

Hypoplastic Left Heart Syndrome: Exploring a Paradigm Shift in Favor of Surgery

Erin A. Paul¹  · Kristina Orfali² · Thomas J. Starc¹

Received: 13 March 2016 / Accepted: 12 August 2016 / Published online: 27 August 2016
© Springer Science+Business Media New York 2016

Abstract We hypothesized that enthusiasm for surgery increased for infants with hypoplastic left heart syndrome (HLHS) at Columbia University Medical Center (CUMC) between 1995 and 2012. We sought to identify factors that engendered this paradigm shift. Confidential surveys were distributed to providers at CUMC in 1995 and 2012 to measure enthusiasm for surgical intervention for HLHS. Surgical preference scores are presented as median [interquartile range]. Surveys were completed by 99/176 providers (56 % response rate) in 1995 and 153/267 (57 %) in 2012. The median surgical preference score for infants with HLHS increased from 35 [25–45] in 1995 to 45 [35–50] in 2012, $P < 0.001$. 53 %, 95 % CI [42, 64] of respondents recommended surgical intervention for a ward of the court in 1995 compared to 81 % [73, 89] in 2012, $P < 0.001$. In 2012, 64 % [53, 75] of respondents were more likely to recommend surgery than 10 years prior. The percentage of respondents who saw good outcomes following three-stage repair increased from 49 % [38, 60] in 1995 to 84 % [78, 90] in 2012, $P < 0.001$. The majority believed that parents should have the option of comfort

care, 91 % [85, 97] in 1995 and 85 % [79, 91] in 2012, $P = 0.06$. In both eras, prematurity and additional surgical problems dissuaded providers from recommending surgical intervention. Despite the fact that most providers have seen good outcomes and now recommend surgery for infants with HLHS, the majority of providers still believe that the option of comfort care should be available to families.

Keywords Hypoplastic left heart syndrome · Comfort care · Congenital heart disease · Counseling · Cardiac surgery

Introduction

Hypoplastic left heart syndrome (HLHS) occurs in approximately 0.016 % of live births and accounts for 4–11 % of congenital heart disease [1–3]. This malformation generally consists of hypoplasia of the left ventricle, mitral and aortic valves and a diminutive ascending aorta. This condition is uniformly fatal without surgical intervention, and prior to 1979, comfort care was the standard therapy for newborns with HLHS. Since the introduction of the Norwood procedure, infant survival rates have steadily increased from 21 to 30 % [4, 5] in the early 1980s to 58 % [6] in the 1990s and 84 % [7] in the mid-2000s.

Although utilization of the Norwood procedure has increased over time, nationwide data between 1988 and 2005 from academic and community hospitals suggested that approximately 50 % of newborns received comfort care only [8]. A more recent database study performed between 2000 and 2012 showed that 32 % of newborns with HLHS did not receive surgical intervention [9]. Some viewpoint papers have proposed that comfort care is no longer a reasonable or even ethical option to offer to

Electronic supplementary material The online version of this article (doi:10.1007/s00246-016-1455-y) contains supplementary material, which is available to authorized users.

✉ Erin A. Paul
eas2188@columbia.edu

¹ Division of Pediatric Cardiology, Department of Pediatrics, New York-Presbyterian Morgan Stanley Children's Hospital, Columbia University Medical Center, 3959 Broadway, CHN-253, New York, NY 10032, USA

² Division of Neonatology and Bioethics, Department of Pediatrics, New York-Presbyterian Morgan Stanley Children's Hospital, Columbia University Medical Center, 3959 Broadway, CHN-253, New York, NY 10032, USA

parents of infants with HLHS [10], while others argue the necessity of presenting all management options to facilitate informed decision making [11–13]. Several surveys have assessed counseling patterns, but factors impacting provider recommendations are not fully understood [14–16]. One survey compared how pediatric cardiologists and cardiothoracic surgeons counseled in 1999 and 2007 and found no significant self-reported difference in management recommendations over that time period [17]. Our study aims to compare trends in attitudes at a single institution over time. We hypothesized that enthusiasm for surgery for infants with HLHS increased at CUMC between 1995 and 2012. Our secondary goal was to explore possible factors affecting this change.

