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Sexual functioning is impaired in adults with congenital heart disease ☆

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ABSTRACT

Background: To investigate the overall sexual functioning and disease specific sexual problems in congenital heart disease (ConHD) patients, for both genders and different cardiac diagnostic groups, and compare these with Dutch normative data. Also disease specific sexual problems were investigated.
Methods: From a longitudinal cohort of patients, operated for ConHD between 1968 and 1980, 254 patients (median age: 40, 53.4% male) were included in this study: atrial septal defect (n = 72), ventricular septal defect (n = 71), pulmonary stenosis (n = 30), tetralogy of Fallot (n = 53) and transposition of the great arteries (n = 28). Patients completed internationally validated, generic questionnaires and also disease specific instruments on sexual functioning.
Results: Patients showed a delay in starting sexual activities compared with peers. Females with ConHD scored significantly worse compared with normative data on all scales of sexual functioning, indicating a broad range of sexual problems and 15% showed clinical levels of sexual dysfunction. Of the males, 14% suffered from erectile dysfunction. Males with ConHD scored worse on erectile function, orgasmic function and satisfaction regarding their sexual life compared with normative data. No differences were found between the different cardiac diagnoses.
The majority of patients reported disease specific worries and fears about the use of contraceptives, heredity, pregnancy and delivery. Patients indicated to have been suboptimally informed about sexuality in early adolescence.
Conclusions: This study shows that sexual functioning is impaired in adults with ConHD. Providing information to patients about sexuality, pregnancy, delivery and heredity should be improved, and given at young age.

1. Introduction

The number of adults with congenital heart disease (ConHD) is steadily increasing due to the successes of pediatric cardiology and open-heart surgery. This nascent demographic phenomenon is creating major issues concerning the optimal medical and psychological management of these patients [1–3]. Adults with ConHD have very specific needs, both on medical and psychosocial topics. Previous studies concerning psychosocial well-being and quality of life in adult ConHD patients have largely neglected sexual functioning and only very few studies have reported on this topic [4].

As part of a longitudinal study following a large cohort of consecutive patients operated for ConHD at young age, we investigated sexual functioning 30–43 years after cardiac surgery. Sexual functioning was assessed with both internationally validated instruments and disease-specific questionnaires, for males and females separately.

The aims of this study are:

1. To investigate the overall sexual functioning, for both sexes and different cardiac diagnostic groups, and compare this with the general population.
2. To investigate disease specific problems in sexual functioning.

2. Methods

2.1. Inclusion criteria

The original cohort exists of all consecutive patients who underwent their first open heart surgery for Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD), Pulmonary Stenosis (PS), Tetralogy of Fallot (ToF) or Transposition of the Great Arteries (TGA) between 1968 and 1980 in the Erasmus Medical Center, and were younger than 15 years at the time of surgery. This cohort has already been investigated in1990/1991 and in

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2000/2001. The baseline characteristics, medical and psychosocial results of these investigations have been reported in detail previously [5–10].

The target population of this third follow-up (2010–2011) consisted of the 412 patients who participated in the previous 2 follow-ups. Of these patients, ten had died (causes: 6 cardiac-related, 3 unknown, 1 accident), 1 underwent heart transplantation and 28 patients were untraceable. Of the remaining 373 eligible patients, 102 refused to participate in this study due to practical reasons (work, distance to hospital), leading to a response rate of 73%.

2.2. Patient sample

Of the 271 patients who participated and completed psychological instruments, 17 patients refused to complete the questionnaires on sexual functioning. Patients were classified into 2 groups of disease severity according to the classification adopted at the American Heart Association Task Force on Adults with CHD [11]. Patients with corrected ASD, VSD and PS were classified as simple ConHD (unless they had complications such as severe ventricular dysfunction), while patients with ToF or TGA (all operated with a Mustard repair) were classified as moderate to complex ConHD.

2.3. Assessment procedure

The research protocol was approved by the institutional ethical committee and complies with the 1975 Declaration of Helsinki. All patients were approached uniformly and invited to visit the hospital for extensive cardiac and psychological examination. All participating patients signed informed consent before participating. During their visit, a cardiologist performed the cardiac and medical examination. The semi-structured interview and psychological questionnaires were completed during the hospital visit. Due to practical reasons (work, children), 20 patients completed the questionnaires at home. The questionnaires were administered verbally for patients who had difficulty reading or understanding the questionnaires.

