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Original Article

Patient Preferences in Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) Screening and ICD Implantation: Canadian ARVC Registry Substudy

Sandra L. Carroll, PhD, RN,[a,b] Daryl Pullman, PhD,[c] Martin Gardner, MD, FRCPC,[d] Andrew D. Krahn, MD, FRCPC, FHRS,[e] and Jeff S. Healey, MD, FRCPC[b]

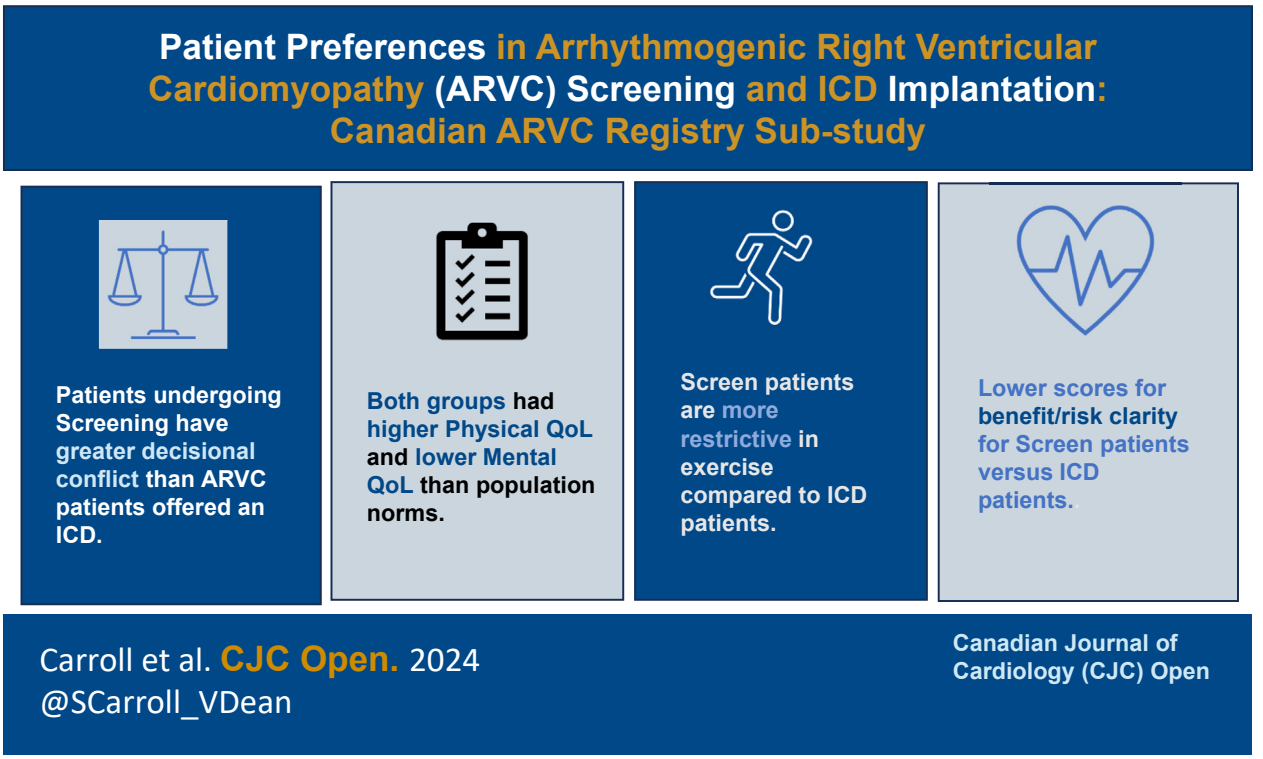
[a] School of Nursing, McMaster University, Hamilton, Ontario, Canada

[b] Population Health Research Institute, Hamilton, Ontario, Canada

[c] Faculty of Medicine, M4M211 Memorial University of Newfoundland St. John's, Newfoundland, Canada

