

Original Article

Employment and work disability in adults with cystic fibrosis

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25 July 2011; 11 October 2011; 25 October 2011

Available online 21 November 2011

Abstract

Background: As a result of prolonged survival, more patients with cystic fibrosis (CF) participate in the labour force. The aim of this study was to evaluate their education, occupation levels and risk factors for work disability.

Method: 207 patients answered a self-administered questionnaire about their educational level and work status. Independently, medical records were reviewed for illness severity indicators.

Results: 39 patients (19%) were students, 117 (57%) were in the labour force, 13 (6%) were seeking employment and 38 (18%) were inactive. CF patients had a higher educational level and were more likely to hold skilled jobs and to work part time than the general population. FEV1 and educational level were the strongest predictive factors of disability.

Conclusion: Many CF patients have access to professional life. Their higher educational levels improve the chances of attaining employment, which highlights the need for career counselling. Working part time helps to maintain employment despite declining health.

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Keywords: Cystic fibrosis; Adults; Disability; Occupational health

1. Introduction

In France, cystic fibrosis (CF) affects nearly 6500 patients [1] and one newborn child in 4136 [2]. With improvement in treatment and care, the life expectancy of CF patients has steadily increased over the past few decades. Their median life span is approaching 40 years and nearly half of the population reaches adulthood [3]. CF, formerly a childhood disease, is now a chronic disease of adults. Adult life confronts patients with new challenges. These patients require age-appropriate care and counselling about adult issues including career choices [4]. Wideman studied the educational needs of adults who were diagnosed with CF in adulthood and showed that employment and insurance issues were topics of high interest, once the basis of the disease and its treatments were known [5].

Employability is associated with better quality of life scores [6] and lower depression scores [7]. The positive association between employment and some quality of life scores was maintained even after controlling for relevant medical parameters [8]. Moreover, some aspects of quality of life reported by patients may be in themselves predictors of survival in CF [9]. This interaction between work, quality of life and possibly survival, makes it essential that caregivers strive to help their patients succeed in their professional lives.

However, very few CF adolescents receive any formal career guidance [10], probably because little is known about the work life of adult CF patients and the data about the factors associated with their employment are equivocal. CF involves a series of issues that could pose a barrier to employability. Respiratory insufficiency or malnutrition may limit the capacity

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to hold a job, whereas their need for treatment time or their absenteeism may be incompatible with working life.

To better characterise the impact of CF on professional activities, we conducted a cross-sectional observational study in our cohort of adult CF patients. The aim of our study was to evaluate their social and demographic characteristics, their education and occupation levels, and to determine risk factors for work disability.

2. Patients and methods

2.1. Design

We conducted a cross-sectional study between January and June 2009 in our adult CF centre in Paris, France. CF physicians asked their CF patients to complete a questionnaire, which was then self-administered during an outpatient visit or hospitalisation at the CF centre. 207 out of 317 CF patients participated in the study. Among the 110 non-participants, 51 patients were not seen at the CF centre during the inclusion period, 58 patients were not asked to answer the questionnaire and only one patient refused because of lack of time. This observational population survey was conducted in accordance with the Declaration of Helsinki and French law and patients gave their written informed consent to participate.

2.2. Data collection

The self-administered questionnaire recorded the demographic characteristics of the patients, their daily management of treatments (physiotherapy time, nebulised treatment time and overall treatment time), their educational level and detailed occupational status. Jobs were classified in occupational activity families according to FAP-2003 [11]. This nomenclature, established by the French Department of Employment, groups occupations that involve common skills based on close professional practices. Each job has a corresponding field of activity and level of qualification. Concerning employment status, we differentiated four groups of patients: students, employed persons, unemployed persons (seeking a job) and inactive persons (including retired persons, housewives and those not seeking a job). Medical records were reviewed independently for the history of the disease, treatments and illness severity indicators.

