

# Perception scores of siblings and parents of children with hypoplastic left heart syndrome

Elizabeth C. Caris MD<sup>1</sup>  | Nicole Dempster PhD<sup>2</sup> | Gil Wernovsky MD<sup>3</sup> |  
Yongjie Miao MS<sup>2</sup> | Melissa Moore-Clingenpeel MA MAS<sup>2</sup> | Trent Neely<sup>2</sup> |  
Rachel Fonseca BS<sup>2</sup> | Holly Miller-Tate RN<sup>2</sup> | Robin Allen RN<sup>2</sup> |  
Samantha Fichtner RN<sup>2</sup> | Jamie Stewart RN<sup>2</sup> | Clifford L. Cua MD<sup>2</sup> 

<sup>1</sup>Children's Hospital of Pittsburgh of UPMC, Pittsburgh, Pennsylvania, USA

<sup>2</sup>Nationwide Children's Hospital, Columbus, Ohio, USA

<sup>3</sup>Children's National Medical Center, Washington, Washington, D.C., USA

## Correspondence

Elizabeth Caris, MD, 4401 Penn Avenue, 5th Floor Faculty Pavilion, Pittsburgh, PA 15224.

Email: elizabeth.caris@chp.edu

## Abstract

**Objectives:** Siblings of children with chronic medical conditions endorse a lower quality of life compared to age-matched peers. Caregiver and sibling-self report of adjustment are often discordant. Congenital heart disease significantly affects family life. To date, there have been no studies addressing the functioning of siblings of children with hypoplastic left heart syndrome, one of the most severe forms of congenital heart disease. The goal of this study was to assess the impact of hypoplastic left heart syndrome on sibling's quality of life as well as the caregiver's perception of this effect.

**Study Design:** Cross-sectional study using a web-based survey distributed via various listservs targeted towards families of children with hypoplastic left heart syndrome. Employed the Sibling Perception Questionnaire, designed to assess sibling and caregiver perceptions of adjustment to chronic illness. A Negative Adjustment Composite Score was calculated for each respondent, with higher values representing more negative adjustment.

**Results:** Thirty-five caregivers responded. Majority of caregivers were female (74%), white (86%) and college educated (54%). Thirty-two siblings participated, ranging in age from 7 to 30 years of age ( $12.5 \pm 6.3$ ). Most children with hypoplastic left heart syndrome (73%) had undergone the third stage of palliation. Forty-two caregiver-sibling pairs were examined. Caregiver Negative Adjustment Composite Scores were significantly higher than sibling scores, with caregivers reporting more adjustment problems ( $2.4 \pm 0.4$ ) than siblings ( $2.3 \pm 0.3$ ,  $P < .05$ ). Sibling age was correlated with worse caregiver and sibling scores ( $r 0.35$ ,  $P < .05$ ).

**Conclusions:** Caregivers of children with hypoplastic left heart syndrome perceive their siblings as struggling more than the children self-report. Siblings tend to report worse adjustment as they get older. These data suggest that programs should include support for the entire family through all ages to optimize quality of life.

## KEYWORDS

congenital heart disease, hypoplastic left heart syndrome, siblings, survey

## 1 | INTRODUCTION

As pediatric medicine continues to advance, there are more children living with chronic diseases that require intensive management. One of

the most common forms of chronic illness is congenital heart disease (CHD), with an estimated 1.4 million individuals living with some form of CHD in the United States.<sup>1</sup> Hypoplastic left heart syndrome (HLHS) is one of the most severe forms of CHD. With advancement in surgical palliation techniques, survival has increased. With improved outcomes,

increased emphasis has been placed on family and patient quality of life.<sup>2,3</sup>

With this shift in focus, there is more awareness of the impact of chronic illness on family and sibling functioning. There have been multiple studies that have looked at the adjustment, coping and quality of life of siblings of children with different chronic diseases.<sup>4-6</sup> The data is mixed, but several recent meta-analyses have suggested that siblings do endorse a lower quality of life compared to age-matched peers, and their parents generally have a more negative view of their children's quality of life than they self-report.<sup>7,8</sup> In these studies, parents report that siblings of children with chronic health issues have more issues with both internalizing (anxiety, depression) and externalizing (behavior problems, aggression) behaviors than controls.

