

Original Article

Advance Care Planning for Adults With Moderate-to-Severe Congenital Heart Disease: Readiness Trial

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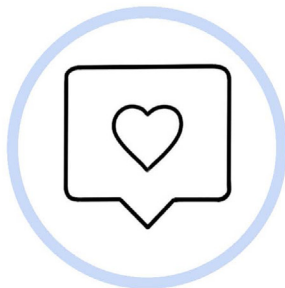
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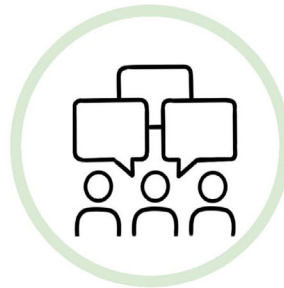
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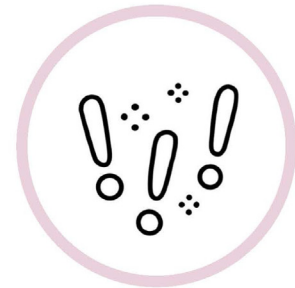
PATIENT VALUES

- Having answers about chances of survival
-
- Understanding choices of treatment and outcomes related to forgoing treatment
-
- Support from family and friends
-
- Being physically comfortable
-
- Not being alone



ACP CONVERSATIONS

- The preferred time to discuss wishes for future healthcare is before getting sick, while healthy
-
- More likely to initiate discussion with spouse/partner when conducting conversations related to wishes for care when faced with life-threatening illness
-
- Most patients have not participated in formal ACP conversations with a healthcare provider



PATIENT FEARS

- Being a burden
-
- Living with great pain
-
- Not being able to communicate with my loved ones in a meaningful way
-
- Total physical dependency on others
-
- Having an illness that significantly impacts your well-being or quality of life

ABSTRACT

Background: Adults with congenital heart disease (CHD) are growing in number and living longer with complex lesions; however, many are

RÉSUMÉ

Contexte : Les adultes atteints de cardiopathie congénitale sont de plus en plus nombreux et vivent plus longtemps avec des lésions

at risk of death in midlife. Conversations about advance care planning (ACP) in this population have been found to be infrequent and not part of regular outpatient care. The intent of this study was to explore readiness of patients with adult CHD (ACHD) to discuss ACP and assess the impact of interventions to support ACP conversations.

Methods: We conducted a prospective, randomized, single-centre trial of adults with moderate to severe CHD. Simply, the control group was asked if they were ready to discuss their wishes if they were to become seriously ill. The intervention group was provided 2 additional resources with the readiness question, the modified Lyon Family-Centered ACP survey and a patient-partner created ACHD-ACP video.

Results: A total of 111 responses (control [n = 59] and intervention group [n = 52]) were collected for analysis. Women represented 59.0% of the participants. The mean age was 39.6 years (standard deviation = 14.8 years). No significant difference was found between the control and experimental groups' readiness (94.9% and 90.4%, respectively). Most participants (92.8%) responded positively towards initiating conversations related to end-of-life and ACP discussions.

Conclusions: We found that adults with CHD are ready to have ACP conversations as part of their outpatient care. Patient preferences and values should guide ACP conversations; further research is needed to determine whether the modified Lyon Family-Centered ACP survey and ACHD-ACP video are helpful adjuncts for ACP in outpatient clinics.

complexes; cependant, nombre d'entre eux présentent un risque de décès au mi-temps de la vie. Il a été démontré que les conversations sur la planification préalable de soins (PPS) sont rares au sein de cette population et qu'elles ne font pas partie des soins ambulatoires habituels. Cette étude avait pour but de déterminer dans quelle mesure les adultes atteints de cardiopathie congénitale étaient disposés à parler de la PPS et d'évaluer l'effet des interventions pour favoriser des conversations sur la PPS.

Méthodologie : Nous avons mené un essai monocentrique, prospectif et à répartition aléatoire auprès d'adultes atteints de cardiopathie congénitale modérée à sévère. Nous avons tout simplement demandé aux membres du groupe témoin s'ils étaient prêts à parler de ce qu'ils souhaiteraient s'ils devenaient gravement malades. Le groupe d'intervention a eu à sa disposition deux ressources additionnelles portant sur la réceptivité des patients, le questionnaire Lyon Family-Centered ACP Survey (ML-ACP) modifié et une vidéo sur la PPS en présence de cardiopathie congénitale créée en partenariat avec les patients.

