“Incompatible With Long Life” Does Not Mean “Allow to Suffer”

“She’s our miracle baby. They told us she had less than one percent chance of being born alive but she’s here! She’s proved all the doctors wrong already.” Amber’s parents were both joyous and defensive looking over their tiny baby in the neonatal intensive care unit at me. “She’s alive, why did they call you?” We started our relationship as I often do, by explaining that palliative care is about living well while facing serious illness, that I have not come to bring death to their child.

I sit on the blue plasticized hospital couch and ask Greg and Leah for their story. “The ultrasound wasn’t right, they ordered more tests, waiting was awful, when they called I was home alone with our toddler, I had never heard of trisomy 18.” Proud and angry moments come from two mouths; quick, out of order, and overlapping memories. “You could tell our doctor felt really bad about it, she looked sad but told us we should terminate the pregnancy, we would never do that!” Tense shoulders start to relax and their fear of my presence seems to slide off of them. “We were only hoping to hold her before she was gone, but everyone was wrong about her. Now we want to focus on helping her eat and making sure she doesn’t stop breathing.”

Amber is tucked under a hat and blankets that take up far more of the crib than she does. A tiny tube coming from her nose is taped to her pink cheek and connected to a syringe pump full of milk. Her bedside nurse steps in to stop the pump as Amber stirs. “She can’t take much before arching and crying, eventually she’ll start coughing. I want to give her more because she hasn’t been growing well enough.”

Dictations stop when I enter the neonatal team room. “Thanks for coming. How sad for this to happen to such a lovely family. When can your hospice team get her home?” I watched the neonatologist’s face take on a confused expression when I asked about a trial of medication for Amber’s acid reflux and about the parents’ concerns about her trouble feeding. “It seems like we’re talking about going the wrong direction. I don’t know that we should be adding more treatments when trisomy 18 is incompatible with life. It’s probably best that her family take her home and spend as much time with her as they can before she dies.”

Trisomy 18 has been described in the past as “incompatible with life” as almost all babies with trisomy 18 died in early infancy. Many physicians are unaware that with increases in medical interventions, particularly the use of medical nutrition, life expectancy has increased such that around one in ten survive to turn five years old. All have difficulty growing and significant brain dysfunction such that they are fully dependent on others for their cares. In addition some are born with severely malformed hearts, suffer seizures, stop breathing multiple times per day or can’t breathe without machines while others have multiple but less severe issues that allow them to survive for years. Amber has a small hole in her heart that is expected to close over time and an abnormally formed left kidney. She has stopped breathing a few times which has required her nurse to rouse her and give her oxygen. The episodes could be life threatening without intervention and she is at high risk of a serious lung infection from her feeding difficulties. On the other hand, she isn’t suffering and her parents are determined to get her home to be part of their family for as long as she is here.

I do hope that my own confusion isn’t as apparent on my face in the team room. This particular neonatologist is one I have worked closely with in the past and know well. I have seen her calmly rescue tiny premature babies in crisis and agonize over those who don’t recover as expected. Only a month ago I challenged her recommendation to place a child on a long-term breathing machine who had severe bleeding in his brain leaving him unable to wake up. While both of us would support Greg and
Leah if they chose hospice and avoiding life prolonging measures for Amber, it is entirely unexpected that she would view treating painful acid reflux or considering a different type of feeding tube as providing excessively aggressive care. As gently as possible, I ask her about it. She replied “I don’t think it will change the outcome for her.”

As a pediatric palliative care physician, I meet many children with trisomy 18 and their families. The experience for those who are less severely affected and survive longer is similar to that of Leah and Greg. Most parents feel that they are discouraged from pursuing interventions to treat symptoms and to prolong life, even while acknowledging that life may still be short. This common experience gives rise to an intrepidly bonded community of parents who in turn reach out to connect with, and prime, new parents to search for bias in every interaction with the medical community, both finding true bias and projecting bias where it isn’t present. During one of my later visits, Leah described the great support she found in an online parent group of trisomy 18 children. She was relieved to have found other families who were advocating for medical interventions to prolong their children’s lives and treat symptoms. Leah understood from the parents in this group that many kids with trisomy 18 had bone issues, and since she was hoping to “get as many appointments done as possible while in the hospital,” she requested an orthopedic consultation. It felt as if the room were elongating as I saw a great distance developing between Leah and Amber’s doctor. I could see his opinion of Leah changing, not only was she not wanting to go home with hospice care but now was asking for unwarranted consultations. Parents described that “every person that comes to see us doesn’t think we understand trisomy 18, just because we want to know what other things are available to help our daughter.” As a palliative care physician, the complexity of trisomy 18 has placed me in the unusual position of advocating for more medical interventions instead of the typical position of limiting therapies that prolong life but are no longer helpful.

In effort to help decrease the distressful interactions between the clinical team and Leah and Greg, we wrote a document together that specified what Amber’s conditions are, parents’ understanding of the prognosis and their goals. We outlined the goal of giving her more quality time with her family by pursuing medical treatments that will help relieve distressing symptoms and treat reversible conditions. It also addressed the plan to decrease interventions when her quality of life declines. This “passport to care” allows Leah and Greg an enhanced way to communicate with all providers and avoid repeated “educational sessions.”

The duty to care does not stop with diagnosing a life-limiting condition. In fact, I could argue that it is during these desperate moments our duty to care is heightened. We owe our patients and their families the best we can offer regardless of the prognosis. Addressing pain and suffering, symptom management, and burden of disease is an ancient canon of medicine that did not come with the stimulation “only for those who are curable.” Trisomy 18 and our understanding of this genetic anomaly has evolved. So must our ability to care for these patients. In the end, they may still die young, but at least the life they live with their families is the best we could help them have.
REFERENCES:


