Towards developing an ethical framework for decision making in long-term ventilation in children

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ABSTRACT

The use of long-term ventilation (LTV) in children is growing in the UK and worldwide. This reflects the improvement in technology to provide LTV, the growing number of indications in which it can be successfully delivered and the acceptability of LTV to families and children. In this article, we discuss the various considerations to be made when deciding to initiate or continue LTV, describe the process that *should* be followed, as decided by a consensus of experienced physicians, and outline the options available for resolution of conflict around LTV decision making. We recognise the uncertainty and hope provided by novel and evolving therapies for potential disease modification. This raises the question of whether LTV should be offered to allow time for a therapy to be trialled, or whether the therapy is *so* unlikely to be effective, LTV would simply prolong suffering. We put this consensus view forward as an ethical framework for decision making in children requiring LTV.

What is already known?

- The number of children needing long-term ventilation is increasing.
- Decision making in long-term ventilation is made on a case-by-case basis, but no standardised framework exists.

What this study adds?

- Standardised processes with multidisciplinary involvement are likely to improve the consistency and quality of decision making in long-term ventilation.
- Novel therapies introduce uncertainty into decision making in long-term ventilation.
- Parallel planning must be undertaken alongside providing long-term ventilation.

INTRODUCTION

Long-term ventilation (LTV) is considered as mechanical support for breathing in a medically stable patient, regardless of interface, for all or part of the 24-hour day.[1] The use of LTV in children has increased exponentially in the UK in the last 20 years.[2] This increase is disproportionate to the rate of population growth, which might suggest that the decision threshold to initiate LTV has become lower. The question 'should a child receive LTV?' is complex, individual and dynamic. A framework for decision making in life-limiting and life-threatening conditions is provided by the Royal College of Paediatrics and Child Health.[3] The same principles should guide decision making in children subject to LTV. However, we advocate that additional considerations should also apply (box 1).

Here we discuss those considerations for the children, their parents and the professionals who have to make decisions about LTV; both practical aspects and parallel planning when LTV is used as a life-sustaining treatment. Most of our data and discussion will pertain to tracheostomy-delivered LTV, which carries a greater requirement for care. We will not be discussing the use of non-invasive ventilation, due to its use to support children with a wide spectrum of disease and care needs. We will also not discuss the use of LTV (usually nasal mask ventilation) to optimise palliative care in children with rapidly fatal conditions; this has been written about elsewhere.[4] The principles of decision making that we describe will apply to children deteriorating despite 24 hours a day non-invasive ventilation support, or where escalation to tracheostomy ventilation is considered.

CLINICAL CONSIDERATIONS

The Children Act 1989 set significant harm as the threshold to intervene in family life in the child's best interests.[5] LTV is never offered with the intention of causing harm; however, the impact of LTV on the child must be accounted for when balancing the benefits and burdens of treatment and disease. The goals of LTV can be divided into: (1) a bridge to either recovery or definitive treatment that is aimed at transition to a state that will not require LTV- for example, LTV allowing the growth and development of a child until recovery occurs with time or treatment, or (2) a destination therapy, where there are no further recovery or definitive treatment options, and life-long LTV will be required to enable a child to be discharged from hospital (ie, LTV is not a bridge to a cure but the destination itself).

Benefit is clear when LTV is a realistic bridge to recovery or definitive treatment. While there are no randomised controlled trials, observational studies provide guidance on situations where LTV will act as a bridge to recovery or treatment (box 2).[2 6–8] Even when used in this manner, consideration must be given to the child's overall functional status and the progress of disease rather than the diagnosis

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Box 1 Specific considerations regarding decision making in long-term ventilation (LTV)

