Barriers and Facilitators of Palliative Care and Advance Care Planning in Adults With Congenital Heart Disease

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Palliative care has potential to improve quality of life and goal-concordant care for patients with adult congenital heart disease (ACHD). However, it is rarely employed prior to critical illness because the best methods for implementation are not well-defined. We qualitatively evaluated ACHD patients' understanding of and opinions regarding palliative care and advance care planning (ACP) to better define the needs of this population. We conducted a thematic analysis of 25 semistructured interviews with patients with ACHD in which we assessed participants' perspectives on the need for, and barriers and facilitators to, the use of palliative care and ACP. In a group of participants with ACHD (mean age 38, 48% male) classified as simple (24%), moderate (32%), or complex (44%), we identified 4 major themes: (1) using knowledge to combat future uncertainties; (2) unfamiliarity with and limited exposure to palliative care and ACP; (3) facilitators and barriers to engaging in palliative care and ACP; and (4) importance of timing and presentation of ACP discussions. In conclusion, participants expressed a desire for knowledge about ACHD progression and treatment. They supported routine incorporation of palliative care and ACP and identified related facilitators and barriers to doing so. Importantly, timing and format of these discussions must be individualized using shared decision-making between clinicians, patients, and their families. **(Am J Cardiol 2020;135:128−134)**

Background

Honoring patients' care preferences surrounding quality of life drives high-quality medical care.[1] This is especially important for patients with adult congenital heart disease (ACHD) who live with chronic illness. Life expectancy has markedly increased with medical and surgical advancements, but many continue to face disability, multimorbidity, and early death.[2] Palliative care improves patients' quality of life through the early identification, assessment, and treatment of pain and other problems that cause suffering.[3] Part of palliative care is advance care planning (ACP), a process that allows individuals to plan for their future health care.[4] Prior studies indicate that patients with ACHD want information about prognosis, view discussions about future care as important, and are willing to participate in such discussions.[5−7] Yet actions involving palliative care and ACP rarely occur prior to the onset of severe illness.[8,9] Practices vary, partly because best methods for implementing them in this population are not well defined.[7,10] In this study, we utilized qualitative methods to assess participants' perspectives on the need for, and barriers and facilitators to, the use of palliative care and ACP.

Methods

We conducted a qualitative study in follow-up to a cross-sectional survey study evaluating patients' views on palliative care with special emphasis on ACP.[7] Following survey completion, respondents were given the option to provide contact information if interested in participating in a subsequent interview. Out of 150 survey respondents, 103 (69%) provided contact information for this purpose. We contacted potential participants in 3 rounds, using random selection and then purposive sampling to ensure a diverse sample. Purposive sampling criteria included diversity in ACHD lesion severity, age, race, and sex. We attempted to contact 42 potential participants; we were unable to reach 14 after at least 3 attempts, 1 declined, and 2 were pending at the time of saturation. This study was approved by the University of Washington Institutional Review Board. All participants gave informed consent and permission for medical record access.

We conducted semi-structured interviews 4 to 6 months after survey completion. A single team member (JMS) interviewed participants either in person (n = 1) or over the phone (n = 24), based on participant preference. Interviews ranged in length from 22 to 59 minutes, mean 43 minutes, and were audio-recorded. Interview questions (see Appendix) were open-ended and assessed knowledge of and views

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regarding palliative care and ACP. The interview guide was created based on research questions and survey findings. We completed 25 interviews, at which point we reached thematic saturation, the point at which no new themes were identified.[11,12] Interviews were transcribed verbatim; identifying information was excluded.

Demographic information and ACHD diagnosis were obtained from electronic medical records and survey questions.[7] ACHD was categorized as being of simple, moderate, or great complexity in agreement with the ACHD guidelines.[13] Data were analyzed using the web-based application Dedoose (Version 8.0.35 (2018). Los Angeles, CA: Socio-Cultural Research Consultants, LLC; www.dedoose.com).

