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ORIGINAL ARTICLE
Adult congenital heart disease

Life expectancy and end-of-life communication in adult patients with congenital heart disease, 40–53 years after surgery

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Aims

Although survival of patients with congenital heart disease (CHD) improved significantly over time, life expectancy is still not normal. We aimed to investigate how adult patients, their partners, and treating cardiologists estimated the individual life expectancy of CHD patients. Furthermore, preferences regarding end-of-life (EOL) communication were investigated.

Methods and results

In this study, we included 202 patients (age: 50 ± 5) who were operated in childhood (<15 years old) between 1968 and 1980 for one of the following diagnoses: atrial septal defect, ventricular septal defect, pulmonary stenosis, tetralogy of Fallot, or transposition of the great arteries. A specific questionnaire was administered to both the patients and their partners, exploring their perceived life expectancy and EOL wishes. Two cardiologists independently assessed the life expectancy of each patient. Most adults with CHD believed their life expectancy to be normal. However, significant differences were found between estimated life expectancy by the cardiologist and patients (female: $P = 0.001$, male: $P = 0.002$) with moderate/severe defects, as well as for males with mild defects ($P = 0.011$). Regarding EOL communication, 85.1% of the patients reported that they never discussed EOL with a healthcare professional. Compared with patients with mild CHD, significantly more patients with moderate/severe defect discussed EOL with a physician ($P = 0.011$). The wish to discuss EOL with the cardiologist was reported by 49.3% of the patients and 41.7% of their partners.

Conclusion

Adult patients, especially with moderate/severe CHD, perceived their life expectancy as normal, whereas cardiologists had a more pessimistic view than their patients. Increased attention is warranted for discussions on life expectancy and EOL to improve patient-tailored care.

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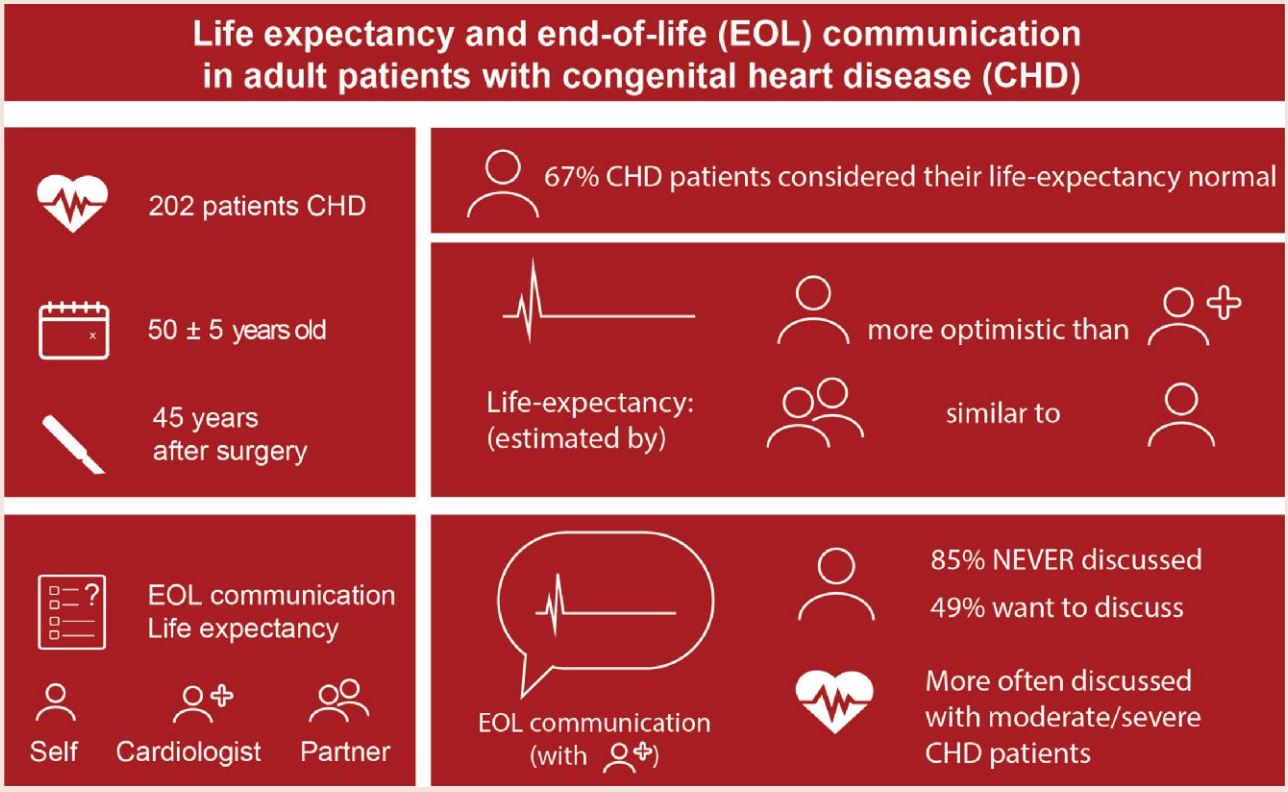
Graphical Abstract



