International Journal of Cardiology 181 (2015) 218–224

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Acceptance and psychological impact of implantable defibrillators amongst adults with congenital heart disease

Radwa Bedair [a], Sonya V. Babu-Narayan [a,b,c], Konstantinos Dimopoulos [a,b,c], Sadia Quyam [a], Anne-Marie Doyle [a], Lorna Swan [a,c], Michael A. Gatzoulis [a,b,c], Tom Wong [a,b,c,*]

[a] Adult Congenital Heart Disease Centre, Royal Brompton Hospital, United Kingdom
[b] National Heart and Lung Institute, Imperial College, London, United Kingdom
[c] NIHR Cardiovascular Biomedical Research Unit, Royal Brompton Hospital and Imperial College London, United Kingdom

ARTICLE INFO

Article history:
Received 22 June 2014
Received in revised form 14 November 2014
Accepted 1 December 2014
Available online 3 December 2014

Keywords:
Implantable cardioverter defibrillator
Adult congenital heart disease
Device acceptance
Quality of life
Anxiety
Depression

ABSTRACT

Background: The psychological impact of implantable cardioverter defibrillators (ICDs) in adults with congenital heart disease (ACHD) has not been established.
Objective: To compare device acceptance, quality of life, anxiety and depression between ACHD patients with ICDs (ICD-Congenital), with pacemakers (PPM-Congenital), with no devices (No Device-Congenital) and non-ACHD patients with ICDs (ICD-Non-Congenital).
Methods: A total of 147 ACHD and 46 non-ACHD patients (age 45.0 ± 14.7 years, 56.5% males) completed the Florida Patient Acceptance Survey (FPAS), the 36-item Short Form Health Survey (SF-36) and Hospital Anxiety & Depression Scale (HADS).
Results: ICD-Congenital patients (n = 59) showed lower device acceptance compared to PPM-Congenital patients (n = 41), p = 0.04, and reported worse quality of life (p = 0.001) and higher prevalence of depression (p = 0.009) when compared to No Device-Congenital (n = 47) patients. ICD-Congenital and ICD-Non-Congenital patients (n = 46) showed similar mental and physical health, device acceptance, anxiety and depression. Within ICD-Congenital, patients with poorest device acceptance (FPAS <67, "Non-Acceptors") showed significantly lower mental health scores (p = 0.008), and higher levels of anxiety (p = 0.02) and depression (p = 0.01) compared to "Acceptors" (FPAS ≥67). "Non-Acceptors" were younger at survey (p = 0.006), younger at ICD implantation (p = 0.01) and were less likely to have received appropriate shocks (p = 0.03).
Conclusion: Younger age and lack of appropriate ICD shocks are risk factors for poor ICD acceptance. Device acceptance is lower in adults with congenital heart disease who receive an ICD than those who receive pacemakers. Appropriate screening for anxiety and depression may be warranted for ACHD patients considered for ICD implantation or already living with ICDs.

1. Introduction

The adult congenital heart disease (ACHD) population is growing both in number and disease complexity [1]. A significant proportion of these patients are at a heightened risk of sudden cardiac death [2], and implantable cardioverter defibrillators (ICDs) are being increasingly considered for primary or secondary prevention of sudden cardiac death in them [3,4]. Studies in patients with acquired heart disease have shown that living with an ICD may have a negative impact on psychological wellbeing, which may be associated with a higher prevalence of anxiety and depression [5] and with poor device acceptance [6,7]. Furthermore, quality of life in ICD recipients can be adversely affected by multiple factors that are clinical, social or device specific [8–13]. The same predisposing factors for poor psychological wellbeing cannot be assumed in ACHD patients [14,15]. Patients living with congenital heart disease and an ICD may additionally become newly aware of their reduced life expectancy, may be younger at the time of device implantation, and have increased device-related morbidity and need for recurrent hospitalisation and interventions [16,17].

We, therefore, aimed to examine and compare device acceptance, quality of life, anxiety and depression in ACHD patients with an ICD, ACHD patients with a permanent pacemaker, ACHD patient with no device and non-congenital heart disease patients with an ICD.

2. Methods

2.1. Patient population and study design

From October 2011 to October 2012, congenital and non-congenital heart disease patients, 18 years or older, attending either the device follow-up clinic or the adult congenital

* Corresponding author at: Heart Rhythm Centre and NIHR Cardiovascular Biomedical Research Unit, Royal Brompton and Harefield NHS Foundation Trust, Sydney Street, London SW3 6NP, United Kingdom.
E-mail address: tom.wong@imperial.ac.uk (T. Wong).

http://dx.doi.org/10.1016/j.ijcard.2014.12.028
0167-5273/

heart disease clinics were invited to participate in a questionnaire study. Consecutive patients with congenital heart disease were included, whilst younger patients with non-congenital heart disease and ICDs were selected for recruitment from every pacing clinic. Patients who had a device-related procedure within three months, who were admitted to hospital within one month or were awaiting a device-related procedure, were excluded. Patients with learning difficulties or limited knowledge of English were not included if it was felt that they would not be able to complete the questionnaires. Eligible patients were given a study pack that included: a patient information sheet, consent form, study questionnaire and an addressed envelope for completion and postal return. Patients were contacted once by telephone if they did not already return the questionnaire.

The study was approved by the National Research Ethics Service and written, informed consent was obtained from all participants. Patients were divided into the following groups at screening to allow recruitment into each for comparison:

- ICD-Congenital: patients with congenital heart disease and an ICD. This included any patient with a defibrillator function in their device, including cardiac resynchronisation therapy (CRT-D).
- PPM-Congenital: patients with congenital heart disease and permanent pacemakers. This included patients with a permanent pacemaker for bradyarrhythmia, cardiac resynchronisation (CRT-P) or for control of atrial arrhythmia.
- No Device-Congenital: patients with congenital heart disease and no implantable device. No patients with simple defects such as uncomplicated atrial septal or small ventricular septal defect were included.
- ICD-Non-Congenital: patients with non-congenital heart disease and ICDs who received their ICD follow-up at our centre.