There are several studies that suggest that caring for a child with HLHS is more stressful than other chronic diseases.<sup>9,10</sup> In studies that compare sibling's quality of life across different chronic illnesses, congenital heart disease demonstrates a more negative effect than cystic fibrosis, cancer and diabetes.<sup>11</sup> Specifically, siblings of patients with tetralogy of Fallot and univentricular heart disease reported more behavioral and internalizing problems compared to other chronic illnesses. To date, there have been no studies specifically addressing the quality of life and functioning of siblings of children with HLHS.

The current study was designed to assess the impact of HLHS on sibling's quality of life as well as the caregiver's perception of this effect. It also sought to identify factors associated with more negative adjustment in both siblings and caregivers.

## 2 | METHODS

This was a prospective cross-sectional study. A web-based survey was distributed via various listservs and social network sites targeted towards families affected by HLHS. Consent by the caregiver and siblings was assumed to occur if the survey was completed. IRB determined that this study did not fit the definition of human subject research under 45 CFR part 46.102(f) therefore IRB evaluation was waived.

The first part of the survey consisted of questions relating to the basic demographics of the caregivers, siblings and children with HLHS. Data was also collected about the medical history of the child with HLHS. The second part of the survey consisted of the Sibling Perception Questionnaire (SPQ).<sup>12</sup> This questionnaire was initially designed to assess the adjustment of siblings of children with cancer but has been used with siblings and caregivers of children with other medical problems.<sup>13-15</sup> It consists of 21 questions addressing communication, intra- and interpersonal relationships as well as fear related to the child's medical condition and contact with other siblings. The first caregiver was given the opportunity to complete the SPQ for each sibling in the family. A second caregiver was then given the opportunity to answer the same questions about each sibling. Then each sibling in turn was asked to complete a parallel version of the SPQ. For each caregiver and sibling that completed the survey, the three scales that reflect negative responses to their sibling's medical problems were

**TABLE 1** Caregiver demographics

	N = 35 (%)
Age	38.7 ± 6.8
Gender	
Male	9 (25.7)
Female	26 (74.3)
Race	
White	30 (85.7)
Hispanic	1 (2.9)
Black	1 (2.9)
Other	3 (8.6)
Completed education	
High school	4 (11.4)
Technical school	5 (14.3)
College/university	19 (54.3)
Graduate/professional school	7 (20.0)
Personal status	
Single	1 (2.9)
Married	30 (85.7)
Divorced	3 (8.6)

summed, as it has been shown to be more internally consistent than the individual scales themselves.<sup>13</sup> This created a Negative Adjustment Composite Score,<sup>13,16</sup> with values ranging from 1 (never) to 4 (a lot), with higher values representing more negative adjustment. For purposes of this study, only siblings older than age five were invited to participate.

Means and standard deviations were calculated for all continuous measures unless otherwise stated. Sample N's and percentages were calculated for categorical measures. To examine the relationship between caregiver and sibling responses, sibling Negative Adjustment Composite Scores were subtracted from the caregivers' scores to generate a discrepancy score.<sup>13,17</sup> This discrepancy score was compared to the standard deviation of the discrepancy score for the sample as a whole. A concordant pair was defined as a caregiver/sibling discrepancy score within 0.5 SD of zero.

The discordant pairs were then further categorized to determine the nature of the difference. Pairs with raw discrepancy scores greater than 0.5 SD above zero indicate that the siblings reported fewer problems than caregivers, and those with raw discrepancy scores greater than 0.5 SD below zero indicate that siblings reported more problems with adjustment than their caregivers. Paired tests or signed-rank tests, where appropriate, were used to compare Negative Adjustment Composite Scores between parents and siblings within each family. One-way ANOVA, Kruskal-Wallis, and Fisher's exact tests were performed to determine differences between the sibling-caregiver pairs with respect to sibling, caregiver and family characteristics.

