Is Treatment Futile for an Extremely Premature Infant With Giant Omphalocele?

abstract

Doctors have no ethical obligation to provide futile treatment. This has been true since the time of Hippocrates who warned physicians not to treat patients who were “overmastered by their disease.” This principle remains valid today but, as the Society for Critical Care Medicine notes, it is difficult to identify treatment as absolutely futile in all but a few clinical situations. Far more common, they note, are “treatments that are extremely unlikely to be beneficial, are extremely costly, or are of uncertain benefit.” These, they say, “may be considered inappropriate and hence inadvisable, but should not be labeled futile.” So what should doctors do when they have a case that seems close to the futility threshold but does not, perhaps, quite cross it? In such cases, is it appropriate to make unilateral decisions to withhold life-sustaining treatment even if the family objects? Or should treatment be provided knowing that it might cause pain and suffering to an infant with no likelihood of benefit? To address these questions, we present a case of an extremely premature infant with a giant omphalocele and ask 3 neonatologists, Dr Dalia Feltman of Evanston Hospital, Dr Theophil Stokes of the Walter Reed Medical Center, and Dr Jennifer Kett, a neonatologist and fellow in bioethics at Seattle Children’s Hospital, to comment. Pediatrics 2014;133:123–128
Doctors have no ethical obligation to provide futile treatment. This ethical precept has been true since the time of Hippocrates, who warned physicians not to treat patient who were over-mastered by their disease. This principle remains valid today but, as the Society for Critical Care Medicine notes, it is difficult to identify treatment as absolutely futile in all but a few clinical situations. Far more common, they note, are “treatments that are extremely unlikely to be beneficial, are extremely costly, or are of uncertain benefit.” These, they say, “may be considered inappropriate and hence inadvisable, but should not be labeled futile.” So what should doctors do when they have a case that seems close to the futility threshold but does not, perhaps, cross it? In such cases, is it appropriate to make unilateral decisions to withhold life-sustaining treatment even if the family objects? Or should treatment be provided, knowing that it might cause pain and suffering to an infant with no likelihood of benefit? To address these questions, we present a case of an extremely premature infant with a giant omphalocele and asked 3 neonatologists, Dr Dalia Feltman of Evanston Hospital, Dr Theophil Stokes of the National Naval Center, and Dr Jennifer Kett, a fellow in both neonatology and bioethics at Seattle Children’s Hospital, to comment.

THE CASE

A neonatologist is asked at 2 AM to consult with a mother admitted for preterm labor at 25 weeks’ gestation. The pregnancy was complicated by a prenatal diagnosis of a large omphalocele on an ultrasound performed at 18 weeks. At that time, the parents spoke to neonatologists and surgeons about the omphalocele. Parents were planning to deliver at a tertiary care center to facilitate repair of the defect.

The mother now presents with spontaneous rupture of membranes, continuing contractions, and cervical dilation. The obstetricians say that she will deliver in the next few hours. Previous discussions with the parents did not broach the possibility of extremely premature birth.

The neonatologist explains that because of the combination of extreme prematurity and severe congenital anomaly, delivery room resuscitation will be futile. She explains that the pediatric surgeon concurs with this assessment. She informs the family that the infant will be given excellent palliative care so that she will not suffer. She encourages the parents to hold their infant after she is born.

The family is irate. The mother says, “This is wrong. You can’t just let her die. Please try to save her life. Do everything that you can.” The father says, “We are calling a lawyer. We demand that you do something to help our baby!”

What should the neonatologist do?

Dr Feltman Comments

Whether a physician seeks limitation permission or simply explains that a futile treatment will be withheld, disagreement regarding the patient’s best interests may arise. The family may argue, plead, or threaten legal action for perceived abandonment. How should the medical team proceed?

First, let’s examine the nature of a “futile” intervention as it pertains to this case.

Youngner2 describes 4 criteria that are often used to support a claim that treatment is futile. These are as follows: (1) that the patient will have a poor quality of life; (2) that the treatment is physiologically ineffective; (3) that, although partially effective, the treatment cannot cure the underlying illness and will only postpone death; and (4) that the treatment has an extremely low probability of being successful.

