CHARACTERIZING DECISION-MAKING SURROUNDING EXERCISE IN ARVC: ANALYSIS OF DECISIONAL CONFLICT, DECISIONAL REGRET, AND SHARED DECISION-MAKING

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Abstract

Background: Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a genetic condition that predisposes individuals to arrythmia, cardiomyopathy, and sudden cardiac death (SCD). Because of associated risk, it is recommended that those diagnosed with or at-risk for ARVC restrict exercise. Guidelines recommend shared decision making (SDM), but there has been little exploration as to whether SDM is happening or its impact on exercise decision-making.

Objectives: To 1) describe the extent to which SDM is happening in the population, 2) characterize for whom SDM is happening more or less in the population, and 3) determine if shared decision-making is associated with decisional conflict, decisional regret, and adherence.

Methods: Adults diagnosed with ARVC or who have tested positive for genetic ARVC-risk enrolled in the Johns Hopkins ARVC Registry were invited to complete a one-time questionnaire that included exercise history, athlete identity, SDM (SDM-Q-9), decisional conflict (DCS), and decisional regret (DRS).

Results: 205/316 invited to the study participated (response rate = 64.8%). 68.0% (n=121) reported clinically significant decisional conflict regarding exercise at the time of ARVC diagnosis or GT (DCS≥25), while 55.1% (n=98) reported clinically significant decisional conflict in the year prior to study completion. Prevalence of decisional regret was also high, with 55.3% (n=99) of participants experiencing moderate to severe decisional regret (DRS≥25). Decisional conflict scores at the time of diagnosis or GT were linearly associated with SDM-Q-9 scores (ß= -.66 R^2=0.567, p<0.01). Those diagnosed at 21 or younger reported significantly more SDM (12.8±5.1, p=0.013) and less decisional conflict (-10.1±4.5, p=0.03) than those diagnosed later.

Discussion: SDM is associated with decreased decisional conflict and decisional regret and is not associated with adherence. This indicates that SDM may be the preferred model of exercise decision-making for those with ARVC and contributes to the literature suggesting that SDM is effective model of decision-making in genetic counseling.

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Part 1: Literature Review

Shared Decision-Making and Exercise Decisions in ARVC

Shared decision-making (SDM) is an increasingly popular model in medicine, including in genetic counseling. While SDM has been defined inconsistently throughout the literature, broadly there are three components to SDM: a patient' values, the various options for a decision, and the risks and benefits of these options [1-3]. SDM focuses on increasing patient involvement in medical decision-making and has been associated with lower decisional conflict and decisional regret for patients, as well as higher satisfaction with the physician facilitating the decision [4,5]. Recently, the call for SDM has extended into the space of inherited heart disease. Some clinicians are now calling for exercise decision making to follow an SDM model for patients with inherited cardiomyopathy and arrythmia syndromes [6-8]. Other clinicians refute the utility of SDM in these exercise decisions [9]. Some are particularly concerned in the case of exercise decision-making with young athletes, citing patient perceptions of SCD risk estimates as low and patients' motivation level to continue sports participation as reasons why SDM might not be a fitting model in this space [9]. While there are many opinions on the matter, there has been little work to describe what clinical support patients are receiving with regards to exercise decision making, and almost none describing the decision-making process adolescent patients are experiencing. Adolescents and young adults are often particularly active and developmentally capable of making many independent decisions, however exercise decision-making in the context of ARVC may be a particularly difficult decision for them adults to make. SDM literature has shown that, in certain cases, SDM is appropriate and desired by adolescents, while other studies show that adolescents prefer less autonomy in medical decision making [10]. However, in our review of the literature we have not come across an evaluation of SDM in

adolescent cardiac decision-making. Adolescents and young adults often have a less conservative understanding of risks than adults and are more likely to engage in risk-taking behaviors, which could impact their preferences surrounding exercise engagement.

Exercise decision making and ARVC

Patients with inherited cardiomyopathies have to weigh the risk associated with exercise against the physical, psychological, and social benefits that exercise can bring. In this realm, it is unclear if and how SDM is taking place [7,9,11,12]. Inherited cardiomyopathy diagnoses often come with recommendations to restrict exercise that vary depending on the diagnosis and the recommending clinician. In one study, the majority of surveyed pediatric cardiologists indicated that the physician should make the ultimate decision on whether patients should participate in athletics, and 11% mentioned that counseling should follow an SDM process [13]. Additionally, lack of physician's own exercise engagement was associated with increased likelihood to restrict patients from exercise, showing that physician recommendation was influenced by the physician's values and lifestyle [13]. From this information, we see that in this setting SDM may only be practiced by some clinicians, and recommendations may depend on physician characteristics.

Furthermore, it is unclear what factors impact exercise decision-making, what information is most valued by the patient, and what processes families use to weigh risks and benefits and arrive at a decision [14]. According to the literature, level of athleticism impacts decision-making post-diagnosis, as adults who considered themselves more athletic implemented fewer exercise restrictions [14]. Unsurprisingly, having experienced symptoms is associated with lower levels of activity in those with genetic cardiac conditions [15]. Little is known about

informational needs in adult or adolescent patients, except that adults are generally unaware of exercise guidelines for their disease [14]. Research on adults also highlights the evolution of these decisions, as they often change over time and are impacted by perceptions of the importance of exercise, cardiac risk, and social cues[14]. There is limited data about how these decisions are made by adolescents, young adults and their families.

Exercise decision-making may be particularly complicated for adolescents with inherited cardiomyopathies and arrythmias. Many adolescents are in a life stage during which involvement in an active lifestyle is an important part of their health, development, and social life. According to the CDC, 57.4% of high school students surveyed in the 2019 Youth Risk Behavior Surveillance survey participated on at least one sports team in the previous 12 months, not including those who participated in intramural or recreational sports [16]. Although there is debate about whether sports participation is a protective factor against adolescent risk-taking behavior, some evidence suggests that adolescents who play sports tend to participate in less cigarette usage, illegal drug use, marijuana use, shoplifting and unprotected sex [17-19]. In addition to potentially protecting against some risk-taking behaviors, adolescent sports participation and exercise involvement is associated with mental health benefits. Adolescents who participate in sports are less likely to have depressive symptoms, suicidal ideation or have attempted suicide [17,20,21]. The mental health benefits of physical activity for adolescents may be particularly important during the current COVID-19 pandemic. Recent studies have shown that adolescents that engaged in more physical activity during COVID quarantines were less likely to have depressive symptoms, while a greater number of COVID cases in an area was associated with higher levels of depressive symptoms in adolescents [22,23].

While there is limited research on the impact of exercise restriction on adolescents with inherited heart conditions, it is reasonable to hypothesize that for adolescents who depend on the physical, social, and mental health benefits of exercise, restriction is not without consequence. Qualitative work has indicated that adolescents that competed in sports at a higher level or are more restricted by their clinical providers have a more difficult time adjusting to their restrictions[24]. In addition to being in a vulnerable life stage socially, adolescents are also in a vulnerable life stage with regards to their cognitive development. Generally, adolescents perceive consequences of many risk-taking behaviors to be less negative than adults do [25]. Because adolescents often do not conceptualize risk the same way adults do, adolescents may not weigh the risk of exercise induced SCD as heavily as adults do.

While generating evidence to inform SDM would be useful for a number of genetic cardiac diseases, it is particularly salient in arrhythmogenic right ventricular cardiomyopathy (ARVC). ARVC is a genetic cardiomyopathy that increases risk of life-threatening ventricular arrhythmias, heart failure, and SCD in affected individuals. ARVC is an ideal condition in which to explore SDM surrounding exercise because patients are disproportionately athletes, it is often diagnosed in adolescence and young adulthood, and ventricular arrhythmias (VTs) are unambiguously associated with aerobic exercise. Furthermore, exercise has been associated with increased risk of developing symptoms at a younger age in individuals who are genotype-positive for ARVC while restricting exercise is protective [26,27]. Based on these associations, both affected and at-risk (G+, P-) individuals are often advised to restrict exercise engagement. ARVC treatment guidelines advocate a shared decision-making process for decisions about exercise [12].

Theoretical Framework: the Ottawa Decisional Support Framework

The Ottawa Decisional Support Framework (ODSF) provides a framework for examining the quality of clinical decision-making centered around the experience of the patient. This framework asserts that decisional outcomes are influenced by whether or not decisional needs are fulfilled by appropriate decisional supports[28]. Decisional outcomes are used to define the quality of the decision, the quality of the decision-making process, and the impact of the decision. Decisional needs include accurate information regarding potential consequences of the decision, emotional support, and clarity regarding one's own values. Decisional supports largely focus on the clinical and personal support that a patient receives when making a decision.

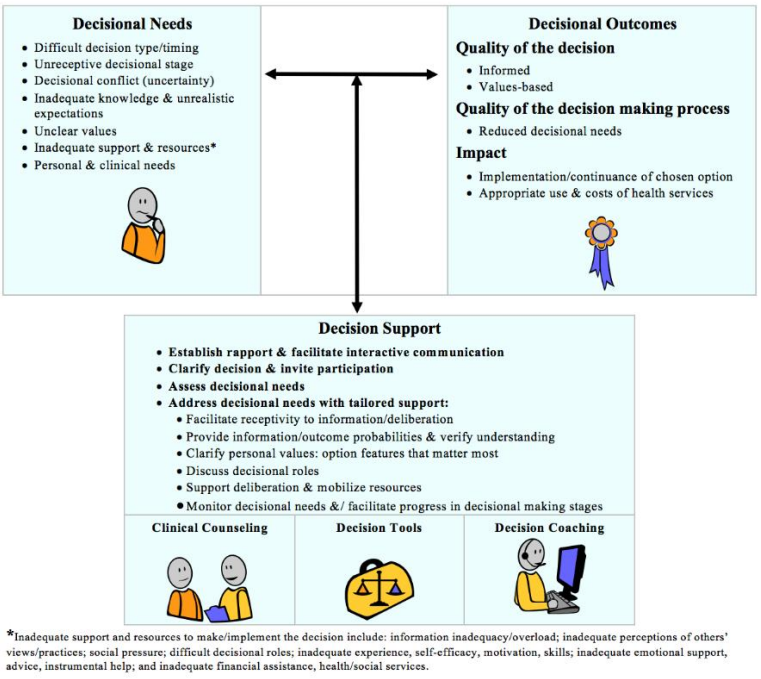

Figure 1: Diagram of the Ottawa Decisional Support Framework. Image courtesy of https://decisionaid.ohri.ca/odsf.html [29].

