The impact of congenital heart diseases on the quality of life of patients and their families in Saudi Arabia

Biological, psychological, and social dimensions

Ahmad S. Azhar, *MD, FRCPC,* ***Zahra H. AlShammasi,*** *MD,* ***Rawan E. Higgi,*** *MD .*

ABSTRACT

الأهداف: تقييم أثر أمراض تشوهات القلب الخلقية على جودة حياة المرضى وأسرهم من نواحي الصحة الجسدية، النفسية، والاجتماعية.

الطريقة: تمت هذه الدراسة المقطعية بين شهر مايو 2014 وأغسطس 2015، وقد شملت الأطفال الذين يعانون من أمراض تشوهات القلب الخلقية والذين لا تزيد أعمارهم على الـ16 سنة والمتابعين في مستشفى جامعة الملك عبد العزيز بجدة، المملكة العربية السعودية. تم تعبئة استبيان شامل لدراسة الأبعاد البيولوجية، والنفسية، والاجتماعية الخاصة بالأطفال المرضى، وكذلك بوالديهم وإخوتهم. حُسبت النتائج على شكل درجات تتراوح من 0 إلى 100% معبّرة عن مدى التأثر لكل بعد من الأبعاد الثلاثة السابقة ولكل فرد من أفراد الأسرة.

النتائج: تم اشتراك 180 طفلاً في هذه الدراسة، 104 (57.8%) منهم كانوا ذكوراً، ومتوسط أعمارهم كانت 5.65 (4.8) سنة. بيّنت الدراسة أن 25% من الأطفال المصابين بتشوه خلقي في القلب يعانون حالات التهابات الجهاز التنفسي المتكررة، و35% يتعالجون في المستشفيات بشكل متكرر، و38.9% لديهم تأخر في المعالم التطورية للطفل، أما 12 (6.7%) منهم فقط كانوا مسجلين في الخدمات الاجتماعية. وأوضحت الدراسة أن نسبة الأمهات اللاتي يواجهن صعوبة في التأقلم مع مرض أبنائهم تبلغ 20% من الحالات و22.2% منهن أصابهن الاكتئاب. بلغت متوسطات قياسات درجات التأثير (SD) لأمراض تشوهات القلب الخلقية على الأولاد المصابين كما يلي: 26.1 ± 26.2 على البعد الصحي الجسدي، 28.7 ± 28.8 على البعد النفسي، و20.2 ± 25.7 على البعد الاجتماعي. تبيّن أن الأمهات كنّ أكثر تأثراً من الآباء. أمراض تشوهات القلب الخلقية المعقدة كان لها تأثير إضافي، كما أبرزت النتائج ارتفاعاً نسبياً في درجات تأثير أمراض تشوهات القلب الخلقية على جودة حياة الأطفال العامة عند الأسر التي تفتقر إلى المعرفة الكافية لما يتعلق بالمرض.

الخاتمة: أمراض تشوهات القلب الخلقية تؤثر على جميع جوانب جودة الحياة عند المريض وأسرته وتتميّز بكثرة الأمراض المصاحبة. يعتبر الدعم الاجتماعي والنفسي، إلى جانب التربية الصحية للمرضى وأوليائهم - من آباء وأمهات - من العوامل الحاسمة لتحسين جودة حياتهم.

Objectives: To assess the impact of congenital heart diseases (CHDs) on bio-psychosocial aspects of the quality of life (QOL) of patients and their families.

Methods: A cross-sectional study was carried out between May 2014 and August 2015, including children aged <16 years, and followed-up at King Abdulaziz University Hospital, Jeddah, Kingdom of Saudi Arabia for CHD. A broad questionnaire was administered to investigate biological, psychological, and social dimensions of afflicted children, their parents, and siblings. Outcomes were computed as impact scores (0-100%) for each dimension and family member.

Results: A total of 180 children (104 [57.8%] males; mean age ± standard deviation [SD] = 5.65 ± 4.8 years) were included. There were 25% children complaining of recurrent respiratory infections, 35% of frequent hospitalizations, 38.9% had milestone delay, and 12 (6.7%) only had a social security registration. Mothers declared difficulty coping with their children's disease in 20% of cases and 22.2% reported being depressed. Mean ± SD impact scores in afflicted children were: 26.1 ± 26.2 (biological), 28.7 ± 28.8 (psychological), and (20.2 ± 25.7) social dimensions. Mothers' impact scores were higher than fathers'. Complex CHDs had an additional impact, and children from families with less knowledge on CHD had relatively greater impact scores.

Conclusion: Congenital heart diseases impact all aspects of QOL of patients and their families, and are associated with high comorbidity. Social and psychological support and education for patients and their parents are crucial factors for improving QOL.