- LTV is provided for a wide, and ever increasing, range of conditions. For many of these indications, the outcome of LTV has not been evaluated, is uncertain or is unknown.
- LTV is never a curative intervention; it cannot alter static or progressive conditions or the risks and burdens of comorbidities.
- In some cases, LTV is a 'bridge' to definitive therapy or recovery following growth. However, in many circumstances, LTV is a 'destination' therapy. Goals of treatment can change over time and need repeated re-evaluation.
- LTV is by definition 'long term'. Dependency on a ventilator can become part of life, rather than a treatment that can be withdrawn. This may occur without re-evaluation of the changing balance between benefits and burdens.
- Although portable ventilators have been available for over 30 years, the relative low numbers of long-term ventilated children means that expertise is still concentrated in tertiary centres, and care needs still involve specialist skills. This can introduce unforeseen burdens beyond the direct effects of attaching a ventilator to a child, such as prolonged stay in hospital and intensive care environments, which may retard development.
- Emerging or experimental treatments provide hope for a cure, which may often be unrealistic. LTV may be sought for a child as a bridge to potential cure. However, the likelihood of a successful cure has to be reviewed regularly, along with treatment burdens.

alone. Follow-up studies show that some children on LTV die due to causes unrelated to the reasons for initiation of therapy.[7]

As a destination therapy, LTV is likely to prolong life to different degrees; for example, in congenital central hypoventilation syndrome, children will progress to adulthood with life-long LTV support; in progressive neuromuscular disorders, life may be prolonged by preventing respiratory insufficiency, but life will still be limited by disease progression. However, LTV comes with its own burdens. Invasive ventilation requires regular airway suctioning as a consequence of the iatrogenic and often disease-related inability to effectively cough. Airway suctioning can cause discomfort; even though not as painful as deep suctioning in intensive care, repetitive tracheostomy suctioning can cause distress.[9,10] Discomfort associated with ventilator dyssynchrony has been described in adults.[11] This risk in children is potentially greater for a number of reasons including less voluntary control of breathing and ineffective triggering due to smaller tidal volumes.[12] Dyssynchrony can occur during manual ventilation prior to airway suctioning. The presence of a tracheostomy tube can alter the colonisation flora with potentially pathogenic microorganisms.[13]

Box 2 Indications for using long-term ventilation (LTV) as a bridge to recovery/definitive therapy (not destination therapy)

Airway

- Airway malacia.
- Upper airway obstruction.
- Subglottic stenosis.

Chest wall and lung parenchymal disease

- Chronic lung disease for which treatment exists or spontaneous recovery is possible.
- Lung hypoplasia.
- Chest wall deformities amenable to surgery.
- Cystic fibrosis with lung transplant as definitive therapy (may be dictated by national transplant programme criteria).

Cardiovascular

- Congenital heart disease where surgical repair is more amenable later in childhood, for example, valve disorders.

Neurological

- Reversible phrenic nerve injury.
- Reversible neuropathy/myopathy, for example, Guillain-Barré syndrome and critical illness neuropathy.

LTV is sometimes requested as a bridge to emerging or experimental therapies. The benefits of providing LTV in this manner are more difficult to evaluate. The decision may have to be made on an individual basis using existing data and based on best and worst case scenarios. In the worst case, LTV is instituted for a progressive disease for which no effective remedy emerges, leading to a 'locked-in' state with no motor activity. An existing framework for the evaluation and ethical assessment of such speculative therapy has been published.[14,15] However, the emergence of novel and emerging therapies for progressive neuromuscular conditions such as gene therapy and nusinersen in Spinal Muscular Atrophy Type 1 (SMA-1) [16,17] offers some families an expectation of disease remission and modulation. Even though current evidence does not support the notion that these therapies may *reverse* the need for LTV in children, the ethics of not offering LTV to these children must be reappraised. In this context of emerging treatments, disagreements between families who want to be afforded time for a 'miracle cure', and professionals who feel expectations are not realistic, are not uncommon and require sensitive handling (below). Professionals need to be mindful that therapies now adopted into the mainstream, such as non-invasive ventilation in Duchenne Muscular Dystrophy, may have once been considered unethical. This evolution often arises out of families challenging medical dogma. Nevertheless, this should not encourage limitless attempts at disease cure at the cost of causing suffering to the child: optimism must be balanced by what has already been tried and what is already known.[18]