Characterizing intervention as futile due to poor quality of life is least appropriate in this case. If this child could survive, but the parents and medical team had different views on whether certain aspects of her life would afford her a “reasonable” quality of life, this would warrant a different analysis, because parental wishes typically trump those of the medical team in such cases. Instead, in this case, the question is whether survival is possible because the infant is likely to have severe lung disease due both to prematurity and to the effects of the giant omphalocele. Aggressive delivery room interventions cannot be characterized as physiologically futile. They may be temporarily (hours to days to weeks) successful. This temporary success doesn’t alter the poor probability of survival. Treatment will be futile for anything but the temporary postponing of death. There are no reports of successful treatment of a giant omphalocele for an infant born at 25 weeks. Instead, experts strongly suggest that aggressive interventions may, at best, only postpone death for this infant for no more than a few weeks.3

Youngner2 argues that any futility arguments outside of physiologic futility (an example being providing cardiopulmonary resuscitation to a decapitated patient) are subject to the values of the patient, family, and physician. This case is not one of such physiologic futility. Instead, it is one in which treatment is deemed likely to cause pain and suffering and to only prolong the dying process. The question, then, is who should decide which action (resuscitation and intensive care or palliative care) is in the patient’s best interests. Lantos and Meadow4 summarize 3 commonly accepted pathways for when such disagreement occurs: (1) continued persuasion efforts, (2)
intervention per family request, and (3) nonintervention by unilateral physician decision. They controversially suggest that, in some cases, a fourth option might be possible: to accede to the parents’ request for resuscitation but only perform cardiopulmonary resuscitation for a few minutes. These parents clearly love their infant and clearly feel betrayed by the doctors. They have prepared for aggressive surgeries knowing that death might occur. They don’t know how to deal with the new circumstances of an extremely preterm delivery. It is unlikely that the doctor will be able to persuade them to accept palliative care in the limited time that remains before delivery. Some argue that there is no need to solicit the parents’ permission to withhold attempts at life-sustaining treatments because such treatments are futile. By this argument, doctors have no obligation to offer or even discuss futile therapies. Thus, parents needn’t decide or even accede. In this case, intubation and ventilation might postpone death for days or weeks. Parents may feel that this is an appropriate goal of care. Perhaps they believe that such treatment is the right thing to do for their infant. If the neonatologist believes that the parents are acting in good faith and trying to do what is best for the infant, she could attempt resuscitation, explaining to the parents that this would be experimental, against physicians’ recommendations, and would inflict suffering without benefit. When one considers the 2 traditional aspects of a child’s “best interests” (present experiences and future interests), painful procedures are often justified because they may afford the patient a future that is better than the present. In our case, however, intubation, blood draws, mechanical ventilation, possible chest tubes, and chest compressions will be the means and the end. The burdensome treatments will not overcome the condition; they will only allow for continued dependence on these interventions for the days or weeks before death. This situation highlights the dilemma that many clinicians feel. Overriding the primum non nocere principle in the absence of patient benefit, regardless of parental benefits, creates moral distress. Burt and Curtis characterize this visceral distress: “clinicians’ personal unwillingness to provide aggressive treatment that they themselves or their families would not want may arise from feelings of distaste, and even repugnance, that administering such treatments without reasonable prospect of success amounts to the infliction of physical abuse on dying or dead people.” Ultimately, intervening per this family’s request hurts both infant and physician, with questionable benefit to the family.

What about a less than vigorous, symbolic effort at resuscitation? Lantos and Meadow argue that a modified resuscitation sometimes may be acceptable. They argue that some families in these situations “feel paralyzed and trapped.” For such families, they propose that doctors give ambiguous explanations of what will be done, that is, “we will continue to do everything we can that we think will help. And we will always do everything we can to make sure that he is not suffering. We will always be there, for him and for you.” For our case, limited ability to assess parents’ genuine inclinations makes this approach potentially dangerous. Particularly under time constraints, it is difficult to judge the parents’ reasons for wanting resuscitation. If judged incorrectly, the family may feel deceived if a less than fully aggressive resuscitation is performed. Additionally, the “slow” code typically refers to a patient who will most definitely die regardless of whether the code is slow or traditional; a slow code would not be appropriate in our case, for as previously discussed, aggressive interventions could actually work for this infant to temporarily postpone death. Thus, this case comes down to a question of whether aggressive intervention that will only postpone death is so clearly against the infant’s best interests that it should not be provided even if the parents demand it. In my opinion, the burdens of treatment and the fact that survival in such a case is unprecedented justify nonintervention. Although unilateral decision-making is justified, this family deserves the “compassion, comfort, attendance, and shared grief” advocated by the rationale behind Lantos and Meadow’s “slow code.” This family has limited time to process inevitable death, perceiving abandonment. Whereas previous specialists delineated aggressive management, the neonatologist to their dismay “won’t do anything.” Assent should not be pursued; rather, condolences should be given because resuscitation would only prolong death. Focus should be redirected from what the physician will not do for the patient to what she will do. Attendance to comfort, elicitation of parents’ preferences regarding holding the infant, religious ceremonies, photography, etc., are the appropriate focus now that death is inevitable.

