Neonatal Palliative Care for Complicated Cardiac Anomalies: A 10-Year Experience of an Interdisciplinary Program at a Large Tertiary Cardiac Center

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Objectives To report the outcomes of a Neonatal Palliative Care (NPC) Program at a large tertiary cardiac center caring for a subset of fetuses and neonates with life-limiting cardiac diagnoses or cardiac diagnoses with medical comorbidities leading to adverse prognoses.

Study design The Neonatal Comfort Care Program at New York-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center is an interdisciplinary team that offers the option of NPC to neonates prenatally diagnosed with life-limiting conditions, including single ventricle (SV) congenital heart disease (CHD) or less severe forms of CHD complicated by multiorgan dysfunction or genetic syndromes.

Results From 2008 to 2017, the Neonatal Comfort Care Program cared for 75 fetuses or neonates including 29 with isolated SV CHD, 36 with CHD and multiorgan dysfunction and/or severe genetic abnormalities, and 10 neonates with a prenatal diagnosis of isolated CHD and postnatal diagnoses of severe conditions who were initially in intensive care before transitioning to NPC because of a poor prognosis.

Conclusions At New York-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center, a large tertiary cardiac center, 13.5% of parents of fetuses or neonates with isolated SV CHD opted for NPC. Twenty-six of 29 newborns with SV CHD treated with NPC died. Of the remaining, 2 neonates with mixing lesions are alive at 3 and 5 years of age, and 1 neonate was initially treated with NPC and then pursued surgical palliation. These results suggest that NPC is a reasonable choice for neonates with SV CHD. *(J Pediatr 2019;214:79-88)*.

Neonatal palliative care (NPC) is a holistic approach for the care of neonates affected by life-limiting or complex medical conditions with adverse prognoses. This interdisciplinary treatment aims to achieve a state of comfort for the neonate during their natural life and to provide support to their families pre and postnatally. NPC can be proposed during pregnancy at the time of the prenatal diagnosis or postnatally as redirection of care when the diagnosis is clarified, or a neonate's prognosis becomes adverse.[1]

Annually, nearly 1% of neonates born in the US have congenital heart disease (CHD).[2-4] Within this group, one-quarter have critical CHD and will require surgery or catheter intervention in the first year of life.[5,6]

The Neonatal Comfort Care Program (NCCP) at New York-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center (NYPMSCH/CUMC) is an interdisciplinary team that offers NPC to neonates prenatally diagnosed with life-limiting conditions, including patients with single ventricle (SV) CHD or with less severe forms of CHD complicated by multiorgan dysfunction and/or severe genetic syndromes.[7]

Hypoplastic left heart syndrome (HLHS) is one of the most severe forms of SV CHD that affects 1000 neonates annually in the US.[8,9] Prior to 1980, HLHS was fatal and NPC was the primary therapy offered to families postnatally. The development of the Norwood technique revolutionized surgical intervention for HLHS allowing for a 3-stage SV surgical palliation.[10] Fetal intervention, including aortic balloon valvuloplasty and atrial septal stenting, has broadened the options for intervention, with varying outcomes.[11-17]

Pursuing SV palliation commits a neonate or child to at least 3 operative procedures, multiple cardiac catheterizations, and in many cases, cardiac transplantation. Significant medical and neurodevelopmental morbidities exist.[18-29] The true burdens of these comorbidities are still being delineated as the earliest patients with HLHS who completed SV palliation are entering their fourth decade of life.

CHD	Congenital heart disease
HLHS	Hypoplastic left heart syndrome
NCCP	Neonatal Comfort Care Program
NPC	Neonatal palliative care
NYPMSCH/CUMC	New York-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center
SV	Single ventricle
VSD	Ventricular septal defect

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Fetal echocardiography allows for prenatal diagnosis of CHD generally between 18 and 22 weeks of gestation, and in some cases as early as 12 weeks.[30-33] Parents given a prenatal diagnosis of SV CHD confront multiple options including termination of pregnancy, continuation of pregnancy with postnatal NPC, or continuation of pregnancy with postnatal surgical palliation.

Parental decision making when faced with a life-limiting diagnosis is complex and influenced by factors such as level of education, family structure, financial resources, religion, and the breadth of options discussed during prenatal counseling. Since the advent of stage I Norwood intervention, the mortality rates for HLHS have drastically improved, from nearly 0% survival to approximately 60% transplant-free survival at 6 years for patients with HLHS.[34] Some practitioners have argued that with significant improvement in survival, postnatal NPC should no longer be offered.[35] However, given the continued mortality risk and significant comorbidities, other practitioners argue that it is still appropriate for the option of continuation of pregnancy with postnatal NPC to be offered and facilitated for families who opt for this therapy.[36-39]

The aim of this study is to describe the outcomes of fetuses and neonates with the diagnosis of severe or complicated CHD treated with NPC over the course of 10 years by the NCCP at NYPMSCH/CUMC, with a focus on a subset of fetuses or neonates with isolated SV CHD. We include families' demographic data.

