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Hypoplastic Left Heart Syndrome: Parent Support for Early Decision Making

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Hypoplastic Left Heart Syndrome is a life threatening congenital cardiac anomaly. After a child has been diagnosed with hypoplastic left heart syndrome, parents must make life or death decisions within days of birth. Healthcare providers must provide appropriate education so that parents are able to make informed, timely decisions. Information regarding the diagnosis, treatment options, and parental decision making process for initial decision making for hypoplastic left heart syndrome are provided to guide nurses who work with these families. The challenging decision making process which parents must go through after diagnosis of hypoplastic left heart syndrome will be described.

Rachel and Evan, a young couple expecting their first child, received unexpected and devastating news at the completion of a routine screening ultrasound at 20 weeks. The ultrasound revealed that their unborn son had a congenital heart defect. After further testing, the

diagnosis of hypoplastic left heart syndrome (HLHS) was confirmed. Rachel and Evan received information about HLHS from several physicians, made an appointment with a pediatric cardiothoracic surgeon to discuss surgical options, and found information from pediatric hospitals and HLHS online support groups. Ultimately, Rachel and Evan had decisions to make. Four options were presented by the healthcare team, including (1) terminating the pregnancy immediately, (2) continuing with the pregnancy and proceeding with staged palliative surgical intervention in the first week of their son's life, (3) continuing with the pregnancy with the hopes of a heart transplant in the first week of their son's life, or (4) choosing comfort care after delivery, and allowing death to occur naturally, likely in the first week of life. The time frame for option one was urgent because of abortion laws in their state and the gestational age of the fetus. After several sleepless nights of reviewing information, statistics, options, and prognosis, they elected to continue with the pregnancy and give their son a chance at life.

Introduction

Hypoplastic Left Heart Syndrome cases which are diagnosed prenatally allow parents time to gather information and carefully weigh the implications of the decision. In the case of a postnatal diagnosis, however, parents have minimal time to process complicated information, and may receive conflicting management options for making a major decision that will affect the life of their child and family for years to come. Nurses who care for these patients and families, regardless of when the diagnosis is made, need to have an understanding of HLHS including pathophysiology, mortality rates, treatment options, and prognosis to thoroughly counsel parents. Nurses must also be able to provide psychological and emotional support to families during this time of high psychological stress.

Background

HLHS is a serious cardiac malformation that involves the left ventricle, aorta, and mitral valve. HLHS occurs in 1 to 3 per 10,000 live births, and it is one of the most severe congenital heart defects. Approximately 1500 children are born in the United States each year with HLHS, and two thirds of cases occur in boys. HLHS carries a high mortality rate that accounts for approximately 25% of deaths related to cardiac problems in the first week of life, and 15% of infant deaths in the first month of life (Soetenga & Mussatto, 2004).

This condition may be diagnosed prenatally as early as 16 weeks gestation, or after birth via echocardiography, usually in the first few hours or days after birth (Kliegman, Behrman, Jenson, & Stanton, 2007). Postnatally, HLHS is most commonly diagnosed two to three days after birth when the child develops cyanosis and respiratory distress (Soetenga & Mussatto, 2004). In postnatal diagnosis, HLHS is usually an isolated finding in an otherwise developmentally normal, average size full term infant (Claxon-McKinney, 2001).

No matter when HLHS is diagnosed, parents are overwhelmed and may be unprepared to make a critical decision about the life of their newborn and must make HLHS management decisions within the first few days of their child's life. Parents have very little time to learn about HLHS or to seek a second opinion; therefore, they rely heavily on the advice of healthcare team members (Kon, Ackerson, & Lo, 2004). Nurses, especially those working in obstetrics and neonatal or cardiac intensive care units need to have the skills to ensure that parents are empowered to make informed decisions. An understanding of HLHS, treatments and prognosis are all necessary in order for nurses to be effective patient advocates.

Pathophysiology

Hypoplastic left heart syndrome (HLHS) is a term used to describe a group of cardiac anomalies including the underdevelopment of the left side of the heart and ascending aorta

hypoplasia. The left ventricle may be small and nonfunctioning, so the right ventricle must maintain pulmonary and systemic circulation (Kliegman, Behrman, Jenson, & Stanton, 2007). Prior to birth, the fetus receives its oxygen supply from the blood supply in the placenta, which bypasses the nonfunctioning lungs.

