


Provider perspective on the role of palliative care in hypoplastic left heart syndrome

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Original Article

Cite this article: Lowenstein S, Macauley R, Perko K, and Ronai C (2020) Provider perspective on the role of palliative care in hypoplastic left heart syndrome. *Cardiology in the Young* **30**: 377–382. doi: [10.1017/S1047951120000128](https://doi.org/10.1017/S1047951120000128)

Received: 15 August 2019
Revised: 26 November 2019
Accepted: 11 January 2020

Keywords:

Hypoplastic left heart syndrome; single ventricle defect; palliative care; goals of care; prenatal diagnosis

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Abstract

Objective: Hypoplastic left heart syndrome is a single ventricle defect. While staged surgical palliative treatments have revolutionised care, patients with hypoplastic left heart syndrome continue to have significant morbidity and mortality. In 2017, the National Pediatric Cardiology Quality Improvement Collaborative recommended all single ventricle patients to receive a prenatal palliative care consult. This study aimed to elucidate provider perspectives on the implementation of prenatal palliative care consults for families expecting a child with hypoplastic left heart syndrome. **Methods:** An online survey was administered to obstetric and paediatric providers of relevant disciplines to assess their experience with palliative care involvement in hypoplastic left heart syndrome cases. **Results:** Nearly, all physicians (97%) and most registered nurses (79%) agreed that the initial palliative care consult for patients with hypoplastic left heart syndrome should occur during the prenatal period. Respondents also indicated that prenatal palliative care consults should also be offered in a variety of other CHD conditions. Participants believed positive aspects of this new referral protocol included an expanded support network for families, decreased family stress during the postnatal period, increased patient education about what to expect during the postnatal period, and continuity of care. **Conclusion:** Multidisciplinary healthcare professionals believe that prenatal palliative care consults provide a variety of benefits for patients and families with hypoplastic left heart syndrome. Additional, multi-centre research is necessary to evaluate whether prenatal palliative care consults should become standard of care for families expecting a child with a single ventricle defect.

Hypoplastic left heart syndrome is a complex CHD that occurs in 0.016 to 0.036% of live births, with total CHD prevalence being approximately 9 per 1000 live births.¹ Currently, children with hypoplastic left heart syndrome have the highest risk of mortality among CHD patients. The majority of hypoplastic left heart syndrome patients undergo staged surgical palliation, which consists of three operations, usually prior to 4 years of age.^{1,2} The surgical treatments available have revolutionised care for hypoplastic left heart syndrome patients. For example, over the past three decades, hospital survival for the Norwood procedure has improved from 0 to over 90% at experienced centres. Patients and families, however, continue to endure lifelong complications, including, but not limited to, arrhythmias, protein-losing enteropathy, plastic bronchitis, and poor functional status.³

Paediatric palliative care is a discipline focused on relieving suffering and optimising quality of life in children with chronic, critical, or terminal illnesses. These services represent a burgeoning field that has been shown to shorten the duration of hospital stays, decrease suffering, reduce the number of invasive procedures, and ultimately, improve the quality of life for children.^{4–7} The American Academy of Pediatrics supports an integrated model of palliative care “in which the components of palliative care are offered at diagnosis and continued throughout the course of illness, whether the outcome ends in cure or death”.⁸ This statement advocates that patients and families engage early in goals of care discussions with medical providers.

Paediatric palliative care services commonly support children and families with genetic, neuromuscular, and oncological conditions.⁴ While most children's hospitals offer comprehensive medical treatment plans for patients and families with hypoplastic left heart syndrome, the involvement of paediatric palliative care services in CHD is not common practice.^{9–11} A recent review of the evolving incorporation of paediatric palliative care with paediatric cardiology emphasised the importance of developing interdisciplinary teams to improve the integration of palliative care into the care of children with heart disease.¹² Another study evaluated the role of postnatal paediatric palliative care for children with advanced heart disease and found that paediatric palliative care involvement at the end of life lead to goal concordant care for patients and families.¹³ For families expecting a child with hypoplastic left heart syndrome, however, engaging in goals of care discussions during the prenatal period may allow them more time

