated to health (6). Furthermore QOL in adults with CHD depends on multiple factors—to a limited extent it is determined by their cardiac

condition but it is very dependent on the individual's perceptions. Traditionally the long-term outcome measures that have been the bedrock of clinical research have been mortality, physical morbidity and functional status. QOL outcomes have been seen as supplementary to “hard” clinical data yet have been shown to strongly correlate with important medical outcomes. Moreover, QOL outcomes provide guidance to health care professionals in terms of the decision to institute specific treatments and to policy-makers allocating resources. It has been demonstrated that when assessing QOL, focus should be on the individual and domains that are specifically important to them rather than an external perception of need. Despite the increasing growth in QOL research in the general biomedical field, it has not been routinely used as a key outcome measure in cardiovascular research (7). The congenital community have recognized and embraced such patient reported outcomes as an important research domain for over 40 years with over 230 published studies assessing QOL. However, there has been criticism over methodological limitations of some such studies owing to such heterogeneous populations, study design and poor consistency of QOL conceptualization and measurements (8,9).

Assessment of patterns of patient-reported outcomes in adults with congenital heart disease—International Study (APPROACH-IS) is a cross-sectional, multi-level study designed to comprehensively assess patient-reported outcomes, including perceived health status, psychological

functioning, health behaviors and QOL, in adults with CHD using a single methodology, including patient- and country-specific criteria, in an attempt to ensure generalizability of findings (10). Apers *et al.* should be commended in their scientifically rigorous attempt to use a uniform, standardized and consistent approach, and in utilizing two well-validated QOL tools: the linear analog scale (LAS) and the satisfaction with life scale (SWLS). In addition, this is the now the largest study and the first global study in this field enrolling 4,028 patients from 15 countries across five continents. Both developed and developing countries were studied although it was not possible to collect data from any unit in Africa. The international basis of the study attempted to address if there was international variation in QOL and to identify factors that are country-specific. The study responder rate was high with data available in 3,952 patients (median age 32 yrs). The majority of patients had CHD defined as moderate complexity (49%) and were in New York Heart Association functional class 1 (54%).

Overall, QOL was reported as being good in the majority of patients in-keeping with previous studies. There was, however, a large variability in QOL. On the 100-point LAS (which sets a score of 50 to be neutral) the median score was 80. Nearly 10% of subjects reported a score less than 50, consistent with a poorer QOL. Thirty percent of patients reported a score between 50–70 and the majority a score greater than 70. The authors demonstrated that a worse QOL was associated with patient-specific characteristics such as older age, those seeking employment, unemployed or classified as disabled, those having never been married and those with a higher NYHA functional class. Patients in Australia reported the highest QOL. This was ten points higher than Japan where patients reported the lowest QOL. The authors conclude that further investigation is required to explain this difference in QOL between countries however there was no relationship between QOL and the national happiness score or cultural dimensions between countries implying that CHD patients with poor QOL can be identified with the same QOL tools irrespective of their country of residence. The authors do however correctly identify that future studies should also include a control population within individual countries to assess if there are variations in QOL that are specific to the general population rather than a specific patient group. Overall, it was concluded that variation in QOL was associated with patient-specific and not country-specific characteristics. Interestingly, increasing disease complexity or severity was not associated with QOL re-iterating previous findings (6).

Often patients with very complex disease report a good QOL.

To date, there has been a paucity of studies undertaken to assess if there are country-specific determinants of QOL. A number of socioeconomic factors are associated with well-being including income per capita, income inequality, social welfare, individualism, democracy and freedom, social capital and physical health (11). It has been suggested that wealthier nations need to focus on determinants of subjective well-being in order to improve QOL for patients with chronic illness, whereas with continued economic growth, the global well-being in poorer nations should improve. Cultural characteristics within countries have also been shown to shape QOL (12) with Japanese and Taiwanese students being less likely to report extremes on a life satisfaction response scale in comparison to American students, although interestingly in this current study by Apers *et al.* there was no relationship between QOL and cultural dimensions. It has previously been shown that across countries there are significant variations in the care experiences of those suffering from chronic illness. For example, in the United States current health care policies were found to have the most marked adverse impact on the experience of chronically ill patients in terms of difficulty in accessing care, poor care co-ordination and efficiency and patient-reported safety concerns, with many patients being unable to afford to follow care recommendations (13). Chronically ill patients have tended to fare better in countries with strong primary care infrastructures, however in the developed countries previously studied, there was still room for improvement highlighted in transitional care, in the co-ordination for patients seen by multiple clinicians or allied health care professionals and in efforts to engage or support patients to manage their conditions. Initiatives have been set up within countries sharing many similar features such as; incentivizing health-care providers, using registries, electronic information systems and telehealth technology and targeting the highest risk patients for outreach and follow-up.

