Difficult to Swallow: Epidermolysis Bullosa, Esophageal Stricture, and the Boundaries of Forgoing Medical Nutrition and Hydration

William Sveen and Nneka Sederstrom

ABSTRACT

A child with epidermolysis bullosa that caused worsening esophageal stricture raised questions about the boundaries of forgoing medical nutrition and hydration (MNH). After pursuing multiple therapies to extend their child's life, the parents considered the increasingly burdensome treatment options unacceptable and asked to stop medical nutrition. Although forgoing MNH is similar to other life-sustaining medical treatments and an acceptable option for parents of children with severe, life-limiting illnesses, most of the literature focuses on forgoing MNH in children who are actively dying, lack appetite, or have total intestinal failure. However, the ethics team determined that forgoing MNH was acceptable because the child had a life-limiting illness and significant burdens were associated with continued existence and available treatments. Various objections are considered.

William Sveen, MD, MA, is an Assistant Professor in the Department of Pediatrics, Division of Pediatric Critical Care Medicine, at the University of Minnesota, in Minneapolis, Minnesota. *wsveen@umn.edu*
Nneka Sederstrom, PhD, MPH, MA, FCCP, FCCM, is the Chief Health Equity Officer at Hennepin Healthcare, Minneapolis. *nneka.sederstrom@hcmed.org*

CASE

The parents of a four-year-old child with recessive dystrophic epidermolysis bullosa and severe esophageal stricture and gastrostomy tube dependence requested an ethics consultation to discuss withdrawing medical nutrition and hydration and allowing their child to die. The patient received an investigational bone marrow transplant in infancy, with an older sibling as a donor. The transplant engrafted successfully and improved the healing time of injured skin, but was not fully curative. The patient continued to have blistering lesions covering large portions of their body. The patient required frequent, painful dressing changes, became dependent on opioids due to chronic pain, and developed multiple joint contractures.

The esophageal stricture occurred prior to transplantation and, unfortunately, progressed after transplantation. After multiple esophageal dilations and stents, the esophagus atrophied to the point of risking perforation with further procedures. Swallowing caused significant pain and anxiety, and eventually panic attacks that occurred multiple times daily for minutes to hours. The patient became dependent on benzodiazepines due to anxiety about the pain of swallowing, despite aggressive symptom management with palliative care and psychiatry. All nutrition was obtained by MNH via gastrostomy tube, but the child occasionally took small volumes orally for taste and comfort. The family saw multiple specialists across different health systems to discuss medical and surgical therapies, but they were not satisfied with the suggested options, that included esophageal reconstruction, esophageal transplant, tracheal diversion with tracheostomy, and increasing sedating medications.

Despite pursuing aggressive and investigational therapies to extend their child's life, the parents considered the current burdens of their child's care to be worse than the benefits, and requested an ethics consult to discuss withdrawing MNH to allow their child to die. The ethics team determined that the parents' request was reasonable. The patient was enrolled in hospice, scheduled MNH was discontinued, and instead MNH was provided orally or per gastrostomy for comfort at the patient's request only. The patient had a steady decrease in the amount of requested nutrition over the course of six weeks and died at home surrounded by family. Anxiety and the use of benzodiazepine associated with saliva management decreased until the week prior to death, when the child required increased anxiolysis. Child and family life specialists and social workers were closely involved to help the child, older siblings, and parents through the dying and bereavement process.

DISCUSSION

Allowing parents to make life and death decisions on behalf of their child is largely consistent with deference toward parental authority throughout pediatric medical treatment. Parents have the responsibility to raise children in the context of their family. They have to live with the consequences of their decisions for the child and family in a more intimate way than any clinical provider does. They also know their child, know their family, and have a stronger affection for both than clinical providers do, making parents likely to be acting in what they perceive to be the best interest of their child.[1]

The best interest principle is a difficult standard to require of parents, since it is inherently subjective. Different parents may reasonably decide what is best for their children based on their strongly held values. Parents also have to balance the interest of their child with the interest of their other children and their family. For example, for parents to focus all of their emotional and financial resources on a hospitalized child while they ignore the needs of other children is not in the best interest of the entire family. Furthermore, we do not revoke parental decision making when parents make a decision that objective data suggest is not in the best interest of the child, for example, exposure to secondhand smoke.

