Parental Refusal of a Liver Transplant for a Child With Biliary Atresia

Pediatricians are required by law to notify child protection agencies if parents are neglecting their child’s medical needs. Sometimes, however, it is difficult to determine when a particular parental choice ought to be reported. If the treatment is standard therapy and the consequences of nontreatment are dire, than parental refusal is usually considered neglectful. Organ transplants, however, represent a unique situation for 2 reasons. First, because there is a dire scarcity of organs, there are more people who need organs than there are organs to transplant. In this situation, it seems odd to force a transplant on a child whose parents do not want it, knowing that other parents are eager to have their child undergo a transplant. Second, transplantation is risky and requires lifelong follow-up and treatment. Thus, parental cooperation is essential. We present a case of a child who needs a liver transplant and whose parents refuse, and ask 2 surgeons, a gastroenterologist, and a child abuse specialist to comment on the case. David C. Cronin is director of the liver transplant program at the Medical College of Wisconsin. Robert Squires is a gastroenterologist, George Mazariegos is a transplant surgeon, and Janet Squires is chief of the child advocacy center, all at the University of Pittsburgh.

The Case

A 10-month-old girl, Baby A, was admitted to the hospital because of worsening jaundice. She had been diagnosed with biliary atresia at 1 month of age, and a Kasai procedure was performed when she was 10 weeks old. Since then, she had been failing to thrive and had worsening jaundice.

On the current admission, the patient’s weight was 8 kg (<5th percentile). Her abdominal girth was 50 cm with significant hepatomegaly. Her total protein was 6.0 g/dL with an albumin level of 2.5 g/dL. Alkaline phosphatase was 355 U/L, aspartate aminotransferase was 150 U/L, and alanine aminotransferase was 61 U/L. Her total bilirubin level was 20.5 mg/dL with a direct bilirubin of 18.0 mg/dL. Her prothrombin time was 17.0 seconds, partial thromboplastin time was 28.5 seconds, and her international normalized ratio was 1.8.

Baby A’s physicians recommended a liver transplant and estimated that, without a transplant, she would not survive another year.

Her parents did not consent to have her listed for a transplant. They believed that God was already healing their infant. They recounted the story of an uncle who had been told that he might need a kidney
The physician requested an ethics consultation, asking whether it was permissible for the parents to refuse a liver transplant in this situation and, if not, whether the ethics committee would support a report to the child protection agency for medical neglect.

David C. Cronin Writes

The field of solid organ transplantation represents an area of medical practice that often must address issues of allocation of a scarce resource, justice, equity, and decisions of who should and who should not be allowed to benefit from the transplantation. In the United States today, a liver transplant is standard-of-care for pediatric patients who have end-stage liver disease secondary to biliary atresia. Among pediatric patients, the most common cause of end-stage liver disease and the most common indication for a liver transplant is biliary atresia. The majority of these patients require liver transplantation before the age of 2 years.

Unfortunately, although liver transplantation represents the single best and only definitive therapy associated with prolongation of life in the setting of end-stage liver disease, organs available for transplantation are most significantly limited for candidates weighing <20 kg. This disparity between candidate need and organ supply is 6 times greater for pediatric candidates compared with adult candidates. Consequently, death while awaiting liver transplantation is a potential and real outcome. Due to the scarcity of organ availability, patients awaiting a liver transplant are triaged for organ allocation on a sickest-first basis. Determination of the sickest patient is represented by a prospectively validated severity-of-illness score referred to as the Pediatric End-Stage Liver Disease score; this scale is valued from 6 to 40, with increasing value representing an increasing risk of death awaiting liver transplantation. Baby A’s Pediatric End-Stage Liver Disease score was 30, representing a significant 30-day mortality risk without a liver transplant.

Medically, this child represents the usual presentation of end-stage liver disease secondary to biliary atresia. In this particular case, the diagnosis of biliary atresia seems to be without question. The child presented early in life, underwent standard surgical therapy (Kasai procedure), and has progressed to end-stage liver disease as demonstrated by the laboratory values, physical examination, and failure to thrive. Although the patient has an indication for liver transplantation, the reason to pursue transplant at this point is her failure to thrive and worsening jaundice. Although the family has demonstrated commitment to good patient care (follow-up with physicians for diagnosis, surgical therapy, and postoperative care), they have a misconception about the difference between liver transplantation and kidney transplantation and the difference between the onset and reversibility of some forms of kidney failure and the progressive nature of liver failure in this situation. To discount the family’s concerns as irrelevant (as they are) would not be medically or ethically appropriate. The health care team needs to present the information about the nature of the illness with and without liver transplantation as an option. The specific discussion that this child will die without a liver transplant must be conveyed. This must be done at a level that is understandable to the family and will most likely need to be conducted over a period of time. Furthermore, there is an opportunity to allay the concerns of “butchering” by reviewing the previous surgery (the Kasai procedure) and outlining the similarities in surgical scar and surgical procedure. The family’s repulsion may be a representation of fear and frustration.

