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REVIEW

How Patient Perceptions Shape Responses and Outcomes in Inherited Cardiac Conditions

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At least one-third of adults living with an inherited cardiac condition report clinically-significant levels of psychological distress. Poorer health-related quality of life compared with population norms is also consistently reported. These outcomes are associated with younger patient age, having an implantable cardioverter defibrillator, and receipt of uncertain clinical test results, and can influence self-management behaviours, such as adherence to potentially critical life-preserving medications. According to the Common Sense Model of Illness, people use information from multiple sources to 'make sense' of their health condition, and how they conceptualise the condition can strongly influence adaptation and coping responses. Previous studies with people with inherited cardiac conditions show that illness perceptions, such as greater perceived consequences and a poorer understanding of the condition, are associated with greater psychological distress and poorer adherence to medication. The Common Sense Model provides one potential framework for identifying patients who may be more vulnerable to adverse health outcomes, and for developing early interventions to reduce the physical and psychosocial burden of these conditions. Interventions based on the Common Sense Model have successfully improved physical and psychosocial outcomes associated with other cardiac conditions, and could be tailored for use with patients with an inherited cardiac condition (ICC).

Keywords Inherited cardiac conditions • Anxiety • Depression • Health-related quality of life • Illness perceptions • Common Sense Model of Illness

Introduction

In many countries, including Australia and New Zealand [1], there has been a concerted effort over the past 20 years to detect and subsequently manage patients with inherited cardiac conditions, such as hypertrophic cardiomyopathy (HCM), Long QT Syndrome (LQTS) and arrhythmogenic right ventricular cardiomyopathy (ARVC). While attention has focussed on reducing patient morbidity and mortality [2], few studies have investigated the psychological and

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social implications of living with these conditions. Here we explore the ways in which adults with an inherited cardiac condition 'make sense' of their condition, and how this may influence physical and psychosocial outcomes. We discuss a framework called the Common Sense Model of Illness, which could be used to inform the development of interventions to reduce the psychosocial burden associated with inherited cardiac conditions.

According to the Common Sense Model of Illness [3], patients' perceptions of their illness, (i.e. how they 'make sense' of their health condition), can influence coping behaviours, such as avoidance or engagement with clinical management. This, in turn, is associated with illness outcomes, including psychological well-being, health-related quality of life, and key disease markers, such as low-density lipoprotein cholesterol control in patients with hypercholesterolaemia [4]. We propose this framework as one way for clinicians and researchers to explore patients' perceptions and experiences of their condition, and as a potential model for better understanding how we can influence longer-term health and well-being after diagnosis of an inherited cardiac condition. We will focus on evidence relating to adult patients only, given the known and complex influence of developmental stage and parental illness perceptions on the ways in which children conceptualise illness [5,6]. There is evidence that children with an inherited cardiac condition (ICC) report poorer health-related quality of life than healthy peers [7], and the emotional adjustment of children with congenital heart disease has been found to be influenced by maternal perceptions of the illness [8,9]. Thus, an in-depth review of illness perceptions in paediatric populations with an inherited cardiac condition would be a valuable addition to the literature.

Psychological Distress During Initial Investigations for Inherited Cardiac Conditions

During initial clinical and genetic investigations, many people will experience acute distress [10]. The intensity and duration of this distress can vary, at least in part, due to the circumstances that led to the cardiac investigation [10,11]. For example, patients referred following an incidental finding during a routine medical exam may experience lower distress than those referred after the sudden death of an immediate family member [12]. Irrespective of the reason for referral, however, it is not uncommon for individuals attending a genetic cardiac clinic to experience confusion, worry, fear, frustration and grief.

Generally, when family members are asymptomatic and are found not to carry the family at-risk gene, this acute distress response dissipates relatively quickly after receipt of test results and few long-term psychological consequences have been reported in the literature [10,13]. This may not be the case for asymptomatic non-carriers who have lost an immediate family member to sudden cardiac death, many of whom experience ongoing grief and psychological distress [14]. Outcome data for asymptomatic gene positive relatives typically indicate relatively low levels of psychological distress; however, comparisons between studies is difficult due to variation in cardiac populations, and in the timing and methods by which psychological outcomes are measured. Asymptomatic relatives at-risk of HCM report elevated worry and disease-related distress during follow-up appointments [15], and disease-related distress has been found to remain high for over 12 months when clinical test results are uncertain [10]. This distress is understandable given the possibility they may develop the condition in the future, and will be reminded of this by the need for continued clinical follow-up.