Decisional conflict is a concept that contributes heavily to the ODSF. Decisional conflict is defined as a state of uncertainty regarding a course of action [30]. Often, decisional conflict arises when a person is confronted with a decision that has many options, involves a high level of

risk, or incites anticipated regret [30]. Decisional conflict can be experienced both before and after a decision is made, as either a lack of decisional needs being met or believing that, for any number of reasons, the decision made was unsatisfactory. Quantitatively, decisional conflict is broken down into five subcategories: how informed a decision was, how clear the decision-maker's values were, how much support a decision-maker had throughout the process, uncertainty surrounding the decision, and how effective the decision was. In the medical decision-making literature, increased decisional conflict has been associated with delaying a decision, changing a decision, fretting or nervousness after a decision, and lower patient satisfaction with the physician [4,31,32]. Level of decisional conflict regarding exercise involvement has, to our knowledge, never been assessed in individuals with ARVC.

Decisional regret is a decisional outcome according to the ODSF. It has been used in the medical literature to describe the regret an individual might feel after making a medical decision. As medicine moves increasingly towards a less paternalistic model of care in many spaces, there is concern that patients will experience more decisional regret as they become more involved with their medical decision-making without the proper clinical supports [33]. Decisional regret has been widely explored in the medical decision-making literature, and increased decisional regret has been associated with dissatisfaction with a patient's perceived role in the decision-making process, less satisfaction with the decision, and less informed decisions (as reported by patients) [34,35]. Decisional regret has also been associated with decreased role and social functioning, higher levels of clinical depression and anxiety, and decreased quality of life [34,36]. While these are important findings to note, it is also critical to acknowledge that they are merely correlational – decisional regret has been associated with all the above outcomes but has not been shown to

cause them. To our knowledge, decisional regret has never been assessed in individuals with ARVC or any inherited heart disease regarding their exercise decision-making.

For patients with ARVC faced with exercise decision-making, decisional needs could include knowledge of empirical risk of a cardiac event, risks specifically associated with exercise, options for managing risk through changing patterns of physical activity, knowledge of risks that cannot be mitigated (role of exercise in progression), and exploration of the value of exercise to the specific individual. Decisional supports are the structures in place to satisfy decisional needs, such as clinical counseling, patient decision aids (PtDAs), and decision coaching. In this study, we will focus on SDM engagement as a decisional support. In ARVC exercise decision-making, decisional support is often provided by clinicians and increasingly by genetic counselors. Decisional outcomes will be quantified as the decision that was made, decisional regret and decisional conflict.

Part 2: Manuscript

Characterizing Decision-Making Surrounding Exercise in ARVC: analysis of decisional conflict, decisional regret, and shared decision-making

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Abstract

***Background**:* Due to concerns for exacerbating or developing symptoms, exercise restriction is recommended for those with or at-risk for Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC). Guidelines recommend shared decision-making (SDM), but there is little evidence regarding the usage or impact of SDM for exercise decision-making. Therefore, we sought to 1) describe the extent to which SDM happens in the population, 2) characterize for whom SDM happens differentially, and 3) determine if SDM is associated with decisional conflict, decisional regret, and adherence.

***Methods**:* Adults diagnosed with ARVC or positive genetic testing (GT) for ARVC-risk enrolled in the Johns Hopkins ARVC Registry were invited to complete a one-time questionnaire that included exercise history, SDM, decisional conflict, and decisional regret.

***Results**:* Of those invited to the study, 205/316 participated (response rate = 64.8%). 68.0% (n=121) reported clinically significant decisional conflict regarding exercise at the time of ARVC diagnosis or GT (DCS≥25), while 55.1% (n=98) reported clinically significant decisional conflict in the year prior to study completion. Prevalence of decisional regret was also high, with

55.3% (n=99) of participants experiencing moderate to severe decisional regret (DRS≥25). Decisional conflict scores at the time of diagnosis or GT were linearly associated with SDM-Q-9 scores (ß= -.66 R^2=0.567, p<0.01). Those diagnosed at 21 or younger reported significantly more SDM (12.8±5.1, p=0.013) and less decisional conflict (-10.1±4.5, p=0.03) than those diagnosed later.

***Discussion**:* SDM is associated with decreased decisional conflict and decisional regret and not associated with adherence. Therefore, SDM may be the preferred model of exercise decision-making for those with ARVC.

Key words: Arrhythmogenic right ventricular cardiomyopathy, genetics, exercise, shared decision-making, decisional conflict, decisional regret

Introduction

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is an inherited cardiovascular condition associated with frequent ventricular arrhythmias, cardiomyopathy, and, of particular concern, increased risk of sudden cardiac death (SCD). ARVC is most often inherited in an autosomal dominant manner with reduced penetrance. Most of the known genetic causes of ARVC are variants in genes associated with the cardiac desmosome, protein structures that link cardiomyocytes. Evidence suggest that frequent, intense aerobic exercise is associated with poorer cardiovascular outcomes, both for people with known ARVC variants (i.e. *PKP2*) and those with gene elusive ARVC[37,38]. For those at risk for ARVC, exercise is associated with increased likelihood of developing clinical disease or having sustained ventricular arrythmias (VT) [37,39]. For those diagnosed with ARVC, exercise is associated with higher arrhythmia burden, dangerous structural changes to the heart, and heart failure [40]. Consequently, although

exercise recommendations for ARVC patients differ somewhat among professional organizations, all agree that patients with definite diagnoses of ARVC should avoid most competitive sports and frequent high intensity aerobic activity[12].

Nonetheless, decisions surrounding exercise participation for ARVC patients and at-risk relatives are complex and ongoing. The ideal level of exercise for a specific patient is uncertain, may vary by genotype, and is based on an ever-evolving evidence base. Patients have to weigh the risks associated with exercise against the physical, psychological, and social benefits that exercise can bring. Because exercise exacerbates the chance of developing ARVC, many of those diagnosed are highly active individuals for whom exercise restriction may be particularly challenging [41,42].

In recognition of this complexity, guidelines recommend that exercise decisions for those with or at risk for ARVC follow a shared decision-making (SDM) model [12]. SDM is an increasingly popular model in medicine that aims to increase patient autonomy and engagement in medical decision making. While SDM has been defined inconsistently throughout the literature, broadly there are two components to SDM: clarifying patient values and exchanging information about options and their risks and benefits [1-3]. The utility of SDM in exercise decision-making for people with inherited heart conditions is disputed. Some clinicians are calling for exercise decision making to follow an SDM model for patients with inherited cardiomyopathy and arrythmia syndromes [6-8]. Still other clinicians refute the utility of SDM in these exercise decisions, with particular concern for young athletes, citing patient perceptions of SCD risk estimates as low and the motivation level to continue sports participation as reasons why SDM might not be a fitting model in this space [9]. While there are many opinions on the matter, there has been little work to describe what clinical support patients are receiving with

regards to exercise decision making, and almost none describing the decision-making process adolescent patients are experiencing. The Ottawa Decisional Support Framework (ODSF), a popular model of decision-making, postulates that unresolved decisional needs (such as uncertainty or lack of support) can lead to poor decisional outcomes (such as decisional regret, lack of adherence to the decision or sustained decisional conflict). Both decisional conflict and decisional regret have been associated with poor psychosocial and medical outcomes. Decisional regret related to medical decisions has been associated with decrease in role and social functioning, increased physical pain, lower quality of life, and increased depression and anxiety [34-36]. Decisional conflict has been associated with delaying medical decisions, lower physician satisfaction, fretting, nervousness, and increased decisional regret [31,34,43]. SDM has been associated with decreased decisional conflict and decisional regret, as well as increased adherence to decisions in some populations [44-48]. In contrast to much of the existing medical decision-making literature, exercise decision making happens throughout the lifespan, rather than at a single decision-making time or time period (such as for a surgical decision or treatment of a time-limited disease). It is uncertain whether the predicted benefits of SDM would be applicable to exercise decision-making for ARVC. Furthermore, the appropriateness of SDM application in adolescents is debated because while they are capable of making many decisions independently, there are concerns about their ability to fully comprehend risk [25]. This is of concern for adolescents with ARVC because the risks associated with ARVC are serious and potentially irreversible.

In summary, exercise decisions are difficult for those with ARVC, and SDM is recommended but there has been no study of either the extent of SDM for exercise decision-making or its consequences. Therefore, via a cross-sectional questionnaire administered to adults

in the Johns Hopkins ARVC registry, we sought to describe exercise decision-making in the ARVC population and to analyze associations between SDM and decisional outcomes. Our aims were to 1) measure the extent to which SDM for exercise in ARVC is occurring , 2) characterize which patients are most likely to engage in exercise SDM with particular focus on adolescent patients and athletes, and most importantly 3) determine if SDM is associated with decisional conflict, decisional regret, and adherence.

