

## **Virtual Mentor**

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### **CLINICAL CASE**

#### **“Please Let Me Hear My Son Cry Once”**

Commentary by Wynne Morrison, MD, MBE

Norma had two goals in life: to become a parent and a gynecologist. She achieved the latter and, to boot, married a classmate, George. Her first dream seemed stymied, however, by their long hospital hours, exhaustion, and career demands. When Norma unexpectedly became pregnant at the age of 35, she and George were overjoyed, but also concerned about the risks associated with her relatively advanced age of conception—concern that turned out to be sadly justified when their unborn child was found to have trisomy 13, or Patau syndrome. George and Norma discussed options, including aborting the fetus, but decided to move forward with the pregnancy. Norma prayed every night, “Oh God, please let me hear my son cry once...that is all I want...the sound of my own child in this world.”

Norma’s prayers were answered and their son was born. Bob had a severe cleft lip and palate, but Norma and George thought their son was the most beautiful child in the world. Due to their medical competence, they were eventually able to take their child home. Because she spent so much time with her son, Norma quickly became familiar with all of his unique sounds and was quick to notice the onset of short apneic episodes. She could not tell whether Bob was experiencing pain during these attacks, but administered the morphine prescribed by Dr. Moy when she believed he was in pain. The events increased in frequency and duration, and, eventually, Dr. Moy began to advise against using morphine out of concern that this pain relief might result in respiratory failure.

When Bob was 3 months of age, conflict between Norma and Dr. Moy surfaced again when Norma requested that Bob be given his normal childhood vaccinations. Dr. Moy explained that he felt these were unnecessary and might increase morbidity. He expressed his regret at not having been trained for situations like this, and he knew there were many clinical and ethical issues at play.

When Norma called Dr. Moy to report that Bob had developed a fever, Dr. Moy told her to take him to the local emergency room immediately. There, after a taking a history and learning of the trisomy 13 diagnosis, the ED physician told Norma, “Your son is dying. There is little we can do to help him.” Dismayed, Norma pushed him to proceed with a physical examination, upon which the physician, to his surprise, found that Bob had an acute otitis media infection. He asked, “So do you want to treat his infection? Also, does your son have DNR orders?”

### **Commentary**

Trisomy 13 syndrome is a rare genetic disorder in which the affected patient carries three copies, rather than two, of chromosome 13. Common clinical features include a cleft lip or cleft palate, cardiac anomalies, scalp defects, microcephaly, developmental delay, seizures, frequent apnea, skeletal anomalies, and other anatomic defects [1]. Life expectancy is, on average, a few months of age, although there are reports in the literature of “long-term” survival into the teenage years [2-4]. With recent advances in medical therapy and our improving ability to support children with chronic illnesses, it is likely that clinicians in many fields of practice will encounter children living with syndromes like trisomy 13 and will have to help families decide what interventions are appropriate to pursue.

In the past, most parents of children with trisomy 13 were told that the syndrome was lethal and that interventions should focus only on keeping the child comfortable. Aggressive support, such as invasive procedures like cardiac surgery or tracheostomy, was generally not offered. Koogler et al. have argued that the ubiquity of this approach made the “lethal” label something of a self-fulfilling prophecy [5]. Clinician attitudes have shifted somewhat over time, especially as it has become much more common to provide interventions like open heart surgery to children with less severe chromosomal defects, such as trisomy 21 (Down syndrome). Many physicians may still feel that it is in the best interests of children with trisomy 13 to provide comfort measures only, but the current standard is to discuss options with the parents rather than making unilateral decisions.

The physicians in this case are struggling with how to discuss these issues with parents. Dr. Moy, the primary physician, is taking a palliative approach, providing morphine for comfort to the infant, a therapy that would be contraindicated for a child with apnea whom one expected to survive. It seems that the family is comfortable with this plan and feels that the morphine is helping, but it is unclear how specific the discussions have been about goals of care. Is comfort at this point more important to them than prolonging life? If Bob stops breathing, would they want intubation or resuscitative attempts? If he is dying, would they be more comfortable having him at home or in the hospital? Having early, explicit conversations to set mutually agreed-upon goals can help ensure that all parties are working toward the same ends and increase the comfort of the primary care physician with providing interventions like morphine.

Even when such conversations have happened, however, communication across the health care spectrum can be a challenge. The treating physician in the emergency department may not be aware of the discussions the family has had with the primary care physician or may not understand the plan that is being pursued. Such lack of information can lead to unwanted interventions, or, conversely, to inaccurate assumptions that no interventions are to be provided, as appears to have happened in this case. A phone call from the primary physician to the emergency department attending physician as the family is on their way in would be extremely helpful in providing context.