[d] Division of Cardiology, Dalhousie University, Halifax, Nova Scotia, Canada

[e] Faculty of Medicine, University of British Columbia, Vancouver, British Columbia, Canada



ABSTRACT

Background: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is typically diagnosed following an arrhythmic event or during screening after a family member experiences sudden cardiac death. Implantation of a defibrillator (ICD) improves survival but can be associated with morbidity and risks, an important consideration within

RÉSUMÉ

Contexte : La cardiomyopathie ventriculaire droite arythmogène (CVDA) est généralement diagnostiquée à la suite d'un événement arythmique ou lors d'un dépistage après la mort subite cardiaque d'un membre de la famille. L'implantation d'un défibrillateur (DCI) améliore la survie mais peut être associée à une morbidité et à certains risques,

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a shared decision-making context. This study examined patient decisional needs and preferences surrounding ARVC screening and prophylactic ICD implantation.

Methods: **This Canadian ARVC registry substudy included 20 new patients and/or families offered ARVC screening (the screen group), and 27 diagnosed ARVC patients who were offered an ICD. Measures included the following: the Decisional Conflict Scale; preference and benefit−risk visual analogue scales; the Medical Outcomes Study Short Form-36 (SF-36); and exercise restriction. Descriptive analysis was employed, and results are reported as mean (standard deviation) or proportions.**

Results: **ICD patients reported having lower decisional conflict scores—19.6 (13.6) compared to the screen group patients—33.1 (32.2). The visual analogue scale results showed lower benefit and risk clarity scores for screen group patients—6.6 (3.6)—compared to those offered ICD implantation—7.4 (2.6). More screen group patients (55%) reported restricting exercise than did ICD patients (30%). In both groups, the Medical Outcomes Study Short Form-36 Physical Component Summary scores were higher than population norms—50 (standard deviation 10): the screen group, 52.0 (8.8); the ICD group, 54.1 (7.4), and the Mental Component Summary scores were slightly lower—the screen group, 47.7 (10.8); the ICD group, 49.7 (8.9).**

Conclusions: **Patients undergoing ARVC screening reported greater decisional conflict and lower benefit and risk clarity compared to patients diagnosed with ARVC who were offered an ICD. Screen group patients were more restrictive in their exercise. Understanding patient preferences and needs during ARVC screening and ICD candidacy can assist in improving decision support with patients and families.**

ce qui constitue un élément important à prendre en considération dans le cadre d'une prise de décision partagée. Cette étude a examiné les besoins et les préférences décisionnels des patients concernant le dépistage de la CVDA et l'implantation prophylactique d'un DCI.

Méthodes : **Cette sous-étude du registre canadien de la CVDA a porté sur 20 nouveaux patients et/ou familles à qui l'on a proposé un dépistage de la CVDA (groupe de dépistage), et sur 27 patients ayant reçu un diagnostic de CVDA à qui l'on a proposé un DCI. Les mesures comprenaient les éléments suivants: l'Échelle de conflit décisionnel, les Échelles visuelles analogiques de préférence et de bénéfice-risque, le Medical Outcomes Study Short Form-36 (SF-36), et la restriction de l'exercice physique. Une analyse descriptive a été utilisée et les résultats sont présentés sous forme de moyenne (écart-type) ou de proportions.**

Résultats : **Les patients porteurs d'un DCI ont déclaré avoir des scores de conflit décisionnel inférieurs − 19,6 (13,6) - à ceux des patients du groupe de dépistage − 33,1 (32,2). Les résultats des Échelles visuelles analogiques ont montré des scores de clarté des bénéfices et des risques inférieurs pour les patients du groupe de dépistage − 6,6 (3,6) - par rapport à ceux à qui l'on a proposé l'implantation d'un DCI − 7,4 (2,6). Un plus grand nombre de patients du groupe de dépistage (55 %) que de patients bénéficiant d'un DCI (30 %) ont déclaré restreindre leurs activités physiques. Dans les deux groupes, les scores de la composante physique du Medical Outcomes Study Short Form-36 étaient plus élevés que les normes de la population - 50 (écart-type 10): groupe dépistage, 52,0 (8,8); groupe DCI, 54,1 (7,4), tandis que les scores de la composante mentale étaient légèrement inférieurs − groupe dépistage, 47,7 (10,8); groupe DCI, 49,7 (8,9).**