Methods

Design

This study utilized a cross-sectional questionnaire design.

Participants

Participants were identified from the Johns Hopkins ARVC Registry. In order to be invited to join the study, registry participants were required to be 1) 18 or older at the time of the study, 2) diagnosed with ARVC per 2010 Task Force Criteria and/or have had a positive genetic test for ARVC, 3) engaged in active registry follow-up (eg. active consent), 4) coded as alive in the registry, and 5) diagnosis or GT since 2011 [49].

Recruitment

Eligible registry participants were invited by email to complete a one-time online questionnaire about their exercise history and exercise decision-making. A $20 Amazon gift card incentive was offered for completing the questionnaire. The initial invitation was followed by two reminders sent at one-week intervals.

Measures

The questionnaire included demographics, measures of athlete identity, exercise participation, shared decision making, decisional conflict, and decisional regret. Participants were asked to reflect on two decision points: the period of time around their diagnosis (in the year before diagnosis and the 6 months after), and their current decision making (in the year prior to study completion). Participants with both an ARVC diagnosis and positive GT were asked to consider the time around when they were diagnosed. A copy of the questionnaire is available in the Supplementary Material.

Demographics and Clinical Variables

Participants were asked to report their age, gender, the education level they had achieved, their current relationship status, who was living in their house at the time they were diagnosed or had GT, who currently lives in their house, their ARVC status (whether they are diagnosed with ARVC or had positive GT only), and the age at which they were diagnosed or had GT. Participant ICD status and whether participants had experienced ventricular tachycardia (VT) or ventricular fibrillation (VF) at the time of presentation were extracted from the Johns Hopkins ARVC Registry REDCap database.

Athlete Identity

Athlete identity was captured using questions pioneered by Subas and colleagues [41]. The questions asked whether or not the participant identified as an athlete or an active individual before their diagnosis, whether or not a person identifies as an athlete or active individual currently, whether or not others viewed the participant as an athlete or active individual before their diagnosis, and whether or not others view the participant as an athlete or active individual currently.

Exercise participation/Adherence to exercise guidelines

Exercise participation was measured at three time points: in the year before diagnosis or GT, in the 6 months after diagnosis or GT, and currently (in the year prior to study completion). At each time point participants were asked to list the three physical activities they spent most of their time doing, categorize each activity as light, moderate, or vigorous intensity level, and report how much time they spent doing each activity. Definitions for light, moderate, and vigorous intensity were provided using language and definitions from the Multi-Ethnic Study of Atherosclerosis (MESA) Typical Week Physical Activity Survey [50]. Self-reported intensity levels were cross-checked with the Compendium of Physical Activities and those who reported vigorous intensity with activities that could not be vigorous were recoded to the appropriate activity level (i.e. those who reported vigorous activity with "golf" were recoded to moderate activity) [51]. In order to be considered vigorous, the Compendium of Physical Activities had to report that the activity was able to be performed at a level equivalent to 6.0 METs or higher. Time spent on each activity was captured by reporting hours per day, days per week, weeks per month, and months per year that the participant engaged in each activity. Participants were also asked if they had ever participated in competitive sports and at what level they had participated. From this data, the hours per week each participant spent at vigorous activity level pre-diagnosis or GT, at the time of diagnosis or GT, and currently was calculated. Adherence to guidelines was operationalized as not engaging in any activity that was considered vigorous intensity after diagnosis.

Shared Decision-Making Questionnaire (SDM-Q-9) [52]

The SDM-Q-9 is a validated measure that captures the extent to which shared decision-making took place, usually in a medical decision. The instrument includes 9-items, each

measured on a 6-point Likert scale. Individuals respond to statements, with answers ranging from "completely agree" to "completely disagree." The questionnaire is reliable, with a Cronbach's alpha value of 0.938. The SDM-Q-9 is scored on a scale of 0-100, with 100 being perfect shared decision making and 0 being no shared decision making. On the SDM-Q-9 participants were asked to consider their decision around the time of diagnosis or GT (in the 6 months after diagnosis or GT).

Decisional Conflict Scale (DCS) [28,53]

The DCS is a validated, 16 item measure of decisional conflict in the context of a medical decision. It has a Cronbach's alpha value that exceeds 0.78. Items are measured on a 5-point Likert scale. The scale has five subscales: the informed subscale, values clarity subscale, the support subscale, the uncertainty subscale, and the effective decision subscale. The DCS is scored on a scale of 0-100, with 0 being no decisional conflict and 100 being substantial decisional conflict. A DCS score of 25 or more is considered to be clinically significant decisional conflict. Participants were asked to reflect on their decisional conflict related to exercise at two time points: first in the 6 months after they were diagnosed or had GT, then in the year prior to study completion.

Decisional Regret Scale (DRS) [33,54]

The DRS is a 5-item scale that measures an individual's regret of a decision, most commonly a medical decision. It is measured on a 5-point Likert scale. The scale is reliable, with a Cronbach's alpha value between 0.81 and 0.92. The DRS is scored on a scale of 0-100. A score of 0 is considered no decisional regret, a score of 1-24 is considered mild regret, and a score greater than or equal to 25 is considered moderate to severe regret [55].

Statistical Analysis

Categorical variables are reported as frequency (%). Continuous variables are summarized as either mean ± SD or median (interquartile range) and compared across groups using students t-tests. The Kolmogorov–Smirnov test and skewness and kurtosis were used to evaluate normality. SPSS (version 28.0.1.1, SPSS Inc., Chicago, IL) statistical software was used for all analyses. A p-value of <0.05 was considered significant. Associations between continuous independent and dependent variables were determined using univariate linear regression. Multivariate linear regression including independent variables with $p<0.05$ in univariate analysis was conducted using the enter method. All scales were scored according to published standard procedures. If any item in a scale was missing that scale/subscale was not scored. No imputation was performed.

Results

Study Population

A total of 316 invitations were sent and 205 individuals completed the questionnaire, resulting in a response rate of 64.8%. Of the 205 responses, two were removed because they did not self-report a clinical diagnosis of ARVC or positive GT for ARVC. Eleven additional participants self-reported more than 11 years since diagnosis. These responses were cross-checked in the registry and those that had date at presentation in 2011 or more recently were included, and the remaining 9 were excluded. This left 194 responses for analysis.

The demographic and exercise history of the population are summarized in Tables 1 and 2. The average age of the population at the time of questionnaire was 43.9±15.0 years with men

and women equally represented. Of note, most of our population had clinical diagnosis of ARVC (76.7%, n=148). Consistent with this, most had an ICD at last follow-up (59.4%, n=111)), and 39.4% (n= 54) presented with sustained VT or VF.

Table 1: Demographic and Clinical Characteristics**		
	range	N (%) or mean±SD
Gender* (# female)		105 (54.1)
Age	18–82	43.9±15.0
Age at time of diagnosis or GT	10–75	38.6±15.2
Age Categories		
# diagnosed 18 or younger		20 (10.6)
# diagnosed 21 or younger		29 (15.3)
# diagnosed 25 or younger		40 (21.2)
Years since diagnosis	0–13	5.0±2.9
ARVC status (# with ARVC diagnosis)		148 (76.7)
ICD at last follow-up		111 (59.4)
VT/VF at presentation		54 (39.4)
Lived alone at time of diagnosis		14 (7.2)
Lived alone at time of questionnaire		23 (11.9)
Education level		
some high school		2 (1.0)
completed high school/GED		9 (4.6)
some college		25 (12.9)
completed college		71 (36.6)
some graduate school		15 (7.7)
completed graduate school		72 (37.1)
Relationship status		
single		20.6 (41)
married or partnered		79.4 (151)

*gender options included male, female and non-binary/third gender. No participants responded that they identified as non-binary/third gender.
VT = ventricular tachycardia
VF = ventricular fibrillation
ICD = implantable cardioverter defibrillator
GT = genetic testing

Table 2: Exercise History**		
	Range	N (%) or mean±SD
Ever participated in competitive sports		143 (77.7)
Athlete Identity		
Identified as an athlete at the time of diagnosis		134 (69.8)
Identified as an active individual at the time of diagnosis		179 (93.7)
Currently identifies as an athlete		27 (15.5)
Currently identifies as an active individual		107 (60.8)
Vigorous Activity		
Engaged in vigorous activity in the year before diagnosis		124 (63.9)
Engaged in vigorous activity in the 6 months after diagnosis		16 (8.2)
Engaged in vigorous activity in the year prior to study completion		13 (6.7)
Hours spent doing vigorous activity per week		
in the year before diagnosis	0.0–44.3	4.9±7.2
in the 6 months after diagnosis	0.0–15.4	0.5±1.9
in year prior to study completion	0.0–6.7	0.2±0.8

**Some items were left blank by some participants. Percentages reflect the proportion of those who answered the items (i.e. those who did not answer were excluded).

Exercise Decision Making

As shown in Table 2, the population was particularly athletic. More than three-quarters (77.7%, n=143) reported participating in a competitive sport at some time during their life, and 69.8% reported that they viewed themselves as athletes in the year before they were diagnosed. Additionally, 93.7% (n=179) of participants viewed themselves as active individuals in the year before they were diagnosed. Overall, participants were highly engaged in vigorous activity before diagnosis or GT. In the year before diagnosis or GT, 63.9% (n=124) of participants participated in some level of vigorous activity and participants averaged 4.9±7.2 hours per week

at vigorous intensity exercise (median=2.8, IQR=[0.0, 6.5]). Participants had overwhelmingly decreased exercise since their ARVC diagnosis or GT. Nearly all (94.6%, n=175) of participants reported that they had decreased their exercise because of their ARVC diagnosis or GT. Only one (0.5%) individual reported increased exercise since diagnosis, and 4.9% (n=9) reported that they had not changed their exercise since diagnosis or GT. After diagnosis or GT, self-reported vigorous activity level also decreased greatly. In the 6 months after their diagnosis or GT, 8.2% (n=16) of participants participated in vigorous activity. In the year prior to study completion, 6.7% (n=13) of participants participated in vigorous activity. In the 6 months after diagnosis or GT, participants averaged 0.5±1.9 hours per week or vigorous activity with a median of 0.0 and IQR [0.0, 0.0]. In the year prior to study completion, the average time spent on vigorous activities was 0.2±0.8 hours per week, again with a median of 0.0 and IQR of [0.0, 0.0].