Materials and Methods

Confidential surveys were distributed to nurses, cardiologists and neonatologists in August 1995 [18] and to nurses, cardiologists, neonatologists and pediatric intensivists from September to November 2012. Management questions were based on provider attitudes toward care of a hypothetical newborn with HLHS who would be an “ideal candidate” for surgery without any complicating medical or psychosocial features. The 1995 and 2012 surveys collected similar demographic data, asked seven overlapping preference questions and eight common Likert scale questions. Demographic data included respondents’ age, gender, clinical role, department, years in practice, parental status and experience with outcomes. Both demographic data and response rates were compared across study periods using Chi-square tests. The surveys can be reviewed as online supplemental information.

Of the seven preference questions, five addressed what providers would discuss with and recommend to families or to a judge in the case of a ward of the court. For most of these questions, management options included the Norwood procedure, transplantation, comfort care or some combination thereof. Comfort care was defined as pain control and comfort measures without life-prolonging intervention. The remaining two shared preference questions addressed how strongly providers felt that surgery should be performed and how strongly they believed that comfort care should be available.

A surgical preference score was devised to compare the seven preference questions shared between the survey periods. For each of these questions, respondents were awarded zero, five or ten points. If respondents omitted one of these questions, they were awarded the average score for that question. Resulting composite scores ranged from zero to seventy. A score of zero reflects a maximal preference for comfort care, and seventy represents a maximal surgical

preference. Preference scores are reported as median [interquartile range] and were compared across survey periods using Mann–Whitney tests. Responses to individual questions that were shared across the survey periods are reported as %, 95 % CI [confidence interval] and were compared using Chi-square tests or Fisher 3×2 tables.

There were eight Likert scale questions shared between survey periods focusing on complicating medical and psychosocial factors that might influence provider advice. For each five-point Likert scale question, respondents received a score ranging from -2 to $+2$ corresponding with factors that would make them “much less likely” or “much more likely” to recommend surgery. Scores were compared between survey periods using *T* tests.

Pediatric intensivists were included in the survey in 2012 alone and were excluded from analyses involving comparison between survey periods.

There were several questions unique to the 2012 survey. Providers were asked to estimate rates of cognitive delay and survival. Rates are reported as mean \pm standard deviation (SD). Linear regressions were used to relate surgical preference score to the aforementioned estimates.

In 2012, providers were also asked what management option they would choose if their own newborn was diagnosed with HLHS. Linear regressions were used to relate providers’ theoretical personal choice with individual surgical preference scores, survival and cognitive delay estimates, gender, age, years of experience, experience with good outcomes and parental status.

All statistical analyses were performed using Epi InfoTM [19].

This study was reviewed by the CUMC institutional review board and was deemed exempt.

Results

Demographics

Surveys were completed by 99/176 providers (56 % response rate) in 1995 and 153/267 (57 %) in 2012. Table 1 shows respondents’ demographics. There was no significant difference in response rates between physicians and nurses in 1995 (61 vs. 54 %, $P = 0.51$). However, in 2012, 74 % of physicians responded compared to 44 % of nurses ($P < 0.001$). There were approximately equal numbers of nurses but a larger number of fellows and attendings in 2012. A female predominance among respondents was preserved in both survey periods. Age distribution was skewed toward both respondents less than 30 years of age and those greater than 50 years of age in 2012 compared to 1995. Number of years of experience was also skewed

Table 1 Respondent characteristics

Year	1995 <i>n</i> (%)	2012 <i>n</i> (%)
Role*	(<i>n</i> = 99)	(<i>n</i> = 153)
Nurses	65 (66 %)	66 (43 %)
Fellows	12 (12 %)	31 (20 %)
Attending	22 (22 %)	56 (37 %)
Gender	(<i>n</i> = 99)	(<i>n</i> = 152)
Male	17 (17 %)	39 (26 %)
Female	82 (83 %)	113 (74 %)
Age in years**	(<i>n</i> = 99)	(<i>n</i> = 146)
<30	8 (8 %)	30 (21 %)
31–40	54 (54 %)	52 (36 %)
41–50	27 (27 %)	22 (15 %)
>50	10 (10 %)	42 (29 %)
Years of experience**	(<i>n</i> = 99)	(<i>n</i> = 145)
0–5	12 (12 %)	47 (32 %)
6–10	32 (32 %)	35 (24 %)
11–15	24 (24 %)	9 (6 %)
16–20	12 (12 %)	14 (10 %)
>20	19 (19 %)	40 (28 %)

Distribution of respondents shown for each survey year as a raw number and percent of total

* $P = 0.002$; ** $P < 0.001$

toward more respondents with zero to 5 years of experience or more than 20 years of experience in 2012.

Surgical Preference Scores

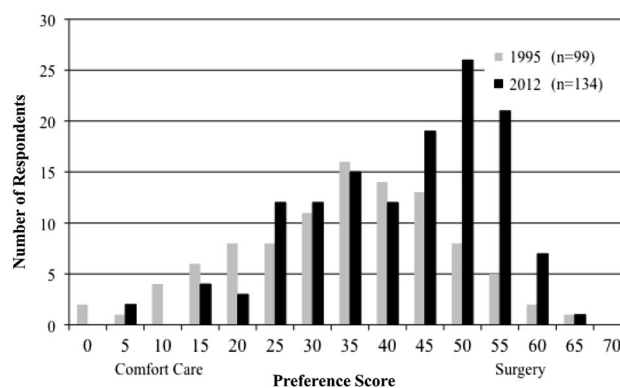
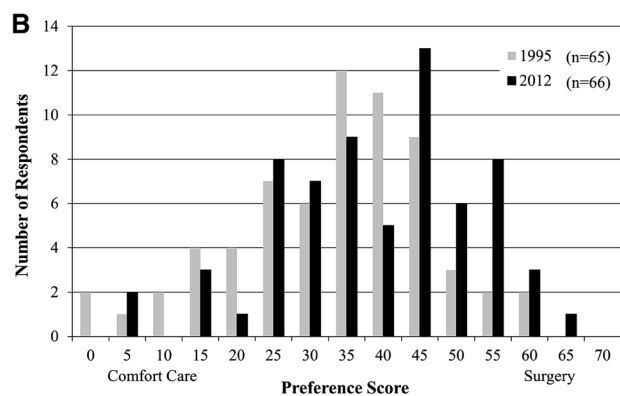
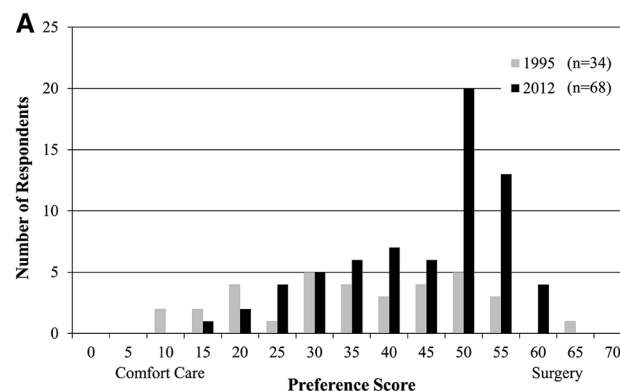
In response to management questions for an ideal candidate with HLHS, the median surgical preference score increased from 35 [25–45] in 1995 to 45 [35–50] in 2012 ($P < 0.001$) (Table 2; Fig. 1). Surgical preference scores increased significantly for physicians from 35 [26–49] in 1995 to 50 [35–51] in 2012 ($P = 0.003$; Fig. 2a), and for nurses from 35 [25–40] to 40 [29–50] ($P = 0.02$; Fig. 2b). This shift was also significant for cardiologists (40 [30–50] in 1995 vs. 50 [40–55] in 2012, $P = 0.03$), neonatologists (30 [21–35] in 1995 vs. 48 [35–50] in 2012, $P = 0.02$) and NICU nurses (29 [20–35] in 1995 vs. 36 [30–45] in 2012, $P = 0.004$). NICU nurses' surgical preference scores were

