French Cystic Fibrosis Registry

Annual data report 2012

VAINCRE LA MUCOVISCIDOSE

ined
INSTITUT NATIONAL D'ÉTUDES DÉMOGRAPHIQUES

Authors:

Gil BELLIS, Institut national d'études démographiques

Lydie LEMONNIER, Vaincre la Mucoviscidose

Marie SPONGA, Vaincre la Mucoviscidose

Neil ZEGHIDOUR, Vaincre la Mucoviscidose

The authors would like to thank the clinicians of the care centers and their teams.

Suggested citation:

French CF Registry - Annual Data Report 2012

Vaincre la Mucoviscidose and Ined

Paris, March 2014

Website:

www.registredelamuco.org

Editorial

1992-2012…

Over the past two decades, the French CF Registry data have illustrated the significant progress of CF care in France, thanks to all joint actions undertaken by healthcare professionals, researchers and the French CF Association Vaincre la Mucoviscidose.

This period was marked by decisive steps such as the accreditation of expert CF care centres (CRCMs) and the implementation of a national newborn screening program in 2002.
Some examples illustrate this evolution:

- Less than 2000 patients were registered in 1992 among whom 20% of adults, more than 6000 patients and 50% of adults in 2012: these figures reflect the improved completeness of data collection and quality of care;
- Median age at diagnosis…fell from 7 months 20 years ago to 3 months in 2012, an early diagnosis being the key to proper care in specialized centers;
- FEV1, a major criterion for evaluating lung function, has been a good indicator of constant progress in healthcare over the past two decades: the mean value of FEV1 of patients in their twenties rose from 40-45% in 1992 to 70% in 2012.
- Nutritional status, which is another important marker of health status, has progressed as evidenced by the significant 0.8 increase in BMI Z-Scores in all age groups.
- Another evidence of the improvement of both health status and quality of life is the annual number of pregnancies, which has continuously increased, rising from 6 in 1992 to 54 in 2012.
- Over the past 20 years, lung transplantation improved dramatically and particularly over the past decade: with less than 20 transplantations a year between 1992 and 2000, the number of transplantations reached 97 in 2012;
- A falling crude death rate – for the first time less than 10‰ in 2012 – and a continuously-growing life expectancy at birth – estimated at 29 years in 1992 and at 50 years in 2012 – are good illustrations of all these advances.

Let us hope that the Registry will have, by its thirtieth birthday, shown new progress for patient care to treat and, someday, heal cystic fibrosis!

Table of contents

Information

Percentages may not add up exactly to 100 due to rounding

Children are patients under 18 years of age, adults are patients aged 18 or more.

Cystic Fibrosis

Cystic fibrosis is a hereditary disease with autosomal recessive transmission: only subjects who have inherited two mutations – one from the father, the other from the mother – are affected.

The gene responsible for the disease was identified in 1989. It is located on the long arm of chromosome 7 (7q31) and codes for the CFTR protein, a protein involved in the regulation of chloride ion transport across the cell membrane. To date, more than 1,900 mutations have been identified, the most common (encountered in 80% of patients) is the F508del mutation.

Before implementation of the systematic newborn screening program, the most common context for diagnosis was as follows: alerted by clinical symptoms (steatorrhoea, bronchial obstruction, recurrent respiratory infections), the physician would carry out a sweat test. An elevated sweat chloride ions concentration would confirm the diagnosis, and this would be followed by molecular analysis of the *CFTR* gene and determination of the disease causing mutations.

Newborn screening has been systematic in France since 2002. This decision was taken by the Ministry of Health, which entrusted the task to the French association for screening and prevention of disabilities in children (*AFDPHE - Association Française pour le Dépistage et la Prévention des Handicaps de l'Enfant*). The screening technique uses measurement of immunoreactive trypsin (IRT) in the blood at age 3 days and detection of *CFTR* mutations. The IRT protein is more abundant when there is pancreatic abnormality during foetal life and in the first few months of life. Measuring IRT concentrations enables 95-98% of newborn children with cystic fibrosis to be detected, though the test is not specific enough (it picks out some children who do not have cystic fibrosis) and is therefore linked with a molecular analysis.

After looking for the main *CFTR* mutations (F508del and about thirty others), three situations can arise:

– two mutations are identified. The newborn baby and its parents are asked to visit a cystic fibrosis care centre (*CRCM - Centre de Ressources et de Compétences de la Mucoviscidose*) to confirm the diagnosis based on a clinical assessment and a positive sweat test, and to initiate the necessary treatment and monitoring ;

– a single mutation is identified (the probability of not identifying a second mutation is around 15%). The sweat test must be carried out in a specialised centre. If the test is positive, the child is treated in the same way as the previous group. If the test results are negative, information concerning the heterozygous nature of the newborn will be given to the parents during genetic counselling ;

– although the IRT level is high, no mutation is found. The risk that the child has cystic fibrosis is, in this case, below 1%. A second blotting paper sample test is carried out at age 21 days. If a raised IRT level persists at D21, the child is referred to a specialised centre for an additional assessment (sweat test).

On the pathological level, functional abnormalities occur in the digestive tract, the respiratory tract, the sweat glands and the genital tract. This wide range of abnormalities is associated with a broad spectrum of clinical expression, both regarding the age when the first symptoms appear and their subsequent evolution. The severity of respiratory symptoms affects life expectancy in the majority of cases.

Lifelong treatment is time consuming, demanding and aimed at symptomatic relief. It is essentially based on respiratory management (physiotherapy, antibiotic treatment, oxygen therapy, lung transplant for end stage respiratory disease) and digestive and nutritional management (pancreatic enzyme supplements and a hypercaloric diet).

The French Cystic Fibrosis Data Registry

In 1992, the medical Council of the association *Vaincre la Mucoviscidose*, set up a national cystic fibrosis observatory, the *Observatoire national de la mucovicidose* (ONM), with the following objectives:

– improving knowledge on medical and social characteristics of the population with cystic fibrosis and the impact of therapeutics;

– gaining a better understanding of the socioeconomic cost of this disease with a view to obtaining sufficient resources to cover constantly growing needs;

– improving information available to help both parents and patients in their personal choices, and associations and other institutional partners in strategic decisions.

Covering the entire population of patients in France, has since been added to the initial objectives. The association has therefore transformed the ONM into a national cystic fibrosis registry, the *Registre français de la mucovicidose*. This initiative was approved in July 2006 by the committee for protection of personal data in medical research (*Comité Consultatif sur le Traitement de l'Information en matière de Recherche dans le domaine de la Santé, CCTIRS*) and in March 2007 by the data protection agency (*Commission Nationale de l'Informatique et des Libertés, CNIL*). At the end of 2008 and then in 2011, the registry was certified by the national committee of rare disease registries (*Comité National des Registres Maladies Rares*), an organ of the *Institut de Veille Sanitaire* (InVS) and of the *Institut National de la Santé et de la Recherche Médicale* (INSERM).

The population is composed of people with cystic fibrosis followed in the care centres participating in the registry in France (metropolitan France, Reunion Island and Guadeloupe). Data are collected once a year by means of a questionnaire transmitted using Web, paper questionnaires or exports from electronic patient files. The information requested refers to the preceding year and includes semi-anonymous patient identification, diagnosis, medical follow-up, treatments used, anthropometric data, respiratory function, bacteriological data, evolution of the condition and social and family situation. Statistical analysis is performed on anonymized data.

Unless otherwise indicated, the results presented hereafter relate to the population seen during the year 2012 and were produced by cross-sectional analysis of data. Data on patients seen during the year in at least two centres were processed differently. Patients in this category (said to have multiple accounts) were counted only once and allocated to the centre they visited most often during the year.

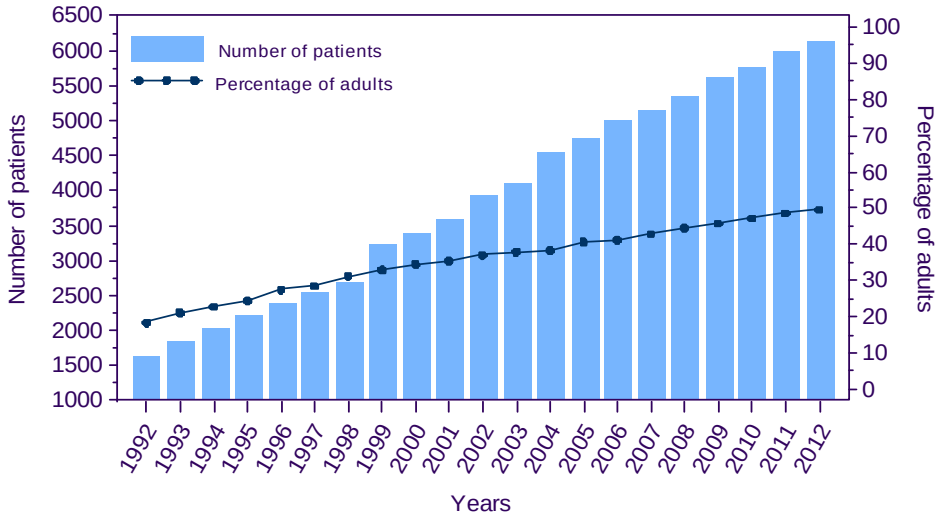
Read precautions before use

Comparisons between the indicators from different countries must be done with care, due to a number of bias coming from the implementation of newborn screening programs, transplantation frequency, socioeconomic status and a limited number of patients in some age groups.

Demographics

Characteristics of the population

Figure 1 - Number of patients seen during the year and % of adults, evolution since 1992



French CF Registry - Annual Data Report 2012

Table 1. Annual evolution of the main indicators

	Years of follow-up									
Indicators	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012
All patients*	4117	4549	4755	5003	5148	5379	5650	5792	6046	6196
Patients seen during the year**	4111	4544	4745	4994	5140	5357	5628	5758	5993	6145
Children	2550	2799	2812	2932	2935	2971	3049	3040	3074	3099
Adults	1561	1745	1933	2062	2205	2386	2579	2718	2919	3046
Over 40 years	124	160	175	196	226	272	329	358	415	469
Men	2157	2368	2497	2595	2686	2786	2916	2958	3100	3171
Women	1954	2176	2248	2399	2454	2571	2712	2800	2893	2974
Mean age (years)	15.8	16	16.3	16.4	16.8	17.3	17.7	18.1	18.7	19.2
Median age (years)	14	14	15	15	15	16	16	16	17	17
Minimum age (years)	0	0	0	0	0	0	0	0	0	0
Maximum age (years)	77	78	74	76	77	78	79	80	87	86

French CF Registry - Annual Data Report 2012

*Patients whose vital status is known, whether they visited or not the CF centre.

**Reference patients for this report, excepted data on survival.

These figures, along with the total number of patients in the registry, compare against a total of 6,095 CF patients registered on 31 December 2012 under the general social security regime (for wage employees) which covers around 88% of the French population.

Demographics

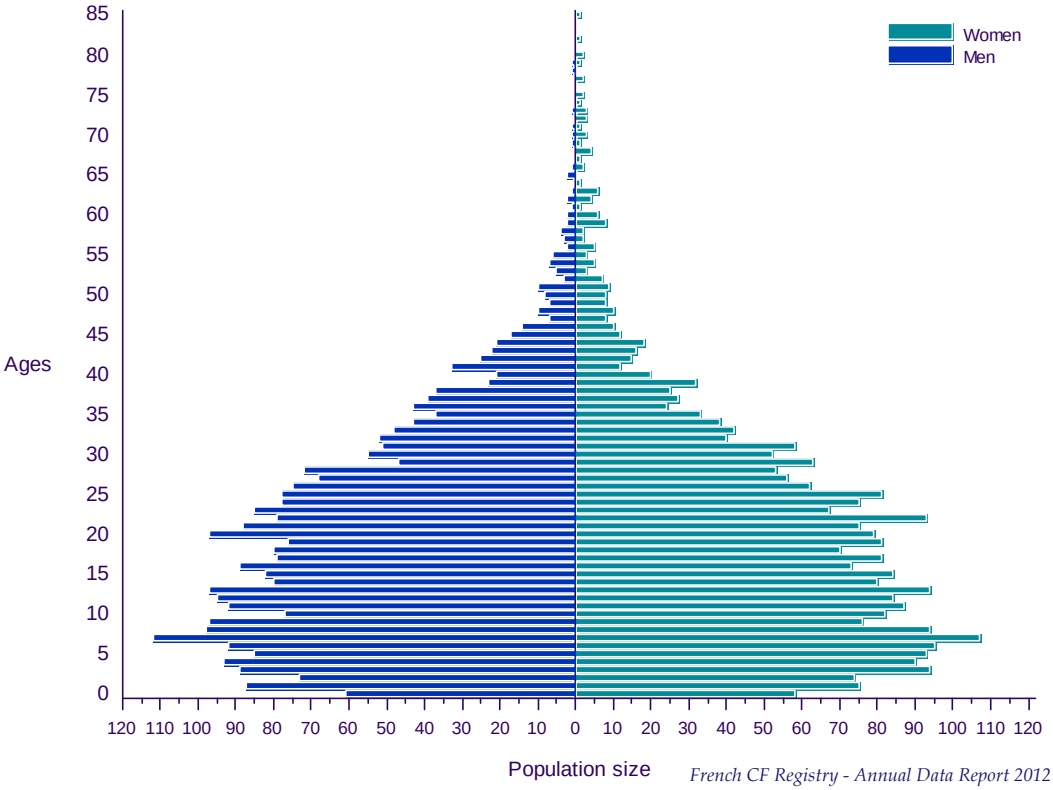
Characteristics of the population

Table 2. Characteristics of the population, by sex and age

	2010		2011		2012	
Characteristics	Men	Women	Men	Women	Men	Women
Patients seen during the year	2958	2800	3100	2893	3171	2974
Children	1532	1508	1567	1507	1578	1521
Adults	1426	1292	1533	1386	1593	1453
Mean age (years)	18.2	18	18.7	18.6	19.2	19.1
Median age (years)	17	16	17	17	18	17

French CF Registry - Annual Data Report 2012

Figure 2. Population pyramid



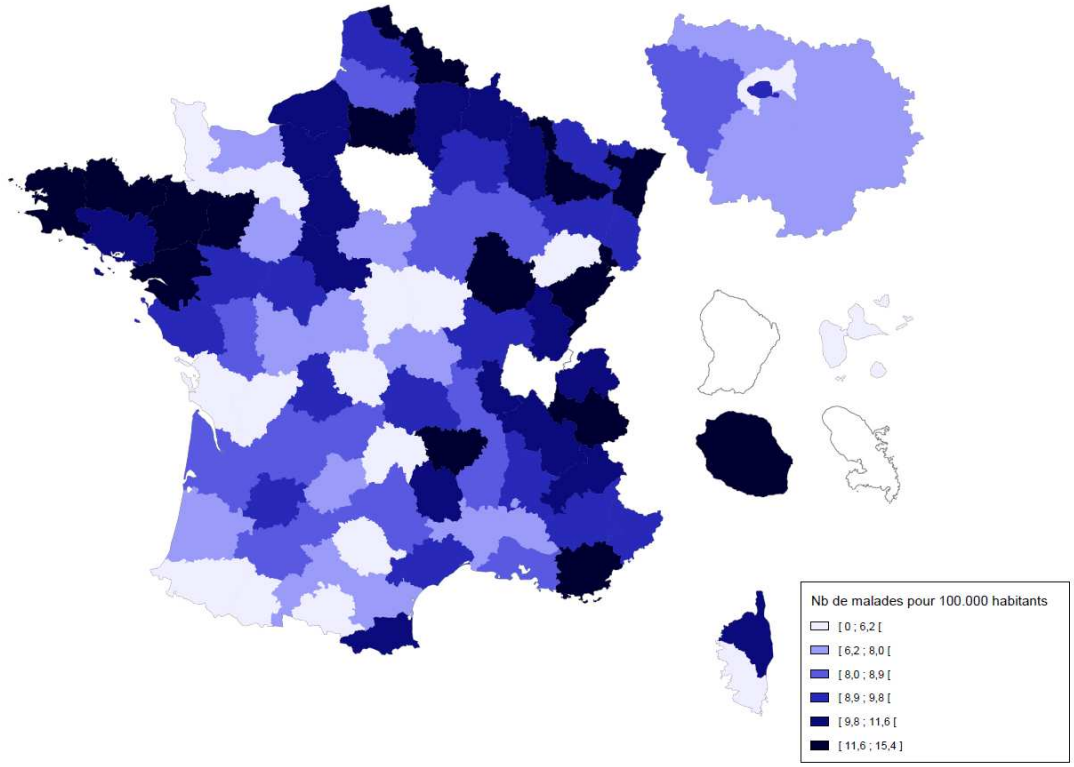
French CF Registry - Annual Data Report 2012

A total of 119 patients are aged below 1 (0 year completed age). Entry into the registry is delayed the first year of life as a certain number of infants diagnosed through neonatal screening in a given year are not registered until the following year.
As an indicative reference, 23 children born in 2011 were diagnosed by neonatal screening in 2012. On the 2011 population pyramid, the number of patients aged 0, which stood at 133, could thus have been 133 + 23 = 156.