Résultats : Au total, 111 réponses (groupe témoin [n = 59] et groupe d'intervention [n = 52]) ont été recueillies à des fins d'analyse. Les femmes représentaient 59,0 % des participants. L'âge moyen était 39,6 ans (écart-type = 14,8 ans). Aucune différence notable n'a été observée entre le groupe témoin et le groupe d'intervention (respectivement 94,9 % et 90,4 %) quant au fait d'être prêts à tenir ces conversations. La plupart des participants (92,8 %) ont répondu positivement, se disant prêts à amorcer des conversations sur la fin de la vie et la PPS.

Conclusion : Nous avons constaté que les adultes atteints de cardiopathie congénitale sont prêts à avoir des conversations sur la PPS dans le cadre des soins ambulatoires. Les conversations sur la PPS doivent être guidées par les préférences et les valeurs des patients, il faut d'autres recherches pour déterminer si le questionnaire ML-ACP et la vidéo sur la PPS en présence de cardiopathie congénitale sont des ajouts utiles pour la PPS dans les cliniques externes.

In recent decades, surgical and medical advances have shifted the demographics of people living with congenital heart disease (CHD). As recent as 50 years ago, the majority of people born with moderate to severe CHD died in childhood.¹ Now, adults with CHD account for over two-thirds of the total CHD population.² Despite medical advances, those with CHD remain at risk of premature death in midlife.^{1,3-6} Although there is a risk of sudden death with CHD, adults with CHD may suffer complications of their disease that significantly impact quality of life before a terminal event. Common complications of adult CHD (ACHD) include heart failure, stroke, endocarditis, arrhythmias, aortic dissection, myocardial infarction, or other noncardiac issues.^{2,4,7,8} Many patients with CHD have high health care utilization throughout their lifetime due to the risk of complications and need for frequent interventions.⁹⁻¹¹ Despite this exposure to the health care system and the risk of early death, conversations about advance care planning (ACP) in this population are infrequent and not

regularly included in outpatient care.^{5,12,13} Current challenges in ACP for those with CHD arise from the paucity of data for how and when to initiate ACP in young people with chronic, life-limiting illnesses other than cancer. Many ACP resources are aimed at older populations, limiting their applicability to and use by patients with CHD. Further barriers include health care providers' discomfort with bringing up end-of-life (EOL) issues with young people and the prognostic uncertainty along the heterogeneous spectrum of ACHD.^{12,14,15} ACP is essential to ensure that interventions align with the person's wishes and promote living well with a life-limiting illness.¹⁶ Current guidelines have emphasized the importance of individualized ACP.^{17,18} A critical component to deciding when to begin ACP is evaluating when a person is ready to discuss ACP. When studied in other populations, early ACP and palliative care have been associated with improved patient satisfaction, similar clinical outcomes, and decreased health care costs.¹⁹⁻²¹ In this study, we evaluated readiness for ACP and whether the use of an introductory video created for adults with CHD improved their readiness to discuss ACP.

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Methods

This project emerged from research priorities set by teens and adults with CHD in a speciality clinic in Alberta,

Table 1. Demographics

Demographic	Standard of care (n = 59)	Intervention (n = 52)
Age (y)		
18-29	21 (35.6)	9 (17.3)
30-39	15 (25.4)	20 (38.5)
40-49	10 (16.9)	5 (9.6)
50-59	5 (8.5)	13 (25.0)
60+	8 (13.6)	5 (9.6)
Identified gender		
Male	23 (39.0)	23 (44.2)
Female	36 (61.0)	29 (55.8)
Other	0 (0.0)	0 (0.0)
Highest level of education attained		
Below high school	1 (1.7)	0 (0.0)
Some high school	1 (1.7)	1 (1.9)
High school	18 (30.5)	15 (28.8)
Trade school	11 (18.6)	12 (23.1)
University degree	24 (40.7)	20 (38.5)
Master's degree	3 (5.1)	3 (5.8)
Medical doctorate	1 (1.7)	1 (1.9)
Postdoctorate	0 (0.0)	0 (0.0)
Occupation status		
Student	6 (10.2)	3 (5.8)
Employed	38 (64.4)	38 (73.1)
Student and employed	0 (0.0)	1 (1.9)
Unemployed or retired	15 (25.4)	10 (19.2)
Legally documented alternative decision maker		
Yes	15 (25.4)	17 (32.7)
No	42 (71.2)	35 (67.3)
Unsure	2 (3.4)	0 (0.0)
Marital status		
Single	23 (39.0)	19 (36.5)
Partnership	2 (3.4)	1 (1.9)
Common-law	7 (11.9)	4 (7.7)
Married	27 (45.8)	26 (50.0)
Divorced	0 (0.0)	2 (3.8)
Widowed	0 (0.0)	0 (0.0)
Congenital heart disease*		
TOF	13 (22.0)	10 (19.2)
Coarctation of aorta	7 (11.9)	9 (17.3)
VSD	10 (16.9)	4 (7.7)
ASD	7 (11.9)	4 (7.7)
Ebstein's anomaly	3 (5.1)	4 (7.7)
Tricuspid atresia	2 (3.4)	2 (3.8)
Fontan	2 (3.4)	5 (9.6)
Transposition of great arteries	2 (3.4)	4 (7.7)
Other	16 (27.1)	13 (25.0)
Unsure	2 (3.4)	1 (1.9)
Medical and surgical health history		
No cardiac surgical history	5 (8.5)	8 (15.4)
Pacemaker involved in surgical history	3 (5.1)	4 (7.7)
1 cardiac surgery	22 (37.3)	24 (46.2)
2 cardiac surgeries	12 (20.3)	9 (17.3)
3 cardiac surgeries	8 (13.6)	5 (9.6)
4 or more cardiac surgeries	11 (18.6)	6 (11.5)
Unsure	1 (1.7)	0 (0.0)
Documented goals of care		
R1	4 (6.8)	8 (15.4)
R2	0 (0.0)	0 (0.0)
R3	1 (1.7)	0 (0.0)