Conclusions : **Les patients soumis à un dépistage de la CVDA ont fait état d'un plus grand conflit décisionnel et d'une moins grande clarté des bénéfices et des risques que les patients chez qui on a diagnostiqué une CVDA et à qui on a proposé un DCI. Les patients du groupe de dépistage étaient plus restrictifs dans leur activité physique. La compréhension des préférences et des besoins des patients lors du dépistage de la CVDA et de la candidature à l'implantation d'un DCI peut contribuer à améliorer l'aide à la décision avec les patients et les familles.**

Advances in clinical testing and diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) have led to the identification of patients and family members who may be at risk for sudden cardiac death (SCD) associated with this familial condition.[1] The process of ARVC diagnosis is multifaceted and consists of several diagnostic modalities, often including genetic testing.[1,2] Diagnosis had been guided by the modified 2010 ARVC Task Force criteria,[3] which include the following 6 major and minor categories: (i) global or regional dysfunction and structural alterations; (ii) tissue characterization of wall; (iii) repolarization abnormalities on electrocardiogram; (iv) depolarization and/or conduction abnormalities on electrocardiogram; (v) arrhythmias; and (vi) family history. In 2023, these guidelines were updated to include those with biventricular and left ventricular cardiomyopathy.[4,5] The prevalence of ARVC is between 1 of 2000 to 1 of 5000, with as many as 20% having atrial arrythmias.[6] About two-thirds of those meeting ARVC Task Force criteria contain a gene mutation.[6] Patients and family members may learn of their ARVC risk after an initial presentation of an arrhythmic episode or the SCD of a family member. These occurrences are frequently the impetus for genetic testing and clinical investigation, with genetic testing recommended for those who meet guideline criteria, as well as their first-degree relatives.

Patients and family members who are navigating the diagnostic process of inherited rhythm diseases may experience uncertainty, decisional conflict, or changes in quality of life when learning of and making decisions related to their inherited arrhythmia risk.[7-9] When clinically indicated, patients may be offered an implantable cardioverter defibrillator (ICD) as prophylaxis for SCD from a life-threatening arrhythmia. For ARVC patients who are offered an ICD, the decision to receive a medical device requires them to consider both benefits and risks.[10] This approach includes

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Corresponding author: Sandra L. Carroll, RN, PhD, Professor, Faculty of Health Sciences, School of Nursing, McMaster University, 1280 Main St. W., Hamilton, Ontario L8S 4L8, Canada. Tel.: 905-525-9140 x22400.

E-mail: carroll@mcmaster.ca

See page 33 for disclosure information.

understanding ICD-specific complications, such as infection, lead or device generator issues, as well as the potential need for ongoing ICD replacements.[11] Both appropriate and inappropriate shocks can occur, and literature suggests a pooled event rate of 9.1%,[11] and an annual risk of inappropriate shocks of 3.9%.[6] Having greater knowledge about patients' perceptions and preferences for genetic testing and ICD implantation can assist in determining how best to facilitate quality decision-making to support this patient population.

In Canada, a prospective ARVC registry was established in 2013 to inform the natural history of ARVC patients, including the annual risk of SCD and/or shocks for persons with the phenotype, as well as those who were gene positive without phenotype evidence of ARVC.[12] The registry, now part of the Hearts in Rhythm Organization (HiRO) (www.heartsinrhythm.ca),[13] offered an opportunity to understand what patients deemed to be important when they are undergoing genetic screening or considering prophylactic treatments for ARVC. This study examined patient and family perspectives regarding ARVC decision-making in an effort to strengthen ARVC treatment discussions within arrythmia clinics.

