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RESEARCH

Initial Counseling Prior to Palliation for Hypoplastic Left Heart Syndrome: 2021 vs 2011

Deipanjan Nandi[1] · Stacey Culp[2] · Andrew R. Yates[1] · Timothy M. Hoffman[3] · Amy L. Juraszek[4] · Christopher S. Snyder[5] · Timothy F. Feltes[1] · Clifford L. Cua[1]

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Abstract

We sought to examine current practices and changes in practice regarding initial counseling for families of patients with hypoplastic left heart syndrome (HLHS) given the evolution of options and outcomes over time. Counseling (Norwood with Blalock-Taussig-Thomas shunt (NW-BTT), NW with right ventricle to pulmonary artery conduit (NW-RVPA), hybrid palliation, heart transplantation, or non-intervention/hospice (NI)) for patients with HLHS were queried via questionnaire of pediatric care professionals in 2021 and compared to identical questionnaire from 2011. Of 322 respondents in 2021 (39% female), 299 respondents were cardiologists (92.9%), 17cardiothoracic surgeons (5.3%), and 6 were nurse practitioners (1.9%). Respondents were largely from North America (96.9%). In 2021, NW-RVPA procedure was the preferred palliation for standard risk HLHS patient (61%) and was preferred across all US regions ($p < 0.001$). NI was offered as an option by 71.4% of respondents for standard risk patients and was the predominant strategy for patients with end-organ dysfunction, chromosomal abnormality, and prematurity (52%, 44%, and 45%, respectively). The hybrid procedure was preferred for low birth-weight infants (51%). In comparison to the identical 2011 questionnaire ($n = 200$), the NW-RVPA was endorsed more in 2021 (61% vs 52%, $p = 0.04$). For low birth-weight infants, hybrid procedure was more recommended than in 2011 (51% vs 21%, $p < 0.001$). The NW-RVPA operation is the most recommended strategy throughout the US for infants with HLHS. The hybrid procedure for low birth-weight infants is increasingly recommended. NI continues to be offered even in standard risk patients with HLHS.

Keyword Hypoplastic left heart syndrome

Introduction

Five initial pathways currently exist for patients born with HLHS: Norwood palliation with a modified Blalock-Taussig-Thomas shunt (NW-BTT), Norwood palliation with a right ventricle to pulmonary artery conduit (NW-RVPA), hybrid procedure (stenting of the ductus arteriosus, bilateral pulmonary artery banding, and balloon atrial septostomy), primary cardiac transplantation, or non-intervention/hospice (NI) care. The advantages and disadvantages of each of these pathways are of some debate, including greater experience of preferred strategies at different institutions, possible survival benefits of the RVPA, and possible neurodevelopmental benefits [1–6].

A 2011 questionnaire of pediatric cardiac providers was undertaken by us. At that time, differences in initial counseling for these families regarding treatment options appeared to vary based on institutional location and patient characteristics. Despite improvements in overall morbidity and mortality for this patient population, our earlier questionnaire reported that nearly half of respondents would offer hospice care as a potential option for all patient with HLHS, even with no other medical issues [7]. Since that questionnaire, a decade has passed with continued refinement in

✉ Deipanjan Nandi
deipanjan.nandi@nationwidechildrens.org

1 Nationwide Children's Hospital, Ohio State University, Columbus, OH, USA

2 Department of Biomedical Informatics, Ohio State University, Columbus, OH, USA

3 University of North Carolina, Chapel Hill, NC, USA

4 Wolfson Children's Hospital, Jacksonville, FL, USA

5 Children's Hospital of Richmond at VCU, Richmond, VA, USA

pre-operative, operative, and post-operative management, as well as a better understanding of long-term outcomes for the various surgical options.

The primary goal of this study was to describe the initial counseling and recommendations provided by caregivers when faced with several anatomic variations or clinical conditions associated with HLHS. The secondary goal was to compare responses with the prior questionnaire administration from 10 years ago.