Taking a position for or against supernatural intervention is often not productive. In the end, we should not decry the possibility of miracles but rather incorporate divine intervention in addition to the appropriate medical/surgical therapy. If after the educational and support measures and social service interventions are unsuccessful in moving the parents to accept transplantation as the appropriate treatment, the option of taking protective custody of the pediatric patient and acting in the child’s best interest is ethically and legally supported. In this situation, the parents have a duty to provide appropriate medical care but do not have the right to prevent appropriate medical intervention. This duty transcends religious objections in almost all circumstances.

Ethically, the child does not have autonomous decision-making ability. As such, the pediatric patient should not be victim to inappropriate medical decisions that have significant life-threatening or life-limiting consequences made by others (including the parents or surrogate caregivers). Parents and surrogate caregivers are responsible for providing appropriate medical care.
Legally, the state has an interest in the overall health and life of the most vulnerable members of society; in this case, pediatric patients. Consequently, there are clear mechanisms for health care providers to protect children at risk by taking protective custody and acting on a best interest basis. Here, the prohibition of liver transplantation on a religious basis would not be supported legally. In fact, because this is a child, many social support options exist to facilitate travel, housing support, and pharmaceuticals for the medical therapy.

In the end, it is better to invest the time and effort to educate the family, answer questions, allay their fears, and work collaboratively to embrace the appropriate medical treatment. If unsuccessful, protective custody and prevention of medical neglect are ethically and legally justifiable but would not be an easy path.

Robert Squires Writes

Most children with biliary atresia have jaundice that begins in the first few weeks of life. Often, families assume that the problem is due to “normal newborn jaundice” that just goes away. It is natural to reject the notion that a child may have a serious liver problem when the child “looks okay.” From the earliest period of care, parents should be offered a thorough discussion, by using laymen’s terms and drawings, that will review the age-specific etiologies and help them understand what future problems and needs their child may have.

It is not clear to me how this child was “diagnosed” with biliary atresia at age 1 month and had a Kasai procedure at age 10 weeks. The diagnosis of biliary atresia is usually made at the time of an operative cholangiogram, and the Kasai procedure is done immediately by using the same anesthesia. A cholangiogram should only be performed in a center with the surgical capacity to do the appropriate intervention at the same time. I would worry, and try to address, the possibility that there were communication problems at that early stage of treatment.

If biliary atresia is a potential diagnosis, the family must understand some general concepts going into the cholangiogram procedure: (1) biliary atresia is a fatal disease in the absence of a Kasai procedure (the natural history for children who do not have this procedure is that virtually all of them die of their liver disease before their third birthday); (2) after the Kasai procedure, careful and close medical follow-up will be necessary to avoid or manage complications related to biliary atresia (there are some children who live into adulthood with a successful Kasai); (3) not all Kasai procedures are successful; and (4) if the Kasai procedure is not successful, and if the jaundice has not cleared within 3 months after the procedure, then the child’s course will most likely be similar to those who did not receive a Kasai procedure, and an early death can only be averted with a liver transplant.

The Kasai procedure should be performed between 4 and 8 weeks of age. The older the child is at the time of the procedure, the less likely that the child will establish satisfactory biliary drainage.

The months after the Kasai procedure are crucial. The family must have a realistic knowledge of possible outcomes. If the procedure is successful, the elevated bilirubin levels will fall to the near-normal or normal range over the next 3 months. In other cases, the liver disease becomes progressive, and the child will need to be considered for liver transplantation. Early referral for a transplant evaluation, even as early as age 5 to 6 months for a child with biliary atresia and a failed outcome on a Kasai procedure, might give the family and child the best opportunity for accessing resources and planning for the future.

If I were consulted on this case, I would want to know the family’s understanding and experiences, as well as their concerns and anxieties. If parents do not seem to grasp the severity of the disease, do not accept responsibility, have inadequate resources, have competing problems that affect their ability to care for the child, or show signs of incapacitating fear or anxiety, supportive strategies are essential.

I would work with other family members, such as grandparents, and other persons important to the parents, such as ministers or friends. A second medical opinion from another center may be important. Support from social services and referrals to available community resources, including attention to financial issues, is necessary. Facilitating meetings with other parents can be very helpful. Luckily, in the current case, there is time for patience, as the child’s deterioration is not imminent. I would do almost anything to gain the confidence of the parents, to support keeping the family together, and to work with the parents to provide life-saving medical care.

However, if the parents remain completely opposed to the option of a liver transplant, I cannot accept the inevitability of the death for a child who could be helped by an available medical procedure with reasonable risks. This infant will die without a transplant. I believe such a case should be referred to the child protection authorities, and the judicial system must help decide what will be done.

