Most patients with cystic fibrosis (CF) carry mutations at the CFTR gene which are associated with malfunction of the pancreas. Since malabsorption due to pancreatic insufficiency causes intolerance of nutritional fat, patients are at risk of low energy intake in addition to increased faecal nutrient loss. In the past malnutrition was considered one of the key clinical features of CF. A low fat diet was recommended by most CF centres until the mid 1980s when acid resistant pancreatic enzymes became available. At that time researchers from Toronto had reported that an energy rich diet not restricted in fat, together with additional (conventional) enzyme supplements, resulted in a better nutritional status and longevity of their patients compared with those treated in Boston, a comparably large and experienced centre. Seventeen years later, with modified dietary recommendations in the US, substantial lower differences in the growth indices of patients with CF between the US and Canada were observed. Thus, there was evidence that improved nutrition was associated with better long term outcome, as had already been stated in an earlier paper by Kraemer et al. A consensus report on nutrition in CF published in 1992 described nutritional management as an important part of the multidisciplinary approach to the disease. Nowadays there is no reason to accept nutritional failure or growth retardation in any patient with CF.

Patient registries for CF permit analysis of data from large cohorts of patients over many years. Results from Canada and the US, the UK, Australia, Europe, and Denmark have shown improved nutritional status, lung function, and survival of CF patients during the last decades. Respiratory function tests, energy expenditure, growth status, and weight were more abnormal with increasing age. Later birth cohorts had improved outcome, and some authors described sex related differences in health. Most of these studies present cross sectional data, however, while the longitudinal course of certain patient groups over years has been addressed in only a few reports.

The relationship between nutritional status and lung function has not been analysed in detail using data from patient registries. The German CF quality assurance project was founded in 1995 and data from more than 5000 patients with CF aged 1 month to 58 years have been documented up to the end of the year 2000. The aim of the present study was to describe the prevalence of malnutrition in these patients and to elucidate the relationship between nutrition and lung function by cross sectional and longitudinal analyses. We speculated that patients with normal weight would be in a better clinical condition than malnourished patients, and that the yearly decline in pulmonary function would be smaller if weight was normal.

METHODS
The German CF quality assurance (CFQA) project is a patient registry which contains relevant clinical and laboratory data,
Body mass index (BMI) was $<19 \text{ kg/m}^2$ and/or stunting—that is, if (1) weight was below 90% of the median normal value for sex and age, or if (2) height was <90% of the median normal value for sex and age. Patients were defined as malnourished if data indicated wasting and/or stunting. Patients were considered increased if Z scores were $>-1.6$.

### Figure 1

Frequency of (A) malnutrition and (B) impaired lung function in patients with and without *Pseudomonas aeruginosa* (PA) colonisation.

### Table 1

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>Weight for height (% pred)</th>
<th>Vital capacity (% pred)</th>
<th>FEV$_1$ (% pred)</th>
<th>IgG (g/l)</th>
<th>PO$_2$ (kPa)</th>
<th>MEF$_{25}$ (% pred)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2–5.9</td>
<td>[19.0]</td>
<td>[85.3]</td>
<td>[86.7]</td>
<td>[10.8]</td>
<td>[8.1]</td>
<td>[9.9]</td>
</tr>
<tr>
<td>6–11.9</td>
<td>[96.9]</td>
<td>[96.5]</td>
<td>[97.3]</td>
<td>[98.2]</td>
<td>[97.6]</td>
<td>[98.1]</td>
</tr>
<tr>
<td>12–17.9</td>
<td>[84.2]</td>
<td>[83.6]</td>
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</tr>
<tr>
<td>&gt; 18</td>
<td>[71.4]</td>
<td>[70.1]</td>
<td>[72.7]</td>
<td>[54.5]</td>
<td>[10.0]</td>
<td>[7.0]</td>
</tr>
</tbody>
</table>

**Note:** FEV$_1$=forced expiratory volume in 1 second; PO$_2$=oxygen tension; MEF$_{25}$=mid expiratory flow at 25% of vital capacity. Numbers in square brackets indicate percentage of patients with abnormal results.

