How Pediatricians Counsel Parents When No “Best-Choice” Management Exists

Lessons to Be Learned From Hypoplastic Left Heart Syndrome

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Background: Hypoplastic left heart syndrome (HLHS) is a life-threatening congenital cardiac defect. Three mutually exclusive management options exist: the Norwood palliative procedure, cardiac transplantation, and comfort care without surgical intervention.

Objectives: To assess which management options are presented to parents of infants with HLHS, and to determine what factors influence physicians’ recommendations.

Design: Cross-sectional survey.

Setting: Fourteen of the largest pediatric cardiac surgery centers in the United States.

Participants: Attending physicians in neonatology, cardiology, critical care practice, and cardiac surgery.

Intervention: A survey was distributed asking physicians what options they present to parents of infants with HLHS and what their recommendations are in general, as well as physician perceptions of HLHS outcomes and demographic information.

Main Outcome Measures: Which options physicians discuss and which they recommend.

Results: Of 454 eligible physicians 257 (57%) responded to the survey, of which 110 make treatment recommendations to parents. Neonatologists were least likely to recommend surgery. Physicians who recommend surgery recommend procedures performed at their own institution over those performed elsewhere (odds ratio, 2.80; 95% confidence interval, 2.24-3.51). Twenty-six percent of physicians do not discuss nonsurgical management, and 25% of those at centers that do not perform cardiac transplantation do not discuss this option. The recommendations physicians make to parents are poorly associated with their predictions of postoperative outcomes.

Conclusions: Physician recommendations to parents are poorly associated with their estimates of outcomes, and some physicians recommend the treatment preferred at their own institution over other options even when they predict better outcomes from another approach. Further, many physicians do not disclose all reasonable management options to parents of infants with HLHS. These findings raise doubts as to whether parents are given adequate information to make truly informed decisions.

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I N THE CARE OF INFANTS, PARENTS or guardians need to be given sufficient information about the treatment alternatives and their likely outcomes to make informed decisions regarding the child’s medical care.1-3 Physicians provide information to parents and frequently make treatment recommendations, which are presumably based on evidence regarding benefits, risks, and outcomes. Little is known regarding the influence of physician characteristics on how options are presented and what recommendations are made. In particular, it is not known how pediatricians handle the informed permission process when there is uncertainty over the preferred course of care and evidence on outcomes is inconclusive.

We studied how physicians obtain informed permission in hypoplastic left heart syndrome (HLHS) when parents of affected infants must make management decisions within the first few days of life. Because parents have little time to educate themselves or to seek multiple medical opinions, they may rely heavily on physicians’ advice. While an in utero diagnosis may afford a greater opportunity for parent-physician dialogue and prenatal counseling, even in the best of circumstances, parents must make decisions quickly and under significant stress. In HLHS, several accepted, mutually exclusive management options exist: the Norwood palliative procedure (PP), cardiac transplantation, and comfort care without surgical intervention. For both of the
surgical alternatives, intention-to-treat 1-year survival is approximately 60% (recent reports at meetings, however, suggest that postoperative outcomes are continually improving) and both require multiple postoperative admissions and repeated surgical procedures. Survivors of either surgical procedure have significantly limited physical endurance and are at substantial risk of cognitive impairment. Approximately 55% of parents forego surgery in their affected infants, and when presented with a hypothetical scenario in which they themselves are the parent of an infant with HLHS, physicians are split between the Norwood PP, cardiac transplantation, and comfort care.

We surveyed physicians who obtain informed permission for treatment of infants with HLHS at high-volume pediatric cardiac surgery centers across the United States. We asked how they counsel these parents and what management recommendations they make. Furthermore, we assessed what factors influence these recommendations. We were particularly interested in the relative importance of perceived outcomes of surgery compared with the physicians’ characteristics and type of practice.

METHODS

STUDY DESIGN

A 10-page survey was constructed and mailed to all attending physicians in the sections of neonatology, pediatric critical care, pediatric cardiology, and pediatric/congenital cardiac surgery at 14 of the largest pediatric cardiac surgery centers in the United States. The survey consisted of closed-response and open-ended questions. It was pretested on a group of pediatric cardiologists, pediatric intensivists, and pediatric cardiac surgeons at a high-volume children’s hospital that was excluded from this study.

