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Original Article

Evaluation of quality of life in patients with congenital heart disease: an observational case control study

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Abstract: Quality of life (QoL) has become an important issue in patients with congenital heart disease (CHD). Accordingly, the focus has shifted from mere survival to a better QoL. The objective of this study is to assess QoL, by examining the World Health Organization QoL-bref (WHOQoL-BREF) questionnaire, in patients with CHD and a control population of a same geographic area matched for age, sex, cardiovascular risk factors and educational level. 154 patients with CHD recruited from a single hospital outpatient clinic and 250 healthy controls were studied between October 2018 and April 2019. Median age in patients with CHD was 27 (20-34) years and 62% were male. 32, 90 and 32 patients with CHD showed mild, moderate, and complex defects, respectively. 131 (53%) controls referred having felt or experienced negative feelings such as bad mood, despair, stress, or depression compared to 53 (34%) patients in the CHD group ($P<0.001$). Despite needing more medical treatment ($P<0.001$), patients with CHD enjoyed more their lives ($P<0.001$), felt their lives made more sense ($P<0.001$), were happier with themselves ($P=0.006$) and with their personal relationships ($P=0.020$), had a greater support from friends ($P=0.031$) and felt safer in their daily lives ($P=0.004$) than patients in the control group. Similarly, patient with CHD felt their environment was healthier, had more opportunities for leisure and were happier with their access to healthcare system ($P<0.005$) than controls. On the contrary, except the need for more medical treatment in patients with great CHD defects ($P=0.019$), no significant differences were seen in the WHOQoL-BREF survey according to the anatomical complexity. In conclusion, overall patients with CHD scored better in the WHOQoL-BREF questionnaire, especially in the psychological and environment domains, than patients in the control group.

Keywords: Quality of life, congenital heart disease, complexity, case-control study

Introduction

Advances in medical diagnosis and cardiac surgery procedures have enabled the increased survival of patients with congenital heart disease (CHD). Uncertainty about the future, fear of death, emotional suffering is common experiences in patients with CHD. Therefore, it is important to understand the impact of the CHD in these patients and explore their functioning and challenges using a screening tool.

Although quality of life (QoL) is not scientifically measurable, as it is very subjective to the individual, this broad multidimensional concept that usually includes subjective evaluations of both positive and negative aspects of life has gained importance in recent years providing complementary information to clinical data letting us identify patients with CHD who are at risk for health problems and need professional assistance.

Over the last years many QoL studies, including multicenter controlled cross-sectional studies in addition to systematic critical reviews [1-4], have been reported and the overall conclusion is that the QoL of adult patients with CHD is good. However, findings have not always been consistent which may hamper the ability to draw firm conclusions [5]. Additionally, QoL varies across countries and this between-country variation has been related to some patient characteristics such as age, marital and employment status or functional class assessment. However, no case control studies related to QoL in patients with CHD have been made in Spain a country were more than half of young people between 25 and 29 years old and a quarter of adults between 30 and 34 years old still live with their parents irrespective of their study levels [6] or family incomes.

The objective of this study is (a) to compare the QoL in Spanish patients with CHD and a control population of a same geographic area and (b) to confront CHD complexity by examining the World Health Organization Quality of Life-bref (WHOQoL-BREF) questionnaire.

Material and methods

Subjects

Cross-sectional study among consecutive CHD patients recruited from a single hospital outpatient CHD clinic between October 2018 and April 2019. Inclusion criteria were patients 18 years or older with a structural CHD verified by cardiac imaging. Patients unable to answer the survey, who did not want to participate or with a life expectancy less than one year were excluded from the study. Controls were selected at random from the same source population in the Grand Canary Island, the second most populous of the Canary Islands, an archipelago off the Atlantic coast of Northwest Africa which is part of Spain and with more than 2.150.000 inhabitants. Recruitment of the control population was completed in the same period and matched for age, gender, cardiovascular risk factors and educational level to patients with CHD. All patients were Caucasian and all of them gave informed consent for participation in the study. The protocol of the study was approved by the Hospital's Ethics Committee.