**Dr. Stokes Comments**

Imagine that the neonatologist in this scenario possesses the ability to travel through time. She has traveled into the future and has seen that this infant died despite her best efforts at resuscitation. Having seen the future, she is absolutely certain that “delivery room resuscitation will be futile.” She is completely confident that her plan to provide “excellent palliative care so that she will not suffer” is ethically justified. Even so, the parents disagree. So what should the neonatologist do? The neonatologist seems to have the best interests of the infant in mind and has recommended a rational and ethical
course of action. Somehow, though, she has failed to adequately communicate with this family. I wonder whether a lack of empathy might be to blame. I would want to know more about her previous conversations with this family. Did she acknowledge the grief and the fear that this family was assuredly feeling? Did she find out if they had a name picked out for their infant? What was it like to learn of the omphalocele? What have their experiences been with doctors? What are they hoping for? What do they fear most?

In taking the time to listen, learn, and feel with these parents, the doctor lays the foundation for a relationship based on trust and a promise to face hardship together. The doctor demonstrates that she is human, that she cares, and that she will be there when times get tough. Such a relationship is critical when agonizing decisions must be made. It does not appear to have been established in this case. This is a family who has been mentally preparing themselves for the birth of an infant with a major congenital anomaly. They have consulted with specialists and have learned about the obstacles facing their daughter after birth. They have been told that everything possible would be done to save their daughter. Now a doctor whom they have never met is telling them that it is hopeless to even try and save their infant.

Is it futile to resuscitate an infant born at 25 weeks with a giant omphalocele? Giant omphalocoeles are often associated with pulmonary hypoplasia, and at 25 weeks’ gestation this might indeed make resuscitation impossible. But pulmonary hypoplasia is difficult to diagnose prenatally. Estimates of gestational age often vary by 1 to 2 weeks. What if the infant was really 27 weeks, and what if the lungs weren’t hypoplastic? What if the omphalocele and intestines were intact at birth and amenable to a delayed surgical repair? These questions cannot be answered until after birth.

Clearly, the prognosis is not good. I believe that this infant will likely die. But this is my opinion, and I would have an honest discussion with the family about this. I would not tell them that delivery room resuscitation is futile. We would discuss both palliative care and delivery room resuscitation, and we would come up with a plan together. I would do my best to help them feel supported in their decision. If they wanted me to attempt resuscitation then I would. If they wanted me to provide palliative care then I would. If I started a resuscitation and it wasn’t working, then I would tell the parents that it wasn’t working, I would stop, and would then provide palliative care. Whichever course of action we decided on, I would assure them that I would be in the delivery room to care for their infant when she was born, and that we would continue to care for their daughter together.

Dr Kett Comments

Medical interventions that are not expected to benefit the patient are commonly referred to as futile. Although the attending physician does not provide her rationale in this case, it is likely that she believes that this infant, with 2 potentially life-threatening medical conditions, is unlikely to survive. If there is no possibility that this infant can survive the immediate perinatal period, then aggressive delivery room resuscitation is indeed likely to be futile, and might even be cruel. However, it is important that our initial assessment of this infant’s prognosis be carefully analyzed.

Extreme prematurity and giant omphalocele are, of course, extremely serious diagnoses. Infants born at 25 weeks face long NICU stays and may experience a number of life-threatening complications. However, the majority of infants born at this gestational age do survive, and many survive without serious impairment. Because of this, the resuscitation of infants born at 25 weeks’ gestation and above is generally routinely recommended. Children born with giant omphalocele also face prolonged hospitalization and may likewise experience a number of serious complications. However, in the absence of other congenital anomalies, most full-term infants born with giant omphalocele survive to be discharged.