Each physician was assigned a unique identifying number. That number was added to the return envelope of each questionnaire before mailing. As each questionnaire was returned, the receipt of the response was noted on the master list. Questionnaires were not marked with identification numbers to preserve the confidentiality of the respondents. Surveys were sent in the fall of 1999 along with a stamped return envelope. All nonresponders were sent a follow-up letter as well as a second copy of the survey via fax. The research protocol was approved by the University of California, San Francisco, Committee on Human Research.

STUDY SITE SELECTION

Major US pediatric cardiac centers were grouped into 3 tiers based on volume of HLHS admissions: modest, moderate, and high volume. Because admission data are unavailable for all US congenital cardiac surgery sites, grouping was done by consensus of an expert committee of neonatologists, pediatric intensivists, pediatric cardiologists, and pediatric cardiac surgeons at the University of California, San Francisco. The 14 centers grouped into the high-volume tier were selected as study sites and are listed in the “Categorization of Study Sites” section.

MAIN OUTCOME VARIABLES

The primary outcome variable was which management option the physician recommends to parents of infants with HLHS.

This variable was dichotomized into those who recommend either of the 2 surgical options and those who either recommend comfort care without surgery or recommend both surgery and comfort care equally. Subgroup analyses were then performed comparing those who recommend the Norwood PP with those who recommend cardiac transplantation, and comparing those who recommend comfort care without surgery with those who recommend both surgery and comfort care equally.

INDEPENDENT VARIABLES

Independent variables included physician specialty, whether the physician provides long-term postoperative follow-up care for children with HLHS, religious affiliation, stated choice if their own child were diagnosed as having HLHS, race/ethnicity, sex, years in practice, parental status, site of practice, and 6 “predicted postoperative outcome” variables: the physician’s predictions of 1-year postoperative survival, cognitive outcome (percentage of long-term survivors with an IQ of ≥70), and physical limitation in survivors (percentage of teenage survivors with only mild or no physical limitations) for both the Norwood PP and cardiac transplantation. Questions regarding religious affiliation and race/ethnicity were open ended. These responses were coded for inclusion in the univariate and multivariable analyses. For the site of practice variable, institutions were grouped into 3 categories based on physician responses: centers that perform the Norwood PP nearly exclusively (Norwood centers); centers in which most patients receive Norwood PP (however, many also receive cardiac transplantation); centers that perform both procedures (transplantation centers).

STATISTICAL ANALYSES

Because multiple physicians were surveyed within each facility, our analyses were designed to consider the likelihood that the responses of physicians within the same facility will be more similar than responses from physicians at different facilities. To adjust for this, all analyses were done using multilevel models using a generalized estimating equation model (STATA 7.0 Special Edition, xige statistical command; StataCorp LP, College Station, Tex). These models modify all error terms by considering the within-facility correlation with clustering at the level of the institution.

Hierarchical logistic regression modeling was used and is reported as the odds ratio with 95% confidence interval. Statistical significance was set at 5%.

RESULTS

CHARACTERISTICS OF RESPONDENTS

A total of 454 physicians met inclusion criteria and were sent surveys. Two hundred fifty-seven physicians (57%) responded. Of these, 110 stated that they generally make treatment recommendations to parents, and they are the subjects of our analyses. Physicians who stated that they do not make treatment recommendations were excluded from the study. Table 1 gives the physicians’ characteristics. The distribution of physician specialty was similar in the responder and nonresponder cohorts. Because respondents in different specialties differ in whether they discuss treatment options with parents, 74% of the surgeons responded that they discuss treatment options with parents and were included in the analyses compared with 65% of the cardiologists, 25% of the neona-
Facilities grouped as Norwood centers were The Alfred I. duPont Hospital for Children, Wilmington, Del; University of California, San Francisco; Cleveland Clinic Foundation, Cleveland, Ohio; Seattle Children’s Hospital, Seattle, Wash; Children’s Hospital of Philadelphia, Philadelphia, Pa; Boston Children’s Hospital, Boston, Mass; Babies & Children’s Hospital of New York–New York Presbyterian Hospital, New York; Texas Children’s Hospital, Houston; C. S. Mott Children’s Hospital, Ann Arbor, Mich; and Miami Children’s Hospital, Miami, Fla. Facilities grouped as providing both surgical procedures were the Mattel Children’s Hospital at the University of California Los Angeles, and St Louis Children’s Hospital, St Louis, Mo. Those grouped as cardiac transplant centers were Loma Linda University Children’s Hospital, Loma Linda, Calif, and The Children’s Hospital, Denver, Colo.