Methods

This is a retrospective descriptive consecutive case series of pregnant women and fetuses or neonates followed by the NCCP from 2008 to 2017 in the setting of a prenatal diagnosis of SV CHD or another CHD with multiorgan dysfunction and/or severe genetic syndrome. This is a selected population of mothers who decided to carry their pregnancy to term and elected for a postnatal plan of NPC or redirection of care from intensive to palliative care in the early postnatal period. Data collected from the electronic medical records included prenatal diagnosis, pregnancy outcome, postnatal diagnosis, postnatal management, neonates' length of life, and maternal demographics.

The Institutional Review Board approved the study and a waiver of consent granted.

The NCCP

Candidates. At NYPMSCH/CUMC, potential candidates for NPC include fetuses or neonates with a diagnosis of SV CHD or those with less severe forms of CHD complicated by multiorgan dysfunction and/or severe genetic syndromes. SV CHDs include a broad range of cardiac lesions that require staged SV palliation including HLHS, unbalanced atrioventricular canal defect, certain anatomic variants of double outlet right ventricle, double inlet left ventricle, pulmonary atresia with an intact ventricular septum, and severe forms of Ebstein anomaly of the tricuspid valve.

NCCP Team. At NYPMSCH/Columbia University Medical Center (CUMC), the NCCP includes a core team (neonatologist who serves as medical director, nurse, social worker, program manager) and other consulting professionals (speech pathologist, lactation consultant, child life specialists, psychologist, and chaplain).

Prenatal Counseling and Birthing Plan. After the maternal fetal medicine physician communicates the fetal diagnosis and the cardiac and cardiothoracic surgical teams discuss the prognosis and surgical options, the core team meets with families interested in a plan of NPC. The medical director further clarifies the diagnosis, discusses the range of prognoses, and offers palliative options for each scenario. During the course of the pregnancy, the core team meets with the family as needed, to provide support to parents and siblings. An NPC plan and an interdisciplinary birth plan are prepared and included in the mother's clinical chart. In case of a fetus with uncertain or pending diagnosis, the postnatal plan of care is contingent upon postnatal confirmation.

Postnatal Care. The medical director assures continuity of care by providing prenatal and postnatal palliative consults and by serving as primary physician for each neonate.

The NCCP team follows each delivery, evaluates the neonate's clinical condition, and manages the postnatal medical plan focused on comfort during the neonate's natural life. When the prenatal diagnosis is certain, no postnatal imaging or laboratory testing is performed. At NYPMSCH/CUMC, fetal echocardiograms are performed by experienced fetal cardiologists, thus, postnatal imaging is done only when anatomic details need to be clarified. Moreover, given the extremely short life expectancy for these neonates, we prioritize time devoted to bonding for the neonate and family. The NCCP operates with previously reported standardized guidelines developed from a policy statement by the American Academy of Pediatrics and evidence-based data obtained in neonatal populations.[40-42]

Bonding, maintenance of body temperature, relief of hunger/thirst, and alleviation of discomfort/pain are the essential elements of NPC. Parent/neonate bonding and the interaction with siblings and other family members are implemented by providing private rooms in labor and delivery and in postpartum where the neonate rooms-in with the mother. Occasionally, neonates treated with NPC are admitted to the neonatal intensive care unit but exclusively in the cases of maternal medical complications, parental request, or in case of redirection from intensive to palliative care.

Skin-to-skin care has been used to facilitate bonding and maintenance of body temperature. Feeding is an essential element of comfort and the NCCP team encourages parents to feed their neonate as long as they are in stable condition. Lactation consultants are available for women who opt to

