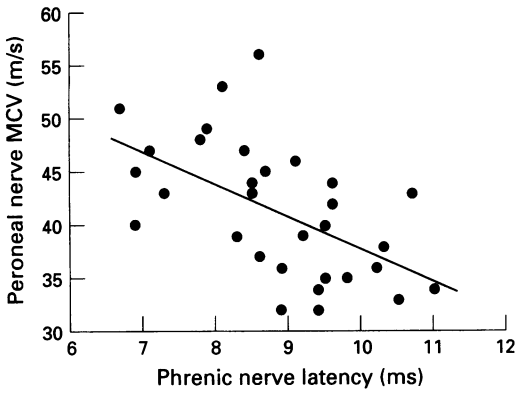

Correlation of right phrenic nerve latency and peroneal nerve motor conduction velocity (MCV) in patients on haemodialysis (n=31); r=−0·56, p<0·001.

peroneal motor conduction. A significant relation existed between these two variables ($r=-0{\cdot}56$, $p<0{\cdot}001$) as shown in the figure.

Discussion

To the best of our knowledge this is the first study to demonstrate the occurrence of phrenic neuropathy in patients on chronic haemodialysis. Phrenic nerve involvement has been described in several neuropathies including critical illness polyneuropathy, the Guillain-Barré syndrome, brachial neuritis, hereditary motor and sensory neuropathy type 1, porphyria, leprosy, and Waldenström's macroglobulinaemia.[9 10]

All our patients with delayed phrenic nerve latency showed electrophysiological signs of peripheral neuropathy. In addition, delayed phrenic nerve latencies correlated significantly with pathological peroneal nerve conduction velocity.

Since inspiratory muscle dysfunction is a frequent clinical feature in uraemic patients[2] and phrenic nerve neuropathy is known to contribute to diaphragmatic weakness,[3] the detection of phrenic nerve involvement is of particular interest.

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Different perceptions of disease severity and self care between patients with cystic fibrosis, their close companions, and physician

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Abstract

***Background* – An investigation was carried out to determine whether patients with cystic fibrosis, their close companions, and physician perceived the severity of the disease and the level of patient self care similarly.**

***Methods* – Sixty adults with cystic fibrosis (16–44 years), their close companion, and physician independently completed scales measuring their perceptions of disease severity and patient self care on three occasions over a two year period. Percentage predicted forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC), Shwachman score, and weight for height were recorded following each assessment.**

***Results* – Patients and close companions considered the disease to be less severe than their physician. Fifty patients (83%) rated their health as "above/well above average", and 49 (82%) close companions rated the patient's health in the same way, but only 21 (35%) patients were considered by their physician to have mild disease. Differences also emerged in the estimation of patient self care; 48 close companions (80%) rated patients as "very good" or "excellent" in their self care, compared with 26 (44%) of the patients themselves. Only nine patients (15%) were considered to achieve this level of self care by their physician. Over the two year period the physician's ratings of severity increased in accordance with the decline in lung func-**

tion. Patients' perceptions remained unchanged.

***Conclusions* – From the physician's viewpoint, patients and close companions underestimate the severity of cystic fibrosis and overestimate patient self care. Patients' perceptions remain constant over time even when their health is clinically deteriorating.**

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Keywords: cystic fibrosis, self care, perception.

Compliance with advice and treatments is a problem common to many medical disciplines.[1] Numerous demographic, medical history, and clinical factors which might characterise nonadherence in patients with cystic fibrosis have been investigated. Most clinical variables which have been studied have little predictive value.[2–4] Compliance is a complex behaviour often influenced by the patients' perceptions and expectations. Patients who do not underestimate the seriousness of their illness have been shown to be more compliant.[5] One reason for poor adherence may be that patients do not share the perceptions of their physician concerning the severity of their disease, or how well they care for themselves. This study aimed to assess and compare the perceptions of patients, their close companions, and their physician concerning the severity of their disease and their general self care. In addition, patients and physician estimated disease severity and self care over a two year period to investigate whether perceptions change as the disease progresses.

