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CASE REPORT

Medical and Day-to-Day Burden on Families of Children with Hypoplastic Left Heart Syndrome Who Have Undergone Single Ventricle Surgical Palliation over a Decade

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Abstract

Background: Children with hypoplastic left heart syndrome (HLHS) and their families face unique medical and non-medical challenges throughout their entire life. The objective of this research is to describe day-to-day burden on families and medical outcomes of children with (HLHS) who underwent surgical palliation.

Methods: A retrospective chart review was performed including 165 infants diagnosed with HLHS who underwent surgical palliation from 2008-2017 in a single institution. Sixteen parents responded to a questionnaire which included yes/no questions to assess child and family burden related to initial hospitalization, discharge process, home routine.

Result: Parents reported a "difference" between prenatal counseling and child's course (64%) and experienced distress and burden by technology (60%) and feeding regimen at discharge (77%). Families reported siblings' distress (67%) and need to quit their job or cut back hours (92%). Mortality at NICU discharge was 14% and 21% at time of review.

Conclusion: The study shows a significant self-reported day-to-day burden on families of children with HLHS undergoing surgical palliation, even in the presence of relatively low incidence of mortality and complications.

Keywords

Neonatology, Hypoplastic left heart syndrome, Pediatric cardiology

Abbreviations

HLHS: Hypoplastic Left Heart Syndrome; NICU: Neonatal Intensive Care Unit; SVSP: Single Ventricle Surgical Palliation CHD: Congenital Heart Disease; NCCP: Neonatal Comfort Care Program; CUIMC/NYPMSCH: Columbia University Irving Medical Center/NewYork-Presbyterian-Morgan Stanley Children's Hospital; SVR: Single Ventricle Reconstruction MBTS: Modified Blalock-Taussig Shunt; RVPAS: Right Ventricle-To-Pulmonary Artery Shunt; ECMO: Extra-Corporeal Membrane Oxygenation

Introduction

Children with hypoplastic left heart syndrome (HLHS) and their families face unique medical and non-medical challenges that persist well beyond the newborn period. Prior to 1981, infants with HLHS died within the first few weeks of life. In 1981, Norwood, et al. published their first case of an infant surviving after the Norwood procedure [1,2]. This procedure has since been practiced widely, with modifications, and the inclusion of 2 additional surgeries: The Glenn and the Fontan procedures [1-3]. These surgeries continue to be referred to as 'Single Ventricle Surgical Palliation' (SVSP) and a transplant is often needed to survive to adulthood [4].

Although there are improving surgical outcomes data, mortality and morbidity remain high when

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compared to other congenital heart lesions [5-7]. Therefore, it is still accepted by many cardiologists, cardiothoracic surgeons, intensivists, and ethicists that it is reasonable to offer the option of palliative care to families of infants with HLHS [5,8,9], especially when other risk factors are present [7]. These practices also vary widely internationally, as D'Udkem and Hutchinson described the prenatal counseling approach of families receiving a fetal diagnosis of single ventricle anatomy in the largest pediatric surgery center in Australia and New Zealand. The authors do not openly offer the option of comfort care as an alternative to surgery but would accept this option as a family choice [9]. This could be as the acceptability of comfort care for the severe forms of congenital heart disease (CHD), including HLHS, varies widely amongst providers. Swanson, et al. asked over 100 pediatric cardiologists and cardiothoracic surgeons whether they would support parents' choice of non-interventions for several types of CHD. Specifically in the case of HLHS, 45% of the responders would agree with a comfort care plan and would support parents in this choice, 33% would disagree with the choice, but still would support parental decision, and 22% would disagree and would not support parents in the choice, but no one would seek legal action to remove child from parental custody [8]. In addition, many families may choose termination when faced with this life-limiting diagnosis.

At our institution, neonatal palliative care has been offered as alternative to surgical palliation or termination of pregnancy to families of children with single ventricle anatomy through a service of perinatal palliative care, the Neonatal Comfort Care Program (NCCP) [10]. The NCCP is a multidisciplinary team of neonatologists, social workers and nurses working in collaboration with the maternal-fetal-medicine team to provide comfort/palliative care to neonates with life-limiting conditions and support their families when parents elect to continue the pregnancy.