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From the Department of Pediatrics, King Abdulaziz University, Jeddah, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Ahmad S. Azhar, Department of Pediatrics, King Abdulaziz University, Jeddah, Kingdom of Saudi Arabia. E-mail: aazhar@kau.edu.sa / azcardio@hotmail.com

Congenital heart diseases (CHDs) are one of the most frequent fetal malformations, with a prevalence of up to 13 per 1000 live births,[1-6] and are associated in 12% of cases with a chromosomal anomaly, such as Down syndrome. In the Kingdom of Saudi Arabia (KSA), although studies do not provide accurate data, the incidence of CHD is reported to be less than 5 per 1000 newborns.[7] Congenital heart diseases are generally classified into 2 categories: acyanotic and cyanotic forms, according to the presence or absence of cyanosis.[3] Another classification categorizes CHD into 2 types: simple CHD, such as septal malformations (atrial septal defect [ASD] and ventricular septal defect [VSD]); and complex CHD combining more than one simultaneous defect, such as the tetralogy of Fallot.[8] In KSA, several studies agree that VSD is the most common type of CHD, occurring in one-third of cases, followed by ASD.[7] In recent years, progress in the surgery for CHD has undeniably improved the outcome of the disease, which has considerably increased the life expectancy of patients; only a few decades ago, the overall mortality was 4 of every 5 infants, which decreased to less than 2 of every 5 in the last decade.[1,2,6,9] Additionally, prenatal diagnostic techniques, including sonograms, have allowed early detection of the malformation and more appropriate post-natal care.[9] Hence, almost 9 of 10 treated patients reach adulthood nowadays. In consequence, 2 decades from now, a larger number of adult patients than children will have CHD.[1,2] From this optimistic situation, novel issues have arisen regarding the quality of life (QOL) of these patients, beyond survival and purely medical concerns.[10] The situation is similar in KSA, as the availability of up-to-date techniques of diagnosis and surgery in cardiology have effectively increased the life expectancy of patients with CHD, creating a new population of adults with operated CHD who remain vulnerable individuals with long-life special medical needs.[7] Furthermore, this population of patients is exposed to neuro-developmental disorders, resulting either from underlying genetic anomalies or from the chronic circulatory insufficiency relative to CHD.[2,11] Despite the notable improvement in diagnostic and surgical techniques, the prevalence of these disorders seems not to have declined over time.[12] On the other hand, the particular psychosocial environment in which CHD children grow-up, namely, recurrent hospitalizations/interventions, an overprotective family attitude, irregular school courses, stigmatization, and persisting functional limitations, and so forth, may aggravate psychosocial and neuro-developmental outcomes. As a consequence, children with CHD may grow-up with a particular trait, including weak cognitive and communication skills, poor sociability, impulsivity, and impaired executive functions.[4,11] This situation raises concern for the QOL for these patients and their respective families, besides medical issues that often become less predominant in the clinical presentation. Therefore, the optimal management of these patients should include particular attention to neuro-psycho-educational aspects of the disease and specialized care.[4,11] Furthermore, due to the burden of the diagnosis of CHD and its stressful feature, families may encounter difficulty in coping with the chronic condition of their afflicted relative, exposing them to significant deterioration in their biological, psychological, and social lives.[13] Besides the limited number of studies that have investigated CHD in KSA, there are very scarce data regarding the QOL of these patients. The aim of the present study was to assess the impact of CHD on the bio-psychosocial aspects of the QOL of both patients and their family members, including siblings. We also investigated the actual access of patients and their families to any type of social or psychological support, and the access of parents to health education programs helping them to manage the disease of their children.

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Methods. This was a cross-sectional study based on a questionnaire that was adapted by the authors, combining qualitative and quantitative assessments. Children followed-up for CHD in King Abdulaziz University Hospital (KAUH), at Jeddah, KSA and their respective family members (parents and siblings) were involved in the study. We only included children aged 5 months to 16 years. The study was carried out between May 2014 and August 2015, to reach a target sample size of 180 children. Eligible children were identified from the hospital's electronic database and parents, both or one representative, were invited for a private interview after providing consent. Parents were received by appointment, as per their convenience for an approximately 30 minute interview, until reaching the study target sample size.

Questionnaires were filled out by the interviewer: a trained medical student. They were written in both Arabic and English, and the interview was conducted in the most appropriate language for the respondents with oral translation into the local dialect by the interviewer, where necessary. The study was approved by the

Biomedical Research Ethics Committee of KAUH and was carried out in accordance to the principles Helsinki Declaration.

