Two of the most ethically complex situations in pediatrics are those involving families whose religious beliefs preclude the provision of life-sustaining treatment and those involving young adults who have reached the age of legal majority and who face decisions about life-sustaining treatment. This month’s “Ethics Rounds” presents a case in which these 2 complexities overlapped. An 18-year-old Jehovah’s Witness with sickle cell disease has life-threatening anemia. She is going into heart failure. Her doctors urgently recommend blood transfusions. The young woman and her family adamantly refuse. Should the doctors let her die? Is there any alternative?
INTRODUCTION

Two of the most ethically complex situations in pediatrics are those involving families whose religious beliefs preclude the provision of life-sustaining treatment and those involving young adults who have reached the age of legal majority and who face decisions about life-sustaining treatment. This month’s “Ethics Rounds” is a case in which these 2 complexities overlapped. An 18-year-old Jehovah’s Witness with sickle cell disease has life-threatening anemia. She is going into heart failure. Her doctors urgently recommend blood transfusions. The young woman and her family adamantly refuse to allow transfusions based on the belief held by Jehovah’s Witnesses that the Bible prohibits blood transfusions. Should the doctors let her die? Is there any alternative?

The following were the ethics consultants for this case: from New York Methodist Hospital, Dr Revathy Sundaram, the chief of hematology/oncology and attending physician for the Sickle Cell Comprehensive Care Unit; Dr Nnenna Ukachi, a third-year resident; and Ms Samantha VanHorn, the research coordinator; and from Children’s Hospital of Philadelphia, Dr Wynne Morrison, a pediatric intensivist, bioethicist, and palliative care physician.

CASE REPORT: PART I

TH, an 18-year-old girl with sickle cell anemia, was born to a family of Jehovah’s Witnesses. Her past history is significant for multiple crisis episodes, including several ICU admissions for acute chest syndrome. She has been transfused multiple times during previous admissions.

Shortly after her 18th birthday, TH was admitted for a pain episode in her lower back, coinciding with the onset of her menstrual period and unrelieved by oral opioid analgesics. In her case, this painful crisis was not triggered by the onset of menses, nor had they been previously. Her flow was not heavy and lasted 3 to 4 days. Her initial vital signs were stable, with an admitting hemoglobin value of 8 g/dL. She was started on intravenous morphine, antibiotics, and supportive care. Her pain continued despite maximal doses of patient-controlled morphine analgesia. By the fourth day, she was noted to be very pale with hemoglobin of 3.8 g/dL and a relatively poor reticular response of 7.9%.

TH was transferred to the PICU and counseled on the need for a blood transfusion. She was alert and responsive, with tachycardia and rising blood pressure. She expressly refused any transfusions citing her religious belief that transfusions would deprive her of eternal salvation. She was started on intravenous iron and darbepoetin. The next day her hemoglobin dropped to 3.2 g/dL. Physical examination revealed a gallop rhythm. Her chest radiograph was suggestive of vascular congestion. She was started on levalbuterol.

What Do You Do Now? Why?

Response From Nnenna Ukachi, Samantha VanHorn, Revathy Sundaram

This case raises many ethical issues. Jehovah’s Witnesses are widely known in the medical community for their refusal of blood and blood products. According to multiple biblical texts, blood is regarded as sacred, representing life, and a gift from God. As a result, blood is acceptable only when used for the atonement of sins, and may not be ingested or transfused in any form. A Jehovah’s Witness who accepts a blood transfusion essentially dissociates him/herself from the religion and is shunned by other members.1 TH cited religious conflicts and made her refusal of transfusions apparent as soon as she was legally able to do so. For members of the clinical team who have cared for TH from infancy, this may have been a difficult decision to accept. In addition, she appeared to be acutely ill and in impending cardiac failure from the description of this crisis episode. The first step would be to maintain her hemodynamic stability and then transfer TH to the ICU for close monitoring with fluid hydration and aggressive pain control. Prevention and treatment of other causes of anemia and diminishing iatrogenic blood loss would be essential. Although not emergent, the use of hormonal contraceptives should be considered to minimize iron deficiency from menstrual loss.

To improve oxygenation, the risks and benefits of alternate and experimental therapies, which have been used in adult patients who refuse the use of blood, should be discussed with TH. Although experience with these therapies is limited in pediatrics, they should be offered to a patient in this situation. It would also be helpful to discuss the case with a physician from an established “bloodless” program. Such centers have experience in the treatment of Jehovah’s Witness patients who refuse transfusions. A consultation with an internist-hematologist might be helpful. In addition to offering medical advice for management, an adult hematologist might also assist the pediatric team in gaining appreciation for an adult-oriented approach. Erythropoietin analogs and nonblood oxygen carriers should be offered to the patient.