Anxiety, Depression and Health-Related Quality of Life in People Living With an Inherited Cardiac Condition

For symptomatic adults living with an inherited cardiac condition, various psychosocial outcomes have been reported (Table 1). Overall, more than one-third of patients report levels of psychological distress indicative of a need for clinical intervention, and a sizeable proportion report reduced health-related quality of life compared with population norms [16–20]. Table 2 outlines a range of individual and environmental factors that have been shown to be associated with psychological distress in adults with an inherited cardiac condition. For instance, studies suggest in LQTS, males appear to be more vulnerable to depression [21], and, in HCM, females appear to be at greater risk of anxiety [22].

Despite the high levels of psychological distress reported by a sizeable subset of adults with an inherited cardiac condition, many also adjust and cope well with the ongoing symptoms and self-management demands (Table 3) [15]. One Australian study of HCM patients found those who were satisfied with their understanding of their condition tended to report better adjustment and less health-related worry [15]. Other protective factors identified in general illness literature include greater perceived social support, optimism, problem-focussed coping, and positive general self-efficacy (e.g. one's belief in their ability to complete a task) [23].

How Psychosocial Factors May Influence Disease-Related Outcomes

Across diverse health conditions, psychological distress is consistently associated with a wide variety of adverse clinical outcomes, including difficulties with treatment

Table 1 Summary of psychological distress and health-related quality of life outcomes in adults with an inherited cardiac condition.

Reference	Cardiac Illness	Country	Sample	Findings	Measure
Morgan, O'Donoghue, McKenna and Schmidt (2008) [16]	HCM	United Kingdom	N = 115 27% had a familial HCM, 33% a family hx sudden death; ≥18 years; Mean age 43 years (SD = 10); Mean time since diagnosis 6 years (range 1-10)	50% of participants met criteria for anxiety and 31% met criteria for depression. 37% of participants met criteria for an anxiety disorder, 21% met criteria for a mood disorder.	HADS SCI
Hintsa, Järvinen, Puttonen, Ravaja, Toivonen, Kontula and Swan (2009) [42]	LQTS	Finland	N = 1267; 595 (47%) were carriers of a LQTS mutation (of which 261 were symptomatic and 334 were asymptomatic) and 672 (53%) were family members who were non-carriers of a LQTS mutation; 16 – 65 years; Mean age 42 years (SD = 13); Mean time since entering the registry 6 years	Symptomatic participants were 1.4 times more likely to report depressive symptoms than asymptomatic patients. Depressive symptoms were not related to LQTS gene mutation, but were significantly associated with the experience of clinical symptoms (both arrhythmic events in the LQTS mutation carrier group and syncope events in the non-carriers).	BDI (revised)
Christiaans, van Langen, Birnie, Bonsel, Wilde and Smets (2009) [22]	HCM	The Netherlands	N = 258; Group 1 = mutation carriers with manifest HCM (135, 52%), Group 2 = predictively tested relatives with manifest HCM detected after DNA testing (34, 13%), Group 3 = predictively tested relatives still without manifest HCM detected (89, 35%); Mean age 49 years (SD = 15); Age range 16 – 86 years; Mean time since genetic testing 3 years (SD = 1)	Group 1 reported greater symptoms of depression compared to groups 2 and 3 and group 2 reported greater levels of anxiety than group 3. Overall Group 1 reported poorer physical quality of life compared to groups 2 and 3 and group 2 reported poorer physical quality of life compared to group 3.	HADS SF-36
Hamang, Eide, Rokne, Nordin and Øyen (2011) [12]	HCM & LQTS	Norway	N = 126; 32 (25%) had a clinical diagnosis (12 had LQTS and 20 had HCM) and 94 (75%) were at genetic risk due to a family history (76 due to LQTS and 18 HCM;	From the total sample 25% and 14% scored above the threshold on the HADS for anxiety and depression respectively. There was no difference in anxiety and depression scores between patients with a clinical	HADS

Table 1 (continued).

