The Cystic Fibrosis Registry of Ireland
Annual Report 2003

Contents

Executive Summary

The field of cystic fibrosis is a complex and intricate one. All of the professionals who work in this field realise that they are dependent on each other in order to be effective. They are also very aware that each and every person with cystic fibrosis is an individual who travels a different route. But it is widely agreed that there are fundamental similarities as well as differences. It follows then, that if we knew more about those similarities, better treatment interventions could be detected. These professionals carry an inherent belief that if **all** that we know about cystic fibrosis was before us in an orderly display, we could do a better job.

We began this project in the summer of 2001, with a rigorous set of aims and objectives in place. It was then necessary to create a useful and relevant management system. It is important for different healthcare professionals to be able to view the results in this system, in order to apply the findings to improve the outcomes of **P**ersons **W**ith **C**ystic **F**ibrosis [PWCF]. This type of system utilises knowledge, measurement, comparison, and analysis. The results should be available to a wide audience.

At the close of 2003, we can confidently say that we have a state-of-the-art system which documents the facts and data from patients charts. This system also yields summaries of those facts and data. We can put many queries to this system to ask it to align and re-align the information in a variety of ways. In other words, we can analyse the past, which is a major first step to knowledgeable change.

And so, we move into the realm of "knowledge management." There are three stages of knowledge management: 1) collecting facts and data; 2) relating those facts to produce information; and 3) 'mining' that information for reliable knowledge.

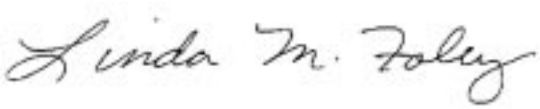
The Irish CF Registry now has the functionality for completing stages one and two of the knowledge management process. It is the responsibility of the Irish CF Registry to keep the database up to date, as complete as possible, and to ensure the data is valid. Examination or 'mining' of the information to produce valuable findings will be done by research people. They will be able to explore many, many aspects of cystic fibrosis through this approach. Conclusions drawn from these exercises will only be reliable if the original data is accurate.

This report describes the activities that took place in 2003. All of the work undertaken to date is bearing fruit. Individual reports of diagnosis, complications, and treatment are now available. Summaries of patient data for individual CF consultants can be printed out. These reports combine information from numerous places in a patient's chart into one easy-to-read format. This information can be sifted and sorted in a variety of ways to shed even more light on what has taken place.

These major steps however are limited to the number of PWCF who are enrolled on the Registry. By the end of 2003, over 20% of PWCF in the Irish population will be enrolled. The next step is to enroll as many more as possible in the coming year. To date, the limiting factor has been time. If more people are involved in compiling the data, the time to acquisition of a complete database becomes shorter and the tool becomes more effective.

Greater progress could be made at a faster pace if there was a permanent endorsement of this registry. It would imbue confidence in those who work tirelessly toward better outcomes.

There is no doubt that there is cohesion and commitment among the professionals, parents, and patients in the field of cystic fibrosis. I hope that this report complements their dedication and enhances the treatments for individuals to ensure longer survival and a better quality of life.



Linda M Foley, B.Sc.
Director, Cystic Fibrosis Registry of Ireland.

Mission Statement & Ethos

Mission Statement

"The national Cystic Fibrosis Registry of Ireland endeavour to collect and analyse information relating to cystic fibrosis in order to improve the quality of care for all of the people with cystic fibrosis in the Republic of Ireland."

Ethos of the Registry

"The Registry must be independent of institutions."

- The Registry shall stand on its own, as a satellite to the hospitals, patients and associations from which it receives its information.
- The Registry Management Committee is committed to up-holding this ethos in carrying out its duties.

Description of the main activities carried out by the Registry in the past twelve months forms the basis of this report. Please note that the abbreviation, **PWCF**, is an internationally accepted term standing for **P**ersons **W**ith **C**ystic **F**ibrosis.

Registry Management Committee

The Registry Management Committee has undergone minor changes during 2003, and these changes are indicated in italics.

