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# Parental Refusal of Surgery in an Infant With Tricuspid Atresia

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We present a case of a fetal diagnosis of tricuspid atresia (TA). The pregnant woman and her husband requested that the baby be treated with only palliative care. The cardiologist did not think it would be appropriate to withhold life-prolonging surgery once the infant was born. The neonatologist argued that outcomes for TA are similar to those for hypoplastic left heart syndrome, and the standard practice at the institution was to allow parents to choose surgery or end-of-life care for those infants. The team requested an ethics consultation to assist in determining whether forgoing life-prolonging interventions in this case would be ethically supportable. In this article, we ask a pediatric intensivist, a pediatric cardiologist, and a neonatologist to discuss the ethics of withholding life-sustaining treatment of a baby with TA.

Advances in fetal diagnosis now allow couples who are having a baby to anticipate the decisions that will need to be made after birth. If the prenatal diagnosis is made early enough in pregnancy, then 1 option may be to terminate the pregnancy. When pregnancies are carried to term, then choices might be necessary about whether to pursue life-sustaining treatment or, instead, to provide only comfort-oriented palliative care. In the abstract, the ethical principles that should guide such decisions are clear. If the treatment is clearly beneficial, then the baby's right to treatment should outweigh parents' right to refuse. If, instead, the outcomes that are anticipated with treatment are ambiguous or uncertain, then the parents' choices determine the course of action. In practice, those principles are difficult to apply. Doctors may disagree about whether a particular treatment of a particular condition is sufficiently successful so that parental refusal should not be permitted. Tricuspid atresia (TA) is 1 such condition. In this Ethics Rounds, we present a case of TA and seek

expert commentary on the ethics from an intensivist, a cardiologist, and a neonatologist, all of whom are also bioethicists.

## THE CASE

A young married woman was pregnant with her first child. During routine prenatal care, an ultrasound was performed and there was concern that the child might have congenital heart disease. A fetal echocardiogram was performed at ~24 weeks' gestation that revealed tricuspid atresia (TA). The cardiologist met with the parents to discuss their son's condition, explaining the standard surgical approach and long-term prognosis. At home, the parents researched the proposed surgery on the Internet and learned that many parents decline this surgery. At their next appointment, the couple told the cardiologist that they did not want their baby to get the surgery and instead would focus on making him comfortable.

## abstract

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The perinatal team, including the cardiologist, neonatologist, obstetrician, and others, met to discuss the case. The cardiologist voiced concerns about the father's decision, stating that she did not think it would be appropriate to withhold life-prolonging surgery once the infant was born. The neonatologist argued that outcomes for TA are similar to those for hypoplastic left heart syndrome (HLHS), and the standard practice at the institution was to allow parents to choose surgery or end-of-life (EOL) care for those infants. Based on the principle that equal patients should be treated equally, he argued that allowing the parents to decline surgery would be appropriate. The team requested an ethics consultation to assist in determining whether forgoing life-prolonging interventions in this case would be ethically supportable.

#### **Alex Kon, MD, Pediatric Intensivist, Comments**

When making life-and-death choices for an infant, parents and providers must consider primarily the infant's best interest.<sup>1</sup> However, such decisions are highly value laden. Different parents, and different providers, may judge the same situation very differently. To some, the benefits of prolonging life, even for a short time in the face of significant morbidity, outweigh the burdens of even significant suffering. For others, minimizing suffering is a more important goal than prolonging life. In such cases, there is often no single right answer. Furthermore, although the best interests of the infant are central in decision-making, the interests of the parents, siblings, and other may also be considered.<sup>2,3</sup>

The American Academy of Pediatrics recognizes that most such decisions fall into a gray area in which several goals of care may be ethically permissible. The academy recommends that providers seek

to overrule parents only when parents make decisions that are clearly contrary to the infant's best interests.<sup>1,2</sup> Merely disagreeing with parents' values and preferences is insufficient. This standard requires that providers intervene only when parents make choices that are inconsistent with decisions reasonable people would make.

Providers lack the authority to unilaterally overrule parents who decline life-prolonging interventions (except in rare, emergent situations). When providers believe that parents are clearly acting contrary to the infant's best interests, they may seek a court order to provide therapy that they believe is necessary and appropriate. The decision to seek a court order should not be made lightly. In general, courts are more willing to authorize treatment over parental objection if the treatment is of short duration, there is consensus in the medical community regarding the medically appropriate treatment, the prognosis with treatment is favorable and there is a high likelihood that the child would have a relatively normal life, and without treatment there is a high likelihood that the child would die. With this understanding, we turn to the case at hand.