It is also important to determine if the patient is psychologically and physically capable of making decisions. For TH, her newly empowered status as a legal adult and the confrontation with a difficult and potentially life-altering
decision such as this may be overwhelming, but the care team should ensure that her decisions are not made under duress or pressure from family or friends, which could potentially invalidate them. In addition to clinical support, she and her family would require continued psychosocial and emotional support. It would be crucial to educate TH and her family on the risks and benefits of the treatment options available, determine the validity of her decision, and to provide optimal supportive care regardless of the decision and its outcome. Involving the Hospital Ethics Committee is crucial, but more important is identifying an individual who TH respects, trusts, and with whom she is comfortable speaking. Typically such an individual is from the medical team and has an established therapeutic relationship with TH. Examples might include a social worker or child life specialist. Such an individual can often advocate for the patient/family and serve as a liaison between the patient/family and the medical team, helping to bridge differences in a nonconfrontational manner.

Pediatric providers seldom face situations in which their decisions to save a life are questioned or not accepted. When the patient is a minor, they usually can seek and obtain a court order for treatment. When the patient is a young adult, everything changes. Pediatricians then feel anger and helplessness. Their personal beliefs and the lifelong training direct them to provide supportive care regardless of the decision and its outcome. Reaching the age of legal maturity may seem like a death sentence. A clinical ethics consultant in this case would want to explore how strenuous TH’s objections had been in the past, what her relationship with her religious community is like, and how well she understands the consequences of refusal. TH should be privately offered the opportunity to accept a transfusion without revealing to her family or community leaders that it took place. Any possible additional therapies to support her without transfusion should be aggressively pursued.

In the United States, adults with decision-making capacity have a right to refuse medical therapy, even if the therapy is thought to be life-saving. This standard applies to Jehovah’s Witnesses who would rather die than receive blood transfusions. A medical team may challenge a refusal only if they think that the patient does not understand the therapy, alternatives, or consequences of refusal or is otherwise impaired (eg, suicidally depressed).

In pediatrics, however, it has become relatively common practice to transfuse children whose parents are Jehovah’s Witnesses when such transfusions are absolutely necessary. This practice is not universal or without controversy, but many pediatricians obtain court orders to override parental refusals when all alternatives to transfusion have failed. A central argument for overriding parents is that the child has not yet made a mature choice to be a member of a particular religious community and to follow that community’s doctrines. We justify transfusing the child because there is a chance the child will decide transfusions are acceptable as he or she grows older. An emotional argument buttresses the philosophical one: clinicians are distressed at the possibility of letting a child die for lack of what is seen as a relatively routine therapy.

We recognize that decision-making capacity emerges over time as an adolescent matures. A 16-year-old patient’s informed opinion may, therefore, deserve more respect and consideration than that of a 12-year-old, depending on the maturity of the child, experience with decision-making, and the implications of the decision. The legal cutoff for adulthood marks an arbitrary, abrupt transition, but it exists nonetheless.

Pediatricians are sometimes unprepared for a patient’s transition to legal adulthood at age 18. We may forget to ask about advance directives, may habitually look to parents to continue to make decisions, and may become accustomed to using the courts to transfuse a patient without a family’s consent. For TH’s physicians in the vignette described, it must be very distressing to realize that they may be legally required to accept her refusal. Sometimes, ethics crises arise that could, perhaps, have been prevented. It would certainly be best for a thorough exploration of these issues to begin before the patient’s 18th birthday and before the patient is in an acute, uncompensated situation. If TH and her long-term physicians had agreed on a working plan before the crisis occurs, all would have been more comfortable accepting it.
If given an opportunity to plan in advance, TH's physician might have said to her: “I understand that you do not wish to accept blood transfusions and that this issue is very important to you. I have been caring for you as your physician for years and am committed to continuing to do so. Many times in the past, I have given you blood, and I am convinced that I have saved your life or helped treat your pain when I did. You are about to turn 18, and when you do, it will be really hard for me if avoiding transfusions puts your life at risk. I therefore want to let you know that I will give you transfusions if I think they are absolutely necessary, but I will not make you sign a consent form and will not let anyone in your family or your community know that you were given blood. If imagining being in such a situation is absolutely horrifying to you, then I need you to send me a letter in writing now saying that you do not want me to ever transfuse you. I would then not give it to you even if the situation became life-threatening. If you were dying because of not getting blood, I would make sure you were comfortable. I also need to let you know that if you go to another physician who has never transfused you before, chances are that doctor will honor your wishes as an adult to refuse blood. I hope that you do not choose this option, but understand if you do.”

Such a strategy forces the patient to take action (writing a letter or finding another doctor) to “actively” refuse blood transfusions. It allows the patient to passively accept blood by simply not writing a letter and continuing to follow up with the same physician. The physician would need to make sure this is a legally tenable option both at the institution and in the state where he or she is practicing by talking with the hospital attorneys and planning how to best document the conversation. Perhaps a judge will have to be identified who is willing to confidentially support such a plan. The need for legal clarity may make such a nuanced approach too difficult to manage.