Table 1. Continued.

Demographic	Standard of care (n = 59)	Intervention (n = 52)
M1	0 (0.0)	1 (1.9)
M2	0 (0.0)	0 (0.0)
C1	0 (0.0)	0 (0.0)
C2	0 (0.0)	0 (0.0)
Not documented	54 (91.5)	43 (82.7)
Legal documents		
Will	17 (28.8)	18 (34.6)
Agent	7 (11.9)	6 (11.5)
Power of attorney	12 (20.3)	11 (21.2)
Advanced directive	9 (15.3)	10 (19.2)

Values are presented as n (%).

ASD, atrial septal defect; TOF, tetralogy of Fallot; VSD, ventricular septal defect.

* Percent values do not sum to 100% because some individuals had several congenital heart diseases.

Canada.²² In focus groups, participants voiced a specific desire to know about life expectancy and felt that these conversations were not a part of their regular care.²² Most with moderate to complex CHD (86%) assumed that they would have a normal life expectancy because this had not been explored with their CHD specialist.²² To better understand readiness of patients with ACHD to discuss ACP with health care providers and assess the impact of directed interventions to support ACP conversations, we conducted a prospective, randomized, single-centre trial of adults with moderate to severe CHD, defined by the Bethesda criteria.²³ This study was approved by the University of Calgary Conjoint Health Research Ethics Board.

Participants were included in this study if they had moderate to severe CHD, were older than 18 years, and able to communicate in English to allow the use of standardized surveys. Participants were excluded if they lacked capacity in health care decision-making. Participants were recruited through an ACHD speciality clinic in Calgary, Alberta. They were asked by their care team during their regularly scheduled appointment whether they were interested in participating in a study about ACP and then met with a research assistant to undergo detailed consent for participation. Demographic information was obtained, including asking participants whether they had a documented goal of care, a shorthand method used in our provincial health care system to communicate to health care providers what the person's wishes for intervention would be if they were seriously ill. Participants were recruited from November 2021 to June 2023 with delays in enrolment occurring because of the COVID-19 pandemic.

We aimed to enroll approximately 100 participants based on the number of patients with moderate to severe CHD who attended our local ACHD clinic in a 1-year period assuming 50% enrolment. We randomized participants using gender stratification to ensure equal representation of those who identify as men, women, or other nonbinary gender representations in the control and intervention groups. We used this strategy to determine if there were sex- or gender-based differences between the groups as CHD is experienced differently by individuals for many reasons, including their sex and gender.

Surveys were loaded on 2 iPads, labeled as iPad 1 or 2. The research assistant was blinded to which iPad was the control