Infants with single-ventricle (SV) physiology such as TA and HLHS whose parents choose life-prolonging interventions follow a similar surgical course. Both cardiac lesions warrant surgical intervention within the first few days of life that includes placement of a surgical shunt to supply pulmonary blood flow (note that experts continue to develop new surgical and hybrid approaches for initial management). The initial surgery is usually followed by  $\geq 2$  additional cardiac surgeries, generally including a bidirectional cavopulmonary anastomosis (Glenn procedure) by 6 months of life and a Fontan procedure before 5 years of age. Many affected infants

have feeding difficulties and high caloric requirements for growth, so a gastrostomy tube is often needed to maintain adequate intake. Potential complications of multiple cardiac surgeries necessitating cardiopulmonary bypass include heart failure, arrhythmias (which may necessitate pacemaker placement), and stroke with resultant neurologic deficits. Furthermore, infants with SVs have a shorter than average life expectancy because of early heart and liver failure. Because infants with TA generally have an adequately sized left ventricle, in contrast to infants with HLHS, there may be reason to believe that infants with TA would be less likely to need cardiac transplantation later in life than infants with HLHS; however, there are insufficient data to draw firm conclusions.

As noted by the obstetrician, the standard for infants with HLHS generally is to allow parents to choose either life-prolonging interventions or EOL care. In the case of HLHS, the option of EOL care is generally considered ethically permissible because surgical outcomes are suboptimal (5-year survival is  $\sim 80\%$ ,<sup>4-7</sup> and there is significant risk of neurodevelopmental and other disorders among survivors<sup>8-15</sup>), experts in the field are divided between favoring life-prolonging interventions or EOL care,<sup>16-18</sup> and many believe that infants with HLHS endure significant suffering throughout their treatment course. For these reasons, although there remains debate about appropriate care for infants with HLHS,<sup>19-21</sup> in general parents are given the choice between life-prolonging interventions or EOL care.

Surgical outcomes for infants with TA are similar when compared with outcomes for HLHS. Although it is less well studied than HLHS, 5-year postoperative survival in TA is  $\sim 80\%$ ,<sup>22-25</sup> and survivors are also

at risk for neurodevelopmental and other disorders.<sup>26</sup> Therefore, when the obstetrician argued that infants with TA should be treated similarly to those with HLHS, his reasoning had some merit.

In this case, however, we must look not only at outcomes for infants who undergo life-prolonging interventions but also at outcomes for those whose parents decline life-prolonging intervention. In the case of HLHS, 95% of infants will die within the first month of life.<sup>27</sup> Because of the rapid demise of these infants, providers can deliver high-quality palliative care, and the pain and suffering of infants during the dying process can be minimized. Therefore, EOL care is generally considered a reasonable alternative to life-prolonging interventions for infants with HLHS. Data also suggest that when given complete information, parents are split regarding the choices they make.<sup>28–32</sup> Furthermore, when experts are asked what they themselves would choose for their own children, they too are split in their decisions.<sup>16–18</sup> Therefore, when parents of an infant with HLHS believe that the potential burdens of interventions outweigh the potential benefits, they are generally allowed to opt for EOL care.

In contrast, infants with TA who do not undergo life-prolonging interventions have a significantly different prognosis. Many untreated infants with TA survive past the newborn period. Data suggest that ~50% of these infants will die in the first year of life, and the remaining children will survive for several years (and potentially into young adulthood).<sup>33,34</sup> The natural course of untreated TA is that children's pulmonary vascular resistance (PVR) gradually increases over months to years. As the PVR increases, children develop worsening hypoxemia. Because over time the increased PVR becomes nonreversible, these children are not good candidates

for delayed surgical intervention. Therefore, left untreated the child with TA will probably experience a slow decline in health during the toddler and school-age years, leading to a long dying process. Because of the length of the dying process, palliative interventions are less likely to be effective, and there is a high risk that the child will suffer significantly.

Because an infant with TA is likely to suffer significantly if surgery is withheld, a decision to forgo life-prolonging interventions cannot be considered consistent with that child's best interest. Although the obstetrician was correct in noting that similar patients should be treated similarly, the difference in the natural course of TA and HLHS leads to different ethically permissible options for affected infants. Therefore, the parents should be educated about the natural course for their son if life-prolonging interventions were not provided, and the providers should explain why they believe that such a decision is not appropriate. If the parents persist in their refusal to give permission for appropriate intervention, the team should seek a court order to authorize medically indicated treatment.

In general, when we consider a child's best interests, we tend to focus solely on the potential benefits and burdens of the proposed intervention. This case illustrates the importance of considering not only the potential benefits and burdens of the proposed treatment but also the potential benefits and burdens of the alternatives, including the option to forgo life-prolonging interventions. In some cases, such as this, the alternatives are so clearly contrary to the patient's best interests that a decision to forgo life-prolonging interventions is not ethically supportable.

