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Quality of life following surgery for congenital cardiac malformations in neonates and infants

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THE PAST TWO DECADES HAVE WITNESSED A MAJOR shift towards repair of most congenital cardiac malformations during the neonatal or infant periods of life.[1] Early anatomic correction or palliation, dramatic improvements in survival, and reduced morbidity due to improvements in perioperative and long-term medical management, have resulted in new populations of children that have reaped the benefits of the best care currently available for treatment of congenital cardiac disease. The impact of the congenital cardiac malformations, however, extends far beyond the walls of the hospital or clinic where we diagnose, treat, and follow our patients. The breakthrough of achieving predictable results with repair or palliation of most lesions during the neonatal and infant periods mandates us to look beyond survival, and to examine the lives our patients lead when they are outside of our care. Our purpose in this review is to discuss the measures of psychosocial outcome that are appropriate for exploration in those neonates and infants who survive cardiac surgery, to explore what is known about the psychosocial outcomes and quality of life for these patients, and what needs exist for future research.

Measuring psychosocial outcomes

There are several different psychosocial outcomes of importance, including quality of life, psychological and behavioural functioning, impact on the family, effects on siblings, and issues of parenting. Exploring these psychosocial variables may contribute unique information to understanding the overall outcome in different groups of patients, or in comparing different strategies of treatment, for example, surgical palliation as opposed to transplantation for those born with hypoplasia of the left heart. Also, asking questions about psychosocial functioning may identify important issues in the child or family that are not readily apparent in our routine clinical evaluations.

Consistently, research has found that the objective clinical condition of the individual patient, or its medical severity, does not reliably correlate with his or her subjective perception of psychosocial functioning or quality of life.[2,3] Quality of life, and psychosocial outcomes, are inherently subjective in nature, and are influenced by many more factors than clinical condition alone. Individual, familial, and sociocultural influences all play a role in how an individual perceives the impact of a given disease.[4,5] Unlike cardiac function, which can be measured fairly exactly, or exercise tolerance, where we can ask someone what he or she can do, and then verify it on the treadmill, when assessing subjective variables like quality of life, we must ask the subjects directly how they are doing. We have no way quantitatively to verify their satisfaction with life. In addition, we must take care not to impose our own judgments of value on their experience.

Subjective reports of quality of life, and/or health-related quality of life, have been used as a representation of overall psychosocial functioning in children with chronic conditions.[6] Although quality of life has been approached from multiple theoretical perspectives, it is generally agreed that it is multidimensional in nature, and includes physical, mental, and social components.[7] It is inherently subjective, and should be measured from the perspective of the subject, or else from the perspective of a reliable proxy. It is dynamic, changing in response to the physical and social demands associated with age, and/or the

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desires, of the subjects themselves. The definition given by the World Health Organization for health[8] as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity" has guided most research on subjective variables such as quality of life. In the past decade, many instruments have been designed to measure quality of life in general samples of children, and in populations of children with acute or chronic illnesses.[6,9] These instruments typically measure several dimensions, including global quality of life, and function in the physical, emotional, social, and educational domains.[10] Measures exist that are both generic and disease-specific. Generic instruments offer the benefit of a global assessment of quality of life, and permit comparisons to be made with the general population. Disease-specific instruments are designed to measure quality of life with a specific focus on areas of concern, which may be unique to a given disease.[11] A combination of both types of instruments likely offers the most comprehensive assessment of quality of life. Recently, surveys of quality of life have become available that are specific to congenital cardiac disease. Uzark et al.[12] have published the psychometric properties of the Cardiac Module of the Pediatric Quality of Life Inventory™, and the Pediatric Cardiac Quality of Life Inventory[13] is currently undergoing validation in a multicentric international study.