After birth, three factors affect the hemodynamic status of children with HLHS; (1) a decrease in pulmonary vascular resistance, (2) the amount of blood flow through the atria, and (3) the degree of patency of the patent ductus arteriosis (Soetenga & Mussatto, 2004). Due to underdevelopment of the left ventricle in HLHS, the child's right ventricle output not only supplies pulmonary circulation, but must also provide systemic circulation via the ductus arteriosis (Claxon-McKinney, 2001). Once the child's ductus arteriosis begins to close, blood is diverted from the systemic circulation to the pulmonary circulation, which results in pulmonary congestion (Soetenga & Mussatto, 2004). Following complete closure of the patent ductus arteriosis, usually around three days after birth, children develop metabolic acidosis, decreased perfusion, circulatory collapse, and die within days (Claxon-McKinney, 2001). Signs of shock and heart failure may appear within hours or days, including dyspnea, hepatomegaly, low cardiac output, weak or absent peripheral pulses, and cardiac enlargement (Kliegman, et al., 2007). Without immediate intervention, an child with HLHS will die (Claxon-McKinney, 2001).

Once HLHS is detected after birth, intravenous prostaglandins are initiated at 0.05 to 0.1Hg/kg/minute in order to maintain the patency of the ductus arteriosis, enabling blood flow to the aorta (Soentenga & Mussatto, 2004). If the child's condition deteriorates either due to heart failure, vascular collapse, or side effects of prostaglandins, the following interventions are initiated; mechanical ventilation, inotropic support, bicarbonate infusion, diuretic infusion, and

sedatives to treat cardiac and respiratory insufficiency (Hoshino, Ogawa, Hishitani, & Vehara, 1999; Soentenga & Mussatto, 2004).

Treatment Options

Twenty five years ago, a diagnosis of HLHS was a fatal one (Claxon-McKinney, 2001). With improvements in anesthesia, intensive care management, and new surgical techniques, the survival of children with treatment of HLHS has improved dramatically over the past 20 years (Osiovich, Phillipos, Byrne, & Robertson, 2000).

When the diagnosis of hypoplastic left heart syndrome is made prenatally, usually at the twenty week ultrasound, parents have the option of pregnancy termination, or continuing with the pregnancy and making a decision after the birth of the infant. In rare cases, HLHS may present on the in-utero sonogram as a progressive lesion that begins with valvular aortic stenosis in midgestation. The decreased blood flow through the stenosed aortic valve results in gradual ventricular chamber hypoplasia because of reduced blood flow through the left ventricle (Kleigman, et al., 2007). Hypoplasia prevention has been achieved via in-utero aortic balloon valvuloplasty, although this procedure is still considered experimental (Kleigman, et al., 2007). When parents are given a prenatal diagnosis of HLHS, decisions must be made quickly if termination of the pregnancy is chosen. Whether the diagnosis is made prenatally or postnatally, once a child is born with HLHS, the parents must choose from one of three options: comfort care without surgery, staged surgical palliation, or immediate heart transplantation (Renella, Chang, Ferry, Bart, & Sklansky, 2007).

If surgery is not chosen, the child is provided “comfort care” without life saving measures. Comfort care is the withholding of surgical treatment, while providing the child with relief from pain and suffering. Comfort care was the only option for children with HLHS until 1980, when surgical palliation techniques were developed (Connor & Thiagarajan, 2007). Comfort care can

involve feeding and holding the child, along with analgesic and anxiolytic administration if the child appears uncomfortable (Kon, 2005). This can be delivered in the hospital or in the home setting with supportive hospice care providers.

If palliative surgery is chosen, the child has the first of several (usually three but sometimes more) staged heart surgeries within a few days of birth. Palliative surgery for HLHS typically involves a three-staged surgical reconstruction of the heart (Ellinger & Rempel, 2010).