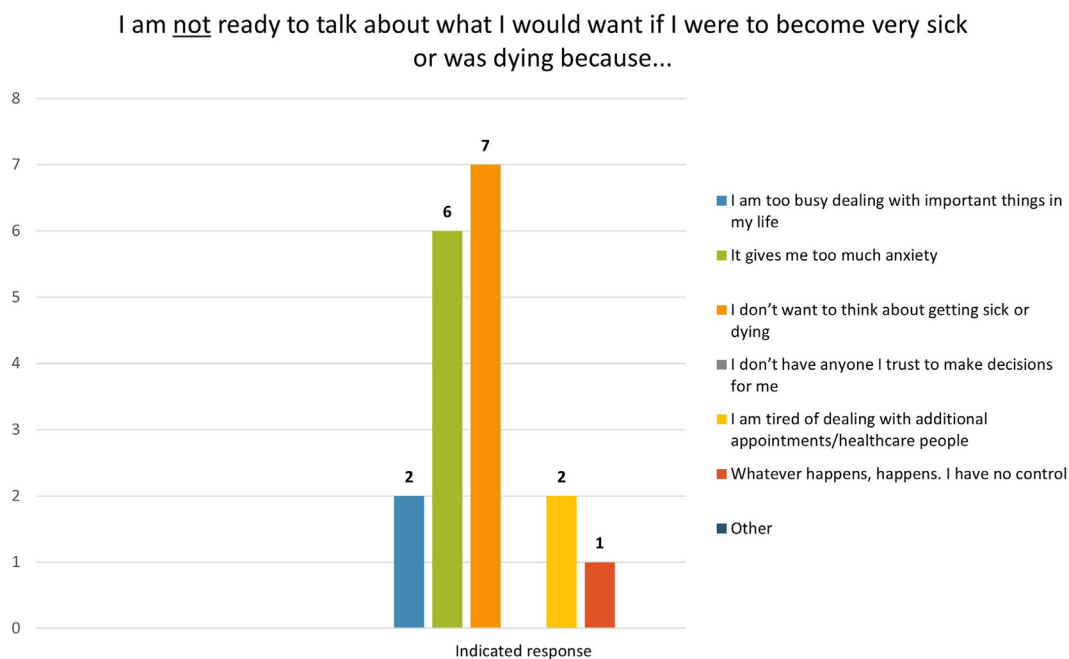


Figure 1. Reasons for avoiding ACP. Eight participants reported not being ready to discuss ACP, with their response represented in each column. Participants were able to select more than 1 answer in response to the survey question. ACP, advance care planning.

and intervention survey. The control group was provided with a demographics form to fill out as well as a “Readiness Survey,” which asked whether the participant was ready to have ACP conversations. If they indicated that they were not ready, they were then asked to provide reasons why they did not feel ready.

The intervention group was provided the demographic form but was also provided with 2 additional resources developed and adapted by members of our research team, including patient partners before completing the “Readiness Survey.” Our team modified the Lyon Family-Centered ACP (ML-ACP) survey, which had been developed to evaluate young adults’ and adolescents’ values, beliefs, and experience with illness and EOL care.²⁴⁻²⁶ This 31-question scale was modified to reflect ACP legislation in our jurisdiction adding a question that included whether the participants knew who their legal decision maker would be. We also adapted some questions to ask the participant what they would want if they were seriously ill and could not speak for themselves, rather than “if they were dying,” reflecting recommendations in the Serious Illness Conversation Guide used in our provincial health care system to guide ACP.²⁷ In addition, an ACHD-ACP video was collaboratively developed with 3 patient advisors to introduce the concept of ACP from the perspective of those living with CHD. The 4-minute video features patients with CHD telling their story of ACP. Topics discussed in the video include what ACP is, why it is important at any age, and the process of ACP. Participants electronically completed the surveys alone in a clinic room.

To evaluate the impact of interventions on the participant’s readiness, the control and intervention groups were asked whether they were ready to discuss their wishes if they were to become seriously ill or were dying. They were also asked to provide reasons for not feeling ready to discuss ACP. The time to

complete surveys took participants between 10 and 25 minutes. Survey data were collected and stored using the REDCap database to ensure a secure, Health Insurance Portability and Accountability Act (HIPAA)-compliant, web-based system for data management. Participants were deidentified using a random number generator, and all identifiable survey data were removed from survey responses. Survey data were analysed with the JASP 0.17.1 (Intel; University of Amsterdam, Amsterdam, The Netherlands) statistical software tool with descriptive statistics.

Results

Demographics

A total of 111 responses (control [n = 59] and intervention group [n = 52]) were collected for analysis, with all participants having moderate to complex CHD (Table 1). The discrepancy between control and intervention enrolment occurred because of the use of preassigned gender stratified file folders, some of which had been pulled in the wrong order during enrolment. In addition, 12 participants who were enrolled declined to complete the surveys, further contributing to the imbalance between control and intervention groups. Sixty-five (59.0%) of the participants were women. The mean age of the total sample was 39.6 years (standard deviation = 14.8 years). Eighty-eight percent of participants had a history of undergoing at least 1 cardiac surgery. When asked about ACP topics, most participants (64.9%) did not have any legal documents consisting of either a will, agent, power of attorney, or an advance directive. When considering all legal document types, a will was most frequently completed by participants (31.5%). Of the participants who had at least 1 completed legal document, those aged older than

