


Embodying a New Meaning of Being At Risk: Living With an Implantable Cardioverter Defibrillator for Arrhythmogenic Right Ventricular Cardiomyopathy

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Abstract

Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D) is a genetic condition that can cause fatal arrhythmias. The implantable cardioverter defibrillation (ICD) is a primary treatment for ARVC/D. Using a grounded theory approach, this study examines the experiences of 15 individuals living with an ICD. The ability to cope with and adjust to having an ICD is influenced by the acceptance of the ICD as something needed to survive, an understanding of the ICD's function, existing support networks, and ones' ability to manage everyday challenges. Coping well requires reshaping ideas about the meaning of being at risk and understanding how the ICD fits into that changing personal risk narrative. A thorough understanding of the unique needs of individuals with ARVC/D and of the specific factors contributing to the psychosocial distress related to having an ICD (vs. having the disease itself) is needed. Nurses must be prepared to provide ongoing support and education to this population.

Keywords

constant comparison, genetics, grounded theory, nursing, research, qualitative

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Nurses play a critical role in providing care to individuals living in families with genetically linked conditions, particularly in regions with limited access to genetic services. Nurses must be knowledgeable about these disorders, the available resources, treatment options, and the unique needs facing this population. Little is known about the needs of individuals and families living with genetically linked conditions in rural areas. Newfoundland and Labrador (NL) is a small province on the east coast of Canada that has a large rural population, is genetically isolated, and has an abundance of genetic conditions (Rahman et al., 2003), including Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D). The incidence of ARVC/D in NL is between 1/500 and 1/1,000, which is estimated to be one of the highest in the world (Etchegary, Pullman, Simmonds, Young, & Hodgkinson, 2015).

ARVC/D is an autosomal dominant genetically linked condition caused by a missense mutation in the transmembrane protein TMEM43 located on chromosome 3p25 (Merner et al., 2008). In ARVC/D, the myocardial is replaced with a fatty fibrous tissue eliciting lethal ventricular arrhythmias that can result in a sudden cardiac death (SCD) and/or

biventricular heart disease (Romero, Mejia-Lopez, Manrique, & Lucariello, 2013). The implantable cardioverter defibrillator (ICD) is the primary treatment for ARVC/D. The ICD is a small device inserted under the chest wall, which in the event of a cardiac arrhythmia delivers an electrical shock to the heart to restore it to a normal sinus rhythm.

The ICD has been found to reduce mortality in patients at high risk of ventricular arrhythmias and SCD. Research in NL looking at the efficacy of the ICD in preventing SCD in ARVC/D has shown that the 5-year mortality post ICD implant was 0 (Hodgkinson et al., 2005). For the ARVC/D population, the ICD is life-saving; without an ICD, the first symptom of heart failure might be death. Several studies have examined the experience of living with ARVC/D in NL (Etchegary et al., 2016; Etchegary et al., 2015; Manuel &

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Brunger, 2014, 2015); however, findings have not specifically reported on what life is like to have an ICD. For nurses to provide care that is tailored to the needs of this population, they must have a fuller understanding of how these individuals adjust to and cope with having an ICD within the context of a genetic condition such as ARVC/D.

Despite a large body of research on the experience of living with an ICD, the psychosocial implications of having the device are still not clearly understood for the general population of ICD recipients (Manzoni et al., 2015; Sears, Matchett, & Conti, 2009) and even less for the ARVC/D population. There is no consensus as to the factors that precipitate psychosocial distress, the onset and duration of psychosocial distress, what coping mechanisms help reduce stress, or what interventions can improve patient health care outcomes. The lack of clarity is compounded by the fact that most research on the ICD has focused on the relationship between anticipating or receiving a shock, correlates of poor device acceptance, and psychosocial distress (Borne, Varosy, & Masoudi, 2013; Manzoni et al., 2015; Pedersen, Van Den Broek, Van Den Berg, & Theuns, 2010; Sears & Kirian, 2010; Thylen, Dekker, Jaarsma, Stromberg, & Moser, 2014). Specifically, research has shown clearly that anticipating or having a shock contributes to poor health care outcomes such as depression, anxiety, or a decrease in quality of life (Bostwick & Sola, 2011; Cesarino, Beccaria, Aroni, Rodriques, & Pacheco, 2011; Ingles, Sarina, Kasparian, & Semsarian, 2013; Jacq et al., 2009; James et al., 2012; Palacios-Cena et al., 2011) in comparison with those who have not received a shock (Suwanpasak & Boonyapisit, 2014). Research has also shown that poor health outcomes are proportional to the number of ICD shocks (Wheeler et al., 2009). However, the correlation between living with an ICD, having a shock, and quality of life is not as straightforward as once thought, warranting more research (Pedersen et al., 2010). That is, the psychosocial implications of living with an ICD beyond the distress related to anticipating or having a shock appear to be far more significant for determining quality of life with the ICD than any distress associated specifically with it firing.