Keywords Congenital heart disease • end-of-life • psychology • long-term follow-up • life expectancy

Introduction

Although the majority of infants born with congenital heart disease (CHD) survive into adulthood, overall life expectancy is reduced, especially in complex CHD.[1] Residual lesions and late complications such as arrhythmias, ventricular dysfunction, and need for re-intervention are common, especially in adulthood.[1,2] However, a recent Swedish study showed that at least 75% of patients who were alive at the age of 18 survived after 50 years of follow-up.[3] Despite constant progress in medical care, mortality is still higher than in the general population, although there is variance between diagnoses and higher mortality rates among patients with more severe residual lesions.[3–5] Previous studies identified heart failure and sudden cardiac death as the main causes of death.[5] In literature, life expectancy varies depending on the underlying diagnosis. Patients with a patent ductus arteriosus, on one side of the spectrum, have a (nearly) normal life expectancy, whereas patients with a univentricular heart clearly have more substantial reduction of their life expectancy. Patients with a systemic right ventricle have a risk of around 60% of developing heart failure by the age of 40 and clearly have a lower life expectancy compared to the normal population, although information on very long-term outcome is not yet available.[6] Despite the reduced life expectancy of patients with CHD, adolescents and young adults with moderate/severe defects expect to have equal life expectancy compared with their healthy peers.[7,8] These relatively young patients are still in the so called 'honeymoon' phase, living a fairly normal life in which complications may not have occurred yet. The older they get the higher the risk of complications. In addition, surgical techniques and newly developed percutaneous procedures have improved over time and these relatively recently operated patients probably will do better than patients operated in the 60s and 70s. These factors could affect their positive view on life perspectives. Until now, no study focused on the estimation of life expectancy in adults with CHD. Self-perceived life expectancy is fundamental for planning life achievements and for setting goals, such as pursuing a career and having a family and children, and for retirement planning. Patients with a reduced life expectancy will also reach end-stage heart disease prematurely when medical care no longer can reverse their deteriorating condition. In this setting, communication regarding end of life (EOL) is fundamental. In the position paper of Schwerzmann *et al.*, it was affirmed that starting early with EOL communications is crucial.[7] These discussions are not only fundamental for managing expectations but also for establishing a written plan. Such a plan holds significant benefits not only for patients and their partners but also for cardiologists, general physicians, family members, and other individuals involved in the care process. Literature on EOL communication studies is scarce and one previous study showed that only 1% of the CHD patients discussed EOL with their healthcare providers.[8]

The purpose of our study was to assess for the first time the self-, partner-, and cardiologist-perceived life expectancy of middle-aged adults with CHD per sex as fundamental for the planning of life of the patient with CHD. In addition, we aimed to assess the perceived

and the desired EOL communication in patients and their partners to optimize future care.

Methods

Patient sample

A longitudinal study was started four decades ago, investigating a cohort of patients with CHD who underwent cardiothoracic surgery between 1968 and 1980 at young age (<15 years) at the Erasmus MC (Rotterdam, the Netherlands). The original cohort consisted of 597 consecutive patients diagnosed with atrial septal defect (ASD), ventricular septal defect (VSD), pulmonary stenosis (PS), tetralogy of Fallot (ToF), or transposition of the great arteries (TGA). During follow-up (>40 years), the original cohort was investigated every 10 years (1991, 2001, 2011, and 2021).

The target population of the current follow-up consists of the 431 patients who were still alive, but only the 343 patients who participated in at least two previous follow-up studies and were traceable were invited to participate in the current study. Of those, 201 participated (59%). (Supplementary material online, *Figure S1*). Patients ($n = 201$) were classified in two groups according to the European Society of Cardiology classification: mild CHD ($n = 138$),[9] including ASD, VSD, and PS and moderate/severe CHD ($n = 63$), including corrected ToF and TGA after Mustard procedure.[9] The research protocol was approved by the local institutional ethical committee (MEC-2019 0465) and followed the ethical guidelines of the 1975 Declaration of Helsinki. All patients were approached uniformly and before participating in the study, written informed consent was obtained.

Study design

All patients received the questionnaires via email and completed them digitally at home via a privacy-protected website (GemsTracker, Copyright©2011, Erasmus MC and Equipe Healthcare companies) before their visit at the outpatient clinic of the Erasmus University Medical Center. Due to delay in producing the digital questionnaires or to personal reasons (e.g. no access to email), 44 patients and their partners completed the paper version. Patients visited our outpatient clinic where a cardiologist performed cardiac and medical examinations.

