The Chronically Critically Ill Patient: Pediatric Considerations

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Whether defined as chronically critically ill, long-term mechanical ventilator dependent (or otherwise chronically medically supported), or medically fragile, a population of infants and children with chronic illness clearly exists. Infants and children with chronic healthcare needs are at an increased risk for physical, developmental, behavioral, and/or emotional conditions and generally require healthcare services of a type or amount beyond that of a general pediatric or adult population. This review will focus on the specific management and psychosocial needs associated with the healthcare of this subgroup of infants and children with chronic illness. Attention will be paid to defining the population, describing trends over time, reviewing their special needs, and discussing outcomes. Increased focus and an increasing quantity of resources for this subgroup of infants and children are needed, as the number of such pediatric patients continues to grow. *Key words: pediatric; mechanical ventilation; tracheostomy; chronic illness; respiratory failure; neuromuscular weakness; long-term care.* [Respir Care 2012;57(6):993–1002. © 2012 Daedalus Enterprises]

Introduction

As a basis for this review, the differences between children and adults in terms of the chronically critically ill patient must be considered. As the proceedings from this Journal Conference have described, a complex population

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of chronically critically ill adult patients clearly exists. However, is the same true for infants and children? Is there a subset of pediatric patients who have the same clinical entity of an inflammatory chronic critical illness, as defined previously in this issue of RESPIRATORY CARE? At first thought, any clinician working in a neonatal or pediatric ICU would answer in the affirmative. However, is the infant or child who remains mechanically ventilated beyond 2–3 weeks really similar to the adult patient with prolonged respiratory failure? Or would this type of patient in the neonatal or pediatric ICU be better described as having prolonged acute critical illness? The answers to these questions cannot be found in the medical literature, as there is a clear void of published information.

Regardless of how one answers the previous questions, there is clearly a subgroup of neonatal and pediatric patients who have a prolonged period of critical illness. At some point in the process, the emphasis of medical care for these patients turns from acute to chronic. For those patients with chronic respiratory failure this arbitrary transition from acute to chronic is often viewed in the perspective of medical providers as the time of tracheostomy placement. For those patients with chronic cardiac or neurologic failure, the transition point is generally much less clear.

Marcin et al[1] attempted to define long-stay versus short-stay pediatric ICU (PICU) patients, and arbitrarily chose a cutoff of 12 PICU days. Based on this definition, long-stay patients comprised only 4.5% of the PICU population but occupied 36% of the total bed days. The long-stay patients were more likely to be technology-dependent upon discharge.

Thus, it must be left to the interpretation of the reader whether this group of neonatal and pediatric patients should be defined as: chronically critically ill; long-term mechanical ventilator dependent (or otherwise chronically medically supported); or, as is often described in the medical literature, medically fragile. This population of *medically fragile* pediatric patients is frequently defined as: technology dependent, including the need for chronic oxygen therapy, invasive or noninvasive ventilatory support, continuous or intermittent cardiorespiratory monitoring, chronic dialysis, a tracheostomy tube, a cerebrospinal fluid shunt, a gastrostomy tube, and/or a central venous catheter; having severe neurodevelopmental impairment requiring ongoing medical support; and/or diagnosed with 3 or more chronic medical conditions. More generally, the Maternal and Child Health Bureau has defined children with special healthcare needs as those who have or are at increased risk of a chronic physical, developmental, behavioral, or emotional condition and require healthcare and related services of a type or amount beyond that required by children generally.[2] Regardless of the specific description or definition, this review will describe the population of high-risk infants and children who require chronic medical care for respiratory, neurologic, renal, and/or cardiac dysfunction/failure.

Defining the Pediatric Patient with Chronic Illness

Infants and children with special healthcare needs are at an increased risk for physical, developmental, behavioral, and/or emotional conditions, and generally require healthcare services of a type or amount beyond that of a general pediatric population.[2] Among this subgroup is a smaller number of medically complex, or medically fragile, patients including those with numerous medical needs resulting from multisystem disease states, technology dependence, and/or complex medication treatments.[3] Advances in neonatal and pediatric critical care, as well as improvements in general medical and nutritional care, have resulted in higher survival rates of these medically fragile infants and children, who are often left with complex systemic health problems.

Beyond the specific name for this subgroup of pediatric patients who require chronic medical care, we must understand the specific pathophysiology of the underlying condition(s), including the overall individual clinical situation. As a basis for this discussion, it must be remembered that anatomy, physiology/pathophysiology, and pulmonary and neurologic development vary greatly based on age. The differences from pediatrics to adulthood become magnified when one considers the psychological, social, and economic factors related to chronic medical illness. Along these lines, it must be stressed that the continuum of pediatric medicine from neonates to adolescents is tremendous. The specific disease processes, their clinical implications, and the needs of these patients are varied.