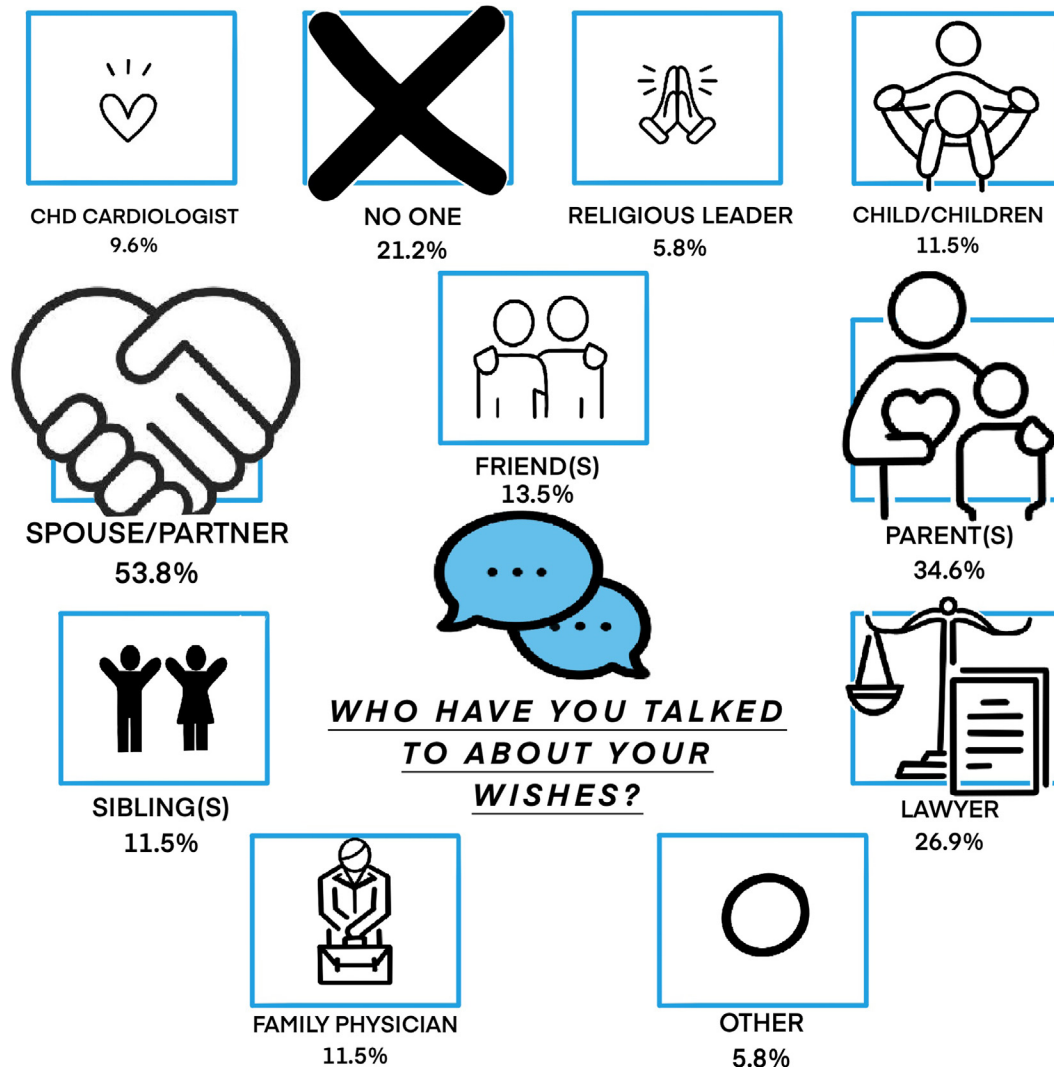


Figure 2. Advance care planning discussions. The figure represents who the intervention group ($n = 52$) have informally or formally discussed their preferences and values around the type of care they would want if they were seriously ill or dying. Data are presented as a percentage. Participants were able to select more than 1 answer in response to the survey question. CHD, congenital heart disease.

50 years were more likely to have a legal ACP-related document ($n = 31$, 80.7%) compared with participants aged less than 50 years ($n = 80$, 17.5%). The majority of participants (97.0%) did not have documented goals of care despite regular health care visits. Participant willingness to discuss ACP topics was not found to be associated with other variables such as marital status, age, or severity of CHD.

Readiness

Most participants indicated that they were ready to discuss ACP (92.8% of the unified population). No difference was found between the control and experimental groups' readiness (94.9% and 90.4%, respectively; $P > 0.05$). Of those in the unified sample who reported that they were not ready to discuss ACP and EOL care ($n = 8$), the factors most often reported were anxiety (75.0%) and "not wanting to think about death or dying" (87.5%) (Fig. 1).

Formal ACP

A minority of participants indicated that they had written down thoughts about future health plans (28.8%), participated in formal conversations about ACP with a health care provider or a lawyer (25.0%), or designated an alternative decision maker (ADM) in a legal document (25.0%).