Demographic and clinical data

Data including sex, age, cardiovascular risk factors (systemic arterial hypertension, diabetes mellitus, hyperlipidaemia, and smoking habit), education level (none, primary, middle or university) and marital status (single, divorced, separated, cohabit, married or widower) were collected in case and control patients. Also, the New York Heart Association (NYHA) functional class was recorded in patients with CHD. The complexity of patients' heart defects (simple, moderate, or complex) was classified into diagnostic groups according to the underlying cardiac anatomy [7].

WHOQoL-BREF questionnaire

The goal of the QoL questionnaire is to assess one's perceived satisfaction or dissatisfaction in the major domains of life. The WHOQoL-BREF comprises 26 items. The first two questions of the survey are two stand-alone questions, one pertaining to the respondents' rated QoL, and one related to their Satisfaction with Health. The rest of questions measure the following broad domains: physical health, psychological health, social relationships, and environment (**Table 3**). A time frame of two weeks was indicated in the assessment of the survey. The WHOQoL-BREF questionnaire was completed by patients with CHD during the physician consultation after ensuring the ability to read and understand it. Conversely, the WHOQoL-BREF was sent online, during the same study period, to control patients. Domain scores were scaled in a positive direction: 1 point meant nothing, 2 points very little, 3 points more or less, 4 points very much and 5 points extremely [8]. The WHOQoL-BREF was used as it is suitable to use in patients with different health conditions and has been validated in the Spanish population [9].

Statistical analysis

Quantitative variables were expressed as mean ± standard deviation or median and percentiles (5-95). Qualitative variables were expressed in percentages. A normal distribution was tested using the Kolmogorov-Smirnov test. Possible associations between categorical variables were evaluated by using the Pearson chi-square test. Continuous data were compared by Student's t-test or Mann-Whitney test for variables with or without normal distribution, respectively. A One-Way Analysis of Variance (ANOVA) was used to test the equality of three or more means at one time by using variances. A p values less than 0.05 was considered statically significant. Data analysis was carried out using the Statistical Package for the Social Sciences (SPSS) 24.0 (SPSS, Chicago, IL).

Table 1. Congenital cardiac classification according to anatomy complexity

Types of congenital heart disease	Number of patients
Simple defects	
Isolated small ASD	1
Isolated small VSD	13
Mild native isolated pulmonary stenosis	2
Previously ligated or occluded ductus arteriosus	1
Repaired secundum ASD or sinus venous defect without significant shunt	7
Repaired VSD	7
Other mild repaired conditions	1
Moderate defects	
Aorto-left unrepaired conditions	1
Anomalous pulmonary venous connection	1
AVSD partial or complete	13
Aortic valve stenosis or bicuspid aorta	13
Congenital mitral valve disease	1
Coarctation of the aorta	16
Ebstein anomaly	2
Pulmonary stenosis with surgery or valvuloplasty	15
Peripheral pulmonary stenosis	1
Subvalvular aortic stenosis	5
Repaired Tetralogy of Fallot	22
Complex defects	
Cyanotic CHD (unrepaired or palliated, all forms)	1
Double outlet right ventricle	4
Fontan procedures	1
Single ventricle	2
Pulmonary atresia	6
Dextro and Levo transposition of the great arteries	17
Truncus arteriosus	1
Total	154

ASD: atrial septal defect, VSD: ventricular septal defect, AVSD: atrioventricular septal defect, CHD: congenital heart disease.

Results

Case and control population

One hundred and fifty four out of 158 (97%) patients with CHD and 250 controls matched for age, sex, cardiovascular risk factors and educational level were included in the study. 4 patients with CHD unable to understand the survey were excluded from de analysis. No patient was excluded due to co-morbidity problems and no patient refused to participate in the study. According to anatomy patients with CHD were classified as having simple (32 patients), moderate (90 patients) or complex (32 patients) defects (**Table 1**). **Table 2** shows demographic and clinical data in CHD patients and the control group. In relation to the NYHA classification patients with complex defects showed a significant worse functional class than patients with mild or moderate defects (mild: 1.0 ± 0.2, moderate: 1.1 ± 0.4, great: 1.75 ± 0.67, P<0.001).