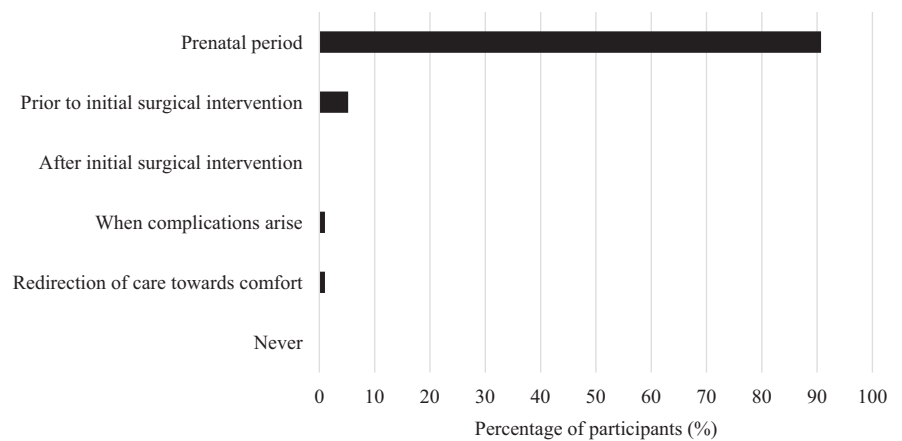


Figure 1. Graphical representation of respondent recommendations for the timing of the initial paediatric palliative care consultation for hypoplastic left heart syndrome patients and families.

to learn about the structural and functional aspects of hypoplastic left heart syndrome, the treatment options available, and the long-term outcomes.^{11,14,15}

At the recommendation of the paediatric cardiology department, in 2017, the Bridges Palliative Care Program at Doernbecher Children's Hospital in Portland, Oregon expanded their service to include automatic prenatal paediatric palliative care consultations to families with a fetus diagnosed with hypoplastic left heart syndrome. The impact of this novel prenatal referral protocol has not yet been evaluated.

The primary aims for this study were to determine provider perspectives on the involvement of paediatric palliative services in the care of patients with hypoplastic left heart syndrome, assess provider perspectives on the timing for consults to paediatric palliative care for patients with hypoplastic left heart syndrome, and determine whether the involvement of paediatric palliative care in the prenatal period alters providers' perspective of the treatment course for patients with hypoplastic left heart syndrome. We also sought to identify additional CHD conditions that may benefit from prenatal paediatric palliative care consults.

Materials and methods

Study participants

Study participants included physicians, nurse practitioners, physician assistants, registered nurses, genetic counselors, social workers, and child life specialists from the fetal therapy, paediatric cardiology, paediatric cardiothoracic surgery, neonatal ICU, and paediatric ICU departments at Doernbecher Children's Hospital and Oregon Health & Science University. There were 320 providers, of a variety of training backgrounds, who were invited to participate starting in July 2018, 1 year since paediatric palliative care consults for patients diagnosed with hypoplastic left heart syndrome in the prenatal period had become standard of care at Doernbecher Children's Hospital.

Study procedures

Eligible providers were identified via the secure online Oregon Health & Science University directory. Provider perspectives on the involvement of paediatric palliative care in the care of children and families with hypoplastic left heart syndrome during the prenatal period were elicited via an online survey (Supplementary material). The online survey was administered through REDCap,

a secure application for managing HIPAA compliant online surveys. Participation in the research study (Approved by Oregon Health & Science University Institutional Review Board: # STUDY00018348) was voluntary, informed consent was obtained, and all responses were confidential. The survey had 13 questions and took approximately 10 minutes to complete.

Data analysis

This study aimed to evaluate the attitudes and experiences of multi-disciplinary providers regarding the implementation of prenatal paediatric palliative care consults for families expecting a child with hypoplastic left heart syndrome. Data are reported as frequencies and percentages (%) for categorical variables. To evaluate the impact clinical training, years of clinical experience and number of hypoplastic left heart syndrome the participant has cared for had on participant responses, and bivariate analyses were performed on Statistical Package for Statistical Sciences (Version 25). Statistical significance was assessed using chi-square analyses, with a significance level of 0.05.

Results

The survey was administered to 320 eligible participants, of which 98 consented for participation, and 90 completed the survey, for a response rate of 28.1%. A variety of healthcare professionals responded to the survey (physician $n = 37$, 37.8%; registered nurse $n = 47$, 48.0%; nurse practitioner $n = 6$, 6.1%; other $n = 8$, 8.0%). Of total respondents, 73.5% had more than 5 years of clinical experience, and over 58% had cared for more than 20 hypoplastic left heart syndrome patients.

