The Cystic Fibrosis Registry of Ireland
Annual Report 2002

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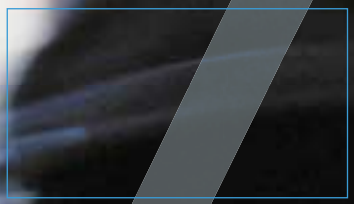

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Executive Summary

Cystic Fibrosis is an inherited condition which requires a multi-disciplinary medical approach to maximise the length and quality of life of those affected. Approximately 30-40 children with Cystic Fibrosis are born in Ireland every year. Because the genetic mutation which leads to Cystic Fibrosis is present in one person in 20, it is a very common and widespread mutation. Thus, children may be found living in any part of the country. At present, there are approximately 1,100 people with Cystic Fibrosis living in the Republic of Ireland.

Care for people with Cystic Fibrosis has progressed remarkably over the last 30 years. Most of those with Cystic Fibrosis will live into adolescence and increasingly into their fourth and fifth decades. This improvement in longevity is due to many factors, but most significantly it is because specialist care centres have evolved (primarily in medical school training hospitals). A multi-disciplinary approach has been adopted at these specialist centres and children's lives have improved and lengthened as a result of this development.

Another factor which has contributed to the overall betterment of people with Cystic Fibrosis is the creation of Registries. These databanks gather information from individual patients over wide geographical regions. Registries provide a pool of authentic data that can be used for research purposes. This has greatly facilitated researchers who are interested in comparing treatment strategies from different centres. In other countries, standard medical practice improvement has raised the overall quality of life of people with Cystic Fibrosis. This has been achieved through the evolution of treatment guidelines that have been made possible through the existence of Registries.

Registries, unlike specific clinical trials, not only hold data on overall morbidity and mortality, but also have the versatility to display one individual's data over time. In that way they can be used in any number of research settings, while also assisting the single consultant with reports of individual patients.

In 1999 the Cystic Fibrosis Association of Ireland facilitated meetings with a number of consultants who primarily treat people with Cystic Fibrosis. This group formulated a proposal to build a national Cystic Fibrosis Registry in Ireland.

Late in 2000, the Minister of Health and Children approved funding for a pilot project for a National Registry. Ireland was the first European country to be awarded government funding for such a proposal, a significant foresight. Ireland is also the first country to use the most up to date technology available on the internet to house such a registry. This technology will make the Irish Cystic Fibrosis Registry a dynamic and timely data resource.

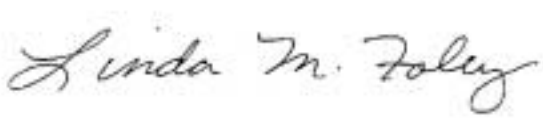
During the initial year the Registry has employed one person to formulate and implement the Registry. This report focuses on the activities and accomplishments of the first year of operation from June 2001 to June 2002.

The database was finalised in June 2002 and the first patient entered into the system in July 2002, thirteen months after work began on the Registry.

Plans are in place to complete the registration of patients over an eighteen month period. Thereafter each enrolee will have annual assessment information added to their data.

There are a great many people who have contributed their time and ideas to this ambitious project and their participation is appreciated and acknowledged at the end of this report.

This report will be available on the registry website, www.cfairegistry.org in pdf format shortly after publication.



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

Mission Statement & Ethos

"The national Cystic Fibrosis Registry of Ireland will 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."

This statement, taken from the original funding proposal submitted to the Department of Health and Children, underpins the ethos of the Registry.

Other words which describe the concept of the Registry are:

- Guaranteed confidentiality
- Responsibility
- Procedures that protect patients
- Guaranteed security

The Registry shall stand on its own, as a satellite to the hospitals, patients and associations from which it receives its information.

The Registry belongs to its constituent members: a type of collective ownership. Each enrolee contributes his/her own information and can control whether that is entered into the database through signing a consent form.

Each patient may have access to their own Registry records. The Data Protection Act 1988 guarantees individuals the right to a printed copy of their information if it is kept as a computerised record. Anyone who is enrolled in the Cystic Fibrosis Registry may apply to the Registry Director for a copy of their own information.