WHOQoL-BREF questionnaire

In relation to the WHOQoL-BREF questionnaire 131 (53%) control patients referred having felt or experienced negative feelings such as bad mood, despair, stress or depression (more or less, very much or extremely) in the past two weeks compared to 53 (34%) patients in the CHD group (P<0.001). Similarly, control patients assessed a worse quality of life ("very poor" or

Table 2. Demographic and clinical data of congenital heart disease and control patients

	Control	CHD	P*
Number of patients, n	250	154	
Age, years	27 (24-33)	27 (20-35)	0.090
Sex, male	132 (53)	96 (62)	0.060
BMI, (kg/m^2)	24 ± 4	24 ± 5	0.891
Arterial hypertension, n	11 (4)	13 (8)	0.095
Diabetes mellitus, n	7 (3)	5 (3)	0.797
Dyslipidaemia, n	13 (5)	4 (3)	0.206
Smoking, n	25 (10)	9 (6)	0.144
Educational levels			0.071
None or primary, n	14 (6)	16 (10)	
Medium or university, n	236 (94)	137 (90)	
Marital status			<0.001
Single, divorced, separated or widower, n	161 (64)	132 (86)	
Married or cohabit, n	89 (36)	21 (14)	

CHD: congenital heart disease, BMI: body mass index. The data are expressed as mean ± standard deviation, median and quartiles (5-95) and number and percentages. *Categorical variables are evaluated by the Pearson chi-square test, continuous data with normal distribution by Student's t-test and continuous data without normal distribution by Mann-Whitney test.

“poor”) [45 (18%) control patients vs. 15 (10%) CHD patients, P=0.022)] and refereed to be less happy with their health (“very dissatisfied” or “dissatisfied”) [59 (24%) control patients vs. 23 (15%) CHD patients, P=0.026] than CHD patients. **Table 3** exhibits the median of each question of the WHOQoL-BREF questionnaire in CHD and control patients. From this table patients with a complex CHD defect had a greater need for medical treatment and a worse acceptance of their physical appearance. The rest of the questions of the survey did not obtain statistically significant difference according to CHD complexity. On the other hand, when all CHD and control patients were compared, the former reported a better quality of life, being happier with their health, having better relationships and support from friends and having fewer negative feelings such as bad mood, despair, stress, or depression than the later. In fact, lives in patients with CHD made significantly more sense than in controls.

Discussion

Health-related quality of life reflects the patient's perception of the impact of the illness and its treatment on their lives [10-12]. Like other patients with chronic illnesses, CHD with repaired or unrepaired defects continue to face physical, psychosocial, and environmental challenges after successful treatment through surgical or catheter intervention [13]. In fact, numerous factors may affect health-related QoL in patients with CHD such as impaired peer relationships, family overprotection, sports activity restriction, delayed progression into independent adulthood, discrimination and bullying or the feeling of being different from healthy people [14-17].

Previous studies measuring different dimensions of behavioural and emotional problems in adolescents and adult patients with CHD have shown contradictory results. On the one hand some authors have found a high frequency of depression and anxiety disorders among CHD patients [18, 19] while other authors have not found that anxiety or depression were greater than in the general population [20]. In our case we found a high incidence of negative feelings such as bad mood, despair, stress, or depression in both groups but surprisingly higher in the control population after matching for age, sex, cardiovascular risk factor or educational level. In fact, our results show that, as a group, patients with CHD were more satisfied with their lives than controls despite pain prevented them from doing things and needed more medication intake. It may seem illogical but having lived their whole life with a chronic disease, which has remained stable for most of the time, may have changed their expectations. In this context, although this may seems strange, researchers offer three possible explanations: the disability paradox, the response shift, and the sense of coherence. The central concept in all of them has to do with establishing and maintaining a sense of balance between the body, mind, and spirit and with the individual's social context and environment [21] that enables coping with their congenital defect. In fact, the more a person is able to understand and