2.4. Instruments and normative groups

The Female Sexual Function Index (FSFI) is a multidimensional and standardized self-report questionnaire used to assess 6 domains of sexual functioning in females [12]. The FSFI has shown good discriminative validity between females with and without sexual problems [12–14].

The Female Sexual Distress Scale-Revised (FSDS-R) is a 12 item self-report questionnaire used to assess sexual related personal distress indicating sexual dysfunction. The FSDSR measures the psychological distress encountered during sexual intercourse [12,15].

If a patient scores within the clinical range on both the FSFI and the FSDSR, this is classified as a DSM-IV sexual disorder. Normative data for both the FSFI and FSDSR were derived from Kuile et al. [12]. Normative data on sexual dysfunctions in the general Dutch population were derived from the Rutgers Foundation [16].

The International Index of Erectile Function (IIEF) is a multidimensional self-report questionnaire that measures erectile function and sexual functioning in males [17]. Normative data for the IIEF questionnaire was derived from Rosen et al. [17]. Normative data on erectile dysfunction in the general Dutch population was derived from the Rutgers Foundation [17].

The ConHD Specific Problems related to Sexual Functioning (CSSP) is a structured self-report specifically designed for this study by two congenital cardiologists and a psychologist specialized in ConHD to assess the impact of ConHD on sexual functioning (unpublished questionnaire Utens et al. 2010). Normative data on general sexual functioning of the Dutch population was derived from the Rutgers Foundation [16,18].

2.5. Statistical analyses

Categorical variables are represented by frequencies and percentages. Where appropriate, a chi-square test or Fisher's exact test was used when comparing frequencies. Data on generic questionnaires (FSFI, FSDSR, IIEF) were analyzed according to international manuals using means and standard deviations [12,17]. Data on disease-specific questionnaires are represented with medians and interquantile ranges because of the skewed nature of the data. Comparison of continuous variables between simple and moderate/complex ConHD was made with Student's T-tests, comparison between ASD, VSD, PS, ToF and TGA groups was made by one-way ANOVA tests. In case of a skewed distribution, Mann-Whitney-U tests resp. Wilcoxon signed rank tests were used. Univariate binary logistic regression (forced entry model) was used to test for effects of medication on erectile function and for the effects of educational level, occupational level and income on sexual dysfunctions in both men and women. Two-tailed probability values of <0.05 were considered statistically significant. The statistical packages IBM SPSS Statistics for Mac version 19.0 (Release 19.0.0) and R (64 bit) for Mac, version 2.14.2 were used to perform the calculations.

3. Results

3.1. Baseline characteristics (Table 1)

The baseline characteristics of the 254 included patients (53.5% male, median age 40 years) are summarized in Table 1. A total of 97% was heterosexual, 2% was homosexual and 1% was bisexual. The majority of patients were married, and 84.5% reported to be sexually active. Patients with moderate/complex ConHD used cardiac medication significantly more often than the patients with simple ConHD ($p = 0.004$) and on average, had a worse systemic ventricular function ($p < 0.001$).

3.2. Gender specific sexual functioning compared with normative data

3.2.1. Females (FSFI and FSDSR) (Fig. 1)

On all sexual functioning scales (FSFI questionnaire), females with ConHD scored significantly worse compared with normative data, indicating a broad range of sexual problems; with younger females showing worse outcome compared with older females (groups based on median age of 39 years) ($p < 0.0001$). However, on the FSDS-R scale assessing sexual distress indicating pathological sexual dysfunction, females obtained similar results compared with normative data. A pathological sexual disorder (defined on both clinical levels on the FSFI and FSDS-R) present in 14.6% of our female patients compared with 17.7% in the reference group ($p = 0.5$) [18].

In-between ConHD diagnostic groups: No difference was found on either of the questionnaires between diagnostic groups.

Effect of systemic ventricular function: Univariate logistic regression showed no significant effects of systemic ventricular function on the prevalence of sexual disorders in females (FSDSR, FSFI and clinical score).

Effects of socio-economic status: Univariate logistic regression showed no significant effects of occupational level, educational level and income on the prevalence of sexual disorders in females (FSDSR, FSFI and clinical score).

