Cystic Fibrosis and the Respiratory Therapist: A 50-Year Perspective

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The role of the respiratory therapist in the care of patients with cystic fibrosis has expanded throughout the years. As key members of the multidisciplinary team, respiratory therapists actively participate in the medical management of patients with cystic fibrosis along the continuum of care, from acute in-patient stays to the out-patient clinic and/or home setting. Through their involvement in diagnostic testing, administering therapy, or direct bedside care, patient and caregiver education, and disease management, respiratory therapists strive to preserve lung function, maintain overall health, and improve the patient's quality of life. *Key words: cystic fibrosis, CF, respiratory therapist, history.* [Respir Care 2009;54(5):587–593. © 2009 Daedalus Enterprises]

Introduction

Cystic fibrosis (CF) is an autosomal recessive genetic disorder that affects the functioning of nearly all of the body's exocrine glands. The pulmonary manifestations are characterized by repeated endobronchial infections, an exaggerated inflammatory response, airways obstruction, and bronchiectasis.[1] CF was first described as a distinct entity in the late 1930s independently and nearly simultaneously by Guido Fanconi and Dorothy Anderson. In 1936, Fanconi, a Swiss pediatrician, described the connection between bronchiectasis, celiac disease, and pancreatic changes associated with CF.[2] Shortly thereafter, Anderson, a pathologist, used the term "cystic fibrosis of the pancreas" to describe the common postmortem pathology finding of mucus plugging of the glandular ducts in the 38 malnourished patients she studied with suspected celiac disease.[3] During that period, diagnostic tests were not readily available. Rather, diagnosis was primarily based on the patient's clinical presentation, such as pulmonary infections, growth failure, and steatorrhea. Prognosis was poor and most patients succumbed to the disease at less than 1 year of age.

Since CF was first described more than 70 years ago, the median survival age of patients with CF has markedly increased. Advancements in the diagnosis and treatment of CF improved patient outcomes (Fig. 1).[4] Newborn screening and sweat and genetic testing facilitated early diagnosis and medical intervention. The development of various drugs and other therapies made it possible to treat CF pulmonary infections, alleviate malabsorption, and relieve airways obstruction. Epidemiologic studies tracked and reported patterns of care and patient outcomes. In a recent report in *Pediatrics*, the investigators and coordinators of

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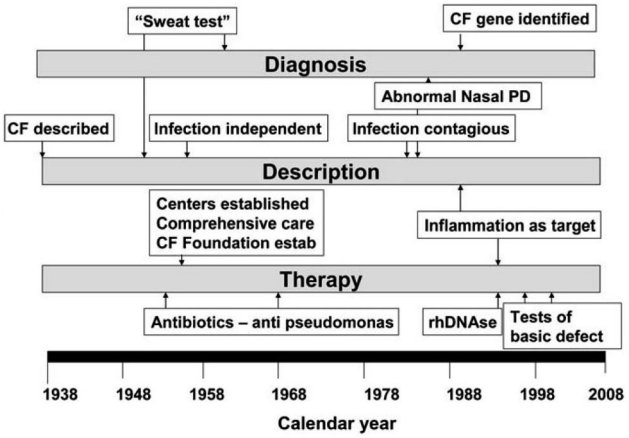

Fig. 1. Chronology of discoveries in cystic fibrosis (CF) diagnosis, clinical description, and therapy. PD = potential difference. rhDNAse = recombinant human deoxyribonoclease (dornase alfa). (From Reference 4, with permission.)

the epidemiologic study of CF identified a positive association between patient outcomes and early diagnosis by prenatal or newborn screening, prompt medical treatment, and proactive approaches to care, such as frequent clinic visits, more sputum cultures, and aggressive antibiotic therapy, which improved nutrition and preserved lung function in children 6–12 years old.[5] CF is no longer exclusively thought of as a childhood disease. In the United States, 45% of the approximately 30,000 individuals diagnosed with CF are more than 18 years of age. Although the currently reported median age of survival is 37.4 years, many patients with CF live well into their fifth decade of life.[6,7]