The Norwood procedure is usually performed in the first week of life, which reconfigures the heart to enable the right ventricle to pump blood to the pulmonary and systemic circulation. Two different techniques for the Norwood are described: the Blalock- Taussig shunt and the Sano. Either a Gore-tex conduit is placed to connect the right subclavian and right pulmonary artery (the Blalock-Tassig shunt) or a tissue conduit is placed to connect the right ventricle to pulmonary artery (the Sano) to allow passive blood flow to the lungs. The atrial septum is opened to allow oxygenated blood from the lungs to flow to the right side of the heart from the left atrium, so that the right ventricle supplies systemic circulation (Ellinger & Rempel, 2010).

The second surgery, the Glenn, is performed between four and six months of age. The original shunt is removed, and a bidirectional cavo-pulmonary connection is inserted. Blood is directed to the pulmonary circulation by connecting the pulmonary arteries and superior vena cava. The third surgery, the Fontan, is completed at two- four years of age. Blood flow is directed from the inferior vena cava into the pulmonary arteries (Ellinger & Rempel, 2010).

Mortality rates vary for each procedure, but the Norwood has by far the highest mortality rate. A recent study of national data of 550 children with HLHS noted a 32% mortality rate for the Norwood procedure (Connor, Arons, Figueroa, & Geddie, 2004). Another study reported a much lower mortality rate of 8% (Sano, Huang, Kasahara, Yoshizumi, Kotani, & Ishino, 2009). Mortality rates for the Glenn and Fontan are much lower, at 2 to 5.4% and less than 5%

respectively (Connor, et al., 2004). Although surgical survival rates have improved greatly in the past 20 years, there is still a high possibility that children with HLHS are at risk for mental and physical abnormalities (Connor & Thiagarajan, 2007). The oldest survivors of surgical reconstruction are in their late twenties, as techniques for survival were pioneered in the early 1980s (Wernovsky, 2008). Thus, life expectancy remains uncertain.

Cardiac transplantation may be offered as a primary or secondary treatment if the child's heart is severely hypoplastic (Claxon-McKinney, 2001). A heart transplant eliminates the need for several staged operations in early life, but is typically not an option because of lack of donor heart availability. Heart transplantation mortality rate is high, at around 41% (Connor, et al., 2004). Other drawbacks of heart transplantation include complications while waiting for an organ, early graft failure and the need for lifelong immunosuppressant administration (Claxon-McKinney, 2001). Some patients who have received palliative surgery may require a heart transplant, depending on results of palliation (Ross & Frader, 2009).

Decision Factors

Because syndromes such as Turner's, Noonan's, Smith-Lemli-Opitz, and Holt-Oram may also be present in children with HLHS (Connor & Thiagarajan, 2007), pre-operative evaluation is important. One-quarter to one-third of children with HLHS will have a major or minor central nervous system abnormality. Thus, careful preoperative genetic, neurologic, and ophthalmologic evaluations should be performed on children being considered for surgery (Kliegman, Behrman,Jenson, & Stanton, 2007).

Establishing a prenatal HLHS diagnosis has important implications for parental treatment choice after birth, along with fitness of the fetus for surgery (Tibballs & Cantwell-Bartl, 2007). Parents who receive a diagnosis earlier have more time to prepare themselves for a child with

HLHS. They also have the opportunity to deliver their child at or near a children's hospital which treats HLHS and meet with healthcare providers before the birth of the child. The timing of HLHS diagnosis has an impact on treatment decisions. A study performed by Tibballs & Cantwell- Bartl (2007) of 201 children with HLHS found that 96% of the study population of parents of children diagnosed prenatally that continued with the pregnancy chose surgery once their child was born, whereas only 47% of parents who were given the diagnosis postnatally chose surgery. The results of this study demonstrate that more parents who are given the diagnosis of HLHS after birth choose comfort care, which is likely contributed to the option of termination to those given the diagnosis prenatally. As HLHS is diagnosed more frequently, the incidence of termination also increases (Tibballs & Cantwell-Bartl, 2008).