3.2.2. Males (IIEF) (Fig. 2)

Males with ConHD scored worse on erectile function, orgasmic function, intercourse satisfaction and overall satisfaction compared with normative data. This result indicates a broad range of sexual problems. A total of 13.7% of males scored within the clinical range of having erectile dysfunction. This is more than twice as high as in normative data stratified by age (5.9%, $p = 0.002$). Patients with erectile dysfunction were not significantly older than patients without ($p = 0.8$). The use of beta-blockers did not have an effect on erectile function in this population ($p = 0.9$).

Diagnostic groups: No difference was found between the different ConHD diagnoses.

Effect of systemic ventricular function: Univariate logistic regression showed no significant effects of systemic ventricular function on the prevalence of sexual disorders in males (IIEF and clinical score).

Effects of socio-economic status: Univariate logistic regression showed no significant effects of occupational level, educational level and income on the prevalence of sexual disorders in males (erectile disorders on the IIEF).

3.3. Disease specific problems related to sexual functioning (Tables 2 and Online Supplemental Tables 3 and 4)

Gender specific sexual aspects are shown in Table 2. Females had their first menstrual period at a median age of 13 years, which is comparable to the general Dutch population. Cardiac complaints worsened in 11% of females during menses. Menstrual complaints mainly consisted of pain (41.7%) and excessive blood loss (35.1%).A total of 19.8% required medical attention for menstrual related problems.

3.4. Problems before, during and after sexual activity (Online Supplemental Table 3)

Patients in our study were on average 18 years old when they first had sexual intercourse. On average, they had 1 sexual partner during

Table 1
Baseline characteristics.

Variable		ConHD classification			ConHD diagnosis					
	Total	Simple	Moderate/complex	p	ASD	VSD	PS	ToF	TGA	p
	n = 254	n = 173	n = 81		n = 72	n = 71	n = 30	n = 53	n = 28	
Age	40 [36–45]	40 [37–45]	38 [34–41]	<0.0001	43 [39–47]	39 [35–43]	39 [37–45]	40 [35–45]	36 [33–38]	<0.0001
Gender										
Male	53.5	50.3	60.5	0.129	37.5	60.6	56.7	56.6	67.9	0.022
Female	46.5	49.7	39.5		62.5	39.4	43.3	43.4	32.1	
Marital status										
Unmarried	16.7	15.6	19.0	0.504	19.4	14.1	10.0	15.7	25.0	0.535
Cohabitants	21.0	20.2	22.8	0.644	19.4	16.9	30.0	25.5	17.9	0.546
Married	62.3	64.2	58.2	0.367	61.1	69.0	60.0	58.8	57.1	0.726
Medication										
No medication	76.5	82.1	64.3	0.004	76.6	84.8	88.5	72.7	50.0	0.004
Aspirin	4.9	3.8	7.1	0.287	4.7	1.5	7.7	9.1	3.8	0.327
Calcium antagonist	0.9	0.6	1.4	0.524	–	1.5	–	–	3.8	0.447
Beta blocker	7.5	5.8	11.4	0.136	7.8	6.1	–	6.8	19.2	0.150
Nitrate	–	–	–	–	–	–	–	–	–	–
Anti-arrhythmics	2.2	1.9	2.9	0.646	3.1	–	3.8	2.3	3.8	0.426
Digitalis	0.9	–	2.9	0.095	–	–	–	–	7.7	0.026
Diuretics	3.1	2.6	4.3	0.680	1.6	4.5	–	–	11.5	0.080
ACE inhibitor	8.0	3.8	17.1	0.001	1.6	7.6	–	9.1	30.8	<0.0001
Cholesterol lowering	3.1	2.6	4.3	0.680	4.7	–	3.8	6.8	–	0.160
Oral anticoagulation	4.9	3.2	8.6	0.083	4.7	3.0	–	2.3	19.2	0.028
Systemic ventricular function										
Good	62.9	86.4	29.8	<0.0001	92.4	80.0	75.0	46.3	–	<0.0001
Mildly impaired	24.8	11.9	42.9	<0.0001	7.6	15.0	25.0	48.1	33.3	<0.0001
Moderately impaired	7.9	–	19.0	<0.0001	–	–	–	–	53.3	<0.0001
Severely impaired	4.5	1.7	8.3	0.035	–	5.0	–	5.6	13.3	0.052

Data are presented as n %, unless indicated otherwise. Continuous data are presented as median [interquartile range (IQR)].
ASD = Atrial Septal Defect.
PS = Pulmonary Stenosis.
TGA = Transposition of the Great Arteries.
ToF = Tetralogy of Fallot.
VSD = Ventricular Septal Defect.

the past 3 months. Roughly a quarter of patients (24.7%) had sexual intercourse between zero and one time per month, 23.3% had sexual intercourse between 2 and 3 times per month, almost a third (28.4%) had sexual intercourse four to nine times per month and the rest (11.4%) had sexual intercourse more than nine times per month.

