

## RECEIVING A CHILD'S DIAGNOSIS OF COMPLEX CHD: PARENTS' PERSPECTIVES AND COMMUNICATION TOOLS

By Debbie Hilton Kamm

This is first in the series of two parent articles by Debbie Hilton Kamm, the co-founder and president of California Heart Connection ([www.caheartconnection.org](http://www.caheartconnection.org)), a nonprofit support network for those with heart defects. She is the creator of the HLHS Information Page ([www.HLHSinfo.org](http://www.HLHSinfo.org)), a site for parents expecting a child with Hypoplastic Left Heart Syndrome (HLHS). She has a 5-year-old son with HLHS and Total Anomalous Pulmonary Vein Return (TAPVR) who underwent the 3-stage surgeries and vein repair. She has a Master of Business Administration and a background in health care marketing.

### Introduction

When presented with their child's diagnosis of complex Congenital Heart Disease (CHD), parents are understandably emotional and often overwhelmed due to lack of knowledge of the heart condition, unfamiliarity with the terminology and medical environment, and grief over the loss of their "healthy" child. They may be intimidated to ask questions, or very often, do not even know what questions to ask. Yet, parents must quickly learn about the diagnosis and make many tough decisions including whether or not to terminate if the diagnosis occurs during pregnancy. Physicians can help parents in this situation by acknowledging their emotions, clearly communicating diagnosis-specific information, and being aware of the manner in which information is presented and interpreted by the parents.

This article is based upon hundreds of interactions with parents through: California Heart Connection, a nonprofit support network ([www.caheartconnection.org](http://www.caheartconnection.org)); The HLHS Information Page, a web site created to help parents receiving the prenatal diagnosis of HLHS ([www.HLHSinfo.org](http://www.HLHSinfo.org)); and online support forums for those with children with CHD. It is also based upon personal experience receiving a prenatal diagnosis of Hypoplastic Left Heart Syndrome (HLHS) for our now 5-year-old son. Handouts and forms referenced throughout the article have been created to help improve communication with parents at diagnosis and can be downloaded for free from the California Heart Connection web site at [www.caheartconnection.org](http://www.caheartconnection.org).

### Receiving the Diagnosis

**Setting and Emotions.** Parents appreciate having privacy and being with a spouse or loved one for support as they get the news.[1] Physicians can acknowledge the emotions by simple statements such as, "I know this isn't what you are expecting." Or, "Let me know if you'd like to stop at any

time and take a break." Having a social worker or other team member involved from the beginning who can help the parents deal with their emotions will help them absorb information on the diagnosis and treatments.

**Demeanor.** Parents sometimes don't recall hearing or understanding much of the diagnosis-specific information after the initial diagnosis is given, but they do recall the manner in which the information is presented. In fact, families often place more emphasis on how they were informed than the specific details about the condition.[1] The same information presented in different ways can also lead parents to very different conclusions and greatly impact decision-making. Roger Mee, M.D., Department Chair of Pediatric and Congenital Heart Surgery at the Cleveland Clinic illustrates this point in the book, *Walk on Water*. The message conveyed by saying, "There's a fifty percent chance we can get your child through," with an upbeat tone and a smile differs greatly from saying, "There's a fifty percent chance your child will die," with a furrowed brow and frown.[2] Physicians should be aware of the subtle messages they may be conveying by their choice of words, tone and demeanor.

**Timeframes.** Some parents leave the initial appointment at diagnosis unclear on what the next step will be. Giving parents written timeframes for any future appointments, follow-ups or tests can be helpful.

**Accessibility of Physicians.** (*Medical Specialties Form*). Having access to physicians as questions arise is important to parents. Allowing parents to e-mail physicians over non-urgent matters allows them to ask questions without "bothering" physicians during their busy work schedules. Many parents are confused by the different medical specialties; therefore, providing written descriptions and contact information for each can help parents ask appropriate questions of the correct medical professional.

**Control.** Lack of control is often mentioned as one of the most prominent feelings parents encounter when receiving their child's diagnosis. Deciding how much information to take in at one time, whether or not (or when) to pursue additional testing, and researching treatment options helps some parents feel more in control.

**Repetition.** The emotional turmoil, new terminology, and lack of knowledge on the subject may lead to many parents repeatedly asking the same questions. Some note that due to parental grief, fear and anxiety, retention of information about their child's condition can be as little as 20%.[3] Parents should be encouraged to ask questions, even repeatedly, to help them absorb the information.

## Prenatal Diagnosis

**Amniocentesis.** Parents receiving a prenatal diagnosis of CHD are sometimes urged to make an immediate appointment for an amniocentesis. Some parents recall the amniocentesis being performed before they had time to consider the possible risks and benefits of the procedure. Others have been told that the results could somehow change the course of treatment, only to find out later that was not true for their child's condition. Parents should be given time to digest the information given at the time of diagnosis, and weigh the risks and benefits of amniocentesis for their particular situation. Allowing this time can make parents feel more in control of the process, and can help them to make more informed decisions.