Shared Decision-Making

The distributions of SDM scores for adults and adolescents are shown in Figure 1. The average score on the SDM-Q-9, reflecting exercise decision-making at diagnosis/GT was 59.64.8±25.0. Scores ranged from no SDM (SDM-Q-9 = 0) to perfect SDM (SDM-Q-9 = 100). Generally, participants reported high SDM on items related to exchange of information (i.e. "my provider made it clear that a decision needed to be made" or "my provider helped me understand all of the information") and lower scores on items that reflected partnering or considering participant opinion (i.e. "my provider asked me which option I prefer" or "my provider and I selected an option together"). SDM-Q-9 mean item scores are presented in Supplemental Table

2. Table 3 summarizes the association of extent of SDM regarding exercise with demographic, clinical, and exercise/athlete characteristics.

Figure 1: Histograms of SDM Scores

Fig. 1a

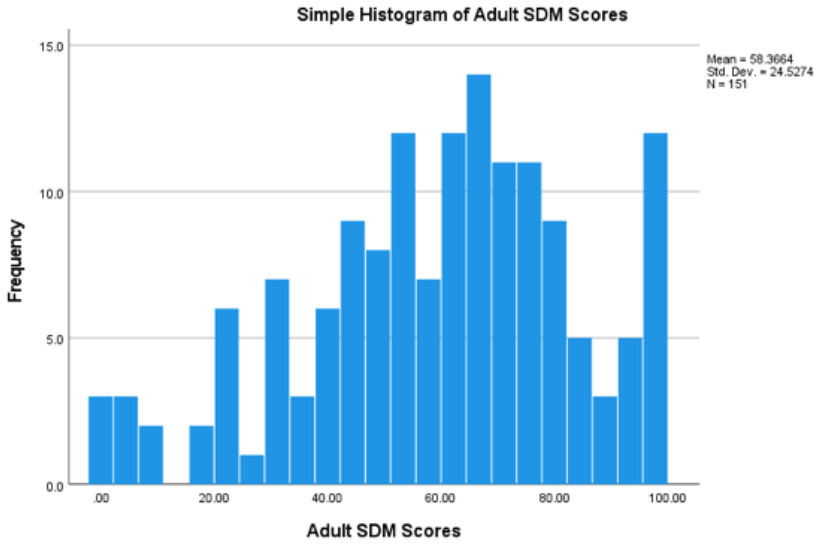


Fig. 1b

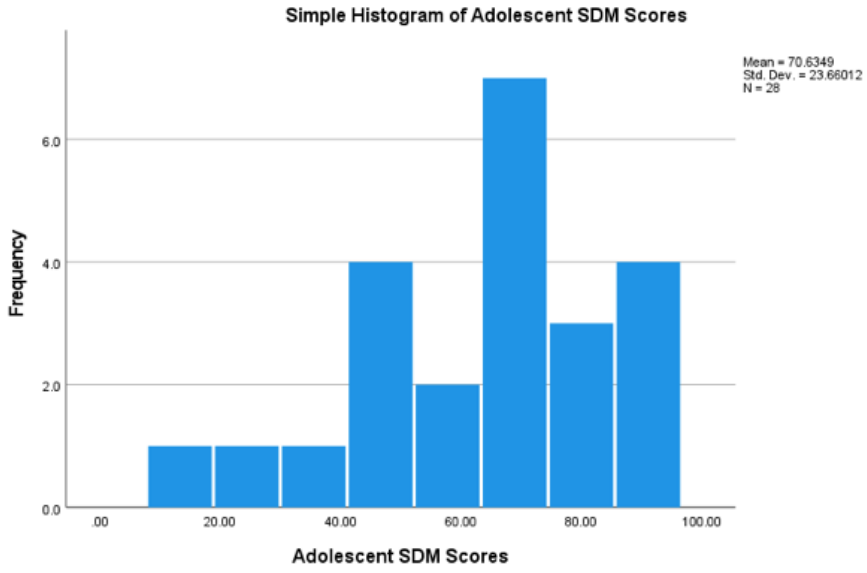


Figure 1: ***1a)*** *histogram of adult SDM-Q-9 scores (those with diagnosis/GT at 22 or later);* ***1b)*** *histogram of adolescent SDM scores (those diagnosed at 21 or earlier)*

Table 3: Summary of SDM-Q-9 scores			
	N	mean±SE or linear coefficient	p-value
Gender			
Male	85	61.2±2.5	
Female	95	58.2±2.7	0.422
Age Categories			
Diagnosed 21 or younger	28	70.6±4.5	
Diagnosed 22 or older	153	57.8±2.0	0.013
Age at diagnosis/GT	178	-0.42±0.12	<0.001
ARVC Status			
Diagnosed with ARVC	141	59.6±2.1	
Genetically at risk for ARVC	41	58.7±3.7	0.841
Exercise history			
Had played a competitive sport	142	60.4±2.2	
Had never played a competitive sport	40	56.9±3.5	0.431
Viewed self as athlete in the 6 months before diagnosis	128	61.2±2.3	
Did not view self as athlete in the 6 months before diagnosis	54	56.2±3.1	0.219
Viewed self as active in the 6 months before diagnosis	170	59.3±1.9	
Did not view self as active in the 6 months before diagnosis	11	61.4±9.5	0.789
Vigorous activity in the 6 months before diagnosis	123	60.9±2.3	
No vigorous activity in the 6 months before diagnosis	60	56.9±3.2	0.305
Clinical history			
ICD at last follow-up	104	58.8±2.4	
No ICD at last follow-up	74	60.1±3.0	0.736
Had sustained VT at presentation	53	55.8±3.5	
No sustained VT at presentation	76	64.8±2.4	0.031
Years since diagnosis/GT (linear regression)	178	0.62±0.67	0.352

SDM-Q-9 = shared decision-making questionnaire
GT = genetic testing
ICD = implantable cardioverter defibrillator
VT = ventricular tachycardia

Being younger in age at diagnosis was associated with higher levels of SDM. The association of younger age at diagnosis with more SDM was evident both when comparing SDM in adolescent (diagnosed or tested 21 or younger) vs. adult patients (diagnosed or tested 22 or older) (difference in means=-12.8±5.1, p=0.013, 95% CI=(-22.8, -2.9)) and modeling age linearly (ß=-0.42, p<0.001, 95% CI=(-0.65, 0.18)). The relationship between SDM and being diagnosed or tested during adolescence as compared to adulthood strengthened when the age category was instead defined as diagnosis or testing at 18 or younger (difference in means=-16.4±5.8, p=0.007, 95% CI=(-28.2, -4.6)). Notably, time since diagnosis was not associated with SDM.

In contrast, athletic history, participation, and identity were not associated with extent of SDM. There was a slight trend in most exercise history categories towards those who were more active or athletic reporting more SDM, but it was insignificant for every variable analyzed. Likewise, clinical and demographic variables were largely not associated with SDM. The exception to this was seen among patients who had experienced a sustained ventricular arrhythmia prior to or at the time of diagnosis. This clinical presentation was associated with significantly less SDM.

When both age at diagnosis and whether the participant presented with VT were added to a multivariate linear model, trends were maintained but neither had a significant effect on SDM ($ß_{age}$= -0.3±0.1, p= 0.06, 95% CI = (-0.6,0.01); $ß_{VT\ at\ pres}$= -8.1±4.2, p= 0.06, 95% CI=(-16.4,0.2)).

Decisional conflict (DCS) and decisional regret (DRS)

Overall, the population had significant levels of decisional conflict and decisional regret regarding exercise decision-making. Two-thirds (68.0%, n=121) of participants reported

experiencing clinically significant decisional conflict in the 6 months after diagnosis or GT. In the year before study completion, 55.1% (n=98) of participants were experiencing clinically significant decisional conflict. Similarly, 16.8% (n=30) of participants experienced no decisional regret, 27.9% (n=50) experienced mild decisional regret, and 55.3% (n=99) experienced moderate to severe decisional regret with regards to the decisions they made about exercise in the 6 months after they were diagnosed. The population levels of SDM, decisional conflict and decisional regret are summarized in Table 4.

