

**Cystic**  
**Fibrosis**  
*our focus*

**UK Cystic Fibrosis Registry Review 2012**

March 2013

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# Introduction

**The UK Cystic Fibrosis Registry is internationally recognised as one of the most successful disease registries; so much so that we are being approached by other charities interested in learning from our model.**

The overall aim of the Registry is to achieve continuing improvements to the quality and safety of care; it achieves this by providing high-quality data and analysis, and by supporting research, including studies observing the safety of new drugs. We also have a role in influencing government policy through demonstrating the value of disease registries.

This review aims to raise awareness of the Registry and its achievements, not least to say a big thank-you to the patients who agree to share their information, and to the clinical teams who collate and enter data for the Registry. We envisage that a review, supplementing the Annual Report, will be published every two years.

“The UK CF Registry has developed into a robust and trusted tool to follow the progress of cystic fibrosis care in the UK and to ensure we continue to improve outcomes for people with cystic fibrosis. This success story directly relates to investment from the Cystic Fibrosis Trust, the agreement of parents and people with cystic fibrosis to have their outcomes entered and the hard work of centre teams to provide accurate and timely data. We want to ensure that all stakeholders are informed about the huge impact the registry is having in commissioning care and in supporting research.”  
Dr Diana Bilton, Chair, UK CF Registry Steering Committee.

# Contents

## **Section 1:**

The Registry: in this section we will describe how information for the Registry is collected and anonymised, the quality assurance process and the work of the Registry Steering Committee which governs access to the Registry. We provide some facts and figures on why the Registry is so well regarded and an example of how it is used for research.

## **Section 2:**

Interpreting the annual report: providing a summary and interpretation of the analysis in the latest annual report, highlighting the main messages, in particular facts and figures illustrating improvements in care over the last several years.

## **Section 3:**

Working with the NHS: The main purpose of the Registry is to drive up standards of clinical care. In this section we will report on how the Registry is being used in partnership with clinical centres and the NHS to improve care and to inform the planning and commissioning of services for patients.

## **Section 4:**

Working with industry: This is a recent development in which we are working with pharmaceutical companies to improve the safety monitoring of new drugs in return for income that will help to provide clinical centres with a Registry Support Grant.

## **Section 5:**

Policy developments: In 2011 the government produced a Life Sciences Strategy in which the Prime Minister included NHS data as one of the five key strengths of UK life sciences. In this section we will explore some of the implications of the initiatives arising from this strategy.

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**Data is well protected**

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**Data is used to support projects which are aimed at improving the safety and care of patients**

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## **Section 1: The Registry**

The Registry is governed by the Registry Steering Committee, chaired by Dr Diana Bilton and with members representing adult CF physicians, CF paediatricians, people with cystic fibrosis and their carers, and commissioners of NHS services.

There are currently 58 CF centres and over 80 CF clinics entering information from 9,700 patients onto the web based Registry; this is 99% of the people with cystic fibrosis in the UK.

- Each person is registered, with their consent, as soon as a patient is diagnosed with cystic fibrosis.
- Entries can be made at each patient encounter and most importantly at their annual review.
- We are very proud of the fact that 89% of data entries are completed. This is very high for this type of database and is a key factor in the success of the Registry, and a testament to the centres' commitment.

The Registry has a dedicated manager employed by the Cystic Fibrosis Trust: Elaine Gunn is an experienced respiratory nurse who has worked in cystic fibrosis research, registries and databases since 1994.

### **How is data protected?**

The Registry is held on computer in a password protected, locked office at the Trust's headquarters in accordance with the ethical requirements for approval of the registry.

The management of the Registry conforms to the Data Protection Act. Each person registered is given a unique ID number and data is anonymised using this number.

The Registry is tested with respect to the encryption and safety of the data held, and there are elaborate encryption protocols governing user access.

Regular monitoring visits to the CF centres ensure that data entry staff are given training and support. All entries are checked to ensure accuracy and random sets of patients' notes are reviewed to ensure no bias in the registry data.

**“The UK CF Registry has developed into a robust and trusted tool to follow the progress of cystic fibrosis care in the UK and to ensure we continue to improve outcomes for people with cystic fibrosis.”**

Dr Diana Bilton, Chair, UK CF Registry Steering Committee.