From a respiratory perspective, children, especially younger children, have smaller airways (both natural and artificial), weaker and less effective cough clearance, increased chest wall compliance, decreased diaphragmatic efficiency, and an overall higher risk for airway occlusion. From a cardiac perspective, most infants and children have both structurally and functionally normal hearts but may have less cardiac reserve than many adults. Special consideration must be given to the relatively large subset of pediatric patients with congenital heart disease, which comprises an important group of pediatric patients with chronic, critical illness.[4] Nutritionally, infants and children, especially those with chronic illness, generally have much lower nutritional reserve than their adult counterparts. Lastly, it must be noted that the majority of chronically critically ill pediatric patients have comorbidities, many of which are congenital—metabolic, respiratory, central nervous system, and/or cardiac—in origin.

Beyond the physical and physiologic differences between pediatric and adult chronically ill patients are the

vast socioeconomic, psychological, and developmental differences. Although a few may debate this point, most would agree that chronically ill pediatric patients impart a greater socioeconomic and psychological toll on their families and other caregivers than occurs in the chronically ill adult population. Being a parent has its challenges, but being the parent of a child with chronic medical illness magnifies these challenges many-fold.[5–11] However, it must also be stressed that the psychological rewards for caring for these children can be tremendous.[5,12] This complex aspect of chronic illness is discussed in more detail later in this paper.

Children are clearly not small adults when it comes to the location for the care provided for this complex population. As has been described in previous papers from this Journal Conference, chronically critically ill adult patients may be managed in the acute care setting (ICU or special care unit), long-term acute care facilities, skilled nursing facilities, or in their homes. However, the pediatric population varies greatly, as the available number of long-term acute care and skilled nursing facilities centers who will care for pediatric patients (especially infants and young children) is extremely limited.

Thus, the population of medically fragile pediatric patients are most commonly found in their home or in the acute in-patient setting. Due to their complex medical conditions and the frequent need for mechanical ventilatory support, the most common in-patient hospital environment is either the pediatric ICU or a pediatric step-down/progressive care unit. Aside from exacerbations, many of these children are managed by their parents (or other family members), with the assistance of home health workers in their own homes, although a small portion of these children for various reasons may require placement into foster care. Although skilled nursing/rehabilitation facilities are often utilized for adult patients, the number of such facilities for pediatrics, especially for infants and young children, is exceedingly small and highly regionalized (Fig. 1). Much less commonly utilized locations are residential homes (generally adolescents) and hospice programs.

A frequent resultant discussion in the care of the chronically critically ill pediatric patient is whether the infant/child would be better managed in the in-patient setting or in his/her own home.[13,14] Children clearly have the right to be cared for in an environment that is most suited to their medical, psychological, and developmental needs. Prolonged hospitalizations can have a very negative impact on infants and children.[14] In this balance, one must carefully weigh the various (and often opposing) factors.

An in-patient, and especially an ICU, setting provides for a substantially higher level of monitoring, observation, experienced staffing, and management than the home or a long-term care facility. On the other hand, such an acute care in-patient admission can be very costly and could be considered by some to be an unnecessary allocation of a limited resource. Care for this patient population in the home setting allows for improved patient and family comfort. The overall medical management of the patient in his/her home is almost always more cost-effective medical care. However, the safe medical care of this vulnerable population in the home setting can be considered more challenging and stressful, especially when adverse events (even minor ones) occur.

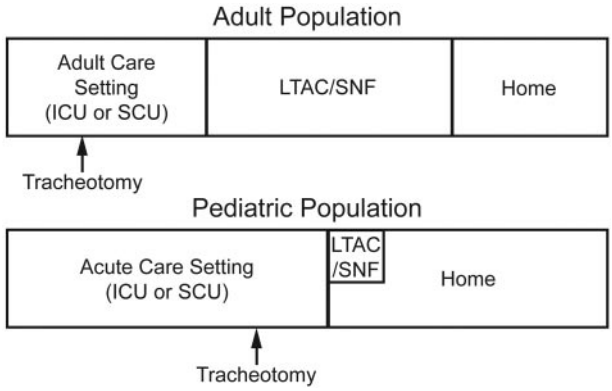

Fig. 1. Care of chronically ill adult and pediatric patients by location. Top: Most chronically ill adult patients are managed across a continuum of an acute care setting (ICU or special care unit [SCU]), long-term acute care (LTAC), or skilled nursing facilities (SNF), and their homes. Bottom: Chronically ill pediatric patients are more likely to be managed in either an acute care setting or their homes.

