CHANGING FACE OF CYSTIC FIBROSIS

Scott Bell

Cystic fibrosis (CF) has rapidly changed from a predominantly paediatric disease with markedly reduced life expectancy to a lifelong disease which places its major burden on adults. More than 90% of children born with CF in Australia are expected to live beyond their 18th birthday. This has resulted in a major burden on the provision of clinical care in hospitals with adult respiratory units, and the provision of multidisciplinary care has been limited by available resources.

As young people with CF grow older, new clinical challenges and complications of the disease are emerging. These include cystic fibrosis-related diabetes, bone disease, the emergence of new bacterial pathogens which are often multi-resistant, and increasing numbers of CF patients pursuing parenthood. A multidisciplinary approach to the management of these emerging complications is required, particularly when associated with advanced lung disease. Treatment of lung disease is dependent on the accurate detection of changing airway biology, infection and inflammation. High resolution CT scanning and lung function testing in the very young child with CF offer new understanding and potential for earlier detection of lung damage. Lung transplantation continues to offer hope for patients with cystic fibrosis who have end-stage lung disease. New and novel therapies include gene transfer therapy, mucolytic therapies including hypertonic saline, new antibiotic therapies including macrolides, and the hope of pharmacological agents which alter the basic CFTR defect. This has led to very complex, expensive and time-consuming therapies, which impose their own burden on the lives of people with CF and the hospitals who provide their care.