Further information on data protection can be obtained from:

Data Protection Commission
Block 4
Irish Life Centre
Talbot Street
Dublin 1
Telephone: 01 874 8544
Website: www.dataprivacy.ie


Objectives & Aims of the Registry

Objectives:

1. To identify, collect, classify, record, store and analyse information relating to the prevalence, incidence and treatment of Cystic Fibrosis in the Republic of Ireland.

2. To collect, classify, record and store information in relation to existing and newly diagnosed Cystic Fibrosis patients.

3. To promote and facilitate the use of the data thus collected in approved research projects and in the planning and management of services.

4. To furnish advice, information and assistance in relation to any aspect of Cystic Fibrosis to the various Health Boards and other service providers and persons with Cystic Fibrosis.

5. To provide data on long term prognosis for Cystic Fibrosis patients in the Republic of Ireland – including treatment outcomes, management regimes, quality of care, international best practice, etc.

6. To provide data on long term prognosis for Cystic Fibrosis patients in the Republic of Ireland and to compare this same information with international data.

7. To publish an annual report based on the activities of the Registry.

Aims:

The Registry will be used to update a website. The Registry will be dynamic rather than static.

1. To register all Cystic Fibrosis patients whose usual residence is in the Republic of Ireland and to record genotypes (genetic information) of all Cystic Fibrosis patients treated in the Republic of Ireland.

2. To ensure that this registration is complete, accurate, timely and confidential; to effectively use the data collected.

3. To provide annual reports on the incidence, prevalence and treatment of patients registered, at a sufficient level of morbidity and geographical detail to make them available to those involved in planning and delivering Cystic Fibrosis diagnosis and treatment services, and to disseminate these to all interested parties.

4. To provide a Cystic Fibrosis information service for the Department of Health & Children, Health Boards, Hospitals and Clinicians.

5. To initiate research into the causes, distribution, treatment and outcome of Cystic Fibrosis, to participate in similar research initiated by others and to publish the findings.

6. To provide individual consultants with trends and updated information in respect of their patient population.

7. To assist in the evaluation of novel treatments and screening programmes.

8. To develop and improve Cystic Fibrosis registration methodology.

9. To compare and contrast Cystic Fibrosis management and treatment with best practice internationally.

Description of the Registry

The Registry is located in offices at:

CF House
24 Lower Rathmines Road
Dublin 6

A Director was appointed by the Cystic Fibrosis Association of Ireland in June 2001, to oversee the setting up and implementation of the Registry. It is the responsibility of the Director to include all possible sources of information for entry into the database. These sources of information include hospital patient charts, doctors, nurses, physiotherapists, dieticians and patients themselves.

Information will be tabulated on a health board/regional basis in order to support planning for local and regional services for the betterment of Cystic Fibrosis patients country-wide.

The Registry will be accessible to permitted users only through the internet. Information that is stored in the database will be taken from hospital medical charts.

Every hospital that will contribute patient data to the Registry must approve of the Registry through its Ethics Committee. This process must be complied with for each hospital and, although the process is similar for all hospitals, each Ethics Committee must decide individually whether it will release hospital chart information to the Registry.

After Ethics Committee approval and the consent form is signed by (or on behalf of) the patient, the Director is notified and arranges to visit the hospital with a lap-top computer. All of the relevant data is transcribed from the medical chart onto the Registry database. Copies of the signed consent form will be kept in the patient's chart and at the Registry office.

Participation in the Registry will be treated as confidential and disclosure of any personal records or results relating to the Registry will be limited to the patient's consultant doctor and/or the Registry Director.

At some point in the future it is hoped that anonymised data will be linked to other national Cystic Fibrosis Registries in Europe and the USA. This will greatly enhance our general knowledge of Cystic Fibrosis and its treatment.

Implementation Activities

The working group for implementation is the Registry Management Committee. It is made up of the Director of the Registry, Cystic Fibrosis Specialist consultants, Cystic Fibrosis Specialist nurses, and three members of the Cystic Fibrosis Association of Ireland.