Often the decision of where to best manage complex pediatric patients with chronic healthcare needs rests with the intensity and frequency of the actual medical care required for an individual infant or child at a given point in time. The need for intravenous medications, intravenous infusions, and/or complex or frequent ventilator or airway manipulations may warrant an in-patient acute care admission. Other potential factors include the route of enteral nutrition (oral, nasogastric tube, or gastrostomy tube), requirements for wound care, and any need for aggressive rehabilitation (physical, occupations, and/or speech).

Trends and Outcomes

The population of chronically critically ill pediatric patients is diverse and encompasses numerous congenital and acquired conditions. Benneyworth et al[15] estimated the prevalence to be 6–14 per 100,000 children. In terms of in-patient admissions, the rate is estimated to be 174 per 100,000 non-newborn pediatric discharges. Infants and young children consume a higher proportion of these healthcare resources than older patients.[15,16] The number of infants and children with chronic respiratory failure requiring long-term ventilatory support is gradually in-

creasing as a consequence of improved overall medical treatment and technologic advances, which have contributed to longer survival of chronically ill patients and the development of improved home medical equipment.[17,18] Cross-sectional studies have attempted to describe the population of children requiring long-term mechanical ventilation, with the prevalence estimated at 6.3/100,000 children ≤ 16 years of age.[19–21]

Recent economic trends in the care of the chronically critically ill pediatric patient are concerning. The frequency of in-patient care has increased for all children with complex chronic conditions over the last decade.[22] From 2000 through 2006, the in-patient discharge rate for infants and children requiring long-term mechanical ventilation has increased by 55%.[15] Although mean stay decreased during this time frame by 18%, the total charges for these admissions increased by approximately 70%. It must be stressed that a limitation to this report is the lack of cost data, as the relationship between charge and cost may have varied over the time frame studied.[15] An interesting trend is that the in-patient discharge rates for patients between 1 and 9 years of age increased more dramatically than for infants, preteens, or adolescents. However, the charges per admission increased most substantially for the infant population. From a financial perspective, it should be noted that this study reported an almost doubling of the population supported with public financing, while the population with private insurance increased by only 12%.

Burns et al[23] assessed hospitalization rates for pediatric patients with chronic illness based on the underlying diagnoses. From 1991 through 2005, the rate of in-patient admission for those patients with a single underlying diagnosis increased by almost 20%. However, the rate for those patients with multiple underlying diagnoses more than doubled. Subgroup analyses for those patients with primary diagnoses of either bronchopulmonary dysplasia or cerebral palsy demonstrated similar findings to the group as a whole. Adjusted odds ratios of death were compared to a child without chronic illness for those requiring a gastrostomy tube, tracheostomy tube, or cerebrospinal fluid shunt. The risk of mortality was clearly greater for those patients in each of these categories who had multiple underlying chronic conditions. The highest risk of mortality (odds ratio 95.5, 95% CI 74.7–122) was for the subgroup of patients with multiple chronic conditions requiring a cerebrospinal fluid shunt (Table 1).[22] This table also demonstrates that the overall risk of mortality for infants and children requiring in-patient admission was significantly higher for those with any chronic medical condition.

Edwards et al[24] reported their 22 year, single center, university-affiliated, home mechanical ventilation program experience, including 228 children. The overall 5-year survival was 80% with a 5-year rate of liberation from mechanical ventilation of only 24%. As would be predicted, the subgroup with chronic respiratory disease were more likely to be weaned from mechanical respiratory support (29.1%, 95% CI 21.2–38.2%) than were those with either neuromuscular disorders (6.4%, 95% CI 1.8–15.7%) or congenital hypoventilation syndrome (4.3%, 95% CI 0.5–14.8%). Of importance, several publications have claimed that mortality of chronically ventilated children in the home environment is primarily influenced by the clinical course of their underlying disease, rather than issues directly related to their respiratory support.[25–27]

Table 1. Adjusted Odds Ratios of Death, as Compared to a Child Without Chronic Illness

Technology	Single Complex Chronic Condition	Multiple Complex Chronic Conditions
Gastrostomy tube	9.2 (8.1–10.4)	27.0 (22.8–31.9)
Tracheostomy	8.8 (5.4–14.3)	31.4 (17.3–56.9)
Cerebrospinal fluid shunt	51.2 (41.6–63.0)	95.5 (74.7–122.0)
In-patient mortality	5.2 (4.8–5.6)	12.5 (11.4–13.8)

Values are adjusted odds ratios and 95% CIs.
(Data from Reference 22.)