The cardiac diagnosis, comorbidities and details of previous procedures were obtained from patient records. Data relating to the device implant, device follow-up and device related complications were obtained from device follow-up records. Congenital heart disease was classified into mild, moderate and high complexity using the Bethesda classification [18].

2.2. Device acceptance: Florida Patient Acceptance Survey (FPAS)

Device acceptance was assessed using the FPAS questionnaire, which is disease-specific to patients with implanted devices [19]. It consists of 18 items rated on a five-point scale from 0 to 5. Total and subscale scores for Return to Life, Body Image Concerns, Positive Appraisal and Device Related Distress are calculated on a scale of 0–100. There is no validated cut-off for categorising device acceptance into poor and good, with previous studies using individual study cohorts to determine cut-offs based on the lower tertile of patients [20]. We used a cut-off score of 67, representing the lowest tertile of all FPAS scores in the three device groups, to categorise patients into "Non-Acceptors" and "Acceptors".

2.3. Anxiety and depression: Hospital Anxiety and Depression Scale (HADS)

Symptoms of anxiety and depression were assessed using the Hospital Anxiety and Depression Scale, which is validated for a number of chronic conditions, including cardiac diseases [21]. HADS is a 14-item scale consisting of a seven-item anxiety subscale and a seven-item depression subscale. Each item is scored on a four-point scale (0–3) giving maximum subscale scores of 21 for depression and anxiety. A cut-off score of ≥ 8 for both subscales indicates clinically relevant anxiety or depression [22,23].

2.4. Quality of life: Short Form 36 (SF-36)

The SF-36 questionnaire is a non-disease-specific questionnaire consisting of 36 questions [24,25] designed to measure eight dimensions of health: physical functioning, role limitation-physical, bodily pain, general health, vitality, social functioning, role limitation-emotional and mental health. The eight health dimension scores are each scaled from 0 to 100. The subscales are computed into two composite summary scores: Mental Component Summary Score (MCS) and Physical Component Summary Score (PCS).

2.5. Statistical analysis

Continuous variables are reported as mean, standard deviation or median, interquartile range as appropriate. Differences in continuous variables between groups were assessed using Wilcoxon rank sum test or Kruskall Wallis test, for non-normally distributed variables and the independent sample t-test for normally distributed variables. The chi-squared contingency table tests or Fisher's exact test were used for categorical variables, as appropriate. All analyses were done using SPSS version 19 and R version 2.15.2.

3. Results

Questionnaires were given to a total of 319 patients: 83 to ICD-Congenital, 69 to PPM-Congenital, 85 to No Device-Congenital, and 82 to ICD-Non-Congenital patients. In total 193 patients (61% response rate) completed the questionnaires. No significant difference in response rate was seen between groups (chi-square test $p = 0.14$): ICD-Congenital −71%; PPM-Congenital −59%; No Device-Congenital −55% and ICD-Non-Congenital −56%. Study participants were slightly older (45.0 ± 14.7 years) than non-participants (41.6 ± 13.7 years), $p = 0.04$, but there were no significant differences in gender (57% versus 63% male, $p = 0.27$), disease complexity (33% versus 33% complex disease) or systemic ventricular function (25% versus 25% severe dysfunction) between participants and non-participants.

3.1. Demographic and clinical characteristics of the study population

The demographic and clinical details of patients in the four groups are summarised in Table 1. ACHD participants represented a wide spectrum of congenital heart disease diagnoses, with high complexity disease being equally prevalent in the three congenital heart disease subgroups. Co-morbidities included stroke, brain abscess, multiple sclerosis, rheumatoid arthritis, Crohn's disease, bowel cancer, sarcoidosis and severe scoliosis. The diagnoses in the non-congenital heart disease group were cardiomyopathy ($n = 25$), channelopathy ($n = 13$), ischaemic heart disease ($n = 6$) and valvar heart disease ($n = 2$).

3.2. Device-related data

Device-related data are summarised in Table 2. Compared to ICD-Congenital patients, PPM-Congenital patients had devices implanted at a younger age. Whilst total, appropriate and multiple shock rates were similar in ICD-Congenital and ICD-Non-Congenital, a significantly higher rate of inappropriate shocks was seen in the ICD-Congenital group (31% versus 6.5%). Of the 18 ICD-Congenital patients who received inappropriate shocks, 10 additionally received appropriate shocks and 8 had only inappropriate shocks. At the time of survey, one patient in ICD-Congenital and one patient in ICD-Non-Congenital had ICD therapies switched off.

3.3. Device acceptance

Device acceptance was significantly lower in ICD-Congenital compared to PPM-Congenital (FPAS 73 versus 83). ICD-Congenital had worse Return to Living scores compared to PPM-Congenital as well as a trend towards higher Device-Related Distress. There were no differences in total or subscale FPAS scores between ICD-Congenital and ICD-Non-Congenital. Positive Appraisal of the device was high across all three device-groups (median >80) and no differences in Body-Image Concerns were observed. These findings are summarised in Table 3.

Within ICD-Congenital, "Non-Acceptors" (FPAS score <67, $n = 22$) were on average a decade younger at survey and younger when they received their ICD compared to "Acceptors" (FPAS ≥ 67, $n = 37$), but otherwise shared similar clinical characteristics (Table 4). "Acceptors" in the ICD-Non-Congenital group were of similar age at survey (48.9 SD 13.7 versus 51.3 SD 14.5 years, $p = 0.56$) and ICD implant (43.8 SD 15.8 versus 46.9 SD 16.3 years, $p = 0.55$) as "Non-Acceptors".