Parents rely heavily on the advice and recommendations of health care providers. When physicians were presented with a hypothetical situation in which they were the parents of children born with HLHS, physicians were divided between surgery, comfort care, and cardiac transplantation (Kon, et al., 2004; Renella, Chang, Ferry, Sklansky, 2007). Two studies indicated that institutional and physician biases towards HLHS treatment methods exist (Karamlou, Diggs, Ungerleider, & Welke, 2009; Kon, et al., 2004). For example, some physicians recommended the treatment preferred at their institution even when better outcomes were predicted from another approach, and many physicians did not disclose all options for HLHS treatment to parents for variable reasons including unfamiliarity with HLHS, facility treatment capabilities, and personal beliefs (Kon et. al., 2004). Some clinicians say that comfort care should no longer be presented as an option due to advances in medicine and surgery (Wernovsky, 2008). Physicians are faced with the ethical dilemma of recommending compassionate care as short-term outcomes are still evolving and long-term outcomes are still

uncertain (Osiovich, Phillipos, Byrne, & Robertson, 2000). These findings raise the question of whether parents are given enough information to truly make informed decisions (Kon et. al., 2004). Even though there is great controversy in pediatric cardiology over whether parents should be given the option of compassionate care, Ellinger & Rempel (2010) noted that in 15 children diagnosed with HLHS between 2003 and 2007, 13 of the parents in the study reported that all treatment options, including comfort care, had been discussed in their counseling.

Nursing Implications

Theory

The uncertainty that parents experience with their child's diagnosis of HLHS include which decision to make and what kind of life their child will have in the future. Parents whose child is diagnosed with HLHS face uncertainty in illness. The concept of uncertainty was examined by Merle Mishel with the Uncertainty of Illness Theory. Mishel defined uncertainty as "the inability to determine the meaning of illness-related events, occurring when the decision maker is unable to assign definite value to objects or events, or is unable to predict outcomes accurately" (Mishel, 1988, p.228). Because of the uncertainties and unpredictability of serious diagnosis, parents often experience high levels of uncertainty and stress as well as family stress (Hoff, et al., 2005; Holm, Patterson, Rueter, & Wamboldt, 2008).

Other antecedents in uncertainty described by Mishel and Braden (1988) are lack of credible authority, lack of social support, and lack of education (Wallace, 2003). Additionally, lack of event familiarity is described as "the degree to which a situation is habitual, repetitive, or contains recognizable cues" (Wallace, 2003 p. 866). Most parents of children with HLHS have never heard of the disease, and have no experience making serious decisions about their child's health. The degree of trust and confidence the parents have in health care providers corresponds

to lack of credible authority (Wallace, 2003). Parents intuitively feel that they know what is best for their child. Trusting someone else who is telling them that their child has a life-threatening illness may be very hard for parents.

A significant relationship between uncertainty and social support was found by Mishel and Braden (Wallace, 2003). Social support is the network of people who help parents cope with their child's illness. Often, children and parents are transferred to larger hospitals away from their home after a diagnosis of HLHS. Distance makes it difficult for family and friends to be present to provide comfort and support during the decision making process.

Loss of the "Perfect Baby"

Dr. T. Berry Brazelton has described the behaviors, feelings and attachment of children and parents including the imagined "damaged baby" and "perfect baby" that parents have during pregnancy. Parents come to terms with a third baby after deliver; the "real baby". Once parents encounter the "real baby" and the fears of a sick child are reality, parents experience overwhelming feelings of responsibility and helplessness. Dr. Brazelton describes three predictable defenses that are elicited by parents in this situation- denial, projection, and detachment. These are natural reactions which help parents adapt to their sick child. Those caring for the parents, including physicians and nurses, should understand that these are natural and necessary defenses which help parents adjust to and accept their sick child. Nurses can help parents by encouraging an optimistic, but still realistic and sensitive approach with parents (Brazelton, 1992). Nurses should also understand the feeling of loss that parents experience after the loss of their "perfect baby". Nurses can encourage parents to bond with their child and build memories by taking pictures and pointing out which parent's features are seen in the child. This will facilitate acceptance and attachment between child and parent.

