Original articles

Hospital care for adults with cystic fibrosis: an overview and comparison between special cystic fibrosis clinics and general clinics using a patient questionnaire

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Abstract

Background – Provision of medical care for adult patients with cystic fibrosis is an increasing problem as the number of patients surviving into adulthood increases. Recent reports have suggested that care is best provided in specialist centres because of longer survival. Recent changes in the National Health Service funding and delivery of service may adversely affect the provision of such a specialist service. The aim of this study was to assess the current pattern of medical service received by adults with cystic fibrosis and to compare the type of care between special cystic fibrosis and general clinics.

Methods – Confidential postal questionnaires were sent to all 1052 members of the Association of Cystic Fibrosis Adults (ACFA) comprising 59% of the UK population of cystic fibrosis patients over 15 years and 80% over 25 years of age. The response rate was 82%.

Results – Two thirds of patients were attending special cystic fibrosis clinics for either adults or adults and children. There were significant differences in the proportion of patients using special cystic fibrosis clinics between regions but not between social class groups. Significant differences between cystic fibrosis and general clinics were noted. Patients attending cystic fibrosis clinics were more likely to have had simple clinical investigations (blood tests, sputum culture, oxygen saturation, chest radiography, weight and lung function measurement) in the previous year. They were also more likely to have received intravenous antibiotics at home, and to have access to paramedical personnel. Patients attending cystic fibrosis clinics were taking higher doses of pancreatic enzyme supplements with respect to quantity and potency of preparation. Such patients also had less severe symptoms irrespective of social class, and were more likely to be satisfied with professional aspects of their care. Regardless

of type of clinic, potential deficiencies were identified in overall medical care with omission of clinical investigations in severely affected patients and evidence of undertreated respiratory and digestive symptoms in patients with moderate and severe disease.

Conclusions – This survey provides evidence that adults with cystic fibrosis attending special cystic fibrosis clinics receive more intensive care, have better symptom control, and are more satisfied with the service provided than those attending general clinics.

(Thorax 1994;49:300-306)

Improvements in survival from cystic fibrosis have resulted in a sustained increase in the prevalence of the disease by approximately 100 patients a year. Most of this increase is in adults,¹ and already over a quarter of patients are over the age of 30.² Recent reports suggest that prevalence and prognosis will continue to improve.^{2,3} The implication of these observations is that health care services for patients with cystic fibrosis, and particularly adults, will need to expand.

The best way to provide appropriate medical care has, however, been the subject of debate in the UK. In countries where care is organised almost exclusively in specialist centres reported survival rates are superior to those of the UK,⁴⁻⁶ suggesting that resources should be invested in specialist clinics. In 1988 only 51% of all patients with cystic fibrosis in the UK attended what the British Paediatric Association defined as a "specialist centre."² Although potentially confounded by effects of social class, survival in these patients was greater than in those attending general clinics.^{7,8}

This evidence prompted both the British Paediatric Association and the Royal College of Physicians to recommend that care should take place in specialist centres, funded by regional health authorities.^{1,2} These would function as treatment centres and sources of expertise and education where care is shared with a local hospital. In addition, the Royal

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Reprint requests to:
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Received 15 February 1993
Returned to authors
18 November 1993
Revised version received
9 December 1993
Accepted for publication
20 December 1993

College of Physicians specified the need for a systematic approach to the development of standards of care, setting of quality targets, and allocation of resources.¹ However, progress towards establishing this model of care for adult patients with cystic fibrosis has been slow. Furthermore, the provision of specialist care is expensive⁹ which, in the light of recent reforms to the National Health Service outlined in *Working for patients*,^{10,11} could act as a disincentive to the purchasing of specialist care for adult patients with cystic fibrosis and further compromise rational service planning and development.

This survey was designed to document the demographic and social characteristics of adults with cystic fibrosis in the UK and, in particular, to identify the medical services they received. Additional aims were to compare the delivery of treatment and patient satisfaction between special cystic fibrosis clinics and non-cystic fibrosis clinics, and to serve as a baseline against which to monitor future changes in hospital care resulting from health service reforms. The social and demographic characteristics of the patients have been published separately,¹² and this paper presents details of the provision of medical care.

