Published in final edited form as:

J Palliat Care. 2012 ; 28(1): 21–27.

End-of-Life Discussions and Advance Care Planning for Children on Long-Term Assisted Ventilation with Life-Limiting Conditions

Jeffrey D. Edwards,

Division of Pediatric Critical Care, University of California, San Francisco, and Moffitt Hospital, Box 0106, 505 Parnassus Avenue, San Francisco, California, USA 94143-0106; edwardsj@peds.ucsf.edu

Sheila S. Kun,

Division of Pediatric Pulmonology, Children's Hospital Los Angeles, and Keck School of Medicine, University of Southern California, Los Angeles, California, USA

Robert J. Graham, and

Critical Care, Anesthesia, Perioperative Extension (CAPE) and Home Ventilation Program, and Division of Critical Care Medicine, Children's Hospital Boston, and Harvard Medical School, Boston, Massachusetts, USA

Thomas G. Keens

Division of Pediatric Pulmonology, Children's Hospital Los Angeles, and Keck School of Medicine, University of Southern California, Los Angeles, California, USA

Abstract

Families of children with life-limiting conditions who are on long-term assisted ventilation need to undertake end-of-life advance care planning (ACP) in order to align their goals and values with the inevitability of their child's condition and the risks it entails. To discuss how best to conduct ACP in this population, we performed a retrospective analysis of end-of-life discussions involving our deceased ventilator-assisted patients between 1987 and 2009. A total of 34 (72 percent) of 47 study patients were the subject of these discussions; many discussions occurred after acute deterioration. They resulted in directives to forgo or limit interventions for 21 children (45 percent). We surmise that many families were hesitant to discuss end-of-life issues during periods of relative stability. By offering anticipatory guidance and encouraging contemplation of patients' goals both in times of stability and during worsening illness, health care providers can better engage patients' families in ACP. As the child's condition progresses, the emphasis can be recalibrated. How families respond to such encouragement can also serve as a gauge of their willingness to pursue ACP.

INTRODUCTION

Families with a child who has chronic respiratory failure must choose between accepting long-term assisted ventilation, often via tracheostomy, or forgoing life-assisting technology for their child. Children with potentially regressive underlying conditions (for example, bronchopulmonary dysplasia) may ultimately be liberated from mechanical ventilation. Other children — those with underlying conditions that are static or progressive — are unlikely to be liberated. Many in this latter group are also at increased risk for critical illness

and/or early death due to life-limiting conditions and medical fragility, despite chronic ventilatory support (1-3). Increasingly, families with children in both circumstances choose assisted ventilation (4, 5).

The families of children with chronic respirator failure and life-limiting conditions need to be prepared to address the issues and decisions they will encounter at the end of the child's life, regardless of whether or not they choose long-term assisted ventilation. Much has been written on the ethics of initiating chronic invasive ventilation and the decision-making process involved (6-8), as well as on the discharge preparation of children and caregivers for home mechanical ventilation (HMV) via tracheostomy (9-12). However, how HMV patients with life-limiting conditions and their families are, and should be, prepared for the end-of-life decisions they will ultimately face, despite this assistive technology, has not been adequately addressed.

In order to investigate prior approaches to end-of-life planning for HMV patients with life-limiting conditions, we reviewed the medical records of our deceased HMV patients and assessed our HMV program's history of such preparation. We hypothesized that post-HMV-initiated end-of-life discussions were frequently prompted by the child's acute deterioration. We used the described circumstances of these discussions to reflect on how and when advance care planning (ACP) ought to occur. We also sought to use this review as a means to raise additional questions that deserve further study.

METHODS

We performed a retrospective chart review of children and young adults who died receiving full- or part-time home mechanical ventilation via tracheostomy and who were followed at Children's Hospital Los Angeles (CHLA) between July 1987 and April 2009. This paper and database review was approved by the CHLA Institutional Review Board. Patients were excluded if the circumstances of their deaths or other pertinent post-initiation medical history was not available.