Research in congenital cardiac disease

Several excellent studies have presented research on quality of life, and/or psychological and behavioural functioning, in subjects with congenital cardiac disease. The majority of these have been single centre studies, and most have represented samples of children from heterogenous diagnostic backgrounds. To date, there are very limited reports on psychosocial outcomes from large multicentric samples of patients, and quality of life has not routinely been incorporated as a measure of outcome in randomized clinical trials involving patients with congenital cardiac malformations. Although there have been inconsistencies in the findings, much can be learned from the extant literature. In several studies, samples of both children and adults with congenital cardiac disease have reported that quality of life is not significantly different from their healthy peers.[14–18] Other studies have reported quality of life to be significantly lower than healthy controls, particularly in the areas of physical, social, and overall functioning.[12,19–21] Factors affecting the interactions of the children with their peers and family members, such as restrictions of activity, and altered relationships, have been found to have the greatest impact on the general state of health.[22] Adolescents with congenital cardiac disease have also displayed positive psychosocial attributes, including lower situational anxiety, and higher superego strength.[23] Exploration into the impact on the family of congenital heart disease has revealed higher levels of family stress, financial strain, and a higher than normal incidence of depression in parents.[24–26] Social support, resiliency, and degree of parental stress have been found to be important moderators of the way both the child and the family adjust to living with congenital cardiac disease.[27–29]

A study is in progress at Children's Hospital of Wisconsin better to understand quality of life, and functional outcomes, in survivors of neonatal and infant cardiac surgery. Preliminary results are presented here. This is a mailed survey study approved by our Human Research Review Committee. To evaluate health-related quality of life, we use the generic module of the Pediatric Quality of Life Inventory™ Version 4.0,[10] which allows for parental reporting on subjects aged from 2 to 18 years, and subjective reporting for those aged from 8 to 18 years. Functional status is evaluated using the Health Utilities Index Mark II.[30] Parents are asked to report on the quality of life and functional state of their child over the past thirty days as seen from the perspective of the child. The Impact on the Family Scale[31] is used to explore five dimensions of influence of the disease, including financial, social, personal strain, mastery, and effects on siblings. Both positive and negative outcomes are examined. Socioeconomic status is calculated using Hollingshead's 2-factor index, which includes parental education and occupation. Age at time of survey, gender, number of open-heart operations, and time since the last operation, are all included as variables.

To date, 182 parents of children with congenital cardiac malformations have completed the surveys. The mean age of the children represented was 8.7 ± 5 years, with a range from 2 to 18 years, and two-thirds of the group was male. The respondents represent three major diagnostic categories, 54 having transposed arterial trunks, 55 a functionally single ventricle, and 73 complex variants of functionally biventricular disease (Fig. 1). On average, each patient had undergone 2 ± 0.9 open-heart operations, with a range from 1 to 5. All subjects had their first cardiac operation during infancy. The mean time from the last cardiac operation to the time of responding to the survey was 3.7 ± 2 years, with a range from 0.4 to 9.1 years.

The Pediatric Quality of Life Inventory™ provides norms for reference samples of healthy children, and children with other chronic conditions, such as diabetes, orthopaedic problems, and arthritis.[10] Parents

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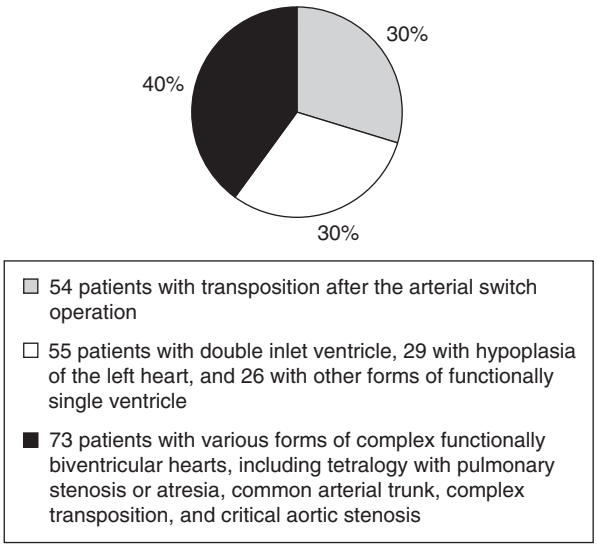

Figure 1.
Diagnostic categories of the 182 patients whose parents responded to the Pediatric Quality of Life Inventory™.