Reference	Cardiac Illness	Country	Sample	Findings	Measure
			Mean age 45 years (SD = 16);	diagnosis and those family members at genetic risk, or between disease types (LQTS compared to HCM). Participants with an HCM diagnosis reported poorer physical functioning, and greater avoidance due to heart-focussed anxiety compared to participants with a LQTS diagnosis.	SF-36 CAQ
James, Tichnell, Murray, Daly, Sears and Calkins (2012) [43]	ARVC with an ICD	America	N = 86; All participants had a clinical diagnosis of ARVC; ICD indicated for secondary prevention in 46 (54%) participants, primary prevention in 39 (45%); 39 (45%) had not experienced an ICD shock; Mean time since ICD implanted 5 years (SD = 5); Mean age 46 years (SD = 13); Age range 18 – 79 years	31% of participants met criteria for anxiety and 9% met criteria for depression.	HADS
Ingles, Yeates, Hunt, McGaughran, Scuffham, Atherton and Semsarian (2013) [17]	LQTS, HCM, FDC, ARVC, CPVT	Australia	N = 409; 251 (62%) were affected individuals with a diagnosis of an inherited cardiac condition (208 had HCM, 43 LQTS, 28 FDC, 17 ARVC and 8 had CPVT) and 158 first-degree at-risk relatives who were clinically unaffected but not previously genotyped; ≥15 years; Mean age 49 years (SD = 16)	Participants with LQTS reported poorer mental functioning than at-risk relatives. Participants with HCM, FDC and CPVT reported poorer physical functioning than at-risk family members.	SF-36
Ingles, Sarina, Kasparian and Semsarian (2013) [44]	HCM, ARVC, FDC, LVNC, LQTS, CPVT, Brugada Syndrome	Australia	N = 90; Participants had a clinical diagnosis of an inherited cardiac condition and had received an implantable cardioverter defibrillator (>12 months); ≥15 years; Mean age 49 years (SD = 14) Mean time since implant was 5 years	38% reported elevated anxiety, 17% reported elevated depressive symptoms. 31% reported symptoms indicative of post-traumatic stress response.	HADS IES-R

Table 1 (continued).

Reference	Cardiac Illness	Country	Sample	Findings	Measure
Ingles, Spinks, Yeates, McGeechan, Kasparian and Semsarian (2016) [14]	Sudden cardiac death due to HCM, ARVC, LQTS and LVNC	Australia	N = 103; Participants were family members of a young sudden death victim; 10% were clinically affected; 19 deaths were caused by HCM, 16 by ARVC, 9 by LQTS and 1 by LVNC; Mean age of 44 years (SD = 16) Mean time since death was 5 years (range 0.5 – 10 years)	20% reported symptoms of prolonged grief. 44% reported post-traumatic stress symptoms indicative of a need for clinical intervention. Family members reported higher depression, anxiety, and stress scores compared to population norms.	PGDS IES-R DASS – 21
Richardson, Spinks, Davis, Turner, Atherton, McGaughran, Semsarian and Ingles (2018) [45]	CPVT	Australia	N = 36 53% were people with a clinical diagnosis, 28% parents of an affected child under 18 years, 19% were at-risk family members; ≥18 years; Mean age 48 years (SD = 16) Mean time since diagnosis was 9 years	26% and 16% of participants reported symptoms of anxiety and depression indicative of a need for clinical intervention, respectively. 18% reported symptoms indicative of a post-traumatic stress response.	HADS IES-R

Abbreviations: HCM, hypertrophic cardiomyopathy; CPVT, catecholaminergic polymorphic ventricular tachycardia; LQTS, long QT syndrome; FDC, familial dilated cardiomyopathy; ARVC, arrhythmogenic right ventricular cardiomyopathy; LVNC, left ventricular noncompaction; BDI, Beck Depression Inventory; CAQ, Cardiac Anxiety Questionnaire; DASS, Depression Anxiety and Stress Scales; HADS, Hospital Anxiety and Depression Scale; IES-R, Impact of Events Scale – Revised; PGDS, Prolonged Grief Disorder Scale; SCI, Structure Clinical Interview; SF-36, Medical Outcomes Study Short Form Health Survey; SD, standard deviation.

decision-making, medication use and cooperation with medical recommendations, suboptimal health service use, and higher morbidity and mortality [24–28]. Increased stress and reduced happiness have been identified as risk factors in the preceding 24 hours of a cardiac event for LQTS patients [29], and lower mental wellbeing predicts non-adherence to medication in HCM patients [30].

