



Review

A part of the team: The changing role of palliative care in congenital heart disease

Naomi Goloff, Brian F. Joy*

University of Minnesota, United States

University of Minnesota Masonic Children's Hospital, United States

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ABSTRACT

Palliative care teams have historically not been involved in the treatment of children with congenital heart disease. When palliative care is involved, most consults occur too late during “crisis” situations when patients are already critically ill, and most had no prenatal involvement of the palliative care team. Early involvement of palliative care teams can reduce patient and family stress as well as allow palliative care to participate in longitudinal care planning and decision making. Palliative care teams should be introduced as part of the larger heart center team to avoid misperceptions that families may have involvement of palliative care is making a judgement on the prognosis of their child. Consideration should be given to developing “triggers” for palliative care consultation to ensure that all patients have access to early palliative care as standard practice, and to help reassure families that a palliative care referral is not synonymous with giving up on their child.

1. Introduction

Pediatric palliative care is medical care that focuses on alleviating the suffering and stress associated with serious illness for children, as well as their families. The aim is to improve quality of life (QOL), facilitate informed decision making and assist with coordination and continuity of care. It is typically provided by a multidisciplinary team of specialists, which can include physicians, nurses, advance practice providers, social workers, child life specialists, chaplains, psychologists, to “provide an extra layer of support” [1] for patients and their families. Tremendous advances have been made in palliative care in the past 20 years, including identifying the role of palliative care for children with serious illness beyond end of life care. As an example, the American Academy of Pediatrics statement on palliative care for children highlights the recommendation for an integrated model of palliative care that continues throughout the course of a serious illness [2], recommends that disease directed therapy and the extra support of palliative care continue concurrently, rather than a mutually exclusive model.

Although few studies have defined the role of palliative care for children with congenital heart disease (CHD), other specialties that care for children with serious illnesses and chronic conditions show that palliative care helps with earlier discussions about prognosis, shorter hospital and intensive care unit stays, less suffering from distressing symptoms and better QOL [3]. Studies focused on adults with advanced

heart disease have shown that involving palliative care helps to improve symptom burden, depression and QOL [3]. Palliative care is also increasingly recognized as being an important part of care in pediatric intensive care units [4].

Palliative care teams have historically not been involved in the treatment of children with CHD [3]. When they have been involved in their care, it was mainly for hospice and when parents have chosen comfort care for a newborn with severe disease [5]. Family interactions with cardiologists, cardiac surgeons, and cardiac intensive care teams often revolve around procedures to “fix” the heart [6], and there is misunderstanding that diseases cannot be treated simultaneously with palliative care involvement [7]. Newborns with severe and immediately life-limiting disease are clear and obvious examples of the role of palliative care, given the expertise in end of life care, but palliative care has a role beyond the end of life period. A recent review article over a 5-year period at a large academic children's hospital reveals the landscape of pediatric palliative care consultations [3]. Most palliative care consults occur too late, with most patients in the intensive care unit, during situations where patients were already critically ill, and most had no prenatal involvement of the palliative care team [3].

The evolution of the surgical and medical management of children with CHD over the last 40 years have resulted in substantial improvements in postoperative survival in CHD with early mortality rates approaching zero [8]. Despite these improvements, one year mortality in patients born with critical CHD remains high at 24.8% [9].

* Correspondence to: B. F. Joy, Department of Pediatrics, University of Minnesota, 2450 Riverside Ave, East Building, MB538, Minneapolis, MN 55454, United States.
 E-mail address: bjoy@umn.edu (B.F. Joy).

Additionally, patients with some of the more complex forms of CHD have frequent and prolonged hospitalizations as well as significant associated comorbid conditions involving the respiratory and gastrointestinal systems [10].

Moreover, advances in medical management have increased the burden of medical care that families and care providers are expected to provide at home. Infants are frequently discharged home from the hospital with feeding tubes, multiple medications and sometimes require respiratory support such as oxygen or in extreme cases chronic ventilation via tracheostomy. This burden of medical care and technology results in significant stressors for families [11].

This review will examine the current practice and data surrounding palliative care involvement in CHD and will make the case for earlier palliative care involvement by including palliative care providers as part of the larger care team. It will utilize a framework focusing on patients with single ventricle heart disease since their disease course and clinical experiences highlight many of the opportunities where palliative care teams can support patients with CHD.

2. Case 1: Robert

Robert was postnatally diagnosed with hypoplastic left heart syndrome (HLHS) when he presented in cardiogenic shock at two days of age. He was critically ill with severe lactic acidosis and seizures. Decision was made to perform a hybrid procedure (bilateral pulmonary artery bands and patent ductus arteriosus stent) to delay the exposure to cardiopulmonary bypass required to perform a Norwood procedure. His initial hospital stay was 26 days but he was readmitted 3 months later after a cardiac arrest during cardiac catheterization. He had a comprehensive stage 2 repair at five months of age and required readmission postoperatively for drainage of a large pericardial effusion. At 18 months of age he had surgery to fix a narrowing in his aortic arch and then had Fontan completion procedure at 4 years of age.