It is imperative that families understand and appreciate what these surgeries entail and what life after SVSP may look like when they choose surgical palliation [11]. Even with improving survival, children with HLHS need continuous medical attention and face frequent complications if they are to survive to adulthood [4,12,13]. After hospital discharge, they require daily medications, frequent doctors' appointments and further invasive procedures and testing [14]. They also face readmissions, emergency room visits and unplanned procedures [15]. These procedures and frequent encounters with the medical system can have a profound impact on both the children and their families.

Parents of children with CHD undergoing surgery face psychosocial challenges [16-18]. They reported high anxiety scores, lower quality of life and greater stress in caregivers of children with HLHS in the settings of morbidities and complications [18-20]. However, little is known about the day-to-day burden on family life.

This study aims to describe both the medical and day-to-day burden on families of children with HLHS who have undergone SVSP over the course of 10 years at Columbia University Irving Medical Center/NewYork-Presbyterian-Morgan Stanley Children's Hospital (CUIMC/NYPMSCH).

Methods

This study is a retrospective descriptive case series of children with HLHS and includes cross-sectional questionnaires of their parents. Children who underwent SVSP for HLHS at CUIMC/NYPMSCH from January 2008 through December 2017 were included. This study was reviewed and approved by the Institutional Review Board at CUIMC and covered under protocol AAAS1099. A waiver of consent was granted for retrospective data collection. Informed consent was obtained from parents who participated in the interviews.

Retrospective analysis to assess medical burden

Patients with HLHS were identified by reviewing the neonatology census from January 2008-December 2017. The diagnosis of HLHS was confirmed through both operative and cardiology notes. In order to assess the medical burden, the following data points were collected for each patient at the time of discharge, at follow-up visits and at subsequent admissions: Demographics (maternal race, gestational age at birth, birth weight, sex, prenatal diagnosis and genetic syndrome), procedures other than cardiac surgery (for example cardiac catheterization or gastric tube), surgical repair (including type of surgery and age at surgery), and complications during the first admission (including delayed chest closure, cardiac arrest, extracorporeal membrane oxygenation (ECMO), arrythmia, thrombotic event, diaphragm or vocal cord paralysis, seizure, stroke, necrotizing enterocolitis and renal failure) in the Neonatal Intensive Care Unit (NICU). The number of chest radiographs, echocardiograms, discharge medications, and discharge appointments were also noted. Feeding regimen and technology use (which includes nasogastric tube, gastric-tube, tracheostomy, or oxygen use) at discharge were recorded. After discharge emergency room visits, cardiac catheterizations, hospital days, mortality, and transplant were noted as well. Summary statistics are reported as medians and interquartile ranges or number and percent.

Cross-sectional questionaire to assess day-to-day burden

To assess day-to-day burden on children and families, parents were offered questionnaires by members of the study team. All English-speaking parents of surviving

children followed in our institution were eligible for the study. Parents were informed of the study during routine follow-up encounters. Those who elected to participate were interviewed by the study team either in person, by phone or completed the questionnaire and returned the results to the study team. Interviews were not recorded.

The questionnaire was investigator-designed and based on the aims of interest. It included several multipart "yes/no" questions to assess child and family burden related to initial hospitalization, discharge process and home routine. It also provided space for comments if desired. It was not validated by external sources, rather was investigator-designed.

Parents were asked whether they perceived surprise and/or distress or burden by the number of interventions (X-rays, echocardiograms, blood draws) during hospitalization, and by the number of medications, follow-up appointments, feeding regimen and technology requirement at discharge. Other questions assessed disruption of family and home routine and perception of prognostic prediction of the prenatal counseling.

The process of designing the questionnaire included focus group meetings with NICU nurses, parents of infants admitted to the cardiac NICU, neonatologists with expertise in neonatal cardiology, and the cardiologist in charge of the single ventricle follow-up program. Face validity was obtained by receiving feedback from non-experts to verify the understandability of the items. Parent interviews are reported with the percent of parents who answered each question in the positive.

Results

There were 165 newborns identified with HLHS who underwent SVSP during the study time period. Patient demographic characteristics are reported in Table 1. Table 2 describes the outcomes and complications at NICU admission, at the time of discharge and long-term. Data at discharge is provided for 132 newborns, as 21 newborns died, and 12 newborns were excluded as they remained either hospitalized through the second surgery or transitioned to pediatric rehabilitation facilities. Data for long term outcomes was available in 109 patients, since 23 children were lost to follow up as they received follow-up medical care at outside institutions.