Instruments

In this study, a structured questionnaire was used to assess life expectancy and EOL communication.[10] This questionnaire was specifically designed for the current study and was based on the work of Lyon *et al.*[11] The questionnaire consists of two parts: one regarding the life expectancy and the other on the experienced and wish need to discuss EOL care with healthcare professionals. A translation of the questionnaire is provided as a Supplemental material online, attachment 2_questionnaire (EN translation).

Patients and their partners assessed the life expectancy of the patients as 'normal', 'mildly reduced (<10 years)', 'reduced (10–20 years)', or 'strongly reduced (>20 years)' and two cardiologists (J.C. and R.K.) also assessed the life expectancy upon clinical judgement. A third cardiologist (J.R.) was involved to solve cases of disagreement. Furthermore, patients and cardiologists were asked about the specific life expectancy in years ('How old do you think you/your patient will become?') (Utens structured interview). Cardiologists assessed life expectancy of the patients in comparison of the general Dutch population of the same age and sex.[12]

Questions regarding experience, timing, and willingness to communicate about EOL were asked to both patients and their partners.[10]

Statistical analysis

Baseline medical characteristics of participants were illustrated with descriptive statistics: categorical data were presented as percentages (frequencies) and continuous data as mean ± standard deviation. Normal distribution was assessed with Shapiro–Wilk test and histograms. In case of non-normal distribution, median and 25th–75th percentile were shown. Differences between diagnostic groups were analysed with independent *t*-test, Mann–Whitney *U* test, or one-way analysis of variance (ANOVA) when appropriate for continuous data. The χ^2 test or, in case of paired data, the Stuart–Maxwell test was used to assess differences between categorical data.

A logistic regression corrected per age and gender was used to assess the association between the patient's life expectancy estimation and the wish to discuss EOL with the cardiologist.

Statistical analyses were processed using IBM SPSS Statistics for Windows 28.0 (Armonk, New York, USA) and R software v. 4.2.1 for Windows.

Results

Group characteristics

Demographic and clinical characteristics are presented in *Table 1*.

Our cohort consists of 201 patients (46% females) with a mean age of 50.0 ± 5.1 years. Compared to patients with mild CHD, patients with moderate/severe CHD were younger ($P < 0.001$), underwent surgery at a younger age ($P < 0.001$), and suffered more frequently from heart failure ($P < 0.001$). In addition, they were taking more often cardiac medication ($P < 0.001$), showed lower exercise capacity on the bicycle ergometer test ($P = 0.002$), were more frequently in New York Heart Association (NYHA) class >I ($P < 0.001$) and had more often diminished systemic ventricular function ($P < 0.001$).

Life expectancy reported by patients, their partners, and cardiologists

Figure 1 shows life expectancy reported by patients and cardiologists. In total, 67.2% of CHD patients considered their life expectancy to be normal, whereas the cardiologists considered the life expectancy to be normal in 45.3%. *Table 2* shows life expectancy per diagnostic group according to patients, partners, and cardiologists, divided per sex.

Female patients with mild CHD had a more pessimistic estimation of their life expectancy when compared to their partners ($P = 0.036$ for all categories), whereas male patients rated their life expectancy similar to their partners. Furthermore Male patients with mild CHD estimated their life expectancy more optimistically than the cardiologists ($P = 0.011$ for all categories).

Most female patients with moderate/severe CHD and their partners perceived their life expectancy to be comparable to their healthy peers. However, this was in contrast with the opinion of the cardiologists who considered that only 20.0% of them have a life expectancy comparable to their healthy peers ($P = 0.001$ for all categories). Similarly, male patients with moderate/severe CHD believed that their life expectancy was significantly higher compared to the cardiologists' estimation ($P = 0.002$ for all categories).

Mean life expectancies per diagnostic group and per sex are shown in *Figure 2*. The (red) horizontal line represents the mean life expectancy of the age- and sex-matched general population (81.4 years for males and 84.7 years for females). No significant difference was reported among the CHD diagnostic groups according to the patient's ratings. However, cardiologists rated life expectancy of the cohort significantly worse than the norm ($P < 0.001$) and different per diagnosis. Furthermore, significant differences were observed between the reports of cardiologists and male patients with both mild and moderate/severe CHD (both $P < 0.001$). Similarly, significant differences were also observed in female patients with moderate/severe CHD ($P < 0.001$) when comparing their reports to those of the cardiologists.