Methods

PATIENTS

Membership of the Association of Cystic Fibrosis Adults (ACFA) extends to approximately 68% of the UK population with cystic fibrosis aged over 16 years, and to over 80% of those over 25 years of age. A confidential self administered postal questionnaire was sent to 1052 eligible ACFA members known to be over 16 years of age on 1 July 1990, and followed by two reminders. The sample included patients attending large special cystic fibrosis clinics and general clinics at local hospitals.

DEFINITION OF CYSTIC FIBROSIS AND GENERAL CLINICS

Those attending clinics exclusively for patients with cystic fibrosis were defined as attending cystic fibrosis clinics, and those who did not were defined as attending general clinics.

QUESTIONNAIRE

The multiple choice questionnaire included questions covering social and demographic information, primary and secondary hospital care, and medication. Severity of disease was estimated using a standard symptom questionnaire with a summary score calculated as the average of five symptoms (breathlessness, cough, sputum, abdominal discomfort, fatigue) each rated on a scale of one to five. This was internally validated against other measures of disease severity such as percentage ideal weight for height, annual sick leave from work, and annual time spent in hospital. A score of ≤ 2.0 represented mild symptoms, 2.1–3.4

represented moderate symptoms, and ≥ 3.5 represented severe symptoms.

Patients were asked to identify the hospital they attended and the type of doctor (paediatrician, chest physician, etc) as well as whether the clinic they attended was only for patients with cystic fibrosis (adults or children) or was a general clinic treating a number of medical or paediatric conditions. They were also asked to recall their contact with paramedical personnel, their current medication, and investigations which had been performed in the previous year at the hospital, reported in the questionnaire. Although there was no direct access to patient records to validate the responses, adults with cystic fibrosis attending hospitals with large cystic fibrosis clinics correctly identified them in 91% of cases.

Patient satisfaction was assessed using several professional and non-professional aspects of care together with an overall rating.

DATA ANALYSIS

Data were analysed by the Epi-Info statistical package and the BMJ Confidence Intervals Analysis package using, where appropriate, χ^2 , Mantel-Haenszel, analysis of variance, and confidence intervals for single proportions and the difference between proportions. Not all respondents answered all questions, and analysis for each question is confined only to those who made valid responses.

Results

The response rate was 82.3% ($n = 866$), representing 56% of the total number of adults with cystic fibrosis in the UK.

TYPE OF HOSPITAL CARE

A total of 669 patients (82%) identified a hospital they were attending for their care. Of these 411 (61%) were attending at least one hospital which provided a large clinic for patients with cystic fibrosis in a major city. More than one hospital was named by 21% of patients, indicating the development of shared care arrangements, usually between local hospitals and special cystic fibrosis clinics.

Using the definition given above, 494 of 746 patients (66%) were attending a cystic fibrosis clinic and 252 (34%) were attending a general clinic (table 1). There were social class differences in both type of clinic attended and type of doctor seen, largely because of the higher proportion of patients from manual social classes who were still attending cystic fibrosis specialist paediatricians. In this study cystic fibrosis clinics included those for adults, children, or both, which removed the difference due to social class leaving only the balance between adult and paediatric care (table 1).

There was a difference in disease severity between patients attending cystic fibrosis clinics and general clinics (mean symptom score 2.33 for cystic fibrosis and 2.51 for general clinics, $p < 0.005$ Mann-Whitney U

Table 1 Type of hospital doctor seen and type of clinic attended by adult cystic fibrosis questionnaire respondents, by social class group

