

# QUALITY OF LIFE OF CHILDREN WITH CONGENITAL HEART DISEASE

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**Abstract:** Progress in cardiovascular surgery led at about 85% of children with congenital heart disease to survive at the adult age. Increased survival rates of patients with congenital heart disease, through improved medical results, has led to intensifying concerns for quality of life of children with this diagnosis. Based on a literature review, the paper describes the main factors influencing the quality of life of children with congenital heart defects and also brings into question the necessity of evaluating the health of children with this medical condition in order to substantiate ways of treatment and measures to increase the quality of life of children and their families.

## INTRODUCTION

Health status is one of the key elements of human welfare, enabling people to perform their duties and to be active members of the society. Good health can bring a number of benefits to individuals through access to education, to the labour market, increase productivity and implicitly, of welfare, cost reduction with medical care, better social relationships and a longer high quality life. Thus, health is one of the 11 dimensions of the Better Life Index (*OECD Better Life Index*), along with housing, income, employment, social support/community, education, environment, civic engagement, life satisfaction, safety and work- life balance.(1)

In the quality of life researches (*Quality of Life - QoL*), there can be distinguished two perspectives on health. The first perspective deals with health as an essential element of quality life, analysed on the basis of the assessments of individuals over their health status, access to health services and their evaluation. The second approach, aimed at the quality of life from the perspective of health (*Health Related Quality of Life - HRQoL*), focuses on how patients with different medical conditions disease experience their quality of life, on their perceptions or survival time of the beneficiaries of medical treatment.(2)

The importance given to studies of quality of life from the perspective of health has increased as a result of certain diseases chronicity with effect on the lifestyle of patients. At the same time, increased survival rates of patients with congenital heart disease (CHD), surgically corrected or not, prompted intensifying concerns for their quality of life, and in particular to children diagnosed with this medical condition, in order to identify ways of treatment, as well as preparation of patients, relatives and even medical or social services to potential difficulties during the life of patients. Under these circumstances, studies on the quality of life of patients with CHD also focus on how certain clinical or medical procedures changes affect the lives of patients.(3)

Congenital heart disease are some of the most common congenital anomalies, representing an increased cause of morbidity and mortality, particularly during the first year of life.(4) In Romania, the actual incidence of CHD is not properly

assessed due to underreporting and lack of epidemiological data on these diseases at national level, which makes even more necessary the health assessment of children with CHD. Study of this problem worldwide has been approached since the 1980s and today, the majority of congenital heart diseases are diagnosed in infancy. This period is the most important, given that approximately 70% cases of CHD can be diagnosed, by the age of one month and 90% of cases by the age of one year.(5) Some patients may fully recover, but others require reintervention, rehospitalisation, therapy and lifelong care. The latter require continuous specialized medical surveillance.

## PURPOSE

This article aims at describing the factors that influence the quality of life of children diagnosed with congenital heart disease and thereby, at providing comprehensive insight on the difficulties they face in order to sustain measures to increase the quality of life for both them and their families.

## MATERIALS AND METHODS

The article is based on specialty literature review regarding the quality of life from the health perspective of patients diagnosed with congenital heart disease, as well as on data on the incidence of this disease among children at European and national level and worldwide.

### The incidence of congenital heart disease

In the United States, approximately 25.000 children are born with CHD, only 60% reaching adult age, due to the nature and severity of the congenital heart disease.(6)

At European level, reports showed an average prevalence of 0.8% of CHD, with variations between different countries. It has been estimated that in the European Union about 35.000 children are born annually with CHD, out of which 3.000 die.(7)

World Health Organization (WHO) statistics on mortality of children under 1 year reveal that in Romania, in 2000, the deaths related to CHD have represented 8.49% of the total 4.370 deaths of children under 1 year of age. In 2002, there was an increase in this indicator, the CHD related deaths

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representing 11.40% of all deaths of children under the age of one.(8)

In 2010, in Romania, a regional register of the CHD in Euro region 7 Center (counties of Alba, Braşov, Covasna, Harghita, Mureş, Sibiu) was implemented. It included 1 249 infants and newborns and showed that CHD diagnosis was late confirmed, around the age of one month (23.59 days) and that the antenatal diagnosis is extremely low (1.52%).(9)

Another study was conducted between 2001-2010 in the north-eastern Romania on a number of 2 211 cases diagnosed with malformations (of the total 62.709 newborns hospitalized during this period). It has identified malformations of the cardiovascular system in the case of 28.9% of the studied patients. Of the patients in the study group, 9.5% died, the highest weight in the number of deaths being occupied by those with cardiac anomalies (53.5%), followed by patients with plurimalformative syndromes (23.3%), central nervous system abnormalities (13%), genitourinary malformations (7%), genetic syndromes (4.9%), thoracic and pulmonary malformations (3.8%), limbs and skeletal malformations (3.2%), malformations of the digestive tract and abdominal wall (2.7%), and other types of malformations (4.9%).(10)

