CHALLENGING CASE: CHRONIC DISEASE—DEVELOPMENTAL AND BEHAVIORAL IMPLICATIONS

“What Can I Do to Enhance the Development of a Premature Infant with Chronic Lung Disease?”*

CASE

Timmy was born at 32 weeks of gestation after an uncomplicated pregnancy until there was a spontaneous rupture of the membranes and preterm labor associated with chorioamnionitis. A 2-month hospitalization in the neonatal intensive care unit (NICU) was associated with pneumonia, a Grade II intraventricular hemorrhage, chronic lung disease, and a slow weight gain in the nursery. He was discharged to home with plans for ongoing care by his pediatrician. The primary care pediatrician attended a multidisciplinary conference with the NICU staff and Timmy’s parents. At the time of discharge from the nursery, at 38 weeks postconceptual age, Timmy still required oral diuretics and supplemental oxygen, as well as other medications such as iron. Timmy’s respiratory rates were between 40 and 60 breaths per minute at rest, with mild intercostal retractions. He was discharged with a cardiorespiratory monitor.

The discharge examination revealed mild to moderate asymmetrical hypotonia with intact deep tendon reflexes, shoulder girdle weakness, and a mild head lag. Timmy would regard a human face and a bright object and would follow them briefly. He became active and would thrash his extremities with minimal tactile, bright light, or auditory stimulation. Typically, he settled slowly with swaddling and a pacifier. Nursing was slow to develop; he was currently receiving one half of his calories at the breast and the remainder of his calories from bottle-feeding of fortified expressed breast milk.

As she prepared for the first office visit with Timmy and his parents, the pediatrician asked herself, “What can I do to enhance the developmental outcome for this child?”

Immature pulmonary function, infections, central nervous system insults such as hypoxia and hemorrhage, and growth demands for energy that can tax their developmental immaturity. In addition, the auditory, visual, and tactile environment of an intensive care unit impacts their ability to thrive.

Most of these very low birth weight premature babies are cared for by neonatologists and other professional colleagues in the neonatal intensive care unit (NICU). When the infant is ready for discharge to home, the responsibility for care then typically shifts to a primary care clinician. For many clinicians, graduates of the NICU are among their most challenging patients with chronic conditions. Coordinating care among multiple specialists while helping families to integrate different professional recommendations requires a focused set of goals that can be communicated effectively.

Commentaries on this challenging case are written by two pediatricians and a parent who has experienced the birth of two premature infants. Dr. Peter Gorski is a developmental-behavioral pediatrician who has extensive clinical experience working with premature infants and caregivers in the NICU. His work has focused on the interactions between the environment and physiological-behavioral changes in premature infants. Dr. Yvonne Vaucher is a neonatologist who has cared for preterm babies for more than two decades. She has a special interest in monitoring developmental outcomes of premature infants who undergo specific interventions as well as those who develop chronic lung disease. Faith Kettel is a mother of very low birth weight premature twins who were discharged from a NICU a decade ago with chronic lung disease.

Dr. Martin T. Stein

As premature infants survive at smaller birth weights and at earlier gestational ages, biological and developmental principles of pediatric practice have been redirected to the unique needs of these young children and their families. Survival associated with acceptable developmental outcomes is expected for the majority of very low birth weight infants. The case of Timmy is not an uncommon course for these infants as they are challenged by immature pulmonary function, infections, central nervous system insults such as hypoxia and hemorrhage, and growth demands for energy that can tax their developmental immaturity. In addition, the auditory, visual, and tactile environment of an intensive care unit impacts their ability to thrive.