Of course, the concern in this case is not each diagnosis as a separate entity, but that their combination would exponentially increase the likelihood of complications and therefore death. To begin with, other genetic or structural anomalies are common in infants with omphalocele. If present, many of these conditions might have a poor prognosis in the setting of extreme preterm birth. However, it may be possible to identify the features of some congenital anomalies on ultrasound before delivery to help make this assessment. Skin integrity is another area in which these conditions may impact one another. Extremely preterm infants have fragile, friable skin and can experience skin breakdown even from pulse oximeters and monitor leads. The care of an omphalocele membrane would be extremely challenging in this setting. In addition, infection risk is always a serious concern for extremely preterm infants, and the presence of an abdominal wall defect would be expected to increase this risk. Finally, these conditions are likely to severely amplify one another with regard to respiratory status. Extremely preterm infants are typically affected by respiratory distress syndrome and commonly require mechanical ventilation and surfactant administration after birth. Many infants born at this gestational age will go on to develop significant chronic lung disease. Respiratory insufficiency is also common in infants with giant omphalocele. If an infant with pulmonary
insufficiency is also born extremely preterm, he or she simply may not have adequate lung function for survival. However, the degree of pulmonary insufficiency present can be difficult to predict prenatally. In addition, even severely preterm infants can have variable lung disease that is difficult to quantify before delivery.

Clearly, the combination of extreme prematurity and giant omphalocele increases the risk of a number of complications. However, this situation does not mean that such complications are certain to occur. Respiratory status, skin integrity, and risk of infection are difficult or impossible to predict before delivery. Although pulmonary insufficiency is common in the setting of giant omphalocele, some infants with giant omphalocele do not have pulmonary insufficiency, and the respiratory complications of extreme prematurity alone are routinely managed successfully in the NICU. Likewise, extremely preterm infants do have surgical procedures on occasion, and caring for their wounds is a regular part of NICU care. Although this patient is at high risk of complications and death, it is not certain that he or she will experience them.

It is important to consider how much suffering this infant would be expected to endure in the pursuit of such a slim chance of survival. In the absence of complications, the most likely sources of discomfort would be mechanical ventilation and wound care. Mechanical ventilation, although certainly uncomfortable, is not generally thought to be painful. In fact, in many NICUs, infants who are mechanically ventilated do not receive analgesia or sedation. In addition, giant omphaloceles are not generally treated with primary closure. Staged bowel reduction or topical agents intended to epithelialize the existing membrane are typically used instead. Although these procedures may also be uncomfortable, they do not involve any type of surgical incision, and therefore seem unlikely to be exceedingly painful. On the other hand, the treatment of potential complications might cause a great deal of suffering. For example, chest tube placement for pneumothoraces or the surgical repair of omphalocele rupture or an abdominal infection might cause significant pain. However, in the context of such complications, life-support measures could be reevaluated.

It seems unlikely that an infant with giant omphalocele who is born at 25 weeks’ gestation could survive to discharge, but it is not impossible. In addition, in the absence of complications, it does not seem likely that this infant would experience a great deal of suffering. In this case, where the likelihood of success is small, but not zero, and where the infant would not necessarily experience great suffering, it seems reasonable to support the family’s strong desire to attempt resuscitation.

In addition, the first several hours and days of life could provide information about lung function and skin integrity that could not be obtained prenatally. The decision to pursue continued life-support measures could be reevaluated as new clinical information became available. However, a careful review of this infant’s imaging studies should be performed before delivery, because the presence of other anomalies might drastically change the prognosis for this infant. In addition, the surgical team must be amenable to this plan, because they will be responsible for much of this infant’s treatment. Finally, this infant’s course must be closely monitored; should a clinical change occur that would cause significant suffering, the benefits of ongoing life support should be carefully reassessed. It is important that the care of this infant be at all times focused on her best interests, rather than the interests of the parents or the medical staff.

**CASE REPORT: PART II**

The neonatologist suggests that she and the family meet together with the surgeon. The surgeon concurs with the futility assessment and says that an operation would not be feasible at this gestational age. The parents recognize that, without the possibility of surgery, resuscitation and neonatal intensive care would only cause pain. They say, “We don’t want baby to suffer if she can’t survive.” When the infant is born later that night, the neonatologist confirms the prematurity and the enormous abdominal defect. She notes a heart rate of 50. She wraps the motionless and apneic infant in a warm blanket and encourages parents to hold her and take pictures. A few minutes later, the infant dies. The family is left with the infant to grieve privately.