Prof. G McElvaney	Professor of Medicine and Consultant Respiratory Physician, RCSI, Beaumont Hospital
Mrs Linda Foley	Director, CF Registry
Dr. C. Gallagher	Consultant Respiratory Physician, St. Vincent's University Hospital
Dr. P. Greally	Consultant Paediatrician, National Children's Hospital, AMNCH, Tallaght
Dr. G. Canny	Consultant Paediatrician, Our Lady's Hospital for Sick Children, Crumlin
Dr. J Gleeson [*replaces Dr B McDonagh*]	Consultant Paediatrician, Sligo General Hospital
Ms. C. O'Connor	CF Specialist Nurse, Beaumont Hospital
Ms. G. Leen	CF Specialist Nurse, National Children's Hospital, AMNCH, Tallaght
Mr. Martin Wickham	Esat and member CFAI
Mr. Carl Rainey	*Chairperson,* CFAI [*formerly Vice-Chairperson, CFAI*]
To be appointed	*CEO,* CFAI

Objectives & Aims

The specific objectives and aims of the Registry were published in the first Annual Report, December 2002. These are available on the Registry website: www.cfairegistry.org

The detailed objectives may be viewed in a broader sense as follows:

1. to provide a reporting system for those who care for PWCF to better view/visualise their patient records;

2. to provide a data repository of all PWCF; and to use that data for detailed analysis of many clinical aspects.

During the past twelve months we have been working to achieve both of these objectives. It is important to deliver a system that has relevance to daily clinic activities (Objective 1); and at the same time to create a databank for research (Objective 2). The style of the reporting system is dealt with in more detail in the "Report Generation" section of this publication.

A process for researchers has been completed. This is to be used by anyone who wishes to extract anonymous data from the Registry. Data Request Forms (from the Registry) have been created. A Data Request Form is then submitted to the Registry Management Committee by the researcher. Once the Registry Management Committee approves of the release of data (in writing), it becomes part of a permanent record held by the Registry.

Registry Website: www.cfairegistry.org

Ireland was the first country in the world to launch its Cystic Fibrosis Registry from a web platform (July '02). Since then, in April, 2003 the USA has moved to data entry from all of its centres via the internet.

Many countries are now considering this format for its convenience and 'on-line' appeal. As soon as data is entered, it becomes 'live' and reports can be printed immediately. Also, in the future, there is the possibility of pooling anonymous data from different countries which would yield larger segments for analysis. This gives strength and validity to conclusions drawn from the data.

The public pages of the Registry Website [www.cfairegistry.org] were launched during 2003. These pages contain the following information:

- Background information about cystic fibrosis
- Full explanation of the Irish CF Registry
- List of all CF hospitals/clinics/consultants
- Useful CF links to other CF Registries and Associations
- Information Booklet and Annual Report downloads available
- Frequently Asked Questions
- Privacy Policy

The rest of the website is restricted to consultants who have patients entered on the Registry. They will have their own usernames and passwords. They will be able to view their own individual patient summaries as well as summaries of **all** their patient data. Training and testing of these functions will take place in 2004.

The CF Registry has the ability to audit all interactions with the database whether they concern new data entry, changes to existing data or report downloads. An auditing report will become part of the annual submission when other users begin to use the database.

Other activities involving the internet that took place during 2003 were:

- Domain name registration for www.cfairegistry.org and Security Certificate renewal.
- Data Protection certification annual renewal granted acknowledging the Director of the CF Registry as a Data Controller

Census 2003

Census of PWCF has been compiled. This is done through the CF Specialist Nurses who return the number of patients attending their clinics and hospitals. Thus, this distribution is based on hospital attendance rather than a person's place of residence.

There is an inherent problem however, in that many PWCF attend more than one hospital. They may attend a tertiary centre for the purpose of Annual Assessment and they may also attend another hospital in their own region for antibiotic treatment for severe infections. Attempts have been made to resolve the possibility of "double-counting", but the accuracy of the census will not be validated until the vast majority of patients are entered onto the CF Registry. Only then will we be able to perform audits and cross-checks of numbers of PWCF against the hospital/ consultant lists.

However, the following graphs give us an overall estimate of the number of PWCF in the country. The pie chart shows this census grouped by hospital within a health board.