**Continuing the pregnancy.** Many parents report great shock and sadness at the mention of termination at diagnosis, interpreting that to mean the condition is fatal or that it is the recommended course of action. Many parents will want to weigh all of the factors involved such as the seriousness and accuracy of the diagnosis, potential treatments and prognosis prior to deciding whether or not to continue the pregnancy. However, time to make these decisions is often very limited. Some parents may also consider putting the child up for adoption, while others may be more willing, or able, to handle a child with medical needs. [4]

For many parents the decision often boils down to a simple question: "What can I live with?" Some could not live with the idea of putting a child through surgery, or with the unknowns of having a child with CHD. Others can only live with themselves knowing they did everything possible for their child. Receiving the diagnosis of a child's CHD is a life-altering experience for parents, no matter what course of action they choose, or what the final outcome may be.

## Diagnosis-specific Information

**Prepared handouts.** Many parents state that printed materials were critical in helping them to understand the diagnosis and treatments. Those who don't receive any printed handouts often leave the office confused, and may not accurately remember what they have been told. Clearly labeled printed diagrams of the normal heart and of the specific defect are beneficial in helping parents understand the condition. Hand-drawn diagrams are often confusing, but may be necessary if the anatomy is unique. Having clear visual aids helps parents follow the physician's explanation of the defect, and allows them to review the information once at home.

**Levels of Information.** Physicians may wish to offer information in levels, starting with basic information on the normal heart to set the stage for parents and act as a review for those who have not recently studied the heart. Basic information on the specific heart defect can then be presented, followed by more detailed descriptions of the anat-

omy, and how it affects the child. Some parents may wish to review the detailed information immediately with the physician; some may prefer to review the printed information later. Physicians can ask parents directly how much information they want during the initial diagnosis, and if they would like to stop at any point.

**Internet Resources.** (*Internet Resources Handout*). Many parents access the Internet for more information, especially if not given written materials at the time of diagnosis. Physicians can help guide parents to reputable Internet resources which provide accurate information; however, even very reputable websites can have outdated or misleading information on a specific diagnosis. Therefore, physicians should review web sites and other information prior to referring parents to them to avoid any confusion or misunderstandings of the diagnosis.

## Information for Decision-Making

**Current Information.** Parents sometimes state frustration that the initial information they received at diagnosis was outdated. For example, parents of children with complex CHD sometimes report that they were given a very negative prognosis at diagnosis, only to discover later that overall success rates or those at major heart centers are better than were originally quoted. In one instance, a woman grieved the loss of her baby shortly after birth, after being told no treatments existed for babies with Hypoplastic Left Heart Syndrome (HLHS). She subsequently viewed a television show that profiled HLHS survivors from infancy through their late teens and was shocked and distraught to learn that surgeries were available when her child was born. The fact that she was not given the option of surgical intervention resulted in extreme anguish, and added to her grief for the loss of her child.

**Treatment Considerations.** (*Hospital Contact Form and What to Ask the Surgeon/Hospital Handout*). To aid in decision-making, parents need clear information on the results of surgeries performed locally and how they compare with results in other centers.[5] Some recommend the use of quantitative tools to compare the quality of care based upon the complexity of the surgical procedures, such as the Aristotle Method.[6] This quantitative information can then be weighed along with the needs of the family's situation, insurance limitations, financial constraints and other variables. Providing a list of questions for parents to ask when considering different hospitals also allows them to weigh the individual variables based upon their own needs. Some parents, for example, may choose a hospital based upon location or services provided and not solely upon surgical experience or outcomes.

**Insurance.** (*Insurance Terms Handout and What to Ask the Insurance Company Handout*) Insurance considerations are one of the major concerns and source of confusion for families. Helping parents understand insurance-

related terms, and providing appropriate questions to ask of the insurance company can be extremely beneficial in reducing their anxiety level and helping them with their decision-making.

**Support.** Connecting with other parents who have children with CHD can provide much needed support and information. Physicians may create their own patient contact list, or refer families to an established support network. Physicians are encouraged to contact support organizations to determine if the information and support provided is appropriate for their patients.

The following support organizations offer information, parent-matching and online support groups in addition to the other listed services:

- California Heart Connection – [www.caheartconnection.org](http://www.caheartconnection.org) – events, downloadable handouts and forms.
- Little Hearts, Inc – [www.littlehearts.org](http://www.littlehearts.org) – newsletter, stories, large annual picnic.
- TCHIN – [www.tchin.org](http://www.tchin.org) – portrait gallery, list of local support groups.