Material and Methods

Nationwide Children's Hospital Institutional Review Board approved a 25-item questionnaire that was previously developed and internally tested for question clarity in 2011 and used again now [7]. The questionnaire was directed to those providing initial counseling to family members of children with HLHS. Respondents were asked regarding palliative approaches offered at their institution (i.e. NW-BTT, NW-RVPA, hybrid procedure, cardiac transplantation, NI) and their "usual" recommendations for an uncomplicated patient with HLHS. An uncomplicated patient with HLHS was defined as not having any of the co-morbid conditions that are subsequently listed. Individuals were then asked to rank their primary recommendation and options discussed with families in the setting of several anatomic or co-morbid conditions: intact atrial septum, ventricular dysfunction (moderate or greater), tricuspid regurgitation (moderate or greater), low birth weight (< 2 kg), prematurity (< 30 weeks gestational age), chromosomal abnormalities, and other end organ dysfunction. Respondents were also asked to rate the mortality quoted to families (if discussed) for the various staged palliations through Fontan completion, as well as long-term outcomes.

The questionnaire was created via SurveyMonkey and anonymous responses were solicited from the PEDIHEART and American Academy of Pediatrics Cardiology email distribution list. All data were included with missing data reported. Descriptive statistics were used for demographics and baseline case responses. In an effort to eliminate local institutional bias, we repeated all statistical measures while excluding all respondents who self-identified as from the Midwest region and from a center where the primary palliation was the hybrid procedure.

Mean years of practice were compared between 2011 and 2021respondents using an independent-samples t-test. Chi-squared tests of homogeneity were used to compare the distribution of the demographic variables of gender, cardiac specialty, setting, HLHS volume, world region, and US region between 2011 and 2021. World regions were categorized as North America, Europe, and Other to meet the assumptions for the chi-squared test. Chi-squared tests were used to determine if there were differences in proportions of respondents who reported ever offering NW-BT, NW-RVPA, hybrid palliation, primary heart transplantation, and NI care between 2011 and 2021. Chi-squared tests were also used to assess the differences between 2011 and 2021 in the proportion of respondents recommending these interventions amongst the above-listed increased risk comorbidities. Data are presented as means and standard deviations unless otherwise stated. IBS SPSS Statistics Version 28.0 was used for all analyses.

Results

2021 Questionnaire Results

Three hundred twenty-two people (197 male, 125 female) responded in 2021. Not all respondents answered every question; thus, the actual number of responses vary by question. Since the actual number of subscribers who received the e-mail was unknown, a response rate could not be calculated.

Two hundred ninety-nine respondents were pediatric cardiologists (92.9%), 17 respondents were pediatric cardiothoracic surgeons (5.3%), and 6 respondents were pediatric cardiology nurse practitioners (1.9%). Respondents were a median of 11 years (5–22) post their ultimate training. Respondents practiced in a university-based hospital (76.4%) more frequently than in a private hospital (13.4%), private practice (6.8%), or other setting (3.4%). The cohort consisted of pediatric cardiologists who were primarily general pediatric cardiologists (34.5%), echocardiographers (28.0%), cardiac critical care (10.9%), interventionalists (7.5%), transplant (3.7%), electrophysiologists (0.9%), adult congenital (0.9%), and other (8.4%). Respondents were primarily from North America (96.9%) with Europe having 8 respondents and Asia and the Middle East both having 1 respondent. Within North America, respondents self-described their location as: East Coast (21.7%), South (28.0%), Midwest (31.7%), West Coast (15.5%).