Table 4: SDM, DCS and DRS summary

	N	mean±SD, median [IQR] or %
Shared Decision-Making *(at the time of diagnosis/GT)* (mean±SD)	183	59.6±25.0
Decisional Conflict		
In the 6 months after diagnosis/GT		
Whole scale (mean±SD)	178	34.3±22.2
Proportion with clinically significant DC (DCS≥25) (%)	121	68.0
In the year prior to study completion		
Whole scale (mean±SD)	178	27.3±20.7
Proportion with clinically significant DC (DCS≥25) (%)	98	55.1
Decisional Regret *(in the 6 months after diagnosis/GT)*		
Whole scale (median [IQR])	179	25 [10,45]
Proportion with no DR (DRS=0) (%)	30	16.8
Proportion with mild DR (0<DRS<25) (%)	50	27.9
Proportion with moderate to severe DR (DRS≥25) (%)	99	55.3

SDM = shared decision-making
DC = decisional conflict
DCS = decisional conflict scale
DR = decisional regret
DR = decisional regret scale

SDM, Decisional Conflict, and Decisional Regret

As shown in Figure 2, SDM had significant, negative linear relationships with both decisional conflict (both in the 6 months after diagnosis and currently) and decisional regret. In other words, a greater extent SDM was associated with lower levels of decisional conflict and decisional regret. SDM at the time of diagnosis or GT had the strongest association with on DCS scores in the 6 months after diagnosis or GT (Fig.2a; ß= -0.66, R^2= 0.567, p<0.001, 95% CI= (-0.75, -0.58)). The association between SDM and DCS in the year prior to study completion was weaker but maintained the same direction of the effect (Fig.2b; ß= -0.41, R^2= 0.247, p<0.001, 95% CI= (-0.49, -0.26)). SDM was significantly, yet weakly associated with DRS (Fig.2c; ß= -0.37, R^2= 0.180, p<0.001, 95% CI= (-0.52, -0.30)). DRS scores were more strongly associated with DCS scores in the 6 months after diagnosis, with higher DCS scores associated with higher DRS scores (Fig.2d; ß= 0.64, R^2= 0.397, p<0.001, 95% CI= (-0.52, -0.75)). This showed that those who had higher decisional conflict in the 6 months after they were diagnosed or tested tended to have higher decisional regret regarding the decisions they made about exercise during that time. The direction of these relationships were maintained when the data was stratified into those with diagnosis or GT at 21 or younger and those with diagnosis or GT 22 and older (see supplementary figures 1 and 2).

Figure 2: Scatterplots of SDM, DCS, and DRS

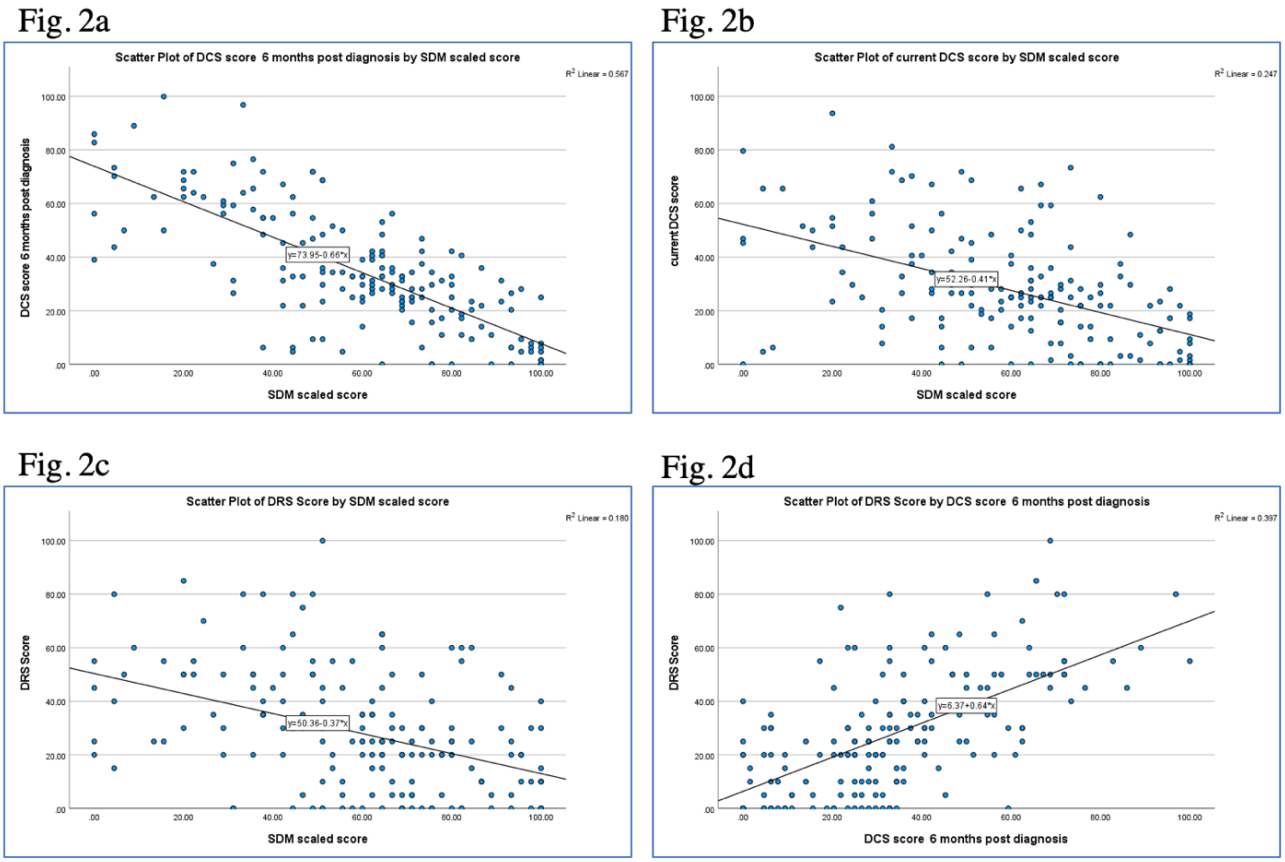


*Figure 2: **2a**: scatterplot of DCS score at 6 months after diagnosis/GT associated with SDM; **2b**: DCS score in the year prior to study completion associated with SDM score; **2c**: DRS score associated with SDM score; **2d**: DRS score associated with DCS score 6 months after diagnosis/GT*

SDM and Adherence to Exercise Guidelines

SDM did not appear to be associated with adherence to exercise guidelines by any measure. Those who engaged in any vigorous activity did not have significantly different SDM scores than those who did not participate in vigorous activity in the 6 months after diagnosis (mean difference= 3.8±6.6, p=0.564, 95% CI=(-9.2, 16.7)) or in year prior to study completion (mean difference= -6.9±7.2, p=0.340, 95% CI=(-21.1,7.3)).

Discussion

In this study we characterized decision-making for exercise among people with ARVC and at-risk relatives with the goals of evaluating the extent and implications of SDM for the decision made, decisional conflict, and decisional regret. We found that participants report a highly variable extent of SDM for exercise, with younger participants more likely to report having engaged in SDM. While participants reported decreasing exercise significantly after diagnosis, they expressed high levels of decisional conflict and decisional regret with respect to making a decision about how much to exercise. Importantly, SDM was associated with less decisional conflict and decisional regret. Adherence to exercise guidelines was high regardless of extent of SDM. Our findings therefore suggest an SDM process for exercise decision making will likely benefit ARVC patients and possibly others with or at risk for inherited heart diseases who must make choices about exercise because of disease-related recommendations.

SDM is recommended in guidelines for exercise decision making for ARVC because of its known associations with positive outcomes of decision-making, such as decreased decisional conflict and decisional regret[12]. While SDM is effective and preferable in theory, with regards to exercise decision-making for those with ARVC it is complicated because the decision is ongoing throughout the lifespan, adverse outcomes can be life-threatening, and there has been little study surrounding its efficacy and implementation. We found that SDM is happening to some extent, but with high variability. Participants reported anywhere from no SDM to perfect SDM regarding exercise. Generally, participants reported high SDM on items related to exchange of information and lower scores on items that reflected partnering or considering participant opinion. This suggests that providers may, in general, sufficiently educate their patients on the

risks and benefits of exercise with ARVC, but not specifically make space for patients to share their values and preferences or work through what might be the best decision for them.

Additionally, we found that SDM is not happening at the same level for everyone. Most demographic and clinical variables were unrelated to extent of SDM reported. However, a few variables did have significant associations with SDM. Unsurprisingly, having had sustained VT/VF at presentation was associated with significantly less SDM. While the reason for this association is uncertain, one could speculate that both the higher risk for recurrent ventricular arrhythmia and the emergent presentation could play a role. More unexpectedly, we found that those who were diagnosed in childhood, adolescence or young adulthood reported significantly more SDM than those diagnosed at older ages. While more research is necessary to determine why this is the case, there are a few possible explanations. First, it is possible that adult cardiologist practice differently than pediatric cardiologists. Furthermore, we know that provider preferences and lifestyle impact the recommendations they give [13]. Another possible explanation is that, while the SDM-Q-9 addresses specifically the decision happening between a patient and provider, participants were reflecting on their decision-making process as a whole, including others who may have been involved in the process. Children and adolescents often make medical decisions with involvement from their parents or other family members, so it is possible that they experienced more robust SDM and more support from their families that was reflected in their SDM-Q-9 scores. Notably, athletes reported similar SDM scores to non-athletes. This was surprising because those who are particularly athletic are often considered more likely to be non-adherent with exercise guidelines, therefore we hypothesized they may be less likely to be engaged in SDM [41,56].

Perhaps most impactfully, we found that higher levels of SDM were associated with lower decisional conflict and decisional regret. This is important because decisional conflict and decisional regret scores were relatively high. This is important because both decisional conflict and decisional regret have been associated with poor psychosocial and medical outcomes.

While SDM was associated with lower decisional conflict and decisional regret, it was not associated with adherence to exercise guidelines. This suggests that those who were engaged in SDM were not more likely to disregard exercise guidelines, at least in this population. This is in line with the existing literature on SDM and adherence, which has overwhelmingly linked SDM to either increased adherence or found no difference in adherence based on SDM, depending on the population [44,47,48,57-59]. This finding is significant because some clinicians refute the utility of SDM in exercise decision-making for those with inherited heart disease, arguing that it could lead to patients deciding not to comply with exercise recommendations [56]. With all of this in mind, it is clear that decisional conflict and decisional regret are significant problems in this population and that following an SDM model is associated with less decisional conflict and decisional regret without being associated with less adherence to guidelines.