### Table 2

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>Vital capacity (% predicted)</th>
<th>FEV$_1$ (% predicted)</th>
<th>MEF$_{25}$ (% predicted)</th>
<th>IgG (g/l)</th>
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</tr>
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<tbody>
<tr>
<td><em>P. aeruginosa</em> positive patients: 6–11.9</td>
<td>-11.3 (-14.9 to -7.7)</td>
<td>-14.5 (-19.5 to -9.5)</td>
<td>-15.4 (-23.5 to -7.4)</td>
<td>2.9 (1.9 to 3.9)</td>
<td>-0.3 (-1.7 to 1.1)</td>
</tr>
<tr>
<td>12–17.9</td>
<td>-21.1 (-24.6 to -17.7)</td>
<td>-23.6 (-27.5 to -19.8)</td>
<td>-20.2 (-26.5 to -13.9)</td>
<td>3.6 (2.6 to 4.6)</td>
<td>-0.5 (-1.5 to 0.3)</td>
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<tr>
<td>&gt; 18</td>
<td>-18.9 (-21.6 to -16.3)</td>
<td>-20.4 (-23.7 to -17.7)</td>
<td>-11.4 (-14.5 to -8.4)</td>
<td>2.1 (1.2 to 2.7)</td>
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<td><em>P. aeruginosa</em> negative patients: 6–11.9</td>
<td>-18.9 (-13.2 to -6.5)</td>
<td>-11.8 (-16.5 to -7.2)</td>
<td>-13.8 (-22.4 to -5.2)</td>
<td>1.3 (0.4 to 2.2)</td>
<td>0.2 (-2.7 to 3.1)</td>
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<tr>
<td>12–17.9</td>
<td>-15.6 (-19.9 to -11.4)</td>
<td>-18.7 (-23.7 to -13.7)</td>
<td>-21.4 (-31.0 to -11.9)</td>
<td>1.3 (0.1 to 2.4)</td>
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<td>&gt; 18</td>
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</table>

**Note:** Example: In patients aged 6–11.9 years colonised with *P. aeruginosa*, vital capacity was 11.3% of predicted lower in malnourished than in normally nourished children. FEV$_1$=forced expiratory volume in 1 second; PO$_2$=oxygen tension; MEF$_{25}$=mid expiratory flow at 25% of vital capacity.

### Cross sectional analysis

The 1997 data from all patients were divided into four different age groups: 2–5.9 years, 6–11.9 years, 12–17.9 years, and 18.0 years of age or older, since severity of disease increases with age. According to a publication by Lai and coworkers, patients were defined as malnourished if data indicated wasting and/or stunting—that is, if (1) weight was below 90% of the predicted normal value for sex and height in children, if body mass index (BMI) was <$19\text{ kg/m}^2$ in adults or if (2) weight was <$80\%$ of the median normal value for sex and age, or if (3) height was <$90\%$ of the median normal value for sex and age. Pulmonary function was considered abnormal if (1) forced vital capacity (FVC) was <$80\%$ predicted and (2) if forced expiratory volume in 1 second (FEV$_1$) was below 75% of the predicted normal value. Mid expiratory flow at 25% of vital capacity (MEF$_{25}$) was rated abnormal if values were less than 60% predicted. The lower limit of normal for oxygen tension (PO$_2$) in oxygenised capillary blood was 10.7 kPa (80 mm Hg). Serum immunoglobulin G (IgG) values were considered increased if Z scores were $>2$. Reference values were used according to recommendations made by the Scientific Advisory Committee of the CFQA project who selected appropriate normal values for the population under consideration. The tabulated reference data for weight were related to a third degree polynomials equation of height. Comparisons between age groups were made for means of nutritional and lung function parameters, and the relative proportion of patients with abnormal results was determined for each age group. In addition, patients were grouped according to the presence or absence of risk factors such as *Pseudomonas aeruginosa* colonisation or malnutrition.

**Table 1 Mean clinical data (95% confidence intervals) in 1997 by age group**

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>Weight for height (% pred)</th>
<th>Vital capacity (% pred)</th>
<th>FEV$_1$ (% pred)</th>
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**Note:** FEV$_1$=forced expiratory volume in 1 second; PO$_2$=oxygen tension; MEF$_{25}$=mid expiratory flow at 25% of vital capacity. Numbers in square brackets indicate percentage of patients with abnormal results.

