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Advance Care Planning and End of Life

Features

The Family Voice

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Advance Care Planning and End of Life

Maurice G. Sholas

The finality of death can be disruptive, destabilizing, and debilitating to the person told that they will prematurely succumb, but it also affects everyone in that individual's support network. This reality is especially prescient when the individual dying is a child. Pediatric advanced care planning is emotionally impactful, and a practice area very relevant to the ethics community practicing in pediatric care centers. It is also complex because the parents, the medical care team, and psychological development governs the child's ability to participate. The challenges around planning for the death of a child have caused barriers in communication between careproviders and families. It has led to moral distress in clinicians, dissatisfaction in the parents, and avoidance of that portion of the arc of care. Also, it is a driver of discord in the treatment care team. There is no way to make the loss of a child feel positive or pleasant. But there are methods to make that process focus on dignity, inclusivity, collaboration, and respect. This issue of the *Journal of Pediatric Ethics* reviews perspectives that encompass end-of-life planning and clinical scenarios, from birth to young adulthood. There are the perspectives of medical providers, clinical ethicists, and parents. Thus, this is an exploration of the arc of a painful process, which for some is unavoidable. It intends to serve as a starting point for continued research, commentary, and active reflection.

Historically, the will of the parents has been overrepresented in pediatric end-of-life scenarios when accommodating the family perspective. Through a sense of paternalism, some children were prevented from being a part of decisions that affect the end of their life because it was thought they were unable to adequately process the situation and constructively contribute. There is not a rigid boundary separating children who are capable of only assent from those who are capable of true consent. There is a range of ages and life experiences that make some minors at various ages mature enough to "act intentionally with understanding."[1] Other children have been excluded from the process through a sense of materialism that is driven by the desire of the parents and careproviders to protect children from distress and sadness. The worry is that there is nothing to be gained by informing children of the circumstances around the foreseen end of their life and the choices that have to be made in the wake of that reality. The adults involved feel this information may cause children to be less hopeful and to be left emotionally traumatized. That view is not supported by research, as the articles in this issue demonstrate. Thus, it is a challenge and often and ethical conundrum to modernize the role of the pediatric patient in end-of-life advanced care planning.

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As we look at the process of including children in advanced care planning, two natural poles emerge. On one end there are neonates. They are defined nearly exclusively through the lens of their family's narrative and the framework of expectations in the minds of their parents. Those children are the passive recipients of the processes chosen for them by their parents, who struggle to realize that death does not represent a "tragic flaw . . . like Icarus flying too close to the sun."[2] In those situations there is no way to divine children's wishes and incorporate them into planning. On the other end of the spectrum there are mature minors who develop the ability to process and participate in end-of-life planning. Research has found that there is a positive response by mature children included in end-of-life planning, with no evidence that "the discussion itself will take away the hope of parents and family members."[3] The sensitive but essential steps of advanced care planning for children must leave parents and families convinced there is a "sense of reverence [and]. . . . human respect."[4] And they must also be honest, gentle, and caring processes that include children in a manner that is consistent with their developmental capacity and autonomy.[5]

Advanced care planning and end-of-life discussions are the medical incarnation of the proverbial spirit that no one wishes to summon. But it is important that clinical ethicists and medical provider communities work proactively with families to dispel that trepidation. To make nearly unbearable circumstances humane, all must do the work that makes a "simultaneously heartbreaking and beautiful journey"[6] as empowering and inclusive as possible. Difficult work this is; but indispensable in a setting that requires it.

NOTES

1. J. Needle, M. Lyon, D. Brunqell, and C. Heith, "Mature Minors, Mature Decisions: Advance Care Planning for Adolescent Patients with Life-Limiting Illness," in this issue of the *Journal of Pediatric Ethics* 1, no. 3 (Spring 2020).

2. L. Freitag, "Parental 'Holding and Letting Go' in End-of-Life Decision Making in the Neonatal Intensive Care Unit," in this issue of the *Journal of Pediatric Ethics* 1, no. 3 (Spring 2020).

3. Needle, Lyon, Brunquell, and Heith, "Mature Minors, Mature Decisions," see note 1 above.

4. E. Beaudry, "Precious Life; Precious Loss ," in this issue of the *Journal of Pediatric Ethics* 1, no. 3 (Spring 2020).

5. A. Lanzel, "Children's Views on Death and Dying: An Overview and Ethical Focus on Advance Care Planning Communication with Children," in this issue of the *Journal of Pediatric Ethics* 1, no. 3 (Spring 2020).

6. K. Olavson, "The Most Difficult Decision We Ever Had to Make," in this issue of the *Journal of Pediatric Ethics* 1, no. 3 (Spring 2020).

Features

Children's Views on Death and Dying: An Overview and Ethical Focus on Advance Care Planning Communication with Children

Ashley Lanzel and Katharine E. Brock

ABSTRACT

Advance care planning is not only for adults who are dying. It is for children as well, especially children with a life-threatening or life-limiting illness. In a pluralistic society, we should also question the ethical implications of communicating with children about death. Communicating about death and advance care planning with children should vary with their age and cognitive and developmental level.

In this article, the concepts of death and cognitive development and emotional responses around death, childhood communication, and coping strategies are used to explore an approach to open communication about advance care planning that involves children, when practically possible and ethically permissible.

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INTRODUCTION

Children and death: this association may bring a shudder to some. Children are not supposed to die and they are not supposed to be exposed to death. Why? Death is taboo. But death is universal and important. As Morley D. Glicken states, "The way in which we approach and cope with death greatly influences our approach to life. It is often out of our deep fear and denial of death that we begin to deal, existentially, with the meaning of life."[1]

As rates of mortality in infants and children decline, their exposure to death may become more limited. Yet children still learn about death as part of their development, understanding the "life cycle" through everyday experiences. Without intimate exposure to death, children may become desensitized. For children who intimately face death, it is imperative to communicate clearly about death in a way that is developmentally appropriate. This article investigates children's developmental understanding of and emotional coping with death. It applies this knowledge to inform clinicians on how to communicate with children about death and advance care planning.

CONCEPTS OF DEATH

There are several theoretical frameworks to describe and evaluate the concepts of death and chil-

dren. Examples include the psychoanalytic, intuitive, and cognitive frames. The psychoanalytic approach delves into children's concepts of death in order to explore and potentially help support their emotional responses to death.[2] This approach can be contrasted with the intuitive approach, that, as Virgina Slaughter and Maya Griffiths write, explores "children's knowledge about death in terms of their developing intuitive, or folk knowledge about the domain of biology."[3] The cognitive framework, specifically that proposed by Jean Piaget, correlates changes in a child's understanding of death with cognitive development (see table 1).[4]

These three approaches often do not account for the diversity in the understanding of death and children in different cultures. Maureen Callanan notes that we often fail to account for the sociocultural, religious, and conflicting beliefs that coexist in children.[5] Difficulties exist in comparing these across cultures. Some of the nuances in understanding death that include environmental and religious exposures may be easier to distinguish once children are able to have abstract thoughts. Future research may be helpful in exploring these ideas.

The three approaches generally agree that for children to have a "mature" understanding of death, they must grasp several concepts. These concepts of death progress with age and include: (1) inevitability or universality (all living things die), (2) irreversibility or finality (the dead body cannot come back to life), (3) cessation or nonfunctionality (all living functions end upon death), and (4) causality (living things die as a result of various biological reasons or bodily breakdown).[6] The concepts of unpredictability and personal mortality[7] are not universally included. These concepts usually solidify between five and 10 years, but may occur earlier in those exposed to death at an earlier age.[8]

Infants and toddlers may have difficulty differentiating death from their separation from a parent.[9]

TABLE 1. Developmental conceptualization of death

Age	Piaget's stage	Piaget's development	Concept of death
Birth-2 years	Sensorimotor	Understands world through senses and motoric manipulations; attachment bond is created.	Has difficulty distinguishing death from separation, but does experience loss, especially in inconsistent environments.
2-3 years	Pre-operational	Uses symbolic reasoning and magical thought; illogical thinking is dominated by perception; is egocentric; fears separation; believes in animism.	Struggles to understand irreversibility. Can deny death exists (dead body could be sleeping). Can believe anger (or other emotion) or an action (going to the hospital) caused death of another. Acts out feelings rather than verbalizes them.
4-6 years	Pre-operational	See above. Often concerned with death, as an attempt to develop a definition of life.	Has partial concepts of inevitability, irreversibility, and cessation. Example: a child who does not yet understand cessation has concrete questions such as "How does a dead person breathe or eat underground?" Causality is not fully formed. May cite nonnatural or violent causes of death.
6-12 years	Concrete operational	Develops logical thought to solve problems; develops distinction between animate and inanimate objects; acquires knowledge and develops peer relationships.	Death is seen as concrete and can be externally or internally caused, such as by illness. Can mention spiritual causes ("It is one's time"). Can personify death (i.e., Grim Reaper, Angel of Death, ghosts, etc.). Can make an oath on own death or parent's death. Can consider funerals to be an ill omen. Can cross fingers and say verses to ward off evil and protect self. May accept finality of death, but often applies it to others, not self.
13-19 years	Formal operations	Abstract thought, scientific reasoning form personal identity and social interests.	Death is final, universal, and becomes abstract, but can still question death.

However, by five to seven years of age, children typically have acquired partial concepts of inevitability, irreversibility, and cessation.[10] Piaget notes, "Questions of children of the ages of 5, 6 and 7 are also very often concerned with death, and show their attempts to find a definition of life. . . . The animism of younger children is much more implicit and unformulated. They do not question whether things know what they are doing, nor whether things are alive or dead, since on no point has their animism yet been shaken."[11]

By 10 years of age, the concept of causality begins to develop and matures further still with age, intelligence, and development. A pattern can be seen in children as they develop the concept of causality. Children ages five to six often cite nonnatural or violent causes of death; children eight to nine years old cite natural or illness-related causes; 11 to 12 year olds may cite spiritual causes such as it "being one's time."[12] Children who consider suicide or homicide may not be fully mature in their understanding of death and its finality.[13]

Notably, predictors of understanding death include cognitive ability and age, but not maternal communicative competence, as proposed by some researchers.[14] Specific cognitive development of a vitalistic causal-explanatory framework assists in children's learning about life and death.[15] Additionally, there is conflicting evidence about socioeconomic status, race, ethnicity, and exposure to death in how children assimilate their understanding of death.[16] Therefore, examining cognitive development may be the most helpful in learning what children understand about death.

While Jean Piaget's theory of cognitive development "emphasizes the active role of the child in discovering and constructing reality, evolving from sensorimotor approach, through stages of concrete reasoning, to the capacity for abstraction,"[17] Erik Erikson's psychosocial approach to development "describes psychosocial 'crises' that the child must negotiate in the formation of his or her sense of self and in relation to others (trust versus mistrust, autonomy versus shame and doubt, initiative versus guilt, and industry versus inferiority)."[18] Both of these approaches posit a linear progression through development, which is similar to other paradigms, such as Elizabeth Kubler-Ross's initial proposal of the stages of grief,[19] and Lawrence Kohlberg's[20] or Carol Gilligan's[21] description of the progression of moral development. However, children may vacillate or cycle among stages. Serious illness and death can challenge the forward progression, especially if a child's emotions and/or environment radically shift.

Children are savvy detectives and pay particular attention to hushed conversations, telephone calls, and "evasive" clues, especially when the topic is "off-limits" or secret.

CHILDREN'S EMOTIONS ABOUT DEATH

Children have emotional responses to death that are similar to those of adults. In addition to denial, anger, bargaining, depression, and acceptance, they can experience sadness, guilt, shame, pain, loneliness, despair, embarrassment, ambivalence, hope or hopelessness, helplessness, confusion, and peace, to name a few. One strong emotion that is intimately linked to death in adults and children is fear.

The fear of death may be innate, acquired, or a combination of the two. Gregory Zilboorg, Nelli L. Mitchell, and Karen R. Schulman, supporters of the idea that this fear is innate, believe that there is a need to fear death in order to preserve a species.[22] Similar to the stress responses of fight and flight, fear may be a response that is needed to avoid the danger of death for as long as possible. Mitchell and Schulman state that the "terror of not being"[23] begins in childhood and often persists. Mitchell and Schulman believe that this emotional response to death, especially the "dread of annihilation and mutilation," transcends the intellectualization of death.[24] However, they write, "the fear of death in children is intensified by the absence of the intellectual equipment and by the absence of necessary defensive mechanisms essential for comprehending

the experience of loss."[25] Therefore, the ability to repress or mindfully embrace this fear of death may be taught by society to aid in coping with loss or death.

Others believe that the fear of death may be acquired during the process of death conceptualization and maturation. For instance, Piaget proposes that young children seem to have less fear of death than older children because they do not truly understand death. However, Piaget remarks that some children "are haunted every night by fears of death, either for themselves or their parents."[26] This may be normal temporarily or may pathologically persist.

Further still, when children have not developed a biological conceptualization of death, rather than a behavior-related conceptualization, their fear of death can be exacerbated. As Slaughter and Griffiths describe, children's "immature conceptualization of death leads them to focus on un-resolvable questions like, 'why do some people I love decide to go live under ground instead? Will he or she come back soon? Isn't it cold down there?' "[27]

Loss, separation, and death are essential concerns for individuals, including children. Having more advanced cognitive and emotional development may help in coping with these essential concerns, notably anticipatory grief in children who are facing their own death or the death of a loved one.

CHILDREN FACING THEIR OWN DEATH: FOCUS ON CHILDREN WITH CANCER

Children with relapsed, progressed, or terminal cancer generally know that they are dying.[28] They are forced to confront mortality. They know this whether or not someone tells them.[29] Studies have found that children are keenly aware of death and often know more than their parents believe they do. Children are savvy detectives and pay particular attention to hushed conversations, telephone calls, and "evasive" clues, especially when the topic is "off-limits" or secret. Often times, children do not talk about death to protect their parents and family members.[30]

Despite communication with children about death, or the lack thereof, children experience anticipatory grief or, as Barbara M. Sourkes describes it, "grief expressed in advance when the loss is perceived as inevitable."[31] Family members may not even notice a child's grief. In order to identify anticipatory grief, family members must pay close attention. Children leave many clues. They begin to make matter-of-fact statements about death, play act, or draw about death.[32] They often times have nightmares, have sleep disturbances, or act out.[33] Those who attend to a dying child may observe the signs of preparation the child makes to confront impending death. As Sourkes writes, "The dying child's anticipatory grief is palpable as he or she lives the intensity of separation in its ultimate form."[34]

In coping with separation, especially a separation as significant as death, a therapist may be helpful. One dying six-year-old child said to a therapist, "Thank you for giving me aliveness."

When a child faces imminent death, it is devastating for all involved. There is a plethora of literature that delves into the physical, psychological, and social layers of complexity in supporting children, their family members, and their healthcare providers during this difficult time. One of the first to investigate how children perceive their terminal prognoses and death was the anthropologist Myra Bluebond-Langner, who completed *The Private Worlds of Dying Children* in 1977.[35] This ground breaking work explores awareness and communication in children with leukemia, at that time a terminal diagnosis. She found that children knew of their prognosis, even when parents and healthcare providers went to extensive lengths to "protect" them from that knowledge. This book began a wave of change that facilitated more open communication with children about their diagnosis and prognosis. It opened the door to provide shared experiences and a meaningful time for children and their family members at the end of life.

Since then, studies have found that parents who talk honestly with their children about death have

less regret than those who don't.[36] Children who are actively dying may experience physical, psychological, social, and spiritual pain.[37] Having an opportunity to address these woes can be beneficial and exceed the discomfort experienced by those who address them. Children fear suffering until the end of life, fear being alone at the time of death, worry about those surviving them,[38] and desire to make the most of the life they have left.[39] Open communication can provide a means to promote familial healing, including healing for well siblings who are often overlooked during a child's cancer journey.