Table I. SV CHD with normal genetics

Pregnancy outcomes	Case no.	Prenatal diagnosis (gestational age at NYPMSCH/CUMC diagnosis) (wk)	Multiple gestation (Y/N)	Gestational age at delivery (wk)	Neonate location after birth	Length of life
Liveborn at NYPMSCH/CUMC	1	HLHS (MS/AS) (32)	N	38	R-in/Home	10 d
	2	HLHS (MA/AA), intact atrial septum (21)	Y (Triplet)	34	NICU	14 d
	3	RV dom AVC, PA, malposed gestational ages, infradiaphragmatic TAPVR; heterotaxy (RAI) (19)	N	38	R-in/Home	45 d
	4	HLHS (MA/AA), decreased RV function (19)	N	38	NICU	2 d
	5	HLHS (MA/AA) (32)	N	39	R-in/Home	1 mo
	6	HLHS (MS/AA) (31)	Y (Twin)	34	R-in	15 d
	7	Dextrocardia, RV dom AVC, TGA, obstructed TAPVR, heterotaxy (RAI) (30)	N	39	NICU	2 d
	8	HLHS (MA/AS), restricted foramen (26)	Y (Twin)	36	R-in/Home	9 d
	9	Dextrocardia, RV dom AVC, DORV, malposed gestational ages, pulmonary stenosis, cardiac TAPVR, heterotaxy (23)	N	40	R-in/Home	Alive and stable at 3 y
	10	HLHS (MS/AA) (26)	Y (Twin)	35	R-in/Home	1 mo
	11	DILV, large VSD, type B IAA (23)	N	37	NICU	13 d
	12	HLHS, restrictive vs intact atrial septum (27)	N	30	DR	2 h
	13	RV dom AVC (MA/AA), interrupted IVC/azygous continuation (heterotaxy, LAI) + Right congenital diaphragmatic hernia	N	38	Resuscitation followed by redirection of care in DR	1 h
	14	PA/IVS, tricuspid stenosis, severe RV hypoplasia, confluent branch PAs	N	37	R-in/Home/4 m elected for surgery	Alive and stable at 3 y
Liveborn at outside hospital	15	HLHS (MA/AA) (32)	N	40	NICU	7 d
	16	HLHS (MS/AA) (32)	N	Unknown	Unknown	5 d
	17	TGA vs DORV, hypoplastic MV, pulmonary outflow obstruction, RV/LV discrepancy (single ventricle) + ventriculomegaly/hydrocephalus (21)	Y (Twin)	40	NICU	5 d
	18	HLHS (MS/AS) (27)	Y (Twin)	40	R-in	3 d
	19	HLHS (MA/AA) (33)	N	40	R-in	3 d
Intra-Uterine Fetal Demise	20	HLHS (MS/AA) (18) + severe bladder outlet obstruction (18)	N	21		
	21	PA/IVS, poor LV function (21)	N	32		
	22	Dysplastic TV, severe TR, dilated RA, cardiomegaly, functional vs anatomic PA (21)	N	36		
	23	HLHS (MS/AS), decreased RV function (22)	N	30		
	24	HLHS (MA/AA), intact atrial septum (26)	N	35		
	25	RV dom AVC, PA, TAPVR, heterotaxy (RAI) (27)	N	29		
	26	Dysplastic TV, severe TR, dilated RA, PA, cardiomegaly (32)	N	35		

AVC, atrioventricular canal; *AS/A*, aortic stenosis/atresia; *DORV*, double outlet right ventricle; *DILV*, double inlet left ventricle; *DR*, delivery room; *Home*, home hospice; *IAA*, interrupted aortic arch; *MS/A*, mitral stenosis/atresia; *NICU*, neonatal intensive care unit; *PAs*, pulmonary arteries; *PA/IVS*, pulmonary atresia with an intact ventricular septum; *PS/A*, pulmonary stenosis/atresia; *R-in*, room-in; *R/LAI*, right/left atrial isomerism; *RV dom AVC*, right ventricular dominant atrioventricular canal; *TAPVR*, total anomalous pulmonary venous return; *TGA*, transposition of the great arteries; *TV/R*, tricuspid valve/regurgitation.

Table II. SV CHD with nonlife-limiting genetic diagnosis

Pregnancy outcomes	Case no.	Prenatal cardiac diagnosis (gestational age at CUMC diagnosis) (wk)	Prenatal genetic diagnosis	GA at delivery (wk)	Location	Length of life
Liveborn at NYPMSCH/CUMC	1	Unbalanced RV dom AVC, critical coarctation (24)	Trisomy 21	39	NICU/Home	Alive and stable at 5 y
	2	Severe Ebstein anomaly, severe TR, hydrops (34)	Trisomy 21	39	DR	1 h
	3	HLHS (MS/AA), intrauterine growth restriction (31)	Del long arm chromosome 11–Jacobsen syndrome	41	R-in	3 d

breast-feed, and speech pathologists are consulted for neonates with a weak suck or physical facial anomalies. Special nipples, bottles, or feeding devices are used to help these neonates feed safely and comfortably during their natural life. For neonates with worsening condition or in a state of impending death, relief of hunger and thirst can be achieved by colostrum care or non-nutritive sucking. Most neonates treated with palliative care because of severe or complex CHD do not experience pain; however, during the dying process some degree of discomfort can occur. The NCCP has a 10-year experience of successfully treating discomfort with opioid and/or benzodiazepines. Most neonates who survive the maternal admission are discharged home with local hospice support.

Results

The NCCP at NYPMSCH/CUMC was established in 2008. Over the course of 10 years, the NCCP cared for 75 fetuses or neonates affected by SV CHD or other CHD associated with multiorgan dysfunction or severe genetic conditions and their families.

A first subset included 29 fetuses or neonates with SV CHD with normal chromosomal analysis, or nonlife-limiting genetic diagnoses (Table I and Table II). The majority of fetuses or neonates (55%) had a diagnosis of HLHS. Three neonates had a nonlife-limiting genetic diagnosis including trisomy 21 (n = 2) and deletion of long arm of chromosome 11 (n = 1). Importantly, all 3 families indicated that the cardiac diagnosis was the primary driver for electing for NPC. All neonates, except for 7 admitted to the neonatal intensive care unit for maternal complications and/or family's preference, were able to room-in with the family. Of the live births (22/29), the median length of life was 8 days and the mode was 2 and 3 days.