Demographics

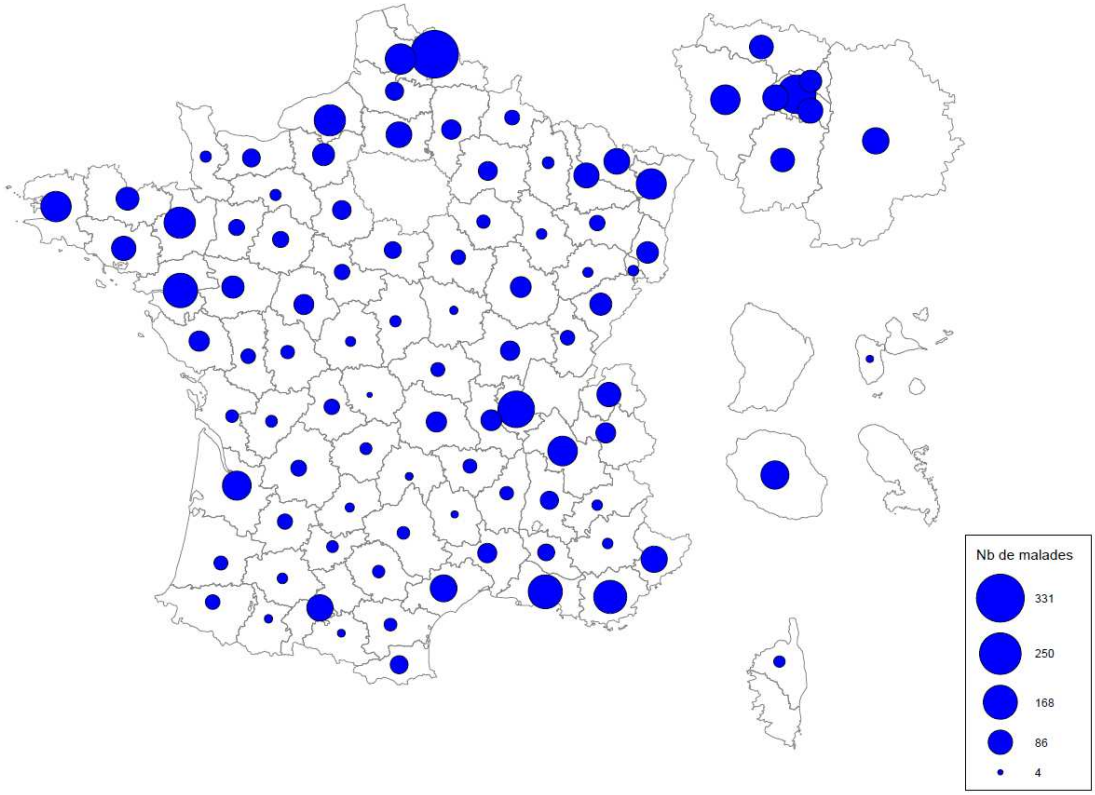
Geographical location

Map 1. Prevalence of cystic fibrosis by « département » of residence (number of patients for 100 000 inhabitants)



French CF Registry - Annual Data Report 2012

Map 2. Localisation of the patients by « département » of residence (absolute numbers)



French CF Registry - Annual Data Report 2012

Demographics

Location by type of centre

Table 3. Patients' characteristics by type of centres

Centres		Patients' characteristics			Age of patients (years)				
Types of centres	Nb	Nb (a)	%	Mean nb by centre	Min	Max	Mean	Median	Inter-quartile
Paediatric CRCMs*	19	2159	35.1	113.6	0	60	9.8	9	9
Adult CRCMs*	12	1929	31.4	160.8	16	80	30.9	29	13
Paediatric/Adult CRCMs*	18	1967	32.0	109.3	0	86	18.1	16	18
Subtotal	*49*	*6055*	*98.5*	*123.6*	*0*	*86*	*19.2*	*17*	*19*
Paediatric local Centres	7	34 (b)	0.6	4.9	0	33	14.8	15	7
Adult local Centres	1	2 (c)	0.0	2.0	32	35	33.5	34	3
Paediatric/Adult local Centres	2	37 (d)	0.6	18.5	4	53	19.0	17	10
Other Centres	4	17 (e)	0.3	4.3	3	50	17.7	16	17
Subtotal	*14*	*90*	*1.5*	*6.4*	*0*	*53*	*17.5*	*16*	*12*
Total	**63**	**6145**	**100**	**97.5**	**0**	**86**	**19.2**	**17**	**19**

French CF Registry - Annual Data Report 2012

* CRCM: Specialised CF Centre (Centre de Ressources et de Compétences de la Mucoviscidose)

Notes :
(a) After checking of patients in the multiple account category (cf page 5)
(b) Including 15 patients also seen by a CRCM.
(c) Including 2 patients also seen by a CRCM.
(d) Including 6 patients also seen by a CRCM
(e) Including 8 patients also seen by a CRCM.

Mortality

Characteristics

Figure 3. Annual number of deaths since 1992

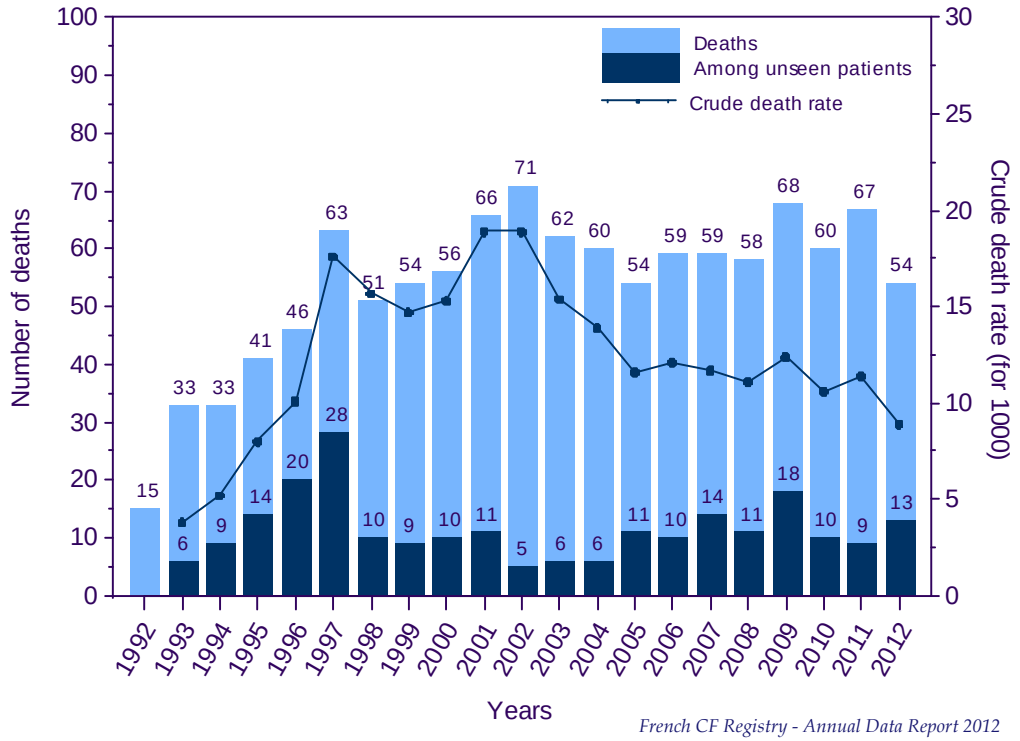


Table 4. Characteristics of mortality

Indicators	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012
Number of deaths	62	60	54	59	59	58	68	60	67	54
- including patients not seen during the year*	6	6	11	10	14	11	18	10	9	13
- including transplanted patients	6	9	9	15	24	26	31	28	34	28
Crude death rate (per 1000)	15.4	13.9	11.6	12.1	11.7	11.1	12.4	10.6	11.4	8.9
Mean age (years)	24.4	22.2	24.3	26.3	27.2	29.1	25.3	29.2	26.4	32
Median age (years)	23	21	22	23	26	28	24	28	25	28
Minimum age (years)	6	0	6	4	2	0	0	0	2	3
Maximum age (years)	65	50	71	76	70	66	73	69	55	88

* Information of the death transmitted while the patient did not visit any centre during the year.

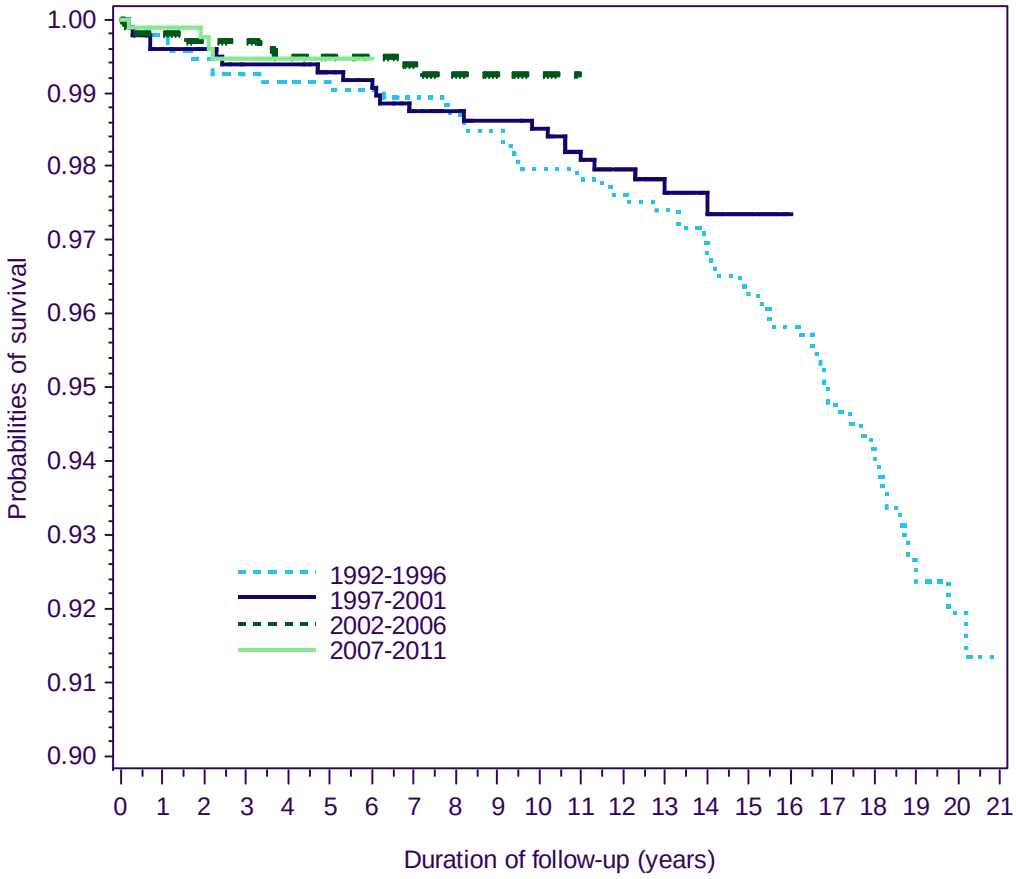
Mortality

Survival analysis

Figure 4. Survival curves by birth cohort (Kaplan-Meier method)

In order to show the evolution of health status of the patients, a survival analysis was performed on 4 birth cohorts; the numbers of patients and of deaths are:

- Births from 1992 to 1996 (in 2012 this cohort was followed during 21 years maximum) : 942 patients, 60 deaths
- Births from 1997 to 2001 (maximum 16 years of follow up) : 965 patients, 22 deaths
- Births from 2002 to 2006 (maximum 11 years of follow up) : 1020 patients, 8 deaths
- Births from 2007 to 2011 (maximum 6 years of follow up) : 887 patients, 4 deaths



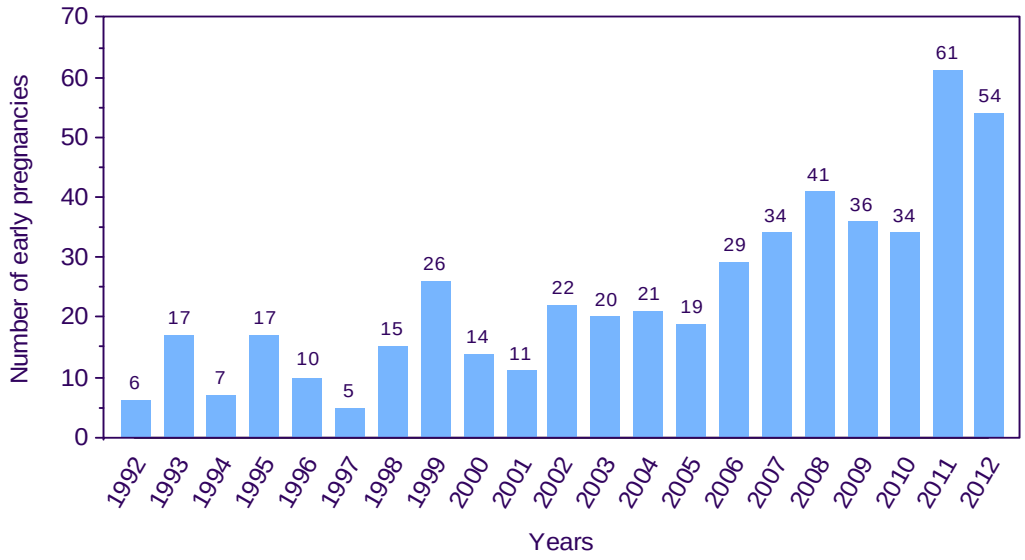
French CF Registry - Annual Data Report 2012

There is no significative survival difference between those cohorts (Log-Rank test = 4.56, p = 0,34).

Survival analysis by genotype and sex are available on annex 1.

Pregnancy

Figure 5. Annual number of early pregnancies, evolution since 1992



French CF Registry - Annual Data Report 2012

Table 5. Early pregnancy characteristics

Characteristics	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012
Number of early pregnancies	20	21	19	29	34	41	36	34	61	54
Pregnancy rates in women aged 15 to 49 years (for 1000)	23.3	22.3	18.2	25.9	29	33.3	27.5	24.5	41.7	35.1
Mean age at onset of pregnancy	26.3	27.2	26.9	27.2	27.8	26.9	27.9	28.3	28.3	27.2

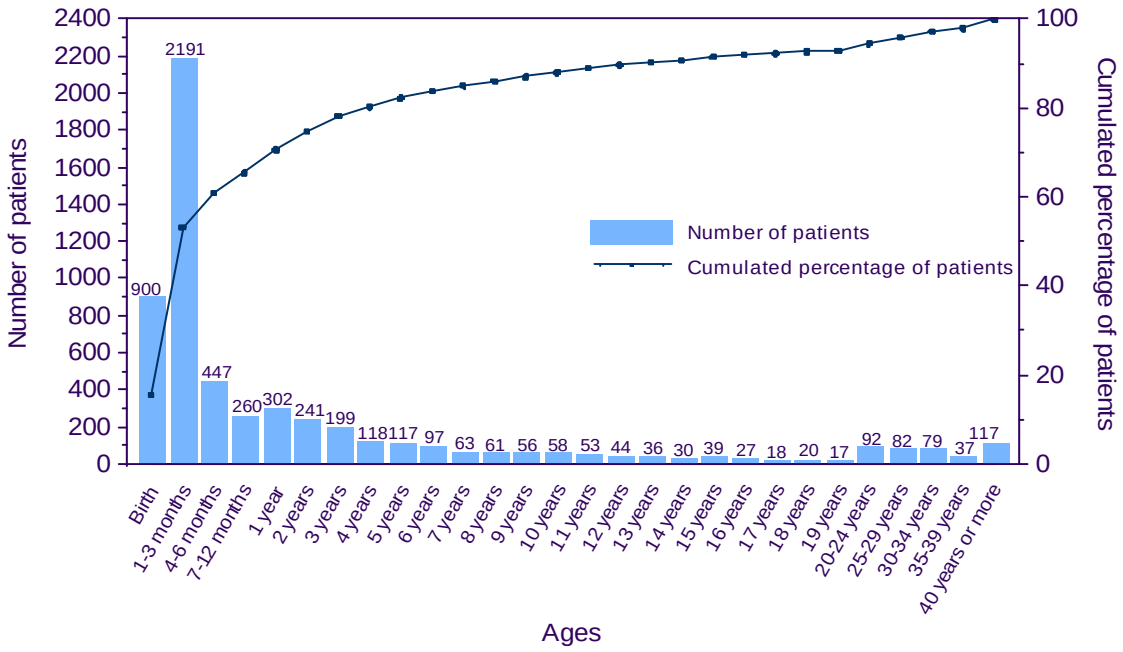
French CF Registry - Annual Data Report 2012

Diagnosis

Main characteristics

Figure 6. Number of patients and cumulative percentage of patients by age at diagnosis

N = 5 801 (number of patients whose age at diagnosis is known).