### Table 1. Physician Characteristics

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value†</th>
</tr>
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<tbody>
<tr>
<td>Specialty</td>
<td></td>
</tr>
<tr>
<td>Cardiologist</td>
<td>72 (65)</td>
</tr>
<tr>
<td>Surgeon</td>
<td>17 (15)</td>
</tr>
<tr>
<td>Neonatologist</td>
<td>16 (15)</td>
</tr>
<tr>
<td>Intensivist</td>
<td>5 (5)</td>
</tr>
<tr>
<td>Site of practice</td>
<td></td>
</tr>
<tr>
<td>Norwood PP center</td>
<td>87 (79)</td>
</tr>
<tr>
<td>Norwood PP and cardiac transplantation center</td>
<td>12 (11)</td>
</tr>
<tr>
<td>Cardiac transplantation center</td>
<td>11 (10)</td>
</tr>
<tr>
<td>Personal choice</td>
<td></td>
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<tr>
<td>Norwood PP</td>
<td>47 (43)</td>
</tr>
<tr>
<td>Cardiac transplantation</td>
<td>4 (3)</td>
</tr>
<tr>
<td>Norwood PP as bridge to cardiac transplantation</td>
<td>2 (2)</td>
</tr>
<tr>
<td>Comfort care</td>
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</tr>
<tr>
<td>Uncertain</td>
<td>39 (36)</td>
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<tr>
<td>Religion</td>
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<tr>
<td>Other religion</td>
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<tr>
<td>No religious affiliation</td>
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<tr>
<td>Race/ethnicity</td>
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<tr>
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<tr>
<td>Asian</td>
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<tr>
<td>Sex</td>
<td></td>
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<tr>
<td>Male</td>
<td>80 (73)</td>
</tr>
<tr>
<td>Female</td>
<td>29 (27)</td>
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<tr>
<td>Parental status</td>
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<tr>
<td>Have children</td>
<td>96 (88)</td>
</tr>
<tr>
<td>Do not have children</td>
<td>13 (12)</td>
</tr>
<tr>
<td>Time in practice, mean (SD), y</td>
<td>12.3 (8.0)</td>
</tr>
</tbody>
</table>

Abbreviation: PP, palliative procedure.
*Data are given as the number (percentage) of respondents unless otherwise indicated.
†Some physicians did not answer all of the questions. Percentages are based on those physicians responding to each question.

### PREDICTIONS OF POSTOPERATIVE OUTCOMES

Predictions of postoperative outcomes varied significantly. Predicted Norwood PP 1-year survival ranged from 30% to 88% (mean [SD], 68% [14%]), predicted percentage of Norwood PP survivors with an IQ of 70 or higher ranged from 50% to 100% (mean [SD], 85% [12%]), and predicted teenaged Norwood PP survivors with only mild or no physical limitation ranged from 0% to 100% (mean [SD], 52% [27%]). Predicted cardiac transplantation 1-year survival ranged from 5% to 90% (mean [SD], 56% [19%]), predicted percentage of transplantation survivors with an IQ of 70 or higher ranged from 5% to 100% (mean [SD], 89% [9%]), and predicted teenaged cardiac transplantation survivors with only mild or no physical limitation ranged from 0% to 100% (mean [SD], 75% [23%]).