	Overall number (%)	Manual social class number (%)	Non-manual social class number (%)	95% CI for difference between manual and non-manual	Mean symptom score of group
Type of doctor seen					
Chest physician specialising in cystic fibrosis	482 (65.2%)	111 (57.5%)	363 (68.0%)	2.5% to -18.5%*	2.40
Non-specialist chest physician	124 (16.8%)	32 (16.6%)	89 (16.7%)	-6.0% to 6.0%	2.51
Non-specialist general physician	31 (4.2%)	13 (6.7%)	18 (3.4%)	-0.4% to 7.2%	2.47
Paediatrician specialising in cystic fibrosis	70 (9.5%)	26 (13.5%)	43 (8.1%)	0.01% to 10.8%*	2.22
Non-specialist paediatrician	18 (2.4%)	6 (3.1%)	12 (2.2%)	1.9% to 3.4%	2.26
Other	14 (1.9%)	5 (2.6%)	9 (1.7%)	1.6% to 3.4%	2.17
Total	739	193	534		
Type of clinic attended					
For cystic fibrosis adults only	376 (50.4%)	85 (43.8%)	282 (52.4%)	-16.8% to -4.4%*	2.37
For cystic fibrosis adults and children	118 (15.8%)	35 (18.0%)	81 (15.1%)	-3.2% to 9.2%	2.23
For patients with all sorts of medical conditions	222 (29.8%)	65 (33.5%)	155 (28.8%)	-2.9% to 12.4%	2.52
Don't know	18 (2.4%)	8 (4.1%)	9 (1.7%)	1.5% to 5.4%	2.40
Other, e.g. attend ward or do not attend hospital	12 (1.6%)	1 (0.5%)	11 (2.0%)	-3.1% to 0.03%	2.40
Total	746	194	538		
All special cystic fibrosis clinics	494 (66.2%)	120 (61.9%)	363 (67.5%)	-13.5% to 2.3%	2.33
All general clinics	252 (33.8%)	74 (38.1%)	175 (32.5%)	-2.3% to 13.5%	2.49

* $p < 0.05$ for difference between manual and non-manual groups.

Figure 1 Proportion of cystic fibrosis adult questionnaire respondents attending special cystic fibrosis clinics by RHA or country of residence.

Table 2 Clinical investigations performed on adult cystic fibrosis questionnaire respondents in the previous year: a comparison of those attending general and special cystic fibrosis clinics. Blood gases include measurement of transcutaneous oxygen saturation

Type of investigation	Overall number (%)	General clinics number (%)	Cystic fibrosis clinics number (%)	95% CI for difference between cystic fibrosis and general clinics	χ^2_{MH} stratified by symptom severity for difference between cystic fibrosis and general clinics
Blood tests	645 (85.8%)	208 (82.7%)	437 (87.4%)	-10.4% to 0.65%	2.32 ($p > 0.1$)
Blood gases	366 (48.7%)	89 (35.3%)	279 (55.7%)	-27.8% to -13.1%*	23.5 ($p < 0.001$)*
Lung function	579 (77.0%)	186 (73.9%)	393 (78.6%)	-11.3% to 1.72%	1.57 ($p > 0.1$)
Sputum culture	653 (86.9%)	207 (81.6%)	449 (89.7%)	-13.1% to 2.4%*	8.88 ($p < 0.01$)*
Weight	702 (93.4%)	224 (89.0%)	479 (95.7%)	-11.2% to -2.7%*	5.61 ($p < 0.02$)*
Chest radiograph	683 (90.8%)	219 (86.9%)	464 (92.9%)	-10.6% to -1.2%*	4.96 ($p < 0.05$)*
n	752	252	500		

* $p < 0.05$.

test). This was mainly because of differences within the non-manual social class group (mean score 2.27 cystic fibrosis clinics, 2.45 general clinics, $p < 0.02$ Mann-Whitney U test), and not to differences in the social class composition of the two groups.

The proportion of patients attending cystic fibrosis clinics also differed between regional health authorities (fig 1). There were differences in distance travelled to hospital; 117 of 493 (24%) patients attending cystic fibrosis clinics had to travel over 50 miles compared with only 19 of 248 (8%) of those attending general clinics (95% CI for difference 11.1% to 21.1%).

OUTPATIENT CARE

Those patients attending clinic more frequently had more severe disease, regardless of clinic type. Patients attending general clinics were more likely to attend very frequently (once a month) (45 of 249 (18%) attending general clinics compared with 52 of 493 (11.2%) attending cystic fibrosis clinics, 95% CI for difference 2.03% to 13.0%), or infrequently (every 5–6 months) (37 of 249 (14%) attending general clinics compared with 39 of 493 (8%) attending clinics, 95% CI for difference 1.9% to 12.0%). A higher proportion of those attending cystic fibrosis clinics had direct access to the consultant or cystic fibrosis team without first having to consult their general practitioner (474 of 485 (98%) compared with 193 of 226 (85%) in general clinics, 95% CI 7.53% to 17.1%).