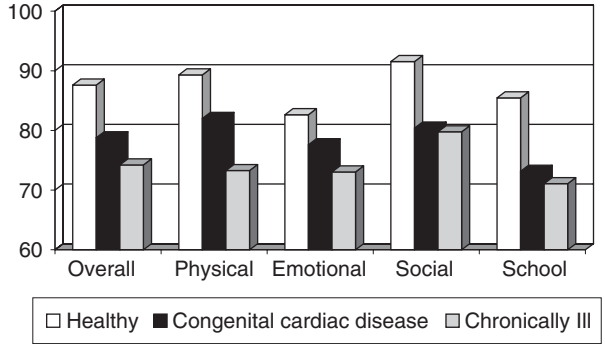

Figure 2.
Parents of 182 children with congenital cardiac disease reported quality of life that was significantly lower than healthy controls on all of the subscales of the Pediatric Quality of Life Inventory™ ($p < 0.001$). Compared to children with other chronic conditions, overall, physical, and emotional quality of life was better ($p < 0.01$), social and educational quality of life were not significantly different for children with congenital cardiac disease.

reported quality of life for their children that was significantly lower than healthy controls on all of the subscales of the instrument: overall, physical, emotional, social, and educational functioning ($p < 0.001$). Scores were higher than children with other chronic conditions on overall, physical, and emotional functioning ($p < 0.01$), but did not differ from the chronically ill sample on social or educational functioning (Fig. 2). Reports of quality of life were not correlated with socioeconomic status, number of open-heart operations, or the time since the last operation. Analysis of age at the time of survey revealed a small but significant negative correlation

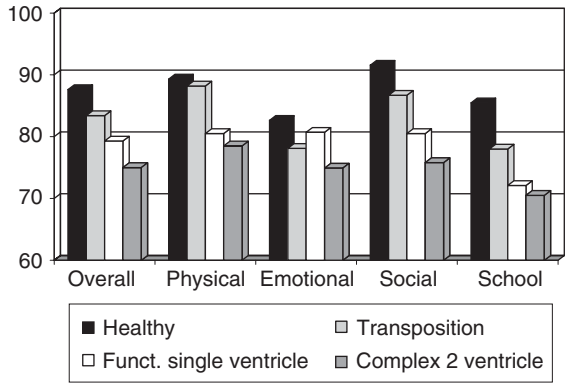

Figure 3.
Reports of quality of life compared by diagnostic category. The group of patients with complex functionally (funct.) biventricular hearts (Complex 2 ventricle) reported the lowest scores for quality of life on all the subscales of the Pediatric Quality of Life Inventory™. Results were significantly lower than those reported by the group for simple transposition in terms of overall, physical, and social functioning ($p < 0.05$).

with all of the subscales of the instrument ($r = -0.2$–0.3, $p < 0.01$). When assessed according to age, those aged from 8 to 12 years consistently reported the lowest quality of life across all of the domains assessed. This span represents a time when surgical reintervention is common, and also a time when children are first beginning to consider their own mortality and vulnerability.

Interestingly, when comparisons were made between the three diagnostic groups, those with complex functionally biventricular hearts, including subjects with tetralogy of Fallot and pulmonary stenosis or atresia, common arterial trunk, complex forms with discordant ventriculo-arterial connections, and aortic valvar disease, consistently reported the lowest scores for quality of life. These were significantly lower than the patients with simple transposition on overall, physical, and social functioning ($p < 0.05$ – Fig. 3). When the sample was divided by quartiles, the subjects reporting the lowest quality of life, the lowest quartile, were found to be significantly more likely to report functional disabilities on the Health Utilities Index in all domains assessed, specifically mobility, cognition, sensory, self-care, emotional function, and pain (all $p < 0.001$ – Table 1). Familial impact was reported as less negative than that of a reference sample of children with other chronic conditions reported by Stein and Jesop.[31] The areas identified as having the greatest negative impact on families were financial concerns, and mastery of the needs of their child for care. Parents of children with hypoplasia of the left heart reported the greatest impact, significantly worse than parents of children with simple transposition for total impact,

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Table 1. Subjects reporting quality of life in the lowest 25th percentile were significantly more likely to report functional disabilities on the Health Utilities Index.