Participants reported having conversations about ACP with their spouse or partner (53.8%), parents (34.6%), and lawyer (26.9%). They were least likely to have ACP conversations with their general practitioner (11.5%), CHD specialist (9.6%), or faith representative (5.8%). Just over one-fifth reported not speaking with anyone about ACP or their preferences for treatment if seriously ill (Fig. 2).

When considering whether participants felt their ADM would know what their wishes would be if they were to become very ill and could not speak for themselves, less than half of participants were very sure that their ADM would know what type of care was desired (42.3%), followed by

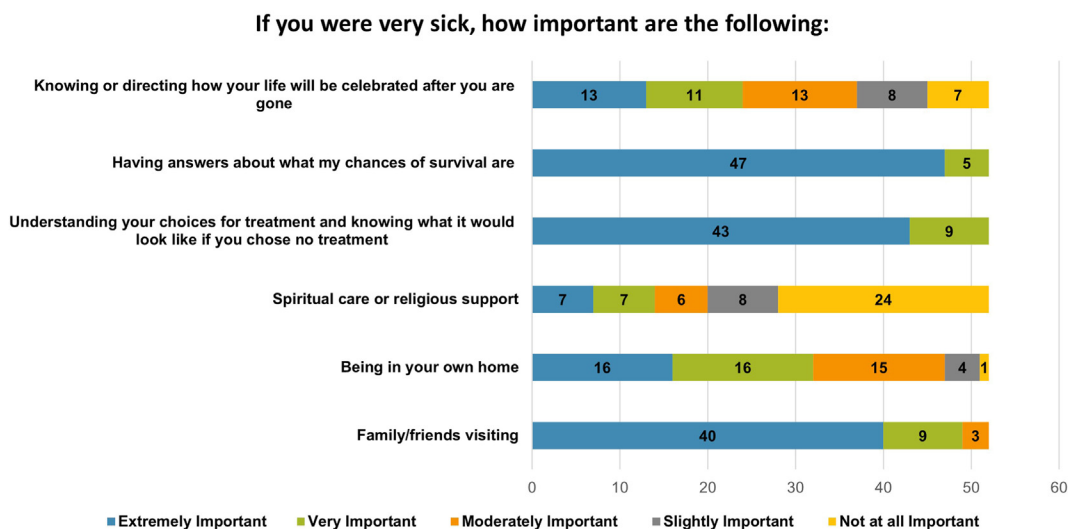


Figure 3. Values and preferences during serious illness. Participants in the intervention group (n = 52) were asked to rate the importance of different preferences if they were faced with a serious illness.

nearly a third of participants who were only “somewhat sure” (34.6%). Participants reported that their CHD specialist best understood their treatment preferences compared with other support networks or health care providers.

ACP conversation content, timing, and patient values

When asked about the best time to conduct ACP discussions, most participants indicated a preference for conversation initiation before getting sick, while healthy (80.8%), followed by when first diagnosed with a serious illness (53.8%).

If they were sick, most participants indicated that having visitation from family and friends (76.9%), understanding choices for treatment and outcomes related to forgoing treatment (82.7%), and having answers about chances of survival rate (90.4%) were extremely important (Fig. 3). Importantly, 73.1% also reported that not being able to communicate with loved ones in a meaningful way would be a worse outcome than death.

The most frequently cited fear when thinking about serious illness was related to being a burden (Fig. 4). This was further supported by 63.5% of participants selecting total physical dependence on others for toileting or dressing as a worse outcome than death. Few participants (15.4%) responded that they were very afraid of an illness that results in not being able to work. Only 11.5% of participants indicated an extreme likeliness to avoid medical appointments in fear of finding a serious health issue.

When considering EOL preferences, 75.0% of participants rated not being alone as “most important,” followed by being pain-free (61.5%), being physically comfortable (46.2%), being at peace spiritually (33.7%), and not being attached to life support (30.7%). Further supporting this preference, 59.6% of participants felt that living with great pain was worse than death.

Discussion

It is critical for health care providers to assess a patient’s readiness for ACP conversations. Our study confirms that

adults with CHD are ready to discuss ACP in an outpatient clinic visit, echoing findings in other studies that most adults with CHD want to discuss ACP while healthy with their CHD cardiologist.^{5,13,15,26,28,29} A barrier frequently cited in the literature is that young adults with CHD are not ready for ACP,¹³ but in reality outpatient cardiologists are well positioned to lead these discussions given their longitudinal relationship with patients and their families. Participants highlighted the value of their relationship with their specialist cardiologist, with 88.5% feeling often understood by their specialist cardiologist, consistent with previous studies showing high trust among this population with physicians.^{15,30} Second, our study looked to explore how to have meaningful ACP conversations in an outpatient setting. Our study used the foundational work by Tobler et al.²⁶ to explore preferences and values of adults with CHD around serious illness and EOL to ultimately integrate meaningful ACP into regular outpatient care. Tools such as the ML-ACP may provide a helpful framework for clinicians, patients, and ADMs to discuss specific preferences in the context of their unique illness experience; however, further research exploring the impact of this tool on ACP in regular outpatient clinic visits is needed.

