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Cystic fibrosis

Series editor: D J Shale

Introduction

The effects of improved survival in patients with cystic fibrosis are being felt by respiratory physicians as young adults seek care for their chronic respiratory disease. This largely reflects the effects of improved care in the neonatal period and infancy 20 years ago; the effects of current paediatric practices will be experienced by adult respiratory physicians in another two to four decades. Most patients seen currently have progressive lung disease and its management demands a considerable proportion of time and resources from respiratory services.

Although respiratory complications account for 98% of deaths from cystic fibrosis in adults, the morbidity is shared with other affected systems, such as the liver and gastrointestinal tract. A respiratory physician caring for patients with cystic fibrosis must be aware of these and other clinical problems, as well as the social and personal consequences of the disorder to the patient (table). The management of adults with cystic fibrosis requires an all round approach with a supporting team of specialists, as recommended by working parties of both the British Thoracic Society and the British Paediatric Association. Debate continues over the benefits of cystic fibrosis centres, usually defined as a clinic with more than 40 patients, though it would seem to be more important for a centre to be defined by the facilities offered than by the number of patients seen. Adequate care of adults with cystic fibrosis requires not only familiarity with the disorder but having the resources to cope with the various manifestations and complications.

Problems of adult patients with cystic fibrosis

Medical
Pancreatic insufficiency
Chronic respiratory infection
Diabetes mellitus
Cirrhosis, portal hypertension, oesophageal varicies
Meconium ileus equivalent
Delayed physical development
Male infertility
Female infertility
Reduced lifespan
Social and psychological
Education
Employment
Effects of chronic disease
Delayed maturity
Family relationships
Death

There are likely to be considerable developments in the management of cystic fibrosis as a result of the identification of the gene defect and of the cystic fibrosis transmembrane regulator protein. This has implications for earlier diagnosis, carrier detection, genetic counselling of relatives, and new forms of treatment. In addition, other therapeutic agents are likely to be tried and may go some way towards slowing the progression of lung disease before gene therapy becomes a reality for use in infancy. These developments will impinge on the adult physician caring for adults with cystic fibrosis because patients and their relatives will want to take advantage of genetic investigations and advice. As in other groups of patients with chronic disease, patients with cystic fibrosis expect a high degree of knowledge from their physician—at least equal to their own.

This series of articles on cystic fibrosis will cover respiratory and non-respiratory clinical topics relevant to physicians caring for such patients. The series should in addition provide insight into fundamental work that will change the pattern of clinical practice in the future. The series begins with a consideration of the problem of *Pseudomonas aeruginosa* infection and its management and a review of the mechanisms that underlie lung injury in cystic fibrosis as these are of immediate relevance to the respiratory complications seen by adult physicians. Further articles will cover heart-lung transplantation, current understanding of the genetics of cystic fibrosis, and the physiology and pharmacology of the cell membrane in cystic fibrosis. Other clinical topics will include the management of gastrointestinal problems and a review of management policies at three treatment centres in different countries.

We hope that this series, by providing an up to date review of topics relevant to the care of adult patients with cystic fibrosis, will increase interest in this disorder among respiratory physicians. In particular, it should provide information for physicians being asked to manage adult patients with cystic fibrosis.

D J SHALE
Respiratory Medicine Unit, University of Nottingham, City Hospital, Nottingham NG5 1PB