Table 2 Shift in surgical preference scores

	1995	2012	<i>P</i>
All respondents	35 [25–45]	45 [35–50]	<0.001
All physicians ^a	35 [26–49]	50 [35–51]	0.003
All nurses	35 [25–40]	40 [29–50]	0.02

Data are presented as median [interquartile range]

^a PICU physicians were only surveyed in 2012 and are excluded from comparisons between the study periods

**Fig. 1** Surgical preference scores in 1995 and 2012 for all respondents**Fig. 2** Surgical preference scores in 1995 and 2012 for **a** physicians and **b** nurses

significantly lower than PICU nurses' scores in 1995 (29 [20–35] vs. 43 [36–45], $P < 0.001$, *T* test), but this difference was no longer significant in 2012 (36 [30–45] vs. 35 [25–49], $P = 0.77$, *T* test).

Provider Counseling and Recommendations

Providers were asked what management options they would discuss with families. The majority of providers discussed both surgical intervention and comfort care with

families in both eras (86 %, 95 % CI [79, 93] in 1995 and 81 %, 95 % CI [74, 88] in 2012). Few providers discussed surgery in isolation (7 %, 95 % CI [2, 12] in 1995 and 16 %, 95 % CI [10, 22] in 2012). Even fewer discussed comfort care in isolation (7 %, 95 % CI [2, 12] in 1995 and 3 %, 95 % CI [0, 6] in 2012). None of these differences were statistically significant ($P = 0.06$).

Providers were asked what they would recommend to families if they were asked for their opinion. Many providers did not feel comfortable dictating management, but of those who offered advice 74 %, 95 % CI [59, 89] would recommend surgical intervention in 1995 compared to 90 %, 95 % CI [84, 96] in 2012 ($P = 0.04$) (Table 3).

This trend was significant when providers were asked to recommend management for a ward of the court; 53 %, 95 % CI [42, 64] of respondents recommended surgical intervention in 1995 compared to 81 %, 95 % CI [73, 89] in 2012 ($P < 0.001$). There was also a shift in how many providers felt comfortable offering any advice to families. Only 35 % of respondents offered advice for an ideal candidate in 1995 and that increased to 60 % in 2012. In contrast, in both survey periods, over 75 % of respondents offered advice for a ward of the court.

Provider Experience and Opinion

If respondents discussed the Norwood procedure, most encouraged surgery in both survey periods (57 %, 95 % CI [47, 67] in 1995, 70 %, 95 % CI [62, 78] in 2012, $P = 0.05$). There was no statistically significant shift in the percentage of respondents who felt strongly that some type of surgery should be performed for most infants with HLHS (52 %, 95 % CI [42, 62] in 1995 vs. 69 %, 95 % CI [61, 77] in 2012, $P = 0.06$). However, 64 %, 95 % CI [53, 75] of respondents in 2012 felt that they would be more likely to recommend surgery currently than they would have been 10 years ago. 85 %, 95 % CI [79, 91] of respondents in 2012 disagreed with the statement that parents who refused surgical intervention should be taken to court.

The percentage of respondents who had seen a patient with a good outcome following three-stage repair increased

from 49 %, 95 % CI [38, 60] in 1995 to 84 %, 95 % CI [78, 90] in 2012 ($P < 0.001$). However, there was no significant correlation between experience with a good outcome and surgical preference score ($R = 0.10$, $P = 0.30$). The surgical preference scores were not significantly different for those respondents who had not seen a good outcome compared to those respondents who had (42 [33–50] vs. 45 [35–50], respectively, $P = 0.17$). Seventeen of the twenty respondents who had not seen a good outcome were NICU affiliated.

