Cystic fibrosis in teenagers and young adults

The millennium will see over 6000 patients with cystic fibrosis in the UK, half of whom will be teenagers and adults, the over 15 age group increasing by about 120 per year.1 We are not providing sufficient resources for this latter group now, nor are we making sufficient commitment to their future needs. There are three reasons for this. Firstly, some fault rests with the apparent failure of the NHS hospital trusts’ management to deploy income derived from purchaser provision for cystic fibrosis care back into the cystic fibrosis service. There is also their persistent failure to meet the staffing levels recommended for cystic fibrosis centres by the British Paediatric Association (BPA), the British Thoracic Society, the Cystic Fibrosis Trust,2 and the Royal College of Physicians3 and their alleged failure to have established a realistic cost for treatments. Secondly, there are a significant minority of professional colleagues who refuse to refer older patients with this multisystem disease to cystic fibrosis centres with appropriate multidisciplinary teams. Finally, some fault also rests with those of us who have a major commitment to cystic fibrosis care failing to initiate a proper training programme with recognised accreditation for junior doctors wishing to care for patients with cystic fibrosis.

Special needs of the teenage patient with cystic fibrosis: the ‘transitional’ clinic

Inevitably close ties develop between the paediatric medical and nursing teams and the patients and their families. Important though these are in giving the child and family emotional security, they must be loosened and eventually broken to allow the patient to grow into independence, and realise responsibility for self. Teenagers with cystic fibrosis can only grow into adulthood in an environment designed to cultivate their independence. Close collaboration between paediatric and adult cystic fibrosis teams is essential so that a system of care is established within which the patient’s transition through the turbulence of adolescence is as smooth as possible. In Leeds we believe that this is best achieved in a flexible ‘transitional’ clinic, run by staff from both the paediatric and adult teams. Patients usually transfer from paediatric to transitional clinic at about 14 years of age, and from transitional to adult clinic, according to individual needs, between 16 and 18 years of age. A survey of our own patients showed that knowledge of the transitional clinic had reduced patients’ anxieties about leaving the paediatric unit, and getting to know the adult carers, before moving on to the adult clinic proper, had reassured them.4

Cystic fibrosis centres for young adults

The teenager with cystic fibrosis must be allowed to leave the paediatric unit and the paediatrician behind. There is no justification for the paediatrician retaining the reins of care either because of the spurious excuse that no adult respiratory specialist will give the level of commitment required, or to maintain ‘patient numbers’ in the NHS market economy. Nor should ‘adult’ clinics run by a paediatrician and chest physician be seen as fulfilling a need outside of the transitional clinic referred to above. The paediatrician has to ‘let go’. But responsibility for continuing care must involve an adult cystic fibrosis centre. If, as paediatricians, we accept the argument for centred care for children with cystic fibrosis, then similarly we must accept centred care for adult patients.

Some with responsibility for older patients with cystic fibrosis believe that all of them should receive total care at the cystic fibrosis centre. Why? Because cystic fibrosis is a multisystem, complex disease and, with present high standards of care in paediatric centres, multisystem complications are often confined to adults. Hence, not only must a critical mass of experience be learned so that one is alerted to complications early, but colleagues in many disciplines (for example obstetrics and gynaecology, endocrinology, rheumatology, gastroenterology, surgery), must become familiar with the problems presented in their own fields by adult patients with cystic fibrosis. A sufficient patient ‘through-put’ can take place only in a cystic fibrosis centre. Access to specialist paramedical personnel and to basic, not esoteric, investigations is significantly easier within specialist clinics.5 Patients cared for by cystic fibrosis specialist staff receive more intensive treatment, especially with regard to nutrition, and are significantly more likely to adhere to physiotherapy regimens and to be self-reliant. Physicians who believe that they can offer optimal care by looking after a few patients with a physiotherapist, dietitian, and social worker unschooled in the special needs of cystic fibrosis, are wrong. Units which believe that they can offer a safe and optimal home based intravenous antibiotic treatment programme without a cystic fibrosis liaison nurse are wrong. Young adult patients have voiced clearly their concerns about the fall in quality service they have experienced on transfer from specialist paediatric cystic fibrosis units to general adult chest clinics.6

There has been a marked increase over the last five years in the numbers of older patients attending specialist clinics. Seventy seven per cent now receive full or shared care from a cystic fibrosis centre. Ninety eight per cent want some input to their care from a specialist centre and 96% of patients not currently receiving centre care would prefer it if they were.7 Existing centres must be properly resourced to cope with these rapidly increasing numbers, and new centres set up in areas of need.

What an adult centre must offer

First and foremost the centre must be adequately staffed and have access to adequate bed numbers, preferably on a dedicated cystic fibrosis ward. It is unlikely that anyone other than a full time cystic fibrosis physician will be able to care properly for more than 100 patients. Therefore most large units will require two consultants. Teenagers and adults should have easy access to significant amounts of their doctor’s time. Issues such as career choices, life
expectancy, problems of pregnancy and lung transplant, cannot be properly discussed at outpatient appointments or during bedside 'chats' within the ward round. Certainly junior doctors must be trained in the complexities of adult cystic fibrosis medicine, but patients cannot be expected to relate intimately and repetitively to a succession of short term staff who may know less about their disease than they do themselves. Units would be ideally served in addition by a permanent staff grade post.

Only those unaccustomed to the myriad problems of cystic fibrosis in an aging patient group would dare to assume ultimate responsibility for their wellbeing without the expert support of a physiotherapist, dietitian, social worker, pharmacist and nursing staff, all with experience of cystic fibrosis in their own field. More such professionals are needed now and this need will increase commensurate with the predicted expansion in the patient population. We should be working in concert with each of these disciplines to ensure that proper numbers of staff are being trained.

What is a realistic bed need? It is clear from first hand experience that the four beds per 50 patients suggested by the BAA are taken up by other commitments, if we are to treat for an adult clinic. In 1994 almost one quarter of patients were refused admission because no bed was available compared with 13% of patients in 1990. We are not responding to this worsening situation. Many patients need two to three monthly courses of intravenous antibiotic treatment. Some on the transplant waiting list will spend one to two weeks a month in hospital. Long and cold winters will further stretch resources. In our own unit in January of this year we regularly had 10–15 patients on a waiting list for admission.

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involved in their early years and appropriate transfer is made to an adult clinic. A commitment to this model is needed from all paediatricians and respiratory physicians to ensure that those with cystic fibrosis have continued access to the highest standards of medical care. The 'bottom line', however, is whether hospital management is prepared to properly resource the service. Unless new staff are appointed, and beds and equipment made available, the structure of care for teenagers and adults with cystic fibrosis is likely to fragment. I believe that this will inevitably, rapidly, and irretrievably be detrimental to patients' health.

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