## 3 | RESULTS

A total of 35 caregivers from 26 unique families responded to the survey. The majority of caregivers that responded were white, married, college educated females (Table 1). Nine families had more than one caregiver respond to the survey and six had two siblings respond to the survey for a total of 32 siblings. The majority of families consisted

TABLE 2 Family characteristics

	N (%)
Number of children in family	
2	12 (46.2)
3	12 (46.2)
4	1 (3.8)
5	1 (3.8)
Sibling age	12.5 ± 6.3
Sibling gender	
Male	12 (37.5)
Female	20 (62.5)
Distance from surgical hospital	
Less than 25 miles	5 (19.2)
25–50 miles	1 (3.9)
50–100 miles	10 (38.5)
100–150 miles	4 (15.4)
More than 150 miles	6 (23.1)
State/country	
USA northeast	5 (19.2)
USA southeast	4 (15.4)
USA midwest	3 (11.5)
USA south	5 (19.2)
USA west	1 (3.9)
Europe	2 (7.7)
Other	6 (23.1)

of two caregivers (87%) and 2–3 children (92%). Thirty-two siblings participated (12 boys, 20 girls). Siblings ranged in age from 7 to 30 years of age (12.5 ± 6.3) (Table 2). The 26 children with HLHS ranged in age from 20 months to 27 years old (8.3 ± 5.7). Most of the children with HLHS (73%) had undergone the third stage of palliation and almost half (42%) of the children with HLHS had other medical issues (Table 3).

With 35 total caregivers responding and 32 total siblings responding, there were 42 total caregiver-sibling pairs to examine. Caregiver mean scores on the Negative Adjustment Composite Score were significantly higher than the siblings scores, with caregivers tending to report more adjustment problems (2.4 ± 0.4) than siblings (2.3 ± 0.3;  $P < .05$ ). Most of the pairs were discordant (N 27) and categorized as  $S < C$  (N 20), indicating siblings overall reported fewer problems with adjustment than caregivers (Table 4).

The sibling Negative Adjustment Composite Score was positively correlated with their age with a trend toward significance ( $P = .08$ ,  $r = 0.28$ ) indicating that siblings tend to report worse adjustment as they grow older. Sibling age was also significantly correlated with caregiver Negative Adjustment Composite Score ( $r = 0.35$ ,  $P = .025$ ), with caregiver Negative Adjustment Composite Score increasing as sibling age increases (Table 5).

There were no significant differences between discordance categories based on caregiver characteristics such as race ( $P = .16$ ), education ( $P = .4$ ), gender ( $P = .12$ ) or marital status ( $P = .09$ ). There were also no significant differences between discordance categories based on sibling gender ( $P = .17$ ), stage of surgical repair ( $P = .10$ ), the presence of additional medical issues ( $P = .07$ ), distance from surgical hospital ( $P = .87$ ), or availability of home nursing care ( $P = .84$ ).

## 4 | DISCUSSION

HLHS is a severe form of CHD that has lifelong effects on all aspects of life for the patients and their families. There is a growing body of literature addressing quality of life, anxiety, and stress experienced by patients and their caregivers coping with this chronic illness. While current literature acknowledges chronic illness has adverse impacts on healthy siblings, no study to our knowledge has included families with complex congenital heart disease, such as HLHS. This study shows that siblings are affected by this form of chronic illness consistent with previous studies in other chronic disease states.<sup>8</sup> In addition, caregiver perception of this effect is often different than sibling self-report, usually with the parents rating a more negative effect than the siblings of children with HLHS.