Table 3. Quality of life questionnaire from the World Health Organization (WHOQoL-BREF)

	CHD complexity			P*	Total CHD	Control population	P**
	Mild	Moderate	Great				
How do you assess your quality of life?	4.0 ± 0.8	4.0 ± 0.8	3.5 ±1.0	0.152	3.7 ± 1.0	3.4 ± 0.9	<0.001
How happy are you with your health?	3.7 ± 1.1	3.7 ± 1.1	3.5 ± 1.0	0.638	3.5 ± 1.1	3.3 ± 1.0	0.031
Physical health domain							
How do you think your pain prevents you from doing things?	1.9 ± 1.2	1.9 ± 1.2	2.2 ± 1.1	0.533	2.0 ± 1.1	1.8 ± 1.0	0.009
How much do you need medical treatment for your daily life?	1.6 ± 1.1	1.6 ± 1.1	2.6 ± 1.4	0.019	2.1 ± 1.3	1.5 ± 1.0	<0.001
Do you have enough energy for your daily life?	4.0 ± 0.8	4.0 ± 0.8	3.5 ± 1.1	0.098	3.6 ± 1.0	3.5 ± 0.9	<0.001
How well are you able to move?	4.3 ± 1.0	4.3 ± 1.3	4.2 ± 1.0	0.945	4.2 ± 1.1	4.3 ± 0.9	0.297
How happy are you with your sleep?	3.3 ± 1.2	3.3 ± 1.2	3.2 ± 1.4	0.911	3.3 ± 1.2	3.1 ± 1.1	0.275
How happy are you with your ability to perform your daily activities?	4.0 ± 1.0	4.0 ± 1.0	3.5 ± 1.1	0.160	3.7 ± 1.0	3.7 ± 0.9	0.469
How happy are you with your capacity to work?	3.6 ± 1.0	3.6 ± 1.0	3.4 ± 1.4	0.844	3.5 ± 1.2	3.7 ± 0.9	0.110
Psychological health domain							
How much do you enjoy life?	4.0 ± 0.9	4.0 ± 0.9	3.7 ± 1.0	0.202	3.9 ± 0.9	3.4 ± 0.8	<0.001
How much do you feel your life makes sense?	3.9 ± 1.3	3.9 ± 1.3	3.7 ± 0.9	0.185	4.0 ± 1.1	3.6 ± 1.0	<0.001
How much can you concentrate?	3.5 ± 0.8	3.5 ± 0.8	3.2 ± 1.0	0.385	3.4 ± 1.0	3.3 ± 0.9	0.358
Are you able to accept your physical appearance?	4.4 ± 0.9	4.4 ± 0.9	3.7 ± 1.2	0.023	3.9 ± 1.1	3.6 ± 1.0	0.006
How happy are you with yourself?	4.1 ± 1.0	4.1 ± 1.0	3.7 ± 1.2	0.284	3.9 ± 1.1	3.6 ± 1.0	0.006
How often do you have negative feelings such as bad mood, despair, stress, depression?	2.2 ± 1.0	2.3 ± 0.9	2.6 ± 1.0	0.174	2.4 ± 1.0	2.7 ± 0.9	0.001
Social relationships domain							
How happy are you with your personal relationships?	4.1 ± 1.0	4.1 ± 1.0	3.7 ± 1.1	0.278	3.8 ± 1.1	3.6 ± 1.0	0.020
How happy are you with your sex life?	3.0 ± 1.9	3.4 ± 1.5	3.1 ± 1.4	0.386	3.2 ± 1.6	3.3 ± 1.7	0.680
How happy are you with the support you get from friends?	3.8 ± 1.0	3.9 ± 1.5	4.0 ± 1.2	0.800	3.9 ± 1.1	3.7 ± 0.9	0.031
Environment domain							
How safe do you feel in your daily life?	3.9 ± 0.7	3.9 ± 0.7	3.5 ± 1.1	0.263	3.7 ± 0.9	3.4 ± 0.9	0.004
How healthy is your physical environment?	3.6 ± 0.8	3.6 ± 0.8	3.6 ± 1.1	0.387	3.7 ± 0.9	3.3 ± 0.8	<0.001
Do you have enough money to pay for your needs?	3.2 ± 1.1	3.2 ± 1.1	3.1 ± 1.1	0.415	3.3 ± 1.1	3.4 ± 1.0	0.542
How available is the information you need?	2.7 ± 0.9	3.7 ± 0.9	3.9 ± 0.9	0.474	3.7 ± 1.0	3.8 ± 0.8	0.496
How much opportunity for leisure do you have?	3.6 ± 1.1	3.6 ± 1.1	3.7 ± 1.0	0.416	3.5 ± 1.0	3.2 ± 0.8	0.001
How happy are you with the situation of the place where you live?	4.1 ± 0.9	4.0 ± 1.2	3.8 ± 1.2	0.612	4.0 ± 1.0	3.8 ± 0.9	0.094
How happy are you with your access to healthcare?	3.8 ± 1.3	3.9 ± 0.9	3.7 ± 1.2	0.816	3.8 ± 1.0	3.6 ± 0.9	0.041
How happy are you with your means of transportation?	3.7 ± 1.1	3.3 ± 1.2	3.3 ± 1.2	0.280	3.4 ± 1.2	3.1 ± 1.3	0.039

