Decisions for Long-Term Ventilation for Children

Perspectives of Family Members

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Abstract

Rationale: The decision whether to initiate or forgo long-term ventilation for children can be difficult and impactful. However, little has been published on the informational and decisional needs of families facing this decision.

Objectives: To assess what families with children with chronic respiratory failure and life-limiting conditions need and want for informed decision-making.

Methods: English- and Spanish-speaking parents who were facing (contemporaneous decision makers) or had previously faced (former decision makers) a decision regarding invasive or noninvasive long-term ventilation for their children were recruited using convenience sampling. Patients who were older and cognitively capable also were invited to participate. We performed semistructured interviews using an open-ended interview guide developed *de novo* to assess parents' decisional needs and experiences. Qualitative data analysis used a thematic approach based on framework analysis, and thematic saturation was a goal.

Results: A sample of 44 parents and 2 patients from 43 families was interviewed. All contemporaneous decision makers ($n = 28$) favored or believed that they would choose long-term ventilation. Fifteen of 16 former decision makers chose long-term ventilation.

Thematic saturation was achieved from the perspective of parents who favored or chose long-term ventilation. Four domains were identified: parents' emotional and psychological experiences with decision-making, parents' informational needs, parents' communication and decision support needs, and parents' views on the option not to initiate long-term ventilation. For most parents, making a decision regarding long-term ventilation was stressful, even though they articulated goals and values that could/did guide their decision-making. In general, parents wanted comprehensive information, including what life would be like at home for the child and the family. They wanted their medical providers to be honest, tactful, patient, and supportive. Parents reported that they felt being presented with the option not to initiate was acceptable.

Conclusions: In this study, we identified specific informational and decision-making needs regarding long-term ventilation that parents facing decisions feel are important. These data suggest that providers should present families with comprehensive, balanced information on the impact of long-term ventilation and, when the child has a profoundly serious and life-limiting condition, explore the option not to initiate long-term ventilation.

Keywords: artificial respiration; child; decision-making

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Each year, hundreds of children develop chronic respiratory failure (CRF) and become dependent on long-term ventilation (LTV), either invasive via a tracheostomy or noninvasive (1, 2). Most of these children have other severe medical conditions that result in functional limitations, medical fragility, and shortened life expectancies (3–5). Although LTV can support respiratory function, it does not mitigate these other conditions. LTV does, however, demand constant, complex home care. For children with life-limiting conditions, it is morally and legally permissible to forgo LTV and allow natural death if LTV is considered ineffective or overly burdensome, even when death is not imminent (6). As in other intense situations in which families are forced to make crucial choices for their seriously ill children (7, 8), the decision of whether LTV is the best option for some children with CRF and their families is difficult and profound.

Despite the gravity of this situation and evidence that families in this situation want more and earlier information about LTV (9, 10), little has been published on the informational and decisional needs of the families facing this situation. To aid providers who counsel and assist families in this decision-making, we interviewed parents who were facing or had faced decisions regarding LTV for their children with CRF to characterize their informational and decisional support needs.

Methods

Subjects and Data Collection

Eligible subjects were parents of children with CRF and a life-limiting condition who either 1) were currently facing the decision whether to initiate their children on invasive or noninvasive LTV (contemporaneous decision makers) or 2) had previously faced such a decision (former decision makers). Although there is no universally accepted definition of CRF, we subscribed to the one in *Rogers' Textbook of Pediatric Intensive Care*: "The diagnosis of CRF is usually made once repeated attempts to wean from assisted ventilation have failed for at least 1 month in a child without superimposed acute respiratory disease or a patient who has a diagnosis with no prospect of being weaned from the ventilator (such as high spinal cord injury)" (11). Examples

of life-limiting conditions included progressive neuromuscular diseases, cerebral palsy with severe motor and cognitive impairment, and congenital brain malformations. Parents of children with CRF and non-life-limiting conditions (e.g., bronchopulmonary dysplasia without other severe comorbidities of prematurity and isolated airway/thoracic abnormalities) were excluded. English- and Spanish-speaking parents were eligible. Cognitively capable adolescent and adult children with CRF also were invited to participate after parental permission and/or patient consent were obtained. When two parents or a parent and the child were both willing to be interviewed, they chose whether to be interviewed separately or together.