Demographic data included sex, age at CF diagnosis and age at time of the survey. Genotypes were classified into three categories according to the probable effect of their mutations on CFTR function [12] regardless of clinical severity. These categories were: severe genotype (two severe mutations), mild genotype (at least one mild mutation) and undetermined genotype (at least one unidentified mutation and no mild mutation). Patients were classified as pancreatic insufficient (faecal elastase < 200 µg/g or faecal fat > 6 g/day) or pancreatic sufficient. Sputum culture results were recorded and chronic bronchial colonisation was defined as the presence of bacteria for at least six months and three positive sputum cultures. Pulmonary function test results (forced vital capacity [FVC] and forced expiratory volume in 1 s [FEV1]) were expressed as

the percentage of the predicted value [13] and the best values achieved during the past six months were noted.

Body mass index (BMI; weight/height²) was also recorded. Other collected data included common comorbidities (e.g., diabetes mellitus, liver cirrhosis) and indications about treatment (number of yearly intravenous [IV] antibiotic courses, oxygen treatment).

2.3. Statistical methods

Quantitative variables are expressed as means with standard deviation, range and median, indicated as appropriate. Descriptive data are expressed as percentages of the patients for whom data were available.

For the comparisons between patients, the Chi-square test was used for qualitative data. For quantitative data, Student's t-test was used when the distribution of the variable was normal; otherwise a Wilcoxon–Mann–Whitney rank-sum test was applied.

To identify parameters associated with disability, the analysis was conducted in two phases. Firstly, in a univariate analysis, socio-demographic and clinical characteristics, and treatment modalities were compared between workers and non-workers. A prevalence ratio (PR) of disability was calculated for each characteristic. Confidence intervals of PR were calculated using normal approximation (Wald). P values were calculated using Fisher's exact test. Disability was defined by having no job at inclusion. Students and retired people were excluded from these analyses. Secondly, multivariate analysis was performed by logistic regression. The regression model included six a priori selected variables: socio-demographic characteristics (age, gender and education), CF severity indicators (exocrine pancreatic insufficiency and FEV1) and diabetes, which has been recognised to have a negative impact on employment independently of CF [14]. Significance was defined as $p < 0.05$ for all analyses. The R packages were used for statistical analyses [15].

3. Results

3.1. Sample characteristics

The main characteristics of the 207 participants are listed in Table 1. The 110 other patients cared for at our CF centre who were not included in the study were not different from the study participants in terms of gender (65 females, 59%), mean age at diagnosis (8.3 years), mean FEV1 (56.9% pred.), mean FVC (71.1% pred.), mean BMI (20.6 kg/m²) and mean number of IV antibiotic courses per year (1.54), although they were slightly younger than the participants (mean age: 28.8 years, $p = 0.03$).

The majority of the patients were single (38% were married or living with a partner), 30% lived at their parents' home and 23% had children. CF was the reason for inactivity for the 28 patients who were not employed and not looking for work. Two persons were retired.

Table 1
Patient characteristics.

N	207
Sex	
Male	107 (51.7)
Age (years)	30.7±8.7
Employment status	
Students	39 (18.8)
Employed	117 (56.5)
Unemployed	13 (6.3)
Inactive	38 (18.4)
Age at diagnosis (years)	7.8±11.6
>20	30 (15)
Genotype	
Severe	149 (72.0)
Mild	46 (22.2)
Unknown	12 (5.8)
FEV1 (% pred.)	54.0±21.8
≥70	43 (22)
<40	57 (29)
FVC (% pred.)	70.1±20.8
Chronic bronchial colonisation	
<i>Pseudomonas aeruginosa</i>	123 (59.4)
<i>Staphylococcus aureus</i>	80 (38.6)
BMI (kg/m ²)	20.6±3.0
PI	171 (83.4)
Diabetes	65 (31.4)
Cirrhosis	12 (5.8)
IV antibiotic courses (number/year)	1.87±2.2
Transplantation	
Lung	11 (5.3)
Liver	1 (0.1)
On the waiting list	12 (5.8)

NS: not significant.

FEV1: forced expiratory volume in one second; FVC: forced vital capacity; BMI: body mass index; PI: exocrine pancreatic insufficiency; IV: intravenous. Mean values±SD for quantitative variables.

N (%) for qualitative variables.