### **What happens to the data?**

- A National Report is published annually, with analysis of the data undertaken by statisticians from the National Heart and Lung Institute at Imperial College, London. The Report is available on the Cystic Fibrosis Trust website ([cysticfibrosis.org.uk](http://cysticfibrosis.org.uk)).
- Researchers submit requests for information from the Registry and these undergo formal review by the Registry Steering Committee.
- A recent example of how Registry data has been used for research of international significance, is summarised in the box, page 5.
- In Sections 3 and 4 we describe how the Registry is being used to improve the quality and safety of patient care, in collaboration with NHS commissioners and industry.

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The Registry Annual Report supports efforts to continually improve care

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There are many influences on lung function, such as age, infection status and socio-economic status, so direct comparisons of clinics without full and careful analysis of all these factors may be misleading

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89%

of the data in the Registry is complete

99%

Details of 99% people with cystic fibrosis in the UK have been added to the Registry

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cystic fibrosis centres and clinics enter patient information on the Registry

## Section 2: Interpreting the Annual Report

The annual report this year, with analysis from data entered on the Registry in 2011, shows

- we have improved the level of complete data to 89%.
- average survival is similar to last year at 41.5 years.
- there has been a significant increase in the numbers of bilateral lung transplants, from 26 in 2010 to 43 in 2011.

### How are we doing in achieving good lung function?

Lung function levels are reported as a percentage compared to the expected normal so that anything above 85% is considered within the normal range for the general population. 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.

### How are we doing in treating *Pseudomonas* infection?

The consensus view in the UK is that 90% of people chronically affected by *pseudomonas* should be prescribed one of the range of nebulised antibiotics available. In the 2011 analysis, the overall figure was 82%.

### How do we compare centres?

We include CF centre-specific data for some of the key outcomes such as lung function and nutritional status. It is important to understand that there are many influences on lung function, such as age, infection status and socio-economic status, so direct comparisons of clinics without full and careful analysis of all these factors may be misleading.

The Registry Steering Committee has teamed up with national experts and gained research funding from the National Institute of Health Research (NIHR). The aim of the research is to develop better ways of comparing centres, to ensure we identify best practice and best outcomes and share them with everyone. We expect this research to allow us to produce future reports with more accurate and detailed centre comparisons.

### The effect of social deprivation on clinical outcomes and the use of treatments in the UK cystic fibrosis population: a longitudinal study<sup>1</sup>

This study, utilising the UK CF Registry, found that children from the most deprived areas weighed less, had a lower body-mass index, were more likely to have chronic *P aeruginosa* infection and a lower %FEV1. After adjustment for disease severity, these children were more likely to receive intravenous antibiotics and nutritional treatments and less likely to receive DNase or inhaled antibiotic treatment, compared with children from the least deprived areas.

In conclusion, children with cystic fibrosis from more disadvantaged areas have worse growth and lung function compared with children from more affluent areas, but these inequalities do not widen with advancing age. Clinicians consider deprivation status, as well as disease status, when making decisions about treatments, and this might mitigate some effects of social disadvantage.

## Section 3: Working with the NHS:

The Cystic Fibrosis Trust has a strategic approach to improving quality of care through:

- setting standards of care,
- reviewing those standards through the Peer Review Programme; and
- monitoring outcomes through the Registry.

The formulation of *Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK* was undertaken in an effort to ensure that all patients have equal access to the highest level of multidisciplinary specialist care. The second edition of the Standards was published in 2011 and is a consensus document approved by the British Thoracic Society (BTS) Specialist Advisory Group.

Since 2006 our peer review work, assessing standards of care, has resulted in massive improvements in care and an additional £20 million in funding to CF centres across the UK.

The programme was revamped in 2012, working alongside the British Thoracic Society and British Paediatric Respiratory Society. The result is a review programme which is recognised by CF teams across the country as an effective, fair and efficient method of assessing CF centres.

The NHS in England and Scotland have contracts with the Cystic Fibrosis Trust for the provision of information from the Registry to help inform the planning and commissioning of services. The purpose is to improve the quality and efficiency of care provided to patients.

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**Information from the Registry helps the NHS to ensure that the funding of services is based on individual needs**

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From April 2013 we will see a new national funding system for cystic fibrosis in NHS England, which should lead to greater consistency and fairness in funding of cystic fibrosis care, nationally. Payment by Results (PbR) will mean that people with cystic fibrosis receive care funded according to individual needs, including the drugs they are prescribed. Information from the Registry makes sure that funding is allocated according to levels of disease severity. The aim is to provide an equitable, transparent, rules-based system.

## Section 4: Working with industry

Drug companies are required to conduct studies looking at the long-term safety of new drugs, known as pharmacovigilance, as a condition of gaining a marketing licence. Recent guidelines from the European Medicines Agency [EMA] are helping to raise standards for these studies by producing a standard study protocol.