Overall mortality at the time of retrospective chart review of our population was 21.2%. During initial hospitalization we report 13.7% mortality, 32% unplanned interventions, 17% thrombotic events, 4.9% seizures and 3.7% strokes. More than half of our patients had delayed sternal closure after their first surgery.

The parents of 29 children were informed about the study. Seventeen consented to participate and 16 completed the interview either in person or over the phone. The age of the children at the time of the interview spanned from ages 2-12. All but one child was prenatally diagnosed. Demographic characteristics of the parents who completed the interviews and of their children are reported in Table 3. Table 4 and Table 5 report the interview results.

Table 1: Patients' demographics (N = 165).

Variable	Median (IQR) or N (%)
Inborn	133 (80.6)
Maternal Age (years)	29 (24,35)
Gestational age (weeks)	38 (38,39)
Birth weight (grams)	3145 (2793,3479)
Male	112 (68)
Prenatal diagnosis	143 (87)
Genetic diagnosis	8 (4.8)
Maternal Race	
White	76 (46.1)
African American	25 (15.2)
Latino/Hispanic	8 (4.8)
Asian	4 (2.4)
Other or unknown	52 (31.5)

Of the 16 families that answered the questionnaire, the majority (93.8%) received a prenatal diagnosis. None had a genetic diagnosis, and 75% of the parents that answered were white. The children's ages ranged from 8-10 years-old and all, but one questionnaire was answered by a mother. Only 25% of the children were feeding fully orally by mouth at the time of discharge with 68.8% requiring nasogastric feeds and one child requiring gastric tube feeds, which is like the feeding regimen of the total studied population. Three of the 16 children had received a transplant at the time of the interview.

Parents reported surprise and/or distress and burden from various aspects of their child's care. Almost half of the parents reported surprise with the amount of blood drawn and one third reported distress and burden. At discharge, 60% of the parents reported distress and burden because of the number of appointments and the technology requirement.

The highest degree of self-reported surprise (75%), distress and burden (77%) were associated with the feeding regimen at discharge.

Most families expressed significant financial and organizational difficulties. More than 90% of the responders reported that one or both parents needed to either quit or cut back hours at work in order to care of their child and more than 80% experienced problems hiring a babysitter who would be comfortable caring for their child due to medical complexity. Moreover, 2/3 of the respondents stated that they needed to stay awake at night to provide attention to the child.

Table 2: Outcomes and complications at NICU admission, at time of discharge and long-term.

Variable	Median (IQR) or N (%)
IN HOSPITAL OUTCOME or COMPLICATION (N = 165)	
Mortality	21 (13.7)
Pre-operative septal intervention	13 (7.9)
Age at surgery (days)	5 (3, 7)
Unplanned intervention	52 (31.5)
Delayed sternal closure	94 (57.3)
Open chest days	4 (3, 5)
Cardiac arrest	16 (9.8)
ECMO	19 (11.6)
Other surgery	20 (12.2)
Post-op mechanical ventilation (days)	6 (4, 7.75)
Total length of any respiratory support (days)	15 (9, 27)
Arrhythmia	43 (26.5)
Thrombotic event	29 (17.2)
Diaphragm paralysis	13 (8.0)
Vocal cord paralysis	21 (12.9)
Chylothorax	14 (8.5)
Stroke	6 (3.7)
Seizure	8 (4.9)
Necrotizing enterocolitis	8 (4.9)
# of X-rays	21 (15, 30)
X-rays per hospital day	0.63
# of ECHOs	8 (6, 11)
ECHOs per hospital day	0.25
DISCHARGE OUTCOME or COMPLICATION (N = 132)	
Discharged home after first stage	132 (80.0)
Length of NICU admission (days)	33 (21, 53)
Feeding regimen at discharge (N = 132)	
PO + NG	72 (54.5)
PO	37 (28.0)
GT	10 (7.6)
NG	13 (8.8)
Technology at discharge	97 (73.5)
Number of discharge meds	5 (3, 8)
Number of discharge appointments	6 (4, 7)
LONG-TERM OUTCOMES (N=109)	
# of emergency room visits in 1st year of life	1 (0,2)
# of catheterizations in 1st year of life	1 (1, 2)
# of surgeries in 1st year of life	2 (2, 3)
Total # Hospital days in 1st year of life	60 (37, 93)
Total # of Hospital days in 1st year of life (not including NICU admission)	14 (6.8, 34.3)
Transplant	16 (14.4)
Age at transplant (years)	3 (1.3, 3.6)
Protein losing enteropathy	7 (8.0)
Plastic bronchitis	1 (1.0)
Mortality at time of chart review of entire cohort	35 (21.2)
Mortality age < 1 year	29 (82.9)
Mortality age > 1 to < 3 years	2 (5.7)
Mortality > 3 years	4 (11.4)