Research suggests that the relationships between having an ICD and health care outcomes are contingent upon the time lapse since the firing of the ICD and the measurement of quality of life being used (Pedersen et al., 2010). Some argue that neither shocks nor years since implantation are associated with device acceptance, suggesting that psychological factors other than those related to it firing might be more indicative of device acceptance (Luyster, Hughes, Waechter, & Josephson, 2006; Pedersen et al., 2011; Versteeg et al., 2012). In fact, one study indicated that having an ICD shock only explained 3% to 5% of the variance in distress levels pre to post ICD implant (Pedersen, Hoogwegt, Jordaens, & Theuns, 2013). Furthermore, ICD recipients do not experience the same responses to having an ICD. For example, in one study 20% of those with an ICD reported increasing signs of depression and anxiety; however, having a shock

was related to depression and not anxiety (Luyster et al., 2006). Others have also noted that receiving an ICD shock is not consistently linked to poor patient outcomes (Borne et al., 2013; Pedersen et al., 2011) such as depression and anxiety (Wheeler et al., 2009). While the number of shocks does shape the experience of having an ICD, this factor has a minor impact on psychosocial wellness compared with other factors, including being older, concerns about the ICD, depression, having a Type D personality, or living with symptomatic heart failure (Pedersen et al., 2013). Similarly, psychological morbidity and disease severity—not the ICD implant—were found in another study to be linked to poor acceptance of the device (Webster et al., 2014). Research has also shown that those having an ICD at a young age reported difficulty coping with the device (James et al., 2012), indicating that early screening for psychological factors that can lead to poor acceptance of the ICD is imperative (Lang et al., 2013).

Evidence is not clear as to when ICD-related distress peaks, its duration, and how it changes over time (Magyar-Russell et al., 2011). For example, those who have the ICD for prolonged periods were found to experience higher rates of depression and anxiety (Luyster et al., 2006). Others suggest that within a year, post ICD insertion, most ICD recipients do adjust to having the ICD (Kapa et al., 2010; Palacios-Cena et al., 2011), particularly in the case of those who engaged in psychoeducational interventions (Cinar, Tosun, & Kose, 2013; Sears et al., 2007). Research also proposes that the immediate post ICD implant period is when people experience the highest level of stress and anxiety (Kapa et al., 2010).

The psychosocial impact of having an ICD beyond that associated with the firing of the ICD has captured the attention of some researchers. Studies have shown that individuals with ICDs can experience alterations in body image (Spindler, Johansen, Andersen, Mortensen, & Pedersen, 2009; Versteeg et al., 2012); restrictions on physical or sexual activities (Berg et al., 2013; Probst et al., 2011); social isolation and anxiety, particularly in relation to losses such as one's driver's license, employment, and insurance (Probst et al., 2011; Zayac & Finch, 2009); stressors related to potential ICD complications (Probst et al., 2011); and an overwhelming feeling of losing control (Morken, Severinsson, & Karlsen, 2010).

To address the challenges of living with an ICD, recipients avail of a variety of coping strategies. These include taking a positive outlook on life, drawing on social support, having faith in the ICD capabilities, and balancing activities of daily living (Flemme, Hallberg, Johansson, & Stromberg, 2011). For some, the firing of the ICD is a constant reminder of mortality and vulnerability that sparks them to engage in lifestyle changes in an effort to reduce its firing. For others, having the ICD was found to instill a sense of reassurance that if they did have a cardiac event the ICD would save them (Probst et al., 2011).