The two groups of subjects were recruited using convenience sampling. Most subjects were parents of children who were receiving care in one of three pediatric intensive care units (PICUs) of Morgan Stanley Children's Hospital. Four former decision makers, whose children were not admitted, were known to the pediatric pulmonology and critical care faculty; these parents were invited to participate via a phone call. Written informed consent was obtained before interviews.

Semistructured interviews were completed in person or over the phone by an investigator (J.D.E.) or by a trained research assistant, who was fluent in Spanish and experienced as a hospice counselor. Parents whose primary language was Spanish were interviewed by the research assistant or by an investigator (J.D.E.) with the assistance of a professional Spanish-speaking interpreter. Contemporaneous decision makers were interviewed after the medical team started to have discussions regarding the diagnosis of CRF and the possible need for LTV and before tracheotomy was performed or before their children were discharged using noninvasive LTV.

In an iterative process among the investigators, open-ended interview guides were developed *de novo* on the basis of published literature and clinical experience. These guides were used to elicit parents' experiences and opinions regarding the following topics:

- How should LTV-related sensitive topics and decisions regarding LTV be broached and discussed?
- What influences/helps/hinders (or influenced/helped/hindered) counseling and decision-making?
- How do/did they feel about and approach this decision?

The interview guides are available in the online supplement. Unscripted probes were used to further elucidate or clarify parents' responses when appropriate. Interviews continued until it was deemed that thematic saturation had been reached.

Interviews were audio recorded and transcribed verbatim by a professional medical transcriptionist. If an interview was conducted solely in Spanish, the audio recording was translated to English and transcribed by the bilingual research assistant. Personal identifiers were removed, and transcripts were reviewed by an investigator (J.D.E.) for clarity. Demographic information was collected from the interviewees; clinical data were collected from the child's medical record.

Data Analyses

Qualitative data analysis was conducted using a thematic approach based on framework analysis (12, 13). The overarching thematic framework was developed beforehand on the basis of what we hypothesized to be the key elements of informed, shared LTV decision-making. On the basis of analysis of interview texts, themes were identified within key domains. Two investigators (J.D.E. and M.C.M.) independently coded each transcript. All coding discrepancies were resolved through discussion and consensus. Quantitative data were presented as simple counts.

NVivo 11 software (QSR International) was used for coding. The Columbia University Institutional Review Board approved this study.

Results

Parents of 51 children met the inclusion criteria and were invited to be interviewed between December 2015 and December 2017. Forty-four parents of 43 children (43 of /51; 84% participation) were interviewed (see Tables 1 and 2). This included 18 contemporaneous invasive LTV decision makers, 10 contemporaneous noninvasive

Table 1. Characteristics of children with chronic respiratory failure

Characteristic	Parent Decision Group			
	Contemporaneous		Former	
	Invasive LTV (<i>n</i> = 17)	NIV LTV (<i>n</i> = 10)	Invasive LTV (<i>n</i> = 8)	NIV LTV (<i>n</i> = 8)
Age, median (range)	11 mo (2 mo–16 yr)	4.5 yr (5 mo–16 yr)	4 yr (6 mo–20 yr)	8.5 yr (22 mo–18 yr)
Female sex	10 (59)	4 (40)	5 (63)	3 (38)
Primary reason for CRF				
Central hypoventilation	6 (35)	3 (30)	5 (63)	5 (63)
Ventilatory muscle weakness	4 (24)	6 (60)	2 (25)	3 (37)
Chronic pulmonary disease	7 (41)	1 (10)	1 (12)	—
Already using/previously used NIV LTV	3 (18)	—	1 (12)	—

Definition of abbreviations: CRF = chronic respiratory failure; LTV = long-term ventilation; NIV = noninvasive.

LTV decision makers, 8 former invasive LTV decision makers, and 8 former noninvasive LTV decision makers. Six eligible contemporaneous decision makers and two former decision makers declined participation. Five interviewees were primarily Spanish speaking. Three interviewees requested not to be recorded but allowed notes to be taken during their interviews. Two former decision makers were interviewed over the phone. Other interviewees included a young woman using invasive LTV and an adolescent girl being initiated on noninvasive LTV.

All contemporaneous decision makers said they favored or were going to choose LTV. Fifteen of 16 former decision makers chose to initiate LTV; one former decision maker declined LTV and instead chose to focus exclusively on the comfort of her child. Another former decision maker had several children using LTV, one of whom died unexpectedly while using LTV.