Determining when to intervene and limit parent decision-making authority typically requires establishing a different threshold of minimal best interest or harm, often considered to be abuse or neglect. A commonly used standard is the harm principle, that more objectively states that parents must harm a child with a decision before their ability to make the decision can be revoked.[2] Others have incorporated versions of both the guidance principle of best interest and the intervention principle of harm reduction into a single concept such as constrained parental autonomy or the zone of parental discretion.[3]

Regardless of the philosophical framework used, the American Academy of Pediatrics (AAP) consistently defers to parents regarding decisions about whether to withdraw or withhold life-sustaining medical treatment (LSMT) such as mechanical ventilation, renal replacement therapy, vasopressors, and so on, for children with severe, life-limiting illness. The AAP considers foregoing MNH as similar to foregoing other LSMT, while it recognizes that the emotional and social associations with eating and feeding often make foregoing MNH subjectively feel different to families and clinicians.[4] A 2009 policy statement by the AAP describes situations when the burden of treatment may outweigh the benefits, making foregoing MNH an option for families.[5] These include patients who are irreversibly comatose and who will not experience the dying process, patients who are terminally ill and lack appetite, and patients with total intestinal failure that requires high morbidity and mortality treatments such as indefinite total parenteral nutrition (TPN) or bowel transplant.

This AAP policy briefly discusses "other conditions that are incompatible with long-term survival and for which significant burden is associated with continued existence or available treatment options." However, the AAP policy then gives the example of infants who have uncompensated heart failure with an inability to tolerate sufficient fluid volumes, which make MNH actually harmful to the child. There is no discussion regarding children for whom MNH continues a burdensome existence, but the MNH itself does not cause the harm.[6] However, since forgoing MNH is considered similar to other LSMT, a reasonable parent may consider the burden of continued existence to outweigh the benefits of MNH. These burdens include not only physical pain but restriction of activity, fear, anxiety, isolation, and other forms of emotional distress that do not need to be directly caused by MNH.[7]

The AAP considers foregoing MNH as similar to foregoing other LSMT, while it recognizes that the emotional and social associations with eating and feeding often make foregoing MNH subjectively feel different to families and clinicians.

The treatments available to the child in this case all had limited benefits or significant harms. Increased salivary management such as botulinum toxin salivary injections and salivary gland removal were unlikely to provide much relief, since reducing saliva production with anticholinergic medication would not significantly reduce anxiety, as the child found even a small amount of saliva painfully difficult, and swallowing it provoked anxiety. A central line for indefinite TPN would have reduced but not eliminated the need to swallow saliva, and would introduce new risks and the burdens of more intensive care, including central-line-associated bloodstream infections for a child who had impaired skin integrity. A tracheal diversion with tracheostomy would have eliminated the need to swallow any saliva, but would have been highly invasive, would have removed the child's ability to speak, and would have required care associated with a tracheostomy. Increasing sedative medication to reduce anxiety might have reduced symptoms, but might have further adverse effects on the child's quality of life.

None of the above treatments would have fixed the child's esophageal structures. Options to repair a severely stenotic and atrophied esophagus are investigational, and have a high degree of failure in children who have normal connective tissue healing. These include esophageal transplant and esophageal reconstruction. Of note, none of the possible treatments would relieve the physical pain of epidermolysis bullosa.

The parents made a reasonable assessment that the burden outweighed the benefits of continued treatment, making this case an example in which MNH was withdrawn due to the significant burden of continued existence. This young child lived at the uncomfortable intersection of incurable pathophysiology and significant daily distress, despite aggressive palliative care. The patient had intestinal failure due to the esophageal stenosis, but the ability to absorb food enterally via gastrostomy made it less severe than the total intestinal failure cited in the AAP guidelines. Few would argue that these parents were obligated to have their child undergo esophageal reconstruction or intestinal transplant, since these measures are highly invasive and have a high risk of failure, and so potentially cause harm.

OBJECTIONS CONSIDERED

The Child Was Not Terminally or Critically Ill

Many discussions about forgoing LSMT occur when patients are terminally ill or experience an acute decompensation and require critical care. Clinicians may have been more comfortable with the parents' decision in this case, as there was no obligation to continue burdensome life-sustaining medical treatment given their child's state of health.

The parents considered waiting for their child to become ill and then to forego any increase in treatment. They described a scenario in which their child had a skin infection or aspiration leading to sepsis, and then no antibiotics or life-sustaining medical treatment would be provided. However, they determined this would prolong the current suffering of their child, lead to increased suffering with acute decompensation, and limit the family's ability to control aspects of the child's death such as dying at home surrounded by family.

The Child Was Neurotypical and Might Starve

Some of the patient's clinicians were concerned that since the patient was developmentally typical, the child would experience significant distress at the end of life compared with those who are minimally conscious or comatose. However, the parents noted the opposite was also true, using the same reasoning: if the patient had more to lose in death, they certainly also experienced more suffering in continued life.