End-of-life communication

The 72.6% CHD patients reported that they never talked about EOL care with anyone close to them and 75.6% of them had no wish to have this discussion with their close ones. In addition, 78.6% did not

Table 1 Biographical and clinical characteristics of patients with congenital heart disease

		CHD classification		P
	Total CHD (*n* = 201)	Mild (*n* = 138)	Moderate/severe (*n* = 63)	
Biographical status:				
Female	46.0% (93)	48.6% (67)	41.3% (26)	0.360
Age at follow-up (years)	50.0 ± 5.1	50.8 ± 5.1	48.3 ± 4.8	0.001
Education level:				
Lower	33.3% (67)	33.9% (45)	34.9% (22)	0.944
Secondary	31.3% (63)	31.9% (44)	30.2% (19)	
Higher	35.3% (71)	35.5% (49)	34.9% (22)	
Medical history:				
Duration of pregnancy (weeks)	40.0 [40.0–40.0]	40.0 [39.0–40.0]	40.0 [40.0–40.0]	0.223
Weight at birth (kg)	3.2 [2.9–3.6]	3.2 [2.7–3.6]	3.3 [3.0–3.6]	0.117
First open heart surgery:				
Age at first open heart operation (years)	4.9 [1.3–7.2]	5.5 [2.1–8.4]	2.4 [0.8–5.4]	<0.001
CVA and TIA	5.0% (10)	4.3% (6)	6.3% (4)	0.728
Heart failure	4.0% (8)	0.7% (1)	11.1% (7)	<0.001
NYHA class 1	87.0% (167)	92.5% (124)	74.1% (43)	<0.001
CPET (%)[a]	98.2 ± 22.3	101.4 ± 21.4	89.6 ± 22.3	0.002
Cardiac medications	39.8% (80)	31.2% (43)	58.7% (37)	<0.001
Systemic function:				
Good	66.5% (127)	82.7% (110)	29.3% (17)	<0.001
Reasonable	22.5% (43)	14.3% (19)	41.4% (24)	
Poor	9.4% (18)	3.0% (4)	24.1% (14)	
Bad	1.6% (3)	—	5.2% (3)	

For continuous variables median [IQR] are reported. For categorical variables, percentages (*n*) are shown. Differences between diagnostic groups were analysed for the continuous data with Mann–Whitney *U* test since not normally distributed. Chi-squared test or, if necessary McNemar test, were used to analyse differences between categorical data.
CPET, Cardiopulmonary Exercise Test; CVA/TIA, cerebrovascular accident/transient ischaemic attack; NYHA, New York Heart Association.
[a]Percentage of expected max exercise capacity for healthy control adjusted per sex, height, and age.

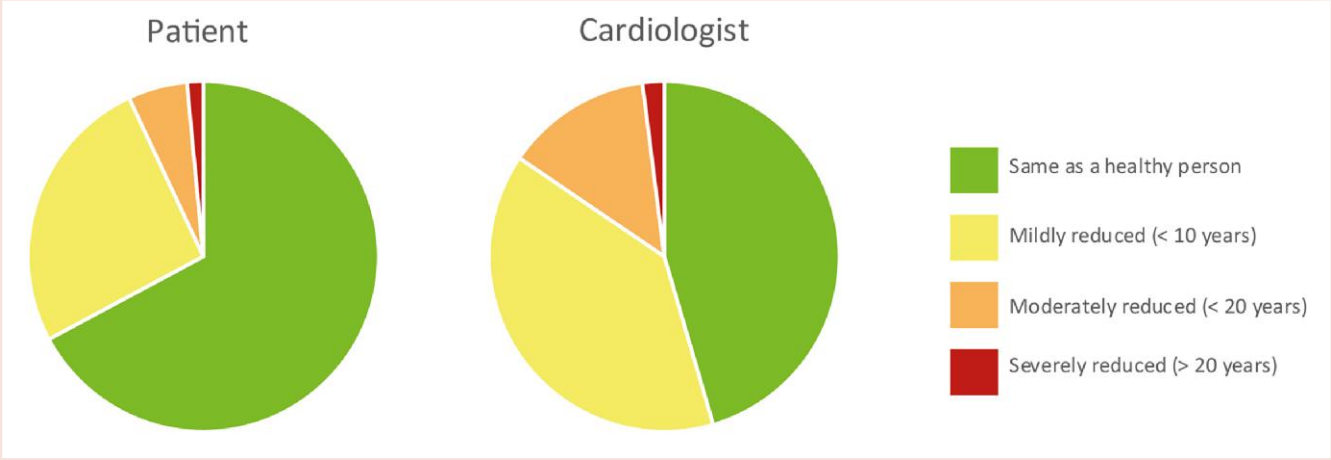


Figure 1 Life expectation according to patients and cardiologists.

think that a member of the treatment team should discuss EOL with them. Moreover, 88.4% of their partners never discussed patients' EOL with anyone close to them and 66.7% thought that a member of the treating team should not bring up questions about EOL. No significant difference between mild and moderate/severe CHD was found (*Table 3* and Supplemental material online, *Figure S2*).