COMMUNICATING WITH CHILDREN ABOUT DEATH

How do children communicate about death? Usually they ask questions like, "Why do people die?". This may occur after exposure to death in the media or experientially.[40] Most parents feel unprepared for such questions, and their responses vary. The best way to communicate with a child about death should be adapted to each child, parent, and situation. Parents must gauge their child's personality, temperament, and situation. While parents' words may vary, a gentle, caring, and responsive manner is better received than a cold, unsympathetic, or unresponsive one.[41] It is important to observe children's reactions during communication. They often provide their own "barometer," and will limit the amount of information they can handle. When they meet their limit, they change the subject, run away to play, or find a distraction. When they are ready to discuss death again, they will bring it to a caregiver or friend's attention.

Each child requires individualized communication, but there are some key concepts that can be helpful when discussing death. Slaughter and Griffiths write, "Researchers and clinicians have advised adults to discuss death in truthful, concrete and unambiguous terms with children."[42] This means using words such as "died," "dead," and "death," as opposed to "sleeping," "passed away," or "was lost." This communication must be developmentally appropriate, and it is appropriate to respond to a child's question with a question, such as "Why do you ask?". As Kenneth J. Doka advises, "Children of the same age are not necessarily of the same [cognitive] and emotional level of development."[43] If responses are out of proportion with a child's developmental level, the child's understanding may not be real. For instance, when children nod their head in silence and walk away, this does not verify understanding as it would in an adult; children often are attempting to have the adult leave them alone.[44] True understanding can be validated when children can explain what they were told in their own words. Sometimes, they may frame the information in terms of a familiar reference to death, such as a book like *Charlotte's Web*.[45] This childhood classic is often a favorite of dying children.

CHILDREN COPING WITH DEATH: PLAY THERAPY AND DRAWING

In coping with separation, especially a separation as significant as death, a therapist may be helpful. One dying six-year-old child said to a therapist, "Thank you for giving me aliveness."[46] Therapists and child-life specialists are trained to use play to approach children at their level. Sourkes states, "Play enables the seriously ill child to 'reenter' childhood."[47] She notes, "Shared imaginative play enables the child to confront the realities of life and death."[48] Other methods to help children cope with death include memorabilia, stuffed animals, and making legacy items.[49] Some children engage in making lists of feelings, drawing mandalas, and writing or reading books.[50]

ETHICS OF COMMUNICATING TO CHILDREN ABOUT DEATH AND DYING

Previously it was thought that withholding information from children about death would protect them, in a paternalistic form of therapeutic privilege. However, more recent research indicates that truthful disclosure in a developmentally appropriate manner is often beneficial to both children and families. Truthful disclosure may decrease children's psychological distress as they approach death and the distress of their bereaved surviving siblings.

Respect for persons is a key ethical concept to honor. For healthcare providers, it is important to ask parents, guardians, or significant family members what they prefer that the child be told, and how. Parents are more likely to allow a discussion of terminal illness or impending death with their child once they realize that the benefits of these discussions outweigh the harms. However, the benefits could be overshadowed by harms if a child is told in an inappropriate manner. It is better that parents help to determine the safest manner to communicate about an illness with their child.

The legal aspects of disclosing information to minors are not evaluated here, but cases exist that support both disclosure and nondisclosure to children. The courts often support the fiduciary duty of

honesty to trusting patients. From a justice perspective, there should be adequate resources provided to children and their family members to aid in their anticipatory grief and bereavement. One way to incorporate children into the process is to help them with age-appropriate advance care planning (ACP). Involving children in ACP may thus become an ethical and professionalism consideration for caregivers and clinicians.

INVOLVING CHILDREN IN ACP

ACP involves making decisions about the care a person would like to receive when the person cannot speak for her- or himself, or when the person's autonomy and capacity are limited. ACP for children, whose development of autonomy and capacity is emerging, involves discussing their hopes, wishes, and worries, and planning ahead for their future care, including the preferences of the child and family members. Making these decisions can be challenging, but it helps to honor the child's preferences and the family's values. These values and preferences may not be known unless they are specifically discussed. The obligation to learn about these values and preferences is not legally binding in many states, but it is an ethical duty for clinicians, especially for those who care for children with life-limiting or life-threatening illness.

Children have the added challenge of being minors; despite good conversations and advance care planning, their wishes may not be followed because they are not their own legal decision makers. Typically in pediatric care, parents, legal guardians, or surrogate decision makers provide permission for the care given to their child. In some cases, children may voluntarily provide assent for care; however, children do not have the power to provide consent, unless they are emancipated minors. Because children do not have the legal power or competence to provide consent or dissent to certain modalities of care, their voices may be lost in decision making, despite recommendations to involve children when appropriate[51] and possible.[52]

Before considering possible barriers to involving children in ACP, the general barriers to conducting advance care discussions for children with life-threatening conditions should be considered. Amy Durall and colleagues surveyed 266 healthcare providers and identified three main barriers to involving children in ACP: parents' unrealistic expectations, the differences between clinicians' and patients'/parents' understanding of prognosis, and parents' lack of readiness to have a discussion.[53] Durall and colleagues found significant differences in the barriers identified by nurses and physicians. Nurses more often identified ethical considerations to be barriers, and the unimportance of these ethical considerations to clinicians. Physicians said that ACP discussions are important, but not knowing what to say during theses discussions was a barrier. On the caregivers' side, potential barriers included cultural or religious beliefs, poor prognostic awareness, a fear of abandonment after a decision was made, feeling alone in making a decision, or fear of regret in making a decision.

Other barriers that parents reported included healthcare providers' reluctance to discuss ACP due to prognostic uncertainty, or because providers were not "willing to face up to the facts," and perhaps because parents had individualized needs, concerns, and coping mechanisms.

ACP does not mean "giving up" on a patient, but rather aligning the medical plan with the patient's and family's goals of care. Because the goals of care can shift over time as an illness evolves, as the side-effects of a treatment change, or when certain experiences with a treatment occur, ACP may need to be readdressed. Therefore, it is important for care-providers to communicate clearly that the patient and family members will not be abandoned, and will be supported in the decision-making process.[54]

Parents may have mixed feelings about ACP. Julia Lotz and colleagues conducted in-depth inter-

views with bereaved parents and found, on the one hand, that some parents reported that ACP helped them to be good parents, facilitated coping, and empowered them to make decisions for their child.[55] Parents believed ACP gave them a sense of control and security by preparing them for what was to come. On the other hand, some parents identified personal barriers of not feeling ready, wanting to focus on the present, or attempting to suppress burdensome thoughts. Other barriers that parents reported included healthcare providers' reluctance to discuss ACP due to prognostic uncertainty,[56] or because providers were not "willing to face up to the facts," and perhaps because parents had individualized needs, concerns, and coping mechanisms. Parents expressed a desire for ACP to involve shared decision making and a communication strategy that is gradual, sensitive, maintains hope, and involves children relative to their developmental maturity—which did not include infants. Specifically, parents "felt that their child should be heard and taken seriously even if unable to make treatment decisions."[57]

Potential barriers included parents' cultural or religious beliefs, or their concerns that their child did not have the cognitive capacity to understand, or the emotional capacity to participate. Another barrier was providers' or caregivers' not knowing what to say or how to answer a child's questions. Some caregivers might desire to have their child participate in ACP discussions, but that the first of these discussions not be with a healthcare provider. Giving caregivers time to discuss ACP with healthcare providers without a child present may help the caregivers process emotionally, freely obtain information, ask difficult questions, and develop an understanding without worrying about the child's response to the information. Some parents may prefer that a family member or a member of the healthcare team, other than the child's physician, discuss ACP with the child. For instance, a child may have a closer relationship with a child-life specialist and prefer to speak with that person.

Inviting a child to participate in ACP can be time consuming. A child may need to take breaks from the discussion, and ACP may need to occur over a series of conversations. Starting the conversations earlier in a child's illness may facilitate the gathering of information and give the child and family the space they need to formulate their thoughts. Because ACP takes into account the wishes, hopes, worries, and preferences of the child and family, the "plan" will be individually nuanced. ACP for children needs to be flexible, as patients and parents may change their minds on what they desire over time.[58]

Despite all of these barriers, knowing the wishes of children at the end of life is helpful. Healthcare providers have a fiduciary duty to protect the welfare of children, especially at the end of life. [59] By involving the preferences of children in ACP, not only are caregivers informed of what the children desire, but the possibility that their preferences are honored increases. When children's preferences are honored, their quality of life improves and family members have improved bereavement outcomes.[60]

There is little research around the involvement of young children in decision making, but guidelines of care continue to support patient- and family-centered or shared decision making. More research has been done with the involvement of adolescents and young adults (AYA) in ACP. By and large, research indicates that AYA want and are able to choose and record (1) the medical treatment they do and do not want, (2) how they would like to be cared for, (3) the information they want their friends and family members to know, and (4) how they would like to be remembered.[61] Some AYA who are undergoing hematopoietic stem cell transplant indicate that they want to continue many medical interventions at the end of life rather than limit them, which may demonstrate the conflict of balancing cure and comfort.[62] Like any patient at the end of life, AYA may change their mind. Jennifer Mack and colleagues found that AYA with recurrent or stage IV cancer initially favored life-prolonging interventions, but later preferred for comfort care.[63] Maureen Lyon and colleagues report on a randomized controlled trial with pediatric oncology patients; they found that the patients and families who completed an ACP program consisting of a survey, a Respecting Choices interview, and the *Five Wishes* program, were more likely to agree to limit treatment at the end of life than a control group who received standard care.[64] Similar results were found with AYA who had HIV/AIDS.[65] Adolescence is a time when separation from patents is normal, and teens are forming a better sense of self, as so AYA may mimic the values and preferences of their parents—or they may not. A transition of preferences and values does not necessarily need to cause familial discord. With open communication, families may come to understand the perspective of AYA, and vice versa.

COMMUNICATING WITH CHILDREN AND FAMILIES ABOUT ACP

Communication with patients and family members is the most important tool in establishing therapeutic relationships. If communication is stilted, im-

paired, misconstrued, or abused, the therapeutic relationship may be lost, and prevent any healing possibility. While communication is essential in all clinical encounters, it is of the utmost importance to be caring, compassionate, and clear when discussing ACP and end-of-life care (see table 2).

Respect for the values of patients and family members is essential to support a therapeutic relationship. This respect involves understanding the preferences of patients and families and aligning care with those preferences, as medically feasible and practically possible.[66] Coercion or an attempt to bias decisions per a healthcare provider's values is not ethically permissible. Inviting children into the discussion may be ethically permissible and is promoted professionally in the field of pediatrics. As the American Academy of Pediatrics states, "The child should participate to the fullest extent possible, given his or her preferences, cultural and spiritual tradition, illness experience, developmental capacity, and level of consciousness."[67]

How does one learn of these wishes or involve children, adolescents, and young adults in ACP? Building on the foundation of excellent communication, helpful guides to ACP for children include *My Wishes* and *Voicing My Choices,*[68] which are similar to the *Five Wishes* ACP guide. These ACP booklets avoid the legal jargon often seen in adult forms. Other interactive modalities include card and board games, such as "Go Wish"[69] and "Shop Talk,"[70] respectively. Another modality to communicate with children about ACP may include an interactive technology called "The Compass."[71] This tool, that is under development, uses technology similar to computer or video games to illicit a child's ideas, goals, priorities, and desires related to end-of life-care.[72] Teaching healthcare providers excellent communication skills and to utilize their support staff and resources may promote including children in ACP.

CONCLUSION

Cognitive and emotional responses to death can be striking, but children of all ages can be well supported in a developmentally appropriate manner. Discussing death may be uncomfortable, but if it is done in an honest, gentle, and caring manner, outcomes are improved. This includes communicating to dying children that they will not be alone. Alleviating fears, making memories, and giving children some control will help them to experience some comfort despite their distress. Leaving a legacy is important to children. For survivors, especially child survivors, open communication may facilitate resilience and decrease maladaptive responses, ranging from disturbed sleep to acting out. Involving children in ACP may not only improve goal-concordant care, but may bring solace to surviving family members. Utilizing this framework, talking about death can become less taboo, scary, and difficult, and instead become an opportunity for providers, families, and children to work together to give each child the best care possible.

NOTES

1. M.D. Glicken, "The Child's View of Death," *Journal of Marriage and Family Counseling* 4 (1978): 75.

2. V. Slaughter and M. Griffiths, "Death Understanding and Fear of Death in Young Children," *Clinical Child Psychology and Psychiatry* 12 (2007): 525-35, 526.

3. Ibid.

4. Briefly, Jean Piaget's theory delineates four linear stages of cognitive development that progress from the sensorimotor stage (typically birth to two years of age), the pre-operational stage (two to seven years of age), the concrete operational stage (seven to 12 years of age) and the formal operational stage (adolescence to adulthood). In the sensorimotor stage, children "understand their world through their senses and motoric manipulations." S.B. Hunter and D.E. Smith, "Predictors of Children's Understandings of Death: Age, Cognitive Ability, Death Experience and Maternal Communicative Competence," *Omega* 57 (2008): 145.

During the pre-operational stage, children use symbolic reasoning and magical thought in approaching the world; "their thinking at this stage is illogical because it is dominated by perception." Hunter and Smith, "Predictors," 145. They are often egocentric, have a fear of separation, participate in magical thinking, and believe in animism. *Animism* is the attribution of nonliving things having life or of living things, such as plants, having souls. Animism is similar to personification. Children older than 12 years see life as a property of animals and plants, but can distinguish animate and inanimate objects. In approaching something that is alive, children in this stage may see it as "functional, active or undamaged." Glicken, "The Child's View," see note 1 above, p. 75.

Those in the operational stage struggle to understand irreversibility and deny that death exists. A dead body may seem to be sleeping to them. They may also believe that their anger caused the death of another. In the operational stage, logical thought is used to solve concrete problems. Thus, death is concrete and can be externally or internally caused. The personification of death is realized. An example of this personification would be the "Grim Reaper," the "Angel of Death," or ghosts. In this stage, children may accept the finality of death, but more often they will apply it to others than to themselves.

Finally, in the formal operations stage, abstract thought and scientific reasoning arises. At this stage, death is final, universal, and becomes abstract. While a child is continuing to mature in this stage, the finality of death

TABLE 2. Communication for children with a serious illness who are approaching death

Age	Needs to be met	Anticipation	Coping and communication strategies	Advance care planning
Infants	Provide a secure, consistent, loving presence and comfortable surroundings.	Will not anticipate death, but may sense the anticipation of family members.	Encourage families to celebrate milestones and make memories. Provide families with requested information. Ensure social and emotional supports are available to families.	While infants will not be involved in ACP, parents, siblings, and family systems have particular needs with the loss of an infant.
Preschool	Provide a safe, loving environment. Help child interpret feelings expressed through actions. Provide realistic choices for child.	May anticipate death, especially if the child knows other children in a similar situation who died after doing a particular action (like going to the hospital or taking a medicine).	Encourage families to continue normal rituals as possible, celebrate milestones, and make memories. Storytelling and creating stories with children may help careproviders learn about a child's worries and hopes.	Invite the child to "tell his or her story" in a way that may aid in developing ACP.
School age	Provide a safe, loving environment and a listening ear. Provide realistic choices for the child. Roles and goals may provide purpose.	Will anticipate death and may have significant fear of separation. May ask details about death and dying. May focus on how unfair it is to be going through this. Will worry about how death will impact other family members.	Provide honest, straightforward explanations to questions; provide illustrations and demonstrations to facilitate communication. Child may desire to plan a funeral, write a will, or plan for special items (like a teddy) or family roles. Ensure connection with peers if desired. Celebrate milestones and make memories. Reassure child that pain will be treated. Let child share personal story with others. Listen to child.	Involving a child in ACP at this age is critical as it can empower the child. Play acting, drawing, or walking through a communication tool may be helpful. Consider the use of the "My Wishes" tool.
Preteen	Provide a safe, loving environment. Roles and goals may provide purpose.	Child will anticipate death, but at times may be emotionally labile, ambivalent, or focused. Expressions and actions may be inconsistent. Will express injustice regarding dying or death.	Honestly answer questions and address child's concerns. Distraction may help at times. Celebrating milestones, making memories, and especially making meaning from experiences will be important. Ensure that the patient is connected with peers, if desired.	Involving child in ACP at this age is critical, as it can empower the child. Asking open-ended questions or using a communication tool may be helpful.
Adolescents	Provide a safe, loving environment. Learn about short-term and long-term goals. Child may need help to balance hopes for the future and to find meaning in the present circumstances. Child will need others to know what is important to her or him.	While teen may be vulnerable while anticipating death, he or she may still make impulsive decisions when feeling invulnerable. Teen may worry that people will forget about him or her after teen dies.	Honestly answer questions and address concerns. Celebrate milestones, make memories, make meaning from experiences, and develop a legacy. Ensure teen has connection with peers and avoids isolation. Help teen maintain activities. Image may be a major focus for teens. Provide support around body image concerns.	Promote open communication with the healthcare team. Ask open-ended questions, listen to preferences and provide choices. Ensure ACP involves the teen. Consider using "Voicing My Choices" booklet.

can still be questioned. However, "the ability of the child at age ten to discuss death often signifies his emancipation from childhood. Children take oaths on their own death or parent's death. Funerals are considered ill omens and children cross fingers and say verses to ward off evil and protect themselves." Glicken, "The Child's View," see note 1 above, p. 76.