Questionnaire. Many authors worldwide have investigated the QOL of children, adolescents, and adults with CHD using either qualitative, quantitative, or mixed methods. Approximately a 100 specific and non-specific instruments have been used, giving discrepant results and conclusions.[13-16] After examining some relevant and validated questionnaires, such as the Pediatric Cardiac Quality of Life Inventory (PCQLI) developed in the USA and the ConQol Index developed in the UK, we concluded that a questionnaire that fits better with the demographic, social, and cultural characteristics of our population was needed. Therefore, we developed our questionnaire based on the most relevant items from the literature, taking into consideration the specificities of our local population. The questionnaire included 4 parts: 1) the child's demographic data, family social conditions, including social security prescription, and different kinds of financial, psychological, and social support received; 2) impact of CHD on the child's QOL; 3) impact on the parents' QOL; 4) impact on the siblings' QOL; and 5) families' needs and expectations.

Parts 2, 3, and 4 are the main parts, related to the assessment of the QOL in the afflicted child, parents, and siblings. Each part investigated a number of parameters grouped in 3 subunits: biological, psychological, and social dimensions. Each item is a dichotomous question (answered by yes or no).

Scoring system. Where applicable, answers for dichotomous questions were scored 0 for "no" relative to the absence of impact, and 1 for "yes" relative to the presence of impact in the concerned parameter. When a question was not applicable to the subject, no score was attributed. The sum of the scores was divided by the number of applicable questions and expressed in percentage, giving the impact score. The bigger the impact score, the higher the impact of CHD on the given dimension and family member. Thus, we obtained a biological impact score (BIS), a psychological impact score (PIS), and a social impact score (SIS) for each family member, as well as a global impact score (GIS) that was calculated by dividing the scores of all the items by the total number of applicable questions. Parents had a common score, as well as individual scores each. Finally, a family score was calculated by dividing the sum of the parents' and siblings' scores (excluding the patient) by the corresponding number of applicable items. The questionnaire underwent face and content validity; and reliability was tested for the core part of the questionnaire (child's assessment), giving a Cronbach's alpha of 0.757.

Statistical methods. Statistical analysis was performed using the Statistical Package for Social Sciences version 20 (IBM Corp., Armonk, NY, USA). Descriptive statistics were used to present the population characteristics and to study the patterns of responses to the questionnaire, as well as the scores described previously. Means and standard deviations (SD) were calculated on continuous variables and frequencies/percentages on categorical variables. Correlations between categorical variables were analyzed using chi-square tests. Impact scores, ranging from 0-100% were analyzed as a continuous variable in statistical comparisons of means, using independent t-tests and one-way analysis of variance, as appropriate. Impact scores were also ranked into 3 levels: low impact (≤30), moderate impact (30-60), and high impact (>60), and were analyzed as an ordinal variable using the Mann-Whitney U test. Linear regression was used to analyze correlations between 2 numeric variables. A $p<0.05$ was considered to reject the null hypothesis.

Results.

Characteristics of the population. A total of 180 families were interviewed for a total of 180 children with CHD: 104 (57.8%) boys and 76 (42.2%) girls, mean age ± SD = 5.65 ± 4.8 years. There were 125 (69.4%) cases of simple CHD and 55 (30.6%) of complex CHD, and 16 (8.9%) of families having another child affected with CHD (who were not followed-up in our center). In 87.2% of families, mothers were the only care-givers of the child or children with CHD, care was given by both parents in 10.6%, and in the remaining 2.2%, the care-giver was someone other than the parents (**Table 1**).

Regarding the support received by families, only 12 families (6.7%) declared having social security and 58.3% declared receiving psychological support from their child's physician. Most families (73.9%) declared having received explanations regarding the disease of their child from healthcare providers; however, 53.3% declared needing further information and advice. Types of explanations received and needed are detailed in **Table 1**.

Quality of life assessment. **Table 2** summarizes the interviewers' answers regarding a selection of questions related to QOL. Assessments of the afflicted children revealed that 25% had recurrent upper respiratory tract infections (URI), which are defined by >9 episodes per year, or complicated by more than 4 episodes of otitis media.[17] In addition, 35% of the children had frequent hospital admissions in relation with CHD, almost

one-third (29.4%) had impaired physical growth and 38.9% had milestone delay. In linear regression, both frequencies of URI (p<0.001) and hospitalizations (p<0.001) were correlated with children's BIS, but not with PIS (p=0.965 and p=0.097), nor SIS (p=0.461 and 0.958).

Parent's assessments showed that 18.9% of the mothers and 9.4% of the fathers had difficulty coping with the disease of their children and the proportion of self-reported depression (as assessed by the item: "I am usually in a low, depressed mood: yes/no") was 3 times higher in mothers (22.2%) than in fathers (7.2%). Siblings' assessments showed that 32.8% of siblings had a feeling of jealousy toward their sick brother/sister and 19.4% of them felt neglected by their parents because of the disease of their brother/sister.

Table 1- Demographic, social, and clinical factors in QOL of children with CHD aand their families.