Robert did relatively well for the six years following his Fontan procedure with no hospital admissions or procedures. At 10 years-old, though, he was admitted in decompensated heart failure and was listed for heart transplant. Robert remained hospitalized on intravenous heart failure medications until his heart transplant 10 months later. His pre- and post-transplant course was complicated by behavioral issues secondary to his frustrations with his prolonged hospitalization. By 11 years of age, Robert had undergone 5 cardiac surgeries, 9 cardiac catheterizations, and 8 hospital admissions for a total of 475 days in the hospital.

There are multiple time points that could have prompted palliative care involvement including at initial presentation in cardiogenic shock, cardiac arrest during cardiac catheterization, presentation in heart failure, or listing for heart transplant. In this case, the palliative care team was not consulted until his presentation in heart failure at 10 years of age.

The case illustrates the significant impact and morbidity of CHD. HLHS is one of the most severe forms of CHD and one of a number of cardiac lesions categorized as single ventricle heart disease. Single ventricle heart disease encompasses a wide variability of cardiac pathologies in which there is no way to ‘fix’ or repair the heart back to normal cardiac physiology. The results of this are a minimum of 2–3 palliative surgical procedures, multiple cardiac catheterizations and frequent hospitalizations. In one study, patients had a mean of 10 inpatient admissions, with 85 days in the hospital [10]. Long term sequelae for those that survived the staged palliative procedures include a high disease burden, including arrhythmias, liver dysfunction, protein losing enteropathy, lymphatic dysfunction, respiratory problems and heart failure. Additionally, adolescents with Fontan circulation have been found to be at risk for a lower QOL when compared to a healthy population [12].

This case also highlights the unpredictability of CHD, particularly single ventricle heart disease. Ambiguity and uncertainty have been

found to be stressors for parents of children with CHD but interestingly, unpredictability was not related to increased parenting stress [13]. The unpredictability of CHD does argue for earlier involvement of palliative care teams so that they have established relationships with patients and families in order to easily re-engage during these ‘crisis’ situations. Palliative care teams can fill many roles in patients with serious illness: advance care planning, continuity of care, emotional support, bridging conflicts between parents and medical team, helping with medical decision making, anticipatory guidance, aligning goals of care and medical information, especially when the prognosis is unclear [14]. Palliative care can play a key role in aligning goals of care and medical information.

2.1. Barriers and Timing of Palliative Care Consultation

Pediatric palliative care should be a natural fit for patients with CHD, with improving survival but still with high morbidity and mortality. However, there continues to be many barriers to palliative care involvement. Clinicians are often misinformed about the nature of palliative care, assuming that it is synonymous with comfort care [7]. During prenatal counseling, care for infants with critical CHD is often described as “palliative versus intensive care,” implying an either-or scenario, and further promoting the idea that palliative care solely means comfort care or “non-intervention” [15]. Studies have investigated unmet holistic needs of children with life-threatening illness and their families [16], but few have specifically focused on children with serious CHD. There is much focus on “curing” the heart disease but not necessarily as much weight given to QOL. There is little understanding that curing a disease and QOL are not mutually exclusive. Given these misperceptions, clinicians are often hesitant to refer patients to palliative care because of concern that referral will undermine parental hope if done too early, or that parents will think the clinician is “giving up” on the child [17].

Although few data exist to guide optimal palliative care consult timing for patients with serious CHD, data from pediatric oncology show that most families are open to palliative care involvement at diagnosis, and feel it would be helpful early in course of illness [18]. A multi-center survey of cardiologists and cardiac surgeon attitudes toward palliative care showed that 85% thought that palliative care consults were helpful, and 60% thought that the consults occurred too late [17]. Multiple adult and pediatric studies in other patient populations show that there is little downside to earlier palliative care involvement: studies show that patients live just as long (or sometimes longer), and have improved QOL, with palliative care involvement [19–21]. Over the past few years there has been a trend toward earlier palliative care consultation primarily seen in serious and life-threatening illnesses in the general pediatric population [16].

2.2. Early Palliative Care Consultation

Many palliative care consults occur during crisis situations once patients are already critically ill, but consults in crisis do not allow for the benefits of a longitudinal relationship [3,21]. For pediatric patients with serious illness there is a role for ongoing palliative care involvement during the entire illness [16]. Those who practice palliative care maintain that it is possible to treat an underlying disease, even with an intent to cure, and have a palliative care consultation, as palliative care focuses on much more than just end of life. Families often continue to hope for life-prolongation concurrently while receiving palliative care. Palliative care can support hope for life-prolongation while at the same time supporting overall wellbeing, alleviating suffering, and helping to make decisions aligned with patients' and families' goals [3,16]. The role of palliative care is not just when goals of care have shifted toward comfort or if death is near; there is an important role for palliative care in CHD regardless of prognosis.

A review of palliative care consultations for children with serious

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