Transition from paediatric to adult care: problems that arise in the adult cystic fibrosis clinic

A K Webb FRCP Andrew W Jones MRCP Mary E Dodd MCSP

*J R Soc Med 2001;***94**(Suppl. 40):8–11 SECTION OF PAEDIATRICS & CHILD HEALTH, 28 NOVEMBER 2000

INTRODUCTION

The superb care of paediatricians has ensured that the majority of cystic fibrosis (CF) patients become adults[1]. Mean survival of CF patients is now into the fourth decade of life, and it is predicted that the CF child born today will have a mean survival into the fifth decade[2]. The adult CF clinic in Manchester was developed in the early 1980s because there was a significant number of patients over the age of 16 still being cared for by paediatricians. The initially small number of patients attending the adult CF clinic now exceeds 200 and, each year another 15–20 are referred from the paediatricians. This continuum of paediatric and adult CF centred care has resulted in better nutrition and pulmonary function, the two main prognostic indicators for survival of patients with cystic fibrosis[3]. It is crucial that when the adult CF multidisciplinary team takes over the care of the paediatric patient the relationship gets off to a good start, which will survive the many hiccups that are part of the CF carer/patient lifetime. This article discusses not only getting it right at the time of transfer, but also the social and medical problems that can be anticipated during the first year of life as a CF adult.

TIME OF TRANSFER

Process

There has been considerable debate but no comparisons as to the best way of transferring paediatric CF patients to adult care[4], and very few structured guidelines that can be used to ensure a seamless path between the two[5].

Joint clinics held either at the adult or the paediatric hospital are the most common method. Adolescent units are a proposed halfway house but may require additional facilities[6]. Some patients transfer simply with a referral letter and just get on with life at their new adult centre, displaying little nostalgia for their paediatric years (personal observation), but there will always be debate about specific patients. Should prepubertal patients be transferred? Is it cruel to transfer the very sick patient who is likely to die within the year to an unfamiliar multidisciplinary team? Some paediatric teams are flexible and transfer between the ages of 16 and 18 when the patient feels ready, but some paediatric hospital trusts insist their patients transfer automatically at the age of 16.

However, although the method of transfer may be immaterial the exchanged social and clinical information about the patient is crucial. Specific detailed information about all aspects of care will help the patient feel more secure in their new surroundings. Multidisciplinary teams include staff who are experts in their own areas, each member having greater depth of knowledge about and training in their specific area of expertise. The specialist nurse will know more about the practical aspects of implantable venous access devices (ports) and home intravenous antibiotics than the physiotherapist, who correspondingly knows more about airway clearance, exercise and inhalation therapy. Both have a greater depth of knowledge than the consultant! This complex inter-dependence of knowledge is channelled into the individual patient. For this reason, members of the paediatric and adult CF teams should probably communicate individually at time of the patient transfer about detailed aspects of care.

Transfer of clinical information

Communicating as much clinical information as possible at the time of transfer will often forestall subsequent problems. Most paediatric and adult CF teams undertake annual reviews and these are an invaluable source of reference at the time of transfer. Some important areas of clinical interest to the adult team are outlined below.

Cross-infection between patients has increasingly resulted in outpatient and inpatient segregation. Essential information provided by the paediatric unit should include the patient's most recent sputum culture: the specific growth of *Burkholderia cepacia* (with strain type), methicillin-resistant *Staphylococcus aureus*, atypical mycobacteria and multiresistant *Pseudomonas aeruginosa* should be highlighted.

For many years the standard measures of disease progression have included spirometry, nutritional status and oxygen saturation: sequential tabulation of these results at the time of transfer will immediately provide an index of disease severity and potential for progression. Osteoporosis has a high prevalence in the paediatric and adult CF population[7,8], and most patients will have had a scan to

Manchester Adult Cystic Fibrosis Unit, Whythenshawe Hospital, Manchester M23 9LT, UK

Correspondence to: A K Webb
E-mail: the5webbs@hotmail.com

assess bone mineral density, which if reduced can be treated appropriately.

It is becoming increasingly important to diagnose and treat diabetes early in CF patients[9], as it may have a direct impact on prognosis[10]. A record of the most recent oral glucose tolerance test is invaluable. Liver status from annual or biannual ultrasound is a useful reference at time of referral for monitoring and potentially treating the progression of liver disease.

Information about levels of habitual activity and sporting interests give clues to fitness, which can often decline when the patient leaves school; the maintenance of interest in activities needs continuing encouragement.

Perhaps the two most difficult and sensitive areas to deal with at the time of referral are male infertility and female contraception, especially if a parent (usually the mother) accompanies the patient at the time of first consultation.