With regard to ICD shocks and device acceptance, in ICD-Congenital, appropriate ICD shocks were significantly lower in the "Non-Acceptors" (9.1%) compared to "Acceptors" (35.1%) (Fig. 1). Of the 22 "Non-Acceptors", 6 had received ICD shocks: 1 had received only appropriate shocks, 4 had received only inappropriate shocks, and 1 had received both appropriate and inappropriate shocks. In ICD-Non-Congenital group, "Non-Acceptors" (FPAS <67, $n = 20$) showed significantly higher rates of any shock (including multiple, appropriate and inappropriate ICD shocks) compared to "Acceptors" (FPAS ≥ 67, $n = 26$). Of the 20 "Non-Acceptors", 11 had received shocks: 8 had received only appropriate shocks, 2 had received only inappropriate shocks and 1 had received both appropriate and inappropriate shocks. In ICD-Congenital, the

Table 1
Demographic and clinical characteristics.

	Overall	ICD-Congenital	PPM-Congenital		No Device-Congenital		ICD-Non-Congenital	
				p[a]		p[a]		p[a]
n	193	59	41		47		46	
Age, y	45.0 SD 14.7	45.5 SD 13.5	42.5 SD 16.2	0.36	41.1 SD 14.1	0.12	50.4 SD 14.1	0.07
Males, n (%)	109 (57%)	35 (60%)	17 (42%)	0.08	23 (49%)	0.2	34 (74%)	0.08
Living with partner, n (%)	124 (64%)	33 (56%)	27 (66%)	0.3	29 (62%)	0.5	35 (76%)	**0.03**
Living alone, n (%)	23 (12%)	9 (15%)	1 (2%)	**0.04**	8 (17%)	0.8	5 (11%)	0.5
Living with parents, n (%)	29 (15%)	11 (12%)	6 (15%)	0.6	10 (21%)	0.7	2 (4%)	**0.03**
Working, n (%)	112 (59%)	31 (54%)	22 (54%)	0.9	37 (79%)	**0.01**	22 (45%)	0.5
NYHA I, n (%)	106 (55%)	29 (49%)	25 (61%)	0.24	31 (66%)	0.08	21 (46%)	0.72
NYHA II, n (%)	60 (31%)	18 (31%)	13 (32%)	0.90	12 (26%)	0.57	17 (37%)	0.49
NYHA III, n (%)	27 (14%)	12 (20%)	3 (7%)	0.07	4 (9%)	0.09	8 (17%)	0.70
Moderate or severe systemic ventricular dysfunction, n (%)	48 (25%)	19 (32%)	8 (20%)	0.16	2 (4%)	**0.000**	19 (41%)	0.34
Co-morbidity, n (%)	23 (12%)	6 (10.2%)	4 (10%)	0.946	10 (21%)	0.113	3 (6.5%)	0.51
Clinical atrial arrhythmia, n (%)	60 (31%)	26 (44%)	14 (34%)	0.32	12 (26%)	**0.048**	8 (17%)	**0.004**
Previous ablation, n (%)	39 (20%)	22 (37%)	8 (20%)	0.06	4 (9%)	**0.001**	5 (11%)	**0.002**
Previous cardiac surgery, n (%)	138 (71%)	55 (93%)	36 (88%)	0.352	40 (85%)	0.174	7 (15%)	<**0.001**
Operations, n	1.85 SD 1.2	2.2 SD 1.2	1.6 SD 1.0	**0.02**	1.6 SD 1.2	**0.02**	0	**–**
Age at last cardiac surgery, y	25.3 SD 17.0	28.7 SD 16.4	21.7 SD 17.5	0.06	23.8 SD 17.1	0.17	39.0 SD 14.5	**–**
Time since surgery, y [b]	17.3 SD 14.0	16.1 SD 12.4	18.6 SD 13.7	0.38	17.9 SD 16.4	0.55	10.7 SD 7.7	**–**
Mild complexity, n (%)	25 (17%)	6 (10%)	8 (20%)	0.185	11 (24%)	0.065	–	**–**
Moderate complexity, n (%)	73 (50%)	31 (53%)	19 (46%)	0.542	23 (49%)	0.7	–	**–**
High complexity, n (%)	49 (33%)	22 (37%)	14 (34%)	0.748	13 (28%)	0.29	–	**–**
Tetralogy of Fallot spectrum n (%)	57 (39%)	31 (53%)	10 (24%)	**0.005**	16 (34%)	**0.03**	–	**–**
Cavo-pulmonary connection, n (%)	8 (5%)	1 (2%)	4 (10%)	0.089	3 (6%)	0.228	–	**–**
Atrial switch, n (%)	9 (6%)	4 (7%)	4 (10%)	0.589	1 (2%)	0.260	–	**–**
Systemic right ventricle, n (%)	23 (16%)	13 (22%)	6 (15%)	0.354	4 (9%)	0.051	–	**–**
Native disease, n (%)	11 (7%)	4 (7%)	4 (10%)	0.589	3 (6%)	0.93	–	**–**
Cyanosis, n (%)	9 (6%)	3 (5%)	2 (5%)	0.483	4 (9%)	0.480	–	**–**

ICD; implantable cardioverter defibrillator. NYHA; New York Heart Association functional class. PPM; permanent pacemaker.
Significant differences are highlighted in bold print.
[a] p values are reported for comparisons to ICD-Congenital.
[b] Time since last cardiac surgery.

absence of appropriate shocks was associated with poor acceptance. The reverse was seen in ICD-Non-Congenital, where ICD shocks (both appropriate and inappropriate) were clearly associated with poor acceptance. "Non-Acceptors" in ICD-Congenital showed significantly higher levels of anxiety, depression and worse overall mental health.