Pediatricians can promote optimal developmental outcome by attending to two primary determinants of recovery from high-risk birth: the child’s nervous system and the child’s family system. The two weigh equally on the course of adaptation and growth. They link functionally as each supports and challenges the other’s regulatory capacity. A few, although not nearly enough, clinical insights are revealed to us in the details of the presentation. Timmy was born at a gestational age coincident with the

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early, still fragile organization of autonomic, sensory, and motoric coordination of respiration, arousal, and activity states.\textsuperscript{1} He developed seemingly moderately severe acute and chronic lung disease necessitating initial antibiotic treatment as well as ongoing use of diuretics and oxygen. A relatively mild ventricular bleed may have resulted from a period (pre-, peri-, or postnatal?) of hypoxia and cerebral ischemia.

Despite these problems, a number of documented strengths support the infant’s prospects for health and development. Timmy gained sufficient weight and physiological stability to merit discharge to home from the NICU at approximately the same time that he would have been expected to arrive home had his birth occurred at full term. His neurodevelopmental assessment at 38 weeks postconceptual age revealed findings consistent with prematurity and nonspecific weakness resulting from illness, effort, and incompletely compensated energy expenditure. Timothy’s mother continues to work at breastfeeding, both parents plan to attend the first office visit, and the pediatrician has been involved in the care of the infant and family during the newborn hospitalization. From her early involvement, the pediatrician has gained a key database about the condition, experience, and coping mechanisms of infant and family while earning the trust that caring inspires.\textsuperscript{2}

The primary care pediatrician can now begin the delicate work required to validate the parents’ observations and interpretations of Timmy’s developmental progress; to gradually build the parents’ sense of security, confidence, and autonomy from the critical care medical system; and to offer early medical, developmental, and/or psychological interventions, when useful, to support the child’s development and the family’s adaptation. Prematurely born infants, especially those with chronic respiratory distress and early neurological insult, often exhibit subtle neurodevelopmental vulnerabilities throughout the first 3 to 6 months after discharge from the NICU. Lower sensory thresholds to sound, light, sudden postural shift, or prolonged handling and eye contact can sometimes cause routine care-giving experiences to exceed the infant’s autonomic controls. The pediatrician should listen to the parents’ stories and make use of her own behavioral assessment to consider strategies for protecting the boundaries of the child’s immature nervous system.\textsuperscript{3} At the same time, the primary care provider should take every opportunity to communicate indicators of developmental progress, including increasing vigor, alertness, balanced muscle strength, quality of movement, and behavioral self-regulation during calming, sleeping, and signaling.

Furthermore, longitudinal follow-up studies unanimously find that supporting parents in their efforts to cope, understand, and interact with their infant contributes a major advantage to the child’s developmental progress.\textsuperscript{4–6} Emotional attachment enhances parental sensitivity, fuels infants’ effort to communicate and to succeed, and inspires the adaptive organization of corresponding neural circuitry.\textsuperscript{7}

REFERENCES


Dr. Yvonne Vaucher

Chronic lung disease (CLD) imparts developmental, behavioral, and educational risk as well as medical risk to both term and preterm infants.\textsuperscript{1–4} The pediatrician plays a central part in promoting an optimal outcome in all of these areas.

Premature infants with CLD are often discharged to home care with complex medical regimes that may include special feeds, nutritional, and electrolyte supplements; fluid restriction; respiratory stimulants; bronchodilators; inflammatory suppressants; oxygen; and monitors.\textsuperscript{5} It is no wonder that parents often feel completely overwhelmed by their infant’s impending discharge and the responsibilities they will abruptly assume. The weeks just before and after discharge are difficult emotionally as well as physically for parents of infants with CLD. Within the first month, parents become more comfortable with medications and equipment and more confident in their ability to accurately assess their infant’s physical status. As fewer trips to medical specialists are needed, parents are encouraged by seeing their child’s health gradually improve under their care. Gradually they realize that they have indeed become their child’s most expert and knowledgeable caretaker and advocate, a role which will be important throughout childhood to ensure that their child receives appropriate evaluation and intervention. The pediatrician can help the family cope with the hospital to home transition by (1) arranging a predischarge conference with the parents; (2) being familiar with the infant’s physical examination and medical needs before discharge; (3) maintaining close communication with the family after discharge; and (4) helping to coordinate the required primary care and subspecialty appointments.\textsuperscript{6} Acknowledging the parents’ emotional and physical exhaustion, reassuring parents that they are capable caretakers, and encouraging parents
to become vocal advocates for their infant will reduce parental anxiety and provide the psychological support needed for parents to develop confidence in their own abilities to care for their child. They will then be free to turn their attention to developmental as well as medical issues.