Of those respondents who discussed comfort care, fewer respondents encouraged it in 2012 (46 %, 95 % CI [36, 56] in 1995 vs. 28 %, 95 % CI [20, 36] in 2012, $P = 0.01$). In both eras, the majority of providers believed that parents should have the option of choosing comfort care (91 %, 95 % CI [85, 97] in 1995 and 85 %, 95 % CI [79, 91] in 2012, $P = 0.06$).

Impact Factors

All psychosocial or medical factors that made providers less likely to recommend surgery in 1995 became less negative in 2012 (Table 4). In the case of an unmarried mother, a mother in poor health or a family who had previously lost a newborn with HLHS, physicians were statistically significantly less likely to be dissuaded from recommending surgery in 2012 than in 1995.

In both eras, providers were more likely to recommend surgery for a newborn of older parents who have no other children and used in vitro fertilization (IVF) for the pregnancy. Prematurity and other surgical problems tended to dissuade providers from recommending surgical intervention in both 1995 and 2012.

Survival and Cognitive Delay Estimates

The mean five-year survival estimate following three-stage surgical intervention was 68 ± 14 % and positively correlated with surgical preference score ($R = 0.20$, $P = 0.01$). On average, respondents estimated that 40 ± 22 % of school age survivors of three-stage repair

Table 3 Percent of providers offering advice who would recommend surgical intervention in 1995 and 2012

	1995		2012		<i>P</i>
	<i>n/N</i>	% [95% CI]	<i>n/N</i>	% [95% CI]	
Ideal patient	25/34	74 % [59, 89]	73/81	90 % [84, 96]	0.04
Ward of the court	40/76	53 % [42, 64]	83/103	81 % [73, 89]	<0.001
Respondent's child	n/a	n/a	62/100	62 % [52, 72]	n/a

n number of respondents recommending surgical intervention, *N* number of respondents giving management advice

Table 4 Factors impacting provider advice

	1995	2012	<i>P</i>
Psychosocial factors			
Single mother	−0.22	0.04	0.002
Parents previously lost child to HLHS	−0.56	−0.27	0.009
Mother in poor health	−0.56	−0.32	0.03
Poor financial status of parents	−0.26	−0.09	0.06
First child of young parents	−0.13	+0.04	0.07
Older parents who used IVF for the pregnancy	+0.38	+0.27	0.28
Medical factors			
Newborn with additional surgical problem	−0.76	−0.65	0.31
Prematurity <32 weeks gestation	−0.98	−0.93	0.68

Average scores for each Likert scale question are shown for both survey periods

would have severe cognitive delays. This cognitive delay estimate negatively correlated with surgical preference score ($R = -0.30$, $P < 0.001$).

Providers' Theoretical Personal Choice

In 2012, respondents were asked what management option they would choose if they personally had a newborn with HLHS. Of the 153 survey participants, 53 were unable to or preferred not to answer, while 100 chose a management option. Of those who chose a management option, 62 % , 95 % CI [52, 72] chose surgery, while 38 % , 95 % CI [28, 48] chose comfort care.

The median surgical preference score was 50 [45–55] for those respondents who would choose surgical intervention for their own infant compared to 30 [20–39] for those who would choose comfort care for their own infant ($P < 0.001$).

Personal choice of surgical management positively correlated with surgical preference score ($R = 0.70$, $P < 0.001$), age ($R = 0.26$, $P = 0.01$) and years of experience ($R = 0.28$, $P = 0.006$) and negatively with estimates of severe cognitive delays ($R = -0.32$, $P < 0.001$). No significant correlation was found between personal choice and gender, experience with good outcomes, parental status or survival estimates.

Discussion

Management for newborns with HLHS has evolved over the last three decades both nationally and at CUMC. Our institution has offered and performed cardiac transplantation, three-staged palliative surgery and comfort care for newborns with HLHS. This study uniquely shows the change in attitudes at our medical center toward surgical intervention for newborns with HLHS over 17 years of experience and identifies factors that may have contributed to this change.