3.4. Quality of life scores: SF 36

In the three groups with implanted devices (ICD-Congenital, PPM-Congenital and ICD-Non-Congenital), there were no statistical differences in the subscale and composite scores for physical and mental health as assessed by the SF-36 questionnaire (Table 3). Compared to

Table 2
Device related data and complications.

	ICD-Congenital	PPM-Congenital	p	ICD-Non-Congenital	p
Age at device implant, y	39.8 SD 14.2	31.7 SD 18.7	**0.02**	44.7 SD 15.9	0.1
Years with device, y	5.7 SD 6.1	10.9 SD 9.1	**0.001**	5.1 SD 4.8	0.7
Device-related procedures, n	2.02 SD 1.61	2.18 SD 1.62	0.629	1.74 SD 0.82	0.308
Infection/erosion, n (%)	1 (1.7%)	2 (4.9%)	0.56	1 (2.1%)	0.65
Lead fracture, n (%)	2 (3.4%)	0	–	3 (6.5%)	0.370
Remote monitoring, n (%)	27 (45.8%)	–	–	15 (32.6%)	0.172
Advisory lead, n (%)	10 (17%)	–	–	5 (11%)	0.522
Lead displacement, n (%)[a]	3 (5%)	1 (2.4%)	0.542	7 (15%)	**0.048**
Box and lead position					
Pectoral/endocardial, n (%)	57 (97%)	33 (81%)	–	45 (98%)	
Femoral/endocardial, n (%)	1 (1.7%)	3 (7.3%)	–	–	
Abdominal/epicardial, n (%)	–	5 (12.2%)	–	–	
Subcutaneous lead ICD, n (%)	1 (1.7%)	–		1 (2%)	
ICD for primary prevention, n (%)	33 (56%)	–	–	22 (48%)	0.4
ICD upgrade from PPM, n (%)	10 (17%)	–	–	4 (7%)	0.2
Age at ICD implant, y	41.5 SD 13.7	–	–	45.5 SD 16.0	0.17
Number of years with ICD	4.0 SD 3.0	–	–	4.2 SD 4.3	0.7
CRT-D, n (%)	16 (27%)	–	–	12 (26%)	0.9
ICD therapy off, n (%)	1 (1.7%)	–	–	1 (2%)	–
Shock, n (%)	23 (39%)	–	–	15 (33%)	0.3
Multiple shocks, n (%)	17 (29%)	–	–	8 (17%)	0.3
Appropriate shock, n (%)	15 (25%)	–	–	13 (28%)	0.7
Inappropriate shock, n (%)	18 (31%)			3 (6.7%)	**0.003**
Only inappropriate shock, n (%)	8 (14%)	–	–	2 (4%)	0.180

CRT-D; cardiac resynchronisation therapy-defibrillator. ICD; implantable cardioverter defibrillator. PPM; permanent pacemaker.
Significant differences compared to ICD-Congenital are highlighted in bold.
[a] Experienced consultant/staff operators only are permitted to perform device implantations in ACHD patients which may have resulted in lower rates of lead displacement.

Table 3
Device acceptance (FPAS), quality of life (SF36), anxiety (HADS-A) and depression (HADS-D) median scores in the four study groups.

	ICD-Congenital	PPM-Congenital	p[a]	No Device-Congenital	p[a]	ICD-Non-Congenital	p[a]
FPAS							
Return to Living (RL)	69 (48, 88)	88 (58, 89)	**0.02**	–	–	63 (31, 80)	0.2
Device-Related Distress (DRD)	20 (9, 45)	10 (0, 30)	0.06	–	–	30 (6, 54)	0.6
Positive Appraisal (PA)	84 (69, 100)	88 (75, 100)	0.2	–	–	88 (81, 100)	0.1
Body Image Concerns (BIC)	25 (0, 50)	25 (0, 50)	0.6	–	–	0 (0, 25)	0.2
Total FPAS score	73 (57, 90)	83 (67, 83)	**0.04**			69 (59, 88)	0.9
SF-36							
Physical functioning (PF)	48 (32, 53)	51 (37, 55)	0.101	51 (44, 55)	**0.017**	42 (29, 55)	0.876
Role limitation-physical (RP)	47 (30, 57)	54 (36, 55)	0.141	54 (45, 57)	**0.024**	37 (28, 56)	0.173
Bodily pain (BP)	50 (37, 62)	51 (42, 62)	0.254	55 (46, 62)	**0.007**	49 (37, 62)	0.790
General health (GH)	36 (28, 51)	43 (33, 53)	0.073	48 (36, 55)	**0.002**	36 (26, 53)	0.846
Vitality (VT)	46 (33, 55)	52 (46, 58)	0.056	52 (46, 58)	**0.012**	40 (33, 52)	0.416
Social functioning (SF)	49 (41, 57)	51 (41, 57)	0.886	57 (46, 57)	**0.034**	46 (35, 57)	0.243
Role limitation-emotional (RE)	56 (33, 56)	56 (33, 56)	0.798	56 (44, 56)	0.225	44 (33, 56)	0.175
Mental health (MH)	47 (36, 56)	50 (40, 59)	0.273	52 (44, 56)	0.123	44 (36, 56)	0.776
Physical Component Summary (PCS)	45 (32, 53)	50 (37, 56)	0.056	52 (43, 57)	**0.001**	43 (30, 53)	0.936
Mental Component Summary (MCS)	50 (37, 58)	51 (41, 58)	0.493	54 (43, 57)	0.408	44 (36, 55)	0.284
HADS							
HADS-anxiety	6 (3, 10)	6.5 (4, 10)	0.878	6 (4, 8)	0.368	6 (3, 11)	0.838
HADS-depression	4 (1, 8)	2 (1, 6)	0.163	2 (1, 3)	**0.005**	4 (1, 8)	0.514
Anxiety, n (%)	25 (42%)	17 (42%)	0.990	18 (42%)	0.671	20 (44%)	0.910
Depression, n (%)	15 (25%)	6 (15%)	0.213	3 (6%)	**0.009**	15 (33%)	0.419