Optimal physical growth and well-being are integral to sustaining developmental progress in early childhood. CLD is ultimately outgrown as healthy lung tissue becomes a progressively greater proportion of total lung volume. Sufficient caloric and nutrient intake, adequate tissue oxygenation, avoidance of excessive energy expenditure, and protection from further lung injury are all essential to promoting normal lung growth. Achieving adequate food intake may require caloric and electrolyte supplementation of breast milk or formula feeds. Fluid intake may be restricted to reduce excess lung water, which impairs pulmonary function. Infants with CLD may have oromotor uncoordination, hypoxia during feeds, and/or nipple aversion. Parents often need help coping with these feeding problems, especially when their infant is difficult to feed or is dissatisfied with a limited feeding volume. Occupational therapy intervention is helpful for oromotor uncoordination and nipple aversion. The pediatrician can support the family in their concerns, help them understand the need to maintain adequate growth and the rationale for nutritional supplements and/or fluid restriction, guide them in adjusting the feeds as their infant’s fluid tolerance and lung function improve, and request interventions (e.g., occupational therapy) if indicated.

When the infant’s growth is poor, hypoxemia may be the culprit. Feeding, sleeping, exertion, and crying all reduce oxygen saturation. The pediatrician must be certain that parents recognize clinical changes and behavioral cues associated with hypoxemia and increase oxygen delivery accordingly. Parents often focus on reducing the amount of oxygen given because reduced oxygen indicates that the lung problems are improving. They may be unwilling or unable to recognize chronic or intermittent hypoxemia, which adversely affects growth, delays recovery and limits their infant’s ability to attend to developmental tasks. The pediatrician can help the family understand that adequate oxygenation is key to their infant’s physical recovery as well as to their developmental progress. Oxygen saturation should be monitored periodically to be certain that tissue oxygenation is adequate. Families should also be counseled that changes in behavior such as restlessness, poor feeding, lethargy, or irritability may be the earliest signs of deteriorating pulmonary status. Prompt recognition of respiratory problems by the parents will enable timely intervention, thereby avoiding severe or prolonged exacerbation of lung disease, which delays development.

Episodes of infection delay both physical recovery and developmental progress. Pertussis and respiratory syncitial virus (RSV) can be especially devastating complications. Routine immunizations should not be delayed; monthly immunoprophylaxis is recommended during the RSV season. Parents can protect their infant from respiratory disease by (1) avoiding large groups of people (such as church gatherings, supermarkets, etc.) for several months after discharge; (2) screening all household visitors for illness; (3) limiting close contact with young children, if feasible; (4) maintaining good hand-washing practices for anyone in physical contact with the infant; and (5) avoiding exposure to second-hand cigarette smoke. Parents need to know that despite reasonable precautions, their infant will inevitably be exposed to viral diseases and is likely to develop at least one respiratory illness during the first year after discharge. The pediatrician should reassure the parents that, despite their best intentions and precautions, exacerbation of symptoms and rehospitalization associated with lower respiratory tract infection is an inherent risk of CLD.