The data are expressed as mean ± standard deviation *A One-Way Analysis of Variance (ANOVA) was used to test the equality of three or more means at one time by using variances. **Continuous data were compared by Student's t-test.

integrate, to handle and to make sense of an experience or disease, the greater the individual's potential to successfully cope with the situation or the disease. Also, as occurs in a large part of the Spanish population in their 20s and 30s, many of our patients with CHD lived with their parents which may increase their strength and resilience.

Also, as previously reported CHD complexity was not related to QoL, indicating that symptoms experienced by patients may be more important contributory factors to QoL than defect complexity itself [10]. Surprisingly, patients with complex defects scored similar in the QoL questionnaire than patients with mild defects. The finding that disease complexity dose not play a role in predicting QoL may be explained by the fact that we measured QoL from a holistic point of view, instead of a merely functional perspective. In this context, Lane et al. [22] found that patients who had received palliative treatment reported QoL scores similar to those who had never required cardiac surgery and to the general population, although both CHD groups had significantly poorer physical functioning than the general population.

There are, however, limitations in our study that may impact our findings. Control patients completed the surveys online while CHD patients did it in clinic. Despite a web version can be an alternative to a paper version for health related QoL evaluation [23] it may imply bias in our results. Also, as it is a cross-sectional research design this study does not allow us to examine the direction of effects linking QoL to other variables of interest.

In conclusion, no significant differences were seen in most of the questions of the WHOQoL-BREF survey attending to CHD complexity. However, unlike controls, patients with CHD had a better perception of their QoL and were happier with their lives. Despite this, we found a high prevalence of negative feelings in more than one third of our patients with CHD.

Disclosure of conflict of interest

None.

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References

[1] Schroder M, Boisen KA, Reimers J, Teilmann G and Brok J. Quality of life in adolescents and young adults with CHD is not, reduced: a systematic review and meta-analysis. Cardiol Young 2016; 26: 415-425.

[2] Kahr PC, Radke RM, Orwat S, Baumgartner H and Diller GP. Analysis of associations between congenital heart defect complexity and health-related quality of life using a metaanalytic strategy. Int J Cardiol 2015; 199: 197-203.

[3] Ladak LA, Hasan BS, Gullick J and Gallagher R. Health-related quality of life in congenital heart disease surgery in children and young adults: a systematic review and meta-analysis. Arch Dis Child 2019; 104: 340-347.