Despite the large body of literature that has examined the psychosocial implications of living with the ICD, limited research has examined this experience within the context of ARVC/D. Given the uniqueness of this particular patient population, whereby death is a certainty in the absence of an ICD implant, research on the experiences of living with an ICD would provide valuable insights to assist nurses and other health care professionals caring for this population. The large numbers of individuals in NL living with ARVC/D and an ICD provide an opportunity to examine the experiences of this unique patient population. This article specifically explores the experiences of individuals who have ARVC/D and are living with an ICD in NL.

Method

Design

This article reports on one aspect of a qualitative study that examined the experiences of 29 individuals living in families at risk of ARVC/D (Manuel, 2013). A grounded theory (Glaser & Strauss, 1967) approach guided that research study, as outlined in previous publications (Manuel & Brunger, 2014, 2015). In grounded theory, qualitative data are collected to gain a fuller understanding of individuals' lived experiences. Drawing on tenants of symbolic interactionism, participants' interactions with the natural world, the meaning attributed to events such as the firing of the ICD, and the symbols that they use to convey meanings are all examined (Mead, 1934).

The substantive theory, constructing the meaning of being at risk, captured participants' experiences throughout the genetic testing process. The three theoretical constructs emerging from the data were as follows: (a) awakening to a new meaning of being at risk, (b) deciphering the meaning of being at risk, and (c) embodying a new meaning of being at risk. This article reports on one aspect of construct (c): how individuals adjusted to and coped with having the ICD.

Sample

A subset of 15 ARVC/D individuals ($n = 8$ men; $n = 7$ females) living with an ICD in the province of Newfoundland and Labrador were interviewed about their experiences of ARVC/D, including their experiences of living with an ICD. Theoretical sampling guided the enrollment of participants (Glaser, 1978). Of these 15 individuals, 13 lived in rural NL and two in urban NL. Participants were recruited by a clinical geneticist at the NL Provincial Genetics Program and snowball sampling. The geneticist made initial contact with potential participants, and informed them of the study and its purpose; those who were interested gave permission to have their contact names released to the research team. The mean age of participants was 41. There were two young adolescents, aged 15 and 16. Participants had been living with an

ICD between 1 and 7 years. One participant had previously had an ICD but because of disease progression subsequently had received a heart transplant.

Data Collection and Analysis

Participants participated in audio-recorded semi-structured individual interviews and/or a focus group lasting between 45 and 90 minutes. Interviews were transcribed into written text, and any personal information was removed to ensure confidentiality. Ethics approval was obtained from the NL Health Research Ethics Board and the Research Proposals Approval Committee of the Eastern Regional Health Authority of NL. Informed written consent was acquired prior to initiating the interviews and focus groups. Throughout the research process, data analysis and collection took place simultaneously to facilitate an understanding of participants' experiences of living with an ICD. Using a constant comparative approach, theoretical coding of the data, memoing, and diagramming, the theoretical constructs emerged from the participants' narratives. Breaks in data collection allotted time to reflect on and compare isolated incidents within and between narratives, and to code for and develop concepts, properties, and categories. NVivo software was used to manage the data. Rigor was addressed by having two researchers with qualitative expertise review the data to ensure that there was an emergent fit between the data, the categories, and constructs. Interpretive summaries were given to participants to confirm that their stories were captured accurately. An audit trail and memos were used to ensure transparency of the research process, to identify any potential bias, and to recognize variations in the data that warranted additional examination.

Findings

Participants' ability to cope with and adjust to having the ICD was influenced by their beliefs about the efficacy and management of the ICD, including (a) accepting that the ICD is needed to survive, (b) ICD function: Anticipating and understanding why the ICD fired, (c) drawing on social support, and (d) living with everyday challenges.

Accepting That the ICD Is Needed to Survive

The majority of participants wanted to have the ICD for one main reason—it was considered to be their only real chance for survival. That is, despite the fact that many of them engaged in a healthy lifestyle, they appreciated that they could not prevent the onset of what could be a fatal cardiac arrhythmia. They spoke of ICD as being an “insurance policy” or a “lifesaver” that regenerated a sense of confidence in their ability to stay “alive and safe” if they had a cardiac event. This aura of feeling safe was reinforced by their lived experiences of the efficacy of ICD to prevent what would

otherwise be imminent death. Hence, participants advocated having the ICD implanted sooner rather than later: "I wanted to have an ICD because of the sudden deaths in the family. If there was a chance that the defibrillator was going to give me another shot at life I wanted it as soon as possible." For one participant, the fatal nature of ARVC/D provided the impetus behind having an ICD: "I have got no choice but to deal with it (having ICD). I'd rather not have it, but I have to stay alive."