After analysis of these 46 interviews, the investigators determined that thematic saturation had been achieved from the perspective of parents who favored or chose LTV. Themes were categorized into four broad domains: parents' emotional and psychological experiences with decision-making, parents' informational needs, parents' communication and decision support needs, and parents' views on the option not to initiate. Domains and their subthemes, with representative quotes, are provided in Table 3.

Parents' Emotional and Psychological Experiences with Decision-Making

All 18 contemporaneous invasive LTV decision makers and 4 contemporaneous

noninvasive LTV decision makers viewed their situation as one in which they had to consent to initiate LTV or decide between initiating LTV (with or without tracheostomy) and not initiating LTV.

Although their responses were nuanced, these parents perceived their decision in two main ways. Seven parents believed that there was no decision to be made, because supporting their child's breathing or

Table 2. Demographics of 44 parental decision makers

Characteristic	Data
Relationship to child, <i>n</i> (%)	
Mother	34 (77)
Father	10 (23)
Age, yr, median (IQR)	35.5 (29–41.5)
Race, <i>n</i> (%)	
White	28 (64)
Black or African American	8 (18)
Asian	5 (11)
Native Hawaiian or other Pacific Islander	1 (2)
Hispanic/Latino	21 (48)
Religion, <i>n</i> (%)	
Christianity	28 (64)
Judaism	5 (11)
Islam	4 (9)
Hinduism	2 (5)
Buddhism	1 (2)
Wiccan	1 (2)
None	3 (7)
Importance of religion, <i>n</i> (%)	
Very important	25 (57)
Fairly important	7 (16)
Not too important	6 (14)
Not at all important	4 (9)
Not answered	2 (5)
Highest education level achieved, <i>n</i> (%)	
Some high school	7 (16)
High school/GED degree	12 (27)
Associate's degree	6 (14)
Some undergraduate	7 (16)
Bachelor's degree	9 (20)
Some graduate	1 (2)
Graduate/professional degree	2 (5)
Number of other adults in household, median (IQR)	1 (1–2)
Number of other children in household, median (IQR)	1 (0–2)

Definition of abbreviations: GED = general educational development; IQR = interquartile range.

Table 3. Thematic framework and selected quotes

Domains	Themes	Selected Quotes
Parents' emotional and psychological experience with decision-making	LTV is the only option	"I'm left with no choice."
	No great options	"It's my baby girl, and I'm going to do whatever to keep her alive." "The thing is, I know he will be on everything his whole life, and I know there is no [other] option..... It's not easy."
	Stressful situation	"It's scary that you might make a wrong decision based on your feelings and your wants, instead of the child's, which is hard to do." "It is difficult. I am not going to lie. It is difficult, but decisions have to be made, and I can't really give any good feeling as to how to describe how we came to the decision, but with how I am and my mind-set, I just think she needs it."
	How parents approached their decision	"I feel like it's the end of the world. I can't do it. I don't want to have a hole in my child's neck." "You have to think rational as a parent. It's not about what you want or what I like and what I don't like. It's about what's best for my child. Because you're just the advocate; you're not the person going through it." "Gather all the information you can before you make a decision, whether you've got to question the nurses, the doctors, the specialists, or you've got to seek the knowledge yourself, whether it's online or in an actual hard copy book or family and friends sending you links to pages or anything like that." "I speak to the doctors, and the doctors will give me their point of view. And I'll give them my point of view. We reach an agreement to see what's the best for my daughter." "A lot of thinking, a lot of praying, a lot of praying. And just a lot of listening."
Parents' informational needs	How much information	"I think it's best that you get all the information so that you can make a sound decision. I don't believe in making decisions where you don't know everything. I feel like you need to be presented with all the facts, the "what-ifs," and the "maybes" so that you can make a good decision." "I think a parent needs to know everything, the good and the bad, whether they want to hear it or not." "I didn't want to know too much about [negative things about my child's condition], and I still don't want to know too much about it as far as, you know, what to expect..... I think a parent needs to know everything, the good and the bad, ... relevant to this day right now."
	Specific information	"What does it take? What is your day like? Can you sleep at night? Is it worth getting the nursing? Things like that. Like, really specific [stuff] on how you can make your household function with somebody with a trach and vent." "They should explain more about the trach and how people have it and live their life." "We did hear the possibility when he gets worse, but we didn't have what would happen when he gets better."
Parents' communication and decision support needs	Communication needs	"We're persons who want [information] raw, unfiltered, and undressed, just give [it to] us as it is.... I just want the information as it is. I will process it. I may not like everything that they're telling me. You're never going to like it, because your child is sick." "They explained certain things, and they've given us our time to research it on our own as well, and they let us talk about it. So, I think those are all helpful because they don't expect a decision right then and now."
	Decision support needs	"But in the hospital, when they say that he needs a trach, they didn't explain to me all these [other] things [about caring for a child who uses long-term invasive ventilation]; they just say he needs a trach." "If everybody tells me, you know, she's going to die, you need to be ready. Well, I don't want to listen. We need to hope for the best." "If a family came up to me, and they're about to go into the same journey that I'm already in, I would tell them to stay strong, stay firm."

