Cardiol Young 2006; 16: 125–127

ISSN 1047-9511
doi: 10.1017/S1047951106000035

Editorial Comment

Beyond survival: What are the outcomes that really matter to our patients?

Kathleen A. Mussato

Herma Heart Center, Children's Hospital of Wisconsin, Milwaukee, USA

THE IMPORTANCE OF EXAMINING QUALITY OF LIFE as an outcome for children with congenital cardiac disease has long been recognized. Despite multiple differences in cultural, and systematic approaches to provision of care for those with congenital cardiac malformations, as clinicians we all hope that the results of our therapeutic choices impart quality, not merely quantity, of life to the children that we treat. Questions remain as to how to measure quality of life accurately, and how to make sense of the findings obtained. The need to answer these questions is becoming increasingly urgent. Parents receiving a prenatal diagnosis of congenital cardiac disease want to know what impact the malformation will have on the quality of life to be enjoyed by their child. When options for treatment are offered, the differences in outcomes relative to this quality of life are rightfully questioned. We are obliged to provide accurate, data-based, answers to these questions, rather than anecdotes or our own personal interpretations of quality of life for such children. Fortunately, quantitative methods to evaluate quality of life, and designs for studies that allow meaningful interpretation of results, are now beginning to evolve.

When exploring reports of subjective outcomes such as quality of life, care must be taken to understand exactly what is being measured, and how. Many studies proposing to investigate quality of life in children with cardiac disease have lacked a sound theoretical foundation.[1] Physiologic measures, and other variables such as functional abilities that can be quantitatively verified, have long been used as proxies for quality of life. Some studies have equated quality of life purely with functional abilities, such as the classification developed by the New York Heart Association, the frequency of symptoms, or the capacity for exercise. Others have categorized variables, such as good ventricular function versus dysfunction, households with married versus single parents, and age-appropriate schooling versus special education, as indicators of "normal" versus reduced quality of life.[2–6] The definition of health proposed by the World Health Organization in 1948, as "a state of complete physical, mental, and social well being, and not merely the absence of disease or infirmity",[7] has frequently been cited as an important guideline for evaluating quality of life. Health, however, may not be interchangeable with quality of life, and researchers must take care clearly to delineate what they intend to measure. Gill and Feinstein[8] emphasized that it is the values and preferences of the individual that distinguish quality of life from other measures of health. Good quality of life can exist despite the burden of disease, and we must elicit the subjective interpretation of the individual in order to understand the true impact of an illness and its treatment. It has been repeatedly demonstrated that professionals involved with the provision of health care rate the quality of life for given health states different than that of survivors and their parents.[9–11] We must suspend our individual biases, therefore, and ask not only how we think our patients are doing, but also how *they* think they are doing.

In this issue of Cardiology in the Young, two manuscripts are presented that explore quality of life in children with congenital heart disease. Macran, Birks, Parsons et al. from the British Heart Foundation Care and Education Research Unit[12] put forward the results of the development and psychometric testing of a disease-specific measure of quality of life for children with congenital cardiac disease, the ConQol.

Correspondence to: Kathleen A. Mussatto RN, Herma Heart Center, Children's Hospital of Wisconsin, PO Box 1997, MS 715, Milwaukee, WI 53201, USA. Tel: +1 414 266 2073; Fax: +1 414 266 2075; E-mail: kmussatto@chw.org

Disease-specific quality of life instruments explore aspects of health, functioning and symptoms that may be unique to a given disease state, allowing comparisons across different diagnoses, treatment strategies and assessment of longitudinal changes.[13] Cardiac disease may often be an invisible ailment in children and adolescents who have undergone repair as infants, yet the burden of ongoing follow-up, treatment and symptoms may present problems for these young people that may not be readily discernable when using generic quality of life instruments alone. Disease-specific measures for use in subjects with congenital cardiac disease have emerged only recently, and have yet to be widely tested across different disease states and cultural settings. For this reason, the development of instruments like the ConQol represents an important endeavor in our continuously emerging science.

The ConQol was designed to represent the perceptions of the impact of congenital cardiac disease on the daily lives of children and adolescents aged from 8 through 16 years as measured from their perspective. The solicitation of self-reports from children and adolescents completing this instrument reflects the "child-centered philosophy" of this endeavour. While the emphasis on the perception of the child is laudable, it should be recognized that parental perceptions may drive health-care seeking behaviours. Parental input on the assessment of quality of life, therefore, cannot be discounted. Using parents as proxy reporters for the quality of life of their children is not ideal in isolation, but in conjunction with self-reports from the children, it often adds important information relevant to the interpretation of the results obtained. The measure includes 3 dimensions: symptoms, activities, and relationships, and an additional dimension for those aged from 12 through 16 years, addressing control and coping. Two separate scores are derived, one representing the quality of life items and the other, the symptom items. Separation of these scales in this instrument adds to its unique contribution, and supports the premise that assessment of symptoms alone does not adequately constitute assessment of quality of life. The ConQol will provide a useful addition to the tools available for both clinicians and researchers to evaluate quality of life and the burden of symptoms in children with cardiac disease. The authors have made a significant contribution to the literature in establishing the reliability and validity of this well-developed instrument.

