

Cystic fibrosis in Melbourne

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The outlook for cystic fibrosis has changed dramatically in the past 30 years. Major clinics report a cumulative survival of patients managed during the past 5–10 years of about 80% to late teenage and 50% to the age of 30. There is considerable uncertainty about which factors are responsible for this improvement as few aspects of the management of cystic fibrosis have been subject to appropriately designed double blind controlled trials. Treatment regimens have evolved over the years largely on the basis of the experience of clinic physicians.

Probably the single most important factor in improving survival has been the development of specialised clinics. The Royal Children's Hospital's cystic fibrosis clinic, established in 1953, currently manages 360 children and adolescents, about 95% of those in Victoria, Australia (population 4 million, annual births 60 000). The associated adult clinic at the Alfred Hospital has 130 patients aged over 18 years. These clinics ensure a continuum in management from childhood to adolescence and into adult life. Patients transfer to adult care when they are established in tertiary education or in the workforce. The clinics are multidisciplinary with thoracic physicians (paediatric or adult), social workers or clinic coordinators, dietitians, physiotherapists, and occupational therapists. Each patient has a specific physician who is ultimately responsible for his or her care. Patients are seen in thoracic medicine outpatients departments, within which the specialised services are provided to avoid the label of cystic fibrosis being too visibly applied. Other medical specialists such as psychiatrists, gastroenterologists, and surgeons are available for specific problems.

An emergency advice telephone service is provided, and patients with acute problems can usually be seen on the same day. Generally, patients do not attend their family practitioner for problems associated with their cystic fibrosis. For paediatric patients living outside the metropolitan area a regional paediatrician is usually involved and helps with acute problems, but the clinic physician retains final responsibility. At three monthly outpatient attendances height, weight, and pulmonary function in patients over 7 years are measured and a chest radiograph is obtained at least annually. Patients are admitted to clinic hospitals for exacerbations of chest infection, for complications of cystic fibrosis, and for emergency and elective surgery to the care of a thoracic physician irrespective of the nature of the problem. A few children with minor flare ups of chest infection are admitted to district hospitals after discussion with the thoracic physician responsible.

The diagnosis

The management of the patient and family at

the time of diagnosis establishes a pattern and an approach that has a fundamental influence on the course of the disease.¹ Once the diagnosis is established both parents are seen by the paediatric thoracic physician responsible for the patient's long term management together with the clinic coordinator or social worker. At the initial interview basic factual information and the realities of cystic fibrosis are discussed, but a hopeful and positive outlook is given. If both parents and caregivers have a positive approach it will play a major part in achieving the best outcome.

After this discussion the child is admitted to hospital, where a three day study of fat balance is undertaken, any chest infection is treated, and considerable time is spent by all team members and nursing staff in educating the parents on all aspects of cystic fibrosis. Two to three weeks after discharge the patient and family are seen again. In the meantime there will be telephone contact with clinic members and the district nurse attached to the clinic will visit the family at home.

The chest

Every effort is made to delay the development of chronic chest infection and once it develops to retard its progress with antibiotic treatment and physiotherapy. A loose cough or expectoration of purulent sputum is treated with antibiotics. Patients who are usually cough free, have normal pulmonary function and have normal results on chest radiography and who develop recurrent infection are treated with an oral antibiotic against *Streptococcus aureus* and *Haemophilus influenzae* for two to three weeks or until the cough clears. If the cough does not clear the patient's condition is reviewed; a different oral antibiotic is given for a further two weeks or the patient is admitted to hospital, particularly if there is a loss of pulmonary function. Patients with chronic cough and sputum will normally be taking oral antibiotics long term and occasionally a high dose inhaled aminoglycoside. Exacerbations in this group of patients are initially treated with a different oral antibiotic. Chloramphenicol is useful and is often combined with a high dose inhaled aminoglycoside. Ciprofloxacin is used in older patients. If the flare up is not controlled within two to three weeks the patient is admitted to hospital. Admission is also indicated if there is deterioration in general health, weight loss, reduced lung function, or new radiographic changes. Routine admissions are not favoured.