Variable	Frequency (%)
Age, mean ± SD	5.65 ± 4.81
Gender	
Male	104 (57.8)
Female	76 (42.2)
Diagnosis (CHD)	
Simple	125 (69.5)
Complex	55 (30.5)
Number of other sick children	
0	164 (91.1)
≥1	16 (8.9)
Pregnancy	
Planned	79 (43.9)
Unplanned	101 (56.1)
Person usually taking care of the child	
Mother	157 (87.2)
Mother & father	19 (10.6)
Other	4 (2.2)
Social registration	
Yes	12 (6.7)
No	166 (92.2)
Child's physician supportive	
Yes	105 (58.3)
No	71 (39.4)
Families who declared having insufficient knowledge	96 (53.3)
Families who declared receiving explanations about…	
The disease	133 (73.9)
Risk factors	50 (27.8)
Symptoms	87 (48.3)
Complications	73 (40.6)
Surgical procedures	74 (41.1)
Medical treatment	48 (26.7)
Families who declared still needing information about…	
The disease	40 (22.2)
Risk factors	33 (18.3)
Symptoms	35 (19.4)
Complications	54 (30.0)
Surgical procedures	45 (25.0)
Medical treatment	33 (18.3)

SD - standard deviation, CHD - congenital heart disease, QOL - quality of life

Impact scores. **Table 3** displays mean ± SD impact scores (BIS, PIS, SIS, and GIS) in each family member, including the afflicted children. In afflicted children, the mean impact scores of CHD were comparable in the 3 dimensions of QOL: biological (BIS=26.1 ± 26.2), psychological (PIS=28.7 ± 28.8), and social dimensions (BIS=20.2 ± 25.7); and mean ± SD GIS was 23.9 ± 17.7.

In parents, mothers' scores were higher than fathers', although comparative analysis was not performed to assess the statistical significance. Results, expressed in mean ± SD impact scores of mothers versus fathers showed: BIS=29.5 ± 32.3 versus 13.9 ± 26.0; PIS=25.0 ± 20.8 versus 15.22 ± 18.8; and SIS=19.5 ± 23.0 versus 11.2 ± 17.7. Similarly, GIS was disproportional between parents: 24.7 ± 17.0 in mothers versus 12.8 ± 12.4 in fathers. In siblings, it was interesting to note that the most important impact was on psychological dimension, with mean ± SD PIS=25.0 ± 24.6; while BIS=7.1 ± 23.8 and SIS=8.3 ± 19.2.

Impact of the severity of CHD on the parents' QOL. according to the severity of the diagnosis, we divided the population into 2 categories: simple CHD group and complex CHD group. Simple CHDs included the following anatomical abnormalities: isolated congenital aortic valve disease; isolated congenital mitral valve disease; isolated patent foramen oval or small ASD; isolated small VSD with no associated lesions; and mild pulmonic stenosis. Complex CHD groups comprised more severe types of CHD, such as conduits, cyanotic CHDs, mitral atresia, and transposition of the great arteries.[18] As reported in **Table 1**, there were 55 (30.6%) cases of complex CHD. Furthermore, impact was categorized into 3 levels with impact scores: low <30, moderate 30-60, and high impact >60. There was no statistically significant correlation between the severity of CHD and the level of its impact on any of the 3 dimensions of parents' QOL; however, some notable results deserve to be highlighted.

In complex CHD, parents are likely to be more biologically impacted than in simple CHD, with 23.6% of mothers and 14.8% of fathers with high biological impact (BIS >60) in cases of complex CHD, versus only 15.3% (p=0.395) and 5.0% (p=0.087), in cases of simple CHD. Parents combined BIS showed the same results: 16.4% of cases with high biological impact BIS >60% in complex CHD versus 6.5% in simple CHD (p=0.061). Similarly, GIS was moderate (30-60) in 21.8% of parents and 21.9% of the families in cases of complex CHD versus 12.2% (p=0.098) and 10.8% (p=0.099), in case of simple CHD. There was no case

of high global impact (GIS >60) in parents or families' assessments (**Table 4**).

Impact of CHD on exclusive care-givers. Descriptive statistics revealed that in most cases (87.2%) mothers were the only care-giver for the afflicted child (group A). In the remaining cases (12.8%), either parents or another individual were the main care-givers (group B). We compared the 2 group scores to assess the impact of being exclusive care-giver on mothers' BIS, PIS, SIS, GIS, spiritual life, and social activities.

The results showed that 50.6% of the mothers from group A (exclusive care-givers) declared having their

Table 2 - A selection of questions related to the afflicted children's parents' and family members' quality of life.