ECMO: Extracorporeal Membrane Oxygenation; post-op: Post-Operative; ECHO: Echocardiogram; PO: Oral; NG:Nasogastric; GT: Gastric Tube.

Discussion

This study offers relevant and innovative knowledge of the substantial day-to-day burden on children with HLHS and their families through parent questionnaire. Moreover, the study provides information regarding the medical burden of children with HLHS, obtained on a large cohort of patients who have been treated and followed in a tertiary cardiac center over 10 years.

Responses from families of children with HLHS indicate that HLHS is a disease that affects not only the child, but has an impact on the entire family, including siblings.

One pivotal aspect of the day-to-day burden is the financial impact. Du Plessis, et al. collected on-line surveys of 107 parents of children who underwent the Fontan operation. While the greatest parental concerns were fear of death and psychological well-being for their children, 2/3 of the parents reported serious financial impact for the family [16]. Our investigations support this finding as most families expressed significant financial and organizational difficulties. The vast majority reported that one or both parents needed to either quit or cut back hours at work in order to care for their child and most experienced problems hiring a babysitter able to take care of the child. This can have a drastic financial and social impact on the entire family.

An important aspect of day-to-day burden associated with the presence of a child with HLHS relates to the impact on the siblings. Parker, et al. performed a literature review spanning 1967 to 2019 and found that having a child with CHD is associated with changes in family life routine and has a negative impact on siblings [21] Caris, et al. showed that siblings of children with HLHS are negatively affected by their sibling's chronic illness, with worse adjustment as they get older [22]. In our study 2/3 of the parents reported "signs of distress" in the siblings of the child with HLHS, possibly associated with disruption of the siblings' routine, which was reported in half of the cases.

It is interesting and important to note that most families we interviewed reported "surprise and burden" about interventions during admission and post-discharge day-to-day family life, even in the presence of the relatively low incidence of morbidities and complications as reported in our academic institution. Notably, the most distressing and burdensome aspect of their child's course was the feeding regimen, as 80% of the children of the responding parents were discharged with either a nasogastric or gastrostomy tube.

Table 3: Demographics and outcomes of the families that completed the interviews (N = 16).

Variable	Median (IQR) or N (%)
Inborn	14 (93.3)
Gestational age (weeks)	39 (38, 39)
Birth weight (grams)	3185 (2827, 3482)
Male	12 (75)
Prenatal diagnosis	15 (93.8)
Genetic diagnosis	0 (0)
Child's current age (years)	9 (8, 10)
Interview completed by mother	15 (93.8)
Maternal age at birth (years)	30 (27, 34.5)
Maternal Race	
White	12 (75.0)
African American	1 (6.3)
Asian	1 (6.3)
Other or unknown	2 (12.5)
Home feeding regimen at discharge	
PO	4 (25.0)
PO + NG	11 (68.8)
GT	1 (6.3)
Transplant	3 (18.8)

Table 5: Parent Interview Results (%varies by # of parents who completed each question, total N = 16).

Question	N (%)
Need to quit job or cut back hours	12 (92.3)
Difficulty hiring babysitter	11 (84.6)
Need to stay awake at night to care for child	7 (63.6)
Siblings showed signs of distress	6 (66.7)
Sibling routine disrupted	4 (50.0)
Difference between prenatal counseling and child's course	9 (64.3)
Offered palliative/comfort care prenatally	9 (60.0)

Table 4: Parent Interview Results (%varies by # of parents who completed each question, total N = 16).

