Cystic Fibrosis our focus

UK Cystic Fibrosis Registry

Annual Data Report 2012: Summary
September 2013

Contents

The full 'Annual Registry Data Report 2012', which also includes performance data from CF centres across the UK, can be downloaded from cysticfibrosis.org.uk/registry

Introduction

We are delighted to present this summary of the UK (CF) Registry's 2012 scientific report, which the Trust produces for the scientific and clinical community.

This annual scientific report contains clinical information drawn from the more than 10,000 people with cystic fibrosis treated in clinics across the UK, who have consented to their information being included in the UK CF Registry.

It's a vital source of information for clinicians to improve standards of patient care, and to those undertaking important research into cystic fibrosis.

This is, however, the first year we are providing this information in a more accessible, user-friendly form to ensure that all those with cystic fibrosis, and their families, as well as others, have full access to this information too; our purpose is to inform and explain.

This report, therefore, sets out the latest statistics on the number of people with cystic fibrosis in the UK, how they are divided by age, gender and genetic type, and how they are faring against a series of clinical measures. It also includes figures on what therapies are being used across the UK and how these have changed over recent years.

The report includes the latest figures for the average survival rate for people with cystic fibrosis too. This should be looked at with some caution, but the good news is that it shows that, overall, people with the condition in the UK continue to live longer.

The Registry is made possible by generous donations to the Cystic Fibrosis Trust and from income derived from the NHS in England and Scotland.

The Trust is deeply grateful to all the people with cystic fibrosis and their families who kindly agree to share their data, and to the staff in the specialist cystic fibrosis centres who collect and enter the data each year.

We hope this report helps improve wider understanding of the condition, and enables all those with cystic fibrosis and their carers to take a more active role, alongside their clinical teams, in the shaping of their treatment.

Ed Owen
Chief Executive,
Cystic Fibrosis Trust

Dr Diana Bilton
Chair,
Registry Steering Committee

The Cystic Fibrosis Trust is making a daily difference to the lives of people with cystic fibrosis and those who care for them. It:

- Invests in cutting-edge research
- Drives up standards of care
- Offers support and advice to people with cystic fibrosis and their families
- Campaigns hard on behalf of the 10,000 people with cystic fibrosis in the UK
- Shouts loud to raise awareness, increase understanding of cystic fibrosis and raise vital funds.

Get involved at cysticfibrosis.org.uk

Summary of findings

These are some of the key findings in the analysis of the registry data for 2012:

- The median survival for the CF population is currently 43.5 years, compared to 38.8 for the population in 2008.
- In children aged 6 to 15 years, 66% had lung function in the normal range.
- There has been an increase in the use of nebulised drug treatment in 2012 compared to 2008 in all age groups, which should result in better long-term outcomes.
- There has been a reduction in chronic pseudomonas infection in 2012 compared to 2008. In the 16-19 and 20-23 age groups this reduction was statistically significant.
- 46 people with cystic fibrosis received a double lung or heart and lung transplant in 2012, compared to 47 and 27 in the previous two years.
- There were 106 deaths reported in 2012, at a median age of 28 years.

About the Registry

The Registry tracks the health and treatment of people with cystic fibrosis; information is collected every year from over 10000 people who receive care at accredited CF centres and agree to participate in the Registry.

The data collected includes area of residence, gender, age, height, weight, CF mutations, pulmonary function test results, medication use and problems [complications] related to cystic fibrosis.

This completely anonymised dataset, held on a secure and confidential computer database, is available to clinicians, researchers, NHS commissioners and to industry, under the scrutiny of the CF Registry Steering Committee.

This data is used to:

- facilitate research and development aimed at improving outcomes for people with cystic fibrosis;
- support the planning and commissioning of cystic fibrosis NHS services at designated centres;
- enable centre-specific analysis in order to identify best care which can then be instituted across all centres; and
- support research protocol development by providing information about people with cystic fibrosis who may meet the criteria for a particular research project.

Information about the governance of the Registry and the Registry Steering Committee is in Annex 1 and 2.

What is 'complete data'?
'Complete data' means the number of complete sets of data from the annual reviews of people with cystic fibrosis – these are used for the production of the Registry annual reports

Key facts about the UK cystic fibrosis population in 2012

- 10078 people were registered, and of these, 87% had complete data (see box, left) entered into the Registry.
- There were 274 new diagnoses in 2012 – five a week.
- Of the 63 children with complete data born in 2012, 50 (79%) were identified by newborn screening.
- More than 57% of those registered were over 16 years old.
- 70% of those aged 16 or over, were working or studying.
- Over 800 people with cystic fibrosis were aged 40 years or older. The oldest man on the Registry was 83 years old and the oldest woman was 81.

Genotypes

Genotype refers to the CF mutations that cause an individual to have cystic fibrosis. To have cystic fibrosis you need to have two CF mutations, one inherited from each parent.