In stability periods, the mean daily treatment time was 55 min (median: 61 min). 183 patients (89%) underwent daily physiotherapy, which required an average of 38 min. 136 patients (66%) received nebulised treatment requiring an average of 32 min daily. Twenty patients received oxygen therapy (continuously in ten patients) and ten had non-invasive ventilation. More than a third of the patients (75 patients, 36%) received no IV antibiotic therapy during the past year, whereas a third received at least three courses. Among diabetic patients, 47 (72%) were treated with insulin. Dietary supplements were prescribed to 78 patients (38%) including 15 patients who received enteral feeding.

3.2. Employment status

Among the 168 patients who had completed their studies, 70% was employed when interviewed and 94% had had a job in the past. Among transplant recipients, one was inactive, two were students, two were seeking employment and seven were employed.

The educational levels of the non-student patients were as follows: 119 (72%) had at least a high school diploma, 94 (57%) had studied at least two years at university and 28

(17%) had a Master's degree. Eighteen patients (13%) declared that they had stopped their studies because of CF.

Activities which were not recommended to CF patients the most frequently were: health care work (n=16), physical work (n=11), work involving contact with children (n=6), work involving exposure to dusts or fumes (n=6), outdoor work (n=6), work involving contact with animals (n=4), hair-dressing (n=4) and work in green spaces (n=4).

Job characteristics were recorded for 116 out of the 117 patients who were employed (Table 2). One patient did not give any details about his job. Jobs were mostly sedentary or involved light physical activity (70%). Very few people had an unskilled job: 29 were professionals (25%), 45 had intermediate positions (39%), 37 were clerks (32%) and only five were blue-collar workers (4%). Seven patients worked at home (3 females and 4 males). Eight patients were self-employed.

More than half of the workers (55%) considered that they had limitations in their job due to CF, 67% thought that CF prevented them from having a career and 37% believed that it was a cause of a lower income. Only 33% had revealed that they had CF at job interviews but 62% had announced their disease to their colleagues. Workstation adjustments had been done for 25 patients (22%), and these most often consisted of a decrease in the hours worked. Part-time workers were more often females and seemed to have a more severe disease than their full-time employed peers (Table 3).

Among the patients who had had at least one antibiotic IV course in the past year, 71% had stopped working during these treatments, mostly for the entire course (81%). Overall, 70% of the workers declared having at least one sick leave for CF during the previous year (the median amount of time spent on sick leave was 15 days/year). These leaves caused a service disruption in 54% of the cases.

Table 2

The occupational domains of employed people (according to FAP-2003) (11).

Key letters	Domains	N
A	Agriculture, navy, fishery	0
B	Construction (private and public)	1
C	Electricity, electronics	1
D	Mechanics, metallurgy	1
E	Manufacturing industry	1
F	Light industry, wood, graphics industry	1
G	Maintenance	1
H	Engineers, industry executives	3
J	Transport, logistics, tourism	6
K	Craftwork	0
L	Management, business administration	19
M	Data processing	8
N	Research and development	1
P	Public service, legal profession	15
Q	Banking, insurance	6
R	Trade	15
S	Hotel trade, catering, food industry	5
T	Individual and community services	1
U	Communication, information, arts and entertainment	8
V	Health, social activity, cultural activity, sports	16
W	Education, teaching	7
X	Politics, religion	0

Table 3
Patients' characteristics according to working hours.

	Full-time	Part-time	p
N	73	43	
Sex: male	50 (68.5)	16 (37.2)	0.002
Age (years)	31.8±8.2	32.3±6.2	NS
FEV1 (% pred.) ^a	56.4±21.4	51.8±23.6	NS
FVC (% pred.) ^a	73.9±19.3	67.5±22.7	NS
IV antibiotic courses (number/year)	1.4±2.2	2.5±1.8	<0.001
Time of daily treatment (minutes)	49.3±43.3	70.9±45.0	0.02
Time of one-way travel between home and work (minutes)	34.2±26.3	23.8±20.2	0.02

NS: not significant.

FEV1: forced expiratory volume in one second; FVC: forced vital capacity; IV: intravenous.

Mean values±SD for quantitative variables.

N (%) for qualitative variables.

^a Excluding lung transplanted patients.