(Continued)

Table 3. (Continued)

Domains	Themes	Selected Quotes
Parents' views on the option not to initiate	Parent's understanding of the option not to initiate	<p>"Yes, I do understand why [a family might choose not to initiate long-term ventilation]. Again, it's very invasive to have a trach, and I think it can clash with religious beliefs and just prolonging what may be the end of the child's life just to have to be on a machine."</p> <p>"I think that there's a point where technology can keep someone alive without regard for the quality of that life. And so, there comes a point where a decision has to be made and say, you know, 'What are we trying to prolong here?'"</p> <p>"I would make sure you know all your options [and] what is being done. All the tools and technology that is provided doesn't mean that it's the right thing to do, just because the doctor is telling you, 'We're gonna do this next, or we're gonna do that next.' [Parents] shouldn't feel afraid if something is really in the back of their minds or a gut feeling.... You're not crazy for having those thoughts; there is nothing wrong with you."</p>

Definition of abbreviations: LTV = long-term ventilation; trach = tracheostomy; vent = ventilation.

preserving their child's life was the "only" option to them, and not doing so was unimaginable. One parent said, "So we must use any chance that can give my baby life, even for 1 hour more, 1 minute more, 1 day, 1 year.... [I]t's God's choice how long she's going to live." Despite favoring LTV, 15 others described the decision as difficult, as if there were no great options and they had to choose between substantial downsides. As another parent stated, "I think it's the hardest decision you can have, really, because you never know what the right decision is. There are so many questions and not enough answers. Really, I don't know how they can ask me to make a decision." Three of these 22 parents said that their first response was to reject LTV and/or deny their child's situation.

The former invasive LTV decision makers echoed similar views about how they originally perceived their decision-making situation—there was "no" other option than LTV, or it was a difficult decision. Two former decision makers believed they were never presented with a choice whether to initiate invasive LTV. Five regretted delaying tracheostomy for their children, because they believed it spared their children from frequent endotracheal intubations, provided more freedom than orotracheal intubation or constant noninvasive LTV, or facilitated care.

The majority of the contemporaneous decision makers felt devastated by their child's condition and/or tremendously stressed about their decision. This included

those parents who believed that LTV was the "only" option: "How does it feel? It's horrible. It's the worst feeling in the whole world." Explanations of why the situation was so stressful included feeling like they did not have all the desired information; worry about the possible downsides of LTV either for their child or, less commonly, their family; and feeling like both options had substantial downsides. Former decision makers spoke more about feeling overwhelmed when they received their child's diagnosis and less about their LTV decision-making being stressful.

Contemporaneous decision makers had various approaches to their decision (or ways to manage their stress) and sometimes had more than one compatible approach (Table 4). Five parents spoke of putting their faith in a higher power and that this higher power would guide their decision-making or dictate how things should be. Four verbalized that they wanted providers' opinions and suggestions about everything, including what would be the best option for their child. Several parents spoke about drawing emotional support from other family members. Four former decision makers recommended that other parents trust their own intuition and experience regarding their child, even sometimes over those of medical professionals.

Parents' Informational Needs

Forty parents emphasized the importance of knowing everything about their child's

condition(s) and LTV, regardless of whether the information was upsetting. This sentiment was spoken in terms of making a well-informed decision for their children and/or being prepared for the future. As one parent put it, "Because I don't think you can make a fair decision without knowing everything and what to expect, whether you want to hear it or not." In contrast, four parents acknowledged that they preferred to receive only positive messages (e.g., the benefits of LTV, the possibility that the child could be weaned from LTV) or did not want to hear negative information (e.g., the risks of LTV, the risk for future deterioration because of medical complexity) unless it was specifically relevant to a decision at hand.

