Hospitalist Care of the Medically Complex Child

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The nature of inpatient pediatrics is changing. Over the past decade, several factors have converged to influence the kinds of children currently being hospitalized. Managed care organizations have been under increasing pressure to control costs and reduce unnecessary prolonged hospital stays. Many emergency departments are using observational units to avoid hospitalizations while reserving inpatient wards for higher acuity and complex patients [1]. There has been a shift in the perception in the minds of clinicians as to what constitutes an appropriate hospital stay and what may be treated on an outpatient basis. Therapies such as home oxygen for certain pediatric conditions (eg, bronchiolitis) and home intravenous therapy for fluids and medications are being used increasingly. These developments have produced a shift in the relative proportion of otherwise healthy children with simple, self-limited acute illness being hospitalized to children with chronic illnesses presenting with acute exacerbations or consequences of their underlying illnesses being cared for in the hospital [2]. This article focuses on hospitalist care of these medically complex children (MCC) and provides an overview on (1) the challenges in defining this population, (2) the unique issues surrounding their inpatient care (using a family-centered care approach that includes coordinated care, minimizing secondary complications, nutritional needs, functional limitations, transdisciplinary collaboration, and pri-

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mary care issues), (3) technology devices commonly found, and (4) a proposal for a research agenda regarding MCC.

Defining the medically complex child

Defining this population has been challenging for health services researchers and clinicians. Several recent initiatives have shaped the current conceptual model. In 2001, the Institute of Medicine recommended that the Agency for Health care Research and Quality identify "15 priority conditions, taking into account frequency of occurrence, health burden, and resource use" [3]. The Institute of Medicine identified children with special health care needs (CSHCN) as a priority population [3,4]. Researchers and experts in this field have published the following definition: "Children with special health care needs are those who have or are at increased risk for a chronic physical, developmental, behavioral, or emotional condition and who also require health and related services of a type or amount beyond that required by children generally" [5]. Although CSHCN comprise between 13% and 18% of all children [6], they account for more than 80% of the cost of health care for children in the United States [5]. CSHCN are two to three times more likely than other children to have unmet health care needs that ultimately lead to belated, dramatic, and expensive interventions [7]. Children with chronic health conditions are three times more likely to have an unscheduled intensive care unit admission than healthy children, and 32% of these admissions are judged to be potentially preventable [8]. Of the potentially preventable events, 64% are related to health care system deficiencies, such as inadequate care coordination. Medical errors are an especially important example of preventable events, and errors in care for CSHCN are likely to be higher than for other children because of the complexity of their care [9,10]. Because these children are approximately four times as likely as children without disabilities to be hospitalized and, once hospitalized spend eight times as many days in the hospital, the total impact of health care system deficiencies in CSHCN is substantial [11].

The importance of understanding and improving care for CSHCN is widely recognized, but research to date has done little to improve measurably the quality of care that these children receive. A major reason for this deficiency is that CSHCN comprise a heterogeneous group with a multitude of conditions and diagnoses, each of which affects a relatively small group of children [12]. Single-center studies focused on children with a particular diagnosis suffer from small sample sizes, and results are rarely generalizable. Researchers recently moved toward a consequence-based definition for CSHCN in an attempt to capitalize on the similarities among diagnoses and allow for more robust statistical studies with greater clinical relevance [13–20]. Development of operational, valid, and easily administered methods of identifying CSHCN is challenging. To identify these children, several studies have used various survey-based tools, categorical lists that use codes as defined in the International Classification of Diseases

(ICD-9 CM), or methods that use administrative databases to predict children who are high resource users (as measured by cost or length of stay). Each approach has its own limitations; survey tools are generally laborious (with the exception of the CSHCN screener designed and validated in the outpatient setting by the Foundation for Accountability) [21,22], and the categorical lists or predictors of high resource use may be performed only after a patient is discharged from the hospital and the data are abstracted from administrative data.

What is urgently needed is a method to identify prospectively the subset of CSHCN who are medically complex. These children share several similar features that may be understood best by examining an example (Box 1).

Prospective, feasible identification would allow hospitalists to maximize the effectiveness of the hospital stay of these patients. For example, imagine a child with neurologic impairments who is repeatedly hospitalized for acute illnesses or elective surgical procedures who presents to the inpatient service. Patients such as these typically require an enormous amount of resources and services that ideally would be engaged at admission, such as social work to see how the family is coping and to review the current financial situation and need for assistance; pharmacy to review medications, look for interactions, and recheck dosing at home compared with directions written upon admission; physical and occupational therapy to assess how a patient is functioning and how best to maximize function and prevent the patient from developing any secondary complications (eg, decubitus ulcer or another aspiration episode). Typically these health care professionals would assess and treat the patient only once the hospitalist wrote the orders. A prospective identification method could be used to activate services

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Box 1. Prototypic example of a medically complex child: the child with neurologic impairments

Diversity of conditions (eg, brain tumors, intraventricular hemorrhage, traumatic brain injury, tuberous sclerosis, congenital brain anomalies, cerebral palsy with global developmental delay)

Multisystemic disease (eg, respiratory, neurologic, gastrointestinal)

Multiple medications (eg, bronchodilators, anti-sialagogues, anticonvulsants, gastric motility agents)

Multiple specialists (eg, pulmonologist for recurrent aspiration pneumonia, otolaryngologist for salivary gland management, neurologist for seizures, gastroenterologist and surgeon for the management of gastroesophageal reflux disease)

Important subsets (eg, children who depend on technology)

Frequent admissions (eg, for recurrent aspiration pneumonia, seizures, antireflux surgeries)

Critical need for optimal coordination of their care in the inpatient and the outpatient settings

to care for these patients, however, and help the hospitalist coordinate inpatient care and ultimate discharge to the primary care provider.