Professor G McElvaney
Chairman, Registry Management Committee; Professor of Medicine and Consultant Respiratory Physician, RCSI, Beaumont Hospital

Mrs Linda Foley
Director, Cystic Fibrosis Registry

Dr. C. Gallagher
Consultant Respiratory Physician, St. Vincent's University Hospital

Dr. P. Greally
Consultant Paediatrician, National Children's Hospital, Tallaght

Dr. G. Canny
Consultant Paediatrician, Our Lady's Hospital for Sick Children, Crumlin

Dr. B. McDonagh
Consultant Paediatrician, Sligo General Hospital

Ms. C. O'Connor
Cystic Fibrosis Specialist Nurse, Beaumont Hospital

Ms. G. Leen
Cystic Fibrosis Specialist Nurse, National Children's Hospital, Tallaght

Mr. Martin Wickham
Esat and member Cystic Fibrosis Association of Ireland

Mr. Carl Rainey
Chairperson, Cystic Fibrosis Association of Ireland

Ms. Ciairín de Buis
Director, Cystic Fibrosis Association of Ireland

Mr. Paul Allen
KPMG Consulting, ad hoc member for duration of set-up period

The Registry Management Committee met on a number of occasions throughout 2001/02 to agree on the following:

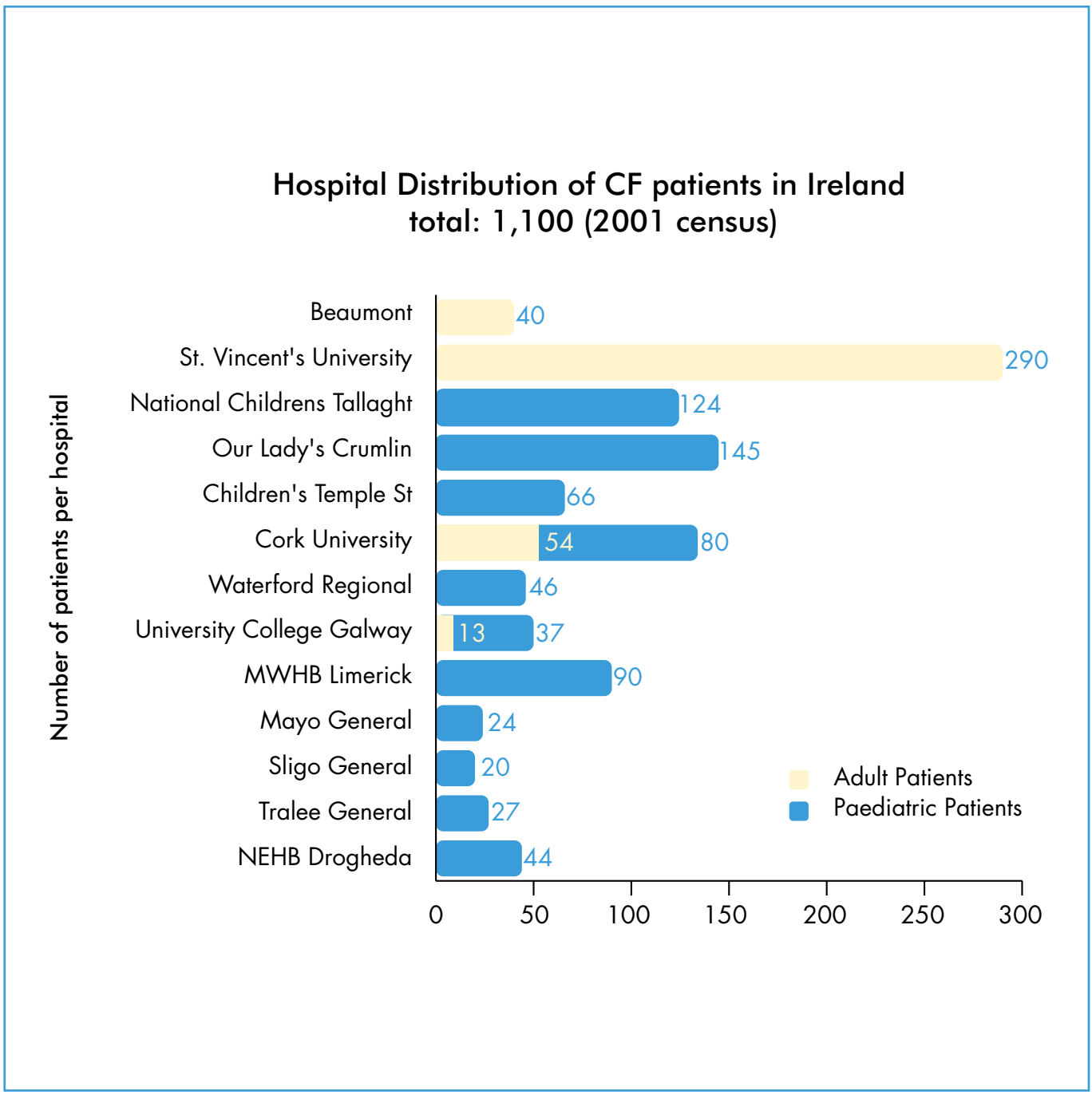
a) the fields that would be included in the database;
b) the process for data collection;
c) the internet service provider;
d) the confidentiality and security procedures.