In 2012, the infant mortality rate in Romania was 9.01 to 1.000 live new-borns, lower compared to previous years (15% in 2005), but still recording one of the highest values among EU Member States. Congenital anomalies were placed among the leading causes of death, determining a mortality of 2.1 per 1.000 live newborns, slightly down compared to previous years (2.4% during 2008-2010). Among congenital anomalies, the congenital malformations of the circulatory system were placed first among the causes of death for children under one year in 2012, with a value of 48.4% of total deaths.(11)

Worldwide, congenital heart disease affects 4-12 per 1.000 live newborns. Over one third of affected children are born with critical heart diseases, which indicate malformations requiring palliative or corrective surgery in early life stages.(12)

The literature mentions environmental and genetic factors and the interaction between them, as influencing the incidence of these diseases (e.g. presence of a child with congenital heart disease in the family, congenital rubella, the use of various medications by the mother during the first trimester of pregnancy, maternal mellitus diabetes etc.).(6,13) Depending on the severity of the condition, of the time of surgery, patients with CHD can be divided into six groups, each with special needs:

- patients with minor cardiac malformations that remain asymptomatic until adulthood;
- patients presenting for surgical correction in adulthood because CHD has been balanced naturally or surgically corrected;
- patients presenting for reintervention after correction in childhood;
- patients who require repair of residual defects after surgical correction;
- patients developing heart failure after CHD surgical/palliative intervention or requiring transplant;
- patients developing heart disease acquired in addition to the CHD.

Outstanding progress in paediatric cardiovascular surgery has determined the decrease of infant mortality due to congenital heart defects, so that about 85% of the patients with CHD can now reach adulthood.(14)

### **CHD effects on the children's quality of life**

The results of various studies regarding the quality of life of patients with congenital heart disease are grouped around two main perspectives, determined especially by the severity of

the disorder. On the one hand, there are studies showing that patients with CHD have a poorer quality of life than the general population. On the other hand, there are studies according to which patients with moderate heart disease have similar quality of life to the general population.(3)

CHD effects on the patient differ depending on his/her age: newborn and infant, young child/preschool age, school age, teenager, adult, especially women diagnosed with congenital heart malformation in personal medical history. The development / growth stages – the entry into community (kindergarten, school), the transition to adolescence and the adolescence - involve additional effort and another perception of the disease from both. Surgery can ensure the survival and increased life expectancy of children with such disorders, but not always allows a normal life following postoperatively treatment. The changes in lifestyle of children diagnosed with CHD, generated by the frequency of hospitalization, refraining from certain activities (e.g. physical exercise) or isolation from the usual environment, have a negative impact on their quality of life. Frequent hospitalizations, medication and various limitations imposed by the disease can cause anxiety and depression. Sometimes, these can even affect children's school performance through the numerous absences from school or through the disease effects on learning ability. However, a poor quality of life is often attributed to the lack of social acceptance in schools.(6)

One of the most important factors influencing the quality of life is the delay in physical growth in terms of height and weight, which varies depending on the type and severity of the disease. In the same category of physical factors also falls the postoperative scar, which can be regarded as a "stigma" which indicates disease and may give rise to comments and questions in the environment in which the child lives. Consequently, children can become introverted or choose to isolate themselves and sometimes may need the psychological support of their family or even of a psychotherapist.(6) The adolescents with CHD have an increased risk of developing behavioural problems and to experience a lower quality of life.(15)

Family environment (relationships between family members and with the child) is an essential factor in overcoming illness and for subjective well-being. A positive family environment, characterized by high cohesion, reduced conflicts between family members, social support, makes it easier for the individual to adapt to his/her the disease, thus leading to an improved quality of life. On the other hand, adverse family relations or parental stress are negatively associated with the quality of life.(3) Also, overprotective parental behaviour can reduce children's ability to take care of themselves and at the same time, their self-esteem.

Depending on the moment (prenatally or after birth) in which parents find out the child's diagnosis with CHD, they will be able to gather information, make decisions and seek medical services for supervision and medical intervention in the shortest time possible. Such a diagnosis can cause changes in the professional trajectory of one or both parents and implicitly social-economic changes, which in turn can have a negative impact on quality of life of the whole family. Family members must fully adapt to the demands posed by a child with a chronic illness. The complexity of diagnosis, rehospitalisation for reevaluations and the complications, the need for surgical re-intervention, are critical moments for the patient respectively for his family. Family concerns (especially of the parents with newborn diagnosed prenatally or after birth) are related to the complexity of CHD, the prognosis and therapy results in time.

