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## The role of palliative care in critical congenital heart disease

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## ABSTRACT

Patients with critical congenital heart disease are exposed to significant lifetime morbidity and mortality. Prenatal diagnosis can provide opportunities for anticipatory co-management of patients between palliative subspecialists and the cardiac care team. The benefits of palliative care include support for longitudinal decision-making and avoidance of interventions not consistent with family goals. Effectively counseling families requires an up-to-date understanding of outcomes and knowledge of provider biases. Patient-proxy reported quality of life (QOL) is highly variable in this population and healthcare providers need to be aware of limitations in their own subjective assessment of QOL.

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### Background

The term “critical congenital heart disease” refers to a set of conditions affecting a cohort of patients with congenital heart disease (CHD) who require a surgical or trans-catheter intervention before initial discharge from the hospital after birth.<sup>1</sup> It is useful to focus on this population when discussing the role of palliative care in CHD as these patients have more clinically significant lesions associated with lifelong morbidity and risk of mortality and so can be considered to have life-threatening illness (LTI).<sup>2</sup> This population experiences frequent clinical interventions and recurrent hospitalizations. Children with LTI often receive fragmented, reactive care

with poor care coordination and insufficient family involvement.<sup>2</sup> Importantly however, patients with critical CHD differ from some other populations of children with LTI in that anatomical cardiac anomalies are associated with predictable physiological derangements and in many cases, a more predictable and well-described natural history.<sup>3–10</sup> This provides clinicians caring for these children with an opportunity to fully discuss current outcomes and prepare families and patients for what the future will likely hold. In the current era, in the developed world, many cardiovascular lesions are prenatally diagnosed by ultrasound, providing opportunities for longitudinal decision-making and involvement of palliative care with the patient and family.<sup>11</sup> The purpose of this

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article will be to broadly discuss the role of palliative care in the management of this cohort of patients.

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### **Prenatal diagnosis and counseling**

Prenatal screening for major CHD has evolved from targeted screening of families with a known risk factor (such as an affected relative of the fetus) to general population-based screening.<sup>11</sup> This alteration in screening came about because of the absence of known parental risk factors in the vast majority of cases of CHD. Reported rates of prenatal detection of critical CHD are as high as 91% in targeted populations in some studies.<sup>11</sup> This practice change has several important implications. Prenatal diagnosis of CHD improves outcomes by reducing mortality and morbidity in affected fetuses.<sup>12-15</sup> Recognition of significant CHD facilitates delivery planning at an appropriate center and early, appropriate resuscitation, which may reduce family stress and trauma, although not all parents experience this effect.<sup>16</sup> Early detection of CHD also allows for prenatal counseling and family grief and acceptance as well as pregnancy termination if an acceptable outcome is unlikely.<sup>11</sup>

Limited data exist on what constitutes effective prenatal counseling for this patient population, suggesting the need for more insight into what constitutes effective counseling approaches. Studies evaluating prenatal counseling in other at-risk fetal populations (extreme preterm delivery) show that focusing on review of complications and outcomes may not help a significant proportion of parents, with parents' ultimate decisions regarding resuscitation being based on hope, faith, coping, and personal assessment of risk.<sup>17,18</sup> The importance of this observation in patients with critical CHD is highlighted by a study of 240 families with a prenatal diagnosis of Hypoplastic Left Heart Syndrome (HLHS) that showed roughly equal distribution of termination and decisions not to intervene between standard risk and high-risk fetuses, suggesting that factors other than short-term outcomes drove decision making.<sup>17</sup>

The content of prenatal counseling, particularly as regards to pregnancy termination or the decision to pursue non-interventional care, varies considerably between individual providers and across institutions. A well-documented era-effect in the literature describes declining numbers of families opting to forgo interventions as surgical outcomes have improved and enthusiasm for interventions has increased. This trend is well described for babies with HLHS.<sup>18,19</sup> Variation in practice spans the spectrum from routine discussion of an option for non-intervention (compassionate terminal care), through a "trial of therapy," to not presenting the option for non-intervention at all. Furthermore, semantic imprecision complicates this landscape with available literature describing "palliative" vs. "intensive" care, promoting the notion that palliative care means non-intervention.<sup>20</sup> As palliative care focuses on longitudinal support through illness and decision-making as well as end-of-life care, a different approach to language describing prenatal options would likely reduce confusion and benefit families as well as staff. In addition, it is likely that the content, methods, and biases of those providing counseling have a meaningful effect

on decision-making. A study of 20 mothers of fetuses with critical CHD showed that those who chose non-intervention perceived that providers had negative attitudes towards surgery, while those who chose intervention viewed it as the only choice.<sup>19</sup> Providers who care for acutely ill neonates and children (e.g., neonatology and cardiac critical care) likely overemphasize treatment burden in their counseling, whereas cardiologists and surgeons who have a more longitudinal view of survivors possibly overemphasize success.<sup>21</sup> Multidisciplinary counseling (e.g., cardiologist, perinatologist/neonatologist, and palliative care physician versed in CHD patients) may help to provide a more balanced discussion of outcomes and expectations than counseling by a single discipline in isolation.

Even as outcomes have improved, surgical and catheter-based interventions involve physiological compromises associated with important neurodevelopmental and other longer-term morbidities and risk of mortality.<sup>22-25</sup> For example, the extension of life gained by surgical single ventricle palliation has come, for many patients, with the morbidities of "failing Fontan physiology."<sup>26</sup> Many of these morbidities pose clinical management challenges and have a significant impact on quality of life. For this reason, we believe that non-intervention remains a rational and ethical choice for single ventricle lesions and should be routinely offered for lesions that are particularly difficult to palliate successfully—e.g., prenatal diagnosis of HLHS with an intact atrial septum; or a postnatal diagnosis of pulmonary atresia/intact ventricular septum (PA/IVS) with a right ventricle dependent coronary circulation (RVDC).

The early involvement of palliative care for fetuses or newborns with more significant cardiac lesions allows palliative care providers to establish a relationship with the family and provide continuity between the obstetrical and pediatric environments, supporting both short and longer-term decision making concurrently with other key subspecialists. For known high-risk patients (premature, low birth weight, and extra-cardiac anomalies), early discussion of goals may help avoid burdensome interventions, frame possible end-of-life decisions, and allow parallel planning. Collaboration with palliative care specialists may also promote acquisition of primary palliative care skill and improve communication training for cardiologists, surgeons, and critical care providers. This is congruent with a philosophy of care that unifies the goals of curing and comforting.<sup>2</sup>

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### **Effect of advances in care and survival**

The treatment of CHD is an area that has seen substantial innovation.<sup>27</sup> These innovations involve both technical modification of surgical technique and the increasing prevalence of the "team concept" of care delivery in the form of congenital heart centers that concentrate specialists and create unified, integrated approaches to the longitudinal care of patients with CHD.<sup>28</sup> The most visible effect of this innovation is substantial improvements in survival and the salvage of patients with lesions once thought to be incompatible with life.<sup>29</sup> Less visible, but equally important, are the increasing use of minimally invasive techniques and a more nuanced understanding of the morbidity suffered by patients

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