Key Implementation Tasks

- Functional specification of the database. Working group led by Paul Allen, KPMG Consulting.
- Agreement of the fields to include in the Irish database; research into other national registries.
- Data Protection Commission certification.
- Research on bespoke databases; Internet service providers; quotes.
- Registry Information Booklet: written primarily for patients who are potential enrolees; but also for anyone who is interested in the concept of registries and particularly in those which may be housed on the internet.
- Ethics Committee approvals from ten hospitals granted. This covers 85% of Cystic Fibrosis patients in the country. The other 15% of patients attend five further hospitals. Three of these submissions have been made; pending approval in those hospitals, 97% of the population will be covered by ethical approval.
- Survey of the number of Cystic Fibrosis patients attending Irish hospitals (See graph on page 6).
- Survey of the number and type of medical staff who cater for Cystic Fibrosis patients.

Implementation Activities

- Proposed report generation: 50 standard reports created in the following categories: Individual Patient Reports (single patient records from the database); Consultant Reports (pertaining to an individual consultants Cystic Fibrosis patients); Global Reports (pertaining to the country-wide Cystic Fibrosis population); Registry Director Reports (pertaining to the status of populating the database). Finalisation and availability of these reports from the website will be an action for the next phase of the project.
- www.cfairegistry.org: A set of public pages (27) to explain and support the activities of the Cystic Fibrosis Registry of Ireland including a "Privacy Policy".
- Funding proposal for Years 2-5 submitted to the Department of Health & Children.
- Patient entry onto the database commenced in July 2002; thirteen months after implementation began.



Security of the Registry Ireland

The Irish Cystic Fibrosis Registry is housed on a server in a secure building in Citywest Business Park, Dublin. The building is alarmed and guarded 24 hours per day, seven days a week.

To gain entry to the database via the internet, one requires both a user name and password which are issued by the Cystic Fibrosis Registry Ireland.

The data held on the database is encrypted. Only an authorised user will have a "key" to unlock the code and view the data.

Every person who is enrolled in the Registry may have a copy of the records kept on the Registry. This is guaranteed by the Data Protection Commission of Ireland and the Data Protection Act, 1988.