Several definitions commonly are used in the literature to describe other subgroups of children with chronic medical conditions, such as technology-dependent children and medically fragile children. This terminology history was reviewed by Newacheck et al [5] in 1998. Although a formal definition is lacking, we prefer the term ''medically complex children''(MCC) to capture the principles as outlined previously. Throughout this article we use the term ''medically complex children'' to be concise; however, we are more correctly referring to children with complex medical care needs.

Unique issues of inpatient care

A comprehensive approach by pediatric hospitalists is essential for delivering effective, efficient, coordinated, and family-centered care that best meets the multifaceted needs of these children and their families. Twenty-two percent of recurrent hospital admissions for children with chronic illness are related to medical controversy regarding the most appropriate treatment strategy, and approximately 33% are associated with medical dependency [23]. In general, parents of children with disabilities are less satisfied with medical care when compared with children with other medical conditions [24]. Increased patient and family satisfaction with in-hospital care may promote adherence with post-discharge care plans, minimize readmissions, and ultimately improve the health and well-being of MCC. When approaching MCC, hospitalists are encouraged to treat the ''whole child'' in the context of function rather than diagnosis-specific or organ-based categories [25–27]. In facilitating the coordination of care, pediatric hospitalists can ensure that the focus remains on the child and his or her function at the level of the individual, the family, and the community.

The American Academy of Pediatrics defined the medical home in 1992 [28] and further developed the concepts that focus on CSHCN. The medical home is characterized by care that is accessible, family centered, continuous, comprehensive, coordinated, compassionate, and culturally effective [29,30]. Although these principles apply primarily to ambulatory care, we believe that this model of care is a valuable way to view key elements of the inpatient care of children with complex medical conditions. For some of these children, hospitalists may find themselves as the inpatient medical home physician (especially for children without a primary care physician).

Family-centered care

Parents of MCC with associated complex medical regimens have responsibilities that differ from those of typical parents. They are responsible not only for the physical care of their children but also for dealing with medical, educational, and other service providers while balancing competing family needs

[31]. Parents of children with cerebral palsy have more chronic physical ailments, including back pain, migraine headaches, and stomach/intestinal ulcers, and higher overall distress when compared with other parents [32]. Caregivers of technology-dependent children experience more anxiety, anger, guilt, frustration, sorrow, social isolation, sleep deprivation, and depression when compared with parents of able-bodied children [33]. Thirty percent of all recurrent hospitalizations for children with chronic illnesses are related to a lack of respite services and community support, and 26% are related to psychological or medical issues that affect other family members [23]. There is an association between the health and well-being of MCC and that of their parents. Given the vital role that families play in providing care to MCC, it is critically important that hospitalists acknowledge parents as caregivers and position them centrally on inpatient treatment teams for their children.

In family-centered systems of care, parents are well informed, supported, and afforded ultimate control over decisions regarding the care of their child. Such care is associated with improvements in parents' emotional well-being, satisfaction with services, and the burden experienced [34]. The concept of family-centered care as described by the American Academy of Pediatrics includes physician recognition of key family members and their values and shared decision making between medical providers and families [35]. Pediatric hospitalists can promote family-centered care by regarding parents as the experts in their child's condition and including them in all facets of inpatient care.

Coordinated care

In inpatient settings, care coordination involves managing treatment planning and outcomes monitoring, coordinating input from subspecialists, organizing care to avoid duplication of services, sharing information among health care professionals and family members, managing discharge planning, and family training [36,37]. Hospitalists can lead transdisciplinary team meetings as a forum for care coordination through the establishment of common goals for hospitalization, the development and revision of care plans, and the defining of discharge criteria [38]. Up-to-date care plans that clearly include a problem list, key elements of a child's history, and therapeutic interventions can serve as an efficient and effective means of communication and care coordination [110]. Care plans should be accessible to the family and all members of the transdisciplinary treatment team [39]. Care coordination offers the additional advantage of minimizing hospital-reported medical errors, which occur more often with MCC [9,10].

Discharge planning is a key element of care coordination. The arrangement of home care services for skilled nurses, allied care providers, and durable medical equipment (eg, ventilators, suction machines, and feeding pumps) requires time and effort [40]. Early anticipation of discharge needs smoothes transitions from hospitals to homes for MCC and their families. Before discharge, the inpatient team should ensure the safety and accessibility of a child's home and the avail-

ability of uninterrupted phone, electrical, and transportation services for children who depend on medical technology [39,41]. In cooperation with parents, discharge planners can facilitate the scheduling of numerous recommended follow-up appointments. Hospitalists should communicate a timely summary of the hospitalization and recommendations for follow-up to the primary care physician to further ease the transition of MCC to their medical home. Novel approaches to maximize this transfer of the complexity of information between settings are urgently needed for this population, because the number and types of errors caused by the transitions of care are likely to be dramatic. One novel approach that focused on shared care plans and clinical care specialists in Whatcom County, Washington, was highlighted at the 2004 Institute for Health Improvement's annual forum, and a series of videos is being prepared to be released for television in 2006 [42].