Technology Dependent Children

Tracheostomy Placement and Care

As shown in Figure 1, the placement of a tracheostomy tube for the infant or child generally occurs much later in an in-patient admission than for adults. The exact timing of tracheostomy placement is determined on an individual patient and/or institutional basis, without reliance on any generally agreed upon guidelines. The performance of a tracheostomy most often identifies the transition from acute care to chronic care, at least as commonly interpreted by neonatal and pediatric medical care teams. It should be noted that tracheostomy placement in the pediatric population is common, with approximately 5,000 procedures annually in the United States.[28]

Graf et al[4] found that the most common reasons for tracheostomy placement in the pediatric population are airway management (63%), chronic respiratory disease (23%), central hypoventilation (9%), and neuromuscular weakness (6%). Other general indications include chronic respiratory disease, central hypoventilation syndrome, and neuromuscular weakness in descending order of occurrence. When identifying specific underlying diagnoses, the most common include congenital heart disease, airway malacia (often secondary to congenital heart disease), mental retardation/cerebral palsy (for either airway control or secretion management), primary lung disease, and gastroesophageal reflux disease (generally as a secondary find-

ing).[4] The majority (66%) of these tracheostomies were placed after a period of prolonged mechanical ventilation. The remaining were placed during an elective admission (26%) or as a result of an emergency (8%).[4] A disproportionate number of the tracheostomy placements that occur within the pediatric population have been reported in those under one year of age.[16,29]

It should be noted that the risks of tracheostomy placement are more pronounced in pediatric patients, as compared to the adult population, due to the smaller caliber of both the airway and the tracheostomy tube. Potential difficulties and risks, which should be anticipated, include an impediment to swallowing and normal phonation, tracheitis, pneumonia, mucus plugging, unintentional decannulation, and granulation tissue formation at the stoma or in the airway. Intense vigilance and training are required for the family members and home health providers caring for infants and children with tracheostomy tubes. Despite the potential hurdles, tracheostomy placement often allows these patients to proceed to rehabilitation and/or continued care in their home.

Noninvasive and Invasive Long-Term Ventilators

The majority of the population of chronically ill infants and children are defined by the need for long-term respiratory support, as previously mentioned. Racca et al[30] described 362 pediatric patients requiring chronic ventilatory support. Of these infants and children, 41% were invasively ventilated, while the remaining 59% were managed with noninvasive respiratory support. Those patients requiring invasive ventilation were significantly younger than their counterparts who were ventilated noninvasively (1 year vs 8 years, $P < .001$), and 81% of the invasively ventilated patients required mechanical support for > 12 hours per day, while only 16% of the noninvasive ventilation group required mechanical support for > 12 hours per day ($P < .001$). Although not reported, one would suspect that the limited number of interface options for infants and children results in an inability to rotate through several options, and, thus, an increased tendency for skin breakdown if used for much more than 12 hours per day. Of note, for a comparison to the adult population, please see the publication by Hess in these proceedings.[31]

As compared to the adult population, the technology options to support infants and children who require chronic mechanical ventilation, either invasive or noninvasive, are limited. In terms of noninvasive ventilation, the biggest hurdle is an inadequate variety of FDA approved interfaces and securing devices, although the number of options has recently increased. The optimal interface is one that allows for patient comfort, an appropriate seal, and avoids skin breakdown.

For invasive ventilatory support, the most common technologic problems encountered with chronic care ventilators are inadequate trigger sensitivity, a high expiratory resistance, an inability to respond to variable air leak, and a slow response time, especially in relation to the relatively high respiratory rates seen in the young pediatric population. Several of the currently available pediatric home ventilators have adequately addressed these limitations. Fauroux et al[32] studied several of the available home ventilators used in the pediatric population in an attempt to determine tidal volume accuracy. In general, the 6 ventilators studied provided reliable tidal volume delivery in the face of increased airway resistance or decreased pulmonary compliance. However, in general, these ventilators were unable to provide accurate tidal volume delivery when an unintentional air leak occurred.

Monitoring

The requirements for home cardiorespiratory monitoring of the pediatric mechanically ventilated patient can be debated. The clinician must determine for each individual infant or child whether he or she requires pulse oximetry (continuous or intermittent) and/or capnography (time-based or volume-based). General clinical guidelines include the need to provide cardiorespiratory monitoring for infants with a recent history of apnea and/or bradycardia, a history of any acute life threatening event, and/or clinically important gastroesophageal reflux. Recommendations for those who require continuous pulse oximetry and/or capnography are less clear.

The optimal home monitor is one that is reliable and accurate, has download capabilities, and includes battery backup. The ability to download data is important to assess patient trends over time as well as to assess any episode(s) of clinical instability (eg, bradycardia, apnea, or desaturation). Optimally, the monitor would have the ability to provide remote download capability (eg, telephone or internet) to allow a medical care provider to quickly assess the data, especially for those patients who may live at a substantial distance from their medical care providers.

