Sometimes a patient’s life can be saved but the price that must be paid, in terms of the burdens of treatment and the painful sequelae, can be considered too high. In this month’s “Ethics Rounds,” we present such a case from a Dutch ICU. We ask 2 experts, 1 from the United States and 1 from the Netherlands, to comment on the case. Dr Peter J. Smith is a developmental pediatrician at the University of Chicago. Professor A.J. van Vught is the director of the pediatric ICU at the Wilhelmina Children’s Hospital, University Medical Centre, Utrecht, Netherlands. Finally, 1 of the doctors involved in the case, Wendela G. Leeuwenburgh-Pronk, discusses the outcome.

PART I

Sophia, an 11-month-old baby, developed high fever, malaise, and a rash. Her parents took her to a nearby hospital. In the emergency department, the pediatrician diagnosed septic shock with purpura fulminans. Antibiotics were started, and she was transported to a PICU 1 hour away. She had been a full-term baby with no health problems. Her immunizations were up to date, including vaccination against Neisseria meningitidis type C.

On PICU admission, she was intubated and started on ventilatory support. She developed a severe refractory septic shock with disseminated intravascular coagulation and rhabdomyolysis. She required intensive inotropic and vasopressor support, fluid resuscitation, and supplementation of blood products. She recovered from the septic shock. However, all 4 extremities and the buttocks showed severe necrotic lesions. Both hands and feet were irreversibly necrotic, most likely necessitating amputation. There was necrosis of the proximal muscles. Her long-term prognosis for motor function was uncertain.

After a series of multidisciplinary meetings that included intensivists, rehabilitation specialists, and plastic surgeons, the doctors agreed that the best case scenario was to amputate both hands and feet. The worst case scenario would require amputation of all 4 extremities (above-knee and above-elbow) leaving only the torso and head. The precise extent of amputation could only be determined during surgery (with exploration of the underlying tissue). The parents ask: What would you recommend?

Peter J. Smith, MD

“This is a nightmare.” Any parent will cringe at hearing this story. No words can change the awfulness of this story or allow us to really understand what these parents and the child are going through. Emily Dickinson wrote eloquently of how impossible it is to experience the pain of another:
Any comments about this story must be built on the knowledge that neither doctors nor bioethicists can know the true perspective of the parents or the child. However, there are a few things we should keep in mind.

First, things now are better for children (and adults) with amputations than at any time in human history. Technology and outcomes continue to improve. This is due, in no small part, to the advances made due to injuries suffered in combat recently by large numbers of soldiers. Importantly, these improvements are not only related to better technology. They also reflect general public acceptance of life with disabilities (it is a current trend in advertising to visibly show prosthetics). As has been the case throughout history, war changes societal perceptions of disabilities.1

Second, any predictions of this child’s future are fraught with uncertainty. Those related to disability and impairment are likely to be overly pessimistic.2 We should remember that an individual without feet is now one of the fastest people on the planet.3 (Were his parents told that he would never walk?)

Third, we should carefully consider the contextual factors that, aside from the child’s medical problems, are likely to have a big impact on her prognosis. What services are available in her community? What sort of social support does her family have? Are there rehabilitation facilities that are easily accessible, could take a child her age, and could help transition her to home? I would answer the parents’ question by recommending that they pray or meditate or introspect deeply before making a decision. I would recommend that they lean on those around them. I would recommend that they talk not just to doctors but also to others in their community who understand their values. I would recommend that all involved treat Sophia as a wise child who potentially has something to teach us. Most especially, I would reassure Sophia’s parents that there are many who will walk with them on the journey that lies ahead, no matter what decisions they make or what events are in their future.

A.J. van Vught, MD

This 11-month-old girl was admitted to the ICU in severe septic shock with disseminated intravascular coagulation and extensive necrotic lesions of the skin and all 4 extremities. After hemodynamic stabilization, both hands and feet appeared to be lost, and amputation was the next logical step in the treatment plan. However, it could well be that the amputations would be much more extensive, leaving the child with virtually no limbs at all, just a torso and head.