The specific information desired by contemporaneous decision makers focused on how LTV would affect their child and their family (see Table 5). When the 16 former decision makers were specifically asked what information they wished they had when they faced their decision, they shared many of the same topics mentioned by the contemporaneous decision makers. In addition, five former decision makers added that they wished they had been better informed about the difficulties of home nursing (e.g., shortages of competent home care professionals); three mentioned the difficulties of travel; and two highlighted the involvedness and inconveniences of other equipment/treatments needed to care for their children (e.g., suctioning and

Table 4. How contemporaneous decision makers approached decisions for long-term ventilation

Approach	Number Who Mentioned (Total of 28)
Focus on what is most important for the child	
Primacy of preserving life	16
Prioritizing quality of life	4
Comfort	1
Get all the information possible first	5
Weigh the pros and cons of each option for the child	5
Get advice/support from others (e.g., extended family members, religious leaders, and medical professionals)	5
Have faith in a higher power and hope for best	5
Also consider the pros and cons of each option for oneself and the larger family	3
Follow your intuition	2
Ignore the unsolicited advice of others (e.g., extended family members)	1

mechanical insufflation–exsufflation). When the former decision makers were asked what they would share with contemporaneous decision makers, seven underscored that caring for a child using LTV is very labor intensive, stressful, and crucial (i.e., the child's life can depend on it); two acknowledged that "it is not for everyone." No former decision maker said that knowing more about any topic would have changed their decision or that they regretted their decision. Similarly, the young woman using invasive LTV was content with her life and appreciated her family's decision to initiate LTV years ago.

Parents' Communication and Decision Support Needs

Parents discussed their preferences for receiving information and sharing decision-making regarding CRF and LTV. Contemporaneous decision makers identified the following provider practices or qualities regarding communication as helpful: being honest ($n=9$), allowing time for information to be processed and questions to be asked ($n=9$), and being tactful and using sensitive language ($n=9$). Other helpful qualities mentioned included being supportive ($n=5$), sharing information before decisions or crises ($n=4$), using lay language ($n=4$), and using an interpreter for non-English speakers ($n=2$). Three contemporaneous decision makers wanted their children to be

informed and included as much as possible. Speaking about what they would tell contemporaneous decision makers, four former decision makers advised them to be at peace with the situation and be resilient.

Regarding unhelpful communication practices, 14 contemporaneous decision makers mentioned insufficient information or information that was not timely enough, particularly with regard to the children's diagnosis or prognosis and lack of detail regarding LTV. Nine said they felt pressured to make a decision. Four believed that frequent changing of medical providers hindered communication or decision-making. A few others perceived overly negative attitudes or statements about their children, depersonalization of their children, and conversations about their children that excluded them.

Parents' Views on the Option Not to Initiate

Forty-three of 44 parents granted that forgoing not initiating LTV could be a legitimate choice. Usually, this acknowledgment was in the context of a hypothetical scenario in which a child was suffering or had poor quality of life and limitation of life support was morally/religiously legitimate to the hypothetical family. Few parents believed that these criteria were (in the case of contemporaneous decision makers) or would have been (in the case of former

decision makers) applicable to their children or themselves. The young woman using invasive LTV likewise agreed that LTV was not obligatory in all situations. When asked, most parents said that they were/would not be offended if/when the medical team raised the option of not initiating LTV for their children. One contemporaneous decision maker acknowledged the possibility that initiating invasive LTV could ultimately be the "wrong" thing to do for her child. Another former decision maker said that invasive LTV was the right thing to do for her child but that she would not make the same decision for herself.

The former decision maker who declined LTV for her child said, like other parents, that she wanted information about all options and wanted it presented in a supportive way. However, she expressed disappointment that she was the one who broached the topic of limiting interventions that she believed were not in her child's best interests, and she delayed doing so because she feared that the medical team would see her as an uncaring parent. "I wish someone sat with me and said, 'You have options.'" Once she felt comfortable to express her thoughts candidly and dispelled the team's assumptions about what she wanted, she felt "relieved," validated, and more in control of her child's future. This parent recommended that all families be apprised of the full range of options, "regardless of if you can or cannot help [the child] get better."