People with cystic fibrosis should know which mutations they have. If you do not know whether you or your child has had your CF mutations identified, talk to your CF centre. This is important because having this information on the Registry will help in identifying people to be included in new trials of mutation-specific drugs, leading to more personalised treatment.

8462 people, that is over 96% of everyone on the Registry in 2012, have been genotyped with a recorded value. Around half of people with cystic fibrosis have two copies of DF508, the most common mutation.

The table below lists the most common CF mutations.

Single mutations and their classes 2012

Mutation	Class	Number	Per cent
DF508	II	7671	90.7
G551D	III	471	5.6
R117H	IV	361	4.3
G542X	I	307	3.6
621+1G->T	I	180	2.1
N1303K	II	111	1.3
1717-1G->A	I	106	1.3
1898+1G->A	I	95	1.1
DI507	II	85	1.0
R560T	III	83	1.0
3659delC	II	82	1.0

New research aimed at learning from care in different centres

We would like to improve how care outcomes across the different CF centres are considered, so that they can learn from each other and ensure optimum care for the community.

It is clear that outcomes in terms of lung function, for example, can be affected by many factors, including those related to care received, such as use of CF medication, and those related to the person with cystic fibrosis – such as genotype, age and socioeconomic status. So 'better lung function' does not necessarily equate to 'better centre'.

Recently we were awarded a research grant from the National Institute of Health Research, for a project aimed at developing better ways of looking at outcomes across centres, with the emphasis on learning from the best processes of care. This work is being led by Dr Stephanie MacNeill, the statistician who works on the Registry, and Professor Paul Cullinan, from the National Heart and Lung Institute at Imperial College, London.

Clinical care of people with cystic fibrosis

The Cystic Fibrosis Trust works closely with the specialist cystic fibrosis centres and clinics across the UK, with professional bodies such as the British Thoracic Society and the British Paediatric Respiratory Society, and with NHS commissioners of care, to drive up standards of care.

This is achieved through the setting of national standards and a process of audit or peer review of those standards in the CF centres and clinics.

Recognised specialist CF centres provide access to a multidisciplinary team of specialist doctors, nurses and allied health professionals, as set out in the nationally recognised standards of care, which can be downloaded from the Trust's website at cysticfibrosis.org.uk/publications.

Research has been instrumental in driving up standards of care, most notably through demonstrating the benefits of screening newborns for cystic fibrosis.

The process of peer review of CF centres, against the recognised standards, includes the provision of a range of information from the Registry on treatment and outcomes. This information is produced as appropriate depending on whether the centre is looking after children or adults.

The purpose is to:

Check the completeness of a centre's records on the Registry

Check whether a particular centre is producing outcomes that are in the range of the national average, and

To look at how all centres are working towards certain goals such as:

- increasing the recording of genetic mutations;
- increasing the number of people with cystic fibrosis being prescribed mucolytics such as DNase to breakdown mucus in the lungs;
- increasing the number of people with cystic fibrosis who are receiving nebulised antibiotics to prevent lung function decline;
- reducing the number of children aged less than 11 years, who have chronic pseudomonas to 0%; and
- reducing the rate of chronic pseudomonas in adults under 30 years, to less than 60%.

What is median?

In the following sections we summarise some of the key findings from the 2012 report and the term 'median' is used in several of the graphs.

Median is a type of average, denoting the value or quantity lying at the midpoint of the distribution of observed values or quantities, so...: "the median duration of hospital stay was 10 days" means that half of the people were in hospital for fewer than 10 days and half for more.

Monitoring outcomes of care

The core purpose of the Registry is to collect routine clinical data that is used to improve clinical outcomes. Each CF centre can analyse their own data at any time, for an individual, or for a group of people with cystic fibrosis.

Each year, data in the Registry is analysed, a report is produced and the findings discussed by the cystic fibrosis community as part of our mission to improve care.

Clinic appointments

At each clinic appointment a set of measures is undertaken to monitor the condition of people with cystic fibrosis. These routine measures are Body Mass Index, lung function and sputum sampling. The purpose is to assess whether there is a decline in function and whether a change is needed in the care being provided. We are also interested in monitoring the effects of any new drug treatment.

Body mass index

Body mass index [BMI] is an important measure because research has shown that people with cystic fibrosis with a good BMI do better in terms of lung function and survival.

BMI is a number calculated from weight and height. This number does not measure body fat directly, but it can be considered as an alternative for direct measures of body fat.

