

Ethics Rounds: When Adolescent and Parents Disagree on Medical Plan, Who Gets to Decide?

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Abbreviations:

DMD: Duchenne Muscular Dystrophy

BiPAP: bi-level positive airway pressure

PICU: pediatric intensive care unit

CRC: The Convention on the Rights of the Child

QOL: quality of life

LST: life-sustaining therapy

Table of Contents Summary: An adolescent with Duchenne Muscular Dystrophy wants a life-extending tracheostomy whereas parents do not want this for him.

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Dr. Hopkins conceptualized and drafted the introduction, case, and affiliated comment, and reviewed and revised the manuscript.

Dr. Ott conceptualized the ethical dilemma, drafted the affiliated comment, and reviewed and revised the manuscript.

Dr. Salih conceptualized the ethical dilemma, drafted her affiliated comment, and reviewed and revised the manuscript.

Dr. Bosslet reviewed drafts of the manuscript, contributed to writing, and reviewed and revised manuscript.

Dr. Lantos reviewed drafts of the manuscript, contributed to writing, and approved the final manuscript.

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Abstract/Introduction

Duchenne Muscular Dystrophy (DMD) is an X-linked autosomal recessive disease affecting 16 to 20 per 100,000 live births.^{1,2} It is characterized by progressive muscle weakness due to a defect in the dystrophin gene. It typically leads to loss of ambulation by age 8-14 years,¹ followed by cardiomyopathy and respiratory failure. Historically, adolescents with DMD died around 20 years of age.¹⁻³ As respiratory compromise occurs, patients are supported with non-invasive ventilation [e.g. nasal bi-level positive airway pressure (BiPAP)].³⁻⁶ When this becomes unsuccessful, they may be candidates for tracheostomy; this often happens in the second or third decade of life.⁷ The decision of whether or not to proceed with tracheostomy is complicated and is most often left to patient and family. Family members do not always agree.

We present a case in which acute illness forced a minor and his family to face this decision earlier than is typical. The adolescent desired a tracheostomy to extend his life. The parents did not believe that a tracheostomy was in his best interest and felt that comfort care was the most appropriate approach. Experts comment on the ethical issues raised by medical decision-making in cases involving adolescents and life-and-death decisions.

The case

A 15-year-old boy (B) with Duchenne Muscular Dystrophy was admitted to the pediatric intensive care unit (PICU) for respiratory distress. Prior to admission, B utilized a power wheelchair for mobility and was able to feed himself. He required total assistance for transfers to toilet and bathtub. He was attending high school and making average grades. He enjoyed watching movies, learning about cars, and listening to rap music.

B was diagnosed with septic shock due to influenza. He developed respiratory failure necessitating intubation and mechanical ventilation. He remained on the ventilator for weeks. A few weeks after intubation, the patient's negative inspiratory force remained nominal, indicating poor chest wall compliance. Extubation to BiPAP was attempted but the patient quickly developed respiratory failure requiring re-intubation. Another attempt the next week led to the same result. The progressive respiratory failure and failed extubation left only two options - tracheostomy with long-term ventilatory support or palliative care at home. Per best practice guidelines, these decisions were discussed with a multidisciplinary team and the family.⁴

Discussions were held with parents regarding whether tracheostomy placement and long-term ventilatory support were the best course forward. They focused on B's interests and based their preference on B's QOL and least amount of suffering for their son. After many conversations and family meetings, parents decided against tracheostomy and wanted to start arranging for hospice care.

Doctors then discussed options with B. They explained that a tracheostomy was possible and that it would mean more weeks in the hospital, a gastrostomy tube, and that complications included more frequent infections and hospitalizations. He was also told that he would likely need to be on a ventilator for the rest of his life. He understood that if he chose not to have surgery, he would be extubated and that he would be provided with medications to make him comfortable until he died. B asked that the tracheostomy be performed.

Given the father's request for hospice without tracheostomy and the adolescent's request for a tracheostomy, the ethical question became: should tracheostomy be performed?