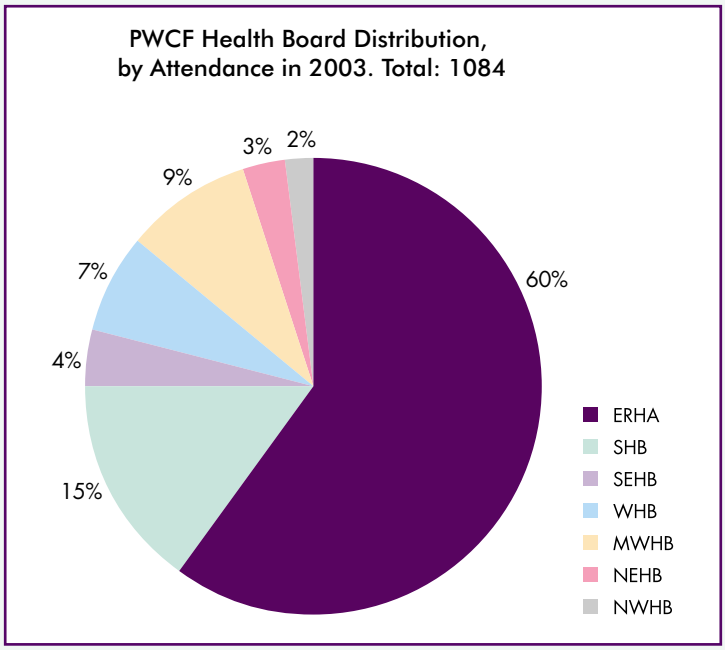
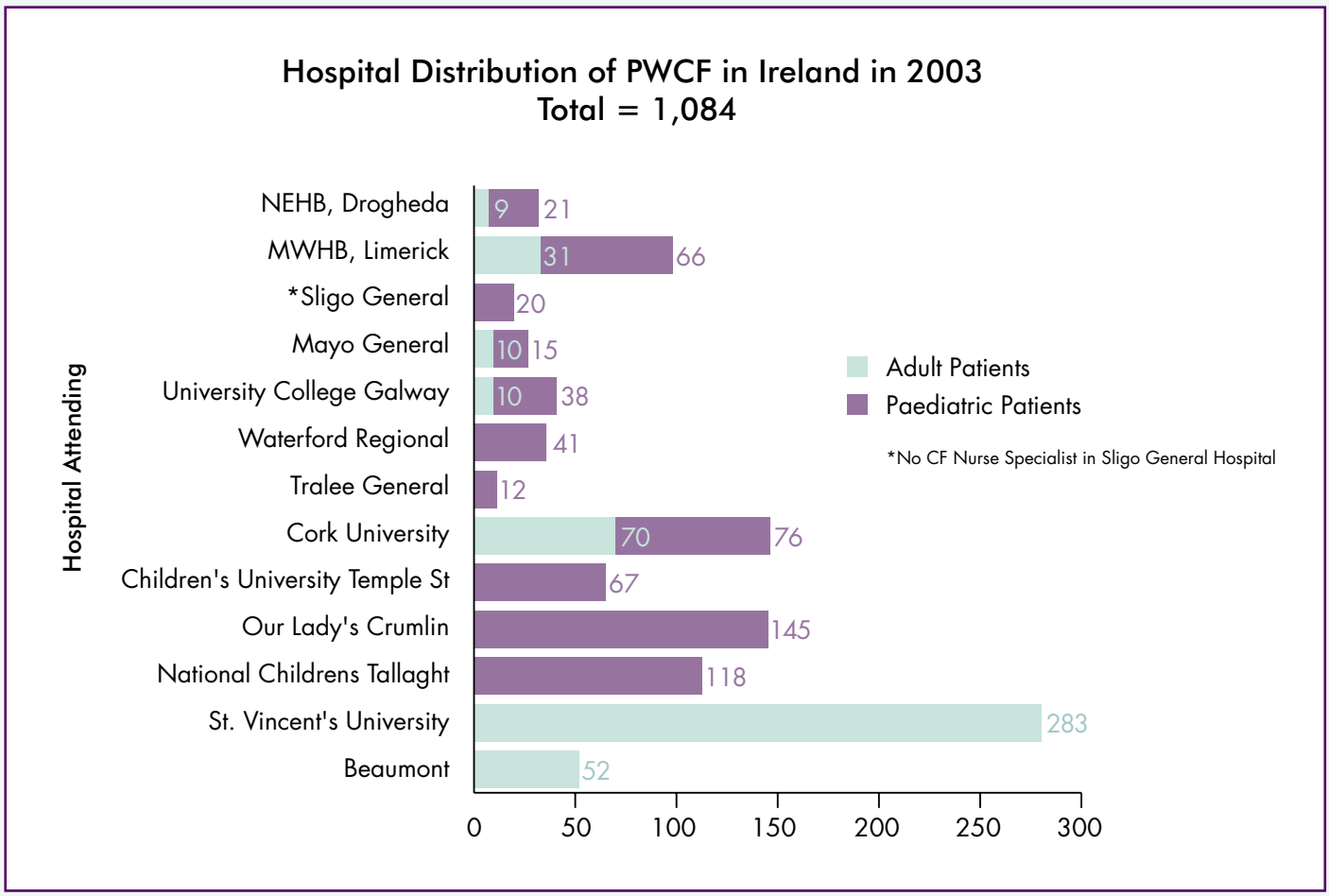


Figure A: (Above) Health Board Distribution determined by hospital attendance.

