

The fears and needs of the dying child: The case of Julianna Snow



Illustration by Steve Derrick

Aishwarya Gautam



Ms. Gautam is third-year medical student at Case Western Reserve University School of Medicine, Cleveland, OH. She is the recipient of the second place award (tie) in the Alpha Omega Alpha Honor Medical Society 2024 Helen H. Glaser Student Essay Award.

If people do not want to think about death, they especially do not want to think about the death of a child. As a result, when families are placed in the unimaginable position of taking care of a child who is terminally ill, they may find themselves with many questions surrounding what appropriate and compassionate care looks like for their child.

AMA code

Until recently, the best-interest standard was the predominant approach to treating minors.¹ Rooted in legal tradition, the best-interest standard considers the child legally incompetent and excludes them from decision-making.² In 2017, the American Medical Association (AMA) updated its Code of Medical Ethics in favor of a more nuanced approach that places greater emphasis on fostering children's autonomy and moral growth. In the update, the AMA states, "Parents (or guardians) are also recognized to have a responsibility to foster their children's autonomy and moral growth, a responsibility clinicians share."¹ The report also states, "Providing information in a developmentally appropriate way that respects the minor patient's cognitive ability, engaging the child in decision-making to the extent possible and

seeking the child's assent to proposed interventions helps to fulfill that responsibility."¹ Although at its surface, this approach seems almost common-sense, a great deal of murkiness arises when it is applied to extreme situations, such as the care of a child with terminal illness.

Julianna Snow was a five-year-old girl with Charcot-Marie-Tooth Disease (CMT), a neurodegenerative condition characterized by both motor and sensory deficits. These include atrophy and weakness in the arms, legs, and fingers, balance problems, nerve pain, and reduced ability to feel heat, cold, and touch. CMT is a heterogeneous hereditary disorder. There are many different gene mutations that can cause CMT, and the severity of symptoms can vary greatly depending on the mutation.³

Julianna's CMT was one of the more severe manifestations of the disease. CMT first robbed Julianna of the ability to chew and swallow food; then it affected the muscles involved in breathing. As a result, mucous would collect in her lungs and she repeatedly developed pneumonia.⁴

The treatment for this was a procedure called nasotracheal suctioning, which involves threading a tube through the nose, down into the throat past the gag reflex, and into the lungs to remove the mucous. Apart from being anxiety-inducing, the procedure can be quite painful. The procedure is so uncomfortable that many children scream and need to be restrained. In fact, most children are sedated for the procedure.⁵ Unfortunately for Julianna, her condition was too delicate for her to be sedated. She had to remain conscious for the entirety of the procedure, and she was too weak to do anything except cry. In Julianna's case, the procedure often had to be repeated in the span of a few hours.⁴

Julianna's condition worsened to the point where she needed to have parenteral nutrition (through a tube inserted directly into her stomach) and a pressurized mask to deliver oxygen to her lungs. Anything as routine as a cold or the flu could put Julianna in a very precarious position.

In October 2014, Julianna's doctor had a conversation with her parents, explaining that her condition was worsening and when she inevitably got sick again, she would likely die. Despite the grim news, Julianna's parents intended to take her to the hospital the next time she was ill. That was, until they thought to ask Julianna what she wanted. By this point, she had gone through hundreds of rounds of nasotracheal suctioning.⁴

The Washington Post article covering this story describes the conversation between Julianna and her mother, Michelle Moon, as follows:

"Julianna, if you get sick again, do you want to go to the hospital again or stay home?" she asked her daughter in a conversation she blogged about on her personal blog and for the *Mighty*.

"Not the hospital," Julianna said.

In another conversation, Julianna asked whether her mother wanted her to get treatment. Moon asked what Julianna wanted.

"I hate NT. I hate the hospital," she said.

"Right. So if you get sick again, you want to stay home," her mom said. "But you know that probably means you will go to Heaven, right?"

"Yes."

"And it probably means that you will go to Heaven by yourself, and Mommy will join you later."

"But I won't be alone," Julianna replied.

"That's right. You will not be alone."

"Do some people go to Heaven soon?" the four-year-old asked.

"Yes. We just don't know when we go to heaven," her mom said. "Sometimes babies go to Heaven. Sometimes really old people go to Heaven."

"Will Alex go to Heaven with me?" Julianna asked, referring to her six-year-old brother.

"Probably not. Sometimes people go to Heaven together at the same time, but most of the time, they go alone," Moon told her. "Does that scare you?"

"No, Heaven is good," her daughter said. "But I don't like dying."⁴

In the end, Julianna's parents decided to honor her wishes. In Moon's view, Julianna had "... made it clear that she doesn't want to go through the hospital again. So we had to let go of that plan because it was selfish."⁶ In her blog, Moon wrote about how Julianna's last months of life in hospice ended up being the happiest time she remembers.

Pushing boundaries

Julianna's family's decision to take her wishes into consideration pushes the boundaries of the AMA's guidance that children participate in their own care in a way that is commensurate with their abilities. It raises all sorts of questions. Given her young age, did Julianna have a good

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enough understanding of her situation? To what extent could she understand death?