This case shows the dilemma that pediatric intensivists occasionally face despite preventive measures as vaccination, improved technology, and medication that allow vigorous support of ventilation and hemodynamics, and sophisticated treatments such as early fasciotomy and sympathectomy. The result of intensive care sometimes appears be catastrophic to the child and the family. What should be done then?

The first thing to do is getting the medical facts clear. Before counseling the parents, I would like to know the expert opinion of the orthopedic surgeon, the plastic and reconstructive surgeon, and in particular the rehabilitation physician. Rehabilitation medicine with its wide-ranged array of prosthetic and ergotherapeutic possibilities is best equipped to reveal the different functional outcome scenarios and its distinct probabilities. In addition, the capability of Sophia’s parents to cope with the handicaps and the support they can receive must be explored. What future can Sophia be offered?

The next question will be what to do with the information? What are the options? One would be to proceed with maximal treatment and amputate the limbs to whatever extent to save Sophia’s life? Another would be to forgo further surgery and leave Sophia in palliative care with eventual death from sepsis? Or is there a way in between? It is clear that if Sophia survives, the burden to her and her family will be substantial.

The key question is what is in the best interest of Sophia? Along with that question, who should decide about her best interest? Can limits be set to the treatment, and if so, what are the limits and who decides about them?

In most legal systems a mentally competent patient is autonomous in the decision about his or her treatment, even if this decision is obviously against compelling medical advice. In children, this is more complicated. The parent’s opinion is highly relevant, but in cases of life and death not decisive. The attending physician and the treatment team have their own responsibility to the child. In most countries the child’s opinion is relevant too, if not by law then in any case morally.

Weighing future handicaps, burden and suffering against Sophia’s potential to develop to a happy member of the society comprises inevitably personal and subjective beliefs and attitudes. This counts for the parents and for the attending physician and the treatment team. This should not be a problem as long as these attitudes and beliefs are clearly communicated.

It is evident that no treatment team will refrain from amputations of limbs if the functional outcome with prostheses for Sophia and vigorous support to her and
her family is good. More complicated is the worst case scenario where all 4 limbs must be amputated, severe necrosis of the buttocks prevents upright positioning, and there is no functional support achievable. Then treatment directed merely to survival but imposing a grim prospect for Sophia and her family with suffering and virtually no developmental opportunities is beyond the aims of medicine.

Consultation of the ethics committee will be helpful to the medical team and the parents in defining the borders of treatment. Following the same principles, those borders may not be the same in every case. In developing countries, the limits may differ from those in the United States or Western Europe.

After gathering thorough expert opinion of the orthopedic surgeon, the plastic and reconstructive surgeon, and the rehabilitation physician and after discussing the limits of treatment with the ethics committee, I would likely advise surgery in this case. If during surgery amputations could not remain within the predefined limits, the operation must be interrupted, and additional treatment be limited to palliative care.

During the whole decision process, the parents of Sophia must be kept fully informed and supported. Their attitude, beliefs, and opinions are part of the decision. However, in my view, the final responsibility for the decision remains within the medical treatment team.

In most cases, parents will follow the advice of the intensive care team.

**PART II**

On day 13 of PICU admission, escharotomy was performed on all extremities because of severe compartment syndrome. During this procedure, it became clear that the muscles of both lower legs and right arm were necrotic, necessitating amputation of both legs above the knee and right arm above the elbow in the future. According to the rehabilitation physician, the skin and muscles of the buttocks were crucial for rehabilitation in case of extensive amputation. However, uncertainty remained about the extent of necrosis in that area because no exploration was performed. Meanwhile, Sophia seemed to recover from the severe sepsis without apparent neurologic sequelae. The majority of the medical team questioned the quality of life, both short- and long-term, if Sophia faced a future of immobility and frequent hospitalizations for decubitus and infections. The medical director of the PICU supported this point of view.

When the medical team discussed the possible extensive amputation with Sophia's parents, her mother stated that this was no reason to withhold or withdraw the medical treatment. However, it was difficult to assess whether the parents fully understood the severity of the physical sequelae. Should the amputations proceed?

**A. J. van Vught**

In this situation, the opinion of the rehabilitation physician is of utmost importance. It seems likely that above-knee amputation of both legs will severely handicap Sophia but that she would be able to learn to walk independently. The above-elbow amputation of the right arm will further impair her independence. However, much will depend on the function of the left arm and the condition of the buttocks.