Inpatient management generally entails intravenous treatment with an aminoglycoside and ticarcillin. Other antipseudomonal antibiotics are not used initially unless the patient was recently admitted to hospital and failed to respond to the standard regimen. If after 10 days the clinical response is inadequate and sputum culture indicates *Pseudomonas aeruginosa* resistant to standard drugs the treatment is changed. *P cepacia* has been seen rarely. Inpatient treatment also entails intensive physiotherapy, nutritional supplementation, and continuation of a regular exercise

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programme. Patients are discharged once their sputum production returns to previous levels, pulmonary function is restored to baseline, and they feel better. Sometimes a new baseline for sputum production and lung function has to be accepted after three to four weeks of treatment. Home intravenous treatment is used in some older patients after six or seven days in hospital and is attractive to patients who are working full time. It is not used in younger patients as there are other reasons for admission to hospital besides intravenous treatment—a break for both parent and patient, more intensive physiotherapy, and an opportunity for re-education all seem helpful.

Removal of tracheobronchial secretions is of great importance. All parents are shown physiotherapy methods and from the age of 10 or 11 children learn the forced expiratory technique so that they can be independent of needing help. Effective coughing is taught as this is probably the most important aspect of physiotherapy.² All patients have once or twice daily physiotherapy for a time after diagnosis and if they have a loose cough. Salbutamol is often inhaled before physiotherapy. If the patient is cough free with no sputum and good lung function regular physiotherapy is not mandatory. The vital role of physical activity is emphasised at diagnosis. From an early age children are encouraged to participate regularly in a range of sporting activities. In older children and adults with minor lung disease daily sessions of aerobic exercise may completely replace physiotherapy. Sessions of aerobic exercise of 20–30 minutes' duration are recommended, after which patients spend a few minutes coughing effectively. Physically active patients seem to maintain better health.

Nutrition

Normal growth during childhood and adolescence and maintenance of nutrition in adults are of great importance, though at present there are inadequate data to confirm the suggestion that poor nutrition predisposes to progressive lung disease. Patients with cystic fibrosis need an energy intake about 120–150% of normal requirements. This is best achieved orally, and the main role of the dietitian is to encourage a high and varied energy intake with supplementation as necessary. The dose of pancreatic enzymes is adjusted to the degree of malabsorption and the patient's preferred food intake. There is little evidence to suggest that very high intakes of pancreatic enzyme are effective in most patients with malabsorption that is difficult to control.³ Recent studies in our clinic showed that some of these patients have a very low postprandial pH in the fourth part of the duodenum that may be corrected by the use of the prostaglandin analogue misoprostol.⁴

In the past 5–10 years enteral and parenteral nutrition have been recommended for patients not maintaining an adequate weight. Though short term weight gains can be achieved, the long term benefits are uncertain. We have found

that overnight nasogastric feeding is poorly tolerated and may lead to exacerbation of lung disease because patients avoid coughing. Permanent gastrostomy for overnight feeding has been more successful in a few children and a larger number of older patients who have been unable to maintain a reasonable weight.

Transition from paediatric to adult care

A specific transition programme has been developed to effect efficient transfer of patients from the paediatric to the adult centre and is a critical component of the overall management plan. Patients and their families are prepared years in advance to expect transfer and to work towards this as a goal of treatment. The programme recognises that at the same time several separate transitions may be occurring for the patient, including (a) transition from paediatric to adult care, (b) transition from adolescence to adulthood, and (c) transition from being well to having more symptoms. Strategies have been developed by the health care team to address these multiple transitions both collectively and as individual issues. The adult care team normally meets the patient once in the paediatric hospital before the formal transfer takes place.

The future

With comprehensive care that gives hope and support to patients and family, survival to adult life should be the normal expectation. Most of our adult patients are in continuing education or full time work. Cystic fibrosis, however, still inevitably results in premature death. Members of the clinic must be as skilled in helping patients and their families with the terminal stages of the illness as they are in promoting good health. Looking after dying patients places stresses on the health care team, and perhaps adult clinics, where most deaths occur, should be limited to no more than about 150 patients.

Unless there are major changes in treatment, perhaps as a result of the identification of the cystic fibrosis gene and of the basic metabolic defect, it is hard to see that there will be further dramatic improvement in life expectancy. There is inherent variability in the severity of the disease, and despite optimal care probably 10–20% of patients will continue to die during childhood and adolescence. We may see further improvement in the survival of patients who reach adult life, but even with heart-lung transplantation none has a normal life expectancy.

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- 3 Robinson PJ, Sly PD. High dose enzymes in cystic fibrosis. *Arch Dis Childh* (in press).
- 4 Robinson PJ, Smith AL, Sly PD. Duodenal pH in cystic fibrosis and its relationship to fat malabsorption. *Dig Dis Sci* (in press).