Drs. Hopkins and Ott Comment – Supporting adolescent decision-making

Adolescents with DMD and their families will all face difficult decisions regarding ventilatory support as their disease progresses. Because of the recent advances in non-invasive ventilation, clinicians recommend that tracheostomy be avoided for as long as possible, and this is usually what patients prefer.⁴⁻⁷ A 2018 consensus report provided guidelines for when to opt for tracheostomy. The report recommended multiple trials of non-invasive ventilation and recommended tracheostomy only if these failed.⁴ Ideally, conversations regarding ventilatory support would occur prior to hospitalizations and situations requiring urgent decisions. In this case, B and his family faced this decision in a relatively urgent situation due to hemidiaphragmatic paralysis that precluded long-term use of non-invasive ventilation. Fourteen-year-old B indicated he wanted a tracheostomy and his parents believed that it was not in his best interest. We argue that B's decision should be supported based on the medical ethical principles of autonomy, beneficence, and justice.

Supporting Adolescents' Developing Autonomy. Recent neuroscience research provides nuance to our understanding of adolescent brain development.⁸⁻¹⁰ Medical decision-making capacity, as defined as understanding the health information, appreciating how the different choices would affect them personally, reasoning (balancing risks and benefits, logical flow) and ability to make a voluntary choice, is similar to adults by the age of 12-14 years.^{9, 11, 12} However, adolescents make decisions differently. Perceptions of risk, cognitive control, susceptibility to distraction and emotion, and ability to simultaneously consider multiple perspectives differ from adults.^{8, 10} These differences in decision-making capacity can influence health decisions, particularly in situations of high emotion and distraction.¹⁰

When appropriate time is provided for making a decision and when emotions are removed or controlled as much as possible, adolescents make decisions as logically as adults.⁸

To support an adolescent's developing autonomy, the medical team has a responsibility to create an environment that is as supportive as possible of the teen. In this case, the obvious emotional distress of making a life-and-death decision can never fully be removed.

Who Determines an Adolescent's Best Interest? The principle of beneficence is also at play in this case. A tracheostomy is a life-changing medical intervention with well described long-term risks. The decision of whether the benefits outweigh those risks will depend upon a judgment about the patient's anticipated QOL with the intervention. Will he be able to enjoy life on a ventilator? Children as young as four years of age can provide some information as to their health status and subjective health-related QOL.¹³ A 15-year-old who has been living with a progressive disease is certainly capable of making a decision based on his own QOL.

The ability of someone other than the individual in question to quantify a person's QOL is more variable. For example, reports by parents and children have been shown to be consistent with regard to physical aspects of life and less consistent with regard to emotional or social aspects, such as autonomy, social functioning, mood, or optimism about the future.^{13, 14} We cannot assume that the parents and adolescent will agree on QOL with and without a tracheostomy.¹³ Given the discrepancies between parent and child, we argue that the patient himself is the best person to determine if his QOL is worth the level of suffering and potential risks that accompanies the tracheostomy. Since he was able to express his desire for tracheostomy, this should be respected.

Justice and Disability. The ethics principle of justice has to do with treating individuals fairly and equally. It applies to children as well as adults. The United Nations Convention of the Rights of the Child (CRC) specifically states that "The Convention applies to all children, whatever their race, religion or abilities."¹⁵ There is no age criterion. The American Academy of

Pediatrics asserts that children who experience disability are at risk for having their rights ignored, and advocates for “equitable access to relevant health services for all children.”¹⁶

It is part of the healthcare team’s responsibility to be aware of this type of unfairness when guiding or participating in decisions of children with disabilities. As John Harris argued, “Life-saving resources should simply be allocated in ways which do not violate the individual’s entitlement to be treated as the equal of any other individual in the society: and that means the individual’s entitlement to have his interests and desires weighed at the same value as those of anyone else.”¹⁷

In this case, consideration of autonomy, beneficence and justice all point in the same direction. Since the patient is the only one who can truly determine his QOL with and without a tracheostomy and since he has the capacity to make an informed decision, the parents and healthcare team should respect his choice.

Dr. Zeynep Salih Comments – Recognizing the importance of the role of parents

The difference between B and his parents was unusual. B wanted to continue with life sustaining therapy (LST) but parents did not. Unless they are abusive or neglectful, parents are the default decision makers for minors in healthcare decisions, especially when a child cannot speak for himself/herself. Parents are expected to use the best interest standard as guidance. They should consider what would be good for the child and not factor in other outside interests.¹⁸

In this case, they came to the conclusion that a tracheostomy was not in B’s best interest. B’s own opinion was different from that of his parents. To further complicate the issue, this was a decision about a life sustaining therapy (LST) that would deeply affect the family as well. Chronic ventilator therapy is enormously burdensome on both the child and the family. It has

emotional, financial, and societal implications. Thus, a reasonable individual might wonder whether the parents should have the decisive vote in this case.