One of the key components of the debate over the need to monitor the chronically ventilated pediatric patient at home involves alarms. Loose leads are a frequent cause of false cardiorespiratory alarms. Patient movement can cause false pulse oximetry alarms. These false alarms, as well as ventilator alarms (both real and false), can quickly lead to alarm (ie, sensory) overload and an inability to distinguish real alarms from false ones. On the other hand, the alarms on the various devices are not always adequately sensitive (ie, an alarm may not occur despite the occurrence of a real problem), and, thus, the lack of a triggered alarm can provide false reassurance to a child's parents and other caregivers in the event of a real problem.

As technology improves, hopefully the reliability and accuracy of alarms will improve as well. But for now the debate over how chronically ventilated patients are monitored, and to what degree, in the home environment will continue for some time.

Neuromuscular Weakness

The clinical expression of pediatric patients with static or progressive disorders of neuromuscular weakness can be quite varied. There is a substantial proportion of these patients who require intense and coordinated care for their respiratory, cardiac, nutritional, and developmental needs. This group spans from infants through adolescents and encompasses patients with spinal muscular atrophy, muscular dystrophy (eg, Duchenne, Becker), and various other congenital or acquired myopathies, as well as victims of spinal cord injury and traumatic brain injury. This population varies in the severity of muscle weakness as well as cerebral function, which can range from normal to severe developmental delay.

When managing this heterogeneous population, the clinician must consider the specific disease process, the anticipated clinical progression, and any other organ systems that may be affected. In most cases of congenital neuromuscular disease, there is involvement of the bulbar muscles, affecting feeding; abdominal and intercostal muscles, affecting cough strength and effectiveness; and skeletal muscles, leading to a decreased ability to ambulate and potentially even stand. Care must be carefully integrated to maintain as much neuromuscular function as possible, promote cerebral development, minimize complications from muscle weakness (eg, aspiration, contractures), and provide emotional and psychosocial support for the patient and his/her family.

Of utmost importance is the recognition of impaired cough, decreased mucociliary clearance, and impaired gas exchange. Aggressive monitoring and intervention with airway clearance techniques and noninvasive ventilatory support have been shown to reduce morbidity and prolong life for muscular dystrophy patients.[33]

Respiratory monitoring in children with neuromuscular weakness includes the assessment of inspiratory and expiratory muscle strength, including maximal inspiratory pressure, maximal expiratory pressure, forced vital capacity, and FEV_1, using spirometry. Polysomnography with blood gas measures of P_{aCO_2} help to assess nighttime, and potential daytime, hypoventilation. FVC has been demonstrated to predict a worsening respiratory status in patients with Duchenne muscular dystrophy.[34]

Static lung volumes using plethysmography will identify restrictive lung disease from a combination of kyphoscoliosis, decreased chest wall compliance, and reduced inspiratory muscle strength.[35] As muscular weakness progresses, the patient will become less able to clear secretions, leading to atelectasis and pulmonary infections. In addition, nocturnal hypoventilation can be monitored with polysomnography and treated with noninvasive or invasive ventilation prior to progression into diurnal hypoventilation and respiratory failure.

Airway Clearance Techniques

For infants and children with neuromuscular weakness, most episodes of acute respiratory failure are due to impaired mucociliary clearance during benign respiratory infections.[36] The therapeutic aim in this population is to reduce mucus viscosity and improve cough clearance on a routine basis, with amplification of airway clearance techniques during periods of acute illness, especially respiratory infections, and perioperatively. The goal is to mobilize the mucus from the peripheral airways to the larger, more central airways for easier expulsion.[35] Commercially available mucolytics that may aid in decreasing mucus viscoelasticity include dornase alfa and N-acetylcysteine. Of note, there are no therapeutic trials to demonstrate reduced pulmonary morbidity in this population with these inhaled therapies, and thus they are generally used by pediatric pulmonologists and intensivists on a case by case basis.

Manual chest percussion and/or vibration with a circumferential chest vest are often clinically prescribed multiple times a day, with escalation during acute illness. Mechanical in-exsufflators are cough assist devices that can be used with a face mask, mouthpiece, or in-line with an endotracheal or tracheostomy tube. These devices deliver deep insufflations with positive pressure until the lungs are inflated, followed immediately by a negative pressure exsufflation, with the goal of facilitating mucus mobilization. Multiple such cycles are provided until no further secretions can be induced.[36]

Discharge Planning

Discharge planning for the chronically ill pediatric patient should start as early in the in-patient admission as possible. This planning should include regular multidisciplinary conferences with a discussion of the various options—continued admission to the acute care setting (ICU or special care unit), transfer to one of the few pediatric long-term acute care or skilled nursing facilities, or discharge to home. Below we discuss the processes involved with discharging a medically fragile pediatric patient to the home environment.