On one hand, it is necessary to protect the child with CLD against adverse influences on growth, development, and energy resources. On the other hand, the infant must experience appropriate developmental stimuli and challenges to progress normally. The pediatrician is key in helping families maintain this delicate balance. Infants with CLD often have substantial gross motor delay until they have attained sufficient muscle mass and energy reserve to engage in gross motor activities. They may resist prone positioning because of shoulder girdle weakness and poor head control. A pediatric occupational therapist, experienced in working with chronically ill infants, can recommend supportive techniques and activities needed to foster age-appropriate developmental skills. It is imperative that each infant’s physical limitations be recognized by anyone working with or planning interventions for a child with CLD, including parents, teachers, and physical/occupational therapists. Excessive energy expenditure associated with vigorous gross motor activity or prolonged crying may precipitate hypoxemia and acute deterioration in respiratory status. Improvement in gross motor skills is often heralded by more rapid weight gain and weaning from oxygen. At this point, there is a rapid catch up in major motor milestones. Despite gross motor delay, infants with CLD are more likely to reach adjusted age-appropriate developmental milestones in areas that require minimal energy expenditure (e.g., adaptive, fine motor, language, and personal/social skills). The pediatrician can help parents focus on those areas in which their infant is most able to meet developmental expectations.

The child with CLD is at risk of being perceived, long after the lung disease is resolved, as a vulnerable child with attendant deleterious effects on independence and developmental progress. Just as parents need help in understanding why their premature infant with CLD must be protected from various adverse influences on lung function and growth initially, they may also need assistance in avoiding overprotection once their child has recovered. Periodic, formal developmental assessments throughout infancy and childhood will help the pediatrician determine how the child is progressing and what resources are currently needed.
To fulfill their potential, infants with CLD need a consistent, readily accessible, primary care provider to address these issues and coordinate care throughout childhood. The pediatrician can enhance developmental outcomes in a child with CLD by (1) promoting optimal physical growth, which will maximize brain growth and allow the child to recover from CLD as soon as possible; (2) ensuring that the infant is adequately oxygenated, because hypoxemia can be deleterious to both lung growth and exercise tolerance; (3) providing immunization and RSV immunoprophylaxis to reduce the risk of additional lung injury; (4) helping the parents offer appropriate developmental stimuli and challenges; and (5) focusing the family on the child’s capabilities and accomplishments, thereby fostering a positive parent-child interaction that will continue to promote optimal child development long after CLD has resolved.9

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REFERENCES

Faith R. Kettel

Parents of children like Timmy were expecting to have an uncomplicated delivery and a beautiful new baby. Instead, their dreams are shattered by prematurity, infection, and a sick, premature infant. This happened to me. I am the proud mother of twin boys, born at 29 weeks, who are now healthy, handsome, and 11 years old.

Being the parents of a premature infant with CLD is difficult. The parents wish that ear infections and tubes were the only things to worry about. Instead, they monitor oxygen percentages, respiratory rates, lack of weight gain, and food intake. They are terrified of infection occurring and the possibility of rehospitalization and even death. They wonder what the future holds for their child, both physically and mentally.

I have much to share from my experience. First, parents must be parents. They should be encouraged to love, hold, and cherish their new baby. To grow and mature, their baby needs special tender loving care. Second, parents should keep in close contact with their pediatrician. They should be given parameters to follow and emergency guidelines. Third, parents should be proactive and advocate for any services they believe their child needs. Parents should ask questions regarding development and what they can expect with their child, keeping the child’s adjusted age in perspective.

My children were involved in an early infant development program. It was designed to monitor early development in areas of fine and gross motor coordination; cognitive, receptive, and expressive language; and other social skills. Sometimes these areas can be overlooked by parents and professionals because of the fragile health of the child. Observing developmental change and discussing these observations helped me to understand and respond to behaviors that may not have been obvious at the time.

The question of future development and neurological damage is always on the mind of parents of children like Timmy. My children were referred for a comprehensive developmental assessment at age 3 years 6 months. The evaluation was very helpful in determining proper educational opportunities for my children.

Timmy’s parents should be encouraged to educate themselves so that they can be better prepared to understand his behaviors during feeding, sleeping, and other times. Sometimes they should be encouraged to talk with other parents who have experienced similar situations.

All in all, being a parent of a premature infant with CLD is stressful, but rewarding. I am thankful for all the medical, physical, and emotional support my children were given.

