

# When Staff and Parents Disagree: Decision Making for a Baby with Trisomy 13

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## Abstract

Parents often make medical decisions for their children. As pediatricians, we must be sure that the decisions are medically informed. At times, we may disagree with the decisions that parents make and must decide if they are acting in the best interest of their child. This article presents a case in which these issues arose regarding a child with trisomy 13.

**Key Words:** Ethics, child, trisomy 13, medical futility.

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## Case

MICHAEL, A 4-MONTH-OLD BABY with trisomy 13, was admitted to the pediatric intensive care unit (PICU) for respiratory distress. He had been born to a 22-year-old mother and a father in his 20s. The parents were married and this was a planned first pregnancy, with good prenatal care. The pregnancy was uncomplicated and the baby was born at term. At birth he was noted to be dysmorphic and small for his gestational age and was admitted to the neonatal intensive care unit (NICU). He was diagnosed with trisomy 13 at 6 days of age. He had ventricular septal and atrial septal defects, right micro-ophthalmia, right-sided choanal atresia, hydronephrosis, undescended testes and microcephaly. He was in the NICU for 3 months, and due to his fragile state and ongoing medical needs, was then discharged to a chronic care facility.

Trisomy 13, which occurs in approximately one in five thousand live births, presents with many associated congenital anomalies. These anomalies include heart defects, brain defects, breathing problems, seizures, severe mental retardation and deafness. About 50% of patients with

trisomy 13 will die within the first month of life, and only 5–10% will survive for more than one year.

Michael's parents were deeply involved in his care. The father worked and the mother was planning on staying home with the baby. They were Jewish, with strong faith and beliefs based on that faith, and good family support. They also relied a great deal on a rabbi to help them make important decisions.

When Michael was readmitted to the hospital, not long after his initial discharge, it soon became obvious that some of his respiratory problems were related to aspiration of his feeds. He had a barium swallow to confirm the aspiration and was then started on nasogastric (NG) tube feeding. The aspiration appeared to be related to his underlying neurologic impairment, and we thought that Michael would never be able to eat by mouth again. Despite the NG tube feeds, his respiratory status deteriorated to the point where he required intubation. The parents were consulted, and they elected to have Michael intubated despite his poor long-term prognosis. Michael had a difficult time while intubated, and required sedation and paralysis to maintain his oxygen levels. He had frequent life-threatening episodes despite being on a ventilator. Six days after intubation, he was diagnosed with a seizure disorder and was started on medication. It soon became clear that Michael would not be able to come off the ventilator quickly, and discussions about long-term plans were held with the family.

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The medical team, together with the social worker, spoke with Michael's parents at length about Michael's long-term prognosis and the options for further care. During the conversations, the father was the one who asked and answered the questions, while the mother was mostly quiet. They were young, but intelligent, and obviously concerned about their son. They were also medically "savvy" after Michael's long NICU stay and had educated themselves about his condition. We discussed Michael's poor prognosis for neurologic function, as well as the likelihood of a very early death. Michael's parents were quite aware of the life expectancy and neurologic prognosis for children with trisomy 13. We talked about quality of life versus quantity of life. The options presented to the parents included withdrawal of support with palliative care, or a tracheostomy and at least short-term ventilator support. We did not feel that Michael would survive a trial of extubation, and withdrawal of support was not an option for the parents. Obviously, long-term intubation, which would require acute care hospitalization, was also not an option.

Ultimately the parents elected to have a tracheostomy placed for airway protection and ventilator support, and a gastrostomy tube placed for feeding. Michael had a somewhat difficult postoperative course, but eventually was able to be discharged on a ventilator to a different chronic care facility, with the hope that he would eventually be able to go home.

### Discussion

Michael's case was difficult for the medical staff in several ways. We saw a baby with a very poor long-term prognosis, who we felt was suffering significantly. He often appeared uncomfortable, especially with suctioning and other medical interventions. At the same time, he did not appear to get any pleasure from life. He did not smile and

did not seem to recognize his family. His prognosis for neurologic function was extremely poor. The parents were involved and concerned, but not at the bedside for extended periods of time, so they did not see the suffering that we saw throughout the day. For the medical team, the burdens of treatment seemed greater than the perceived benefits.

In addition to those concerns, there was a sense that the father was making the decisions and not including the mother. The ultimate goal for the father was to take Michael home, a plan with which the mother did not seem quite comfortable.

Although Michael's long-term prognosis was poor, we could not say for certain that he would die within the next few months, which made it more difficult to argue for limitation of care. The parents had educated themselves about trisomy 13 and understood the poor prognosis, but at the same time they had read on the Internet about children who had survived with the condition for more than a year. For obvious reasons, they were holding onto that image very closely. While we were able to have frank discussions with them about Michael's future, they remained firm in their hopes for him.

As medical professionals, we see many neurologically impaired, chronically ill children who suffer through recurrent hospitalizations and medical interventions, only to die a premature death. So while treating Michael may not have been absolutely futile, it seemed that way for many on the medical team. As pediatricians, we must constantly balance the prognosis, needs and well-being of the child with the hopes, wishes and needs of the parents.

Ultimately, we agreed to what the family wanted, and were comforted by the fact that they were making an educated choice and a decision that they felt was in their child's best interest. In the end, the parents will carry the weight of their decision with them forever. When the benefits of treating or withholding treatment are uncertain, we should respect the family's informed decision.

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