Palliative Care for the Family Carrying a Fetus with a Life-Limiting Diagnosis

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The prenatal diagnosis of a lethal condition creates a crisis in the lives of the prospective parents and family of an affected fetus. The family is subject to shock, anger, disbelief, and despair similar to parents of children who face a life-limiting diagnosis. Although there may be debate about viewing a fetus as an individual, it is clear that families receiving the news of a lethal condition in their fetus need help. The construct of palliative care with its focus on emotional, spiritual, social, and symptom support can provide a model for caring for these families [1–6]. Some of the decisions that a family of an affected fetus faces are unique to the prenatal situation, but a palliative care model nonetheless can be applied.

In spite of improvements in technology in obstetrics and neonatal intensive care, the neonatal mortality rate remains at 4.7 per 1000 live births in 2003 and the infant mortality rate is 6.9 per 1000 live births. Congenital malformations are the leading cause of death in 20% of these cases [7]. Although a lethal diagnosis overall is rare, it is clear from these data that clinicians and families are faced with these circumstances thousands of times a year. In addition, many families who receive a lethal diagnosis in their fetus in the first trimester choose to terminate the pregnancy. All of these
families are faced with difficult emotions and choices and could benefit from proper clinical guidance and support.

Perinatal hospice has been proposed as an alternative to termination of a pregnancy [8]. Calhoun and colleagues reported a substantially lower rate of termination at their institution where they have an active perinatal hospice program compared with national estimates. Breeze and colleagues [5] reported 40% of families offered palliative care made a decision not to terminate. The proposed explanation surrounds how a perinatal hospice program alters the way the choice between termination and carrying to term may be presented. Without a perinatal hospice program, families may be presented the choice of termination or carrying to term in incomplete terms. As Calhoun states, “A bare presentation of these options may leave parents with the perceived choice of futilely watching their infant die…versus actively doing something to end this new and emotionally wrenching dilemma” [9]. Describing in more detail what kind of support will be provided to the families and infants through the remainder of gestation and delivery may help them make an empowered choice of what is best for the baby and family.

Palliative care in newborns has been supported in three general situations: (1) congenital anomalies incompatible with life, (2) newborns born at the limits of viability, and (3) infants who have overwhelming illness not responding to life-sustaining intervention. It seems a logical extension to consider a palliative care approach when a lethal diagnosis is made prenatally. There is a significant challenge in decision making that is related to the degree of certainty of the diagnosis and the certainty of lethality [4]. Palliative care principles can be used to guide supportive conversations. If one likely outcome is perinatal death, preparing for that outcome, along with the other possible outcomes, is critical to a family’s experience of the remainder of the pregnancy and in setting the stage for a normal grieving process.

The general tenets of palliative care can form the basis of compassionate care for families in this situation (Box 1). Clear communication, an exploration of values, the role of spirituality, and what role the parents want to play in creating meaning and memories all should be explored. The purpose of this article is to lay out a framework for engaging families in these discussions and to help clinicians and families make decisions that can begin the process of emotional healing and grieving. There is little research available that has assessed the specifics of this approach. Rather than shy away from giving any guidance in the absence of data, however, this article offers specific advice about commonly encountered problems. It is our hope to increase the comfort level of obstetricians, pediatricians, and neonatologists in helping these families.

Case

It was with complete shock that Mrs. Brown learned her fetus had trisomy 18. The decision to undergo an amniocentesis had seemed fairly routine. It
simply was a test to make sure everything was going well, not to discover something had gone terribly wrong. She and her husband agonized about what to do. They were advised to terminate because this was a lethal diagnosis, and they felt a tremendous pressure to “do something.” But Mrs. Brown already has felt the fetus moving, and she carried a tremendous sense of attachment. She could not help but want to meet her baby. After much discussion with their family, Mrs. and Mr. Brown decided to carry the baby to term or at least to try, because they knew there was a significant chance the fetus might not survive even that long.

Things became only more complicated as they learned about the challenges facing their unborn daughter. They learned at their level 2 ultrasound that she had a large ventricular septal defect and a large meningomyelocele. They met with a geneticist, maternal fetal medicine specialist, and neonatologist. They struggled to make sense of the information. They were told that all babies who had meningomyelocele were born by cesarean section to improve the level of spinal function. This advice did not seem to fit with the idea that the baby most likely would die within the first few days of life.

