Case 3—When the Family Won't Decide

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TN was a baby boy born with multiple congenital anomalies, including alobar holoprosencephaly (HPE), where the brain does not develop nor divide into right and left lobes.[1] These cerebral anomalies were diagnosed in utero. He also had a cleft lip/palate, feeding problems, severe gastro-esophageal reflux (GER), failure to thrive, and seizure disorder. TN had the abnormal muscle tone and impaired motor abilities present in virtually all individuals with HPE. TN's mother (age 16) and father (age 20) lived separately with their respective parents. TN was conceived during their very brief romantic relationship, which neither desired to continue. TN's mother dropped out of high school when she found out she was pregnant, and her attempts to complete a High School Equivalency Diploma Program (GED) were not consistently productive, no doubt complicated by the birth of a seriously ill baby. TN's father had also dropped out of high school before graduating, and was working part-time as a pizza delivery driver.

During a follow-up visit to the hospital's pediatric clinic when TN was two months old, the neonatologist recommended that his parents consider allowing a PEG tube[2] to be placed to help insure TN was receiving adequate nutrition and hydration. TN's parents refused to provide consent, and his mother said he was drinking a small bottle of formula (approximately five ounces) every three hours or so; however, TN had marginal and sometimes no weight gain documented during subsequent clinic follow-up visits.

When TN was six months old, he was admitted to the hospital after a clinic visit for a three-day calorie count, which would help re-evaluate the benefits of having a PEG tube placed. The planned three-day hospital stay was extended due to TN's temperature instability, electrolyte imbalances, and increased needs for care. TN's young mother visited her baby almost every evening, but was not there in the early morning hours when her son's doctors made their rounds. His father visited irregularly and infrequently. On day eight, TN began aspirating formula, and he was transferred to the Pediatric Intensive Care Unit (PICU). The pediatric gastroenterologist strongly recommended placing a PEG tube, and after several attempts to contact TN's mother about his deteriorating condition, was finally able

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to reach her by telephone. "I don't want my baby to have any more things done to him," she said, "We just want him back home with us." She reluctantly agreed to the PEG tube placement, and told her parents that the doctor kept saying it was urgent and that she needed to decide right away.

The PEG tube was immediately placed, but it did little to prevent TN's continued decline over the course of his hospitalization. He required intubation and became ventilator dependent, and despite the PEG tube, his feeding issues continued. He developed life-threatening anemia (hemoglobin <4; normal range is 10.5–14). The attending physician left numerous messages for multiple family members over the course of nearly 20 h, attempting to obtain consent for an emergency blood transfusion. Risk Management was contacted, and they stated that if TN's life was in danger, the indicated blood transfusion should be given unless the patient's mother could be reached and she specifically and emphatically said "No." If his mother could not be reached, or if she could not make an immediate decision, the patient should be transfused as necessary to save his life. TN's family did finally come to his bedside that evening and met with the attending physician. TN's mother agreed to a "one-time only" blood transfusion, which was ordered.

The blood transfusion stabilized the baby's hemoglobin, but the next day pediatric neurosurgery was consulted because of TN's worsening neurological status. The neurosurgeons found that TN had no gag, cough or corneal reflexes, and only minor respiratory effort. Their assessment, recorded in the baby's electronic medical record, stated: "This baby has severe anomalies with hypothalamic failure. Prognosis is dismal. The anomaly is trying to run its natural course. Intervention will be to no avail." TN was then 10 months old.

TN's mother was only minimally interactive at that point; her earlier regular visits to the hospital had become almost non-existent, and his father stopped visiting all together. The attending physician again attempted to encourage TN's parents to come to the hospital so his worsening condition could be better explained to them. After many phone calls and voice mail and text messages, TN's parents agreed to come to the hospital to meet with their son's physicians. During the initial meeting, his parents explained that TN had been doing "fine" until they were "forced" to consent to the PEG tube placement and that they believed the trauma from that procedure had caused the complications leading to TN's present decline. Subsequent meetings were scheduled, but TN's father refused to attend and would also not answer his phone. TN's maternal grandparents and maternal aunt eventually attended, along with the baby's mother, who said little. The baby's family members were antagonistic and hostile, and they continued to blame the hospital and physicians for TN's declining health and precarious status.