On a four-tiered Likert-type scale of strongly agree, agree, disagree, and strongly disagree, more than 93% of respondents either strongly agreed or agreed that all patients diagnosed before birth with hypoplastic left heart syndrome should have a paediatric palliative care consult. Most participants agreed that the initial paediatric palliative care consult should occur during the prenatal period for hypoplastic left heart syndrome patients and families (prenatal period $n = 88$, 90.7%; prior to initial surgical intervention $n = 5$, 5.2%; after initial surgical intervention $n = 0$, 0%; when complications in treatment regimen occur $n = 1$, 1.0%; when redirection of care towards comfort is being considered $n = 1$, 1.0%; never $n = 0$, 0%) (Fig 1). Of physician respondents, 97.0% ($n = 36$) indicated that the initial paediatric palliative care consult should occur during the prenatal period, compared with 79.1% ($n = 37$) of

Table 1. Respondent free responses on the positive aspects of implementing prenatal paediatric palliative care consults as standard of care for hypoplastic left heart syndrome patients and families

Theme	Participant responses
Expanded support network for families	<ul style="list-style-type: none"> • “Anytime we expand the support network we provide these families; we are improving the quality of the overall care we offer, and the addition of palliative care in the fetal period for these families has significantly strengthened that foundation of support” • “Families that are able to develop a relationship with the palliative care team at initial diagnosis are able to gain trust and feel supported through their child’s journey” • “Improved psychosocial support for families and connectedness to resources and personnel that they will encounter during their hospitalization” • “Provides family support and then if redirection of care is ever indicated, the palliative care team is already familiar to the family”
Decreased family stress during the postnatal period	<ul style="list-style-type: none"> • “Families have an ability to connect and form relationships with the palliative care team prior to more intense conversations and care requirements in the postnatal period” • “Initiating these care relationships with the families prior to birth is incredibly helpful in the stressful postnatal period, as it allows the parents to lean on an established support network after delivery”
Increased patient education about what to expect and how to prepare for the postnatal period	<ul style="list-style-type: none"> • “Families will have the ability to discuss their hopes for their child and make sure that aligns with their decision to terminate, do palliative care, or proceed with surgical intervention” • “Help guide families through the expected course so they can be as best prepared as possible to make difficult decisions in the postnatal period” • “Helps families understand the concept that their baby will have a chronic condition that will not be ‘fixed’ in the neonatal period”
Continuity of care	<ul style="list-style-type: none"> • “Helps to transition between NICU and PICU care – one team travels with them – provides nice continuity” • “Very helpful that families already know the team if and when hard decisions need to be made about the patient’s care”