3.3. Factors associated with employment status

Univariate analysis showed that the patients who were living with their parents, those who were single and those who had a low educational level worked less frequently than the others (Table 4). IV antibiotic courses (PR of disability=1.92 [1.06–3.48], $p<0.05$, for at least one course per year), oxygen treatment and FEV1 (PR for disability=3.55 [1.18–10.71], $p<0.01$, if less than 70% predicted) were associated with being employed. In the multivariate analysis, FEV1 and educational level were the only statistically significant factors associated with employment status (Table 4).

4. Discussion

We provide data about the employment status of 207 adults with CF, which is the largest CF cohort for whom their field of professional insertion was studied.

4.1. Work status

Our results showed that most of our CF adult patients had participated in the labour force at some point in their lives, as 94% of them had had a previous job and many of them were employed at the time of the study (70% of the non-student patients). This encouraging finding is consistent with the employment rates of CF patients found in studies conducted in other countries: 55% of 49 CF adult patients in California [16], 48% of 183 patients in North Carolina [7], 72% of 50 patients in Australia [6] and 56% of 130 patients in Canada [17]. In France in 2007, among people between the ages of 25 and 39, the labour force participation rate and the employment rate were respectively 89% and 82% for the general population, and 63% and 51% for people with an officially recognised disability [18].

We also highlight the good workforce participation of CF patients after transplantation: among ten non-student lung transplant recipients, seven were employed. This result is in line with previous studies, which have shown that CF is a lung transplantation indication associated with a good prognosis for subsequent employment [19,20].

The key factor for successful professional insertion is education and the educational level of our patients was higher than that of the general population. Indeed, whereas, in 2007, 24.8% of the women and 20.3% of the men in France between the ages of 25 and 34, and ~15% of those of either sex aged 35–44, studied for more than 2 years at university, 32.6% of our patients achieved this level of study. [21]. Due to this high level of education, it is not surprising that many of our patients had access to skilled jobs. Very few were blue-collar workers (less than 5% versus 23% in the general population), whereas technicians, associate professionals, engineers and managers combined were overrepresented in our CF patients (25% versus 16% in the general population) [21]. Burker et al. made a similar observation among 88 employed CF patients [22]: 73% were skilled workers, 21% were semi-skilled workers and only 6% were unskilled labourers (but 31% of the CF patients who had stopped working were unskilled). It is also likely that the skilled jobs attained by patients with higher educational levels were jobs that could more easily accommodate declines in health. Generally, the labour market is more favourable to persons who have scholarly degrees.

We showed that many CF patients succeed in study and work. However, two-thirds thought that CF was an obstacle to their career and cited some of the difficulties they encountered in the workplace. Over half of the patients reported being limited in their work by their disease. Absenteeism often disrupted the functioning of the companies, which is particularly problematic for workers with responsibilities. Patients who were not working also illustrated the employment difficulties related to CF, since three quarters of them reported that their disease was the main cause for their inactivity.

Staying in employment may require an adaptation at the workplace. A third of the workers had expressed interest in or had benefited from an adjustment of their workstation. In most cases, this adjustment involved working time. The percentage of CF patients who worked part-time was two to three times higher than in the general population (37% in CF patients versus 15.9% in workers aged 25–49 in 2009) [21] and was more common among women CF patients. This may be explained by the fatigue of these patients and/or their need for time to complete their care. The disease of patients working part-time also seemed to be more severe than that of patients who worked full-time: their respiratory functions tended to be worse; they received more IV antibiotic courses; and they spent more time on their treatment. Unfortunately, part-time work often leads to a decline in the individual's resources, as social assistance compensates for only part of the income losses related to working less than full-time.

The exercise capacity of adults with CF is often reduced. They are often able to perform mild physical activity (ground walking at a normal pace), but their ability to withstand greater physical activity is reduced [23]. These results can be extrapolated to their professional activities and suggest that CF workers in a tertiary position are likely to be only slightly limited or not limited at all. This relationship is not surprising, since almost 70% of our workers actually had a tertiary activity, such as management and administration, data processing,

Table 4
Factors associated with disability.