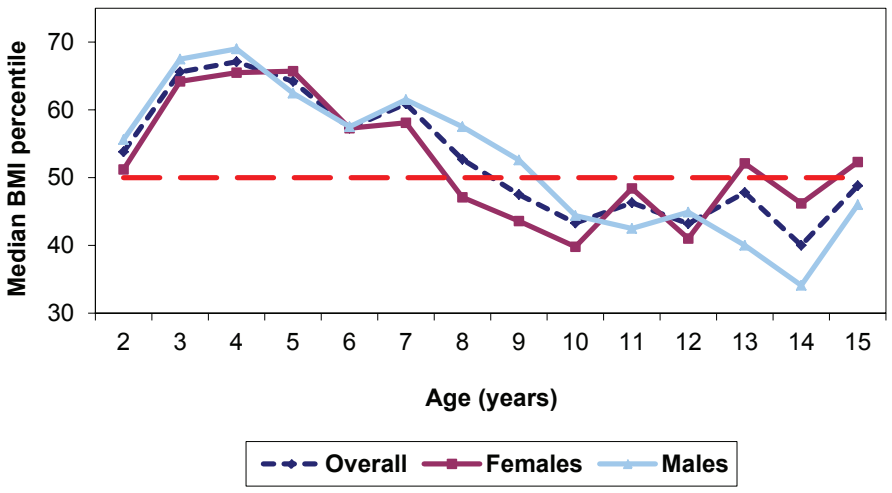
Although the BMI number is calculated the same way for children and adults, the criteria used to interpret the meaning of the BMI number for children and teens are different from those used for adults. For children and teens, BMI age- and sex-specific percentiles are used for two reasons:

- The amount of body fat changes with age.
- The amount of body fat differs between girls and boys.

So for children and teens, BMI is age- and sex-specific. For this reason percentiles are the most commonly used indicator to assess the size and growth patterns of individual children because the percentile indicates the relative position of the child's BMI number among children of the same sex and age.

Median BMI percentiles among children in 2012 (n=3164): The red dotted line indicates the 50th percentile – the aim is to monitor and maintain weight for height as close to this target as possible.

Figure 1.12

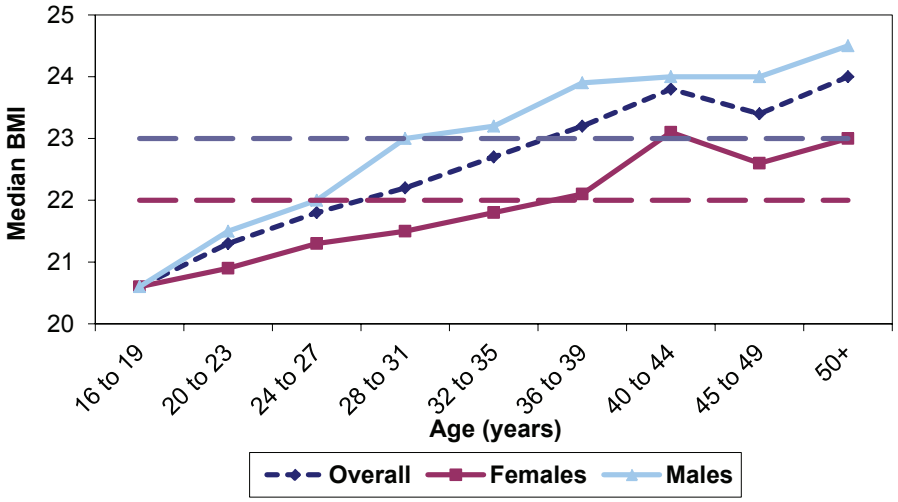


"I've learnt over the last year or two just how powerful the Registry data is in helping all the CF specialist centres and clinics analyse outcomes of care and treatments provided. That this Registry data is being actively used to the advantage of all of us dealing with cystic fibrosis: the CF Trust, CF teams and researchers, is hugely reassuring for now and the future."

Dominic Kavanagh, Cystic Fibrosis Trust Clinical Care Patient Adviser, who has cystic fibrosis

Median BMI values among adults in 2012 (n=4907): The purple dotted line indicates a BMI of 22 which is a marker used to target BMI in adult women; the blue dotted line indicates a BMI of 23 which is a marker used for adult men

Figure 1.13



Lung function

At routine CF clinic appointments, a lung function test is performed and the key measure is called the forced expiratory volume in 1 second (FEV_1), see box, below. This measures how much air comes out of the lungs in the first second when someone blows out hard and fast. An FEV_1 of 100% means that lung function is exactly the same as that of an average person of the same age and height who does not have cystic fibrosis – in other words, 100% equals totally normal. However, as we are all slightly different, values between 86% and 115% are considered to be within a normal range.

The aim of good cystic fibrosis care is to preserve normal lung function for as long as possible. This is important for adults, as lung function with a FEV_1 of 50% and above, will enable people to live normal lives, including attendance at work and college.

What is FEV_1?