This study found that caregivers rate siblings as having more difficulties with overall adjustment than the siblings self-report. This in contrast to previous work with siblings of children with cancer, which suggested that siblings experience a more serious burden from illness than their caregivers perceive.<sup>16,18</sup> In both studies, the authors note that the correspondence between parent-proxy and child self-report measures of quality of life are low to moderate at best. Furthermore, a recent meta-analysis looking at fifty-one published studies pertaining to the siblings of children with a chronic illness overall suggested that caregiver reports of adjustment and quality of life were more negative than child self-reports.<sup>7</sup> The reasons for the discrepancy between caregiver and sibling-self report of adjustment and quality of life are likely multifactorial. It is possible that caregivers tend to generalize the deficits or challenges faced by the child with HLHS to the sibling, or the healthy sibling may be used to minimizing their needs when compared to their sibling with HLHS. Previous studies have suggested that the discrepancies observed are related to lack of parental availability to the well sibling,<sup>18</sup> shifting family dynamics in the face of an ill sibling,<sup>19</sup> and adjustment of parental expectations for the healthy sibling's behavior.<sup>20</sup> Future studies are needed to better understand these perceptions and

TABLE 3 Characteristics of the child with HLHS

	N (%)
Age of child with HLHS	8.3 ± 7.7
Gender of child with HLHS	
Male	15 (57.7)
Female	11 (42.3)
Last completed surgery	
Stage II	6 (23.1)
Stage III	19 (73.1)
Transplant	1 (3.8)
Other medical issues?	
Yes	11 (42.3)
No	15 (57.7)
Estimated lifetime total in-hospital stay	
Less than 3 months	7 (26.9)
3–5 months	8 (30.8)
5–7 months	4 (15.4)
7–9 months	2 (7.7)
More than 9 months	5 (19.2)

discrepancies to ensure that both the healthy and chronically ill child's needs are being met.

The perception of adjustment was further modified by the age of respondents. There was a trend towards a positive correlation between siblings' Negative Adjustment Composite Scores and sibling age. There was also a significant positive correlation between sibling and caregiver Negative Adjustment Composite Scores, which would indicate that as siblings experience more difficulty with adjustment, caregivers do tend to recognize this. Previous studies show that siblings are most affected by chronic illness soon after their family member is diagnosed, and as time passes, adjustment and quality of life improve.<sup>18</sup> However, age does seem to play a role in reported adjustment and quality of life. Younger siblings tend to report more physical complaints compared to adolescent siblings who tend to report more emotional and psychosocial issues.<sup>8</sup>

In the current study, a wide age range of siblings responded about a wide age range of HLHS patients. Younger children may be less able to perceive negative effects and judge adjustment than their parents, whereas older children have more personal insight into these issues. As HLHS patients grow older, once they have completed the initial three stages of palliation, more developmental delays and long term medical side effects of their altered hemodynamics may become evident. This may create a long term unpredictable environment for families and siblings that can pose more challenges to positive adjustment and warrant intervention.

No significant differences based on family and patient characteristics were found between the different discordance groups. The survey administered focused on issues that seem pertinent to family functioning such as distance from their surgical hospital and availability of home nursing help. Parental education and socioeconomic status have been shown to be important predictors of overall morbidity and mortality in children with HLHS.<sup>21,22</sup> Families and siblings affected by HLHS most likely have additional, issues that affect their day to day life that the current survey did not elicit. Future studies should evaluate sibling-specific characteristics and other family dynamics that influence adjustment such as their own comorbidities, cognitive struggles or other

**TABLE 4** Sibling and caregiver Negative Adjustment Composite Score (NACS) by discordance category

	Total sample	Concordant pairs S = C	Discordant pairs		
			S ≠ C	S < C	S > C
Sample size	42	15	27	20	7
Sibling report	2.3 ± 0.3	2.4 ± 0.3	2.3 ± 0.4	2.3 ± 0.3	2.3 ± 0.4
Caregiver report	2.4 ± 0.4	2.4 ± 0.3	2.5 ± 0.5	2.6 ± 0.4	2.0 ± 0.4
P = .03					

NACS score ranges from 1–4; higher scores indicate more adjustment problems. Abbreviations: SPQ, Sibling Perception Questionnaire; S, sibling; C, caregiver.