The educational level of parents is also correlated with

the quality of life of children diagnosed with CHD. Low educational attainment is often associated with delays or lack of request for medical assistance and, therefore worsening the child's health. Time factor is, however, extremely important for treating these disorders: it is preferably that the surgical intervention to be performed prior to the deterioration of cardiac function or at the occurrence of respiratory or circulatory system complications. The financial situation of the family is also correlated with the quality of life of the sick child and his family: health care costs may create financial difficulties, and these in turn can destroy the relationships between parents.

Educational level determines a different response, by adapting to the new family situation that can be dramatic (for example, blaming mothers who discover the CHD diagnosis after birth). These emotions can exert a negative influence on the development of the newborn. The attitude of the mother, in particular, but also of the whole family, regarding a child with CHD is difficult to predict, but the high level of stress, time limited hopes or socio-economic changes that may be caused by a professional change, have a negative impact on his quality of life of the child.

### **Studies on the quality of life of children with CHD**

In the studies on quality of life of patients with various disorders, attention for their physical condition must be accompanied by the one for their psychological and social welfare. An important factor in evaluating the quality of life of patients with CHD is their perceptions on the impact of the disease and of its treatment on their lives, the more so as the objective indicators, traditionally used for determining the quality of life of patients with cardiac disease, as cardiopulmonary exercise capacity, effort resistance, have proved insufficient.(12)

Quality of life studies are a relatively new field in research in paediatric cardiology and measuring the quality of life of children, not just those diagnosed with CHD, is proving more difficult than for adults. Ethical issues, such as confidentiality and protection of the respondents, essential in any research, are even more important in children. For example, children are more vulnerable in power relations with the adult researcher and, perceiving him as an authority figure they can try to answer so as to satisfy him, out of fear of a certain reaction. With regard to consent to participate in the study, it must be obtained from parents, and even then the process involves gathering information from both the child patient and from his parents.(16) At the same time, the instruments for measuring the quality of life must be adapted to the child's age and often in such studies creative methods are used involving photographs or drawings.

It is increasingly recognised the role of child's involvement in decisions concerning his treatment in order to improve the quality of his own life; however the younger the child is, the stronger the role of his parents is. Although, the perceptions of parents and children on the disease are different in terms of distinct levels of awareness and information regarding the disease, both perspectives are useful for the researcher. For example, discussions with children can provide information about their emotions and feelings, physical limitations or side effects of medication, which might not always result from the dialogue with the parents.

Quality of life assessment should be an integrated part of patient's treatment, helping professionals to identify children's preferences, differences between them and to develop tailored treatment plans for their patients. Counselling and psychosocial support given to patients and to their families in these cases have a decisive role in the family integration, preventing social isolation and increasing the ability to control

and possess child care needs.(17)

If in studies with adult respondents, the research methods are more diverse, when it comes to children, the most commonly used are interviews and observation. Nevertheless, questionnaires are also used, an example being the paediatric cardiology PedsQL model for measuring quality of life from the perspective of health in healthy children and adolescents as well as in those with chronic and acute diseases. The PedsQL model has available modules for a range of illnesses, including asthma, rheumatologic disorders, diabetes or cancer. The questionnaire contains 23 items and four main dimensions: physical, social, emotional and educational. In the case of parents, children aged 8 to 12 years and adolescents (13-18 years) the questionnaire can be self-administered, after a previously training, while for children aged 5 to 7 years, the questionnaire is administered by a specialist.(18)

A study on methods applied in research of quality of life associated with health, for the period 2001-2005, identified only 30 generic and 64 specific instruments to measure quality of life associated with health, most of them covering elements regarding the physical, mental state and social state of the respondent.(19) Whichever method is used, in case of children one essential aspect is represented by adjusting the instruments to their age.

## **DISCUSSIONS AND CONCLUSIONS**

CHD are the most common structural and functional abnormalities of the heart for children, having a major impact on morbidity and general mortality. Evaluation of the impact of these diseases on patients and their families, by knowing their expectations regarding therapy and prognosis, along with their accurate information, can lead to measures that contribute to lower the level of stress and of pre/ post-interventional anxiety. The increased level of stress of these families can influence physical and psychological development of patients with CHD.

Constant evaluation of these patients can help medical personnel in making decisions regarding medical procedures, in the process of informing the patient or his family on possible effects of the chosen procedure, in determining the effectiveness of chosen treatment and the development of cardiac rehabilitation programs.

Educating and informing the patients with CHD and their families regarding the need of supervision and continuous medical care throughout life, requires the existence of specialized centres to meet the concerns for improving the quality of life of these patients. To ensure high quality care, given the increasing population with these conditions towards adult age, the problems faced by patients must be evaluated and based on these evaluations, appropriate psychological support must be provided.

Quality of life from the perspective of health (HRQoL) must be a constant concern in medical practice. Thus, it requires evaluation of both medical therapies effects on the lives of patients and their effectiveness, as well as of how patients experience their quality of life. Both components are an important support for clinical assessment, medical decision making and even improved medical care.

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