George Mazariegos Writes

This scenario is a challenging situation. Multidisciplinary evaluation and working closely with the hepatologist seeing the child will help present a unified message to the family. As Dr Squires has emphasized, the key to helping the...
family make an informed decision is to be sure the family understands the child’s medical problem and the current data available regarding the benefits and the risks of organ transplant. I would evaluate for potential surgical and medical contraindications to transplant. These factors include a severe progressive systemic disease, a life-threatening malignancy, or rare cardiopulmonary conditions that limit anesthesia and surgical options. There could be a temporary condition, such as sepsis, coagulopathy, or severe malnutrition, which might require treatment while waiting for a future transplant opportunity. From a technical point of view, there are few absolute contraindications. With improved surgical techniques and experience, previous relative contraindications to transplant in biliary atresia such as portal venous thrombosis or situs inversus have been eliminated.

In this case, despite the fact that this is a young child who is small and who has had a previous failed outcome on a Kasai procedure, this infant would meet the criteria of a child who should do well in an experienced transplant center. Surgical possibilities include a deceased donor transplant, a living donor transplant, and a deceased donor split transplant. Thus, the prospect that this child will attain a suitable transplant in a reasonable time is high. Pediatric liver transplantation has excellent short- and long-term patient and graft survival outcomes. I would review with the family that the early (1-year) survival is expected to be >90%; in fact, our recent experience shows survival at 98%. Studies of long-term survival, typically defined at 5- and 10-year marks, show survival rates at ~90%, with most children having high-quality life parameters. I would remind parents that biliary atresia is a disease that does not recur after transplant, so that the child is cured of the original problem. The major focus of the care after transplantation is managing the complications of the medicines used for immunosuppression. There are major advances in this area, and many transplant recipients have a reasonable expectation to live with low-level immunosuppression. With a functioning liver, the child is expected to improve her nutrition and growth, which are essential for neurologic development. Finally, I would stress to the family the almost certain poor outcome without transplantation.

There is no reasonable expectation that a very symptomatic infant can improve her nutrition and growth or to thrive with the current liver disease. Without a liver transplant, Baby A would not be expected to live beyond age 2 years.

I do note the parents’ use of the word “butchered.” There is no question that transplant surgery is major and can be traumatizing. However, experienced centers understand the importance of treating the “whole child,” and great efforts are made to help the child and family cope. Our center has developed many resources to support families. Our major focus is return to normalcy for the child and family. A great resource for these parents would be to talk with other parents who have experienced a child having a transplant. Although there are clearly challenges and burdens, most families delight in the development and accomplishments of their child who has survived a lethal condition and who now has a real chance for a bright future.

If all of our efforts fail and the parents of this child would continue their refusal of a liver transplant, I would call our hospital’s child protection team. In my assessment, with the known outcome of death for the child without a transplant, the case should be reviewed by child protection authorities, and I would be willing to participate in a legal process in which a judge might make the ultimate decision. I would hope such action would not be necessary.

Janet Squires Writes

Parents are usually the appropriate surrogate medical decision-makers for their own children. We assume that they have the child’s interest at heart and are in the position to decide the best medical treatment. But there are limits. If parents choose a course of treatment that puts the child in imminent danger of harm, then physicians have a legal obligation to report such acts to the child protection system. Few pediatricians hesitate to report findings indicative of physical child abuse or sexual child abuse.

Medical neglect is hard to define. Textbook discussions of medical neglect usually focus on time-limited interventions in which the benefits and risks are fairly easy to describe, such as a lifesaving blood transfusion. This case presents a myriad of more complex issues. Just a few of the complicating factors to be considered include:

- Organ transplant involves complex technology. As new procedures and therapies become available, at which point do such interventions go from being “cutting edge” and research, to becoming standard of care?
- Organ transplants are not typically curative. The recipient will have a chronic medical condition requiring ongoing medical care, often for a lifetime.
- The burden of treatment is significant. Surgery is invasive, and complications are common.
- Burden for a family of a child receiving a transplant is very high. Transplant centers may be geographically far from family homes. Parents must often give up work and take time from their other children.
• Courts are reluctant to mandate medical treatments that do not have an end point. If a court were to mandate an organ transplant despite parental refusal, who will care for the child afterward?
• In this scenario, parents mention a religious belief that God will heal their infant as 1 motive for their decision. Does it matter what parental reasons are given?
• Risk/benefit ratios may depend on the center providing the medical care. Outcome measures are not uniform for all centers.

