FEATURE

Medically Complex Children and Early Intervention for Comprehensive Medical Care at Home

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Disclaimer: Identifying details have been modified to protect the confidentiality of the family.

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Abstract

We present the case of a 4-year-old with multiple congenital anomalies. These included complete tracheal rings with agenesis of the right lung and missing right ribs. He was hospitalized from birth until he was age 3 years and 6 months. At age 4 years and 4 months he had yet to move to a family home or experience his first day of school because he was in a transitional care facility. The intensity of his care was initially high, slowly declined over time, and eventually entered the range where he would be potentially manageable in a home envi-

ronment. Despite the countless medical teams treating him, he did not receive a referral either to the early intervention or educational services that he had a right to under the Individuals with Disabilities Education Act. While his case is extreme, there is a disconnect between the United States health care and educational systems, which negatively impacts both the health and development of children with chronic conditions who require frequent and prolonged hospitalizations. **[*Pediatr Ann.* 2015;44(1):36-39.]**

Medical care for young children has improved dramatically over the past 25 years. These improvements have led to an increasing number of children discharged from the hospital setting who previously would not have survived. At the same time, society has improved in its systems of support of children with specialized needs in the community. However, these two developments have not been formally linked, and because of the lack of connection, opportunities are currently being missed. We present one dramatic example of a scenario that highlights a significant trend in current care models: intensive inpatient care does not guarantee an equal amount of developmental and family support.

In 2009, an infant male was born after a full-term pregnancy by uncomplicated, spontaneous vaginal delivery to a mother who had no prenatal care. His birth weight was 3.3 kg and length was 49.5 cm. It was immediately recognized that the baby had multiple congenital anomalies and required immediate intubation for respiratory distress. He was soon diagnosed with 12-15 complete tracheal rings and agenesis of the right lung. Seventeen days after birth, he received a slide tracheoplasty to repair his complete tracheal rings.

During his surgical recovery, he had multiple respiratory arrests that led to frequent bronchoscopies. He was medically paralyzed and sedated for 36 days while being orally intubated. He experienced oral aversion and oral, pharyngeal, and esophageal dysphagia. He was started on chronic medications for gastroesophageal reflux disease. He had bilateral ear anomalies, and auditory brainstem response results demonstrated a severe to profound hearing loss in both ears within the 500 Hz-4,000 Hz frequency range. At age 12 months, he underwent a laryngoscopy and bronchoscopy followed by the insertion of a tracheostomy. A gastrojejunostomy was also performed at that time due to his swallowing difficulties and his oral aversion. At age 21 months, he developed seizures, leading to status epilepticus and he required medical paralysis and multiple medications to stop his seizures. After this episode, he developed gastrointestinal (GI) dysmotility, which required slow-continuous feeding through the gastrojejunostomy tube. He was weaned off mechanical ventilatory support by age 3 years and 5 months.

Throughout all these steps of his hospitalization, he was cared for in isolation because he was a carrier for pseudomonas multiresistant organisms. At age 3 years and 6 months, he was finally discharged from inpatient hospitalization and transferred to a transition care unit. In this homelike setting, he became more mobile and started walking. However, his increased mobility led to complications with his gastrojejunostomy tube for which he had multiple operations to repair the dislocation of the equipment (when walking, he would step on it and accidentally pull it out). At age 4 years and 3 months his gastrostomy and jejunostomy tubes were separated. This led to further improvement in both his GI motility and overall mobility. Due to family stress, his parents were unable to take care of him, and he became a ward of the state with ultimate placement in a skilled nursing facility.

CURRENT CONDITION

He has become playful and interactive, and his physical and developmental growth are good. He was not exposed to sign language training and cannot formally communicate in that way; however, he communicates informally with very robust gesturing. He still cannot receive any food by mouth. He requires close monitoring at all times due to potential dislocation of the multiple tube sites—tracheostomy, gastrostomy, and jejunostomy.

During his stay at the transition care center he was treated with multiple daily medications including inhaled steroids, erythromycin, sildenafil, omeprazole, multivitamins, albuterol, triamcinolone (topical), hydrocortisone (topical), and protective barrier ointment. His durable medical equipment included three home ventilators (a "main" one, a "transport" one, and an "emergency back-up" one), a specialized wheelchair, tubing for gastrostomy and jejunostomy stoma, tubing and suction equipment for tracheostomy, bilateral supramalleolar orthoses, wrist splints, hearing aids, and eyeglasses.

DISCUSSION

It is clear that this child is medically complex due to his multiple congenital anomalies. Agenesis of the lung is a very rare occurrence: only about 1 in 100,000 births.[1] Lung agenesis in conjunction with congenital tracheal stenosis is even rarer. Untreated, this unique combination of lesions has a 33%-65% mortality rate.[1] This patient received the best possible care in a tertiary care medical center with surgeons, intensivists, and the general health team being particular experts in his condition.