Barriers to participant readiness

Despite participants actively being followed in an ACHD clinic, 89.0% did not have a goal of care designation, similar to the findings by Tobler et al.,²⁶ who found that 77% of participants had never written down thoughts about their future health plans. Participants cited not wanting to think about getting sick or dying (n = 7, 87.5%) and anxiety (n = 6, 75.0%) as reasons not to participate in ACP. Similarly, Greutmann et al.¹⁵ found that barriers to ACP included patients not wanting to think about death. Steiner et al.¹² found that avoidance was described by participants as easier than dealing with difficult conversations. Further, in adults with CHD who were pregnant, Steiner et al.²⁹ found that reasons for not participating in ACP included fear and anxiety about

How afraid are you of...

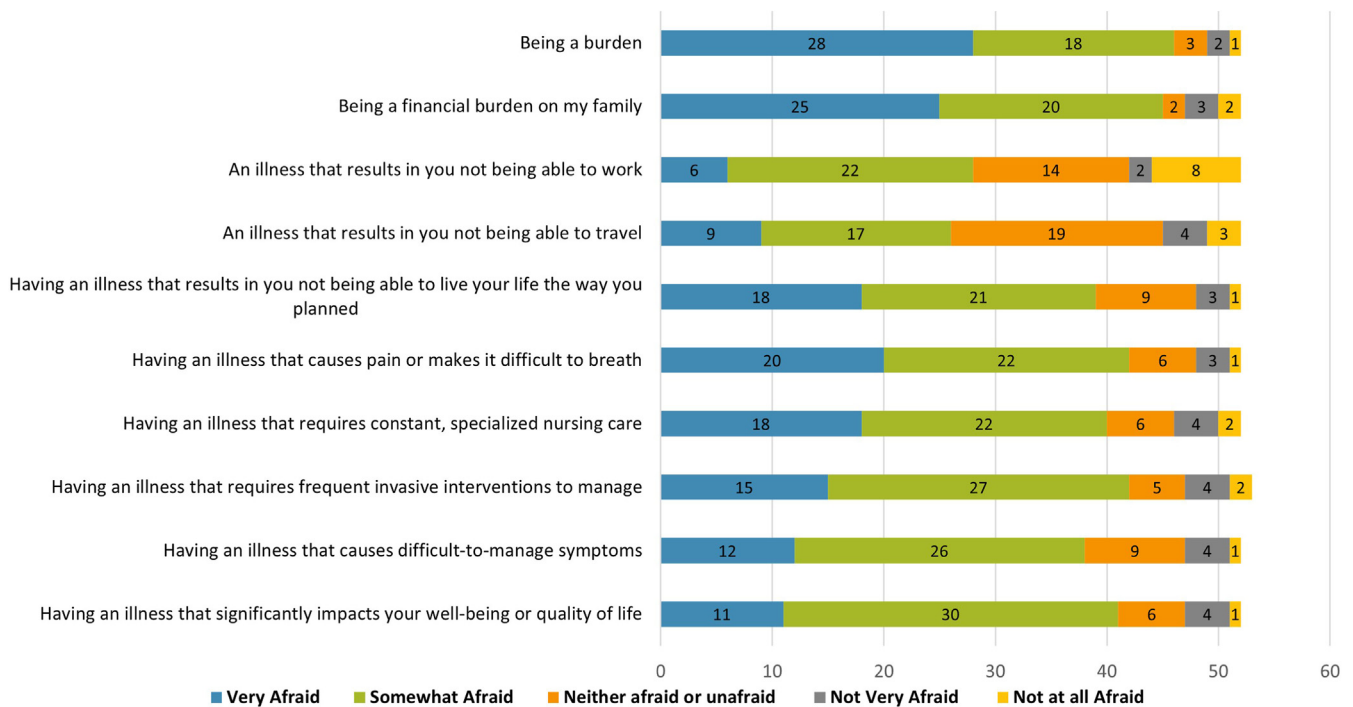


Figure 4. Expressed fears with serious illness. Participants in the intervention group (n =52) were asked to rate their fears when faced with a serious illness.