As treatments became available and longevity improved, the need for skilled respiratory therapists (RTs) emerged. In the mid-1950s the development of comprehensive centers that specialized in CF care helped physicians become familiar with the clinical manifestations of the disease and gain experience with treatment. Research led to a better understanding of the disease process and the mechanisms of lung-function decline. The centers used a multidisciplinary team approach to patient care, which continues today. Collaboration between researchers and clinicians provided the framework for the establishment of best practices, the development of practice guidelines, and the implementation of evidence-based treatment protocols. From a purely pulmonary perspective, this practice change facilitated a paradigm shift in the management of patients with CF. Rather than a focus on the treatment of ongoing pulmonary disease, the new strategy was a proactive approach of early intervention, maintenance of health, and preservation of lung function. This new focus brought opportunities for role expansion for RTs, which parallels that for other chronic respiratory conditions such as asthma and chronic obstructive pulmonary disease. I will discuss the role of the RT in CF care and how that role has spanned the breadth of CF care.

Administration of Cystic Fibrosis Therapies

The comprehensive CF care centers adopted a multidisciplinary team approach and holistic management of this complex disease. Clinicians administered the essential therapies or "pillars of treatment," which included nutritional support, relief of airways obstruction, treatment of recurrent pulmonary infections, and suppression of inflammation.[4] Although many new medications and therapies are now used to treat CF, the "pillars of treatment" principle remains unchanged, and RTs are intricately involved in the treatment of the pulmonary manifestations of this disease. Bland aerosol and supplemental oxygen were mainstays during the initial development of CF care. Bland aerosol was delivered by large-volume nebulizers and "mist tents," to improve bronchial hygiene by liquefying tenacious secretions and alleviating airways obstruction.[8] A low to moderate concentration of oxygen (30−50%) was administered for hypoxemia in the mist tents,[9] but research found little benefit from mist-tent therapy, and some risks such as bacterial contamination, nosocomial infection, and increased airway resistance.[10] Today a low-to-moderate oxygen concentration is delivered by nasal cannula or reservoir mask.

A better understanding of the effect of the gene defect on the airway epithelium and the nature and composition of CF mucus led to the development of new CF drugs. Clinical trials complemented basic and laboratory research and offered practitioners an opportunity for professional role expansion. In addition to direct care responsibilities, RTs routinely performed pulmonary function testing and specimen collection for in-patient and out-patient clinic visits. Their skills made them a valuable asset to the research team. The RT's role naturally expanded to include collection of sputum specimens, measurement of vital signs and other physiologic variables, and the performance of pulmonary function tests or other diagnostic tests. Practice changes resulted from this research. Various inhalable agents, including β agonists, hypertonic saline, and mucoactive drugs, were developed and used to increase hydration of the airway surface and thereby improve clearance of tenacious secretions and cellular debris and decrease airway obstruction. The evolution of CF pharmacology also included replacement of some medications, such as acetylcysteine with dornase alfa, whose efficacy was supported by the literature.[11]

Initially there were few options for secretion clearance. Postural drainage and clapping/vibration became the accepted standard treatments for mobilizing secretions.[12] Technological and clinical advances provided new airway-clearance techniques, including various breathing tech-

niques and devices. RTs were uniquely positioned as airway-care experts to participate in the review of the evidence on secretion-clearance practices and establish best practices. However, systematic reviews of the airway-clearance research pointed out various methodological limitations, including small sample sizes, lack of reproducibility, sparse use of sham therapy, and measurement of only short-term outcomes (eg, a single treatment session), so the evidence could not support the use of any particular airway-clearance devices and/or techniques.[13-15]

Evaluation of Airway-Clearance Devices and Techniques

Choosing the best airway-clearance device or technique for a patient requires understanding the various clinical and technical factors such as the patient's cognitive ability, CF's effect on mucociliary transport, the severity of pulmonary impairment,[16] how the airway-clearance device/technique works, and its limitations, hazards, and precautions.[17-20] Vague instructions from the manufacturer and inadequately detailed device specifications may hinder a clinician's ability to develop a successful airway-clearance care plan.[21] RTs must rely on their investigative skills to conduct laboratory studies that will determine the performance characteristics of the device or technique of interest. Unlike a clinical trial, product evaluations may not necessitate a large investment of financial or human resources. However, investigators must maintain scientific rigor when conducting this type of research. One consideration is to carefully select the data-acquisition tools used in the experiment.