PATIENT AND FAMILY CONSIDERATIONS

The ability to breathe may be considered to be something any rational human being would want. However, this may not be so if the support required to deliver breath itself causes discomfort or pain. In most cases, children on LTV, both via tracheostomy and mask interfaces, report the burden of care as mild or moderate.[19] While adult qualitative studies suggest acceptance among patients in whom LTV offers symptomatic relief or is essential for survival,[20] some patients with neuromuscular disease have expressed hopelessness, related to their diagnosis rather than to the burden of LTV.[21]

However, the instigation of LTV is often most contentious when patients are unable to voice their opinions through disability or developmental age. Physical expressions of pain and discomfort may provide a guide to their experience of LTV. Despite this, the discomfort caused may still be preferable to having their life limited. Adult patients who had negative views of LTV accepted it once they realised the consequence of refusal.[20] It is also important to remember physicians[4,21,22] and

Box 3 Overlapping interests between parents and children

Determinative
Parents' capacity of care and desire to do so likely is an important determinant of whether their child has an interest in continuing life.

Evaluative
A child is more likely than not to adopt the values of the parent.

Interdependence
A child may have an interest in promoting the parents' interests as much as the parents are interested in promoting the child's interests.

Epistemic
The parents are best placed to assess the interests of a child on account of knowing their child best.

even caregivers[19] tend to underestimate quality of life scores in patients on LTV.

The Children Act of 1989 defines parental responsibility as being concerned with 'bringing the child up, caring for him and making decisions about him'.[5] Parents and children have overlapping interests[23] (box 3). Parental surveys highlight the demanding nature of looking after a child needing LTV.[24 25] Parental attitudes towards LTV will have a determinative effect on the child's life. A child is likely to adopt their parents' values towards LTV, just as they are likely to develop similar moral and religious values, for example, values regarding sanctity of life. Importantly, though, it is the child that has the personal experience of their condition—one which is unique and which, in time, will have a stronger effect on their values than the experience of their parents. While children depend on their parents, parents also depend on children for their own emotional welfare. Parents are likely to have insightful knowledge of their child's best interests by virtue of knowing their children as individuals. This is still invariably true, even among children who spend lengthy periods of their lives in hospital, being cared for by large teams of professionals. The balance of overlapping interests between child and parent are changeable: regular review of these, through multidisciplinary input, is necessary to safeguard against the parental interests overtaking the child's best interests that must always be regarded as paramount. Rarely this may mean a child's best interests have to be protected through fostering or adoption.

In addition to parents, the impact of LTV on siblings and family life must also be considered. Siblings who are well may be relatively neglected due to the disproportionate care needs for an LTV child. Families also can become increasingly isolated, leading to breakdown in relationships. The presence of carers in the household, especially if they change regularly, can be intrusive for the whole household, with loss of privacy. Therefore, family views and expectations are important considerations.[26]

PUBLIC HEALTH CONSIDERATIONS

LTV in children is expensive. This may play a role in clinician decision making, but more widely, in service provision of health and social care. This expense extends beyond the individual patient for whom LTV is being considered. Data from 2006 demonstrated an average cost of £239 855/year for a 24-hour home care package, with much higher costs for children being cared for in specialist and intensive care units.[27] Although currently very few children receive 24-hour care packages, and trained carers rather than nurses are often used (data from the UK Hospital to Home database), costs are still substantial. Novel drugs of comparable cost (eg, ivacaftor in cystic fibrosis[28]) are subject to health economic analyses by bodies such as the National Institute of Health and Clinical Excellence (NICE). However, a similar analysis of LTV has not been conducted, possibly given the heterogeneous indications and relative low volume of children requiring it.

There are considerable hospital costs in addition to home care costs. The median waiting time to be discharged from hospital, following the decision to establish LTV and once the child is fit for discharge, is 75 days (unpublished data, Hospital to Home). This time is often spent in a high dependency or intensive care facility where bed costs range from £800 to £3200 a day. Two-thirds of children discharged home with an LTV care package are readmitted to hospital in the following 12 months[27] (although these data precede the increase in numbers of dedicated LTV specialist teams). Beyond the financial cost, there is also an opportunity cost to other patients who require intensive care, given that this is a scarce resource. The LTV nursing workforce is often drawn from a common pool: community nurses and home carers may be recruited from hospitals, which leaves gaps in hospital staffing rotas. Some children on LTV currently receive respite and clinical support from third sector children's hospices. In the current economic climate, many such services are under threat, further reducing opportunities for these children outside of their own homes.