Patient data were collected on age of initiation, comorbidities and neurodevelopmental disability, place of residence, and primary cause of chronic respiratory failure (CRF). Causes of CRF were classified into three groups: chronic pulmonary diseases (CPD), ventilatory muscle weakness (VMW), and central hypoventilation syndromes (CHS).

Records were reviewed for documented discussions with patients and/or family members that occurred after HMV was initiated and that explicitly addressed end-of-life goals, options, or advance directives. Data were collected on the number of discussions, reasons for the discussions, timing of the discussions in relation to the initiation of HMV, cardiopulmonary arrest (if applicable), and death; data were also collected on whether a decision to limit or forgo care resulted (for example, an advance directive or a do-not-resuscitate order). Reasons for the discussions were categorized as: acute deterioration, development of a new comorbidity, or the progression of an underlying condition. Conferences in which families were informed of their child's poor prognosis were excluded if no discussion of end-of-life options was documented.

Finally, post-initiation records were reviewed for number of hospitalizations and cardiopulmonary arrests. Arrests were defined as events that resulted in closed chest compression of any duration, emergency defibrillation, or administration of cardiac rescue medications. Events that involved only rescue positive-pressure breaths or non-cardiac emergency medications (for example, anti-epileptics) were excluded. We noted the outcome and timing of each event in relation to HMV initiation and, if applicable, the patient's first and last discussion of end-of-life options. Outcomes of arrests were classified as survival to

discharge, with or without new neurologic morbidity, or death. Summary statistics are presented, including median and interquartile ranges (IQR) when appropriate.

RESULTS

In all, 70 (22 percent) of 315 HMV patients died between June 1987 and April 2009; 23 (33 percent) were excluded because of insufficient information. The remaining 47 patients (67 percent) comprised our study cohort and had 148 person years of follow-up. Table 1 details the clinical characteristics of our cohort, as well as their incidence of ACP discussions. The entire cohort had underlying conditions that were either progressive or conferred a level of fragility that would decrease the child's lifespan relative to that of a healthy child. A documented discussion of end-of-life options and wishes occurred for 34 patients (72 percent); 24 patients (51 percent) had two or more discussions. Progression of an underlying condition prompted the first discussion for 16 (47 percent) of these 34 patients. Acute deterioration was the impetus in 14 (41 percent) first discussions; the remaining 4 (13 percent) were prompted by a new comorbidity. Discussions took place in the pediatric intensive care unit (ICU) (59 percent), ventilator ward (23 percent), clinic (15 percent), and neonatal ICU (3 percent). Intensivists documented 47 percent of the discussions, followed by pulmonologists (35 percent), combined (15 percent), and other sub-specialists (3 percent). A social worker was present and wrote an accompanying note in 88 percent of all discussions. Eight of nine patients who resided in a chronic care facility had a documented discussion.

Table 2 describes the time between birth, initiation of HMV, first discussion, last discussion, first cardiopulmonary event, and death. When times are shown as zero, it means that the two events occurred within days of each other. First discussions occurred at the time of, or within a few days of, death for nine children — 26 percent of those who had a documented discussion. When discussions occurred at or around the time of death, all were prompted by acute deterioration.

Discussions resulted in advance directives or orders to forgo or limit extraordinary interventions — such as cardiopulmonary resuscitation and escalation of therapies that were unlikely to result in survival — for 21 children (45 percent). Such decisions were made in the setting of acute deterioration or arrest for 19 of these children. In three of the cases, families elected to withdraw continuous inotropic and/or vasoactive medications after their children suffered an acute, severe decompensation. Ventilatory support was not withdrawn for any patient. In five cases, orders limiting future extraordinary care were later rescinded and not reinstated after the children recovered from their acute illness. There were no planned or temporary suspensions of orders to limit or forgo care — for surgeries, for example.