FPAS; Florida Patient Acceptance Survey. HADS; Hospital Anxiety and Depression Scale. ICD; implantable cardioverter defibrillator. PPM; permanent pacemaker. SF-36; Short Form Questionnaire. Values reported as median (interquartile range).
[a] p reported for differences between ICD-Congenital and other subgroups. Significant differences are highlighted in bold.

the No Device-Congenital group, ICD-Congenital patients showed significantly worse physical functioning, role limitation-physical, bodily pain, general health and Physical Component Summary Score (PCS). Additionally, two subscales of mental health were lower in ICD-Congenital, with significantly lower vitality and social functioning compared to No Device-Congenital. The overall mental health reflected in the Mental Component Summary Score (MCS), however was not statistically different.

3.5. Anxiety and depression: HADS

Participants in all four groups had high anxiety scores (median HADS-A 6), with a 40% overall prevalence of clinically relevant anxiety (i.e. HADS-A $\geq$8). Compared to No Device-Congenital, ICD-Congenital patients had significantly higher depression scores and higher prevalence of depression (25% versus 6.5%). There were no significant differences in the rate of depression observed in the three device-groups (Table 3).

Table 4
Characteristics of the "Non-Acceptors" versus the "Acceptors" of ICDs in ICD-Congenital group.

	Non-Acceptors (FPAS <67)	Acceptors (FPAS ≥67)	p
n	22	37	
Age, y	39.3 SD 13.0	49.2 SD 12.7	**0.006**
Males, n (%)	13 (59.1%)	22 (59.5%)	0.978
Living alone, n (%)	1 (4.5%)	8 (21.6%)	0.077
Working, n (%)	9 (42.9%)	22 (61%)	0.182
NYHA class	1.7 SD 0.8	1.7 SD 0.8	0.909
High anatomical complexity, n (%)	10 (45.5%)	12 (32.4%)	0.317
TOF-spectrum	11 (50%)	20 (54%)	0.763
CRT-D, n (%)	5 (22.7%)	11 (29.7%)	0.559
Indication—primary prevention, n (%)	12 (54.5%)	21 (58.3%)	0.777
Age at last cardiac surgery, y	19.6 SD 12.7	34.5 SD 16.0	**0.001**
Age at first ICD implant, y	35.7 SD 13.0	44.9 SD 13.0	**0.011**
Years with ICD	3.6 SD 2.8	4.22 SD 3.15	0.438
Moderate or severe systemic ventricular dysfunction, n (%)	4 (18%)	15 (41%)	0.08
ICD shock, n (%)	6 (27.3%)	17 (45.9%)	0.155
Multiple ICD shocks, n (%)	6 (27.3%)	11 (29.7%)	0.840
Appropriate ICD shocks, n (%)	2 (9.1%)	13 (35.1%)	**0.033**
Inappropriate ICD shocks, n (%)	5 (22.7%)	13 (35.1%)	0.317
SF-36—Physical Component Summary	40.9 SD 10.7	42.5 SD 12.6	0.62
SF36—Mental Component Summary	40.5 SD 11.8	49.7 SD 12.6	**0.008**
HADS-A	8.91 SD 4.5	5.9 SD 4.6	**0.018**
HADS-D	7.0 SD 4.8	3.8 SD 4.2	**0.01**
Anxiety, n (%)	13 (59.1%)	12 (32.4%)	**0.045**
Depression, n (%)	11 (50%)	4 (10.8%)	**0.001**

CRT-D; cardiac resynchronisation therapy-defibrillator. FPAS; Florida Patient Acceptance Scale. HADS-A Hospital Anxiety and Depression Scale-Anxiety subscale. HADS-D; Hospital Anxiety and Depression Scale-Depression subscale. ICD; implantable cardioverter defibrillator. NYHA; New York Heart Association functional class. TOF; tetralogy of Fallot. Significant differences are highlighted in bold.