**Accuracy of the Diagnosis.** Parents often report that they assumed (and were not told otherwise) that diagnostic tests are 100% accurate. In the case of a prenatal diagnosis where termination may be considered, it is imperative that parents understand the potential error rate of the tests. In the case of HLHS, for example, Richard Jonas, MD, Chief of Cardiovascular Surgery and Co-Director of the Children's National Medical Center Heart Institute states: "It is important to remember, however, that although prenatal echo is sensitive to the diagnosis of HLHS, it is not highly specific and can over diagnose the problem. We have seen a number of cases where babies required only coarctation or aortic valve intervention, and on occasion, no intervention at all despite a prenatal diagnosis of HLHS." [7]

### Presenting Balanced Views

**"Rare" Condition.** Many parents assume that "rare" means one in a million, or that it occurs so infrequently that there are no survivors of the condition. Giving quantifiable information on the incidence of the condition helps parents gain perspective on how many others have experienced it, giving

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***"This article will be a great help to physicians and parents. When explaining the diagnosis of fetal heart disease, it is important to communicate accurately and honestly, both verbally and otherwise. I do not believe that it is ever appropriate for me, as the fetal cardiologist, to recommend termination of pregnancy, or to attempt to answer the question, 'What would you do if you were me?' It is also important for parents to be connected to support organizations upon receiving a diagnosis, and I am happy to refer patients to California Heart Connection."***

***~Mark Sklansky, MD  
Director, Fetal Cardiology  
Children's Hospital  
Los Angeles***

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them reassurance and a possible pool of resources.

**Best Case Scenarios.** The diagnosis understandably focuses on what is wrong with the child, possible risks of treatments, and possible limitations the child may have. However, parents appreciate physicians who focus on their child as a whole, rather than

solely the negative aspects of the condition.[1] One family reported that after answering questions about the complications, risks, and possible negative outcomes, the physician said, "Now let me tell you about the best-case scenario." He then proceeded to talk about the children who had survived, and were living normal, active lives, which gave the family more hope and a more balanced view of the possibilities for their child.

**Personal or Professional Preferences.** Physicians' preferred treatment for a particular CHD may vary significantly based upon their views and experiences. One study showed that physicians placed greater emphasis on long-term survival with good heart function in their preferences for treatment. However, parents may use different criteria when considering treatment options. They may put more emphasis solely on short-term or medium-term survival, for example, with the hope that treatments or cures will become available in the future. The same study found that physicians who did not have direct experience with adults with a specific heart defect were more pessimistic about the outcomes for children diagnosed with that particular type of CHD.[8] Therefore, it is vital that parents understand physicians' preferences, and the reasons for them to provide context and reduce confusion upon hearing conflicting views from other physicians. Parents can then evaluate their own views and beliefs and make the decisions that are most appropriate for their situation.

**"What Would You Do?"** Seeking advice, many parents ask this question of physicians. However, a hypothetical response may not be relevant; a physician's response may be very different if his child was actually diagnosed with CHD. Many parents assume that the physician's opinion is based upon medical fact, and view the answer as a professional recommendation. They may be offended or alienated if the physician's stated course of action varies from that which the parents are considering. Therefore, a more appro-



appropriate and reasonable response to this question is: "Since I'm not in your situation, I honestly can't answer that. But, let me put you in contact with other parents who have made these tough decisions." Allowing parents to connect with others who have actually been faced with the diagnosis and have chosen different courses can help them gain a realistic view of the condition and what it entails. Each family can then make treatment decisions based upon their own financial situation, health insurance issues, emotional state, morals and values, and beliefs about the diagnosis and treatments.

### Conclusion

When receiving a diagnosis of complex CHD, parents must deal with a myriad of emotions while learning about the diagnosis and the treatment options. It is important for physicians to acknowledge the parent's emotions, and understand that the manner in which information is presented and interpreted by parents can greatly influence their decision-making. Physicians can help parents better understand the diagnosis by presenting clearly written information and guiding them to reputable and accurate sources of information and support. The use of the handouts and forms referenced above can help improve communication and ease the anxiety of parents receiving their child's diagnosis of CHD.

The following handouts and forms can be downloaded for free from the California Heart Connection web site at [www.caheartconnection.org](http://www.caheartconnection.org):

- Internet Resources Handout – selected online resources for heart diagrams and information on specific diagnoses of CHD

- Hospital Contact Form – a form for listing hospitals which treat the specific diagnosis.
- What to Ask the Surgeon and Hospital Handout – a list of questions to help compare hospitals.
- Insurance Terms
- What to Ask the Insurance Company Handout – a list of questions to determine specific benefits, restrictions, out-of-pocket costs, etc.
- Medical Specialties Form – a list and description of different medical specialties with space to provide contact information for each.
- Resource Guide – an important reference tool to help parents find resources and ask appropriate questions regarding how to care for their child. Includes information on support networks, medical resources, helpful products, Early Intervention services, insurance/financial information, and more.

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