During the 10-year time period, 185 families of fetuses or neonates with similar diagnoses opted for stage I surgical palliation. Therefore, 13.5% of our population opted for palliative care over surgical palliation.

At the time of publication, 3 neonates whose families opted for NPC are still alive. The first (Table I; patient 9) was diagnosed at 23 weeks gestation with heterotaxy syndrome. The neonate was delivered at 40 weeks gestation and was discharged home with hospice care. The child is currently 3 years old with pulse oximeter oxygen saturation to the mid-80s. The second patient (Table II; patient 1) was referred to CUMC at 24 weeks of gestation where a diagnosis of a right ventricular dominant atrioventricular canal and trisomy 21 was confirmed. The neonate was delivered at 39 weeks of gestation and was discharged home with hospice care. The child is currently 5 years old in compensated heart failure with neurodevelopmental delay consistent with trisomy 21.

Lastly, 1 patient with pulmonary atresia with an intact ventricular septum, and tricuspid stenosis was initially treated with NPC following diagnosis at 23 weeks and delivery at 37 weeks of gestation (Table I; patient 14). The neonate was discharged home with hospice care. At 4 months of life, the family opted to pursue SV surgical palliation including first a Blalock-Taussig shunt, followed by a bidirectional Glenn, Blalock-Taussig shunts take down, atrial septectomy, mitral valve annuloplasty, and left pulmonary artery stent placement. The child is currently 3 years old, however, the clinical course has been complicated by multiple reinterventions, endocarditis, thrombosis, arrhythmia, and hypothyroidism.

A second subset included 36 fetuses or neonates with CHD who had multiorgan dysfunction and/or life-limiting genetic diagnoses including trisomy 18 (75%), trisomy 13 (8%), unbalanced translocation of chromosomes 15;17, 47 XXX and large encephalocele, severe hydrops, renal agenesis, pentalogy of Cantrell, conjoined twins, or extreme prematurity. The most common cardiac anomalies included ventricular septal defect (VSD) (33%) and complete atrioventricular canal defect (28%). Fifteen fetuses died in utero. Most of the 21 neonates born alive died in the early postnatal period with a mean age at death of 30 days, a median and a mode of 1 day.

A third subset included 10 neonates with prenatal diagnosis of isolated CHD and postnatal diagnosis of an associated severe condition. These neonates were initially treated with intensive care before transitioning to NPC because of an adverse prognosis. Postnatal findings included trisomy 18 (50%), trisomy 13, CHARGE syndrome (coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities, and ear abnormalities), VACTERL association (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities), DiGeorge syndrome, severe brain anomalies, and extreme prematurity. The most common cardiac anomalies included

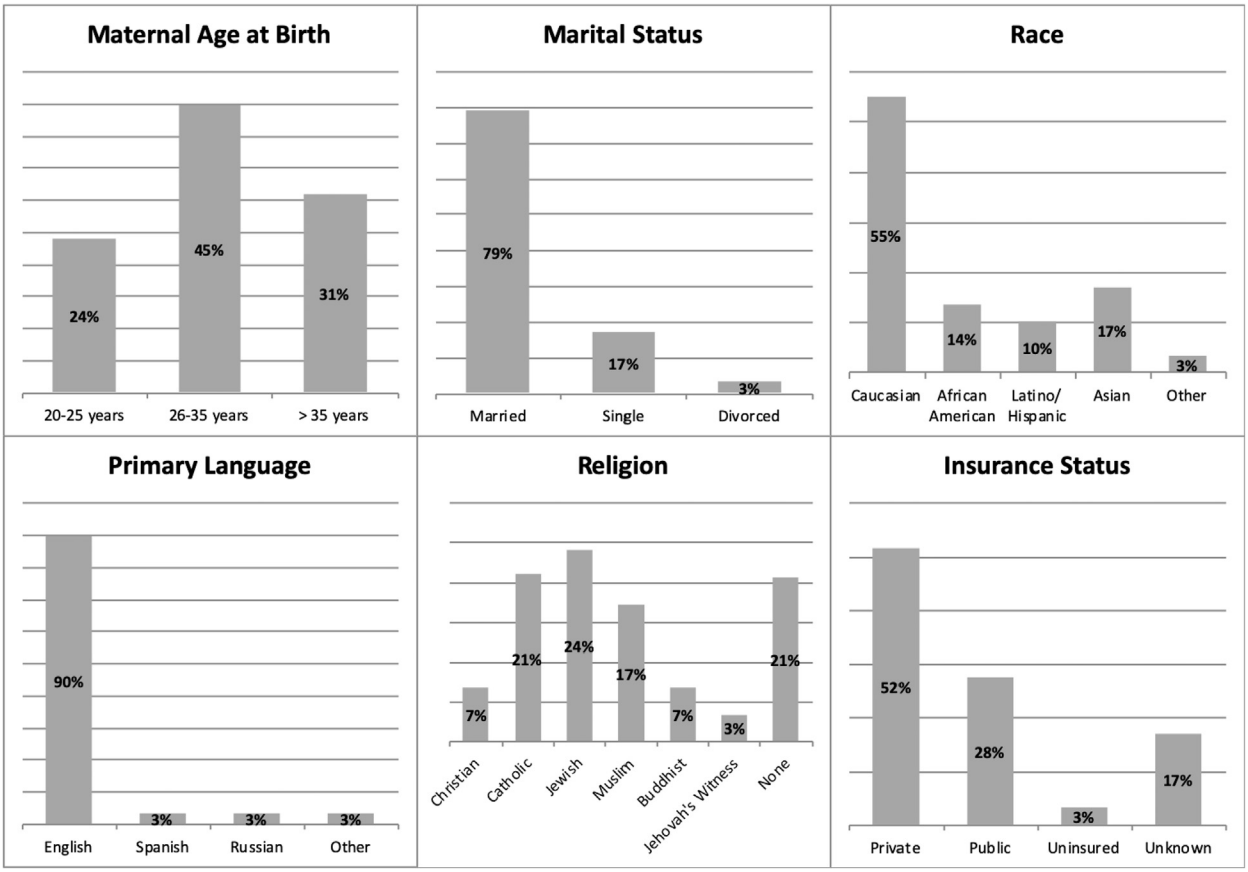

Figure 1. Maternal demographic data for fetuses or neonates (n = 29) with complex SV CHD with normal genetics or nonlife-limiting genetic diagnoses.

VSD (40%), double outlet right ventricle (20%), and truncus arteriosus (20%). Most neonates died within a few days after redirection of goals of care, but 1 neonate with trisomy 18 and VSD was discharged home with hospice care and died 5 months later.

The maternal demographic information is found in Figures 1-3.

Discussion

Our main findings show that a significant number (13.5%) of parents of fetuses or neonates with isolated SV CHD opted for palliative care over SV palliation, despite the fact that NYPMSCH/CUMC has excellent surgical outcomes and survival.

This study also highlights the important finding that some neonates with mixing lesions may survive past the neonatal period under the care of an NPC program. At the time of publication, 2 children whose families had opted for NPC are alive at ages 3 and 5 years with hypoxemia and congestive heart failure, respectively. Both children have had so far, reasonable quality of life by parental report and medical follow-up. Although this long-term outcome is much less likely for a cardiac defect that is ductal dependent for systemic blood flow, this highlights the importance of counseling families about all possible postnatal outcomes when a family opts for either NPC or SV palliation.

Our maternal demographic data (Figures 1-3) demonstrates that families who opt for NPC come from a broad range of backgrounds, highlighting that there is no one type of family that makes this choice.

SV CHD, particularly HLHS, was previously a universally fatal diagnosis. Through the advent of new surgical procedures in the 1980s, survival has drastically improved. Early series of stage I surgical palliation from 1984 to 1999 reported 10- and 15-year survival of 39%, with marked improvement in 3-year survival from 28% in 1984 to 1988 to 66% in 1995 to 1998.[43] Studies investigating new surgical strategies demonstrated improved survival to discharge following stage I palliation from 53% in 1992-1996 to 93% in 1996-2001 with survival to stage II palliation increasing from 44% to 81% in the same cohort.[44] The Single Ventricle Reconstruction Trial followed 549 patients with single ventricle randomized to receive either a Sano or Blalock-Taussig shunt during stage I palliation. In this cohort, 3-year mortality was 33%.[45] The overall transplant free survival at 5 years ranged from 60% to 64%.[34] More recently, a single institution assessment of