Figure B: (Below) Hospital and Age Distribution of PWCF in Ireland.
N.B: An Adult is defined as a person who has achieved their 18th birthday.



The most important aspect of the expected changes over time is the increased proportion of Adult PWCF. This will approach 50% by 2008, as is already the case in many other countries.

Based on the 2003 census, projections have been made for future estimates of the PWCF population. Projections of the CF population will be useful in resource planning.

The following assumptions have been made regarding PWCF population projections:

1. The Paediatric portion will stay relatively static as it will lose approximately the same number of PWCF to the Adult group as it assumes from diagnosis at birth protocol (newborn screening).

2. The Adult portion of the population will grow and assume a larger proportion of the whole.

Figure C: (Below) Projected CF Population increase to 2008, showing growth in Adult portion as well as overall growth

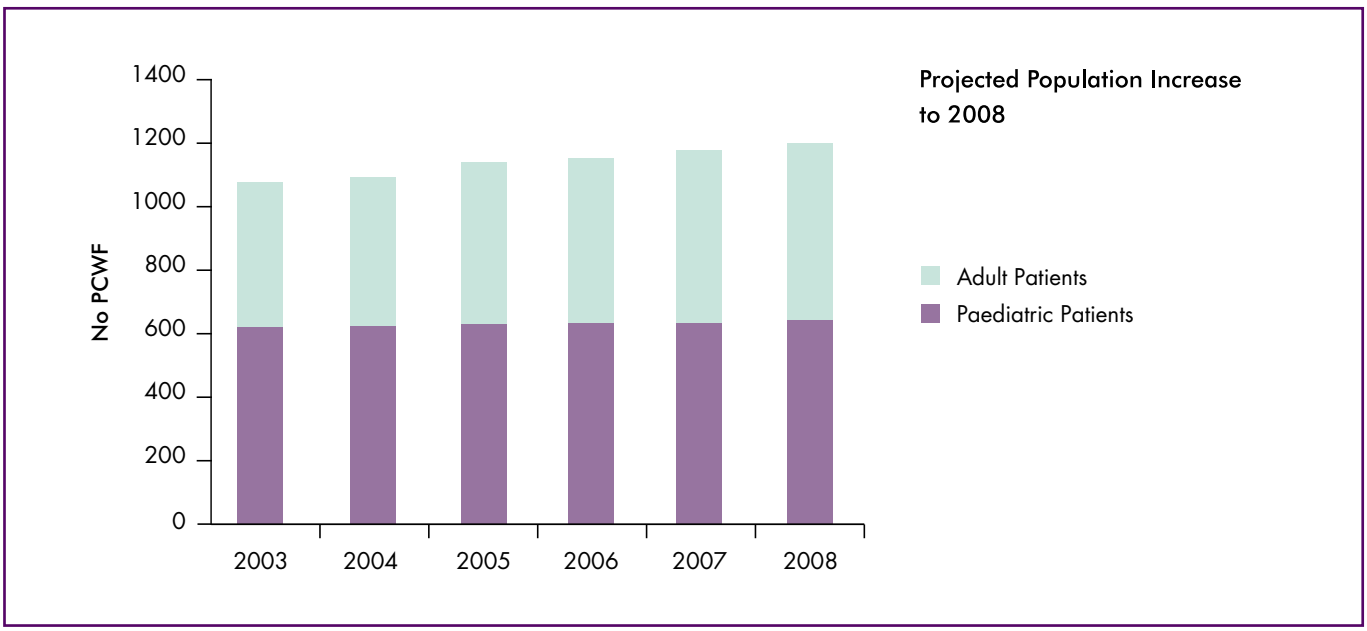
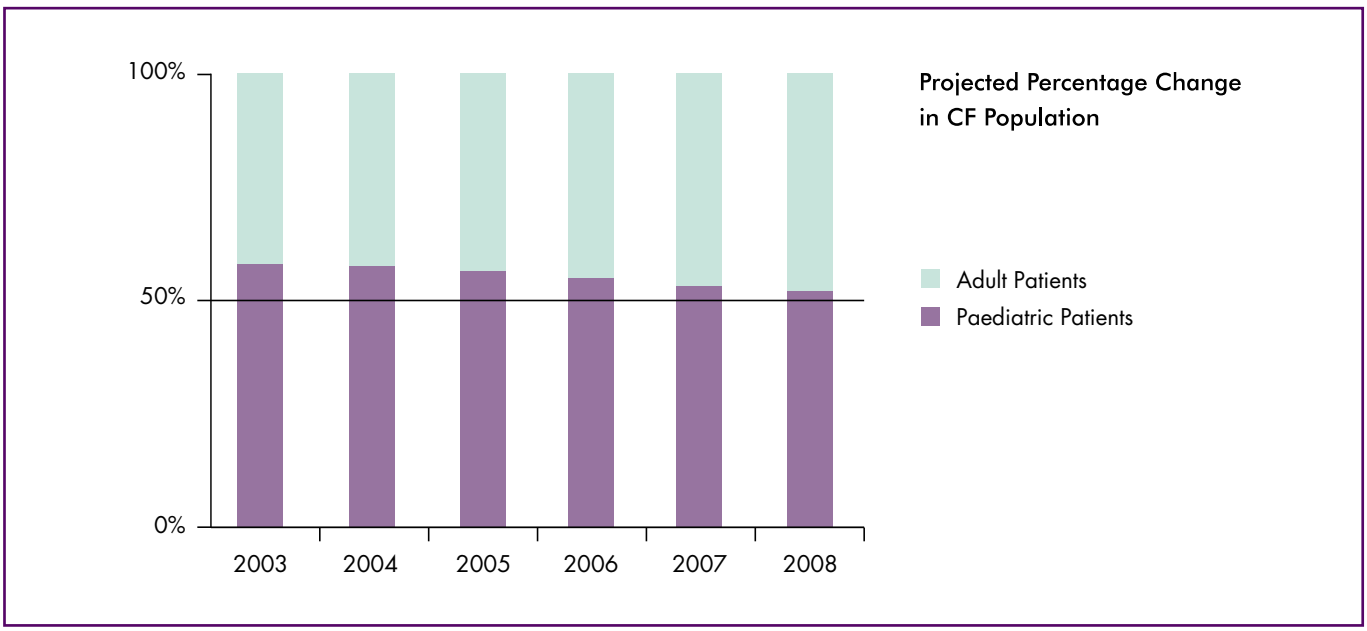