The best interests of parents and children in families are intertwined. Children are the most vulnerable individuals in the society and families are tasked with providing shelter, safety, food, basic education, and healthcare, so that children can reach their full potential. In other words, “parents have prerogatives that flow from the rights of their children”¹⁹. How does LST fit in this paradigm? Parents have their own individual rights to enjoy “Blessings of Liberty” as well; they are entitled to act without restraints.¹⁹ Based on these factors, parents may have the right to reject a tracheostomy and life-long mechanical ventilation for their son as too restrictive of their own personal liberties.

Parents are obligated to provide the basic needs of their children.²⁰ They are not obligated to consent for medical treatments that are experimental, treatments for which denial would not cause any of the child’s basic needs to fall below a certain threshold, or a treatment society would not want for every child.^{20, 21} This case raises the question of whether they have the right to refuse based not on the experimental nature of the treatment but, instead, based on the limits of what society can rightly obligate parents to do for their children.

When the child can communicate, the paradigm for decision-making for LSTs shifts to include the perspective of the child.²² If B has decision making capacity, he should have the right to make this decision for himself. For him, as for all people with serious illness, the illness may affect decision-making. We know that B is ill enough to be intubated and ventilated and, under those circumstances, is being asked to make a life or death decision. One may ask if it is fair to a 15-year-old to be the decision-maker in this situation. Westman argues that, “Children, especially adolescents need to be protected by their parents from consequences of unwise

decisions making during their minor years.”¹⁹ Is B’s decision to have a trach an unwise decision? If so, is it unwise for him? Or for his family?

Parents consider many factors while making such difficult decisions.^{24, 25} In this case, we do not have information on the family’s decision-making process to decline tracheostomy and home ventilation for their son and instead opt for comfort care. However, there is no evidence that B’s parents were making their decision on anything other than his best interest. I assume they have considered his prior wishes and how their son would have viewed his QOL on a ventilator.

I would suggest further discussions with the parents to clarify the basis for their decision. These discussions should consider how either decision would affect B’s family’s life and ultimately B’s life. These parents will need support no matter what decision is ultimately taken while taking care of B and this will not be the last time they would face the difficult decision of continuing LST for their son.

Dr. Gabriel Bosslet comments- DMD decision making is complicated and requires thoughtful decisional muddling

Decision-making in Duchenne’s (DMD) represents a unique departure from the usual difficulties posed by adolescent decision-making- there are several reasons for this. First, because DMD is an early onset childhood disease, the family unit has been the functional decisional agent throughout the disease process and remains so during, and often after, adolescence. Additionally, many men with DMD have cognitive dysfunction that calls into question the individual’s capacity for complex decision-making. Because of these issues, the decisional conundrum for clinicians and ethicists is not how to proceed with B’s care, but

whether or not to directly ask his preference in the face of life-threatening illness. While I think it ethically permissible to do so, we must understand that “to ask is to honor.” That is, it would be wrong to ask B his wishes if we were not going to respect the preferences that he expresses.

DMD is a disease that has dramatic consequences for a family unit. The usual cadence of adolescence as a time of growing independence is impaired (if not absent). Parents must be actively involved in the adolescent’s activities of daily living in a way that most other parents cannot even comprehend. This physical dependence represents an intrinsic limitation on what some have called “executive autonomy” (as opposed to “decisional autonomy”).²⁶ This impairment of executive autonomy means that the patient with DMD must rely on his caregivers for implementing his autonomous choices- they must acquiesce to those choices and agree that they are reasonable. Intertwine this with the fact that for most patients with DMD, the caregivers are the parents. They generally have strong feelings regarding what choices are right for their son. Deciding where autonomy lies gets, well, messy. Most parents in these situations will act in accordance with their son’s decisions (within reason), but it does require their assent. Full autonomy (decisional AND executive), therefore lies within the family unit rather than with the individual.

Patients with DMD have functional (although not global) cognitive impairment with lower IQs than age-matched peers.²⁷ They also have abnormalities in information processing, complex attention, and sequential executive functioning.²⁸⁻²⁹ This is important to realize when assessing a DMD patient’s capacity for decisional autonomy, especially in the setting of the respiratory distress and existential angst faced by B during his illness and discussion.