**TABLE 5** Pearson correlation coefficients

	Age of Patients with HLHS	Sibling Age	Caregiver NACS	Sibling NACS
Age of Patients with HLHS	–	0.75**	0.13	0.17
Sibling age		–	0.35*	0.28
Caregiver NACS			–	0.73**
Sibling NACS				–

N = 42 \*P < .05; \*\*P < .01.

health and social issues. There is a need for more comprehensive and routine screening to identify families and siblings that are struggling.

There are several limitations raised by the current study. Given that the data was collected via anonymous web-based survey and social media sites, we are unable to evaluate the response rate. Therefore, there was a selection bias among caregivers that chose to respond. The caregiver sample that chose to respond were predominantly white, married, college educated women. The questionnaire was also only administered in English. All of these factors make it difficult to generalize our findings to a broader demographic. Most of the children with HLHS had completed their third palliative surgery, so perceptions of caregivers and siblings during the earlier stages of palliation are less well delineated. The overall sample size for both caregivers and siblings was small, thus this should be considered a preliminary analysis of these familial perceptions. More robust statistics could not be performed due to the small sample size. In addition, this analysis did not account for family clustering. A more systematic sampling of families with two parents and multiple siblings would be required for this type of analysis. Despite these limitations, this study represents a first step in recognizing the issues facing families affected by HLHS, and future studies will be needed to further evaluate sibling and family functioning. Currently, clinic based screening practices are being developed at our institution to identify at-risk families and inform them of these dynamics.

In conclusion, siblings of children with HLHS tend to report less negative adjustment compared to parental assessment. Both caregivers and siblings tend to report more negative adjustment over time. These data suggest that interventions that aim to improve functioning and quality of life in families affected by HLHS should include siblings, and continue throughout childhood and adolescence. Families may benefit from strategies to improve communication early on, and siblings may benefit from ongoing discussions about future medical issues and challenges that their family member may face in the future.

#### CONFLICT OF INTEREST

None.

#### AUTHOR CONTRIBUTIONS

Elizabeth Caris, Nicole Dempster, and Clifford Cua contributed to the conception and design of the project. Gil Wernovsky, Trent Neely, Robin Allen, Jamie Stewart, Holly Miller-Tate, Rachel Fonseca,

Samantha Fichtner, Melissa Moore-Clingenpeel, and Yongjie Miao contributed to the acquisition, analysis, and interpretation of the data. All authors were involved in the drafting and revision of the manuscript as well as final approval. All authors take full accountability for the contents of the manuscript.