5. M.A. Callanan, "Diversity in Children's Understanding of Death," *Monographs of the Society for Research in Child Development* 79 (2014): 142-50.

6. A.B. Sood et al., "Children's Reactions to Parental and Sibling Death," *Current Psychiatry Reports* 8 (2006): 115-7; M. Cuddy-Casey and H. Orvaschel, "Children's Understanding of Death in Relation to Child Suicidality and Homicidality," *Clinical Psychology Review* 17 (1997): 33-45; Slaughter and Griffiths, "Death Understanding," see note 2 above; A.T. Bates and J.A. Kearney, "Understanding death with limited experience in life: Dying children's and adolescents' understanding of their own terminal illness and death," *Current Opinion in Supportive and Palliative Care* 9 (2015): 2; Hunter and Smith, "Predictors of Children's Understandings," see note 4 above, 143-4.

7. Personal mortality is often realized initially around six years of age, but develops more between the ages of eight to 11.

8. Slaughter and Griffiths, "Death Understanding," see note 2 above, p. 526; Sood et al., "Children's Reactions," see note 6 above, p. 115; Cuddy-Casey and Orvaschel, "Child Suicidality and Homicidality," see note 6 above, pp. 40-1.

9. Children in the sensorimotor stage do not have the cognitive and expressive skills for adults to investigate and classify as clearly as those of older children.

10. A child who has not yet understood cessation might ask a concrete questions such as, "How does a dead person breathe or eat underground?".

11. J. Piaget, *The Child's Conception of the World* (London, U.K.: Routledge and Kegan Paul, 1929), 210.

12. Bates and Kearney, "Understanding Death," see note 6 above, pp. 2-3.

13. Cuddy-Casey and Orvaschel, "Child Suicidality and Homicidality," see note 6 above, p. 42.

14. Hunter and Smith, "Predictors of Children's Understandings," see note 4 above, pp. 143-62.

15. V. Slaughter and M. Lyons, "Learning about life and death in early childhood," *Cognitive Psychology* 46 (2003): 1-30.

16. M. Tallmer et al., "Factors Influencing Children's Concepts of Death," *Journal of Clinical Child Psychology* 3 (1974): 17-9.

17. B.M. Sourkes, *Armfuls of Time: The Psyhological Experience of the Child with a Life-Threatening Illness* (Pittsburgh, Penn.: University of Pittsburgh Press, 1995), 8.

18. Ibid., 8-9.

19. Per Kubler-Ross, the stages of grief include the following: denial, anger, bargaining, depression, and acceptance. E. Kubler-Ross, *On Death and Dying* (New York, N.Y.: Macmillan, 1969).

20. The stages of moral development delineated by Lawrence Kohlberg focus on justice and include the following: (1) pre-conventional level, stage 1—obedience and punishment; (2) pre-conventional level, stage 2—individualism, instrumentalism, and exchange; (3) conventional level, stage 3—"good boy/girl"; (4) conventional level, stage 4—law and order; (5) post-conventional, stage 5—social contract; and (6) post-conventional level, stage 6—principled conscience. School-age children usually achieve the first level, whereas the general public achieves the second level, and only a minority of adults achieve the third level. There is a steady progression without jumping past the different stages. L. Kohlberg, "The development of children's orientations toward a moral order. I. Sequence in the development of moral thought," *Vita Humana Internationale Zeitschrift für Lebensaltersforschung* 6 (1963): 11-33; R.N. Barger, "A Summary of Lawrence Kohlberg's Stages of Moral Development," University of Notre Dame, 2000, *https://www.csudh.edu/dearhabermas/kohlberg01bk.htm.*

21. Carol Gilligan argued that Kohlberg's classification favors males. From a feminist and, some would say, ethics of care approach, Gilligan developed a schema for moral development that also has a progression through stages. She later states that she views moral development through histories or cultural framework rather than in stages, but the focus is the idea of linear developmental progression. The stages Gilligan delineates include: (1) preconventional with a goal for survival, and transition from selfishness to responsibility to others; (2) conventional with a goal of self-sacrifice and transition from goodness to truth that the other person is a person too; (3) postconventional with a goal to not hurt others or self (principle of nonviolence). C. Gilligan, "New Maps of Development: New Visions of Maturity," *American Journal of Orthopsychiatry* 52 (1982): 199-212; G. Jorgensen, "Kohlberg and Gilligan: Duet or duel?" *Journal of Moral Education* 35 (2006): 179-96.

22. G. Zilboorg, "Fear of Death," *Psychoanalytic Quarterly* 12 (1943): 467; N.L. Mitchell and K.R. Schulman, "The Child and the Fear of Death," *Journal of the National Medical Association* 73 (1981): 963-7.

23. Mitchell and Schulman, "Child and Fear," see note 22 above, p. 963.

24. Ibid., 964.

25. Ibid.

26. Piaget, *Child's Conception,* see note 11 above, p. 136.

27. Slaughter and Griffiths, "Death Understanding," see note 2 above, p. 527.

28. There have been great gains in cancer therapy and supportive care, leading to a survival rate of 80 percent for some cancers, but cancer is still a life-threatening illness. Notably, the outcomes vary among the different types of cancer, with survival rates greater than 90 percent in some forms of acute lymphoblastic leukemia and about 70 percent for some solid tumors and brain tumors. R. Siegel et al., "Cancer Statistics," *CA: A Cancer Journal for Clinicians* 62 (2012): 10-29; E. Ward et al., "Childhood and adolescent cancer statistics, 2014," *CA: A Cancer Journal for Clinician*s 64 (2014): 83-103.

29. Parents and society may try to hide the impending death of a person, including that of the child his- or herself. For instance, Isabel T. Gutierrez et al. found among "highly educated, mostly European American parents" that "35% of the parents . . . shielded their young children from representations of death in books, but twice as many parents (75%) . . . shielded their children from representations of death in television and movies." However, in the same study, "21% of parents said that they used a book about death as a resource for helping their young children cope with death." I.T. Gutierrez et al., "Affective Dimensions of Death: Children's Books, Questions, and Understandings," *Monographs of the Society for Research in Child Development* 79 (2014): 43.

30. Callanan, "Diversity in Children's Understanding," see note 5 above, p. 145.

31. B.M. Sourkes, "The Broken Heart: Anticipatory Grief in the Child Facing Death," *Journal of Palliative Care* 12 (1996): 56.

32. Some of the play acting may resemble a battle or fight, as war imagery is often portrayed by society when it references the "fight against cancer." This idea began many years ago; for example, Kearney wrote in 1977: "a former colleague Dr. R.K. Whyte liked to quote Shakespeare when initiating cancer treatment in children, 'Cry "havoc!" and let slip the dogs of war,' clearly indicating the nature of effective treatment. It really is a war and the child is the battlefield. For the parents it must be like living through a blitz. After a time a modus Vivendi emerges, but always there are reminders of the threat." P.J. Kearney, "Ethics, Cancer and Children," *Medical Hypotheses* 3 (1977): 178.

33. Doka found at Oakland Children's Hospital that "children exhibit their feelings through sleep disorders and acting out (personal communication) . . . [they] also engage in little adult behaviors: . . . acting like the deceased, experiencing psychosomatic illnesses, fearing illness, fearing losing control, regressing, being unable to separate, refusing to mention the deceased, losing appetite or overeating, bed-wetting, and feeling depressed." K.J. Doka, *Children Mourning, Mourning Children (*Washington, D.C.: Hospice Foundation of America, 1995), 128-9.

34. Sourkes, "Broken Heart," see note 31 above, p. 58.

35. M. Bluebond-Langner, *The Private Worlds of Dying Children* (Princeton, N.J.: Princeton University Press, 1978).

36. U. Kreicbergs et al., "Talking about Death with Children Who Have Severe Malignant Disease," *New England Journal of Medicine* 351, no. 12 (2004): 1175-86.

37. C. Saunders, "The symptomatic treatment of incurable malignant disease," *Prescribers' Journal* 4, no. 4 (1964): 68-73.

38. Bates and Kearney, "Understanding Death," see note 6 above, p. 5.

39. Ibid., 5-6.

40. Gutierrez et al., "Affective Dimensions," see note 29 above, p. 50.

41. Doka, "Children Mourning," see note 33 above, p. 20; K. Palma, "Talking to young children about death," *Boston.com,* 2015, *https://www.boston.com/culture/parenting/2015/07/28/talking-to-young-children-about-death.*

42. Slaughter and Griffiths, "Death Understanding," see note 2 above, p. 534.

43. Doka, "Children Mourning," see note 33 above, p. 90.

44. Ibid., 91.

45. That *Charlotte's Web* is a favorite among children, especially dying children, is not new. Parents were asked to list children's favorite books, and 3 percent of the books (six books) depicted death. Three of these six books were winners of the Caldecott Medal: *Charlotte's Web; Fables;* and *Sam, Bangs & Moonshine.* The other three books were *The Wonderful Wizard of Oz; Hansel and Gretel;* and *The Big Book of Dinosaurs.* Gutierrez et al., "Affective Dimensions," see note 29 above, p. 45. A. Wilkes, *The Big Book of Dinosaurs* (New York, N.Y.: DK Penguin Random House, 2015); E.B. White, *Charlotte's Web* (New York, N.Y,: HarperTrophy, 2016); A. Lobel, *Fables* (New York, N.Y,: HarperTrophy, 1983): J. Grimm and W. Grimm, "Hansel and Gretel," in *The Original Folk and Fairy Tales of the Brothers Grimm: The Complete First Edition,* reprint, trans. J. Zikes (Princeton, N.J.: Princeton University Press, 2014), 43-8; E. Ness, *Sam, Bangs & Moonshine* (Markham, Ont., Canada: Fitzhenry and Whiteside, 1966); L.F. Baum, *The Wonderful Wizard of Oz* (Chicago, Ill.: George M. Hill, 1900).

Bluebond-Langner observes that *Charlotte's Web* is often requested by dying children to be read to them in their last few days. Bluebond-Langner, *The Private Worlds of Dying Children,* see note 35 above.

46. Sourkes, "The Broken Heart," see note 31 above, p. 59.

47. Sourkes, *Armfuls of Time*, see note 17 above, p. 5.

48. Ibid., 7.

49. Examples of legacy items could include ink prints of hands or feet, photos, scrapbooks, video and or audio recordings, artwork, letters to read for the future, memory boxes, and locks of hair.

50. Some helpful books for bereavement include, but are not limited to, L.K. Brown and M. Brown, *When Dinosaurs Die: A Guide to Understanding Death* (Boston, Mass: Little, Brown, 1996); M. Heegaard, *When Someone Very Special Dies: Children Learn to Cope with Grief* (Minneapolis, Minn.: Woodland Press, 1988); J. Silverman, *Help Me Say Goodbye: Activities for Helping Kids Cope When a Special Person Dies* (Minneapolis, Minn.: Fairview Press, 1999); R. Temes, *The Empty Place: A Child's Guide through Grief* (Far Hill, N.J.: New Horizon Press, 1992); A.D. Wolfelt, *How I Feel: A Coloring Book for Grieving Children* (Fort Collins, Colo.: Companion Press, 1996); M. Gellman and T. Hartman, *Lost and Found: A Kid's Book for Living Through Loss* (New York, N.Y.: Harper Collins, 1999); P. Karst, *The Invisible String* (Camarillo, Calif.: Devorss, 2001).

51. What does the appropriate involvement of children mean? Discussions about death, dying, and the care that a person wants to receive at the end of life are very sensitive topics. Children in the sensorimotor and the pre-

operational stages of development—children typically below seven years of age—are rarely cognitively capable of contributing to an ACP discussion formally. Children in the pre-operational stage do not understand irreversibility and have magical thinking. Thus, in discussions of ACP with children who are in the pre-operational stage, the children should be assured that discussing the possibility of death does not cause death to happen.

52. V. Larcher et al., "Making decisions to limit treatment in life-limiting and life-threatening conditions in children: A framework for practice," *Archives of Disease in Childhood* 100 (2015): s1-23; S.E. Zinner, "The Use of Pediatric Advance Directives: A Tool for Palliative Care Physicians," *American Journal of Hospice and Palliative Medicine* 25, no. 6 (2009): 427-30: G. Villanueva et al., "End of life care for infants, children and young people with life limiting conditions: Summary of NICE guidance," *BMJ* 355 (2016): i6385; World Health Organization, "WHO definition of pediatric palliative care," n.d., *https://www.who.int/cancer/palliative/definition/en/.*

53. A Durall et al., "Barriers to Conducting Advance Care Discussions for Children with Life-Threatening Conditions," *Pediatrics* 129, no. 4 (2012): e975-82.

54. Villanueva et al., "End of life care," see note 52 above, p. i6385.

55. J.D, Lotz et al., " 'Hope for the best, prepare for the worst': A Qualitative interview study on parents' needs and fears in pediatric advance care planning," *Palliative Medicine* (2016): 1-8.

56. A tool that may be helpful to clinicians in determining whether a prognosis is poor is asking, "Would you be surprised if this patient died in the next 3- and 12-months?" K. Burke et al., "The 'surprise' question in paediatric palliative care: A prospective cohort study," *Palliative Medicine* (2017): 1-8.

57. Lotz et al., " 'Hope for the Best,' " see note 55 above, p. 4.

58. E. Beecham et al., "Keeping all options open: Parents' approaches to advance care planning," *Health Expectations* 20 (2016): 675-84.

59. Clinicians and caregivers may be more apt to invite children to join discussions about care when their illness has been refractory to standard treatment, the available treatment options offer little curative promise or life-extending benefit, and they are nearing the end of life. R.C. Barfield and E. Kodish, "Pediatric ethics in the age of molecular medicine," *Pediatric Clinics of North America* 53, no. 4 (2006): 639-48.

60. J.D. Lotz et al., "Pediatric Advance Care Planning: A Systematic Review," *Pediatrics* 131, no. 3 (2013): e873-80.

61. L. Wiener et al., "Allowing Adolescents and Young Adults to Plan Their End-of-Life Care," *Pediatrics* 130, no. 5 (2012): 897-905.

62. J. Needle and A.R. Smith, "The Impact of Advance Directives on End-of-Life Care for Adolescents and Young Adults Undergoing Hematopoietic Stem Cell Transplant," *Journal of Palliative Medicine* 19, no. 3 (2016): 300-5; C.K. Ullrich et al., "End-of-Life Care Patterns Associated with Pediatric Palliative Care among Children Who Underwent Hematopoietic Stem Cell transplant," *Biology of Blood and Marrow Transplantation* 22 (2016): 1049-5.

63. J.W. Mack et al., "Care in the Final Month of Life among Adolescent and Young Adult Cancer Patients in Kaiser Permanente Southern California," *Journal of Palliative Medicine* 19, no. 11 (2016): 1136-41.

64. M.E. Lyon et al., "Family-Centered Advance Care Planning for Teens with Cancer," *AMA Pediatrics* 167, no. 5 (2013): 460-7.