The physicians and nurses involved in TN's care believed that his family had never been realistic about the severity of the baby's condition. At the end of another lengthy meeting between members of the extended family and the medical team, including the palliative care service, the maternal aunt stated she felt the decisions had been taken out of their hands when TN's mother was "forced" to consent to the PEG tube; she had, after all, declined it in the past. She also reiterated that the family would have preferred for the baby to be at home, and she said they were never given

that option. The aunt then said that the family would need to pray for guidance before making any decisions to limit or change TN's care to comfort measures only, despite the neurologists' note about the baby's dismal prognosis. The family members further stated that they did not believe that TN was suffering, and they said, "God would not let our baby suffer and will take him when it is time."

One month later it became clear TN was near death. Urgent calls were placed one afternoon as well as the next morning with no success in reaching any of the family members. Convinced there would be no response from them, the attending physician removed the ventilator, and TN died within minutes. The family finally came in later that day, and were given private time at TN's bedside and support from pastoral care and child life services.

Discussion Questions

1. How might better communication between the medical staff and TN's family have improved his care?
2. Did the medical team handle TN's case appropriately?
3. Under what circumstances might it be appropriate for medical staff to make end-of-life decisions for a patient whose decision makers cannot be reached?
4. How should hospitals deal with parents or other family members who refuse to take on an active surrogate decision-making role?

A Bioethicist Responds

Little TN died at less than one year of age after what must have been a very unpleasant life suffering from feeding problems, severe gastro-esophageal reflux, failure to thrive and a seizure disorder, not to mention having to undergo PEG tube placement as well as frequent heel-sticks for blood-level monitoring. Add to this physical discomfort, his parents' general unresponsiveness to calls from physicians and hospital staff, and their apparent lack of interest in working with the health care team in their son's best interests, the overall portrait of TN is a sad one indeed. One wonders if it could not have been better.

HPE is a serious condition, most often fatal, and it is important to understand the facts surrounding it. Mortality is high for all newborns with HPE, although some of them survive beyond the neonatal period, with a few even surviving many years. Alobar HPE is the severest form, but studies have shown that some 20–30% of this group may even survive beyond one year. The higher mortality rates among the alobar group tend to correlate with the severity of concomitant factors such as brain malformation, facial malformation, chromosomal abnormalities and multiple congenital anomalies. Developmental disabilities are present in virtually all persons with HPE, but they are severest in the alobar group, where children generally make little progress in development and tend to have profound global impairments. Children with HPE also commonly suffer from other problems such as hydrocephalus, seizures, motor impairment, dysphagia, pulmonary and gastrointestinal problems, and hypothalamic and endocrine dysfunction (Levey et al. 2010).

It is evident from the case narrative that TN had numerous significant deficits from birth. Without knowing more specific medical details about his case, it would be difficult to say whether he might have survived beyond one year of life, yet it is understandable why the neonatologist recommended PEG tube placement at two months, and then again at six; the fact that TN was surviving, albeit with inadequate nutrition, was probably sufficient reason for the neonatologist to want to offer him any reasonable means toward continued life on both occasions. What TN did not have in his favor, it appears, was the kind of family support that was needed. His parents refused to give consent for the PEG at two months; his mother refused again at six months when first asked to consent, but she eventually agreed "because of the urgency of the doctor's request," as she reportedly told her own parents. Once the PEG was in place, TN's mother and father visited him less and less, and they became less responsive to phone calls. When the baby's situation became critical and the parents were summoned to a meeting with the medical team, though only after repeated efforts, TN's mother and father blamed his problems on the trauma of PEG tube placement. Subsequent meetings were found to be even less successful, with the father refusing to attend at all and other family members becoming even more hostile to the medical care team. Given the attitudes and behaviors of his family as he entered his tenth month of life, TN's chances of survival certainly could not have appeared good. Furthermore, there was no promising home life into which he would be likely to return. It is little wonder that things developed as they did in the eleventh month of TN's life.