French CF Registry - Annual Data Report 2012

Diagnosis

Main characteristics

Table 6. Diagnosis characteristics

Characteristics	2010	2011	2012
ALL PATIENTS			
Patients whose age at diagnosis is known - N (%)	5455 (94.7 %)	5670 (94.6 %)	5801 (94.4 %)
Age at diagnosis			
- Median age (months)	3	3	3
- Mean age (years)	4.1	4.3	4.3
- Minimum age (years)	0	0	0
- Maximum age (years)	79	87	79
NEW PATIENTS DIAGNOSED DURING THE YEAR			
Number of patients			
New patients - N (%)	181 (3.1 %)	215 (3.6 %)	188 (3.1 %)
- Including newborn patients	111	133	119
Age at diagnosis (a)			
- Median age (months)	2	1	1
- Mean age (years)	6.4	6.9	5.5
- Minimum age (years)	0	0	0
- Maximum age (years)	69	69	72
Context of diagnosis			
- Prenatal diagnosis - N (%)	8 (4.4 %)	10 (4.7 %)	6 (3.2 %)
- Meconium ileus - N (%)	11 (6.8 %)	26 (12.1 %)	13 (6.9 %)
- Neonatal screening - N (%)	112 (61.9 %)	131 (60.9 %)	124 (66 %)
- Symptoms (excluding MI):			
-- N (%)	56 (30.9 %)	58 (27 %)	41 (21.8 %)
-- Mean age at diagnosis (years)	18.8	22.0	17.5

French CF Registry - Annual Data Report 2012

Notes :
(a) Including family history and antenatal diagnosis.
(b) Patients diagnosed before the results of neonatal screening were excluded.

Among the 188 new patients, 119 were born in 2012. The method used to compile this report (patients seen in a care centre in 2012) means that infants born in 2012 and seen for the first time in 2013 are excluded (cf note page 7).

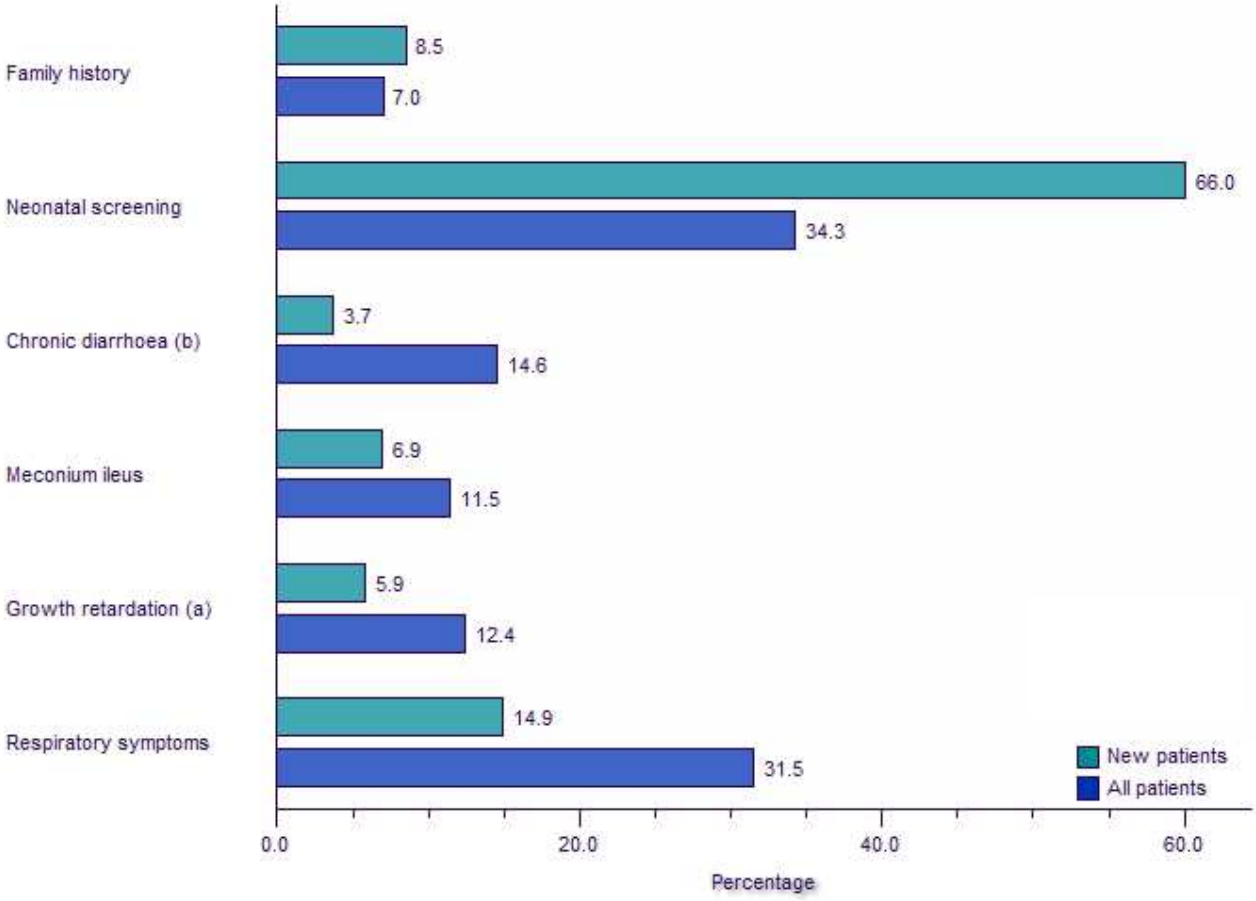
The number of patients diagnosed by neonatal screening (124) given in this report is not the total for France, but represents the patients for whom screening resulted in diagnosis. Patients diagnosed with CF before the screening result was known, (e.g. throught meconium ileus), are not included in the total.

By comparison, 364 new patients were registered in 2012 under the general social security regime (for wage employees).

Diagnosis

Diagnosis signs

Figure 7a. Diagnosis signs (most frequent ones)



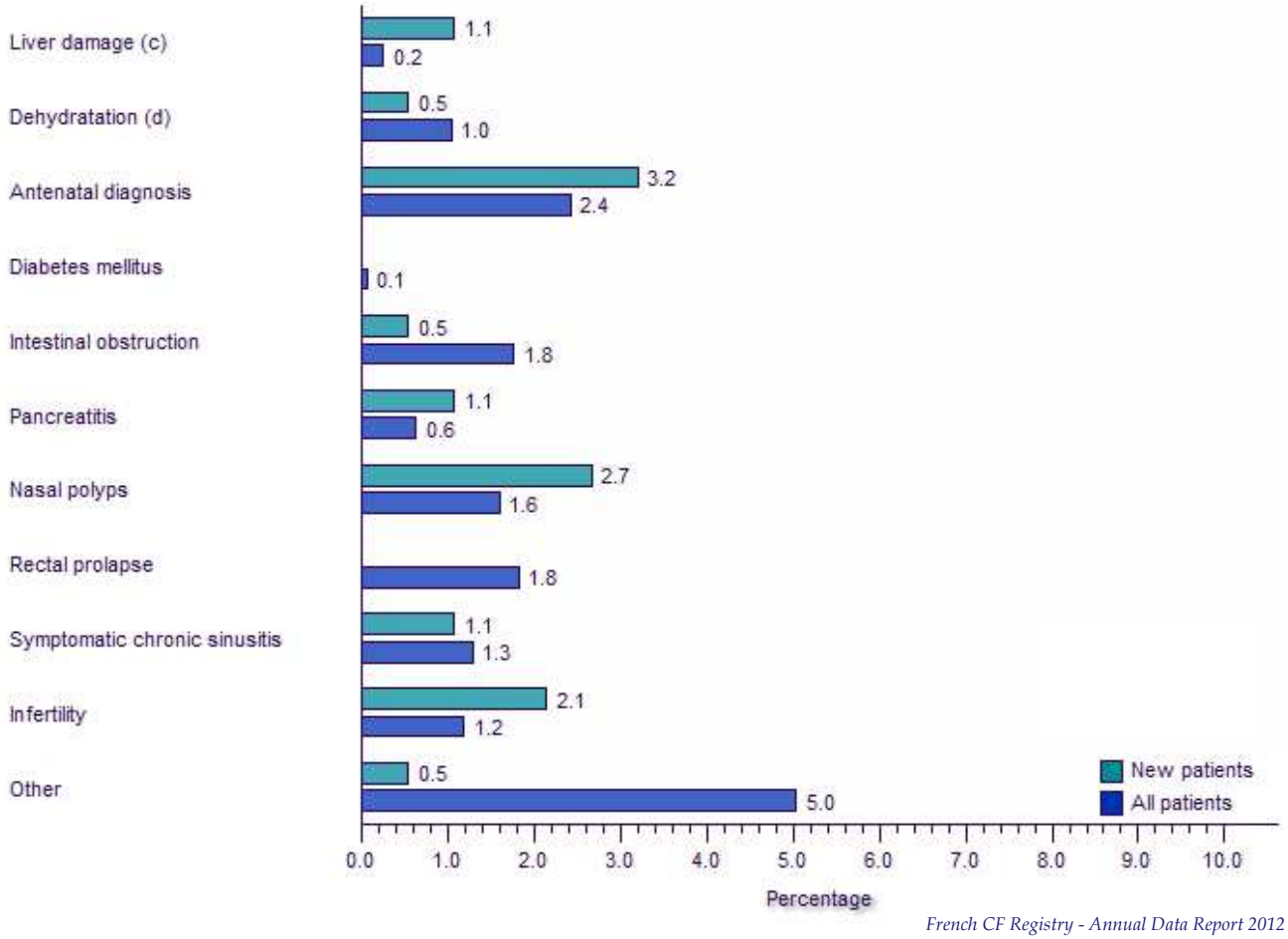
French CF Registry - Annual Data Report 2012

(a) Growth retardation / Malnutrition
(b) Chronic diarrhoea / Steatorrhoea / Malabsorption

Diagnosis

Diagnosis signs

Figure 7b. Diagnosis signs (less frequent ones)



French CF Registry - Annual Data Report 2012

(c) Liver damage / Jaundice / Portal hypertension
(d) Dehydratation / Electrolyte inbalance

Diagnosis

Genotypes

Table 7. Number and proportion of genotypes

Genotypes	Number of patients	%
F508del / F508del	2597	42.3 %
F508del / G542X	195	3.2 %
F508del / N1303K	144	2.3 %
F508del / 2789+5G->A	100	1.6 %
F508del / 1717-1G->A	93	1.5 %
F508del / R117H	95	1.5 %
F508del / R553X	66	1.1 %
F508del / G551D	63	1.0 %
F508del / 3849+10kbC->T	49	0.8 %
F508del / 3272-26A->G	48	0.8 %
F508del / W1282X	41	0.7 %
F508del / Y122X	45	0.7 %
F508del / [delta]I507	43	0.7 %
F508del / L206W	46	0.7 %
F508del / R347P	39	0.6 %
F508del / D1152H	38	0.6 %
F508del / 2183AA->G	35	0.6 %
F508del / 5T	23	0.4 %
F508del / R1162X	29	0.5 %
F508del / A455E	31	0.5 %
F508del / 1078delT	24	0.4 %
F508del / R347H	23	0.4 %
F508del / Y1092X	24	0.4 %
F508del / R334W	21	0.3 %
F508del / 3659delC	20	0.3 %
N1303K / N1303K	21	0.3 %
F508del / 711+1G->T	21	0.3 %
G542X / G542X	21	0.3 %
F508del / S945L	22	0.4 %
F508del / 394delTT	18	0.3 %
F508del / E60X	18	0.3 %
F508del / S1251N	17	0.3 %
F508del / W846X	19	0.3 %
F508del / G85E	19	0.3 %
F508del / R1066C	15	0.2 %
F508del / 1811+1.6kbA->G	18	0.3 %
F508del / 3120+1G->A	16	0.3 %
Y122X / Y122X	14	0.2 %
711+1G->T / 711+1G->T	14	0.2 %
F508del / 621+1G->T	12	0.2 %
F508del / Q220X	10	0.2 %
F508del / I148T	9	0.1 %
Other CFTR genotypes	1596	26.0 %
Subtotal (known genotypes)	5812	94.6 %
F508del / Missing	104	1.7 %
Other / Missing	54	0.9 %
Missing / Missing	175	2.8 %
Subtotal (partial genotypes / Missing)	333	5.4 %
Total	6145	100.0 %

French CF Registry - Annual Data Report 2012

Diagnosis

Genotypes

Table 8. Age of patients by genotype

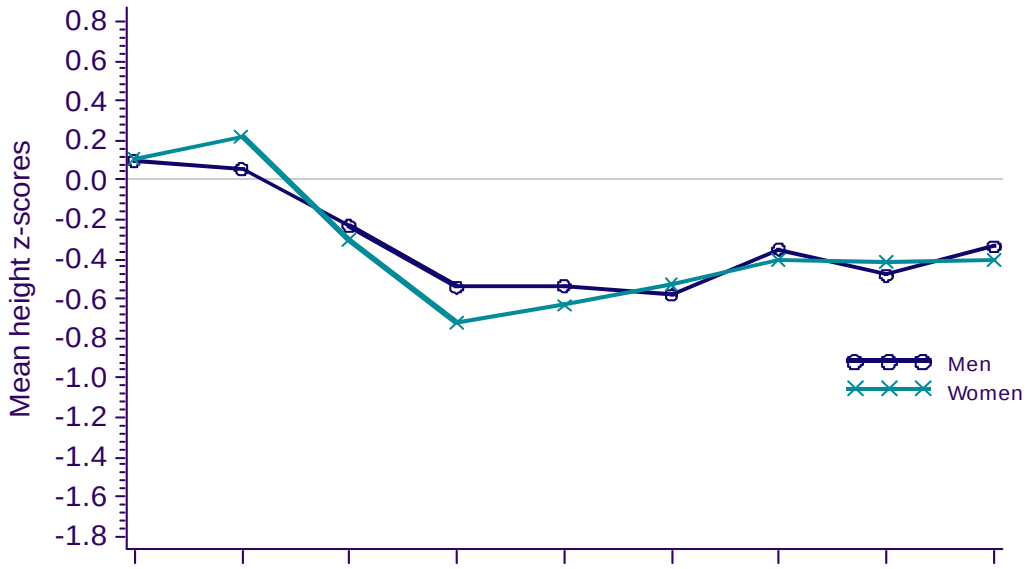
Genotypes	Number	%	Age (years) Mean	Median	Max
F508del / F508del	2597	42.3	18.1	17	59
F508del / Other	2391	38.9	19	16	86
Other / Other	824	13.4	18.7	16	80
F508del / Missing	104	1.7	30.7	28.5	77
Other / Missing	54	0.9	26.9	25	82
Missing / Missing	175	2.8	29.5	26	79

French CF Registry - Annual Data Report 2012

Anthropometry

Height and Weight

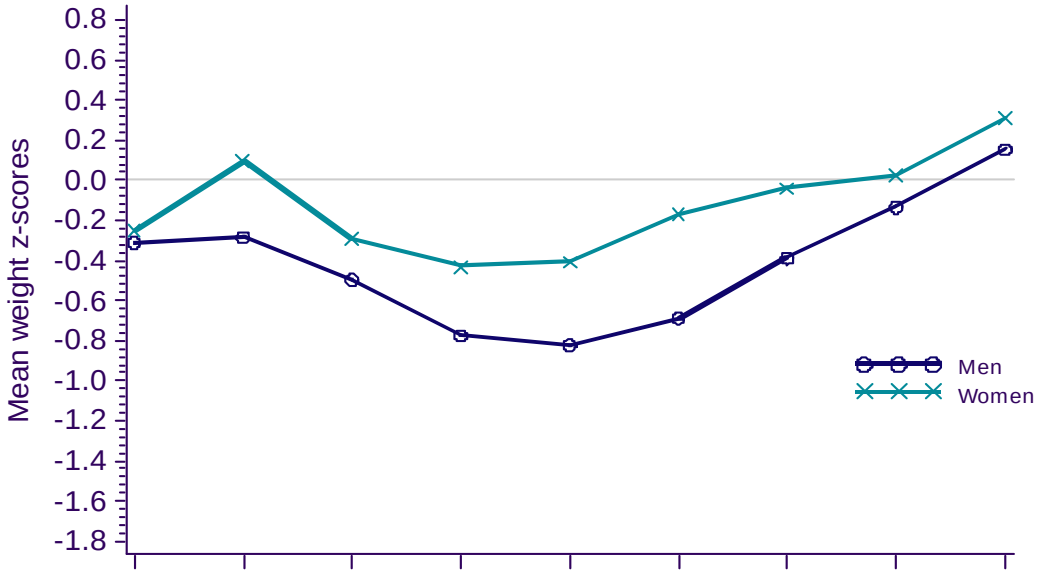
Figure 8. Mean height z-scores, by age group



Age groups (years)	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40 +
Men	0.10	0.06	-0.23	-0.54	-0.53	-0.58	-0.35	-0.48	-0.33
Women	0.11	0.22	-0.30	-0.72	-0.63	-0.53	-0.40	-0.41	-0.40

French CF Registry - Annual Data Report 2012

Figure 9. Mean weight z-scores, by age group



Age groups (years)	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40 +
Men	-0.31	-0.28	-0.49	-0.77	-0.82	-0.69	-0.39	-0.13	0.15
Women	-0.25	0.10	-0.29	-0.43	-0.41	-0.17	-0.04	0.03	0.31

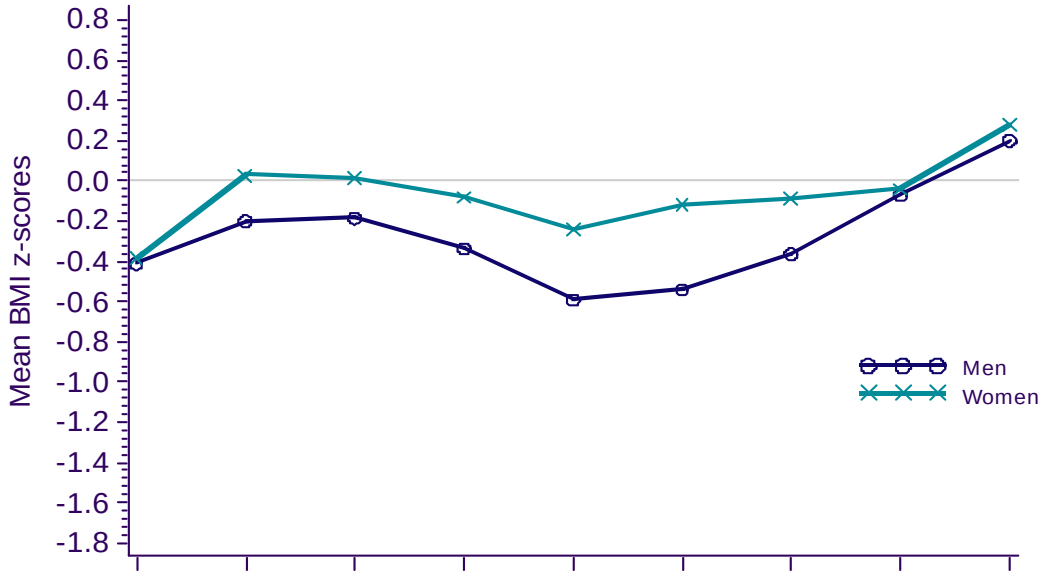
French CF Registry - Annual Data Report 2012

Height and weight z-scores have been calculated with respect to the French reference population (Sempé M., 1997, *Auxologie – Méthode et séquences*, Méditions, Lyon, 205 p.).

