

Special Series: Science in Action: Evidence and Opportunities for Palliative Care Across Diverse Populations and Care Settings

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Palliative Care Opportunities Among Adults With Congenital Heart Disease—A Systematic Review



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Abstract

Context. Little is known about advance care planning (ACP) and palliative care needs among adults with congenital heart disease (ACHD).

Objectives. The objective of this study was to identify and synthesize studies concerning palliative care among ACHD patients.

Methods. We searched five electronic databases (PubMed, Embase, SCOPUS, Web of Science, and CINAHL) using the keywords palliative care and congenital heart disease. Inclusion criteria were adults (age > 18 years) with congenital heart disease and publications in English through March 3, 2019.

Results. Our search yielded 2872 studies, and after removal of duplicates, we screened 2319 abstracts and identified seven for inclusion. Study findings were grouped into three domains: ACP, symptomatology, and end-of-life care. Among the five cross-sectional studies, only 1%–28% of ACHD patients recalled participating in ACP discussions with their doctors but 69%–78% reported a strong interest and desire to participate in ACP. In one study, 46% ($n = 67$) of patients had elevated anxiety symptoms (Hospital Anxiety and Depression Scale [HADS-A] ≥ 8) and 11% ($n = 15$) had elevated depressive symptoms (HADS-A ≥ 8). ACHD patients who had a documented goal of care conversation before cardiac decompensation had a lower incidence of resuscitation and aggressive treatments at end of life (12% [$n = 3$] vs. 100% [$n = 12$], $P < 0.001$).

Conclusion. While few ACHD patients complete advance directives, our findings support that many ACHD patients recognize the value of initiating end-of-life and goals of care conversations early on in the course of illness. Future studies investigating communication and implementation strategies of ACP as well as the symptom experience of patients with ACHD are needed. *J Pain Symptom Manage* 2019;58:891–898. © 2019 American Academy of Hospice and Palliative Medicine. Published by Elsevier Inc. All rights reserved.

Key Words

Advance care planning, palliative care, adult congenital heart disease

Introduction

Advances in cardiothoracic surgical techniques over the last 5 decades have enabled more adults to live longer despite significant congenital heart disease,

resulting in a population of adults with congenital heart disease (ACHD) greater than 1.4 million in the U.S.¹ Yet, even with optimal surgical and medical ACHD management, resultant medical and surgical complexities

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still lead to frequent hospital admissions, significant psychological stress, and persistent risk of premature heart failure—related mortality.^{2–5} In a large national Dutch Cohort ACHD registry, the overall median age of death was 48.8 years, while the median age of death from sudden cardiac arrest was 39 years.⁶

Given the risk of comorbidities and premature death among adults with ACHD, proactive advance care planning (ACP) and end-of-life discussions among ACHD patients and their surrogates are already a Class IIa recommendation in the American Heart Association/American College of Cardiology 2018 ACHD Guidelines.⁷ Nevertheless, the extent of dissemination and implementation of palliative care and ACP in ACHD populations remain unclear. Consequently, we conducted a systematic review of the literature to identify and synthesize available data on the impact of ACP and palliative care on patient- and family-reported outcomes and end-of-life care among ACHD populations (Table 1).

Methods

Search Strategy

In conjunction with a medical librarian (C. W.), we queried PubMed, Embase, SCOPUS, Web of Science, and CINAHL. The search used the keywords and medical subject headings of “palliative care,” “congenital heart defect,” “heart malformation,” “heart defect,” “hospice care,” “end-of-life care,” and “advance care planning.” Searches were limited to studies of adults aged 18 years and older, published in English from database inception until March 3, 2019, and contained original, quantitative, or qualitative data related to patient- and family-centered outcomes. We excluded gray literature, case reports, case series, and commentaries or perspective pieces. We registered the study on Prospero (ID# CRD42019116675).