Clinical Implications

While SDM for exercise decision making has been recommended for inherited heart disease, data has been unavailable on the efficacy of SDM for this complex and ongoing decision. The results of this study suggest that SDM may be the preferable model of decision making for individuals with ARVC considering exercise modifications. Importantly, this study provides evidence that indeed SDM is associated with more positive decisional outcomes for ARVC patients and at-risk relatives without being associated with less adherence to exercise

guidelines. These findings have implications for the care of ARVC families and possibly more broadly for discussions of exercise in inherited heart disease clinics. Specifically, based on our findings it, seems likely that SDM for exercise will benefit ARVC patients and families by reducing decisional regret and decisional conflict. Importantly, we saw no evidence high SDM was associated with poorer adherence to guidelines related to avoiding competitive sports or frequent vigorous aerobic exercise. It is also worth noting that multidisciplinary heart disease clinics are well-placed to engage in SDM for exercise. Cardiology providers are familiar with and capable of implementing SDM. For example, the decision to implant an ICD often follows an SDM model, and recently there has been advocacy to establish professional recommendations for SDM in ICD implantation [60-62].

Genetic counselors are important members of the healthcare team for individuals with inherited cardiovascular disease. While genetic counselors in many settings are do not make medical recommendations, guidelines in cardiology are often well established and widely accepted. Thus, in cardiology genetic counselors are often put in the unique position of facilitating decision-making in the context of clear medical guidelines, such as in the case of exercise decision-making for those with ARVC and other inherited heart conditions. This study offers evidence that genetic counselors may be able to use SDM as a tool to facilitate exercise decision-making while simultaneously providing the appropriate guidelines. Further research could focus on how genetic counselors implement SDM, decisional aids to assist patients in exercise decision-making, and further exploration of the reasons that SDM appears to be more robustly implemented for adolescents than adults.

Limitations

The cross-sectional nature of the study prevents us from establishing directionality of the relationships discussed. The population of this study was recruited through the Hopkins ARVC registry, which may not be representative of all people with ARVC. People in the registry are those who are highly motivated to be part of research initiatives and who have been referred to the study or have found the study independently. The retrospective nature of the study introduces some limitations on the ability of participants to accurately recall their experiences of exercise decision-making around the time they were diagnosed. We acknowledge that our data is a limited representation of the nuanced exercise histories of these individuals. Our population reported high exercise guideline adherence (almost no participants reported engaging in vigorous aerobic activity after diagnosis), which limited our ability to analyze the effect of SDM on adherence. Further research could focus on non-adherent individuals.

Supplemental Materials

Supplemental Material 1: Recruitment Email

Dear XXXXXXX,

You are receiving this email because you are enrolled in the Johns Hopkins ARVC Registry (*IRB NA_00041248 , Characterizing Decision-Making Surrounding Exercise, PI Hugh Calkins, MD)*. We invite you to complete a questionnaire to help us learn more about exercise history and decisions made by those impacted by ARVC.

The questionnaire will take about 20 minutes to complete. It is completely fine if you do not wish to participate in the study or choose not to answer all the questions and will by no means affect the care you receive. Your responses will not become part of your medical record.

Everyone who completes the questionnaire will receive a $20 Amazon gift card.

The first question will ask for your ARVC study ID.

Your ARVC study ID is: XXXXXXXXX

Click here for your questionnaire:

Please contact Crystal Tichnell (ctichne1@jhmi.edu) with any questions.

Thank you so much in advance for your continued partnership and helping us to understand the impact of exercise in ARVC.

Sincerely,

Crystal Tichnell, MGC, RN
Genetic Counselor / Registered Nurse
Johns Hopkins Hospital - ARVD Program
600 North Wolfe Street, Blalock 545
Baltimore, Maryland 21287

On behalf of

Jessica Sweeney
Genetic Counseling Trainee
Johns Hopkins/NHGRI

Supplemental Material 2: Questionnaire

Sweeney ScM GC Thesis

Start of Block: Introduction

Q39 Thank you for completing this questionnaire.

Questionnaire name: Characterizing Decision-Making Surrounding Exercise
Principal Investigator: Hugh Calkins, MD
IRB Number: NA_00041248

Key Information: You are being asked to complete this questionnaire as part of your participation in the Johns Hopkins ARVC Registry (NA_00041248, PI Hugh Calkins, MD). The purpose of this questionnaire is to learn how individuals with or at-risk for ARVC make decisions about exercise. We also hope to better understand your past and current participation in exercise and sports. This questionnaire will take about 20 minutes to complete. No identifying information will be collected and your answers will not become part of your medical record. Everyone who completes this questionnaire will have the option to receive a $20 Amazon gift card.

Contact Information: If you have any questions, please feel free to contact the research coordinator Crystal Tichnell at ctichne1@jhmi.edu.

Page Break

Q19 Please enter your ARVC study ID number

_____

Q1 What is your age?

_____

Skip To: End of Survey If Condition: What is your age? Is Less Than 18. Skip To: End of Survey.

End of Block: Introduction

Start of Block: Demographics

Q15 What is your gender?

- ○ Male (1)
- ○ Female (2)
- ○ Non-binary / third gender (3)
- ○ Prefer not to say (4)

Q16 What is the highest level of education that you have achieved?

- ○ Some high school (1)
- ○ Completed high school/GED (2)
- ○ Some college (3)
- ○ Completed college (4)
- ○ Some graduate school (5)
- ○ Completed graduate school (6)

Q17 What is your relationship status? (Check all that apply)

- ☐ Single (1)
- ☐ Married (2)
- ☐ Partnered (3)
- ☐ Living with a partner (4)

Q41 Who lives in your household? (Check all that apply)

- ☐ I live alone (1)
- ☐ I live with roommate(s) (2)
- ☐ I live with friend(s) (3)
- ☐ I live with my parent(s)/guardian(s) (4)
- ☐ I live with my partner (5)
- ☐ I live with my child(ren) (6)
- ☐ Other (7)

Q43 Who was living in your household **in the 6 months** after your diagnosis or being told you were at-risk for ARVC? (Check all that apply)

(If you are diagnosed with ARVC, please think about the 6 months after you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, please think about the 6

months after you were told you were at risk.)

- ☐ I was living alone (1)
- ☐ I was living with roommate(s) (2)
- ☐ I was living with friend(s) (3)
- ☐ I was living with my parent(s)/guardian(s) (4)
- ☐ I was living with my partner (5)
- ☐ I was living with my child(ren) (6)
- ☐ Other (7)

End of Block: Demographics

Start of Block: ARVC Status

Q28 This questionnaire will ask a series of questions about your exercise decision-making throughout your lifetime. Specifically, we will ask about two time points: the decision you made at the time you were diagnosed with ARVC or had a positive genetic test result, and the decision you are currently making.

Q29 Now, we will ask you about your ARVC status

Page Break

Q2 What is your status with regards to ARVC? (check all that apply)

- ☐ Diagnosed with ARVC (1)
- ☐ Positive genetic test for ARVC (2)
- ☐ Not diagnosed with ARVC and no positive genetic test for ARVC (3)

Skip To: End of Survey If What is your status with regards to ARVC? (check all that apply) = Not diagnosed with ARVC and no positive genetic test for ARVC

Display This Question:

If What is your status with regards to ARVC? (check all that apply) = Diagnosed with ARVC

Q3 At what age were you diagnosed with ARVC?

_____

Display This Question:

If What is your status with regards to ARVC? (check all that apply) != Diagnosed with ARVC

And What is your status with regards to ARVC? (check all that apply) = Positive genetic test for ARVC

Q4 At what age were you told you were at risk for developing ARVC?

_____

End of Block: ARVC Status

Start of Block: Athletic History

Q33 Here, we will ask some questions about your exercise and athletic history.

Page Break

Q5
In the year just before my diagnosis or test result..

(If you are diagnosed with ARVC, please think about the year just before you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, think about the year just before you were told you were at risk.)

	Strongly agree (1)	Agree (2)	Neither agree nor disagree (3)	Disagree (4)	Strongly disagree (5)
I viewed myself as an athlete (1)	○	○	○	○	○
I viewed myself as an active individual (2)	○	○	○	○	○
Others viewed me as an athlete (3)	○	○	○	○	○
Others viewed me as an active individual (4)	○	○	○	○	○

Page Break

Q40 Now, we would like to ask about all your leisure-time exercise. This includes exercise for conditioning/working-out, hobbies, and competitive and recreational sports.

Exercise
Think about the 3 exercise activities you spent most of your time doing **in the year before your diagnosis or learning that you were at risk of developing ARVC.** For each activity, enter information about the intensity and duration.

Light intensity are activities that require little effort and are easy to do.

Moderate intensity refers to effort that is harder than light intensity but not all out effort. These activities cause small increases in breathing or heart rate and are done for at least 10 minutes continuously.

Vigorous intensity is a very hard activity that requires close to all-out effort. Vigorous activity causes large increases in breathing or heart rate and is done for at least 10 minutes continuously unless the sport precludes this duration (eg football, sprinting).

(As a reminder, if you are diagnosed with ARVC, please think about the year just before you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, think

about the year just before you were told you were at risk.)

	Indicate light, moderate, or vigorous intensity (L=light, M=moderate, V=vigorous) (1)	How many hours a day did you spend doing that activity? (2)	How many days a week did you do this activity? (3)	How many weeks of the month did you spend doing this activity? (4)	How many months of the year did you spend doing this activity? (5)
List Activity #1 Below (1)					
List Activity #2 Below (2)					
List Activity #3 Below (3)					

Q26
Think about the 3 exercise activities you spent most of your time doing **in the 6 months after your diagnosis or learning that you were at risk of developing ARVC.** For each activity, enter information about the intensity and duration.