[4] Apers S, Kovacs AH, Luyckx K, Thomet C, Budts W, Enomoto J, Sluman MA, Wang JK, Jackson JL, Khairy P, Cook SC, Chidambarathanu S, Alday L, Eriksen K, Dellborg M, Berghammer M, Mattsson E, Mackie AS, Menahem S, Caruana M, Veldtman G, Soufi A, Romfh AW, White K, Callus E, Kutty S, Fieuws S and Moons P; APPROACH-IS consortium and ISACHD. Quality of life of adults with congenital heart disease in 15 countries: evaluating country-specific characteristics. J Am Coll Cardiol 2016; 67: 2237-2245.

[5] Moons P and Luyckx K. Quality-of-life research in adult patients with congenital heart disease: current status and the way forward. Acta Paediatr 2019; 108: 1765-1772.

[6] Continuous Household Survey (ECH). Methodology. National Statistics Institute (INE). Spain Madrid 2019: 1-9.

[7] Arnett DK, Blumenthal RS, Albert MA, Buroker AB, Goldberger ZD, Hahn EJ, Himmelfarb CD, Khera A, Lloyd-Jones D, McEvoy JW, Michos ED, Miedema MD, Muñoz D, Smith SC Jr, Virani SS, Williams KA Sr, Yeboah J and Ziaeian B. 2019 ACC/AHA guideline on the primary prevention of cardiovascular disease: a report of the American college of cardiology/American heart association task force on clinical practice guidelines. J Am Coll Cardiol 2019; 74: e177-e232.

[8] WHOQOL Group. Study protocol for the World Health Organization Project to develop a Quality of Life Assessment Instrument (WHOQOL). Qual Life Res 1993; 2: 153-159.

[9] Lucas-Carrasco R. The WHO quality of life (WHOQOL) questionnaire: Spanish development and validation studies. Qual Life Res 2012; 21: 161-165.

[10] Gill TM and Feinstein AR. Acritical appraisal of the quality of quality-of-life measurements. JAMA 1994; 272: 619-626.

[11] Moons P, Budts W and De Geest S. Critique on the conceptualisation of quality of life: a review and evaluation of different conceptual approaches. Int J Nurs Stud 2006; 43: 891-901.

[12] Bratt EL and 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.

[13] Kim GB. Psychosocial adjustment and quality of life of adolescents and adults with congenital heart disease. Korean J Pediatr 2014; 57: 257-263.

[14] Dalieno L, Mapelli D and Volpe B. Measurement of cognitive outcome and quality of life in congenital heart disease. Heart 2006; 92: 569-574.

[15] Nakou S. Measurement of quality of life in the health care field. Applications in child birth. Archives of Hellenic Medicine 2001; 18: 254-266.

[16] McMurray RL, Parsons JM, Quirk J, Veldtman GR, Lewin RJ and Sloper P. A life less ordinary: growing up and coping with congenital heart disease. Coron Health Care 2001; 5: 51-57.

[17] Lee S and Kim SS. The life of adolescent patients with complex congenital heart disease. J Korean Acad Nurs 2010; 40: 411-422.

[18] Horner T, Liberthson R and Jellinek MS. Psychosocial profile of adults with complex congenital heart disease. Mayo Clin Proc 2000; 75: 31-36.

[19] Bromberg JI, Beasley PJ, D'Angelo EJ, Landzberg M and DeMaso DR. Depression and anxiety in adults with congenital heart disease: a pilot study. Heart Lung 2003; 32: 105-110.

[20] Saliba Z, Butera G, Bonnet D, Bonhoeffer P, Villain E, Kachaner J, Sidi D and Iserin L. Quality of life and perceived health status in surviving adults with univentricular heart. Heart 2001; 86: 69-73.

[21] Albrecht GL and Devlieger PJ. The disability paradox: high quality of life against all odds. Soc Sci Med 1999; 48: 977-988.

[22] Lane DA, Lip GY and Millane TA. Quality of life in adults with congenital heart disease. Heart 2002; 88: 71-75.

[23] Chen WC, Wang JD, Hwang JS, Chen CC, Wu CH and Yao G. Can the web-form WHOQOL-BREF be an alternative to the paper-form? Soc Indic Res 2008; 94: 97-114.