After initiation of HMV, all study patients had follow-up clinic visits and a subsequent hospitalization. The median number of hospitalizations per subject was five (IQR 3-8). Of 296 total admissions, 79 percent were unscheduled and 60 percent included an ICU stay.

Twenty-nine patients (62 percent) had a total of 39 cardiopulmonary arrests resulting in resuscitative efforts; eight (17 percent) had more than one arrest. Eight (20 percent) of these arrests were due to airway obstruction, decannulation, or ventilator disconnection. In all, 31 arrests (79 percent) occurred in hospital — 17 in an ICU, 6 on the ventilator ward, 5 in the emergency department, 2 in the operating suites, and 1 in a clinic; 8 events (21 percent) occurred out of hospital — 6 in a private residence and 2 in a chronic facility. All children who arrested outside of hospital were taken to one. In 20 arrests (51 percent), death was the outcome; in 19 (49 percent), survival to discharge was the outcome. Three surviving patients

(16 percent) experienced an appreciable decline in neurological function from their previous baseline.

DISCUSSION

Children on HMV via tracheostomy are a heterogeneous child population with complex chronic conditions and reliance on invasive ventilation. Although this technology aids in sustaining the child's respiratory function, it does not eliminate the need for continued ACP for those with severe comorbidities or life-limiting conditions. ACP is the process that guides patients to an understanding of their diagnosis — and, if possible, their prognosis — and then to consider the decisions and treatment options they will face in the future. In the case of someone whose personal goals and values cannot be known, a surrogate decision-maker should make choices in accord with that person's best interests (13). While ACP should be goals- and values-oriented, discussions of possible choices at the end of life are also necessary. Previous studies have demonstrated that parents of children with chronic conditions feel that ACP is or would be beneficial and wish to know of available options at the end of their child's life, including those that forgo interventions (14-16).

Our review demonstrated that unscheduled hospitalizations, ICU stays, and cardiopulmonary arrests were relatively common among our cohort of children on HMV. In addition, an earlier review showed that many of their deaths were unexpected and not the result of the progression of their underlying diseases (3). Thus, ACP is especially relevant to this population. End-of-life discussions were documented in about three-fourths of our cohort. Many of these discussions were first documented after the child experienced an acute deterioration and during a hospitalization. Some of the discussions took place after the child suffered an arrest or just prior to the child's death. This may suggest that earlier opportunities were missed, although it is also likely that more discussions occurred than were documented. This timing also reflects our experience that many families were not open to candidly discussing end-of-life issues during times of relative stability. However, some families were willing to limit future care that was unlikely to alter their child's course. These decisions were most commonly made after the children had already experienced a decline in clinical condition. Our review also demonstrated that close to one quarter of those families who chose to limit treatment efforts later elected to rescind the limitation. These two factors — resistance to discussing future end-of-life decisions before deterioration and the reversal of earlier limitations after the child survives a critical illness — may indicate the existence of a survival plateau. Such a plateau was first described by Simonds and colleagues, who observed it in patients with Duchenne muscular dystrophy after initiation of non-invasive ventilation (17). Birnkrant and Noritz later referenced this phenomenon as an obstacle to gaining families' assent to palliative care for their children with progressive neuromuscular disease on non-invasive ventilation (18). It is important to note, as well, that ventilatory support was not withdrawn from any child; this may suggest that the option is not being offered, or it may highlight the difficulty of reversing prior fundamental directions of care.

In part due to this review and other reflections on our practices, the CHLA HMV program has refocused its efforts on engaging families in ACP at the time of and after initiation and before critical illness. Ideally, ACP should begin prior to CRF, initiation of chronic ventilation, and critical illness (19, 20). However, CRF often results after an acute illness forces an at-risk child into non-compensable respiratory insufficiency and it is no longer possible to wean the child from mechanical ventilation (6, 19, 21). In either situation, we consider conversations about HMV to be part of ACP for children with CRF. ACP is no less important after HMV has been initiated than it is when HMV is addressed well in advance.

