

Quality of Life and Congenital Heart Disease in Childhood and Adolescence

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Abstract

Advances in cardiac surgery techniques and early diagnosis have enabled the increased survival of individuals with congenital heart disease. The investigation of the quality of life in children and adolescents with congenital heart disease provides complementary information to clinical data that can assist in decision making on the part of health professionals. Although many studies have been conducted to investigate the quality of life of children and adolescents with congenital heart disease, the results prove to be contradictory; while some studies show that congenital heart disease can impact the quality of life, others describe a better perception of quality of life among children and adolescents who suffer from the disease when compared with healthy control subjects. The purpose of this study is to review the literature on the assessment of health related quality of life in children and adolescents with congenital heart disease, in order to systematize the existing knowledge on this topic today. It is observed that research seeks to investigate aspects of personality in cardiac patients, their coping strategies used and perceived social support, aiming at better understanding the association of these variables with the level of quality of life in this population.

Introduction

Advances in treatments and the effective possibilities of control of chronic-degenerative and congenital diseases have resulted in longer life time to people afflicted by these diseases¹. In relation to cardiac malformations, it is estimated that about 10 out of 1000 newborns alive are affected by some sort of congenital anomaly, being one-third of these with critical diagnoses requiring surgical intervention^{1,2}. In the last decades, early diagnosis has contributed to the increase of incidence in the findings³, and the advance of surgical techniques has led to the increase in the number of heart disease children and adolescents.

Keywords

Quality of life; heart disease; congenital abnormalities; childhood; adolescence.

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In face of this context, it is important to consider the repercussions of physical, psychological and social order that accompany congenital heart disease (CHD) and that can bring losses to these patients' quality of life. The Health related quality of life (HRQoL) initially investigated further in the adult population, has gained special attention in childhood and adolescence due to the impact that its injury can cause in long-term, in addition to the benefit that early interventions focusing on its improvement can provide, especially in chronic diseases.

The assessment of HRQoL in children and adolescents with heart disease has been used as an important health outcome, to identify groups and subgroups that are at risk, track health habits and to better understand the impact of different diagnoses of heart disease on the quality of life of this population. The objective of this study is to review the current literature on the evaluation of health related quality of life in childhood and adolescence, with emphasis on the patient with congenital heart disease.

Method

A search was conducted in Pubmed and Scielo databases for articles published between 2000 and 2012. The following key-words were used: "quality of life", "congenital heart disease", "children", "adolescents". 350 articles were found, of which 58 were selected for this review according to the following selection criteria: cross-sectional studies, longitudinal and systematic reviews, whose methodological procedures were well described, as well as statistical analysis, studies using measurement instruments valid and recognized; studies performed with children and adolescents with significant sample size. Additionally, references were sought in books authored by specialists in the field of literature review articles with adequate theoretical quality for the discussion of the topic, in order to complement and deepen the analysis of the current literature.

Review results and discussion

Health related quality of life in children and adolescents

While the quality of life (QoL) in adults has been investigated in the past decade, this study among children and adolescents is a recent area. The difficulty of defining and conceptualizing QoL becomes a constant challenge for researchers in this area, especially in the construction of valid and reliable instruments, including all its scales and considering the perception of the individual under development.

In general, the QoL assessment tools in childhood have little congruence in relation to the content covered, with different conceptions about normal childhood development in different age groups and the importance of family function and social context as factors necessary for their well being. The broad spectrum of contents and the variety of scales and items envisaged in the QoL instruments in childhood reflect the differences in their process of preparation, in the theoretical line used, in the target population and in the objectives of application of the instrument^{4,5}.

Some areas of discussion of an ideal model of QoL in childhood and adolescence, as highlighted by Kuczynski⁴, suggest that it is important to assess the global sphere (related to society and its macro environment) and external sphere (related to socioeconomic conditions) on top of personal and interpersonal aspects as children need, overall, to develop themselves in adequate and health-promoting conditions. Reinforcing this idea, Matza et al.⁶ consider that in general children do not have the choice to significantly change an environmental problem, unlike adults, who have financial resources and adequate social support, for example, to leave a job or an unsatisfactory marriage.