### TREATMENT OPTIONS PRESENTED TO PARENTS

Sixty-four percent of the respondents discuss all options. All respondents discuss the Norwood PP. Twenty-five percent (n=22) of respondents at Norwood PP centers do not discuss cardiac transplantation as an option. Twenty-six percent of physicians (n=29) do not discuss with parents the option of comfort care without surgery. In fact, 7 physicians (18%) who were uncertain if they would choose surgery or comfort care without surgery for their own child do not discuss comfort care with parents, and 1 physician who would choose comfort care for his own child stated that he does not discuss this option with parents. This physician wrote on the survey form: “It’s not politically possible to discuss [comfort care without surgery] once the baby has been born. My suspicion is that any family requesting [comfort care without surgery] would be told by the Chief of Cardiac Surgery and the Chief of Cardiology that they could not sanction such a request.”

### VARIABLES ASSOCIATED WITH RECOMMENDING SURGERY

Eighty-five percent of the physicians (n=94) recommend surgery over comfort care. Physician specialty was most highly associated with the recommendation made—100% of the surgeons and intensivists recommend surgery compared with 90% of the cardiologists and 44% of the neonatologists. This strong association remained after adjusting for the other variables in the multivariable model (Table 2). Site of practice, religious affiliation, race/ethnicity, sex, and parental status were not associated with the recommendation made in the multivariable analysis; years in practice was weakly associated (odds ratio [OR], 0.99; P=.03).

Among those physicians who would choose surgery for their own affected child, 96% recommend surgery to parents, 4% recommend both options equally, and none recommend comfort care over surgery. Among those who would choose comfort care without surgery, 53% recommend surgery over comfort care to parents, 29% recommend both options equally, and 18% recommend comfort care over surgery. In univariate analysis, respon-
Of the 94 physicians recommending surgery, 78 recommend the Norwood PP, 11 recommend cardiac transplantation, and 5 recommend both options equally. At cardiac transplantation centers, 100% of physicians (n=10) recommend transplantation over the Norwood procedure. At centers performing both operations, 89% of physicians (n=8) recommend the Norwood procedure, none recommend transplantation, and 11% (n=1) recommend both procedures equally. Univariate analyses revealed that the strongest predictor of physician recommendation was site of practice (OR, 2.68; 95% CI, 2.50-2.87), and this association remained in the multivariable analysis (Table 3). Physician specialty, religious affiliation, race/ethnicity, sex, parental status, and years in practice were not associated with the recommendation made. Furthermore, physician recommendation was not associated with predicted outcomes of either surgical option, nor with the perceived difference in outcome between the 2 procedures. In fact, 10 physicians (4 at Norwood PP centers and 6 at centers that perform both procedures) recommend the Norwood PP to parents despite their prediction of better outcomes in all areas (survival, physical, cognitive) posttransplantation compared with the post–Norwood procedure.

**SUBGROUP ANALYSES**

**Variables Associated With Recommending Cardiac Transplantation vs Norwood PP**

Of the 94 physicians recommending surgery, 78 recommend the Norwood PP, 11 recommend cardiac transplantation, and 5 recommend both options equally. At Norwood PP centers, 93% of physicians recommending surgery (n=70) recommend the Norwood procedure over transplantation, 1% (n=1) recommend transplantation over the Norwood procedure, and 5% (n=4) recommend both equally. At cardiac transplantation centers, 100% of physicians (n=10) recommend transplantation over the Norwood procedure. At centers performing both operations, 89% of physicians (n=8) recommend the Norwood procedure, none recommend transplantation, and 11% (n=1) recommend both procedures equally. Univariate analyses revealed that the strongest predictor of physician recommendation was site of practice (OR, 2.68; 95% CI, 2.50-2.87), and this association remained in the multivariable analysis (Table 3). Physician specialty, religious affiliation, race/ethnicity, sex, parental status, and years in practice were not associated with the recommendation made. Furthermore, physician recommendation was not associated with predicted outcomes of either surgical option, nor with the perceived difference in outcome between the 2 procedures. In fact, 10 physicians (4 at Norwood PP centers and 6 at centers that perform both procedures) recommend the Norwood PP to parents despite their prediction of better outcomes in all areas (survival, physical, cognitive) posttransplantation compared with the post–Norwood procedure.