Variable		% of disabled	Univariate analysis			Multivariate analysis ^a		
			PR	95% CI	p	OR	95% CI	p
Age (years)	<30	30.3	1					
	≥ 30	29.8	0.95	[0.59–1.53]	NS	1.46	[0.64–3.31]	NS
Sex	Female	31.1	1					
	Male	28.3	0.91	[0.56–1.46]	NS	0.89	[0.40–1.93]	NS
Age at diagnosis (years)	<18	30.3	1					
	≥ 18	26.5	0.87	[0.47–1.62]	NS			
Pancreatic insufficiency	No	21.4	1					
	Yes	30.7	1.43	[0.67–3.04]	NS	2.11	[0.68–6.56]	NS
BMI (kg/m ²)	≥ 18.5	27.3	1					
	<18.5	38.2	1.4	[0.84–2.34]	NS	1.47	[0.57–3.75]	NS
Diabetes	No	29.6	1					
	Yes	29.4	0.99	[0.59–1.66]	NS	0.76	[0.31–1.83]	NS
Colonisation with <i>PA</i>	No	27.4	1					
	Yes	30.8	1.12	[0.68–1.85]	NS			
FEV1 (% pred.) ^b	≥ 70	9.7	1					
	55–69	26.8	2.77	[0.84–9.10]	0.08	5.68	[1.24–26.0]	0.026
	40–54	42.9	4.43	[1.41–13.87]	0.005			
	30–39	28.6	2.95	[0.83–10.52]	0.13			
	<30	39.3	4.06	[1.26–13.08]	0.013			
FVC (% pred.) ^b	≥ 70	24.1	1					
	50–69	28.6	1.19	[0.64–2.19]	NS			
	<50	45.2	1.87	[1.09–3.23]	0.029			
Daily treatment time (minutes)	<30	24.3	1					
	30–59	38.3	1.57	[0.80–3.10]	NS			
	>59	26.8	1.1	[0.56–2.16]	NS			
Nebulised treatment	No	25.0	1					
	Yes	32.1	1.28	[0.75–2.18]	NS			
IV antibiotic courses (number/year)	0	18.6	1					
	1–2	36.0	1.93	[1.00–3.70]	0.05			
	>2	35.7	1.92	[1.00–3.63]	0.05			
Oxygen treatment	No	26.0	1					
	Yes	55.0	2.11	[1.30–3.42]	0.016			
Family	Single	35.9	1					
	Married or living together	22.2	0.62	[0.37–1.04]	0.06			
Housing	Living without their parents	21.4	1					
	Living with their parents	53.8	2.51	[1.61–3.92]	<0.001			
Educational level	Less than A level	43.5	1					
	A level to 2 years at university	29.2	0.67	[0.40–1.11]	0.16	0.60	[0.24–1.45]	NS
	≥ 3 years at university	16.7	0.38	[0.19–0.76]	0.004	0.25	[0.08–0.74]	0.013

PR: prevalence-ratio of disability; CI: confidence interval; OR: odds-ratio of disability; NS: not significant; BMI: body mass index; *PA*: *Pseudomonas aeruginosa*; FEV1: forced expiratory volume in one second; FVC: forced vital capacity; IV: intravenous.

^a Logistic regression including the eight variables listed in the table.

^b Excluding lung transplanted patients.

commerce, banking, insurance, socio-cultural action and education. The same trend was noted by Burker et al. who reported that 26% of the CF patients in their study had sedentary work, 45% had light work, 17% had an average job and only 2% performed heavy work [22].

The health professions and occupations involving contact with children are often not recommended for CF patients, due to the higher risk of being exposed to respiratory pathogens. However, 10% of our patients were employed in these sectors.

4.2. Factors associated with work status

Our analysis strategy allowed the associations between work status and many individual factors to be analysed. Most of these parameters did not appear to affect employment status. For

example, work status was unaffected by age, gender or daily time of treatment. This last result was unexpected, but the continued activity of individuals requiring higher levels of treatment was most likely permitted by them being allowed to reduce their working hours. However, this should be confirmed in subsequent studies.