### Angira Patel, MD, MPH, Pediatric Cardiologist, Comments

Congenital heart diseases consisting of a functional SV (eg, HLHS, TA) require staged palliation concluding with a Fontan operation. Over the last 40 years, surgical technique has evolved and significant strides have been made to reduce mortality. Historically, HLHS has been technically more challenging with higher mortality than other types of SVs, and EOL care has been accepted an ethically permissible option. Contemporary results, especially at high-volume technically excellent centers, show long-term survival for HLHS approaching that of other forms of SV such as TA. Data are difficult to extrapolate because of center-related and era effects, but best estimates range from 80% to 85% for 10-year survival for both HLHS and TA.<sup>7,35</sup> In actuality, all functional SVs have a similar long-term burden of intensive surgical and medical therapies. The difference in mortality between HLHS and TA is negligible and no longer sufficient to treat the 2 diagnoses as different entities for an ethical analysis. Specifically, life-prolonging treatment involves  $\geq 2$  surgeries in the first 3 years of life, cardiac catheterization and interventions, and lifelong need for monitoring and treatment of complications including premature death, ventricular failure, thromboembolic disease, arrhythmia, liver disease, protein-losing enteropathy, and potential need for heart transplantation. These interventions are palliative and not curative. However, the timing of death without intervention for HLHS may be different than for TA; infants with HLHS generally die within 2 to 4 weeks without intervention, but a small minority of infants with TA (depending on underlying anatomy) can survive longer.<sup>36</sup>

Given surgical and medical advances leading to similar survival outcomes for HLHS and TA with the same

burden of long-term morbidities, the question now becomes, “Is it ethically permissible to allow a family to forgo life-prolonging interventions for a child with any SV diagnosis? Do we honor the choice of these parents that is probably based on their own family’s individual values and preferences?” I say yes.

Parents are appropriately tasked with the role of surrogate decision-makers for their children because they are in the best position to consider the relative weight of risks and benefits of therapies. Disagreements within the medical community occur when there is a question of whether the parents are acting in the child’s best interest. For this child with TA, there are both early and ongoing morbidities resulting in what parents perceive to be suffering and pain for their child. In defining boundaries of what is medically and ethically acceptable, we must weigh the morbidity and burden of invasive and intensive therapy against the potential benefits of those interventions. An SV palliation by definition involves lifelong invasive medical care that may not be the right choice for some families. These parents, understanding both known and unknown risks and benefits and incorporating their own values and preferences, have decided that an SV palliation is not in the best interest of their child. There is no “right” answer here. More leeway to parental decision-making should be allowed when the burdens of expected therapy are real and significant, as they are in this case. Allowing the family to forgo life-prolonging therapies is ethically supportable.

However, the parents of this infant ought to be counseled specifically on natural history of TA without intervention (which may be different from HLHS). If the infant survives >6 months, a reevaluation of options would be anticipated, including consideration of surgical

therapies or continued EOL care. The subtleties of the diagnosis and the possibility that death may not come quickly necessitate a more nuanced discussion.

I acknowledge the moral distress of the cardiologist regarding what is best for the child in the setting of high probability for survival (despite the intensive medical and surgical interventions and potential morbidities). In this case, the cardiologist may have to endure this distress yet stand with the family. More importantly, the role of the treating cardiologist continues to remain crucial: to provide ongoing support, conveying known information about how their child will die without treatment and what can be done to provide comfort to optimize their time together.

#### **Steven Leuthner, MD, Neonatologist, Comments**

As the ethics consultant, I would begin by clarifying 2 issues. The first is the accuracy of the prenatal diagnosis. One would not want to counsel and have the parents make this prenatal decision unless there is a reasonable degree of accuracy in the diagnosis. Although there are a variety of anatomic situations with TA, the published accuracy of the diagnosis is excellent, at 97%.<sup>24</sup> Other independent predictors of poorer outcome, such as chromosomal or syndromic findings, might be of value in the prenatal counseling period as well.

The second issue to clarify would be the Internet information the parents are basing their decision on, for the single reason of making sure they are basing their decision on appropriate data and similar cases. It would not be surprising if this Internet information was more about HLHS than TA. If there was some misunderstanding or inappropriate comparisons were being made, simply reviewing this information might lead to consensus.

So let us assume the parents’ information is about SV issues and more likely HLHS, the diagnosis is correct, and there is no chromosomal abnormality or other syndrome. The question then is whether HLHS and TA are equivalent medically and therefore ethically. Some might argue that the presentation of the infant and the different forms of TA are applicable. For instance, approximately one-third of TA cases have accompanying transposition of the great arteries that would necessitate a Norwood palliation in the newborn period, and two-thirds are not transposed and have a pulmonary outflow track problem that would necessitate a Blalock–Taussig shunt or pulmonary band. The fact remains that no matter what the initial surgery would be, the eventual surgical goal is for an SV Fontan procedure. So what is the outcome difference for the Fontan physiology if a patient has a morphologic right rather than left ventricle, as well as long-term outcome of the Fontan procedure? This gets at the justice issue the neonatologist is suggesting, that if for HLHS one would support providing palliative care, then why not for TA?