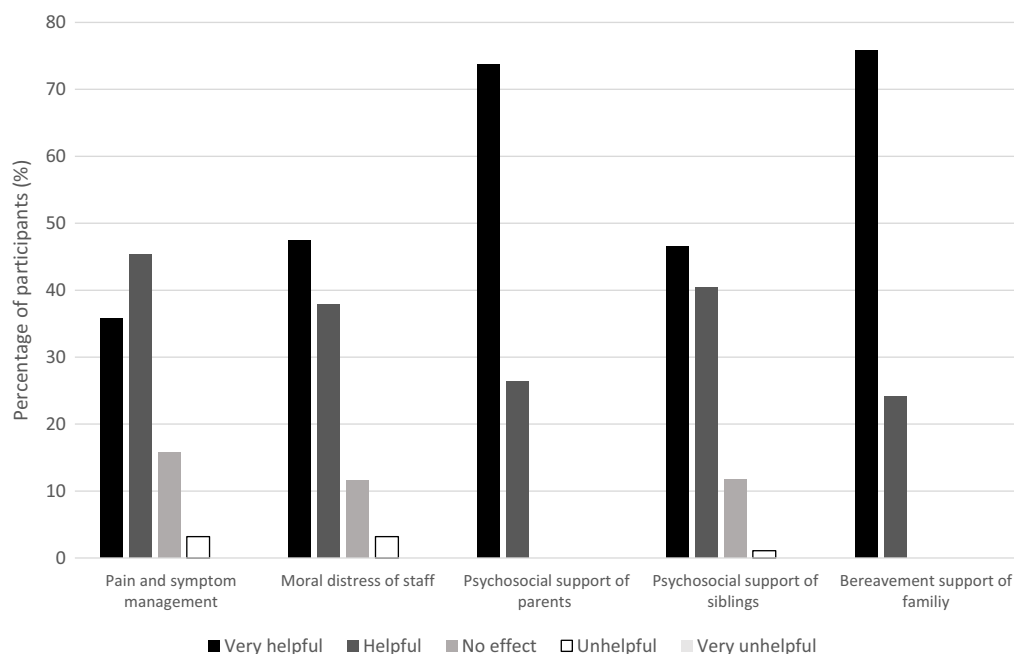


Figure 2. Graphical representation of respondents’ perspectives on the usefulness of paediatric palliative care services.

registered nurse respondents ($p = 0.02$). There was no statistically significant difference in provider responses based on years of clinical experience or the number of hypoplastic left heart syndrome patients they have cared for in terms of the recommended timing for the initial paediatric palliative care consult.

Most respondents strongly agreed that palliative care consults are beneficial for advance care planning for patients and families ($n = 79, 81.4\%$), quality of life for patients and families ($n = 71, 73.2\%$), and transition from hospital to home care ($n = 63, 64.9\%$). Palliative care consults were considered to be very helpful

or helpful for pain and symptom management of the patient ($n = 77, 81.1\%$), moral distress of staff ($n = 81, 85.3\%$), psychosocial support of parents ($n = 95, 100\%$), psychosocial support of siblings ($n = 82, 87.2\%$), and bereavement support of family ($n = 95, 100\%$) (Fig 2).

When asked what participants believe are the positive aspects of this new referral protocol, common themes included an expanded support network for families, decreased family stress during the postnatal period, increased patient education about what to expect and how to prepare for the postnatal period, and continuity of care

Table 2. Respondent free responses on their concerns about implementing prenatal paediatric palliative care consults as standard of care for hypoplastic left heart syndrome patients and families

Theme	Participant responses
Family may interpret the consult as the medical team not exhausting both medical and surgical management options for their child with hypoplastic left heart syndrome (HLHS)	<ul style="list-style-type: none"> • “Some families may misinterpret the concept of introducing the palliative care team as the team ‘not doing everything’ in regards to medical/surgical management of the complex heart disease” • “I think the palliative care team has to coordinate closely with the cardiology and surgical teams in order to make sure that messaging is consistent to the family”

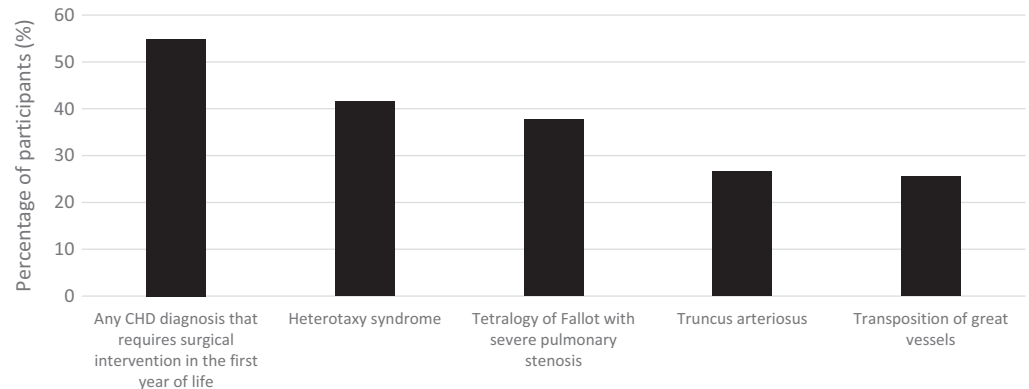


Figure 3. In addition to hypoplastic left heart syndrome, providers recommend other CHD conditions that receive prenatal paediatric palliative care consults.

from the neonatal ICU to the paediatric ICU and beyond (Table 1). When asked about concerns related to the new referral protocol, respondents primarily indicated that the family may interpret the consult as the medical team not exhausting both medical and surgical management options for their child with hypoplastic left heart syndrome (Table 2). The majority of respondents, however, stated no specific concerns with the new referral protocol ($n = 72$, 80.1%).