Variables	Surprised–N (%)	Distressed or Burdened–N (%)
#X-rays	4 (26.7)	5 (35.7)
#Echocardiograms	4 (26.7)	2 (14.3)
#Blood draws	7 (46.7)	5 (35.7)
# Medications at discharge	5 (31.3)	5 (35.7)
#Appointments at discharge	3 (20.0)	8 (57.1)
Feeding regimen at discharge	12 (75.0)	10 (76.9)
Technology requirement at discharge	6 (40.0)	3 (60.0)

In the study population only a quarter of the children were successfully fed by mouth at the time of discharge, as is to be expected for infants with HLHS. Children with HLHS have a high need for assisted feeding strategies or gastrostomy tube placement [23]. Moreover, feeding difficulties have been reported as one of the most significant stressors among parents of children who underwent cardiac surgery. Tregay, et al. conducted interviews with the families of 20 infants who had undergone cardiac surgery. While only half of the children in the study required some form of assisted feeding, 80% of the families expressed concern and stress, sometimes overshadowing other medical concerns [24].

While mortality of patients with HLHS has decreased over time [22], morbidity after single ventricle palliation is still commonplace [4,10,14,25]. The single ventricle reconstruction (SVR) trial compared the outcomes of children who underwent a modified Blalock-Taussig shunt (MBTS) versus a right ventricle-to-pulmonary artery shunt (RVPAS). The trial followed children at 3 and 6 years after single ventricle reconstruction and reported similar transplant-free survival in the 2 groups (RVPAS = 64%, MBTS = 59%, P = 0.25). Notably, cumulative incidence of morbidities included thrombotic events (20%), seizures (15%), and strokes (7.5%) [15,26].

The medical burden reported in our study is similar or better to previously reported outcomes. Fifty-seven percent of children in our study had delayed sternal closure, which is lower than the rate of 80% reported by Mills on 193 patients following stage 1 palliation [27]. Children in our cohort had similar rates of imaging studies per day (X-rays and echocardiograms), median 1st admission length of stay, ECMO and transplant of other children with HLHS as other published reports [28,29].

It is important that parents have a full understanding of the disease course, of the complex needs of their children and the challenge for the entire family in order to make an informed decision about their child's care [11].

In our study, 64% of responding parents answered that there was a "difference" between prenatal counseling and their child's actual clinical course. Some parents stated that their child's clinical course was more complicated than they expected, while others felt that the physicians that provided prenatal counseling were "too negative" and their child's clinical course was less complicated than they expected. Some unsolicited comments exemplified families opposite experiences of what they perceived as contradictory information. About half of them reported unexpected better outcomes with unsolicited comments: *"We were given a much worse scenario than what we got. Glad for that"; "They were pretty negative. Doctors go by statistics, I guess my son is not the typical and he did so well all along."*

Contrastingly, other parents expressed that their child's clinical course was worse than they had been told: *"We were not prepared for anything. Anything bad that could have happened to my baby, happened"; "The experience was far more traumatic than ever expected. Prenatally we were just told he would have Norwood/Glenn/Fontan, made it seem like it was just 'boom/boom/boom', was false security based on statistics."*

One reason for the perceived difference between what parents heard at prenatal counseling and the actual child's clinical course could be that the shock of the diagnosis and the way it was presented might have negatively affected parental understanding. Bertaud, et al. conducted a qualitative study by interviewing parents of surviving children with HLHS. Some mothers reported being overwhelmed by the amount of information received and others perceived strong pessimism regarding the prognosis, possibly leading to a detrimental impact in their understanding [30].

Parents in our study reported wide variation in the options offered at prenatal counseling as only 60% reported being offered a palliative care plan. One possible explanation for this finding is that the parents in our study received initial counseling with many different providers at different institutions prior to presenting at our institution. This is in line with the variability in the content of prenatal counseling described in prior literature. A survey of 201 pediatric cardiologists found that palliative care, or non-intervention, was discussed by only 71% of respondents [31]. Comfort care is offered at our institution to children with life-limiting diagnoses, including HLHS, especially when complicated, and has been studied by our team. Prior research showed that in our institution, during the same period in which the current data were collected, 13.5% of families with a single ventricle diagnosis chose comfort care [32].

One strength of this study includes the collection of long-term outcome data of a large population of infants with HLHS (165) over a decade with only 23 patients lost to long-term follow-up because moved care to other institutions.

Another strength relates to the survey portion of the study. Data from interviews were collected from parents of infants of older age (8- to 10-year-old), who therefore had the opportunity to experience long-term day-to-day challenges of having a child with HLHS.