**Table 2 Mean differences (95% confidence intervals) between malnourished and normally nourished patients by age groups and by *Pseudomonas aeruginosa* colonisation**

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>Vital capacity (% predicted)</th>
<th>FEV$_1$ (% predicted)</th>
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**Note:** Example: In patients aged 6–11.9 years colonised with *P. aeruginosa*, vital capacity was 11.3% of predicted lower in malnourished than in normally nourished children. FEV$_1$=forced expiratory volume in 1 second; PO$_2$=oxygen tension; MEF$_{25}$=mid expiratory flow at 25% of vital capacity.
Multifactorial variance analysis was performed to investigate the association between lung function and malnutrition and the confounders P. aeruginosa infection and sex. The Student’s t test for independent samples was performed to compare means between groups with normal or abnormal results. \( \chi^2 \) and Fisher’s exact tests were used to evaluate proportions of patients.

**Longitudinal analysis**

Patients who had complete data sets for 1995 and 1996 were eligible for the first part of the longitudinal analysis. 536 children aged 6–11.9 years and 477 adolescents aged 12–17.9 years were analysed in detail. For each patient the differences in weight for height and FEV\(_1\) between 1996 and 1995 were calculated.

Three longitudinal groups were defined according to the resulting differences: “better” if the results in 1996 were at least 5% higher than those in 1995: “no change” if the differences were between –5% and +5%; and “worse” if the results were at least 5% lower than in 1995. For each of these groups mean changes in FEV\(_1\), and weight for height between the two years were determined. When means from the three longitudinal groups were compared, one way analysis of variance with the Student-Newman-Keuls test was performed. All calculations were made using SPSS (SPSS Inc, Chicago, USA).

In a second longitudinal analysis the equality of FEV\(_1\), slopes and intercepts in different risk groups was tested for 1995, 1996, and 1997. Mixed model analysis was performed using SAS Proc Mixed version 6.12 for Windows. A multiple comparison adjustment for the p values and confidence limits for the differences of least squares means was performed with a Tukey test. An approximate F test was used to assess the effect of deleting a variable from the model. A level of p=0.05 was used to indicate statistical significance.

**RESULTS**

**Cross sectional data**

Clinical and lung function data were available for 3298 patients above 2.0 years of age (table 1). As expected, mean weight for height, vital capacity, FEV\(_1\), and PO\(_2\), decreased with age, and the proportion of patients with abnormal results was larger in older patients. Mean weight for height was below 90% predicted in 19.0% of children aged 2–6 years and in 31.5% of adolescents. In children below 12 years of age, PO\(_2\) had the highest frequency of abnormal results. More than 50% of adolescents had abnormal MEF\(_25,75\), IgG, and PO\(_2\), while FEV\(_1\) was decreased in only 38.5%. A multifactorial analysis of variance revealed that FEV\(_1\) was related to both malnutrition (p<0.001) and P. aeruginosa colonisation (p<0.001) but was independent of sex (p>0.05) for all age groups.

Pulmonary function was decreased in 56.0% and 85.6% of P. aeruginosa positive adolescents and adults, respectively, whereas only 37.4% and 66.3% of non-colonised patients had abnormal lung function (p<0.001, fig 1). An association between P. aeruginosa colonisation and malnutrition was also observed. However, most patients had a weight for height above 90% predicted or a body mass index above 19 kg/m\(^2\), irrespective of bacterial status.

Patients with malnutrition had significantly lower vital capacity and FEV\(_1\), and higher serum IgG. As shown in table 2, this was independent of P. aeruginosa infection. For example, in malnourished children aged 6–11.9 years who were P. aeruginosa positive, mean vital capacity and FEV\(_1\), were 11.3% and 14.5% predicted lower, respectively, than in normally nourished children (table 2). A similar difference between children of normal and decreased weight was observed in the absence of P. aeruginosa colonisation—for example, FEV\(_1\), was 11.8% predicted lower. In P. aeruginosa positive and negative adolescents, malnutrition was associated with even larger differences in lung function, IgG, and PO\(_2\). The differences between normally nourished and malnourished adults with CF were comparable to those in adolescents, and the presence or absence of P. aeruginosa had no additional effect.