Study Selection

Search results were uploaded into Covidence software (V. 1182, Covidence; Alfred Hospital [Melbourne, Australia] and Instituto de Efectividad Clínica Y Sanitaria [EROS, Buenos Aires, Argentina]), and duplicates were removed and screened based on the criteria set forth in Table 1. Two members of the study team independently screened each title and abstract and then full articles, and a third study team member adjudicated any disagreements. The study was completed and results are reported consistent with the Preferred Reporting Items for Systematic Reviews and Meta-analyses statement.⁸

Data Extraction

Data were abstracted from the full-text articles into a preformatted template table by one team member and checked by the other members. The preformatted template table included the following: first author,

Table 1
PICOTS Question

PICOTS Question	
Among adults with congenital heart disease, what are the patient- and/or family-reported outcomes on advance care planning and end-of-life care?	
P—Adults with congenital heart disease	
I—None	
C—None	
O—Qualitative or quantitative patient-reported outcomes, family-reported outcomes, and/or end-of-life care	
T—Any	
S—Any	
Inclusion criteria	Exclusion criteria
1. Published before March 3, 2019	1. Published in the gray literature
2. Human participants aged over 18 years with congenital heart disease	2. Commentaries, case reports, or case studies
3. Published in English	3. Studies that did not incorporate patient- or family-reported outcomes and/or end-of-life-related outcomes
4. Original quantitative or qualitative research	

year of publication, country, objective, study design, study setting, sample size, inclusion/exclusion criteria, intervention and comparison/control group (if applicable), outcome measures, and results. All extracted data and discrepancies were reviewed, and adjudication was performed by group consensus. The final results were presented in a tabular summary and summarized in a qualitative synthesis.

Data Synthesis

Data were synthesized into a descriptive summary. Owing to study type and heterogeneity, we could not conduct a meta-analysis.

Quality Appraisal

All studies were descriptive and thus not appropriate for quality review using existing tools.

Results

Our search of five databases yielded a total of 2872 studies (Fig. 1), with 2319 articles remaining after removal of duplicates. After title and abstract screening, 20 studies remained for full-text screening and seven studies met final criteria for inclusion in this review (Fig. 1). Among these seven included studies (Table 2), four were conducted in Canada and three in the U.S. Two of the studies were retrospective cohort studies analyzing ACHD patients' hospitalization records, while the remaining five were cross-sectional surveys administered to ACHD patients and/or clinicians in the outpatient setting. Across all included studies, the mean patient age ranged from 33 to 37 years and 39%–54% of patients were female. In the three studies that recorded patient race/ethnicity, Caucasians made up 83%–89% of study