Anthropometry

Body Mass Index (BMI)

Figure 10. Mean BMI z-scores, by age group



Age groups (years)	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40 +
Men	-0.41	-0.20	-0.18	-0.34	-0.59	-0.54	-0.36	-0.07	0.20
Women	-0.38	0.03	0.01	-0.08	-0.24	-0.12	-0.09	-0.04	0.28

French CF Registry - Annual Data Report 2012

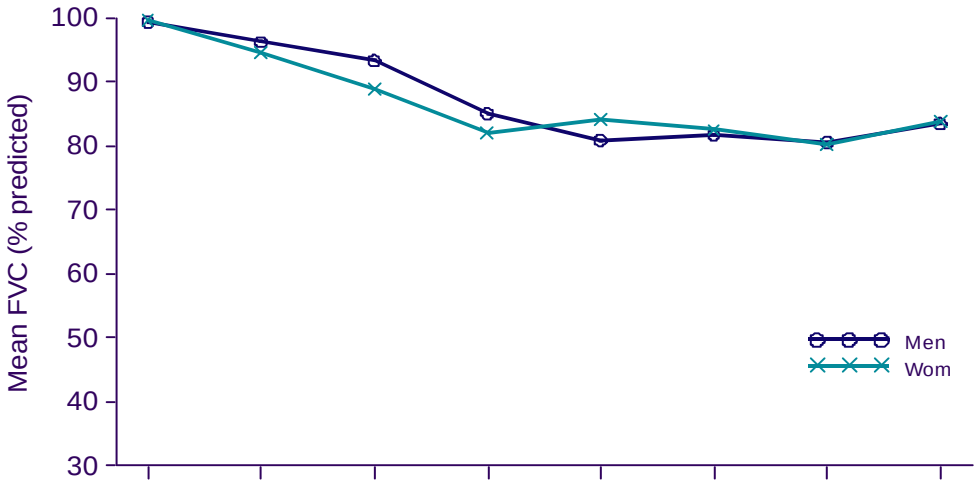
The z-score is a anthropometric reduced centered variable (Z = [measure-mean]/standard deviation), ajusted for sex and age; the mean and standard deviation are taken from the French reference population with the same sex and age as the patient. This index measures the difference with population norms and a negative score means growth retardation.

The BMI z-score was calculated with respect to the French reference population (Rolland-Cachera MF *et al.* A. Body Mass Index variations: centiles from birth to 87 years. Eur J Clin Nutr 1991;45:13-21).

Spirometry

In 2012, 94.6 % of the patients aged 6 or more had at least one spirometry (as in 2011 and compared with 92.4% in 2010).

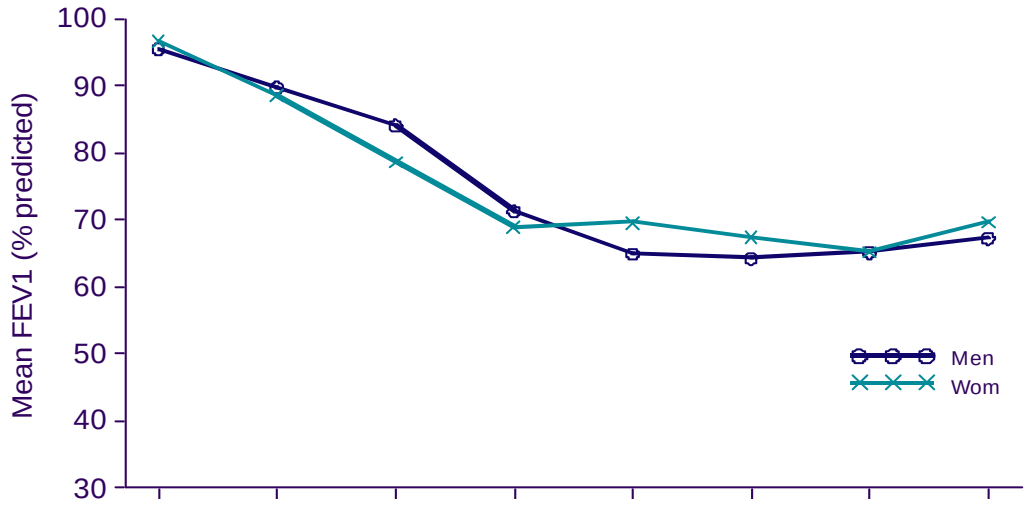
Figure 11. Mean FVC (% predicted), by age group



Age groups (years)	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total
Men	99.5	96.3	93.4	85.1	81.0	81.9	80.6	83.5	88.9
Women	99.8	94.7	89.0	82.1	84.2	82.6	80.3	83.8	88.2
All patients	99.6	95.5	91.2	83.7	82.5	82.2	80.5	83.7	88.6

French CF Registry - Annual Data Report 2012

Figure 12. Mean FEV_1 (% predicted)*, by age group



Age groups (years)	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total
Men	95.6	89.8	84.1	71.3	65.0	64.3	65.2	67.3	77.5
Women	96.7	88.8	78.7	69.0	69.7	67.4	65.3	69.7	77.6
All patients	96.1	89.3	81.5	70.2	67.2	65.8	65.2	68.5	77.5

French CF Registry - Annual Data Report 2012

* Since 2011, the best FEV1 of the year is collected instead of the last of the year.

The pulmonary function tests need an active participation of the patient, difficult to obtain before 6 years of age.
The forced vital capacity (FCV) and the forced expiratory volume in the first second (FEV1) are given in % predicted (Knudson *et al*. Changes in the normal maximal expiratory flow-volume curve with growth and aging. *Am Rev Respir Dis 1983*, 127, pp. 725-734).

See appendix 2 for additional information on spirometry and transplantation.

Spirometry

Figure 13. FEV_1 (% predicted) classes

Values of FEV1% predicted are classified in four « functional » groups according to various degrees of bronchial obstruction.

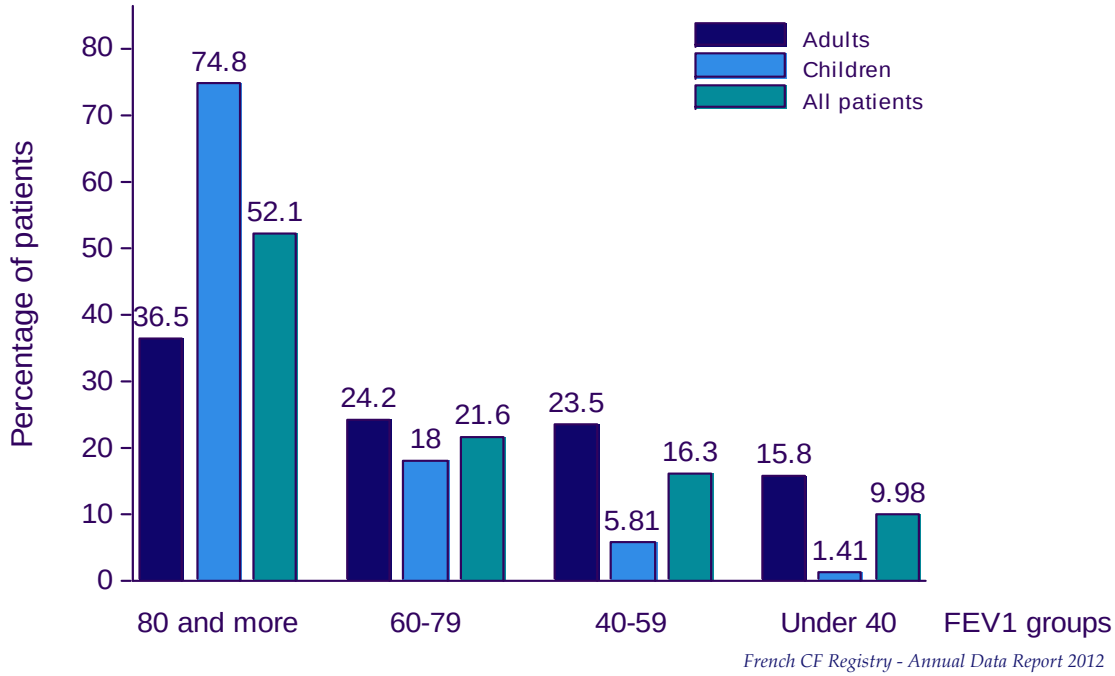
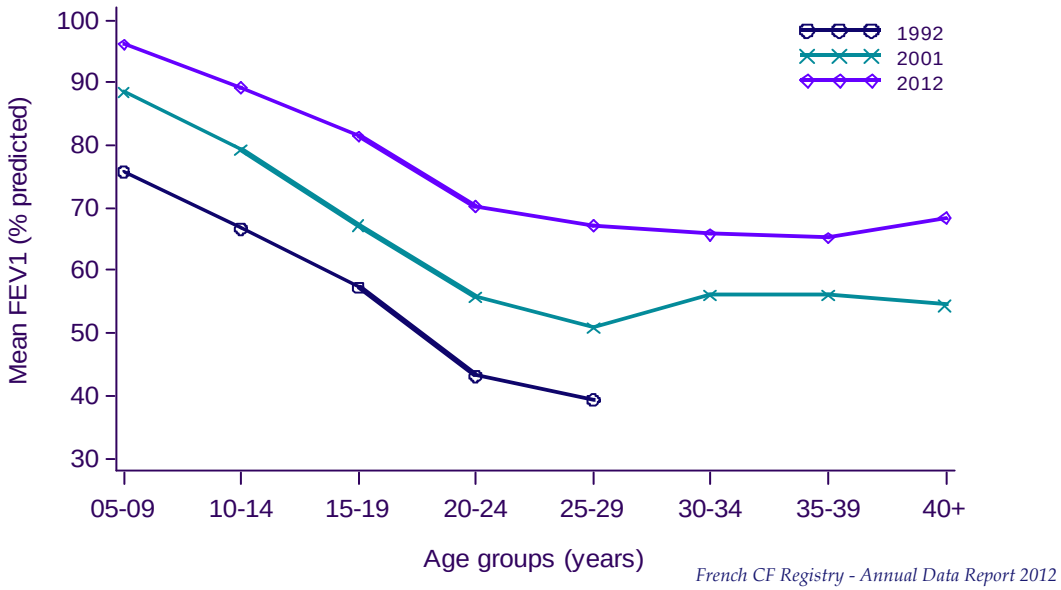


Figure 14. Mean FEV_1 (% predicted)* in 2012 compared with 1992 and 2001, by age



* In 1992 and 2001 was collected the last FEV1 of the year. In 2012, it was the best value of the year.

Microbiology

Table 9. Sputum cultures

Patients with at least one sputum	N	*Proportion (%)*
All patients	5603	*91.2 %*
Children	3024	*97.6 %*
Adults	2579	*84.7 %*

French CF Registry - Annual Data Report 2012

In 2012, 91.2% of the patients had at least one sputum culture; this proportion remains steady as it was 91.7% % in 2011 and 92,9 % in 2010. Among the patients without sputum culture (N=542), 55.7 % of them were transplanted.

Table 10. Distribution of the germs

	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%*
All patients	*794*	*949*	*868*	*795*	*816*	*655*	*479*	*320*	*469*	*6145*	.
Patients with at least one sputum	**751**	**929**	**862**	**777**	**731**	**563**	**377**	**259**	**354**	**5603**	**91.2%**
Normal culture	438	467	281	196	97	72	58	36	58	1703	27.7 %
Achromobacter xylosoxidans	12	31	51	55	58	47	32	16	27	329	5.4 %
Aspergillus	24	121	241	283	288	202	131	74	99	1463	23.8 %
Burkholderia cepacia	2	6	12	24	24	10	13	9	5	105	1.7 %
Haemophilus influenzae	314	356	251	161	144	60	43	30	51	1410	22.9 %
Atypical mycobacteria	.	3	21	33	28	20	7	7	16	135	2.2 %
Pneumococcus	96	94	19	18	12	7	7	2	6	261	4.2 %
Pseudomonas aeruginosa, including:	142	223	307	376	459	383	271	190	219	2570	41.8 %
- Chronic P. aeruginosa	4	39	93	177	313	266	173	134	148	1347	21.9 %
- Multidrug resistant P. aeruginosa	1	4	28	44	92	92	65	45	59	430	7.0 %
Staphylococcus, including:	445	648	725	595	512	337	210	117	152	3741	60.9 %
- MSSA	428	615	662	544	435	267	170	94	128	3343	54.4 %
- MRSA	14	56	85	63	79	65	47	23	20	452	7.4 %
Stenotrophomonas maltophilia	75	68	121	118	97	58	28	14	28	607	9.9 %
Streptococcus (non pneumoniae)	43	69	38	45	47	50	25	9	36	362	5.9 %

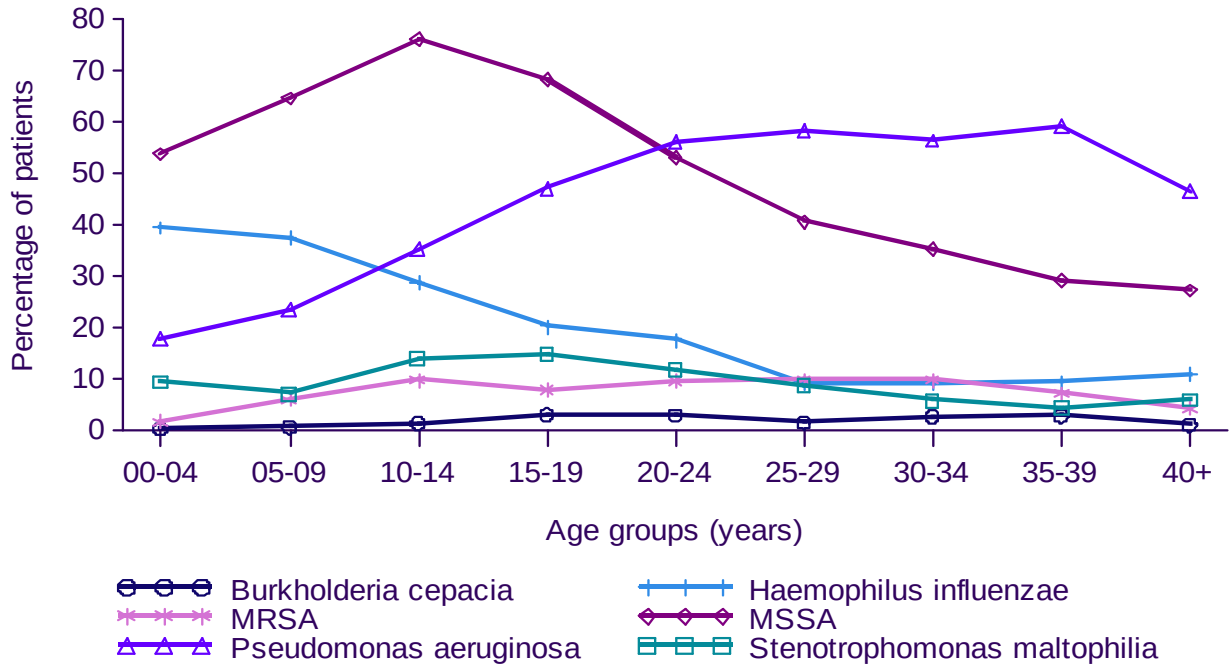
French CF Registry - Annual Data Report 2012

* Percentage with respect to the entire population.