In all cases, we now provide patients' families with anticipatory guidance related to the limitations and risks of HMV (see Box 1) beginning with initial conversations about the nature of HMV and whether it is a good option for their child. To help ensure that expectations are realistic, we attempt to ensure that caregivers receive and understand this guidance before HMV initiation. We highlight these points again during the final discharge-planning meeting and, as part of completing the written discharge checklist, caregivers must sign a document acknowledging that they have heard the points discussed.

While much of ACP revolves around an exploration of families' goals for their children and the values that shape those goals, candid discussions of likely clinical scenarios should also be undertaken. Admittedly, it is difficult to address all such scenarios; the choice of which ones to cover depends largely on the individual child. For instance, if the child has chronic renal insufficiency, chronic renal replacement therapies should be discussed before the onset of end-stage renal failure. At minimum, for children at risk of acute deterioration, CPR should be discussed. In order for families to assent to or forgo these measures, they must understand that chest compressions with possible defibrillation and cardiac medications are the default in the face of cardiac arrest, that CPR is often successful only when the arrest is triggered by an acute reversible cause, and that non-fatal arrests can lead to new neurologic morbidity (22). After initiation, we assume that a family's choice of HMV is an implicit decision for future interventions and resuscitation in the face of acute, unexpected deterioration. Teaching CPR and management of airway emergencies is an explicit recognition of this choice. Since the causes of acute deterioration are often initially unclear and may be reversible, we believe that this conclusion is pragmatically and morally defensible, even for children who are medically fragile or who have life-limiting illnesses.

Reconciling the preference for resuscitation and extraordinary care can become a challenge when the child's underlying condition declines more significantly or the care team concludes that the child's critical illness is likely not reversible. Thus, this preference, while initially appropriate, often needs to be gently recalibrated as circumstances and family readiness change. This approach is reflected in our study finding that clinical instability often prompts end-of-life discussions. However, our review has also compelled us to proactively, rather than reactively, strive to gauge our families' readiness for ACP and to document all discussions, even those that do not result in a decision. When we engage in reactive discussions, we risk limiting ourselves to addressing narrow, immediate medical concerns during crises rather than exploring larger goals of care in anticipation of likely or inevitable outcomes (23). Other potential prompts for readdressing end-of-life goals include more frequent or more severe acute illnesses, development of a new comorbidity, and the child approaching the upper range of the anticipated lifespan. During periods of instability, the situation's gravity trumps family readiness, and the physician must alert the family of significant concerns related to morbidity or death.

If the child is clinically stable, it is not necessary for families to pre-emptively articulate their preferences for future scenarios. Rather, it is sufficient for them to gain a realistic understanding of their child's condition and risks and think through the values and goals that will likely shape their decisions. To help with the latter, one approach is to regularly recommend to families that they contemplate what they want for their child when he or she is in a period of relative health and, in the opposite scenario, what they want when the child is threatened with serious acute illness or worsening of an underlying condition. This approach permits the physician to gauge a family's readiness for further ACP — if, of course, the family is willing to share their thoughts with the physician. It also encourages families to reflect on the inevitability or the serious risks of their child's condition while also contemplating positive and hopeful goals or scenarios. The amount of time and effort medical caregivers should expend discussing scenarios of relative health versus illness

depends on the child's condition and trajectory. As a child's condition worsens, ensuring that families are contemplating end-of-life goals and eliciting more specific decisions become more crucial. Regular encouragement to contemplate and possibly discuss these goals or decisions would ideally be offered during regular out-patient visits, but it could also be provided during hospitalizations.