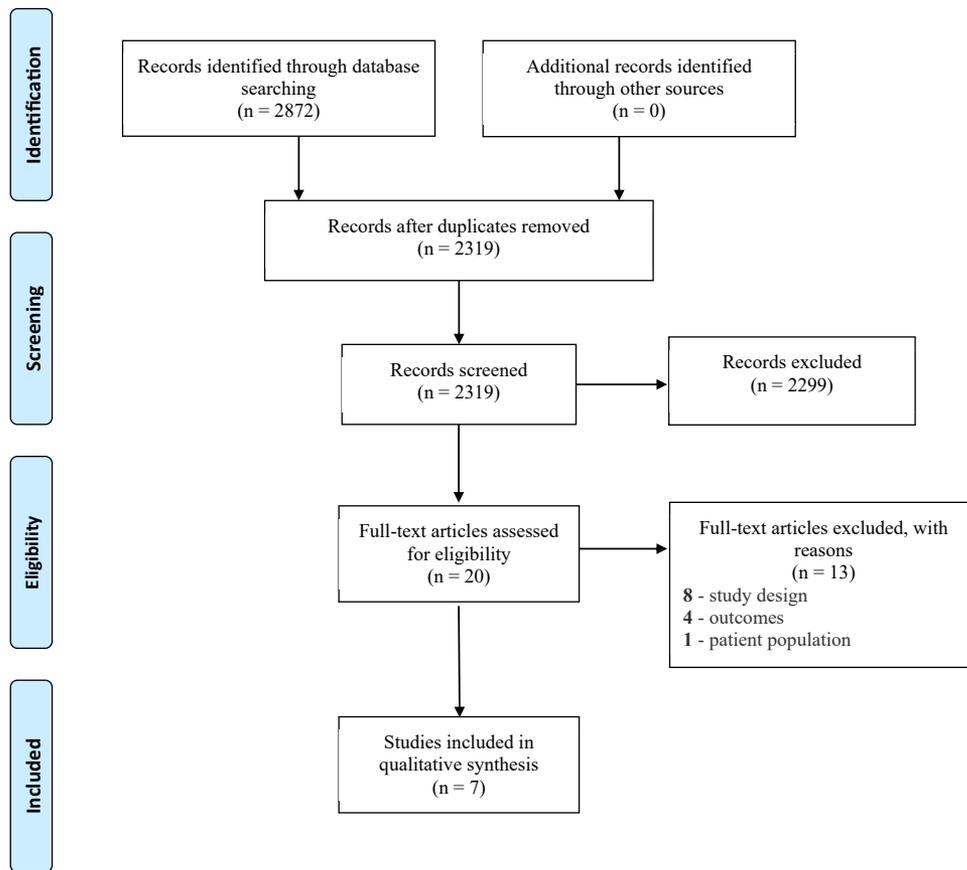


Fig. 1. PRISMA diagram. PRISMA = Preferred Reporting Items for Systematic Reviews and Meta-analyses.

patients. No studies addressed the integration of primary palliative care clinicians among ACHD populations. Study findings can be grouped into three domains: ACP, symptomatology, and end-of-life care.

Advance Care Planning

All five of the survey studies assessed ACHD patient and/or ACHD clinician beliefs concerning different aspects of ACP. In two studies, 1% to 29% of ACHD patients recalled participating in ACP,^{9,10} though 50% of clinicians in one of the studies reported offering ACP.⁹ While only 5%–21% of surveyed patients had completed an advance directive,^{11,12} 69%–78% reported a strong interest and desire for ACP and advance directive discussions.^{10–12} Furthermore, 58%–68% of ACHD patients worried about their future quality of life,^{10,13} although, in one study, only 37% of ACHD patients believed their life expectancy to be lower than average.¹⁰

Multiple studies identified existing barriers to ACP in ACHD populations. Steiner and colleagues noted that 42% of patients did not feel sick enough to discuss end-of-life care,¹⁰ whereas in the study by Greutmann et al., 63% and 79% of ACHD clinicians did not believe in offering ACP if the patient's estimated life expectancy was greater than 5 or 10 years, respectively.¹³

However, a study by Deng et al. reported that 78% of ACHD patients preferred to initiate ACP while still healthy and before hospitalization.¹²

One single-site, retrospective cohort study of 48 ACHD patients noted that ACP conversations were documented among 10% of patients ($n = 5$) and 41% of surrogates ($n = 21$).¹⁴ Yet, in another retrospective cohort study involving 65 ACHD patients cared for in multiple hospitals across a single geographic region, 42% had a documented advance directive and, as compared to patients with nonhematologic cancers, ACHD patients were more likely to have documentation of ACP in their health records (RR 1.46, 95% CI 1.09–1.96).¹⁵

Symptomatology

Two studies examined ACHD patient symptoms as related to their overall health. In the study by Tobler et al., patients completed a cross-sectional survey that included the SF-12, a validated questionnaire evaluating perceived mental and physical health status.⁹ Both the physical (49) and mental score (48) components of the SF-12 were within 1 point of the general U.S. population SF-12 averages.⁹ Deng and colleagues administered the Hospital Anxiety and Depression