A higher proportion of patients attending cystic fibrosis clinics had sputum culture, blood gases, oxygen saturation, weight and chest radiographs performed in the previous year than those attending general clinics (table 2). Such differences were irrespective of disease severity.

PARAMEDICAL PERSONNEL

Those patients attending cystic fibrosis clinics were more likely to have seen paramedical personnel (table 3) irrespective of disease severity (χ^2_{MH} for dietitian 9.00, $p < 0.005$; physiotherapist 21.20, $p < 0.001$; medical social worker 8.74, $p < 0.005$). The mean symptom score was significantly greater in those who had seen both dietitians and physiotherapists regardless of clinic type. Patients who had recently seen a dietitian were more often receiving food supplements (301 of 462

Table 3 Paramedical personnel seen by adult cystic fibrosis questionnaire respondents in the previous year: a comparison between those attending cystic fibrosis clinics and general clinics

Person seen	Overall number (%)	General clinics number (%)	Cystic fibrosis clinics number (%)	95% CI for difference between cystic fibrosis and general clinics
Dietitian	462 (61.7%)	137 (54.4%)	322 (65.3%)	-18.4% to -3.5%*
Physiotherapist	543 (72.5%)	157 (62.3%)	383 (77.7%)	-22.4% to -8.4%*
Medical social worker	180 (24.0%)	46 (18.3%)	133 (27.0%)	-14.9% to -2.55%*
Cystic fibrosis clinical nurse specialist	144 (19.2%)	20 (7.9%)	123 (24.9%)	-22.1% to -11.9%*
n	749†	252	493	

* $p < 0.05$.

† Some patients not attending any clinics included in overall total.

(65%) *v* 82 of 287 (29%), 95% CI difference 29.8% to 43.3%) and to have received advice to increase fat intake (278 of 455 (61%) *v* 88 of 276 (32%), 95% CI difference 22.1% to 36.3%) irrespective of disease severity (Mantel-Haenszel χ^2 for both $p < 0.001$). Patients who had recently seen a medical social worker were more likely to have applied for mobility allowance (113 of 162 (67%) *v* 192 of 492 (39%), 95% CI difference 19.9% to 36.5%) and attendance allowance (113 of 162 (70%) *v* 203 of 490 (42%), 95% CI difference 20% to 36.8%) irrespective of disease severity, with no change in the refusal rate.

INPATIENT CARE

Admission to hospital had not been required for 379 of 747 (51%) patients in the last year. The mean (SD) number of hospital admissions was 1.65 (2.77). There were no differences in the frequency of hospital admissions between clinic types or social class groups. Inpatient facilities differed with more cystic fibrosis clinics providing single rooms for patients with cystic fibrosis (302 of 448 (67%) *v* 46 of 222 (21%), 95% CI difference 39.8% to 53.6%) and having activity rooms (198 of 427 (46%) *v* 34 of 214 (16%), 95% CI 23.7% to 37.3%). Patients admitted from cystic fibrosis clinics were more likely to be allowed to keep and administer their own medication (359 of 411 (87%) *v* 141 of 199 (70%), 95% CI 9.4% to 23.6%).

HOME INTRAVENOUS ANTIBIOTIC THERAPY

Those attending cystic fibrosis clinics received more self administered intravenous therapy at home (260 of 491 (53%) *v* 78 of 252 (31%), 95% CI difference 14.8% to 29.2%). This difference was independent of differences in disease severity (Mantel-Haenszel χ^2 32.39, $p < 0.001$). Those receiving home intravenous therapy had more severe disease regardless of type of clinic attended (symptom score 2.52 *v* 2.30 in those not receiving home intravenous therapy, 95% CI difference 0.116 to 0.322), and spent more time in hospital in the preceding year, with 95 of 339 (28%) patients who received home intravenous therapy spending four weeks or more in hospital compared with 54 of 408 (13%) patients not receiving home intravenous therapy (95% CI difference 8.99% to 20.6%). Home intravenous antibiotic therapy does not appear to prevent hospital

admission but is used as an adjunct to hospital treatment in a group with more severe disease.