Chronic colonization: more than 50 % of positive test results in the last 12 months (with at least 4 tests during this period) and/or significant increase in anti-PA antibodies (according to the laboratory).
Multi-resistant colonization: resistant to all the antibiotics in at least two antibiotic classes.

Microbiology

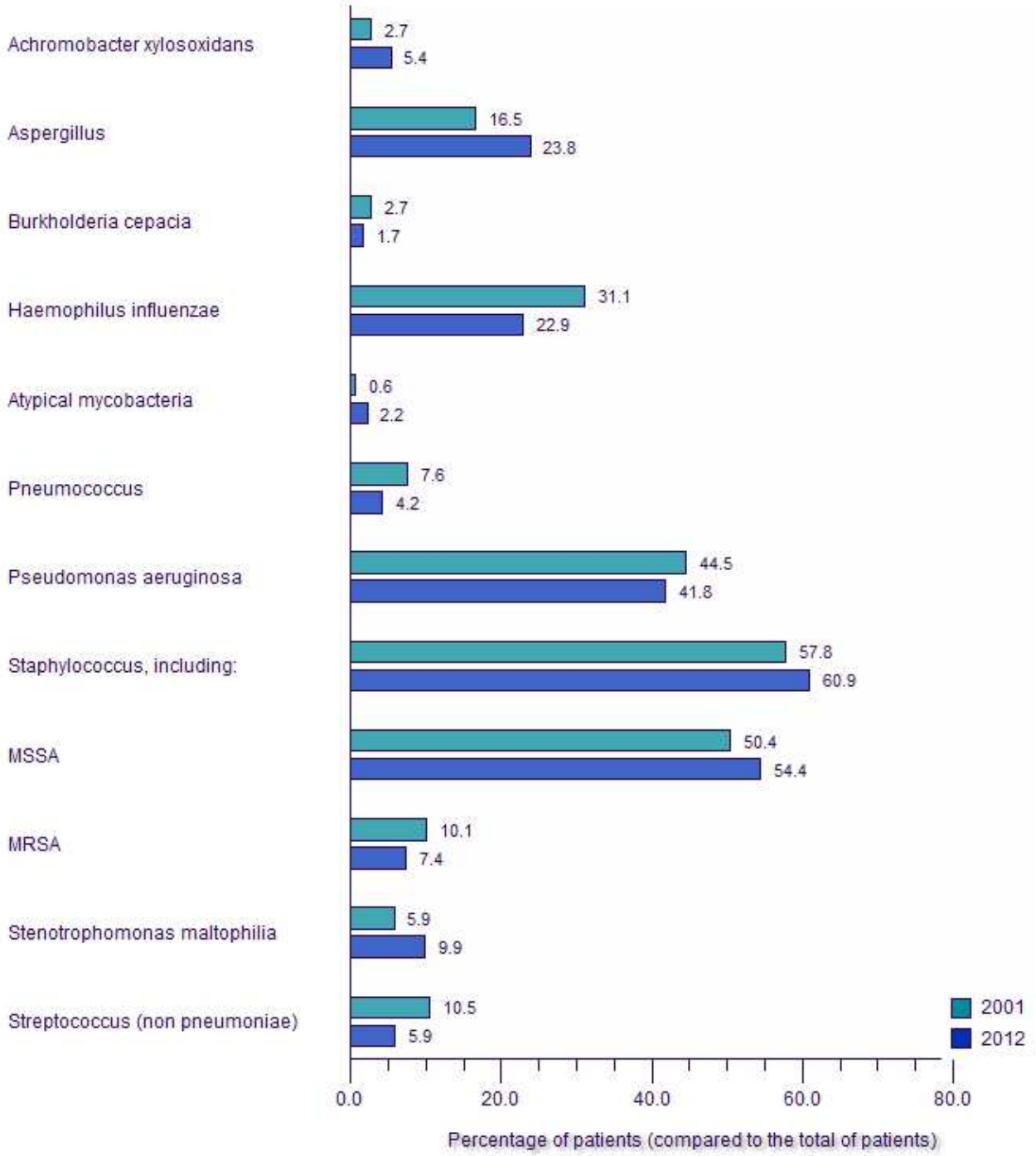
Figure 15. Clinically important bacteria



French CF Registry - Annual Data Report 2012

Microbiology

Figure 16. Comparison of germs in 2012 and in 2001



Complications

Respiratory

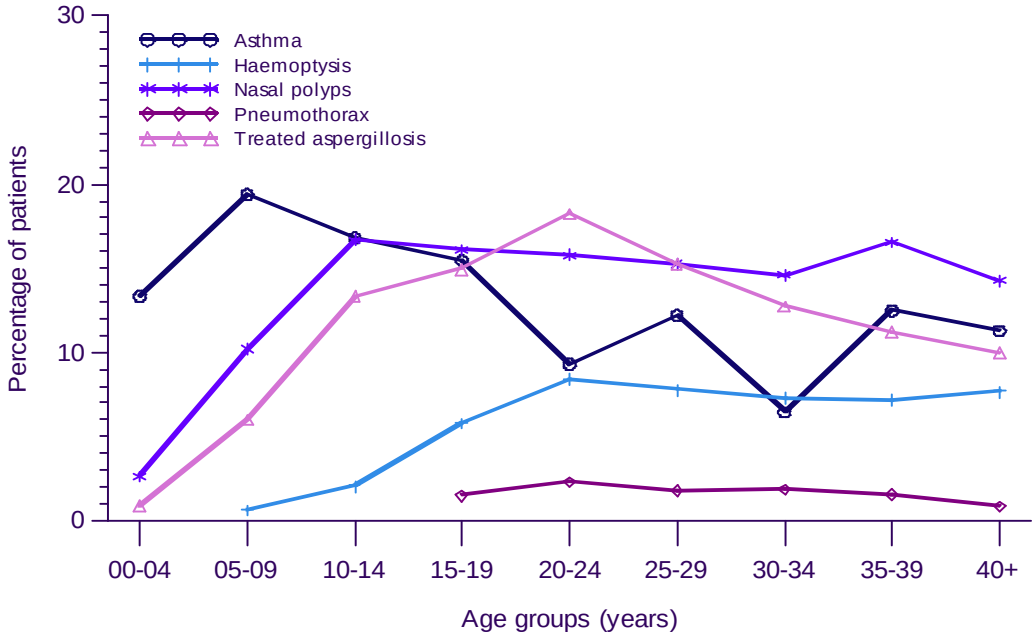
Table 11. Respiratory complications

	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%
All patients	*794*	*949*	*868*	*795*	*816*	*655*	*479*	*320*	*469*	*6145*	.
Treated aspergillosis	7	57	116	119	149	100	61	36	47	692	11.3 %
Asthma	106	184	146	123	76	80	31	40	53	839	13.7 %
Haemoptysis	.	6	18	46	69	51	35	23	36	284	4.6 %
Pneumothorax	.	.	.	12	19	12	9	5	4	61	1.0 %
Nasal polyps	21	97	145	128	129	100	70	53	67	810	13.2 %

French CF Registry - Annual Data Report 2012

Figure 17. Respiratory complications

Percentage of age groups reporting complications



French CF Registry - Annual Data Report 2012

Complications

Gastro-intestinal

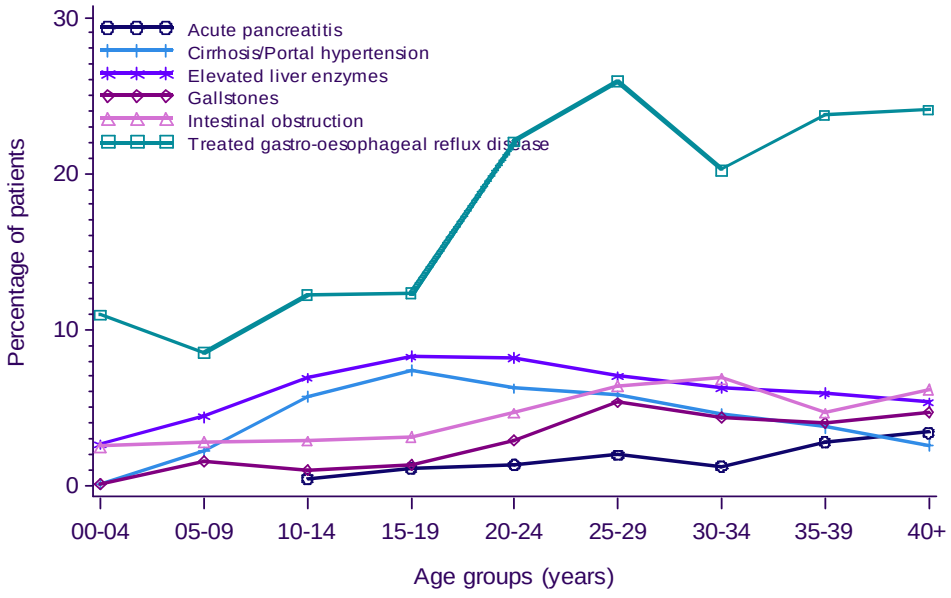
Table 12. Gastro-intestinal complications

	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%
All patients	*794*	*949*	*868*	*795*	*816*	*655*	*479*	*320*	*469*	*6145*	.
Gallstones	1	15	9	11	24	35	21	13	22	151	2.5 %
Cirrhosis/Portal hypertension	1	21	50	59	51	38	22	12	12	266	4.3 %
Elevated liver enzymes	21	42	60	66	67	46	30	19	25	376	6.1 %
Abnormal exocrine pancreatic function	625	757	748	690	716	570	397	259	295	5057	82.3 %
Intestinal obstruction	20	27	25	25	38	42	33	15	29	254	4.1 %
Acute pancreatitis	.	.	4	9	11	13	6	9	16	68	1.1 %
Treated gastro-oesophageal reflux disease	87	81	106	98	180	170	97	76	113	1008	16.4 %

French CF Registry - Annual Data Report 2012

Figure 18. Gastro-intestinal complications

Percentage of age groups reporting complications



French CF Registry - Annual Data Report 2012

Complications

Diabetes mellitus

Table 13. Diabetes mellitus and degenerative complications of diabetes

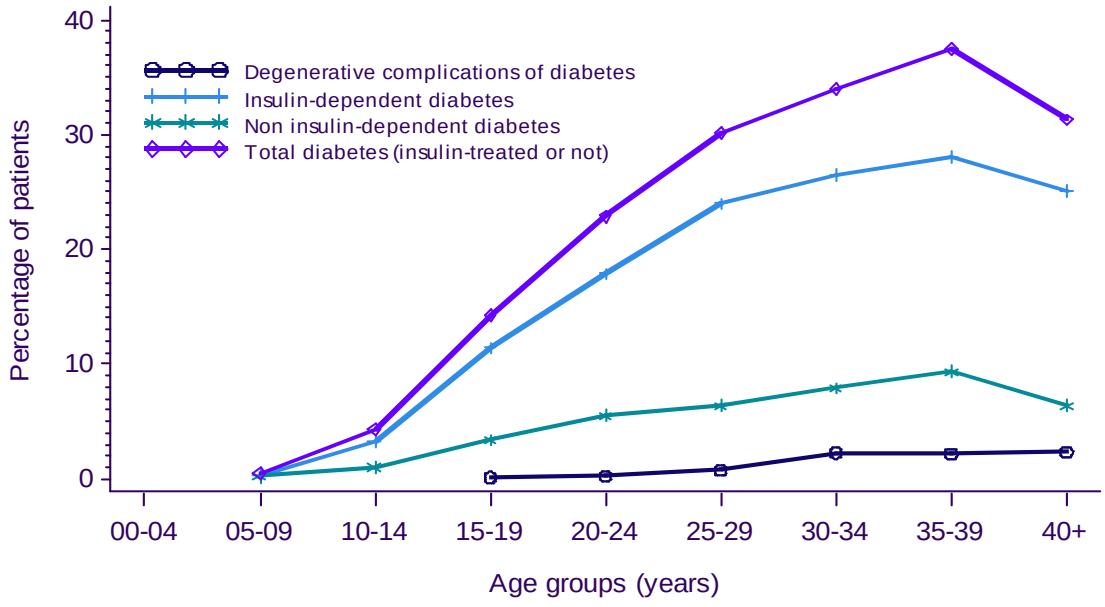
	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%
All patients	*794*	*949*	*868*	*795*	*816*	*655*	*479*	*320*	*469*	*6145*	.
Degenerative complications of diabetes	.	.	.	1	2	5	11	7	11	37	0.6 %
Non insulin-dependent diabetes	.	2	9	27	45	42	38	30	30	223	3.6 %
Insulin-dependent diabetes	.	3	29	91	146	157	127	90	118	761	12.4 %
Total diabetes (ID and non ID diabetes)	.	5	38	114	187	198	163	120	147	972	15.8 %

French CF Registry - Annual Data Report 2012

The line « Total diabetes » sums the number of patients having at least one type of diabetes. Among the 972 patients, 12 patients presented with both types of diabetes during the year.

Figure 19. Diabetes mellitus and degenerative complications of diabetes

Percentage of age groups reporting complications



French CF Registry - Annual Data Report 2012

Complications

Other complications

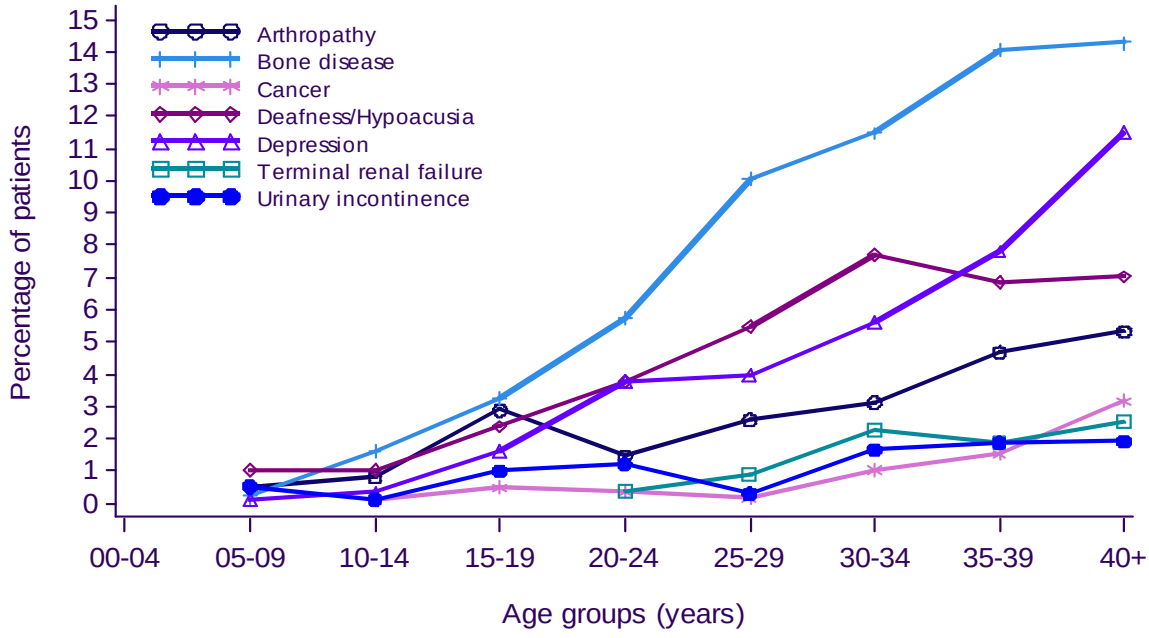
Table 14. Other complications

	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%
All patients	*794*	*949*	*868*	*795*	*816*	*655*	*479*	*320*	*469*	*6145*	.
Arthropathy	.	5	7	23	12	17	15	15	25	119	1.9 %
Cancer	.	.	1	4	3	1	5	5	15	34	0.6 %
Depression (evaluated and followed)	.	1	3	13	31	26	27	25	54	180	2.9 %
Urinary incontinence	.	5	1	8	10	2	8	6	9	49	0.8 %
Terminal renal failure	.	.	.	.	3	6	11	6	12	38	0.6 %
Bone disease	.	2	14	26	47	66	55	45	67	322	5.2 %
Deafness/Hypoacusia	.	10	9	19	31	36	37	22	33	197	3.2 %

French CF Registry - Annual Data Report 2012

Figure 20. Other complications

Percentage of age groups reporting complications



French CF Registry - Annual Data Report 2012

Organ transplant

Tableau 15. Characteristics of the patients on waiting list and of transplant recipients