Most respondents reported that prenatal integration of paediatric palliative care has not altered the treatment course for hypoplastic left heart syndrome patients ($n = 53$, 66.3%). For participants who had observed or implemented modifications in the treatment course for hypoplastic left heart syndrome patients following prenatal palliative consults, the common theme was better informed decision-making for families during discussions about goals of care.

In a multi-select question, respondents indicated that prenatal paediatric palliative care consults should be routinely offered in a variety of other CHD conditions (any CHD diagnosis that requires surgical intervention in the first year of life $n = 45$, 54.9%; heterotaxy syndrome $n = 34$, 41.5%; tetralogy of Fallot with severe pulmonary stenosis $n = 31$, 37.8%; truncus arteriosus $n = 30$, 36.6%; transposition of great vessels $n = 21$, 25.6%) (Fig 3). Of providers who indicated prenatal paediatric palliative care consults should routinely occur for any patient with CHD that requires surgical intervention in the first year of life, 40.0% were physicians ($n = 14$) and 51.4% were registered nurses ($n = 20$). Other suggestions written in by participants included any CHD associated with pulmonary hypertension, dilated cardiomyopathy, hypoplastic right heart syndrome, truncus arteriosus with interrupted aortic arch, and pulmonary atresia with ventricular septal defect and multiple aortopulmonary collaterals.

When asked if including paediatric palliative care consults as standard of care for hypoplastic left heart syndrome patients and families negatively impacts the relationship non-palliative care

clinicians have with the patient and family, 57.8% strongly disagreed ($n = 46$) and 33.3% disagreed ($n = 27$). Nearly, all participants strongly agreed or agreed that implementing required paediatric palliative care consults for fetuses diagnosed with hypoplastic left heart syndrome decreases the barriers for providers to make a referral to paediatric palliative care services ($n = 83$, 88.9%). Almost all participants strongly disagreed or disagreed that palliative care is at odds with simultaneous delivery of therapies directed at cure or long term survival ($n = 84$, 92.2%).

Discussion

This study provides the first assessment of multidisciplinary provider perspective on prenatal involvement of paediatric palliative care in the management of patients and families with hypoplastic left heart syndrome. We demonstrate that providers overwhelming support this new referral protocol as it expands the support network for families, decreases family stress during the postnatal period, and increases patient education about what to expect during the postnatal period, along with providing continuity of care from the neonatal ICU to the paediatric ICU and beyond.

Much of the current literature – which examines postnatal rather than prenatal palliative care consultation – demonstrates provider resistance to early paediatric palliative involvement in a variety of childhood illnesses. In 2013, a focus group-based study showed paediatric oncology providers were concerned early paediatric palliative care involvement might negatively impact their relationships with patients and families by provoking anxiety, creating a sense of diminishing hope, and being at odds with continued pursuit of therapy.¹⁶ In a more recent cross-sectional online survey, nearly 50% of paediatric cardiology providers caring for patients with advanced heart disease believed palliative care services may “undermine parents’ hope” and give the impression that the paediatric cardiologists are “giving up on their child”.¹⁷ A third study

Table 3. Summary of consult recommendations based on survey results gathered at Doernbecher Children's Hospital

Consult specifics	Recommendations
Patient population eligible for consultation	<ul style="list-style-type: none"> • Patients with a prenatal diagnosis of a single ventricle defect
Consultation timing and subsequent follow-up consults	<ul style="list-style-type: none"> • First consultation during prenatal period after the family has met with medical teams • Subsequent consults during every inpatient admission
Content to be addressed during initial consultation	<ul style="list-style-type: none"> • Educate family about the definition of palliative care⁸ • Assess family members understanding of their child's illness with respect to prognosis, treatment options, and goals of therapy • Assess and provide psychosocial support to families
Documentation of consultation	<ul style="list-style-type: none"> • Document consultation in electronic medical record

highlighted a belief among paediatric palliative care providers that paediatric cardiologists may be resistant to recommending palliative care involvement for children with advanced heart disease.¹⁸ Taken together, these studies indicate opportunity for improvement in the interdisciplinary approach to treating children with chronic, critical, or terminal illnesses.