Minimizing secondary complications

Although an acute illness prompts hospitalization for MCC, there are always concomitant comorbid conditions. Hospitalists must remain alert to the potential for exacerbation of these underlying conditions caused by the interdependent nature of organ systems and not focus solely on resolution of the acute illness. A complete medical history should include details of daily schedules, medications, diet, patterns of sleep, typical stool output, functional status, and usual behaviors and activities. This depth of information allows unfamiliar health care providers to identify and address promptly any secondary complications that may prolong hospitalization and increase stress for families. Most MCC require many medications for various standard and "off-label" indications on a long-term basis, which renders them at increased risk for drug interactions and medical errors [9]. Hospitalists should review with parents the dose, route, frequency, and indication of each medication, including homeopathic and over-the-counter medications. For example, valproic acid may be prescribed for aggressive outbursts and gabapentin may be prescribed for neuropathic pain syndromes. The assumption that either of these medications is administered to treat seizures would be an error. During the usual review of food and drug allergies, the hospitalist should explore each child's history of latex reactions, especially when caring for children who have had multiple reactions. A review of the home medication schedule reveals meaningful information, such as administration of phenytoin with meals rather than nonadherence as an explanation for subtherapeutic blood levels.

Nutritional needs

The nutritional needs of children with chronic conditions warrant special consideration during hospitalizations. Upon admission, hospitalists should dis-

cuss with families the details of their child's feeding program including route (eg, oral, tube feedings), content (eg, altered texture, preferred foods), and mealtime schedule. When children are fed at home with adapted cups, spoons, nipples, oral stimulation techniques, or positioning strategies, similar feeding approaches should be providing during the hospitalization. Nutritional assessments and periodic reassessments are indicated during every hospitalization. In addition to plotting heights and weights on standard growth curves, for certain populations (eg, children with Down syndrome) growth parameters can be plotted on specialized growth curves. The intake of calories and fluids should be monitored regularly. Intervention with enteral feedings or hyperalimentation is started when nutritional intake or status is deemed inadequate, particularly for children with dysphagia and a decreased likelihood of regaining weight lost during acute illness.

Functional limitations

Many children with chronic conditions also experience functional limitations and benefit from adaptive equipment, including standers, wheelchairs, and orthotics, during hospitalization to promote independence and mobility. Children should participate in their usual activities as much as possible to minimize complications of immobility (eg, constipation, atelectasis, joint contractures, muscle deconditioning). Hospitalists can consult with physical, occupational, and speech therapists to ensure that the necessary equipment and support are available for each child.

Transdisciplinary coordination

Hospitalization can be stressful and disruptive for MCC and their families. Pediatric hospitalists can collaborate with child life specialists, chaplains, social workers, and other treatment team members to address their emotional needs. Efforts to minimize disruption of schoolwork are encouraged, and schoolwork should continue when possible. Strategies for return to school after discharge should be developed. Educational consultants should guide parents in negotiating the school services best suited to their child's unique needs, including special educational programming through an individualized educational plan. For children unable to return to school in the short term, homebound education can be prescribed for a period of time.

Primary care issues

CSHCN have more unmet medical needs than typically developing children [43]. Because hospitalists and specialists provide most of their care, well-child

care issues may be overlooked. Hospitalists should provide missed immunizations or prophylaxis against influenza, respiratory syncytial virus, or pneumococcal infections as needed. Hospitalization may be a time to consider programs of early intervention, discuss eligibility for social security, or explore community-based services for recreation and socialization.

Application of the concepts

Because the scope of diagnoses involved when considering children who are "medically complex" or "technology-dependent" or have "special health care needs" is broad, we chose children with cerebral palsy as the model of MCC. Children with cerebral palsy present a heterogeneous group of disabilities that range from mild motor impairments to complex developmental and functional limitations. The breadth of experience of children with cerebral palsy and their families is expected to generalize to various developmental disabilities [44].

Consider a child with quadriplegic cerebral palsy. She has global developmental delays, spasticity, gastrointestinal dysmotility, dysphagia with gastrostomy tube dependency, failure to thrive, epilepsy, neuromuscular scoliosis, and functional limitations. Let us assume that she is hospitalized with acute respiratory distress secondary to interstitial viral pneumonia. Because of her tachypnea, feedings are held for 3 days, after which she improves with a regimen of chest physical therapy, postural drainage, and supportive care. She then experiences a generalized tonic clonic seizure, however, which is associated with vomiting and significant aspiration pneumonitis. Was this an avoidable event? On careful review, it is noted that when gastrostomy tube feedings were resumed, formula was delivered continuously rather than in periodic boluses. Although she had stable epilepsy with therapeutic phenytoin levels upon admission, a random serum phenytoin level at the time of her seizure was grossly subtherapeutic. In hindsight, it is evident that her parents provided intermittent gastrostomy tube feedings at home and gave her phenytoin at least 1 hour before or 2 hours after meals to ensure adequate absorption [45,46]. Familiarity with all details of her care should alert the hospitalist to potential food and drug interactions in this child.

After recovering from the seizure and aspiration pneumonitis, the girl with quadriplegic cerebral palsy is clinically improving and discharge is anticipated in 1 to 2 days. She then develops recurrent emesis with feeding intolerance. On physical examination, her abdomen is moderately distended but nontender. Radiographs confirm fecal impaction with obstipation, which is not surprising in a child with gastrointestinal dysmotility exacerbated by immobilization, interrupted enteral feedings, and analgesic medications. Constipation in children with cerebral palsy can present as anorexia, abdominal pain, sleep disturbances, irritability, urinary retention, nausea, and vomiting. Because these nonspecific symptoms could suggest several illnesses, a standardized comprehensive care process for MCC is a primary strategy to the anticipation,

prevention, and early recognition of common complications of hospitalization such as this.