Furthermore, 85.1% of the patients reported to have never discussed EOL and life expectancy with their cardiologist. However, a

Table 2 Life expectation per congenital heart disease group according to the patient, partner of the patient, and the cardiologists and differences between expectancy of life between simple and moderate/complex congenital heart disease

	Mild CHD			*P* (patient vs. cardiologist)	*P* (patient vs. partner)	Moderate/severe CHD			*P* (patient vs. cardiologist)	*P* (patient vs. partner)
	Patient (*n* = 138)	Partner (*n* = 100)	Cardiologist (*n* = 138)			Patient (*n* = 63)	Partner (*n* = 51)	Cardiologist (*n* = 63)		
FEMALE (*n*=93)				0.622	0.036				0.001	0.655
Same as a healthy person	70.1% (47)	84.9% (39)	67.2% (45)			65.4% (17)	61.1% (11)	20.0% (5)		
Mildly reduced (<10 years)	23.9% (16)	10.9% (5)	25.4% (17)			30.8% (8)	33.3% (6)	40.0% (10)		
Moderately reduced (<20 years)	4.5% (3)	—	7.5% (5)			3.8% (1)	5.6% (1)	32.0% (8)		
Severely reduced (>20 years)	1.5% (1)	4.3% (2)	—			—	—	8.0% (2)		
MALE *(n=108)*				0.011	0.215				0.002	0.256
Same as a healthy person	74.6% (53)	79.2% (42)	53.5% (38)			48.6% (18)	53.1% (17)	8.1% (3)		
Mildly reduced (<10 years)	16.9% (12)	16.7% (9)	42.3% (30)			43.2% (16)	25.0% (8)	56.8% (21)		
Moderately reduced (<20 years)	5.6% (4)	3.7% (2)	12.8% (2)			8.1% (3)	18.8% (6)	32.4% (12)		
Severely reduced (>20 years)	2.8% (2)	—	1.4% (1)			—	3.1% (1)	2.7% (1)		

Stuart–Maxwell test was used to assess differences between categorical data.