ACP, that ACP was “not a priority,” or that they were overwhelmed already with multiple appointments. Adults with CHD have higher risk of early childhood medical trauma from serious illness³¹ and higher incidence of neurocognitive disorders.³² This underscores the need for clinicians to independently assess a patient’s readiness to discuss ACP before inviting them to participate in interventions such as the ML-ACP or ACHD-ACP video to promote autonomy and self-determination in their care. ACP discussions are dynamic and iterative. Our study results reflect the growing body of literature supporting early conversations before a life-threatening or serious event. Proactive discussions between patients and their specialist care team should occur as a standardized part of the transition to ACHD clinics from paediatric care and be readdressed with any invasive procedure, hospitalization, or with any new or serious health concern.

Modifying the survey to have the readiness question upfront to determine the continuation of the survey may better promote patient-led interventions, in this study we assessed readiness after the survey and video to determine whether these interventions improved participant readiness but did not find any difference between the control and interventions group.

ACP preferences and values

Participants in our study commonly cited being a physical burden and having to rely on others for care as unacceptable outcomes of treatment. This fear may reflect the younger age of participants, who may perceive their duty and value within their family structure to be the provider. It also reflects societal

values, that often young people with disability face discrimination with social, economic, and health inequality. Openly addressing these worries and how they can be mitigated may benefit patients and family’s anxiety around illness and disability.

To promote individualized ACP, knowing the patient’s preferences, values, and unacceptable outcomes upfront can help steer conversation about which interventions best reflect the person’s goals. Greutmann et al.¹⁵ cited that 79% of providers feel it would be beneficial to have additional resources and information related to ACP, and 87% are interested in further exploring communication methods for EOL conversations. Shared documentation of preferences, through the use of tools such as the ML-ACP, between patients, their families, and providers can guide ACP conversations. Documentation of ACP conversations and their outcomes is a crucial part of outpatient care and should be reassessed at regular intervals as determined by the patient and their specialist team.

Future research to explore how the ML-ACP survey can be used in regular visits as well as clinician and patient perception of ease of use will be helpful to further tailor its use in this population. Also cited in the literature is the variability between jurisdictions for legal accommodations for ACP.³³ The ML-ACP can be adapted to include legal ACP/ADM definitions to reflect regional legal differences.

Limitations

The data collected in this study represent a population that consented to participate in a study about ACP, which may over-represent those ready or willing to discuss ACP.

Nonbinary participants were absent among those who participated, resulting in a lack of representation of all gender identities. In addition, we did not capture the diversity of the population through racial identity in this study. Beyond the observation that current ACP resources appear largely directed towards the elderly population, many ACP tools lack representation of Canada's racial diversity. In the same way that lower representation of patients with ACHD in ACP resources might discourage younger adults from engaging in ACP, equity-diversity-inclusion research may better address representation barriers to ACP.

Next steps—integrating ADM into ACP

The experience of adults with CHD is complex. There are life-long connections between those with CHD, their families, and health care providers. In infancy and childhood, initial discussions about outcomes and ACP occur primarily with parents or caregivers, but ADMs may shift over time from parents to partners to children of adults with CHD.^{30,33} To better reflect these multiple transitions, ACP needs to be iterative and include those who are essential to making decisions with and for those with CHD.^{15,33} Only 76.2% of participants in our study were very sure or somewhat sure that a legally designated substitute decision maker would know what type of care they want. Future research exploring how to best identify and integrate ADMs into outpatient appointments to ensure that values and preferences of patients with CHD are known to not only health care providers but their ADMs is needed.

Conclusions

Assessing patient readiness is a critical component to effective ACP. We found that young people are ready to have ACP conversations as part of their regular outpatient care. Patient preferences and values should guide ACP conversations; further research is needed to determine whether the ML-ACP survey and ACHD-ACP video are helpful adjuncts for ACP in outpatient clinics.

Clinical Perspectives

- Clinical competencies: The clinical implications of this work align with the patient care and interpersonal and communication domains to be used by readers to enhance their competency as caregivers.
- Translational outlook: A challenge that requires future research is how to integrate alternate decision makers into outpatient ACP. Only 76.2% of participants in our study were very sure or somewhat sure that a legally designated substitute decision maker would know what type of care they want. This supports the need for future research including the patient-family dyad in survey completion to support shared decision-making.

Acknowledgement

We would like to acknowledge Dr Maureen Lyon for her support, mentorship, and development of the Lyon Family-Centered ACP survey.

Ethics Statement

This study was approved by the University of Calgary Conjoint Health Research Ethics Board. This research has adhered to the relevant ethical guidelines.

Patient Consent

The authors confirm that patient consent forms have been obtained for this article.

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Disclosures

The authors have no conflicts of interest to disclose.

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