Questions	Frequency	%
Part 2: Child's QOL assessment		
The child has recurrent URTI (>9 URTI/year or >4 OM/year)	45	(25)
The child has so many hospital admissions	63	(35)
The child physical growth is affected	53	(29.4)
The child's illness causes delay in receiving some vaccines	31	(17.2)
The child is missing too many school classes	15	(8.3)
The child is receiving overprotective care	101	(50.1)
The child has milestone delay	70	(38.9)
Part 3: Parents QOL assessment		
I find it so difficult to cope with this event		
Mothers	34	(18.9)
Fathers	17	(9.4)
I feel guilty because of performing my duties as a mother/father		
Mothers	41	(22.8)
Fathers	20	(11.1)
I stay awake in the night with my child - not having enough sleep		
Mothers	61	(33.9)
Fathers	28	(15.6)
I am usually in a low-depressed mood		
Mothers	40	(22.2)
Fathers	13	(7.2)
My family plan - next childbearing - has changed		
Mothers	51	(28.3)
Fathers	38	(21.1)
My relationship with my other children has been affected		
Mothers	18	(10.0)
Fathers	9	(5.0)
My spiritual/prayer life has improved		
Mothers	4	(2.2)
Fathers	1	(0.6)
Our life is financially affected because of our child's disease		
Mothers	40	(22.2)
Fathers	27	(15.0)
Part 4: Siblings' QOL assessment		
They feel jealousy	59	(32.8)
They feel neglect	35	(19.4)
Their school performance has been affected	20	(11.1)

QOL - quality of life, URTI - upper respiratory tract infection, OM - otitis media

Table 3 - Impact of congenital heart disease (CHD) on afflicted children's and family members' quality of life.

Family member	BIS Mean ± SD	PIS Mean ± SD	SIS Mean ± SD	GIS Mean ± SD
Child	26.1 ± 26.2	28.7 ± 28.82	20.21 ± 25.7	23.93 ± 17.7
Mother	29.5 ± 32.26	25.01 ± 20.82	19.54 ± 23.02	24.71 ± 17.0
Father	13.28 ± 26.01	15.22 ± 18.80	11,15 ± 17.76	12.8 ± 12.4
Both parents	21.07 ± 24.73	19.93 ± 17.20	13.90 ± 15.96	18.21 ± 12.33
Siblings	7.09 ± 23.79	24.96 ± 24.6	8.28 ± 19.15	13.62 ± 14.27

BIS - biological impact score, PIS - psychological impact score, SIS - social impact score, GIS - global impact score, The bigger the impact score, the higher the impact of CHD on the given dimension and family member.

spiritual life improved versus 13% only from group B (p=0.007). On the other hand, 34% of mothers from group A declared missing important social activities to care for their child versus 9.1% only in group B (p=0.014). Furthermore, a greater proportion of mothers from group A had severe impacts (scores >60%) on all 3 dimensions (biological, psychological, and social), as well as the global impact, as compared with those

Table 4 - Correlation between congenital heart disease (CHD) severity (complex versus simple) and level of impact of the disease on the parents' quality of life.

Impact score	CHD severity		Total	*P*-value
	Complex CHD (n=55[a])	Simple CHD (n=125[b])		
	Frequency (%)			
BIS				
Mother				0.395
Low	34 (61.81)	83 (66.93)	117	
Medium	8 (14.543)	22 (17.74)	30	
High	13 (23.63)	19 (15.32)	32	
Father				0.087
Low	42 (77.77)	103 (85.83)	145	
Medium	4 (7.40)	11 (9.16)	15	
High	8 (14.81)	6 (5.0)	14	
Both parents				0.061
Low	34 (61.81)	95 (76.61)	129	
Medium	12 (21.81)	21 (16.93)	33	
High	9 (16.36)	8 (6.45)	17	
PIS				
Mother				0.088
Low	31 (56.36)	89 (71.77)	120	
Medium	20 (36.36)	26 (20.96)	46	
High	4 (7.27)	9 (7.25)	13	
Father				0.635
Low	45 (81.81)	105 (84.67)	150	
Medium	7 (12.72)	10 (8.06)	17	
High	3 (2.42)	6 (4.83)	9	
Both parents				0.735
Low	40 (72.72)	96 (77.42)	136	
Medium	13 (10.48)	23 (18.85)	36	
High	2 (1.61)	5 (4.03)	7	
SIS				
Mother				0.621
Low	43 (78.2)	89 (71.20)	132	
Medium	9 (16.36)	27 (21.6)	36	
High	3 (5.45)	9 (7.2)	12	
Father				0.223
Low	48 (87.27)	108 (89.26)	156	
Medium	7 (12.7)	9 (7.44)	16	
High	0 (0.0)	4 (3.31)	4	
Both parents				0.402
Low	49 (89.10)	104 (83.2)	153	
Medium	6 (10.90)	18 (14.4)	24	
High	0 (0.0)	3 (2.4)	3	
GIS				
Parents				0.098
Low	43 (78.18%)	108 (87.80)	151	
Medium	12 (21.81%)	15 (12.19)	27	
High	0 (0.0)	0 (0.0)	0	
Family				0.099
Low	32 (78.05%)	74 (89.16)	106	
Medium	9 (21.95%)	9 (10.84)	18	
High	0 (0.0)	0 (0.0)	0	

[a,b] Total sample size was not reached for all parameters due to unanswered questions, BIS - biological impact score, PIS - psychological impact score, SIS - social impact score, GIS - global impact score

from group B, but the differences were not statistically significant (**Table 5**).