Interview data were analyzed using thematic analysis, an approach which allowed us to identify themes contained in the interviews.[14–16] We created an initial codebook beginning with interview questions themselves as well as themes previously identified in prior literature.[5,6,9] We then revised the codes and themes as they emerged from the interview data. Codes were reviewed and revised by 2 team members (JMS and RAE) using 2 interviews, and then 3 team members (JMS, AD, CEB) iteratively revised the codebook by coding 5 additional interviews. We ensured that we reached consensus, defined as at least 80% agreement among team members, before finalizing the codebook. The remainder of the interviews were then coded by 1 team member (AD) and reviewed by JMS to ensure consensus in code assignment. Themes and the links between themes were derived by JMS and AD. Team members did not have a prior relationship with study participants.

Table 1.
Participant demographics

Characteristic	Statistic (n = 25)
Age (years),* median, range	38 (21-63)
Male*	12 (48%)
White	17 (68%)
Hispanic	3 (12%)
Asian	3 (12%)
Other	2 (8%)
Adult Congenital Heart Disease Severity*	
Simple	6 (24%)
Moderate	8 (32%)
Complex	11 (44%)
Education Level	
High School Diploma/Equivalent	8 (32%)
4-year College	10 (40%)
Graduate	7 (28%)
Religious (self-report)	9 (36%)
Physical/Emotional Symptoms	
None	8 (32%)
Nonlimiting	11 (44%)
Limiting	6 (24%)
Prior experience with advance care planning or palliative care	10 (40%)

* Part of purposive sampling criteria

Results

Twenty-five patients with ACHD took part in the study (Table 1), 12 (48%) of whom were male. Mean age was 38 years (range 21 to 63 years). Six participants (24%) had simple, 8 moderate (32%), and 11 complex (44%) ACHD lesions. The majority (76%) reported not feeling limited by physical or emotional symptoms related to ACHD. Ten (40%) had some prior exposure to ACP or palliative care, most frequently in relation to a surgery or cardiovascular intervention. Participants qualitatively reported good functional status despite living with significant ACHD, describing it as their own version of normal. However, they all described concerns that can be categorized as "palliative care needs" as well as barriers and facilitators to the use of palliative care and ACP.

At some point in the interview, 72% of participants described having experienced negative emotional impacts of living with ACHD. Feelings of uncertainty and worry about the future as it relates to having ACHD were ubiquitous. Participants wanted to know as much as possible about their condition and prognosis, feeling this might provide some degree of control over the unknown. They frequently described knowledge as power, and they cited ACP as a way to facilitate educated decision-making "if things got bad." "I need to know everything, because I need to know, 'Oh, this could happen.' 'This is normal. . . . I want to be able to, if a decision comes up, have a sense of knowledge about it already to help make that decision" (43-year-old female, complex disease). Participants also hoped having knowledge could lead to better outcomes: "I just want to know what my options are and be knowledgeable about the disease that I have and what I can do to either help it or make it better so I have a better quality of life overall." (21-year-old female, complex disease).

Feelings of uncertainty were exacerbated by not having others like them (i.e., with similar severity ACHD) with whom to discuss their feelings. Being part of a support group or ACHD community was endorsed by more than half of participants as a desirable way to learn what to expect as they grow older with ACHD. "Even if it's just to learn about who are some people that have a condition like I do that are older than me, and what are some of the things that they go through when they're older. Like what are some of the things I can expect as I get older? Because frankly, I don't really know" (44-year-old male, complex disease).

Participants were notably unfamiliar with the definition and elements of palliative care. Seven participants (28%) stated they did not know what palliative care entailed, and another 6 (24%) participants asked for further clarification. Those who reported having heard the term poorly understood its meaning. Participants described it using phrases such as "giving up" and "hospice": "I'm dying. We have exhausted all of our options" (63-year-old female, moderate disease). Another equated it to their healthcare team giving up on them: "It sounds scary, and I would be scared if I hear that, because that's like hearing my doctor give up on me" (39-year-old female, complex disease). Participants with complex ACHD seemed relatively more likely to report palliative care familiarity than those with milder disease (5 compared to 2 each for simple and moderate disease).

Similarly, most participants were only vaguely familiar with the term "advance care planning." Despite not being able to define it clearly, approximately two-third of participants described encountering some form of ACP in the past.