Coming to a Decision

Parents of very sick children both want and need to make decisions about their child's treatment. Parents of a child diagnosed with HLHS have many needs while in the initial decision making process, even if the diagnosis was made prenatally. The parents who experience the most stress are those whose child was not diagnosed with HLHS in-utero because they are not prepared for the diagnosis after birth (Tibballs & Cantwell-Bartl, 2008). The initial decision making process is the most difficult as it is the basis of all future medical decisions. Parents are often not able to be at the same facility as their child in the first few days after birth, especially if the diagnosis is made postnatally. If the correct diagnosis is HLHS, parents do not have much time to consider the risk/benefit ratio of each treatment (Ziegler, 2003).

Healthcare professionals are obligated to use medical technology only when the benefits outweight the burdens for children with life threatening conditions (American Academy of Pediatrics, 2000). Childhood diseases which may require palliative care are different than those that adults experience, and are usually rare. These diseases are considered either a life limiting illness, which is a condition where premature death is expected, or a life threatening illness, where premature death probability is high, but there is also a chance of survival into adulthood (Chang & Johnson, 2012). HLHS could be considered either category, depending on case by case physical characteristics. Parents of children with life threatening conditions must assume many roles including caretaker, decision maker, counselor, and informant, along with the roles of parent and spouse. Parents have a legal right to make medical decisions for their child. Healthcare professionals who explain medical interventions to parents must be extremely knowledgeable about all treatments that are in the best interest of the child (Chang & Johnson, 2012). Parents should be offered all reasonable treatment options in order to give informed

consent. Informed consent requires the following: (1) the treatment option believed to be the most appropriate by the provider, (2) all reasonable alternatives, and (3) what to expect if life-prolonging measurements are declined (Kon, 2008.)

Raising a child with HLHS requires a lifelong commitment by the family. Additionally, parents opting for cardiac transplantation must relocate to the transplant center for two years, adding stress and financial constraints (Ziegler, 2003; Kon, 2005). Giving a child with HLHS a chance at a happy and meaningful life requires much family sacrifice; parents must spend a great deal of time at hospitals and clinics in addition to the emotional and financial burden of having a child with a lifelong illness (Kon, 2005). The child's life will also be filled with frequent trips to clinics for medical tests and physical examinations. Parents need this information early in their decision making process to be aware of future sacrifices.

Differences in Care

There is a disparity between socioeconomically advantaged and disadvantaged families when health care providers present only life-prolonging options to parents (Kon, 2008). In postnatal diagnosis, advantaged families sought out more information through books, internet, support groups, and by speaking with other families on children with HLHS; this may lead to the discovery of the option of comfort care if it was not offered. Disadvantaged families may not have access to these resources and feel less comfortable asking the physician any questions along with questioning the physician's recommendations. Parents who are more educated, are more familiar with the healthcare system, and those with a higher socioeconomic status are more likely to choose comfort care for their child. Interestingly, parents who are less educated, and sometimes less financially stable are more likely to opt for expensive surgeries and lifelong treatment of their child (Kon, 2008).

Treatment options given to parents of children with HLHS vary by facility. Approximately 90% of centers that provide care for children with HLHS recommend staged surgical reconstruction, and none of these centers offer comfort care for children with HLHS, which does not allow for informed decision making by parents at these centers (Wernovsky, 2008).

Team Members

Many different team members can provide a supportive role for parents during decision making. Cardiothoracic surgeons, neonatal intensivists, nurses, social workers, chaplains, and other allied healthcare professionals all have a role supporting parents (Cantwell-Bartl & Tibballs, 2008). Psychological counseling should be made available to parents at the time of diagnosis (Cantwell-Bartl & Tibballs, 2008). It is vital that members of the healthcare team provide unbiased, consistent information so that parents are not confused and more overwhelmed (Kodadek & Feeb, 2002). Social support should be provided for all families with a child born with a congenital heart defect (Cantwell-Bartl & Tibballs, 2008).

Team members who routinely work with critically ill patients need specialized training in stress and grief counseling, including information on child rights, informed consent, privacy, and knowledge of behaviors to care for children at risk for death (Chang & Johnson, 2012). Counseling parents who are shocked, unprepared, and grieving is demanding. Perhaps the most important caregiver is the nurse. Nurses are available at all times to answer parent questions and provide care, whereas physicians are only available at certain times of the day. Nurses are also more likely to speak in terms that parents can understand. Nurses should establish an empathetic relationship with parents, and small acts of kindness such as providing a warm blanket or a beverage are greatly appreciated. Nurses can help parents feel more secure in decision making

by examining the treatment options along with each family's unique situation (Kodadek & Feeb, 2002).