Despite the many strengths of this current study it has identified some limitations requiring to be addressed in order to further progress this field. QOL was often only assessed in patients from a single center within a country. Although this center was likely to have been a national tertiary referral center and perhaps providing specialist care for all congenital patients throughout the country, it raises concerns if reported QOL is representative of patients with CHD that are perhaps seen in non-specialist centers or lost

to follow-up completely. It could be argued that patients attending a tertiary referral regularly are those likely to have access to the greatest support. This includes best medical practice but also support surrounding disease education, lifestyle advice and the provision of psychological services.

Previous work has identified that there is a major issue with CHD patients, even those with severe lesions, being lost to follow-up especially during transition from pediatric to adult care. Amongst young adults, there is an increase in the proportion with CHD being admitted to the emergency department (14) and an overall increase in the number of admissions for CHD (15). It is concerning that a previous study revealed that 1 in 5 young adults with severe CHD were not even seen by a cardiologist during the ages of 18–22 years (16). This lapse in care has been shown to be associated with adverse outcomes such as further cardiac complications, a greater need for urgent cardiac intervention and an adverse impact on QOL (17). The barriers to ongoing lifelong care and a smooth transition, such as a patient's poor comprehension of their condition, difficulty adjusting to an autonomous adult healthcare environment from the more paternalistic setting of pediatric care and lack of parental input, must be addressed if we are to solve this perturbing trend.

When robust care is in place, the congenital heart disease team (including the cardiologists and specialist nurses) has the privilege of getting to know their patients over many years of follow-up. This therapeutic relationship places clinical staff in an ideal position of being able to identify patients at risk of “detrimental” personal health and non-health related perceptions.

Healthcare models, therefore, need to develop a more holistic approach to life-long care. Although time consuming, there should be a focus on nurturing positive personal perception especially in older children and young adults with CHD. One could argue that this investment is of equal long-term importance to ensuring effective long-term hemodynamics! The relationship between the clinical team, the patient and their family/carers is key to this process: this safe environment can foster an ethos of enablement and personal autonomy in a way that works with the patient's complex medical issues and not against them.

Adult congenital heart disease units have an obligation to prioritize and invest in comprehensive transition programs if they wish to maximize the wellbeing of the patients under their care. Although much of this work will be focused on the adolescent years this enabling principle should permeate

care at every age and stage of the congenital heart disease pathway.

In contrast to most patients where illness is a factor of an advancing age, the majorities of CHD patients have lived their entire lives with their condition and will have undergone at least one; if not multiple interventions by the time they reach adulthood. In the current study by Apers *et al.*, QOL was assessed amongst CHD patients without a control group for comparison. This would be the logical next step in their programme. Previous studies have revealed inconsistencies regarding the QOL amongst adults with CHD and healthy controls. However in a previous study utilizing the same methodology adults with CHD reported a better QOL than healthy controls (18). It may seem incongruous that CHD patients should report better QOL; however, having lived with a condition lifelong it is conceivable CHD patients have adapted to life and changed expectations accordingly. This is a key determinant of QOL and is termed sense of coherence. This is a measure of one's outlook on life and expresses the extent to which the individual perceives the world as structured and predictable (i.e., comprehensibility), that resources are available to meet demands posed by situations (i.e., manageability) and that the demands are worthy of investment (i.e., meaningfulness) (19). Patients with congenital heart disease have been reported to have an increased sense of coherence (20). Another explanation of enhanced QOL in CHD patients is the disability paradox where individuals with a disability may perceive higher QOL by acknowledging their impairment, remaining in control and having a high level of self-satisfaction (21). By contrast, poor illness perceptions are predictive of poor QOL (22). With the assistance of psychology colleagues congenital clinicians we can help change illness and non-health related perceptions and can identify and exploit coping strategies.

A sub-group of CHD patients with complex needs and management issues are those with learning difficulties and cognitive impairment. They are often excluded from research studies especially if traditional tools for assessing QOL are used, as was the case in this current study. Inclusion in studies will remain difficult especially from an ethical standpoint but nonetheless, active assessment of QOL has been shown to be possible, even amongst patients with profound learning difficulties, with input from their carers (23). This sub-group represents a significant proportion of the workload of congenital cardiologists who often have significant unmet needs and are identified as an at-risk group for impaired QOL so it is all the more

pertinent that we attempt to assess QOL, it may however require ingenuity to do so.

The importance of specialist lifelong follow-up in CHD cannot be overstated, not only in identifying potential new cardiac lesions and planning re-intervention in a timely fashion, but in order to understand the issues important to our patients, to advocate for them and to empower them to achieve the best QOL possible. The adult congenital team often inherits patients where treatment decisions made in childhood have a huge bearing on adult QOL. Closer collaboration and shared decision-making between pediatric and adult teams can facilitate proactive planning to target QOL as another treatment domain.

Looking to the future, congenital cardiologists will be tackling the uncharted territory of many CHD patients achieving a near-normal life expectancy. The elderly CHD patient will bring a new set of healthcare issues (24) and we should be anticipating new challenges to QOL related to the physical and psychosocial aspects of aging. This represents another area for further longitudinal research.

