Cystic Fibrosis in Focus

In the first year of the new millennium the thematic review series in *Respiration* is focusing on cystic fibrosis (CF), a disease which may be as old as modern man. The six issues will cover the pathogenesis of lung disease, modern centralised CF care, CF in the adult, the relationship between genotype and phenotype, future pharmacological treatment, and methods for early detection of airway disease in infants and children with CF.

There are at least three explanations for the great interest in CF currently. Firstly, the life expectancy today of an infant with newly diagnosed CF is certainly very much longer than only a few decades ago, when many children did not reach school age. Now over 80% of the CF patients attending some CF centres may expect to reach at least the age of 40 years [1], and soon we can expect to see as many adults as children with CF. This dramatic change is the result of improved traditional treatment based on controlling airway infection, mobilisation of secretions, reduction of inflammation, and improved nutrition. CF is no longer the concern only for the paediatricians, but also for chest physicians, internists, and even obstetricians!

Secondly, the improved knowledge about the pathophysiology of the disease will provide new ways to attack the disease pharmacologically. It started with the discovery by Di Sant'Agnese et al. [2] in 1953 that the sweat from subjects with CF was abnormally rich in salt, leading to the conclusion that CF is a general exocrinopathy. In 1983 Quinton [3] reported that the primary defect in CF is the reduced Cl^- transport across the cell membranes. Further research has demonstrated a variety of distur-

bances resulting from the primary defect, and CF has become a model disease of disturbed transmembrane ion transport.

Thirdly, the spectacular advances in the delineation of the genetics of the defect has brought hope that a cure for CF may not be too far away. The gene coding for the defect chloride channel regulating protein (CFTR) was identified by Riordan et al. [4] in 1989. Today over 800 mutations in the CF gene have been reported.

Finally, the impressive improvements in CF treatment and research are the results from collaboration between dedicated people with widely differing professional backgrounds. In this sense too CF is a model disease.

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References

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