Original Article

Patterns and Outcomes of Care in Children With Advanced Heart Disease Receiving Palliative Care Consultation

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Abstract

Context. Although access to subspecialty pediatric palliative care (PPC) is increasing, little is known about the role of PPC for children with advanced heart disease (AHD).

Objectives. The objective of this study was to examine features of subspecialty PPC involvement for children with AHD.

Methods. This is a retrospective single-institution medical record review of patients with a primary diagnosis of AHD for whom the PPC team was initially consulted between 2011 and 2016.

Results. Among 201 patients, 87% had congenital/structural heart disease, the remainder having acquired/nonstructural heart disease. Median age at initial PPC consultation was 7.7 months (range 1 day–28.8 years). Of the 92 patients who were alive at data collection, 73% had received initial consultation over one year before. Most common indications for consultation were goals of care (80%) and psychosocial support (54%). At initial consultation, most families (67%) expressed that their primary goal was for their child to live as long and as comfortably as possible. Among deceased patients ($n = 109$), median time from initial consultation to death was 33 days (range 1 day–3.6 years), and children whose families expressed that their primary goal was for their child to live as comfortably as possible were less likely to die in the intensive care unit ($P = 0.03$) and more likely to die in the setting of comfort care or withdrawal of life-sustaining interventions ($P = 0.008$).

Conclusion. PPC involvement for children with AHD focuses on goals of care and psychosocial support. Findings suggest that PPC involvement at end of life supports goal-concordant care. Further research is needed to clarify the impact of PPC on patient outcomes. J Pain Symptom Manage 2018;55:351–358.

Key Words

Pediatric palliative care, pediatric end-of-life care, advanced heart disease, goals of care

Introduction

Pediatric palliative care (PPC) has been associated with improvements in multiple domains of care for children with complex chronic conditions. In fields such as oncology and stem cell transplant, PPC involvement is associated with earlier prognostic[1] and end-of-life (EOL) care[1–3] discussions with families. Additionally, patients receiving PPC have shorter durations of hospital[4,5] and intensive care unit (ICU)[6] stays. Across a variety of conditions, PPC involvement is associated with less suffering due to symptoms,[3,7] better quality of life,[7–9] and fewer invasive interventions.[1,6,10,11]

Historically, PPC teams have rarely been involved in the care of children with cardiovascular diseases,[10,12]

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despite the significant morbidity[13,14] and mortality[15] of these diagnoses. The limited investigation to date into PPC involvement for children with heart disease likely reflects the emphasis on surgical or interventional procedures to "cure" structural heart disease and the variability of the clinical progression and, thus, uncertainty regarding prognosis.[13] Although studies have begun to assess the EOL experience for children with advanced heart disease (AHD) and their families[13,14,16] and explore physician perspectives regarding PPC involvement,[17] little is known about which patients receive PPC and the current role of PPC for this population, not only at EOL but also much earlier in the illness course. In adults with AHD, palliative care involvement has been associated with improvements in symptom burden, depression, and quality of life.[18,19]

This study aims to characterize patients with heart disease who receive subspecialty PPC consultation, describe the circumstances leading to PPC involvement and the interventions provided by a PPC team, and assess the relationship between family goals and patterns of EOL care.

Methods

Patient Population

This was a retrospective medical record review of all patients with a primary diagnosis of AHD for whom the PPC team at Boston Children's Hospital was initially consulted between January 1, 2011 and October 31, 2016. Patients were excluded if 1) the PPC team was unable to complete or document an official visit with the family; 2) the patient did not have a significant cardiac diagnosis or the reason for the consultation pertained to a different primary diagnosis (e.g., severe prematurity); 3) the PPC consultation occurred prenatally without postnatal follow-up; or 4) the patient was older than 30 years at the time of initial PPC consultation as these individuals had limited information available through the electronic medical record system. Patients older than 18 years were included so as to capture the involvement of pediatric providers in the care of young adults with complex childhood-onset disease. The Boston Children's Hospital Institutional Review Board approved the study and waived the requirement for informed consent.

Data Collection

Patients were identified from the PPC team database. Patient medical records were reviewed for demographic and clinical characteristics. In addition to details regarding cardiac diagnosis, measures of lifetime disease burden (i.e., history of gastrostomy tube, tracheostomy, heart transplant, or ventricular assist device [VAD]) were recorded.

Topics addressed during the initial PPC consultation and during follow-up PPC visits were abstracted from consultation notes, which include sections for "goals of care," "decision-making/advance care planning," "symptom management," and "support for the child and family." For cases in which goals of care were discussed during the initial PPC consultation, family goals were categorized as 1) to live as long as possible: the family wishes to pursue life-prolonging interventions even if these may defer focusing on child comfort; 2) to live as long and as comfortably as possible: the family continues to pursue life-prolonging interventions and incorporates comfort and quality of life into their decision-making; and 3) to live as comfortably as possible: the family prioritizes comfort and quality of life and does not consider life-prolongation to be a goal. In addition, features of the consultation (e.g., reason for, timing, and location of consultation) were abstracted from the consultation note. To capture the clinical circumstances leading up to the initial PPC consultation, features of the most recent hospitalization (current hospitalization if consultation occurred while inpatient or last hospitalization if consultation occurred while outpatient) were recorded.

For the subset of patients who died before the end of data collection, characteristics of death and indicators of the EOL experience were recorded. Mode of death was defined based on the following criteria: 1) died during resuscitation: the patient was undergoing cardiopulmonary resuscitation (CPR) when a decision was made to cease; 2) died while receiving mechanical or noninvasive ventilation: respiratory support was not withdrawn but CPR was withheld; 3) died after withdrawal of life-sustaining interventions (LSI): death occurred after the decision was made to withdraw LSI; and 4) died while receiving comfort care: the decision was made to provide comfort measures only, although LSI may have been pursued during a different phase of treatment. Final resuscitation status was determined based on the last official code status in the medical record; absence of documented status was treated as "full code," as per hospital policy.

Statistical Analysis

Statistical analysis was performed using SAS, version 9.4 (SAS Institute Inc., Cary, NC). Descriptive statistics were used to summarize clinical and demographic characteristics. Fisher's exact test was used to determine whether differences in family goals of care were associated with timing of PPC consultation relative to death or EOL outcomes. Two-sided P-values <0.05 were considered statistically significant.