Methods

Design

This prospective descriptive substudy was from the larger observational ARVC registry,[12] which enrolled patients and families and followed them over time. The primary aim of this substudy was to document the decisions, decisional needs, and generic quality of life in patients offered ARVC screening and patients offered an ICD as prophylaxis for SCD. All participants consented to join the ARVC registry, providing an additional written consent for this substudy. Ethics approval was received from all participating sites (HiREB #13-183). The research reported in this paper adhered to the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement (https://www.equator-network.org/reporting-guidelines/strobe/.

Sample

The study population consisted of 2 groups who were included in the Canadian ARVC registry[12]: (i) new ARVC registry patients and/or family members attending an arrhythmia clinic and undergoing ARVC screening (screen participants); and (ii) diagnosed ARVC patients who were offered an ICD (ICD participants). The recruitment target was 50 participants across groups. In both cases, only English-speaking patients aged > 18 years were included. Those who met the 2010 ARVC Task Force criteria,[3] with diagnosed or possible ARVC, as well as first-degree relatives that were affected and unaffected, were invited to join the ARVC registry. Baseline data were collected, and previous testing results were included so that both prevalent and incident cases could be noted.[12] In this substudy, ICD participants were those who already had a diagnosis of ARVC or probable ARVC and either had been offered an ICD or in some instances, already had an ICD. Screen participants were those undergoing testing for ARVC. Although the total size of the ARVC registry at the time of the study is not known, 365 patients were enrolled in the initial registry publication,[12] with this substudy taking place at around the same time.

Setting

This study took place at inherited arrhythmia cardiology clinics in 3 provinces in Canada (Ontario, British Columbia, and Nova Scotia). These clinics are part of the current Canadian Genetics Heart Rhythm Network of Inherited Heart Rhythm Clinics located in hospital settings, with data being stored in academic institutions.[12] A clinic in each province was invited to take part in the substudy; however, the majority of participants were from the Ontario site, with 1 from British Columbia, and 3 from the Nova Scotia site. At the time of the substudy, 15 clinics were available across Canada, and since then, with the formation of the National Hearts in Rhythm Organization Registry in 2016, 20 cardiogenetic clinics have become available.[13]

Procedures

Participants who consented to enroll in the ARVC registry were provided with information about the substudy. Those who wanted to learn more about the study were then contacted by a research assistant who asked them to discuss the study and review consent documents. Patients and families who agreed to take part were given a study package of surveys to complete, following their specialist appointment.

Measures

Demographics and characteristics. Question topics included level of education, age, sex, marital status, employment status, dependents, completion of genetic counselling, family history of SCD, exercise restriction and whether it was recommended or self-imposed, and final decisions about whether the participant had been offered or had accepted screening or an ICD.

Decisional Conflict Scale (DCS). The conceptual construct of decisional conflict is intended to reflect uncertainty related to an action. The DCS was developed to examine uncertainty regarding choices in a healthcare decision, and it has demonstrated good reliability and validity.[14] The DCS can be used either before or after a decision is made,[15] and it has been used as an outcome measure in intervention studies to examine the effects of a decision-support intervention. Clinically significant decisional conflict is recognized as being either < 25 or > 37.5. Lower DCS scores (< 25) are associated with a higher likelihood of implementing a health-related decision.[14] Two versions of the DCS were used in this study. The screen group received the traditional DCS, and the ICD group received the low-literacy version, in an attempt to inform the use of a more accessible form of the tool. The low-literacy version of the DCS has been found to have good internal consistency and discriminant validity, along with moderate construct validity.[16] Scoring for both versions results in scores that range from 0 (no decisional conflict) to 100 (extremely high decisional conflict).[17]

Benefit–risk visual analogue scale. The following question—"Are you clear about which benefits and risks about

receiving an ICD/undergoing screening for ARVC matter most to you?"—was rated on a 10-point scale, with scores ranging from 0 = not clear at all, to 10 = extremely clear. Higher scores indicate more clarity. This rating scale was developed by investigators for the present study.