Physicians reported the number of patients with HLHS during the preceding year who received their initial palliation at their institution were: 0 (8.4%), 1–10 (30.7%), 11–20 (32.9%), 21–30 (15.5%), 31–40 (5.0%), 41–50 (2.8%), and greater than 50 (4.7%). The respondents offered the NW-BTT (84.2%), NW-RVPA (89.1%), hybrid procedure (77.3%), cardiac transplantation (31.4%), or hospice care (71.4%) as options for care for an uncomplicated patient with HLHS. The primary procedure of choice for an uncomplicated patient with HLHS was NW-RVPA (59.0%), followed by NW-BTT (32.6%), hybrid (4.7%), non-intervention (0.6%), and primary transplant (0.0%) with 3.1% not responding. There was no difference in preferred initial palliation based upon region of the United

States with the NW-RVPA being the preferred procedure ($p < 0.001$). Hospice/non-intervention was offered by 71.4% and transplant by 31.4% of respondents, although no individual respondent offered hospice care or primary transplantation without also offering at least one surgical approach to palliation.

The primary recommendation for palliation and options discussed with families in the setting of the anatomic or co-morbid conditions studied are displayed in Fig. 1. Non-intervention/hospice care was most recommended if a patient had end organ dysfunction (51%) or a chromosomal abnormality (45%). Non-intervention was also the main recommendation if the patient was premature (45%), though the hybrid procedure was also frequently recommended as well (38%). For low birth weight patients, the hybrid procedure was the main recommendation (54%). For patients with tricuspid regurgitation, ventricular dysfunction, or an intact/restrictive atrial communications, recommendations widely varied. If confronted with a fetal diagnosis of intact or highly restrictive atrial communication, only 22.7% of the respondents (n = 73) would refer the patient to a center that performs a fetal intervention.

Expectations for survival after palliation was solicited based upon individual counseling for families based upon several factors: expected mortality after the first palliative approach, interstage mortality, and 2nd stage mortality. Figures 2A–C outline the mortality ranges that physicians quoted caregivers for each stage by each specific surgical approach assuming no major comorbidities.

Two hundred thirty one respondents stated that they counseled families that neurological outcomes after palliation (assuming no major morbidities throughout) were expected to be below average (79% of respondents), or average (20% of respondents) compared to individuals without heart disease. When asked about physical outcomes compared to peers without heart disease, 79% reported below average status compared to 19% reporting average physical outcomes compared to peers without heart disease. The remainder of respondents did not discuss long-term physical or neurological outcomes.

Of note, none of the above comparisons changed significantly when self-identified respondents from the US Midwest with center primary palliation being the hybrid procedure were excluded.