MEDICATION AND DIET

The dose of pancreatic enzymes taken with meals and snacks was greater in cystic fibrosis than in general clinics, and stratified analysis showed this to be independent of disease severity (Mantel-Haenszel χ^2 $p < 0.05$). The mean dose with meals was 6.7 capsules in patients attending cystic fibrosis clinics and 5.50 in those attending general clinics (95% CI difference 0.34 to 1.76). Comparable mean doses for snacks were 2.97 and 2.36 capsules respectively (95% CI 0.12 to 0.91). The dose of pancreatic enzymes taken was associated with perceived severity of digestive disease ($p < 0.001$, χ^2 test) and negatively correlated with percentage predicted weight for height ($r = -0.14$, 95% CI -0.21 to -0.07).

Of those attending cystic fibrosis clinics 454 of 494 (88%) were taking microsphere pancreatic preparations as opposed to older less effective preparations, compared with 225 of 252 (81%) at general clinics (95% CI difference 0.7% to 9.7%), and this difference was independent of disease severity (Mantel-Haenszel χ^2 4.78, $p < 0.05$). Patients attending cystic fibrosis clinics and those taking microsphere pancreatic enzyme preparations had slightly greater body weight, but comparisons are likely to be confounded by patients with more severe digestive problems preferentially receiving microsphere pancreatic enzyme preparations. Newer high lipase preparations were not available at the time of this survey.

There was no difference between cystic fibrosis clinics and general clinics in the proportion restricting dietary fat intake (126 of 490, 26%), or in the proportion who had been recently advised to increase their fat intake (363 of 728, 50%). However, patients attending general clinics were more likely to restrict their fat intake because of unacceptable symptoms rather than other reasons (44 of 252 (18%) compared with 54 of 494 (11%) at cystic fibrosis clinics, 95% CI 1.1% to 12.0%). Of the adults with cystic fibrosis 13% inappropriately restricted their fat intake as a result of general health promotion messages aimed at the non-cystic fibrosis population. A proprietary food supplement was taken by 368 of 746 (49%) patients, but there was no difference between cystic fibrosis and general clinics.

Nebulised antibiotics were taken by 256 of 746 (34%) patients with or without bronchodilator, and the proportion was slightly greater in those attending cystic fibrosis clinics. The use of nebulised medication was related to severity of symptoms of cough, breathlessness, and sputum production (χ^2 $p < 0.001$), and also to the patient's own perception of chest disease as severe.

SATISFACTION WITH HOSPITAL CARE

Overall, 156 of 686 (23%) patients did not rate their care as good (table 4). In general the professional aspects of care received a more

Table 4 Satisfaction of adult cystic fibrosis questionnaire respondents with various aspects of hospital care: a comparison between general and special cystic fibrosis clinics

Aspect of care	Grade given number (%)					Mean grade		95% CI difference between cystic fibrosis and general clinics	n
	1 (very poor)	2 (poor)	3 (fair)	4 (good)	5 (very good)	General clinics	Cystic fibrosis clinics		
Hospital accommodation	12 (1.9%)	39 (6.2%)	205 (32.5%)	238 (37.7%)	135 (21.4%)	3.64	3.74	-0.26 to 0.06	636
Hospital food	101 (16.1%)	146 (23.3%)	216 (34.5%)	123 (19.6%)	36 (5.8%)	2.73	2.76	-0.22 to 0.16	631
Consultant's knowledge of cystic fibrosis	5 (0.7%)	9 (1.3%)	49 (7.1%)	125 (18.2%)	498 (72.6%)	4.33	4.74	-0.53 to -0.29*	690
Consultant's understanding of your problems	12 (1.8%)	32 (4.7%)	105 (15.4%)	197 (28.8%)	337 (49.3%)	3.93	4.31	-0.54 to -0.23	687
Junior doctors' understanding of cystic fibrosis	20 (3.1%)	71 (11.1%)	220 (34.3%)	226 (35.3%)	101 (15.8%)	3.13	3.65	-0.68 to -0.36*	645
Nurses' understanding of cystic fibrosis	17 (2.6%)	50 (7.6%)	180 (27.4%)	245 (37.1%)	133 (19.9%)	3.27	3.93	-0.81 to -0.51*	662
Physiotherapy advice you receive	8 (1.3%)	20 (3.1%)	79 (12.4%)	223 (35.0%)	297 (46.6%)	3.97	4.27	-0.47 to -0.13*	642
Dietary advice you receive	29 (4.9%)	39 (6.6%)	125 (21.2%)	195 (33.1%)	182 (30.8%)	3.23	3.86	-0.83 to -0.36*	694
Social work advice you receive	72 (19.7%)	31 (8.5%)	83 (22.7%)	84 (23.0%)	52 (14.2%)	2.24	2.89	-1.00 to -0.30*	369
Overall rating of hospital care	12 (1.8%)	24 (3.5%)	120 (17.6%)	278 (40.8%)	247 (36.3%)	3.76	4.20	-0.58 to -0.29*	686

* $p < 0.05$.

