

Virtual Mentor

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CASE AND COMMENTARY

Palliative Care for an Infant with Short Bowel Syndrome and Advanced Liver Disease, Commentary 2

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Case

After 5 months of routine NICU Care, and treatment for malabsorption and malnutrition, Mary was discharged from the NICU and allowed to go home with her parents. Eight hours after discharge, her parents brought her back to the hospital with fever and vomiting. During this second hospitalization, Mary had bacterial and fungal infections, multiple changes in her vascular access sites, and complications, including advanced liver disease, from the total parenteral nutrition (TPN). Her liver dysfunction was characterized by abnormal coagulation, only partially corrected by blood products and vitamin K, hypoalbuminemia, and hypoproteinemia. She bled from her nose and mouth after crying or sneezing, and extensively from her ostomy site. Mary had multiple episodes of hypovolemic shock that required blood transfusions. Her massive hepatosplenomegaly interfered with respiration.

Early in her NICU stay Mary's physicians discussed with the Janes the possibility of transporting Mary to another medical center for an intestinal transplant. Mary's parents appeared to understand the seriousness of their daughter's condition and wanted the doctors to do "everything possible" for her. Mary's liver dysfunction progressed and she became more edematous, had skin breakdown, and had to be more frequently volume resuscitated and transfused. Her tenuous condition now made it impossible to consider moving her to another location for a transplant. Mary's physicians considered her condition terminal and could see that she was suffering.

Dr. Andrews and her colleagues tried to talk to the Janes about palliative care and the imminent death of their daughter. In one instance Dr. Andrews approached Mrs. Jane, who never left the hospital unless her husband or mother came to relieve her, but Mrs. Jane stopped Dr. Andrews in mid-sentence.

"I see where you're going with this, Dr. Andrews, but my baby is strong. Children are resilient. Mary got well enough to go home once and she'll do it again, we've just got to give her a little time."

*The patient's name has been changed to protect her privacy and that of her parents.

Commentary 2

Mary Jane was indeed a resilient infant, and, over the course of her short life, her parents, particularly her mother, not unexpectedly bonded with her and likely contributed to that resilience. Unfortunately her short bowel syndrome and advanced liver disease made it impossible to nourish her adequately by either parenteral or enteral means. Intestinal transplant was considered but quickly became an unreasonable option in light of the severity of Mary's liver disfunction. Even in the best of circumstances, however, likelihood of 1-year survival after intestinal transplant is only approximately 50 percent,¹ making the decision to operate a difficult choice for parents and a difficult recommendation for physicians to make.

In this infant's case other choices were equally difficult, and ethical discussions began to assume as much of a role in her care as the medical discussions. Nonmaleficence was the first consideration, given that the total parenteral nutrition, initiated to sustain the child's life, was ultimately hastening her death by destroying her liver. Withholding this therapy was therefore ethically permitted if not obligated.²⁻³

The physicians, not the parents, ultimately concluded that the greatest benefit to Mary, and the most appropriate therapy, was to offer comfort care. In this they were supported by the American Academy of Pediatrics' Committee of Bioethics which in 1994 concluded that "continuing non-beneficial treatment harms many patients and may constitute a legal, as well as moral, wrong."⁴

As consideration of the appropriateness of palliative care continued, some members of Mary's health care team wondered whether it might be illegal to withdraw nutrition and hydration, citing the 1984 Department of Health and Human Services amendments to the Child Abuse Prevention and Treatment Act. These regulations, commonly known as the "Baby Doe Regulations," require that, except under certain specified conditions, all newborns receive maximal life-prolonging treatment. While still criticized by some, the Baby Doe Regulations continue to influence decision making for terminally ill newborns.⁵⁻⁶ There is clear consensus, however, that withdrawal of artificial nutrition and hydration when they are more harmful than helpful and when the child's parents or legal guardians are in agreement, is fully acceptable and justifiable both legally and morally.

For a variety of reasons Mary's parents (and others in similar circumstances) were unwilling to consider palliative care. Health professionals are often unwilling to embark on this course due to concern about hastening death. Ethical consideration of aggressive palliation often includes a discussion of the principle of double effect.⁷ This principle, which stems from the moral theology of Thomas Aquinas, states that an action with both a good and bad effect is ethically permissible if the following conditions are met:

1. The action itself is morally good or at least indifferent.
2. Only the good effect is intended (even though the bad or secondary effect may be foreseen).
3. The good effect must not be achieved by way of the bad.
4. The good effect must outweigh the bad.

Often the most difficult aspects of initiating such an approach to care is reluctance on the part of health professionals,⁸ and the lack of understanding by patients, parents, or relatives. It is of course not a decision to be made lightly, and in the case of Mary Jane it is clear that additional discussion was required between Dr. Andrews and the parents before such an approach could be accepted.

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