

Respiratory Care and Cystic Fibrosis

Cystic fibrosis (CF) is a multisystem disease with pulmonary infection and inflammation causing the greatest morbidity and mortality. Respiratory therapists have been involved in the care of patients with CF since this was first described as a distinct disease nearly 60 years ago; teaching and applying airway clearance techniques, administering therapeutic aerosols, helping to perform bronchoscopy and pulmonary function testing, and caring for patients requiring noninvasive ventilation and after lung transplant. Respiratory therapy and research respiratory therapists continue to be on the forefront of new discoveries and the application of new therapies for the treatment of CF.

Our knowledge of CF respiratory disease has evolved tremendously since the identification of the primary gene defect and cystic fibrosis transmembrane regulator (CFTR) protein 2 decades ago. This 43rd RESPIRATORY CARE Journal Conference is a state-of-the-art review of current knowledge of CF pulmonary disease and respiratory care. This conference emphasized the role of the respiratory therapist, while also discussing methods of diagnosis, applications of therapy, and scientific understanding of the underlying pathophysiology.

Dr Rubin has disclosed relationships with Pfizer, Ventaira, Trudell Medical International, Monaghan Medical, GlaxoSmithKline, Medihale, Pharmaxis, Syntaxin, Bayer, Hill-Rom, Boehringer Ingelheim, and RegeneRx.

Dr Geller has disclosed relationships with Novartis, Bayer, Mpex, Pari, Aerogen, NanoBio, Aradigm, Genentech, CSL Behring, Boehringer Ingelheim, Trudell Medical International, Monaghan Medical, and Respironics.

Internationally recognized experts in the science of CF and the care of persons with this disease gathered to review the pulmonary pathophysiology of CF, including bacterial infection and microbiology, secretion retention, aerosol deposition, and chronic inflammation, and how each of these influences disease severity and selection and use of appropriate therapies. New guidelines related to CF in the adult, including late complications, mechanical ventilation, and lung transplantation, were also emphasized. In all cases the role of the respiratory therapist in the care and education of persons with CF was the central focus. Respiratory care protocols, infection-control measures and education, the application and teaching of airway clearance, and the respiratory care of the hospitalized person with CF have been fully reviewed, and guidelines are offered for the bedside caregiver. We also welcomed the participation of Dr Bruce Marshall, Vice President of Clinical Affairs for the Cystic Fibrosis Foundation, who reviewed the history and current activities of the CF Foundation and the special role that respiratory therapists play in the CF Foundation.

Bruce K Rubin MEngr MD MBA FAARC

Department of Pediatrics
Wake Forest University School of Medicine
Winston-Salem, North Carolina

David E Geller MD

Nemours Children's Clinic
Orlando, Florida