Faith in the ICD capabilities, and that they had made the right decision to have it inserted, was reinforced each time the ICD fired and saved a life: "I woke up in the morning and my heart was racing. The ICD gave me a shock and brought me back to life." Adding to this sense of confidence in the ICD was the feeling that even if one were to engage in a "risky" behavior such as physical exercise that could invoke a cardiac event, the ICD would deliver a shock that would reset the heart back into a normal heart rhythm, as this participant recalled,

Another time the ICD fired was when I was helping my husband bring in some wood. I was only out there five minutes, bending over picking up one chunk of wood at a time and throwing it in the wheel barrel, and the defibrillator fired.

This strong sense of faith in the technical capabilities of ICD prompted some women to approach health care providers to have an ICD implanted as soon as possible, as this woman stated,

If there was a chance that the defibrillator was going to give me another shot at life I wanted it. So, I fought to get the defibrillator because I didn't really have any symptoms other than the inverted T waves.

Despite the positives associated with living with an ICD, for some, the initial experience of having the device was not a pleasant one or an easy decision. Several participants spoke of the absence of physical symptoms as a key factor that led them to question the need to have an ICD; however, these impressions were typically short lived, lasting only until the first firing of the ICD. For the majority of participants, the fact that they were ARVC/D positive was enough to warrant treatment, regardless of whether they had had symptoms of ARVC/D. Those who were symptomatic welcomed the news of a treatment.

For many participants, it was the realization that they needed the ICD to manage ARVC/D, coupled with the fear of it firing, which caused the most distress. Some felt that having the ICD took away their once "carefree" attitude and replaced it with a continuous state of anxiety that affected the formerly taken-for-granted aspects of their everyday lives: "When I take my shower . . . I don't want to have water going full force because I want to be able to get away from the water (if the ICD fires)." For others, being offered a

defibrillator meant that their risk of having a cardiac event was escalating; they were getting sicker, they could no longer be managed with medication, and their life expectancy was reduced: "Having the ICD has changed the way I see my future because I think I am going to die young . . . ten-years from now I will be gone, anything more than that is a bonus." Adding to participants' stress was knowing that there was a good chance that the ICD, at some point, would deliver an electric shock, but the "when" remained unknown.

ICD Function: Anticipating and Understanding Why the ICD Fired

All participants shared a common sense of dread in anticipation of the first ICD shock: "When I had my ICD put in I was fine. After I got home I got nervous about having the ICD." For the nine participants whose ICD had already fired, the first incident was the most memorable. It left them with such feelings of worry that it made them fear the outcomes of subsequent shocks as well as when the next shock would occur and what it would be like: "Nobody realizes what you go through when you have these shocks . . . I was scared to death" and "That's what scares me: having another shock." For one person, the anticipation of having a shock caused panic attacks: "I started to have panic attacks, at least one a night there for a while."

Participants reported that gradually the apprehension associated with the ICD firing subsides, as they become accustomed to its firing and recognize that this is to be expected given their condition. Living with an ICD eventually becomes an accepted part of life, as described in this comment:

To me, after I had the first shock all the rest of them did not seem to be so bad. It is probably because you are used to it after it happens. The last one was when I was on the deck of the boat and I knelt down by a fellow. He asked me, "What happened?" I replied, "The defibrillator just went off," and I just got up and went on with my work.

Over time, participants begin to compile a kind of etiology of firing. For some, the firing of the ICD more than twice warranted treatment: "If the ICD fires twice in a row you have to go to the doctor and get checked out, to see if there is anything wrong with it." For others, it was the events surrounding the firing of the ICD that were significant: "If you are not passed out it is probably a problem with the machine; if you are passed out and it happens twice, it is probably something with your heart." As participants became more familiar with the workings of the ICD, they started to assign their own meanings to the shock, to the number of shocks requiring medical attention, and to their risk:

The first shock, we used to panic and go right on to town. Then it kind of mellowed. Now it's, we'll call the cardiologist in the

morning, and whenever he can get us in. So we've kind of eased off.