The discharge planning process should include an evaluation of the patient's (if age appropriate) and his/her family's psychosocial readiness for discharge from the acute care setting, an assessment of the family's economic read-

iness for discharge, the availability of pediatric trained home healthcare resources (nurses and/or respiratory therapists), the need for physical, occupational, and/or speech therapy, and plans for schooling as age appropriate. An on-site home safety evaluation is essential. The child's home environment must have appropriate electricity to support a mechanical ventilator, a reliable clean water supply, reliable telephone access, and a reliable means of transportation. Overall, the home environment must be considered safe for the medical needs of the individual infant or child.

Although quite variable by location, additional requirements for discharge to home often include at least 2 trained adult family (immediate or extended) members. Training in the care of the child should include videos, printed material, and, most importantly, hands-on experience. The necessary equipment (with backup as clinically indicated) must be arranged well in advance of the planned discharge date. In addition, one may consider designing a "care contract" with the family (and child as age appropriate) to clearly set expectations for both the family and the medical care team who will provide primary care for the patient after discharge.[24]

Transition From Pediatric to Adult Healthcare

With the increasing number of chronically ill and medically complex children in our society, an important part of primary and subspecialty medicine is the transition from pediatric to adult healthcare. Improvements in medical therapies, including intensive care, technology, and approaches to rehabilitation, have increased the life expectancy in many chronically ill pediatric populations, including cystic fibrosis, chronic respiratory failure, and neuromuscular weakness.

Transition to adult-based healthcare services may allow for increased independence for developmentally appropriate adolescents and young adults, but this transition can be associated with real angst among these patients and their families. Additionally, many adult physicians have little experience in caring for these traditionally "childhood illnesses." As described in a recent review on this topic by Crowley et al,[37] there can be adverse events on healthcare and health service use around the time of transition. Successful categories of intervention to improve the transition experience from pediatrics to adult medicine include focused education programs; skills training sessions; special staffing, including transition coordinators and combined clinics with both pediatric and adult physicians (ie, transition clinics); and targeted services (eg, young adult clinics, telephone support programs).

Pediatric Palliative Care

With an increasing ability to support various healthcare conditions with medical technology, a patient and family centered approach must be provided for our pediatric patients with chronic critical and deteriorating health conditions. This includes, where possible and when appropriate, the input and consultation from pediatric palliative care teams and/or hospice programs. Feudtner and colleagues[38] reviewed data from the National Center for Health Statistics from 1989–2003 and determined that the adjusted odds of pediatric patients with "a complex chronic condition" dying at home has increased significantly from 1989 to 2003 in all age groups. Since these children are increasingly dying at home, introducing the concept of palliative care earlier in the course of chronic illness may be appropriate for many of these patients.

In a prospective multicenter cohort study by Feudtner et al,[39] demographic, clinical characteristics, and outcomes were described for all patients served by the pediatric palliative care teams in 6 North American hospitals from January through March, 2008, with 12-month follow-up. Of the children who received a consult, 17.1% were < 1 year of age, 37.5% were 1–9 years of age, 30% were 10–18 years of age, and 15.5% were 19 years of age and older. The majority of patients (55%) had more than one principal diagnosis. The breakdown of the most common clinical conditions were genetic/congenital (40.8%), neuromuscular (39.2%), cancer (19.8%), respiratory (12.8%), gastrointestinal (9.9%), and cardiovascular (8.3%). Patients were receiving a mean of 9 different medications at the time of consultation. Eighty percent of the 515 patients used at least one form of medical technology, with the most common being a feeding tube (68.2%), followed by central venous catheter (22.3%) and tracheostomy (10.1%). In addition, 9.5% required some form of noninvasive ventilation, and 8.5% were listed as being ventilator dependent. The main goals of consultation were symptom management, facilitating communication, decision making, and assisting with coordination of overall care. During the 12-month follow-up, 30.3% of the patients in the cohort had died with almost two thirds dying in the hospital. Approximately one quarter died at home, and a small minority died in a hospice care facility. In summary, pediatric palliative care consults occur most often among patients with complex, chronic medical conditions, and can aid with symptom management and transition to comfort care measures. Such an approach allows the patient and family control of medical interventions, resuscitation plans, and an opportunity for optimal quality of life whether in the hospital or at home.