Table 2
Summary of Included Studies

First Author	Study Objective	Study Design	Sample Description	Setting	Outcome Measures	Results
Tobler et al., 2011, Canada ¹⁴	To describe end-of-life preferences of an ACHD cohort	Retrospective	N: 48 Diagnosis: ACHD with death due to cardiac causes Age (mean): 37 Gender: F-42% Race: NR	Single hospital	<ul style="list-style-type: none"> • End-of-life discussions • End-of-life services 	End-of-life discussions occurred with five patients, and 21 (41%) surrogate decision-makers. Patients with documented end-of-life conversations were less likely to have attempted resuscitation (12% vs. 100%, $P < 0.001$).
Steiner et al., 2018, USA ¹⁵	To describe end-of-life care among ACHD patients	Retrospective	N: 65 (ACHD), 10,784 (cancer), 3809 (heart failure) Diagnosis: ACHD, cancer, heart failure Age (mean): 45 (ACHD), 63 (cancer), 71 (heart failure) Gender: F-38.5% (ACHD), 44.6% (cancer), 40.5% (cancer) Race: white-88.5% (ACHD), 84.5% (cancer), 78.3% (heart failure). Black-1.9% (ACHD), 4.5% (cancer), 8.6% (heart failure)	Multihospital in Seattle	<ul style="list-style-type: none"> • ICU admission • Location of death • Documentation of advance care planning 	Among ACHD patients, 40% had ICU admission, 64% died in the hospital, and 42% had documentation of an advance care planning document. ACHD patients were more likely to have an ICU admission (RR-1.57, 95% CI 1.12–2.18), die in the hospital (RR-1.75, 95% CI 1.43–2.13), and have documentation of advance care planning (RR 1.46, 95% CI 1.09–1.96) compared with the cancer cohort. In-hospital death, ICU admission, and advance care planning were similar among the ACHD and heart failure cohort.
Tobler et al., 2010, Canada ⁹	To explore patient and clinician preferences and experiences toward end-of-life-specific ACP	Cross-sectional survey	N: 200 (patients), 48 (clinicians) Diagnosis: ACHD Age (mean): 35 (patients), ≤ 45 (clinicians) Gender: F-48% (patients), 44% (clinicians) Race: NR	Outpatient clinic	<ul style="list-style-type: none"> • End-of-life planning discussion • Preferences for timing of end-of-life communication 	2 (1%) patients discussed end-of-life planning with the medical team, while 24 (50%) of clinicians reported they routinely discuss end-of-life care with patients. 156 (78%) patients thought that the

Tobler et al., 2012, Canada ¹¹	To explore knowledge regarding advance directives among the ACHD population	Cross-sectional survey	N: 200 (patients), 48 (clinicians) Diagnosis: ACHD Age (mean): 35 (patients), ≤45 (clinicians) Gender: F-48% (patients), 44% (clinicians) Race: NR	Outpatient clinic	<ul style="list-style-type: none"> • End-of-life planning discussion • Knowledge about advance directives 	<p>medical team should discuss end-of-life planning.</p> <p>Clinicians were more likely to discuss end-of-life planning with patients who have more complex cardiac lesions.</p> <p>70% of patients were interested in learning more about life expectancy.</p> <p>5% of patients had completed an advance directive and 56% had never heard of it.</p> <p>87% of patients thought an advance directive is important.</p>
Greutmann et al., 2013, Canada ¹³	To explore factors that affect ACP in the ACHD population	Cross-sectional survey	N: 200 (patients), 48 (clinicians) Diagnosis: ACHD Age (mean): 35 (patients), ≤45 (clinicians) Gender: F-48% (patients), 44% (clinicians) Race: NR	Outpatient clinic	<ul style="list-style-type: none"> • End-of-life planning discussion • Preferences for timing of end-of-life communication 	<p>24% of patients were not ready to discuss advance care planning, and higher likelihood of avoidance with greater lesion complexity.</p> <p>63% of clinicians thought patients were not ready for end-of-life discussions if estimated life expectancy >5 years and 79% if >10 years.</p>
Deng, et al., 2017, USA ¹²	To explore ACHD patient preferences for ACP	Cross-sectional survey	N: 152 (patients) Diagnosis: ACHD Age (mean): 33 Gender: F-50% (patients) Race: white-83%	Outpatient clinic	<ul style="list-style-type: none"> • Advance care planning discussion • Preferences for timing of end-of-life communication 	<p>13% of patients had prior advance care planning conversations with physicians and 21% completed advance directives.</p> <p>78% of patients reported a preference for initiation of advance care planning conversations while healthy.</p> <p>61% of patients desired estimated life expectancy information.</p>

