

This month we are pleased to publish the second group of papers from the RESPIRATORY CARE Journal Conference, "Respiratory Care and Cystic Fibrosis."

Mucus and retained secretions are the first things to come into the minds of most respiratory therapists when cystic fibrosis (CF) is mentioned. It is interesting to learn from the paper by Rubin that CF lung disease is *not* largely due to hypersecretion of very viscous mucus, and that there is almost no intact mucin in the sputum with CF. In fact, sputum associated with CF has lower viscosity when compared to asthma or bronchitis sputa, but is highly tenacious and closely resembles pus. It is these characteristics that lead to decreased cough clearance of infected phlegm. Medications aimed at reducing mucin secretion or severing mucin polymers are likely to be of no value and might even be dangerous. Medications meant to decrease the tenacity of sputum containing DNA and F-actin polymers are more likely to be of benefit. Because phlegm tenaciously sticks to the airway epithelium, medications that unstick secretions from the epithelium, such as surfactant or drugs that promote water secretion into the airway, should also benefit patients.

There is a wide range of airway-clearance therapies that can be used for secretion removal in patients with CF. The CF Foundation developed clinical practice guidelines related to airway-clearance therapy, which we published in the April 2009 issue of RESPIRATORY CARE. These guidelines recommend that airway-clearance therapy should be performed by all patients with CF. However, no form of airway-clearance therapy is superior to another and patients may express a preference for one therapy over another. It is also important to determine which therapy is best for patients of different ages and at different stages in the disease. Respiratory therapists and physical therapists help patients and families develop individualized airway-clearance routines that aid in the removal of the secretions. Aerobic exercise is beneficial to patients with CF and should be a component to their overall health routine. One of the most useful parts of the paper by Lester and Flume is the Appendix, which details the proper technique for the various airway-clearance therapies that can be used for patients with CF.

A number of medications are administered by inhaled aerosols to patients with CF. These include bronchodilators, airway wetting agents, mucus-active agents, antibiotics, and others. Until recently, the aerosol devices used for treatment of CF were limited to metered-dose inhalers, dry-powder inhalers, pneumatic jet nebulizers, or ultrasonic nebulizers. A major challenge for patients is dealing with the time burden required to fit all of their treatments into the day. Moreover, novel inhaled drugs that target the genetic defect of CF as well as the pathophysiologic consequences may require a device that is more efficient than those used in the past and at present. New aerosol delivery systems are being developed to address these and other issues. As described by Kesser and Geller, these include slow mist devices, vibrating mesh devices, and smart devices such as adaptive aerosol delivery and breath-control devices. Increasingly, new formulations are released to the market that require the use of a specific nebulizer device. Novel dry powder formu-

lations are also being developed and may be available in the near future.

The leading cause for hospital admission in a patient with CF is an acute worsening of signs and symptoms. The reasons for admission are usually the need for intravenous antibiotics and aggressive airway clearance with good nutritional support. Caring for the hospitalized patient with CF requires the skills to perform and teach airway-clearance therapies and to understand the medications and delivery devices used for a CF treatment. In addition, skills such as the application of noninvasive ventilation and for the care of the end-stage patient awaiting lung transplantation are needed. As described by Newton, an important aspect of good therapy is to be a great coach. This involves coaching patients in infection control, proper use of the nebulizer, and proper use of airway-clearance therapy.

Lung transplantation has become a viable option for CF patients with end-stage lung disease. In fact, survival after lung transplantation is more favorable in patients with CF than for patients with chronic obstructive pulmonary disease or patients with pulmonary fibrosis. As discussed by Rosenblatt, in May 2005 the United Network of Organ Sharing instituted a lung-allocation score to better distribute donated lungs to those patients who would achieve the most benefit. This has resulted in a decrease in the median waiting times until transplantation, and the mortality on the waiting list has decreased. However, the number of CF patients transplanted has not changed. The limiting factor in lung transplantation is the number of organs available. An area that has recently received increasing attention relates to mechanical ventilation strategies to improve gas exchange in the donor, thus allowing more patients to be transplanted. Clearly more work is needed to determine the best way to ventilate the donor, both to increase the likelihood of the lungs being acceptable for transplantation but at the same time to minimize the risk of ventilator-induced lung injury.

Substantial progress has been made in the care of patients with CF. There is no better evidence for this than the marked increase in the median predicted age of survival for persons with this disease. The mission of the CF Foundation is "to assure the development of the means to cure and control cystic fibrosis and to improve the quality of life for those with the disease." Consistent with this mission, the CF Foundation supports programs in basic science, drug discovery, drug development, clinical care, patient education, and advocacy. As described by Marshall, one of the programs instituted by the CF Foundation is a mentoring program to teach respiratory therapists new to CF about the nuances of care for these patients. Kudos to the experienced respiratory therapists who have stepped forward to serve as mentors in this program.

The 43rd RESPIRATORY CARE Journal Conference brought together experts from the United States, Canada, and the United Kingdom to review the art and science of CF. This is the first time that CF was the topic for a Journal Conference, and it came about 6 decades after the disease was named, and 20 years after the gene was discovered. The May and June 2009 issues of RESPIRATORY CARE reflect how diligently the faculty worked to provide up-to-date reviews and lively discussions of these topics.