The Intangibles

There are numerous intangibles that factor into the care of the medically fragile pediatric patient in the home environment. One of the biggest of these factors is the experience of the primary pediatric care providers for this challenging population, as well as the pediatric experience

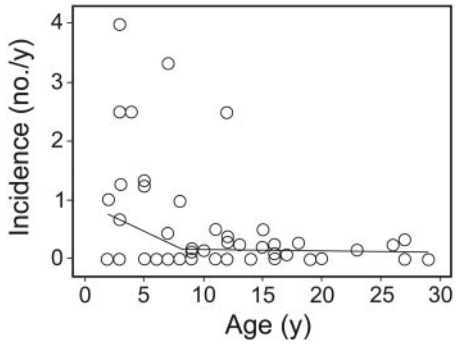

Fig. 2. Incidence of severe emergencies (number of severe emergencies per patient-year of home mechanical ventilation) versus patient age ($P < .002$). The line represents the locally weighted polynomial regression curve. (From Reference 41, with permission.)

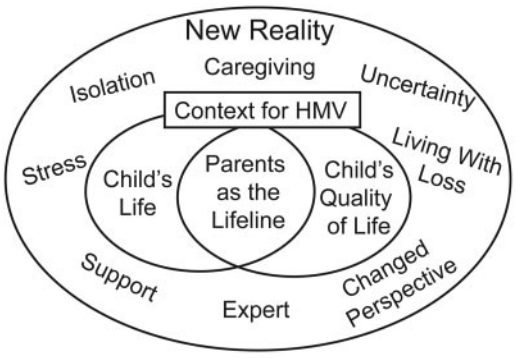

Fig. 3. Themes representing the experience of parents with a child on home mechanical ventilation (HMV) due to neuromuscular disease. (From Reference 5, with permission.)

and training of the home health agency personnel. Kun et al[40] assessed the knowledge of primary care providers in terms of tracheostomy and home ventilator management. Overall, the providers were able to answer 80–85% of the questions accurately; however, there were knowledge gaps in some critical areas. Sixty-three percent of the primary care providers did not know that a low pressure alarm may not sound in the event of tracheostomy decannulation, especially if the ventilator is still connected. Fifty-two percent did not know the association between a high pressure alarm and mucus plugging. Thirty-seven percent relied on ventilator alarms to diagnose mucus plugs. Forty percent did not know the importance of low pressure or low minute ventilation alarms. These knowledge deficits were not related to language barriers, as English and Spanish speaking providers scored similarly. Also of interest is that scores were similar regardless of the years of experience of the medical care providers.

The knowledge of the primary care providers, as well as the experience and training of the home health agency personnel, is essential, as emergencies in the home setting for these chronically ventilated patients are uncommon but do occur, especially for infants and younger children.[41] As such emergency situations are relatively rare, it should be noted that both the parents and the home healthcare personnel probably have never had hands-on training/experience in such situations. Reiter et al[41] studied 295 patient-years of home mechanical ventilation in 54 pediatric patients. These authors found 68 severe emergencies, representing a rate of 0.2 per patient-year (Fig. 2). Respiratory causes were determined in 48 of these cases, including 15 related to tracheostomies, and 3 ventilator failures. Unfortunately, this report does not clearly define *severe emergency*. It should be noted that these emergency situations resulted in 4 deaths.

The care of the medically fragile child in the home environment has a series of socioeconomic and psychological sequelae. These effects are generally considered to be greater for the chronically ill pediatric patient than for the corresponding adult population. Infants and children, by definition, are less able to provide self-care than their adult counterparts. As previously mentioned, the home healthcare personnel and primary care medical providers tend to have less training and be less experienced with technology-dependent pediatric patients. Parental stress is common, especially as at least one of the parents is generally removed from the workforce, thus exacerbating any financial difficulties related to the expenses of prolonged in-patient admissions and the overall medical support of the child with a chronic illness. Routine education and normal development of the medically fragile child can be challenging. A failure to allow for education and psychosocial development of the chronically ill child can further compound the complex situation.[42] Also, it should be noted that, although many would recommend normalizing the environment for these patients as much as possible, integrating these vulnerable children into the routine daycare/school environments often exposes them to the expected viral infections of infancy and childhood. The risk/benefit ratio of such decisions can be difficult and should be individualized.