(Continued)

Table 2
Continued

First Author	Study Objective	Study Design	Sample Description	Setting	Outcome Measures	Results
Steiner et al., 2018, USA ¹⁰	To explore ACHD patient preferences and barriers toward ACP	Cross-sectional survey	N: 150 Diagnosis: ACHD Age (mean): 37 Gender: F-54% Race: white-85%	Outpatient clinic	<ul style="list-style-type: none"> Advance care planning preferences 	<p>28.7% had participated in prior advance care planning and 69.1% of patients were interested in participating in advance care planning.</p> <p>37.4% of participants believed their life expectancy was lower than average.</p> <p>The most common barrier to advance care planning was not feeling sick enough to discuss end-of-life care with a physician (42% of participants).</p>

ACHD = adults with congenital heart disease; ACP = advance care planning.

Scale (HADS) to their ACHD patient cohort.¹² Among the study participants, 46% ($n = 67$) had elevated anxiety symptoms (HADS-A ≥ 8) and 11% ($n = 15$) had elevated depressive symptoms (HADS-D ≥ 8). In addition, univariate analyses revealed that higher ratings for the importance of ACP were noted among patients with elevated anxiety symptoms ($P = 0.001$).¹²

End-of-Life Care

In a single-site retrospective cohort study, ACHD patients who had a documented goals of care conversation before cardiac decompensation, as compared to those who did not, had a lower incidence of resuscitation and aggressive treatments (12% [$n = 3$] vs. 100% [$n = 22$], $P < 0.001$).¹⁴ In a different multisite retrospective study of ACHD patients, 40% had an ICU admission in the last 30 days of their life and 64% died in the hospital.¹⁵ As compared to patients with nonhematologic cancers, patients with ACHD were more likely to have an ICU admission (RR 1.57, 95% CI 1.12–2.18) or to die in the hospital (RR 1.75, 95% CI 1.43–2.13).¹⁵

Discussion

In this systematic review of ACP and palliative care among adults with ACHD, we identified only seven studies. All studies were observational in nature, and only two were not cross-sectional. In addition, five of the studies were coauthored by Kovacs and colleagues, and two were coauthored by Steiner and colleagues, emphasizing the paucity of researchers in this important area. To date, no prospective studies on ACP in ACHD are available. However, other cardiac conditions, such as heart failure (HF), have more data guiding palliative interventions; a recent systematic review of palliative care in heart failure populations had a total of 15 studies, of which seven were randomized controlled trials.¹⁶ These findings highlight the lack of available evidence-based data to inform ACP and palliative care practices in the ACHD population.

Across the reviewed studies, patients expressed a strong interest in ACP, yet only a small percentage of patients had completed an advance directive; interestingly, this finding differs markedly from previous studies in heart failure populations.^{17,18} In one Minnesota cohort study, 41% of heart failure patients had completed an advance directive at study enrollment.¹⁷ The low prevalence of advance directive and ACP discussions among ACHD patients illustrate an existing need for more and better strategies to educate patients and clinicians about these important topics. For example, to address the low prevalence of ACP among pediatric and adult congenital heart disease patients, a heart failure clinic in Texas instituted a rigorous quality improvement ACP training program for clinicians¹⁹; after one year of education, training sessions, and

Table 3
Key Points

- Few ACHD patients and ACHD clinicians have participated in advance care planning conversations (1% to 28.7%).
- Few ACHD patients complete advance directives (5%–21%).
- Most surveyed ACHD patients are interested in discussing goals of care in the outpatient setting with their ACHD team and expressed concern about their quality of life in the future.
- The most common barrier to advance care planning was not feeling sick enough to discuss end-of-life care with a physician (42%).
- Many ACHD clinicians (69%) noted reluctance to initiate ACP conversations among patients with expected life expectancy greater than 5 years.
- Among ACHD patients, there are currently no randomized controlled trials concerning ACP and palliative care and no studies evaluating presence and/or effectiveness of primary palliative care.