Figure 2 Cystic fibrosis adult questionnaire respondents' perceptions of the change in medical care they have received over the past five years – general and cystic fibrosis clinics compared. Overall $p < 0.001$, χ^2 .

favourable rating than the hotel aspects, in particular hospital food. There were differences between cystic fibrosis and general clinics for professional but not hotel aspects of care. Patients attending cystic fibrosis clinics were more likely to report that their care had improved over the previous five years (fig 2).

Discussion

This survey was planned to answer two questions: (1) to what extent do adult patients with cystic fibrosis receive the type of care recommended by the Royal College of Physicians,² and (2) in what ways do the processes of medical care and satisfaction with medical care differ between patients attending special cystic fibrosis clinics and general clinics?

SAMPLE AND BIAS

Although a high proportion (69%) of adults with cystic fibrosis in the UK are known to ACFA (UK), this sample is still likely to be biased towards those attending large cystic fibrosis clinics where membership tends to be encouraged. Responders were more likely to be female, and although there might be an overall bias in responders towards those of higher social and educational status, in the responders there was no social class bias in

those attending cystic fibrosis clinics. It remains likely that this survey overestimates the proportion of patients attending special clinics, and underestimates the differences in care between clinic types.

The internal validity of responses was high. Although the type of care received relied on recall by patients in this study, adults with cystic fibrosis are generally very aware of their disease, the types of treatment and investigations required, and are responsible for a complex daily regimen of care which requires them to be fully aware of their medication. Most patients have many years' experience of hospital treatment and care. It is possible that socially disadvantaged patients attending general clinics are less accurate in response or recall, but this could not be evaluated.

ACCESS TO SPECIALIST CARE

The British Paediatric Association and the Royal College of Physicians have recommended specialist centres as the basis of medical care for patients with cystic fibrosis.¹² This survey identified a substantial minority of patients who were not receiving specialist care through cystic fibrosis clinics and significant inequalities in access to such services throughout the UK. One quarter of patients in this sample did not attend a cystic fibrosis specialist doctor or clinic. Patients from manual social classes and in certain areas of the country have lower access, particularly to services for adult patients. Some patients attending special cystic fibrosis clinics still attended paediatricians, which may not be appropriate either for the adult patients or other children on the paediatric wards.

PROCESS OF MEDICAL CARE

This study highlights differences in the delivery of medical care between cystic fibrosis and general clinics. The care given in cystic fibrosis clinics appears to be more intensive, with more direct access to medical advice, more access to home intravenous therapy, more access to dietitians, physiotherapists and other paramedical personnel, more access to basic investigations to monitor progress, and a higher level of therapy, particularly with pan-

creatic enzyme supplements. Such differences were independent of overall disease severity. Cystic fibrosis clinics also had more amenities such as activity rooms, and were more likely to allow adult patients to keep and administer their own medication. Patients attending general clinics were more likely to see the consultant, which probably reflects the increased availability of dedicated junior medical staff and research fellows in cystic fibrosis clinics, and did not have any impact on patient satisfaction.

Whilst there is evidence that care in both clinic types is being directed towards those in most need, this study has shown that there are patients who could benefit from more treatment or care, but are not currently receiving it. For example, in the group of patients who were under 85% predicted weight for height, 38 were taking fewer than four enzyme capsules a meal and 41 had not seen a dietitian in the last year.

Many of these process measures relate to aspects of care considered desirable either by learned bodies,² individual physicians,¹³ or the adult patients themselves.^{14,15} While not all clinical investigations or treatments are appropriate for all patients, it seems reasonable to expect that basic investigations such as weight, lung function testing, chest radiography, blood tests, oxygen saturation and sputum culture should be performed in the course of a year. Many patients, even those with severe symptoms, could not recall these being performed in the last year.