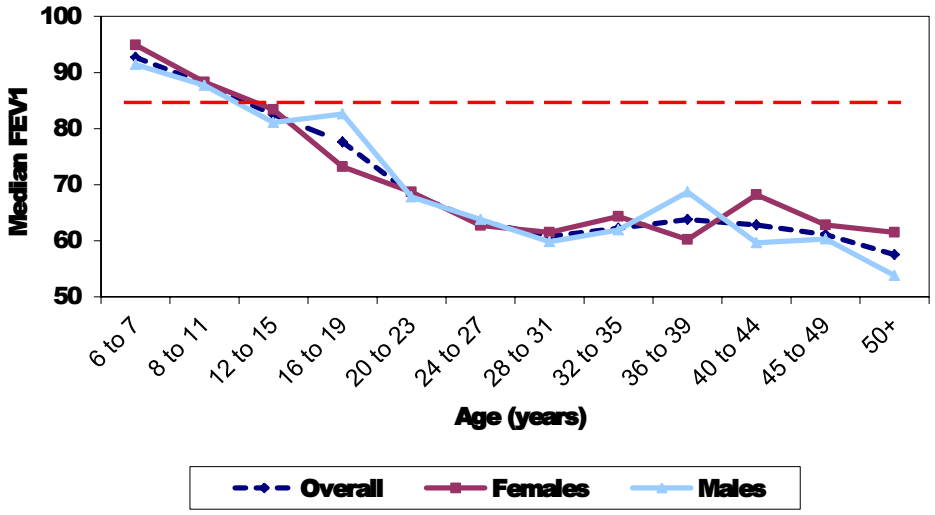
For an ADULT, the following is a guide to FEV_1 percentage predicted measurements and what they may mean:

- FEV_1 greater than or equal to 85% predicted is considered as normal or near normal for the general population.
- 70–84% predicted shows mild lung disease.
- 40–69% predicted shows moderate lung disease.
- Less than 40% predicted can be a sign of severe lung disease.

On the Registry we look at FEV_1 % predicted across the different age groups and you will see from the graph below that we are moving towards the goal of keeping lung function in children as near normal as possible.

Lung function by age group – Median FEV_1 (% predicted) among people with CF aged six years and older (n=6935)

Figure 1.14



The red dotted line in this figure illustrates a target FEV_1 % predicted of 85%. Anything above this indicates normal or near-normal lung function values.

In 2012, 66% of children aged 6 to 15 years had a lung function in the normal range.

For an individual we are interested in change in function – a lower measure does not necessarily mean there is a problem but it may signal a need for further investigation.

Over the years we want to see the average lung function of all the people with cystic fibrosis at different ages to increase as a result of excellent care and introduction of new therapies.

Keeping lungs healthy – airway clearance

Medication alone cannot keep cystic fibrosis lungs healthy. Getting the thick mucus out of the lungs is key. Children can be introduced to breathing exercises in the form of a game from the age of two or three. From the age of nine, most children can start doing part of the physiotherapy for themselves.

Why is chest physiotherapy important?

Chest physiotherapy helps prevent thick, sticky mucus in the lungs from blocking the airways, which can reduce infection and prevent lung damage. Parents are taught how to do physiotherapy with their child by the physiotherapist in the CF clinic. Adults with cystic fibrosis can learn to carry out their own physiotherapy.

Airway clearance techniques (ACTs) help move mucus out of the lungs. The body's normal and basic ACT is a cough, which is a reflex that clears mucus with high-speed airflow. However, in cystic fibrosis, the mucus cannot be cleared by coughing alone. People with CF should do airway clearance every day to keep lungs healthy, even when they are feeling well. When sick, airway clearance should be done more often.

There are lots of different airway clearance techniques, and a specialist CF physiotherapist will conduct an individual assessment, and advise on the most appropriate technique to use, as well as the length and frequency of treatment sessions.

This table shows the number and percentage of people with cystic fibrosis who are using the different airway clearance techniques recorded on the Registry in 2012. Individuals may use more than one technique.

ACTs recorded on the Registry in 2012

Technique	Overall 8794 Number (%)	< 16 years 3732 Number (%)	≥16 years 5062 Number (%)
Active cycle of breathing techniques [ACBT]	3525 (40.8)	1861 (51.6)	1664 (33.1)
Autogenic drainage (including assisted autogenic drainage)	971 (11.2)	137 (3.8)	834 (16.6)
Any form of Positive Expiratory Pressure [PEP]	4330 (50.1)	2207 (61.2)	2123 (42.2)
VEST	157 (1.8)	94 (2.6)	63 (1.3)

Why is exercise important?

Exercise is particularly important for people with cystic fibrosis because it helps to clear mucus from the lungs and improves physical bulk and strength.

Children with cystic fibrosis should be encouraged to take part in as much physical activity as possible – ideally types of exercise that leave you out of breath, like running, swimming, football or tennis. It is important to let teachers at school know that exercise should be encouraged because they may not know if it is good or bad for someone with cystic fibrosis.

Your hospital physiotherapist can advise on the right exercises and activities for you or your child.

Keeping lungs healthy – medication

Medication can be taken through various methods, including inhaling into the lungs using nebulisers, taking orally or injected.

- Bronchodilator drugs open your airways by relaxing the surrounding muscles, relieving tightness and shortness of breath.
- Antibiotics help treat or control infection.
- Steroids to reduce inflammation in the airways can be used in specific circumstances.
- Mucolytics such as DNase breakdown mucus, making it easier to clear.