Finally, the patient is ready for discharge. When her parents are dressing her in anticipation of going home, they are upset to find a decubitus overlying her left ischium. Pressures sores can develop quickly in children with compromised nutrition, depleted fat stores, and bed rest [47]. A standardized care process approach would anticipate and reduce the increased risk during hospitalization of this preventable complication.

Technologic devices for medically complex children

Hospitalists should be familiar with enterostomy tubes, tracheostomy, indwelling central venous catheters, noninvasive ventilatory support, and ventricular shunts in terms of indications, complications, and the hospital-based evaluation of these devices.

Enterostomy tubes

Children who depend on feeding technologies have a long-term gastrostomy, gastrojejunostomy (GJ), or jejunostomy tube. Indications for enterostomy tubes include oral motor feeding problems, mechanical esophagopharyngeal occlusion/ stricture/atresia for any reason, inadequate oral caloric intake for growth, altered adsorption or metabolism that requires constant infusion nutrition, unpalatable diets as disease treatment, excess unpalatable medications (such as in HIV), and conditions requiring venting of the stomach for obstruction. GJ or jejunostomy tubes are used for patients with severe gastroesophageal reflux and individuals at high risk for aspiration. Neurologically impaired children comprise the largest category of patients who depend on enterostomy tube feedings, with failure to thrive and risk of aspiration being two primary indications [48].

Tubes are placed surgically with laparotomy or laparoscopy or are placed percutaneously with endoscopic or radiologic guidance. Surgical placement is usually accomplished with another primary surgery, such as fundoplication for gastroesophageal reflux. GJ tubes simply add guided placement of a tube into the jejunum.

Complications of gastrostomy are divided into "early or late," and "major or minor." Early major complications are more common with surgical placement (19.9%) than percutaneous endoscopic gastrostomy (9.4%) or percutaneous radiologic gastrostomy (5.9%), which offers the least risk [49]. Minor and late complications are independent of the approach. Overall, complications that lead to gastrostomy tube revisions occur in 6% of children [50]. Tube design and location can influence risk of late complications; GJ tubes have higher risk [51,52]. Major complications include procedure-related aspiration pneumonia [53], dislodgement before tract maturation, gastrointestinal bleeding, peritonitis,

severe wound/abdominal wall infection, intussusception, gastrocolocutaneous fistula, sepsis, and death. Major complications are heralded by typical signs and symptoms that may be more difficult to recognize in the neurologically impaired enterostomy population. Pneumoperitoneum is common with tube placement, may be a complication in very small infants [54], and can obscure the diagnosis of some major early complications in all children. Worsening of gastroesophageal reflux is reported in some studies, particularly in neurologically impaired children, but not all studies support this finding [55]. Careful consideration of the need for fundoplication surgery should precede enterostomy.

Minor complications include tube dislodgement after tract maturation, tube blockage, migration, leakage, gastrostomy tube site infection, granulation tissue formation and "buried bumper syndrome." Recognition of minor complications can be straightforward for tube dislodgement and blockage, whereas other presentations may be obscure. Vomiting with a gastrostomy tube may indicate migration with blockage of the duodenum by the balloon or malposition; with GJ tubes, it may indicate leakage in the gastric coil, back-migration or malposition into the stomach, and intussusception. Diarrhea "like formula" or aspiration of fecal material may indicate a gastrocolocutaneous fistula. Redness at the site may be caused by tape sensitivity, leakage, granulation tissue, or infection. Tube leakage should be distinguished from gastric contents leakage around the tube. Excess mobility of the tube may enlarge the tract and lead to gastric leakage, which can be difficult to treat. For gastric contents leakage, air drying, barrier agents, and sucralfate powder may help. Using an acid-reducing agent, placing a temporary GJ tube, ordering no oral intake for the child with gastrostomy tube suction, and removing the tube for hours or days to allow the tract to heal and shrink may be necessary. Granulation tissue tends to be friable and bleed easily. If present, it can be treated with warm saline compresses and may need cautery with silver nitrate. Buried bumper syndrome occurs when excess traction leads to the internal bumper eroding through the stomach wall, with re-epithelialization covering or burying the bumper. Symptoms may include abdominal pain with feedings, resistance to flow through the tube, and inability to rotate the tube. Treatment requires tube removal and replacement.

Approximately 20% of patients experience infection; it is usually local but may progress to cellulitis and, rarely, necrotizing fasciitis [49]. Local infection can be treated with cleaning and use of local antibacterial agents and oral antibiotics. Cellulitis requires systemic antibiotics, and necrotizing fasciitis is a surgical/infectious emergency.

Tube blockage is best treated by prevention. Treatment of blockage includes using water, pancreatic enzymes, and carbonated drinks. Tube dislodgement before 4 weeks is a major complication because tract maturation occurs between 4 weeks and 3 months. It likely requires a repeat procedure, with peritonitis being a potential complication caused by separation of the stomach wall from the inner abdominal wall. A mature stoma still may close within hours to days if the tube is dislodged. Parents may be taught to insert gently a temporary Foley catheter, with the balloon deflated, until its location can be determined radio-

logically if it occurs within 8 weeks of placement. After that, a correct sized Foley catheter or a spare gastrostomy tube can be reinserted gently followed by balloon inflation. If gastric contents are aspirated and air injection verifies the correct position, feedings can be reinstituted until proper replacement. Many gastrostomy tubes are currently replaced by a low-profile "button" within 2 to 4 months. Removal and replacement of a button requires special expertise and equipment that may necessitate consultation with the service that placed the gastrostomy tube. A dislodged button still necessitates placement of a Foley catheter to maintain patency of the ostomy until the button can be replaced.