The majority (76.5%) of patients felt satisfied with their overall physical appearance. Obesity was the main reason why some of the patients felt dissatisfied. When rating the appearance of their scar, patients scored a 6 on a scale from 1 to 10 (1 = ugly, 10 = beautiful). The surgical scar made 11.4% of patients feel ashamed and less attractive during sexual activity. This was reported significantly more often by females (17.5% versus 6.2% in males, p = 0.004).

Of all patients, 11.7% reported having problems with sexual activities because of their ConHD. This was observed more frequently in females compared with males (16.2% versus 7.7%, p = 0.037), and more frequently in moderate/complex ConHD patients compared with

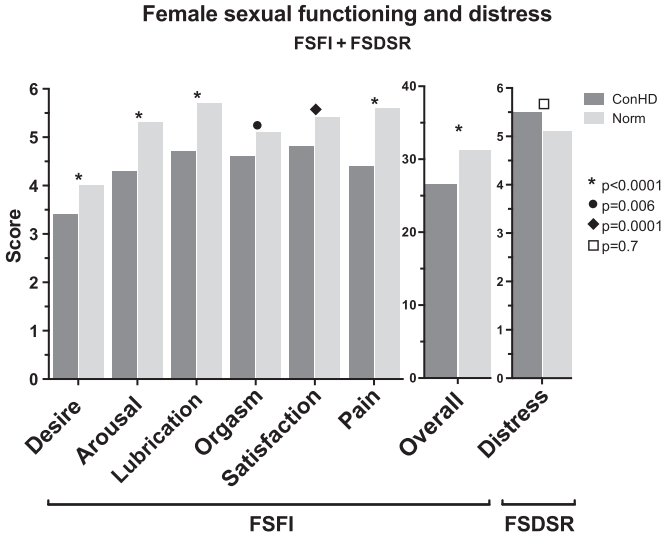

Fig. 1. ConHD = all congenital heart diseases together; Norm = normative data. A high score indicates favorable sexual functioning. On the distress scale, a high score indicates high distress. Data are presented using means.

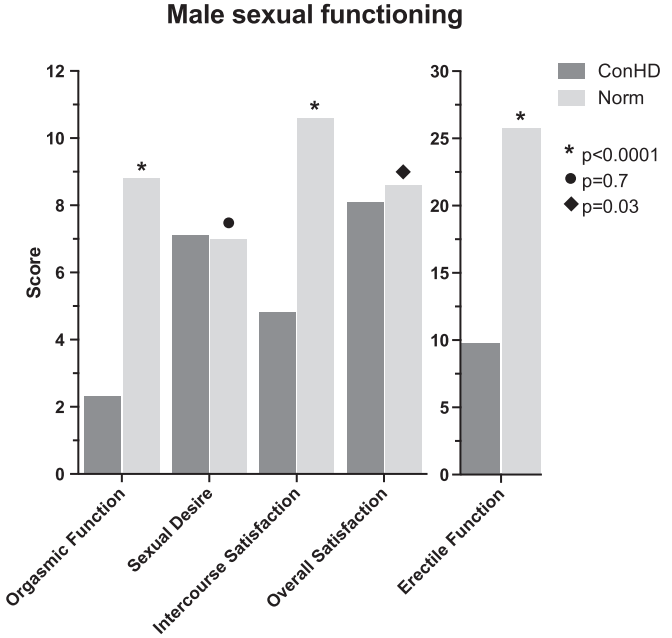

Fig. 2. ConHD = all congenital heart diseases together; Norm = normative data. A high score indicates favorable sexual functioning. Data are presented using means.

Table 2
Female disease specific sexually related aspects, fears & worries.