Twenty-eight percent of participants had previously signed official documents such as wills or advance directives. Some remembered filling out ACP paperwork prior to a surgery (3 participants), and 1 had done so during a heart failure hospitalization. Those with complex ACHD seemed relatively more likely to have had prior ACP discussions regarding their own care (4 participants compared to 1 each for simple and moderate disease), but not more likely to be familiar with the concept in general. The majority of participants (56%) had at some point thought about or talked to family members about ACP, but "just never got around to" taking any formal action such as completing an advance directive. Older participants, in particular, mentioned it had "come up over the years" or felt like it was discussed more recently because of their age, with most participants over 35 years of age reporting prior ACP knowledge.

Many participants familiar with palliative care or ACP learned about it from having experienced the illness or death of a loved one (5 participants) rather than having encountered it with regard to their own condition. One remarked, "Honestly, it was brought to my mind less from my own treatment and more when my dad got cancer. Prior to that, it wasn't really something I thought about much, even... when I was having surgeries" (27-year-old male, simple disease). A middle-aged participant with advanced complex disease described in detail the extensive planning he had done for his elderly father, but then acknowledged never having thought to do the same for himself. Some participants learned about it through their jobs in medical or scientific positions, or at school (5 participants). These participants seemed most likely to associate palliative care with comfort and pain management as opposed to only hospice or end-of-life care. They were also receptive to these services.

When discussing palliative care as a tool to alleviate suffering, participants were interested in incorporating it into their future care. Many voiced that they would want assistance being comfortable and able to focus on their priorities if their medical condition deteriorated: "If I was bedridden, or things were just going downhill, then yeah, I'd want to be in a situation where I was getting the support I needed" (31-year-old male, complex disease). One participant described seeing a role for palliative care in potentially increasing longevity through the provision of comfort-focused care: "If you're in pain all the time, you don't want to live. Your quality of life sucks. So if you can get really good interim care, then you will want to stick around more. You'll live longer" (56-year-old female, moderate disease).

Similarly, participants acknowledged the importance of ACP as a way to plan for care in the future. Half of the study participants said something like "anything can happen" at any time. One participant said, "Well, people have car accidents and end up on life support. And you never know when that sort of thing's gonna happen. You don't have to be old... and how healthy or unhealthy you are is not necessarily a good indicator of when you're gonna go" (63-year-old female, moderate disease). Overall, ACP was described as a tool to be prepared for the unexpected. "We may go through our whole lives never needing that seatbelt, but because we know it, and we use it, that 1 time in a million, it will save our lives... Maybe we'll never need [palliative] care or hospice, or we'll never need a counselor, but we know it's there" (56-year-old female, moderate disease). Other facilitators to ACP are described in Table 2.

In contrast to aspects that encouraged participants to seek out palliative care and ACP, they also noted barriers. For palliative care, participants' association of palliative care with end-of-life care or hospice clearly led to a reluctance to be referred to palliative care services: 2 participants indicated they were surprised to be asked about palliative care at this time, given their relative good health. For ACP, participants primarily discussed feeling unsure about the

Table 2.
Facilitators and barriers to participation in advance care planning

Facilitator	Supporting Quote
Arranging logistics	"You're able to spend whatever quality time that you have with your friends and family, rather than having to worry about things that you shouldn't have to worry about." − 36-year-old male, complex disease
Lessening burden on loved ones	"I want to make sure that I have everything laid out, and that I'm not a burden to my children... I don't want to put them in situations where they'll have to make decisions. I would like those decisions already made for them." − 36-year-old male, moderate disease
Ensuring preferences are honored	If I was walking and got hit by a bus and got put in a state where the decision would need to be made for me, there is a strong chance that they would not be decisions I would want." − 27-year-old male, simple disease
Barrier	**Supporting Quote**
Cost of creating legal documents	People just try to get by day by day, let alone spend $100 on a living will. I think it's costly... You have to have a lawyer prepare an advance directive. Maybe if they were given a discount if they did it sooner than later, that might be something that they could think about. − 39-year-old female, complex disease
Lack of resources to make informed decisions	My main impediment is just lack of information and lack of resources... The main impediment is no one has handed me a packet before. − 30-year-old female, complex disease I feel like I'm a layman trying to make that decision, and I just don't know enough. − 40-year-old male, complex disease
Denial and avoidance are easier	Maybe it's a part of denial to say that nothing is ever going to happen, so I don't actually have to put this in writing. − 36-year-old female, simple disease Sometimes it's better to live life with rose-colored glasses. − 37-year-old male, complex disease