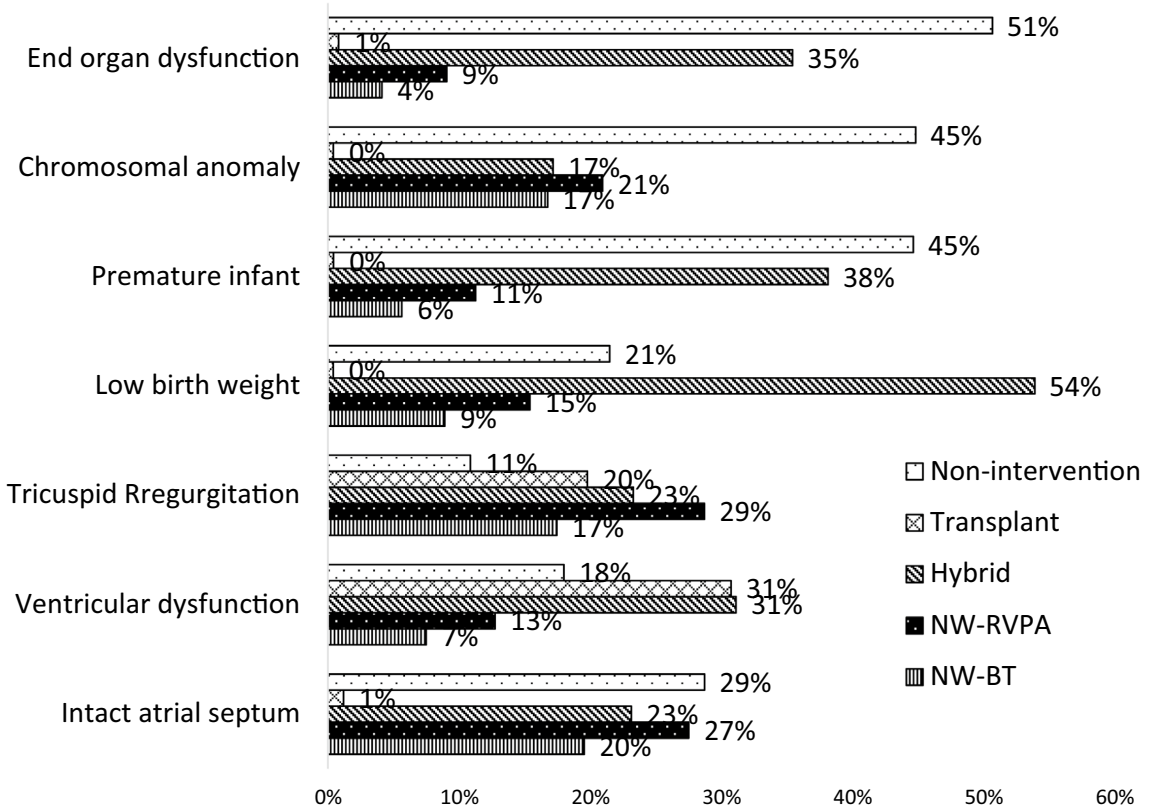

Fig. 1 Primary recommendation based upon anatomic or co-morbid risk factor, defined as end-organ dysfunction, chromosomal abnormalities, pre-maturity less than 30 weeks gestational age, birth weight less than 2 kg, tricuspid regurgitation of moderate or greater degree, ventricular dysfunction of moderate or greater degree, or intact atrial septum. *NW-RVPA* Norwood with right ventricle to pulmonary artery conduit; *NW-BT* Norwood with BT shunt