Nebulised drug treatment by age – DNase

	2008		2012	
	Number	(%)	Number	(%)
0–3 yrs	46	(7.6)	97	(10.4)
4–7	125	(20.1)	306	(31.8)
8–11	227	(34.2)	445	(51.6)
12–15	359	(46.4)	612	(62.5)
16–19	377	(49.5)	587	(59.1)
20–23	319	(44.0)	626	(61.9)
24–27	288	(47.6)	475	(58.2)
28–31	182	(43.4)	395	(58.2)
32–35	108	(41.5)	237	(51.6)
36–39	83	(35.0)	141	(46.5)
40–44	147	(35.7)*	156	(47.0)
45–49			105	(46.3)
50+			104	(43.0)
Overall	**2261**	**(37.2)**	**4286**	**(48.7)**

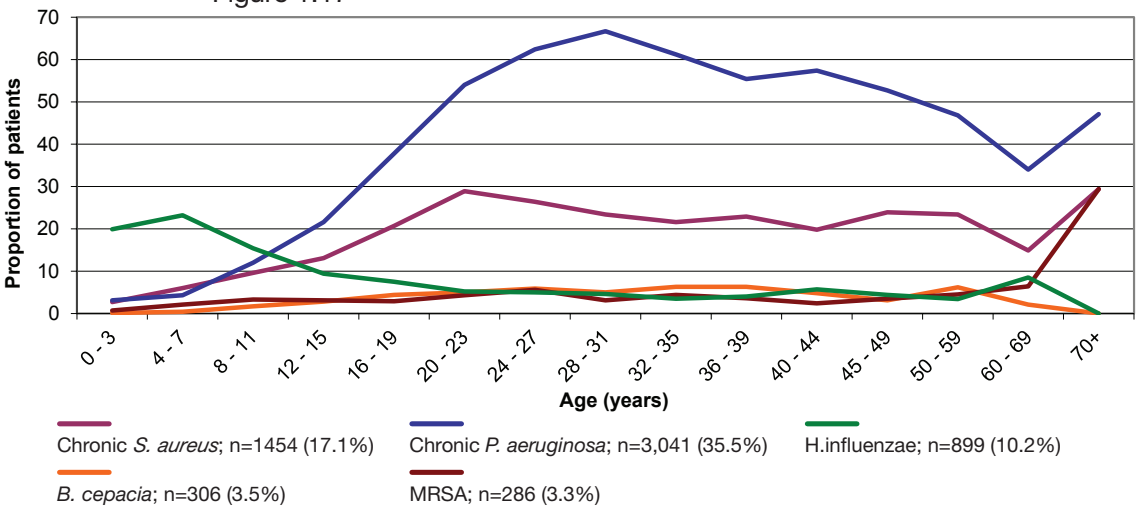
*In 2008 those aged 40 years and older were grouped together

The figures in the table indicate an increase in the use of nebulised drug treatment in 2012 compared to 2008 in all age groups, and this is something we hope will result in better long-term outcomes.

Infection

Lung infections by age group in 2012

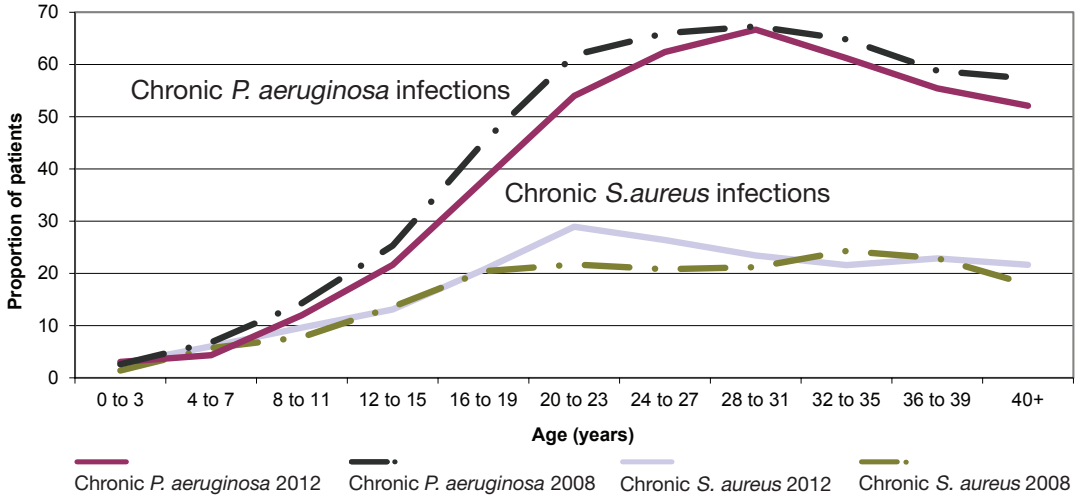
Figure 1.17



Current treatments and good cross-infection measures mean that we can aim to reduce the number of people with cystic fibrosis transferring from paediatric to adult care with chronic *Pseudomonas aeruginosa* infection. The current objective is for fewer than 30% of the paediatric population to be chronically infected at the time of transfer. A future aim is to see this reduce to less than 20%.

