

## **UK CF REGISTRY ANNUAL DATA REPORT 2009 - FAQs**

### **How do I understand what the report means?**

The CF Registry Report is a comprehensive set of clinical outcome data from all Specialist Cystic Fibrosis Centres in the UK and over 80 network clinics that provide shared care in conjunction with a Centre. The information is presented in chart form to allow for ease of reading and contains information at an individual service level on average FEV<sub>1</sub> (forced expiratory volume in 1 second) which is a measure of lung function, average BMI (Body Mass Index), which is a method of measuring overall nutritional status, and average rate of chronic (established) infection with bacteria such as *Pseudomonas aeruginosa*.

Each Centre or clinic has been allocated a number, and you can find out which number corresponds to your Centre or clinic by referring to the key at the back of the report. Although Centres and clinics are identified patient information is anonymised.

### **What does complete data mean?**

Complete data in the context of the CF Registry refers to the submission of a minimum set of data required to produce the information in the report. The data set is taken from a patient's annual review, whether this occurs as a 'one stop' annual MOT with the CF clinical team or the same data set collated over the period of a year.

### **Why is the number of patients registered higher than the number of patients with 'complete' data?**

The number of patients registered on the database represents the total number of live patients for whom there are full demographic (name, address, date of birth) details in 2009. The number of patients with 'complete' data represents those for whom the minimum clinical data set has been submitted by their clinical team over the preceding 12 months. The gap between these two statistics is closing year on year as services are working hard to make sure that a full data set is submitted for every patient registered.

### **What does median predicted survival mean?**

Median predicted survival is the age beyond which half of current UK CF Registry patients would be expected to live. It takes into account the ages of patients on the Registry and the distribution of deaths in the same year. So for those people on the CF Registry (up until the end of 2009), half of them would be expected to live longer than 34.4 years, and sadly, half of them would not. This statistic should be interpreted with caution, acknowledging the fact that this figure is likely to fluctuate from year to year for a variety of reasons, including the proportion of patients with complete data and the

number of deaths in that year for example. It will be more helpful to analyse this particular statistic over a longer period of time, e.g. every 5-10 years in order to mitigate any random fluctuations that may occur and identify any trends.

### **What is the difference between median predicted survival and median age of death?**

The median age of death means that during the reporting year (in this case 2009), of the reported deaths, half were below 27 years and half were above. The statistic for median predicted survival is higher because this includes **all** people on the Registry including those born more recently who, since birth, have benefitted from the greater understanding of CF, a better level of specialist care, earlier and more effective treatments than were available in the past.

### **I am worried that median predicted survival has gone down and the number of deaths has gone up.**

It is true that the 2009 report suggests that the number of deaths has risen since 2008 and median predicted survival is lower. It is important to note that the report is a snapshot of data from a single year and we expect that statistics will fluctuate from year to year. In 2008, there were fewer patients on the registry with complete data, so it may be that this affected the data from that year. It will be more helpful for us to look at both of these statistics over a longer period of time to mitigate any random fluctuations that may occur from one year to the next.

### **How can I tell from this data if I/my child is doing well?**

The information in the report is useful as it helps us build up a general picture of the CF population in the UK. However it does not take into account an individual's experience with CF. For example some people are very well and have a healthy FEV<sub>1</sub>, others will have had infection recently that will have affected their health. CF can affect people differently so there is no 'average' person with CF. We suggest you talk to your clinical team if you have any questions or queries about your own data or that of your Centre/clinic.

### **My Specialist CF Centre or clinic is near the bottom in some of the tables. Does this mean I am not getting good care?**

The charts presented in the CF Registry report are not meant to present as league tables and should not be read as such. There may be many reasons why a particular CF Specialist Centre or clinic has produced its averages for such measures as predicted lung function or body mass index.

Examples could include such reasons as a small clinic only having a few, very young children, all with high lung function, or conversely, a Centre that is looking after a great many older people with CF who have a lower lung function, but who are being kept in a stable condition. Indeed, as people with CF are living longer, this is increasingly likely to be the case. There may also be cases where people who have not been doing as well in a network clinic setting have been transferred to a CF Centre to help improve their health.

It is also true, however, that the care of those with Cystic Fibrosis in some Centres and clinics is provided by an under-resourced service and this may contribute to the underlying reasons why there is lower lung function or BMI averages. The charts therefore need to be interpreted with caution. We would suggest that if you have concerns relating to the figures you discuss them with your clinical team.

### **Should I move my care to another hospital?**

As there are so many factors that can affect the average levels of BMI, lung function and other measures in the CF Registry report, it does not necessarily mean that care at a particular CF Centre or clinic is of a higher or lower standard than another service. The report should not be read as a league table as the individual outcome data charts do not consider many other factors.

It is, however, a right within the NHS that people are able seek care wherever they choose. For a specialist and complex condition like Cystic Fibrosis, it is accepted that the best care is provided by a multidisciplinary team comprising CF doctor, nurse dietitian, physiotherapist, psychologist and social worker, supported by other professionals such as pharmacists and microbiologists. The team need to be experienced and up-to-date in CF care, and this comes not only through specialist training, but also by working in an environment dedicated to Cystic Fibrosis and maintaining their professional development with regard to the most up-to-date clinical practice and treatments. Without this expertise, details that would be picked up in a specialist environment may be missed.