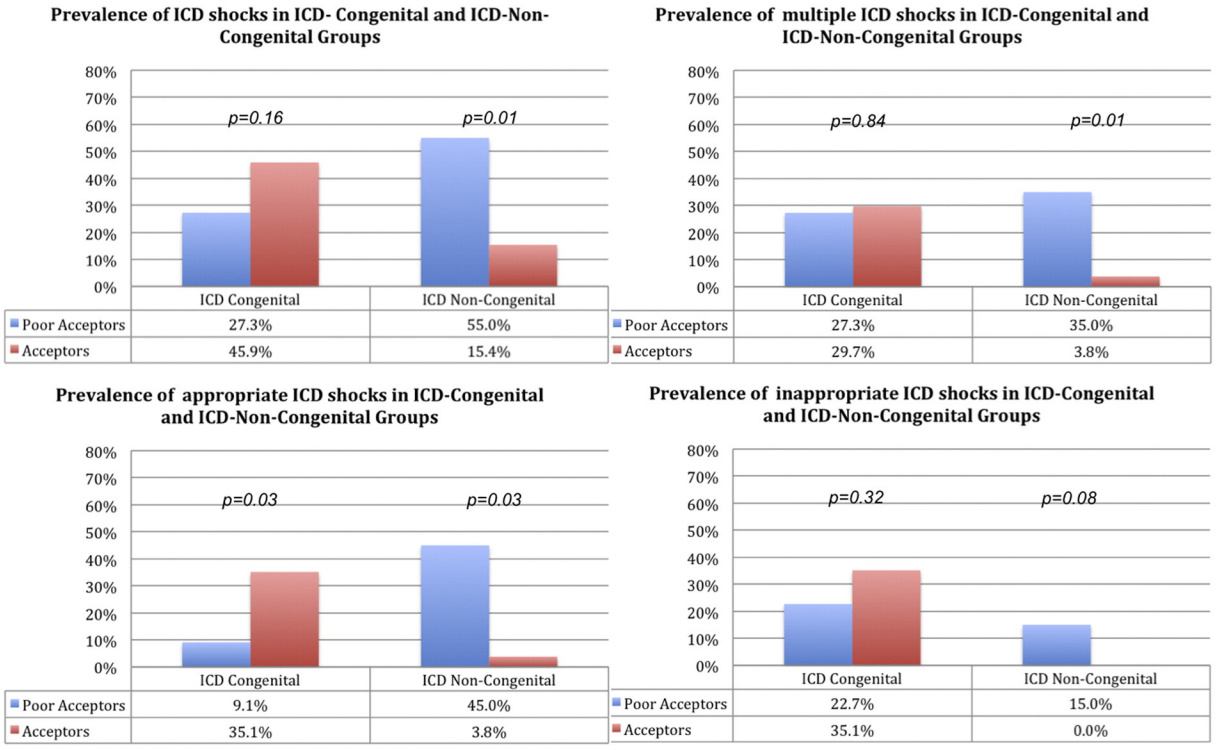

Fig. 1. Bar charts showing the prevalence of ICD shocks in Acceptors (FPAS >=67) and Poor Acceptors (FPAS < 67) of the ICD-Congenital and ICD-Non-Congenital groups. ICD: Implantable Cardiovertor Defibrillator. FPAS: Florida Patient Acceptance Survey.

4. Discussion

Younger adult age and lack of appropriate shocks are associated with poor ICD acceptance in congenital heart disease patients, which in turn, is associated with high levels of anxiety and depression. Patients with congenital heart disease and ICDs appear to have a worse quality of life and are more likely to be depressed compared to patients without ICDs, whilst the psychological wellbeing of patients with congenital or non-congenital heart disease and ICDs is similar.

Although ICDs can save lives, the need for heightened awareness of the potential negative effects from implantable defibrillators and the need for further studies relating to different patient populations receiving ICDs have been previously emphasized [26]. To our knowledge, this is the first study assessing ICD acceptance in ACHD patients in depth and given that 147 ACHD patients participated, one of the largest studies examining the psychological wellbeing of ACHD patients living with an ICD.

4.1. Risk factors for poor acceptance of ICDs by ACHD patients

Although device acceptance in the ICD-Congenital patients was comparable to the non-congenital ICD cohorts, it was significantly lower than in our PPM-Congenital group, with ICD-Congenital patients additionally showing lower 'Return to Living' scores. The poorer acceptance in congenital patients with ICDs compared to that with pacemakers might be related to a number of factors.

Pacemakers were implanted in symptomatic ACHD patients. ICD implantation was for primary prevention in half of the ACHD patients that will have understood a potential rather than actual need for an ICD. As the majority of these primary prevention ICD ACHD patients have not been symptomatic from an arrhythmia perspective, acceptance of the device may be more difficult. For patients, a heightened awareness of the risk of dying suddenly, the potential for appropriate but also inappropriate shocks and uncertainties about their individual requirement for ICD protection could also lead to lower acceptance levels and, thus, make 'Return to Living' arduous.

We did not find any differences in device acceptance or psychological well-being between congenital and non-congenital heart disease ICD recipients. Our findings contrast with those of Opic et al., who found that tetralogy of Fallot patients showed more anxiety and less satisfaction with life when compared to an older group of patients with acquired heart disease and ICDs [14]. Our findings were in a similar number of tetralogy of Fallot patients (n = 31) but we included a wider spectrum of ACHD patients of which one third had complex disease. Our non-congenital ICD group was also different than in the Opic et al. study. We included predominantly similarly younger aged patients with cardiomyopathy and channelopathy, with a minority having ischaemic heart disease. Psychological well-being may also be negatively affected in cardiomyopathy and channelopathy patients related to the familial nature of these conditions [6].

By comparing our "Non-Acceptors" to the remaining congenital-ICD recipients, we identified risk factors for poor device acceptance. This group of "Non-Acceptors" showed very high levels of anxiety, depression and worse mental health. Age emerged as a risk factor, with younger adult patients showing worse acceptance. Multiple studies in acquired heart disease patients also suggest that younger age is associated with worse psychological functioning with ICDs [5,27]. This is an important finding as, not only does it identify a 'target group' for heightened awareness and possible intervention, but also concerns a group of patients who are likely to be living with ICDs for longer.

A recent prospective study showed that adults with congenital heart disease had more shock-related anxiety than that reported for ICD recipients in the general population [15]. Fewer appropriate shocks also emerged as a risk factor for poor device acceptance in ACHD patients. We postulate that receiving an appropriate shock may be perceived by ACHD patients as confirmation of the need for ICD implantation and that is will work if required to prevent sudden cardiac death. Furthermore, a higher ratio of inappropriate to appropriate shocks was associated with poor acceptance, as perhaps expected. Inappropriate shock is of concern given the relationship between receiving shock therapy and anxiety. The strikingly higher rate of inappropriate shock in the ICD-Congenital group (31%) is consistent with previous

descriptions, and in particular the significant proportion (14%) of patients receiving only inappropriate shocks. This may be related to multiple factors including ECG discrimination and atrial arrhythmia—sufficient detailed previous device interrogation data are unavailable for defining the relative contribution of each.

