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Original Article

Disease-related difficulties and satisfaction with level of knowledge in adults with mild or complex congenital heart disease

Mascha Kamphuis,[1,2,3,5] S. Pauline Verloove-Vanhorick,[1,4,5] Ton Vogels,[5] Jaap Ottenkamp,[1,2,6] Hubert W. Vliegen[1,3]

[1]Leiden University Medical Centre, [2]Department of Paediatric Cardiology, [3]Department of Cardiology, [4]Department of Paediatrics, Leiden; [5]TNO Prevention and Health, Leiden; [6]Emma Children's Hospital, AMC, Amsterdam, The Netherlands

Abstract *Objectives:* To evaluate difficulties in daily life, and satisfaction with level of knowledge about their disease, in patients with congenital cardiac disease in order to improve counselling. *Methods:* A self-administered questionnaire was completed by 80 patients with mild, and 76 with complex, congenital cardiac disease. They were aged from 17 to 32 years. *Results:* Even patients with only mild malformations experienced difficulties related to their disease, but being found in only 11%, these were significantly less than those uncovered in 87% of those with complex disease ($p < 0.001$). Those patients with complex malformations frequently felt restricted in choices because of their disease in areas such as sport (59%), employment (51%), and education (34%). Other difficulties reported were: paying a higher premium for life insurance (29%), having to give up on a sport (28%), and being excluded from a job (18%). Depending on the item, between one-fifth and two-thirds of participants reported gaps in knowledge, most frequently for "causes of congenital cardiac disease", "future consequences", and "family planning". For 53% of those with mild anomalies, and 93% of severely affected patients, the cardiologist is the most important source of information. *Conclusions:* A minority of adults with mild, and a majority of those with complex congenital cardiac disease report difficulties in daily life. A substantial number of these patients feel that they have an inadequate level of knowledge about their disease. Our results suggest the need for a specific programme of counselling.

Keywords: Disease-related difficulties; knowledge; congenital heart disease; adults

Since the first operation for correction of a congenital cardiac malformation was performed in 1945, technologies have developed, and survival of patients has increased, so that now there is a substantial number of adults with congenital cardiac disease.[1] Consequently, restrictions related to the disease, and future perspectives, have become increasingly important. Few previous studies have evaluated difficulties in daily life of such adults with congenital cardiac disease. These studies were mainly focussed on employability and insurability.[2–10] Difficulties in other areas of life, such as sport or having children, have hardly been studied.

Understanding of the chronic illness is associated with less distress and confusion, more satisfaction with medical care, better compliance with treatment, and an improved emotional state.[11] With new laws, and increasing emancipation of patients, the demand for education increases. New sources of information, such as the Internet, have changed the needs of adults with congenital cardiac disease. The level of knowledge of such patients with congenital cardiac anomalies has also been examined in only a few studies. Some evaluated what the patient was supposed to know according to the researchers or

Correspondence to: J. Ottenkamp PhD MD, Leiden University Medical Centre, Dept J-6 S188, PO Box 9600, 2300 RC Leiden, The Netherlands. Tel: +31 71 526 2838; Fax: +31 71 524 8110; E-mail: j.ottenkamp@lumc.nl

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physicians,[11–18] while others focussed on the satisfaction of the patients with their own level of knowledge.[9,10,16,19,20]

We carried out our cross-sectional study in patients with mild and in patients with complex malformations with the following objectives:

- To assess difficulties in daily life related to the cardiac anomaly.
- To assess the satisfaction of the patients with level of knowledge about their disease.
- To assess the sources of information concerning their health used by these patients.

Methods

All participating patients gave written informed consent. The study was approved by the local medical ethical committee.

Table 1. Participating patients: main congenital cardiac disease groups.

Mild lesions (80)
Spontaneous resolution (36)
Persisting lesions (44)
Ventricular septal defect (19)
Pulmonary stenosis (7)
Atrial septal defect (6)
Aortic stenosis (6)
Bifoliate aortic valve (3)
Abnormal pulmonary venous drainage (2)
Prolapsing mitral valve (1)
Complex lesions (76)
Systemic morphologically right ventricle (38)
Conduit or mechanical prosthesis (22)
Univentricular atrioventricular connection – Fontan (11)
Palliative operation – shunt (5)