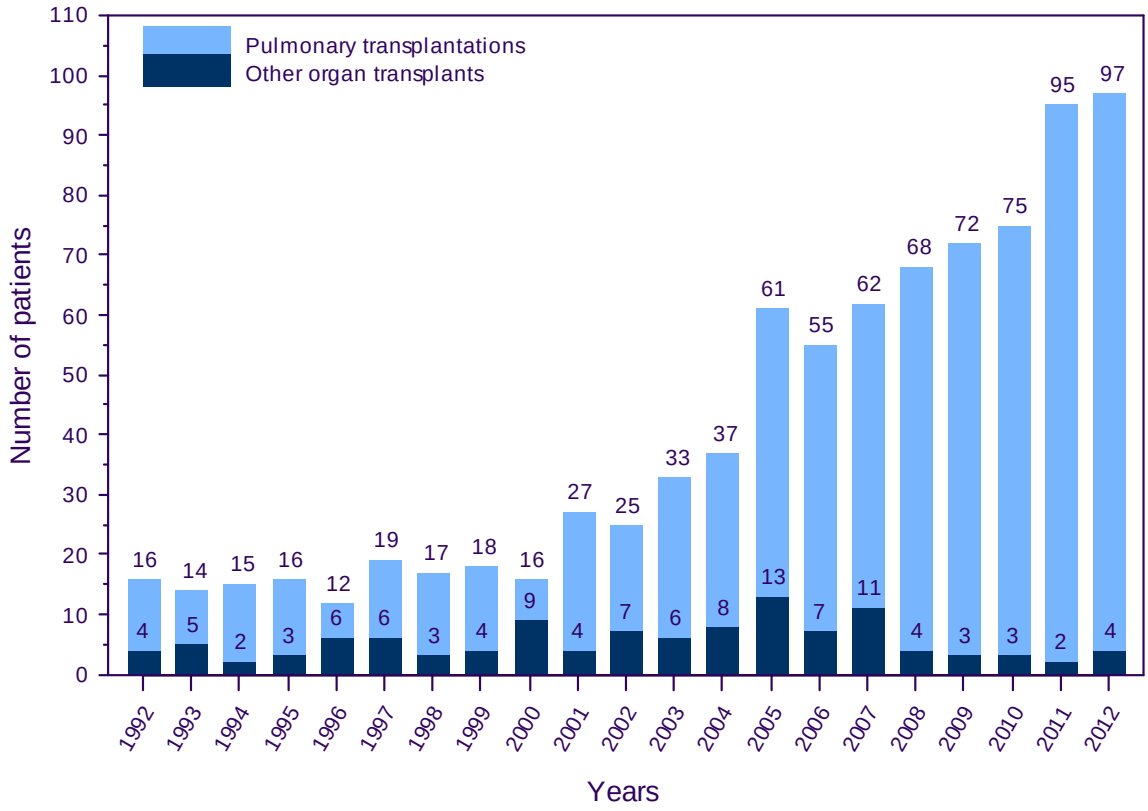
Characteristics	All years	In 2012
WAITING LIST	All waiting patients	Listed in 2012
Nb of patients	**153**	**96**
Mean age (years)	28.8	27.6
Extremes of age (years)	6-55	6-55
Deaths on waiting list	4	3
TRANSPLANTATION	All transplanted*	Transplanted in 2012
Nb of patients	573	101
Transplant type:		
- bilateral lung - N (%)	514 (89.7 %)	92 (91.1 %)
- liver - N (%)	20 (3.5 %)	2 (2.0 %)
- kidney - N (%)	24 (4.2 %)	
- other organs - N (%)	9 (1.6 %)	2 (2.0 %)
Combined transplantations:		
- heart-lung - N (%)	30 (5.2 %)	1 (1.0 %)
- heart-lung / liver - N (%)	2 (0.3 %)	
- bilateral lung / liver - N (%)	13 (2.3 %)	3 (3.0 %)
- bilateral lung / kidney - N (%)	3 (0.5 %)	1 (1.0 %)
- liver / kidney - N (%)	2 (0.3 %)	
- other combined transplant - N (%)	6 (1.0 %)	
Mean age (years)	31.7	29.3
Extremes of age (years)	6-60	6-55
Post-transplantation deaths	20	7

French CF Registry - Annual Data Report 2012

**45 patients underwent two or more organ transplants.*

Transplantations

Figure 21. Annual number of transplanted patients, since 1992



French CF Registry - Annual Data Report 2012

Inpatient and outpatient visits

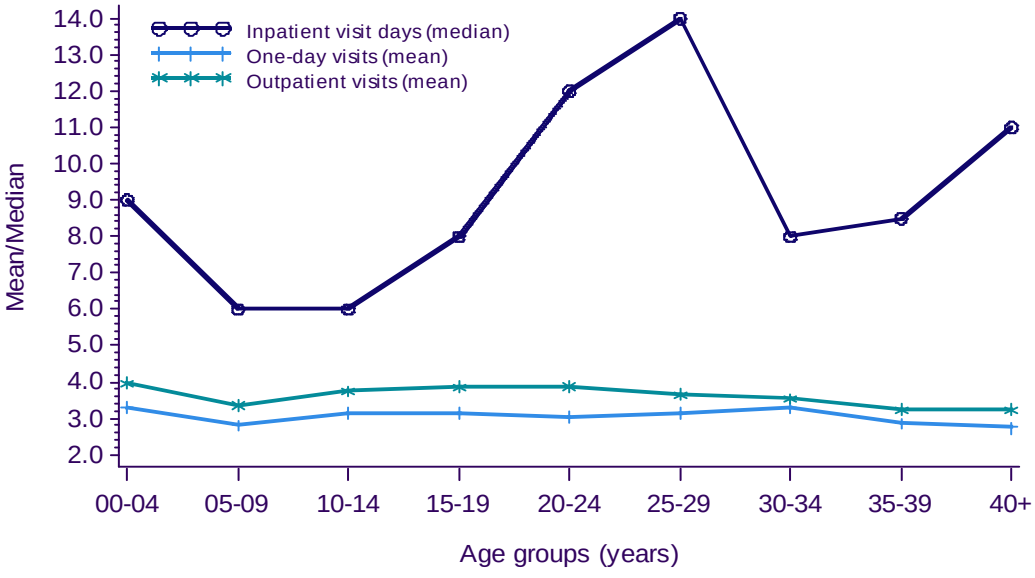
Table 16. Characteristics of the visits

	Age groups (years)									
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total
All patients	*794*	*949*	*868*	*795*	*816*	*655*	*479*	*320*	*469*	*6145*
< 4 visits* per year	154	256	129	169	217	198	147	103	191	1564
>= 4 visits* per year	640	693	739	626	599	457	332	217	278	4581
Outpatient visits	.	.	.	.	.	.	.	.	.	.
Median	4	3	3	3	3	3	3	3	3	3
Mean	4	3.4	3.8	3.9	3.9	3.7	3.5	3.2	3.2	3.7
One-day visits										
Median	3	2	2	2	2	2	2	2	2	2
Mean	3.3	2.8	3.1	3.1	3	3.1	3.3	2.9	2.7	3.1
Inpatient visits										
Median	1	1	1	2	2	2	1	1	2	1
Mean	1.6	1.7	2.2	2.2	2.3	2.6	2.2	2.2	2.4	2.2
Days (median)	9	6	6	8	12	14	8	8.5	11	9
Days (mean)	19	11.5	14.6	15.6	24.6	23.1	17.8	19.9	23.9	19.2

French CF Registry - Annual Data Report 2012

* Outpatient, One-day and Inpatient visits.

Figure 22. Visits, by age



French CF Registry - Annual Data Report 2012

Therapeutic management

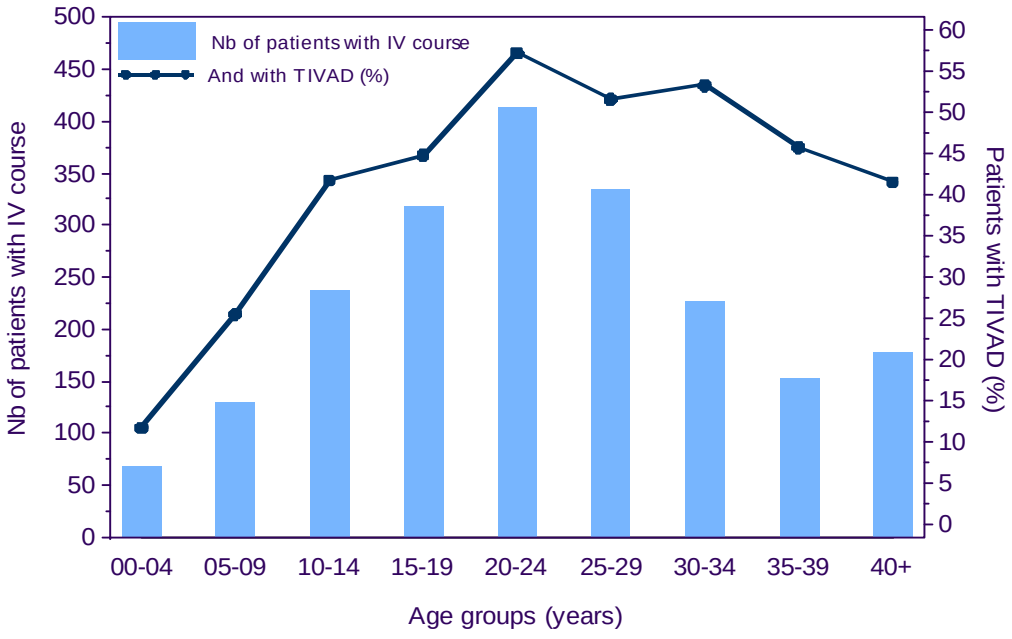
IV antibiotic courses – TIVAD

Table 17. Patients with IV antibiotic courses

	Age groups (years)									
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total
All patients	794	949	868	795	816	655	479	320	469	6145
Nb of patients with at least 1 course	68	129	237	319	414	335	227	153	178	2060
- and with TIVAD*	8	33	99	143	237	173	121	70	74	958
Nb of courses	83	204	484	734	1008	761	524	409	435	4642
Nb of days of courses incl:	1511	3528	6980	10882	15434	11535	8119	5382	5355	68726
- at hospital	898	1283	2011	2893	2746	2793	1324	1562	1563	17073
- at home	299	2284	4923	7752	11856	8749	6521	3368	3458	49210
TIVAD* (with and without course)	11	40	119	156	290	219	162	90	110	1197

French CF Registry - Annual Data Report 2012

Figure 23. Patients with at least one IV antibiotic course and a TIVAD*, by age



* TIVAD: Totally Implantable Vascular Access Device

French CF Registry - Annual Data Report 2012

Therapeutic management

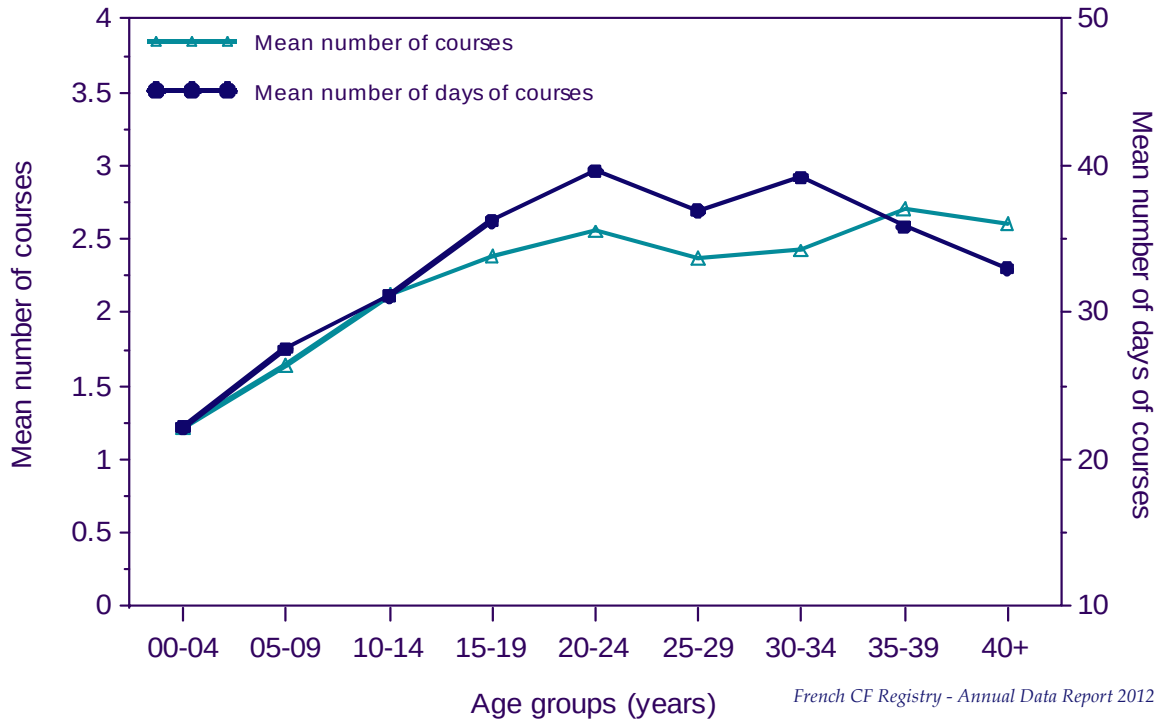
Antibiotic courses

Table 18. Repartition of courses

	Age groups (years)									Total
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	
Courses										
Mean	1.2	1.6	2.1	2.4	2.6	2.4	2.4	2.7	2.6	2.4
Standard deviation	0.5	1.2	1.5	1.5	1.9	1.6	1.6	3.7	4.0	2.2
Median	1	1	2	2	2	2	2	2	2	2
Days of courses										
Mean	22.2	27.6	31.2	36.3	39.7	37.0	39.2	35.9	33.1	35.4
Standard deviation	35.2	37.8	29.9	25.8	37.7	32.5	33.9	29.9	30.1	32.9
Median	14.5	15	28	30	30	28	30	30	27.5	28

French CF Registry - Annual Data Report 2012

Figure 24. Mean number of courses and of days of courses



French CF Registry - Annual Data Report 2012

Therapeutic management

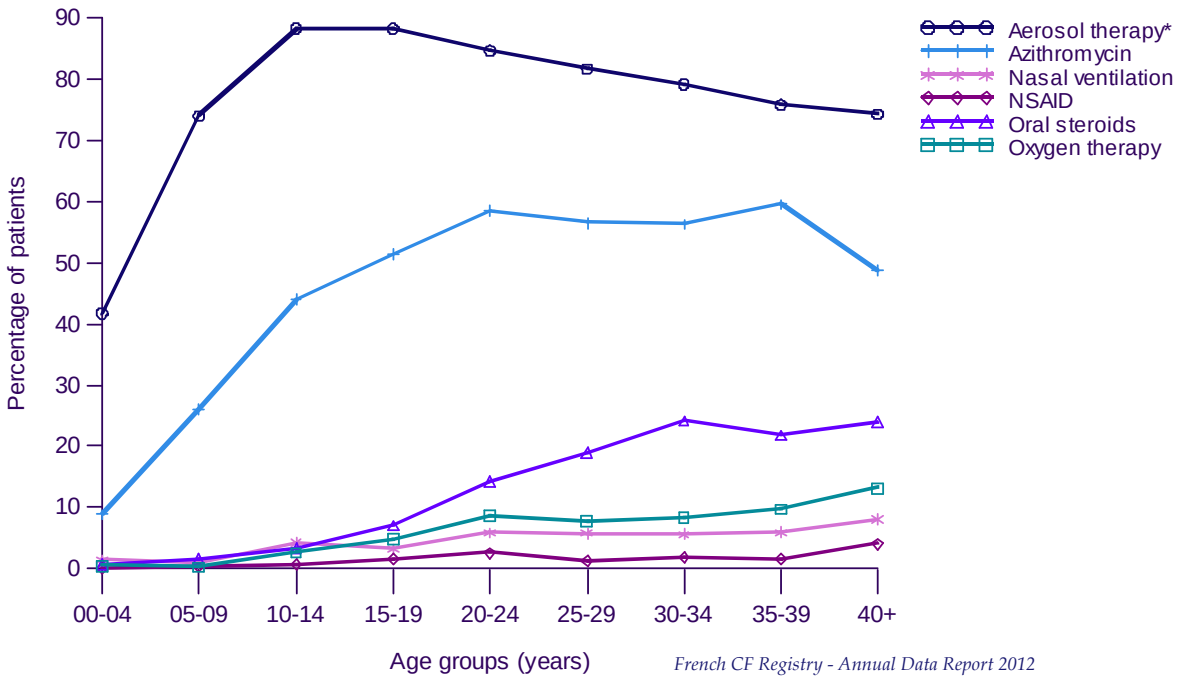
Respiratory

Table 19. Respiratory therapeutics

	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%
All patients	*794*	*949*	*868*	*795*	*816*	*655*	*479*	*320*	*469*	*6145*	.
Aerosol therapy*	332	703	768	703	692	536	380	243	349	4706	76.6 %
NSAID	1	3	6	13	21	8	9	5	19	85	1.4 %
Azithromycin	70	247	383	409	478	371	270	191	229	2648	43.1 %
Oxygen therapy	4	3	23	38	71	51	40	31	62	323	5.3 %
Oral steroids	6	15	29	56	117	125	116	70	113	647	10.5 %
Nasal ventilation	11	10	37	26	48	38	27	19	38	254	4.1 %