ACHD = adults with congenital heart disease; ACP = advance care planning.

standardization of ACP discussions and documentation, the number of advance directives increased from 0% to 42%. A similar approach could be taken in other ACHD and primary care clinics, with the hope of increasing ACP awareness, completion of advance directives, and overall improvement in communication.

While our findings support that many ACHD clinicians recalled initiating ACP discussions, reluctance to initiate conversations was noted for ACHD patients expected to live longer than five years. Reasons for this clinician reluctance included the lack of clinician comfort with initiating end-of-life conversations, difficulty prognosticating end-of-life likelihood for ACHD patients, and clinician concern that such discussions might minimize patient hope and trigger patient depression.²⁰ However, other data from patients with HF support that early ACP is actually associated with improved quality of life measures, reduced hospital admission, and decreased patient anxiety, without any significant effect on mortality.²¹ For example, the PAL-HF study randomized heart failure patients to usual care vs. an interdisciplinary palliative care intervention, and after 6 months of the intervention, the study group receiving proactive palliative care reported higher quality of life metrics, less depression, less anxiety, and greater spiritual well-being.²² Although that study focused exclusively on HF patients, its intervention and successful results might reasonably be applied to the ACHD population.

Unlike the general adult HF population, ACHD patients are unique in that they die at a much younger age, live their entire lives with chronic illness, and many have parents as surrogate decision-makers. In addition, the ACHD population is heterogeneous and each patient needs to be individualized when considering appropriate timing of ACP discussions. An asymptomatic patient with a repaired secundum atrial septal defect is very different from a patient with a failing Fontan with multiorgan dysfunction. Moreover, ACP conversations may have potential negative effects including difficulty understanding and accepting the prognosis, lack of understanding of end-of-life care and life-sustaining treatment, reluctance to engage with the health system, and disagreement among patients and family members.^{23,24} Hence, novel approaches to palliative care need to be considered as

some of the traditional models for adult HF patients may not equally apply for the ACHD population.

Our study generated several important clinical implications for palliative care clinicians (Table 3). Overall, despite expressing interest in ACP, few ACHD patients actually participate in ACP discussions or complete advance directives. The included studies revealed that ACHD clinicians were reluctant to initiate conversations among patients with expected life expectancy greater than five years. Unlike the palliative HF literature, which emphasizes both primary and specialist palliative care, none of the studies in our review evaluated the role of palliative care specialist clinicians in the care of ACHD patients. In addition, the included studies did not highlight the role primary care clinicians may play for ACHD patients. Understanding the palliative care needs for the unique ACHD population is critical not only for ACHD clinicians but also for primary care clinicians providing primary palliative care. This significant gap in the clinical and research literature could be addressed through further research as well as through creation and dissemination of standardized resources and/or guidelines addressing recommended ACP and/or palliative care tailored to ACHD patients.

Our study has several limitations. Although broad, our search criteria may have missed select articles, including those published in a non-English language. Moreover, all our recovered studies were observational and thus subject to potential bias and of limited potential generalizability.

Conclusions

Our findings support that ACHD patients and clinicians are interested in exploring and participating in ACP, yet few ACHD patients actually complete advance directives and many ACHD clinicians note existing barriers to initiating ACP conversations. No existing studies explore the prevalence and/or impact of palliative care specialists in ACHD patient care. Given this lack of data, future studies investigating the integration of palliative care clinicians as well as better ACP communication and implementation strategies tailored to ACHD populations are needed.

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