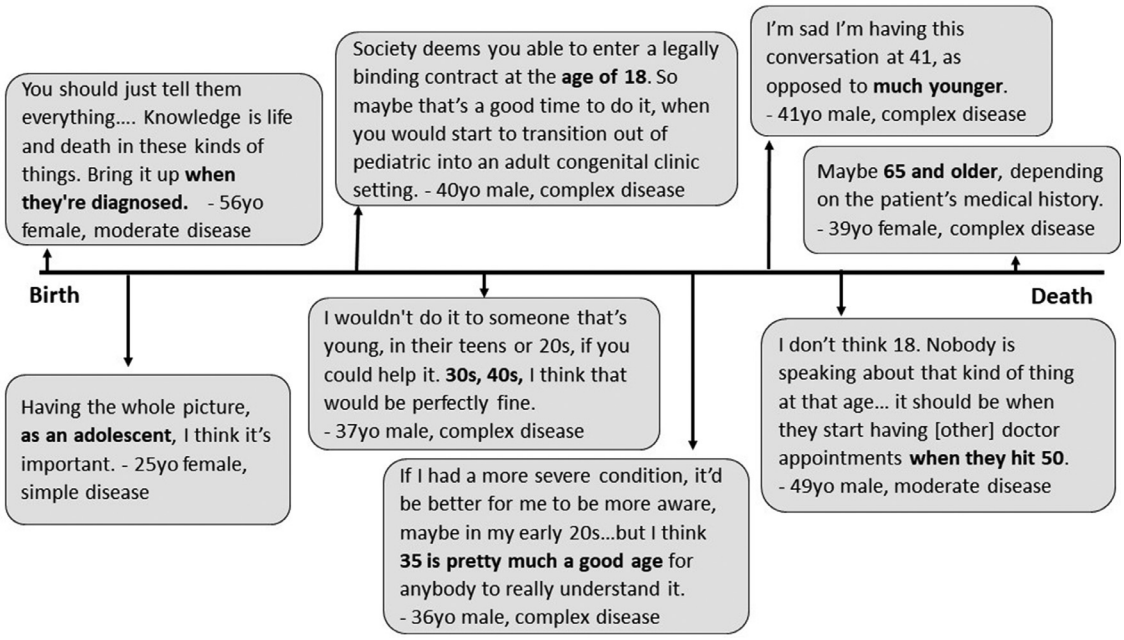

Figure 1. . Participants' opinions about the ideal age to begin ACP discussions.

personal relevance of ACP. "It's hard to feel like you want to talk about it if everything's going OK. It doesn't feel like a priority" (52-year-old male, moderate disease). Remembering a prior ACP discussion, 1 participant remarked, "It didn't feel as real, talking about those things, because I was a teenager. . . You're not really worried about what's going to happen to you later on in life" (25-year-old female, complex disease). Other barriers to ACP are described in Table 2.

Most participants expressed a desire for early ACP discussions, prior to the development of serious complications. "When you're well is kind of the time to make those decisions or start planning out those decisions" (27-year-old male, simple disease). They expressed concern that delaying decision making until health deterioration might be stressful, or even too late. "It would be kind of hard to do something like that in the heat of the moment of crisis, and I think it would just add to the stress and anxiety involved" (33-year-old female, moderate disease). However, there were some who preferred delaying ACP conversations until a certain time or event, such as if the healthcare provider became concerned about an acute decline. This was particularly the case for the few who had concerns that ACP discussions might negatively affect their emotional quality of life and who preferred to focus on positivity. Participants' suggestions as to a specific age when these discussions might be appropriate ranged almost the whole life span (Figure 1). Participants also acknowledged that certain factors, such as clinical status, maturity, and emotional or mental health issues, could impact the "best" time for discussions.