Studying an oscillatory positive-expiratory-pressure (OPEP) device, for example, requires software that is sensitive enough to detect pressure and air-flow oscillation changes as flow and expiratory resistance through the device varies.[22] In addition to providing clinicians with the information needed to better understand device performance, the knowledge gained can be applied clinically to best match the device to the therapeutic goal. This can be exemplified by 2 recently published reports on OPEP devices. Alves et al studied the performance of the Flutter VRP1 OPEP device.[21] Volsko et al[23] compared the performance the Flutter VRP1 and the Acapella OPEP device. Both those studies sought to determine how flow affected the expiratory pressure, pressure amplitude, and oscillation frequency. Both study designs were based on clinical knowledge of the research population.

Volsko et al used pulmonary function data from healthy subjects and CF patients to decide what flow range to use in the laboratory investigation. The flow range represented various degrees of pulmonary impairment, from no detectable decrement in lung function to severe pulmonary impairment. The flow range chosen was validated by measuring expiratory flows during simulated OPEP maneuvers with one healthy subject and 3 patients with CF (with mild, moderate, and severe airflow obstruction).[23]

Alves et al used that same theoretical premise to choose their experimental flow range. Both studies found differences in the oscillation frequency at different flows, device inclinations, and device settings. The researchers also commented on the stability of the pressure waveforms under the various test conditions, which affect the OPEP device's ability to consistently generate the short bursts of flow required for enhanced secretion transport. That knowledge may be useful when selecting an OPEP device for a specific patient, or to develop an airway-clearance protocol. Device evaluations performed by RTs also encourage other RTs to participate in this expanded role in device evaluation.

Development of Airway-Clearance Protocols

The lack of empirical evidence of superiority of any airway-clearance device or technique supports the need for protocols to guide device/technique selection. Volsko et al used the information from their study of OPEP devices to modify the airway-clearance protocol (Figure 2) incorporated in their CF center's in-patient care path for the treatment of exacerbations.[24] The original protocol used only one OPEP device, but the amended protocol included both the Flutter and the Acapella and an algorithm for OPEP device selection. Data collection and analysis of outcome variables were essential and followed each protocol change. Subsequent protocol revisions were based on their unpublished data. The amended protocol limited the use of OPEP to patients with moderate-to-severe dyspnea or documentation by the CF care team that OPEP was previously used and determined to be ineffective for the patient.

This is but one example of how RTs have been instrumental in the development and implementation of protocol-based care. RT-driven protocols improve appropriate allocation of respiratory services by reducing over-ordering and under-ordering of respiratory therapies.[25-28] Many studies have addressed the benefits of airway-clearance protocols.[29-31] Appropriate matching of therapy to clinical need improves patient adherence to therapy and thus reduces missed therapies.[32,33] Adherence is maximized when patient preference is weighted in the device selection. User-friendly tools such as documentation sheets (Figure 3) may enhance the RT's ability to follow the protocol and facilitate the collection of outcomes data. The airway-clearance algorithm is embedded in the respiratory therapy documentation sheet. Data regarding appropriate use of and adherence to the device-selection guidelines can also be tracked on one form.