Impact of social security registration, physician's support and medical information on the child's and parent's QOL. Although the proportion of children/families benefiting from social security registration was very small (6.7%), we attempted to assess the effectiveness of this social support in reducing the impact of CHD on QOL. The following parameters were investigated: children's BIS, PIS, and SIS, and both parents' SIS and GIS. Results were not statistically significant, but globally showed lesser proportions of high impact scores in the group with social security (**Table 6**).

We also analyzed the impact of physicians' support (including emotional, spiritual or educational support, and/or referral to adapted services) on the QOL of parents. A statistically significant difference was only observed in the biological dimension, where the proportion of parents (10.4%) with moderate to severe biological impact (BIS >30%) was lesser in families who declared being supported by the physician, versus those (21.1%) who declared not being supported (p<0.001; **Table 6**).

Impact of knowledge and lack of knowledge on the disease on QOL. Of the total interviewed families, 83 (46.1%) estimated that they lacked of, or still needed information regarding the disease of their children. We assessed the impact of knowledge and lack of knowledge on QOL. In families lacking knowledge, 43.7% of children had a medium or high impact of CHD on their global QOL (GIS >30%) versus only 22.5% in families who had sufficient knowledge on the disease (p=0.016). Distinct analysis for each dimension (biological, psychological, and social) revealed no significant correlations (**Table 6**). In linear regression, the amount of medical information needed showed a positive correlation with both BIS (B=0.014, p=0.049)

Table 5 - Impact of congenital heart disease (CHD) on quality of life (QOL) of mothers, according to being exclusive care-givers or not.

Mother's QOL parameter	Mother exclusive care-giver		Total	*P*-value
	Yes (group A; n=156[a])	No (group B; n=24[b])		
BIS				
Level of impact				0.405
Low	99 (63.9)	18 (62.07)	117	
Medium	26 (16.8)	4 (13.79)	30	
High	30 (19.35)	2 (6.9)	32	
PIS				0.469
Level of impact				
Low	102 (65.4)	18 (78.26)	120	
Medium	42 (26.9)	4 (17.39)	46	
High	12 (7.69)	1 (4.38)	13	
SIS				0.305
Level of impact				
Low	112 (71.8)	20 (83.3)	132	
Medium	34 (21.8)	2 (8.3)	36	
High	10 (6.41)	2 (8.3)	12	
Spiritual life				0.007*
Improved	78 (50.6)	3 (13.0)	81	
Unchanged	73 (47.4)	19 (82.6)	92	
Worsened	3 (2.0)	1 (4.4)	4	
Social activities impacted				0.014*
No	101 (66.0)	20 (90.9)	121	
Yes	52 (34.0)	2 (9.1)	54	
GIS				
Level of impact				0.465
Low	105 (67.7)	18 (78.3)	123	
Medium	44 (28.4)	5 (21.7)	49	
High	6 (3.9)	0 (0)	6	

[a,b] Total sample size was not reached for all parameters because of unanswered questions, BIS - biological impact score, PIS - psychological impact score, SIS - social impact score, GIS - global impact score, * significant result (p-value =<0.05)

Table 6 - Impact of social registration, physician's support, and lack of knowledge related to the disease on children's and parents' quality of life.