	Total N = 118		ConHD classification				
			Simple N = 86		Moderate/complex N = 32		p
	N	%	N	%	N	%	
At what age did you get your first menstrual period?							
Age (median [IQR])	13 [12–14]		13 [12–14]		13 [12–14]		0.5
Do you get more complaints from your ConHD or do the complaints change before or after your menstrual period?							
Yes	13	11.0	9	10.5	4	12.5	0.7
Have you ever experienced one of the following complaints?							
>5 weeks between menses	11	9.5	10	11.8	1	3.2	0.3
<5 weeks between menses	7	6.1	7	8.4	–	–	0.2
Excessive blood loss	40	35.1	27	32.5	13	41.9	0.3
Painful menses	48	41.7	36	42.9	12	38.7	0.7
6 months no menses	14	12.4	12	14.5	2	6.7	0.3
Spotting	5	11.6	10	12.2	4	12.9	1.0
Did you ever need medical attention for menstrual complaints?							
Yes	23	19.8	17	20.0	6	19.4	0.9
Are you (pre)menopausal?							
Yes	19	16.5	17	20.5	2	6.3	0.1
Age (median [IQR])	45 [42–47]		45 [42–47]		44 [42–44]		0.5
Are you receiving medical treatment for your (pre)menopause?							
Yes	3	2.6	2	2.4	1	3.1	1.0

Data are displayed as n %, unless indicated otherwise.
IQR = Interquantile Range.

simple ConHD patients (21.8% versus 7.1%, p = 0.001). In addition, 11.6% of all patients reported that their ConHD had a great influence on starting and maintaining a relationship. This was reported significantly more often in the moderate/complex ConHD group compared to the simple ConHD group (17.5% versus 8.9%, p = 0.048). Overall, patients did not have to prematurely cease with sexual activity due to heart complaints.

About 1 in every 5 patients reported not having enjoyed sexual activity (18.6%), having feelings of insecurity (15.7%) or felt unable to achieve sexual arousal during sexual activity (19.5%) within the last month. Not enjoying sexual activity was reported more often by females compared with males (25.5% versus 13.1%, p = 0.015).

About 1 in 4 patients worry about their sex life (23.8%). This was reported more frequently by females (30.8% versus 18.0%, p = 0.021).

A total of 7.8% of patients reported having been forced to perform sexual activity against their will. This finding was higher in females compared with males (11.3% versus 4.6%, p = 0.05).

When asked to rate the information provided by physicians about sexuality and ConHD, patients scored a 2.0 on average on a 1–10 Likert scale (1 = not being informed about the influence of sexuality on ConHD, 10 being optimally informed). Almost half of the patients (45%) rated the provided information on the possible side effects of sex on their heart condition as 1, and 68% of patients scored ≤5 (unsatisfactory).

3.5. Contraceptives, pregnancy and heredity (Online Supplemental Table 4)

Patients began using contraceptives at a median age of18 years. The most used contraceptive was the condom, followed by oral contraceptives and sterilization. 10.9% of males were sterilized, versus 5.9% of females (p = 0.166). A minority of patients (4.7%) became pregnant while using contraception.

The vast majority of patients (86.2%) never discussed the possible side effects of different methods of contraceptives with their physicians. There was a strong gender difference in information received from the physician; with females asking and receiving more information. Despite this, only 20% of women reported to have received information from their physicians regarding the use of contraceptives. Of this group, 56% had to actively ask for this information. Females reported having a greater fear of being infertile compared with males (26.2% versus 12.0%, p = 0.007). In addition, females reported experiencing more feelings of fear concerning the heredity of their ConHD (64.5% versus 41.0%, p < 0.0001). The fear of harmful effects of their heart disease on the outcome of their child was reported by 34.5% of patients. This was reported significantly more often in patients with moderate/complex ConHD (43.7% versus 30.3%, p = 0.05). Patients with moderate/complex ConHD also had significantly more fears of not having enough energy to raise children (16.9% versus 4.6%, p = 0.002) and fears of a lower life expectancy as a disability to fully raise children (19.7% versus 5.9%, p = 0.002). ConHD was a limiting factor in the decision of having children in 14.6% of patients, mostly in females compared with males (22.2% versus 7.6% p = 0.002) and in patients with moderate/complex ConHD compared with simple ConHD patients (22.2% versus 11.0%, p = 0.027).