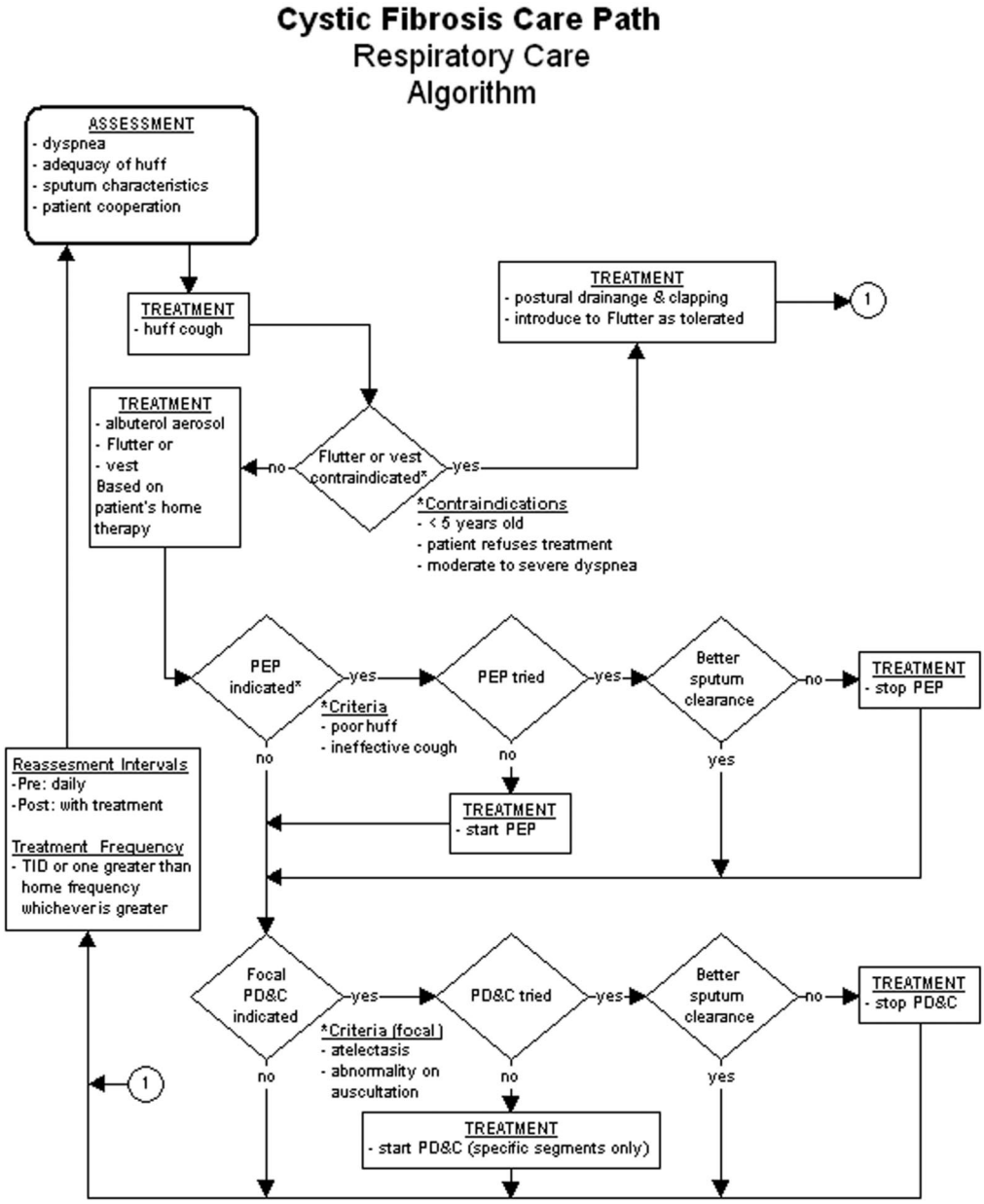

Fig. 2. Airway-clearance algorithm for selecting airway-clearance therapies for patients with cystic fibrosis hospitalized for cystic fibrosis exacerbation. PEP = positive-expiratory-pressure therapy. TID = 3 times a day. PD&C = postural drainage and percussion.

Education of Patients and Families

A CF treatment regimen can be somewhat complicated and time-consuming. Many patients with CF are prescribed a series of aerosol therapies in addition to airway-clearance therapy, and the RT's role in CF care also includes ongoing patient and caregiver education on CF treatments (Table 1).[34] This education should cover how to perform the therapy, how to clean the equipment, and how to integrate the therapy sessions into the patient's daily schedule.[35,36] The patient and caregiver training sessions may include sequencing of inhaled medications. Currently there is lack of evidence of superiority of any particular sequencing strategy. Downs et al combined the principles of self-management with detailed training on how and when to use airway-clearance and aerosol medications, and children 6–12 years old felt more positively about and knew more about their treatment regimens immediately following the intervention and at a 12-month follow-up.[37]

Early recognition of pulmonary exacerbation and prompt treatment of pulmonary infection are important components of the education, aimed at reducing the rate of de-