Many members of the public had doubts about Julianna's judgment given her age. In response to reading about her story, bioethicist Arthur Caplan stated "This doesn't sit well with me. It makes me nervous. I think a four-year-old might be capable of deciding what music to hear or what picture book they might want to read. But I think there's zero chance a four-year-old can understand the concept of death. That kind of thinking doesn't really develop until around age nine or 10."⁴

Comprehending the unknown

From a developmental perspective, it does seem unlikely that a child as young as Julianna could fully grasp the concept of death. Grasping the full meaning of death includes understanding that: death is guaranteed to all humans ("inevitability"); death applies to all living beings ("universality"); death is permanent ("irreversibility"); death halts all physical and psychological processes ("cessation"); and death is caused by the dysfunction of bodily processes ("causality").⁷

Children as young as five- or six-years-old can comprehend the inevitability and irreversibility of death. The notion of cessation of bodily processes and universality of death happens around ages six or seven. Comprehension of the causality component of understanding death happens last. Grasping this concept happens as late as eight-10 years old. Causality is the toughest concept to grasp because it involves appreciating complex biological processes.⁸

Because they cannot grasp the causality component, children may view death as a form of punishment for something they did or thought about. They may not understand how their parents could not have protected them from this illness.⁹ Thus, while Julianna may have understood that dying would be irreversible and she would no longer be with her parents, there were nuances about her death that she likely did not comprehend.

It should be noted, though, that there is a great deal of individual variation in when children reach the milestones mentioned above. Factors that influence a child's evolving view of death include maturity in thinking and processing information, past experiences with death such as the death of a grandparent or a pet; exposure to books, cartoons, television shows and movies depicting death; and parents' views of death including cultural, spiritual, and religious beliefs.⁹

Julianna certainly had a great deal of experience entering the hospital, and medical interventions kept her alive

for the majority of her short life. Thus, despite perhaps not fully grasping the concept of death, Moon argued that Julianna certainly knew what nasotracheal suctioning was.

Members of her care team agreed that her experience mattered and that she should have some input into the end of her life. Bioethicist, Chris Feudtner, director of the Department of Medical Ethics at the Children's Hospital of Philadelphia agreed, stating "To say her experience is irrelevant doesn't make any sense. She knows more than anyone what it's like to be not a theoretical girl with a progressive neuromuscular disorder, but to be Julianna." Feudtner's comments suggest that the subjective experience, even of a minor who may not understand the details of her situation, should not be discounted.⁴

In the book *How Terminally Ill Children Come to Know Themselves and Their World*¹⁰ Myra Bluebond-Langner suggests that experience plays an important role in children's understanding of their disease. The book, which focuses on children with leukemia, describes what they understand about their disease and examines how they come to that understanding. In her description, Bluebond-Langner identifies five chronological milestones in the child's understanding of the progression of their illness: seriously ill; seriously ill and will get better; always ill and will get better; always ill and will never get better; dying (terminally ill). In progressing through these milestones, Bluebond-Langner suggests that a child's experience rather than cognitive ability play a larger role.

The role of experience in developing awareness also explains why age and intellectual ability are not related to the speed or completeness with which the children pass through the stages. Some three- and four-year-olds of average intelligence know more about their prognosis than some very intelligent nine-year-olds.¹⁰

The case of another child, Hannah Jones, has parallels to Julianna's case. Hannah was 12-years-old when, after many years of pain and procedures to deal with a failing heart, she refused a potentially life-saving heart transplant. There was controversy over whether she should be legally allowed to make this decision. Though Hannah was much older than Julianna, there are similarities in the way both mothers took their daughter's feedback seriously and placed a great deal of weight on their experiences. Hannah's mother states, "I wanted them to understand that Hannah knew her own mind after so many years of ill health, which is why I trusted her to make this decision. Hannah taught me to have the courage of my convictions and it was only now as it was tested beyond concrete reason into innate faith that I realized this."¹¹

Both Julianna and Hannah's mothers described, in their own words, the amount of courage it took to take guidance from their child, indicating the challenges of relinquishing control.

Shared decision-making

Despite the difficult nature of sharing decision-making, best practices for caring for a terminally ill child mirror the AMA's focus on granting children autonomy wherever appropriate. For instance, if a child does not want to talk about death, their wishes should be respected as much as possible. If, however, the child is open to discussing, parents should try their best to be open to engaging in conversation about death, as it can help allay a child's fears.⁹

In having conversations with Julianna about her imminent death, Julianna's parents were able to soothe psychological fears as well as physical ones. In her blog, Moon recounts how, shortly after their first conversation about Heaven, Julianna entreated her mother, "remember me always."¹² Children facing death often have a fear of being forgotten—not unlike adults. The dying child often looks to parents or caretakers for reassurance that they will be missed, and that when they die, they will not be alone. Some children also seek permission from caretakers to die. They may have a fear of hurting those whom they leave behind. In fact, they may cling to life until they feel that they have permission to let go. This is a phenomenon that is also mirrored by adults.⁹

Death comes to adults and children alike. There are perhaps more parallels in the thoughts and feelings of the dying adult and the dying child than have been formally explored. However, the case of a dying child involves unique challenges for parents and medical providers. In considering Julianna's case, it becomes evident that pediatric decision-making requires a delicate balance between respecting a child's autonomy, considering their developmental stage, and acknowledging the value of their subjective experiences. It highlights the importance of listening to a child's fears—both psychological and physical—in order to best prepare them for death.

As we continue to learn about how best to work with, and serve, children, the case of Julianna Snow serves as a compelling reminder of the value of listening, discussing, and comforting as profound acts of care.

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The author's E-mail address is aag140@case.edu.