65. M.E. Lyon et al., "A randomized clinical trial of adolescents with HIV/AIDS: Pediatric advance care planning," *AIDS Care* (2017): 1-10.

66. While clinicians may have more training and medical expertise, the values of patients and their families may not align with the medical diagnostic and treatment options. Navigating conflicts can be challenging and may require an ethics consultation.

67. American Academy of Pediatrics, "Section on hospice and palliative medicine and committee on hospital care: Pediatric Palliative Care and Hospice Care Commitments, Guidelines, and Recommendations," *Pediatrics* 132 (2013): 967.

68. Wiener et al., "Allowing Adolescents," see note 61 above.

69. M.R. Potthoff, "Go Wish-Pediatrics: Pilot Study of a Conversation Tool in Pediatric Palliative Care," *Theses and Dissertations* (2015): 1140.

70. "Shop Talk" was designed specifically for youth living with cancer, and is not for all children and advance care planning. Lori Wiener et al., "Shop talk: A pilot study of the feasibility and utility of a therapeutic board game for youth living with cancer," *Support Care Cancer* 19, no. 7 (2011): 1049-54.

71. R. Barfield et al., "Mind the Child: Using Interactive Technology to Improve Child Involvement in Decision Making About Life-Limiting Illness," *American Journal of Bioethics* 10, no. 4 (2010): 29.

72. Ibid., 29.

Parental "Holding and Letting Go" in End-of-Life Decision Making in the Neonatal Intensive Care Unit

Lisa C. Freitag

ABSTRACT

By the time an infant is born, his or her life has already been woven into a family narrative that forms part of the identities of parents and family members. The parents have often begun holding an identity for the expected child, and begun processing their own new identity as a parent. Philosopher Hilde Lindemann calls this narrative process "holding in identity." When a neonate is born prematurely or with severe illness, this emerging narrative can be altered or even interrupted, leaving parents in a situation, not just of fear and uncertainty, but also questioning their own identity as parents.

This article builds on Lindemann's theories of family holding infants in identity at the beginning and end of life to explore the work that must be done by all parents of a newborn, and to consider the effect that family narrative might have on decisions at the end of life in the neonatal intensive care unit (NICU). I argue that, even early in life, conflicts over a child's best interest might be framed as misunderstandings or misconceptions about the child's and the parents' identities. This article highlights parents' difficulties adapting to their new parenthood in the NICU environment, examines the common narrative of the baby as "fighter," and explores the need for parents to "let go" as part of consenting to the withdrawal of aggressive treatment at the end of life. In order to support family in the difficult task of choosing a premature ending to their severely ill child's life narrative, we must understand the relationship between parent and infant as they struggle to get to know each other in the alien NICU environment.

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INTRODUCTION

According to feminist philosopher and bioethicist Hilde Lindemann, a person's identity is formed, at least in part, from narratives constructed by family and close social contacts.[1] This narrative identity is begun before birth, and continues throughout a lifetime. Lindemann believes that maintaining and upholding a unique identity for each family member is an important function of family relationships. Parents in particular are called to guard and encourage their children's emerging, unique identity as they grow and develop their own life narrative.

At times when a person is unable to maintain her or his identity for her- or himself, such as during severe illness or dementia, families can use this ongoing narrative to uphold the identity of their loved one. In her recent book *Holding and Letting Go*, Lindemann suggests that proxy decisions at the end of life that are made by family or close friends should take into account the lifelong identity of the patient.[2] In this way, medical decisions made by family surrogates can become not just decisions about what the person might have wanted, but affirmations of who the person has been. Conflicts about withdrawal of aggressive therapy at the end of life can thus be framed as arguments about the person's iden-

tity and the most appropriate way to end the person's life narrative.

Lindemann also suggests that families continue to hold an identity for a family member after death. Family members "hold in memory" the deceased person, remembering who the person was, what the person did, and, often, how the person died. I believe that family members also take on some responsibility for the events leading up to the death, particularly if they are called upon to "let go" of their loved one by actively making an end-of-life decision. The family narrative of this time might include negative feelings that make it difficult for family members to hold onto their own narrative identity. In order to cope with the death and their responsibility for it, family members must craft a new narrative that will allow them to honor and remember the person who has died in a way that is consistent with all their identities.

For infants, the creation and upholding of identity has only just begun, but still may have an impact on parental medical decision making. Because infants have not yet had a chance to emerge as the narrators of their own story, the family's holding of the infant's identify forms an even more significant part of the infant's identity. Prior to a birth, a family will often construct a deep picture of the child's personality and life story, based on their own life situation and expectations. During the newborn period, the infant's identity remains largely in the hands of the parents, as they gather information about their newborn's personality through caring for him or her. At the same time, parents must reconstruct their own identities to become the parent of this particular child. They are simultaneously learning who their child is and examining who they themselves are becoming as a result of the new addition to the family.

When a neonate is born prematurely or with severe illness, this process is confused and interrupted. Parents are effectively thrown into a narrative that does not match what they have been preparing for. In the midst of finding themselves as new parents to a new and unique person, they must also adapt to a new and unfamiliar life situation. If the baby is critically ill or not expected to survive, they must absorb complex medical information while their own lives have been altered.

End-of-life decisions are always difficult, particularly in the NICU, where life has barely begun. It is especially difficult when parents and members of the healthcare team disagree about the best interests of the baby. There are times when the NICU team understands that it is their duty to maintain life, but parents refuse resuscitation or other procedures. It can also go the other way, when parents insist on continued efforts and neonatologists recognize that further treatment is unlikely to be of benefit. These two instances are very different, both in moral reasoning and in parental understanding and motivations, so we will consider here only the latter, when parents will not accept recommended withdrawal of aggressive treatment.

Parents and NICU staff have different levels of knowledge about the available treatments and their likelihood of success. The importance of adequate communication to narrow this gap is obvious. But despite the best of communication, parents don't always come to the same conclusion as the NICU staff do about the best interests of the baby. I argue that these differences may have very little to do with knowledge level, and might, instead, be due in part to different ways of viewing the baby's identity in the context of his or her family.

This article reviews Lindemann's theories of family holding identity and identity formation in infants. Through narratives written by parents of NICU babies and statements made by parents whose infants died in the NICU, I will explore the ways that the NICU environment may alter or inhibit the identity formation of infants and parents. Parents must adapt to this environment and achieve their identities as parents despite the obstacles. We will then discuss two common narratives that might inform parental decisions at the end of life: the infant as a "fighter," and the making of memories that become an important part of "letting go."

IDENTITY FORMATION IN INFANCY

On the surface, it would seem that a newborn baby in the NICU has very little to offer in the way of a unique personal identity. Babies don't do very much that allow us to recognize their unique personalities. However, according to Lindemann, each individual's personal identity is partly formed by a narrative process she calls family "holding."[3] This holding often begins before a baby is born, as the expectation of a new member is added to existing family narratives. All infants are born not only into a time and place, but also a culture and family that will inevitably mold the person they will become. The family "holds" an identity for each member, constructed from societal expectations and, eventually, the stories told about him or her by family and close contacts. The family "maintains" this identity for each member, and it becomes the basis from which each person's individual narrative and unique

identity grows. Lindemann states, "we can't be who we are without the other persons who initially hold us and maintain us in personhood."[4]

Families who are "holding well" can maintain a person's identity when it is threatened, thus maintaining their personhood for them. The most obvious example of this is in dementia in the elderly; when a family holds the person's identity for them, reminding them (and themselves) who they are through stories and photographs.[5] But this is true at other times as well. The process of maintaining an identity begins in infancy, and an identity can likewise be held for newborns, nonverbal children, and other individuals who cannot (yet) tell their own story. Eventually most children become able to tell their own story, adding to or correcting the identity formed by family. Those who cannot do so—as in severe intellectual disability—must continue in the identity crafted for them by family.[6]

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In the early stages of infancy, the formation of identity takes place in the context of caregiving. As parents do routine care such as bathing, feeding, or changing, they remain open to physical cues from the infant that are clues to his or her emerging personality. They might determine oral likes and dislikes, mood or temperament, and ways of receiving comfort. Even a diaper change can become a sharing of individual attitudes. At the same time, the parents are learning their own new role as they begin to construct stories about the emerging person in their care. This process is part of parenting, and can be a source of joy as well as labor. As Noddings puts it, "When my infant wriggles with delight as I bathe or feed him, I am aware of no burden but only a special delight of my own."[7] A verbal response is not required, just responsive physical interaction.

In some cases, even this physical interaction is not required. Hilde Lindemann points out that the process of identity formation can include an individual who cannot, and may never, take part in this reciprocal responsiveness. Her example is her sister Carla, who was born with anencephaly, yet was held by her family as a valued daughter and sister. Lindemann is not alone in this. Philosopher and bioethicist Carl Elliott has suggested that we all tend to imbue even the most neurologically devastated children with personhood by treating them as persons deserving of respect, and expecting that the "child will be a part of a family like any other child, that her life will have a narrative like that of an ordinary human being."[8] He calls this "taking an attitude toward a soul." Every baby, even the most impaired, is a valued individual with a life narrative taking place within his or her family.

Thus, we can assume that parents in the NICU will begin the process of discovering a unique identity for their child, even if they are unable to interact physically with a very sick baby. It is unlikely that parents will interrupt this process on being told that a poor neurological outcome is anticipated. The expected family narrative for both parents and child has been altered by the illness or disability, but parents will not hold such an infant as less of a person.[9]

Parents seem to be in agreement that each infant has a unique identity, no matter how undeveloped, when they understand that one child is not replaceable with another. In a Wisconsin study based on interviews with the parents of infants who had died in the NICU, for example, although the parents reported that focusing on their other children was "helpful" in coping with the death, all 19 of the parents who were questioned said that "having another child did not replace the child who had died."[10] I suspect that both the impact that the deceased child's existence made on the family story, and the unique identities conferred on family members by the child's life and death, contributed to this irreplaceability.

To consider an infant at the end of life in the NICU, however, we must make one more observation—that identities are reciprocal. Taking an attitude toward a soul not only creates personhood, but confers a certain identity on the person doing it. Lindemann's sister Carla, without even being aware of it, conferred on little Hilde the identity of sister,

and on their mother the identity, not just of mother, but of *Carla's* mother. Thus, every child is not only presumed to have a unique identity that might emerge given time, but also gives all of the people with whom the child interacts irreplaceable identities of their own. In constructing and holding an infant's identity, family members find a new identity for themselves. I maintain that this new identity—as mother, father, sibling, grandparent—is actually more important than the infant's as yet unformed identity. Parents who must make end-of-life decisions must then defend their infant's existence and their own emerging identities in relationship to the infant.

At the end of life, "holding well" includes knowing when and how to "let go." In the final chapters of her book, Lindemann talks in detail about the ways in which an end-of-life decision might inform and be informed by a person's life narrative. She provides the example of an elderly man in an intensive care unit (ICU) following a devastating heart attack, and discusses the ways in which the medical decisions made by the patient's proxy decision maker might affirm or deny his lifelong identity. She points out that medical decisions can thus become not just decisions about what the person might have wanted, but affirmations of who the person has been.[11] Thus, conflicts about withdrawal of aggressive therapy at the end of life can be framed as arguments about the person's identity and the most appropriate way to end her or his life narrative. The events surrounding the death become part of the person's life story, and may, to some extent, redefine it.

Parents who are faced with making an end-of-life decision in the NICU must also make a decision that will define their child's life. It may be that the shortness of the life in question makes the importance of their infant's identity more, rather than less, urgent. We often speak of parents' needing to "let go" when aggressive measures are failing, but we rarely define what, exactly, it is that they are supposed to let go of. They must let go of the story they have constructed to welcome a new life into the world. I suspect that in order to properly let go, parents must first discover who they, themselves, have become as a result of the child's short existence.

Letting go is not only an action that eases the end-of-life decision. It is also the beginning of a new phase in the family's holding, in which the family holds their loved one in a new way—in memory.[12] An infant does not have a long life story to hold, but the memories of an infant's short life in the NICU are particularly intense.

We often speak of parents' needing to "let go" when aggressive measures are failing, but we rarely define what, exactly, it is that they are supposed to let go of.

The stories lived by the family, together with the act of treating the infant as though it is a full person (taking an attitude toward a soul), identify the infant as a person of value, whose short life has had an enormous impact on the stories of the family members. That impact cannot be fully encompassed in the fraught circumstances of an unexpected birth or end-of-life decision for an infant who ought to be at the beginning of life. Nonetheless, I believe that examining the way parents form an identity for their new baby, and rework their own identities in response to the birth, may be helpful in guiding parents through the most difficult decision they are likely ever to make.

IDENTIFYING AS A PARENT IN THE NICU ENVIRONMENT

In the NICU the family's narrative about what to expect at childbirth has gone wrong, often suddenly and unexpectedly. In this era of excellent prenatal care and widespread genetic screening, parents, particularly mothers, are led to believe that the outcome of their pregnancy is under their direct control. When a baby is born with serious medical issues or extreme prematurity, the family's expectations can be shattered, and the child's story is launched into a new and uncharted trajectory. The stories about what to expect can no longer be relied upon.

According to anthropologist Gail Landsman, societal pressure in the wake of improving prenatal care has created a mandate for parents to have a "perfect baby" and to become perfect parents. This expectation creates difficulties for those mothers whose babies are born with illness or disabilities, framing them as failures who "must have done something wrong." A sick baby means guilt and blame, and the parents must work doubly hard to be seen as good parents.[13] Some struggle with identifying themselves as parents at all.

Identifying as a parent is made more difficult by the NICU environment. The NICU is no place to work out the issues faced by parents who are meeting their baby for the first time. For most it is an alien place of harsh lights, urgent beeps, hushed voices, and inexplicable equipment. The expected baby's birth transmutes from an anticipated joyous event—sometimes one that wasn't due to happen for weeks or months yet—to a time of uncertainty and emotional upheaval. In this place, parents feel lost, unimportant, and perhaps not yet a parent at all. In this place a parent must go through the transition in thought and attitude that is becoming a parent.

Hendricks and Abraham, researchers in Switzerland, conducted interviews with parents who had made an end-of-life decision for their extremely premature infant.[14] Parents were asked to describe the events around their decision, recalling their experiences in the NICU one to two years after their infant's death. Some of the parents described being in a sort of mental fog of confusion and alienation. These parents were uncertain about what their contribution to the decision had been. Some reported being so dissociated from the activities in the NICU that they felt there had been no decision at all; that there never had been any choice to be made. Other parents reported being confused or in shock, and unable to make decisions. One mother stated, "I so to speak just watched as if I was not involved. . . . I was not really aware, and I could not really perceive the situation. It was as if in a dream, it could not be true, everything was fine. It took a while before I completely understood that it was my child, my child who was dying."[15]

Hendricks and Abraham state that there had been ongoing efforts in their NICU to involve parents actively in decisions. This demonstrates a level of confusion and dissociation that interferes not only with decision making, but also with coming to recognize oneself as a parent. The Swiss researchers felt that good communication between parents and staff was key, but I wonder how this can be attained if parents are in a fog of disbelief so deep that they doubt their own parenthood, if not the reality of the whole situation.

Even familiarity with the NICU environment does not immunize against this feeling of being lost and confused. When my son was born at 33 weeks, as a pediatrician who had dealt with many sicker babies, I knew that he was in no danger. I knew the function of every one of the things surrounding him, and the meaning of every monitor beep and lab result. Yet there was nothing that I could do that even resembled my idea of motherhood. Picking him up and breast-feeding him, as I was eventually urged to do, seemed preposterous. I couldn't learn how to be a mother. Instead I fell back on my role as pediatrician, examining the charts and watching the monitors until the nurses shooed me away.