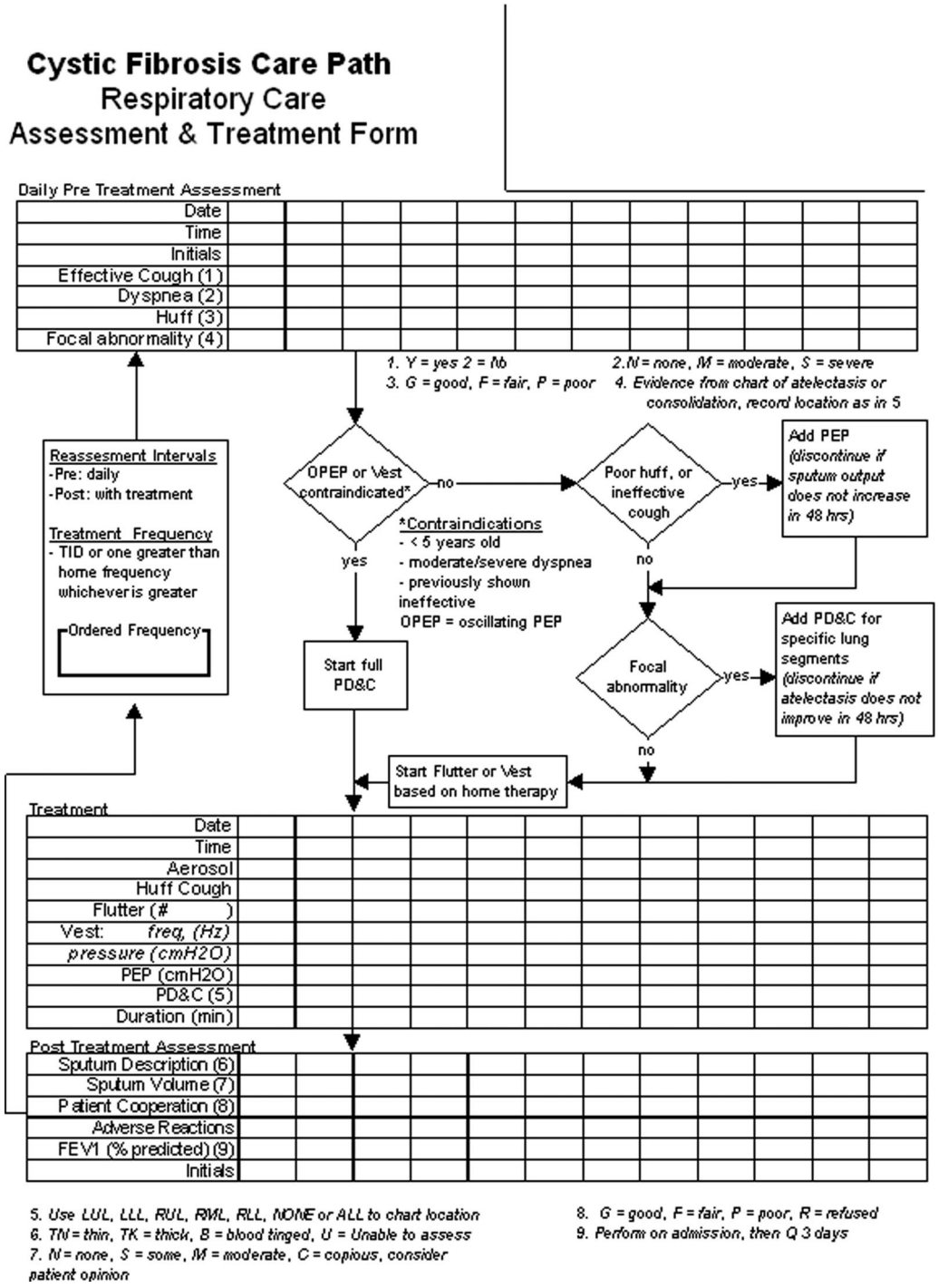

Fig. 3. Assessment and treatment (airway-clearance protocol) form for respiratory care of patients with cystic fibrosis (CF). OPEP = oscillatory positive-expiratory-pressure therapy. TID = 3 times a day. PD&C = postural drainage and percussion.