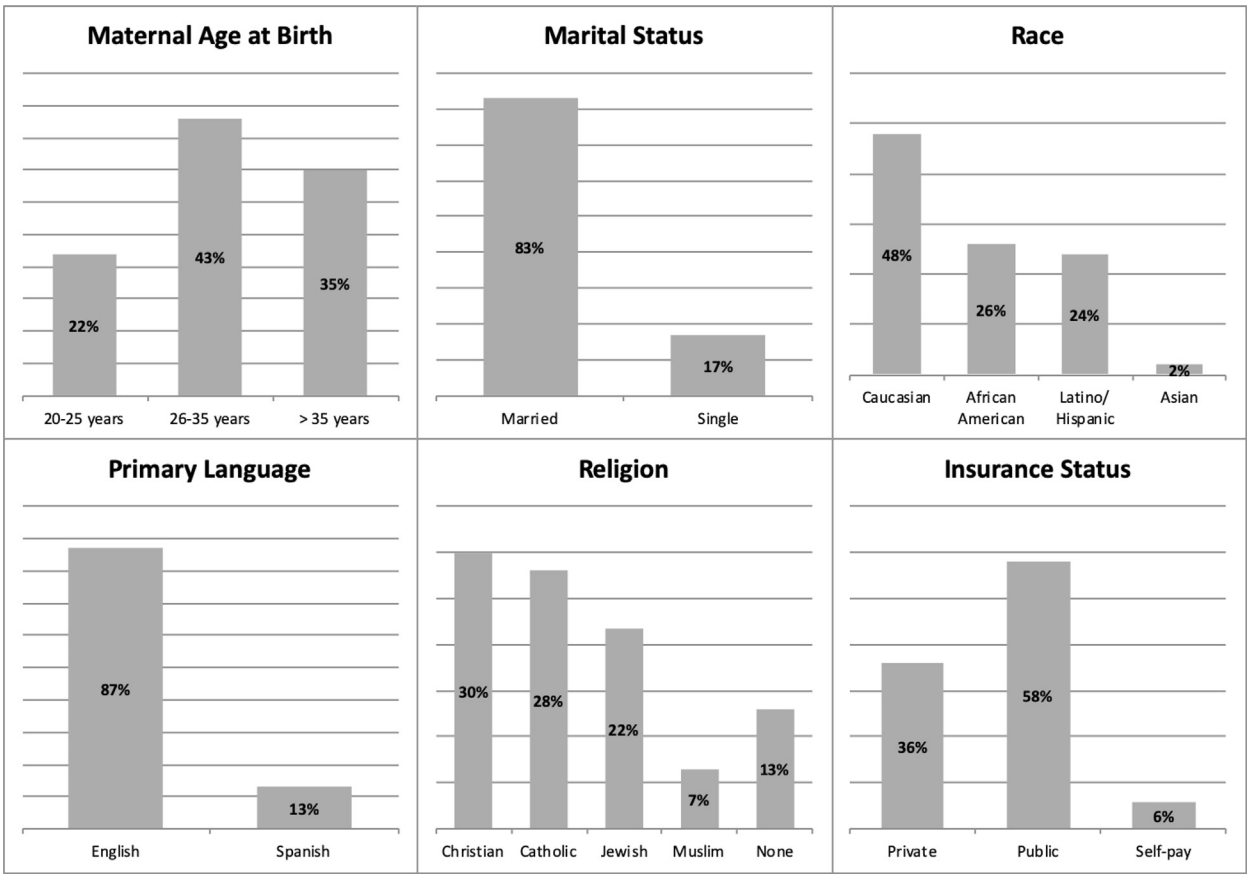

Figure 2. Maternal demographic data for fetuses or neonates (n = 36) with CHD and multi organ dysfunction and/or a severe genetic abnormality.

outcomes from prenatal diagnosis through Fontan operation of 502 patients with SV type CHD demonstrated a 67% survival rate at least 6 months post Fontan surgery in those neonates that survived to birth with intention to treat.[46]

With improvements in survival, larger cohorts of patients with SV are surviving into their third and fourth decade, which has prompted investigation of the medical and neurodevelopmental comorbidities. Early investigations of school-age patients who had undergone stage I palliation prior to 1992 yielded medical comorbidities including 7% with malabsorption or protein-losing enteropathy, 12% with pacemaker, and 64% with chronic medication use.[47] Follow-up studies have documented further comorbidities, including plastic bronchitis, liver fibrosis, cirrhosis and hepatic neoplasm, abnormal bone density, and shorter stature.[18-21,23,48] Multiple studies have demonstrated abnormal neurodevelopmental outcomes including lower Mental Development Index scores, correlating with lower full scale IQ, and even greater decreases in Psychomotor Development Index scores. Rates of intellectual disability range from 6% to 18% and cerebral palsy from 6% to 17%. Neurologic evaluations of children surviving stage III palliation have documented fine motor abnormalities in 49%, gross motor abnormalities in 39%, and speech problems in 30%. Behavioral disorders including attention disorders and mood disorders such as anxiety and depression are common.[24,26,28,29,47,49] Multiple etiologies for the abnormal neurodevelopmental outcomes have been postulated including genetic factors, early postnatal management, and surgical factors such as cardiopulmonary bypass time, deep hypothermic arrest compared with antegrade cerebral perfusion, hemodilution while on cardiopulmonary bypass, and intra- and perioperative monitoring techniques. Emerging research suggests fetal brain growth may be impaired, particularly with HLHS, because of reduced antegrade blood flow through the hypoplastic aorta.[27,50-55] Studies have also documented decreased quality of life scores in a significant percentage of teenage and young adult Fontan patients compared with age-matched controls without chronic health conditions.[56]

Despite improvements in survival, following the prenatal diagnosis of HLHS, there is still a significant percentage of families that opt for termination of pregnancy, ranging from 11% to 49% depending on the gestational age at diagnosis, era, and series.[57-60] Continuation of pregnancy with NPC and no surgical intervention has decreased from nearly 100% in the pre-Norwood era, to 57% in one series from