Selection of patients

There is information on 4383 patients with congenital cardiac disease, born between 1968 and 1982, in the archives of the Department of Paediatric Cardiology at Leiden University Medical Centre. Of these patients, 2280 were randomly selected. Only those with mild or complex malformations were included in the study, resulting in a sample of 500 patients. Patients with mild anomalies had not been submitted to surgery, either because their cardiac anomaly was insignificant, or because it had disappeared spontaneously. Patients with complex malformations had undergone operative procedures that had not led to an anatomically normal heart as described by segmental analysis.[21] Information on this sample of 500 patients was received from hospitals, general practitioners, and local authorities. Of those 500 patients, 128 had died before the study started. Additional criterions for exclusion were:

- Mental retardation in 51 patients.
- Not speaking Dutch or not living in the Netherlands in 46.
- Joined a previous study in 44.[22]

Another 7 patients were lost to follow-up. This left 224 patients who were invited to participate in the study. Their diagnoses and characteristics are given in Tables 1 and 2.

Medical data

Information on the medical history was searched for in the medical files. If clinical information was older than one year, patients were re-examined to confirm their cardiac diagnosis.

Table 2. Characteristics of patients.

	Mild lesions (80)	Complex lesions (76)
Age: mean, median, range	24.6, 25, 17–31 yr	24.3, 24.5, 18–31 yr
Male	30 (38%)	43 (57%)
Medical care > 1 year ago	67 (84%)	6 (8%)
Highest education finished		
Primary education	21 (26%)	32 (42%)
Continued education	59 (74%)	44 (58%)
Daily life		
Employed (more than 12 hours/week)	61 (76%)	43 (57%)
+ disablement benefit partial		2 (3%)
Student	13 (16%)	11 (15%)
+ disablement benefit fully		4 (5%)
Disablement benefit, no activities	–	10 (13%)
On sick leave (whiplash injury/pregnancy)	–	2 (3%)
Housewife	2 (3%)	1 (1%)
+ disablement benefit fully	2 (not related to cardiac disease)	1 (1%)
Unemployed	1 (1%)	2 (3%)
+ disablement benefit partial	1 (not related to cardiac disease)	–

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Questionnaire

All participants completed a self-administered questionnaire. It encompassed the following topics:

Difficulties in daily life. We included 26 items based on previous studies,[7–9,19,23] our pilot study,[22] and clinical practice. The items covered different subjects such as life/health insurance, education, employment, sport, and other daily activities (Table 3). If a difficulty was noted, it was specifically asked whether this was due to the cardiac disorder. Results are presented in total numbers and percentages of patients who felt restricted because of their congenital heart disease (Table 3).

Table 3. Difficulties in daily life related to the congenital cardiac malformation. All differences between those patients with mild and complex lesions were highly significant: $p < 0.001$.

Items	Mild lesions N (%)	Complex lesions N (%)
Did you ever feel restricted in the choice of		
An educational course?		26 (34)
A job?		39 (51)
A sport?	5 (6)	45 (59)
A hobby?		9 (12)
A house?	1 (1)	15 (20)
A holidays destination?		20 (26)
Did you ever give up		
An educational course?		4 (5)
A job?		12 (16)
A sport?	2 (3)	21 (28)
A hobby?		2 (3)
A holiday?		7 (9)
Did your physician ever advised you not to have children?		11 (14)
Did you ever thought it was better not to have children?		22 (29)
Were you ever excluded from		
An educational course?	1 (1)	4 (5)
A job?		14 (18)
Possible promotion?		2 (3)
A job after medical examination?	1 (1)	6 (8)
Sport after medical examination?	1 (1)	1 (1)
Where you ever prevented from		
Applying from a driving licence?		6 (8)
Taking out a mortgage policy?		6 (8)
Taking out a life insurance policy?		13 (17)
Taking out a health insurance policy?	3 (4)	3 (4)
Did you ever had to pay higher premium than other people for		
A mortgage?	1 (1)	6 (8)
A life insurance?	2 (3)	22 (29)
A health insurance?	1 (1)	7 (9)
Total patients, who experience difficulties for at least one of the items	9 (11)	66 (87)

Table 4. Satisfaction with level of knowledge.