cline in lung function.[38] Patients, families, and caregivers need to understand the importance of regimen adherence in preserving health and improving longevity. To uncover knowledge gaps and address adherence barriers, the health-care team should establish a sense of partnership with the patient, and maintain open and frequent communication with the patient, family, and/or caregiver. It is important to acknowledge and seek ways to alleviate the time and labor burden of the regimen,[39] and to proactively be a rich source of information, partly to minimize the risk that a patient, family member, or caregiver might obtain and use information or opinions from potentially questionable sources,

Table 1. The Role of the Respiratory Therapist in Patient and Caregiver Education[34]

Therapy regimen
Medicated and/or bland aerosols
Airway-clearance techniques
Sequencing of treatments
Regimen schedule (adjustment to fit lifestyle)
Equipment decontamination and storage
Hand hygiene
Recognition of signs/symptoms of an exacerbation
Importance of regularly scheduled clinic visits
Evaluate lung function
Sputum cultures
Socialization practices

such as the Internet.[40] The printed information you provide should conform to the reading standards of the United States Department of Health and Human Services: materials are considered "difficult" to read if written above a 9th-grade level, of "average difficulty" if written between a 7th and 9th grade level, and "easy" to read if written below a 6th-grade level.[41] Illustrations, photographs, and drawings may substantially improve the patient-education materials and may obviate or at least explain recondite medical terms and concepts.

Disease Management

RTs also have an important role in optimizing health-care-resource utilization. A 1996 study in Oakland, California, found a mean annual cost of $13,300 for CF care (range $6,200 to $43,300),[42] and the cost paralleled the disease severity. Fifty-three percent of the cost was for medications and out-patient or clinic care, and 47% was hospital costs.[42] A more recent estimate of the lifetime cost of CF care was $200,000 to $300,000.[43] That analysis included the medications and therapies of current practice, but was based on the 1996 cost-of-care values.[43] Although the latter lifetime-costs estimate is an underestimate of the cost of today's standard of care, the study highlights the profound economic impact of CF.

The literature documents the important role of RTs in the management of chronic diseases such as asthma and chronic obstructive pulmonary disease. RTs have developed and implemented self-management and patient-education programs that decreased the cost of care by decreasing emergency-department visits, in-patient days, school absences, and missed work days.[44-46] RTs can coordinate care and attend to the complex needs of the CF patient and family. As part of the CF care team, RTs also function as patient advocates. The RT can support the use of services and patient-assistance programs (eg, from the Cystic Fibrosis Foundation) recommended by the CF care team members. The partnerships RTs develop with patients and families along the continuum of care enhance their ability to assess the patient's education, social-support, and home needs, and to employ strategies that build effective self-management skills and behaviors. RTs' specialized training and skill sets allow them to adapt to and flourish in this dynamic new health-care environment.[47]

Summary

CF is a life-shortening genetic disorder caused by mutations of the CF transmembrane conductance regulator gene. Although nearly all of the body's exocrine glands are affected, profound pancreatic, gastrointestinal, and respiratory manifestations occur. Diagnostic tools, drugs and other therapies, and standardized interdisciplinary care has improved patient outcomes and longevity. The RT plays a key role in the care of the CF patient, from performing therapies to educating patients and families on early detection of exacerbations and establishing disease-management strategies. RTs' commitment to helping patients and families effectively self-manage CF improves patients' overall health status and quality of life.

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Discussion

O'Malley: In the CF community we have what is known as "steals shamelessly." I like your care plan; are you going to publish that?

Volsko: I haven't planned to, but we'll definitely share it.

Rubin: Terry, why don't you include it in your manuscript? That way it will be available to all the RTs.

Volsko: That's a wonderful suggestion. I will do that.

Geller: On most CF care teams there's at least one RT, and the RTs on the in-patient side often have the most patient interactions over a long period, and that's where a lot of information is exchanged between families, patients, and clinicians. But sometimes misinformation is exchanged. How do you educate the other RTs on the extended CF team to make sure the information transmitted to families is consistent?