Methods

PATIENTS

Sixty patients who regularly attended the Adult Cystic Fibrosis Unit in Manchester and their close companions (either mother or partner) who regularly accompanied them took part in the study. The patient group comprised 25 women and 35 men with a mean age of 21 years (range 16–44). At the beginning of the study disease severity measured by percentage predicted forced expiratory volume in one second (FEV_1) ranged from 28% to 105% (mean 61%). Disease severity and self care were rated by patients, their close companion, and physician at assessment 1 (time 0), and by the patients and physician at assessments 2 (one year follow up, n=56) and 3 (two year follow up, n=51).

RATINGS OF DISEASE SEVERITY AND SELF CARE

Patients completed scales concerning their perceptions of their disease severity and level of self care. Disease severity was rated on a five point scale in answer to the question "Compared with others with cystic fibrosis, do you think your health is poor (1), below average (2), average (3), above average (4), well above average (5)". It was explained to the patients and close companions that self care referred to the overall self management of cystic fibrosis which included chest physiotherapy, the taking of oral medication (antibiotics, pancreatic enzymes and vitamins), and exercise therapy. Patient self care was rated on a six point scale in answer to the question "Overall, do you think that your care for yourself is very poor (1), poor (2), slightly poor (3), good (4), very good (5), excellent (6)". Close companions and physician were independently asked similarly phrased questions. To ensure that the physician's ratings were valid at assessment 1, another cystic fibrosis physician independently rated each patient for severity of disease and self care. Very high positive correlations emerged between the two physicians for both disease severity ($r=+0{\cdot}91$) and self care ($r=+0{\cdot}88$).

To investigate whether demographic, medical history, or current clinical status variables were associated with perceptions of disease severity or self care, data were obtained concerning age, sex, employment status, frequency of contact with the clinic, inpatient or outpatient status, and age at diagnosis. Information was collected by patient interview and checked with the patients' notes where appropriate. Weight for height, FEV_1 and FVC % predicted and Shwachman scores were obtained following completion of the scales for each assessment. The Shwachman score is a cystic fibrosis score combining physical symptoms and signs, activity, and nutritional and chest radiographic status.

STATISTICAL ANALYSES

Analyses of data were performed by Mann-Whitney U tests to compare ratings of disease severity or self care between raters. Spearman rank correlations were used to assess the associations between ratings and clinical or demographic variables. Clinical variables and ratings by patients and physician over time were examined using analyses of variance with repeated measures.

Results

DISEASE SEVERITY

At assessment 1, mean (SD) ratings of disease severity were similar for patients and close companions (4·05 (1·10) and 4·20 (0·79), respectively). The physician's judgement was much lower (2·92 (1.15)) and significant differences occurred between the physician's ratings and the two other groups ($p<0{\cdot}01$). Compared with others with cystic fibrosis, 50 (83%) patients' ratings and 49 (82%) close companions' ratings of disease severity were in the "above average" and "well above average" categories. Only 21 (35%) patients, however, were considered by the physician to have such mild disease (table 1).

Physician's ratings of disease severity were positively correlated with clinical findings (FEV_1, $r=0{\cdot}89$, FVC, $r=0{\cdot}79$, weight for height, $r=0{\cdot}91$, Shwachman score, $r=0{\cdot}86$, $p<0{\cdot}001$). Correlation coefficients indicate that those patients with the greatest differences between patient (close companion) and physician scores concerning disease severity had the lowest values (FEV_1% predicted, $r=-0{\cdot}59$; FVC% predicted, $r=-0{\cdot}48$; Shwachman

Table 1 Ratings of disease severity and patient self care by patients, close companions, and physician

	Patients n (%)	*Close companions n (%)*	*Physician n (%)*
Perceptions of disease severity			
Well above average	24 (40)	24 (40)	3 (5)
Above average	26 (43)	25 (42)	18 (30)
Average	8 (13)	9 (15)	20 (33)
Below average	2 (3)	2 (3)	9 (15)
Poor	0 (0)	0 (0)	10 (17)
Perceptions of self care			
Excellent	4 (7)	13 (22)	1 (2)
Very good	22 (37)	35 (58)	8 (13)
Good	28 (47)	12 (20)	20 (33)
Slightly poor	4 (7)	0 (0)	14 (23)
Poor	2 (3)	0 (0)	12 (20)
Very poor	0 (0)	0 (0)	5 (8)