Illness-related behaviours, such as medication adherence, are likely to be influenced by numerous and complex factors; however, studies show psychological variables, such as anxiety, low self-efficacy (particularly related to one's perceived ability to take medication as prescribed) and doubts about the necessity of medication, are associated with medication non-adherence in New Zealand and Australian inherited cardiac condition patients [30,31]. Psychological variables, such as anxiety and self-efficacy, represent potentially modifiable targets for interventions to improve patient health and wellbeing.

A Model for Thinking About the Role of Health-Related Beliefs and Perceptions — The Common Sense Model of Illness

Many successful psychosocial interventions have focussed on illness 'representations', defined in the Common Sense Model of Illness [3] (Figure 1), as how patients 'make sense of' their health condition. According to this model, individuals are not passive observers in medical consultations; rather, they are constantly seeking, receiving and processing illness-related information from multiple sources. While information provided by health care teams is one important source, personal experience of symptoms, past illness experiences, information from family and friends, and information from media and social media may also shape the ways in which an individual conceptualises his or her health and illness.

Table 2 Risk factors for psychological distress and lower health-related quality of life in adults with an inherited cardiac condition.

Risk Factor	Condition	Psychological Distress or Health-Related Quality of Life	Reference
Sociodemographic			
Women	HCM	Lower physical functioning	[17]
	HCM	Greater anxiety symptoms	[22]
Men	LQTS	Greater depressive symptoms	[21]
Younger age	CPVT	Greater anxiety, depression and post-traumatic stress symptoms	[45]
	ARVC with an ICD	Greater device-related distress and body image concerns	[43]
Lower (area level) socioeconomic and education and occupation Indexes	HCM	Poor psychological wellbeing and health-related quality of life	[30]
No follow-up or follow-up in non-specialist cardiac genetic service	HCM	Poorer adjustment to living with HCM and greater worry	[15]
Clinical			
Resuscitated cardiac arrest	Sudden cardiac arrest patients	Greater general and cardiac-specific anxiety symptoms	[46]
ICD implanted	ICC & non-ICC patients (includes ischaemic heart disease)	Greater anxiety, depression and post-traumatic stress symptoms	[44,47]
Uncertain clinical diagnosis	LQTS	Greater disease-related anxiety and depression	[10]
Daily medication regimen	LQTS	Lower health-related quality of life	[38]
SCD of young family member	HCM, ARVC, LQTS & LVNC	Greater post-traumatic stress and prolonged grief symptoms	[14]
Psychological			
Greater perceived uncertainty	Coronary artery disease & cardiomyopathy	Poorer health-related quality of life	[48]
Greater perceived risk of sudden death and greater perceived symptom severity	HCM	Greater chance of being diagnosed with a mood or anxiety disorder	[16]
Greater perceived consequences of carriership	HCM	Greater generalised anxiety and poorer physical quality of life	[22]
Poor ICD device adjustment	ARVC with an ICD	Greater generalised anxiety and depression	[43]
Social			
Poorer social and sexual adjustment	HCM	Greater chance of being diagnosed with a mood or anxiety disorder	[16]

Abbreviations: HCM, hypertrophic cardiomyopathy; ARVC, arrhythmogenic right ventricular cardiomyopathy; ICD, implantable cardioverter defibrillator; LQTS, long QT syndrome; LVNC, left ventricular noncompaction; ICC, inherited cardiac condition.

According to the Common Sense Model of Illness, information is processed along two parallel pathways; one pathway takes into account the role of cognitive processing and the other depicts emotional processing (Figure 1). These two pathways are hypothesised to interact to form a person's mental model (or conceptualisation) of illness, which consciously or unconsciously guides health-related actions and coping responses. The model proposes a continuous feedback loop, in which coping responses are appraised in light of effects on illness-related outcomes and this can lead to adjustments in perceptions and coping behaviours over time. This process is also thought to be influenced by an individual's personal and socio-cultural contexts. Within the cognitive pathway, patients' perceptions of illness are said to cluster around six dimensions, including ideas about disease causes, consequences, timeline, controllability, coherence (how well an individual understands their illness), and identity (symptoms and labels) (Table 4 and Figure 2) [32].