Figure 2 shows cross sectional mean values of FEV\(_1\), by age and bacterial status. FEV\(_1\), values of at least 90% predicted were observed in children and adolescents with normal weight and no P. aeruginosa infection. P. aeruginosa positive patients with normal weight had mean FEV\(_1\), values which were 10–20% predicted lower, but they remained at above 80% predicted except at age 13. In contrast, malnutrition was associated with a severe decline in FEV\(_1\), of about 20% predicted between 12 and 18 years of age, irrespective of bacterial status. Malnourished patients colonised with P. aeruginosa had the worst results at any age. For example, at ages 8 and 17 mean FEV\(_1\), was only 63% and 46% predicted, respectively. These differences between the four groups were both clinically relevant and statistically significant (p<0.001).

**Longitudinal analysis**

Three patient subgroups were formed with respect to the patients’ individual courses from 1995 to 1996. Malnourished adolescents who experienced a decrease of ≥5% predicted in weight for height during that year had a concomitant mean loss in FEV\(_1\), of 16.3% predicted, whereas patients who gained relative weight had a parallel increase in FEV\(_1\), of 2.1% predicted. These differences were statistically significant (p<0.001). In adolescents with normal weight in 1995, FEV\(_1\), declined by 7.6% or 0.16% of predicted if weight decreased by at least 5% or remained stable, respectively, whereas patients who improved their weight by more than 5% had a concomitant increase in FEV\(_1\), of 4.3% (p<0.001). Figure 3 shows the respective data for 6–11.9 year old children.

In a further step a mixed model analysis was performed to identify differences between sexes and between risk groups (patients with or without malnutrition or P. aeruginosa infection, respectively) with respect to the decline in FEV\(_1\), within a year. Malnourished patients of all age groups had significantly worse lung function than their normally nourished counterparts (table 3). At the initial visit in 1995, FEV\(_1\), was 11.7–23.9% predicted lower if weight for height was decreased. During the 2-year follow up period, adolescents with malnutrition experienced a decline in FEV\(_1\), whereas lung function remained stable in adolescents with normal body...
weight (p<0.01, fig 4). The FEV₁ slopes of patients colonised with *P aeruginosa* were significantly worse in children and adolescents with a 1.14–2.79% predicted faster decline in lung function (p<0.001). The only association with sex was found in adolescents, with females showing a slower progression of lung disease than males—that is, a 1.58% predicted difference in FEV₁ within a year (table 3).

### DISCUSSION

The negative impact of underweight on the long term outcome of patients with CF has long been recognised. The results of the present longitudinal analysis from a large cohort of patients confirm that nutrition and lung function are co-dependent variables in CF. Patients with normal weight had a significantly smaller decrease in lung function over a 2 year period than those with malnutrition, and this was shown in all age groups. A fall in weight for height of 5% predicted or more within 1 year was associated with a parallel decrease in FEV₁, whereas patients with improved nutrition showed constant or even improved FEV₁.

Cross sectional analysis revealed that the prevalence of malnutrition was 19% in children aged 2–6 years and increased considerably with age up to 38% in adults. Other disease related parameters, particularly PO₂ and MEF₂₅, were abnormal in even larger proportions of patients, and showed a similar deterioration with age. Of the 858 adolescents aged 12–18 years, more than one third had abnormal MEF₂₅ (59%), serum IgG (53%), PO₂ (50%), FEV₁ (42%), and VC (39%). The figures for FEV₁ are comparable to data from other patient registries. For example, the US registry found that FEV₁ was less than 70% predicted in 68% of adult patients.

There are no data from other patient registries with respect to IgG, PO₂, or MEF₂₅ grouped by age.

In the present study, adolescent girls had better FEV₁ results and a smaller yearly decline in FEV₁ than boys. Younger children and adults showed no sex related differences by

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**Figure 3** Mean values of individual differences in FEV₁ (% predicted) between 1995 and 1996 in 536 schoolchildren aged 6–11.9 years of age. (A) Normally nourished children. (B) Children who were malnourished in 1995 (weight for height (W/H) <90% predicted). If W/H decreased by 5% or more, a concomitant decline in FEV₁ of (A) 12.8% and (B) 7.5% predicted was observed.