The diagnosis of child medical neglect is mainly dependent on the level of harm the child will suffer from the lack of medical care that a reasonable person would choose; the definition of “reasonable” is obviously challenging. The diagnosis of medical neglect is generally independent of the motivations of the adult. Lack of resources should be recognized and addressed. Understanding other reasons that parents do not choose or provide “reasonable” medical care (e.g., religious beliefs, mental health issues, competing family obligations) is important for attempts to work with the family. Fundamentally, however, pediatricians must assess when a child is being significantly harmed by medical choices of a parent or omissions of a parent’s care, and must call it medical neglect.

In this particular case, I also would talk extensively with the parents and try any tactic to incorporate their wishes for their child. If they refused the transplant option, I also believe this is an appropriate case for a judge to decide. I would explain to the parents that a referral to the child protection system is not a criticism of them as parents, nor a challenge to the belief that they love their child. Rather, physicians have a legal obligation to recognize and report cases in which a child is at risk for significant harm. Like the 2 transplant experts, I would do “almost anything” to avoid legal involvement but feel the certain death sentence for this child without intervention is not acceptable.

John D. Lantos Comments
In 1990, there was a case in Saskatchewan, Canada, in which parents refused a liver transplant for their 10-month-old who had biliary atresia. The physicians sought a court order for transplant. The court sided with the parents.7 I can find no reported cases, since then, in the United States or Canada, in which physicians sought protective custody because parents were refusing a transplant. I suspect that not all parents consent to transplant. Instead, I think that physicians generally do not report such parents for neglect.

Why?
The child-centered arguments for overriding such parental refusals, as presented here, are compelling. Without treatment, the child will surely die. With treatment, the chances for a good outcome are excellent. Our reticence to seek protective custody comes, I think, from the dire scarcity of organs. Because there are not enough organs to go around, some people on waiting lists will die awaiting transplant. It seems wrong to force a transplant on an unwilling family if it means that a willing family might go without one. I suspect that such concerns, rather than the complete absence of parental refusals, is the reason why we have not seen court cases involving this situation. This is one of the very few situations in which such concerns about the dire scarcity of a life-saving resource might appropriately tip the balance against a decision that reflects what is clearly in the child’s best interest. As long as people are dying on the waiting list, this is wise health policy.

Dr Mazariégos Responds
I do not believe organ scarcity is a major issue here. Wait list mortality has significantly improved over the past decade (Fig 1), even for the group with the highest usual death rate on the wait list; that is, infants aged <1 year. For example, of 126 children listed for transplant aged <1 year in 2009, a total of 7 on the waiting list nationally died, the fewest number that have ever died in 1 year over the past decade (Figs 1 and 2 present data from the 2010 report of DHHS’s Organ Procurement and Transplantation Network and Scientific Registry of Transplant Recipients).8 Split and live donor transplantation, along with improvements in allocation policy and pretransplant medical care, have made wait list mortality rare (but not yet zero) for children

FIGURE 1
Pretransplant mortality rates among pediatric patients wait listed for a liver transplant; grouped according to age.

FIGURE 2
Pediatric liver transplant rates according to age.
in the United States. In fact, the most significant improvement in transplant rate has been in children aged <1 year (Fig 2). Therefore, based on the current data, I strongly disagree that organ scarcity might appropriately tip the balance against transplant for this child or be wise health policy given the scenario described here: an otherwise healthy child with a reasonable chance for long-term success.

REFERENCES

7. Saskatchewan (Minister of Social Services) v. P.(F.) (‘Kaila’), 1990, 69 Dominion Law Reports (4th) 134 (Saskatchewan Provincial Court)

LESS ANXIETY AT THE HEAD OF THE PACK: We tend to think that people in leadership positions—whether in business, politics, or the military—are under more stress than those who are not. For some, the perceived stress of a leadership position makes them leery of accepting such a position. However, contrary to popular sentiment, a recent study reported in Forbes (Leadership: September 27, 2012) showed that leaders seem to feel less stress than their subordinates. Researchers recruited leaders and non-leaders from the Boston area, including adults enrolled in an executive education program at Harvard University. Researchers compared salivary cortisol levels (a biologic marker of stress) and self-reported anxiety levels (a psychological marker of stress) of leaders and non-leaders. Not only did leaders exercise more, smoke less, and wake up earlier, they also had significantly lower cortisol levels and self-reported stress. In a separate study of adults in Harvard executive education programs, those in higher leadership positions had both lower cortisol levels and anxiety when compared to lower level leaders. This effect was significantly moderated by the leaders’ perception of control. The results replicate findings in non-human primates. What is not known is what comes first: being worry free and therefore better able to become a leader, or the leadership position and concomitant reduced stress. The researchers postulate that those holding leadership positions perceive greater control in their lives and this perception is responsible for reducing anxiety. So, the next time you resist taking the reins on a project because of potential increased stress, it might be worth recognizing that once you settle into the position, you might find your concerns to be unfounded.

Noted by Leah H. Carr, BS, MS-III
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