Decisional outcomes. Information about patient decisional outcomes of acceptance of screening and being offered an ICD was extracted from medical records.

Current patient priorities. Patients were asked the following: "At this time in your life, what are your most important priorities?," and they were asked to rank their top 7 from a list that included: family, career, employment, sports, income, well-being, health symptoms, absence of symptoms, peace of mind, absence of worry, living a long life (despite their health), living a long life (with disability), and other priorities (noted by patient).

Medical Outcomes Study Short Form-36 V2 (SF-36). The SF-36 is a self-report measure of generic quality of life. The survey consists of 36 items across 8 subscales (vitality, physical functioning, bodily pain, general health perceptions, physical role functioning, emotional role functioning, social role functioning, and mental health), which provide overall physical component summary (PCS) and mental component summary (MCS) scores.[18] For ease of comparison, scores are converted to *t*-scores, with a mean of 50, and a standard deviation of 10. Zero represents maximum disability, and 100 represents no disability. The SF-36 is used widely and has been shown to have sound validity and reliability.[18,19]

Analysis

Descriptive analyses were completed to examine patient decisions, decisional conflict, and device acceptance. Due to the small sample size, no tests of significance were performed for these variables, nor were analyses conducted to examine clinical significance. Categorical variables were summarized using frequency and percentage; continuous variables were summarized using measures of central tendency, means, and standard deviations (SDs). Patient priorities were examined using frequencies, and the top 3 priorities were extracted, as these likely represented what was most important to patients.

Results

Forty-seven patients participated in the substudy, 20 patients (43%) participated in the screen group, and 27 patients participated in the ICD group (57%). Seventy percent of the ICD group (n = 19) and 50% of the screen group (n = 9) were male participants, with at least 70% in each group indicating that they were married and/or had a common law marriage. A total of 45% of the screen group (n = 9), and 52% of the ICD group (n = 14), reported having dependents at home. Similar proportions of participants in both groups reported having a family member who had an SCD (25%; screen = 5; ICD = 7). Table 1 summarizes the characteristics of the samples.

As highlighted in Table 2, ICD participants had mean DCS score of 19.6 (SD = 13.6), and screen group patients

Table 1. Participant characteristics

Characteristic, n (%)	Screen group n = 20	ICD group n = 27
Sex		
Female	10 (50)	8 (30)
Male	9 (45)	19 (70)
Not reported	1 (5)	
Age, y, mean (SD)	n = 20*	n = 27
	45.0 (15.52)	48.5 (18)
Highest level of education		
High school	5 (25)	10 (37)
College and/or technical and/or trade	6 (30)	7 (26)
University degree	7 (35)	7 (26)
Graduate and/or professional	2 (10)	3 (11)
Marital status		
Married or common law marriage	14 (70)	20 (74)
Living with a partner	1 (5)	0 (0)
Not in a union	0 (0)	1 (4)
Single and/or never married	5 (25)	6 (22)
Employment status		
Full-time	11 (55)	13 (48)
Part-time	5 (25)	5 (19)
Seeking employment	2 (10)	2 (7)
Retired	2 (10)	4 (15)
Disability	0 (0)	2 (7)
Not reported		1 (4)
Family history of sudden cardiac death		
Yes	5 (25)	7 (26)
No	15 (75)	20 (74)

ICD, implantable cardioverter defibrillator; SD, standard deviation.

*Three participants only had year of birth (date and month chosen as June 6th).

33.1 (SD = 32.2). DCSs < 25 indicate a greater likelihood of implementing the decision.[15] Most patients in both the screen and the ICD groups preferred to share decision-making with someone else, rather than making this decision on their own. The majority reported that they wanted to make their decision with a family member or their physician. No participants indicated that they wanted someone else to make their decision for them.