Contextual factors, therefore, may have an influence in the long-term on social and psychological development of children, and it is important to consider variables such as family, peers, school and community when assessing their QoL. In the case of children with chronic disease, there are few attempts at developing methods and tools for assessment of HRQoL, and it still is uncommon concern in apprehending the perception and impact of the disease from the point of view of the patient, resulting in large discrepancies as to the response and clinical evolution⁴.

To Lawford and Eiser⁷, the child's ability to adapt to their experiences and how they interpret adverse experiences will have impact on their QoL. The reactions of each child to face adverse experience, such as dealing with the disease, show that there is a level of individual adjustment and coping styles that are related to their perception of QoL. The concept of resilience, understood as the process of positive adaptation of the individual in contexts of extreme adversity, has been used to explain individual differences found in children who show high levels of action in some domains of QoL despite facing adverse life conditions.

In a systematic review of instruments to assess the QoL for children and adolescents, Solans et al.⁵ have identified the existence of 30 generic and 64 specific-disease instruments, published between years 2001 and 2005. Samples of generic instruments for pediatric population are: "Pediatric Quality of Life Inventory (PedsQL)⁸", "Autoquestionnaire de Qualité de Vie Enfant Image" (AUQUEI)⁹, "Child Health Questionnaire (CHQ)¹⁰ and Kidscreen¹¹". The specific measures are tools capable of detecting the improvement or worsening of symptoms and functioning of the health condition under study, with higher sensitivity to measure changes to effects of the treatments of certain diseases^{12,13}. In the pediatric population, health conditions that have a higher number of specific measuring instruments are asthma, cancer and epilepsy, and the countries that mostly develop this type of instrument are the United States, UK and Canada⁵.

Although the study of Solans et al. where most of the instruments present acceptable standards in relation to psychometric properties, few presented criterion validity analyses ($n = 5$), structure validity ($n = 15$) or sensitivity to change ($n = 14$). The challenge in accessing child perception is also revealed in the study, since 26% of the disease-specific instruments are destined exclusively to the parents as respondents (Proxy)⁵.

With regard to measures of HRQoL for children and adolescents, Matza et al.⁶ point out some methodological aspects that must be considered in the preparation of the instruments. According to the authors one must consider the minimum age at which children can answer questions about their quality of life as well as the appropriateness of the format of the instrument to the level of understanding of each age group. In addition, there is disagreement among researchers about the need to evaluate the agreement between the assessment of parents and that of children as respondents, especially when evaluating younger children who may not be able to respond on more subjective domains. Thus, before drawing a measure of HRQoL for children, it is essential for researchers to assess the adequacy of the tool to age, including vocabulary, instructions, question structure, content and response options.

In congenital heart disease, specific HRQoL measures have begun to emerge recently and are still in the process of being tested and validated in different cultural contexts. In the UK, the development of ConQoL Index (CQI) occurred from a multicenter study involving 730 children from five pediatric cardiology centers, funded by the British Heart Foundation. It is an instrument to measure HRQoL for children with congenital heart disease at the age of 8 to 16 years old, and seeks to assess the impact of heart disease on daily life measured from the perspective of the child or adolescent. Two versions are available for different age groups: the first, for children 8-11 years old, it includes the scales, symptoms, activities and relationships, and second, for teens 12-16 years old, it adds a scale to the others on coping and control¹⁴.

In the United States, the project of creating the Pediatric Cardiac Quality of Life Inventory (PCQLI) aimed at the development of an inventory covering a wider age group - children 8-12 years old and adolescents aged 13 to 18 years old - including versions for patients and parents (Proxy), having the ability to discriminate the different types of congenital and acquired heart diseases. The results of the pilot study, conducted in three cardiac clinics with 655 pairs of parents and patients, showed content validity, good internal consistency and construct validity of the instrument, with a good correlation between the components that measure the impact of the disease and the psychosocial impact¹⁵.