Most paediatricians inform their male CF patients about infertility before transfer. If the subject has been discussed, this information should be passed on to the adult unit. Questionnaires to male CF patients reported that they would like infertility discussed at about the age of 14[11]: in a recent study 26% of males learnt about their infertility only when they were over 20 years old[12].

Several years ago, at first consultation with a 23-year-old male accompanied by his mother, it became clear to the patient that he was infertile; the mother's guilty feelings that she had never told him were obvious. At the same time the consultant felt that he had been insensitive, having assumed the patient already knew about infertility.

Do CF paediatricians inform their adolescent male patients that they may be infertile because they have a blocked *vas deferens*, but that they are not sterile because they have sperm? Or should this be the role of the adult physician? At least this more positive message has given some hope to CF males that they can become fathers.

Do paediatricians discuss contraception and safe sex with their patients? It is extremely difficult to discuss this for the first time at an adult clinic. A small number of CF adolescents have not reached puberty at time of transfer. Are these conversations too early for them? Correspondingly, a large number of adolescents are sexually active before the age of 16. Chlamydial infection, which is sexually transmitted, is increasing in our female population, and the majority (68%) of pregnancies on our CF unit were unplanned. Clearly, the message about safe sex and contraception is not getting across to the patient from either the paediatricians or the adult physicians.

Social history

The social history (Box 1) sets the scene for the potential difficulties in all aspects of care. Observation of parents and patient together at the first interview will often give a clue to family dynamics. The parent is transferring care to the patient, who is probably also going to leave home in the near future. When the patient passes their driving test and receives a car through mobility allowance at 16 years old, they will no longer be dependent upon parents for transport, and nor will they want them to come to the clinic with them any longer. The parents are probably those who suffer most at the time of transfer.

Box 1 **Social issues**

- Education/ambitions
- Family interactions
- Relationships/puberty/fertility
- Smoking/alcohol/drugs
- Behaviour
- Benefits/allowances

It is important to discuss with the patient their education and ambitions. What do they enjoy in terms of social life? They will often be developing relationships: these distractions will impinge significantly on compliance.

Money or lack of it is a big issue for the adolescent population and at this time their allowances will transfer to them as adults. Disability allowances are usually reviewed about this time and can be withdrawn. It is important that social workers review these at transfer and ensure they are maintained and the review forms comprehensively completed.

It is important to know whether the patient or anyone else in the family smokes. A small number of patients take recreational drugs: both nicotine and marijuana act as bronchial irritants. Patients with cystic fibrosis are exposed to an enormous amount of passive smoking, as active smoking occurs in more than 50% of their families, usually by the parents[13].

PROBLEMS THAT CAN ARISE IN THE FIRST YEAR OF TRANSFER

It is essential that the adult CF team make the new patient welcome to the unit at time of transfer. On the Manchester unit we consider that it usually takes about a year for patient and team to get used to each other. A smooth transfer will often forestall future problems. Joint clinics prior to transfer will help familiarize the patient with future carers; prior to transfer the patient should know as much as possible about the workings of the unit. At the Manchester unit each adult patient receives a comprehensive introductory package which includes a handbook, a contact card with phone numbers of all the team members, and a recent copy of the patient-led news bulletin (Figure 1). Recently we have introduced a website for the unit. Each patient should be shown around the unit and introduced to as many staff as possible. Each patient will usually have their own

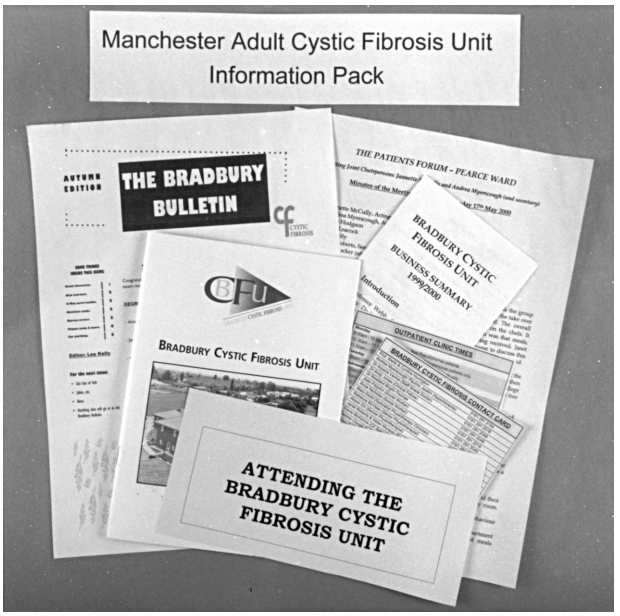

Figure 1 **Information provided for the new CF patient prior to transfer**

room, some facilities for recreation and a designated kitchen.

Most adult units will have protocols for various aspects of care, with which the patient can become familiar. At the Manchester unit patients are given an out-of-clinic appointment for their first two visits; this gives time in a relaxed atmosphere to discuss future care plans.