	Percentages of patients answering "not enough"		
Do you know enough about …	Mild lesions	Complex lesions	Significance ($p < 0.01$)
Future consequences of your cardiac disease?	55	64	
Consequences of your cardiac disease for family planning?	45	51	
Causes of your cardiac disease?	61	43	Yes
What kind of cardiac disease you have?	51	37	
Consequences of your cardiac disease for occupation/career?	39	32	
Consequences of your cardiac disease for exercise/sport?	45	30	Yes
Consequences of your cardiac disease for your education?	39	26	Yes
Things to do to remain healthy?	36	24	
Consequences of your cardiac disease for spending free time?	35	20	Yes
Percentage of patients who feel that their level of knowledge is insufficient	35–61	20–64	

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Level of knowledge about disease. Satisfaction with level of knowledge: Patients were asked if they felt that they had sufficient knowledge concerning 9 different subjects (Table 4). The subjects were again based on our pilot study.[22] At the end of the questionnaire, an open question was added: "What subject concerning your heart disease would you like to know more about?".

Sources of information. We asked which sources of information were used to obtain information about personal health.

Statistics

Frequencies were run with the SPSS 10.0 package. The group of patients with complex malformations was compared with that with mild disease using the Mann-Whitney U test for comparison between groups, and the Kruskall-Wallis test for comparison of ordinal data from multiple groups.

Results

Patients

Of the 224 selected patients, 160 agreed to participate (71%). Of these, 4 patients did not return their questionnaire, so results were based on a sample of 156 patients. The response rate in the group of patients with complex disease (88%) was higher compared with that in the group of patients with a minor malformation (59%). Diagnoses and characteristics of the participants are listed in Tables 1 and 2.

Difficulties in daily life

The difficulties identified as directly related to the congenital malformation are listed in Table 3. In total, 11% of the patients with mild malformations reported one or more difficulties compared with 87% in patients with complex anomalies. All differences in percentages per item between those with mild and complex anomalies were highly significant ($p < 0.001$).

In the group with complex anomalies, only 10 patients did not mention any difficulties related to the disease. These were 7 patients with a systemic morphologically right ventricle, accounting for 18% of the sample, one with a conduit or mechanical prosthesis, accounting for 5%, one with a functionally univentricular heart after a Fontan procedure, 9% of this group, and one of the five who had undergone a palliative shunt procedure.

For each subject, we present most relevant difficulties for the group of patients with complex lesions due to this congenital malformation.

Life insurance. Overall, 27 patients (36%) had problems with taking out a life insurance policy: 5 were rejected, 14 had to pay a higher premium, and 8 experienced both problems.

Employment. Besides 14 patients (18%) who were excluded from a job, 12 patients (16%) had given up on a job because of their cardiac disease. Six of them were excluded after medical examination. Since 6 patients were both rejected and had given up, altogether 20 patients (26%) had been restricted in employment. In addition, two patients were excluded from a possible promotion. Of the group, 45 (59%) were in paid employment, compared with 61 (76%) of those with mild malformations ($p < 0.001$).

Education. Altogether, 6 patients had either given up on an education, or had been rejected because of their cardiac anomaly (8%).

Sport. In total, 21 patients had given up on sport (28%), with the highest percentages in patients who had undergone a palliative shunt procedure or else those with a functionally univentricular heart. Only 1 patient was rejected after medical examination.

Children. Of the patients, 22 (29%) think that they "better not have children", while 11 (15%) were "advised not to have children by their physician". More than one-quarter of patients with functionally univentricular heart were advised not to have children.

Other subjects. Restriction in the choice of their holiday destination was reported by 20 patients (26%), 6 patients were rejected for a driving licence (8%), and 2 gave up on a hobby (3%). Altogether, 15 patients felt restricted in the choice of a house (20%). Six of them were also rejected for mortgage and life insurance, and these factors could have played a role in the restriction.

Level of knowledge about disease

Satisfaction with level of knowledge (Table 4). Depending on the subject, from one-third to three-fifths of the patients with mild malformations indicated insufficient knowledge, with the item "causes of your cardiac disease" as the most important to know more about, with 61% wanting to know more. This is followed by "future consequences", with 55% looking for more information. Some additional remarks from those with mild anomalies were "Will the defect increase with pregnancy?"; "How long do I need medical care?"; and "What about the anaesthesia at the dentist?".

Between one-fifth and two-thirds of those with complex malformations reported problems. The items "future consequences" and "family planning" were the items most identified as the topics for more information, identified by 64% and 51%, respectively. These items were also mentioned in the answers to the open question by about one-fifth of the responders. Remarks from the patients with complex malformations were: "Is there an association for patients with congenital heart disease?"; and "Do you have information on heart transplantation, living together, insurance?". For all subjects, except for

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"future consequences" and "consequences for family planning", patients with mild anomalies made greater demands for thorough information than those with complex lesions. This difference is significant for 4 of the items (Table 4).