Figure D: (Below) Projected change in CF Population to 2008, showing estimated shift in population: the Adult portion will assume a greater portion of the whole.



Collaborations

Collaborative research in cystic fibrosis across countries will bring new data to light in a more expeditious manner than if small groups of doctors work within the boundaries of their own countries. It is for this reason that discussions regarding the best methods for pooling anonymous data have begun.

Combining information from separate databases is only possible if common standards are adopted. It is important to create standards for registries over the next number of years. Much of this work involves how data is collected and stored electronically. Collaboration among many countries will strengthen the development of standards and data collection practices.

International Collaboration

Contact has been established with the following registries: United Kingdom, Germany, Denmark, Canada, Australia and the USA. These groups have shared their experiences, reports and information; and this input has been used to build the CF Registry of Ireland.

There is now a registry group that meets at the European CF Society annual meetings, and this provides a forum for sharing best practice. New countries who are about to join the European Union are looking for guidance in the setting up of their own domestic registries. Countries with older databases are investigating new technologies. All countries are interested in comparing their experiences with others and are keen to discuss best practice.

University of Wisconsin

The Dean of the UW Medical School and Vice Chancellor for Medical Affairs, Professor P M Farrell, MD, PhD, is a world expert in newborn screening and long-time advocate of screening for cystic fibrosis. Some of his group's research published in 2001 reported on a 13-year study which showed that early diagnosis of PWCF screened at birth was associated with significantly greater growth throughout childhood.

More recent research from Prof Farrell's group has concentrated on the analysis of costs of a newborn screening programme versus traditional sweat testing. This evaluation considers the benefits of diagnosing PWCF at birth versus later clinical diagnosis when more persistent signs and symptoms arise. By that time, several things may have occurred, among them preventable malnutrition and persistent chest infections. It is generally agreed nowadays that newborn screening (1-5 days after birth) will not only help to prevent this malnutrition, but will also inform the paediatrician who can begin prevention strategies to counter the persistent chest infections.

A pilot study was carried out during June/July 2003 which gathered cost data for sweat tests from the Irish hospitals. This information may later be used in a larger research project to compare costs of screening against the overall costs of late diagnosis. This work, while not directly involving the Registry in 2003, may develop further and the researchers may request information from the Registry about patients with late diagnosis.

The CF Registry is keen to remain closely aligned with this research and since Ireland is noted to have the highest prevalence of the cystic fibrosis genes in the world, it is an ideal place to carry out this research. All information released to researchers will be anonymous and will have prior approval of the Registry Management Committee.