### **What is the difference between a CF Centre and a CF network clinic?**

A Specialist CF Centre can treat either children or adults. It usually has at least 100 patients and often many more. The CF Centre will have a full multidisciplinary team of specialists and a full range of other services that people with CF often require access to, e.g., specialists in diabetes, liver disease, bones, ENT (ear, nose and throat) etc. A CF network clinic is smaller than a CF Centre and treats only paediatric patients. It provides some aspects of care to children with CF in a local setting under the guidance of the CF Centre to which they are linked and may hold joint visits with the team from the Specialist CF Centre.

### **I have been told that CF Centre care is better than shared care. Some clinics appear to have higher averages, so is this true?**

For people with Cystic Fibrosis, best care is provided by a multidisciplinary team of doctors, nurses, dietitians, physiotherapists, psychologists, social workers and other health professionals. For the care of children with CF two models of care are recognised: full care at a Specialist CF Centre, or shared care between the Specialist CF Centre and a local CF network clinic. The child may be seen at their local CF clinic by a local team, and by the Specialist CF Centre team. For the care of adults with Cystic Fibrosis, it is recognised that only care provided by a multidisciplinary team working at

a Specialist CF Centre will be appropriate. Due to particular geographic challenges some patients in the Highlands and Islands of Scotland receive 'outreach' care which is provided by the Specialist CF Centre team who travel to a local hospital to ensure specialist input for these patients who would find it incredibly difficult to travel frequently to the Specialist CF Centre.

A network clinic may appear to be in a higher position on the charts, but as they are **not** league tables, this should not infer that care at the clinics is better. A particular clinic may only have few very well patients or a clinic may only be used as a location where the person with Cystic Fibrosis receives care from a Specialist CF Centre multidisciplinary team. Some clinics may also hand over care of people who are particularly unwell to a Specialist CF Centre. It is not always possible to draw conclusions about quality of care from the data presented as this represents a snapshot of raw data that does not take into account many other variables.

**My Specialist CF Centre or clinic is at the top of some of the tables. Does this mean I'm getting the best care?**

The charts presented in the CF Registry report are **not** league tables and should not be read as such. There may be many reasons why a particular CF Specialist Centre or clinic has their specific averages for such measures as lung function or body mass index.

Examples could include such reasons as a small clinic only having a few, very young children, all with high lung function, or conversely, a Centre that is looking after a great many older people with CF who have a lower lung function, but who are being kept in a stable condition. Indeed, as people with CF are living longer, this is increasingly likely to be the case. There may also be cases where people who have not been doing as well in a smaller clinic setting have been transferred to a CF Centre to help improve their health.

**The hospital where I get my care in is not in the report. Why is this?**

The CF Registry contains data from all of the UK's Specialist CF Centres, over 80 CF network clinics and 13 stand alone services. We are working to extend the database to any remaining CF services. It may be that your hospital, particularly if you receive shared care, has pooled its data with the Specialist CF Centre with whom it forms part of a network service.

Only information from known CF network clinics and Centres will be on the database however, as all people with Cystic Fibrosis should be attending one of these services to ensure they receive the best care; this is simply not available from a District General Hospital alone that does not have access to specialist CF treatment, care and advice.

**What is the CF Trust doing to help improve care?**

The CF Trust is undertaking a comprehensive series of peer reviews of CF services. A team of expert CF health professionals and a CF Trust representative assesses the effectiveness and resourcing of each centre or clinic and prepares a report for the

hospital team, managers and commissioners. Since implementing the process of peer review in 2006, this has led to an increase in NHS funding of over £17 million dedicated to CF care.

At the request of the Department of Health, the CF Trust is also developing a new system for funding CF care via an annual banded tariff. This will allow appropriate funding to follow the patient to where they choose to receive their care as long as it is a designated CF Centre or clinic.

The CF Trust also makes start up grants to fund clinical posts in CF care and funds a training scheme for CF Consultants of the future.

The CF Trust highlights inequalities in the provision of CF care and particular treatments to commissioning bodies and lobbies for access to all recommended CF drugs where appropriate.

### **Why are only 94% of patients genotyped?**

There are some people for whom a specific genotype has not been conclusively determined; this may relate to older people whose genetic analysis was undertaken some time ago when certain mutations were classified as 'unknown'. There also continue to be new and emerging mutations which take some time to be fully classified.

### **Why do there appear to be more males than females over the age of 16 years?**

From the summary statistics you will note that 53% of patients registered on the database are males, therefore there are more males than females to begin with. Research has and continues to be undertaken to determine whether sex is a determinant of survival but currently there is no conclusive data available to prove or refute this.

### **Why has it taken so long to get this data?**

The UK CF Registry has only been in operation in its current form since 2007 and producing the reports each year has taken a considerable amount of time. We are constantly refining and improving the process and expect the 2010 report to be available later this year.

### **Who can I talk to if I have further questions?**

If you have any questions about the information in the registry you can email the CF Trust at [helpdesk@portcf.org.uk](mailto:helpdesk@portcf.org.uk) or call the helpline on 0300 373 1000. To discuss how the findings relate to you or your child or to discuss the results achieved by your Centre or clinic we suggest you talk to your clinical team who will be able to help you.



11 London Road, Bromley, Kent BR1 1BY

Tel: 020 8464 7211 • Fax: 020 8313 0472 • [www.cftrust.org.uk](http://www.cftrust.org.uk)

Patron: HRH Princess Alexandra, the Hon. Lady Ogilvy, KG, GCMG President: Duncan Bluck CBE

Cystic Fibrosis Trust registered as a charity in England and Wales (1079049) and in Scotland (SC40196)

A company limited by guarantee registered in England and Wales number 3880213

Registered office: 11 London Road, Bromley, Kent BR1 1BY