In Brazil, Soares et al.¹⁶ analyzed the national scientific production on QoL of children and adolescents, finding 30 articles published between 1990 and 2008. The results showed predominance of quantitative approach research (70%) and a higher index of research with quality of life evaluation instruments related to health (66.7%) than generic instruments (28.6%). It was also observed a greater number of articles published in the field of Medicine

(40%), followed by Nursing (30%) and Psychology (26.6%). Regarding the assessment of QoL in chronic diseases, Brazil stood out in pioneering studies conducted by the Faculty of Medical Sciences, State University of Campinas (UNICAMP) on QoL and epilepsy, which served as a starting point for the advancement of discussions on the theme.

Although 50% of national publications found in the study by Soares et al.¹⁶ the QoL of children and adolescents from the perspective of their parents and caregivers, the authors concluded that the scientific community is looking to enhance the perception of children and adolescents as reporters of their experience, considering the particularities of this period of life in the application of adequate research methodologies. Despite all the difficulties encountered in developing tools for the pediatric population, it is important to remember, as pointed out by Kuczynski¹⁷, that "the individual under development can express himself, it is we who cannot (or are not willing) to understand him."

Congenital heart diseases in childhood and adolescence

Cardiac malformations are considered one of the most frequent forms of congenital anomalies at birth. Studies performed in the Brazilian population evaluated the predominance of CHD in live newborns, identifying the ratio of 5.5:1000 live newborns between the years of 1889 and 1998 in the state of Paraná¹⁸, and 9.58:1000 newborns between the years of 1990 and 2003 in the state of Minas Gerais¹. To Huber et al.³, early diagnosis and detection of cardiac malformations in the prenatal period have contributed to the increase of incidence in the findings, both in national and international studies.

In childhood, there is evidence that the presence of CHD can affect physical and ponderal development, motor, cognitive and neurological operation^{19,20}, and there is even a higher incidence of academic difficulties, behavioral problems, speech delay, lack of attention and hyperactivity in patients with complex congenital heart disease^{21,22}. In addition to such clinical evidence, the need for ongoing medical monitoring, use of medication and recurrent hospitalizations may affect self-esteem and self-image of this group of children and adolescents, resulting in delays in normal development tasks²². The maintenance of health in children with CHD still requires care from parents and medical staff with nutrition, immunization, winter disease prevention and prophylaxis of bacterial endocarditis, all these aspects vital to their long-term survival²³.

Upon entering adolescence, patients with CHD face the challenge of autonomy in the face of parental overprotection, usually exercised during childhood. Furthermore, adolescence is a crucial consolidation phase of health healthy behavior, such as for instance, the adoption of good food practices and exercises, but also a period in which risk behavior is manifested for the first time, such as the use of drugs and alcohol and risk sexual practices^{24,25}. Some studies highlight the importance of a transition program in assisting health^{22,26-28}, with the objective to educate adolescents with CHD on their medical condition and guide them with regard to physical exercises^{29,30}, sexuality³¹ and entrance into the labor market.

Some qualitative studies conducted with adolescents with CHD^{32,33}, using semi-structured interview and in-depth techniques, pointed out some difficulties faced by these patients, such as: coping with the illness and physical limitations, social exclusion, discrimination and bullying, in addition to the challenge of normality, becoming independent, the uncertainty about the future and how to use coping strategies to develop self-esteem, among other dilemmas. Thus, health care to patients with congenital heart disease should include the care of their psychological and social needs as well as the physiological, as a means of improving their quality of life.

Although many cardiac patients acquire a stable health condition, some of them have residual defects and important sequelae in adulthood, even after numerous surgical corrections. CHDs are considered chronic conditions, due to the factors that accompany them in the long term and that interfere with the daily life of these patients, including uncertainty about the course of the disease, prognosis, signs and symptoms, and restrictions on physical activity³⁴. The investigation of the quality of life in this population in all age groups, has gained increased attention as an important health outcome, not only related to symptoms and clinical conditions, but also in terms of their level of well-being and satisfaction with life as a whole.