Newborn Screening for cystic fibrosis

As a Newborn Screening Programme is likely to be initiated in Ireland in the near future, it is important that newly diagnosed PWCF enter the Registry as soon as possible after birth. This affords many advantages such as prevention of PWCF being lost to follow-up and active management of cystic fibrosis from birth.

It will be necessary to devise a procedure for CF Registry registration at diagnosis and this will improve CF census data. The paediatricians will find value in tracking their new patients which will be facilitated by the CF Registry.

This project will be part of the programme for the CF Registry in 2004.

References

Farrell PM, Kosorok MR, Rock MJ, Laxova A, Zeng L, Lai HC, Hoffman G, Laessig RH, Splaingard ML, *Pediatrics* (2001)Jan; 107(1): 1-13

Lee DS, Rosenberg MA, Peterson A, Makholm L, Hoffman G, Laessig RH, Farrell PM, *J Pediatr* (2003)Jun; 142(6): 617-23.

Report Generation

Since cystic fibrosis affects a large number of vital systems, recurrent hospitalisations are often a feature of the condition. Hospital charts grow in thickness and the number of volumes increases over time. It is not unusual for even a young patient to have several chart volumes in the same hospital. Earlier information may not be readily available during an out-patient visit or an admission.

Also, this information does not pass in total to another hospital when the PWCF progresses from a separate paediatric CF centre to an adult CF centre.

Through the CF Registry, a printout of all Annual Assessments (as well as diagnostic information) is available to the patient's doctor no matter where he/she sees the patient, provided there is internet access. These hold all pertinent clinical information in a chronological fashion. The paediatric centre may keep a set of Annual Assessments and the adult centre may print out duplicates of all previous information for that person. This will be far more informative for the CF Adult consultant who might not have access to the older files which remain in the paediatric hospital.

All reports are 'on-line' and 'live'. That is, they include all previous data in the databank. Reports may be used to longitudinally track progress of each patient.

There are both tables and graphs (e.g., all cultures in the year; no. of days on each antibiotic, etc.) as part of each report. Later, longitudinal summary data over several years will be available. These will be summarised in a cumulative fashion so that each year another set of data is added and the report will demonstrate a picture of progress from the time the patient first entered the Registry. Individual patient reports will be updated (i.e., printed) as each unit sees fit. Each unit will have the ability to download their own patient reports on site. Examples of these reports can be found in the Appendix.

Each consultant will also have several summaries of his own set of patients. These will take the form of summary tables and graphs which also provide links to the specific data or patient from which they are derived.

Regarding long term research, the data will reside in the Registry and will be available for use in research projects. There will be data for use in epidemiology, microbiology, dietetics, physiotherapy, respiratory medicine, genetics, CF specialist nursing and health economics / resource studies. Several Global Data reports encompassing the entire database population will be available in 2004.

To date, we have created nine Individual Patient Reports and seventeen Consultant Reports. Each report has multi-links to original data or graphic representations. These are all available on-line, on-demand. In addition to these there are also eight Director Reports on-line.

Data Entry

Data entry is the time limiting factor in the achievement of a complete database. Initially it involves three steps: Registration, Diagnosis and Annual Assessment.

Recently a Clinical Research Associate has been employed to assist in data entry and this task will now accelerate.

In some hospitals this task is also being completed by CF Specialist Nurses. Time resource prevents them from completing this at a faster rate, but the benefits become apparent immediately. Because the Registry has the functionality to produce comprehensive summaries of patient care over a 12 month period, these can be seen and printed as soon as data entry is finished.

Other Activities

Other activities that have been completed during 2003:

- All Hospital Ethics Committee approvals from CF Centres have been granted.
- Training of all CF nursing personnel for administration of Informed Consent completed.
- The Committee has finalised the fields that will be used in the database.

  The fields that are currently listed in the Annual Assessment may be expanded in time. There is no restriction to this and new fields will be added as the Registry Management Committee sees fit.

- Two additional in-house databases have been created:

  Consent Form database:

  This is used to track consent forms and compare with the census from a particular hospital. This database will probably become redundant after a number of years as the registry itself will become the source for the census figures.

  National CF Medical staff database used for contact purposes:

  Data input from several disciplines is required to complete the annual assessment. Among them are nursing, dietetics, physiotherapy, microbiology, and transplant services. This database is updated at the time of the census each year.

Continuity & Plans: The Future

The Department of Health & Children has published a "National Health Information Strategy". It contains many clear objectives to "…promote, protect, restore and maintain the health of individuals and the population." The Cystic fibrosis Registry of Ireland has closely aligned it's objectives to those of the health information strategy.