Jejunal tubes have unique potential complications. Because of the size of the small bowel, a smaller internal bumper is needed, which leads to difficulties anchoring the tube and incurs higher risk of dislodgement. Gastrointestinal complaints are also higher, including abdominal distention, pain, tenderness, and diarrhea. Small intestinal ischemia and necrosis can develop as a consequence of direct small bowel feeding, particularly in hemodynamically unstable patients. An ostomy care nurse can be invaluable in troubleshooting enterostomy tube problems for the pediatric hospitalist who cares for such patients.

Tracheostomy

Tracheostomy tubes are indicated in cases of upper airway obstruction, for children who cannot protect their airway, and for patients with long-term mechanical ventilation. Children usually remain hospitalized until the first tube change, which allows some maturation of the stoma. Optimal tracheostomy care starts with the proper tube of appropriate size and shape to fit the airway without exerting pressure on the tracheal mucosa. It also must fit well enough to prevent aspiration (if that is its primary purpose) or loosely enough to allow translaryngeal air escape for vocalization and mucous clearance. It must have an adequate inside diameter to prevent airflow restriction. This should be tailored to each child's specific circumstances.

Standards of care in the hospital should include 24-hour 1:1 care from a provider trained in acute troubleshooting of tracheostomy complications. Frequent respiratory therapist care and input, attention to clearing of secretions, humidification, and ready availability of a replacement tracheostomy tube that matches the patient's tube—and one size smaller—are also essential. Most pediatric tracheostomy tubes are not cuffed; if a cuffed tube is used, that fact should be apparent to all caregivers who might provide emergency management. One should notify the otolaryngology or general surgery services of the patient's admission and the potential for emergency airway management needs.

Complications include accidental decannulation, creating of a false passage, obstruction, infection, hemorrhage, pneumothorax, pneumomediastinum, peritracheal cellulitis, and lower airway infection. Although accidental decannulation and obstruction are the most common overall complications, they are rarely encountered in the inpatient setting. Because of their acuity, they are rapidly addressed by bedside caregivers and rarely impact the hospitalist directly. Pe-

ritracheal infection usually can be treated with oral antibiotics and local wound care, but it can lead to mediastinitis if not addressed. Lower respiratory tract infections are common. Children with tracheostomies are colonized with multiple pathogens, including *Staphylococcus aureus*, *Pseudomonas aeruginosa*, and *Candida albicans*. Empiric antibiotic therapy should reflect this. A rare (1%–2%), life-threatening complication is erosion into the innominate artery, which leads to massive hemorrhage. Extensive hemorrhage in the postoperative period or significant hemorrhage after the first 48 hours should prompt evaluation for this critical complication.

Perhaps the single most important preparation for safe home tracheostomy management is good training of at least two adult caregivers. Although the mortality rate of children with tracheostomy is 11% to 40%, death from a tracheostomy complication is rare. In January 2000, the Pediatric Assembly of the American Thoracic Society published a consensus statement to serve as a standard of pediatric tracheostomy care that provides detailed guidelines and the evidence supporting it [56].

Long-term intravenous access catheters

Many patients currently require extended or long-term intermittent intravenous medication, frequent blood draws, or parenteral hyperalimentation. There are three general categories of central venous catheters (CVCs): (1) external partially implanted (Broviac, Hickman, Groshong), (2) totally implanted with a subcutaneous port (Portacath, Mediport, Infusaport), and (3) percutaneously inserted (PICC line). Whereas the first two CVCs are inserted for long-term access, with reported durations of 12 to 32 months [57,58], PICC lines are used for briefer periods, with reported average durations varying from 16.6 to 72.7 days [59,60]. Complications associated with CVCs include thrombosis, infection, malfunction (discussed later in detail), fractures or breaks, dislodgement, noninfectious phlebitis, and air embolism. Cardiac dysrhythmia, mediastinitis, cardiac tamponade, hemorrhage, and pneumothorax can occur more commonly as acute complications of line placement and must be considered in the first hours after placement. Factors associated with removal of a line are primarily completion of therapy but also include infection (eg, gram-negative bacilli and yeast), migration, use for obtaining blood samples rather than infusion alone, catheter material (polyurethane $>$ silicone), and mechanical failure.

Thrombosis occurs with all CVCs. Symptomatic vascular thrombosis is less common, with reported rates of 4.6% to 9%, but is still the primary cause of thrombosis in children [61]. Thrombosis may involve only the catheter (obstruction) or the insertion vessel (subclavian vein) with ipsilateral limb venous outflow obstruction and associated symptoms, or it may extend centrally into the superior vena cava and lead to clinical signs and symptoms of superior vena cava syndrome. Pulmonary embolus is an underdiagnosed complication [62]. Asymptomatic pulmonary embolus was diagnosed indirectly by ECG or

echocardiographic criteria in 12 of 21 (57%) children with a long-term CVC in one study [63]. The clinical relevance of asymptomatic pulmonary embolus remains unstudied, however.