As they struggled with all of the information they had been given and with the sadness that frequently overtook them, they were struck by the difficulties of continuing with their normal life. Well-meaning strangers met Mrs. Brown with the usual ebullient, “You must be so excited! When are you due?” At routine prenatal visits, she felt awkward waiting with all of the other women who were there to complete the unfolding of their normal pregnancies.

Eventually, the Browns found an obstetrician and a neonatologist who seemed interested in helping them devise a birth plan that met their needs.
They supported the family in their choices, including a vaginal delivery and postnatal interventions aimed at maximizing comfort and time with the parents. The Browns wanted nothing more than a chance to meet their daughter and to hold her. The neonatologist agreed to a limited resuscitation to assist her through the transition period only. There was to be no endotracheal intubation or cardiovascular resuscitation. They all agreed that if their daughter did survive the initial days of her life that the team would assist them in finding resources to care for her at home. All other interventions, such as closing the myelomeningocele, would be discussed in the context of whether or not it would add quality to their baby’s life. They knew that they would have only a limited time to spend with their daughter and looked to whatever time she would spend with them as a gift.

The decision to carry on

Often, families are advised to terminate a pregnancy when a major congenital abnormality is found. Sometimes the options are expressed as terminate or “do nothing.” It often is with the best intentions that medical professionals give this advice. There has been a general view that carrying an affected fetus to term unnecessarily prolongs an emotionally difficult time. It has been known for decades, however, that women who terminate a wanted pregnancy, after receiving a severe diagnosis in their fetus, experience an acute grief reaction comparable to that experienced by women who have had a spontaneous pregnancy loss or even the loss of a neonate after birth [10]. It is, therefore, not clear that termination achieves the goal of optimizing the grief process for families. Every family grieves differently. For some, terminating a pregnancy is the right decision. Others may benefit from an alternative approach. A perinatal palliative care program could provide support to families who make either of these decisions.

The first step in using a fetal palliative care approach is to create a safe place for discussion without assumptions. In helping a family decide whether or not termination, induction of labor, or carrying to term is right for them, an open dialog needs to occur. This dialog needs to address specifics about this pregnancy and also the family’s philosophic, theologic, and even political feelings about termination. Families take these decisions seriously and struggle as this decision suddenly moves from the political to the personal. What does this pregnancy mean to them? What does this diagnosis or prognosis mean to them? What are the challenges they will face if they choose to terminate the pregnancy? What are the challenges they will face if they choose to continue the pregnancy? What might it look like if they deliver the baby near term? What can they expect after birth?

Painting a clearer picture for a confused family can enable them to make a choice that makes sense for them. For many, termination is the swiftest road to healing, and these families need to be supported and given access to resources to help them through their grief. For others, creating a birth
plan may offer a chance to create needed meaning and memories. Because it is unclear which approach facilitates healthier emotional recovery, both options should be made available.

Support during the pregnancy

As illustrated in the fictional case described previously, it can be difficult to cope with normal interactions while carrying a fetus with a potentially life-limiting medical condition. In the United States, perfect strangers feel at liberty to offer congratulations or comments when they encounter a pregnant woman in public. Such encounters can be magnified in particular situations. The prospective parents already may have one or more children. Gatherings for their friends’ birthday parties may be a concentrated experience of pregnancy-oriented discussions. Clinicians should acknowledge these difficulties. Even the business of routine pregnancy care may be challenging for families. Some obstetricians accommodate these families by offering after-hours visits or make them the first clinic visit of the day to avoid a crowded waiting room. Similar efforts can be made around follow-up ultrasounds or other tests [1]. Many families want to prepare for the delivery but feel uncomfortable joining a group birthing class. Private birth classes could be offered, or perhaps a family would benefit from the support of a doula [2]. Finally, some families may reach a point at which they feel their mental health is stretched and they cannot take the pregnancy much further. Early induction of labor for maternal mental health reasons can be considered just as induction for other physical maternal health reasons [11].