Congenital Heart Diseases and Quality of Life

Studies conducted to evaluate the QoL of children and adolescents with CHD have yielded inconclusive results, possibly due to a lack of conceptual and methodological rigor in research, according Moons et al.³⁴. Methodological differences among the studies, such as outline, inclusion criteria, evaluation tools used, follow-up and outcome measures make it difficult to compare results.

However, one can ascertain that a large part of the studies performed is at a lower level of HRQoL among children and adolescents with CHD when compared to healthy control³⁵⁻³⁷. According the findings of a systematic review conducted by Dahan-Oliel, Majnemer and Mazer³⁸, such results are related in part to the physical limitations - actual or perceived - in subjects with CHD, which can lead to unnecessary restrictions and overprotective behavior that possibly interfere in their perception over QoL.

Janiec et al.³⁵ report that children and adolescents with mitral valve prolapse have a lower QoL than the healthy population only in the scale of physical well-being, although among the group with CHD the frequency and severity of clinical symptoms have a negative impact on other scales assessed. The study of Krol et al.³⁶ found that children with CHD had worsening in motor functioning and autonomy compared to healthy children; however, the severity of the diagnosis was not associated with the level of QoL of the heart disease population.

In the study by Uzark et al.³⁷, one in five children with CHD refers significant worsening in psychosocial functioning when compared to the control group, including those with mild or corrected heart disease; on the other hand, in the CHD group, children with more serious injuries presented

lower results only in physical function, and there is no relation between the disease severity and psychosocial functioning. These findings show that the association between severity of diagnosis and QoL has not yet been elucidated among children and adolescents with CHD, possibly due to the methodological differences mentioned above.

Studies by Landolt et al.³⁹ and Spijkerboer et al.⁴⁰ investigated the QoL of children and adolescents after cardiac surgery and/or invasive treatment by catheterization for CHD, respectively, using the same measuring instrument. Their results were similar, indicating worse motor cognitive, social and emotional functioning than healthy children. Multivariate analysis in the study Landolt et al.³⁹ further disclosed that the duration of cardiopulmonary bypass, time of hospitalization, need for medication and adverse family relationships had a negative impact on QoL of the children, only in accordance with the evaluation of their parents and caregivers.

In the systematic review conducted by Latal et al.⁴¹ studies published between 1990 and 2008 on psychological adjustment and QoL in children and adolescents submitted to cardiac surgery were included in order to check the current knowledge on the outcomes in this population. Although there is great variability between studies, the results show that QoL is impaired in some children, especially those with more complex CHDs. The results also show that there are differences in the perception of parents and children regarding the psychological adjustment: while children are evaluated positively in this aspect, parents in general consider that psychological adjustment of their children has impairments, relating them mainly to the severity of the CHD and developmental delays.

Other studies corroborate and expand these findings, indicating that the QoL of parents and caregivers of children with CHD can also be affected due to the impact of the diagnosis on the level of stress and adjustment of these families⁴²⁻⁴⁵. Current literature on QoL of children with special health care needs indicates important factors of positive adaptation to motivation and attitudes of the child as well as the resources and social support available to the family. Such factors are considered more important predictors for a good QoL than the complexity of the medical condition, so strategies that strengthen family welfare and building of social support networks should be valued and offered in health care services⁴⁵.

Contrary to studies that showed impairment of QoL in children and adolescents with CHD, some authors found surprising results. Teixeira et al.⁴⁶ evaluated adolescents and young adults with CHD and their findings indicated a level of QoL higher than the general population, especially in the environmental and social relationship scale. Culbert et al.⁴⁷ studies evaluated the QoL of 306 children diagnosed with Transposition of the Great Arteries (TGA) using the Child Health Questionnaire, and found significantly higher results between the group with CHD when compared to the general population, with the exception of self-esteem scale. Other studies found a small difference between the QoL of congenital heart disease patients, when compared to population standards⁴⁸⁻⁵⁰.