Shifts in Surgical Preference

Significant shifts in surgical preference scores suggest a change in provider attitude at CUMC between 1995 and 2012. Although this study was not designed to illustrate an individual's shift of opinion, it shows an institutional evolution over time. NICU nurses had the lowest median score of any group in 1995 and a median score similar to that of other groups in 2012. NICU providers at CUMC care for patients preoperatively and following the Norwood procedure. We speculate that their exposure to poorer outcomes in earlier eras contributed to their relatively pessimistic outlook in 1995 and that improved outcomes have extinguished the difference between NICU nurses and their PICU counterparts in 2012.

Counseling Trends

Most but not all providers discuss all available management options with families, and this has persisted over time. The survey results suggest that providers are more comfortable offering management recommendations to a judge in the case of a ward of the court compared to offering an opinion directly to a family. In the absence of a legal guardian, a physician may feel free to offer input without jeopardizing their therapeutic relationship or feeling paternalistic. One may conjecture that in the case of the ward of the court, there is no conflict with parental autonomy, and the medical provider may feel free to identify the plan of action that most closely meets the best interest standard for that patient.

Provider Experience and Opinion

Although we did not find a relationship between having seen a good outcome and respondents' surgical preference scores, this may be secondary to a small effect size and limited power. The question only captures whether or not

respondents have seen a single good outcome. Multiple positive experiences may be necessary to change one's outlook. The majority of providers who had not seen a good outcome were NICU affiliates consistent with this population's lack of exposure to older survivors.

Impact Factors

In 2012, respondents were less likely to be deterred from recommending surgery in the face of complicating psychosocial factors as compared to 1995. This may correspond with respondents' growing preference for surgical intervention. Medical complications, however, persistently dissuaded providers from recommending surgery.

Survival and Cognitive Impairment Estimates

Respondents' average five-year survival estimate of $68 \pm 14\%$ approximates recently published 5-year survival of 60 or 64% [20] depending upon shunt type in a North American cohort enrolled between 2005 and 2008.

On average, respondents estimated that $40 \pm 22\%$ of survivors have severe cognitive delay at school age. Fourteen months following the Norwood procedure up to 44% of survivors may have severe developmental delay as defined by the psychomotor disability index [21], but longer-term studies of children following the Fontan procedure suggest a more optimistic outlook with 9–18% having severe impairment [22, 23]. While a national survey of neonatologists, pediatric cardiologists, intensivists and surgeons had previously failed to show a relationship between estimates of surgical survival or rates of cognitive delay and counseling practices [15], our study showed weak but statistically significant relationships between surgical preference scores and both survival and cognitive impairment estimates. It is essential to recognize provider perception of outcomes as a factor impacting counseling and to underscore the need for continuous reeducation as outcomes evolve.

Personal Choice

When providers were asked what management option they would pursue if their own newborn were born with HLHS, the majority of respondents chose surgical intervention.

Counseling practices, as reflected by surgical preference scores, correlated with providers' personal choice. However, a smaller proportion of providers chose surgical intervention for their own child as compared to the proportion of respondents who recommended surgery when giving advice to families. Table 3 shows percentages of providers recommending surgery in the case of an "ideal" patient, a ward of the court or their own newborn. A prior

study of cardiologists and pediatric cardiothoracic surgeons also showed a correlation between personal choice and counseling trends [14], while another survey with a smaller sample size did not show this relationship [15].

Providers who were older or who had more years of experience were more likely to choose surgery for their own newborn than younger providers. The statistically significant but low R-values correlating age and years of experience with surgical preference underscore that these factors may only account for a small portion of the variation in management decisions. Those who predicted poor cognitive outcomes were more likely to choose comfort care. Choosing surgical intervention for one's own newborn did not significantly correlate with having seen a good outcome. These results suggest that although providers are less willing to pursue surgical intervention for their own child than they are to recommend it to families, preference for surgical intervention may vary with age. This shift may be secondary to the cumulative effect of observing several positive patient experiences or from seeing outcomes improve over time.