## ORCID

Elizabeth C. Caris MD  <http://orcid.org/0000-0002-2138-4328>

Clifford L. Cua MD  <http://orcid.org/0000-0002-9172-3874>

## REFERENCES

- [1] Botto L. Epidemiology and Prevention of Congenital Heart Defects. In: Allen H, Shaddy R, Penny D, Feltes T, Cetta F, eds. *Moss & Adams' Heart Disease in Infants, Children and Adolescents, Including the Fetus and Young Adult*. 9th ed. Philadelphia: Wolters Kluwer; 2016:55–81.
- [2] Feinstein JA, Benson DW, Dubin AM, et al. Hypoplastic left heart syndrome: current considerations and expectations. *J Am Coll Cardiol*. 2012;59(1):S1–42.
- [3] Williams DL, Gelijns AC, Moskowitz AJ, et al. Hypoplastic left heart syndrome: valuing the survival. *J Thorac Cardiovasc Surg*. 2000;119(4):720–731.
- [4] Woodgate RL, Edwards M, Ripat JD, Rempel G, Johnson SF. Siblings of children with complex care needs: their perspectives and experiences of participating in everyday life. *Child Care Health Dev*. 2016;42(4):504–512.
- [5] Lavigne J, Ryan M. Psychologic adjustment of siblings of children with chronic illness. *Pediatrics*. 1979;63(4):616–627.
- [6] Houtzager BA, Grootenhuis MA, Last BF. Adjustment of siblings to childhood cancer: a literature review. *Support Care Cancer*. 1999;7(5):302–320.
- [7] Sharpe D. Siblings of children with a chronic illness: a meta-analysis. *J Pediatr Psychol*. 2002;27(8):699–710.
- [8] Vermaes IPR, Van Susante AMJ, Van Bakel HJA. Psychological functioning of siblings in families of children with chronic health conditions: A meta-analysis. *J Pediatr Psychol*. 2012;37(2):166–184.
- [9] Caris EC, Dempster N, Wernovsky G, et al. Anxiety scores in caregivers of children with hypoplastic left heart syndrome. *Congenit Heart Dis*. 2016;11(6):727–732.
- [10] Brosig CL, Mussatto KA, Kuhn EM, Tweddell JS. Psychosocial outcomes for preschool children and families after surgery for complex congenital heart disease. *Pediatr Cardiol*. 2007;28(4):255–262.
- [11] Havermans T, Croock Vercruyse DI, Goethals T, Diest E, Van IV. Belgian siblings of children with a chronic illness: Is their quality of life different from their peers? *J Child Heal Care*. 2015;19(2):154–166.
- [12] Sahler O, Carpenter P. Evaluation of a camp program for siblings of children with cancer. *Am J Dis Child*. 1989;143(6):690–696.
- [13] Guite J, Lobato D, Kao B, Plante W. Discordance between sibling and parent reports of the impact of chronic illness and disability on siblings. *Child Health Care*. 2004;33(1):77–92.
- [14] Lobato D, Kao BT. Integrated sibling parent group intervention to improve sibling knowledge and adjustment to chronic illness and disability. *J Pediatr Psychol*. 2002;27(8):711–716.
- [15] Hodapp RM, Wijma CA, Masino LL. Families of children with 5p-(cri du chat) syndrome: Familial stress and sibling reactions. *Dev Med Child Neurol*. 2008;39(11):757–761.
- [16] Schulte F, Wurz A, Reynolds K, Strother D, Dewey D. Quality of life in survivors of pediatric cancer and their siblings: the consensus between parent-proxy and self-reports. *Pediatr Blood Cancer*. 2016;63(4):677–683.
- [17] Garber J, Van Slyke DA, Walker LS. Concordance between mothers' and children's reports of somatic and emotional symptoms in patients with recurrent abdominal pain or emotional disorders. *J Abnorm Child Psychol*. 1998;26(5):381–391.
- [18] Houtzager BA, Grootenhuis MA, Caron HN, Last BF. Sibling self-report, parental proxies, and quality of life: the importance of multiple informants for siblings of a critically ill child. *Pediatr Hematol Oncol*. 2005;22(1):25–40.
- [19] Cordaro G, Veneroni L, Massimino M, Clerici CA. Assessing psychological adjustment in siblings of children with cancer: Parents' perspectives. *Cancer Nurs*. 2012;35(1):E42–E50.
- [20] Yang H-C, Mu P-F, Sheng C-C, Chen Y-W, Hung G-Y. A systematic review of the experiences of siblings of children with cancer. *Cancer Nurs*. 2016;39(3):E12–E21.
- [21] Bucholz EM, Sleeper LA, Newburger JW. Neighborhood socioeconomic status and outcomes following the norwood procedure: An analysis of the pediatric Heart Network Single Ventricle Reconstruction Trial Public data set. *J Am Heart Assoc*. 2018;7(3):e007065–e007010.
- [22] Taylor LC, Burke B, Donohue JE, et al. Risk factors for interstage mortality following the norwood procedure: impact of sociodemographic factors. *Pediatr Cardiol*. 2016;37(1):68–75.

**How to cite this article:** Caris EC, Dempster N, Wernovsky G, et al. Perception scores of siblings and parents of children with hypoplastic left heart syndrome. *Congenital Heart Disease*. 2018;13:528–532. <https://doi.org/10.1111/chd.12619>