Infections may involve the skin at the exit site, the subcutaneous tunnel, or systemic line sepsis. Reported rates of infection vary from 4% to 60%, with rates of line sepsis ranging from 4% to 9%. Factors associated with a higher risk of infection include frequency of accessing the line, the first month after placement or more than 24 months of use, and thrombosis/fibrin sheath formation [64–66]. The most common organisms are skin flora, but in immunocompromised children, gram-negative bacilli and yeast are recovered. Treatment initially involves empiric antibiotics and then proceeds to appropriate antibiotics based on culture results. Most gram-positive bacteremias/sepsis can be treated successfully without CVC removal. If bacteremia recurs after treatment, the CVC can be sterilized successfully with an "antibiotic lock," usually with vancomycin and a thrombolytic agent, such as urokinase. A prospective, randomized, double-blind trial compared prophylactic "antibiotic lock" with vancomycin and heparin for 1 hour every 2 days to heparin lock alone. The hub colonization and bacteremia rates were 15.8% and 7% with heparin, 0% for both with treatment [67]. Gram-negative line sepsis and yeast line sepsis generally require line removal as part of treatment, but there is recent evidence of successful line salvage with antibiotics and a thrombolytic agent together [65,68,69]. A Cochrane review of dressings used to cover CVC exit site and infection risk found no risk difference among dressings and recommended that dressings reflect patient preference [70]. A 1-year prospective study that monitored the difference in infection rate with or without an inline filter found no difference [71]. Little information exists to document the infectious risks from PICC lines, which are low overall because of shorter duration of use, although a sepsis rate of 25.7% (7/25 catheters) was reported in one study of patients with solid tumors [72]. Minor exit site infections, including yeast infections, have been reported.

Malfunction of CVCs is common and presents as total occlusion, lines that infuse but do not withdraw blood, and lines that are intermittently nonfunctional. Malfunction can be evaluated by Trendelenburg positioning, raising the ipsilateral arm over the head, or hydrating the patient. If these approaches result in a normally functioning line, one should obtain a radiograph to check tip location. Inability to withdraw blood may be caused by a one-way valve-like thrombus on the tip. Complete occlusion is usually the result of thrombus. Treatment with thrombolytic agents, such as urokinase, 2500 to 5000 U for 30 minutes, or tissue plasminogen activator (tPA), 0.5 to 1 mL, left to rest in the line for 2 hours before attempting aspiration can restore function [73]. A study that evaluated the efficacy of tPA in clearing CVCs showed 86% success with a first dose and 95% with two attempts [74]. Other line-associated complications are less frequent and include air embolism at the time of placement, which was shown to be less frequent in well-sedated patients, and complications associated with general anesthesia [75], which also occur rarely with home infusion mistakes [76]. Catheter breaks and leaks occur; each manufacturer has catheter-specific repair

kits that can be used to repair them. Accidental dislodgement usually requires replacement of the catheter; a radiograph should be obtained to check for migration of the tip. PICC lines sometimes can be replaced over a wire. Complications with PICC lines are reported to range from 0 to 40.7%, with occlusion and infection being the most frequently reported. Complications more common with PICC lines include external breaks, shoulder pain, phlebitis without infection, exit site irritation, dislodgement, and occlusion. A study that compared PICC line function with non–central tip location versus central tip location showed comparable results and complication rates, with the exception that non-central PICC lines failed sooner (11.4 days versus 16.6 days), and fewer patients completed their course of therapy (69% versus 73%) [60].

Noninvasive positive pressure ventilatory assistance

Noninvasive positive airway pressure (NiPAP) assisted ventilation can be continuous (CPAP), bi-level, or automatically adjusted (auto-CPAP). NiPAP is indicated in obstructive sleep apnea, chronic neuromuscular disease, apnea of prematurity, syndromes with facial/pharyngeal malformation and dysfunction, acute chest syndrome in sickle cell disease [77], poorly controlled epilepsy with concurrent obstructive sleep apnea, bilateral diaphragm paralysis after surgery, and some instances of chronic and acute respiratory disease. Tracheostomy-delivered CPAP has been used to reduce the incidence of aspiration of saliva with its consequences in neurologically impaired children [78]. Bi-level positive airway pressure delivers two levels of positive pressure that vary by inspiration/expiration to improve comfort and tolerance.

NiPAP may be delivered by full face mask (covers mouth and nose), nasal route (nasal CPAP prongs, nasal mask), or tracheostomy. Automated CPAP is new to pediatrics and has been tested only in a monitored setting, but is likely to see more widespread use [79]. It essentially automatically titrates CPAP to an optimal pressure and adjusts for changes in sleep variables that affect frequency of apneas and hypopneas. Algorithms specific to pediatrics for recognizing apneas and hypopneas are yet to be validated. In all forms of NiPAP, intolerance of the intervention is the most common cause of failure. Mask fit is essential. When a patient on NiPAP is hospitalized, the successful continuation of their respiratory intervention is optimized with use of their home mask and delivery device. A process in place in the hospital to expedite this is useful. In some hospitals, the use of NiPAP ventilation assist necessitates admission to the pediatric intensive care unit. In many patients with chronic NiPAP ventilation, admission to the floor is allowed, but if home equipment is used, a caregiver (parent) trained on that equipment is asked to be available around the clock.