Without formal health economic analyses, it is difficult to undertake policy level 'rationing'. As with other health economic analyses, this would need to be made on the basis of whether the benefit offered by the treatment is worth the cost: currently, NICE set a cost per quality-adjusted life year threshold between £20 000 and £30 000 a year.[29] The cost of LTV is often far in excess of this. Still, LTV may be in the best interests of the child, and as with ivacaftor, the public pressure to pay for it will be difficult to overcome. The emergence of social media has increased the potential for public lobbying, which can lead to considerable pressure on professionals who may not have the necessary recourse to informed consultation, as seen in several recent high-profile cases.[30 31] Despite these difficulties, public health strategy should provide for a growing population of children needing LTV, in order to prevent de facto 'rationing'. Investment in training a larger proportion of nurses and carers to look after children needing LTV, to minimise time spent in hospital and improving continuity at home, could be one public health strategy that positively affects the balance between burdens, fiscal cost and benefits of LTV. In the meantime, individual decision making must follow due process to ensure LTV will be in a child's best interests, regardless of whether society is able to pay.

A more detailed ethical discourse has been put forward by Fraser *et al*.[32] We outline the following practical guidance regarding decision making below.

DECISION MAKING

Major decision making in LTV occurs in two situations: (1) whether to withhold LTV in already intubated patients, or withdraw already established LTV support, and and (2) whether to initiate invasive LTV (via a tracheostomy tube) as an escalation from non-invasive support. The former is always difficult as the withholding or withdrawal of LTV in many instances would precipitate death.

Box 4 Good practice in shared decision making

- Decision making is a process, and decisions should not be made in a single meeting.
- Combine the clinician's knowledge and experience with the understanding, values, beliefs and expectations of the child and parent(s) to achieve optimal outcomes for the child.
- Communication between these stakeholders must be open, empathetic and non-judgemental.
- Initial discussions must centre around obtaining relevant information and discuss diagnosis, prognosis and, crucially, the goals of care—what will success look like?
- It is important to assess the family's current understanding and perceptions concerning the illness and the treatment options.
- It is paramount that the family feel that the healthcare team empathise with them and their child, acknowledging the difficulty in the decision making.
- Physicians should present an honest account of what life entails for a family caring for a child on LTV. Where possible, families should be given an opportunity to contact other families with children on LTV. This may be done through video diaries or remotely, but ideally should attempt to give a realistic perspective of local services available. However, with variation in underlying pathology, dependency and outcome, difficulties can arise in managing expectations appropriately.

Shared decision making is the prevalent approach to reaching a consensus when a decision crossroad is reached in patient care. This child-centred, collaborative process encourages active participation between families and healthcare professionals in reaching a decision together, based on all treatment options available.[33] Shared decision making may not necessarily ensure moral validity: medical teams and parents may reach a consensus, but the decision may not be in the child's best interests.[34]

Medical paternalism can influence shared decision making through the various treatment options presented. Information should be presented in an open, consistent and transparent way, although this does not preclude the healthcare professionals from recommending a management plan. Consistency of approach is best achieved via a core group of clinical professionals. This should include the acute care/current care consultant, the respiratory consultant who will follow-up the child in the long term and a member of the local paediatric team who will be the first port of call for the family. Relevant specialty team members must be present to provide information about the natural course of the underlying disorder and other care needs. Non-medical team members such as acute and specialist nurses, physiotherapists, play therapists and psychologists should be represented. In all cases, palliative care should also be represented to provide that vital interface with advance care planning when life limitation is anticipated.[35]

The participation of children in decision making can improve autonomy and moral validity. However, as stated above, where the decision regarding LTV initiation is often the most contentious, the child may not be physically or developmentally able to partake in the decision-making process. All parties must be cognisant of this, with mechanisms in place for any dissenting voice to challenge a decision, for example, through ethics committees.