Despite the wide range of responses surrounding timing, almost everyone suggested that ACP discussions should be presented as standard practice. Normalizing ACP discussions as "an item on the agenda" (29-year-old male, simple disease) was popular. "It becomes a rite of passage for someone with a congenital heart defect. . . That's just what you do. . . Everybody does it." (40-year-old male, complex disease). However, participants also pointed out that discussions should not be forced. They suggested starting with a basic introduction to "at least just get it on their radar and just let them know that this is something that you may have to think about, and that we will be having conversations about this more and more as time goes on" (21-year-old female, complex disease). Many felt that decisions should not be made during the first discussion. Participants also recognized that care preferences could evolve, indicating the need to revisit them.

Participants made a variety of suggestions regarding the materials through which ACP information should be made available (Figure 2). The majority (68%) felt written or electronic materials should be accompanied by a verbal introduction. Twelve participants (48%) expressed interest in written materials such as pamphlets or brochures, while 5 (20%) expressed a preference for links to online materials. Two participants suggested using both modalities concurrently, specifically to accommodate patients with less technological comfort or access. Five participants (20%) explicitly wanted a checklist as part of their resources.

Regardless of the platform, it was important to participants that materials be tailored. "If you give them something that has a gray-haired person on the front, I think it implies that 'Well, these are things to think about, but I don't really need to do it right now, because it's not going to affect me yet.' Which, something can happen at any time. . . So I think if you want people to have the conversation earlier and feel like it is relevant to their lives, then even changing the pictures on the front could be helpful" (21-year-old female, moderate disease). Participants acknowledged the important role their parents have played in decision making surrounding their heart condition, and several expressed that they would want their parents and/or spouses involved in meetings about ACP. They also commented on the importance of a strong physician-patient relationship for the success of ACP discussions.

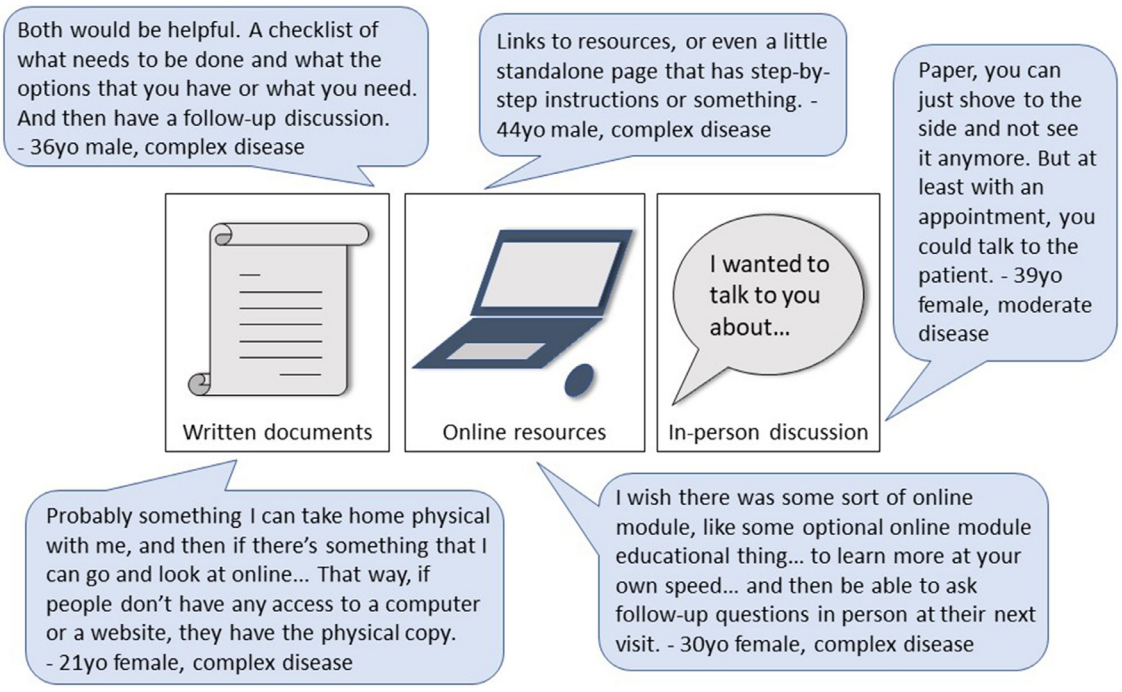

Figure 2. Participants' suggestions regarding preferred format for receiving PC/ACP materials.