Data Collection

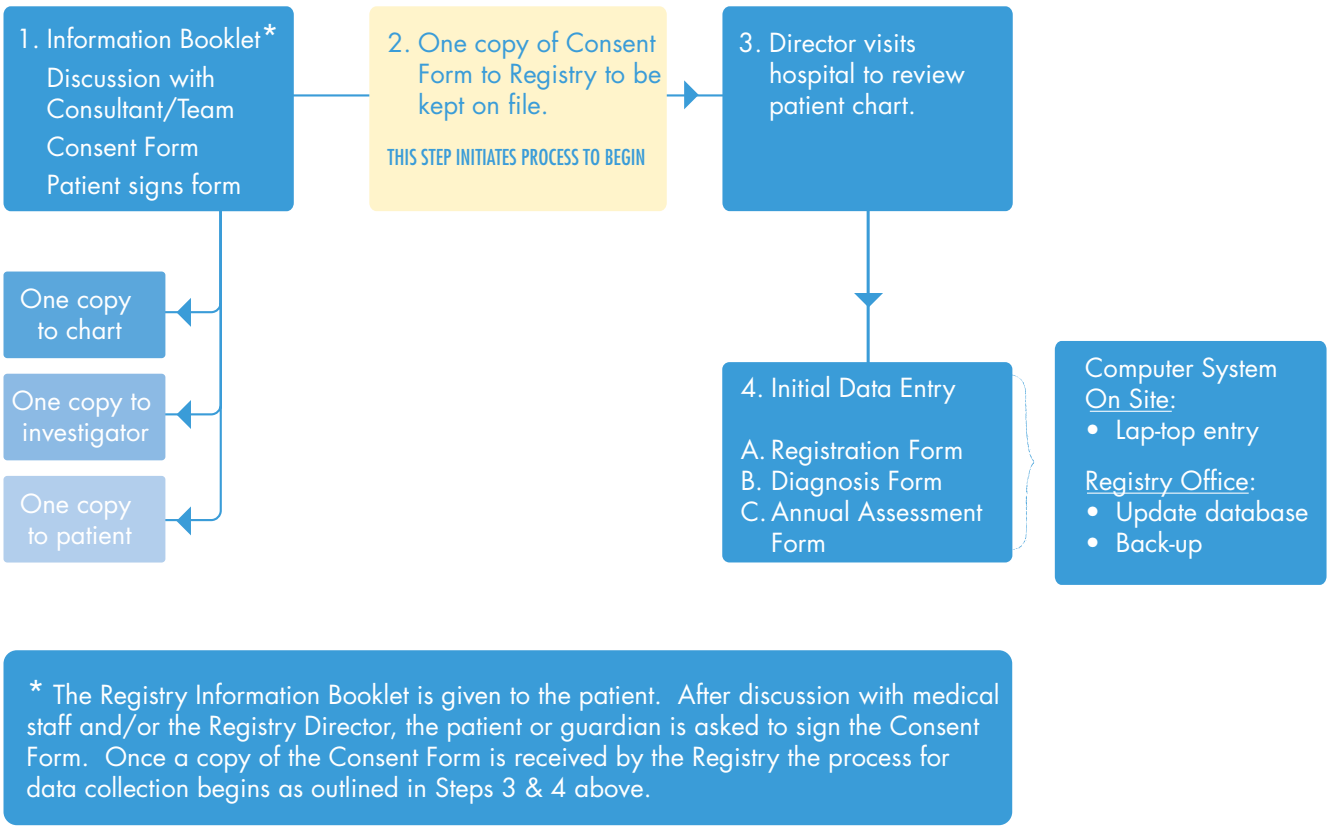
The process for data collection for the first phase of operation (Initial Data Entry) is diagrammed below.

Due to the nature of Cystic Fibrosis, many patients have frequent hospital admissions. This generates a large volume of documentation. Many patients would have several chart volumes in one hospital, and additional information at other hospitals. Vital information on diagnosis for example, may be kept in archived charts or in paediatric charts at other hospitals as would be the case for adult Cystic Fibrosis enrolees.

For all of these reasons, the Initial Data Entry combining Registration, Diagnosis and the first Annual Assessment is very time consuming. At present it is not possible to enter more than four patients per day. Once patients are registered it may be possible to enter up to six per day for Annual Assessments only. This is a very labour intensive exercise. Based on these rates and the current estimate of the total number of patients, it will take approximately 1.5 years to enter all patients once. Thereafter it will take approximately a year to enter all patients' Annual Assessments.

The data entry screens are very comprehensive, including approximately 110 fields to be entered for each Annual Assessment. The tables opposite outline the major categories of information that are collected on each patient.

Process For Data Collection



Data Collection

Primary categories of data collection are Registration, Diagnosis and Annual Assessment

Registration Details

Personal details including name and address
Date of birth
Health Board
Ethnicity
Name of consultant and family doctor

Diagnosis Details

Diagnostic tests, e.g. Sweat Test results
Genotype
Symptoms/Method of Diagnosis
Age at diagnosis

Annual Assessment Details

Number of hospitalisations between annual assessments
Complications
Pulmonary function tests
Chest X-ray reports
Clinical Chemistry
Infections: cultures and treatments
Long term therapies
Vaccinations/Immunisations
Physiotherapy regimes
Nutritional regimes
Transplant status
Social details such as number of days off work or school in previous 12 months

While the data tables may be criticised for too much detail in the collection phase, they offer powerful versatility for a broad range of reports. In later years these data will provide opportunities for extensive research into many different aspects of the Irish Cystic Fibrosis population.

Data Analysis and Reports

Standard reports will be available on the website from live data after Initial Data Entry of at least 50% of patients.