A total of 21.3% of females feared that pregnancy would have a negative influence on their heart or on their overall clinical condition. This was found more often in females with moderate/complex ConHD (p < 0.0001). In addition, some of the females feared that delivery would have a negative effect on their heart (28.0%) and overall health (21.5%). Both of these findings were higher in females with moderate/complex ConHD group compared with females with simple ConHD (p < 0.0001).

4. Discussion

The present study shows a broad range of sexual problems in both men and women with ConHD. Concern over sexual health appeared to be stronger in women. Sexual functioning was significantly impaired compared with normative data. Concern over sexual health appeared to be stronger in women. Our female patients reported a wide range of problems, including a lower sexual desire, less arousal, and more pain during sexual intercourse. The proportion of male patients with erectile dysfunction was more than twice as high as in the general population. Male sexual functioning was clearly impaired as to satisfaction regarding intercourse and orgasmic function. Our female patients reported a wide range of problems, including a lower sexual desire, less arousal, and more pain during sexual intercourse.

No differences were found between cardiac diagnostic groups on generic questionnaires into sexual functioning. However, disease-specific instruments showed that patients with moderate/complex ConHD viewed their ConHD as a limiting factor in having children. Concerns

included not having enough energy in raising children, having a lower life expectancy, potentially harmful effects of the pregnancy on their child, their heart and overall health.

4.1. Disease specific problems related to sexual functioning

An important issue is whether the surgical scar influences personal feelings and hereby indirectly the sexual functioning of patients. Some patients indicated that the scar was part of their body and a symbol that their life had been saved. Earlier in life about half of patients from this same cohort reported to have been troubled by their scar [8]. Now, 10 years later, during the third follow-up of this cohort, a minor proportion of patients still felt ashamed because of their scar and felt less attractive. This was especially seen in females. However, compared with the two previous follow-ups, these numbers seem to have declined. A possible explanation is that with older age, patients seem to have accepted the scar and find it less important. This hypothesis is supported by Horner et al. [19] who described that patients reported to conceal or hide their scars mostly during adolescence. Most problems were observed around 20 years of age, the age that many people start a relationship. This may imply that the desire of young females to correct their scar by plastic surgery should be taken seriously, but does seem age-related and is often not a problem after 10 years.

Patients from our study lost their virginity at a significantly older age compared with the general Dutch population [18]. It has been reported that a lower educational level is associated with an early age of losing virginity [18]. Taking into account the lower educational level of the present cohort [7] the significantly older age when losing virginity is even more noticeable. In our opinion this finding can be explained by possible overprotection from parents [20]. In addition, the later age at which patients become independent and gain autonomy could play a role [21–23]. Finally, feelings of uncertainty or feelings about being less attractive might play a role here. Previously, during the second follow-up of this study at a younger age, our patients reported feeling limited due to their surgical scars. This could also have contributed to fear of rejection, which may cause patients to become sexually active at a later age.

4.2. Gender-specific sexual functioning

In our study we found that female patients had their first menstrual period at a median age of 13 years, which is in line with literature and normative data [18,24]. There was no difference between simple and moderate/complex ConHD patients.

Although females obtained significantly worse outcomes on the FSFI compared with normative data, the prevalence of sexual disorders in females (14.6% had clinical range on both the FSFI and FSDSR) did not differ from the general Dutch population. A possible explanation for this is that the FSDSR instrument is not sensitive enough to measure distress in females with ConHD. In contrast to the study of Reid et al. [25] we did not find that females with a moderate/complex ConHD had significantly more sexual partners indicating promiscuity. This could however be explained by the age difference: the patients in our study are older.

Of the male patients, 13.7% experienced a clinical level of erectile dysfunction, which is higher than reported previously using the same instruments in ConHD patients [26], and over twice as high as in the general Dutch population [16].

4.3. Contraceptives, pregnancy and heredity

This study also shows areas in outpatient care to which more attention should be paid. Only a minority of patients (13.8%) reported to have been fully informed about how sexuality, pregnancy, and contraceptives can influence their heart and overall general health [27–30]. 20% of females reported to have been informed regarding contraceptives by their physician. Studies have shown poor knowledge about contraceptives with many misconcepts [31,32]. Of course at the age of 40 this probably is not a big issue anymore, but there seems to be a clear need for counseling provided at a younger age, preferably before puberty. Information could be provided by pediatric cardiologists, but also by nurse practitioners or patient organizations.