Limitations

Our study focuses on provider attitudes as reported in a survey, which may differ from actual counseling and management practices. In fact, some of our respondents are not routinely in a position to counsel families. Much of our discussion is theoretical due to the paucity of data on real-time counseling practices for newborns with HLHS.

Our response rates were approximately 55% in both survey years, and this may have led to a potential sampling bias. Previously published physician surveys have similar response rates [24, 25] including those regarding HLHS [14–16].

As a tertiary care center, CUMC sees a referral base that is likely skewed toward families pursuing surgical intervention, and this study may not be generalizable to other centers. Between 2006 and 2014, just 8% of HLHS patients at CUMC were diagnosed postnatally. Because this survey is designed to assess counseling practices for a postnatally diagnosed newborn who is an ideal candidate for surgical intervention, it does not address the questions that arise with prenatal diagnosis.

Conclusion

As outcomes have improved over the past 17 years, there has been a significant shift in provider attitudes at CUMC toward surgical intervention for newborns with HLHS. In addition, more providers have had personal experience with good three-stage surgical outcomes. Although most

providers may not recommend comfort care at this time, the majority of providers believe that the option of comfort care should still be provided to families. We speculate that as outcomes continue to improve, provider opinion will continue to shift, and comfort care may become a reasonable option for high-risk patients alone. Further research is needed to better elucidate factors affecting counseling and to uncover recent trends in management.

Acknowledgments This study was supported in part by an internal grant from the pediatric residency program in the Department of Pediatrics at New York-Presbyterian Morgan Stanley Children's Hospital, Columbia University Medical Center.

Compliance with Ethical Standards

Conflicts of interest None.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this type of study, formal consent is not required. This article does not contain any studies with animals performed by any of the authors.