In summary, patient reported outcomes such as QOL are powerful tools to inform clinicians of the, often hidden, morbidity and “suffering” associated with a chronic disease and provide information on the ‘patient experience’. Previous work, including this current study by Apers *et al.*, has highlighted the need to move away from observational studies assessing QOL and to look towards development of targeted interventional studies aiming to improve QOL. The heterogeneous population of patients with CHD, with ever accumulating events will require such interventions to be tailored to the needs of specific populations and individuals.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

References

1. Apers S, Kovacs AH, Luyckx K, et al. Quality of Life of Adults With Congenital Heart Disease in 15 Countries: Evaluating Country-Specific Characteristics. *J Am Coll Cardiol* 2016;67:2237-45.
2. Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. *Circulation* 2014;130:749-56.
3. Baumgartner H. Geriatric congenital heart disease: a new challenge in the care of adults with congenital heart disease? *Eur Heart J* 2014;35:683-5.
4. Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J* 2010;31:2915-57.
5. Nishimura RA, Carabello BA, Faxon DP, et al. ACC/AHA 2008 Guideline update on valvular heart disease: focused update on infective endocarditis: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines endorsed by the Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *J Am Coll Cardiol* 2008;52:676-85.
6. Moons P, Van Deyk K, Marquet K, et al. Individual quality of life in adults with congenital heart disease: a paradigm shift. *Eur Heart J* 2005;26:298-307.
7. Anker SD, Agewall S, Borggrefe M, et al. The importance of patient-reported outcomes: a call for their comprehensive integration in cardiovascular clinical trials. *Eur Heart J* 2014;35:2001-9.
8. Bratt EL, Moons P. Forty years of quality-of-life research in congenital heart disease: Temporal trends in conceptual and methodological rigor. *Int J Cardiol* 2015;195:1-6.
9. Fteropoulli T, Stygall J, Cullen S, et al. Quality of life of adult congenital heart disease patients: a systematic review of the literature. *Cardiol Young* 2013;23:473-85.
10. Apers S, Kovacs AH, Luyckx K, et al. Assessment of Patterns of Patient-Reported Outcomes in Adults with Congenital Heart disease - International Study (APPROACH-IS): rationale, design, and methods. *Int J Cardiol* 2015;179:334-42.
11. Jorm AF, Ryan SM. Cross-national and historical differences in subjective well-being. *Int J Epidemiol* 2014;43:330-40.
12. Diener E, Diener M, Diener C. Factors predicting the subjective well-being of nations. *J Pers Soc Psychol* 1995;69:851-64.
13. Schoen C, Osborn R, How SK, et al. In chronic condition: experiences of patients with complex health care needs, in eight countries, 2008. *Health Aff (Millwood)* 2009;28:w1-16.

14. Gurvitz MZ, Inkelas M, Lee M, et al. Changes in hospitalization patterns among patients with congenital heart disease during the transition from adolescence to adulthood. *J Am Coll Cardiol* 2007;49:875-82.
15. Agarwal S, Sud K, Menon V. Nationwide Hospitalization Trends in Adult Congenital Heart Disease Across 2003-2012. *J Am Heart Assoc* 2016;5. pii: e002330.
16. Mackie AS, Ionescu-Ittu R, Therrien J, et al. Children and adults with congenital heart disease lost to follow-up: who and when? *Circulation* 2009;120:302-9.
17. Yeung E, Kay J, Roosevelt GE, et al. Lapse of care as a predictor for morbidity in adults with congenital heart disease. *Int J Cardiol* 2008;125:62-5.
18. Moons P, Van Deyk K, De Bleser L, et al. Quality of life and health status in adults with congenital heart disease: a direct comparison with healthy counterparts. *Eur J Cardiovasc Prev Rehabil* 2006;13:407-13.
19. Apers S, Luyckx K, Moons P. Quality of life in adult congenital heart disease: what do we already know and what do we still need to know? *Curr Cardiol Rep* 2013;15:407.
20. Müller J, Hess J, Hager A. Sense of coherence, rather than exercise capacity, is the stronger predictor to obtain health-related quality of life in adults with congenital heart disease. *Eur J Prev Cardiol* 2014;21:949-55.
21. Albrecht GL, Devlieger PJ. The disability paradox: high quality of life against all odds. *Soc Sci Med* 1999;48:977-88.
22. Schoormans D, Mulder BJ, van Melle JP, et al. Illness perceptions of adults with congenital heart disease and their predictive value for quality of life two years later. *Eur J Cardiovasc Nurs* 2014;13:86-94.
23. Sines D, Hogard E, Ellis R. Evaluating quality of life in adults with profound learning difficulties resettled from hospital to supported living in the community. *J Intellect Disabil* 2012;16:247-63.
24. Tutarel O, Kempny A, Alonso-Gonzalez R, et al. Congenital heart disease beyond the age of 60: emergence of a new population with high resource utilization, high morbidity, and high mortality. *Eur Heart J* 2014;35:725-32.

Cite this article as: Hunter AL, Swan L. Quality of life in adults living with congenital heart disease: beyond morbidity and mortality. *J Thorac Dis* 2016;8(12):E1632-E1636. doi: 10.21037/jtd.2016.12.16