Discussion

LTV can be life sustaining for children with CRF, but it entails risks for them and tremendous responsibilities for families. Families are not obliged to choose LTV for their children with another life-limiting condition when LTV is perceived to be unhelpful or overly burdensome. This choice normally means the child will die of his or her CRF. Thus, the decision whether to initiate LTV is often difficult and consequential on many levels.

Medical providers are charged with informing families of all their options and the possible consequences of those options, as well as with guiding them through shared decision-making. However, providers do not always meet families' needs for timely and sensitive communication to support decisions (9, 10, 14, 15). Numerous studies

Table 5. Specific information that contemporaneous long-term ventilation decision makers wished to know

Modality	Desired Information	Number Who Mentioned (Total of 28)
Nonspecific	What is home/family life like with a child who uses LTV?	14
	Diagnosis or prognosis of other complex chronic condition	6
	How will LTV help my child?	6
	Can my child leave the home while using LTV?	5
	What are the downsides of LTV?	5
	What will happen to my child if LTV is not initiated?	4
	What if my child gets better?	3
	Home nursing (e.g., how it works, pros/cons)	3
	What other additional equipment/treatments will my child need?	2
	How to care for other children in the home while caring for a child using LTV	2
	How does having a child who uses LTV affect the siblings?	2
Invasive	How to prepare for the death of one's child	2
	What are the risks of a tracheostomy?	7
	How will a tracheostomy affect my child's quality of life?	6
	How does a tracheostomy affect speech and oral intake?	4
Noninvasive	Is a tracheostomy permanent?	4
	Is a tracheostomy uncomfortable?	3
	Will my child ever tolerate it, or how long will it take for my child to tolerate it (because child was not tolerating it well in the hospital)?	3
	What if noninvasive LTV is not enough support?	2
	If my child gets intubated, does this mean he or she has to have a tracheostomy?	2

Definition of abbreviation: LTV = long-term ventilation.

have shown that providers have varying practices/opinions regarding providing LTV to children with life-limiting conditions and/or severe disabilities (16–24). When families are not fully informed or shared decision-making is suboptimal, families are at risk of unrealistic expectations, decisional distress or regret, and complicated grief. Seeking to aid providers who care for these children and families, we conducted what is the first study, to our knowledge, to interview parents of children with CRF and life-limiting conditions to assess their decisional needs regarding LTV.

We found that, in general, parents wanted to know everything, both to make well-informed decisions and to be prepared for the future. Beyond general information about LTV and diagnostic/prognostic information specific to their children, they wanted to know what life would be like at

home, for both the child and the family. They were clear that this meant learning of potential risks and burdens. Parents wanted their medical providers to be honest as well as tactful, patient, and supportive. Their approach to decision-making fit with shared decision-making, in which treatment options are weighed in the context of patient/family values and goals (e.g., primacy of preserving life). However, knowing their motivating goals and values, or even knowing what they would choose, seemingly did not mitigate the stress of the decision and possible consequences. Almost all parents did not see forgoing LTV as the right option for their own children, though they indicated a wish to know all their options and acknowledged that this option could be acceptable in some situations.

Other studies of familial caregivers facing situations related to LTV or end-of-

life decision-making for children also have identified a desire for honest and complete information; supportive, sensitive counseling; and ample time to make decisions (10, 25–28). This includes information on treatment options' potential short-term and long-term consequences for the child and family (10, 29). Furthermore, the decisional needs identified in this study overlap with those perceived by directors of pediatric home ventilation programs (30).

On the basis of these findings and in accordance with calls to improve the consistency and quality of decision-making regarding LTV through standardized processes or tools (10, 31, 32), families in the PICUs of Morgan Stanley Children's Hospital with children who may need LTV are given an LTV information and decision support booklet. Informed by our interviews of parents and directors of pediatric home ventilation programs (30), this booklet includes topics pertinent to both child and family and to both the short and long terms. It also broaches potentially sensitive topics, such as the option to forgo LTV and the potential for failure of noninvasive LTV to support some children (33). Materials such as this can augment provider counseling and ensure that all families receive the same basic information and anticipatory guidance before a decision is made. Others have published articles on their efforts to rigorously improve communication skills and relational abilities of pediatric providers who must counsel families in situations of this sort of gravity (34, 35).