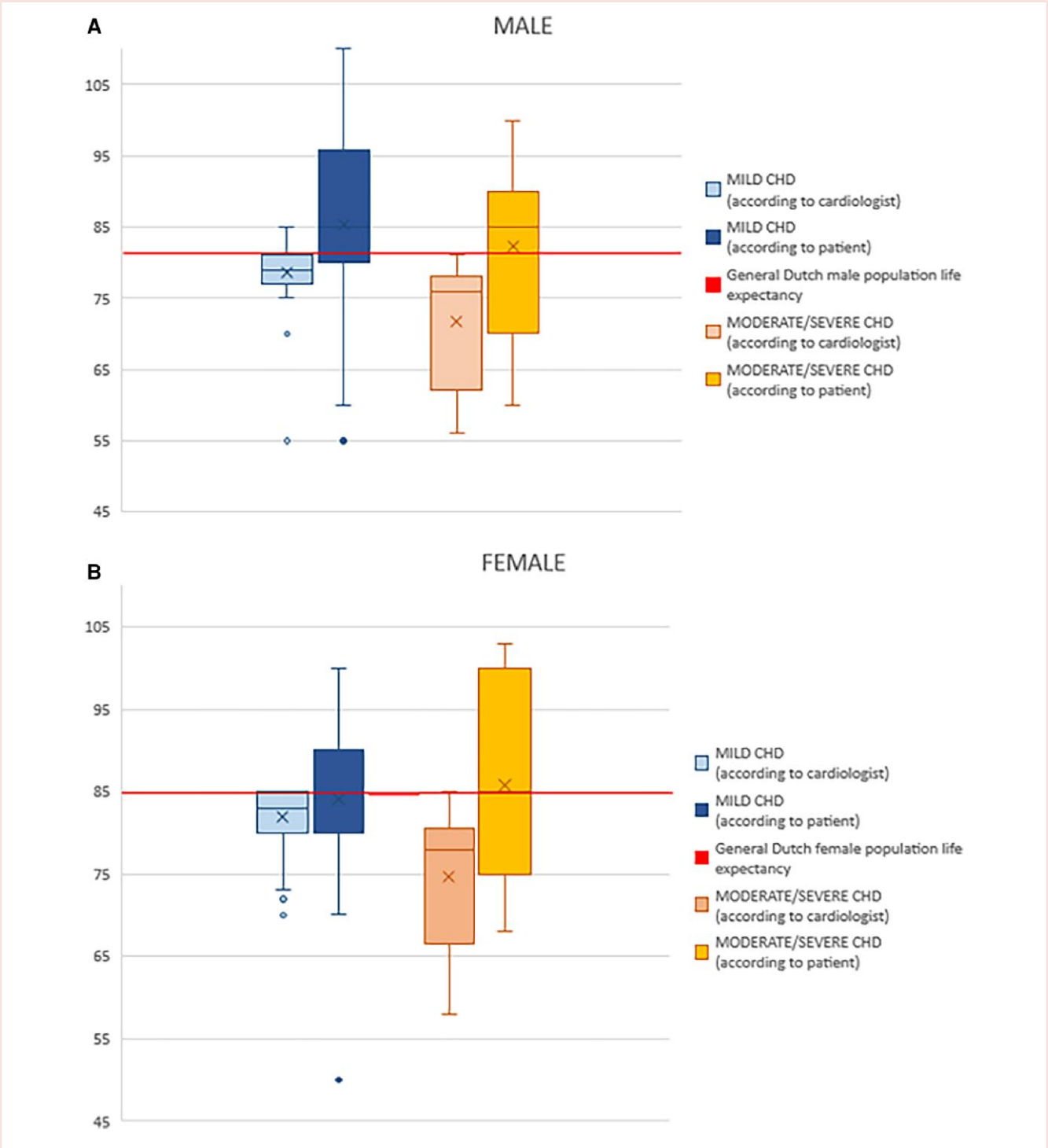


Figure 2 Whisker plot of the patient and cardiologist rated life expectancy per diagnosis: (*A*) male and (*B*) female. The (red) horizontal line represents the weighted mean of the general population life expectancy for a healthy person of same sex and age divided in mild and moderate/severe congenital heart disease.

significant difference was reported between mild and moderate/severe CHD ($P = 0.011$ for all categories: 10.1% of mild vs. 25.4% of moderate/severe CHD patients have discussed EOL and/or life expectancy with their cardiologist). Similar results were reported by their partners. Significant differences were reported between CHD groups ($P = 0.009$ for all categories). Specifically, it was found that 18.3% of partners of patients with mild CHD compared to 33.3% of partners of patients with moderate/severe CHD have discussed EOL and/or life expectancy of their partners with a cardiologist. Despite these results, 49.3% of patients and 41.7% of their partners believed that the cardiologists should

Table 3 Life expectancy and end-of-life communication

	Total group ($n = 201$)	Simple CHD ($n = 138$)	Moderate/complex CHD ($n = 63$)	*P*
Patient				
Have you ever talked to someone close to you about your wishes regarding EOL?				
Yes	27.4% (55)	27.5% (38)	27.0% (17)	0.935
No	72.6% (146)	72.5% (100)	73.0% (46)	
Would you feel comfortable talking to people close to you about your life expectancy and EOL?				
Yes	24.4% (49)	23.9% (33)	25.4% (16)	0.820
No	75.6% (152)	76.1% (105)	74.6% (47)	
Would you appreciate it if a member of your treatment team supported you in discussing your life expectancy and EOL with people close to you?				
Yes	61.2% (30)	63.6% (21)	56.3% (9)	
No	38.8% (19)	36.4% (12)	43.9% (7)	0.619
Do you think that a member of the medical team should bring up questions regarding EOL?				
Yes	21.4% (43)	23.2% (32)	17.5% (11)	0.358
No	78.6% (158)	76.8% (106)	82.5% (52)	
In your opinion, which is the best moment for one of the members of the treatment team to begin discussing EOL?				
When you are still in good health	25.6% (11)	31.3% (10)	9.1% (1)	0.414
When a life-threatening illness/cardiac complication is diagnosed	37.2% (16)	37.5% (12)	36.4% (4)	
At the first hospitalization for a life-threatening illness/cardiac complication	14.0% (6)	12.5% (4)	18.2% (2)	
During the stage of dying	—	—	—	
In every of the mentioned moments	23.3% (10)	18.8% (6)	36.4% (4)	
Have you ever discussed with your cardiologist about your questions and wishes regarding your life expectancy and EOL?				
Yes, about my life expectancy	14.4% (29)	9.4% (13)	25.4% (16)	0.011
Yes, about end of life	—	—	—	
Yes, about both	0.5% (1)	0.7% (1)		
No	85.1% (171)	89.9% (124)	74.6% (47)	
Do you think that your cardiologist should discuss your life expectancy with you?				
Yes	49.3% (99)	47.8% (66)	52.4% (33)	0.549
No	50.7% (102)	52.2% (72)	47.6% (30)	
Partner				
Have you ever talked to someone close to you about the wishes of your partner regarding his/her EOL?				
Yes	11.6% (18)	11.5% (12)	11.8%(6)	0.967
No	88.4% (137)	88.5% (92)	88.2% (45)	
Do you think that a member of the medical team of your partner should bring up questions regarding EOL?				
Yes	33.3% (6)	33.3% (4)	33.3% (2)	1
No	66.7% (12)	66.7% (8)	66.7% (4)	
In your opinion, which is the best moment for one of the members of the treatment team to begin discussing EOL of your partner?				
When you are still in good health	31.8% (7)	21.4% (3)	50.0% (4)	0.051
When a life-threatening illness/cardiac complication is diagnosed	31.8% (7)	50.0% (7)	—	