Several mothers of extremely premature infants have attested to the difficulty of accepting motherhood in the NICU. Vicki Forman, in her memoir about her 23-week premature twins, confesses that she needed several days to start to claim them as her own. Her first acknowledgment of her motherhood, seeing her helpless infants in their isolettes, was to ask herself, "Who will love them if not me?"[16] Another mother, Deanna Fei, whose daughter was born at 23 weeks, writes in her memoir, "The only way to brave this limbo is simply to bear witness. To bear witness is to know her as she is, no more and no less. To know her is to love her, because she is mine, because I am her mother. The more I love her the harder it gets."[17] Later Fei attests to the ongoing learning process of motherhood: "Each day she survives is another day she has survived. Each day I hold her is another day that I'm learning to be her mother."[18] Both of these women had difficulty accepting their premature motherhood, as well as figuring out exactly what that new identity meant to them.

Many mothers, both in memoirs and in research studies, describe the sensation of standing by helplessly as others converge on the infant to provide care. The baby seems to need doctors and nurses, not the clueless parents. Another study investigated how women experience becoming a mother in a NICU in Sweden.[19] Interviews with mothers of preterm infants who had been in several different NICUs in Sweden revealed that mothers felt a sense of separation that kept them from feeling like mothers. Some thought that they were extraneous—their baby required medically trained staff, not the sort of caregiving they had anticipated. One mother felt that she was actually in the way when she visited.[20] Another mother in the same study said, "Well, it's

like having a baby but still not having . . . it's not really my baby. Because without the hospital and the incubator this baby would not live one day. So, you're having a baby in a glass cage that you can visit."[21] The baby needs so much more than the parents can provide, and nurses often must take over even basic care tasks.

Parents in both the Swiss and Swedish cohorts reported that their role as parents was made more difficult by the lack of physical contact with their baby.[22] As I mentioned earlier, much early information about a baby's personality comes through caregiving, as the baby reacts to new stimuli, foods, and experiences. The things that cause crying or give comfort become part of a developing identity. If identity formation for both parents and infant does occur in the context of physically caring for a newborn, it should not be surprising that parents in the NICU have trouble both getting to know their baby, and learning who they have become as parents.

Recognizing this, several strategies for involving parents in NICU care have been proposed. Breastfeeding is often suggested as an important maternal activity that will involve the mother and perhaps boost maternal confidence. If breast-feeding is going well, mothers did report pride in "being such good mothers." But mothers required a lot of support and reassurance to negate bad feelings if the feedings were not going so well.[23] And there are some hints that this optimistic and positive action might prove to be an additional burden if the baby dies. Mothers whose baby had died in the NICU reported that attempts to express milk had been stressful, and that lactating during the baby's death increased their suffering.[24] This is a way for mothers to connect and "do something" for their baby, but it seems inadequate if not burdensome if the infant does not ever get the milk.

In a study from the U.K., researchers observed parents' interactions with infants who were still in the NICU. Parents were actively taught to read their baby's signs of discomfort so that they could participate in comfort care. Parents were able to overcome their hesitancy to touch their infant, make acute observations about the infant's behaviors, and sometimes even contribute information valuable to the staff in providing care. This allowed parents to "develop a unique knowledge of their own infants," and "establish their own roles as caregivers."[25]

Fathers also experience difficulty with separation and are frustrated by their inability to give care. The father of a 24-week preemie writes in his memoir, "Intent on making us active, if symbolic, participant's in Josie's care, [the nurses] showed us how to change her doll-sized diapers, how to put lotion on her skin if it was dry, and how to hold her hands and feet in tight to her body to calm her if she was stressed. . . . This was as much as we could do for Josie at the time, and there were many days when she was so agitated and so stressed that we could do nothing."[26]

I suspect that any measures taken by NICU staff to increase parents' involvement can be only partly effective. Parents will have difficulty accepting their new circumstances in this alien environment where they are, essentially, helpless. Perhaps they can come to terms with this frustration as the NICU stay extends, but it certainly delays the process of feeling like one has become a parent.

Perhaps this is the reason that homecoming figures so strongly in parents' memoirs. Longing for home is a frequent theme in parents' narratives of their NICU experiences. Vicki Forman likely speaks for all parents when she writes, ". . . all you ever wanted when your child had been in the hospital for prolonged periods of time: to be home."[27] Part of this longing is undoubtedly due to the impression that the ordeal will finally be over when the baby comes home. "Graduating" from the NICU is the final step in a series of NICU achievements, and signals an end to the relentless NICU "roller coaster."[28] But home is also the place where parents are finally given charge of their own infant, and can engage in all of the activities of parenthood. They have full responsibility for caregiving, and full access to their infant. Forman states that she did not dare fall in love with her son until she got him home.[29]

There is very little we as practitioners can do to alter the NICU environment so that it is more comfortable and engaging for parents. The birth of a sick baby will remain a time of disorientation and fear. And babies will continue to need scarily dramatic and invasive care in order to survive. I believe most NICUs do well in supporting parents through the disorienting experience that must take place in this foreign and intimidating space.

But parents, particularly mothers, are searching desperately to identify themselves, not just as parents, but as good parents for their infant. There is no instruction book for becoming a parent of a baby who is very ill or dying. This is not what they were expecting when they were "expecting." They no longer know how to act or who they must become. Perhaps all we can do is be aware of the enormous shift that the altered conditions of their baby's birth has made to their life story. Rewriting this narrative is by necessity a slow process, which will likely continue well beyond the infant's NICU stay. Many

parents must revise their identity as parents, and must be affirmed in that role—slowly, patiently, and persistently—while they do so.

THE BABY AS "FIGHTER"

Finding a new identity as a parent does not rely on specific personality traits of the baby. I found no evidence of parents who claimed specific knowledge of "what the baby wanted" or "who the baby is." But there is an identity specific to the infant that NICU babies often acquire—that of being a "fighter."

The baby is a fighter because he or she wiggles during an exam, or resists an IV insertion, or even is able to go down a notch on ventilator settings. The praise transfers from infant to parent.

The language of illness as a personal battle is everywhere. It is high praise given to people with a serious illness—adults and children alike. It is so common that there is practically a moral mandate to be a "fighter." Woodwell says of his premature daughter, "We had a real fighter there, someone special."[30] Charlie Gard's mother, who achieved international attention and sympathy for refusing doctors' recommendations to withdraw treatment, said, "We don't know until we try. He's still fighting, and we're still fighting."[31] The moral overtones of being a "fighter" are clear. It is so special and admirable that it can be equated to a fight for justice.

It may be worth examining how babies become "fighters." I have frequently heard the term used by NICU staff as encouragement or reassurance for parents. The baby is a fighter because he or she wiggles during an exam, or resists an IV insertion, or even is able to go down a notch on ventilator settings. The praise transfers from infant to parent.

Anthropologist Linda Layne, after having her baby at 30 weeks, studied the language used with parents in the NICU. She found the metaphor of the "roller coaster ride" to be both prominent and helpful for understanding the "alternating moments of hope and despair that [she] experienced" during the "seven long weeks" that her baby was in the NICU.[32] No matter their eventual outcome, NICU babies do indeed have both progress and setbacks during their NICU stay. I believe that it is during those rebounds, at the top of the roller coaster's progress, that babies become "fighters."

Nurse researchers in Norway have been studying a quality in NICU babies that they call "vitality." They observed infants and interviewed both nurses and doctors, and conducted interviews with parents who had recently experienced making end-of-life decisions. The nurses define vitality as an elusive but observable quality that is "used synonymously with the verbal expressions 'spark of life' and 'fighting spirit.' "[33] This seems very like identifying a NICU baby as a fighter. Vitality was detectable by parents, neonatologists, and experienced NICU nurses as a certain strength, level of activity, and reactivity. Infants who demonstrated it were felt to be "able to protest and struggle in resistance, to be active and decisive," to be "demanding," "gutsy," and even "angry."[34] The researchers felt that the babies who showed vitality were more likely to survive, and proposed the use of vitality as a moral addition to the factors that should be taken into account when making end-of-life decisions.

Interestingly, the researchers ascribed several moral qualities to those infants who demonstrated vitality, as though vitality was attained not by survival instincts but by strong moral fiber. Babies with vitality had "an ability to 'decide' their own fate" and babies without it seemed "as though they would rather not live." Babies with vitality were "different from all the others" and "someone special." Vitality was "a signal that they would not give up, that they were little Vikings. The infants decided themselves how things would turn out."[35] They had "the will to come out of a hopeless situation."[36]

The study did not, unfortunately, record the adoption of the term vitality by parents, although a few of the parents who were interviewed did report on the presence or absence of it in their baby. The researchers defended its use as an instinctual measure that could and should be used to aid the making of end-of-life decisions, but they did not report

on its ultimate effect on such decisions in their NICU. In particular, they did not report instances when a baby's level of vitality diminished or when there were differing opinions on an infant's vitality that might have increased the difficulty of making end-of-life decisions.

I suspect that impressions of vitality or the assignment of the identity of "fighter" cannot be easily cast aside if an infant's condition declines. As the parents ride the roller coaster of the NICU experience, they are encouraged by the term. They seem to take pride in their baby's identity as a fighter, and in themselves as good parents for their part in his or her continued survival. Once a baby has been identified as a fighter, possibly merely by surviving numerous setbacks, it is perceived that he or she has declared a will to live.

Linda Layne postulated that, because our culture prefers linear narratives with a known ending, parents tend to choose between a narrative of survival or one of death. In order to cope with the day-to-day disappointments and advances, they tend to focus on and prepare for one outcome or the other.[37] However, I believe that parents alternate between these two narratives, alternately planning futures and funerals as they ride the ups and downs NICU roller coaster. When a baby overcomes multiple setbacks, thus becoming a fighter, parents are perhaps more likely to begin to insist that a positive outcome is assured.

At the end of life, an identity as a fighter can be toxic. To the parents, approached for permission to withdraw aggressive care, the idea of withdrawal looks very much like giving up. Not only is this culturally and morally impermissible, but the infant "fighter" has shown his or her preference for survival by surviving so far. He or she could not be giving up. Good parents may feel that it is their parental duty to protect their infant and stand against withdrawal of aggressive care.

At such times, NICU staff often resorts to the language of suffering. But once a baby becomes a fighter, suffering becomes inconsequential. Several of the words used to describe vitality can also be used to describe discomfort; protesting and struggling perhaps in response to painful procedures. Parents have probably been reassured that none of the intrusive things that have been done so far are hurting their baby and that any pain was treated with narcotics. So, for a fighter baby, either suffering has been negated by comfort measures, or any suffering is a sign of life, a ticket that has already been at least partially paid. The baby has consented to it by the fact of survival, demonstrating a will to live.

When we ask parents for permission to withdraw aggressive care, we are telling them that it is time to give up, and perhaps implying that their baby is not a fighter after all. Perhaps it signifies that all of the suffering the baby (and they) have been through was a wasted effort. If so, we need to provide a more appropriate identity that does not negate the struggle so far. Perhaps the metaphor of a battle valiantly lost would suffice, although I rather dislike war metaphors in medicine. Perhaps a tragic destiny too strong to be overcome would be more poetic. Or perhaps we should just own up to the fact that medicine cannot make good on all of its perceived promises. Death is no one's fault, dying is not a tragic flaw, and our mistake is in overstepping our bounds once again, like Icarus flying too close to the sun.

HOLDING IN MEMORY

I mentioned above that Hilde Lindemann's theories of family holding suggest that, after the death of any family member at any age, the survivors must learn to hold their loved one in memory. This holding encompasses grieving for the death and remembering the ways in which the dead person held you, while alive. The person's life story can be told and retold at funerals, ceremonies, and by other memorials.[38]

The families of neonates who have died also have their infant's life story, however brief, to remember and to tell. In the Wisconsin study of parents' experiences with end-of-life care in the NICU, the participating parents all brought mementos of their deceased baby to show the researchers. These families saved and cherished artifacts from the infant's NICU stay: photographs, footprints, and clothing. Many still celebrated the infant's birthday, or acknowledged their child's absence at holidays to keep the infant's memory alive. Several wished to make donations or participated in volunteer activities as a legacy to their child.[39]

A study of parents' narratives following the death of a baby with trisomy 13 or 18 done in Saint Louis, Missouri, found that telling the child's story was very important to parents. Many reported that telling this story was the reason they'd agreed to participate in the study, that they found the telling helped to support themselves, and, they hoped, would have an impact on others.[40] Their infants had spent an average of 74 days in the NICU, and some had gone home before dying. The infants had an identifiable genetic trisomy known to have dire consequences on survival. However, I doubt that this

significantly alters the importance to parents of the infant's life story.

The researchers reported that parents "saw their child as having a name, a personality, meaning and purpose. They understood them as being woven into a framework of relationships that included their family, their friends and their God."[41] This echoes Lindemann's theories about family holding, and the way in which we actively bestow personhood through taking "an attitude toward a soul." These are babies who would not have been resuscitated a decade ago. Perhaps resuscitation both creates and confirms their identities as persons or "souls." The NICU made it possible for them to have a life story, but their families are responsible for living the story and holding it in memory or otherwise.

The NICU made it possible for them to have a life story, but their families are responsible for living the story and holding it in memory or otherwise.

The way in which a person dies, in my experience, is a large part of the story that is eventually told about them. The family must deal with, not just the loss of their loved one, but the memory of the part they may have played in the death and the events leading up to it. Survivors often accept guilt that is not rationally theirs, and continue processing the experience for years after a death, making statements like, "If only I had called the ambulance sooner," or "Maybe we should have transferred to a different hospital." I have concluded that all of the survivors (including medical staff) need to arrive at a story they can live with. That story is likely to be less painful if they do not feel personally responsible for causing or contributing to the death.

Anthropologist and nurse Jacquelyn Slomka has reported that end-of-life discussions between physicians and families often become a form of negotiation during which "moral responsibility" for the death is shared. Everyone desires to avoid responsibility for causing the death. In her study of adult patients in the ICU, she observed that physicians attempted to shift responsibility to the family or to the patient, who would be encouraged to decide to withdraw aggressive treatment. Or all parties might wait for an unresponsive patient to "declare himself." Both the family and the staff needed eventually to accept some of the moral responsibility for the failure of medicine to prevent the death.[42]

It is perhaps not surprising that some parents, faced with tragedy at a time that should have been the beginning of a life, will refuse to share that responsibility. Perhaps their confusion and denial will allow them to avoid the decision, or their part in it, altogether. However, in my experience, some perceive a request for permission to withdraw aggressive treatment as an effort to make them help "kill their baby." If the baby has previously declared him- or herself to be a fighter, the parents might take an even more active stance, insisting that the baby deserves to stay alive no matter what support might be required to maintain that life.

When parents are studied regarding their perspectives on end-of-life care, they agree that parents should participate in end-of-life decisions, although the actual extent of their participation varies.[43] In addition, several parents in the Swiss study who had fully accepted their part in making a decision to withdraw aggressive therapy pointed out that making that decision required a high level of parental responsibility.[44] It is likely that some parents are not ready to accept this responsibility. Some are in shock or denial, and perhaps do not recognize the decision they are helping to make. Others, however, seem to refuse to participate.

I believe that many parents who refuse to allow withdrawal of aggressive therapy fully understand what they have been told about their child's dire illness, the story as told by the physicians and the medical records. Even when parents do not understand or are in a fog of disbelief, it is unlikely that it will be effective to simply repeat the medical staff's story. Instead, parents need to be given a different story, a different way of being a parent, rather than fiercely guarding life. This story is not told by the infant's medical course, but must be found in the parents' stories of becoming parents in a hostile environment and accepting the child as their own.

They must remember that they were good parents, standing by their child in crisis, and giving him or her every chance. They must remember that their child—whose identity and life narrative will always be held only by themselves—was resuscitated, lived in the NICU, and ultimately could not stay with them.

Vicky Forman asked for, and was reluctantly granted, withdrawal of life support for her daughter Ellie, who was born at 23 weeks and lived for three days on maximal life support before having a grade IV intraventricular hemorrhage. In her memoir written 10 years after Ellie's death, Forman wrote, "I will never have the answers to the questions surrounding Ellie and her short life. What if we hadn't insisted—would she have survived? What if I had been more ready to raise a profoundly disabled child—would that have made us better people? What if another set of parents had been in the same situation—would they have made the same decision?"[45] Her memoir attempted to find an answer to these questions, but did not entirely succeed. She lived those 10 years with sole responsibility for her daughter's death.

