

Hypoplastic Left Heart Syndrome: Current Conflicts and Hope for a Brighter Tomorrow by Tara Swanson

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lieve her future looks promising, and it is getting brighter every day.

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Hypoplastic Left Heart Syndrome and the Myths of Informed Consent by Debra Hilton-Kamm, MBA



Ms Hilton-Kamm has a background in healthcare marketing and is the co-founder of California Heart Connection, a nonprofit support network for those with congenital heart disease (CHD). She co-authored the recently-published article, "Prenatal Diagnosis of Hypoplastic Left Heart Syndrome: Impact of Counseling Patterns on Parental Perceptions and Decisions Regarding Termination of Pregnancy" based upon a nationwide survey of parents. Through the websites www.HLHInfo.org and www.CHDResources.org she provides resources to parents and physicians. This article is based upon her research, communication with parents, and personal experience with her 12-year-old son who has Hypoplastic Left Heart Syndrome.

A mother recently contacted me, astounded to learn of teenage and young adult survivors of Hypoplastic Left Heart Syndrome (HLHS). She had given birth to a child with HLHS a few years prior and was told that "nothing could be done." Consequently, she watched helplessly as her baby died. Realizing that surgical intervention was available when her child was born, she expressed shock at the lack of information given and anguish that she did not pursue a second opinion. She asked, "Why didn't the doctor tell me about the surgeries?"

Hypoplastic Left Heart Syndrome is a serious congenital heart anomaly in which the left side of the heart is underdeveloped, and without surgical intervention is fatal within the first weeks of life. In the 1980's the development of two different surgical approaches gave hope to these infants: heart transplantation, and a 3-stage surgical procedure.^{1,2} Despite a myriad of risks and unknowns, some surgeons and parents took a chance. Today, some of those early survivors have reached adulthood, attended college, married and even successfully given birth.³ The surgical outcomes have improved dramatically in recent years,⁴⁻⁸ with current expectations that 70% of those born today with HLHS may reach adulthood.⁹ These milestones could barely have been conceived of just a few short decades ago.

Why, then, are some parents still not adequately informed of the potentially life-saving surgeries?

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Myth 1: Informed consent is applied equally to surgical intervention and comfort care

The process of informed consent must include: the nature of the treatment, the probability of its success, the risks and benefits of the treatment, and risks and benefits of alternatives. In addition, informed consent needs to be free from coercion and manipulation.¹⁰ Most articles written about informed consent and HLHS focus on nonintervention (comfort care), with many authors vehemently arguing that this option should be presented to families.¹¹⁻¹⁵ Some concede that the surgical survival rates for HLHS have increased dramatically, but allude to the potential for neurological issues as a rationale to offer comfort care.¹⁴ Some point out the disparity in treatment options, with comfort care offered for HLHS but not for extremely premature infants, even though both are fatal without aggressive intervention.^{16,17} Wernovsky also refers to transposition of the great arteries and pulmonary atresia, arguing that because comfort care is not offered for these life-threatening congenital heart anomalies, it should no longer be offered for those with HLHS.¹⁷ Dr. Norwood, who made it possible for those with HLHS to survive by developing of the first stage surgery, expressed his views about comfort care stating, "I simply cannot understand why those who will potentially do well and prosper are sentenced to death just because *some* will not do well."¹⁸

While this impassioned debate over comfort care continues, very little attention is focused on the lack of informed consent regarding surgical intervention for HLHS. Parents receiving a prenatal diagnosis may have only weeks to consider termination; those receiving the diagnosis after birth may have just days or even hours to choose between surgical intervention and comfort care. To my knowledge, no

research has been conducted to determine the number of parents who terminated the pregnancy or "chose" comfort care without knowing of the surgical options, or after being given inaccurate or incomplete information on surgical intervention. This paper focuses on the inconsistent, sometimes biased, and often negatively-skewed information given to parents at diagnosis which clearly violates the principles of informed consent.