## **Understanding Family Treatment Goals and Preferences**

It would be a mistake to assume that a family's preferences will remain unchanged as a child's clinical condition changes; however, it would still put the family more at ease, and probably the emergency department physician as well, if he could say to them, "Dr. Moy tells me that you have decided to keep Bob at home if at all possible rather than rehospitalizing him. Is that still your preference?" Their wishes at this time still need to be ascertained, but the conversation does not need to begin in a vacuum.

This family's goals may also shift as they spend time with their child and bond with him. They will be more familiar than anyone on the health care team with their child's day-to-day life, particularly whether he is conscious of his surroundings and whether he is suffering. Physicians may assume that the parents are simply waiting for their child to die, but they may very well also treasure what little time they have with him. It might be a perfectly reasonable goal to try to extend his life as long as possible if the interventions required to do so do not cause too much pain or distress.

It is important for many families of children with severe disabilities that their child be accorded the same respect that a healthy child would. Bob definitely deserves a physical exam in the emergency department to attempt to diagnose a minor illness. His parents may choose to use antibiotics for an easily treatable condition like otitis media if they believe that he currently experiences more contentment than suffering. Some families may agree to a feeding tube for a child with trisomy 13 who has difficulty with oral feeding. Such decisions must be constantly reexamined as the clinical course progresses. Many families draw the line at invasive procedures, deciding not to repair congenital heart defects or undertake airway interventions such as tracheostomies. Even for such invasive procedures, however, the balance of burdens and benefits should be weighed and discussed. While some surgeons hesitate to operate on children with severe congenital anomalies due to the higher risk of complications, many centers now offer surgery to such children, particularly if the procedure in question may improve the child's quality of life.

It is wonderful that this family has identified a primary physician for their child, even though the case is a challenging one for Dr. Moy. Bob may live several months, if not more, and having a physician to provide continuity of care is vital. Routine child care, however, like immunizations, should be provided unless the family and physician together assess that Bob is imminently dying. For some children with trisomy 13 syndrome, the most life-threatening conditions, such as apnea, resolve as the child ages. The question of whether to give immunizations can be used as an opportunity to discuss goals and expectations. The family and physician alike will benefit if the health care system appropriately reimburses Dr. Moy for time spent in such discussions.

Because Dr. Moy may not have much experience talking with families about end-of-life decision making, resources in the community should be assessed. There are likely to be hospice agencies available that may be able to offer an additional layer of

support for the family at home. Hospice staff can clarify goals of care with the family so that they do not need to rehash the same conversations again and again, and they can be called to the home at any hour to avoid unnecessary emergency department visits. At the same time, hospice involvement does not preclude returning to the hospital if necessary for treatable illnesses.

### **Pediatric Hospice Care**

There may be a hospice with pediatric experience in the area—if not, Dr. Moy may be able to partner with a local agency to provide the pediatric expertise while the hospice provides the expertise in end-of-life care. Some hospitals also have pediatric palliative care teams that work with the local hospice agencies to provide this expertise. Having a hospice staff member at the bedside to assess Bob’s degree of pain will also add to Dr. Moy’s comfort with increasing the dose of morphine if necessary. If it is being used to treat discomfort, there should be no upper limit on the dose of narcotic used, and it can be titrated to effect.

The fact that the family decided to continue the pregnancy despite a prenatal diagnosis of a severe congenital syndrome may give some insight into their values. Perhaps this decision was made because of their personal views about the morality of abortion, or perhaps it was based on a desire to see their child or give him as good a life as possible despite its likely shortened span. Many parents torture themselves over decisions such as this one after the fact, so, now that it has been made, they need to be supported and told that choosing to carry Bob to term was a very loving decision in the context of their family values. Some centers are beginning to develop programs in “fetal palliative care” as more and more severe congenital anomalies are diagnosed *in utero*. Consultation with a fetal palliative care team can help a family make decisions about termination, support them if they decide to carry a fetus to term, and help ensure that goals of care regarding delivery room interventions or resuscitation are clear and communicated to the team [6].

Although the parents in this case are both physicians, it is important that they be approached as parents rather than as dispassionate medical practitioners. They have a lot more knowledge than the typical family, but may have little experience with end-of-life care. Even if they do, discussions of orders not to attempt resuscitation (DNR) or the dying process are weighty, emotional experiences when it concerns their own child. The team should approach such conversations in the same careful manner used with any family and not assume that the physician-parents have thought everything through. Rather than asking, “Does your son have DNR orders?—which seems to imply that it might be a problem if he doesn’t—the emergency department physician might have asked what conversations the parents had had with their primary physician or with each other regarding how aggressive they wanted medical interventions to be. He could have followed by expressing the desire to assure that their wishes were honored.

The family may need a lot of help regarding the range of possible choices in this case, as well as reassurance that they are making good decisions. With calm, gentle

guidance, the family can survive this difficult process and carry away cherished memories of their son's life rather than traumatic recollections of strained encounters with the health care system.

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