4.4. Clinical implications

Since this cohort has reached the age of 30–56 years, and most patients have children, the need for sexually oriented information may have disappeared, and may be higher in younger patients. Patients reported to have had worries and fears in the past about the heredity of ConHD, harmful effects on the unborn child and infertility. Patients were also worried about the harmful effects of pregnancy and delivery on their own heart and overall health [19,25,28]. Caregivers should bear in mind that patients wish to receive information about contraceptives and sexual activities [32]. Therefore, we would recommend that patients receive disease specific information regarding sexual activities at young age, before planning to have children. Considering that most hospitals have combined transfer of patients from pediatric to adult clinics, this would provide a perfect opportunity to counsel patients about this problem, for instance by a nurse practitioner. Highlighted topics should at least include the safe use of contraceptives, sexual activities, pregnancy and delivery.

4.5. Strengths and study limitations

This study is the first to report on gender-specific sexual functioning in a systematically followed consecutive series of 30–56 year old patients. Both internationally validated generic questionnaires and disease specific instruments were used to measure sexual functioning for both genders. Where possible, outcomes were compared with normative data. Unfortunately, data on sexual functioning in males is scarce. There was no Dutch IIEF data of male reference groups for The Netherlands. Therefore, IIEF normative data from the USA was used. The patients included in this study all had the diagnosis of ASD, VSD, PS, ToF or TGA, and all were followed in an adult tertiary care center in The Netherlands. Therefore, the obtained results may not be applicable for all ConHD patients, nor in all countries worldwide. The FSDSR instrument may not have been specific enough to detect clinical levels of problems with sexuality in females.

4.6. Future recommendations

With increasing age, it can be expected that erectile disorders and menopausal complaints in ConHD patients may worsen. Systematic follow-up of sexual function is therefore recommended in this population since the present patients with ConHD already show high levels of sexual dysfunction. Attention for and information on sexual functioning should be organized for adolescents with ConHD. The extent in which these findings can be extrapolated to other heart conditions is unknown and should be investigated.

5. Conclusions

Sexual functioning in ConHD adults has largely been neglected in psychosocial research. Our study shows that sexual functioning is hampered substantially in adults with ConHD. Importantly, providing information to patients about sexuality, pregnancy, delivery and heredity should be improved and given at a younger age to assure the best holistic care.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at http://dx.doi.org/10.1016/j.ijcard.2013.06.029.