It is important to make special note about the concept of facilitating attachment. Many clinicians talk about an affected fetus in dispassionate tones and focus exclusively on the diagnosis and the associated risks. Inherent in the warnings that a fetus will not survive is a benevolent attempt to protect the family from growing too close to the baby. By preventing a strong bond, clinicians believe they can lessen the pain of the loss. This may be the driving force behind the recommendation to terminate a pregnancy. Unfortunately, this is not how grief works. In 1984, White and colleagues noted that “to establish normal grieving the bereaved must accept the reality of the person who has died” [12]. The way to facilitate healthy grieving is to help create meaningful memories and enhance the sense of reality of the person who is at risk for dying. The families at the focus of this article are at particular risk for an exaggerated grief response precisely because there is limited time for bonding with their offspring. They will not have long to stare at their child to imprint the image or absorb the baby’s scent and feel. Precisely because of this lack of time, some women see the time of continued pregnancy as extending the time they have with their child [13]. We are biologically driven to bond with our children. This process begins in utero [14]. Our goal should be to facilitate the formation of that bond (Box 2).
It follows that when a family chooses to continue a pregnancy, they are interested in using the time for bonding and memory building with their as-yet unborn child. Caring clinicians honor this choice when they treat a fetus as a real and complete future child rather than as an unfortunate event [1]. There are a few ways this process can be facilitated prenatally. Clinicians should ask families if they have chosen a name, and if so can they refer to the baby by that name. Families find this to be affirming and reassuring. This action conveys to families that they will be real parents of a real baby rather than the unfortunate observers of a terrible event. Families also may benefit from follow-up ultrasounds. This chance to “see” their baby is one more piece of reality and memory building [1]. This can be important especially if a fetus is at high risk for in utero demise. For these families, they may not have the chance to meet their baby alive. The fetal movements and images are all they have to construct their memories.

Box 2. Tips for prenatal care

Prenatal care should include a focus on facilitating attachment, creating memories, and helping families cope with experiences that accentuate grief:

1. Ask the parents if they have chosen a name and if you may refer to the fetus by it. Families find this affirming and it demonstrates respect for their view of the expected baby as a whole person, not just a diagnosis.
2. View any needed follow-up ultrasounds not only as monitoring and diagnostic studies but also as opportunities for a prospective family to interact with their unborn child.
3. Try to minimize waits in crowded waiting rooms by offering off-hour visits or making the family the first visit of the day.
4. Explore private options for birthing classes, such as a doula or a private Lamaze class.
5. Assist the family in connecting with any needed resources. If the baby might survive the immediate postnatal period, investigating hospice support services ahead of time may be helpful. If a family thinks they would benefit from counseling or grief specific counseling, help identify local resources.
6. “In all this, there is no substitute for real compassion...” [12]. Listen to the family’s experiences with a sympathetic ear. We cannot stop normal encounters that may be hurtful, but we can help ease the hurt by listening and reassuring.
Intrapartum care

Intrapartum care should be well thought out ahead of time. The involved clinicians should spend time with families to generate a birth plan (Box 3). Such a plan can begin by asking a family to create a wish list. It can and should include medical and nonmedical goals and questions.

Clinicians need to explore what kinds of medical interventions they consider open for negotiation during the delivery. Although there certainly are cases where clinicians feel pushed to make an intervention that they typically would not feel comfortable doing, most families who elect to carry to term in this situation are seeking reasonable and well thought-out options. One area of potential conflict is in the selection of a mode of delivery.

A standard recommendation for method of delivery has been to avoid cesarean section when a fetus has a lethal diagnosis. Because there is no beneficence-based obligation to the fetus, who will die regardless of the mode of delivery, then the risks to the mother of a cesarean section should be the driving force in this decision. It has, however, been argued eloquently that in some cases, a woman may have a powerful desire to meet her infant alive. The risk for a stillbirth after carrying an infant to term is experienced as an extreme emotional burden and an expectant mother may be willing to accept the risk to herself for any limited time with her infant alive [15].

Pediatricians or neonatologists who consult with families are faced with similar challenges. Families seeking comfort care may be clear that they do not want an endotracheal tube placed for mechanical ventilation. They may, however, want the infant to receive a few minutes of positive pressure ventilation in the hope that initial respiratory difficulties are related to problems with transitioning rather than a consequence of an underlying disease. Such a request may seem counter to the usual conception of an advanced Do Not Attempt Resuscitation plan. In some cases, however, the reassurance that every possible chance was given to their infant will make a huge difference in a family’s grieving process.

A frank and open discussion about the medical options available at birth forms the beginnings of a birth plan. The details of the planned resuscitation should be discussed carefully in the context of meeting a family’s goals. Most elements of routine newborn care likely are optional. For example, if death truly is imminent, the application of eye ointment serves no purpose. Similarly, a vitamin K shot or the state newborn screen becomes only an uncomfortable experience for the baby with no benefit and would be inconsistent with truly “comfort care only.” If the baby is likely to survive the immediate postnatal period, then these interventions might fit better with parental goals.