Table 2 Mean FEV_1% predicted, Shwachman score, and ratings of disease severity and self care over the two year period

	Time 0	*1 Year*	*2 Years*
FEV_1 (% predicted)	61	57	50**
Shwachman score	69	66	62*
Patients' ratings of disease severity	4·1	4·1	4·0
Physicians' ratings of disease severity	2·9	2·7	2·3**
Patients' ratings of self care	4·3	4·5	4·4
Physicians' ratings of patients' self care	3·4	3·2	3·2

** $p<0.01$; * $p<0.05$.

score, $r=-0.51$; and weight for height, $r=-0.61$; $p<0.01$). This indicates that patients with the most severe disease rated their cystic fibrosis as "above/well above average" compared with others with cystic fibrosis. Eight of these patients have undergone assessment for heart-lung transplantation, yet the severity ratings of this small subgroup were similar to the whole study population. The physician rated six patients in the "poor" category and two patients in the "below average" category. Each patient had identical ratings to their close companion, rating themselves as "well above average" (n=4) and "above average" (n=4). The perceptions of disease severity of the patients, physician, and close companions were not associated with demographic or medical history variables at any assessment time (age, sex, employment status, frequency of clinic visits, and age at diagnosis).

SELF CARE

At assessment 1, mean (SD) ratings of patient self care differed significantly between patients (4·30 (1·10)), physician (3·37 (1·23)), and close companions (5·02 (0·66)), $p<0.01$. Forty eight of 60 (80%) close companions placed patients in the "very good" and "excellent" categories compared with 26 (44%) of the patients themselves. Only nine patients (15%) were considered to achieve this level of self care by their physician (table 1). The perceptions of self care of the patients, physician, and close companions were not correlated with demographic or disease severity variables. The perceived disease severity was not related to the assessment of self care for any of the three groups.

PERCEPTIONS OF DISEASE SEVERITY, SELF CARE, AND DISEASE PROGRESSION

There was a significant decrease in FEV_1% predicted ($p<0.01$), Shwachman score ($p<0.05$), and physician's ratings ($p<0.01$) over the two year period. No differences in the patients' ratings of disease severity were observed during this time, thus widening the gap between the patients' and physician's estimations as the disease progressed. This supports the cross sectional finding in assessment 1 that the largest discrepancies occur between physician and the most severe patients. There were no changes concerning the ratings of self care made by patients or physician, therefore the perceptual differences between them remained constant over time (table 2).

Discussion

Discrepancies exist between patient, close companion, and physician in their perceptions of disease severity and self care. From the physician's viewpoint, generally, patients and close companions underestimate the severity of the disease and overestimate patient self care. Patients' perceptions of their disease appear to remain constant over time, even when their health is deteriorating. The patients assessed for heart-lung transplantation also perceived their health to be "above average".

These findings have implications not only for understanding and improving compliance in chronic disease populations where a high level of self care is necessary, but for the overall management of patients and their families in a specialist unit, or by their general practitioners and other health care staff. Patients with cystic fibrosis and their carers may view the severity of the disease and self care from a wider perspective than their physician, incorporating into their perceptions the social and mental health aspects of their life quality in addition to the physical limitations imposed by cystic fibrosis. Since the patients' perception of disease severity was not associated with how well they cared for themselves, other factors which may influence treatment adherence require investigation.

It is unclear whether quality of life of the patients and their families or disease progression are influenced by these perceptions. Denial or underestimation of the seriousness of their disease may afford emotional protection to the patients and their families, but difficulties in communications are likely to arise when preparing the patient and family for heart-lung transplantation assessment or in decisions relating to preterminal care. This work forms part of an ongoing multidisciplinary research project concerned with the ways in which patients cope with cystic fibrosis, adherence to treatments, and disease progression which may alter and influence our current clinical practice.

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