The information parents receive regarding HLHS is highly variable and dependent upon where they receive the diagnosis and from whom.¹⁹ One study found that neonates admitted to noninterventional hospitals generally do not undergo surgical intervention.²⁰ Since the majority of hospitals in the U.S. do not perform these complex heart surgeries, this may represent a significant number of infants. The study also found that those admitted to rural hospitals or to hospitals with lower surgical case volume were also less likely to undergo surgical intervention.²⁰ Among hospitals that do provide these surgeries, there are large variances in mortality, with higher volume facilities attaining significantly higher survival rates.^{21,22} Some found that center and individual surgeon volume influence outcomes.²³ In a study of the top 14 pediatric cardiac facilities, Kon et al found large variations in physicians' predictions of 1 year survival: 30%-88% for the Norwood, and 5%-90% for heart transplant. The predicted percentage of survivors with an IQ of 70 or higher ranged from 50%-100% for the 3-stage surgeries, and 55%-100% for those undergoing heart transplant. The study also found that physicians are likely to recommend treatment options offered at their hospital, even when they believed other options offered higher survival rates.²⁴ Prsa et al found that physicians were more likely to recommend surgical intervention if they: estimated higher post-operative survival, believed they

would choose surgery for their own child, and if they practiced at an active cardiac surgery program. Physicians' recommendations for comfort care were strongly associated with physicians' own hypothetical choice of non-intervention for their own child.²⁵ Many parents are unaware of these major variances in survival rates – information that is vital to these life-and-death decisions. Some argue that the disclosure of physician success rates should be a mandated element of informed consent, and point to states that have favorable legal environments in which success rates may be required by law in the future.²⁶

Internet use by parents of children requiring heart surgery is increasing,²⁷ allowing some parents to find information on the surgical options on their own. Some even travel great distances to high volume centers to give their children a better chance of survival. In 2010, we conducted a nationwide survey of parents of children with congenital heart disease (CHD), with 841 respondents reporting their experiences and perceptions when receiving the diagnosis. Of the 211 who had children with HLHS, some were not initially told about surgical intervention. One parent wrote, "Everything discussed around HLHS was negative and made us feel like we had no choice other than termination. Only after I Googled on my cell phone while waiting did I determine that people actually can survive HLHS." Another parent stated, "The primary doctor who first diagnosed the condition did not give termination as an option; he told us we had no other choice. We found out that night through internet searches that there were other options."²⁸

Myth 2: Physicians are often overly optimistic, giving "false hope" to parents

The results of our study indicate that

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parents receiving a diagnosis of HLHS receive a preponderance of negative information and may interpret certain terms negatively. Of the 211 respondents, 20% reported being told by the first pediatric cardiologist they consulted that the likelihood of the child's death was "very likely;" 28% were told that the likelihood of major complications from surgery was "very likely." Only 18% of those prenatally diagnosed with HLHS, and 8% of those diagnosed after birth reported receiving information on support groups from the physician. Only 41% reported receiving information on ages of HLHS survivors. Eighty-eight percent of parents were told the condition was "rare" which parents interpreted to mean: "few or no other survivors" (30%); "occurs in less than one in a million births" (27%); and/or "little or no chance of survival" (23%).²⁹

Of the 138 parents whose children were diagnosed prenatally with HLHS, 32% said termination was mentioned again after they declined and 22% felt "pressure to terminate" the pregnancy by the pediatric cardiologist. Both of these situations resulted in significantly lower optimism regarding their child's life expectancy, and an increased likelihood of choosing a new pediatric cardiologist. Additionally, when the term "rare" was used in conjunction with the continued mention of termination, or when parents felt pressure to terminate, parents perceived a lower chance of survival for their child.²⁸ The respondents in our study overwhelmingly chose surgical intervention, with only one choosing termination. They were well educated, had access to outside information, and pursued surgical intervention – sometimes despite pessimistic predictions and/or pressure to do otherwise. But what about parents who do not possess these traits or do not have access to outside resources?