NiPAP ventilatory assistance has been shown to reduce hospital days by up to 85% in children with neuromuscular disease, with documented improvements in apnea/hypopnea index and transcutaneous PCO_2 level [80]. Three neurologically impaired children with tracheostomies and documented chronic saliva aspiration

saw reduction in frequency of aspiration by radionuclide salivagram and only "rare hospitalization" after tracheostomy CPAP [78]. In children with obstructive sleep apnea who are successfully started on long-term NiPAP, approximately 96% can be expected to have measurable improvements [81]. There are no published guidelines for amount of pressure support by age or disease. Published studies do not show a consistent relation between degree of CPAP and clinical parameters. It is reasonable to institute therapy at 4 cm H_2O in infants and 6 cm H_2O in older children and titrate to response while monitoring for complications. In most patients cared for on the pediatric medical floor, NiPAP is at a pre-established level and no changes would be anticipated in the hospital. In acute illness, there may be a need for more support; generally this necessity should prompt a transfer to the pediatric intensive care unit. In children chronic disease, the measures of response to therapy include days hospitalized, frequency of pneumonia, measures of ventilatory function, daytime sleepiness scales, and measures of school performance, and the goal is the minimal long-term NiPAP that will maintain the clinical response. Titration of NiPAP is not indicated during an acute hospital stay.

Complications of NiPAP ventilation assist include pneumothorax, skin irritation and pressure injury, nasal drying, nasal pain, drying of the throat, claustrophobia, and nasal deformities in preterm infants. A high index of suspicion for pneumothorax, particularly with acute respiratory illness, attention to potential complications when hospital policy dictates use of standardized equipment while hospitalized, extra care by ancillary services in mask or nasal prong fitting, a protocol for early identification of pressure-induced skin injury with appropriate treatment, use of humidification, and acclimation to NiPAP use likely will reduce the risk of complications.

Cerebrospinal fluid shunts

Shunts are placed to treat increased intracranial pressure. Shunts are named based on their proximal and distal insertion locations. Most originate in the lateral ventricles; other locations include the third or fourth ventricles or an intracranial or spinal cyst. Most shunts terminate in the peritoneal cavity; other locations include the right atrium, pleural space, gall bladder, ureter, urinary bladder, bone marrow, mastoid, thoracic duct, and fallopian tube. Each shunt has three basic parts: (1) the proximal catheter, which exits the central nervous system through a bur hole, (2) a one-way valve that allows unidirectional flow when a specific pressure differential is exceeded, and (3) the distal catheter, which is tunneled under the skin to its destination, sometimes with extra length to allow for growth. Modern shunts also may incorporate reservoirs for cerebrospinal fluid sampling and drug instillation, antisiphon devices to prevent excessive run-off, and on-off valves. They also may be externally programmable. Shunts are placed by neurosurgical procedure, and the neurosurgical service should be consulted whenever a complication is suspected.

Complications include malfunction that leads to inadequate drainage and increased intracranial pressure, infection that may lead secondarily to malfunction, and overdrainage that may lead to the ventricle syndrome. Headache is a common long-term clinical challenge.

Malfunction eventually complicates 30% to 40% of all shunt procedures. Nearly half of all shunt revision procedures are for malfunction, and up to 71% of all patients experience a malfunction during their lifetime [73,82,83]. Symptoms of shunt malfunction are those of increased intracranial pressure, including headache, nausea, and vomiting, irritability, increased seizures, neck pain, back pain, blurred vision, lethargy, and "just not acting right" [84]. Fever is also seen in 22% of uninfected malfunctions [85] but actually may signal infected hardware despite negative cultures [86,87]. Signs include a bulging fontanelle, separating sutures, increased occipital-frontal circumference, papilledema, sun-setting eyes, redness and tenderness along the shunt tubing route, and altered mental status, although these signs may be subtle [88]. Sterile shunt malfunctions may be immunologically mediated and associated with cerebrospinal fluid eosinophilia [89–93]. Eighty-five percent of results of head CT scans are abnormal in shunt malfunction when compared with a baseline CT; more sensitive methods of comparison are being developed [94]. Approximately 15% of cases of malfunction are caused by discontinuity of shunt tubing or kinks and can be excluded by obtaining a plain radiographic "shunt series" [95]. A radionucleotide shunt function study can demonstrate patency of the proximal and distal tubing, but it involves accessing the shunt reservoir. Other evaluations include manipulation of the shunt reservoirs/valve and tapping the shunt to confirm or exclude obstruction of the proximal or distal catheters. These maneuvers are best performed by the consulting neurosurgeon. One should be aware that some patients have more than one shunt, which may or may not be connected. Symptoms can be vague and nonspecific. In one study, the average time between onset of symptoms and diagnosis of shunt malfunction was 11.5 days [73]. In another study, vomiting, lack of fever, and parental suspicion were the most sensitive clinical features. Parents have been shown to be as accurate as physicians in diagnosing shunt malfunction before diagnostic testing [73].