The healthcare sector is moving rapidly into the field of "knowledge management". This may be described as the use of computer technology to organise facts and data, bring together all sources of information into one system; and then interrogate it for relationships that superficially may not seem to exist. This knowledge management technique is being used to bring down costs in healthcare and increase efficiency in health systems. The same technology can be applied to all health conditions. In this case, efficiencies naturally result because more orderly information is at the disposal of the doctor when he/she makes a crucial treatment decision.

No analysis is complete without a detailed and exhaustive look at the past. Technology allows us to do this task almost instantly. One might be able to see why one group of patients responded to a treatment while another group did not. An understanding of clinical history will assist us to predict future response to a treatment. Now, if one creates a computer system that organises this clinical history, then one can look at endless comparisons which will help us to answer the question of what are the best treatments for the patient.

With a system like the CF Registry of Ireland, we can simultaneously look at history, while building and testing hypotheses for improved care. As the bank of historical data grows, so too does the power of observation of any present population. The testing of a change in therapy can take place within a shorter time span.

This is why, now that the system has been assembled, it is so important to keep it continuing. Within only a few months, tangible products are already issuing from the registry.

It is difficult to create plans for the future without continuous funding. A permanent commitment has been sought from the Department of Health & Children.

In order to speed up enrolment and add subsequent annual assessments, further staff is required. This can be satisfied by an addition of one or two data entry persons.

Further interrogation of the database should be the territory of full-time researchers. This can be fulfilled by doctoral or post-doctoral candidates (at universities and medical schools) who are self-financing from academic grants.

A recent publication (2002) on Disease Registers from the UK recommended that the physical location for a research register should be in an academic environment, with statistical and epidemiological expertise available, where all potential users feel they have equal access and where those with legitimate scientific interests feel on an equal footing. To fulfil these objectives the present location of the CF Registry of Ireland would need to change.

If the Department of Health & Children sees fit to offer future funding to the Registry it is hoped that the following will be considered:

1. Permanent commitment to the Registry;

2. Support for an increase in staff when needed;

3. Support for a change in location.

Financial Summary

The following financial summary of the accounts illustrates the expenditure for the first 18 months of operation of the CF Registry of Ireland, excluding salaries. The accounts were audited by Ryan Glennon & Co, Trinity House, Dublin 6.

Financial Summary: *18 Months to end of December 2002*	
INITIAL SYSTEM SPECIFICATION Domain registration, Set-up fee, Six months hosting fee, Security Certificate	€11,554
DEVELOPMENT COSTS Database Application, Installation & Training	€12,357
SET-UP HARDWARE Lap-top computer, Printer, Zip drive, Port Replicator, Monitor, etc.	€6,991
TRAVELLING EXPENSES	€3,460
ADMINISTRATION COSTS Telephone, Heat, Electricity, Printing, Office Supplies, Insurance	€7,943
TOTAL	**€42,305**


Acknowledgements

The Cystic Fibrosis Registry of Ireland is grateful to the Minister of Health & Children, Mr Micheál Martin. His staff have all been generous and constructive with advice. We appreciate their foresight and support of this project from the outset.

The Cystic Fibrosis Association of Ireland and its Chairperson, Mr Carl Rainey, have also been sterling in support. They conceived and encouraged this Registry and I hope that it lives up to their expectations.

The Registry Management Committee under Chairman Professor N. G. McElvaney have voluntarily devoted time and energy to the Registry by attending meetings and reviewing activities. Their strategic thinking ensures that the Registry operates at the highest possible level.

As always the parents and adult PWCF have contributed to the Registry and have been very positive. They should be commended for their strength and compassion under stressful circumstances.

Mr Rahul Rana, of ecom-ireland, continues to lend his most creative talents to the Registry. Not only is he always available, he also consistently delivers undertakings to agreed timelines.

The work that we have done to date would not be where it is without the universal support of the CF Nurse Specialists. They put in remarkable efforts and yet manage to incorporate Registry responsibilities into their daily workload. The CF consultants, physiotherapists and dieticians also work under challenging conditions and all of them have welcomed the Registry by contributing their ideas and encouragement to it.