Benefit—risk visual analogue scale ratings (scores of 0-10 (mean [SD]) revealed that the screen group participants were less clear about the benefits and risks that mattered most to them—score of 6.6 (SD, 3.6), compared to ICD participants—score of 7.5 (SD, 2.6). When asked about their preference at the time of study participation, approximately 11% of ICD group participants (n = 3) and 5% of screen group participants (n = 1) reported that they were unsure of whether they wanted a screening or to receive an ICD. Medical records indicated that all screen group participants (n = 20), and 89% of ICD group participants (n = 24) accepted screening, with 96% of ICD patients (n = 23) and 15% of screen group patients (n = 3) being offered an ICD. Some screen group participants (n = 3) did not meet the criteria for receiving an ICD, and the remaining participants did not have their ICD status recorded at the time of their chart review.

In examining the current patient priorities of participants, family was noted as the top priority for both the ICD and the screen groups, followed by well-being for both groups. The

Table 2. Decisional needs and preferences

Measure of decision-related need or preference	Screen group* n = 20	ICD group N = 27
Decisional conflict scale, mean (SD)	(n = 19)	(n = 27)
Total score	33.2 (32.2)	19.6 (13.6)
Uncertainty subscale	35.5 (42.8)	21.0 (19.7)
Informed subscale	38.6 (41.6)	21.6 (15.6)
Values clarity subscale	35.5 (32.6)	21.3 (15.0)
Support subscale	24.6 (25.7)	17.9 (16.3)
Effective decision subscale*	—	17.1 (17.8)
Preferred role in making health choices, n (%)		
Missing	1 (5)	0
Share decision with someone else	11 (55)	17 (63)
Decide myself after hearing other's views	8 (40)	10 (37)
Prefer that someone else decides	0	0
Values, mean (SD)	n = 19	n = 27
How important is receiving an ICD/ knowing risk of ARVC	9.0 (2.4)	6.5 (4.1)
How clear about benefits & risks that matter most when receiving an ICD/ undergoing screening	6.6 (3.6)	7.5 (2.6)
Preference about ICD and/or screening, n (%)		
Missing	1 (5)	0
Receive ICD and/or screening	18 (90)	15 (56)
Not receive ICD and/or screening	0	9 (33)
Unsure	1 (5)	3 (11)

ARVC, arrhythmogenic right ventricular cardiomyopathy; ICD, implantable cardioverter defibrillator; SD, standard deviation.

*The screen group used the low-literacy version of the Decisional Conflict Scale, which does not include the effective decision subscale.

third priority for the ICD group was health symptoms, and peace of mind was the third priority for the screen group participants.

More than half of the patients in the screen group (55%; n = 11) stated that they were restricting their exercise, compared to 30% in the ICD group (n = 8). A follow-up prompt asked whether exercise restriction was initiated because of their specialist recommendation, and 81.8% of the screen group patients (n = 9), compared to 37.5% of the ICD participants (n = 8) indicated that they were restricting their exercise at the suggestion of their specialist. In our sample, 30% of participants in each group (screen group = 6; ICD group = 8) self-reported receiving genetic counselling at the time of the study.

When considering norm-based SF-36 scoring, with which scores reported as < 50 represent health status as being below average[18]; PCS scores were > 50 in both groups; and MCS scores were lower than PCSs (see Table 3).

Table 3. Short-form-36 scale and component summary scores

Scale and component summary scores	Screen N = 20	ICD N = 27
Physical functioning	87.0 (17.3)	90.7 (13.9)
Role, physical	78.8 (27.8)	84.5 (19.6)
Bodily pain	76.9 (29.0)	82.8 (20.6)
General health	59.7 (21.6)	69.8 (17.9)
Vitality	59.4 (18.6)	59.0 (23.5)
Social functioning	82.5 (23.8)	85.7 (20.1)
Role, emotional	79.6 (30.9)	84.3 (23.6)
Mental health	69.8 (20.2)	76.7 (15.5)
Physical component score	52.0 (8.8)	54.1 (7.4)
Mental component score	47.7 (10.8)	49.7 (8.9)

Values are mean (standard deviation).
ICD, implantable cardioverter defibrillator.