Majnemer, Limperopoulos, Shevell and their colleagues from Montreal Children's Hospital[14] present an assessment of quality of life and stress of parenting in a cohort of children who underwent open heart surgery during infancy, and have now reached approximately 5 years of age. Assessment of preoperative, as well as both early and later postoperative neurologic function, was used in this analysis as a variable in a regression model to predict quality of life and the outcomes for the parents in terms of stress. Parents of 49 of 94 potentially eligible subjects completed well-established instruments evaluating quality of life and parental stress, the Child Health Questionnaire and the Parenting Stress Index. While the small, heterogenous, sample reported here is a limitation to the generalizability of these findings; it is the unique predictive and correlational analyses that bring merit to this manuscript.

Once again, it was found that severity of disease was not a reliable predictor of quality of life, or the stress of parenting. In general, good quality of life was reported for these children, with means for physical and psychosocial well-being above the norms for the instrument used. Social, emotional, and general health were the areas where deficits were most commonly reported. Developmental delays, and problems with anxiety and attention, were also more commonly reported than in the healthy population. This is consistent with other recent findings in those with congenitally malformed hearts. Interestingly, developmental delays did not result in reports of reduced quality of life. Adaptation to chronic illness in a child is a familial process. Successful adaptation likely results in a readjustment of expectations that are consistent with the potential of the child, and reflect the efforts of the family with coping. The complex relationship between behavioural problems in the child and parental stress is also highlighted. The stress of parenting has been found to be an important modifier of psychological outcomes during childhood. Efforts to modify this stress, and provide parents with realistic anticipatory guidance, may contribute to improved long-term outcomes for both the child and family. The authors of this manuscript[14] have presented a well designed, comprehensive follow-up study. Their analyses will help guide future assessment of risk factors and psychosocial outcomes in children and families living with congenital cardiac disease.

In summary, both manuscripts assessing quality of life in this issue of the journal[12,14] offer important and unique information to our understanding of outcomes for children with cardiac disease. Cardiology in the Young has established itself as a source of quality information on psychosocial outcomes in this population. This research goes beyond the traditional medical model of measuring predetermined quantifiable variables, and instead relies on the fundamentally idiosyncratic responses of individuals. This inherent subjectivity and unpredictability make this program of research ceaselessly interesting

and uniquely challenging. While we may never completely understand what factors determine the perceptions of the individual of the impact of congenital cardiac disease, guidance for practice implications and future research are explicit. We are mandated to continue to explore these important outcomes in order better to understand the impact of cardiac disease in children. Only then can we begin to design interventions and programmes for treatment that optimize quality of life and psychosocial functioning for our patients. In short, we must look beyond survival to the outcomes that our patients value most. Only they can tell us what really matters.

References

1. Moons P, Van Deyk K, Budts W, De Geest S. Caliber of quality-of-life assessments in congenital heart disease: A plea for more conceptual and methodological rigor. Arch Pediatr Adolesc Med 2004; 158: 1062–1069.
2. Casey FA, Craig BG, Mulholland HC. Quality of life in surgically palliated complex congenital heart disease. Arch Dis Childhood 1994; 70: 382–386.
3. Casey FA, Sykes DH, Craig BG, Power R, Mulholland HC. Behavioral adjustment of children with surgically palliated complex congenital heart disease. J Pediatr Psychol 1996; 21: 335–352.
4. Miyamura H, Eguchi S, Asano K. Long-term results of the intracardiac repair of tetralogy of Fallot: a follow-up study conducted over more than 20 years on 100 consecutive operative survivors. Surg Today 1993; 23: 1049–1052.
5. Elkins RC, Knottcraig CJ, McCue C. Congenital aortic valve disease – improved survival and quality of life. Ann Surg 1997; 225: 503–510.
6. Gersony WM, Hayes CJ, Driscoll DJ, et al. Second natural history study of congenital heart defects. Quality of life of patients with aortic stenosis, pulmonary stenosis or ventricular septal defect. Circulation 1993; 87: I52–65.
7. World Health Organization. Constitution of the World Health Organization basic document. Geneva, Switzerland: The Organization, 1948.
8. Gill TM, Feinstein AR. A critical appraisal of the quality of quality-of-life measurements. JAMA 1994; 272: 619–626.
9. Cadman D, Goldsmith C, Bashim P. Values, preferences and decisions in the care of children with developmental disabilities. J Dev Behav Pediatr 1984; 5: 60–64.
10. Bar RD, Pai M, Weitzman S, et al. A multi-attribute approach to health status measurement and clinical management – illustrated by an application to brain tumors in childhood. Int J Oncol 1994; 4: 639–648.
11. Harrison H. Making lemonade: A parent's view of "Quality of Life" studies. J Clin Ethics 2001; 12: 239–250.
12. Macran S, Birks Y, Parsons J, et al. The development of a new measure of quality for children with congenital heart disease. Cardiol Young 2006; 16: 163–170.
13. Wolinsky FD, Wyrwich KW, Nienaber NA, Tierney WM. Generic versus disease-specific health status measures. Evaluation & The Health Professions 1998; 21: 216–243.
14. Majnemer A, Limperopoulos C, Shevell M, Rohlicek C, Rosenblatt B, Tchervenkov C. Health and well-being of children with congenital cardiac malformations, and their families, following open-heart surgery. Cardiol Young 2006; 16: 155–162.