In contrast, one or more courses of IV antibiotics and low FEV1 values were risk factors for disability. Although FEV1 values less than 70% of predicted were associated with disability (PR=3.55 [1.18, 10.71], $p<0.01$), this association was not strengthened at lower FEV1 values. Frangolias et al. also reported doubts about the relevance of a poor FEV1 value as a predictor of disability, as they found no difference in mean FEV1 between ‘employed-limited’ and ‘unemployed-limited’ groups [17]. In addition, an FEV1 of less than 40% of the

predicted value appeared a poor cut-off value for predicting work status. A similar trend was found with the number of IV antibiotic courses. This means that receiving at least one course of IV antibiotics per year or having an FEV1 value of less than 70%, indicates an increase in the risk of disability, but this risk does not grow linearly as their clinical condition deteriorates. On the contrary, educational level was a factor that was clearly associated with disability: the higher it was, the greater the chance of getting a job was.

In the multivariate analysis, FEV1 and educational level remained the only statistically significant factors associated with employment status. This result demonstrates that CF patients should be encouraged to reach the highest level of education possible, because this level of achievement is likely to determine their employability at least as much as the severity of their disease.

Four previous studies investigated the predictors of disability in CF patients [6,7,16,17]. All were cross-sectional studies, but their analytic strategies and results differed. Gillen et al. used logistic regression without adjusting for educational level and found that decline in FEV1, diagnosis of CF in adulthood and female gender were all risk factors for work disability [16]. Burker et al. found that employed people had a higher mean educational level than inactive individuals, but found no difference in age, gender and FEV1 between the two groups [7]. However, many of these patients were being evaluated for lung transplant surgery and their mean FEV1 was already quite low (31.9% of predicted). Hogg et al. chose to evaluate disability by a score of 0–4 and to calculate the correlations between this disability index score and quantitative variables [6]. Their disability index depended on age, quality of life indices and hospital admission rate, but not on FEV1. Frangolias et al. found FEV1 to be the best predictor of disability, but did not indicate which variables were included in their model [17]. Altogether, taking into account the methodological differences, our results are consistent with these previous studies and illustrate the major impact of respiratory function and education in the disability of CF patients.

Our study's limitations include its single-centre design and the lack of information about the employment status of non-participants. We nevertheless studied employment status of more CF patients than any previous study. These patients accounted for two-thirds of our active files within the adult CF centre and since no significant group differences with respect to medical variables were found between the study group and non-participants, they can be considered representative of their peers. Another limitation may be that the patients' quality of life was not evaluated with standardised questionnaires but this can be addressed in further studies and related to the employment status of CF patients.

5. Conclusion

If the employability of CF patients is a new challenge of their adult life, our results show that it is mainly conditioned by their level of education. Higher levels of education should be encouraged because they are likely to allow access to skilled jobs. These types of jobs are generally more amenable to the

workplace adjustments that must accompany their long-term degradation in health if they are to remain employed over a long period of time. It is therefore desirable that all adolescent CF patients and their parents be aware of the importance of counselling in the framework of therapeutic education.

Once patients are integrated into the workplace, working hour reductions offer a way to maintain employment as the disease evolves and the constraints of treatment increase. Therefore, rather than choosing a specific trade, patients with CF should seek careers that allow some flexibility.