Good practice in shared decision making is shown in box 4.

CONFLICT AND RESOLVING DIFFERENCES

There are several challenges to shared decision making regarding LTV (box 5). If there is disagreement or conflict, it is essential to seek one or more second opinions and encourage the family to do the same. If a consensus cannot be reached on the best interests of a patient, there are three next steps for consideration:

- Implement a decision that is not in keeping with the expressed choice of one of the decision makers, for example, a further trial of weaning invasive ventilation, even though previous trials have been unsuccessful, rather than committing to LTV. Any such decision must be morally defensible and should be preceded by consultation with colleagues, including palliative care teams and legal teams. Clearly stated and transparent end-points for example, time limits or goals of care should be agreed at the outset.
- Attempt to resolve the disagreement through negotiation, involving independent second opinions, ethics committees and trained mediators. Mediation removes the confrontational nature of the legal process, aims to reach mutual agreement between parties and may be quicker and less expensive than resolution through the courts.[36 37] Third parties must be fully informed about the clinical condition of the child and the wishes of the child and family.
- If resolution is not possible through the above, the decision must be referred to the courts. In the UK, the Family Division of the High Court will appoint an independent guardian ad litem to represent the child's interests and will then hear evidence and arbitrate between differing positions on the basis of a best interests calculus.[38 39] The court can and must make a final and binding decision about what should be done, according to the judge's view about what is in the child's best interest. Although deemed adversarial for some parents, who may be bound by strong religious or cultural beliefs, deferring decision making to an arm's-length body such as a court may be beneficial.

Box 5 Challenges to decision making in LTV

- Dealing with uncertainty is a fundamental problem: it is very difficult to assess prognosis and quality of life on an individual basis.
- The family may have an idealistic, positive outlook on the child's prognosis because he or she has survived previous critical illness.
- There is a risk of physical or emotional depletion after long admission/s in intensive care.
- Consistency may be lacking in messages from multiple healthcare professionals over a long stay in hospital.
- Healthcare providers may avoid raising sensitive, difficult issues with family members and project their own lack of preparedness to discuss these issues with them.

PARALLEL PLANNING TO LTV

When deliberating the decision to initiate or withhold LTV, the clinical team should also discuss what palliative care realistically entails in their local area. Parallel planning should include ceilings of treatment, for example, use of advanced ventilation techniques or cardiopulmonary resuscitation.[28] LTV may not alter the progression of disease and may introduce new comorbidities, for example, tracheostomy breakdown. Parallel planning is a continuous process that guides treatment options as the child's condition progresses. A decision, via the same multidisciplinary team approach, about whether to admit a child to an intensive care unit in the face of a progressive disease and increasing ventilation requirements should be taken before a crisis supervenes. It is good practice to agree an advanced care plan with the family. Once agreed, a written care plan should be circulated widely to all health care providers involved. This should include those likely to provide acute care.[35]

In addition to these considerations, regular re-evaluation is needed for children on LTV to ensure that the burdens of care do not exceed the benefits of treatment. For example, the trajectory of illness may change; problematic mechanical ventilation as a consequence of a severe viral infection or hypoxic brain injury following a cardiorespiratory arrest may completely shift the balance of benefit and burden of care. The direction and speed of progression of the child's condition is just as important as the starting point, and regular, detailed multi-disciplinary team (MDT) review of burden and benefits is mandatory. Planned withdrawal of LTV, providing a more dignified death compared with an ongoing struggle awaiting progressive deterioration, can be undertaken following established principles for palliative extubation.[40]

CONCLUSIONS

The principles of decision making in LTV require specific considerations due to the evolving landscape of novel therapies, the impositions on the child and the long-term care commitment required by family and carers. We have outlined these from clinical, family and public health perspectives. Such considerations should be used in LTV decision making. They will also require constant review, as experience increases with childhood LTV, technology and care provision changes and new evidence emerges.

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