French CF Registry - Annual Data Report 2012

Figure 25. Respiratory therapeutics



French CF Registry - Annual Data Report 2012

**by nebulization, spray or powder*

Therapeutic management

Aerosoltherapy

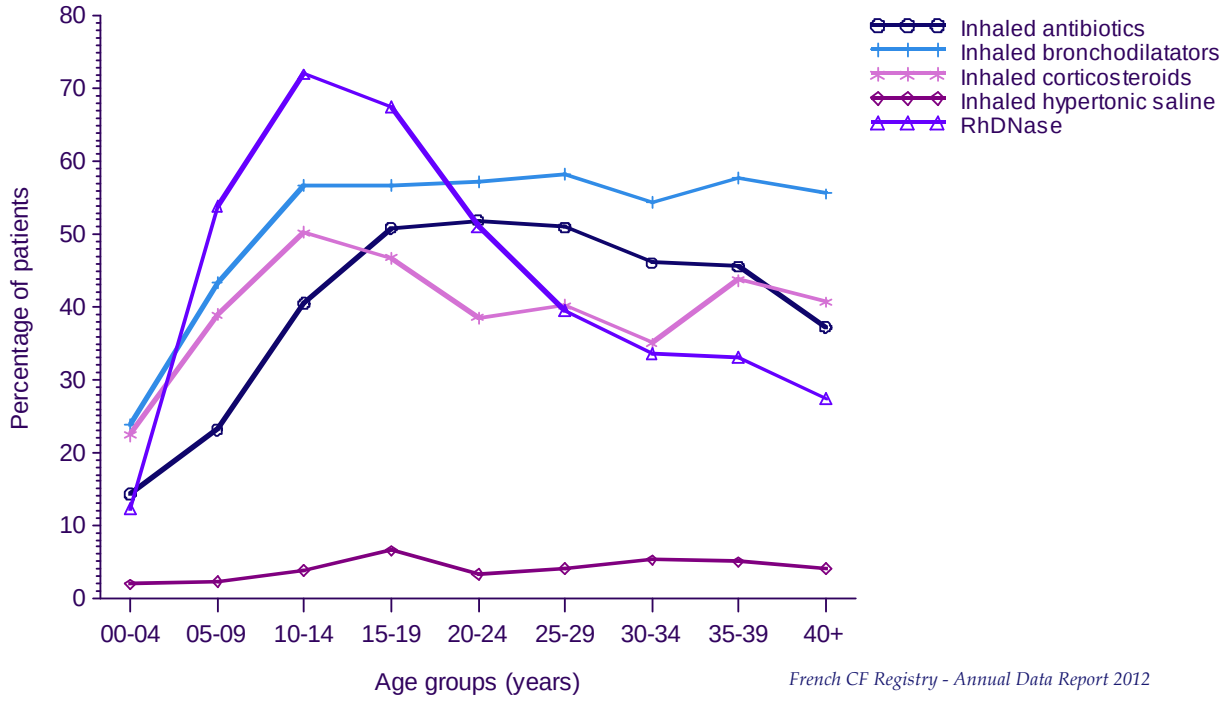
Table 20. Aerosoltherapy treatments

	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%
All patients	794	949	868	795	816	655	479	320	469	6145	.
Patients under aerosol therapy*	332	703	768	703	692	536	380	243	349	4706	76.6 %
Inhaled antibiotics	114	220	352	405	424	334	221	146	175	2391	38.9 %
Inhaled bronchodilatators	190	412	493	452	468	382	261	185	261	3104	50.5 %
Inhaled corticosteroids	178	370	437	372	315	264	168	140	191	2435	39.6 %
Inhaled hypertonic saline	15	22	33	52	27	27	25	16	19	236	3.8 %
RhDNase	98	512	626	537	416	259	161	106	129	2844	46.3 %

French CF Registry - Annual Data Report 2012

**by nebulization, spray or powder*

Figure 26. Aerosoltherapy treatments



French CF Registry - Annual Data Report 2012

Therapeutic management

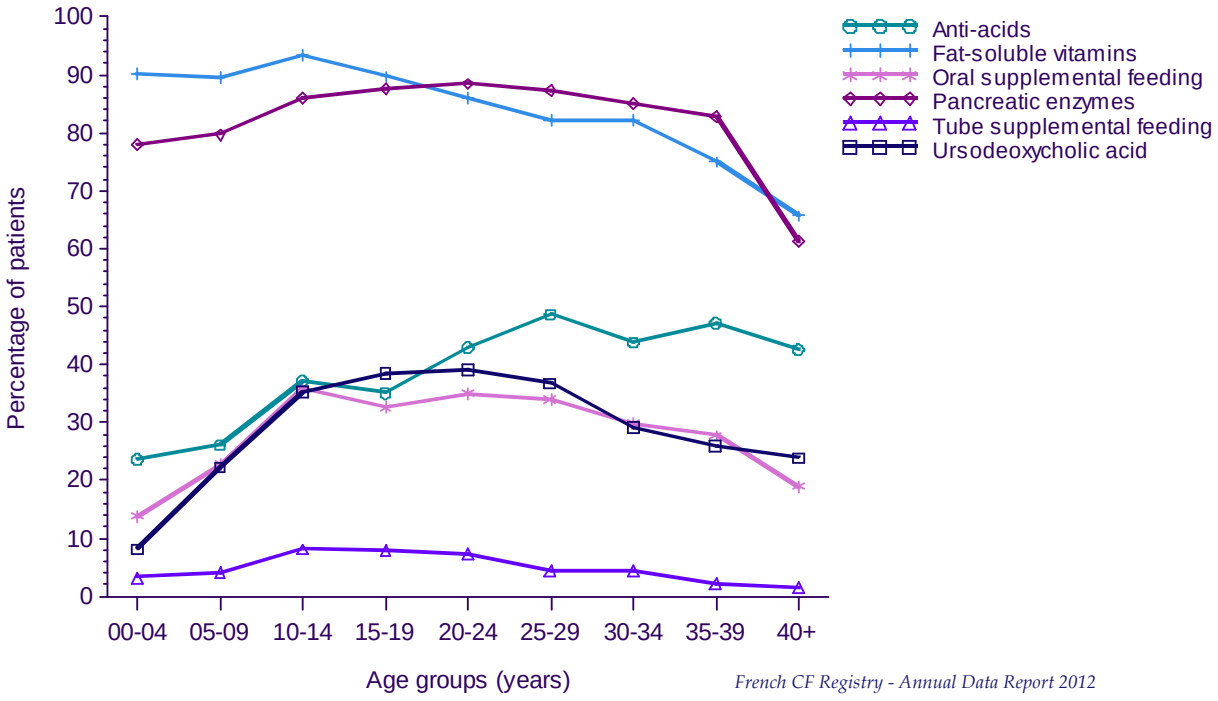
Digestive and nutritional

Table 21. Hepatic, digestive and nutritional treatments

	Age groups (years)										
	00-04	05-09	10-14	15-19	20-24	25-29	30-34	35-39	40+	Total	%
All patients	794	949	868	795	816	655	479	320	469	6145	.
Ursodeoxycholic acid	65	212	307	306	319	241	140	83	112	1785	29.0 %
Anti-acids	188	248	323	279	351	319	210	151	200	2269	36.9 %
Pancreatic enzymes	620	757	747	697	722	572	408	265	288	5076	82.6 %
Tube supplemental feeding	26	39	71	63	60	29	21	7	7	323	5.3 %
Oral supplemental feeding	110	217	311	260	286	223	143	89	89	1728	28.1 %
Fat-soluble vitamins	717	850	811	714	703	538	394	240	308	5275	85.8 %

French CF Registry - Annual Data Report 2012

Figure 27. Hepatic, digestive and nutritional treatments

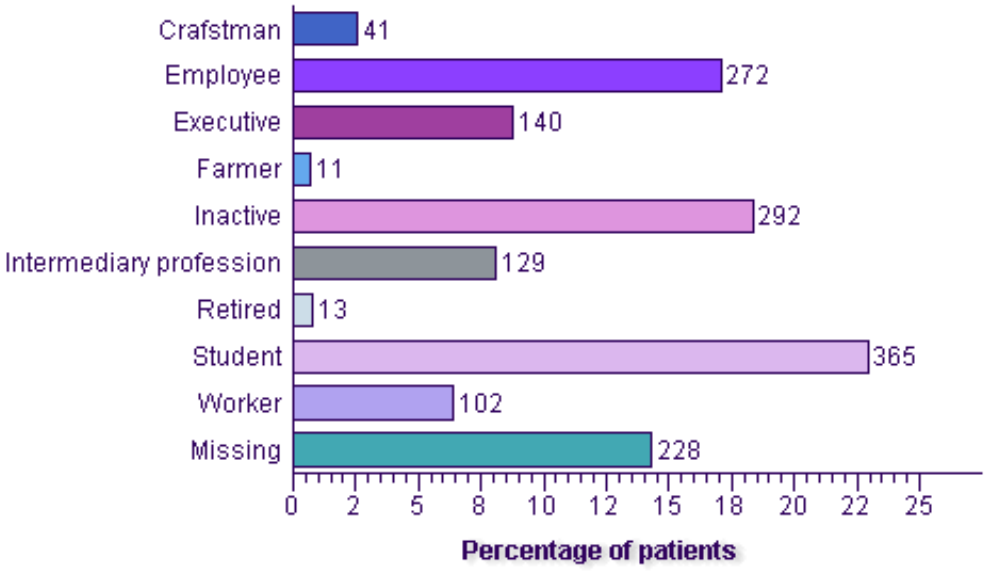


French CF Registry - Annual Data Report 2012

Social data

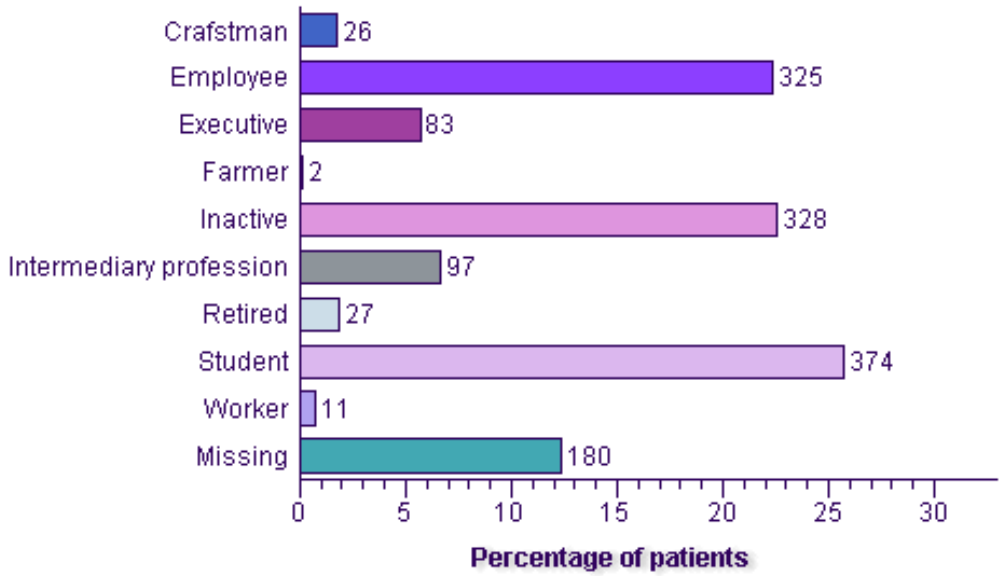
Employment

Figure 28. Employment of men ≥ 18 years



French CF Registry - Annual Data Report 2012

Figure 29. Employment of women ≥ 18 years

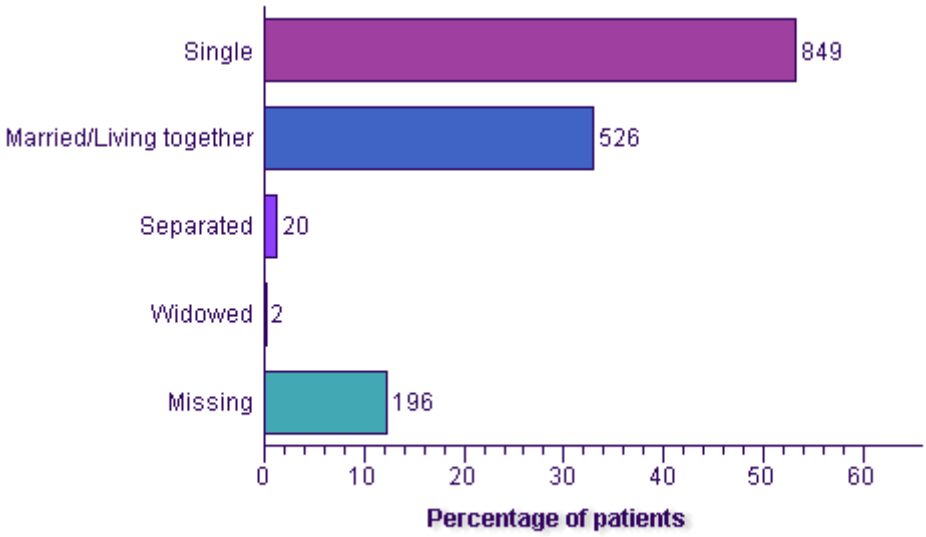


French CF Registry - Annual Data Report 2012

Social data

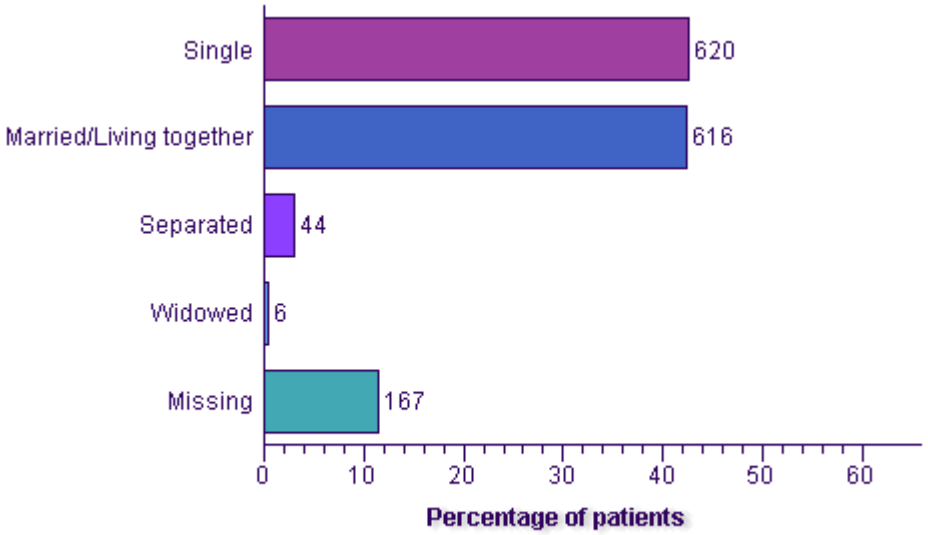
Marital status

Figure 30. Marital status of men ≥ 18 years



French CF Registry - Annual Data Report 2012

Figure 31. Marital status of women ≥ 18 years



French CF Registry - Annual Data Report 2012

Annex 1 (1/2)

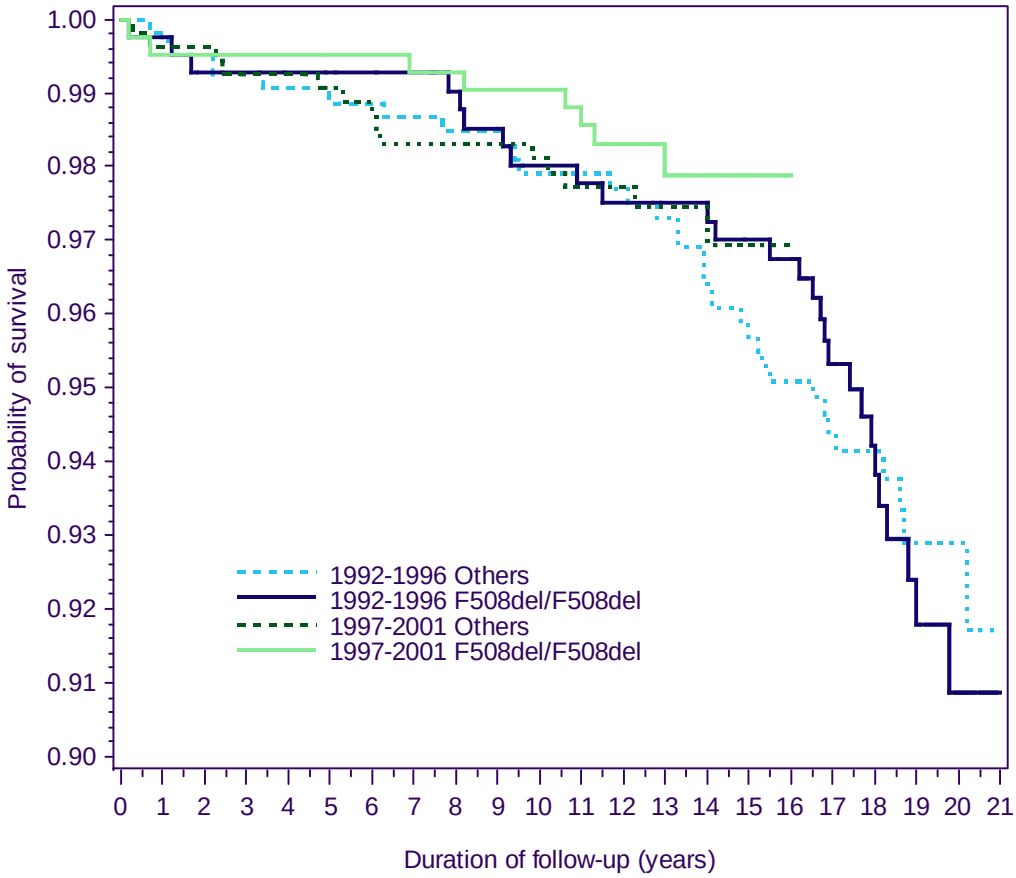
Complement on survival analysis – stratification by genotype

Figure 32. Survival curves by birth cohort and genotype (Kaplan-Meier method)

The survival analysis (fig. 4) was completed for the oldest birth cohorts, stratified according to the **genotype:**

- Births from 1992 to 1996:
  - *F508del/F508del* : 411 patients, 27 deaths;
  - *other genotypes*: 531 patients, 33 deaths

- Births from 1997 to 2001:
  - *F508del/F508del* : 419 patients, 8 deaths;
  - *other genotypes*: 546 patients, 14 deaths



French CF Registry - Annual Data Report 2012

There is no significative survival difference between those cohorts (Log-Rank test = 1.47, p = 0,689).