Although end-of-life issues and options will have been raised earlier (for example, during discussions about initiating HMV, or during previous critical illness), ACP after HMV initiation is no less challenging. In the case of children on HMV who have life-limiting conditions or severe comorbidities, balancing the risks or inevitability of their condition with their family's wish to continue to support them through subsequent critical illnesses is a delicate matter. Constantly reminding families of their child's serious diagnosis or overwhelming them with negative anticipatory guidance risks destroying their hope and sabotaging the family-physician relationship. Yet, by communicating belatedly or in a manner that is too tempered, physicians risk under-informing families. Box 2 lists potential challenges to ACP that we and other medical caregivers of children with complex chronic conditions have encountered (17, 21, 24-27). Challenges can stem from both families and health care providers. Becoming aware of these challenges is the first and most important step in overcoming them, and eliciting families' hopes, goals, and fears is always beneficial. Some families refuse limitation of extraordinary measures at the end of their child's life. While some health care professionals may believe that this refusal prolongs the child's suffering, such decisions should be viewed respectfully if they are consistent with the family's ACP goals (28, 29). The emphasis of ACP should be placed not on eliciting a particular decision but on informing the family about events that could occur and on reassuring them that caring for their child will be the primary goal under all circumstances.

Our review of ACP for children on long-term assisted ventilation raises a number of questions that deserve further investigation. Future studies are needed to: survey HMV families and other health care providers to obtain their views on how and when ACP and anticipatory guidance are best undertaken; survey HMV families for information on any written advance directives they might have prepared before initiation of HMV, and on how HMV altered these directives; determine whether family readiness for ACP is complicated by a period of relative stability for the child after HMV initiation and/or by an expectation that HMV will be life-sustaining or prolonging; determine whether and how chronic ventilation alters perceptions of, and decisions for, subsequent invasive or extraordinary interventions during critical illnesses; ascertain whether the perceptions and decisions of these families are different from those of families whose children have complex chronic conditions but are not on long-term ventilation; and establish whether families of children on HMV because an acute illness has rendered them incapable of being weaned from invasive ventilation have different readiness for ACP or perceptions of subsequent interventions than families whose children have been proactively started on HMV due to their known chronic respiratory insufficiency. All of these questions would be best addressed prospectively and with a multi-institutional and diverse cohort.

While our review helps both to frame the importance of ACP and the difficulties it poses for children on HMV and to highlight several additional questions that deserve further study, it does have some notable limitations. First, we only reviewed HMV patients who died. We felt that these patients represented a reasonable convenience sample of patients who ought to have had ACP. However, there were certainly more non-deceased HMV patients with life-limiting conditions for whom ACP was necessary. Our convenience sample may have introduced selection bias to our review; and because 23 deceased patients were excluded, we may have introduced additional selection bias. Second, our review of ACP largely focused on end-of-life issues, though there can certainly be more to ACP. This is the consequence of

end-of-life discussions being more consistently documented in the medical records than other ACP topics. Nevertheless, it skews our presentation of ACP and diverts the accompanying discussion away from other important ACP topics. Third, our lengthy study period covers a time during which the relative complexity of complex chronic conditions in children increased, home ventilators with continuous flow were introduced, and patient autonomy in decision making gained even greater prominence. However, we surmise that these developments did not significantly change the underlying situation or challenges for HMV patients with life-limiting illness. Throughout the period, a subset of HMV patients lived and died at the limits of medicine at that time. Fourth, as this was a retrospective study, included discussions were limited to those documented in our institution's records. It is likely that more discussions and, possibly, more arrests occurred than we could identify. Finally, it is difficult to know whether all of our recorded arrests were in fact events that warranted cardiopulmonary resuscitation, or if some were confounded by caregiver overreaction.