The survey portion of the study has some limitations. A sampling bias is possible for multiple reasons. First, a small number of parents were informed about the study, and participants included only English-speaking parents and 93.3% of parents that were interviewed were mothers. This limited number of participants is mainly due to the fact that during the occurrence of the COVID-19 pandemic our institution paused investigations and did not allow contact of patients and families for research purposes. Moreover, a few parents declined

to be contacted by the study team, some parents agreed to be contacted, and then did not respond to investigator inquiries. Another possible selection bias relates to demographic difference between the entire population and the subset of parents that agreed to be interviewed as the latter were mostly white (75%) and not representative of the total cohort (only 46% identified as white).

Finally, the investigator-developed questionnaire used in this study has had limited opportunity for validity.

While information about the medical burden of children undergoing SVSP is already well known in medical literature, the novelty of this study is the identification of the day-to-day burden for families and children. Our study offers an innovative contribution in describing the complexity of the life of children with HLHS and their families as it focuses on outcomes that may be equally or more important to parents than medical or surgical complications that have not previously reported in literature.

Although this is a small, investigator designed study, the results offer suggestions to help guiding clinicians in counseling parents with a fetal diagnosis of HLHS in deciding options of care. It is important to give consistent preoperative information that includes both the surgical and social outcomes of this disease.

Further qualitative studies or quantitative studies with larger sample size and less sample bias will be helpful to fully gauge parental expectations and understanding at the time of prenatal counseling, admission and after discharge and to investigate interventions that will help families and patients to improve the medical and day-to-day burden of this disease.

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Conflict of Interest

All authors declared no conflict of interest.

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Statement of Equal Authors' Contribution

Erin Hanft, Elvira Parravicini and Anne Ferris conceptualized the research and developed the survey. Erin Hanft and Elena Abascal collected and analyzed the data. Erin Hanft and Elvira Parravicini wrote the original draft, and all 4 authors edited the final version of the manuscript. All authors have read and agreed to the submitted version of the manuscript.

References

1. Norwood WI, Lang P, Castaneda AR, Campbell DN (1981) Experience with operations for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 82: 511-519.
2. Norwood WI, Lang P, Hansen DD (1983) Physiologic repair of aortic atresia - hypoplastic left heart syndrome. N Engl J Med 308: 23-26.
3. Yabrodi M, Mastropietro CW (2017) Hypoplastic left heart syndrome: From comfort care to long-term survival. Pediatr Res 81: 142-149.
4. Diego R, Xiaoya Z, Abraham GP, Fernando GPJ, Sylvia CBR (2023) Hypoplastic left heart syndrome: A review. Cardiology in Review 31: 149-154.
5. Kane JM, Canar J, Kalinowski V, Johnson TJ, Hoehn KS (2016) Management options and outcomes for neonatal hypoplastic left heart syndrome in early twenty-first century. Pediatr Cardiol 37: 419-425.
6. Liu MY, Zielonka B, Snarr BS, Zhang X, Gaynor JW, et al. (2018) Longitudinal assessment of outcome from prenatal diagnosis through fontan operation for over 500 fetuses with single ventricle-type congenital heart disease: The Philadelphia fetus-to-fontan cohort study. J Am Heart Assoc 7: e009145.
7. Metcalf MK, Rychi J (2020) Outcomes in hypoplastic left heart syndrome. Pediatr Clin North Am 67: 945-962.
8. Swanson TM, Patel A, Baxter AJ, Morris SA, Maskatia SA, et al. (2020) Pediatric Cardiology 41: 1160-1165.
9. D'Udkem Y, Hutchinson D (2022) Being born with a single cardiac ventricle: What do we tell prospetive parents. Prenat Diagn 42: 411-418.
10. Wool C, Parravicini E (2020) The neonatal comfort care program: Origin and growth over 10 years. Front Pediatr 8: 588432.
11. Mery CM, Well A, Taylor K, Carberry K, Colucci J, et al. (2023) Examining the real-life journey of individuals and families affected by single-ventricle contenital heart disease. J Am Heart Assoc 12: e027556.
12. Lewis M, Rosenbaum M (2017) The miracle baby grows up: Hypoplastic left heart syndrome in the adult. Curr Cardiol Rep 19: 74.
13. Simsic JM, Phelps C, Kirchner K, Carpenito KR, Allen R, et al. (2018) Interstage outcomes in single ventricle patients undergoing hybrid stage 1 palliation. Congenit Heart Dis 13: 757-763.
14. Wald RM, Mertens LL (2022) Hypoplastic left heart syndrome acrros the lifespan: Clinical considerations for care of the fetus, child, and adult. Can J Cardiol 38: 930-945.
15. Newburger JW, Sleeper LA, Gaynor JW, Hollenbeck-Pringle D, Frommelt PC, et al. (2018) Transplant-free survival and interventions at 6 years in the SVR trial. Circulation 137: 2246-2253.
16. Wei H, Roscigno CI, Swanson KM, Black BP, Hudson-Barr D, et al. (2016) Parents' experiences of having a child undergoing congenital heart surgery: An emotional rollercoaster from shocking to blessing. Heart Lung J Acute Crit Care 45: 154-160.
17. Caris EC, Dempster N, Wernovsky G, Butz C, Neely T, et al. (2016) Anxiety scores in caregivers of children with hypoplastic left heart syndrome. Congenit Heart Dis 11: 727-732.

18. Rempel GR, Rogers LG, Ravindran V, Magill-Evans J (2012) Facets of parenting a child with hypoplastic left heart syndrome. Nurs Res Pract 2012: 714178.

19. du Plessis K, Peters R, King I, Robertson K, Mackley J, et al. (2018) "Will she live a long happy life?" Parents' concerns for their children with Fontan circulation. Int J Cardiol Heart Vasc 18: 65-70.

20. Harvey KA, Kovalesky A, Woods RK, Loan LA (2013) Experiences of mothers of infants with congenital heart disease before, during, and after complex cardiac surgery. Hear Lung J Acute Crit Care 42: 399-406.

21. Parker R, Houghton S, Bichard E, McKeever S (2020) Impact of congenital heart disease on siblings: A review. J Child Heal Care 24: 297-316.

22. Caris EC, Dempster N, Wernovsky G, Miao Y, Moore-Clingenpeele M, et al. (2018) Perception scores of siblings and parents of children with hypoplastic left heart syndrome. Congenit Heart Dis 13: 528-532.

23. Prodhan P, Tang X, Gossett J, Beam B, Simsic J, et al. (2018) Gastrostomy tube placement among infants with hypoplastic left heart syndrome undergoing stage 1 palliation. Congenit Heart Dis 13: 519-527.

24. Tregay J, Brown K, Crowe S, Bull C, Knowles R, et al. (2017) "I was so worried about every drop of milk" - feeding problems at home are a significant concern for parents after major surgery in infancy. Matern Child Nutr 13: e12302.

25. Fernandez RP, Joy BF, Allen R, Stewart J, Miller-Tate H, et al. (2017) Interstage survival for patients with hypoplastic left heart syndrome after ECMO. Pediatr Cardiol 38: 50-55.

26. Newburger JW, Sleeper LA, Frommelt PC, Pearson GD, Mahle WT, et al. (2014) Transplantation- free survival and interventions at 3 years in the single ventricle reconstruction trial. Circulation 129: 2013-2020.

27. Mills KI, Van Den Bosch SJ, Gauvreau K, Allan CK, Thiagarajan RR, et al. (2018) Physiologic effects of delayed sternal closure following stage 1 palliation. Cardiol Young 28: 1393-1403.

28. Hamzah M, Othman HF, Elsamny E, Agarwal H, Aly H (2020) Clinical outcomes and risk factors for in-hospital mortality in neonates with hypoplastic left heart syndrome. Pediatr Cardiol 41: 781-788.

29. Kogon BE, Kanter K, Alsoufi B, Maher K, Oster ME (2015) Outcomes and hospital costs associated with the Norwood operation: Beyond morbidity and mortality. Cardiol Young 25: 853-859.

30. Bertaud S, Lloyd DFA, Sharland G, Razavi R, Bluebong-Langner M (2020) The impact of prenatal counselling on mothers of surviving children with hypoplastic left heart syndrome: A qualitative interview study. Health Expect 23: 1224-1230.

31. Walsh MJ, Verghese GR, Ferguson ME, Fino NF, Goldberg DJ, et al. (2017) Counseling practices for fetal hypoplastic left heart syndrome. Pediatr Cardiol 38: 946-958.

32. Haxel C, Glickstein J, Parravicini E (2019) Neonatal palliative care for complicated cardiac anomalies: A 10-year experience of an interdisciplinary program at a large tertiary cardiac center. J Pediatr 214: 79-88.