Education and Options

Children born with HLHS can initially look healthy. Parents may see their new child and wonder if the diagnosis is correct. They may not understand what is wrong with their child's heart and may not understand medical jargon. Parents also lack information and/or access to information about HLHS. Physicians at smaller institutions may not be familiar with the disease, and may not be able to provide detailed information. A decision about the child's care must be made in a short amount of time, so parents may not have time for a second opinion, or investigation of the disease on their own.

It is important for nurses to understand and be able to clarify the advantages and disadvantages of surgical interventions (Claxon-McKinney, 2001). Nurses may also need to arrange a second opinion for parents requesting one. Parental decision making enhances parental feelings of control when there is a lack of control occurring around them (Pearson, 1997). To help with social support, parents should be encouraged to have frequent contact with family and friends and join an online support group when possible. Access to appropriate websites, written materials, diagrams, and picture describing HLHS and its management are needed in order to facilitate the decision making process (McConnaha, 1997). Education materials and time to "take in" information, along with teaching by neonatology, cardiology, and cardiovascular surgery are beneficial to parents when time allows (McConnaha, 1997). Some HLHS online resources for parents can be found at littlehearts.org, hlhs.homestead.com, congenitalheartdefects.com, and babysamson.com. These websites provide facts and personal stories of diagnosis, treatment, and current lives of parents and children living with HLHS.

Support and Interventions

Parents of critically ill children have rated support from nurses as an important part of their experience (Miles, 2003). Miles (2003) developed the Nurse-Parent Support Model to describe ways that nurses can help parents of critically ill children. The model includes four components: communication and information, emotional support, parental self-esteem, and instrumental caregiving (Miles, 2003). These components help parents cope with a critically ill child.

Encouragement and assurance from nurses gives families courage in their decision making ability (Claxon-McKinney, 2001). Parents need to be educated with terms that are understandable, and allowed to make their own decisions (Zeigler, 2003). Parents also need support and reassurance that there is not one single "right" decision.

Nurses must apply the American Nurses Association Code of Ethics, which describes the nurse's primary role as advocate of the health of the patient, which includes family (American Nurses Association, 2001). Decisions must be value-centered and made on an individual basis, while taking personal, cultural, and religious values into account (Pierce, 1997). In order for parents to make autonomous decisions, the nurse's role should be (a) to help parents understand the uncertainties of HLHS and treatments and provide accurate, up-to-date information (b) to allow parents time to make a treatment decision, unless the child is in imminent danger of dying, (c) to arrange other healthcare specialties for further consultation, and (d) to remember that accepting or rejecting the recommended treatment options is the parents' decision (Ariff & Groh, 1996).

The behaviors of nurses that help parents feel more in control and confident include providing information, preparing parents for what to expect, assisting with decision making, and

including other family members and allowing them at the bedside if death is expected (Claxon-McKinney, 2001). The most helpful behaviors and interventions of nurses are those that increase the family's sense of privacy and comfort (Claxon-McKinney, 2001). Nurses should help parents feel as physically comfortable as possible, remembering that the child's mother has just given birth days earlier, and explain things in a way that parents are able to understand. Siblings should be able to briefly visit the child when feasible. Current technologies such as Skype and videophones allow for parents to see their child during their time at separate facilities.

Once parents have made a treatment decision, they should be supported in their decision, even if it not the decision healthcare providers would choose. It is also important to educate parents about the physical differences they will see in their child after surgery. Their child who previously looked healthy may be surrounded by monitors and equipment. The child may have several intravenous lines, chests tubes, an endotracheal tube, bandages, and a Foley catheter. These changes should be described to parents before they are reunited with their child, to avoid the shock of how their child may look after surgery.

Counseling and Grief Management

Effective counseling skills for nurses include:(a) clear communication with parents and other members of the healthcare team, (b) ability to inform parents of decisions that must be made and in what time frame, (c) communication of changes in the child's condition that may potentially affect treatment plans, (d) initiation of referrals to appropriate resources, and (e) realization that the parents are the ones who must live with the decision that is made (Savage, 1997).