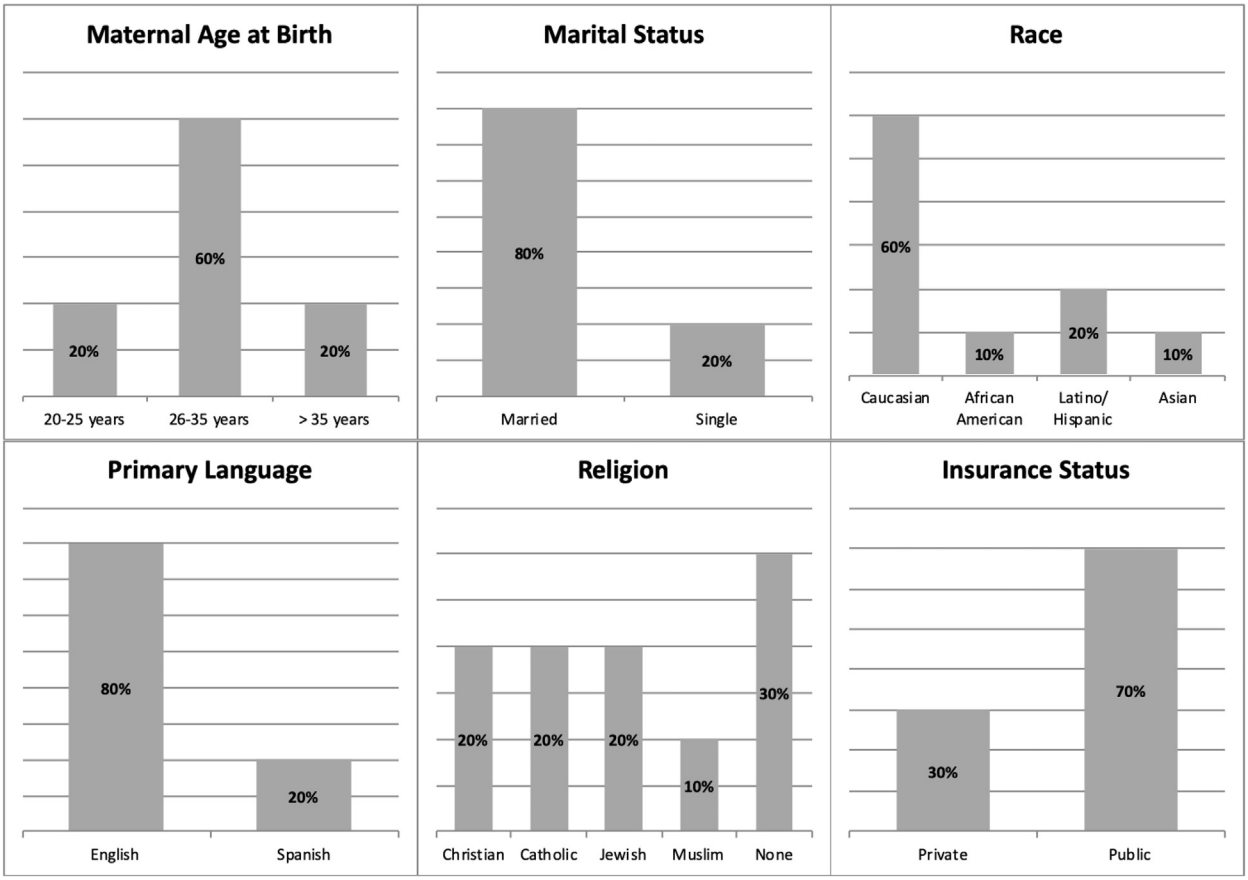

Figure 3. Maternal demographic data for fetuses or neonates (n = 10) with CHD and postnatal diagnosis of a severe genetic diagnosis. Initially intensive care before transitioning to NPC because of an adverse prognosis.

1988-2005, 15% in a series in 2012, and 6% in a series from 2004 to 2011 out of a single institution.[46,58,61,62]

Some practitioners argue that NPC should no longer be offered pre- or postnatally to families with fetuses or neonates with SV CHD because of the improved survival and management strategies for the medical and neurodevelopmental comorbidities.[35] Kon et al have written in support of continuing to discuss NPC with families based on the doctrine of informed consent, which requires that practitioners discuss the treatment options they believe are most appropriate, all reasonable alternatives and what to expect if life-prolonging treatment is declined. He further argues that if only life-prolonging options are presented to families, an inherent disparity develops between advantaged or well-educated families who may pursue additional information and counseling and disadvantaged families who may not do so.[36] In addition, professionals need to discuss with families what treatments would be considered beneficial vs those that would impose an extraordinary burden. Attention must be given to the neonate's 'best interest', given the potential suffering that these neonates will endure over their lifetime. As the survival of children with complex SV defects has improved with surgical and medical advancements, an increasing number of concerning long-term medical, developmental, neurocognitive, and social complications have been identified. However, the full extent of the burden of these complications for these children and their families is only beginning to be delineated.[37-39,63-67]