Modern children's hospitals, especially surgical and intensivist teams, are

better now than ever in saving the lives of critically ill children like this patient. However, because of these successes, a new problem has emerged: developing systems of educational, developmental, and family-support systems that are integrated into our medical systems. Children who are quite ill are increasingly going back into their communities, but our health care systems are not yet prepared to help these children access their mandated support services and educational/developmental therapies. We now know that any critically ill child's long-term outcomes will be impacted by the developmental and educational supports that are provided to assist in managing his or her health impairments.

The child in this case, along with many others in the United States, did not receive timely and appropriate developmental and educational referrals, which resulted in a breakdown of the mandated supports and therapies. He was required to live in the hospital not because of his developmental delays, but because his respiratory and nutrition status required specialized medical interventions. He was treated by hundreds of doctors, had dozens of operations and procedures, and spent more than the first 3 years of his life at an inpatient tertiary care medical center in the intensive care unit. However, he started school 482 days (1 year and 4 months) after he was eligible, because the process for special education referral was never initiated.[2] No one would disagree with the idea that his developmental outcomes would be maximized by transitioning to preschool and living in a permanent home with stable parents.[3,4] However, due to inherent disconnections between health and educational systems, when at age 3 years he was eligible for full educational evaluation, he was not referred. To be clear, there was no specific person or group who is clearly at fault; his care was truly exceptional. The high level of care and initial "investment" of time, money, and compassion are only "worth it" if we make our downstream efforts as equally exceptional.

Additionally, he never received early intervention services that could have begun shortly after birth. Early intervention services include physical, occupational, and speech/language therapy (he did receive some physical and speech therapy sporadically, but focused exclusively on feeding and positioning). Parent training, social work services, and formal transition into the educational systems should have also been a part of the early intervention strategy.[5] Now that he is older than age 3 years, the Department of Education must provide an Individualized Educational Program (IEP) specific to his special needs. This IEP should provide support services like speech and audiology, psychological, physical and occupational therapy, social work, transportation, preschool, counseling, and medical for diagnostic and evaluation purposes.[5] Starting this process earlier would reduce the amount of catching up he would need to do later on in life. He has not developed to his full potential because there is currently no formal link between the health care and educational systems for children with his degree of medical necessity requiring prolonged hospitalization.

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Taking early initiative with respect to preschool services, access to distance learning materials, and facilitation of his IEP would further improve his already impressive developmental progress. Consideration of what technology he would require for his hearing and communicative impairments would be an appropriate first step. This case study suggests that because surgeons and intensivists are becoming increasingly effective at helping critically ill children survive very difficult illnesses, there is an increasing need for early intervention programs to initiate developmental and family support services while the child is in the hospital.

However, this is a component of medicine that is often missed because most health professionals lack knowledge, skills, and attitudes related to education programs. Further, as many professional organizations update the training requirements for pediatricians and subspecialists in pediatrics, this important area of knowledge is being overlooked as a training requirement for residents and fellows.

There are increasing numbers of young children who require prolonged hospitalization and long-term medical supports. Currently, children with complex chronic conditions account for approximately one-third of total US health care costs for children, and 80% of inpatient costs, reflecting a doubling of hospital days used by these children.[6] From 1999 to 2012, the cost of inpatient care increased about $13.7 million.[6] These children utilize these medical supports due to complex life-threatening disorders after prematurity, due to known genetic syndromes, and due to significant neurological, cardiac, or pulmonary disorders. As surgical techniques, intensive care units, medications, and other parts of modern medical systems continue to improve, the odds of going home are increasingly favorable. Thus, early intervention should have a role from the earliest points of hospitalization. It is important for health care professionals to partner with the child's family to work on developmental activities that are part of the child's day, no matter the setting, including general inpatient units and long-term intensive care units.

Investigating the longitudinal trends of children with complex chronic conditions after discharge from the hospital is important in educating health care professionals on how to improve current comprehensive care strategies. Studies looking at special outpatient services (SOSs) and highest categories of expenditures have expounded the need for a more proactive and timely response with the coordination of resources for children with complex medical conditions. In an environment of state budget constraints, many policymakers lump all health care expenditures, which means as inpatient care becomes increasingly expensive, children with chronic conditions will inevitably face problems in long-term health care financing. Further, looking at a population consisting of extremely preterm infants who were born at <1,000 g and <28 weeks gestation, there is evidence of significant disproportionate resource use that is not just limited to the inpatient setting but continues well after hospitalization. Medical specialties, neurodevelopmental services, and occupational and physical therapy were widely used; speech and language and social worker services were used the least.[7] Moreover, approximately 37% reported the need for SOSs that were not being received.[7] Thus, less than one-half of the highest-risk infants and toddlers who stayed in the hospital for more than 3 months on average were receiving sufficient SOSs.

CONCLUSION

It seems prudent that if a health care system invests hundreds of thousands of dollars in the care of one child, it would be important to also invest the money needed to see that the initial investment pays off in the long term. Early child developmental stimulation has been proven to be a good investment.[8] Children's hospitals (including increasing their inpatient and intensive care units) need to develop systems to ensure that these programs are engaged to their full potential.

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