Within the emotion processing pathway, emotional aspects of illness are taken into account and can strongly influence how patients conceptualise their health condition, and how they cope and adjust, including physical,

Table 3 Tasks patients with an inherited cardiac condition may need to manage.

Regular and correct medication use
Lifelong medical surveillance
Distress tolerance and stress management skills
Informed consent and shared decision-making
Symptom management strategies
Invasive procedures (e.g. ICD implantation)
Identification of symptoms warranting clinical investigation
Lifestyle modification, including potential exercise and activity restrictions
Adjustment to changed social and economic circumstances
Communication with physicians, family members (including children or grandchildren, and potential partners), and other caregivers
Grief and bereavement
Survivor guilt
Transmission guilt

SOURCE: Adapted from Marks and Allegrante, 2005 [49].
Abbreviation: ICD, implantable cardioverter defibrillator.

psychosocial and occupational functioning (Figure 1) [33]. Fear, for example, can motivate some people to be vigilant with self-management behaviours, such as attending recommended medical appointments and adhering to prescribed medications. If fear becomes overwhelming, however, some people may cope by becoming hypervigilant regarding disease signs and symptoms, potentially leading to overuse of medical resources. For others, fear may generate attempts to avoid all reminders of the illness, resulting in non-attendance at recommended appointments or cessation of prescribed medications against medical advice.

Coping strategies, such as avoidance, cognitive reappraisal and emotion venting (defined in Figure 1), can directly influence illness outcomes in diverse ways, depending on the way and context in which they are used [4]. For patients with a condition that affords little personal control, such as Huntington's disease, psychological distress may be reduced by avoiding reminders of the condition. Conversely, in the context of conditions where significant personal control is possible, such as diabetes, avoidance of checking blood sugar levels would likely lead to adverse outcomes. Hagger and colleagues suggest psychological interventions should not only focus on assisting patients to develop healthy illness perceptions, but should also bolster adaptive coping strategies [4]: assisting inherited cardiac condition patients, for example, to understand how beta blocker medication works and how important it is in minimising the possibility of sudden cardiac death, then supporting patients to develop practical strategies to remember to take their medication and adjust to any side effects.

Risk perceptions also influence how patients make sense of their condition and may be particularly important in inherited cardiac conditions because of the sudden and dangerous nature of symptoms. Realising one is at increased risk of sudden cardiac death may elicit fear and distress prompting a need for heightened professional and social support [34]. Importantly, a recent study suggests patients' perceptions of risk for dangerous symptoms often do not align with those of the physician [35], suggesting an area in which psychological and social support could provide benefit.

'Making Sense' of an Inherited Cardiac Condition

According to the Common Sense Model of Illness, information about an illness is often processed in the context of pre-existing knowledge and experiences of other illnesses. Inherited cardiac conditions tend to challenge many widely-held assumptions about disease; for example, that illnesses have a distinct symptom profile, with a largely predictable timeframe, where risk increases with age, and treatment can cure or manage the illness [36]. In contrast, inherited cardiac conditions, particularly the cardiac ion channelopathies, are typically diagnosed in young and otherwise healthy people, with few if any symptoms, and treatment is about mitigating the risk of cardiac arrest which, for many, is an abstract concept. In addition, beta blockers can leave some patients feeling worse by inducing fatigue and, in some patients (such as males with LQTS type 1), risk may decrease later in life [37]. Without a pre-existing schema to support adaptive illness perceptions, it is unsurprising that many patients report confusion about symptoms, risks and medication side-effects [38]. For people with a positive genotype and no detected phenotype, the terms 'at-risk relative' or 'asymptomatic carrier' may be experienced as new and perplexing. Being informed of the risks associated with carriership in the absence of clinical signs of the condition is not a typical illness model people can readily make sense of. If we consider perceptions of illness identity as central to patients' conceptualisation of their illness it is easy to see that patients with a genetic diagnosis but no clinical signs of disease may struggle to comprehend and adapt to their condition. Research to better understand how patients perceive various aspects of inherited cardiac conditions is much-needed.