Mah et al[12] reported that the Pediatric Quality of Life Inventory scores (both physical and psychosocial) for those chronically ill children who required home mechanical ventilation were significantly lower than similar patients who did not require chronic respiratory support. However, in this specific subpopulation of families of children with neuromuscular disease, no significant difference was seen in total stress scores. These authors postulated that "for parents living with the constant demands of caring for their child with neuromuscular disease requiring home mechanical ventilation, these caretaking demands, over time, had become part of 'normal' life and were not identified as creating additional stress."[12] Parents have reported that the presence of medical equipment in their home was both

supportive and disruptive.[43] The complex themes representing the experience of parents with a child on home mechanical ventilation due to neuromuscular disease are represented in Figure 3.[5]

Summary and Thoughts for the Future

Whether defined as chronically critically ill, long-term mechanical ventilator dependent (or otherwise chronically medically supported), or medically fragile, a population of infants and children with chronic illness clearly exists. Infants and children with chronic healthcare needs are at an increased risk for physical, developmental, behavioral, and/or emotional conditions, and generally require healthcare services of a type or amount beyond that of a general pediatric or adult population. Specific management strategies and psychosocial support for these patients and their families are essential to optimal outcomes. Increased focus and an increasing quantity of resources for this subgroup of infants and children are needed as the number of such pediatric patients continues to grow.

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Discussion

MacIntyre: It sounds like CCI [chronic critical illness] in pediatric patients is really mostly mechanical lung issues, as opposed to these chronic inflammatory states we've been hearing about. Is that a fair statement?

Cheifetz: In terms of how many pediatric providers would view it, the answer is most likely yes. After yesterday's excellent presentations and discussions, I have been thinking more about the pediatric population. In pediatrics there are probably two distinct populations. One includes patients who require long-term mechanical ventilation. But I believe there is another subset of patients who practitioners view as acutely ill and remain so for weeks. Does this second population fit the definition for CCI as described yesterday? I believe the answer is yes, but often this category of patient is viewed as having "prolonged acute illness" rather than being "chronically critically ill." The difference may be one of nomenclature. Either way, this topic in pediatrics has not been well studied. I searched PubMed again last night for CCI and pediatrics and found a void.

MacIntyre: Does what Jeff Mechanick presented this morning on homeostatic and allostatic overloads[1] apply to the pediatric population?

1. Schulman RC, Mechanick JI. Metabolic and nutrition support in the chronic critical illness syndrome. Respir Care 2012;57(6):958-977; discussion 977-978.

Cheifetz: I'm not sure. I believe it may apply, but this is such a new area for pediatrics, with a lack of published data.

Muldoon:* The observation I have from your talk is this circle-of-life concept, where taking care of the pediatric patient is very much like taking care of the 70-year-old type-1 CCI patient. All those psychosocial stressors are the same. We have something to learn from the pediatrician. These children are surviving, so what happens to them when they're no longer pediatric?

Cheifetz: More generally, when does an adolescent with chronic illness become an adult? That is, when should care be transferred from pediatric to adult services? Most children who are chronically ill, especially those with neurologic injury and/or requiring prolonged mechanical ventilation, are transferred to the adult world in their late teens to mid-20s. Many are then placed in the same adult facilities that were discussed yesterday. The transition point is variable; much depends on the child, or adolescent at that point, and the resources available.

Bertuola:† We have a couple of ICFMR [independent care facilities for the mentally retarded] facilities. We are seeing that the patients are aging, they're requiring ventilation, and we're providing the care in the ICFMR centers. It might be a different avenue for pediatric patients; there are more skilled nursing facilities opening up for pediatric patients as well.

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Cheifetz: Good point. The trend is that this population is really growing. It is unclear whether the hospital discharge data represent additional patients or simply that technology is keeping children alive longer to become adolescents and young adults. Either way, I agree with you: we will have more and more need for additional chronic care resources.

White: We see some of these late-teens/early-20s cases in our LTAC [long-term acute care facility]. They're typically people with cerebral palsy who have airway issues, some very young neuromuscular patients, and occasionally some patients with Down syndrome who have airway issues and need a tracheotomy. It's usually more of the population of patients who just have respiratory issues: upper airway or secretion issues or lung issues. In our LTAC we're not seeing the catastrophic CCI cases that we see in the adult population. These patients are presumably at the pediatric hospitals and not coming to LTACs.

Cheifetz: The issue in pediatrics is much less clear. There is the subset of such patients who are not chronically ill in the inflammatory sense, as described yesterday, and who require prolonged mechanical ventilation for chronic respiratory failure and/or neurologic injury. Then there is the subset who are either chronically critically ill or have a prolonged acute critical illness. Either way, the pediatric practitioner frequently manages this type of patient as acute until a tracheostomy is placed, which could be weeks or sometimes months later.

Regardless of whether an inflammatory CCI process plays an active role, there is a point at which these children truly are defined solely by their need for chronic respiratory and/or neurologic support. I would say that most, if not all, of these pediatric patients have made this transition by the time of discharge. Thus, the best description of this chronically ill population may be the third in the potential subtitles for my presentation: *the medically fragile child*. This is probably the best description for this population once beyond the ICU setting.