Revised interpretations of the ICD shocks come after trips to the hospital post ICD firing, which often result in no medical intervention. Participants reported that they soon realized that the ICD fires because it is doing what it is designed to do—to prevent an arrhythmia. In this way, the experience of living with the constant threat of having the ICD fire any time becomes a part of everyday life: “I was afraid at first (of the ICD firing) but now I just let it slide by.”

Participants emphasized the time and energy that goes into preventing firings of the ICD. They explained that this entails an understanding of the function of the ICD and significance of each shock: Was the shock inappropriate (the result of device malfunction), or was it appropriate (fired in response to a cardiac event) and, if an appropriate firing, what were the precipitating factors? For participants, successful disease management was measured by their effectiveness at identifying modifiable factor(s) that triggered the ICD to fire and their ability to abstain from the “risky” activity. The need to engage in self-surveillance and in the self-regulation of lifestyle factors to prevent the ICD from firing was emotionally draining, as it required participants to make a decision as to whether there was a correlation between the ICD firing and their behavior, and whether the incident warranted a trip to the hospital.

For many participants, successful identification of the factors that had caused the ICD to discharge was described as bringing relief, particularly when the precipitating factor was something that could be easily modified, and thus give them some sense of control over the disease. On the downside, knowing that there are factors that could be controlled also led some participants to experience an enormous amount of anguish, as they spent a lot of time and effort monitoring themselves or others for modifiable factors that might cause the ICD to discharge. Participants frequently juxtaposed the factors thought to have triggered the ICD to fire to create some frame of reference as to which activities to avoid, as captured in this participant's narrative:

The first time I triggered the ICD I was running with my dog. If I had not been running with my dog, it would never have happened. I know exercise can trigger the ICD to go off. The second time I set the ICD off was 5 months later. I triggered it through fatigue, and I think fatigue is well documented in the literature as causing heart problems. I could have prevented the first shock; and if I had been more careful with sleep, I wouldn't have had the second one.

Some participants reported that despite modifying the factors identified as “risky,” the ICD continued to fire. This was discouraging and added to participants' escalating sense of being at risk. It also caused substantial anxiety, as noted in this narrative: “Since I've had these appropriate ICD firings,

all I do is spend my time monitoring my heartbeat, and it's driving me crazy . . . You get negative reinforcement every time you get shocked.”

For some, restricting physical activity to avoid firing of the ICD made sense as many had either personally experienced, or heard stories of a relative who had experienced, an ICD shock that was precipitated by physical activity. Second, health care providers had advised participants of the strong correlation between physical activity and the triggering of a cardiac event. In keeping with the available scientific knowledge (upon the advice of health care providers) and experiential knowledge (witnessing or experiencing a cardiac episode while exercising), many elected to forego physical activity. The following narrative captured this experience:

My other daughter who had an ICD was a jock. All of her sports activity and her lifestyle have changed because of this condition. She was exercising and her pulse rate went up to 208. The defibrillator went off. Next thing, she was at the hospital. She has given up the marathon idea. When she is scuba diving she cannot go below 50 feet, and she will not exercise without a pulse meter on.

Participants reported that as they grew more accustomed to living with an ICD, became increasingly familiar with its role in disease management, and gained trust in its capability to respond to a cardiac event, they cautiously began to introduce light to moderate physical activity back into their routines. For some participants, the reintroduction of physical activity represented the fact that they were regaining control over their lives. In this case, reintroducing physical activity was part of adapting to living with the ICD, having faith in its capabilities, and incorporating the uncertainty as to when the ICD would fire next into one's sense of what is the norm: “I can still be active, go out, socialize, go dancing and pretty well do everything that a normal person can do. I go to the gym once a week. I walk about two to three miles a day.”