Psychological Interventions Targeting Illness Perceptions

Numerous psychological interventions have demonstrated effectiveness in improving illness perceptions in other medical conditions [39–41]. One successful trial randomised myocardial infarction patients along with their spouses to usual care or usual care plus an illness perception

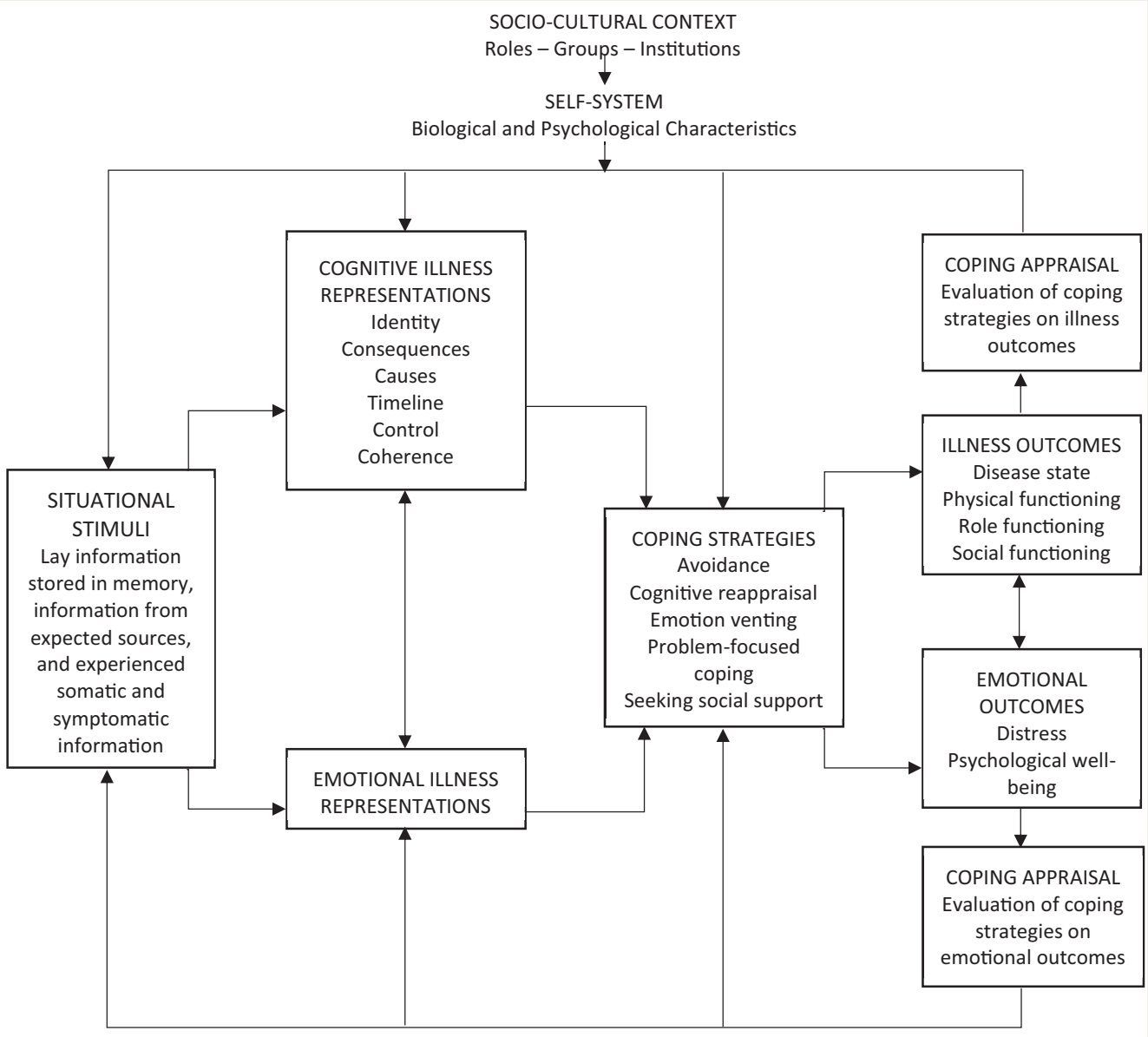

Figure 1 Schematic representation of the Common Sense Model of Illness Representations based on Leventhal et al.'s [54] original illustration. The coping strategies and illness outcomes dimensions are based on work by Hagger and Orbell [51]. Coping strategies include *avoidance:* keeping away from things that act as reminders of the illness; *cognitive reappraisal:* re-interpreting the meaning of something to mitigate the emotional response; *emotion venting:* sharing or expressing feelings with another person; *problem-focussed coping:* actively working towards managing or resolving the source of the stress; seeking *social support:* actively pursuing others who can care for you or assist with the management of stress. Illness outcomes include *disease state:* markers of disease stability or progression, e.g. cholesterol control in hypercholesterolaemia; *physical functioning:* the ability to undertake everyday tasks; *role functioning:* the ability to execute existing positions in one's life (e.g. at work or within the family); *social functioning:* the ability to interact easily with other people or in social situations.

intervention. The intervention consisted of four 30-minute, face-to-face sessions with a psychologist prior to hospital discharge [40]. Illness perceptions (e.g., consequences, treatment control, and coherence) were assessed at baseline (prior to intervention participation) and again at discharge, and 3 and 6 months post-hospitalisation. During sessions, patients were supported to explore their perceptions of myocardial infarction, challenge inaccurate beliefs and attributions, develop strategies to enhance their sense of self-efficacy in managing their condition, and develop an action plan during recovery. Patients in the intervention group, on average, reported greater engagement in physical activity and exercise, fewer contacts with their general practitioner (primary care physician), lower anxiety about returning to work, and better rates of return to work compared to those who received usual care only.

Table 4 Cognitive representations of Illness from the Common Sense Model of Illness.

Illness Perception Dimension	Description	Example	Examples of questions to ask patients to elicit illness perceptions [50]