Parameter	Social registration		*P*-value	Physician's support		*P*-value	Lack of knowledge		*P*-value
	No (n=168[a])	No (n=12)		No	Yes		No	yes	
Child's BIS									0.093
Level of impact									
Low	108 (65.0)	9 (75.0)	0.680	-	-	-	61 (73.5)	57 (59.4)	
Medium	33 (19.9)	1 (8.3)		-	-		14 (16.9)	20 (20.8)	
High	25 (15.1)	2 (16.7)		-	-		8 (9.6)	19 (19.8)	
Child's PIS									0.187
Level of impact									
Low	105 (46.4)	10 (83.4)	0.560	-	-	-	60 (73.2)	57 (60.6)	
Medium	40 (24.5)	1 (8.3)		-	-		14 (17.1)	26 (27.6)	
High	18 (11.1)	1 (8.3)		-	-		8 (9.7)	11 (11.8)	
Child's SIS									0.929
Level of impact									
Low	102 (76.1)	9 (75.0)	0.691	-	-	-	52 (77.6)	60 (75)	
Medium	22 (16.4)	1 (8.3)		-	-		10 (14.9)	13 (16.25)	
High	10 (7.5)	2 (16.7)		-	-		5 (7.5)	7 (8.75)	
Child's GIS									0.016*
Level of impact									
Low	-	-	-	32 (58.2)	63 (70.8)	0.348	52 (77.61)	45 (56.25)	
Medium	-	-		21 (38.2)	21 (23.6)		14 (21)	29 (36.25)	
High	-	-		2 (3.6)	5 (5.6)		1 (1.5)	6 (7.5)	
Parent's PIS									-
Level of impact									
Low	-	-	-	46 (65.7)	87 (82.9)	0.105	-	-	
Medium	-	-		21 (30.0)	14 (13.3)		-	-	
High	-	-		3 (4.3)	4 (3.8)		-	-	
Parent's SIS									-
Level of impact									
Low	139 (83.7)	12 (100)	0.613	56 (78.8)	94 (89.5)	0.001*	-	-	
Medium	24 (14.5)	0 (0)		13 (18.3)	11 (10.4)		-	-	
High	3 (1.8)	0 (0)		2 (2.8)	0 (0)		-	-	
Parent's GIS									0.731
Level of impact									
Low	138 (84.1)	11 (91.7)	0.653	56 (81.2)	92 (87.6)	0.437	70 (84.3)	81 (86.2)	
Medium	26 (15.9)	1 (8.3)		13 (18.8)	13 (12.4)		13 (15.7)	13 (13.8)	
High	0 (0)	0 (0)		0 (0)	0 (0)		0 (0)	0 (0)	

[a] Total sample size was not reached for all parameters because of unanswered questions, BIS - biological impact score, PIS - psychological impact score, SIS - social impact score, GIS - global impact score, Low: <30; Medium: 30-60; High: >60, * Statistically significant result (p<0.05)

and PIS (B=0.030, p=0.003), but an inverse correlation with SIS (B=-0.018, p=0.027).

Efficacy of the medical information given to families. In order to assess the efficacy of the medical information, we compared the proportion of families still lacking knowledge among those who had already received information (group A) versus those who did not receive any information (group B). We detailed this analysis as per the type of medical information, as follows: 1) generalities of the disease, 2) risk factors, 3) symptoms, 4) complications, 5) surgical procedures, and 6) medication.

Briefly, despite information received in each respective area, 15.1% of families still needed to be informed regarding the disease, 12.0% regarding its risk factors, 10.3% regarding its symptoms, 16.4% regarding its complications, 21.6% regarding the surgical procedures, and 16.6% regarding medication. On the other hand, providing specific information regarding general aspects of the disease (p<0.001), symptoms (p=0.004) and complications (p=0.001) seemed to significantly reduce the lack of knowledge.

Discussion. Survival of children with CHD after cardiac surgery is associated with long-term physical and developmental sequelae, impacting the QOL of patients, their families, and care-givers. Therefore, in the past few decades, health-related QOL has become a recognized item within the program of care of these children.[4,19,20] Although it relied principally on parents-reported QOL, our study highlighted some major issues facing families related to the management of the disease of their children, as well as their perception as relatives and care-givers. It also shed light on areas in the follow-up and management of these patients and their families that should be considered for improvement by all involved, including the medico-social teams and decision-makers.

A study by Amedro et al[21] has compared afflicted children's QOL scores, as reported by the parents or by the children themselves, with those of normal controls. They concluded that even with a relative underestimation by parents, the QOL of children with CHD is significantly affected, as compared with normal children's.[21] These conclusions support the reliability of parent-reported QOL of the afflicted children. Conversely, a European study[16] reported better QOL indicators among grown-up patients with CHD, as compared with the general population.

The first parameter we investigated in children's QOL was the recurrence of respiratory infections. Besides their particular severity in children with CHD and their life-threatening character,[22,23] recurrent respiratory infections may significantly contribute to the deterioration of QOL of the afflicted child and family. In accordance with our results, many authors report respiratory infections to be a common comorbidity in CHD, associated with frequent and prolonged hospitalizations.[23-25] Some authors even report an accentuated stress caused by these episodes of hospitalization even in families; otherwise coping well with the disease.[13] The main diagnoses of respiratory infections reported in other studies are bronchiolitis, URI and pneumonia, including otitis episodes.[17,25] Respiratory syncytial virus is reputed to be the most frequent pathogen, notably in bronchiolitis, where an immunoprophylaxis with palivuzumab has interestingly shown efficacy in reducing the severity of these episodes and the frequency of related hospitalizations.[23,26]