(As a reminder, if you are diagnosed with ARVC, please think about the 6 months just before you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, think

about the 6 months just before you were told you were at risk.)

	Indicate light, moderate, or vigorous intensity (L=light, M=moderate, V=vigorous) (1)	How many hours a day did you spend doing that activity? (2)	How many days a week did you do this activity? (3)	How many weeks of the month did you spend doing this activity? (4)	How many months of the year did you spend doing this activity? (5)
List Activity #1 Below (1)					
List Activity #2 Below (2)					
List Activity #3 Below (3)					

Q27
Think about the 3 exercise activities you have spent most of your time doing **in the past year**. For each activity, enter information about the intensity and duration.

	Indicate light, moderate, or vigorous intensity (L=light, M=moderate, V=vigorous) (1)	How many hours a day did you spend doing that activity? (2)	How many days a week did you do this activity? (3)	How many weeks of the month did you spend doing this activity? (4)	How many months of the year did you spend doing this activity? (5)
List Activity #1 Below (1)					
List Activity #2 Below (2)					
List Activity #3 Below (3)					

Page Break

Q7 Have you ever participated in competitive sports of any kind in your life?

Competitive sports are defined as an organized team or individual sport that requires systematic training and regular competition against others that places a high premium on athletic excellence and achievement. Characteristics of competitive athletics is a situation in which the athlete has a strong inclination to extend themselves to extremely high levels of exertion, often stretching their native physical limits sometimes for prolonged periods of time, regardless of other considerations.

- ○ Yes, currently (1)
- ○ Yes, previously (2)
- ○ No (3)

Display This Question:

If Have you ever participated in competitive sports of any kind in your life? Competitive sports are... != No

Q9 At what level of competition have you competed (check all that apply)?

- ☐ Grade/Middle School (1)
- ☐ Recreation League or Club (2)
- ☐ Individual Sport (i.e. road running, cycling, swimming, triathalons) (3)
- ☐ Club High School (4)
- ☐ Junior Varsity High School (5)
- ☐ Varsity High School (6)
- ☐ Club College (7)
- ☐ Junior Varsity College (8)
- ☐ Varsity College (9)
- ☐ Semi-professional (10)
- ☐ Professional (11)
- ☐ International/Olympic (12)
- ☐ Other (please specify) (13)

_____

Q23 Have you modified your exercise because of your ARVC diagnosis or ARVC genetic test result?

- ○ Yes - decreased exercise (3)
- ○ Yes - increased exercise (4)
- ○ No (5)

End of Block: Athletic History

Start of Block: Block 14

Q34 Now, we will ask about the time you were diagnosed with or told you were at risk for ARVC.

If you are diagnosed with ARVC, please think about the time you were diagnosed.

If you are not diagnosed with ARVC but are at risk for developing ARVC, think about the time you were told you were at risk.

Page Break

Q22
Approximately how long after your diagnosis or test result did a health care provider talk to you about the exercise recommendations associated with ARVC?

(As a reminder, if you are diagnosed with ARVC, please think about the time you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, please think about the time you were told you were at risk.)

- ○ A healthcare provider has never talked to me about exercise recommendations (1)
- ○ Immediately after my diagnosis or test result (2)
- ○ Within 6 months of my diagnosis or test result (3)
- ○ Within 1 year of my diagnosis or test result (4)
- ○ More than 1 year after my diagnosis or test result (5)

Q24
Approximately how long after your diagnosis or test result did you modify your exercise?

(As a reminder, if you are diagnosed with ARVC, please think about the time you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, please think about the time you were told you were at risk.)

- ○ I have not modified my exercise (1)
- ○ Immediately after my diagnosis or test result (2)
- ○ Within 6 months of my diagnosis or test result (3)
- ○ Within 1 year of my diagnosis or test result (4)
- ○ More than 1 year after my diagnosis or test result (5)

End of Block: Block 14

Start of Block: SDM-Q-9

Q14
Think about your decision regarding exercise **in the 6 months after** your ARVC diagnosis or being told you were at risk.
Please select how much you agree with each of the following statements.

(As a reminder, if you are diagnosed with ARVC, please think about the time you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, think

about the time you were told you were at risk.)

	Completely agree (1)	Strongly agree (2)	Somewhat agree (3)	Somewhat disagree (4)	Strongly disagree (5)	Completely disagree (6)
My health care team made it clear that a decision needed to be made about my physical activity level (1)	○	○	○	○	○	○
My health care team wanted to know exactly how I wanted to be involved in making the decision about my physical activity level (2)	○	○	○	○	○	○
My health care team told me that there are different exercise options for treating my medical condition (3)	○	○	○	○	○	○
My health care team precisely explained the advantages and disadvantages of my exercise options (4)	○	○	○	○	○	○

My health care team helped me understand all the information (5)	○	○	○	○	○	○
My health care team asked me which exercise option I prefer (6)	○	○	○	○	○	○
My health care team and I thoroughly weighed the different exercise options (7)	○	○	○	○	○	○
My health care team and I selected a exercise option together (8)	○	○	○	○	○	○
My health care team and I reached an agreement on how to proceed (9)	○	○	○	○	○	○

End of Block: SDM-Q-9

Start of Block: Decisional Conflict Scale - retrospective

Q12
Think about the decision you made about exercise **in the 6 months after** you were diagnosed with ARVC or told you were at risk for developing ARVC. Please show how you feel about these statements on a scale of 1 (strongly agree) to 5 (strongly disagree).

(If you are diagnosed with ARVC, please think about the 6 months just after you were diagnosed.

If you are not diagnosed with ARVC but are at risk for developing ARVC, think about the 6 months just after you were told you were at risk.)

	Strongly agree (1)	Agree (2)	Neither agree nor disagree (3)	Disagree (4)	Strongly disagree (5)
I knew which exercise options were available to me. (1)	○	○	○	○	○
I knew what the benefits of each exercise option were. (2)	○	○	○	○	○
I knew what the risks of each exercise option were. (3)	○	○	○	○	○
I was clear about which benefits mattered most to me. (4)	○	○	○	○	○
I was clear about which risks mattered most to me. (5)	○	○	○	○	○
I was clear about which was more important to me (the benefits or the risks). (6)	○	○	○	○	○
I had enough support from others to make a choice. (7)	○	○	○	○	○
I chose without pressure from others. (8)	○	○	○	○	○

I had enough advice to make a choice. (9)	○	○	○	○	○
I was clear about the best choice for me. (10)	○	○	○	○	○
I felt sure about what to choose. (11)	○	○	○	○	○
The decision was easy for me to make. (12)	○	○	○	○	○
I felt that I had made an informed choice. (13)	○	○	○	○	○
My decision showed what was important to me. (14)	○	○	○	○	○
I expected to stick with my decision. (15)	○	○	○	○	○
I was satisfied with my decision. (16)	○	○	○	○	○

End of Block: Decisional Conflict Scale - retrospective

Start of Block: Decisional Regret Scale

Q11
Now, we are going to ask you about how you feel **looking back on the decision you made about exercise in the 6 months after** you were diagnosed with ARVC or told you were at risk for ARVC.

Think about the decision you made about exercise **in the 6 months after** you were diagnosed with ARVC or told you were at risk for ARVC. Please show how you feel about these statements on a scale of 1 (strongly agree) to 5 (strongly disagree).

(As a reminder, if you are diagnosed with ARVC, please think about the 6 months after you were diagnosed. If you are not diagnosed with ARVC but are at risk for developing ARVC, think about the 6 months after you were told you were at risk.)

	Strongly agree (1)	Agree (2)	Neither agree nor disagree (3)	Disagree (4)	Strongly disagree (5)
It was the right decision (1)	○	○	○	○	○
I regret the choice that was made (2)	○	○	○	○	○
I would go for the same choice if I had to do it over again (3)	○	○	○	○	○
The choice did me a lot of harm (4)	○	○	○	○	○
The decision was a wise one (5)	○	○	○	○	○

End of Block: Decisional Regret Scale

Start of Block: Block 15

Q35 Now, we will ask about your current exercise decision making.

End of Block: Block 15

Start of Block: Decisional Conflict Scale - Current

Q13
Think about the decision you are currently making about exercise participation, specifically **in the past year**. Please show how you feel about these statements on a scale of 1 (strongly agree) to 5 (strongly disagree).

(If you were diagnosed or told you were at-risk in the past year, please think about the time since you were diagnosed or told you were at-risk.)

	Strongly agree (1)	Agree (2)	Neither agree nor disagree (3)	Disagree (4)	Strongly disagree (5)
I know which exercise options are available to me. (1)	○	○	○	○	○
I know what the benefits of each exercise option are. (2)	○	○	○	○	○
I know what the risks of each exercise option are. (3)	○	○	○	○	○
I am clear about which benefits matter most to me. (4)	○	○	○	○	○
I am clear about which risks matter most to me. (5)	○	○	○	○	○
I am clear about which is more important to me (the benefits or the risks). (6)	○	○	○	○	○
I have enough support from others to make a choice. (7)	○	○	○	○	○
I am choosing without pressure from others. (8)	○	○	○	○	○
I have enough advice to make a choice. (9)	○	○	○	○	○

I am clear about the best choice for me. (10)	○	○	○	○	○
I feel sure about what to choose. (11)	○	○	○	○	○
The decision is easy for me to make. (12)	○	○	○	○	○
I feel that I have made an informed choice. (13)	○	○	○	○	○
My decision shows what is important to me. (14)	○	○	○	○	○
I expect to stick with my decision. (15)	○	○	○	○	○
I am satisfied with my decision. (16)	○	○	○	○	○

End of Block: Decisional Conflict Scale - Current

Start of Block: Current Identity as an athlete

Q25
Please indicate how much you agree with each of the following statements.