	% of subjects reporting some disability	
Results for Health Utility Index	*Lowest* reports on quality of life (≤25th percentile)	Reports on quality of life (>25th percentile)
Mobility	75*	19
Cognitive	65*	21
Sensory	46*	17
Self-care	25*	4
Emotion	45*	15
Pain	45*	11

*Chi-square; $p < 0.001$

effects on siblings, personal strain, social restrictions and financial impact ($p < 0.05$).

Discussion

Exploration of psychosocial outcomes in children with cardiac disease has the potential for several limitations, as well as issues of methodology. All of the results cited here have been collected under the auspices of research. Inherent to the nature of research is that participation is voluntary, and there is a definite possibility that those who choose to participate in research may be different from those who do not, thus introducing an element of bias for the sample. There is also the possibility of Hawthorne effects, this being the phenomenon of the research subject telling us what they think we want to hear, and responding with what they believe to be the most socially desirable answer. Subjects and parents may even fear judgment by the medical community or society if they were to report negative outcomes.[32] Questions have also been raised concerning the use of parents as proxies in reporting the quality of life of their children.[6,9] For young children, however, this may be the only viable option. The subjective nature of these variables also makes difficult both generalization and interpretation.[7,11]

Despite these limitations, the findings to date demand attention, and indicate that there is the potential for both positive and negative psychosocial impacts when congenital cardiac disease is repaired in infancy. Reliable and valid instruments are available systematically to measure these outcomes, and permit comparisons to be made with both healthy and diseased populations.[7,9,10] The research to date demonstrates that these instruments are easy to use, and acceptable both to subjects and their parents. Professionals involved in health care should take care to avoid making any assumptions about quality of life or other psychosocial outcomes based on clinical assessment alone. Severity of disease does not predictably correlate with psychosocial outcomes. It is not safe, therefore, to assume that children with minimal symptoms have adapted well to their cardiac disease, nor that children with symptomatic cardiac disease will necessarily report negative outcomes. Intrapersonal factors, such as temperament, previous experience with stress, and skills of coping, as well as the influence of family, society, religion, and culture surrounding any individual child will have a huge effect on how the impact of the cardiac disease is interpreted.[4,5,28,33]

In the sample presented here, parents reported quality of life for their children which was significantly lower across multiple dimensions than healthy controls. The responses, however, were as good, or better, than those reported by children with other common chronic conditions. Interestingly, the greatest negative impact on quality of life was reported in the areas of social and educational functioning, despite the perception that congenital cardiac disease primarily has physical effects. Psychosocial sequels are being increasingly recognized as one of the greatest areas of long-term morbidity in children with complex congenital cardiac malformations.[34,35]

Quality of life is dynamic, and requires longitudinal assessment to measure the impact of changing physical and social demands as this complex group of patients progress through childhood and into adulthood. The negative correlation between quality of life and increasing age demonstrates that the impact of the cardiac disease is perceived differently at different developmental stages. The need for reintervention in later childhood, such as surgical replacement of valves, evaluation of arrhythmias, or changes in regimes for medication, may represent the first time the child perceives him or herself as having a chronic condition, or being different than healthy peers. Regardless of the complexity of the intervention, we must acknowledge the stress it poses to the child and his or her family.

In our sample, functional disability was found to be strongly correlated with quality of life. Efforts to reduce morbidity, and maintain an optimal functional state, therefore, may have very positive effects. Restrictions on social and physical activity should be carefully considered, as they also have the potential for significant negative impact on satisfaction with life, particularly in the population of adolescents.[22] Families face several issues of concern related to having a child with congenital cardiac disease, particularly social and financial burdens. Anticipatory guidance for parents and families may help to reduce these problems, and promote effective coping and growth as a family.[4,33]

These facts mandate the systematic assessment of multiple short and long-term outcomes for children

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and families living with congenital cardiac disease. This ongoing follow-up should include both objective clinical parameters, and subjective psychosocial variables. Quality of life, psychological and behavioural function, impact on the family, and issues of parenting, are measures that can and should be included in our routine follow-up of children with congenital cardiac disease. We need this information not only to provide realistic counselling to new and existing families, but also to add an important measure to how we assess the impact of our interventions for these children.

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