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The Importance of Advance Care Planning for Children with Chronic Respiratory Failure

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Medical advances are improving survival for pediatric patients with chronic, severe lung diseases and medical complexity. (1, 2) Longer survival both warrants celebration, and presents new challenges.

Consider the following cases:

Case 1: A child with myotubular myopathy and hypoxic encephalopathy requires around-the-clock ventilator support. His family lives 300 miles from a tertiary care center in a region without medical services that would permit him to be cared for at home. He spends his first 2 years of life in a group home for medically fragile children. At 2, he is discharged home when support services become available. However, insurance coverage for these services is poor, the 300-mile commute to access specialist care is taxing, and one parent stops working to provide the continuous care he requires which puts further financial strain on the family. At age 4, the family moves >1,500 miles to a region where they hope nursing, durable

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medical equipment services, and medical subspecialists to be more affordable and reliably available.

Case 2: An infant delivered at 37 weeks gestation following induction of labor for intrauterine growth retardation is transferred to the neonatal intensive care unit shortly after birth due to seizures and severe episodes of cyanosis related to central apnea. She is intubated and mechanically ventilated, and evaluation reveals pontocerebellar hypoplasia. After discussing their child's prognosis and care options with their NICU team, pediatric neurology, the palliative care team, social work, and a hospital chaplain, the family chooses to forgo respiratory support and take their daughter home with the help of a community hospice service.

These cases reflect the incredible challenges faced by a growing population of parents of children with medical complexity. Their children are heterogeneous in diagnosis but they have a few things in common. First, their families and medical providers face emotionally difficult decisions about how best to care for them throughout their lives. Second, they often depend on advanced technologies for survival. Finally, their prognosis and expected disease course is impacted by decisions made by their families and providers over time. For clinicians, challenging aspects of caring for children with severe, chronic lung disease include making highly consequential recommendations (e.g. whether to initiate home mechanical ventilation), and advance care planning.

The literature suggests that neonatal and pediatric ICU doctors struggle when asked to participate in shared decision-making for children with chronic severe illness (3). There is disagreement about how much influence an individual physician should exert. Some NICU and PICU attendings report making recommendations without consulting other team members, others obtain team consensus before making recommendations, and some provide families with information but do not make recommendations. When families prefer to rely on the professional expertise of physicians, declining to make recommendations may be viewed as an abdication of responsibility (3-4).

The first major decision faced by families of children with severe chronic respiratory disease is often whether to initiate home mechanical ventilation. This decision is complex and emotionally challenging. Some parents in this position report feeling that there was no true choice, as initiating long-term ventilation was the only way to maintain their child's life (5). Families making this decision need comprehensive information about what day-to-day life is expected to be like for both the parents and the child. Social context, defined as the physical, geographic, economic and cultural environment in which a child and family lives, should be integrated into these conversations. Caregivers want to know the risks and burdens of long-term ventilation for their child (5). Families who choose home mechanical ventilation need to acquire skills and social support to care for their child, and be able to devote significant time to their care. Regional differences in available resources (home nursing, medical transportation, etc.) lead to variability in families' ability to care for their children at home (6). The financial burden of supporting a child on home mechanical ventilation should also be disclosed. Families of children with home mechanical ventilation report significant out-of-pocket costs, and it is common for one or more family members to take unpaid days,

reduce paid work hours, or stop working entirely (7). For the decision to initiate home mechanical ventilation to truly be a choice, families must also know they have the option to decline home mechanical ventilation and be educated regarding what this path entails for their child (8).

While the decision to initiate home mechanical ventilation is particularly emotionally challenging, it is far from the only decision these parents will face. Advance care planning includes making decisions regarding accepting or declining available life-sustaining therapies when diagnosed with life-limiting illness, sharing values with loved ones, completing advance directive paperwork and choosing a health care agent in the event that an individual was incapacitated (9). These discussions can be extremely valuable in the setting of complex and chronic disease, however, advance care planning discussions for children with medical complexity and chronic respiratory disease often first occurs in times of crisis. Too often, we forego questions about the rituals, people, and environment that a family values at the time of death until an acute decline, or cardiorespiratory arrest. This suggests we are missing opportunities to initiate these discussions during times of relative stability (10).

Opportunities for advance care planning exist at any point along the disease trajectory. It is worthwhile to explore with families what brings their child joy or makes their life meaningful, and what circumstances might change their perception. Are there levels of medical technology that would be unacceptable? Is there a point at which the need for multiple procedures or hospital admissions would prompt them to reconsider the care trajectory? What changes or sacrifices are unacceptable or infeasible for their family? While these types of questions may be challenging for families to answer in advance, the process of exploring them over time allows a provider to gain a deeper understanding of a family's value system and will assist in future counseling.

Advance care planning early in a child's disease course can improve parent-reported outcomes. For example, parents who participated in early advance care planning were able to plan the location of death, and perceived their children to have better quality of life during their final months (11). Parents in one study unanimously supported discussing Medical Orders for Life-Sustaining Treatment (MOLST), and believed routine conversations about MOLST would decrease stigma, clarify decision-making, and increase utilization of palliative care services. In contrast, only 17% of physicians and 33% of nurse practitioners felt these discussions were warranted (12).

Physicians looking for assistance with both shared decision-making and advance care planning for children with medical complexity will benefit from involving palliative care professionals. Pediatric palliative care works to provide relief from the symptoms and stress of serious illness and focuses on improving quality of life for both patients and their families (13). They also help elucidate what outcomes and principles a family values and clarify how various choices relate to those values. This is done most effectively over time to allow the palliative care team to build relationships, understand families' values, needs, ways of processing information, and stressors. Initiating relationships with palliative care professionals during times of stability lays a foundation to assist with exigent decision-

making when required. Nascent palliative care research and resources for pediatric patients are expanding. These services are increasingly available in hospitals, but are under-utilized in the care of children with medical complexity and severe pulmonary disease who require highly-coordinated, on-going care to ensure a high quality of life (14). Palliative care can be integrated into the Pediatric Intensive Care Unit, where many patients with severe pulmonary disease and/or home mechanical ventilation receive care (15). Criteria have been developed to prompt automatic palliative care consultation in adult ICU patients, and similar criteria could be developed for pediatric patients (16).

Resources are also available for clinicians at institutions without pediatric palliative care specialists. The National Institute of Nursing Research's Palliative Care: Conversations Matter® campaign provides free brochures, videos, and fact sheets specifically designed for pediatric patients to assist providers with difficult conversations (17). Seattle Children's Hospital designed and tested a decision-making tool to help families faced with challenging decisions, finding that this tool improved some aspects of quality of life and family satisfaction (18). There are also decision-support websites for families considering home ventilation (<http://family-reflections.com>) informed by interviews with families and decision-support brochures in English and Spanish for families considering long-term ventilation for their child (20). Overall however, we see a need for more widely available pediatric palliative care as well as earlier use of palliative care teams in centers where they do exist. As our ability to care for children with chronic, severe lung disease continues to grow, our skill and comfort with palliative medicine for these patients will need to keep pace.

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