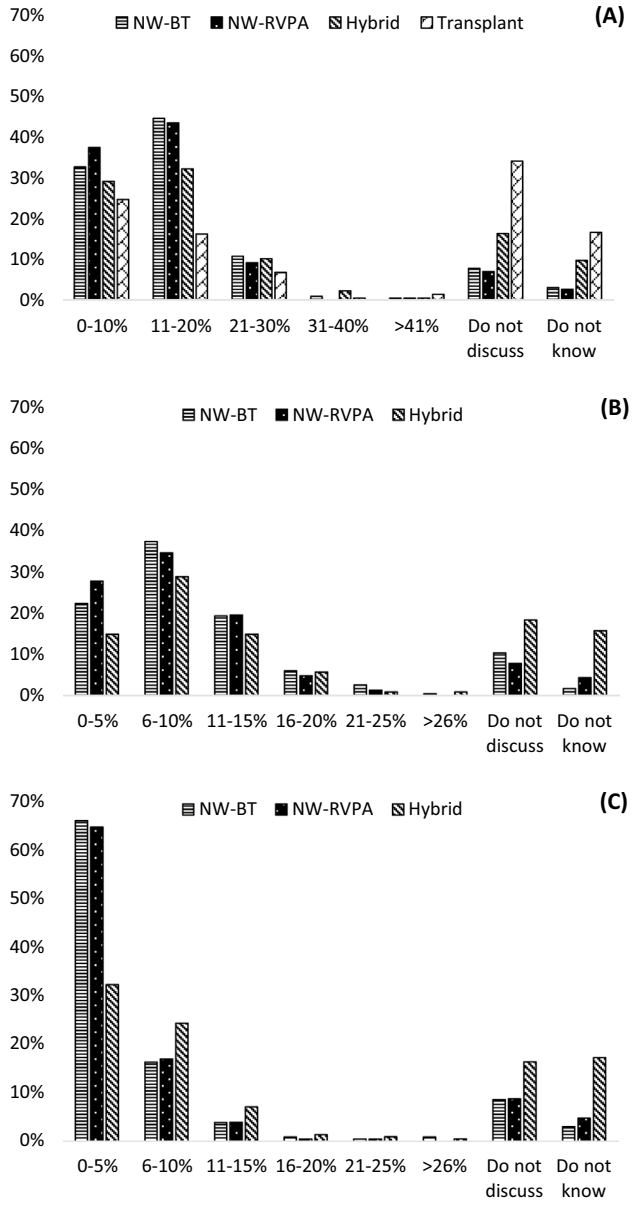

Fig. 2 Percent of respondents counseling of mortality of **A** initial interventions, **B** inter-stage, and **C** post-stage II. *Norwood-BT* Norwood with BT shunt; *Norwood-RVPA* Norwood with right ventricle to pulmonary artery conduit

Comparison to 2011 Questionnaire Findings

When comparisons were made between the questionnaires, respondents from 2021 were in practice significantly longer ($p = 0.03$). There were significantly more female respondents in 2021 ($p = 0.01$). In addition, cardiac specialty and practice setting were significantly different between the two questionnaire years ($p = 0.05$ for both specialty and setting). There was no significant difference in HLHS surgical volume or location of US region amongst the respondents (Table 1).

Table 1 Respondent demographics

Characteristics	2011		2021		p-value
	n	%	n	%	
Total	200		322		
Years of Practice (mean)	12.3		14.3		0.044
Female	42	21.0	125	38.8	< 0.001
Cardiac Specialty					
General	74	37.0	111	34.5	< 0.001
Echocardiography	40	20.0	90	28.0	
Intensive Care	17	8.5	35	10.9	
Interventional	42	21.0	24	7.5	
Cardiothoracic Surgeon	6	3.0	17	5.3	
Transplantation	2	1.0	12	3.7	
Electrophysiologist	11	5.5	3	0.9	
Other	8	4.0	27	8.4	
Setting					
University Hospital	134	67.0	246	76.4	0.001
Non-university hospital	31	15.5	43	13.4	
Private	33	16.5	22	6.8	
Other	2	1.0	11	3.4	
HLHS Volume					
0	28	14.0	27	8.4	0.096
1–10	73	36.5	99	30.7	
11–20	56	28.0	106	32.9	
21–30	20	10.0	50	15.5	
31–40	13	6.5	16	5.0	
41–50	5	2.5	9	2.8	
50+	5	2.5	15	4.7	
World Region					
North America	163	81.5	312	96.9	< 0.001
South America	4	2.0	0	0.0	
Europe	20	10.0	8	2.5	
Asia	11	5.5	1	0.3	
Middle East	0	0.0	1	0.3	
US Region					
East	40	20.0	70	21.7	0.855
Midwest	47	23.5	102	31.7	
South	45	22.5	90	28.0	
West	23	11.5	50	15.5	

HLHS hypoplastic left heart syndrome

The NW-RVPA, hybrid, and hospice care were options which were offered more often in 2021 than in 2011 (Table 2). The NW-RVPA was offered as the primary choice more often for an uncomplicated patient with HLHS in 2021 versus 2011. The hybrid procedure was offered more often as an option for a patient with any co-morbid condition in 2021 versus 2011. The NW-RVPA was also offered as an option more often in 2021 versus 2011 for patients with HLHS and a chromosomal abnormality (Table 2).

Table 2 Changes in procedures ever offered, followed by primary recommendation between 2011 and 2021 based upon anatomic or co-morbid risk factor, defined as end-organ dysfunction, chromosomal abnormalities, pre-maturity less than 30 weeks gestational age, birth weight less than 2 kg, tricuspid regurgitation (*TR)* of moderate or greater degree, ventricular dysfunction of moderate or greater degree, or intact atrial septum

	Year	NW-BTT	NW-RVPA	Hybrid	Transplant	Non-intervention	2021 vs 2011
Ever Offered	2021	84%	89%	77%	31%	71%	$p < 0.001$
	2011	89%	79%	40%	28%	46%	
Non-complicated	2021	34%	61%	5%	0%	1%	$p = 0.066$
	2011	37%	52%	10%	1%	2%	
End organ dysfunction	2021	4%	9%	35%	1%	51%	$p < 0.001$
	2011	13%	8%	12%	1%	68%	
Chromosomal anomaly	2021	17%	21%	17%	0%	45%	$p < 0.001$
	2011	15%	9%	9%	0%	69%	
Premature infant	2021	6%	11%	38%	0%	45%	$p < 0.001$
	2011	20%	11%	22%	0%	47%	
Low birth weight	2021	9%	15%	54%	0%	21%	$p < 0.001$
	2011	30%	21%	24%	0%	24%	
Mod TR	2021	17%	29%	23%	20%	11%	$p < 0.001$
	2011	39%	22%	14%	12%	14%	
Mod ventricular dysfunction	2021	7%	13%	31%	31%	18%	$p < 0.001$
	2011	23%	17%	18%	23%	20%	
Intact atrial septum	2021	20%	27%	23%	1%	29%	$p < 0.001$
	2011	43%	24%	10%	4%	19%	

NW-RVPA Norwood with right ventricle to pulmonary artery conduit; *NW-BTT* Norwood with BTT shunt

Comment

Currently, five pathways exist for patients with HLHS with each having advantages and disadvantages.[1, 2, 4, 7–9]. In our previous questionnaire from 2011, options for and counseling of caregivers of patients with HLHS appeared dependent on regional and institutional preferences as well as any associated co-morbid conditions [7]. In this follow-up study, there appears to be a clear trend across the US for the NW-RVPA as the primary choice for patients with HLHS without co-morbid conditions. Variability continues to exist in recommendations for patients with HLHS that have co-morbid conditions. In spite of improvements in overall morbidity and mortality for this patient population, non-intervention/hospice care is still offered by a majority as an option for uncomplicated patients with HLHS.

Previous studies documented a wide variability in pre-operative management patients with HLHS with no clearly defined "best course" [7, 10–12]. In this questionnaire, it appears that the NW-RVPA procedure has become the procedure of choice across the US, though the NW-BTT procedure is still widely performed. Previously, the NW-RVPA was recommended more often in the US South and West Coast, whereas the NW-BTT was recommended more often in the US East and Midwest for uncomplicated patients with HLHS [7]. The change in counseling in regions and over time for the NW-RVPA may possibly be due to the perceived improved results with the NW-RVPA procedure over the alternative pathways. Some studies have documented overall improved survival with the NW-RVPA procedure [1, 13], though other data show that there is no difference in overall mortality between the NW-RVPA or NW-BT procedure [4].

Though the NW-RVPA procedure appears to be the primary option recommended for an uncomplicated patient with HLHS, there continues to be wide variability when co-morbid conditions co-exist. When there is significant end organ dysfunction, chromosomal abnormality, or pre-maturity, hospice care was the primary recommendation. However, the hybrid procedure was also frequently recommended for premature infants and it was the primary choice for low birth weight patients. For the other possible co-morbid conditions evaluated, there was wide variability in primary recommendations with no clear primary recommendation seen. There has been a clear trend towards the use of the hybrid procedure as recommended pathway for these high-risk patients when compared to the questionnaire in 2011. This is consistent with current publications that show that most institutions perform the hybrid procedure for their high-risk patients with HLHS [1, 9, 14–16]. Only time will determine if these trends continue with the NW-RVPA and hybrid procedures.

The majority of respondents quoted mortality for either the NW-RVPA or NW-BTT procedure as generally highest for the initial procedure and decreasing with subsequent procedures, which is largely consistent with published data [1, 4, 9], and these mortality quotes are similar to the responses in the 2011 questionnaire. However, if 2021 respondents, significant proportions of respondents did not know or discussing mortality of hybrid palliation and subsequent stages. However, 2021 respondents did know and discuss rates of mortality for the hybrid pathway more than in the 2011 questionnaire. This is likely due to the fact that in 2011, the hybrid procedure was a relatively new procedure with limited follow-up data available. Ten years later, more data are available for the hybrid procedure, though it may not be as widely disseminated compared to the more common procedures [6, 15, 17].

