



Cystic Fibrosis: A Commitment to Research and Treatment

ON THE COVER: Cystic fibrosis patients Ellie Schore, 17, and Sarah Yourman, 32 months, can expect to live longer and better lives because of progress in both treatment and basic research. P&S has made important contributions in both areas. Page 3.
Photo by Jonathan Smith.

Dr. Lynn Quittell with CF patients Sarah Yourman, 32 months, and Ellie Schore, 17.

In August 1992, researchers at the University of North Carolina at Chapel Hill produced genetically altered mice that suffer many of the symptoms of CF, a development that could speed the testing of new treatments for the disease. These and other developments have made clinicians such as Dr. Quittell increasingly optimistic.

"I remember one of my patients, a high school girl, talking to me a few years ago about how she didn't think there was any point in going to college and putting in all that hard work if she was going to die soon anyway," says Dr. Quittell. "At the time I understood how she felt and wasn't sure I could in good faith persuade her otherwise. Today, given all the new treatment modalities on the horizon, I feel very differently."

The CF approach at CPMC is a strong meshing of basic research and clinical medicine. Dr. Quittell says the hospital is deeply committed to increasing services to patients at the CF center, which currently has 125 patients.

Dr. Robert Mellins, professor of pediatrics and director of the pediatric pulmonary division, says the team of physicians treating CF patients provides outstanding care. "The doctors who work with CF patients here not only have far-reaching expertise but also are extremely caring individuals who treat with their hearts as well as their minds," says Dr. Mellins. "Dr. Celia Ores, assistant professor of clinical pediatrics, who bridges Columbia's involvement in CF from Dorothy Andersen's time to the present; Dr. Anastassios Koumbourlis, assistant professor of pediatrics, who specializes in critical care, especially post-lung transplantation; and Dr. Lynn

Quittell all do their best to improve life for people with this disease."

Five pediatric pulmonary fellows conduct research in the research and development program headed by Dr. Samuel Silverstein, the John C. Dalton Professor and Chairman of Physiology and Cellular Biophysics and principal investigator and coordinator of the research program.

Because the disease has a number of different physical components, treatment takes a multidisciplinary approach and includes pulmonologists, gastroen-

terologists, nutritionists, and nurses. A CF social worker, Winsome Sewell, deals with the many psychological problems that arise among patients and their families burdened with a disease that makes staggering demands on time, energy, and financial resources.

about the way the disease is treated there. During this visit and in two subsequent visits to Moscow and St. Petersburg, the team was amazed and dismayed to find that modern methods for treating CF have virtually bypassed the country. Instead of receiving an antibiotic the bacteria affecting their lungs are sensitive to, patients may be treated with whatever antibiotic is available. Perhaps not surprisingly, the median life expectancy for CF patients in Russia is seven years—20 years lower than in the United States.

The team showed Russian doctors how to use a simplified sweat test machine to diagnose CF and taught the importance and techniques of chest physical therapy to parents.

Because of the crying need to help Russian CF patients, Dr. Quittell's group plans to continue its work by devising means to supply medicines continuously and to send U.S.-trained physicians and nurses to visit the CF community there on a regular basis.

When the progression of CF is particularly virulent and the lungs are destroyed, lung transplantation is the final option to save a life. At



CPMC, the first lung transplant for CF was performed in 1989 by Dr. Craig Smith, associate professor of surgery. Since then, six other CF patients have undergone transplantation. Several patients are undergoing evaluation and others have been evaluated and await organ donation.

"Our goal is for all our patients to lead normal lives, to not get so sick, and to prevent the progression of the disease," says Dr. Quittell. Increasingly, she feels, this is becoming possible.

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In 1989, Dr. Quittell and a delegation that included Dr. Lisa Saiman from Columbia and Dr. Robert Beall and Robert Dressing of the CF Foundation, traveled to what was then the Soviet Union to meet with doctors to learn