**Dr Stokes Comments**

This case illustrates the power of hope. Hope is a primal emotion, perhaps even a survival instinct. It is why discussions of futility are so fragile. The neonatologist’s statement that “delivery room resuscitation will be futile” seems to have triggered a flight or flight type of response in these parents. I wonder if the family’s response would have differed had the neonatologist said instead, “I think that delivery room resuscitation will be futile.” This simple rephrasing might have allowed the family to hear the conversation, while not completely overwhelming them with hopelessness.

I recently counseled a family whom I was told was “unwilling” to accept their son’s diagnosis of trisomy 18. The diagnosis had been strongly suspected prenatally, and the infant had been admitted to the NICU after birth due to his small size, a small omphalocele, and an imperforate anus. An echocardiogram after birth had revealed a ducal dependent heart lesion. The NICU team was exasperated that the family would
not “accept reality” and were demanding that “everything be done.”

I sat down with the mother and father and asked them to tell me how they were doing. For the next half hour the family told me the story of their pregnancy. The prenatal ultrasound and maternal blood tests were suggestive of trisomy 18. At each of their subsequent visits they heard that their son would be deformed and retarded and that there was no hope for survival. Eventually this made them angry, and they stopped going to their prenatal visits at ~30 weeks’ gestation. They told me that they couldn’t tolerate the negativity and hopelessness of the doctors any longer.

The mother smiled when she told me about hearing her son cry for the first time in the delivery room. The father beamed when he recounted first seeing his son. Their son had proven all of the doctors wrong. He was alive. He had survived. Eventually, the mother asked me for my opinion, and I told them that I was pretty certain that their son had trisomy 18. Chromosome tests were pending, and these were likely to confirm the diagnosis. They nodded knowingly, and we talked further about trisomy 18 and how it had affected their son. I told them that their son was likely going to die of this condition, and we ultimately agreed that a palliative care plan would be best for him if the chromosomes confirmed our suspicions. At the end of our conversation, the father became tearful and thanked me profusely for my care of his son.

In reality, I hadn’t really done anything for their son. I conveyed the same grim prognosis and recommended palliative care just as the other doctors had. All I’d done differently was to listen and to feel. I was realistic while at the same time respectful of their hope. This was seemingly enough to allow them to hear what I had to say.

It is not rational to maintain hope in the face of a hopeless situation, but that is the point that I am trying to make. Humans are rational and emotional creatures, and doctors must recognize this if we are to effectively communicate.

Dr Feltman Comments

As this patient illustrates, even in the “best” case scenario in which parents agree with the physician’s care plan, end-of-life decisions for a child whose extraterine life has not yet begun are extremely difficult. Time constraints such as imminent delivery curtail opportunities for processing feelings and preferences, challenging relationship-building between family and physician. Encounters in other high-velocity, high-stakes settings such as the emergency department may present similar difficulties. When a treatment is deemed quantitatively or physiologically futile and therefore medically inappropriate, honest assertion of this and redirection to palliative interventions may strike the best balance between the patient’s best interests, physician integrity, and attendance to family grief.

Dr Lantos Comments

This case illustrates one of the most difficult moral dilemmas in all of medicine. Without intervention, a patient will die. Intervention is unlikely to be successful, although it could be partially effective and, as a result, prolong the process of dying and increase the suffering of the dying patient. Should the doctor follow her own moral compass and refuse to provide a treatment that seems overwhelmingly likely to cause more harm than good? Or should she follow the parents’ requests and provide the treatment?

Most of the time, such dilemmas arise because of the way we have talked about diagnoses and options. We need to be careful about how we describe what can or cannot be done, and what we will or will not do. Sometimes, inadvertently, we err by either offering more hope than is justified or by limiting interventions too prematurely and absolutely. Decisions about resuscitation should not be made before the infant is born. Even with an excellent prenatal diagnosis, there can be surprises in the delivery room. It is appropriate to assure the parents that a final decision will not be made until the doctor has a chance to examine and assess the newborn.

The decision to withhold or withdraw treatment can happen before or after intubation, before or after chest compressions, and before or after resuscitation medications or cardioversion. In making the decision, it is crucial to consider the parents’ preferences and values. But it is not always right to allow those preferences to be determinative. Ultimately, the attending physician must make the decision about when and whether further intervention should be discontinued.

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