Myth 3: Physicians' views accurately predict parental choices

Physicians' beliefs and perspectives often vary considerably from parents receiving a diagnosis of HLHS, and these beliefs may greatly influence how they counsel parents. Physicians who diagnose HLHS are predominantly male with high education levels (specifically in medicine), high income levels, and some knowledge of or experience with HLHS. The population of parents receiving the diagnosis is comprised of all females who are all midway through pregnancy or have just given birth. They come from all income and education levels, and usually have no previous knowledge of HLHS. These two diverse populations certainly represent divergent demographics, experiences and perspectives. One such difference concerns termination; approximately 50% of physicians and nurses say they would terminate the pregnancy if they received a prenatal diagnosis of HLHS.^{25,30,31}

However, actual termination rates in recent years have been reported to be approximately 12%.³²

One study found that physicians' views of quality-of-life (QOL) were more negative and narrowly focused on medical procedures, while parents and children with CHD expressed a broader view of QOL. Parents and children also attributed positive aspects to their CHD while the clinicians did not.³³ Some parents in our survey addressed how physicians presented QOL. One wrote, "We were not told until 40 minutes into the discussion about how our child would have 'no quality-of-life' that there were surgical treatment options with decent success rates." Another commented, "The first cardiologist we saw told us we were selfish if we allowed our son to live with HLHS because he wouldn't be accepted by society."²⁸

Byrne states that the 3-stage surgical option and heart transplantation "are associated with serious acute complications, with considerable pain and suffering, with repeated prolonged hospitalizations, and with significant risks

of long-term physician and mental disability."¹¹ Certainly parents who are presented with this information alone would view the prognosis extremely negatively and may be swayed to terminate the pregnancy or choose comfort care. In fact, parents who choose comfort care are more likely to believe that the child would "suffer" with the surgeries,^{34,35} that QOL was diminished, and that the chance of survival was poor.³⁵

Physicians who answer the question, "What would you do?" disregard that this hypothetical proposition is mere speculation on their part, and fail to realize the potential damage to the physician/parent relationship that may result. As Truog discusses, physicians in this situation should be facilitative, not directive. He cites the example of a marital therapist who obviously would not directly answer the question, "Now tell me doctor, if you were me, would you divorce my wife?"³⁶ In the case of HLHS, physicians who state they would opt for surgical intervention risk alienating parents who are considering termination or comfort care. Likewise, physicians risk offending parents if they state they would terminate the pregnancy or choose comfort care to parents who would not consider those options.³⁷ Parents may also question the level of care they would receive from physicians who believe they would not choose surgical intervention for their own children.

To give parents "appropriate hope" and obtain true informed consent physicians should:

- Give ALL options and success rates at major centers in addition to local hospitals.
- Avoid using terms like "rare" which could be misconstrued. Instead, use quantifiable data to put information in context such as, "Each year, approximately 1,000 babies are born with HLHS in the U.S."³⁸
- Refrain from repeated mention of

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termination if parents decline.

- Discuss successful cases in addition to worst-case scenarios, and refer to support groups so parents can get a realistic view of raising a child with HLHS.
- Give pre-screened, accurate internet resources and printed materials on HLHS.
- Decline to speculate on what they may do in the situation.

The current environment of inconsistent and biased information on HLHS can lead to serious negative consequences for physicians and parents. Physicians who give inaccurate or incomplete information risk their reputations and the loss of future patients as parents relate their negative experiences to others. More importantly, until parents receive more uniform, accurate, and balanced information, some - like the mother who discovered the surgeries too late - will continue to be haunted by how different their lives might have been had they known all available options.

The AAP should take a leadership role in upholding the principles of informed consent by creating guidelines for physicians on counseling parents regarding HLHS, and proactively educating parents on all of the treatment options.

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