Here the data seem mixed. One study suggests that 10-year survival is  $\leq 85\%$  of those born with a dominant left ventricle, as opposed to only 65% for those born with a dominant right ventricle.<sup>37</sup> A much larger 40-year follow-up of the Fontan operation, which includes a large group of patients with TA, shows the 10-, 20-, and 30-year freedom from death or cardiac transplant being 73%, 59%, and 40%, respectively.<sup>35</sup> They describe the many complications of the Fontan, including premature death, ventricular failure, thromboembolic disease, arrhythmia, liver disease, and protein-losing enteropathy. It must be acknowledged that this is a significant chronic illness with physical and neurocognitive



limitations, necessitating procedures beyond the 3-stage repair if the child survives the surgeries. Acknowledging these issues, the data suggest that at this time it is reasonable to consider HLHS and TA as equivalent SV lesions for ethical decision-making.

If the lesions are equivalent, then the ethical principle of justice, or treating equal patients equally, does come into play. Interestingly, as the survival of infants with HLHS undergoing the staged repair has improved, there continues to be debate about whether parents should still be offered palliative care.<sup>19,20</sup> Essentially using the justice argument, it is often suggested that with improved HLHS survival and outcomes there are other cardiac cases with worse outcomes, yet palliative care might not be offered in those. When thinking of justice in this way, we should beware of faulty reasoning, because 2 wrongs would not make a decision right. The ethical literature continues to show although the outcomes for HLHS have improved, and there might be a recommended medical plan, they have not yet reached the level for which palliative care is not an acceptable choice that parents should be informed about.<sup>21,38,39</sup> In this case, and at this institution, the standard practice is to allow parents of infants with HLHS to choose surgery or EOL care. The neonatologist is correct to suggest that based on justice, because the medical conditions are reasonably equivalent, if it is reasonable to offer EOL care in cases of HLHS, it is reasonable to offer it in cases of TA.

The underlying ethical question is whether the survival and quality of life for these infants meet a threshold for which the medical team should consider intervening legally to override parental authority. In the case of known complex staged repair, ethical consistency would require that a decision to override

parental choice is not only for the initial neonatal surgery but for all anticipated procedures. For the SV path should this also include transplantation if the Fontan failed? Overriding parental authority early also compromises issues of trust and long-term care for a medically complicated child. Ethical frameworks including the best interest standard, reasonable person standard, or parental discretion all support a family having the right to make a reasonable decision, even if they are in the minority view. They maintain that family values and family impact are reasonable to consider. These are high-risk procedures with significant burdens for the infant and family, not only in the neonatal period but throughout life. If we would not mandate things all the way through, then EOL care at any stage seems a permissible choice for a family.

Remembering that this is a prenatal case, it is important to appreciate that a prenatal mandate to intervene after birth could put the family in a position to consider other obstetrical options. These could include termination of pregnancy, although this can only occur in a few places nationally at this gestational age, or even arranging a delivery plan to avoid the institution. This would not serve the infant best, because in the end there must be a trusting relationship to help develop a palliative care plan for this infant who, depending on the outflow track anatomy, could have different care needs and projections of neonatal death.

#### **John D. Lantos, MD, Bioethicist, Comments**

Bioethics is often criticized for not only having no right answers but for not even having a method of getting to a right answer in difficult cases. Instead, the critics say, there are just equally powerful arguments on both sides. That is sometimes true. These

commentaries illustrate the ways in which thoughtful people can look at the same data or the same case and come up with different responses about the appropriate course of action. That does not always happen. Arguments by bioethicists have changed the ways in which we respond to a wide variety of cases. We used to permit parents to refuse life-saving surgery for babies with trisomy 21. We used to refuse to perform life-saving surgery on babies with trisomy 18. The borderline of viability has shifted slowly but steadily and with it the threshold for mandating life-sustaining treatment of premature infants. When disagreements persist, it suggests a lack of consensus in the professional community. In such cases, the proper thing is to defer to parents. Careful consideration of the arguments can help us counsel parents and ensure that their decisions are informed decisions. In that sense, disagreements between bioethicists are no different, and perhaps no more common, than disagreements between cardiologists, policymakers, or other experts. They signal the limits of our collective ability to know what is best and the intensity and integrity of our efforts to keep finding out.

#### **ABBREVIATIONS**

EOL: end of life  
HLHS: hypoplastic left heart syndrome  
PVR: pulmonary vascular resistance  
SV: single ventricle  
TA: tricuspid atresia

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Alexander A. Kon, Angira Patel, Steven Leuthner and John D. Lantos

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