CONCLUSION

Advance care planning for children with life-limiting conditions is a necessary part of the care for these children and their families. ACP is a complicated process when a child's prognosis is known; it is even more so when the prognosis is not predictable but the child is at risk of acute critical illnesses. Our experience is that assisted ventilation can support children with chronic respiratory failure. But it can also complicate the process of pre-emptively deliberating the goals of care and, thus, the decisions that will be faced in the future given the child's life-limiting condition. Care providers should therefore offer measured anticipatory guidance and proactively gauge family readiness for ACP. In order to better engage families in ACP, care providers must identify the potential familial and professional barriers to ACP. Moreover, they should encourage families to contemplate and articulate their goals for their children in scenarios of relative health and worsening illness. As the former wanes and the latter waxes, emphasis can gradually be shifted toward end-of-life issues. While imperfect, this approach is superior to delivering frequent or forceful reminders of the child's fragility and risk for early death, which can impair the family's healthy alliance with health care professionals.

Acknowledgments

The authors would like to thank Dr. Cheryl Lew for her review of the manuscript.

This work was supported by the Department of Anesthesiology Critical Care Medicine, Children's Hospital Los Angeles. Dr. Edwards is supported by a NICHD Pediatric Critical Care Scientist Development Program grant (K12 HD 047349).

Abbreviations Key

ACP	advance care planning
CHLA	Children's Hospital Los Angeles
CHS	central hypoventilation syndromes
CPD	chronic pulmonary diseases
CRF	chronic respiratory failure
HMV	home mechanical ventilation
IQR	interquartile range

VMW ventilatory muscle weakness

REFERENCES

1. Wheeler WB, Maguire EL, Kurachek SC, et al. Chronic respiratory failure of infancy and childhood: clinical outcomes based on underlying etiology. *Pediatr Pulmonol.* 1994; 17(1):1–5. [PubMed: 7509054]
2. Gilgoff RL, Gilgoff IS. Long-term follow-up of home mechanical ventilation in young children with spinal cord injury and neuromuscular conditions. *J Pediatr.* 2003; 142(5):476–480. [PubMed: 12756376]
3. Edwards JD, Kun SS, Keens TG. Outcomes and causes of death in children on home mechanical ventilation via tracheostomy: an institutional and literature review. *J Pediatr.* 2010; 157(6):955–959. e2. [PubMed: 20713294]
4. Gowans M, Keenan HT, Bratton SL. The population prevalence of children receiving invasive home ventilation in Utah. *Pediatr Pulmonol.* 2007; 42(3):231–236. [PubMed: 17262859]
5. Graham RJ, Fleege EW, Robinson WM. Chronic ventilator need in the community: a 2005 pediatric census of Massachusetts. *Pediatrics.* 2007; 119(6):e1280–e1287. [PubMed: 17485451]
6. Shneerson JM. Home mechanical ventilation in children: techniques, outcomes and ethics. *Monaldi Arch Chest Dis.* 1996; 51(5):426–430. [PubMed: 9009634]
7. Perkin RM, Orr R, Ashwal S, Walters J, et al. Long-term ventilation in children with severe central nervous system impairment. *Semin Neurol.* 1997; 17(3):239–248. [PubMed: 9311066]
8. Glass KC, Carnevale FA. Decisional challenges for children requiring assisted ventilation at home. *HEC Forum.* 2006; 18(3):207–221. [PubMed: 17650760]
9. Graf JM, Montagnino BA, Hueckel R, et al. Children with new tracheostomies: planning for family education and common impediments to discharge. *Pediatr Pulmonol.* 2008; 43(8):788–794. [PubMed: 18613098]
10. Warren ML, Jarrett C, Senegal R, et al. An interdisciplinary approach to transitioning ventilator-dependent patients to home. *J Nurs Care Qual.* 2004; 19(1):67–73. [PubMed: 14717150]
11. Make BJ, Hill NS, Goldberg AI, et al. Mechanical ventilation beyond the intensive care unit. Report of a consensus conference of the American College of Chest Physicians. *Chest.* 1998; 113(5 Suppl):S289–S344.
- 12*. Tearl DK, Cox TJ, Hertzog JH. Hospital discharge of respiratory-technology-dependent children: role of a dedicated respiratory care discharge coordinator. *Respir Care.* 2006; 5(7):744–749. 1. [PubMed: 16800908]
13. American Academy of Pediatrics. Guidelines of forgoing life sustaining medical treatment. *Pediatrics.* 1994; 93(3):532–536. [PubMed: 8115226]
14. Friedman SL. Parent resuscitation preferences for young people with severe developmental disabilities. *J Am Med Dir Assoc.* 2006; 7(2):67–72. [PubMed: 16461246]
15. Hammes BJ, Klevan J, Kempf M, et al. Pediatric advance care planning. *J Palliat Med.* 2005; 8(4):766–773. [PubMed: 16128650]
16. Wharton RH, Levine KR, Buka S, et al. Advance care planning for children with special health care needs: a survey of parental attitudes. *Pediatrics.* 1996; 97(5):682–687. [PubMed: 8628607]
17. Simonds AK, Muntoni F, Heather S, et al. Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy. *Thorax.* 1998; 53(11):949–952. [PubMed: 10193393]
18. Birnkrant DJ, Noritz GH. Is there a role for palliative care in progressive pediatric neuromuscular diseases? The answer is “Yes! *J Palliat Care.* 2008; 24(4):265–269. [PubMed: 19227018]
19. Simonds AK. Respiratory support for the severely handicapped child with neuromuscular disease: ethics and practicality. *Semin Respir Crit Care Med.* 2007; 28(3):342–354. [PubMed: 17562504]
20. Sritippayawan S, Kun SS, Keens TG, et al. Initiation of home mechanical ventilation in children with neuromuscular diseases. *J Pediatr.* 2003; 142(5):481–485. [PubMed: 12756377]
21. Fraser J, Henrichsen T, Mok Q, et al. Prolonged mechanical ventilation as a consequence of acute illness. *Arch Dis Child.* 1998; 78(3):253–256. [PubMed: 9613357]