Identity	Name (or label) assigned to a disease, and the signs and symptoms an individual associates with the condition. Perceived identity is hypothesised to be at the centre of the way an individual conceptualises his or her illness [32], and is not necessarily based on medical models.	Some patients who report many symptoms and attribute these symptoms to their illness, are more likely to deny their illness and may avoid engaging with important self-management behaviours, such as attending medical appointments [51].	*We know patients can often find it difficult to work out whether symptoms are due to their heart condition or treatment side effects. Can you tell me about the symptoms you experience that you think might be due to your heart condition?*
Cause	Beliefs about what causes the disease (e.g., genetics, stress, pollution, lifestyle choices). Causal attributions can be multifactorial, and may include both individual and/or environmental factors. How a person interprets the cause(s) of his or her condition can strongly influence coping responses.	Patients who attributed their myocardial infarction to a lack of exercise were more likely to exercise post-discharge than those who did not attribute their myocardial infarction to a lack of exercise [40].	*It's natural to wonder what caused your heart condition. Do you have any thoughts or theories about what has caused your condition?*
Timeline	Beliefs about whether the disease is acute, chronic or cyclical in nature. Timeline beliefs can vary from acute (e.g. a common cold) to chronic (e.g. diabetes) or cyclical (e.g. migraines), and can influence self-management behaviours, such as adherence to medication or medical advice.	Timeline beliefs have been shown to predict whether asthma patients take their medication daily or only when they feel they need to [52].	*How long do you think you will have your heart condition for?*
Consequences	Beliefs about the consequences the disease has had (or may have) on a person's life. Beliefs may centre on consequences for personal experiences, relationships, financial hardship, mental health, social roles, occupational opportunities, as well as other areas important to the person.	A young adult male with LQTS and an ICD may feel his social functioning has been limited because he is unable to continue playing competitive football with his friends. A meta-analysis (using many different illness groups) found perceived consequences are one of the strongest predictors of depression, anxiety and health-related quality of life [53].	*We know having a heart condition can impact many aspects of a person's life. What have been the consequences of this condition for you?*
Controllability/ Cure	The cure/control dimension includes perceptions of both personal control (i.e., how much a person believes he or she can manage the illness), and treatment control (i.e., the extent to which medication or other forms of treatment can control the illness).	Patients with an inherited cardiac condition who believed beta blockers did not control their condition well, were less likely to adhere to this medication [31].	*We have prescribed you a medication for your heart condition. How effective do you feel this medicine has been in managing your heart condition?* *How much control do you feel you have personally over your heart condition?*

Table 4 (continued).