A more desirable outcome was expressed by one of the parents who participated in the study in Switzerland. Recalling the decision she shared with her physicians at the end of her baby's life in the NICU, the mother said, "I did not experience this moment as a freedom but rather as a responsibility of course because the baby could not decide for herself. We are her parents and we should make this decision. . . . Now in retrospect, I regard that as a great act of love. But in those hours, I thought I would die. But you do not die and you go on and you have to decide."[46] This is a statement full of sorrow, but also of strength. It acknowledges the difficulty and responsibility of her decision to allow the neonatologists to withdraw therapy. She can carry forward this singular act of love as she holds in memory the infant who is still, and always will be, hers.

We must find a way to promote the sort of strength that makes allowing death into an act of profound parental love, although I suspect that, since each parent's narrative is different, there is no consistent was to do this. Perhaps we, too, must learn to accept the inevitability of death. Perhaps we must assume more responsibility for death ourselves by admitting the limitations of medicine more often. Or perhaps we can allow death to occur without assigning fault or failure to anyone. At best, we can hope to stand by the parents' side while they begin rewriting the narrative of their baby's life. We can acknowledge that, whatever happens to their baby, they remain forever the mother or father of that particular life. Even if the baby dies, the parents will be coming to terms with their NICU story, and holding their infant in memory for the rest of their lives.

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CONCLUSION

Infants have a unique identity that begins to be formed by their families before birth. This identity is relational, and the infant's identity grows along with his or her parents' identities as the parents of that particular child. In the NICU, some of the identity work that must be done by the parents is interrupted by the suddenness of the birth, the presence of illness, and/or the assumption of specialized care by the NICU staff. When a baby is unlikely to survive and withdrawal of aggressive care is recommended, the fragility of the parents' identities may add to the difficulties with parental decision making.

Each infant's identity is unique, and every story must be different, but there are common themes. The NICU environment is almost universally foreign and overwhelming. This, as well as the unexpectedness of a sudden birth, contributes to a sense of disorientation and unreality. Parents are thrown into this new narrative and don't know how to act. Some have difficulty assigning themselves the identity of par-

ents, and will need to be shown the way to becoming good parents despite the infant's setbacks.

It is perhaps unfortunate that parents and neonatal staff alike have adapted the language of battle and assign to infants the title of "fighter." This is both a descriptive and moral term. It is given to babies who survive multiple setbacks, and identifies them as having an admirable desire to live. However, we must be aware that it may become a difficult label to overcome if medical treatment is not sufficient. There are times when the fighter narrative must be set aside and a new story created. This narrative is only partly found in medical facts, so the identity work cannot be done by bluntly repeating a dismal prognosis. It must be done by gently reconstructing the parents' ideas of what it means to be parents.

Parents can perhaps be guided toward a story that works for them, acknowledging the infant's difficult life while still preserving its meaning. In the midst of confusion and grief, parents need to find a story where the best and most loving thing they can do for their baby is accept death. They need to be shown how to hold their child and themselves in their identities, and also to let go, so that they can continue to hold their child in memory. Those identities must not burden them with guilt or label them as bad parents. They need help to craft memories that celebrate the child's existence, yet permit that existence to end.

They need to create a story they can live with.

NOTES

1. H. Lindemann, *Holding and Letting Go: The Social Practice of Personal Identities* (New York, N.Y.: Oxford University Press, 2014).

2. Ibid.

3. Ibid.

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Mature Minors, Mature Decisions: Advance Care Planning for Adolescent Patients with Life-Limiting Illness

Jennifer Needle, Maureen Lyon, Donald Brunquell, and Catherine Heith

ABSTRACT

Advance care planning (ACP) for adolescents is not yet a standard of care; as such, adolescents with life-limiting disease are often left out of important discussions about end-of-life (EOL) care, namely, advance care planning. This narrative review will describe the ethical, legal, developmental, and biological concepts and perceived barriers to including adolescents in EOL decision making. We will briefly explore adolescents' autonomy, the legal definition of capacity, and how these evolve through adolescence as minors mature and become more experienced with their illness. We argue that adolescents' participation in ACP not only supports adolescents who want to participate, but also assists the parties who are legally responsible for making decisions. This participation leads to goal-concordant care and reduced conflict. Finally, we address common misconceptions about EOL discussions and argue that EOL care does not diminish hope in patients or families and is still possible in the face of prognostic uncertainty. Involving adolescents in ACP respects the autonomy and growing capacity of adolescents and promotes patient- and family-centered care at EOL.

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Nearly 400,000 children in the United States live with potentially life-limiting diagnoses, including cancer, cystic fibrosis, heart disease, and progressive neurologic disease.[1] Many older children and adolescents with life-limiting illness have a long history with their disease and understand its potential complications, including the possible need for life-sustaining treatment. For many of these patients, initial discussions about their preferences for end-of-life (EOL) care happen late in the course of their disease, when there is often inadequate time for deliberative discussion.[2] The patients' perspectives, preferences, and goals for future medical care may not be elicited because their parents or guardian are their legal decision makers. Exploring the perspectives of adolescent patients demonstrates respect for their evolving autonomy and can enhance patient-centered care related to decisions at the EOL.

Advance care planning (ACP) is a process that supports individuals' choice of treatment options and their right to refuse unwanted medical interventions. ACP promotes patients' autonomy, a fundamental principle of clinical ethics. When persons are not in a position to make or communicate their own healthcare choices, ACP is critical to help guide healthcare professionals (HCPs) and surrogate decision makers in making patient-centered medical

decisions. The advance directive documents that may follow ACP are legally binding for patients who are 18 years and older. ACP in pediatrics is complicated by several factors: uncertain prognoses, legal requirements to obtain patients' informed consent and determine their capacity to make decisions, and the need to balance the evolving autonomy and capacity of minors with the need to protect them from emotional and difficult conversations. Because they lack the legal right to make decisions for themselves, adolescents have not traditionally been included in the ACP process, even when there is evidence they have well-considered goals and preferences. Although there is increased attention to the rights of adolescents as autonomous decision makers, significant barriers remain that have ethical and practical implications for patients, families, and HCPs.

There is a growing research interest in the experience and preferences of adolescents, as well as a growing consensus among professional societies[3] that ACP is an important and necessary component of adolescents' care and treatment. Despite this, adolescent ACP is not yet the standard of care. There are only a handful of formal pediatric ACP programs in the U.S., and research on adolescent ACP is even more sparse. Unsubstantiated beliefs and biases among HCPs that preclude adolescents from participating in ACP may result in a lack of, or late, referrals to palliative care, and to potentially undesired treatment at the EOL.[4]

The purpose of this narrative review is to address the concepts, potential barriers, and evidence related to adolescent ACP in the framework of key bioethical principles. Our goal is to foster a professional discussion about the benefits of ACP for adolescent patients and their families.

ETHICAL PRINCIPLES AND LEGAL CONSTRUCTS RELEVANT TO ADOLESCENTS' DECISION MAKING

Autonomy is a key bioethical principle related to decision making. The principle of autonomy requires that patients have the capacity to act intentionally, with understanding, and without controlling influences that would mitigate against a free and voluntary act.[5] Autonomy is tied to the legal concept of decision-making capacity, which requires (1) an understanding of necessary information, (2) an appreciation of the situation and its likely consequences, (3) the ability to reason about treatment options, and (4) the ability to communicate a choice.[6] From a legal standpoint, with few exceptions, once they reach the age of majority—18—adults are assumed to be competent to make medical decisions. On the other hand, adolescents are presumed to be incompetent to make autonomous medical decisions, and a "burden of proof" rests on individual patients to demonstrate their capacity to make autonomous decisions. Importantly, the ethical and legal requirements for autonomy do not preclude adolescents from being aware of their medical condition or prognosis, nor their contributing to decisions regarding their care. We advocate for individualized determinations of autonomy based on the definition above, not chronological age or state law, as the determinant of whether or not an adolescent can, and should, participate in EOL decision making.

While parents retain the legal right to make decisions for adolescents, at this age patients often have capable, stable preferences regarding their own medical care.

Parents have the legal and ethical duty and responsibility to make medical decisions for their child. This duty is best understood as a right in trust that is held to provide a child's "right to an open future."[7] For young children, the *best interest standard* for decision making is used, since these children do not yet have the intellectual or developmental capacity to make decisions about EOL care. The capacity for mature thought processes and decision making grows with time. Parents may use the principle of *substituted judgment* to make decisions for older children, based on what they believe their child would choose. While parents retain the legal right to make decisions for adolescents, at this age patients often have capable, stable preferences regarding their own medical care. Adolescents should be included in EOL discussions to the extent of their

capacity, which appropriately acknowledges their autonomy.

Many states have established some version of the "mature minor doctrine," either by statute or in the courts, which allows adolescents to be emancipated to make their own decisions or provides a degree of freedom for minors to provide informed consent or to refuse medical treatment. Specific regulations vary by state. The circumstances around which this consent is considered are the following.

1. The minor is an older adolescent (14 years or older).
2. The minor is capable of giving informed consent.
3. The treatment will benefit the minor.
4. The treatment does not present a great risk to the minor.
5. The treatment is within established medical protocols.[8]

It is beyond the scope of this article to address the conflicts that exist between the principle of autonomy and the law as it relates to adolescent decision making. However, HCPs are encouraged to consider the necessary elements of autonomy and decision-making capacity in granting minors a voice in their healthcare decision making. Hospital ethics committees and legal counsel should be utilized should conflict arise in unique cases.

THE EVOLUTION OF CAPACITY AND AUTONOMY IN ADOLESCENT PATIENTS

For adolescents, maturity and decision-making capacity increase along a spectrum. Some of this variation is neurobiological, some is due to adolescents' environment and social structure, and some is experiential. Prior illness experience may give adolescents insight into their treatment preferences and a better understanding of the context of their decisions. The American Academy of Pediatrics supports the recognition that "some pediatric patients, especially older adolescents and those with medical experience because of chronic illness, may possess adequate capacity, cognitive ability, and judgment to engage effectively in the informed consent or refusal process for proposed goals of care."[9]

Theories of cognitive development suggest that by the time adolescents are 15, they possess a capacity to make informed decisions that is similar to that of adults.[10] It is known that the appraisal of risk and reward may lead adolescents to make decisions differently than adults do.[11] The Dual Systems model suggests that risk taking, and thus risky decision making, is the result of competition between the early maturing affective system and the more slowly maturing cognitive control system.[12] Cognitive control, which is responsible for planning and judgment, is critical for informed medical decision making and is believed to be not fully mature until individuals reach their middle 20s.[13] This developmental model may be different for adolescents with chronic illness, due to the alteration of their normal developmental processes due to school interruptions, altered social experiences, changes in self-image due to illness, and changes in family dynamics. Hinds and colleagues found that 90 percent of patients with cancer aged 10 to 20 years understood their treatment options and the consequences of EOL decisions in which they participated.[14] Among the most commonly cited factors influencing EOL decision making by adolescents with cancer was "having previous experience with life support measures."[15] These findings suggest that some children, and most adolescents, possess the competencies needed to make informed decisions.

Decision-making processes in adolescents are more significantly influenced by peers and social networks than in mature adults.[16] Peers play a large role in social support for healthy adolescents, however those with chronic illness rely more on their family members (particularly their mother) during treatment and find them more supportive than their friends.[17] It is recommended that ACP involve parents to open the lines of communication about these sensitive topics and to enhance adolescents' support systems. Additionally, given the variability with which adolescents develop the capacity to make complex decisions, we recommend ACP programs with an interdisciplinary focus (that include HCPs, social workers, chaplains, ethicists) that enable highly individualized conversations and ongoing inquiry into adolescents' interests and capacity to participate in decision making.

ADOLESCENTS HAVE A DESIRE TO PARTICIPATE IN ACP AND HEALTHCARE DECISION MAKING

Several research studies have found that the majority of adolescents with chronic illness desire to participate in making their own healthcare decisions[18] and to be informed of their illness and treatment options when they are terminally ill.[19] These researchers report that adolescent patients with possibly life-limiting illness prefer to have discussions about EOL care early in the course of their illness.[20] But the data also suggest that adolescent patients'

preferences for participation in healthcare decisions, as well as receiving prognostic information, is highly individual and may change over time.[21]

Studies with older adults found that ACP provides benefits to patients and their surrogate decision makers: ACP has been found to reduce stress, anxiety, and depression by engaging surrogates in discussions regarding their loved one's preferences,[22] and to reduce decisional conflict.[23] Hinds and colleagues report that, of the parents they studied who had children with terminal cancer, 94.7 said that "choosing as the patient would want or as the patient previously directed the parent to choose" was a factor in EOL decision making.[24] Research conducted using the Family-Centered Advance Care Planning for Teens with Cancer (FACE-TC) intervention found that a significant majority of patients said early discussions about potentially serious outcomes was "helpful."[25] A study that used the FACE intervention with adolescent patients who had HIV reported that 25 percent of the patients said ACP made them "feel sad," but 98 percent said "it was worthwhile," and 94 percent felt that "it was something that I needed to do."[26]

One of the primary barriers to EOL discussions in pediatrics is a concern that the discussion itself will take away the hope of patients and family members, but research findings do not support this concern.

ACP DOES NOT DIMINISH HOPE AND CAN BE USED IN THE FACE OF PROGNOSTIC UNCERTAINTY

One of the primary barriers to EOL discussions in pediatrics is a concern that the discussion itself will take away the hope of patients and family members,[27] but research findings do not support this concern.[28] Mack and colleagues report that parents who receive greater disclosure of prognostic information about their child's cancer were less likely to find the information extremely or very upsetting. For children with cancer with a low likelihood of cure, honest and open communication about prognosis and decision making made parents feel more, not less, hopeful.[29] Numerous studies report that parents and adolescents want information from HCPs that is complete, honest, and delivered with sensitivity.[30] ACP interventions should explore patients' wishes when cure is not possible, including discussion of care and services such as pain control and dying at home.

Prognostic uncertainty is another barrier to initiating ACP.[31] HCPs may provide overly optimistic prognostic estimates to patients,[32] which may lead to ineffective aggressive treatment at the EOL.[33] This may be more reflective of HCPs' discomfort in discussing EOL issues rather than patients' lack of readiness.[34] Sharing best estimates of prognosis is an important element of ACP;[35] however, eliciting goals, values, and preferences regarding a possible negative outcome can be done with an estimate of the likelihood of that outcome. By conducting discussions early in the disease course and working toward normalizing these conversations as a standard of care for all adolescents with potentially life-limiting illness, ACP can foster trusting communication and establish patients' and family members' preferences for sharing information, and not diminish their hope.

CONCLUSION

Despite evidence that many adolescents possess decision-making capacity that is similar to that of adults, adolescents' participation in medical decision making has been limited. From the perspective of pediatric HCPs, the legal and ethical constructs surrounding adolescent decision making may appear to be in conflict. HCPs may rightfully be concerned about the legal consequences of supporting adolescents' EOL preferences, especially if they are in conflict with those of their parents.[36] By facilitating frank discussions during a time of relatively good health and utilizing a framework for determining adolescent patients' maturity and capacity for decision making, ACP has the potential to reduce con-

flict between patients, their parents, and HCPs. To this end, ACP gives patients a voice in the medical care they receive, helps family members to better understand the values and preferences of their loved one, and provides an extra layer of support to HCPs. By normalizing discussions about goals and values, models of care that routinely include ACP can reduce possible social and systems barriers to adolescents' participation in decision making, and reduce the possibility that conversations about future decisions do not reduce patients' hope for desired outcomes. The inclusion of adolescents in ACP supports a patient- and family-centered approach to decision making that is desirable for all stakeholders.