Helping parents come to a decision for their child requires asking very difficult questions such as "What makes a life worth living?" and "What constitutes a fate worse than

death?" (Kon, 2008). Counseling parents must also reflect that although they may have faced many adversities, most individuals living with HLHS are living productive lives; some are athletes and some are in college (Kon, 2008).

Parents of a child with HLHS will experience grief regardless of which treatment is choosen for their infant. They will grieve over their sick child, over the choice they must make, and if they made the right decision. They may also grieve if the outcome is not what was expected, such as surgical or intensive care unit complications. Parents are not given time to grieve the loss of the "normal" child they may have pictured because of the time limit for decision making. Nurses should provide empathy and reassure parents that grieving is expected. They should also give families privacy if death is imminent.

Stages of grief include shock, disorganization, volatile emotions, guilt, loss and loneliness, relief, and re-establishment (Morrison, 1987). Expected reactions to grief are crying, expressions of self-blame, desire to talk about the loss, and expression of ambivalent feelings towards the child (Morrison, 1987). Factors which may block the grieving process include early bad memories, difficulty in coming to terms with reality, and negative feelings about the circumstances of death (Morrison, 1987). Nurses should recognize signs of normal and abnormal grieving in order to facilitate the mourning process for parents.

In the case of child death, nurses play an important role. Parents who experience the death of a child are at risk for short and long-term negative outcomes. The loss of a child may have a significant impact on a couple's relationship and parenting skills (Flenady & Wilson, 2011). Although grief is an expected response to loss, it is vital that nurses provide quality and compassionate care during this time (Flenady & Wilsom, 2011). Psychological support should be offered to families, along with arrangements for post-mortem care (Flenady & Wilson, 2011).

Parents need nurses who are empathetic and demonstrate that their child mattered. Some facilities create "memory boxes" for parents after the death of a child, which may include a lock of hair, foot and handprints, and a picture of the child. Parents should be allowed to name and hold the child if desired. Nurses may attend memorial services, as this may help with their own grieving, and show families that they care.

Even though Rachel and Evan had months to process the information given to them about HLHS before the birth of their son, nothing could have prepared them for the day that Rachel went into labor during her 39th week of her pregnancy. The couple was overjoyed at the thought of the birth of their child, but was anxious and uncertain about what would occur after delivery. Sixteen hours later, Luke was born. In the midst of the flurry of doctors, nurses, and other healthcare personnel, they had a brief moment with their son, who had monitors surrounding him and several lines and tubes attached to his tiny body. As their newborn son was whisked away a few short hours after birth to a children's hospital to prepare for surgery in the coming days, Rachel and Evan could not help but question if the decision they made to give their son a chance at life was the right one. The one thing that they were sure of was that they could not have made it through the first days of their son's life without the support of the nurses. The labor and delivery nurses arranged phone calls between Luke's nurses and Rachel and Evan during the time they were separated when Rachel was recovering. Once Rachel and Evan were able to drive to the children's hospital to see their son before surgery, the nurses in the cardiac intensive care unit were extremely knowledgeable about Luke's condition, and helped Rachel to stay comfortable. They were updated frequently during Luke's Norwood Procedure by his nurses, and were relieved when Luke's cardiothoracic surgeon appeared in the waiting room with the news that Luke's surgery went well. After a weeklong stay in the cardiac intensive care unit,

Luke was transferred to a step-down unit, and was discharged after ten days of recovery. Just a few short months later at the time of Luke's Glenn operation, Rachel and Evan were amazed that some of the nurses remembered their family. Luke is now a healthy three year old, approaching his final Fontan procedure. Even though Rachel and Evan faced difficult decisions early on in Luke's life, they have never regretted their decision of palliative surgery for Luke's HLHS.

Conclusion

HLHS is a serious congenital heart defect with varying treatment. Parents need complete information about HLHS statistics and treatment options. All options should be fully discussed with parents throughout the early decision making process. Nurses caring for families facing HLHS have the ability to help parents feel more comfortable and confident with decision making and are advocates for the family during this process.

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