Currently...

	Strongly agree (1)	Agree (2)	Neither agree nor disagree (3)	Disagree (4)	Strongly disagree (5)
I view myself as an athlete (1)	○	○	○	○	○
I view myself as an active individual (2)	○	○	○	○	○
Others view me as an athlete (3)	○	○	○	○	○
Others view me as an active individual (4)	○	○	○	○	○

End of Block: Current Identity as an athlete

Start of Block: Block 15

Q37 Are you interested in receiving a $20 Amazon gift card?

○ No (1)

○ Yes (2)

Q38 If yes, please enter the email you would like the gift card to be sent to.

_____

End of Block: Block 15

Supplemental Table 1: DCS Subscales

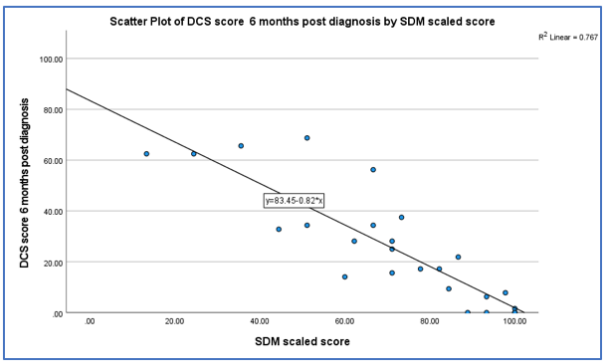
Supplemental Table 1: DCS Subscales		
	N	mean±SD
DCS in the 6 months after diagnosis/GT		
Informed subscale	180	34.0±26.4
Values clarity subscale	179	32.0±26.0
Support subscale	182	31.6±23.1
Uncertainty subscale	179	42.3±26.6
Effective decision subscale	180	31.9±22.8
DCS in the year prior to study completion		
Informed subscale	177	27.9±24.2
Values clarity subscale	175	25.8±21.7
Support subscale	178	27.8±20.9
Uncertainty subscale	178	37.4±24.1
Effective decision subscale	178	27.2±20.3

Supplemental Table 2: SDM Item Average Scores

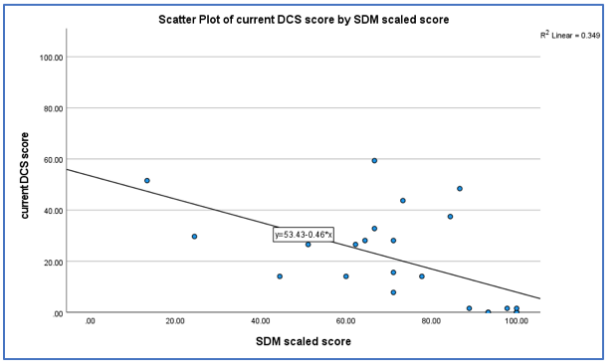
Supplemental Table 2: SDM-Q-9 Average Item Scores		
Item	N	mean±SD
My health care team made it clear that a decision needed to be made	184	4.1±1.3
My health care team wanted to know exactly how I wanted to be involved in making the decision	185	3.1±1.5
My health care team told me that there are different options for treating my medical condition	182	2.9±1.5
My health care team precisely explained the advantages and disadvantages of my options	184	3.3±1.5
My health care team helped me understand all the information	183	3.5±1.5
My health care team asked me which treatment option I prefer	185	2.6±1.5
My health care team and I thoroughly weighed the different options	185	2.6±1.6
My health care team and I selected a treatment option together	185	2.2±1.5
My health care team and I reached an agreement on how to proceed	185	2.7±1.5

Supplementary Figure 1: Scatterplots of SDM, DCS, and DRS: diagnosis or GT at 21 or younger

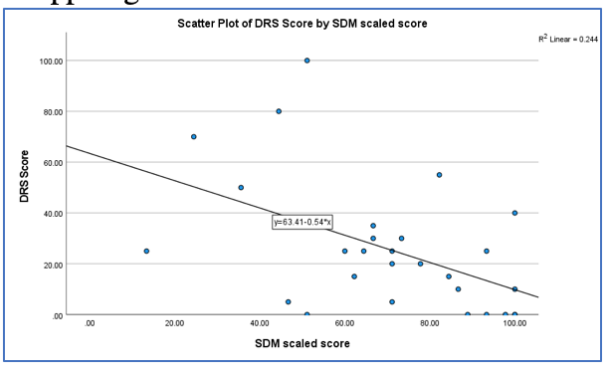
Supp. Fig. 1a



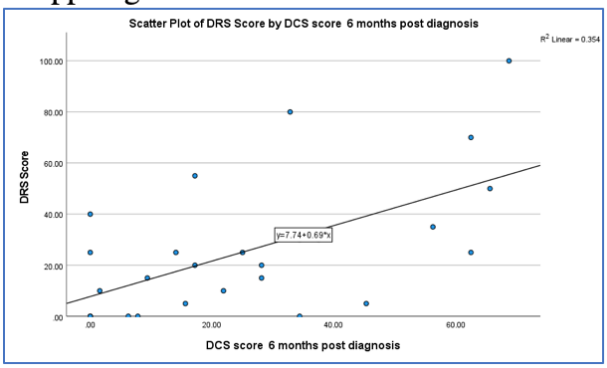
Supp. Fig. 1b



Supp. Fig. 1c



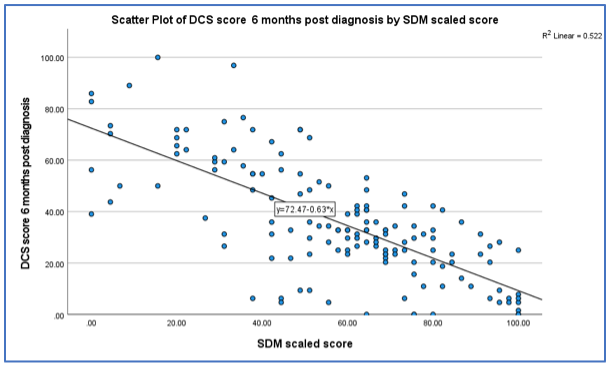
Supp. Fig. 1d



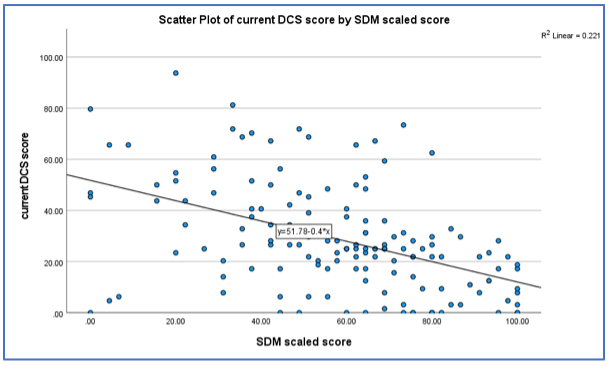
Supplementary Figure 1: SDM, DCS and DRS relationships for only those with diagnosis or GT at 21 years old or younger. ***1a****: scatterplot of DCS score at 6 months after diagnosis/GT associated with SDM;* ***1b****: DCS score in the year prior to study completion associated with SDM score;* ***1c****: DRS score associated with SDM score;* ***1d****: DRS score associated with DCS score 6 months after diagnosis/GT*

Supplementary Figure 2: Scatterplots of SDM, DCS, and DRS: diagnosis or GT at 22 or older

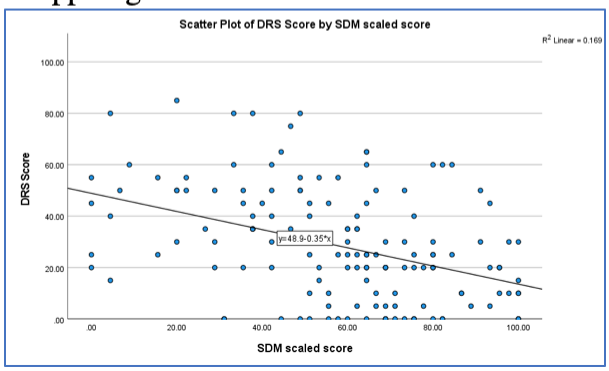
Supp. Fig. 2a



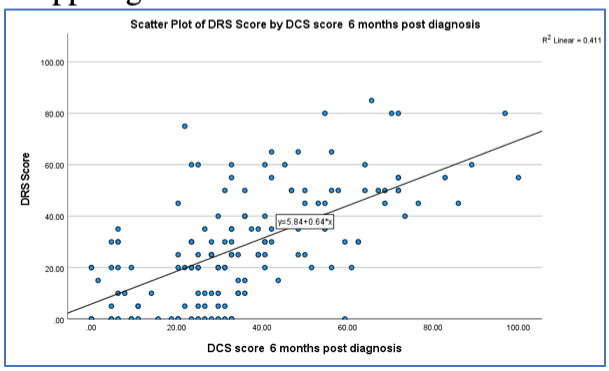
Supp. Fig. 2b



Supp. Fig. 2c



Supp. Fig. 2d



Supplementary Figure 2: SDM, DCS and DRS relationships for only those with diagnosis or GT at 22 years old or older. ***2a****: scatterplot of DCS score at 6 months after diagnosis/GT associated with SDM;* ***2b****: DCS score in the year prior to study completion associated with SDM score;* ***2c****: DRS score associated with SDM score;* ***2d****: DRS score associated with DCS score 6 months after diagnosis/GT*

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