We did not find a relationship between ICD duration and acceptance, but with the lifetime risk of shocks increasing with time, and the complex relationship between shocks and psychological health, we appreciate that our assessment of psychological state at a single point in time has obvious limitations. Awareness for the potential for both improvement and decline in psychological health with time is paramount.

4.2. Impaired quality of life in ACHD patients with ICDs

Our data suggest that ACHD patients with ICDs have worse quality of life than their ACHD counterparts without any device. We cannot establish a causative effect of ICDs on the worse perceived health status and, as recipients of ICDs may be at more advanced stages in their disease process, the observed differences in subscales of physical health may reflect this. It is clear though, that patients with an ICD report lower quality of life, on a physical and emotional level. This further translates into a limitation in their ability to function socially and take on their 'roles' at an emotional and physical level. These multiple factors are highlighted in the SF-36 subscale score and are reflected in the smaller proportion of ICD-Congenital patients working compared to the No-Device group.

Within the congenital heart disease subgroups, those with an ICD showed high levels of depression, with 25% reporting clinical depression, significantly higher than in patients without any devices. A prevalence of 8%–50% for mood and anxiety disorders in patients with congenital heart disease has been previously reported [17,28]. This strong relationship between the presence of the ICD and depression underpins the importance of taking the adverse psychological effects of ICD implantation into consideration when prescribing such therapy and identifies ACHD patients as potential subjects for routine psychological screening.

4.3. Clinical implications

Whilst the majority of patients with congenital heart disease with ICDs cope well, the findings from this study indicate that a significant proportion of these patients have anxiety and depression. A number of psychological and educational interventions can be instituted at counselling, pre-implant and during follow-up stages of ICD therapy [26]. Our findings suggest that further research in ACHD patients to better define factors leading to poor psychological adjustment is warranted. Meanwhile, cardiologists caring for patients with congenital heart disease may benefit from the regular use of simple screening procedures to identify those with psychological needs, including younger patients that may be particularly affected, to enable referral to appropriate psychological support services, where clinically indicated. Given that inappropriate shocks are associated with problems with anxiety and depression, our data justifies studies of interventions such as optimized ICD programming and more aggressive management of atrial arrhythmia to investigate whether inappropriate shock rates and psychological well-being can be improved. Risk stratification to identify patients who warrant sudden cardiac death primary prevention with ICD implantation amongst ACHD patients remains challenging, but as it continues to evolve, the inclusion of findings from our study into resource provision and the counselling process should be considered.

4.4. Limitations

Due to its cross-sectional design, our study cannot establish cause and effect relationships in ACHD patients with ICDs and limitations in psychological functioning. To obviate for this, comparison groups of patients with and without congenital heart disease and ICDs were used. Also related to the study's cross-sectional nature is the absence of serial data for the individual patient, which could strengthen the findings if shown to be reproducible over time. This has been described in patients with permanent pacemakers [29] and may warrant a further follow-up study in our cohort. The differences in SF-36 that we report between the groups may not be clinically relevant if used for longitudinal studies to assess changes for the same patient. For longitudinal studies, published clinically important differences for SF-36 should be referred to [30]. Our cut-off for device acceptance of 67 also needs further validation in larger studies. Our response rate to the questionnaires was 61%. This is not unexpected in a study of this nature [31], particularly given the limitation of re-contacting patients only once, as advised by the ethics committee. We submit that potential biases such as relatively increased participation of patients at both ends of the spectrum of psychological functioning, may be evenly distributed across the four groups and not only confined to the ICD-Congenital group. Heterogeneity in the spectrum of congenital heart disease, even within individual diagnoses, makes delineation of the precise effects of ICDs versus other clinical factors associated with impaired quality of life, challenging.

Nevertheless, to our knowledge, this is one of the largest studies looking specifically at ICD recipients in an ACHD population and there were clear findings that patients with ICDs and ACHD may have unmet psychological needs.

5. Conclusion

Whilst most patients with congenital heart disease cope well with implantable cardiac devices, recipients of ICDs have lower device acceptance compared to ACHD counterparts with permanent pacing. Physicians should be aware that a number of ACHD patients receiving ICD therapy will have difficulties adjusting to and living with their device. This is more likely in younger patients who receive inappropriate shocks, particularly in the absence of appropriate shocks. Appropriate counselling for ACHD patients subjected to or referred for ICD therapy may be necessary and the potential of intervention to improve acceptance and psychological well-being warrants study.

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

Acknowledgements

This work was supported in part by an unrestricted educational grant from Medtronic. The project was supported by the NIHR Cardiovascular Biomedical Research Unit at the Royal Brompton and Harefield NHS Foundation Trust and Imperial College London. SVBN is supported by the British Heart Foundation (FS/11/38/28864). We are grateful to our patients who generously donated their time to make this work possible.

References

[1] B.J. Mulder, Epidemiology of adult congenital heart disease: demographic variations worldwide, Neth. Heart J. 20 (12) (2012) 505–508.
[2] J.E. Deanfield, S.Y. Ho, R.H. Anderson, et al., Late sudden death after repair of tetralogy of Fallot: a clinicopathologic study, Circulation 67 (3) (1983) 626–631.
[3] P. Khairy, L. Harris, M.J. Landzberg, et al., Implantable cardioverter-defibrillators in tetralogy of Fallot, Circulation 117 (3) (2008) 363–370.
[4] P. Khairy, L. Harris, M.J. Landzberg, et al., Sudden death and defibrillators in transposition of the great arteries with intra-atrial baffles: a multicenter study, Circ. Arrhythm. Electrophysiol. 1 (4) (2008) 250–257.
[5] S.F. Sears Jr., J.B. Conti, Quality of life and psychological functioning of ICD patients, Heart 87 (5) (2002) 488–493.
[6] C.A. James, C. Tichnell, B. Murray, et al., General and disease-specific psychosocial adjustment in patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy with implantable cardioverter defibrillators: a large cohort study, Circ. Cardiovasc. Genet. 5 (1) (2012) 18–24.