References

- Fyler DC (1980) Report of the New England regional infant cardiac program. *Pediatrics* 65(2 Pt 2):375–461
- Samanek M, Slavik Z, Zborilova B, Hrobonova V, Voriskova M, Skovranek J (1989) Prevalence, treatment, and outcome of heart disease in live-born children: a prospective analysis of 91,823 live-born children. *Pediatr Cardiol* 10(4):205–211
- Morris CD, Outcalt J, Menashe VD (1990) Hypoplastic left heart syndrome: natural history in a geographically defined population. *Pediatrics* 85(6):977–983
- Gutgesell HP (1985) Current policies for management of infants with hypoplastic left heart syndrome. *Am Heart J* 110(3):703
- Sade RM, Crawford FA Jr, Fyfe DA (1986) Symposium on hypoplastic left heart syndrome. *J Thorac Cardiovasc Surg* 91(6):937–939
- Gutgesell HP, Gibson J (2002) Management of hypoplastic left heart syndrome in the 1990s. *Am J Cardiol* 89(7):842–846
- Ghanayem NS, Allen KR, Tabbutt S, Atz AM, Clabby ML, Cooper DS, Eghtesady P, Frommelt PC, Gruber PJ, Hill KD, Kaltman JR, Laussen PC, Lewis AB, Lurito KJ, Minich LL, Ohye RG, Schonbeck JV, Schwartz SM, Singh RK, Goldberg CS (2012) Interstage mortality after the Norwood procedure: results of the multicenter single ventricle reconstruction trial. *J Thorac Cardiovasc Surg* 144(4):896–906
- Karamlou T, Diggs BS, Ungerleider RM, Welke KF (2010) Evolution of treatment options and outcomes for hypoplastic left heart syndrome over an 18-year period. *J Thorac Cardiovasc Surg* 139(1):119–126
- Kane JM, Canar J, Kalinowski V, Johnson TJ, Hoehn KS (2016) Management options and outcomes for neonatal hypoplastic left heart syndrome in the early twenty-first century. *Pediatr Cardiol* 37(2):419–425
- Wernovsky G (2008) The paradigm shift toward surgical intervention for neonates with hypoplastic left heart syndrome. *Arch Pediatr Adolesc Med* 162(9):849–854
- Kon AA (2008) Healthcare providers must offer palliative treatment to parents of neonates with hypoplastic left heart syndrome. *Arch Pediatr Adolesc Med* 162(9):844–848
- Ross LF, Frader J (2009) Hypoplastic left heart syndrome: a paradigm case for examining conscientious objection in pediatric practice. *J Pediatr* 155(1):12–15
- Feudtner C (2008) Ethics in the midst of therapeutic evolution. *Arch Pediatr Adolesc Med* 162(9):854–857
- Prsa M, Holly CD, Carnevale FA, Justino H, Rohlicek CV (2010) Attitudes and practices of cardiologists and surgeons who manage HLHS. *Pediatrics* 125(3):e625–e630
- Kon AA, Ackerson L, Lo B (2004) How pediatricians counsel parents when no “best-choice” management exists: lessons to be learned from hypoplastic left heart syndrome. *Arch Pediatr Adolesc Med* 158(5):436–441
- Renella P, Chang RK, Ferry DA, Bart RD, Sklansky MS (2007) Hypoplastic left heart syndrome: attitudes among pediatric residents and nurses towards fetal and neonatal management. *Prenat Diagn* 27(11):1045–1055
- Kon AA, Prsa M, Rohlicek CV (2012) Choices doctors would make if their infant had hypoplastic left heart syndrome: comparison of survey data from 1999 and 2007. *Pediatr Cardiol* 34(2):348–353
- Starc TJ, Peacock J, Driscoll J, Gersony WM (1996) Attitudes towards treatment of infants with hypoplastic left heart syndrome. *Pediatr Res* 39(42A):237
- Dean AGAT, Sunki GG, Friedman R, Lantinga M, Sangam S, Zubieta JC, Sullivan KM, Brendel KA, Gao Z, Fontaine N, Shu M, Fuller G, Smith DC, Nitschke DA, Fagan RF (2011) *Epi Info™*, a database and statistics program for public health professionals, 7th edn. CDC, Atlanta
- Newburger JW, Sleeper LA, Frommelt PC, Pearson GD, Mahle WT, Chen S, Dunbar-Masterson C, Mital S, Williams IA, Ghanayem NS, Goldberg CS, Jacobs JP, Krawczeski CD, Lewis AB, Pasquali SK, Pizarro C, Gruber PJ, Atz AM, Khaikin S, Gaynor JW, Ohye RG, Pediatric Heart Network I (2014) Transplantation-free survival and interventions at 3 years in the single ventricle reconstruction trial. *Circulation* 129(20):2013–2020
- Newburger JW, Sleeper LA, Bellinger DC, Goldberg CS, Tabbutt S, Lu M, Mussatto KA, Williams IA, Gustafson KE, Mital S, Pike N, Sood E, Mahle WT, Cooper DS, Dunbar-Masterson C, Krawczeski CD, Lewis A, Menon SC, Pemberton VL, Ravishanker C, Atz TW, Ohye RG, Gaynor JW (2012) Early developmental outcome in children with hypoplastic left heart syndrome and related anomalies: the single ventricle reconstruction trial. *Circulation* 125(17):2081–2091
- Mahle WT, Clancy RR, Moss EM, Gerdes M, Jobes DR, Wernovsky G (2000) Neurodevelopmental outcome and lifestyle assessment in school-aged and adolescent children with hypoplastic left heart syndrome. *Pediatrics* 105(5):1082–1089
- Brosig C, Mussatto K, Hoffman G, Hoffmann RG, Dasgupta M, Tweddell J, Ghanayem N (2013) Neurodevelopmental outcomes for children with hypoplastic left heart syndrome at the age of 5 years. *Pediatr Cardiol* 34(7):1597–1604
- Asch DA, Jedrzejewski MK, Christakis NA (1997) Response rates to mail surveys published in medical journals. *J Clin Epidemiol* 50(10):1129–1136
- Cummings SM, Savitz LA, Konrad TR (2001) Reported response rates to mailed physician questionnaires. *Health Serv Res* 35(6):1347–1355