Others were concerned about obtaining assent to the treatment and to what degree the patient could refuse withdrawal of MNH. The parents actively sought assent from the patient. They informed the patient in a developmentally appropriate way, using child life specialists, that they intended to stop scheduled feeds and that the child would soon die but be kept comfortable. The child was also given the option to request food or liquids orally or enterally whenever they desired, which gave the child control in a situation that previously left the child feeling powerless. They ensured that their child's values and wishes were heard, and, in this case, the patient was relieved to discontinue MNH and assented to comfort care, allowing natural death. Typically, reasonable parents are considered to be most suited to determine the best interest of a young child, even for serious decisions. But in matters of life and death, proceeding against the explicit wishes of a child would be ethically problematic and psychologically damaging to all involved.

Specific concerns were raised that the child would be "starved to death." *Starvation* is a provocative term that connotes symptoms of long-term protein malnutrition such as muscle wasting, edema, infections, and skin breakdown.[8] While the process of providing nutrition has strong emotional, social, and familial associations that must be addressed, palliative care can address the symptoms associated with starvation in such a way that patients who die from foregoing MNH do not experience the grotesque symptoms of starvation. Due to the stigma surrounding starvation and the inaccuracies in describing the actual process of death, AAP policy recommends the term not be used.[9]

The Child May Require Palliative Sedation and Euthanasia

Some clinicians were concerned that aggressive symptom management would result in palliative sedation, which could be construed as euthanasia. *Palliative sedation* is the use of medications to decrease awareness of severe refractory symptoms at the end of life. Palliative sedation may hasten death in some situations, but not all, and is not considered a cause of death when the intent is to treat symptoms.[10] This patient would have been a candidate for palliative sedation if the proportional increase in medications required to adequately treat symptoms resulted in a comatose state. However, although the child needed an increase in benzodiazepines in the last week of life, full sedation was never required, which further demonstrated that children who die from foregoing MNH can avoid the symptoms associated with starvation.

Euthanasia is an active means of causing death to relieve suffering, typically administered by a caretaker or medical professional. Euthanasia is distinct from a passive means of allowing natural death such as foregoing MNH, regardless of whether palliative sedation is involved. That the patient was able to request food or liquids for comfort further distanced this case from actively causing death.

CONCLUSION

This case illustrates the need for further discussion on foregoing MNH in children with life-limiting illness and burdensome treatment. When limited options exist to correct the underlying cause of suffering, symptoms remain refractory to aggressive palliation, and access to multidisciplinary palliative care at the end of life is available, reasonable parents may decide to allow natural death by foregoing MNH. The option to forego MNH should be discussed with the families of patients who experience a burdensome continued existence, and their reasonable decisions to forego MNH should be respected.

BLINDING OF THE CASE

Details of this case were changed to protect the privacy of the family.

NOTES

1. C.L. Cummings and M.R. Mercurio, “Ethics for the pediatrician: Autonomy, beneficence, and rights,” *Pediatric Review* 31, no. 6 (June 2010): 252-5.

2. D.S. Diekema, “Parental refusals of medical treatment: The harm principle as threshold for state intervention,” *Theoretical Medicine Bioethics* 25, no. 4 (2004): 243-64.

3. L. Gillam, “The zone of parental discretion: An ethical tool for dealing with disagreement between parents and doctors about medical treatment for a child,” *Clinical Ethics* 11, no. 1 (2016): 1-8; L.F. Ross, “Better than Best (Interest Standard) in Pediatric Decision Making,” *The Journal of Clinical Ethics* 30, no. 3 (Fall 2019): 183-95.

4. K.L. Weise et al., “Guidance on Forgoing Life-Sustaining Medical Treatment,” *Pediatrics* 140, no. 3 (September 2017): e20171905, doi: 10.1542/peds.2017-1905.

5. D.S. Diekema, J.R. Botkin, and the AAP Committee on Bioethics, “Forgoing Medically Provided Nutrition and Hydration in Children,” *Pediatrics* 124, no. 2 (August 2009): 813-22.

6. Ibid.

7. Weise et al., “Guidance on Forgoing,” see note 4 above.

8. J.C. Ahronheim and M.R. Gasner, “The sloganism of starvation,” *Lancet* 335, no. 8684 (3 February 1990): 278-9.

9. Diekma and Botkin, “Forgoing medically provided nutrition,” see note 5 above.

10. H. ten Have and J.V. Welie, “Palliative sedation versus euthanasia: An ethical assessment,” *Journal of Pain Symptom Management* 47, no. 1 (January 2014): 123-36.