Annex 1 (2/2)

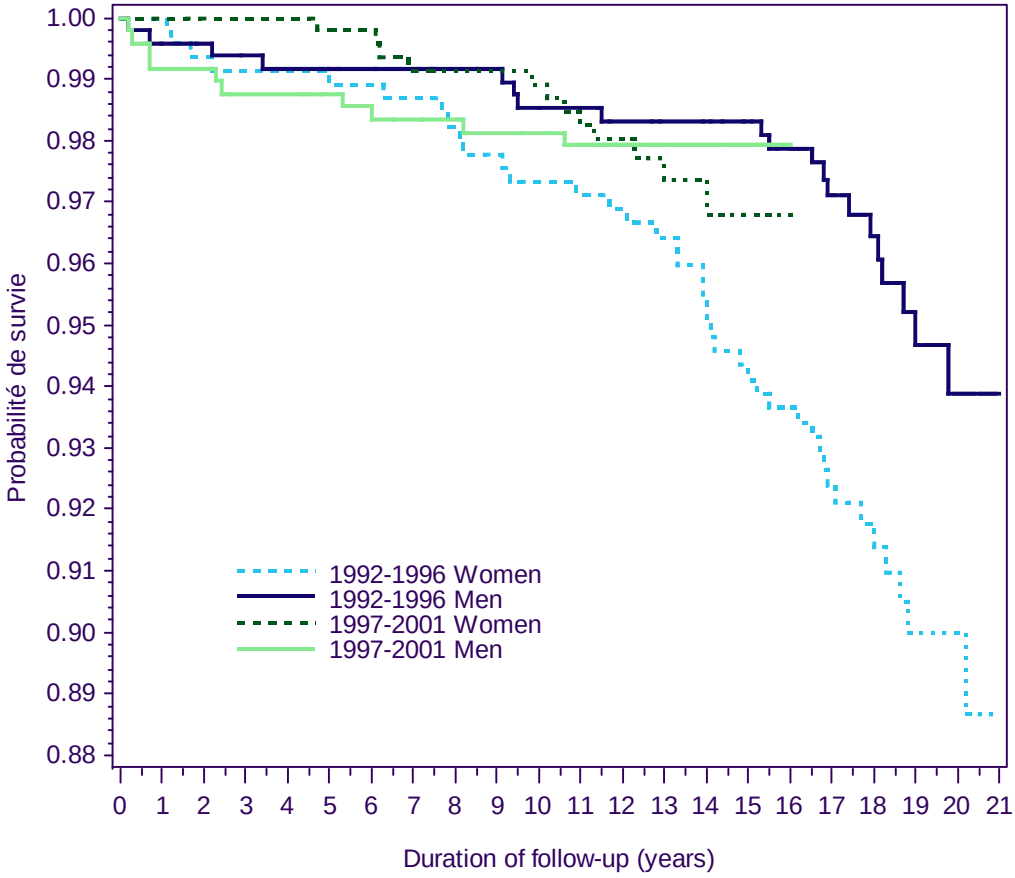
Complement on survival analysis – stratification by sex

Figure 33. Survival curves by birth cohort and sex (Kaplan-Meier method)

The survival analysis (fig. 4) was completed for the oldest birth cohorts, stratified according to the **sex:**

- Births from 1992 to 1996:
  - *men*: 484 patients, 20 deaths;
  - *women*: 458 patients, 40 deaths

- Births from 1997 to 2001:
  - *men*: 487 patients, 10 deaths;
  - *women*: 477 patients, 12 deaths



French CF Registry - Annual Data Report 2012

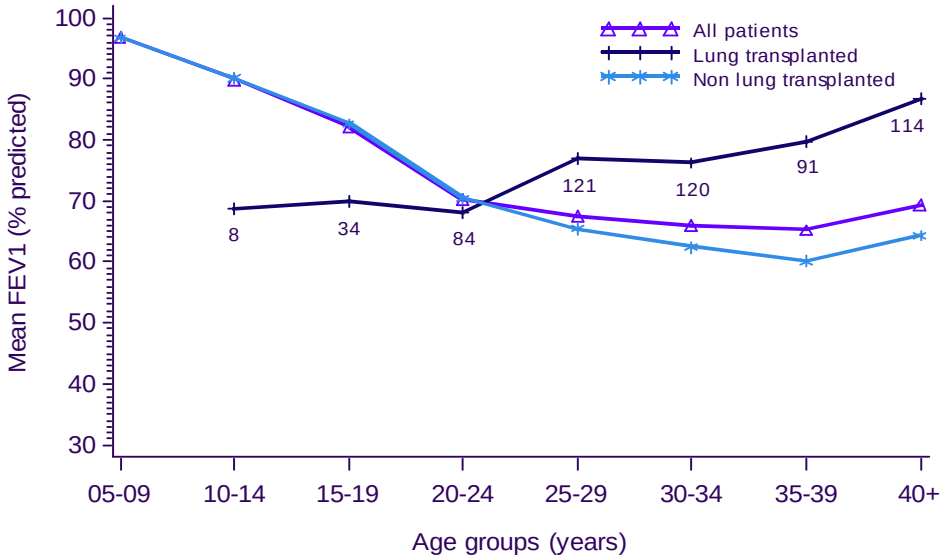
There is a significant survival difference in the 1992-1996 cohorts, men vs women (Log-Rank test = 8.32, p = 0,039) but no difference in the 1997-2001 cohorts.

Annex 2

Spirometry and transplantation

Figure 34. Mean FEV_1 (% predicted) and transplantation

To provide a more comprehensive picture, further comparisons were made using the curves of FEV_1 by age: FEV_1 (% predicted) of all patients was compared to that of patients who had or had not received a heart-lung or bilateral lung transplant.



French CF Registry - Annual Data Report 2012

The curves of the whole population and of non-transplanted patients are identical up to age 20-24. Above 25 years, FEV_1 (% predicted) of non-transplanted patients drops more sharply than that of the total population, with a difference of almost 5 percentage points at ages 35-39. Among older patients (aged 40 or above) an upward trend is observed for both patient categories, suggesting a selection effect of patients with the mildest forms of CF at these ages.

Lung transplanted patients' FEV1 (% predicted) values are close to 69% at ages 10-24, and increase to 77% for older patients.

Note:
- Values in figure 34 are the number of lung transplanted patients in each age group.
- one patient present in the 05-09 age group was not included in the graph.

Annex 3

Participating centres

Table 22. List of the participating CRCMs

CRCMs	Number of patients*
Paediatric CRCMs	
AMIENS Hôpital Nord	92
BORDEAUX Groupe Pellegrin Hôpital d'Enfants	162
GRENOBLE Hôpital de la Tronche Pédiatrie	110
LE HAVRE Hôpital Flaubert	27
LILLE Hôpital Jeanne de Flandres Pédiatrie	192
LISIEUX Centre Hospitalier Robert Bisson	20
LYON Hôpital Mère-Enfant / Groupt Hosp. Est	298
MARSEILLE Hôpital La Timone Pédiatrie	119
NANCY Hôpital d'enfants	132
NANTES Hôpital Mère-Enfant	105
NICE CHU Lenval - Hôpital Pasteur	81
PARIS Hôpital Armand Trousseau	85
PARIS Hôpital Necker	147
PARIS Hôpital Robert Debré	165
RENNES - ST BRIEUC Pédiatrie	146
ST DENIS DE LA REUNION Hôpital d'Enfants	61
TOULOUSE Hôpital des Enfants	140
TOURS Hôpital de Clocheville Pédiatrie	123
VERSAILLES Hôpital Mignot Pédiatrie	67
Adults CRCMs	
BORDEAUX-PESSAC Groupe Sud Hospitalier	103
GRENOBLE Hôpital de la Tronche Pneumologie	103
LILLE Hôpital Calmette Pneumologie	178
LYON SUD Centre Hospitalier	265
MARSEILLE CHU Nord	182
NANCY Hôpital de Brabois Pneumologie	108
NANTES Hôpital Laënnec	184
PARIS Hôpital Cochin	374
RENNES Hôpital Pontchaillou Pneumologie	93
SURESNES Hôpital Foch	333
TOULOUSE Hôpital Larrey Pneumologie	152
TOURS Hôpital Bretonneau Pneumologie	55
Paediatric and Adults CRCMs	
ANGERS - LE MANS	125
BESANCON	117
CAEN	98
CLERMONT FERRAND CHU d'Estaing	88
CRETEIL Centre Hospitalier Intercommunal	113
DIJON Hôpital d'Enfants du Bocage	117
DUNKERQUE Centre Hospitalier	60
GIENS Hôpital Renée Sabran	224
LENS Centre Hospitalier	46
LIMOGES Hôpital Mère/Enfant	56
MONTPELLIER Hôpital Arnaud de Villeneuve	197
POITIERS Hôpital La Milétrie	40
REIMS American Memorial Hospital	119
ROSCOFF Centre de Perharidy	142
ROUEN	167
ST PIERRE DE LA REUNION Groupe Hosp. Sud	79
STRASBOURG	228
VANNES-LORIENT	80

Annex 3

Participating centres

Table 23. List of the participating centres (CRCMs excepted)

Centres	Number of patients*
Paediatric local centres	
BREST Hôpital Augustin Morvan	6
COLMAR CHG Louis Pasteur Pédiatrie	8
DAX Centre Hospitalier	12
MONTLUCON Centre Hospitalier	9
MULHOUSE Centre Hospitalier Pédiatrie	14
ST ETIENNE Hôpital Nord	2
ST TROJAN LES BAINS Centre Hélio Marin	5
Adult local centres	
MULHOUSE Centre Hospitalier Pneumologie	4
Paediatric and Adult local centres	
BRIVE Centre Hospitalier	14
ST NAZAIRE Centre Hospitalier	24
Other centres	
DIEULEFIT Centre Médical/Climatique Bellevue	1
MONTARGIS Centre Hospitalier	1
PARIS Hôp. Européen G.Pompidou Greffes	8
POINTE A PITRE CHU	8
ST QUENTIN Centre Hospitalier Général	8

French CF Registry - Annual Data Report 2012

* Number of patients who visited the centre during the year. Patients followed by a centre and who did not visit it in 2012 were excluded from those statistics.

Annex 4

Table 24. Summary of data (1/2)

	2010	2011	2012
Patients seen during the year and centres participating to the registry			
- Patients registered* (N):	5792	6046	6196
- Patients seen during the year in a centre** (N):	5758	5993	6145
- Centres (N) :	64	65	63
Paediatric CRCMs***:	19	19	19
Adult CRCMs***:	12	12	12
Paediatric and Adult CRCMs***:	18	18	18
Other centres:	15	16	14
Demographics			
- Male patients (%) :	51,4	51,7	51,6
- Age of patients, in years (mean):	18,1	18,7	19,2
- Age of patients, in years (median):	16	17	17
- Age of patients. in years (min-max):	0 – 80	0 – 87	0– 86
- Patients aged 18 years and over (%):	47,2	48,7	49,6
- Early pregnancies during the year (N):	34	61	54
- Pregnancy rates in women aged 15 to 49 ans (for 1 000):	24,5	41,7	35,1
- Age of patients at onset of pregnancy, in years (mean):	28,3	28,3	27,2
- Deaths (N):	60	67	54
Including death of patients not seen during the year:	10	9	13
- Crude death rate (for 1 000):	10,6	11,4	8,9
- Age at death, in years (mean):	29,2	26,4	32
- Age at death, in years (median):	28	25	28
Diagnosis and genetics			
- Age at diagnosis, in months (median) :	3	3	3
- New patients diagnosed during the year (N):	181	215	188
Including by neonatal screening:	112	131	124
- Age at diagnosis of the new patients, in years (median):	2	1	1
- Age at diagnosis of the new patients, in years (min-max):	0 – 69	0 – 69	0– 72
- Full genotypes identified (%):	93,9	94,9	94,6
F508del / F508del:	43,6	43,3	42,3
F508del / Other:	37,5	38	38,9
Other / Other:	12,8	13,1	13,4
F508del / Missing:	1,8	1,9	1,7
Other / Missing:	1,1	1,2	0,9
Missing / Missing:	3,2	2,5	2,8
Anthropometry and spirometry			
- Height z-score, patients aged 17 years and less (mean):	- 0,13	-0,08	-0,10
- Height z-score, patients aged 18 years and over (mean):	- 0,53	-0,52	-0,51
- Weight z-score, patients aged 17 years and less (mean):	- 0,37	-0,33	-0,30
- Weight z-score, patients aged 18 years and over (mean):	- 0,46	-0,41	-0,34

French CF Registry - Annual Data Report 2012

* Patients whose vital status is known, whether they visited or not a centre during the year.

** Reference patients for the statistics of this report, with the exclusion of survival data.

*** CRCM: Specialised CF Centre (Centre de Ressources et de Compétences de la Mucoviscidose)

Annex 4

Table 24. Summary of data (2/2)

	2010	2011	2012
Spirometry			
- FEV_1 (% predicted) - Knudson, patients aged 17 years and less (mean):	85,4**	89,7*	90,6*
- FEV_1 (% predicted) - Knudson, patients aged 18 years and over (mean):	62,9**	67,3*	68,6*
Microbiology			
- Patients with at least one sputum during the year (%):	93	91,7	91,2
H. influenzae:	23,4	22,1	22,9
MSSA:	53,5	54,7	54,4
MRSA:	8	7,7	7,4
P. aeruginosa:	42,9	42,6	41,8
S. maltophilia:	8,5	9,7	9,9
B. cepacia:	2	1,9	1,7
Aspergillus:	21	22,6	23,8
Complications and transplantations			
- Haemoptysis (%):	5,8	5,0	4,6
- Cirrhosis / portal hypertension (%):	3,9	4,1	4,3
- Insulin-dependent and non insulin-dependant diabetes (%):	14,8	15,2	15,8
- Transplanted patients (N):	440	529	573
Including patients transplanted during the year:	78	97	101
- Patients on waiting list (N):	136	177	153
Including patients listed during the year:	75	98	96
Deaths on waiting list:	1	0	3
Therapeutic management			
- IV courses (%):	34,4	34,6	33,5
- Oxygenotherapy (%):	6,5	5,9	5,3
- Nasal ventilation (%):	3,9	4,8	4,1
- Azithromycin (%):	42,4	42,1	43,1
- Inhaled antibiotics (%):	38,8	37,3	38,9
- rhDNase (%):	45,1	46,9	46,3
- Inhaled bronchodilatators (%):	48,8	49	50,5
- Inhaled corticosteroids (%):	40,8	39,2	39,6
- Pancreatic enzymes (%):	82,8	83,2	82,6

French CF Registry - Annual Data Report 2012

* Best FEV_1 of the year.

** Last FEV_1 of the year.

Conception and realization : Medical Department of Vaincre la Mucoviscidose and Ined / March 2014

Vaincre la Mucoviscidose

181, rue de Tolbiac – Paris 13e

Telephone : 00 33 1 40 78 91 95

Email: registre@vaincrelamuco.org

www.vaincrelamuco.org

Institut national d'études démographiques

133, boulevard Davout – Paris 20e

Telephone : 00 33 1 56 06 20 00

www.ined.fr