22. Haque IU, Udassi JP, Zaritsky AL. Outcome following cardiopulmonary arrest. *Pediatr Clin N Am*. 2008; 55(4):969–987.
23. Wharton RH, Levine KR, Buka S, et al. Advance care planning for children with special health care needs: a survey of parental attitudes. *Pediatrics*. 1996; 97(5):682–687. [PubMed: 8628607]
24. Feudtner C, Christakis DA, Zimmerman FJ, et al. Characteristics of deaths occurring in children's hospitals: implications for supportive care services. *Pediatrics*. 2002; 109(5):887–893. [PubMed: 11986451]
25. Steele RG. Trajectory of certain death at an unknown time: children with neurodegenerative life-threatening illnesses. *Can J Nurs Res*. 2000; 3(3):49–67. 2. [PubMed: 11928133]
26. Graham RJ, Robinson WM. Integrating palliative care into chronic care for children with severe neurodevelopmental disabilities. *J Dev Behav Pediatr*. 2005; 26(5):361–365. [PubMed: 16222176]
27. Forbes T, Goeman E, Stark Z, et al. *J Paediatr Child Health*. 2008; 44(7-8):392–398. [PubMed: 18638330]
28. Truog RD. Is it always wrong to perform futile CPR? *N Engl J Med*. 2010; 362(6):477–479. [PubMed: 20147712]
29. Meyer EC, Burns JP, Griffith JL, et al. Parental perspectives on end-of-life care in the pediatric intensive care unit. *Crit Care Med*. 2002; 30(1):226–231. [PubMed: 11902266]

Box 1 Anticipatory Guidance for Families with Children on Long-Term Assisted Ventilation

- Home mechanical ventilation is only an assistive, never a curative, technology for respiration; it cannot alter progressive conditions or the risks and burdens of co-morbidities.
- Home ventilators are often inadequate for assisting respiration during acute illness.
- Re-hospitalization is often necessary for patients with these acute illnesses.
- Tracheostomy complications and accidents – such as airway obstruction, decannulation, ventilator disconnection, false track replacement, aspiration, and tracheal bleeding – are potentially life-threatening. Caregiver competence in tracheostomy care, bag-tracheostomy ventilation, and cardiopulmonary resuscitation can be life-saving.
- The child's goals of care will likely need to be re-addressed when the child's condition worsens, which can happen without warning.