Illness Perception Dimension	Description	Example	Examples of questions to ask patients to elicit illness perceptions [50]
Coherence	Coherence relates to how well an individual believes they understand their illness.	People who believe they understand their illness well are less likely to experience psychological distress and are more likely to be able to function physically and in their roles at home and work than those who do not believe they have a good understanding of their illness [4].	*These conditions are complex and can be tricky to wrap your head around, how well do you feel you understand your heart condition?* NB: To check their understanding is accurate, ask them to tell you their understanding of their condition.

Abbreviations: LQTS, long QT syndrome; ICD, implantable cardioverter defibrillator.

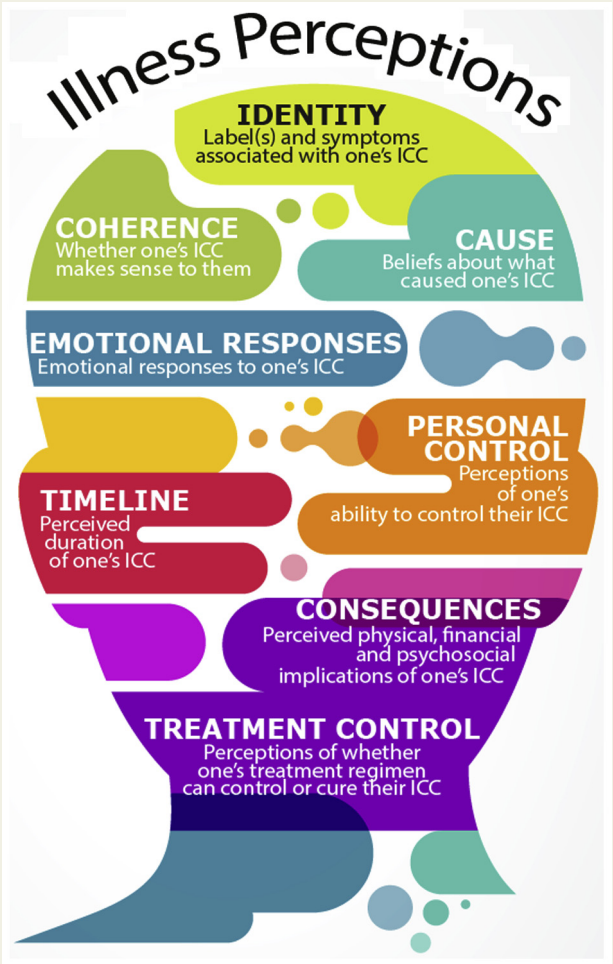

Figure 2 Summary of the illness perceptions from the Common Sense Model of Illness.
Abbreviation: ICC, inherited cardiac condition.

While, to our knowledge, there have been no such interventions developed for people with an inherited cardiac condition, it seems likely that similar interventions would be beneficial in this population given evidence that inherited cardiac condition patients' illness perceptions are associated with adherence [31], risk perceptions [35], psychological distress and health-related quality of life [22]. Qualitative studies [38] have also identified several targets for future interventions, including personal control (e.g., increasing patients' capacity to tolerate uncertainty and enhance self-efficacy with medication use), treatment control (e.g., increasing patients' perceptions of the necessity and efficacy of medication), consequences (e.g., supporting management of treatment side effects and adjustment to physical and role limitations), and emotional responses (e.g., assisting patients in processing significant or traumatic events and making difficult health-related decisions).

Conclusion

A substantial subset of people living with an inherited cardiac condition report high levels of anxiety and depression, and lower health-related quality of life compared with population norms. Current knowledge suggests patients who are younger, are from lower socio-economic groups, and perceive themselves at greater risk of sudden death may be more vulnerable to difficulties adjusting to their health condition. In addition, those with worse perceptions about the consequences of their inherited cardiac condition report greater psychological distress, and patients with a poorer understanding of their condition and less confidence that their treatment is effective are at higher risk of non-adherence. To support patients who are struggling to adhere to their medication, for example, you could use the Common Sense Model of Illness framework and elicit patient perceptions about treatment efficacy, explore their general

understanding of their condition, and their emotional responses to it and this may help to highlight certain beliefs that could be driving the non-adherence (Table 4). Further research is needed to consolidate risk and resiliency factors across different demographic groups and conditions, and to develop interventions to bolster healthy adaptation within this patient population. Research to date suggests that the Common Sense Model of Illness could be a useful framework for such interventions.

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