In the following graph we show the improvement in *Pseudomonas aeruginosa* and *Chronic S.aureus* infections between 2008 and 2012

Figure 1.18



This graph shows a small reduction in chronic pseudomonas in 2012 compared to 2008, across many age groups. When focusing on young adults: 16–19 years and 20–23 years, we found that the reduction was statistically significant. A difference described as ‘statistically significant’ means that it is very unlikely that the difference observed is purely down to chance.

This improvement may be due to aggressive early treatment to eradicate the infection and an increasing awareness of cross-infection leading to segregation.

Medication: The consensus view in the UK is that 90% of people with cystic fibrosis, who are chronically infected with *Pseudomonas aeruginosa*, should be prescribed at least one of the nebulised antibiotics in this table.

Nebulised antibiotic use in 2008

People with chronic Pseudomonas	Overall 2098	<16 years 299	≥16 years 1799
	Number (%)	Number (%)	Number (%)
Tobramycin solution	412 (19.6)	48 (16.1)	364 (20.2)
Other aminoglycoside	43 (2.0)	5 (0.2)	38 (2.1)
Colistin	914 (43.6)	174 (58.2)	740 (41.1)
Promixin	490 (23.4)	73 (24.4)	417 (23.2)
At least one of above in 2008	**1597 (76.1)**	**257 (86.0)**	**1340 (74.5)**

Nebulised antibiotic use in 2012

People with chronic Pseudomonas	Overall 3041	<16 years 377	≥16 years 2664
	Number (%)	Number (%)	Number (%)
Tobramycin solution	1018 (33.5)	120 (31.8)	898 (33.7)
Other aminoglycoside	104 (3.4)	11 (2.9)	93 (3.5)
Colistin	1326 (43.6)	214 (56.8)	1112 (41.7)
Promixin	810 (26.6)	133 (35.3)	677 (25.4)
At least one of above in 2008	**2444 (80.4)**	**340 (90.2)**	**2104 (79.0)**

"Collecting and inputting of data can be a thankless task, but the diligence of the CF centres has now brought together a powerful body of knowledge that is being used to further treatment and improve outcomes in cystic fibrosis. It is in the power of numbers that trends can be seen and the benefit of newer treatments understood."

Marian Dmochowska, parent of two children with cystic fibrosis

Cross-infection, or cross-contamination, occurs when one person spreads an infection to another, either directly or indirectly. For people with cystic fibrosis, cross-infection can be very harmful and poses a particular threat, which is why people with CF should not meet face to face. The risk of cross-infection increases the longer people with cystic fibrosis are in close proximity to one another. There is less risk of transmission of 'bugs' in an outdoor environment, but meeting indoors, travelling with other people with cystic fibrosis, or spending time with them socially has a high level of risk.

Cystic fibrosis centres and clinics are advised to ensure segregation measures are in place.

What is the difference between prevalence and incidence?

Prevalence is the proportion of the cystic fibrosis population on the Registry with a condition whereas the incidence is the proportion of people with CF for whom the year in question was the first year in which the condition was reported.

Complications

Complications are problems related to CF such as cystic fibrosis-related diabetes (CFRD).

CFRD is one of the most common CF complications and is different from diabetes in people without cystic fibrosis. In 2012, 29% of people aged 16 years and older were receiving treatment for CFRD.

Because research shows that early diagnosis and treatment of CFRD leads to better nutrition and health, people with cystic fibrosis aged ten years and older should be tested every year for CFRD. The test is called an oral glucose tolerance test (OGTT).

Number and % of people with cystic fibrosis receiving treatment for CFRD

	Overall (n=8772) Number (%)	<16 years (n=3715) Number (%)	≥16 years (n=5057)Number (%)
Treatment for CF-related diabetes	1641 (18.7)	153 (4.1)	1488 (29.4)

Prevalence (see box, left) of other complications reported in 2012 in percentage of people with cystic fibrosis

	Overall (n=8794) Number (%)	<16 years (n=3732) Number (%)	≥16 years (n=5062) Number (%)
ABPA	909 (10.3)	278 (7.4)	631 (12.5)
Nontuberculous mycobacteria or atypical mycobacteria	465 (5.3)	94 (2.5)	371 (7.3)
Pneumothorax requiring chest tube	39 (0.4)	1 (0.03)	38 (0.8)
Nasal polyps requiring surgery	205 (2.3)	47 (1.3)	158 (3.1)
Cirrhosis with portal hypertension	164 (1.9)	24 (0.6)	140 (2.8)
Gallbladder disease requiring surgery	32 (0.4)	2 (0.1)	30 (0.6)
Fibrosing colonopathy/ colonic stricture	5 (0.1)	2 (0.1)	3 (0.1)
Cancer confirmed by histology	18 (0.2)	3 (0.1)	15 (0.3)
Osteoporosis	493 (5.6)	7 (0.2)	486 (9.6)
Arthropathy	463 (5.3)	18 (0.5)	445 (8.8)