Discussion

This ARVC registry substudy sought to explore the decisional needs and preferences of patients identified with ARVC or a family history of inherited arrhythmias who were either undergoing ARVC screening or had been offered an ICD for prophylaxis. Greater understanding of patients' perceptions and preferences for genetic testing and ICD implantation can assist with how best to facilitate quality decision-making to support this patient population and their families. We found that participants undergoing ARVC screening reported higher levels of decisional conflict, compared to those with ARVC who were offered an ICD. Also, patients offered screening were less certain about the benefits and risks of screening than were ICD patients about getting an ICD. Decisional conflict and decisional regret also have been reported in the context of exercise and shared decision-making.[20] Sweeney and colleagues found that shared decision-making was associated with lower rates of decisional conflict ($P < 0.01$).[20] Our findings suggest that patients may benefit from having access to additional support and information at the screening stage, and when they are considering an ICD, prior to possible implantation.

Several qualitative studies have examined the experience of patients with inherited arrhythmic conditions. Manuel and Brunger[9] interviewed patients who were involved with decision-making related to predictive genetic testing. They found that decision-making was experienced as either a process or a "fait accompli" and was mediated by a number of factors, including an experience of death within the family, a sense of responsibility for others, and disease progression, among others.[9] Patients' perceptions resulted from the interplay between scientific knowledge and their own experiential knowledge. Given that genetic counselling is often initiated after an adverse event experienced by the patient or a family member, the findings that those in the screen group exhibited higher DCS scores and more uncertainty about ICD risks and benefits are not surprising. Emerging evidence also suggests that those who have access to a genetic counsellor report having higher empowerment scores than those who do not.[21] As well, evidence demonstrates that patients exhibit cognitive framing biases that favour a treatment, downplaying the possible harms that can occur when making decisions about ICD implantation.[22] This literature is in line with our findings in which the ICD group reported having less decisional conflict compared with to the screen group.

Generic quality-of-life (SF-36) scores showed that our sample reported having MCS scores that were below population norms.[18] This finding is perhaps not surprising, as the decision to undergo ARVC screening or to receive an ICD can cause fear and uncertainty, aligning with literature indicating that patients want more support and information.[23] A total of 25% of the study sample reported having a family member who had a SCD, which adds to the fear and uncertainty as family members and/or children await results to find out if they too are at risk if the screening determines that they are positive for ARVC. Patient priorities were similar across groups; however, the third-ranked priority for the screen group patients was health symptoms and peace of mind for ICD patients, highlighting the psychological

impact of having an ICD for those that receive one.[23] As well, a review of studies examining the impact of psycho-educational interventions on quality of life found a statistically significant impact on PCS scores but not MCS scores,[24] suggesting that psychological adaptation may be a longer-term process that requires regular check-in and support.

Although not the major focus of this study, patient-reported exercise restriction was explored, and we found variability in the messaging that patients were receiving, with more screen-group patients restricting their exercise, based on specialist recommendation. A number of recent studies have examined exercise in athletes with arrythmias, reinforcing the need for evidence-based guidelines to support safe exercise levels. The avoidance of endurance exercise, and limiting exercise to 2.5 hours per week and to exercise during which maintaining a conversation is possible, has been suggested.[25] Other work examining genotype-positive family members of patients with ARVC was able to establish a dose-response for exercise, and found that those meeting the 2010 ARVC Task Force diagnostic criteria had a greater average intensity level and duration of exercise compared to those who did not meet these diagnostic criteria.[26] The need for a shared decision-making approach regarding exercise has been advocated.[6]