Infection complicates between 2% and 30% of shunt procedures, although the incidence has been declining over time [73,96]. Risk may be higher in infancy and in the recent postoperative period after a shunt procedure. Half of shunt infections occur in the first 2 weeks, 70% to 80% within 2 months, and 80% to 90% within 4 months [73,97]. Lumbar puncture, ventricular taps, cerebrospinal fluid withdrawal, and ventriculograms are not independently associated with an increased risk of shunt infection. Common pathogens in postoperative infection up to 6 to 9 months include coagulase-negative staphylococcal species, *Staphylococcus epidermidis,* and *S. aureus*. Gram-negative bacilli are isolated in 6% to 20% [97], and other pathogens commonly associated with meningitis have been isolated in late shunt infections (>6–9 months), such as pneumococcus and *Haemophilus influenzae* [98]. Symptoms can be vague and nonspecific but often include fever and may include irritability, feeding problems, nausea, vomiting,

lethargy, headache, and change in sensorium. Symptoms of meningeal irritation are often absent. Signs include erythema, swelling, cellulitis, or wound infection overlying the shunt tract, fever, altered mental status, and—with a ventriculo-peritoneal shunt—abdominal pain, diarrhea, and clinical signs of peritonitis. Approximately 50% of infected shunts also malfunction. Evaluation includes a head CT scan and shunt series. Tapping the shunt to obtain a cerebrospinal fluid sample for laboratory evaluation and culture is essential, although there is controversy about the number of white blood cells and protein level that are "acceptable," particularly close to the time of a shunt procedure [89,99]. A Gram stain can be particularly helpful if the results are positive. Multiple studies have shown that use of systemic antibiotics and removal of the infected shunt hardware have a high probability of resolving the infection [100]; however, the ideal length of treatment is unknown [101]. Empiric therapy should begin with vancomycin and a third-generation cephalosporin [102]. Ventriculoatrial shunts may be associated with systemic sepsis symptoms when infected, and complications can include endocarditis, glomerulonephritis, hypocomplementemia, and thromboembolic events.

Rare complications of ventriculoperitoneal shunts include development of an inguinal hernia (up to 16.8%), migration of the distal tip, and perforation of a wide variety of intra-abdominal organs, the incisions, and the diaphragm. An abdominal pseudocyst also can develop with symptoms that can be a diagnostic dilemma. Other rare complications attributed to shunts include intussusception, intractable hiccup, omental cyst torsion, and volvulus around the catheter.

Slit ventricles on head CT occur within 6.5 months in 21% of patients and 48% after 6 years, but symptomatic slit ventricle syndrome is uncommon [103]. One successful approach to treatment has included lumboperitoneal shunting [104].

Future research agenda

Research on MCC as a subgroup of CSHCN is critical to study and improve the quality of care they currently receive. Challenges to studying this population include the enormous diversity of conditions and absolute small number of patients at a single hospital. One study that examined the impact of a pediatric hospitalist system for MCC (as defined by a positive response to the CSHCN screener, transfers from an intensive care unit or an All Payer Refined Diagnosis-Related Group of three or four [those of the highest severity of illness]) found that those children had proportionally lower length of stay and total costs, compared with the already lower length of stay and total costs of the cohort of all children who were cared for by hospitalists, as compared with the traditional academic attending model or community physicians [105]. Future research must determine, however, how to measure effectively the quality of care MCC receive (in terms of processes and outcomes of care). It is important to determine

how a new inpatient system of care may best integrate the care of these children across the continuum of inpatient and outpatient care and across several disciplines and specialties.

For these children to receive the highest quality of care, three critical steps in research must occur to improve the current system of inpatient care. First, prospective methods of identifying MCC that are feasible, valid, and reproducible must be developed and allow for comparative studies across institutions. Recent novel studies are currently being undertaken to accomplish this goal [106,107,111].

Second, measures of inpatient processes of pediatric care must be defined, validated, and tested explicitly (disease-dependent and disease-independent). Process measures have been particularly challenging in inpatient pediatrics because few diseases have well-defined quality measures with strong evidence to link particular processes of care with improved outcomes. In particular, disease-dependent measures require the evidence base to be developed, tested, and refined and are largely lacking in pediatrics (with a notable exception of inpatient asthma care). The challenge is particularly great in assessing the quality of care of MCC who have few well-studied process measures. For example, children who are neurologically impaired have various conditions that may be responsible for their hospitalization, including seizures, aspiration pneumonia, and fever, but research in their care is sparse, at best. Disease-independent process measures are one method of addressing quality of care in the face of such a large range of infrequent conditions. MCC are an ideal group to study given their vulnerability that leads to repeated hospitalizations and allows researchers to understand better the aspects of their care. For example, when using parental expectations with care as an outcome measure, examining how parents of MCC rate their inpatient care compared with parents of children who are not medically complex allows for a more in-depth analysis of the system of inpatient care in the eyes of a frequent user, exemplifying aspects of poor- and high-quality care.

Third, pediatric hospitalists must be studied in their natural laboratory (ie, the hospital) to understand the mechanisms for the successful and unsuccessful models of translating research findings into clinical practice using quality improvement techniques. Various aspects of inpatient care can be studied, tested, and retested after changes in serial interventions using quasi-experimental study designs. For example, at Primary Children's Medical Center in Salt Lake City, we have undertaken a quality improvement study that evaluates the optimum method of improving written medication orders for newly admitted MCC. Pharmacists have been reviewing the initial orders, comparing them to home medication lists provided by parents, and contacting the outpatient pharmacies, the primary care physicians, and the hospital pharmacy database using the last hospital discharge as a comparison point. These medication reconciliations of new admissions from the emergency department and transfers from within the hospital provide insight into the system of care that allows for such errors to occur [108]. Hospitalists are often inpatient unit medical directors who work closely with hospital administration and are poised to see the implementation of and ongoing impact on

outcomes of quality improvement and research findings translated into clinical inpatient practice.

As these three things occur, the care being delivered to hospitalized MCC will undergo a paradigm shift, because that care will be able to be measured, benchmarked against best performers, and improved on to deliver care that is effective, efficient, safe, patient centered, and timely [109].

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