The other important aspect we investigated is the impairment of physical and psychosocial growth. Our study revealed an important proportion of children with impaired physical growth (29.4%) or milestone delay (39%), assessed by questioning gross motor, fine motor, social, and language skills of the child with reference to his/her appropriate milestone fixed schedule. The American Heart Association (AHA), in collaboration with the American Academy of Pediatrics, has stated that children with CHD are at high risk of developmental disorder and delay, which concerns intelligence, communication, and executive functioning, as well as both fine and gross motor skills; it also affects behavior, academic achievement, and psychosocial profile.[11] An interesting meta-analysis reported 42% of cases of neuro-developmental delay in CHD children, which is consistent with our findings. The same study reported up to 50% of cases of brain lesions found on neuroimaging of children with CHD.[27] In a literature review, the AHA estimated that 5-50% of CHD children had significant developmental impairment correlated to the degree of severity of CHD.[11] Besides the physical limitations imposed by the disease, this correlation can also be explained by the impact of the disease on the families' QOL, secondarily impacting the child's social and psycho-affective emancipation.[28] As per the AHA, effective diagnosis and management of such a prevalent and disabling condition implies periodic surveillance and screening, along with the deployment of specific educational measures for these children and their families.[11] A further improvement in operatory techniques and postoperative care could also contribute to the prevention of such disabilities.[29]

The improvement in spiritual life reported by 50.6% of the mothers who were exclusive care-givers suggests the recourse to religion as a refuge from distress, which indicates a conceivable benefit in including spiritual support in the management of CHD patients and their families. This, however, should not disregard the importance of deploying concrete solutions for supporting these families in the daily care of the afflicted child, such as specialized auxiliary agents, education centers, or recreation facilities. These solutions would further help re-socialization of the afflicted children, as well as providing respite to the person exclusively dedicated to daily care.

Regarding the social security issue, although the results were not statistically significant (due to the small number of families socially registered), we postulate that social security registration could be very helpful in improving the QOL of both afflicted children and their families. A study by Loup et al in Switzerland,[16] where health insurance is obligatory, reported that almost 15% of grown-up patients with CHD had problems with their health insurance; however, they did not report the correlated impact on QOL.[16]

Beyond these specific dimensions, the assessment of the global QOL revealed disproportional scores within family members, especially between fathers and mothers. Mothers displayed remarkably higher impact scores that were closer to the afflicted children's scores. These observations should take into account that in most cases, mothers were the only interviewed person in the family and responded to all parts of the questionnaire. This forcibly introduced subjectivity to the answers. On the other hand, the predominant position of the mothers in the category of exclusive care-givers may contribute to this discrepancy.

Regarding the impact on the family, it has been reported that most families cope well with the disease of their child.[13] Our study confirmed that only 18.9% of the mothers, and 9.4% of the fathers had difficulty

coping with the disease of their child, while no case of high impact was found in both the parents or families global scores. A Saudi study by Almesned et al,[30] investigated the impact of the severity of the disease on the families' QOL and revealed an increased impact in cases of complex CHD. Although not statistically significant, our results suggest a similar conclusion for both children's and families' parameters, particularly in the biological dimension and global aspect of QOL. To draw solid conclusions, this aspect should be further investigated using a disease-specific instrument, such as the PCQLI that showed significant discrimination in the QOL scores across levels of CHD severity.[15]

The last important dimension that we investigated was the parents' level of knowledge on the disease, in parallel with the specific information provided by the healthcare professionals. The lack of knowledge on the disease was significantly associated with a higher impact on the child's global QOL. By contrast, providing information regarding the disease, its symptoms, and complications contributed significantly to improving the knowledge of the families. Understanding CHD and all related medical issues probably contributes to parents coping better with the disease of their children, offering them more appropriate care. Several studies[31,32] in parents of CHD children have demonstrated that many of them seek information even through the internet, with the risk of relying on non-accurate sources. These findings highlight the importance of communicating with these parents and regularly assessing their level of knowledge, in order to provide them with accurate, timely, and useful information.

The main limitation of our study is the use of a non-consensual questionnaire, although qualitatively, most items used were obtained from either generic or disease-specific questionnaires, and reliability measurement gave an acceptable level of Cronbach's alpha. Furthermore, the interpretation of the impact scores lacks accuracy because there was no control group. The other limitation is the reliance on mother-reported assessments for all the family members.

In conclusion, CHD is associated with a high prevalence of neuro-developmental delay and psychosocial disabilities with significant impact on all dimensions of QOL of the afflicted child and his family members. As a consequence, the investigation of QOL becomes a crucial element in the management of children with CHD and their families. Healthcare providers and politicians involved in healthcare are invited to review routine, medically based cost-efficacy, and to enlarge the care strategy to include non-medical care, giving priority for the improvement of the QOL of these children and their families. Physicians should allocate adequate time and efforts to raise the parents' awareness and education regarding the adequate management of the disease of their children, and to listen to their concerns and worries in order to absorb their anxiety and provide them with the useful advice. The government should encourage and implement systemic social and psychological support to these families, including social security registrations; and set up community-based educational facilities to help social integration of the afflicted children with their families. Spiritual support can be of great help in our local population and could be integrated in a multidisciplinary care approach.

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