Multiple studies have investigated the options offered by physicians and surgeons during pre- and postnatal counseling, and the choice practitioners would make for their own hypothetical fetuses or neonates diagnosed with SV CHD. The findings support that NPC remains a reasonable choice. Prsa et al collected nearly 750 anonymous surveys from pediatric cardiologists and cardiac surgeons in North America. Following prenatal diagnosis of HLHS, 57% of practitioners discussed the option of continuation of pregnancy with postnatal NPC, and of the 64% that reported making a recommendation to families, 7.5% recommended either termination of pregnancy or NPC. However, given the hypothetical situation that the practitioner's own fetus was prenatally diagnosed with HLHS, 47% reported they would choose termination of pregnancy or NPC, compared with 24% of whom would pursue surgical palliation. Furthermore, of those given the hypothetical scenario that the practitioner's neonate was postnatally diagnosed with HLHS, 19% would pursue NPC compared with 42% of whom would

pursue surgical palliation. Importantly, most estimated 5-year survival rates in children with HLHS after staged repair at 60%-80% and that most children would have only mild neurodevelopmental delays.[68] Kon performed a study comparing responses of pediatric cardiologists and congenital cardiac surgeons from his prior study in 1999 to the study conducted by Prsa in 2007. When given the hypothetical scenario of their own fetus prenatally diagnosed with HLHS, there was no difference amongst cardiologists over time with 44% reporting they would elect for surgical palliation and 17% pursuing NPC in 1999 compared with 45% and 20%, respectively, in 2007. However, amongst congenital heart surgeons there was a nonstatistically significant trend away from choosing surgery, with 77% reporting they would elect for surgical palliation in 1999 compared with 56% in 2007. Interestingly, among surgeons, 5% reported they would elect for NPC in 1999, which increased to 8% in 2007.[69] Yates surveyed 200 physicians in North America and Europe, of which 45% of institutions offered NPC for HLHS. A statistically significant portion of the surveyed population reported recommending NPC over surgical palliation in HLHS with prematurity (<30 weeks of gestation), chromosomal abnormalities, end organ dysfunction, or low birth weight (<2 kg). In regions outside North America, physicians were more likely to offer NPC for HLHS neonates in the setting of ventricular dysfunction, moderate or greater tricuspid regurgitation, or low birthweight (<2 kg).[70] Paul obtained surveys from nurses, neonatologists, pediatric intensivists and cardiologists in 1995 and 2012 at a single institution. The majority of providers discussed both surgical intervention and NPC with families, including 86% in 1995 and 81% in 2012. The majority also believed that parents should have the option of NPC, including 91% in 1995 and 85% in 2012, however, fewer respondents that discussed NPC encouraged it in 2012 (28%) compared with 1995 (46%). In the 2012 survey, 38% of respondents would choose NPC in the hypothetical scenario that their neonate was born with HLHS.[71]

NPC is a reasonable option as supported by studies of families who have opted for nonsurgical care. Vandvik interviewed 20 mothers whose neonates were born with HLHS, 10 of whom chose NPC and 10 of whom chose surgical palliation and found those mothers who chose NPC had more years of education, reported better childhood environments, and were more likely to be in healthcare services. The primary motive for choosing surgery was a perception that it was the only acceptable choice, whereas the motive for NPC was to prevent suffering.[72] Kuebelbeck outlined her family's decision to pursue NPC for their unborn son prenatally diagnosed with HLHS, describing how she felt that putting him through the pain and long-term suffering associated with surgery was simply too much to ask of their son.[73] The choice for a nonsurgical approach has also been advocated via social media by families who desire to honor the natural life of their neonates while avoiding what they felt was excessive and prolonged suffering.[74]

Our recent mortality data for the Norwood procedure from 2014 to 2017 was 6.6% compared with 13.9% mortality for the Norwood among all congenital heart surgery centers reporting to the Society of Thoracic Surgeons Congenital Heart Surgery Database.[75] Despite these outstanding results, 13.5% of parents opted for NPC over surgical palliation for their neonate diagnosed with isolated SV CHD. Moreover, narratives offered by parents who elected for NPC for their neonate in the NCCP demonstrated great appreciation for the opportunity to be given the nonsurgical option and for the care provided.[42]

Limitations

There are limitations in our conclusions because of the retrospective nature of this study. This is a selected population of families who were given the choice for NPC; however, it is unknown whether all families expecting a fetuses or neonates with SV CHD were given this option. At NYPMSCH/CUMC as many as 6 independent fetal cardiologists perform prenatal counseling. Although practitioners are expected to discuss the option of NPC to the appropriate families, the counseling is not identical and, therefore, may have unknown effects on the options for which families elect. Thus, this study may underestimate the potential number of families electing NPC for their neonate, if given the choice. Moreover, practitioners could consider a prospective study of parental decision-making comparing the effects of standard counseling vs standard counseling plus a formal NPC consult.

This study shows that a significant number of parents, if given the option, elect for a palliative care plan for their neonates diagnosed with complex CHD, including SV CHD, even in a large tertiary cardiac center with excellent surgical results and survival. These findings, along with occasional quality-life longevity in patients with SV CHD mixed lesions treated with palliative care, suggest that NPC is still a reasonable choice and this option should be discussed and offered to families. The demographic data presented highlight that there is no typical family that opts for the NPC choice.

Further studies are warranted to identify the full extent of the life-long medical, social, and emotional burden for children treated with surgical palliation and their families that would justify such a choice. ■

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Data Statement

Data sharing statement available at www.jpeds.com.

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