Essential to determining the appropriateness of an ICD shock was having a good understanding of how the ICD functions. Participants spoke with ease about the day-to-day functioning of the ICD. This included an understanding of when the ICD should discharge and within which heart-rate parameters: “Your electrical system misfires and you go into ventricular fibrillation and the ICD will pick up on this and it will fire and knock you back into a normal heart rhythm.” Participants also had a good understanding that technical difficulties with the ICD, such as displacement of wires or reprogramming, could cause an inappropriate shock.

Social Support

Family and friends were identified as having a significant role in balancing feelings of risk with the normalcy of everyday life. For many, knowing that other family members were readily accessible if needed was helpful, as this participant

described, “It was six in the morning, so I phoned my sister and told her she’d better get over here because my defibrillator just fired . . . So she got ready and came over.” Similar accounts of support provided by spouses were common. For the younger participants, it tends to be parents, siblings, or cousins who have had similar experiences that are called upon for support and to gain a better understanding of prescribed treatments such as the ICD: “Before (Son) went in and had his defibrillator he use to talk to (cousin) about it.” The importance of relying on informal support from family members contributes to family cohesion and its importance in the management of the ICD.

Living With Everyday Challenges

Living with an ICD at times means coping with restrictions such as loss of a driver’s license, employment, and educational opportunities that can cause social isolation. Participants reported that living with restrictions such as the removal of one’s driver’s license made them feel as if they were being punished for something that was out of their control: “It’s a classic example of negative reinforcement . . . every time you get shocked, and then you get punished when they take your driver’s license away.” For some, not having a driver’s license made them feel socially isolated, as they were no longer able to be independent in getting to work, or to do the practical activities of running a household, such as picking up groceries.

Knowing that having this disease puts others at risk, participants imposed driving restrictions on themselves. It was a common practice to drive only when absolutely necessary, or to drive slowly and with extreme caution. The following commentary summarizes the stress linked to having one’s driver’s license removed, getting it back, and driving:

I had my license back for nine days before I even drove. I wasn’t afraid to drive, but I think you lose your confidence in your own body. I was only driving back and forth to school, keeping on the inside lane, going slow, and always being cautious if I see a pedestrian; making sure where can I pull off the road.

In such cases, participants were more than willing to relinquish driving to another family member.

Participants’ social lives had also been restricted to varying degrees. For some, social isolation, self-imposed out of fear of the ICD firing, has become a key issue of concern: “I was afraid to go out in public. I didn’t want to go on the bus; I didn’t want to go to school; I didn’t want to go to work because I was afraid it was going to happen.” The impact of this self-imposed isolation was captured in one participant’s narrative: “There is no spontaneity in my life anymore. I am always worried and concerned. I find it embarrassing for people to know that I have had an incident (ICD firing). I have gone into avoidance.” Similar to other participants, this self-imposed isolation was in response to a “loss of confidence in

one’s body,” and the fear of “being embarrassed” that the ICD would fire causing a “scandal.” For another person, it was an alteration in body image that posed the biggest challenge. The weight of the ICD, its visibility under the skin, feelings of it grinding against the breast bone, and presence of the surgical scar caused so much stress that this person had ICD removed and later replaced with a new lighter one. On receipt of the new ICD, the participant expressed his contentment in this narrative:

The new ICD does not bother me as much, where it is less visible. I put on makeup to cover up the scars, so it makes it a bit easier. It’s not too bad but if I touch it I can feel it. I can actually cover it up a lot easier now. I put my arm in a certain position.

Participants also described their accounts of psychosocial distress related to the restrictions that having an ICD imposes on employment and educational opportunities. Acquiring gainful employment and having the education to secure employment were critical to those for whom it was important to be able to provide financially for the family. This concern was obvious in the case of an ARVC/D-positive fisherman who fished alone and had to hire another laborer to handle the boat in case the ICD fired, and he was rendered unconscious. Adding to the stress was that this position was difficult to fill, as potential candidates were reluctant to take the job, knowing that they would be expected to respond to a cardiac event: “I got a guy fishing with me and he is frightened to death all the time.” A lack of opportunities to enter into a field of interest was noted by one participant who felt that he would not pass a required medical examination: “I’d like to be a personal trainer but you have got to be able to teach other people to be fit and how can you when you are not healthy yourself?”