Consider what symptoms the baby may experience. An infant who has severe pulmonary hypoplasia might exhibit respiratory distress and signs of air hunger. Options for management of these symptoms could include giving sublingual morphine, placement of a nasogastric tube to administer
Box 3. Creating a birth plan

Encourage families to create a wish list. This should include any ideas they might have of what could make the birth experience and immediate postnatal period meaningful and memorable. They should then use the wish list in conversations with their obstetrician or midwife and consulting neonatologist or pediatrician to create a birth plan. The ultimate birth plan may not be able to accommodate all the wishes depending on medical limitations, but the involved clinicians should make every effort to accommodate wishes or explain limitations.

1. Mode of delivery. In most cases, spontaneous vaginal delivery without monitoring is appropriate. Occasional families may benefit from a scheduled delivery or cesarean section. If clinicians are uncomfortable offering this and cannot dissuade a family, they should assist the family in finding another obstetrician.

2. Resuscitation. A pediatrician or neonatologist should explain all of the steps in routine resuscitation. They then should discuss which pieces fit with a family’s goals and which do not.

3. Symptom management. Anticipate the kinds of symptoms an infant may experience. If there is a chance of significant air hunger or pain, discuss the different options from sublingual morphine, to nasogastric tube placement, to placement of an intravenous line for more direct medication administration.

4. Spiritual considerations. Does the family want a baptism or other religious ceremony? Do they want to bring their local spiritual leader or do they want support from a hospital chaplain?

5. Pictures or video. The family should consider if they want to take pictures or videos themselves or if they want to have external help with this. There are professional photographers available in many communities who volunteer their time to help create memories for families facing the death of their infant.

6. Mementos. Most nurseries offer additional mementos, such as handprints, molds of hands or feet, collection of locks of hair, and so forth. Ask the families to consider these possibilities, and know what your local institution offers.

7. Participation in care. Are there certain activities that a family might find meaningful, such as giving an infant a bath or dressing her? Are there other family members, including older siblings, who should have a chance to hold her?
medications, or placement of an intravenous line or umbilical line for the administration of intravenous medications. The correct approach depends on balancing the perceived risks (such as delay in getting an infant into a parent’s arms, the pain of a needle stick, and so forth) and benefits and must fit with parents’ overall goals of care.

In addition to a discussion about medical interventions, involved clinicians should assist families in planning for memory building and participation in the baby’s care. Are the prospective parents interested in photographs? If so, do they want a member of the family responsible for taking pictures or do they want to seek outside help? There are professional photographers who volunteer their time for this kind of service. A program called Now I Lay Me Down to Sleep [16] can be helpful in identifying local resources for this.

Nurseries generally offer additional mementos that prospective parents might want to have or help create. Hand or foot molds commonly are available. Parents also might want to collect a lock of hair. Footprints or handprints can be made easily. Nurses also should plan on helping families participate in the care of their baby if there are any activities that the families want to pursue. They may want to bathe or dress the baby. Some families have specific desires of whom they wish to hold the baby and in what order. They may want to include older siblings in some of these activities.

A birth plan also should incorporate any spiritual ceremonies that families want to occur. If they want a baptism, are they content to have a hospital chaplain involved, or do they want to make arrangements for someone from their local organization to participate? Discussing whether or not a ritual needs to be done before a baby dies is important to make necessary arrangements to achieve the goal. Openly reviewing the possibilities of fetal demise before or during labor or a baby being born alive but living for a short time or the possibility of survival to discharge all may influence what may be arranged.

As families explore their wish list with the involved clinicians, a true birth plan will emerge. This plan needs to be shared with all of the clinicians who might attend the delivery and the nursing staff who will care for the mother and baby. Some areas of the country have active perinatal hospice organizations. They may be able to supply a hospice nurse to be present and assist in the planning and execution of the details.

It is important to discuss the details of a delivery plan with as many of the clinicians as possible who could be involved at the delivery. A multidisciplinary meeting between obstetrics and neonatology is the ideal setting. All of these cases are emotionally challenging and may challenge the ethical constructs of some of the clinicians. It is critical to make sure everyone who might be involved is comfortable with the plan. If individuals are not willing to participate in the plan as outlined, arrangements should be made for them to opt out. For example, if one neonatologist in a group insists that he or she would feel obligated to resuscitate an infant with the disease process...
described, then a backup physician could be made available for the delivery. Conflicts need to be anticipated and dealt with in advance. A well-laid plan is for naught if parents perceive genuine discomfort and lack of support from a clinical team. If the conflict is too great to resolve, it may be in a family’s best interest to seek delivery plans with a different clinical group or at an alternative institution.