Volsko: We have held brown-bag lunches for RTs who were interested in being part of the in-patient team; we fed and educated them. We also had monthly team meetings so that the RTs who were a part of the CF care team and would see the patients on an out-patient basis could interact with me and the RTs who were interested in providing in-patient care. We discussed cases and care-path outcomes, and identified areas of improvement. Education doesn't stop with patients and families; RTs need it too.

Flume: Terry, coordination is obviously important in the in-patient setting, because you've got dietary, nursing, and others involved in the patient's care. At some centers, and certainly in other countries, it's the physical therapists [PTs] who do the airway clearance therapies. At our center the RTs do that, but in other places there's one more person or one more discipline that's involved in the care plan. What about incorporating PTs into that care team, and how to coordinate that?

Volsko: We had a PT on the in-patient side, who saw our patients. We got a grant and some exercise equipment. The director of nursing gave us some space on the in-patient unit to set up a small exercise room. As the patients progressed during their in-patient stays we had a PT work with them on exercise. I think that's extremely important; exercise is an airway-clearance modality.

Marshall: I want to ask about the protocol development, which is so important for standardizing what we do. The power of standardization in improving outcomes is now well recognized, but, of course, it often needs to be customized to the individual patient: the term that's thrown around is "mass-customization." No protocol will serve 100% of people perfectly, but hopefully 85–90%, and then it's up to the intelligent clinician to be able to modify it. I was pleased to hear you say that your protocol became more patient-centric, because we want flexibility to meet *patient* needs and preferences, not practitioner need and preferences. We ought to be able to figure out what's right to do and in what sequence, but also to be able to customize it to the patient need and preference.

You showed bar graphs with one time point. One thing we use in healthcare quality-improvement is called a "simple run chart," where you decide on your metric and then follow it over time. The power of that is that it gives you a visual of the dynamics of the situation. You can put in a line for median tendency; you can put a goal line in there, say, for medication misses; you set a target of 5%, and you can work toward that over time. I'd encourage you to think about that.

Another aspect of protocol development is that, as you pointed out—and I congratulate you on that—it is a dynamic process. Another aspect of quality-improvement is looking for protocol violations or deviations and learning from those to improve the protocol. That's where you brought in the patient-centered approach and it sounds like it is very useful.

Volsko: Thank you.

Lester: You mentioned RTs who were scheduled to do 18 chest-physiotherapy sessions a day. That's an incredible workload. That did not get done, obviously.

Volsko: It didn't, and we really beat our RTs to a pulp.

Lester: Being the RT on both the adult and CF team at our center, it's my job to teach the new employees. I get to spend about 2 hours with them in the orientation. We've been using the Respiratory 101 Mentoring document (available at PortCF.org) as a competency tool in our RT department. CF is included in our RTs' annual competencies. After a 30-minute review of the RT 101 document, they take a test that I made up. I find that being on the CF team also requires spreading that knowledge throughout the RT department.

O'Malley: What do you do when an RT is working with the patient and family—and RTs do spend a lot of time with the patients and parents—and the patient or a family member asks a question that is beyond the RT's expertise? Our CF center director mentioned that an had RT incorrectly answered a family's question about genetics. We addressed that problem by doing a presentation on CF, in which we clearly stated that RTs are experts, but if they're asked about growth, nutrition, medicine, or other topics that aren't clearly in their expertise, they must refer the question to the appropriate professional. It's important for RTs to know their limitations.

Volsko: I agree. We wanted to monitor that, so at our center we developed and implemented a supplement to our patient-satisfaction survey, which poses questions that try to identify any little "roadblocks" or "speed bumps" that happened during the stay. There are questions about whether the RTs or other team members embraced their boundaries. And we caught a few, but our process is not 100% foolproof. This a very dynamic process, and we need to look at how we monitor our progress, with the right tools, and constructively deal with the data we obtain.

Davies: I agree with you, Cathy, but it's very much a 2-way process. Patients often ask me questions that are way outside my sphere of knowledge but that Terry and you would be much more knowledgeable about. And this is one of the benefits of multidisciplinary-team CF care—that the team members have diverse expertise. And we should have no problem allowing our patients to know where our knowledge finishes and where that of another team member might start.

O'Malley: Right: the dynamics of a good team.