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Re-Examining the Ethics of Clinical Trial Approval in a Gene-Targeted Era through a Case-Based Approach: Exploring the Implications of the *n* = 1 Batten Disease Trial

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ABSTRACT

Using a case-based approach, this article highlights the necessity of re-examining the ethics of clinical trial approval in a gene-targeted era. It centers on the *n* = 1 Batten disease drug trial, in which an antisense oligonucleotide drug was introduced as a novel drug therapy for an individual child with a neurodegenerative illness, and probes the ethics of equity, safety, clinical utility, and parental autonomy of the trial as a means to raise more global questions about the ethical underpinnings of clinical trail approval. With the promise of the widespread application of personalized medicine on the horizon, it is imperative that the greater scientific community think critically about the ethical foundation upon which drugs make their way to market.

For M.M.'s parents, there was nothing but excitement when they welcomed their first child in 2010. "When I think about [M.M.] as an infant, I think about smiles . . . and laughter . . . and health," her mother recalls.[1] When M.M. was a toddler, her father described her as being "very physical," noting that she "always loved to ride her Strider [rocking toy] . . . play in the snow . . . [and] go sledding," in their hometown in Colorado. Her parents had no sense early on that their daughter was in the throes of a serious neurodegenerative decline. But when M.M. turned three, subtle signs emerged that suggested otherwise.[2]

M.M. began to get "stuck on her words." She would stop mid-sentence, as if unable to complete her thoughts. Given that she had consistently met all of her developmental milestones up to that point, her medical care team was not initially concerned. But with each passing month, her behavior grew stranger, and her parents began to ask questions. By age four, M.M. started pulling books close to her face to make out the images. At five, she began moving her feet in an unusual "pit-patter" manner, repeatedly stumbling and falling. At six, M.M. displayed worsening gait, language, and behavioral regression, and a complete loss of vision. She was declining rapidly, and her doctors did not know why.[3]

An extensive evaluation performed at Children's Hospital Colorado in 2016 revealed multisystem deficits. M.M. was found to have severe vision loss, in association with bilateral macular and retinal dystrophy, and resultant trace optic disc pallor on the right. An electroencephalogram (EEG) showed severe bilateral cerebral dysfunction, with multifocal and generalized epileptiform discharges suggesting subclinical seizure activity. Magnetic resonance imaging (MRI) of her brain and spine revealed significant cerebellar atrophy. After an extensive molecular workup, an answer as to M.M.'s predicament

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emerged. It was the not the answer her parents had hoped to hear.[4]

M.M. was diagnosed with Batten disease, or neuronal ceroid lipofuscinosis, a rare, fatal, inherited condition caused by lysosomal accumulation of ceroid lipofuscin. Swaiman and colleagues explain that patients with the disease suffer from "cognitive impairment . . . [progressively] worsening seizures . . . [and] loss of sight and motor skills."[5] According to the National Institute of Neurological Disorders and Stroke, Batten disease affects two to four of every 100,000 live births in the United States, and about 14,000 children worldwide.[6] It has no definite cure.

M.M.'s parents were determined to change that—for their daughter and for all patients afflicted with the disease. Their first step was to clarify the genetic underpinnings of the disorder. M.M.'s original Batten gene panel was sent from Children's Hospital Colorado after a skin biopsy showed characteristic changes. The panel identified a heterozygous change in the CLN7 gene, which meant that there was a second gene at play in her disease. This was because the CLN7 disease variant of Batten disease is classically recessive, requiring a homozygous change in two alleles at the particular gene locus. Seeking more answers, M.M.'s parents forged a special partnership with Claritas Genomics, a diagnostic laboratory then associated with Boston Children's Hospital. Claritas offered to provide whole exome sequencing for M.M., with the hope that the second gene that contributed to her Batten disease presentation would come to light.

The results indicated that M.M.'s specific mutation was caused by a retrotransposon insertion in the CLN7 gene. Retrotransposons are components of the genome that utilize a "copy and paste" mechanism to insert to target gene sites, and are abundant in the DNA of eukaryotic organisms (organisms that include animals and plants). In M.M.'s case, the insertion was acting as an exon trap, in that it caused alternative splicing and consequent gene truncation and loss of protein function of the gene.

A new question arose as to how this newly identified genetic defect could be addressed in a therapeutic context. Timothy Yu, MD, PhD, a researcher affiliated with Boston Children's Hospital, proposed using the Claritas data to develop an oligonucleotide drug that would block the specific abnormal splicing process that was occurring in M.M.'s genome and restore the normal CLN7 gene product.[7]

This endeavor sought to capitalize on the recent successes of similar therapy approaches for other childhood diseases. Oligonucleotide drugs that target mRNA splicing mechanisms had recently been introduced. In December 2016, the U.S. Food and Drug Administration (FDA) approved nusinersen (Spinraza), an antisense oligonucleotide that was designed to alter irregular SMN2 splicing, the first definitive therapy for spinal muscular atrophy (SMA).[8] The drug had been accepted for FDA priority review in November 2016 "to address the urgent need for an effective SMA treatment" in light of the positive statistical data on improvement in motor milestones demonstrated in the drug's two Phase III studies (see table 1 for descriptions of the various phases in experimental drug trials).[9] In September 2016, the FDA approved the oligonucleotide drug eteplirsen (Exondys 51) for treatment of Duchenne muscular dystrophy, based on efficacy data collected from four different Phase I through Phase III trials that were conducted between January 2009 and September 2014.[10] The clinical trials for nusinersen and eteplirsen involved sample sizes far greater than what would be utilized in M.M.'s case, which would be a clinical trial with a research population of one subject: $n = 1$.

Yu agreed to lead the research efforts, and, faced with M.M.'s increasingly rapid neurologic decline, her parents launched an online advocacy platform to raise the million dollars necessary for undertaking the $n = 1$ clinical trial for their daughter, and to seek FDA approval for moving forward with the project. Within a few months, the idea for a new drug therapy had a fast-tracked timeline to begin a clinical trial.[11]

TABLE 1. Phase I - II - III versus $n = 1$ trial design

Phase	Sample size (n)	Length of phase	Purpose of phase
I	20 - 100	Several months	Safety (determination of doses tolerated without signficiant side-effects).
II	100 -1,000	Several months to ~ 2 years	Efficacy of varying safe dosages.
III	1,000+	1 to 4 years	Safety. Efficacy on a large scale.
I-II-III	1	Several months	Safety (determination of dosages tolerated without signficiant side-effects). Efficacy of varying safe dosages.

The development of the clinical trial broke new ground in numerous ways, not only due to its highly accelerated timeline, but also due to its sample size. As a trial initiated for an individual, there was an inherent blurring of the classic structure of the phases of clinical trials, in which periods of active treatment with an experimental drug are followed by "washout" periods, when the experimental drug is "washed out" of the system of research subjects. In the Batten disease trial, the safety of various dosages of the experimental drug would be monitored alongside the efficacy of the drug over the period of a few short months. In the usual structure of a clinical trial for an experimental drug that involves human subjects, a small research population (usually fewer than 100 people) participates in Phase I of the trial, and the subjects are carefully monitored for side-effects at a range of drug dosages over a period of several months (see table 1.) In Phase II of the trial, hundreds of human subjects are strictly monitored to assess the efficacy of the experimental drug at varying "safe" dosages over a period of months to a period of two years. Significantly larger cohorts participate in Phase III of the trial, in which data are collected regarding the safety and efficacy of the drug over a period of one to four years, utilizing the dosage information that was acquired in Phase II.

The stark contrasts between the design of the $n = 1$ trial and that of traditional Phase I, Phase II, and Phase III trials indicate the need to develop and/or, at the very least, reexamine the ethical framework for approving clinical trials with human subjects, especially as gene-targeted therapies overtake nontargeted therapies. In 1966, Henry Knowles Beecher published his seminal work "Ethics and Clinical Research" in the *New England Journal of Medicine*, in which he urged the scientific community at large to adopt a significantly more stringent ethical standard for scientific experimentation involving human subjects, and shed light on what he deemed "troubling practices" in the U.S.[12] This occurred on a backdrop of an already evolving federal framework that regulated how new drugs were brought to market, but it was Beecher's resolve that ultimately paved the way for the development of the carefully designed I-II-III phase system for clinical trials by as early as the 1970s. As widespread personalized medicine is imminently on the horizon, the ubiquitous use of the $n = 1$ trial design will require a reconsideration of the ethical principles on which Beecher's imperatives rested. The remainder of this article will consider the significance of the Batten disease trial in the larger context of clinical trials, with an examination of the key ethical principles of equity, clinical utility, safety, and parental autonomy.

EQUITY

Massive budget cuts to the U.S. National Institutes of Health (NIH) proposed by the Trump administration in March 2017 threaten to reduce NIH's funding by roughly six billion dollars, and continued monetary provisions for biomedical research are at risk.[13] Particularly at risk are funds for orphan diseases, defined as "a rare disease or condition . . . that affects less than 200,000 persons in the US."[14]

The Orphan Drug Act (1983), however, has functioned for more than 30 years to help offset the costs of drug-related research for rare diseases. This legislation comes from an era in when the costs of drug research skyrocketed in response to the increased safety and efficacy data required for the approval of new drugs. The Kefauver-Harris Amendment of 1962 was the U.S. response to the drug thalidomide's highly publicized role in contributing to defects in human embryonic development.[15] As a result of the 1962 ruling, independent researchers and pharmaceutical companies were required to comply with significantly more stringent guidelines to get new drugs to market. Cost effectiveness became an even more important factor in the pharmaceutical industry's selection of projects to pursue, and rare diseases were progressively cast to the side. Patients with rare diseases and their families banded together to form the National Organization for Rare Disorders (NORD) in 1982, an advocacy organization that was instrumental in the passing of what ultimately became the Orphan Drug Act of 1983.[16]

The Orphan Drug Act facilitated the revitalization of drug research for rare diseases in a number of ways. The act provided seven years of market of exclusivity for companies that developed a drug to treat a rare disorder, which limited competition and allowed a window of profitability. It offered exemptions from certain FDA fees, tax credits for various research-related expenditures, and governmental aid for research.[17]

These federal policies supporting drug research for rare diseases help to counterbalance the high cost of developing treatments for the diseases. The concept of equity, as outlined below, plays a significant role in solidifying governmental support of the development of treatments for rare diseases. In some ways, M.M.'s drug trial raises questions about the durability of that justification.

The current policy-influencing stance on equity centers on a rights-based approach, which is derived

from a notion of "natural rights" put forth by the 17th century philosopher and physician John Locke.[18] Hughes, Tunnage, and Yeo argue that a rights-based approach to healthcare is grounded in the idea that "individuals in a society are entitled to a decent minimum of health care . . . [and are deserving of] the same quality of treatment as other patients."[19] This is juxtaposed with a consequentialist view of equity, which contends that, as a society, we are obligated to make decisions that center on "bringing the greatest good to the greatest number."[20] The rights-based argument is supported by the concepts of "fair innings" and the "rule of rescue."[21] In his article "Intergenerational Equity," Williams defines the concept of "fair innings" as the "feeling that everyone is entitled to some 'normal' span of health."[22] Jonsen, in his seminal article "Bentham in a Box: Technology and Healthcare Allocation," defines the "rule of rescue" as "a perceived duty to save endangered life where possible."[23] The "duty" Jonsen refers to highlights the influence of the Kantian imperative for beneficence, in that it underscores the obligation of a civil society to protect the most vulnerable and to uphold the collective morality of its citizens.[24]

The equity arguments that support the Orphan Drug Act are problematic when applied to the clinical trial approved for the oligonucleotide drug designed for M.M., due to the neurodegenerative aspects of her disease. Williams's "fair innings" argument includes the notion of a " 'normal' span of health," but this is a challenging concept in the context of M.M.'s illness, which had already caused significant neurodegenerative decline. The drug promised to restore the gene product so as to prevent further neurologic change, but there was no likely component of reversibility.[25] There are also questions regarding the validity of an equity-based position; from the standpoint Jonsen's "rule of rescue," this concept is applicable when an individual faces "a real threat of avoidable death."[26] But can neurodegeneration be equated with "avoidable death" here?

Thus, rights-based equity arguments do not provide strong ethical support for the Batten disease trial. Approval of the trial also does not appear to be sound from a consequentialist approach to equity and/or a look to cost effectiveness. A consequentialist approach (that is, an approach that holds that what is right or best is what makes the world better in the future) would be valid, however, in the context of an $n = 1$ trial as a "proof of concept," in which the trial itself could prove (or disprove) that the approach proposed by the researchers has widespread downstream applicability. Hughes argues that the broader societal implications fostered by data that are collected from a trial on an individual could be argument enough to satisfy the requirement of delivering "the greatest good to the greatest number."[27] But in the case of this trial, there was a lack of novelty—and consequently a lack of "proof of concept"—because the FDA had already approved nusinersen and eteplersin in the market of antisense oligonucleotide as exon-skipping therapy.

The neurodegenerative aspects of M.M.'s disease and the lack of "proof of concept" for the n = 1 trial do not provide an ethical justification for the trial based on the principle of equity.

The neurodegenerative aspects of M.M.'s disease and the lack of "proof of concept" for the $n = 1$ trial do not provide an ethical justification for the trial based on the principle of equity. More generally, the trial provides fodder for a broader discussion of the definition of "orphan disease," as disease processes will increasingly be identified by the gene mutations that create them. If an "orphan disease" is defined as one that affects 200,000 or fewer individuals in the U.S., how will new subsets of more common diseases be identified as they emerge? Where will the line be drawn when resources for the development of new drug therapies are allocated? Who will draw these lines, as new federal guidelines permit the chairs of institutional review boards (IRBs) to approve $n = 1$ trials for novel therapies without the standard review process, thereby bypassing the committee and its deliberations?[28] Such expedited review may eliminate the community voice and suppress the ability of the public to have a stake in determining how governmental dollars are spent.

There may be a point at which leveraging the notion of equity will be become unsustainable, monetarily and otherwise.

CLINICAL UTILITY

The question of equity is relevant within the frame of clinical utility, as a clinical trial that may result in a widely beneficial treatment may be easier to support than a trial that offers the possibility of a more limited treatment. For example, if M.M.'s $n = 1$ trial was the first antisense oligonucleotide (ASO) drug, it could serve as a catalyst in the arena of ASO drug development. This was not the case, however, as nusinersen and eteplirsen had already been brought to market.

The clinical utility of M.M.'s $n = 1$ trial was under scrutiny due to its small sample size. Halpern, Karlawish, and Berlin argue in "The Continuing Unethical Conduct of Underpowered Clinical Trials" that underpowered trials for rare diseases are not ethically justified unless "investigators document explicit plans for including their results with those of similar trials in a prospective meta-analysis."[29] But it was not realistic to expect that the drug designed specifically for M.M.'s CLN7 mutation—a small, perhaps unique, subset of mutations causing Batten disease—could be incorporated into a prospective meta-analysis. And in conjunction with this, from an analysis standpoint, the $n = 1$ design of M.M.'s trial introduced additional challenges; there is little research regarding the appropriate assessment of $n = 1$ trial data. As Lillie and colleagues note, even though "washout" periods are intended to mitigate the effects of prior interventions, "accounting for carryover effects is not trivial," and these periods may serve as a barrier to producing data that can be extrapolated for use in other contexts.[30] This may further complicate the incorporation of data collected from M.M.'s trial into a larger analysis pool, whatever that may be.

Other ethicists have offered different perspectives on the ethical justification of underpowered trials. In their article published in *Lancet,* "Why Underpowered Trials Are Not Necessarily Unethical," Edwards, Lilford, Braunholtz, and Jackson maintain that equipoised trials ought to be "acceptable to both [conquentialists], who are concerned simply to achieve the most good, and to non-[consequentialists], who are concerned that the interests of future patients generally should not be allowed to override the rights of present patients."[31] (The term "equipoised" refers to *clinical equipoise*—the ethical basis for the conduct of randomized clinical trials involving human subjects—in which there is uncertainty regarding which arm of the clinical trial is therapeutically better than the other.)

For M.M.'s trial, could it have been anticipated that the new drug treatment would be better than no treatment at all? Was the halting of disease progression without the promise of reversibility argument enough for implementation of the trial? This most likely is not the equipoise upon which the above authors sought justification.