Both opting for inherited arrhythmia screening or ICD implantation involves consideration of benefits, risks, and preferences alongside input from clinicians and family members.[27] In this study, patients preferred making their decision with their physician or family member, noting also that family was their top-ranked priority. This finding reinforces research indicating that most patients seek a collaboration with family and medical professionals when deciding to undertake screening or have ICD implantation.[8,9] The literature also highlights that approximately 20% of patients had not wanted to receive an ICD when it was implanted,[28] and that some patients were unaware that they had the option of not having an ICD implanted.[29] Clearly, the exploration of patient values and needs is critical, and its absence can lead to decisional regret.[28]

Results from this ARVC registry substudy can inform decision support to *prepare* patients identified to be at risk for ARVC (patients considering), or those offered a prophylactic defibrillator (meeting Task Force evaluation criteria), to make informed decisions in partnership with electrophysiology specialists, genetic counsellor team members, advanced practice nurses, or other healthcare professionals in their circle of care. Specialized inherited arrhythmia teams,[13] including advanced practice nurses and genetic counsellors, play a key role in supporting patients in decision-making, especially in providing patients with tailored ICD education prior to implantation.[30] Shared decision-making has been suggested for those offered an ICD, with some agencies, such as the Centres for Medicare and Medicaid Services, requiring the use of a decision aid prior to device implantation,[31] recognizing the need to be attuned to each person's individual preferences for information and education. Ultimately, proactive measurement of these patient-centered outcomes could moderate decisional conflict and improve shared decision-making.

Study limitations and strengths

These findings offer some insight into the decisions, preferences, and experiences of patients undergoing screening or ICD implantation in the context of inherited arrhythmic conditions. However, a number of limitations should be considered. First, our small sample size limited the ability to complete inferential statistics, impacting the generalizability of the findings. This limitation also restricted our ability to examine sample characteristics, such as gender and sex.

As well, in each of the screen and ICD groups, only 30% of patients had received genetic counselling. Whether any patients went on to receive counselling after meeting with the research team, or had been offered genetic counselling but had declined it, was not known. Further, in specialized arrhythmia clinics in which electrophysiologists work alongside advanced practice nurses, a level of expertise is present that may not be found in other clinics. Healthcare organizations would benefit from having a standardized approach to offering and tracking services, such as genetic counselling.

Finally, we do not have detailed information about whether patients had had a prior cardiac arrest, which may have made them more amenable to treatment. Further, the time since diagnosis also was not available, and those with a less-recent diagnoses may have had more time to consider and understand their condition.

Despite these issues, the present study design has several merits. Capturing the experiences of patients soon after their specialist appointment lends credibility to the findings, as their experiences had no time to be influenced by other external factors. As well, this study adds to the literature through the inclusion of decisional outcome measures. Two versions of the DCS[15] were used, with no differences in completion rate. This finding supports the use of the low-literacy version in clinical research, which contributes to making research accessible.[16] It is acknowledged, however, that each cohort's use of a different version of the DCS carries potential bias. In future work, half of each cohort could be randomized to either version, to minimize bias. A 2017 scoping review focused on decision-making related to complex cardiac devices noted the scarcity of studies that included decisional outcome measures, such as decisional conflict.[5] This work could inform future studies and the determination of measures that merit consideration.

Conclusions

By considering and gleaning an understanding of the perspectives of patients and families during ARVC screening and/or ICD candidacy in the context of ARVC, new directions to enhance educational support for patients in practice can be considered. This study revealed that higher levels of decisional conflict are present in ARVC screen group patients, and lower mental health scores, both of which could be ameliorated by the provision of further information and/or support by health professionals in this field.

Implications for practice

- Additional support and education are needed for patients undergoing ARVC screening.
- Discussion and clarity regarding exercise restriction are warranted in both groups.
- SF-36 MCS scores reinforce the need for additional support in both groups.

Ethics Statement

Ethics approval was received from all participating sites (HiREB #13-183). The research reported in this paper adhered to the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement (https://www.equator-network.org/reporting-guidelines/strobe/.

Patient Consent

The authors confirm that a patient consent form has been obtained for this article. All participants consented to join the ARVC registry, providing an additional written consent for this substudy.

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Disclosures

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Supplementary Material

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