OUTCOME OF MEDICAL CARE

It is not possible to determine outcome from a single survey such as this, although a follow up study of this cohort is planned in 1994–5, four years after the survey reported here, to assess this more fully. Patients attending specialist clinics had less severe symptoms irrespective of social class, which may reflect more intensive monitoring, treatment and access to specialist personnel, although this difference may be due to psychological as well as physical benefits. There is also evidence that those attending cystic fibrosis clinics have better control of digestive symptoms, avoiding the need for dietary fat restriction because of unacceptable symptoms. Although the lower symptom score in patients attending cystic fibrosis clinics may reflect self selection of patients – patients living a long way from such a clinic may only be able to undertake the journey if they are fit enough – one might also expect the more severely affected individuals to attend cystic fibrosis clinics as tertiary referrals.

PATIENT SATISFACTION

Patients attending cystic fibrosis clinics were more satisfied with the professional aspects of their care and perceived their care as having improved over the previous five years. Adult patients with cystic fibrosis frequently have good understanding of their disease and are critical of poor standards of knowledge or care.

Overall, over one third did not rate the hospital care they received as good, with particular dissatisfaction expressed with hospital food and social work advice.

HOSPITAL CARE FOR ADULTS WITH CYSTIC FIBROSIS

Expert consensus supports care for adults with cystic fibrosis at specialist centres with defined quality standards,¹² a view shared by most patients themselves,¹⁵ and supported by this study. Whilst such care is available to most patients, there are still substantial numbers for whom it is not. This study has identified inequalities in access to cystic fibrosis clinics, differences in intensity of care between cystic fibrosis and general clinics, and evidence that some patients are receiving suboptimal care.

Although it may be desirable, care which takes place wholly within specialist cystic fibrosis clinics has potential disadvantages. Some adults with cystic fibrosis prefer locally based treatment. Patients have to travel further to attend special cystic fibrosis clinics. Concentration of care in a single centre may lead to reduction in general awareness in other hospitals of cystic fibrosis in adults. If care is centralised there need to be mechanisms for disseminating expertise to local clinicians so that the centre acts as a resource for the region it serves. This might be achieved by the use of formal shared care arrangements with local hospitals, and educational programmes.

Regional health authorities have an average of 100–150 resident adult patients with cystic fibrosis, whereas district health authorities have only 10–20. With such small numbers, specific purchasing of care by district health authorities may be difficult. Leaving their care to the extracontractual referral mechanism may lead to further difficulties in access to specialist clinics and may compromise the planning and development of specialist services. This study has shown some evidence that care in special cystic fibrosis clinics may be of benefit to patients, with improved symptom control and more intensive treatment and monitoring. Whether the latter confers significant clinical benefit and improves social outcome requires prospective evaluation in the planned follow up survey. Patients are certainly more satisfied with this type of care. If purchasing decisions are to be made in favour of cystic fibrosis clinic care, patient numbers suggest that this might be most appropriately done above single district level.

This study provides evidence that a substantial minority of adults with cystic fibrosis still do not receive care in special cystic fibrosis clinics as recommended by the Royal College of Physicians.² There is evidence that the type of care differs between special cystic fibrosis clinics and general clinics, with greater intensity of treatment and investigation, greater access to paramedical team members, and better symptom control in those attending special cystic fibrosis clinics. Patient satisfaction is higher in those attending cystic fibrosis clinics. This provides supporting evidence for

the concept of specialist centres of treatment, and highlights areas for improvement, although the question of whether specialist care confers benefit in terms of clinical status, social outcome, and survival remains to be answered. It is hoped that those responsible for purchasing health care, and those who provide care for adults with cystic fibrosis, will recognise the challenges it poses to provide high quality care for all adult patients with cystic fibrosis.

This study was funded by the Cystic Fibrosis Research Trust. The authors would like to thank Sarah Sweetland at the Cystic Fibrosis Research Trust for help in administering the questionnaire, Laurence Skermer for help with coding and entering of data for the questionnaire, Dr Estelle Gilman, Regional Epidemiologist at West Midlands Regional Health Authority for statistical advice, and the Association of Cystic Fibrosis Adults for permission to use their mailing list and for encouraging a high response rate.

A copy of the questionnaire can be provided on request.

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