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Web Site Discussion

A participant in the cyberspace discussion of the case on the Developmental and Behavioral web site9 provided thoughtful comments that complement those of Drs. Gorski and Vaucher and Ms. Kettel:

Dr. Jean Struthers of Sacramento, California, commented: “I would encourage the family to put the child on his tummy as much as he can tolerate so he can begin to build up those weak shoulder and trunk muscles early. There are multiple ways to do this

9 A bimonthly discussion of an upcoming challenging case takes place at the Developmental and Behavioral Pediatrics web site. This web site is sponsored by the Maternal and Child Health Bureau and the American Academy of Pediatrics section on Developmental and Behavioral Pediatrics. Henry L. Shapiro, M.D., is the editor of the web site. Martin Stein, M.D., the Challenging Case editor, incorporates comments from the Web discussion into the published Challenging Case. To become part of the discussion at the Developmental and Behavioral Pediatrics home page, go to http://www.dbpeds.org.
with fragile premies, including having them lie on the parent as the parent leans back into a supine position (with the child prone) and placing the child on a blanket with something he enjoys in front of him. They should also avoid all types of jumpers, walkers, and exersaucers which can encourage extensor patterns and do nothing to build up his trunk. Reaching is helpful in developing the shoulder muscles and can be encouraged by having his shoulders “rolled” forward (e.g., by placing soft blankets or diapers behind them while he is in an infant chair or car seat) with interesting items to grab in front of him. Paying attention to these types of problems when young will go far in helping to avoid problems later.”

Dr. Martin T. Stein

A premature infant such as Timmy, in the process of transition from hospital to home care, is a moment of opportunity for a pediatrician. Promoting pulmonary growth by providing adequate nutritional intake, ensuring sufficient oxygenation, and attending to all aspects of health supervision is the foundation of excellent pediatric medical practice. To achieve these goals, the pediatrician must form an active therapeutic alliance with the parents. Drs. Gorski and Vaucher make it clear that an optimal developmental outcome for a premature infant with CLD occurs when the pediatrician applies medical knowledge while supporting the adaptation of the parents as they assume a more active role in their infant’s care after discharge from the NICU.

My experience with children like Timmy has taught me the value of a close working relationship with members of the NICU staff and those professionals who participate in the infant’s care after discharge. There is enormous value in examining an infant in the hospital before discharge and reviewing important aspects of medical care, neurodevelopmental status, and behavioral responses to various events. Even a single visit to the NICU around the time of discharge provides an opportunity to understand the unique features of the infant’s medical course and behaviors. Communication with the nurses who care for the child yields important information that guides future care. Finally, parents appreciate the interest in their child that is demonstrated by the pediatrician at this time; not surprisingly, the therapeutic alliance is often strengthened during these predischarge encounters. Timmy’s pediatrician should be commended for her attendance at the discharge conference.

The question raised by Timmy’s pediatrician, “What can I do to enhance the development of a premature infant with chronic lung disease?” was addressed by Drs. Gorski and Vaucher, who made numerous practical recommendations. In addition, they both emphasized the significance of continuous support for the parents in their role as primary caregivers. This work is challenging when, in Mrs. Ketel’s words, “the parents wish that ear infections and tubes were the only things they had to worry about. . . . they wonder what the future holds for their child, both physically and mentally.” It is the parents’ perception of vulnerability that can promote a belief that the infant is more fragile, physically and developmentally, than an objective medical assessment might conclude. The recognition that all infants who graduate from an NICU after multiple life-threatening events are at risk for a vulnerable child syndrome should guide pediatric care. Monitoring and interpreting strengths in the infant and the family at each office visit models a positive and optimistic point of view. Discovering developmental and behavioral strengths during a physical examination or while observing the manner in which a parent holds, communicates with, and responds to a premature infant is the value of excellent pediatric care. Communicating these observations to parents in precise and explicit language is a therapeutic event. It boosts their perception of themselves as effective parents and is a preventive strategy for a vulnerable child syndrome.
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