In this case, if the medical team, support staff, and ethics consultants (who are hopefully involved) are convinced that she understands the consequences of her decisions and is expressing her true wishes, then they should respect her refusal. They should talk with her about what other options make sense in light of her refusal of blood. Should the care at this point focus on her comfort rather than invasive measures like intubation?

It may be difficult for the medical and nursing team if TH dies. They should be offered support. If possible, any who are uncomfortable caring for her given the circumstances should be able to change assignments.

This case reminds us that we should avoid becoming too comfortable overriding our patients’ wishes when we disagree with their decisions, just because we are sometimes able to do so. It also reminds us to be proactive in anticipating the legal and ethical shifts that occur when our patients turn 18, no matter what the clinical circumstances. And, as always, we should continue to care for and about our patients, even when forced to acknowledge that we disagree.12 If TH dies in this case, it will be because she chose to die for her beliefs.

CASE REPORT: PART II

The risks and benefits of an urgent transfusion were explained to TH and her parents at the onset of severe anemia, including the risk of cardiac failure, renal failure, and brain damage. Given the fact that she had previously been administered multiple life-saving blood transfusions without parental consent, several meetings were held with the patient, her parents, the social worker, child psychologist, Risk Management, and the Ethics Committee to determine the validity of her refusal of life-saving therapy. The medical team also met with TH in private and offered her the opportunity to discuss her opinions in a confidential manner. Although the medical team felt that she was capable of making the decision to refuse a blood transfusion, validation by the Ethics Committee was helpful. The Ethics Committee concluded that she had the right to make her own decisions as an adult. Her parents were supportive and agreed with the decision. TH was lucid and understood the risks but persistently refused transfusion.

TH was monitored closely throughout her stay, with blood drawing restricted to complete blood and reticulocyte counts every other day. By the end of the first week, her pain improved and she was weaned to room air and oral analgesics. Her hemoglobin improved slightly to 3.6 g/dL with a reticulocyte response of 14.1%. She received daily physical therapy and was encouraged to ambulate. On day 13, she was transferred out of the ICU and by day 17 she was discharged with a hemoglobin of 7.1 g/dL on digoxin, furosemide, folic acid, hydroxyurea, iron tablets, and levalbuterol.

Response From Wynne Morrison

The outcome of this case has one additional lesson for the medical team: be careful how certain we are in our prognostication. The team in this case walked the medical tightrope with the patient to reach a good outcome, both clinically and ethically. The careful medical management, respect for the patient and for values that may differ from our own, and a bit of luck brought them to this resolution. Hopefully she will continue to do well through the next medical crisis. Whatever that next chapter brings, it sounds like this
patient and this health care team have come to an understanding that all can accept.

Response From Nnenna Ukachi, Samantha VanHorn, Revathy Sundaram

Red cell transfusion is currently the most studied and accepted therapy for most acute and many chronic complications of sickle cell anemia.1,2 However, alternative therapies cannot be ruled out, especially when transfusion is prohibited. In this instance, recombinant human erythropoietin and hydroxyurea may have saved TH’s life. Erythropoietin has been shown to increase red cell production within days to weeks of use.3 A combination of erythropoietin, iron supplementation, and hydroxyurea has been shown to increase fetal hemoglobin and ameliorate sickling crises.4 Early aggressive intervention with these agents can sometimes prevent significant anemia and the ensuing complications.

This case illustrates the importance of using all potentially beneficial therapies, even if they are not first-line therapies or the standard of care, in situations in which those first-line therapies cannot be used.

Response of John D. Lantos

This case illustrates 2 crucial aspects of clinical ethics for pediatricians. First, we must get better at preparing our patients for the transition from childhood and adolescence to adulthood. When they turn 18, they will need to take responsibility for their own health care decisions. Preparation to do so must begin long before their 18th birthday. In this case, 2 aspects of that preparation were not done well. The patient was not, apparently, adherent to the best medication regimen to prevent life-threatening anemia. If she had been, this issue might not have arisen. Second, nobody had, apparently, had conversations with her about her impending new responsibilities to make decisions for herself or outlined a process of shared decision-making. Dr Morrison’s template for such a discussion should be pinned to the wall in the office of every pediatrician who cares for teens or young adults with life-limiting illness. As we prepare our patients, we must also be prepared ourselves for the whirl that with a positive response we will experience when our patients turn 18 and the guiding principle of beneficence gives way to the guiding principle of autonomy. Adults are permitted to make bad decisions. They may choose to die even when life-saving treatment is available. And we need to not abandon them, even if they make decisions that we find morally problematic.

The second lesson from this case is that we are not omniscient. In such a case, we may think that our prognosis of impending death is uncontestable. Had TH been younger, doctors would likely have gone before a judge and given sworn testimony that she would die without a transfusion. A transfusion would certainly have been the standard of care. Yet she did not get a transfusion. And she did not die. Some things, it seems, are both out of our hands and, perhaps, beyond our understanding. Troubling cases like this one can offer painful but useful lessons in humility.

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