Although the adult and paediatric teams are very concerned that the transfer process should be a success, it is comforting to know how positive the patients are about transferring[14]. Interestingly, they are most concerned about exposure to infection, and the features identified by patients as important in terms of information were education about adult CF issues, ready phone access to a nurse, and special care with obstetric/fertility issues and transplantation.

Despite the best of intentions, problems often arise during the first year. Maturity at time of transfer will vary enormously between patients; however, upon transfer the patient will become the youngest rather than the oldest person on the unit. The age range of patients on a large adult unit is 16–50 years, with a mean of 25–28, and many CF adults are now older than their carers. Older patients can influence the new annual intake for both better and worse. They may corrupt younger patients in their behaviour patterns, introducing them to a wilder social life. This pattern may continue out of hospital. Smoking of nicotine and cannabis is relatively common in CF patients, and undoubtedly this can influence the younger generation.

Perhaps the most difficult change for the new adult patient after transfer is taking responsibility for their own care. Compliance with a demanding regimen of self-care has always been a challenge for the most sensible of patients[15]. With the parents no longer dictating self-care and the competition of further education and an expanding social life, it is not surprising that the health of CF patients often declines in adolescence.

Sometimes when new patients are admitted to the ward, older patients ridicule their level of compliance. Parents will comment that compliance has only deteriorated since the patient transferred to the adult unit, and may blame the centre for not being strict enough with their children. During the first year it is very important that the team tailors care and maintains compliance by close attention to details.

Perhaps the most difficult aspect for the younger CF patient on being admitted to the adult ward is the sudden realization that significant numbers of the adult population are chronically unwell, needing a transplant, and each year, some, with whom they may have become good friends, will die. Out of loyalty CF patients will often attend each others' funerals, with the sharpened realization that they will die of the same disease as their friends. Some handle this situation with surprising stoicism, but others, not surprisingly, find it very difficult. The presence in the Manchester unit of a clinical psychologist experienced in CF care has provided enormous help with the fears and anxieties of our patients.

It is not always appreciated by the adult team that parents often feel deprived once they are no longer involved in the continuing care of their children: they may feel deliberately marginalized. Most CF teams do not mind the continuing involvement of parents: an extra dimension of knowledge is often added about compliance; subjects are introduced by parents which the patient is too shy to broach. In reality, the separation from parents is usually made by the maturing adult, who wants to be completely independent.

Many years ago a mother stood up in a public meeting and stated: 'You have separated me from the care of my child'. The child was a 25-year-old married woman!

Sometimes neither the paediatric nor the adult team can know of what might be considerable strife going on in a family. Clearly this can take many forms, and marriage breakdown is relatively common among parents of CF children. Family background can involve crime, incest, neglect, indifference and alcoholism. It is not surprising that a disruptive background can manifest itself with behavioural problems after transfer to the adult unit.

Many years ago, a newly referred female with alcoholic parents accused patients and staff of sexual harassment, and said that she had been raped by a male nurse at her paediatric hospital. Neither fact was correct, but required skilled handling by an experienced social worker to defuse a potentially difficult situation.

Criminal involvement, particularly with hard drugs, can make life extremely difficult for a CF unit, not least because

