

2018 AAP and APA Ethics Essay Contest - 1st Place

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

William Sveen, MD, MA

Case

The parents of a four-year-old girl with recessive dystrophic epidermolysis bullosa and severe esophageal stricture and gastrostomy tube dependence request an ethics consultation to discuss withdrawing medical nutrition and hydration (MNH) and allowing their daughter to die. The family relocated from Europe when the patient was an infant to under-go an investigational bone marrow transplant 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 her body. She required frequent, painful dressing changes and became dependent on opioids due to chronic pain. She also developed multiple joint contractures.

The esophageal stricture occurred prior to her transplant, and unfortunately she experienced progressive symptoms after her transplant. After multiple esophageal dilations and stents, her esophagus atrophied to the point of risking perforation with further procedures. Swallowing caused significant pain and anxiety and eventually panic attacks which occur multiple times daily for minutes to hours. She became dependent on benzodiazepines due to the anxiety of swallowing despite aggressive symptom management with palliative care and psychiatry. All nutrition was obtained by MNH via gastrostomy tube but she occasionally took small volumes orally for taste and comfort. The family has seen multiple specialists across different health systems to discuss medical and surgical therapies but was not satisfied with suggested options including esophageal reconstruction, esophageal transplant, salivary fistulas, tracheal diversion with tracheostomy, and increasingly sedating medications.

Her parents state they are "doing more to her than for her" and despite pursuing aggressive and experimental therapies to extend her life in the past, they are now considering withdrawing MNH to allow her 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 by patient request only. The patient had a steady decrease in the amount of requested nutrition over the course of six weeks and died in her home surrounded by family. She had decreased anxiety associated with saliva management and decreased benzodiazepine use until the week prior to her death when she required increased anxiolysis. Child and family life specialists and social workers were closely involved to help the patient, her older siblings, and parents through the dying and bereavement process.

Discussion

The American Academy of Pediatrics (AAP) considers fore-going MNH as similar to foregoing other life-sustaining medical treatment (LSMT) such as mechanical ventilators, renal replacement therapy, pressors, etc. while recognizing that the emotional and social associations to eating and feeding often make foregoing MNH subjectively feel different to families and clinicians.¹ The AAP 2009 policy statement describes situations where the burden of treatment

may outweigh the benefits, making foregoing MNH an option for families. These include patients who are irreversibly comatose and do not experience the dying process, patients who are terminally ill and lack appetite, and patients with total intestinal failure requiring high morbidity and mortality treatments such as indefinite TPN or bowel trans-plant.²

The policy briefly discusses “other conditions that are in-compatible with long-term survival and for which significant burden is associated with continued existence or available treatment options.” However, the paper then gives the ex-ample of infants with uncompensated heart failure with an inability to tolerate sufficient fluid volumes, making MNH actually harmful to the child. No discussion is made regarding children in whom MNH continues a burdensome existence but the MNH itself does not cause the harm.² 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 activity restriction, fear, anxiety, isolation, and other forms of emotional distress and do not need to be directly caused by MNH.¹

This young girl 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 her ability to absorb food enterally via gastrostomy makes it less severe than total intestinal failure cited in the guidelines. Few would argue that the parents were obligated to have her undergo esophageal reconstruction or intestinal transplant since these measures are highly invasive and have a high risk of failure and potential to increase harm.

Surgical salivary management such as fistulas and tracheal diversion with tracheostomy or increasing sedative medication may reduce symptoms but have further adverse effects on her quality of life. Furthermore, no treatment would relieve the physical pain associated with the underlying and progressive cause of epidermolysis bullosa. The best interest of a child is highly subjective, but typically parents are best suited to determine between reasonable options.¹ 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.

Objections Considered

She is not terminally or critically ill. Many discussions about forgoing LSMT occur when patients are terminally ill or experiencing an acute decompensation and requiring critical care. While clinicians may be more comfortable with the parents’ decision in this context, there is no obligation to continue burdensome care even if she remains at her cur-rent state of health.

The parents did consider waiting for the patient to become ill and foregoing any increase in treatment at that time. They described a scenario in which she could have a skin infection or aspiration leading to sepsis and then no antibiotics or pressors would be provided. However, they deter-mined this would prolong her current suffering, lead to in-creased suffering with her acute decompensation, and limit the family’s ability to control aspects of her death such as dying in her room with her sisters present.

She has typical development and may starve. Some of the patient’s clinicians were concerned that since she is developmentally typical, she would experience significant dis-tress at the end of life compared with those who are minimally conscious or comatose. However, her parents noted the opposite was also true using the same reasoning. If she had more to lose in death, she certainly also experienced more suffering in continued life.

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 break-down.³ While the process of providing nutrition has strong emotional, social, and familial associations that need to 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, the AAP recommends that the term not be used.²

Others were concerned about the need to obtain her assent to the treatment and to what degree she should be able to refuse withdrawing MNH. The parents informed the patient in a developmentally appropriate way using child life specialists that the scheduled feeds were stopping and that she would soon die but be kept comfortable. She was also given the option to request food or liquids orally or enterally whenever she wanted which gave her back control in the situation that previously left her feeling powerless. They ensured that her values and wishes were heard, and in this case, the child was relieved to discontinue MNH. Even if she had disagreed, reasonable parents are typically considered most suited to determine the best interest of a young child, even in serious decisions.¹ However, in matters of life and death, these decisions need a high level of scrutiny, which is why the parents requested an ethics consult.

She 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.⁴ This patient would be a candidate for palliative sedation if the proportional increase in her medications required to adequately treat her symptoms resulted in a comatose state. However, although she needed an increase in benzodiazepines in the last week of life, full sedation was never required for this patient, further demonstrating that children who die from foregoing MNH can avoid 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 passive means of allowing natural death such as foregoing MNH, regardless of whether palliative sedation was involved. Allowing the patient to request food or liquids for comfort further distanced her case from others actively causing her death.

Conclusion

This case illustrates the need for further discussion on fore-going 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 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 families of patients who experience a burdensome continued existence and reasonable decisions to forego MNH should be respected.

References

1. Weise KL, Okun AL, Carter BS, Christian CW; Committee on Bioethics; Section on Hospice and Palliative Medicine; Committee on Child Abuse and Neglect. Guidance on Forgoing Life-Sustaining Medical Treatment. *Pediatrics*. 2017 Sep;140(3).

2. Diekema DS, Botkin JR; Committee on Bioethics. Forgoing medically provided nutrition and hydration in children. *Pediatrics*. 2009 Aug;124(2):813-22.
3. Ahronheim JC, Gasner MR. The sloganism of starvation. *Lancet*. 1990 Feb 3;335(8684):278-9.
4. ten Have H, Welie JV. Palliative sedation versus euthanasia: an ethical assessment. *J Pain Symptom Manage*. 2014 Jan;47(1):123-36