Continued

Table 3 *Continued*

	Total group ($n = 201$)	Simple CHD ($n = 138$)	Moderate/complex CHD ($n = 63$)	P
At the first hospitalization for a life-threatening illness/cardiac complication	—	—	—	
During the stage of dying	—	—	—	
In every of the mentioned moments	36.4% (8)	28.6% (4)	50.0% (4)	
Have you ever discussed your questions and wishes about the life expectancy and EOL care of your partner with your treating cardiologist, either alone or together?				
Yes, about their life expectancy	14.2% (22)	7.7% (8)	27.5% (14)	0.009
Yes, about their end of life	3.9% (6)	3.8% (4)	3.9% (2)	
Yes, about both	3.9% (6)	5.8% (6)	—	
Yes, but without my partner	1.3% (2)	1.0% (1)	2.0% (1)	
No	76.8% (119)	81.7% (85)	66.7% (34)	
Do you think that your cardiologist should discuss the life expectancy of your partner with you?				
Yes	41.7% (63)	42.0% (42)	41.2% (21)	0.923
No	58.3% (88)	58.0% (58)	58.8% (30)	
Do you find difficult to discuss life expectancy and EOL of your partner with him/her?				
Yes	19.9% (30)	17.0% (17)	25.5% (13)	0.216
No	80.1% (121)	83.0% (83)	74.5% (38)	

discuss EOL. However, it was found that the odds of patients' willing to discuss EOL with the cardiologist increased in patients who considered their life expectancy to be reduced [exp(B) = 2.05, 95% confidence interval (CI) (1.11–3.78), $P = 0.021$].

Finally, 80.1% of partners did not report difficulties in discussing EOL and life expectancy with their partners with CHD. No significant difference between CHD groups was found.

Discussion

This is the first study focusing on both the self- as well as the partner- and cardiologist-predicted life expectancy of middle-aged adults with CHD. Reid *et al*. already showed that patients with CHD expected to live only 4 years shorter that their healthy peers.[12] In our study, we found that whereas female patients with mild CHD had a similar view of their life expectancy as the cardiologists. Significant differences were found between cardiologists and respectively the ratings of female patients with moderate/severe lesions and male patients with both mild and moderate/severe CHD. In fact, even though moderate/severe CHD patients have higher mortality, the majority of them considered their life expectancy to be normal.[4,12–14] These results are in line with previous studies reporting unrealistic optimism in patients with other chronical conditions.[15,16] Patients with CHD seem not fully aware of their expected long-term outcomes. Of course, we have to bear in mind that possibly the cardiologists are too pessimistic and might still have a more conservative view on the life expectancy of this group of patients and it may result in a more negative estimation. Obviously, the patients in our study are still alive, representing a selected sample compared with deceased patients and with survival reports in literature. In fact, those patients might be the ones with better prognosis compared with the whole group. It must be considered that it was previously reported that patients can name their condition correctly and that they are aware of the importance of visits and treatments related to their diagnosis, but they had poor knowledge regarding symptoms of deterioration and description of their defect.[17,18] The poor awareness in the adult population could be the direct consequence of the poor awareness in children and adolescents with CHD. In fact, poor or wrong understanding of their condition was reported in younger patients. This might be related to an inefficient communication between physicians and patients, as during childhood, most communication is with the parents of the patients.[19–21] Since several decades, we are used to discuss the diagnosis and future expectations extensively during the transition to the adult outpatient clinic. However, information on future expectations may be expressed in veiled terms and a previous study showed that adults with CHD would like to have more information.[22] Another explanation of our findings could be that the patients and their partners are overly optimistic. They may not want to accept the negative messages and just hope and expect to live a long life. Of course, many reports have described the great achievements in CHD care in the last decades. This was also stated in the lay press and may have influenced the hopes and expectations in the future of medicine of patients with CHD. Self-perceived life expectancy plays a crucial role in shaping life plans such as starting a family, developing a career, and preparing for retirement. However, only one-fourth of the patients would have lived their life differently if they would have expected to live shorter (see Supplementary material online, *Table S4*). In addition, it should be noticed that CHD patients are a specific group, not easily comparable with other groups of patients with reduced life expectancy. Since the enormous success in both medical and clinical care in this field, patients with CHD are now reaching ages above the expectations at their birth. This makes it difficult for both patients and healthcare providers to predict future complications and life expectancy with great certainty. Interestingly, it has to be taken into account that studies showed that patients who are more optimistic reported better quality of life and live a

longer life.[23,24] Therefore, the optimism of this group of patients might have positive impact on their lives.