References

[1] van der Linde D, Konings EE, Slager MA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol 2011;58:2241–7.
[2] Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. First of two parts. N Engl J Med 2000;342:256–63.
[3] Warnes CA. The adult with congenital heart disease: born to be bad? J Am Coll Cardiol 2005;46:1–8.
[4] Moons P, Van Deyk K, Marquet K, De Bleser L, Budts W, De Geest S. Sexual functioning and congenital heart disease: something to worry about? Int J Cardiol 2007;121:30–5.
[5] Utens EM, Verhulst FC, Erdman RA, et al. Psychosocial functioning of young adults after surgical correction for congenital heart disease in childhood: a follow-up study. J Psychosom Res 1994;38:745–58.
[6] Utens EM, Verhulst FC, Meijboom FJ, et al. Behavioural and emotional problems in children and adolescents with congenital heart disease. Psychol Med 1993;23:415–24.
[7] van Rijen EH, Utens EM, Roos-Hesselink JW, et al. Psychosocial functioning of the adult with congenital heart disease: a 20–33 years follow-up. Eur Heart J 2003;24:673–83.
[8] van Rijen EH, Utens EM, Roos-Hesselink JW, et al. Medical predictors for psychopathology in adults with operated congenital heart disease. Eur Heart J 2004;25:1605–13.
[9] Roos-Hesselink J, Perlroth MG, McGhie J, Spitaels S. Atrial arrhythmias in adults after repair of tetralogy of Fallot. Correlations with clinical, exercise, and echocardiographic findings. Circulation 1995;91:2214–9.
[10] Roos-Hesselink JW, Meijboom FJ, Spitaels SE, et al. Decline in ventricular function and clinical condition after Mustard repair for transposition of the great arteries (a prospective study of 22–29 years). Eur Heart J 2004;25:1264–70.
[11] Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in collaboration with the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol 2008;52:e143–263.
[12] ter Kuile M, Brauer M, Laan E. De Female Sexual Function Index (FSFI) en de Female Sexual Distress Scale (FSDS): Psychometrische eigenschappen in een Nederlandse populatie. Tijdschr Seksuologie 2009;33–3:207–22.
[13] Meston CM. Validation of the Female Sexual Function Index (FSFI) in women with female orgasmic disorder and in women with hypoactive sexual desire disorder. J Sex Marital Ther 2003;29:39–46.
[14] Wiegel M, Meston C, Rosen R. The Female Sexual Function Index (FSFI): cross-validation and development of clinical cutoff scores. J Sex Marital Ther 2005;31:1–20.
[15] Derogatis L, Clayton A, Lewis-D'Agostino D, Wunderlich G, Fu Y. Validation of the female sexual distress scale-revised for assessing distress in women with hypoactive sexual desire disorder. J Sex Med 2008;5:357–64.
[16] de Graaf H, Kruijer H, van Acker J, Meijer S. Seks onder je 25ste. Rutgers WPF/Soa aids Nederland. 1st ed. Delft: Eburon; 2012.
[17] Rosen RC, Riley A, Wagner G, Osterloh IH, Kirkpatrick J, Mishra A. The International Index Of Erectile Function (IIEF): a multidimensional scale for assessment of erectile dysfunction. Urology 1997;49:822–30.
[18] Bakker F, Vanwesenbeeck I. Sexuele gezondheid in Nederland 2006. Rutgers Nisso Groep. 1st ed. Delft: Eburon; 2006.
[19] Horner T, Liberthson R, Jellinek MS. Psychosocial profile of adults with complex congenital heart disease. Mayo Clin Proc 2000;75:31–6.
[20] Ong L, Nolan RP, Irvine J, Kovacs AH. Parental overprotection and heart-focused anxiety in adults with congenital heart disease. Int J Behav Med 2011;18:260–7.
[21] Kovacs AH, Sears SF, Saidi AS. Biopsychosocial experiences of adults with congenital heart disease: review of the literature. Am Heart J 2005;150:193–201.
[22] Simko LC, McGinnis KA, Schembri J. Educational needs of adults with congenital heart disease. J Cardiovasc Nurs 2006;21:85–94.
[23] Kokkonen J, Paavilainen T. Social adaptation of young adults with congenital heart disease. Int J Cardiol 1992;36:23–9.
[24] Vigl M, Kaemmerer M, Niggemeyer E, et al. Sexuality and reproductive health in women with congenital heart disease. Am J Cardiol 2010;105:538–41.
[25] Reid GJ, Siu SC, McCrindle BW, Irvine MJ, Webb GD. Sexual behavior and reproductive concerns among adolescents and young adults with congenital heart disease. Int J Cardiol 2008;125:332–8.
[26] Vigl M, Hager A, Bauer U, et al. Sexuality and subjective wellbeing in male patients with congenital heart disease. Heart 2009;95:1179–83.
[27] Hargrove A, Penny DJ, Sawyer SM. Sexual and reproductive health in young people with congenital heart disease: a systematic review of the literature. Pediatr Cardiol 2005;26:805–11.
[28] Uzark K, VonBargen-Mazza P, Messiter E. Health education needs of adolescents with congenital heart disease. J Pediatr Health Care 1989;3:137–43.
[29] Kamphuis M, Verloove-Vanhorick SP, Vogels T, Ottenkamp J, Vliegen HW. Disease-related difficulties and satisfaction with level of knowledge in adults with mild or complex congenital heart disease. Cardiol Young 2002;12:266–71.
[30] Verstappen A, Pearson D, Kovacs AH. Adult congenital heart disease: the patient's perspective. Cardiol Clin 2006;24:515–29.
[31] Rogers P, Mansour D, Mattinson A, O'Sullivan JJ. A collaborative clinic between contraception and sexual health services and an adult congenital heart disease clinic. J Fam Plann Reprod Health Care 2007;33:17–21.
[32] Kaemmerer M, Vigl M, Seifert-Klauss V, et al. Counseling reproductive health issues in women with congenital heart disease. Clin Res Cardiol 2012;101:901–7.