Sources of information. For 53% of those patients with mild lesions, and 93% of those severely affected, the cardiologist is the most important source of information concerning their health. In addition, general practitioners inform about one-third of both groups. Parents and friends also play an important role in the dissemination of information in from one-tenth to one-fifth. Television was the source of information for 10% of those with mild, and 15% of those with complex anomalies. For patients with complex malformations, the patient's association was the source of information for 18%, and the Internet for 7%. Other sources that were mentioned were magazines/papers, the psychologist, and the library.

Discussion

Our study has shown that a minority of patients with mild lesions, and a majority of those with complex anomalies, experience difficulties in daily life directly related to their cardiac disease. In addition, a substantial number of patients felt that they were insufficiently informed about their disease.

Difficulties in daily life

Studies that have focussed on difficulties in daily life for patients with congenital cardiac disease are few. Data published has mainly concerned problems related to employment or availability of insurance.[2–10] In our study, nearly one-third of patients with complex malformations reported difficulties in employment, and just over half felt restricted in the choice of a job because of their cardiac disease. Previous studies concluded that between one-tenth and two-fifths of the patients, depending on the lesions studied, were not able to have the job they wanted. The percentage mentioned in the study by Ghisla et al.,[5] describing a group of patients with complex malformations (tetralogy of Fallot) accounting for over two-fifths, is comparable to that found in our study. For life insurance, just over one-third of our patients with complex malformations were either rejected for cover, or had to pay higher premium than usual. Other groups have reported about one-third of patients being rejected for life insurance.[4,7] It has to be taken into account that not all patients had applied for life insurance yet because of the age range chosen for the study and, therefore, this proportion may well increase with advancing age. This might apply for all other items as well.

Level of knowledge about disease

In our study, from one-fifth to two-thirds of the patients, depending on the item, felt themselves insufficiently informed. Other studies have also shown a substantial percentage of patients wanting more information about their disease. Mühler et al.,[9] and Otterstad et al.,[19] reported that one-third and three-fifths of the patients, respectively, felt insufficiently informed. Wright et al.[10] found proportions between one-third and three-quarters depending on the item. Differences may be explained by the use of slightly different items. Percentages for the items: "… consequences of your cardiac disease for family planning"; "… causes of your cardiac disease?/what kind of cardiac disease do you have?" and "… consequences of your cardiac disease for exercise/sport?" in our study, nonetheless, were remarkably similar to those found by Wright et al.[10] We can, therefore, conclude that, over a period of 14 years, the need by patients for knowledge has not decreased, and their education has still to be improved.

A more recent study showed that a high percentage of 96.8% of patients was satisfied with the information given about their congenital cardiac malformation.[16] This cannot be explained by better objective understanding, since the study also identified important gaps in their knowledge. The high percentage of satisfaction might be the result of more attention for counselling, with the help of a nurse practitioner who is working in the University Hospital. Another explanation may be that the reported high satisfaction is biased by the fact that the researcher questioned the patient directly. Although it is not possible to prove which argument is true, these results emphasise the difference between true understanding of the illness and satisfaction with knowledge about the disease. Both need to be improved.

How to prevent difficulties and improve satisfaction with level of knowledge?

To prevent unnecessary difficulties, and to close gaps in knowledge, counselling should be optimised. Subjects, such as vocational choice, should be discussed at an early age. Specific advice should be given to the patients. They should be encouraged to apply for insurance from several companies, or to seek assistance from an independent insurance agent. Job training, or career counselling, can be offered. In case of doubts about advice, re-assessment has to take place.[24–26] Moreover, all information should be reinforced, since the recall of the patients is disappointing.[17] Regular follow-up, and a "health passport" in which information about diagnosis and operation, exercise prescription, family and career planning is summarised, could prove particularly helpful.[27]

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Methodological issues

The sample of patients included in our study may not be fully representative for the overall population of patients with congenital cardiac disease. Only three-fifths responded from the group of patients with mild lesions. Non-participants lived further away from the hospital than participants, but this is not likely to have influenced the results in this study. Since the percentage of participating women and men responding was different, the bias of those not responding may have affected the results. During cardiac re-examination, it was observed that most patients asked questions, or expressed worries, that they had lived with for several years. It is possible that patients with relatively more difficulties or questions joined the study, influencing results towards more difficulties and gaps in knowledge in the group of patients with mild disease.

In conclusion, if in future we are to prevent difficulties and improve satisfaction with level of knowledge about their disease, we must develop a dedicated programme of counselling for adults with congenital cardiac disease.

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