This study has several limitations. First, we used convenience sampling and, although we tried to recruit all eligible parents, 16% of those approached could not be interviewed. Despite achieving thematic saturation for decision makers who would choose LTV, our sample may not be representative of all caregivers in this group. In addition, we were able to interview only one parent who declined LTV, so it is highly likely that additional information could be gleaned from interviewing more such parents. Second, it was not possible to interview all contemporaneous decision makers at the same stage of decision-making. We did interview them either before their children underwent tracheotomy or before discharge of their children using

noninvasive LTV. Third, although a sizable number of children are represented in the study and all had CRF and a life-limiting condition, they were heterogeneous in terms of their conditions, severity, and functional abilities. Such characteristics may impact decisional needs and how parents view and approach their decisions. Fourth, we did not address the informational needs of parents with children with CRF but without life-limiting conditions. Finally, although two investigators performed thematic coding independently, we did not assess interrater reliability, because discrepancies were rare, and neither coder emerged as dominant.

Decision-making regarding LTV for children with life-limiting conditions can be complex for families and providers. Knowing the needs and approaches of parents facing these decisions can help providers who must guide them in a shared decision-making process. Our findings suggest that parents want to know about the pros and cons of LTV, both for the child and the family. They also indicate that families are receptive to hearing about the option to forgo LTV, though this must be communicated with sensitivity and with the recognition that many parents do not consider it a legitimate option. Although challenging and time consuming, such practices should be the standard and

could help ensure informed decisions that provide families comfort and satisfaction and minimize distress and decisional regret. ■

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References

- Edwards JD, Houtrow AJ, Lucas AR, Miller RL, Keens TG, Panitch HB, et al. Children and young adults who received tracheostomies or were initiated on long-term ventilation in PICUs. *Pediatr Crit Care Med* 2016;17:e324–e334.
- Fine-Goulden MR, Ray S, Brierley J. Decision making in long-term ventilation for children. *Lancet Respir Med* 2015;3:745–746.
- Gregoretti C, Ottonello G, Chiarini Testa MB, Mastella C, Ravà L, Bignamini E, et al. Survival of patients with spinal muscular atrophy type 1. *Pediatrics* 2013;131:e1509–e1514.
- Ishikawa Y, Miura T, Ishikawa Y, Aoyagi T, Ogata H, Hamada S, et al. Duchenne muscular dystrophy: survival by cardio-respiratory interventions. *Neuromuscul Disord* 2011;21:47–51.
- Rane S, Shankaran S, Natarajan G. Parental perception of functional status following tracheostomy in infancy: a single center study. *J Pediatr* 2013;163:860–866.
- American Academy of Pediatrics, Committee on Bioethics and Committee on Hospital Care. Palliative care for children. *Pediatrics* 2000;106:351–357.
- Feudtner C, Schall T, Hill D. Parental personal sense of duty as a foundation of pediatric medical decision-making. *Pediatrics* 2018; 142(Suppl 3):S133–S141.
- Sullivan J, Monagle P, Gillam L. What parents want from doctors in end-of-life decision-making for children. *Arch Dis Child* 2014;99: 216–220.
- Ferguson E, Wright M, Carter T, Van Halderen C, Vaughan R, Otter M. Communication regarding breathing support options for youth with Duchenne muscular dystrophy. *Paediatr Child Health* 2011;16: 395–398.
- Murrell DV, Lotze TE, Farber HJ, Crawford CA, Wiemann CM. The experience of families with children with spinal muscular atrophy type I across health care systems. *J Child Neurol* 2017;32:917–923.
- Keens TG, Kun SS, Davidson Ward SL. Chronic respiratory failure. In: Nichols DG, editor. *Rogers' textbook of pediatric intensive care*. 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2008. pp. 753–766.
- Ritchie J, Spencer L. Qualitative data analysis for applied policy research. In: Bryman A, Burgess RG, editors. *Analyzing qualitative data*. London: Routledge; 1994. pp. 173–194.
- Pope C, Mays N. *Qualitative research in health care*. 3rd ed. Malden, MA: Blackwell; 2006.
- Michelson KN, Clayman ML, Haber-Barker N, Ryan C, Rychlik K, Emanuel L, et al. The use of family conferences in the pediatric intensive care unit. *J Palliat Med* 2013;16:1595–1601.
- Hebert LM, Watson AC, Madrigal V, October TW. Discussing benefits and risks of tracheostomy: what physicians actually say. *Pediatr Crit Care Med* 2017;18:e592–e597.
- Ramelli GP, Hammer J. Swiss physicians' practices of long-term mechanical ventilatory support of patients with Duchenne muscular dystrophy. *Swiss Med Wkly* 2005;135:599–604.
- Kinali M, Manzur AY, Mercuri E, Gibson BE, Hartley L, Simonds AK, et al. UK physicians' attitudes and practices in long-term non-invasive ventilation of Duchenne muscular dystrophy. *Pediatr Rehabil* 2006;9: 351–364.
- Ryan MM. The use of invasive ventilation is appropriate in children with genetically proven spinal muscular atrophy type 1: the motion against. *Paediatr Respir Rev* 2008;9:51–54. [Discussion, pp. 55–56.]
- Gray K, Isaacs D, Kilham HA, Tobin B. Spinal muscular atrophy type I: do the benefits of ventilation compensate for its burdens? *J Paediatr Child Health* 2013;49:807–812.
- Mariguzzi S, Wassmer E. Mechanical ventilation in children with severe neurological impairment. *Dev Med Child Neurol* 2010;52: 1068–1069.
- van Gestel JP, Robroch AH, Bollen CW, Van Der Ent CK, Van Vught AJ. Mechanical ventilation for respiratory failure in children with severe neurological impairment: is it futile medical treatment? *Dev Med Child Neurol* 2010;52:483–488.
- Benson RC, Hardy KA, Gildengorin G, Hsia D. International survey of physician recommendation for tracheostomy for spinal muscular atrophy type I. *Pediatr Pulmonol* 2012;47:606–611.
- Tassie B, Isaacs D, Kilham H, Kerridge I. Management of children with spinal muscular atrophy type 1 in Australia. *J Paediatr Child Health* 2013;49:815–819.
- Oskoui M, Ng P, Liben S, Zielinski D. Physician driven variation in the care of children with spinal muscular atrophy type 1. *Pediatr Pulmonol* 2017;52:662–668.
- Meyer EC, Ritholz MD, Burns JP, Truog RD. Improving the quality of end-of-life care in the pediatric intensive care unit: parents' priorities and recommendations. *Pediatrics* 2006;117:649–657.
- October TW, Fisher KR, Feudtner C, Hinds PS. The parent perspective: "being a good parent" when making critical decisions in the PICU. *Pediatr Crit Care Med* 2014;15:291–298.
- Qian Y, McGraw S, Henne J, Jarecki J, Hobby K, Yeh WS. Understanding the experiences and needs of individuals with spinal muscular atrophy and their parents: a qualitative study. *BMC Neurol* 2015;15:217.
- Contro NA, Larson J, Scofield S, Sourkes B, Cohen HJ. Hospital staff and family perspectives regarding quality of pediatric palliative care. *Pediatrics* 2004;114:1248–1252.