The most frequently cited barriers to care identified by participants were the shortage of health care professionals, the lack of knowledge by health care providers about the ICD, the high turnover rate of physicians and nurses, poor medical coverage for emergencies, and long wait times in local emergency departments in rural communities. In fact, many participants felt that they were often left to coordinate their own care. This was particularly noted in rural areas of the province and evident in the following comment:

I just get in the car and go (into the city) if the ICD fires. The bottom line is they (health care providers) are not educated in it, or they are just learning about it. I want the best there’s around.

Many narratives alluded to the fact that even if there were adequate health care professionals to meet the population’s needs, physical resources were scarce. Some smaller local facilities did not have the technology to download (nor health care personnel to interpret) information from the ICD. Therefore, individuals who went to a local facility either found themselves being told to go home and to come back if

the ICD fires again or were sent by ambulance to another center. Upon arrival in the city, unless there was some technological issue with the ICD itself (such as loose wiring or reprogramming), individuals are typically sent home with limited treatment changes. For many, these trips to local medical facilities become futile efforts, and individuals learn to draw on their own experiential knowledge about the ICD to decipher why it fired and to make decisions regarding their actual risk.

Although participants did appreciate those resources that were available, they also recognized that there is only so much that can be done given the resource limitations. Notwithstanding, the consensus was that participants were pleased with health care providers' bedside manner, support, and knowledge about and explanations of treatment regimes, as summarized in this narrative: "(Cardiologist) gives you the feeling that, okay, you got this thing (ICD) in, and you can feel good and safe." Visits to the cardiologist for most participants were described as ending on a positive note and providing reassurance as to the function of the ICD; to deliver a shock in the event of a cardiac episode.

To maintain a positive mind-set, individuals aligned their perception of health with having a "good quality of life," often defined as accepting that one cannot control all aspects of the disease, or have 100% control over one's fate. Many participants were of the opinion that they must appreciate what life has to offer and move on with their lives. This appreciation meant that participants had to reassign the meaning of the firing of the ICD from being something "risky" to something "normal" that saves their lives:

I am living life the way that I want to. If the ICD is going to go off, it's going to go off anyway. I am not going to sit at home and grieve myself to death because I got this problem. I'm going to do what I can do until the day comes that I can't.

As the ICD becomes a normal part of their lives, participants come to accept it as being critical to maintaining what they considered a "good" level of health. This includes having the confidence in their ability to manage a cardiac episode. This is evident in the voices of two younger participants who described their reaction when their cousin received a shock: "We knew what to do (when ICD fired)."

Discussion

While participants in this study did experience psychological distress analogous to that described in other studies with ARVC/D participants (Ingles et al., 2013; Jacq et al., 2009; James et al., 2012), they primarily emphasized the positive process of adjusting to and accepting the ICD to make it feel like a "normal" part of their everyday lives. Central to this process of adjusting and accepting was finding ways to manage the ICD, while moving forward with activities of everyday life. Similar to findings in other studies (Kantor,

Bullinger, & Gal, 2012; Morken et al., 2010; Zeigler & Nelms, 2009), the ICD was described as providing participants with a newfound sense of safety that did not exist prior to its implantation. The ICD shifted from being something that symbolized an increase in at-risk status to a technology that prevented them from being at risk. For some participants, this shift happened quickly, but for others, it took some time. This process is similar to that described in Lupton's (1999) work on the embodiment of pregnancy; risk perceptions are constructed, shaped, reshaped, and regulated by technology and experts.

Participants in this study spoke of the constant surveillance of their bodies and their efforts to modify their lifestyle to anticipate and decipher what was making the ICD fire. Similar accounts of the psychological stress that participants endured as they tried to predict when the ICD would fire have been reported in the cardiovascular literature (Hallas, Burke, White, & Connelly, 2010; Morken et al., 2010). Efforts to prevent the ICD from firing also caused anxiety, as participants associated the firing of the ICD as a sign that they were getting sicker and at an increased risk for death (a point also made by Palacios-Cena et al., 2011). In alignment with the literature (Zeigler & Nelms, 2009), participants in this study experienced anxiety prior to and post-shock. As explained in other research, a large part of the ICD-related distress reported in this study could be explained by participants' resistance to relinquish control over their lives to the ICD (Flemme et al., 2011). That is, they could not understand how their health could be so dependent on a machine and so resistant to modifiable lifestyle factors. Knowledge of the factors that contributed to the ICD discharge was welcomed, as it provided them with a list of what behavior needed to be modified or regulated. The finding that having a sense of control over the conditions that influence risk facilitates coping has been reported in other genetic studies (Hallas et al., 2010; McAllister et al., 2007; Vodermaier, Epslen, & Maheu, 2010).

