

## CASE STUDY

### Imperiled

Minutes after arriving at the hospital, a pregnant woman is told by her obstetrician that "the baby is in trouble." The placenta had begun prematurely to separate from the uterus wall, thereby cutting off the baby's supply of blood and oxygen. An emergency C-section was performed.

On the baby's seventeenth day of life, the neonatologist explained that the feeding program was not going well. X-ray revealed that portions of the intestine were dying from lack of adequate blood supply. The doctor said that emergency surgery was necessary at a neonatal intensive care unit at another hospital.

In the first surgery, two large sections of her small intestine were removed; other adjoining areas had also been damaged and repair attempts were made. The surgeon could not predict how much of the remaining intestine would survive. Meanwhile, she was to be treated for sepsis and shock. A later surgery was necessary to remove additional pieces of dead intestine.

A third operation revealed that yet another section of intestine had died. Moreover, surgery left her without an ileocecal valve, the muscle that controls passage of fluid from the small to the large intestine. The pediatric surgeon explained to the parents that there was not sufficient intestine remaining for the baby to survive without feeding by vein. He said that babies could be fed by vein for a long time, and that a few children had survived until ten years of age. Following several weeks of consultation with surgeons and gastroenterologists around the country, the parents learned that the only hope for their daughter's survival was through feeding by vein.

The parents struggled with difficult questions: Were they morally obligated to their daughter to continue treatment? Since she could live indefinitely with a catheter, could they decide to begin experimental therapy? Could feeding by vein be discontinued if it was the only way to provide nourishment? All these questions became horribly real when the baby again developed sepsis and shock; the parents had been informed that this was "a predictable complication" associated with a permanent venous catheter. Every time the sepsis and shock recurred, a new location for the catheter would have to be found. The parents, along with the baby's nurse and neonatologist, decided not to treat the infection, to "let her go, now." The Infant Care Review Committee convened in emergency session.

*Adapted from a case study by William G. Bartholome, M.D., M.T.S.*

## ANALYSIS: The "Clinical Moment" in Short Bowel Syndrome: What Can We Do, What Should We Do?

The parents of Baby Girl X face a tragic circumstance: imminent death from sepsis in an infant with extreme Short Bowel Syndrome, complete dependence upon total parenteral nutrition (TPN), and progressive liver failure. The physicians of Baby Girl X must provide her parents with the available treatment options, risks and benefits of each therapy, and guidance through the decision-making process. Three issues require discussion in this case: (1) Does institution of TPN in neonates with extreme Short Bowel Syndrome constitute established or experimental therapy? (2) Which therapeutic options for Baby Girl X are ethically defensible, and medically appropriate? and (3) Can TPN be withheld or withdrawn from neonates with extreme Short Bowel Syndrome?

The Short Bowel Syndrome in infants results from a substantial loss of small intestine caused by congenital or acquired conditions. Baby Girl X developed the Short Bowel Syndrome as a consequence of an acquired, devastating intestinal illness of sick premature infants, necrotizing enterocolitis. Inadequate small intestinal length leads to malabsorption of nutrients, diarrhea, weight loss, and protein-calorie malnutrition. Fortunately, most infants who sustain small intestinal loss have enough remaining bowel to eventually achieve normal or near-normal intestinal function. For such infants, TPN is a temporary therapy in which a specialized intravenous catheter delivers a solution con-

taining sufficient calories and fluids for normal weight gain. The infant's remaining small intestine undergoes a complex process of adaptation, in which it acquires the ability to absorb nutrients almost as efficiently as small intestine of normal length. The period of time for the remaining bowel to achieve complete adaptation depends upon the length and quality of the intestine and the presence of the ileocecal valve. Infants whose remaining small intestinal length is at least 15-20 percent of normal can be expected to eventually achieve adaptation and eat a normal diet. However, the process of adaptation may take months or years in some infants, during which time TPN is the major source of nutrition. Institution of TPN has become established therapy for infants with Short Bowel Syndrome and sufficient remaining small intestine for eventual intestinal function.

While TPN has eliminated malnutrition as the major morbidity of the Short Bowel Syndrome, it can directly or indirectly cause most of the complications. The specified intravenous catheter that delivers the TPN solution can become infected, causing life-threatening systemic sepsis, or induce thrombosis of the central veins. Progressive liver disease, the most serious complication of prolonged TPN, can result in end-stage biliary cirrhosis and death.

Survival of infants with Short Bowel Syndrome is estimated to be between 75 and 85

percent with most patients weaned off TPN by two years of age. The majority of infants who are successfully treated by prolonged TPN attain normal growth parameters and developmental performance.