I am not arguing here that DMD patients should not be included in medical decisions. They should, and their input should generally carry the day. However, these conversations are

best carried out in a calm and thoughtful manner, preferably long before an acute illness complicates an already difficult decision.

In B's situation, the ethically difficult decision is whether or not to include him in the discussion of end-of-life plans. Given the family-centric nature of autonomy for patients with DMD, it is expected that his parents would share their hopes and worries, logistical and cost concerns, and the effect such a decision would have on the family unit openly and honestly, acknowledging that all of these are variables that are relevant with the path forward. But, since B and his parents had not previously discussed preferences and he was coherent and cooperative on the ventilator, it would have been nearly impossible to not include him. If he is included, then we must follow his wishes. It was not surprising that he did not want to forego life-sustaining treatment.

When, then, would it be acceptable to not discuss the plan with a patient in a situation analogous to B's? If B and his parents had previously had discussions in which he expressed wishes that either substantively outlined what he would want in the situation or (more likely) that he wished his parents to be his decision-makers, it would be reasonable to exclude him from the decision-making and only inform him of the decision (if he were coherent and able to understand the conversation).

The bottom line here is that adolescent decision-making is always complicated and is even more so in the setting of early onset childhood diseases. Perhaps the best we can do is muddle through as best we can – trying to balance family interests and adolescent autonomy.

Outcome of case

The parents ultimately agreed that B should be able to make the decision and they supported his desire for a tracheostomy. He underwent both tracheostomy and gastrostomy. After

some time for recovery, the patient underwent speech studies and was eventually able to eat under supervision. He was discharged to home with home health services two months after the surgeries. B remained at home for six months free from serious infections. Unfortunately, one evening, he was found unresponsive. B was unable to be resuscitated and passed away.

John D. Lantos comments

This case is difficult because there is a conflict between what is best for the child and what is best for the family. Clearly, tracheostomy and mechanical ventilation are reasonable choices in these circumstances. Many teens and young adults with DMD choose that course of treatment and claim that mechanical ventilation was “the best thing that had happened to them.”³⁰ So, B’s decision makes sense.

The parents’ concerns may also be valid. We don’t know the basis for their decision, and need to find out, but they may be thinking both of B’s best interests but also of what is best for their family. No family should be obligated to provide home ventilator care for a teen. But such care could be provided either in skilled nursing facilities or in a foster home. Alternatively, as occurred in this case, parents can be supported with home nursing and intermittent respite care to give them needed breaks from the round-the-clock responsibilities of caring for a child on a ventilator.³¹

The key to the case is, as much as possible, to separately analyze B’s interests and decisional capacity and the parents’ and family’s interests.³² To the extent that B has decisional capacity, his wishes must prevail.