[7] S.S. Pedersen, H. Spindler, J.B. Johansen, et al., Correlates of patient acceptance of the cardioverter defibrillator: cross-validation of the Florida Patient Acceptance Survey in Danish patients, Pacing Clin. Electrophysiol. 31 (9) (2008) 1168–1177.
[8] J. Irvine, P. Dorian, B. Baker, et al., Quality of life in the Canadian Implantable Defibrillator Study (CIDS), Am. Heart J. 144 (2) (2002) 282–289.
[9] S.F. Sears, T.S. Lewis, E.A. Kuhl, et al., Predictors of quality of life in patients with implantable cardioverter defibrillators, Psychosomatics 46 (5) (2005) 451–457.
[10] R. Passman, A. Kadish, S. Sears, Defibrillators in heart failure and quality of life, N. Engl. J. Med. 360 (2) (2009) 188–189 (author reply 89).
[11] S.S. Pedersen, S.F. Sears, M.M. Burg, et al., Does ICD indication affect quality of life and levels of distress? Pacing Clin. Electrophysiol. 32 (2) (2009) 153–156.
[12] D.L. Carroll, G.A. Hamilton, Quality of life in implanted cardioverter defibrillator recipients: the impact of a device shock, Heart Lung 34 (3) (2005) 169–178.
[13] H.C. Kamphuis, J.R. de Leeuw, R. Derksen, et al., Implantable cardioverter defibrillator recipients: quality of life in recipients with and without ICD shock delivery: a prospective study, Europace 5 (4) (2003) 381–389.
[14] P. Opic, E.M. Utens, P. Moons, et al., Psychosocial impact of implantable cardioverter defibrillators (ICD) in young adults with Tetralogy of Fallot, Clin. Res. Cardiol. 101 (7) (2012) 509–519.
[15] S.C. Cook, A.M. Valente, T.M. Maul, et al., Shock-related anxiety and sexual function in adults with congenital heart disease and implantable cardioverter-defibrillators, Heart Rhythm. 10 (6) (2013) 805–810.
[16] D.A. Lane, G.Y. Lip, T.A. Millane, Quality of life in adults with congenital heart disease, Heart 88 (1) (2002) 71–75.
[17] A.H. Kovacs, A.S. Saidi, E.A. Kuhl, et al., Depression and anxiety in adult congenital heart disease: predictors and prevalence, Int. J. Cardiol. 137 (2) (2009) 158–164.
[18] C.A. Warnes, R. Liberthson, G.K. Danielson, et al., Task force 1: the changing profile of congenital heart disease in adult life, J. Am. Coll. Cardiol. 37 (5) (2001) 1170–1175.
[19] J.L. Burns, E.R. Serber, S. Keim, et al., Measuring patient acceptance of implantable cardiac device therapy: initial psychometric investigation of the Florida Patient Acceptance Survey, J. Cardiovasc. Electrophysiol. 16 (4) (2005) 384–390.
[20] H. Versteeg, A. Starrenburg, J. Denollet, et al., Monitoring device acceptance in implantable cardioverter defibrillator patients using the Florida Patient Acceptance Survey, Pacing Clin. Electrophysiol. 35 (3) (2012) 283–293.
[21] K.Z. Bambauer, S.E. Locke, O. Aupont, et al., Using the Hospital Anxiety and Depression Scale to screen for depression in cardiac patients, Gen. Hosp. Psychiatry 27 (4) (2005) 275–284.
[22] R.P. Snaith, A.S. Zigmond, The hospital anxiety and depression scale, Br. Med. J. 292 (6516) (1986) 344.
[23] A.S. Zigmond, R.P. Snaith, The hospital anxiety and depression scale, Acta Psychiatr. Scand. 67 (6) (1983) 361–370.
[24] J.E. Ware Jr., C.D. Sherbourne, The MOS 36-item short-form health survey (SF-36). I. Conceptual framework and item selection, Med. Care 30 (6) (1992) 473–483.
[25] C.A. McHorney, J.E. Ware Jr., A.E. Raczek, The MOS 36-Item Short-Form Health Survey (SF-36): II. Psychometric and clinical tests of validity in measuring physical and mental health constructs, Med. Care 31 (3) (1993) 247–263.
[26] S.B. Dunbar, C.M. Dougherty, S.F. Sears, et al., Educational and psychological interventions to improve outcomes for recipients of implantable cardioverter defibrillators and their families: a scientific statement from the American Heart Association, Circulation 126 (17) (2012) 2146–2172.
[27] L.D. Vazquez, E.A. Kuhl, J.B. Shea, et al., Age-specific differences in women with implantable cardioverter defibrillators: an international multi center study, Pacing Clin. Electrophysiol. 31 (12) (2008) 1528–1534.
[28] J.I. Bromberg, P.J. Beasley, E.J. D'Angelo, et al., Depression and anxiety in adults with congenital heart disease: a pilot study, Heart Lung 32 (2) (2003) 105–110.
[29] E.O. Udo, N.M. van Hemel, N.P. Zuithoff, et al., Long term quality-of-life in patients with bradycardia pacemaker implantation, Int. J. Cardiol. 168 (3) (2013) 2159–2163.
[30] K.W. Wyrwich, J.A. Spertus, K. Kroenke, et al., Clinically important differences in health status for patients with heart disease: an expert consensus panel report, Am. Heart J. 147 (4) (2004) 615–622.
[31] J.E. Fincham, Response rates and responsiveness for surveys, standards, and the Journal, Am. J. Pharm. Educ. 72 (2) (2008) 43.