Discussion

This is the first study to use qualitative methods to examine the barriers and facilitators regarding implementation of palliative care and ACP in ACHD patients. It adds depth to the small body of existing quantitative literature on palliative care in ACHD,[5–9,17] and it expands the existing literature on living with ACHD.[17,18] The following 4 themes arose from interviews: (1) using knowledge to combat future uncertainties; (2) unfamiliarity with and limited exposure to palliative care and ACP; (3) facilitators and barriers to engaging in palliative care and ACP; and (4) importance of timing and presentation of ACP discussions.

Palliative care is infrequently utilized in ACHD prior to critical illness,[6,17,18] despite ACC/AHA guidelines[13,19] recommending its inclusion and prior work suggesting that patients want more exposure to palliative care and ACP.[5–7] Findings in this study were similar. However, participants viewed planning for the future as difficult due to the distress associated with living without a clear prognosis, also in line with prior ACHD literature.[20] Despite interest in palliative care and ACP, participants lacked understanding of these concepts, a similar finding as in cystic fibrosis, an analogous life-long illness where ACP discussions are infrequent.[21,22] Ongoing patient education about disease progression and palliative care options will be important to address some of the misconceptions identified as barriers in this study. Building on familiarity with palliative care gained from experiences with others (i.e. family members with cancer) and the concept that ACP may be most useful in cases of prognostic uncertainty may be helpful.

Early adulthood has been suggested as the optimal time for the initiation of palliative care or ACP discussions in patients with ACHD and in similar populations.[6,23–25] However, our participants provided a wide range of responses regarding best timing; they suggested that the clinical situation as well the maturity level of the person were important considerations. Their responses suggest that healthcare providers do not need to be constrained by a single "best time" - that using clinical judgement and patient-centered communication might help to identify appropriate opportunities. In addition, participants in our study supported normalizing palliative care and ACP discussions by including them as part of routine adult care, and everyone agreed that avoiding them altogether was not the correct approach. Similarly, in cystic fibrosis, patients indicate that ACP discussions are best addressed as part of a routine outpatient visit, when they are healthy and without acute health stressors, and that waiting for limited prognosis is too late.[22,25]

These findings suggest that clinicians should introduce basic education on palliative care as a routine part of patient care in ACHD starting early in adult life, while keeping in mind that actual decision-making is a process that will vary in type and timing from person to person. Materials, including text and images, should be tailored to a younger audience with chronic disease to address participants' concerns about generic materials. Ultimately, timing and format preferences remained highly individualized, reflecting the need for health care providers leading these discussions to be equipped with flexibility and diverse resources. Furthermore, preferences may change over time, and innovations in therapies have and will continue to impact prognosis and therefore palliative care and ACP.

In addition to clinicians' roles in introducing palliative care, peer support communities may facilitate learning about ACP, symptom management, and working though difficult decisions. Support groups or peer gatherings were suggested as desirable means to provide a type of "mentorship" in living with ACHD. Similar findings regarding the utility of peer support have been reported in adolescents and young adults

with cancer.[26] However, it should also be acknowledged that support groups are not a preferred social environment for everyone, and that potential for dissemination of misinformation exists. Nonetheless, these findings suggest support groups should be investigated as a means of addressing palliative care needs in ACHD.

Several study limitations may limit the generalizability of these findings. Participants were recruited from a pool of respondents to a survey about ACP and palliative care who indicated their interest in participating in an interview. It is possible that these participants had heightened interest in this topic and that their responses could have been influenced by survey participation. In addition, participants were well-educated, reported reasonably good health, and received care in a health system with a relatively advanced palliative care program. Ethnic minorities were underrepresented. Finally, this study did not examine the role of specific clinicians in providing this type of care. Future studies should address these issues.