I would like to know more about Sophia's prognosis for cognitive function. If her cognitive function is impaired, this could seriously limit her rehabilitation potential.

For the moment, I can understand that the parents insist on continuation of the treatment. If there is some doubt whether they fully understand Sophia's condition, it must be explained to them in more detail.

If Sophia's condition and the consequences of treatment have been fully explained, and the parents fully understand, then I can see 3 possible outcomes for this case. The treatment team and the parents may (1) agree to continue treatment, (2) agree to discontinue treatment and provide palliative care, or (3) they may disagree about what is in Sophia's best interest. Agreement, of course, would be the best situation both for the parents and for the treatment team. However, it can only be considered best for Sophia if what they agree on meets the current professional standard for children in Sophia's condition. Professional standards, in this context, should be interpreted as what either the majority or an important minority of health care professionals would do. The usual sources for professional standards such as relevant literature, case conferences, and published expert opinion must be used to evaluate those standards. In this case, I think that professional standards would allow either a decision to proceed with amputations or a decision to shift to palliative care.

More complicated is the situation in which Sophia's parents and the treatment team disagree on the best interest of Sophia.

The treatment team could consider amputations as foreseen for Sophia to be beyond the limits of a burden that ought to be imposed on her. If that is their opinion, they cannot be forced to carry out the amputations. In fact, they would have an obligation not to do so. However, they have to motivate their position thoroughly with professional arguments, communicate these carefully to the parents, and document their objections clearly and explicitly. If there are physicians who do not share their opinion and who would be willing to perform the amputations, those physicians should assume responsibility for Sophia's continuing medical treatment.
If such a medical team cannot be found, Sophia’s current physicians continue to have responsibility to care for her.

If the disagreement goes in the other direction, and Sophia’s parents believe that withdrawal of additional treatment is in her best interest but the doctors feel otherwise, then the doctors would be obligated to seek judicial intervention to sanction such treatment. As in the earlier situation, if they can find doctors who would feel comfortable supporting the parents’ request, they should transfer Sophia’s care to those doctors. Often, in such disagreements, a second opinion will help the parents.

Full support of Sophia’s parents requires that they be given proper information at every stage of the disease process. Full disclosure and honesty can usually prevent intractable disagreement cases like this.

Peter J. Smith

One of the most difficult things for any medical team to do is to assess a patient’s future “quality of life.” This is especially difficult when the patient is a child. Every doctor has seen cases in which our predictions are dire and the outcome is surprisingly good. We’ve also all been wrong in the other direction. Prognostication requires humility. We must remind ourselves, again and again, that we cannot know the future.

What are the alternatives for Sophia and her parents? Her parents, at this point, seem to want to continue with efforts to save her life. I don’t think the health care team should question the insight of these loving parents who simply want their daughter to live, even though they know that her survival may be accompanied by suffering for her and also for them.

Sophia is clearly a child who is deeply loved by her parents. They all face an uncertain and unwanted future. The fact that they do not want the future is not the same thing as saying that they now do not want Sophia. Our obligation as providers is to make sure that they are making an informed choice. Given Sophia’s problems, decisions about her treatment are clearly in the gray zone. It would be permissible to withdraw life-support and shift to palliative, rather than curative care, but it is also permissible to continue to try to save her life. Sophia’s parents seem ready to do their very best to care for her. We should do the same.

I pray for her comfort, for the courage of her family, and for increasing wisdom in all of those who serve her as health care providers. I would follow Sophia’s family’s wise wishes and do whatever is needed to save her life and get her back home.

PART III: OUTCOME OF THE CASE

(WENDELA G. LEEUWENBURGH-PRONK, MD)

The PICU medical team held several multidisciplinary meetings to discuss what would be in the best interest of Sophia, both short-term and long-term. One PICU staff member (CB) who ran a follow-up clinic for all meningococcal septic shock survivors stressed that long-term health-related quality of life was not associated with physical outcome variables like amputation in these patients. Her studies showed that, after a life-threatening illness such as meningococcal septic shock, many parents and patients reported a greater appreciation of life. Some felt that the event had made them stronger and that they tried to make the best of their lives. This phenomenon is referred to as “resilience.” Important, however, was that most of the patients with more extensive amputation (feet, leg, or arm) had long-term morbidity because of significant functional impairment and the need for surgical reintervention(s) in the years after meningococcal septic shock.

Overall, total scores of intellectual functioning in meningococcal septic shock survivors were comparable to those of the reference groups. As to parents themselves, significantly higher scores were found on the majority of health-related quality of life scales (both physical and psychosocial) compared with Dutch normative data.

The PICU team did not consult the clinical ethics committee to reach a decision because the ethics committee serves as sounding board. Given the extensive nature of Sophia’s problems, the majority of the medical team felt it was not in Sophia’s best interest to continue treatment. The medical director of the PICU supported the majority view and opted to withdraw life support and shift to palliative care.

In the Netherlands, we generally regard such medical decisions as ones that should appropriately be made by the medical team to relieve parents from the burden and responsibility to decide in matters of life or death for their child. We then communicate this decision with the parents and ask for their approval. We also offer them the option of seeking a second opinion.

Sophia’s parents agreed with our views. On day 19 of PICU admission Sophia died in her mother’s arms. During the follow-up interview with the parents several weeks after the death of their child, the medical director invited them to reflect on the course of events during hospital stay. The parents stated they were very content with and grateful for the attitude and guidance offered by the PICU medical team.

John D. Lantos

People argue about whether ethical judgments differ in different cultures. This case, from the Netherlands, highlights some similarities and some differences in the approach to tough cases in the Netherlands, compared with the United States. The fundamental questions
are the same everywhere. How do doctors and parents decide, in tough cases, whether to continue life-sustaining treatment or to switch to palliative care? How sure do we have to be about a poor prognosis before we withhold or withdraw life-sustaining treatment? When do we err on the side of life? The decision, in this case, is one that might have been reached in any country. The approach to doctor-parent communication, however, would likely have differed. In the Netherlands, the doctors try to shoulder the burden of tough decisions. They don’t leave the decision up to the parents. In the United States, doctors would more likely offer parents’ explicit choices. Neither approach is obviously preferable. Both have problems. Such differences suggest the need for additional study of the effects of different approaches on health professionals and parents.

REFERENCES


DRIVING FOR PIZZA: All the pizza at the table was gone. I had eaten more pizza than I thought possible. My only excuse was that it was incredibly good. I was recently in Rome and, surprisingly, the best meals I had were pizza. My first night in Rome, our host drove us from our hotel in the outskirts of the city to a pizzeria in the center of the city, close to the Mausoleum of Augustus. Initially, I thought it odd that we had taken such a long trip for a pizza. Expecting a small personal pizza, I, as with the rest of the members of our group, was stunned to be served what seemed to be a massive pizza - for sure a large pizza back home. We all thought that we should have just ordered a few for the table as it seemed unlikely we could possibly eat that much. We were wrong.

As most know, there are many, many types of pizza and each region claims to make the best. The origins of pizza are a bit murky; but Greeks living in southern Italy before the rise of the Roman Empire baked round, flat breads that they topped with oil, garlic, and various foods. The bread often had a rim of crust that allowed the bread to be carried. Tomato-based toppings for flat breads are a relatively recent phenomenon, as the vegetable was unknown until brought back from the Americas and was initially thought poisonous. The first shops dedicated to making what we now know as pizzas probably opened in Naples in the early to mid-1800s. Pizza Alla Marinara, flat bread topped with tomato sauce, oil, garlic, and oregano was the standard until pizza Margherita, topped with tomato, mozzarella cheese, and basil was developed to honor the first queen of Italy, Queen Margherita. Regardless of the origin, my pizza Margherita was superb. The Roman style is to make the crust extremely thin and crispy, and use buffalo mozzarella. The pizza is cooked in a very hot oven in which the floor is quite a bit warmer than the space - even just a few inches above the floor - and only for a minute or two. The taste is divine and the pizza neither heavy nor too filling. While sure to provoke strong responses from those in southern Italy, California, or Chicago, I am going to have to say that the Romans make the best pizza in the world.

Noted by WVR, MD
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