Incidence of other complications reported in 2012 in percentage of people with CF

	Overall (n=8794) Number (%)	<16 years (n= 3732) Number (%)	≥16 years (n=5062) Number (%)
ABPA	169 (1.9)	64 (1.7)	105 (2.1)
Nontuberculous mycobacteria or atypical mycobacteria	159 (1.8)	30 (0.8)	129 (2.5)
Cancer confirmed by histology	10 (0.1)	1 (0.02)	9 (0.2)
Cirrhosis with portal hypertension	18 (0.2)	6 (0.2)	12 (0.2)

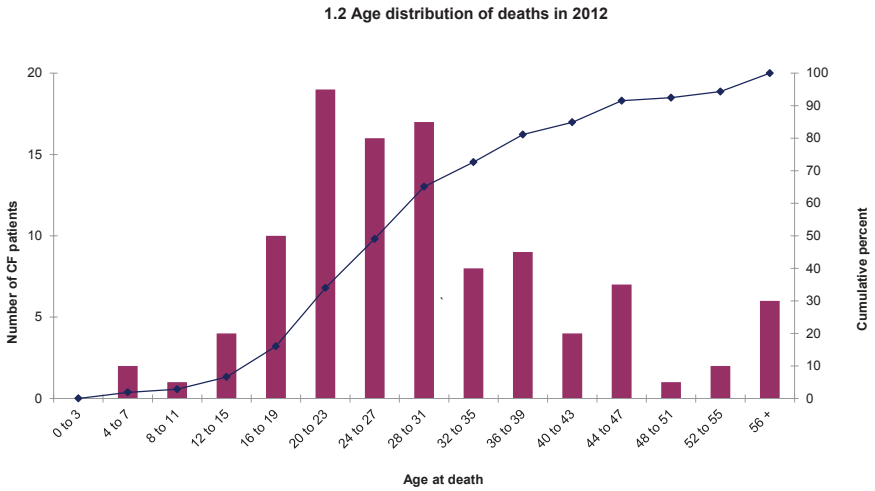
Survival

The median predicted age of survival for people with cystic fibrosis has risen steadily over the last 25 years, due to a number of factors. These include specialist centre care, better nutritional support and improved treatment of lung complications with aggressive use of antibiotics. The median survival for the CF population is currently 43.5 years, compared to 38.8 for the population in 2008.

There were 106 recorded deaths in 2012. The median age at death was 28 years, with a range from 5 to 72 years. Year-to-year fluctuations are normal when measuring health outcomes for any disease or medical condition. What is important is the steady increase in survival over time.

Age distribution of deaths in 2012

Figure 1.2



There were 106 recorded deaths in 2012. The median age at death was 28 years (min = 5 yrs; max = 72 years; 95% confidence interval: 29 years).

Is there an association between newborn screening and survival?

The Registry works closely with the CF UK Newborn Screening Programme Centre, providing data to ensure the best audit of outcomes of screening for CF is performed. Since October 2007 all newborns in the UK are screened for cystic fibrosis. It is thought that early diagnosis may play an important role in improving survival. Research shows that people with cystic fibrosis who are diagnosed because of newborn screening have better weight and healthier lungs later in life than those diagnosed because of symptoms.

The earlier cystic fibrosis is diagnosed, the sooner treatment can begin.

Of the 63 children with complete data born in 2012, 50 (79%) were identified by newborn screening.

To help CF doctors and nurses care for babies diagnosed with cystic fibrosis because of newborn screening, the Cystic Fibrosis Trust has worked with experts in CF and infant care to develop care standards. The CF infant care standards outline cystic fibrosis care to keep babies with CF as healthy as possible. 'The Standards of Care of Children and Adults with Cystic Fibrosis in the UK' can be downloaded from cysticfibrosis.org.uk/publications.

Transplantation

Great steps forward in specialist care and treatment have meant that people with cystic fibrosis are living longer and healthier, but some will reach a point where they require a lung transplant to prolong their life.

The following table shows the number of transplants in recent years.

	2012	2011	2010
Number of PwCF that year with annual review data evaluated for transplants	225	204	169
Number accepted on the transplant list	120	121	82
Number receiving transplants	55**	51*	29
Types of transplants received:			
Bilateral lung	45	43	26
Heart and lung	1	4	1
Liver	7	2	1
Kidney	3	2	1
Pancreas	1	1	-

* One person received two transplants
** Two people received two transplants

Research exploring the potential for improving the quality of donor lungs

DEVELOP-UK is a study at the University of Newscastle, led by Professor Andrew Fisher, which is seeking to increase the number of lungs available for transplant. The research is co-funded by the Cystic Fibrosis Trust and the National Institute for Health Research. The study involves assessing and improving the quality of donated lungs that are not immediately suitable for transplant to allow them to be safely used. The results could change the way lung transplants are performed in the UK and internationally, and reduce the number of patients that die on the transplant list due to a shortage of suitable donor organs.