In our study, 97% of physicians agreed that the initial paediatric palliative care consult for hypoplastic left heart syndrome patients and families should occur in the prenatal period. Multidisciplinary providers also noted that paediatric palliative care improved psychosocial and bereavement support for families without negatively impacting the therapeutic relationship. These results favouring prenatal paediatric palliative care consults are in contrast to previous literature demonstrating provider resistance to postnatal paediatric palliative care involvement. The strong support of prenatal paediatric palliative care involvement in hypoplastic left heart syndrome may be due to implementing prenatal paediatric palliative care consults as standard of care at Doernbecher Children's Hospital, thus decreasing the barriers providers face when discussing paediatric palliative care referrals with families, particularly during the prenatal period.

Providers overwhelming agreed that palliative care can coexist with simultaneous pursuit of surgical intervention for hypoplastic left heart syndrome. Most participants reported that they had not appreciated a decrease in families pursuing surgical interventions for children with hypoplastic left heart syndrome since implementation of prenatal palliative care consults at Doernbecher Children's Hospital. Multidisciplinary providers have, however, observed improved informed decision-making during goals of care discussions with families. Beyond hypoplastic left heart syndrome, 40% of physicians suggested that any patient with CHD requiring surgical intervention in the first year of life should receive a prenatal palliative care consult. Our results therefore suggest that surgery and palliative care, beyond being compatible components of therapy, may have a synergistic effect in treating children.

Based on our results, we would advise that prenatal paediatric palliative care consults be available for all hypoplastic left heart syndrome patient and families, as prenatal involvement of paediatric palliative care allows families the time to comprehend a hypoplastic left heart syndrome diagnosis and prepare for the extensive trajectory of care ahead. Initiating paediatric palliative care relationships with the families prior to birth is also a valuable resource for families in the postnatal period, as it allows them to lean on an established therapeutic relationship and support network while they are navigating challenging situations and care decisions.

At our institution, prenatal paediatric palliative care consults occur after fetal therapy, paediatric cardiology, and neonatal ICU

appointments with the family. During those appointments, medical providers introduce prenatal paediatric palliative care consults as standard of care for hypoplastic left heart syndrome and all other single ventricle patients. This protocol has ensured that the paediatric palliative care team is aware of the proposed prognosis and treatment options prior to meeting with the family. In addition to hypoplastic left heart syndrome, children diagnosed with dilated cardiomyopathy, hypoplastic right heart syndrome, truncus arteriosus with interrupted aortic arch, and pulmonary atresia with ventricular septal defect and multiple aortopulmonary collaterals also receive prenatal paediatric palliative care consults. Lastly, following the prenatal paediatric palliative care consults, the paediatric palliative care team is informed by the neonatal ICU, paediatric ICU, and/or paediatric cardiothoracic surgery team when the patients are hospitalised, to ensure continuity of care (Table 3). At present, the paediatric palliative care department at Doernbecher Children's Hospital is growing in size and has been able to accommodate the increased volume of consults.

The primary limitation of this research study is that it was conducted at a single institution. Additional research is required to understand provider perspectives across multiple institutions to ultimately make strong recommendations about whether prenatal palliative care consults for families expecting a child with hypoplastic left heart syndrome should become standard of care. A second limitation is the lack of assessment of parent/guardian perspectives on the prenatal involvement of paediatric palliative care in hypoplastic left heart syndrome. Since our results also reveal immense multidisciplinary provider support of the new referral protocol for hypoplastic left heart syndrome patients and our response rate for the survey was higher than the national average, it may indicate that respondents may have had strong opinions on the topic and thus positively skewed our results.

Future directions include surveying both providers at multiple institutions and parents/guardians on the involvement of prenatal palliative care for hypoplastic left heart syndrome patients. In addition, a retrospective chart review to more accurately determine the treatment modifications that occurred in the care of hypoplastic left heart syndrome patients following the initiation of prenatal paediatric palliative care consults is indicated. This would include a chart review of hypoplastic left heart syndrome patients before and after the referral protocol was implemented in 2017 at Doernbecher Children's Hospital.

In conclusion, multidisciplinary healthcare professionals believe that prenatal palliative care consults provide a variety of benefits for patients and families with hypoplastic left heart syndrome, and we recommend children's hospitals consider making prenatal palliative care consults available for families expecting a child with a single ventricle defect.

Supplementary Material. To view supplementary material for this article, please visit <https://doi.org/10.1017/S1047951120000128>

Acknowledgements. None.

Financial Support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest. None.

Ethical Standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (National Commission for the Protection of Human Subjects of Biomedical and Behavioral Research) and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committees (Oregon Health & Science University Institutional Review Board: # STUDY00018348).

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