This study presents a qualitative evaluation of the perspectives of patients with ACHD on the need for, barriers and facilitators to the use of palliative care and ACP. Participants expressed a desire for knowledge about disease progression and treatment options, including the incorporation of ACP and palliative care into routine adult ACHD care. When and how to appropriately and effectively address these needs requires careful consideration and shared decision-making between clinicians, patients, and their families. Support groups present an intriguing way to address knowledge gaps and facilitate ACP. This study further suggests that palliative care interventions should build on participants' general appreciation of the importance of palliative care. Studies should also incorporate the perceived benefits of lessening burden on loved ones and ensuring preferences are honored.

Authors' Contribution

Jill M. Steiner, MD, MS: conceptualization, investigation, formal analysis, software, data curation, validation, writing - original draft, writing - review and editing, visualization, project administration; Alysha Dhami, MD: investigation, formal analysis, software, data curation, validation, writing - original draft, writing - review and editing; Crystal E. Brown, MD, MA: formal analysis, validation, writing - review and editing; Karen K. Stout, MD: conceptualization, writing - review and editing; J. Randall Curtis, MD, MPH: conceptualization, writing - review and editing, supervision, funding acquisition; Ruth A. Engelberg, PhD: conceptualization, validation, writing - review and editing; James N. Kirkpatrick, MD: conceptualization, writing - review and editing, supervision.

Disclosures

The authors have no conflicts of interest to disclose.

Supplementary materials

Supplementary material associated with this article can be found in the online version at https://doi.org/10.1016/j.amjcard.2020.08.038.

1. Khandelwal N, Curtis JR, Freedman VA, Kasper JD, Gozalo P, Engelberg RA, Teno JM. How often is end-of-life care in the United States inconsistent with patients' goals of care?Available at: www.liebertpub.com. Accessed January 6, 2020.
2. Steiner JM, Kovacs AH. Adults with congenital heart disease – facing morbidities and uncertain early mortality. *Prog Pediatr Cardiol* 2018; 48:75–81. https://doi.org/10.1016/j.ppedcard.2018.01.006.
3. Anon. WHO | WHO definition of palliative care. *World Heal Organ* 2012. Available at: https://www.who.int/cancer/palliative/definition/en/. Accessed January 2, 2019 .
4. Anon. Definition of advance care planning. *Natl Hosp Palliat Care Organ* 2020. Available at: https://www.nhpco.org/patients-and-caregivers/advance-care-planning. Accessed January 6, 2020.
5. Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach SL, Kovacs AH. Knowledge of and preference for advance care planning by adults with congenital heart disease. *Am J Cardiol* 2012;109: 1797–1800.
6. Deng LX, Gleason LP, Khan AM, Drajpuch D, Fuller S, Goldberg LA, Mascio CE, Partington SL, Tobin L, Kim YY, Kovacs AH. Advance care planning in adults with congenital heart disease: a patient priority. *Int J Cardiol* 2017;231:105–109.
7. Steiner JM, Stout K, Soine L, Kirkpatrick JN, Curtis JR. Perspectives on advance care planning and palliative care among adults with congenital heart disease. *Congenit Heart Dis* 2019;14:403–409. Available at: http://www.ncbi.nlm.nih.gov/pubmed/30575286. Accessed December 24, 2018.
8. Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach LS, Kovacs AH. End-of-life in adults with congenital heart disease: a call for early communication. *Int J Cardiol* 2012;155:383–387. Available at: http://www.ncbi.nlm.nih.gov/pubmed/21094550. Accessed January 19, 2019.
9. Greutmann M, Tobler D, Colman JM, Greutmann-Yantiri M, Librach SL, Kovacs AH. Facilitators of and barriers to advance care planning in adult congenital heart disease. *Congenit Heart Dis* 2013;8:281–288. Available at: http://doi.wiley.com/10.1111/chd.12025. Accessed September 10, 2019.
10. Kovacs AH, Grace SL, Kentner AC, Nolan RP, Silversides CK, Irvine MJ. Feasibility and outcomes in a pilot randomized controlled trial of a psychosocial intervention for adults with congenital heart disease. *Can J Cardiol* 2018;34:766–773.
11. Guest G, Bunce A, Johnson L. How many interviews are enough? an experiment with data saturation and variability and a good number of journals in the. *Field methods* 2006;18:59–82.
12. Hennink MM, Kaiser BN, Marconi VC. Code saturation versus meaning saturation: how many interviews are enough? *Qual Health Res* 2017;27:591–608.
13. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, Nido P del, Fasules JW, Graham TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. *J Am Coll Cardiol* 2008;52:e143–e263.
14. Attride-Stirling J. Thematic networks: an analytic tool for qualitative research. *Qual Res* 2001;1:385–405. Available at: http://journals.sagepub.com/doi/10.1177/146879410100100307. Accessed January 2, 2019.
15. Ando H, Cousins R, Young C. Achieving saturation in thematic analysis: development and refi nement of a codebook. *Compr Psychol* 2014;3:1–7. Available at: https://journals.sagepub.com/doi/pdf/10.2466/03.CP.3.4. Accessed April 29, 2019.
16. Vaismoradi M, Turunen H, Bondas T. Content analysis and thematic analysis: implications for conducting a qualitative descriptive study. *Nurs Heal Sci* 2013;15:398–405.
17. Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach LS, Kovacs AH. End-of-life care in hospitalized adults with complex congenital heart disease: care delayed, care denied. *Palliat Med* 2012;26:72–79.
18. Steiner JM, Kirkpatrick JN, Heckbert SR, Sibley J, Fausto JA, Engelberg RA, Randall Curtis J. Hospital resource utilization and presence of advance directives at the end of life for adults with congenital heart disease. *Congenit Heart Dis* 2018;13:721–727. Available at: http://www.ncbi.nlm.nih.gov/pubmed/30230232. Accessed December 24, 2018.
19. Stout KK, Daniels CJ, Chair V, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, Crumb SR, Dearani JA, Fuller S, Gurvitz M, Khairy P, Landzberg MJ, Saidi A, Marie Valente A, Hare GF Van. 2018 AHA/ACC guideline for the management of adults with