Infants as critically ill as TN simply cannot be expected to survive without proper care and support on multiple levels. High early mortality is associated with severe cases of HPE, and the case of TN would surely qualify as one of them. As Kaliaperumal et al. commented recently, "Care of children surviving with HPE requires multidisciplinary input from different medical and surgical specialties including rehabilitation to ensure optimal patient care and parental support" (2016, 808). Gupta et al., suggest that parents should be counseled prenatally about the poor prognosis of babies with HPE—only 50% of babies with alobar HPE will survive four to five months, and only 20% of those cases will live to twelve months of age. If the pregnancy is continued, the baby should be immediately referred for physical and occupational therapies (2010). Redlinger-Grosse and her colleagues have pointed out that even when HPE is detected prenatally and parents are told of the poor prognosis for their baby, it is often given as an uncertain one (2002). Likewise, they say, parents of children born and diagnosed antenatally with the condition are often told their children will die within days or weeks of birth (Redlinger-Grosse et al. 2002). Redlinger-Grosse et al., focus on the complex decision-making process that parents of a prenatally diagnosed HPE infant go through in deciding whether to continue or terminate the pregnancy, and the many informational, emotional and supportive needs parents will require in order to help ensure optimal outcomes for their HPE children. They note, in any case, that the majority of parents receiving a prenatal diagnosis of a serious abnormality terminate their pregnancies. Once again, the case narrative of TN amply demonstrates that neither he nor his parents had the kinds of support they needed, if for no other

reason than his parents' refusal to accept the advice and counsel of those providing medical care for their son.

The most obvious question to be asked in this case is whether the health care team made the correct decision in removing the ventilator as TN lay near death and urgent calls to the family went unanswered. One might suggest that nothing would have been lost, and no harm done, had comfort measures been initiated, or titrated upwards, while further efforts were made to locate the mother, perhaps even with police assistance. We do not have all the medical facts of the situation at that time, however, and there may have been reasons that this approach was not feasible. On the other hand, even if it were, TN's physicians had by this time come to know enough about his parents and extended family to know that there was every likelihood of pushback, rancor, hostility, or another failed attempt at meaningful communication resulting from continued efforts, perhaps even the possibility that they would be accused of having caused the infant's crisis or of wanting to "kill" him were they to propose removing the ventilator. Perhaps they chose just to go ahead and withdraw the ventilator without securing parental consent since it seemed clear TN was nearing the end of his life anyway, and it was just as convenient the parents were difficult to reach this time?

A more likely explanation, however, and one which can be supported ethically, is that the decision to withdraw was likely made on a best interests standard, acting to do the most good for, and to cause the least amount of harm to, an infant who had already suffered greatly throughout his entire life, knowing that there was no possible way to cure or even ameliorate his situation, and knowing also that death would occur within a matter of days. There is simply no reason under the circumstances of this case to continue what has become futile care.

The only other option open to the team would have been to ask for a *guardian ad litem*, but that would not have been a wise choice, at least in this writer's opinion, for this reason: *guardians ad litem* generally act very conservatively on the part of the patients in whose interests they serve. And there is nothing at all wrong in this approach, for that is their job. This approach may not always produce the most ethical result, however, and if a guardian had been appointed in the case of TN, there is every chance he or she would have refused to consent to the withdrawal of the ventilator while yet further attempts to locate the parents were made. There would have been nothing wrong with this in principle, but if much time had passed, and the medical team had every reason to think that it could well have, the effect would only have been to add to the already great suffering of TN. It is true, in fact, that the family came to the hospital later that day, but given the way they had behaved in the past to their son, as well as toward the medical team throughout his hospitalization, the decision to withdraw life support was entirely justified at the time it was made.

A Health Communication Scholar Responds

This case involves two young people—not a couple—attempting to cope with a likely unplanned pregnancy and a very sick baby. Young people often face

hardships in adjusting to parenthood, including lack of education, limited financial resources, inadequate parenting skills, unstable relationships, and transportation and housing issues. The present case adds to these common challenges the particularly difficult circumstances of coping with the severity and irreversibility of TN's medical condition.

TN was diagnosed with a rare congenital abnormality, holoprosencephaly (HPE), estimated to affect between 1 in 5000–10,000 live births. Risk factors include maternal diabetes, infections during pregnancy (syphilis, toxoplasmosis, rubella, herpes, cytomegalovirus), and various drugs taken during pregnancy (alcohol, aspirin, lithium, thorazine, anticonvulsants, hormones, retinoic acid), but often no cause can be determined.[3] Most pregnancies with a fetus diagnosed with HPE end in miscarriage; only 3% survive to delivery and the majority of those babies do not survive past the first six months of life. It is very unlikely that TN would have lived to 11 months of age without the aggressive interventions proposed by the medical team, to which his young parents only reluctantly consented.