Other pathways

In spite of advances in diagnostic technology, the ability to prognosticate with certainty remains limited. It is important to discuss all possible pathways with a family before delivery. For example, an infant who has severe hydrocephalus may have enough brainstem dysfunction so as not to survive the immediate perinatal period. The prospective parents would benefit tremendously from the palliative care approach laid out previously. Physicians would be remiss, however, to also not help a family plan for the possibility of a normal respiratory drive. A family might choose to take their infant home with hospice for a period of time, anticipating that he may yet succumb to his abnormal neurologic status. The same family may decide later that they want an infant to have a ventriculoperitoneal shunt to optimize the baby’s potential function and improve comfort. Another family might decide that if the baby breathes well in the first hours that they then are committed to aggressive intervention from that moment on. To the extent that various pathways can be anticipated, the clinicians involved should help families anticipate what to expect and plan for the different decision points. It is not be possible to plan for every eventuality nor is it critical to arrive at a decision for all of the “what ifs.” Simply knowing in advance can arm a family with the information they need to explore their hopes and goals of care.

As the possibilities for experimental fetal interventions are developed, they may be offered at a time in the pregnancy where it is deemed a compassionate-use basis because a fetus is dying. This seems to be when palliative care concepts can go hand in hand with aggressive experimental procedures. As these procedures come with a high risk for fetal death, potential maternal complications (including pregnancy loss), and just a sliver of hope, having palliative care as a backup plan offers families support if a procedure does not work. Some families may be referred to or search out a fetal program only to find out that the fetus has a lethal condition and there is no applicable fetal intervention. Palliative care principles should be incorporated into any center performing fetal interventions.

After the infant’s death

Consideration of further diagnostic information, such as parental testing, placental tissue for cytogenetics, cord blood or skin biopsy for fibroblast
culture for cytogenetic testing, radiographs, postmortem examination by a geneticist, and autopsy should be considered, often planned ahead but performed after the death of the baby. Encouraging autopsy is important particularly as it adds new information at least 10% to 40% of the time [17]. This information may confirm or enhance the prenatal diagnosis and subsequently supports parents’ decision to provide palliative care for their child. The decision for autopsy may alleviate guilt and helps families feel altruistic in giving back to the medical community and perhaps other future families. It can aid in predicting recurrence risk and the decision of whether or not to seek a subsequent pregnancy or further diagnostic testing for any other children [11]. Parents holding their baby’s body for any length of time after death has minimal to nonexistent impact on postmortem pathology studies [18]. Although a skin biopsy should be done within the first few hours after death, other evaluations can be delayed for hours or a day without significant consequence to their being informative. Discussions with the local pathologist are recommended.

Bereavement follow-up begins by supporting families in making burial and memorial service arrangements and, whenever possible, staff should be supported in personally attending services. Follow-up with families during their maternal postpartum examination again can show that an infant’s life was meaningful to others. A scheduled appointment to review autopsy or genetic results, typically a few months post death, offers another opportunity to meet with a family. During either of these visits, an assessment of parental coping, grief and depression, and a review of the events of the pregnancy, delivery, time with the baby, and memorial service can be performed. Families often enjoy the opportunity to retell their story to an audience that is accepting of people talking about the death of a child. They may have questions about how to discuss the events with their neighbors, employers, friends, family, or other support systems, and this gives an opportunity to guide them in how to ask for help from these sources.

Bereavement follow-up also might include continued contact with families by phone at scheduled times throughout the year after a baby is born and on anniversary dates, such as the due date, birth date, or day of death. Having some available service for families to find further support in dealing with their grief is critical. Many hospitals have specific bereavement ceremonies and programs that may may serve patients in this function.

Summary

Palliative care constructs for families who have a fetus at risk for dying are every bit as meaningful, and should be as clinically and socially acceptable, as the provision of continued life-extending endeavors. Prenatal diagnosis of a lethal anomaly is a monumental moment in a family’s life. It requires extensive team counseling and planning about complex neonatal and obstetric medical management. The construct of palliative care with
its focus on emotional, spiritual, social, and symptom support can provide a model for caring for these families.

References