Counseling for neurological and physical outcomes for patients with HLHS has not changed over 10 years with the majority of respondents stating that patients with HLHS will likely have some deficits compared to peers. This continues to be consistent with current data available [18–22]. These responses are remarkably similar to the responses 10 years earlier where approximately 70–80% of those respondents also counseled that patients with HLHS would have neurologic and physical deficits compared to peers [7]. The questionnaire did not ask if these outcomes would differ depending on the initial palliative procedure, thus no comment can be made on this issue. Preliminary data, in at least one center, did not show any differences in neurological outcome between NW versus hybrid procedures [23, 24].

Ethical arguments for and against hospice care for patients with HLHS or single ventricle anatomy in general have been offered in the literature [11, 25–29]. There is little doubt that certain associated comorbidities in a patient with HLHS portend worse outcome and hospice care may be the primary recommended option for these patients. However, in this questionnaire, approximately 70% of respondents also offered NI care for the "routine" patient with HLHS without other comorbidities, albeit not the primary recommendation. This percentage was higher than the previous 45% of respondents that offered NI care in the questionnaire a decade earlier. Even with improvements in overall morbidity and mortality results in patients with HLHS [3, 4, 8, 30, 31], it is interesting to note that the percentage of respondents offering NI care has increased over the past decade. This suggests that the pediatric cardiac community still has concerns over the overall outcome in these patients, as well as potentially less paternalistic approaches to counseling than in prior eras. The discussion of hospice care and non-intervention in the present era is appropriate considering the differences in opinions in the medical field, the evolving outcomes in this patient population, and the ongoing decline in access to elective termination [11, 25–29]. One study has noted possible adverse effects of counseling of termination of pregnancy in fetuses with HLHS when the parent has decided not to terminate [12]. However, for families caring for a child with HLHS, providing counseling and education of all potential options would seem to be appropriate. This is especially the case in HLHS whereby an overall survival rate to age 5 approaches at best 70% currently, with three major cardiac procedures to achieve survival. With expected further decline in legal elective termination in the United States, there may also be a greater shift towards hospice care and non-intervention after birth. We have no data on families currently choosing elective termination when carrying a fetus with HLHS. Future research is needed to assess if any shift in care occurs, especially in regards to disparate effects on families caring for children with HLHS located within communities that may have variable access to healthcare options [32].

This study has several limitations. We cannot calculate a response rate because the total number of recipients of the questionnaire request is unknown. The voluntary subscribers to the various listservs employed may not reflect a true cross section of the pediatric cardiology community as a whole and may bias the data; in particular, the limited number of surgeons, who may be largely responsible for center-level practice patterns, may confound some of the analysis. Also, given survey anonymity, there is a potential for duplicate responses which we cannot easily assess for. There were significant differences in some baseline respondent demographics between this questionnaire and the previous questionnaire 10 years ago. We could not determine if these baseline differences in respondent demographics may partially explain the differences in responses between the two questionnaires. That said, there were no significant differences in HLHS surgical volume or location of US region of the respondents between questionnaires, which we think would have made a larger difference in the responses.

In conclusion, the NW-RVPA operation is the most recommended strategy for patients with HLHS, regardless of location. Greater variability exists in strategy when additional risk factors are present, although the use of the hybrid procedure, in low birth-weight infants specifically and in high-risk patients in general, is increasingly being employed. Hospice care continues to be offered even in standard risk patients with HLHS. Further long-term data collection will help determine eventual outcomes of the various procedures in regards to underlying risk.

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Nandi. The first draft of the manuscript was written by Drs. Nandi and Cua, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Declarations

Conflict of interest None of the authors have any relevant financial disclosures related to the content of this manuscript.

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