We have no information about the pre-natal care TN's mother received, or if she was exposed to any of the possible risk factors for HPE. We only have the vague comment that "cerebral anomalies were diagnosed in utero." Was TN's mother apprised of the very serious nature of these anomalies, including that they were sure to seriously foreshorten TN's life? Was she offered counseling to determine her willingness to continue the pregnancy, or given the option to terminate the pregnancy? Surely these young parents would have benefitted from pre-delivery counseling so they would have been in a better position to contribute to decisions about fetal and neonatal management of their baby. The case narrative gives us no clues about whether or to what extent such counseling took place. This might have been the first point at which decision-making authority was transferred from these young persons to the medical team.

Given TN's likely prognosis of death within six months, the pediatric intensive care team's recommendation to place a PEG tube was curious. Even without assisted feeding, TN had outlived his probable prognosis of death within 6 months, but how much longer did the medical team expect him to live, and at what quality of life? TN's mother had refused PEG tube placement previously, and she only reluctantly agreed the second time the issue arose, when TN began aspirating formula and the pediatric gastroenterologist determined that a decision needed to be made urgently. The parents had a right at this point to refuse this intervention due to the seriousness of TN's continued medical challenges and the inability of aggressive medical care to address them. With the right support, they may have opted for comfort or palliative care in the PICU. Given his prognosis of likely death within 6 months, another option might even have been hospice care delivered at home, especially since the family indicated at a later meeting their desire to have TN home with them. Maybe TN's mother was hesitant to agree to the PEG because she realized that one intervention inevitably led to subsequent interventions, none of which would contribute materially to her baby's quality of life or address his underlying medical condition. Maybe the family had started to accept his limited life expectancy and his diminished quality of life. It is difficult to determine the

motivations of the pediatric intensivists—did they see TN's feeding issues as just another challenge that must be met at all costs, simply because TN was a baby in the PICU as Chiswick (2008) suggested? The third time this young family faced pressure to make an "emergency" medical decision for TN involved the blood transfusion, which once again solved a short-term problem but did little more than prolong the baby's suffering. We do not know TN's mother's health insurance status, but she was young enough to still be covered under her parents' medical insurance policy. We can fervently hope that financial incentives on the part of the medical care team were not a significant factor in this case. Nonetheless, neonatal and pediatric intensive care units have become increasingly important profit centers since the 1990s (Harrison 2008; Simpson 1999; Silverman 1993). Hospital systems have become, in the words of pediatrician Lantos, "hooked on neonatology," with clear incentives to expand medical care options to this new population (2001). This fact, coupled with the advances in technology for premature babies as well as those born with disabling conditions, certainly contribute to the overall ethic of aggressive intervention that characterizes U.S. hospital care.

We want to believe that parents have the autonomy and authority to make decisions for their seriously ill children, but there is evidence that shows families often have little informed input in decisions regarding resuscitation or treatment (Harrison 2008; Keenen et al. 2005; Partridge et al. 2001). Although most of these studies concerned infants born prematurely, their findings also apply to infants born with serious disabilities. There are many players in neonatal and pediatric intensive care dramas, and the parents may be the least informed about their child's medical condition or of their legitimate medical and ethical options. A survey of 149 practicing neonatologists in New England showed more than half saw their role as providing information in a neutral manner; far fewer saw their role as helping parents balance the risks and benefits of treatment or the familial or social consequences of their decisions (Bastek et al. 2005). Counseling was found to be largely 'directive' and focused on short-term issues, and parental options to forego resuscitation or other aggressive interventions were rarely mentioned. Another study showed that neonatologists who provided information in a neutral manner left parents feeling isolated; parents preferred support and engagement with the medical care team in the decision-making process (Payot et al. 2007).

It is not surprising that there was frustration and hostility on both sides. The frustration of the medical staff is understandable. Decisions needed to be made and the rightful parental decision makers could not be reached. In the absence of consistent parental or extended family involvement, who should take on that responsibility? The family's level of involvement was just enough to make a referral to a child protective services agency unnecessary. The limited parental involvement left the medical staff to make decisions for this vulnerable patient.