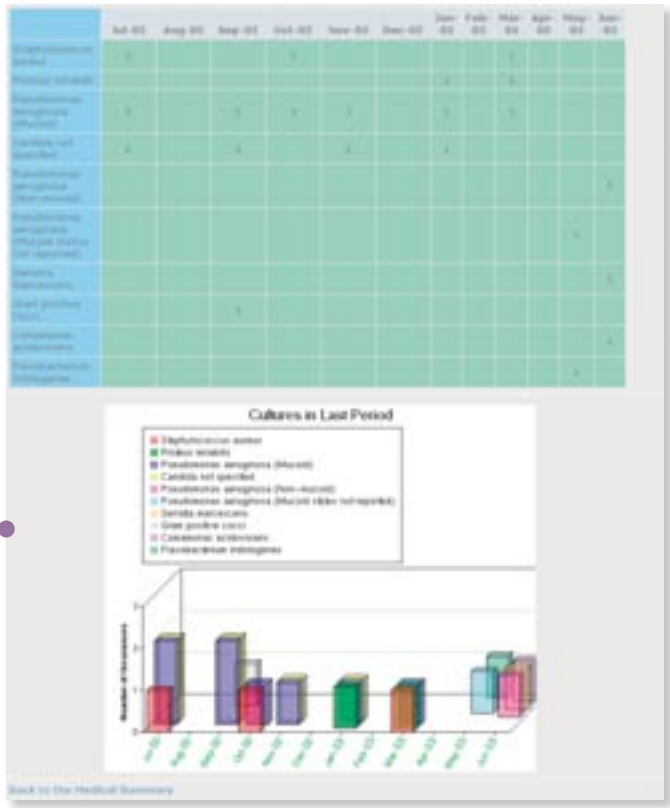
There were many CF registers in operation before we began. Certain individuals have encouraged us, in particular, Dr Anil Mehta, Director of the UK CF Database at the University of Dundee. Dedicated people in many different countries have also conveyed their experiences: Australia, Canada, Germany, Denmark, Israel, and the USA.

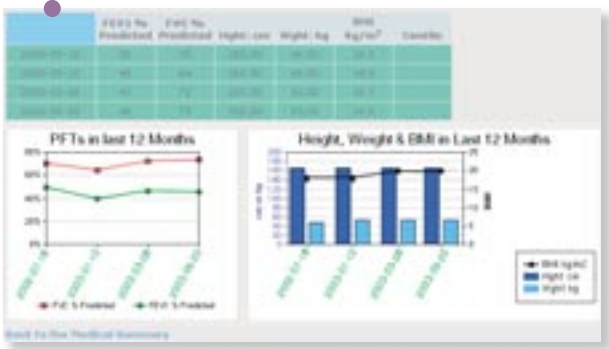
Finally, Professor Farrell at the University of Wisconsin has given his time and advice to this project and should be recognised for his expert contribution. In addition, his staff have shared their knowledge and experience and continue to do so.

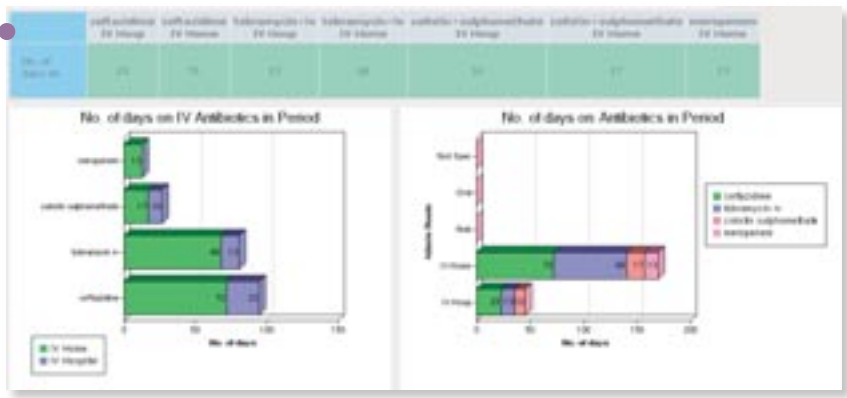
Appendix

Selected Illustrations of Individual Patient Reports

*All reports and links are available for each patient enrolled.







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Cystic Fibrosis is an inherited condition that affects many body functions such as breathing, digestion, and reproduction. This lifelong condition usually becomes more severe with age and can affect both males and females. The symptoms and severity of cystic fibrosis differ from person to person. The majority of people have both respiratory and digestive problems, while some may have only respiratory infections. Common symptoms include chronic cough, wheezing, sinus problems, recurrent pneumonia, poor growth, and skin that is salty to the taste. New discoveries have led to better treatments for cystic fibrosis in recent years. Twenty years ago life expectancy was low, but that trend is now reversed. More and more individuals lead full adult lives. Better treatment strategies help to improve the length and quality of life of people with CF by controlling their symptoms.

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