References

1. Ryder S, Leadley RM, Armstrong N, et al. The burden, epidemiology, costs and treatment for Duchenne muscular dystrophy: an evidence review. *Orphanet J. of Rare Dis.* 2017;12(79).
2. Birnkrant D, Bushby K, Bann CM, et al. Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management. *Lancet Neurol.* 2018;17(3):251-267.
3. Passamano L, Taglia A, Palladino A, et al. Improvement of survival in Duchenne Muscular Dystrophy: retrospective analysis of 835 patients. *Acta Myol.* 2012;XXXI:121-125.
4. Birnkrant D, Bushby K, Bann CM, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management. *Lancet Neurol.* 2018;17(4):347-361.
5. Bach J, Martinez D. Duchenne Muscular Dystrophy: Continuous Noninvasive Ventilatory Support Prolongs Survival. *Respir Care.* 2011;56(6):744-750.
6. McKim D, Griller N, LeBlanc C, et al. Twenty-four hour noninvasive ventilation in Duchenne muscular dystrophy: A safe alternative to tracheostomy. *Can Respir J.* 2013;20(1):e5-e9.
7. Bach J, Goncalves MR, Hon A, et al. Changing Trends in the Management of End-Stage Neuromuscular Respiratory Muscle Failure. *Am J Phys Med Rehabil.* 2013;92:267-277.
8. Blakemore S, Robbins TW. Decision-making in the adolescent brain. *Nat Neurosci.* 2012;15(9):1184-1191.
9. Weithorn L, Campbell SB. The Competency of Children and Adolescents to Make Informed Treatment Decisions. *Child Dev.* 1982;53:1589-1598.
10. Steinberg L. Does Recent Research on Adolescent Brain Development Inform the Mature Minor Doctrine? *J Med Philos.* 2013;38:256-267.
11. Nelson LR, Supiansky NW, Ott MA. The Influence of Age, Health Literacy, and Affluence on Adolescents' Capacity to Consent to Research. *J Empir Res Hum Res Ethics: JERHRE.* 2016;11(2):115-21.
12. Hein IM, De Vries MC, Troost PW, Meynen G, Van Goudoever JB, Lindauer RJ. Informed consent instead of assent is appropriate in children from the age of twelve: Policy implications of new findings on children's competence to consent to clinical research. *BMC Med Ethics.* 2015;16(1):76.
13. Matza L, Swensen AR, Flood EM, et al. Assessment of Health-Related Quality of Life in Children: A Review of Conceptual, Methodological, and Regulatory Issues. *Value Health.* 2004;7(1):79-92.
14. Eiser C, Morse R. Can Parents Rate Their Child's Health-Related Quality of Life? Results of a Systematic Review. *Qual Life Res.* 2001;10(4):347-357.
15. UN Committee on the Rights of the Child (CRC), *General comment No. 9 (2006): The rights of children with disabilities*, 27 February 2007, CRC/C/GC/0, available at: <https://www.refworld.org/docid/461b93f72.html> [accessed 6 December 2018].
16. Council on Community Pediatrics and Committee on Native American Child Health. Health Equity and Children's Rights. *Pediatrics.* Apr 2010, 125 (4) 838-849. DOI: 10.1542/peds.2010-0235.
17. Harris J. QALYfying the value of life. *J Med Ethics.* 1987;13:117-123.
18. Beauchamp T, Childress JF. *Principles of Biomedical Ethics.* New York: Oxford University Press; 2009.

19. Westman, J.C. Children's Rights, Parents' Prerogatives, and Society's Obligations. *Child Psychiatry Hum Dev*. 1999; 29: 315-328.
20. Ross LF. *Children, Families, and Health Care Decision Making*. New York: Oxford University Press; 1998.
21. Goldstein J, Freud A, Solnit AJ. *Before the best interests of the child*. New York: The Free Press; 1979.
22. Weiss KL, Okun, AL, Carter BS, et al. Guidance on Foregoing Life-Sustaining Medical Treatment. *Pediatrics*. 2017;140(3):e20171905
23. Walter J, Ross LF. Relational Autonomy: Moving Beyond the Limits of Isolated Individualism. *Pediatrics*. 2014;133:S16-S23.
24. Sharman M, Meert K, Sarnaik A. What Influences Parents' Decisions To Limit or Withdraw Life Support? *Pediatric Critical Care Medicine*; 2005; 6(5):513-518
25. Carroll KW, Mollen K, Aldridge S. et al. Influences on Decision Making Identified by Parents of Children Receiving Pediatric Palliative Care. *AJOB Prim Res*. 2012; 3(1): 1-7.
26. Naik A, Dyer CB, Kunik ME, et al. Patient autonomy for the management of chronic conditions: a two-component reconceptualization. *Am J Bioeth*. 2009;9(2):23-30.
27. Bresolin N, Castelli E, Comi GP, et al. Cognitive impairment in Duchenne muscular dystrophy. *Neuromuscul Disord*. 1994;4(4):359-369.
28. Perumal A, Rajeswaran J, Nalini A. Neuropsychological profile of duchenne muscular dystrophy. *Appl Neuropsychol Child*. 2015;4(1):49-57.
29. Wicksell R, Kihlgren M, Melin L, et al. Specific cognitive deficits are common in children with Duchenne muscular dystrophy. *Dev Med Child Neurol*. 2004;46(3):154-159.
30. Dreyer PS, Steffensen BF, Pedersen BD. Life with home mechanical ventilation for young men with Duchenne muscular dystrophy. *J Adv Nurs* 2010; 66:753-62.
31. Meltzer LJ, Boroughs DS, Downes JJ. The relationship between home nursing coverage, sleep, and daytime functioning in parents of ventilator-assisted children. *J Ped Nurs* 2010;25:250-57
32. Lindemann H. Why families matter. *Pediatr* 2014;134:Suppl 2: S97-103.