The table on page 10 is intended to give an idea of the type of report which can be generated by the database. The report section of the Registry will be discussed and finalised by the Registry Management Committee during the second phase of the project. These will be 'read-only'. Accessibility and report content are likely to follow the criteria set out in the table.

Data Analysis and Reports

Type of Report	Contents	Accessible to:
Individual Patient Reports	*These reports are restricted to the consultant of the patient.* • Personal details • Most recent update, i.e., Annual Assessment • No. of hospitalisations and exacerbations in last period • Cultures in last period • Antibiotics in last period • Lung Function in last period • Other Treatments in last period • Social Summary	Consultant to the patient
Consultant Patient Reports *(i.e., all patient summary for a particular Consultant)*	*These reports are restricted to the patients of a single consultant.* • List of consultants patients on Registry • Method of diagnosis • Genotypes • Hospitalisations & exacerbations in last period • Transplant status • Total cultures in last period and type of culture • Antibiotics in last period • Other treatments in last period • Vaccinations in last period • Shwachmann Scores summary • Social summary	Consultant of his/her patients ONLY
Global Patient Reports *All data in this section of Reports will be Anonymised*	*These reports contain no personal details of any patient.* • Total number of patients enrolled, mean age, gender split, etc. • Summary of genotypes • Diagnostic method summaries • Types and rates of complications in previous 12 months • Total number of hospitalisations, exacerbations, transplant status • Means for age groups, height, weight, BMI • Numbers of cultures and types of cultures • Antibiotics: number of courses, IV or oral, intermittent, prophylactic, etc. • Nutritional treatment summaries • Physiotherapy summaries • Other drug treatment summaries • Shwachmann score summaries over age groups • Social data summaries	Users of these data will be approved by the Registry Management Committee after review of submitted protocol(s)
Registry Director Reports	• Distribution of patients by Consultant, hospital, region • Age distribution and other demographics • Transplant status summaries	Registry Director ONLY

The Future

There is a wealth of information that can be gleaned from the Registry. Much of this information will be summarised and included in each Annual Report.

Further analysis of the data will be done through various research projects and expert epidemiology advice will be sought.

Future Activities and Objectives

- The number one priority is to populate the database as soon as possible.

  Certain other activities should also be undertaken while the above task is continuing:

- Complete Ethics Committee approval to 100% of hospitals/patients.
- Signed informed Consent Forms for 100% of patients.
- Standardise Annual Assessment tests throughout the country; it will be possible to make data entry far more efficient if all units follow a standard annual protocol.
- Write a Training Manual for the database.
- Deliver standard reports to the website.
- On-site hospital training for standard report down-loads and data entry.
- Begin to compare data with other national registries.

Acknowledgments

The Cystic Fibrosis Registry of Ireland would like to gratefully acknowledge the commitment and support of the Minister for Health & Children, Mr Micháel Martin, T.D., as well as many of the officers in the Department of Health and Children.

Other groups of people have contributed enormously to the Registry. These include the Cystic Fibrosis Association of Ireland and the members of the Registry Management Committee.

There are many consultants who work tirelessly in the area of Cystic Fibrosis care. They include paediatricians and respiratory physicians, as well as microbiologists and medical geneticists. Many of them have provided suggestions and recommendations to the setting up of the Cystic Fibrosis Registry.

Other healthcare professionals whose ideas have been incorporated into the implementation phase are the Cystic Fibrosis Nurse Specialists, dieticians, physiotherapists, social workers and pharmacists.

Ethics Committees from the many hospitals must be thanked for their earnest review of the protocol.

A project such as this benefits enormously from management expertise and that was enthusiastically donated by Mr Paul Allen of Bearing Point Ireland (formerly KPMG Consulting).

The database application and web page design was devised by Mr Rahul Rana of Ecom-Ireland whose advice is always lucid and constructive.

It is important to note that almost every Registry relies on one that pre-dates it, and in this case, Dr Harry Comber of the National Cancer Registry has been generous in sharing his experience and ideas for implementation. In addition, Ms. Eileen Williamson of the National Parasuicide Registry of Ireland has been supportive. Specific to Cystic Fibrosis, researchers who developed registries in the following countries have shared their wisdom: the UK, USA, and Germany.

Finally, the encouragement of parents and patients (particularly adult cystic fibrosis patients) is gratefully acknowledged. They have been instrumental in driving this project from the outset.

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