Silva et al.⁵¹ investigated the variables that would have a negative impact on the Quality of Life in adolescents and young adults with CHD, applying the WHOQoL-BREF tool, and found a better perception of QoL in this group when compared to the general population. The presence of cyanosis was not associated with worsening of QoL; however, the number of surgical procedures and moderate to severe residual lesions had a significant impact on it. It was also highlighted, among the findings, the importance of social support as a variable related to increased resilience among heart disease patients, which consequently promotes better adaptation to the disease.

But how to explain these findings? Surprised by research that found a higher QoL in CHDs, Moons et al.⁵² introduced the term "sense of coherence" as a hypothesis to understand why patients that grow with chronic diseases may be satisfied with their life. The term Sense of Coherence (SOC) was developed by Antonovsky in 1987 to explain why some people get sick when they are under stress, while others remain healthy. Contrary to the pathogenic model with focus on disease, it was based on the premise that the individual should focus positively on his resources and capabilities to maintain his health and well-being.

So, the term Sense of Coherence can be defined as an expression of the world view of the individual, a global orientation based on a dynamic and enduring feeling of confidence in themselves and in their environment. It consists of three components: 1) understandability, or how much the internal and external stimuli received during childhood are structured, predictable and explicable; 2) management, which refers to the perception of the resources available to deal with stressful situations, such as for instance living with heart disease and 3) meaning, or the ability of the individual to believe that his life has meaning, find motivation and assume the control over his life⁵².

In a longitudinal study performed by Apers et al.⁵³, with 429 adolescents with congenital heart disease, the Sense of Coherence showed up as a positive predictor of all domains of perceived general health (physical, emotional, social and school functioning), also positively associated with specific aspects of the disease, such as symptoms, physical appearance and cognitive problems. In another study on the individual and contextual determinants of quality of life in adolescents with heart disease, Luyckx et al.⁵⁴ concluded that the perceived level of health, the Sense of Coherence and parental support correlated positively with quality of life over time. These results confirm the importance of considering the biopsychosocial functioning of the adolescent with heart disease in the evaluation of their QoL, and the Sense of Coherence emerges as a valuable resource to be developed.

Studies performed by Luyckx et al.⁵⁵⁻⁵⁷ have advanced in the investigation of QoL as a variable of the outcome of other predictors, such as perceived parental styles⁵⁵ and the process of formation of identity of the adolescents with CHD⁵⁶, to explain their functioning and describe the main variables that interact in their perception of QoL.

With regard to parental styles, no significant differences were found between adolescents with heart disease and

the control group. The democratic style obtained better results among adolescents with heart disease, while the controlling style showed the worst among the evaluated outcomes (depressive symptoms, loneliness, quality of life, health status, and substance abuse)⁵⁵. The process of identity formation in adolescents with heart disease proved to be similar to the one of control subjects, in a standard within average. Individuals with a strong sense of identity presented better results in terms of quality of life and psychosocial functioning, while those with a diffuse sense of identity had higher scores on depressive symptoms and loneliness, and lower in quality of life⁵⁶.

It is possible that children with chronic diseases, which include children with cardiac disease, are challenged to develop a Sense of Coherence earlier than healthy children, by experiences that derive from the disease and require a great capacity for adaptation. Growing with CHD requires attention to medical treatment and often living with some restrictions on activities that are part of any child's life. Perhaps because they do not know a different reality these children acquire a greater sense of appreciation for life and expectations consistent with their capabilities and limitations, which will influence their perception of QoL in the course of their development.

Conclusion

The number of studies on quality of life in children and adolescents with congenital heart disease has increased in recent years due to increased survival in this population.

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Studies show conflicting results, and currently, there is a tendency to investigate factors such as parental styles, social support and coping strategies to better understand the quality of life in these patients. All of these variables that interact in the perception of quality of life are difficult to grasp by a single measuring instrument.

Author contributions

Conception and design of the research and Writing of the manuscript: Bertoletti J, Pellanda LC; Acquisition of data, Analysis and interpretation of the data, Statistical analysis and Critical revision of the manuscript for intellectual content: Bertoletti J, Marx GC, Hattge Jr SP, Pellanda LC; Obtaining funding: Pellanda LC.

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