- 29 Nageswaran S, Golden SL, Gower WA, King NMP. Caregiver perceptions about their decision to pursue tracheostomy for children with medical complexity. *J Pediatr* 2018;203:354–360.e1.
- 30 Edwards JD, Morris MC, Nelson JE, Panitch HB, Miller RL. Decisions around long-term ventilation for children: perspectives of directors of pediatric home ventilation programs. *Ann Am Thorac Soc* 2017;14:1539–1547.
- 31 Ray S, Brierley J, Bush A, Fraser J, Halley G, Harrop EJ, *et al*. Towards developing an ethical framework for decision making in long-term ventilation in children. *Arch Dis Child* 2018;103:1080–1084.
- 32 Henderson CM, Wilfond BS, Boss RD. Bringing social context into the conversation about pediatric long-term ventilation. *Hosp Pediatr* 2018;8:102–108.
- 33 Koncicki ML, Zachariah P, Lucas AR, Edwards JD. A multi-institutional analysis of children on long-term non-invasive respiratory support and their outcomes. *Pediatr Pulmonol* 2018;53:498–504.
- 34 Wilson A, Hurwitz CA, Smith M, Patino T, Kudalmana AS, Gallas M. Parents as teachers: teaching pediatrics residents the art of engaging in difficult conversations. *J Grad Med Educ* 2019;11: 60–65.
- 35 Meyer EC, Sellers DE, Browning DM, McGuffie K, Solomon MZ, Truog RD. Difficult conversations: improving communication skills and relational abilities in health care. *Pediatr Crit Care Med* 2009;10: 352–359.