The Cystic Fibrosis Trust has been approached by several companies wishing to utilise the Registry to conduct their long-term safety studies after the introduction of new therapies; the use of independently held disease registries for these studies is a practice encouraged by the EMA.

The Cystic Fibrosis Trust is a leader in this development; our primary objectives are to

- improve the quality of these observational studies and
- ensure that patients gain early access to new drugs that have been through a rigorous safety assessment.
- secure funding to help in the running costs of the Registry

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## Providing companies with anonymised data reports from the Registry is helping to improve safety studies

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## Income received from working with industry is providing grants to CF centres to help support staff inputting data to the Registry

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### Public opinion on researchers accessing patient records to improve healthcare

- In 2009 a **survey of 1,179 UK adults** found that 74% were willing to allow access to their personal medical records for medical research.
- An **Ipsos MORI poll of nearly 1,000 adults in 2011** found that 80% would like their doctor to offer them the opportunity to allow a researcher confidential access to their medical records with no other involvement from them.
- The **British Heart Foundation got a similar result when they asked just over 2,000 people in a YouGov poll this year** – 72% said they would be happy to share their entire medical records with researchers under certain conditions, such as if the data were anonymised or key-coded – and an additional 7% were happy for records to be shared with identifiable elements of the data left in.

This programme of work is being conducted under the aegis of the Registry Steering Committee, with professional support on the design of the study protocols provided by Imperial College, London (ICL).

The Trust has sought legal and contractual advice to ensure that the service level agreements with the companies are appropriate. The approach of the Trustees has been to follow guidelines for working with industry, produced by the Association of Medical Research Charities: the guiding principles are Independence, Integrity and Openness.

This work with industry has been discussed with the National Research Ethics Service who have confirmed that our current patient consent procedures cover this kind of activity. The Cystic Fibrosis Trust has produced an information leaflet to ensure that clinicians and patients are fully informed as its work with industry develops. Copies can be downloaded from the Trust's website ([cysticfibrosis.org.uk/registry](http://cysticfibrosis.org.uk/registry)).

The first contract is signed and we are hoping to reach agreement with two further companies in the next few months.

Each company will pay the full costs of their study and the income received by the Trust has provided an opportunity, for the first time, to provide a Registry Support Grant to each clinical centre. We are delighted that we now have a source of funding to reimburse centres for the time spent inputting data to the Registry.

## Section 5: Policy developments

### Access to NHS data

The UK Life Sciences Strategy, published in 2011, announced a consultation on some changes to the NHS Constitution that sets out what patients, staff and the public can expect from the NHS in England.

Recognising the need to support patients' access to new treatments, and the need to protect the right of an individual to opt out, the default position being proposed is that data collected as part of NHS care can be used for approved research, with appropriate protection for patient confidentiality. The implications for the UK CF Registry is that we would still inform patients about the Registry and provide information on how Registry data is being used. Patients would still have the right to opt out.

The results of this consultation are due in 2013 and our responses will be published on the Trust's website and we will keep you informed.

### Why is the government making this proposal?

- A recognition that the UK is under-utilising its strengths, including the incredibly valuable mine of information held by the NHS
- Regulation can deter people from doing research if it is overly bureaucratic and can also reduce the speed with which we can benefit from research, so it is important to keep it under review
- The NHS is seen as a slow adopter of innovation – things we know work, from the research evidence – can take ages to reach the patient
- We need to give patients more opportunities to take part in research which may benefit them, or future generations

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**The Cystic Fibrosis Trust is promoting the benefits of Registries as collections of anonymised data that can improve patients care**

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### **Promoting disease registries**

The Cystic Fibrosis Trust continues to use every opportunity to promote the benefits of disease registries with policy makers in the UK and Europe, and with other charities.

For example, in summer 2012, the Trust was invited to present a poster about the Registry at an *All Party Parliamentary Group on Medical Research* event at the House of Commons – an opportunity to showcase the CF Registry on the theme – *How data saves lives – Unlocking the research potential of information*.

### **Looking forward**

During 2013, we will be undertaking a strategic review of the Registry which will consider the next steps in development, setting goals for the next 3–5 years. One aspect of this will be working with CF clinicians, researchers, industry and NHS commissioners, to look at opportunities to refine the content of the Registry so that it continues to provide the essential information needed to improve care and to support research.



For more information, visit:  
**[cysticfibrosis.org.uk](http://cysticfibrosis.org.uk)**