**Table 3** Longitudinal results: mixed model regression estimates for forced expiratory volume in 1 second (FEV₁) in 1995 (intercept) and for the mean decline in FEV₁ per year (slope)

<table>
<thead>
<tr>
<th></th>
<th>Estimated difference (95% Cl) in FEV₁ (% pred) in 1995</th>
<th>p value</th>
<th>Estimated difference (95% Cl) in change in FEV₁ per year</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>6–11.9 years</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Malnutrition (yes v no)</td>
<td>-11.7 (-17.9 to -5.6)</td>
<td>0.0002</td>
<td>-1.57 (-3.8 to +0.6)</td>
<td>0.160</td>
</tr>
<tr>
<td><em>P. aeruginosa</em> (yes v no)</td>
<td>-5.8 (-10.2 to -1.6)</td>
<td>0.008</td>
<td>-2.77 (-3.9 to -1.7)</td>
<td>0.0004</td>
</tr>
<tr>
<td>12–17.9 years</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Malnutrition (yes v no)</td>
<td>-11.6 (-17.7 to -5.5)</td>
<td>0.0002</td>
<td>-1.63 (-3.3 to -0.3)</td>
<td>0.009</td>
</tr>
<tr>
<td><em>P. aeruginosa</em> (yes v no)</td>
<td>-5.8 (-10.1 to -1.3)</td>
<td>0.008</td>
<td>-2.79 (-4.3 to -1.3)</td>
<td>0.0004</td>
</tr>
<tr>
<td>Sex (female v male)</td>
<td>+3.6 (+0.7 to +6.7)</td>
<td>0.1</td>
<td>+1.38 (+0.1 to +2.7)</td>
<td>0.042</td>
</tr>
<tr>
<td>≥18.0 years</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Malnutrition (yes v no)</td>
<td>-23.9 (-28.2 to -19.6)</td>
<td>0.0001</td>
<td>+1.21 (+0.1 to +2.4)</td>
<td>0.032</td>
</tr>
<tr>
<td><em>P. aeruginosa</em> (yes v no)</td>
<td>-10.1 (-15.1 to -5.1)</td>
<td>0.0001</td>
<td>-1.14 (-2.4 to +0.1)</td>
<td>0.08</td>
</tr>
</tbody>
</table>

Estimates are shown as mean differences between the subgroups malnutrition present v absent and *Pseudomonas aeruginosa* infection present v absent, respectively, and between male and female adolescents.
mixed model analysis. Data from the literature concerning the influence of sex on the course of CF are controversial. Zemel et al reported better weight (% predicted) in boys and higher Z scores for height and age in girls. In contrast, the growth of boys with CF was impaired on the basis of height, fat free mass, and fat mass when observed longitudinally. In the study by Corey et al, male patients had milder disease than females, as reflected by a smaller decline in FEV₁, decreased mortality, and higher survival. No significant differences between males and females have been found in other studies.

Chronic infection with *P. aeruginosa* had a negative impact on pulmonary function, *P* < 0.05. In the study by Corey et al, male patients had milder disease than females, as reflected by a smaller decline in FEV₁, decreased mortality, and higher survival. No significant differences between males and females have been found in other studies.

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**Figure 4** Mixed model analysis for the course of FEV₁ between 1995 and 1997 in the three age groups by risk factors (malnutrition and *P. aeruginosa* [Pa] colonisation) and sex.
available on possible pathophysiological processes in CF. In particular, a cause/effect relationship remains to be proven. In the malnourished mouse model of respiratory infections in CF, nutritional deficiency contributed to compromised innate lung defences, bacterial colonisation, and excessive inflammation in the CF respiratory tract. A recent review summarised clinical studies on the relationship between lung function and nutritional status. Correlations were found between FEV1 and lean body mass, the body compartment which includes skeletal muscle, suggesting an influence of muscle wasting on pulmonary function. The detrimental effect of malnutrition on immune function in general must also be considered as a relevant mechanism in CF. Treatment with intravenous antibiotics was associated with a concomitant decline in inflammatory parameters (C reactive protein, tumour necrosis factor α), energy expenditure, and an increase in body weight. In summary, the results of this study suggest that normal body weight is associated with better lung function in CF. A significantly smaller decline in FEV1 can be expected in all age groups if patients are well nourished, and this is independent of a concomitant colonisation with P. aeruginosa.

ACKNOWLEDGEMENTS

The authors thank the Scientific Advisory Board of the German CFQA project (Chairman: Professor Dr Martin Stern) for permission to analyse data from the registry, and acknowledge the large amount of work by staff from the 97 CF centres who reported patient data to the project (available on website www.thoraxjnl.com).

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