**Variables Associated With Recommending Comfort Care**

Of the 16 physicians who do not recommend surgery over comfort care without surgery, 13 recommend both options equally and 3 recommend comfort care over surgery. On univariate analysis, the only variable associated with recommendation was the Norwood PP 1-year survival, which was weakly associated with recommending both surgery and comfort care without surgery equally (OR, 1.01; 95% CI, 1.0004-1.02). Because of small numbers, multivariable analysis was impossible.
Under the standard model of informed parental permission, the physician discloses all feasible options to the parents and discusses the risks, benefits, and likely outcomes of each option, basing their information and recommendations on the best available evidence on outcomes. Parents then make an informed decision based on what they feel is best for their child. Our study shows that in the case of treatment decisions for HLHS, physician recommendations are poorly associated with their estimates of outcomes. It, therefore, seems as if these physicians do not follow this model of informed permission.

We found that many physicians do not discuss surgical interventions that are not offered at their own institution. Furthermore, physicians who recommend surgery recommend the surgical procedure performed at their own institution; however, these recommendations are not associated with predicted postoperative outcomes. Indeed, some physicians recommend the surgical option preferred at their center even when they predict better outcomes for another approach. As illustrated by the quotation from the physician who would choose comfort care for his own child but does not discuss this option with parents, there is likely some pressure within each institution to recommend options performed locally.

Although previous work has shown that many parents and physicians prefer nonsurgical management for their child with HLHS, 18-20 many physicians, even those who would personally consider this option, omit comfort care without surgery in their discussions with parents. This is not to say that physician recommendations should necessarily mirror their personal preferences. Indeed, what parents believe is in their child’s best interests may differ significantly from the treating physician’s opinion, and physician disclosure of personal preferences may be irrelevant or coercive in parental decision making. 22 However, one would expect that, in general, physicians would at least discuss the treatment option that they believe they themselves might choose.

There are several possible explanations for our findings. On the one hand, the standard model of informed consent may be regarded as impractical in a situation in which parents are likely to be emotionally overwhelmed and need to absorb a great deal of technical information quickly. Furthermore, rigorous outcomes data are lacking, and surgical procedures are in continual evolution. In addition, specialists may assume that infants were referred to their center to have the approach generally used there. On the other hand, it could be argued that parental preferences might be particularly important in a “toss-up” situation in which experts hold different and strong recommendations, approaches differ in invasiveness and short-term morbidity and mortality, and there are no definitive outcomes data. 23 In such situations, parental preferences such as attitudes toward risk and multiple surgical procedures, rather than physician expertise, might be crucial.

There are several limitations to our findings. Because study subjects were all chosen from quaternary referral centers, our findings may not be generalizable to physicians who practice in community hospitals or who counsel parents after HLHS is diagnosed prenatally. Choices physicians say they would make for their own children, which was used as a predictor variable in our analyses, may differ from the actual choices they would make. In addition, what physicians do in practice may differ from what they reported. Furthermore, our small sample size reduced our power to detect associations. Additionally, with a 57% response rate, there may be selection bias; it is possible that those physicians who recommend a certain treatment option might have a higher response rate than others. Finally, we classified centers as modest, moderate, and high volume on the basis of expert opinion, rather than on actual data on surgical volumes.

Our data have several implications for clinical practice. First, in the spirit of informed consent, physicians should discuss with parents the full range of options. In the case of HLHS, this would include the Norwood PP, cardiac transplantation, and comfort care without surgical intervention. Options that are not readily available at that center ought to be presented if transfer to another facility is feasible. Parents cannot choose among different options according to their own values if they are not told about some options. Second, physicians need to consider the limits of their expertise. In discussions with parents, physicians should acknowledge the disagreements among specialists and the lack of conclusive outcomes data. Third, physicians need to consider how they arrive at their recommendations to parents. There may be good reasons for making a recommendation to a family that differs from what the physicians would want for their own child. For example, parents may be more accepting of surgical risks, may have a strong religious commitment to take whatever steps are feasible to prolong the child’s life, or may have already decided on a surgical option before coming to a referral center. However, in other cases in which the parents turn to the physicians for advice, the physicians should clarify in their own minds their reasons for making a recommendation that differs from what they would want for their own child. Presumably physicians would base choices for their own child on considerations that other parents would also find pertinent.
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