Gradually, as found in other studies (e.g., Wheeler et al., 2009), the apprehension associated with the ICD firing subsides, as participants become accustomed to its firing and recognize that this is to be expected. Living with an ICD eventually becomes "normalized." Participants soon became accustomed to the fact that having an ICD means that there is a high probability it will fire in response to a cardiac event. Many were of the opinion that the ICD is an insurance policy and having a shock is just part of this policy. Living with the ICD means that getting on with one's life, accepting the restrictions imposed by the ICD, and appreciating life as it is lived. As the use of the ICD becomes normalized during the illness process, participants come to accept it as critical to maintaining "good" health, a finding that is also supported in the literature on the ICD (Flemme et al., 2011). Nurses can play a significant role in helping these individuals to (re) conceptualize what it means to be healthy while being "at-risk."

For two younger participants in this study, acceptance of the ICD meant being able to understand and manage its firing in response to a cardiac event. Although the literature has linked being younger to psychosocial distress (Bedair et al., 2015; James et al., 2012), this study found that this distress was short lived. Although these participants initially described being apprehensive about the ICD firing, once they had experienced it themselves both as the recipient of a shock and as the manager of a person having a shock, they reported a new sense of confidence in their abilities that decreased their anxiety. Given that this finding is based on the accounts of only two participants, additional research is needed to explore the relationship between age and psychosocial distress in young ARVC/D individuals.

The findings in this study support other research, indicating that individuals with an ICD who have had restrictions placed on their driver's license experience feelings of resentment, anger, loss of independence, worry that sudden incapacitation while driving might pose risk to themselves or others, and in some cases, frustration with not being able to get daily taken-for-granted chores completed, such as grocery shopping (Shea, 2004). Participants also described psychosocial distress related to the restrictions that having an ICD imposes on employment and educational opportunities, a phenomenon also reported by others (Probst et al., 2011; Shea, 2004). Individuals living with chronic heart disease have reported similar life challenges with employment and social interactions (Nordgren, Asp, & Fagerberg, 2007). This finding highlights the fact that nurses and other health care professionals must be prepared to provide counseling in these areas.

This study supports the literature that individuals who have an ICD as a primary treatment for a genetically linked heart condition, as with ARVC/D, do have unique needs (Day, 2012; Ingles et al., 2013). Not only is this population dealing with the fact that they are at risk of a lethal heart condition, but if they wish to survive a cardiac event, they must have an ICD, even if they are asymptomatic. Participants did exhibit psychosocial difficulties; however, over time, they began to integrate management of the ICD into their everyday lives.

Conclusion

This study adds to preliminary discussions surrounding the needs of individuals living with ARVC/D who have an ICD. This study highlights the significance of offering targeted interventions to this population pre, during, and post ICD insertion. Findings suggest that individuals who have an ICD for ARVC/D have diverse needs that emerge at different points. Knowing this, ongoing assessment of these individuals is imperative to ensure that their psychosocial needs are met over the long term. This is not a straightforward path, as for most integrating the ICD into one's life as something "normal" comes with time. As nurses are often the point of entry into the health care system in rural regions, they need

to be prepared to address the unique needs of these individuals as they adjust to having the ICD. Having knowledgeable and consistent health care providers in rural communities to provide care to this cohort is the first step. Furthermore, as noted by others (Day, 2012) we need to consider the uniqueness of genetic conditions such as ARVC/D, and address the question of precisely what factors contribute to the psychosocial distress related to having an ICD, versus those associated with the actual disease itself. On a broader level, nurses need to be aware of prevalent genetic conditions in regions that they serve and coinciding treatments. Given that a genetic condition like ARVC/D does not exist in silo but across generations, it is important that nurses engage family members in discussions about how to support each other as they adjust to prescribed treatment regimes in anticipation of others being ARVC/D positive.

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