Box 2 Potential Family and Health Care Provider Challenges to Advance Care Planning and End-of-Life Discussions for Children on Long-Term Assisted Ventilation

Family

- There is a period of relative stability for the child after home mechanical ventilation (HMV) is initiated (termed a “survival plateau” by Simonds and colleagues [17]).
- The family has an idealistic, positive outlook on the child's prognosis because they see the ventilator as life-prolonging.
- The family has an idealistic, positive outlook on the child's prognosis because the child has survived previous critical illness.
- A mutually dependent relationship develops between the child and family members who have devoted so much energy to the child's care.
- After choosing HMV, the family presumes that they cannot or should not discontinue mechanical ventilation or forgo subsequent interventions when the child's condition deteriorates.

Health care providers

- Health care providers have difficulty prognosticating death for a particular child.
- A shift takes place in the medical home of the child, who moves from generalist to potentially intervention-oriented specialist.
- Health care providers erroneously presume that because a family has chosen HMV, they will always choose extraordinary measures to prolong the child's life.
- Health care providers avoid raising sensitive, difficult issues with family members and project their own lack of readiness to discuss these issues onto them.
- Health care providers experience difficulty or failure in attempting to integrate cure-oriented care and palliative care.
- Health care providers make conscious or unconscious attempts to impose their own values on the family.
- Health care providers may be unable or unwilling to accept the patient's fate.

Table 1

Characteristics and Advance Care Planning Discussions of 47 Expired Children on Long-Term Assisted Ventilation

Characteristics	n (%)
<i>Males</i>	26 (55)
<i>Primary cause of chronic respiratory failure</i>	
chronic pulmonary disease	23 (49)
ventilatory muscle weakness	12 (28)
central hypoventilation syndrome	11 (23)
<i>Conditions/co-morbidities</i>	
additional chronic medical device dependence	40 (85)
neurodevelopmental delay	37 (79)
epilepsy	19 (40)
airway obstruction (e.g., tracheobronchomalacia, vocal cord paralysis)	16 (34)
bronchopulmonary dysplasia	15 (32)
complex structural heart disease	10 (21)
hypoxic ischemic encephalopathy	10 (21)
reactive airway disease	8 (17)
myopathy/muscular dystrophy	7 (15)
prematurity (<37 weeks gestation)	7 (15)
genetic syndrome/chromosomal abnormality	6 (13)
pulmonary hypoplasia/thoracic dysplasia	6 (13)
scoliosis/kyphosis	6 (13)
bronchiolitis obliterans/bronchiectasis	4 (9)
spinal muscle atrophy	4 (9)
<i>Non-pulmonary comorbidities, median (interquartile range)</i>	3 (2-5)
<i>Resided in a chronic care facility</i>	9 (19)
<i>Discussion occurred</i>	34 (72)
before arrest occurred	3 (6)
after arrest	16 (34)
no arrest occurred	15 (32)
<i>No discussion occurred</i>	13 (28)
no arrest occurred	3 (6)
arrest occurred	10 (21)

Table 2

Time Intervals between Birth, Initiation of HMV, Advance Care Planning Discussions, Cardiopulmonary Arrests, and Death in 47 Children

Events	Applicable patients	Intervening time (months)		
	n	median	IQR ^a	range
Birth to initiation of HMV ^b	47	13	6-28	1-178
Initiation of HMV to death	47	22	8-60	1-176
Initiation of HMV to first discussion	34	6	0-25	0-105
First arrest to first discussion	23	0	0-12	0-178
First discussion to death	34	8	0-23	0-178
Last discussion to death	34	1	0-6	0-23

^aInterquartile range.

^bHome mechanical ventilation.