The success rate of lung transplants for people with cystic fibrosis is encouraging, and recipients will enjoy a good quality of life, but the procedure does carry considerable risks, including rejection or infection. It is also important to remember that a transplant is not always the most appropriate form of treatment for someone who is severely ill with cystic fibrosis.

The transplant process is a lengthy one, and people with cystic fibrosis and their families can often face a range of physical and emotional issues. To help you deal with some of these problems, it is routine practice to offer assessment and care at every stage. This is particularly important during the pre-transplant phases.

Liver transplants

A small proportion of people with cystic fibrosis will have severe CF-related liver problems leading to the need for transplantation.

Most people with CF-related liver problems show little or no deterioration over time and often a clinic review about once a year with a scan and blood tests is all that is required. Complications of CF-related liver disease can be managed with monitoring and treatment, however a small minority of people do deteriorate and then need to be considered for liver transplantation.

Liver transplantation in people with cystic fibrosis has proved to be a success, despite early concerns about the outcome of the procedure and the effects of immunosuppression on respiratory infection.

Key messages

These are some of the key messages from the analysis of the registry data for 2012. Increasingly, we are able to compare figures over time in order to show where outcomes are improving.

- The 2012 data demonstrates a rise in the use of therapies designed to maintain lung function and reduce exacerbations. The increase in the use of nebulised drug treatment in 2012 compared to 2008 is in all age groups and should result in better long-term outcomes.
- There has been a reduction in chronic pseudomonas infection in 2012 compared to 2008. In the 16-–19 and 20-–23 age groups this reduction was statistically significant. This suggests that implementation of strategies of aggressive early treatment of pseudomonas infection, coupled with adherence to segregation guidelines, are successful in reducing chronic PA infection. In the future we hope this will lead to better outcomes and lower requirements for intravenous antibiotic therapy.
- It is good to see median survival steadily improving in keeping with registry data from around the world. The median survival for the UK CF population is currently 43.5 years, compared to 38.8 for the population in 2008

Further information about how the Registry is managed, how we look after the data entered by the CF centres, and how this information is put to good use, can be found in the Registry Review 2012 which can be downloaded from the Cystic Fibrosis Trust website at: cysticfibrosis.org.uk/registry

ANNEX 1 Governance of the Registry

The protocol governing the Registry was approved by the National Research Ethics Service in 2007 and the Registry Steering Committee was set up to oversee how the Registry is managed. It meets at least twice a year and is chaired by Dr Diana Bilton.

The Committee is responsible for:

- ensuring compliance with the protocol governing the conduct of the Registry;
- providing direction on the strategic development of the Registry;
- screening applications for access to Registry data;
- monitoring of progress with projects; and
- the production of annual reports and information on Registry developments which are made available to clinicians, people with cystic fibrosis and parents.

The membership of the Committee comprises representatives from the Cystic Fibrosis Trust, people with CF and their carers, CF centre clinical directors, NHS commissioners and the Trust's academic partner, Imperial College. The full membership of the Committee is detailed on the next page.

Annex 2: Registry Steering Committee membership

Dr Diana Bilton (Chair)	Adult CF Centre Director, Royal Brompton Hospital, London
Dr Caroline Elston	Adult CF Centre Director, Kings Hospital, London
Dr Iolo Doull	Paediatric CF Centre Director, Cardiff Hospital, Wales
Dr Keith Brownlee	Paediatric CF Centre Director,St James Hospital, Leeds
Dr Siobhan Carr	Paediatrician, Royal Brompton Hospital, London
Dr Steve Cunningham	Paediatrician, Edinburgh Royal Infirmary, Scotland
Dr Martin Wildman	Adult CF Centre Director, Northern General Hospital, Sheffield
Professor Stuart Elborn	Adult CF Centre Director, Belfast, NI & Trustee of CF Trust
Dr Stephanie MacNeill	Biostatistician, Imperial College, London
Mrs Marian Dmochowska	Parent Representative
Mr Dominic Kavanagh	Patient Representative
Ms Katherine Collins	Director NSD, Scotland
Ms Carrie Gardner	Specialist Commissioner, London
Dr Kim Cox	Lead Specialist CF Commissioner, London
Dr Lisa Davies	Specialist Commissioner, Wales
Mr Ed Owen	Chief Executive, Cystic Fibrosis Trust
Ms Joanne Osmond	Director of Clinical Care, Cystic Fibrosis Trust
Mr Stephen Williams	Director of Finance, Cystic Fibrosis Trust
Dr Janet Allen	Director of Research, Cystic Fibrosis Trust
Ms Elaine Gunn	Registry Manager, Cystic Fibrosis Trust

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