congenital heart disease. *Circulation* 2019;139:e698–e800. Available at: http://ahajournals.org. Accessed July 18, 2019.
20. Berghammer MC, Brink E, Rydberg AM, Dellborg M, Ekman I. Committed to life: adolescents' and young adults' experiences of living with fontan circulation. *Congenit Heart Dis* 2015;10:403–412.
21. Sawicki GS, Dill EJ, Asher D, Sellers DE, Robinson WM. Advance care planning in adults with cystic fibrosis. *J Palliat Med* 2008;11:1135–1141.
22. Linnemann RW, Friedman D, Altstein LL, Islam S, Bach K-T, Georgiopoulos AM, Moskowitz SM, Yonker LM. Advance care planning experiences and preferences among people with cystic fibrosis. *J Palliat Med* 2019;22:138–144. Available at: https://www.liebertpub.com/doi/10.1089/jpm.2018.0262. Accessed June 16, 2019.
23. Wiener L, Zadeh S, Battles H, Baird K, Ballard E, Osherow J, Pao M. Allowing adolescents and young adults to plan their end-of-life care. *Pediatrics* 2012;130:897–905.
24. Christenson K, Lybrand SA, Hubbard CR, Hubble RA, Ahsens L, Black P. Including the perspective of the adolescent in palliative care preferences. *J Pediatr Heal Care* 2010;24:286–291. https://doi.org/10.1016/j.pedhc.2009.07.001.
25. Kazmerski TM, Weiner DJ, Matisko J, Schachner D, Lerch W, May C, Maurer SH. Advance care planning in adolescents with cystic fibrosis: a quality improvement project. *Pediatr Pulmonol* 2016;51:1304–1310.
26. Pennant S, C. Lee S, Holm S, Triplett KN, Howe-Martin L, Campbell R, Germann J. The role of social support in adolescent/young adults coping with cancer treatment. *Children* 2019;7:2.