At the third operation when Baby Girl X was determined to have near-total intestinal loss, her prognosis changed from having a good potential for intestinal adaptation to dismal prospects for a functional intestinal tract. The length of her remaining small intestine was insufficient and even an extended period of adaptation would not be expected to allow absorption of nutrients. In essence, Baby Girl X became a "gastrointestinal cripple"; a patient with virtually no hope of having a normal intestinal tract. Treatment with TPN for Baby Girl X changed from being a supportive, temporary therapy to a chronic, lifelong therapy. Most importantly, Baby Girl X's underlying basic disease of shortened intestine switched from a good prognosis for eventual intestinal function to a condition unable to be altered by an existing medical or surgical therapy. While institution of TPN is clearly an established treatment in and of itself, utilization of TPN "for life" in a neonate whose underlying disease cannot be altered remains an experimental application of TPN.

The parents of a neonate with extreme Short Bowel Syndrome need to be counselled about the inherent medical uncertain-

ties of a lifelong TPN recipient: the acknowledged risks of catheter-related sepsis and liver disease, the potentially catastrophic risk of thrombosis of all available central veins for infusion of TPN, and the unforeseeable effects of TPN from infancy through maturity. The parents should be informed of small intestinal transplantation, a technique which may provide definitive treatment for patients with extreme Short Bowel Syndrome. At the present time, small intestinal transplantation is an experimental procedure which has not been proven to be clinically successful in humans.

The therapeutic options to be considered for a neonate with extreme Short Bowel Syndrome include two realistic options: treatment with TPN or oral feeding. The first therapeutic option, institution of TPN, upholds the principle of beneficence by seeking to allow the long-term survival of Baby Girl X. The principle of beneficence, to help a patient further his or her legitimate interests, assumes that it is in Baby Girl X's best interest to have long-term survival. However, inherent in implementing beneficence is the balancing of benefits and potential harms. The benefits are clear: survival and the hope that definitive treatment for Baby Girl X's underlying disease (i.e. intestinal transplantation) becomes clinically efficacious in the near future. The possible

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burden of this therapeutic option (sepsis, liver disease, unanticipated complications) must be weighed against the obvious benefits of long-term survival, prospects for normal growth and development, and potential definitive therapy by a technique such as small intestinal transplantation.

The second therapeutic option for a neonate with extreme Short Bowel Syndrome would begin oral feeding (breast or bottle), continue routine nursing care, and would expect death from malnutrition within several days. This option of allowing "nature to take its course" is ethically defensible because parents are not required to submit their children to treatment which cannot alter the underlying disease process. At the present time, a definitive treatment for extreme Short Bowel Syndrome is lacking and a patient with this condition will not be able to eat normally. Parents are also not strictly obliged to enter their children into experimental treatment programs. Commit-

ment of a neonate with extreme Short Bowel Syndrome to lifelong TPN is an experimental application of an established therapeutic modality.

The "clinical moment" — making the choice between what can be done and what should be done for a given patient, as described by Edmund Pellegrino, — is now faced by the caretakers of Baby Girl X. Although the withholding of TPN from this infant is ethically defensible for the reasons specified in the second therapeutic option, withdrawal of TPN becomes problematic. First, we must assume that the parents of Baby Girl X have already agreed to the first therapeutic option — institution of TPN — after her third operation documented near-total intestinal loss. They and the physicians have contacted other pediatric experts; they have met another child with extreme Short Bowel Syndrome receiving prolonged TPN. Relatively early in the course of her therapy, Baby Girl X has experienced two complications: catheter-related sepsis and impaired liver function. The question now becomes: at what point can a chosen treatment plan, i.e. lifelong TPN, be changed to another plan, i.e. the second therapeutic option of no aggressive intervention? Once again, the principle of beneficence can be applied to the decision-making process: does long-term survival remain in Baby Girl X's best interest and how should the potential benefits (survival) and harms (sepsis and liver dysfunction) be balanced? Presently, neither of Baby Girl X's burdens constitute an absolutely irreversible situation. Appropriate antibiotic treatment should be successful in eradicating the sepsis. Possibly, the catheter delivering her TPN solutions may need to be removed if the sepsis cannot be eliminated by the antibiotics. A new catheter would subsequently be required after the sepsis is treated and Baby Girl X's clinical condition has improved. While liver disease is frequently progressive in severity, it can stabilize for long periods of time by careful manipulation of the contents of the TPN solutions, small amounts of specialized enteral feeding, and prevention of systemic sepsis. Since Baby Girl X's immediate life-threatening complication of sepsis can be treated, she should be offered continued medical support.

The onset of irreversible burdens in Baby Girl X would warrant re-evaluation of her clinical and prognostic status by her physicians and parents. Irreversible liver disease would be an indication to reconsider implementation of the first therapeutic option. The clinical course of infants with extreme Short Bowel Syndrome is marked by multiple setbacks, each of which often demand a multidisciplinary approach. Once an infant is placed in the therapeutic plan of prolonged TPN, irreversible complications should be the major indication for withdrawal of treatment.

**Case Analysis:** *Dr. Donna A. Caniano, is Assistant Professor of Surgery, Ohio State University College of Medicine*

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