It is also important to notice that females with mild CHD were significantly more pessimistic than their partners in terms of life expectancy. This trend was seen also in male patients, however it was not significant. We can speculate that partners were not aware about the reduced life expectancy of their partners and therefore they never questioned it. Another explanation could be related to the so-called 'protective buffer'. This behaviour may represent the denial or the attempt to minimize the illness or avoidance of communication related to the disease.[25,26]

Importance of good knowledge of life expectancy is fundamental for life planning, therapy adherence as well as discussing timely EOL, and advance care planning. Since patients with (very) mild CHD have near normal life expectations, they do not think that a member of the medical team should bring up questions regarding EOL. In fact, patients who consider their life expectancy to be reduced are also the ones who think that the cardiologist should discuss EOL with the patients. Despite their wish, only a small percentage reported to have had this discussion. Higher percentages of discussions about EOL were reported by their partners and by patients with moderate/severe CHD. Discrepancy between patients' and partners' reports might be explained as misrecognition. In fact, a previous study comparing patients and healthcare providers regarding EOL communication showed that 50% of the healthcare professionals reported to have discussed EOL with their patients, whereas only 1% of the patients recalled the discussion.[8] Understandably, EOL might be a difficult discussion to bring to the table in the early stage of illness; however, evidence is accumulating showing positive effects. Currently, it still seems to be a taboo. A delayed discussion on EOL is reported to have impact on the feeling of guilt in the decision maker (mostly partners or other family members) and reduces the quality of life in patients.[27–29] Furthermore, a late EOL discussion is related to more prolonged and/or aggressive treatment strategy and perhaps even unwanted treatments.[30] End-of-life discussion represents a difficult topic for the clinicians as well. In fact, the difficulty to recognize the good moment for discussion, the fear of causing stress in the patients, and the feeling of the health professionals of being unprepared are important barriers faced by clinicians discussing this topic.[31] To facilitate this communication, Kovacs *et al*. published recommendations for healthcare providers for a better discussion of EOL with CHD patients. It is important to invite the patients to discuss EOL taking into consideration their wishes and cultural and religious background.[8,32] Planning a specific visit and noting in the patient's file that this discussion has taken place are also important.[32] Interestingly, in our study, we found that half of our cohort did not find it necessary to discuss EOL with their cardiologist. This was in contrast with the previous study of Tobler *et al*., which showed that 78% of patients with a CHD would like to discuss EOL with a health professional. This discrepancy might be explained by a very optimistic view of our cohort on their life expectancy.[32] However, this might be an indication that this group of adults may not consider themselves as CHD patients but as ordinary adults with a CHD diagnosis. This might be especially true for patients with mild diagnosis who see a medical specialist (cardiologist) sporadically. Therefore, EOL might not be considered as a topic to be discussed with the cardiologist and even less with the treating team. This is particularly true if we consider that 49.3% of patients thought that a cardiologist should discuss EOL, whereas only 21.4% of them considered that a member of the treating team should discuss it. In fact, we can speculate that patients may see the treating team as an entity in the hospital setting and the cardiologist might be considered in an ambulatorial setting. Whereas the majority of patients see a cardiologist on a regular basis, hospitals are not part of their routine anymore; therefore, the treating team should not discuss EOL as it is not involved in their lives.

Strengths and limitations

This study included a relatively large number of adults with CHD in their middle adulthood. To our knowledge, this is the first study focusing on life expectancy of middle-aged adults. In addition, not only patients but also their partners and cardiologists were included.

The findings refer only to five diagnostic groups (ASD, VSD, PS, ToF, and TGA) who underwent surgery a long time ago; therefore, the results cannot be extrapolated to other CHD diagnosis and other age groups. In addition, this study is focusing on the patients who survived, so per definition they have better outcomes and therefore, a more positive view on their life expectancy. Finally, the cardiologists evaluated the life expectancy per patient based on their current clinical cardiac status that might be influenced by subjective judgment. However, to reduce this bias, two cardiologists assessed the life expectancy separately and disagreement was discussed with a third cardiologist.

Conclusions

Patients with CHD and their partners rated their life expectancy to be normal, while, overall, cardiologists had a more pessimistic view. In addition, the majority of patients reported that they have never discussed EOL with their cardiologist, although half of them would appreciate it. Discussing EOL expectations should be encouraged to make the patients aware of their condition and enable attentive life choices. Furthermore, conversations on EOL should be encouraged and integrated in the routine follow-up of CHD adults on an individual basis and after consent as not all patients are open to these discussions. Since the difficult and emotional nature of the topic, physicians and healthcare providers should be trained to discuss the topic in a sensitive and effective manner.

Lead author biography

Chiara Pelosi, MD, is a PhD student at Erasmus Medical Centre, Rotterdam, the Netherlands. She is currently working on her doctoral thesis investigating long-term psychosocial and cardiovascular outcomes of adults with congenital heart disease who underwent open-heart surgery during childhood.

Data availability

The anonymous data that support the findings of this study are available from the author, upon reasonable request.

Supplementary material

Supplementary material is available at *European Heart Journal Open* online.

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