SAFETY

A determination of clinical equipoise, necessary for the FDA approval of a new drug protocol, includes analysis of the risks and benefits to patients and the safety of the experimental drug at the time of its administration. The safety of antisense oligonucleotide drugs is well documented in the literature.[32] Chan, Lim, and Wong note that much of the data collected thus far indicates that the majority of "toxic effects are dependent on the ASO backbone chemistry . . . [and are] sequence independent."[33] The most significant toxicities derived from multiple studies include complement and coagulation cascade activation, thrombocytopenia, hyperglycemia, and hypotension. Many of the effects have been associated with dose-dependence, and could therefore be mitigated by lowering doses.[34]

Despite the generally sequence-independent nature of the identified toxicities, it is important to consider each ASO drug as a new compound when reviewing its safety data, given the specific targeting of distinct gene exons. Wilton and colleagues found that although "the possibility of off-target annealing" is probabilistically low, it remains a risk.[35] In the most recent safety review application released for the SMA drug nusinersen (Spinraza), for example, the data suggest the drug contributes to inducing thrombocytopenia (in five of 56 of patients), proteinuria (in 33 percent of patients with infantile-type SMA and in 69 percent of patients with later-onset SMA and longer duration of treatment); hyponatremia (severe in one patient); decreased growth; rash (in two of 173 patients); and possible vasculitis (one patient).[36] Yasuda and colleagues reported to the FDA that while there is an overlap with the toxicities identified in other ASO studies, new drug-specific toxicities are also apparent. Further, the overarching safety analysis underscores the very real possibility that more significant toxicities will reveal themselves in the future. Yasuda and colleagues note: "The magnitude of the potential for serious harm after approval is unknown. Because

of limitations due to the small number of patients exposed and duration of exposure in the clinical trials, it is likely that adverse reactions not identified to date, or of a magnitude not observed to date, will occur in the postmarketing setting."[37]

Thus, the fast-tracking of an experimental drug through a clinical trial, coupled with a small population size, may limit the perceptibility of adverse effects. This has explicit relevance to M.M.'s trial, for which the Orphan Drug Act facilitated a similarly accelerated clinical trial approval process and for which the sample size was even smaller. If toxicities reveal themselves downstream—primarily in a postmarketing context—will it be necessary to rethink the existing framework of drug safety monitoring and regulation overall? Will there be the monetary and staff-based wherewithal to ramp up widespread post-market analysis?

Thus, the fast-tracking of an experimental drug through clinical trial, coupled with a small population size, may limit the perceptibility of adverse effects.

The ethical conundrum posed in the context of drug safety for this experimental treatment also centered on the challenges introduced by the $n = 1$ study population. In M.M.'s case, these challenges were largely based on her age and her specific neurodegenerative condition. In "Research Ethics and *N*-of-1 Trials," Crowden, Guyatt, Stepanov, and Vorhra argue that when "*N*-of-1 trials are undertaken as part of clinical care . . . the clinician and patient enter a partnership."[38] It is through this partnership that treatment targets are monitored. In addition to concrete health data, this system relies heavily on patient-reported symptoms. But M.M.'s ability to communicate was limited by her age and her disease process. Given this, was it possible that M.M. could have entered this partnership fairly? To what extent could the patient have accurately self-advocated with regard to monitoring treatment targets and symptoms? Could M.M.'s parents have served as appropriate stand-in spokespersons for her?

PARENTAL AUTONOMY

Donovan and Pelligrino note that "parental preferences may or may not reflect the good of the child." They state, "although we act as if a suitable proxy can exercise the autonomy of the nonautonomous child, this is not altogether an unreasonable fiction."[39] This is why, in defining autonomy as it pertains to decision making for a child, McCullough describes the necessity of recognizing the "pediatrician and parents . . . [as] co-fiduciaries of the child who is the patient."[40] He underscores the role of the physician in guiding the decision-making process, serving as a trusted clinical expert and source of scientifically rooted judgment.

With regard to M.M.'s trial, we would like to assume that her parents acted as a "suitable proxy" when they gave permission for her to participate in the experimental drug trial. Numerous studies have questioned the ethical nature of the consent process as it pertains to clinical trials involving children—perhaps making the entire enrollment process unethical, particularly without the clear directorial role of the child's physician as co-fiduciary. In "Parental Perceptions and Attitudes about Informed Consent in Clinical Research Involving Children," Harth and Thong present what they call "worrisome" data regarding parental naiveté surrounding their comprehension about their child's participation in a clinical trial.[41] Harth and Thong argue that, in the majority of cases, most parents do not have a solid understanding that drug trials examine the efficacy and safety of a treatment at different times. The authors also identify a lack of parental awareness regarding the general risks involved in participation of a clinical trial, and a parents' lack of a fundamental understanding about the consent document's role in helping "protect their rights . . . [and allowing them to] withdraw their child unconditionally from the trial at any time [if desired]."[42] Barfield and Church, in "Informed Consent in Pediatric Clinical Trials," draw attention to the notion that many parents who give consent for their child to participate in a clinical trial do not understand the basic tenets of trial design such as randomization or the differences between the clinical phases (see table 1).[43] In an analysis of informed consent processes for an oncologic

randomized controlled trial (RCT) among pediatric and adult populations, Simon and colleagues found that the adult oncologic patients were "more fully informed and more actively engaged by their oncologists" than the pediatric surrogate decision makers were.[44] The authors found that 92.5 percent of the adult patients accurately identified the implications of randomization in the selection of treatment options in the clinical trial, versus 40 percent of the pediatric surrogates. Six to eight months after the beginning of the trial, the pediatric surrogates reported a much higher, statistically significant, overall level of regret about the decision to participate in research than the adult subjects did (p = .002). Interestingly, Simon and colleagues found that the adult subjects indicated a much higher baseline level of trust in their oncologist compared with the pediatric surrogates.[45]

In "Two Concepts of Clinical Optimism," Jansen considers the role of therapeutic optimism on the ethical validity of the consent process in clinical trials involving children.[46] Jansen distinguishes dispositional optimism from unrealistic therapeutic optimism, and explains that unrealistic therapeutic optimism may impair parents' ability to ethically provide consent for their child to participate in a clinical trial. In Jansen's view, dispositional optimism is "ethically always tolerable because hope does not compromise autonomy of a decision to participate in research."[47] She contrasts this with unrealistic therapeutic optimism, which is a "hope for an unlikely cure . . . [that] can reduce participants' autonomy" through its "overestimate[ion] of the likelihood or magnitude of medical benefit."[48] What was the nature of M.M.'s parents' optimism for M.M.'s trial? Was it unrealistic? If so, was their proxy decision making unethical? As previously noted, there was uncertainty as to how much the drug could contribute to neurocognitive reversal in M.M.'s case. It was more realistic to think that it would halt further neurodegeneration, leading to questions regarding M.M.'s quality of life following the clinical trial, given her continued impairment.

CONCLUSION

Thus, from the perspectives of equity, clinical utility, safety, and parental autonomy, it seems that the FDA's fast-tracking and ultimate granting of approval for M.M.'s clinical trial may have been ethically unjustified—or at least ethically tenuous. While many of the arguments put forth and many of the questions raised in this article were driven by M.M.'s specific neurodegenerative process and her young age, the emphasis on reassessment of clinical trial approval from numerous ethical standpoints is warranted more generally as we move into an era of gene-targeted treatments, in which the possibility of individualized drug development becomes a reality. The principle of distributive justice will take on greater importance in a heavily budget-constrained context, as will the necessity to derive clinical utility from n = 1 studies, and the need to appropriately monitor drug safety. Emerging studies with children will need to be evaluated much more systematically for the reliability of consent made by surrogate decision makers. Upholding these ethical standards will allow us to facilitate and propel therapeutic advancement.

PRIVACY

To protect her privacy, the patient's name has been changed to "M.M."

NOTES

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The Family Voice

The Most Difficult Decision We Ever Had to Make

Kari Olavson

ABSTRACT

The author describes her family's experiences with hospice at home for their young son.

Our journey with Jacob in many ways felt like a lifetime and like just a few quick moments simultaneously. Our son was diagnosed with a rare genetic mutation that caused countless hurdles. During his life, he acquired a gastrostomy tube, a wheelchair, and eventually a tracheostomy.

Even before his diagnosis at age six, we knew his life could be short, and we did our best to give him a life full of meaningful and exciting experiences. At a monster truck event, shortly before being diagnosed, he went into shock due to his inability to regulate his body temperature; he was resuscitated after becoming hypothermic. A similar event happened a few months later, at which point the hospitalitist in the pediatric intensive care unit sat down with me. Her words were kind and we knew the day would come; we were at the crossroads of having to face that we had run out of options and his body was tired. Our next chapter in Jacob's journey was transitioning to hospice through the Children's Palliative and Hospice program.

Kari Olavson is the Mother of Baby Jacob, who was born with a rare genetic illness.

The choice to begin hospice was a very difficult one. Thankfully, the palliative care physician and the team who cared for our son were constantly reminding us of the love and care that lead to decisions like hospice, and that it was not failing in any way. At the beginning of our hospice journey, I had a difficult time letting go of solving every "medical mystery."

After a very complex and mysterious life followed by a rare diagnosis, it was difficult to not constantly look for the answer to every ailment and illness that affected our son. We had endlessly advocated for him and it felt wrong to not do so. Yet our primary hospice nurse reminded us time and time again that hospice is not about solving mysteries, it is about comfort and quality. She gently reminded me many times when I would ruminate about figuring out what was causing trouble that day, until I was able to take it to heart. It helped us so much.

Shortly after beginning hospice, we wanted to attend a family holiday party about two hours from our home. In the past we would have most likely not attended, telling ourselves it was too risky, too scary; but they encouraged us to get him there. Our hospice team helped us problem solve and prepare what was needed so we could make the trip with him.

He slept much of the day, but he opened his eyes to see Santa and he got to see cousins, aunts, uncles, and extended family. We didn't know it then, but it

would be the last time he would see many of them, and the encouragement to go was a perfectly timed gift. Our amazing hospice team also helped us to prepare for an epic roller skating seventh birthday party for him; another event where he saw many people who loved him and was able to find joy in the day.

We continued to make plans, no longer having to worry about burning all of his energy up for medical appointments. The appointments were done. The last one was his initial meeting with his palliative care doctor. After that visit, we were able to do televisits with her in the comfort of his bedroom and with our hospice nurse present. These visits were amazing. The convenience of not having to physically go anywhere was a blessing I can't even put into words. We were able to focus our energy toward joy. Early on, I had concerns the tele-visits would not be thorough enough, but having someone physically present along with the conversation made each appointment feel complete. So much of our hospice journey was in what we discussed versus the physical aspects of medicine.

Eventually, days came when he was very uncomfortable and we were almost constantly on call with his team. Nurses came out when we needed them; they guided us on keeping him comfortable with medications and other measures. They never failed to remind us that this was about quality and comfort. A few short months after the birthday party, the day came that we knew it was time to let him go. He was in a lot of pain, he was tired; it was the moment they prepared us for.

We decided, along with his care team, to help him be comfortable and remove the machines supporting his life. When we told his palliative care physician that we were ready, she quickly decided she would come to be with us to assure his comfort and help guide the hospice and homecare nurse. They held our hands and prayed with us when we prayed over our beautiful son; it just felt so perfect and right.

After he passed, we were able to just be with him and grieve. His nurse took care of every detail and guided us as we tried to process this unimaginable situation; we had support, care, and guidance right up until his small, seven-year-old body was rolled down the ramp outside our front door for the last time. Even after he was gone, they continued to care for us for months. Child-life visited our daughter and helped us try to understand her grief. A chaplain visited and spent an afternoon with me, talking about the little boy I missed and seeing his bedroom.

The care and support our son needed increased until his last day. We could not have done that last leg of the race without his hospice team. They were there supporting us through every step of the simultaneously heartbreaking and beautiful journey that is hospice.

Precious Life; Precious Loss

Ellen Beaudry

ABSTRACT

The author, the mother of a tiny newborn, shares her story at the end of her son's life in the neonatal intensive care unit.

The news was delivered abruptly, almost violently. The neonatologist flew in the room, launched himself up on a stool next to Truman's isolette, and said, "We just received your son's white blood cell count. It is 600. He is not going to win against whatever infection he has. You need to hold your son now."

Little one-pound, two-ounce Truman's heart rate and oxygen saturation continued to fall. The chaplain arrived with shells and holy water. She asked for our son's full name. I spoke carefully, "Truman Mark Beaudry."

There was an almost palpable sense of reverence in the room. I was grateful for the professional, human respect. He was baptized, as everyone in the room paused with heads bowed. I whispered, "Amen."

Truman's nurse worked quickly to disconnect him from all support. The respiratory therapist opened the sides of the isolette and carefully placed him in my arms. I cradled my son as tears began to roll down my cheeks, onto his raw, pink skin. Most of the staff cleared the room, leaving us alone.

I realized that while Truman had struggled mightily for 10 hours, this was my one wish: I just wanted to hold him. I whispered how much we loved him, how badly I wanted him, how sorry I was—for everything. I pulled him in close and began to sing softly,

Terry asked the nurse to listen for Truman's heart. She brought over her stethoscope and listened carefully, allowing Terry to listen, too. Every once in a while there was still a soft beat. I tried my best to rock him in the wheelchair. I kissed his little head again and again. He never moved or seemed in pain. At some point, Truman silently slipped away.

We thought our four older boys deserved a chance to see their brother. So Terry called our family and explained what happened, asking my dad to bring them to the hospital. We asked to speak to the child-life specialist. She offered to help Terry meet the boys and to talk to them before they would come in Truman's room. She planned some activities. We requested that a lot of the big equipment be moved from the room. Also, my wonderful postpartum nurse traveled back and forth all day from her unit to the neonatal intensive care unit (NICU) to care for me. She brought me a clean nightgown and even styled my hair to make me more presentable.

The daytime NICU chaplain was present and took tons of photos. While we were extremely grate-

Ellen Beaudry is the Mother of Baby Truman, who died shortly after his birth.

ful for those, we were sad our repeated requests for a professional photographer from Now I Lay Me Down to Sleep[1] were unfulfilled. We also would've loved to have video footage of Truman in life and in death. What a treasure that would have been.

I was holding Truman as the boys entered his room. They were mostly excited to see the baby. They each took turns holding him. The boys completed a handprint project and colored pictures. Then they went with Terry and my family to have lunch at the Ronald McDonald house in the hospital.

Alone with my son, I cradled him close, rocking back and forth, and crying. Sometimes I could hum a tune. Mostly I just breathed him in, taking deep breaths of the son who was growing cold in my arms despite the love I felt in my heart. I prayed he would forgive me.

Two hours later, the boys came to say good-bye. I didn't tell them they wouldn't see Truman again.

Once our family left, my nurse convinced me to go back to my room to lie down. My ankles were so swollen and I had never even been cleaned up after surgery. My sister was there and offered to stay with Truman. I reluctantly went back to my room.

Later, I asked to go back to the NICU. Terry's mom and sister were on their way and my nurse had suggested that I give Truman a bath. I unwrapped Truman, took off his diaper and removed all his bandages. It was good to see all of him. The nurse helped me fill a tub with warm water and she held Truman's head while I bathed him. It felt good to wash clean some of the wounds left from his IV and respirator. I sang "Rubber Duckie" to him, just like I do for all the boys at bath time. It made me feel like his mother and I am grateful his nurse suggested it.

When the last of our family left, Terry suggested I say good-bye before Truman's body changed even more, but I just wasn't ready to let him go. I knew this was the last time I would ever hold him in my arms, kiss his tiny face, caress his tiny hands and feet. I held him close and rocked so vigorously. I pulled back his blankets and kissed him a thousand times on every tiny speck of his body. I handed him over to the nurse. I thanked her for all she did for us. I said good-bye to our son forever.

NOTE

1. The mission of Now I Lay Me Down to Sleep is "to introduce remembrance photography to parents suffering the loss of a baby with a free gift of professional portraiture." According to its webpage, the 501(c)3 organization has "approximately 1,500 active photographers [and] reaches every state in the United States and . . . 40 countries worldwide." *https://www.nowilaymedowntosleep.org/about-us/our-mission-vision-reach/mission/*