References

- [1] Bellis G, Cazes MH, Parant A, Gaimard M, Travers C, Le Roux E, et al. Cystic fibrosis mortality trends in France. *J Cyst Fibros* 2007;6:179–86.
- [2] Munck A, Roussey M. The French nationwide cystic fibrosis newborn screening program: strategy and results. *Arch Pediatr* 2008;15(Suppl 1): S1–6.
- [3] Vaincre la Mucoviscidose. Registre Français de la Mucoviscidose — Bilan des données 2008. Available from Paris, France: INED; 2011 <http://www.vaincrelamuco.org/>.
- [4] Duguépéroux I, Tamalet A, Sermet-Gaudelus I, Le Bourgeois M, Gérardin M, Desmazes-Dufeu N, et al. Clinical changes of patients with cystic fibrosis during transition from pediatric to adult care. *J Adolesc Health* 2008;43:459–65.
- [5] Widerman E. Knowledge, interests and educational needs of adults diagnosed with cystic fibrosis after age 18. *J Cyst Fibros* 2003;2:97–104.
- [6] Hogg M, Braithwaite M, Bailey M, Kotsimbo T, Wilson JW. Work disability in adults with cystic fibrosis and its relationship to quality of life. *J Cyst Fibros* 2007;6:223–7.
- [7] Burker EJ, Sedway J, Carone S. Psychological and educational factors: better predictors of work status than FEV1 in adults with cystic fibrosis. *Pediatr Pulmonol* 2004;38:413–8.
- [8] Havermans T, Colpaert K, Vanharen L, Dupont LJ. Health related quality of life in cystic fibrosis: to work or not to work? *J Cyst Fibros* 2009;8(3): 218–23.
- [9] Abbott J, Hart A, Morton AM, Dey P, Conway SP, Webb AK. Can health-related quality of life predict survival in adults with cystic fibrosis? *Am J Respir Crit Care Med* 2009;179:54–8.
- [10] Demars N, Uluer A, Sawicki GS. Employment experiences among adolescents and young adults with cystic fibrosis. *Disabil Rehabil* 2011;33:922–6.
- [11] DARES. Direction de l'Animation, de la Recherche, des Etudes et des Statistiques. Ministère du Travail. La nomenclature des Familles Professionnelles : FAP-2003; 2008 <http://www.travail-emploi-sante.gouv.fr/etudes-recherche-statistiques-de-76/statistiques-78/metiers-et-qualifications-83/les-familles-professionnelles-231/la-nomenclature-des-familles-2709.html>.
- [12] Welsh MJ, Smith AE. Molecular mechanisms of CFTR chloride channel dysfunction in cystic fibrosis. *Cell* 1993;73:1251–4.
- [13] Knudson RJ, Lebowitz MD, Holberg CJ, Burrows B. Changes in the normal maximal expiratory flow-volume curve with growth and aging. *Am Rev Respir Dis* 1983;127:725–34.
- [14] Herquelot E, Guéguen A, Bonenfant S, Dray-Spira R. Impact of diabetes on work cessation: data from the GAZEL cohort study. *Diabetes Care* 2011;34:1344–9.
- [15] R Development Core Team. R: a language and environment for statistical computing. Vienna, Austria: R Foundation for Statistical Computing; 2009.
- [16] Gillen M, Lallas D, Brown C, Yelin E, Blanc P. Work disability in adults with cystic fibrosis. *Am J Respir Crit Care Med* 1995;152:153–6.
- [17] Frangolias DD, Holloway CL, Vedal S, Wilcox PG. Role of exercise and lung function in predicting work status in cystic fibrosis. *Am J Respir Crit Care Med* 2003;167:150–7.
- [18] DARES. Direction de l'Animation, de la Recherche, des Etudes et des Statistiques. Ministère du Travail — Enquête complémentaire à l'Enquête Emploi 2007, calculs DARES; 2011 <http://www.travail-emploi-sante.gouv.fr/etudes-recherche-statistiques-de-76/>.

- [19] Paris W, Diercks M, Bright J, Zamora M, Kesten S, Scavuzzo M, et al. Return to work after lung transplantation. *J Heart Lung Transplant* 1998;17:430–6.
- [20] Cicutto L, Braidy C, Moloney S, Hutcheon M, Holness DL, Downey GP. Factors affecting attainment of paid employment after lung transplantation. *J Heart Lung Transplant* 2004;23:481–6.
- [21] INSEE. Institut National de la Statistique et des Etudes Economiques. Available from Enquêtes Emploi; 2011 <http://www.insee.fr>.
- [22] Burkner EJ, Trombley C, Sedway J, Parker Yeatts B, Carone S. Vocational attainment of adults with CF: success in the face of adversity. *J Rehabil* 2005;71:22–7.
- [23] Troosters T, Langer D, Vrijnsen B, Segers J, Wouters K, Janssens W, et al. Skeletal muscle weakness, exercise tolerance and physical activity in adults with cystic fibrosis. *Eur Respir J* 2009;33:99–106.