From the perspective of TN's parents and extended family, it appears that several times during their baby's short life they either did not have sufficient information about their options, or decisions were forced because of an emergent medical crisis or perceived urgency on the part of the medical staff. TN's parents might have felt that since the physicians insisted on the PEG tube placement

(instead of allowing a natural death at that point)—that decisions had in fact been taken out of their control. Instead of a blood transfusion, to which his mother only reluctantly agreed, why not discuss palliative care, since the very next day the neurologists documented TN's prognosis as "dismal?" During one of the last family meetings, the maternal aunt stated they wanted TN home and that they would pray over the decision to accept comfort care for TN. Why wasn't that allowed? Hospice care could surely have been provided in the home to support TN and his family, medically, psychologically, and spiritually. It seems that TN's prognosis was always poor, so it must have been confusing for these young parents to receive messages compelling them to consent to the PEG tube, and to the blood transfusion, and almost simultaneously to have his prognosis described as "dismal". And the medical staff members' decision to remove TN from life support without his family present seems particularly precipitous.

There are lessons from this sad case that might be helpful in minimizing the suffering and frustration of all involved in future similar cases. Chiswick reminds us to foreground the best interests of the infant (2008). Actions and decisions can focus on issues that infants could have an interest in, such as the degree of pain and suffering involved in their care, the futility of medical intervention, and the likelihood of survival free of serious disability. The best interests of an infant are inevitably based on the perceptions of others, including parents and medical staff, and family-centered neonatal care should be based on open and honest communication between parents and professionals on medical and ethical issues (Harrison 1993). It is not acceptable to give information vaguely or euphemistically; even young parents should not be shielded from information about uncertainties or controversies relevant to their child's care. The young age of TN's mother was a factor in this case, but it appears that she did have the support of her extended family to help interpret the information given and assess the choices to be made. Perhaps the social worker involved in this case should have visited TN's mother and her parents at their home. Hospitals, especially intensive care units, are foreign and hostile territories to those unaccustomed to their sounds, smells, and technology.

We should also expect some disconnects in terms of attributions and expectations when we confront a young family with a very difficult set of choices and decisions to make. Patients and medical professionals inhabit different social worlds. The nurses and doctors in the NICU and PICU routinely encounter babies and young children with serious, life threatening, and disfiguring medical conditions. Those outside of the medical professions are unlikely ever to be in contact with a baby as sick or as disfigured as TN.[4] The young parents here needed to be made to feel a part of the medical care team, working together with medical professionals to alleviate their baby's pain and suffering, ensuring the safety and efficacy of treatments, and learning parenting and decision making skills. Above all, parents need to have the option and support to say 'no' to burdensome treatments for their children. In this case, TN's mother should have been given a chance to decide whether to carry the pregnancy to term, as well as the risks and benefits of PEG tube insertion and other medical interventions. Neonatologists should be

realistic about the resources that are or are not available to parents in decision making and caring for seriously ill newborns. The default option in the hospital setting, even more so in intensive care units, is always more treatment. In some cases, the better alternative, the one that adequately accounts for the baby's best interests and the parents' preferences, might be comfort measures, which could have been started in the delivery room if not sooner.

Notes

[1]Holoprosencephaly (HPE) is a birth defect that occurs during the first few weeks of intrauterine life. HPE is a disorder in which the fetal brain does not grow forward and divide as it is supposed to during early pregnancy (incomplete cleavage of the embryonic forebrain/failure of the prosencephalon to cleave into the cerebral and lateral hemispheres). The most severe form of HPE is alobar, where the brain is not divided and there are severe abnormalities, including absence of the interhemispheric fissure; a single primitive ventricle; fused thalami; and absent third ventricle, olfactory bulbs and tracts and optic tracts.

[2]Percutaneous endoscopic gastrostomy (or PEG) is an endoscopic medical procedure in which a tube (PEG tube) is passed into a patient's stomach through the abdominal wall, most commonly to provide a means of feeding when oral intake is not adequate.

[3]For more information, see The Carter Centers for Brain Research in Holoprosencephaly and Related Malformations (https://www.carterdatabase.org/hpe/about/).

[4]Other facial abnormalities present in many children diagnosed with HPE include a flat single-nostril nose (cebocephaly), close-set eyes (hypotelorism), or just one upper middle tooth (single maxillary central incisor). More severe facial deformities may include a single central eye (cyclopia), a nose located on the forehead (proboscis), or missing facial features.

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