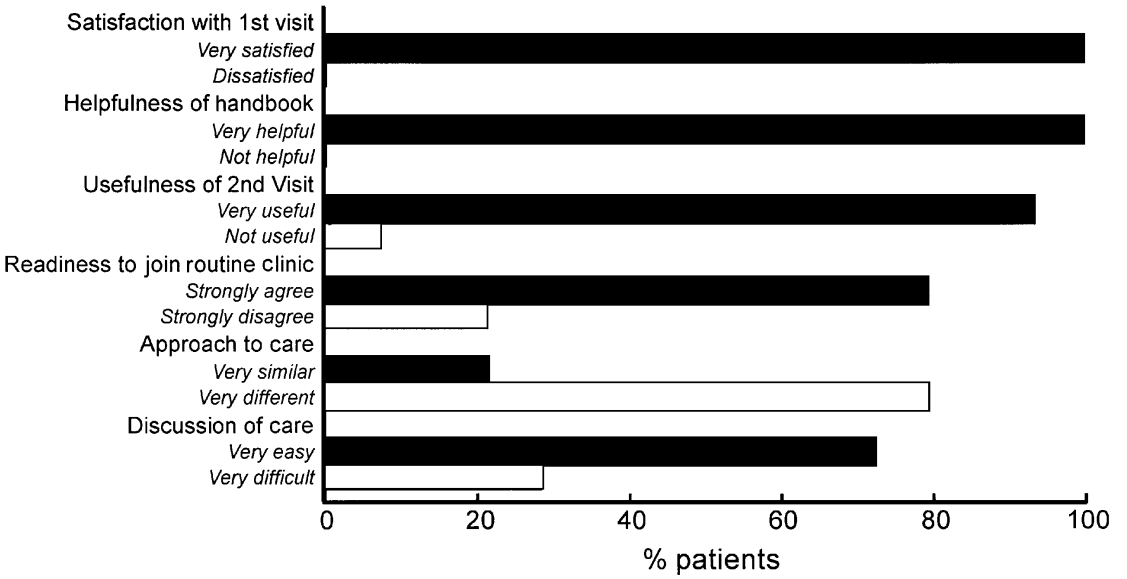

Figure 2 **Replies to a post-transfer satisfaction questionnaire**

of the criminal element that may accompany the patient. Criminal partners will threaten both staff and patients, and it may be necessary to take legal advice as to whether care can continue in such cases.

Good career advice is essential for 16–18-years-old CF patients when they are trying to organize their future. Supporting the patient with advisory letters to potential employers is a sensitive issue: should they tell them about their cystic fibrosis or conceal it? Getting it wrong may prejudice work prospects. Social work support during this period is invaluable.

Many problems can arise during the first year of transfer to adult CF care. However, the majority of patients adapt very well, as is shown by their positive responses to a post-transfer questionnaire (Figure 2).

CONCLUSION

It is only too easy for the adult team to blame the paediatric team for any problems that arise following the transfer of an adolescent, but it should be remembered that the adult CF teams only exist because of the superb care delivered by paediatric teams. The adult team receives a patient whose priorities are changing fast: freed from parental constraints, health may take second place to myriad more interesting distractions.

'On a Friday morning recently a mother rang about her son. Very worried about Paul, who has blood in his sputum, vomiting and breathless, and back pain. She feels it's life-threatening but he won't come in until after Monday, because he has got his driving test.'

The lives of patients with cystic fibrosis are changing rapidly at the time they transfer to adult care. It is part of the job description of an adult CF team to take on the mixture of healthcare and consequent problems that will arise during the first few years of life of the new patient on the adult unit.

REFERENCES

1 Dodge JA, Morison S, Lewis PA, *et al*. Incidence, population, and survival of cystic fibrosis patients in the UK, 1968–95. UK Cystic Fibrosis Survey Management Committee. *Arch Dis Child* 1997;**77**:493–6

2 Elborn JS, Shale DJ, Britton JR. Cystic fibrosis: current survival and population estimates of the year 2000. *Thorax* 1991;**46**:881–5

3 Mahadeva R, Webb AK, Westerbeck RC, *et al*. Clinical outcome in relation to care in centres specialising in cystic fibrosis: cross sectional study. *Br Med J* 1998;**316**:1771–5

4 Conway S. Transition from paediatric to adult-oriented care for adolescents with cystic fibrosis. *Disability Rehab* 1998;**20**:209–16

5 Cystic Fibrosis Research Trust. *Transition from Paediatric to Adult Care: A Guide for Purchasers, Providers and Clinical Teams*. London: Cystic Fibrosis Research Trust, 1997

6 Madge S, Carr SB. Is there a need for a dedicated cystic fibrosis adolescent outpatient service. *Pediatr Pulmonol* 1996;**13**(suppl):338

7 Haworth CS, Selby PL, Webb AK, *et al*. Low bone mineral density in adults with cystic fibrosis. *Thorax* 1999;**54**:961–7

8 Conway SP, Morton AM, Oldroyd B, *et al*. Osteoporosis and osteopenia in adults, adolescents with cystic fibrosis: prevalence and associated factors. *Thorax* 2000;**55**:798–804

9 Milla CE, Warwick WJ, Moran A. Trends in pulmonary function in patients with cystic fibrosis correlate with degree of glucose intolerance at baseline. *Am J Respir Crit Care Med* 2000;**162**:891–5

10 Cystic Fibrosis Foundation National Patient Registry. *1997 Annual data report*. London: CFF, 1998

11 Sawyer SM. Reproductive health in young people with cystic fibrosis. *Curr Opin Pediatr* 1995;**7**:376–80

12 Fair A, Griffiths K, Osman LM, *et al*. Attitudes to fertility issues among adults with cystic fibrosis in Scotland. *Thorax* 2000;**55**:672–7

13 Gillam H, Stenlund C, Ericsson-Hollsing A, Strandvik B. Passive smoking in cystic fibrosis. *Resp Med* 1990;**84**:289–91

14 Boyle MP, Nosky ML. Strategies for improving transition to adult care based on patients' views. *Pediatr Pulmonol* 2000;**S20**:547

15 Abbott J, Dodd M, Bilton D, Webb AK. Treatment compliance in adults with cystic fibrosis. *Thorax* 1994;**49**:115–20