



## **BLUEPRINT FOR THE FUTURE**

*This is a very important report.*

*It summarises the findings of Dr Pollock's study of the services in the Republic of Ireland for people with cystic fibrosis and points out how satisfactory levels of service should be delivered.*

*This is the first time that the staggering distance between the way cystic fibrosis services are currently delivered and the way they should be delivered has been so accurately measured.*

*While Dr Pollock's analysis of current services makes disturbing, and at times uncomfortable reading, his recommendations are cause for optimism.*

*They represent a blueprint for the future of care for people with cystic fibrosis and must be acted on immediately.*

*The Association's primary objective is to make sure that the nine Specialist Cystic Fibrosis Centers recommended in this report are established without delay.*

*This is the only way that the chasm between current services and acceptable services can be bridged. I urge you to become directly involved in delivering this vision.*

*I would like to thank Dr Pollock, Association members, medical staff and members of the Association's Medical and Scientific Council for their co-operation and valuable contribution towards completing this study. I would also like to thank Solvay Healthcare for contributing towards the cost of commissioning this report.*

*“While Dr Pollock's analysis of current services makes disturbing, and at times uncomfortable reading, his recommendations are cause for optimism.”*

**CARL RAINEY**

*Chairman*

*The Cystic Fibrosis Association of Ireland*

*February 2005*



## **FOREWORD**

*In presenting this Report to The Cystic Fibrosis Association of Ireland, I would like to express my gratitude to all of the people who gave so generously of their time and advice in contributing to it. They are named in Appendix 1.*

*I would like to thank, in particular, Jackie Frances, the Paediatric Cystic Fibrosis Nurse Specialist at the Royal Brompton Hospital, who gave me a great deal of her time, and who made available the excellent transition documentation which she had prepared for her unit.*

*I also wish to express my immense admiration for all of the people I have met, working in this field, for their unstinted dedication to the provision of services to the patients and families in their care.*

**DR R.M. POLLOCK**

*MPA Health Strategy & Planning Ltd*



## INTRODUCTION

This Review has been commissioned by The Cystic Fibrosis Association of Ireland with the following remit:-

1. To review existing hospital services for Cystic Fibrosis, and to relate these to accepted international standards.
2. To comment on the ability of the service, as at present organised, to deliver care to currently acceptable clinical standards.
3. To review the measures which might be taken in structuring the service, so as to improve outcomes nationally.

## METHODOLOGY

The approach adopted has been:-

1. To conduct a Selective Literature review.
2. To visit units which are held in the highest regard in the UK and to discuss their operation and ethos with their lead clinicians and other Cystic Fibrosis staff.

The units visited were Royal Brompton Hospital (Children's Unit), Great Ormond Street Children's Hospital and Royal Brompton Hospital (Adult Unit).

3. To visit Cystic Fibrosis Units in Ireland, discuss the local situation with the lead clinician and with the other staff of the Unit and to tour the available facilities (10 of the 13 'Centres' on the Cystic Fibrosis Registry of Ireland were visited: Mayo, Sligo and Tralee were not visited).
4. To have telephone discussions with expert specialists on the Subject (the list of centres visited and people interviewed appears as Appendix 1).

*“...Ireland displays a very much higher number of deaths than does England and Wales...”*

## BACKGROUND

Of all European countries, Ireland has the highest incidence of Cystic Fibrosis (1:1600 cf 1:25000 in Finland) and there are also strong indications that outcomes in Ireland are worse.

Comparison of survival between countries is complicated by the absence of a European Registry recording individual age and vital statistical data for all cases (as is the case in the USA). However, trends in survival may be inferred from the trends in median age at death (Fogarty et al, Chest 2000)<sup>1</sup>. Clearly caution has to be exhibited in interpretation (e.g. some variation in diagnosis and coding may occur) nevertheless Ireland displays a very much higher number of deaths than does England and Wales, and the same is true for the comparison with Northern Ireland. Fogarty et al also observe that socio economic status appears to be an important determinant of survival. Curtis et al (1997)<sup>2</sup> also report that health insurance and increased social economic status were independently associated with increased survival.



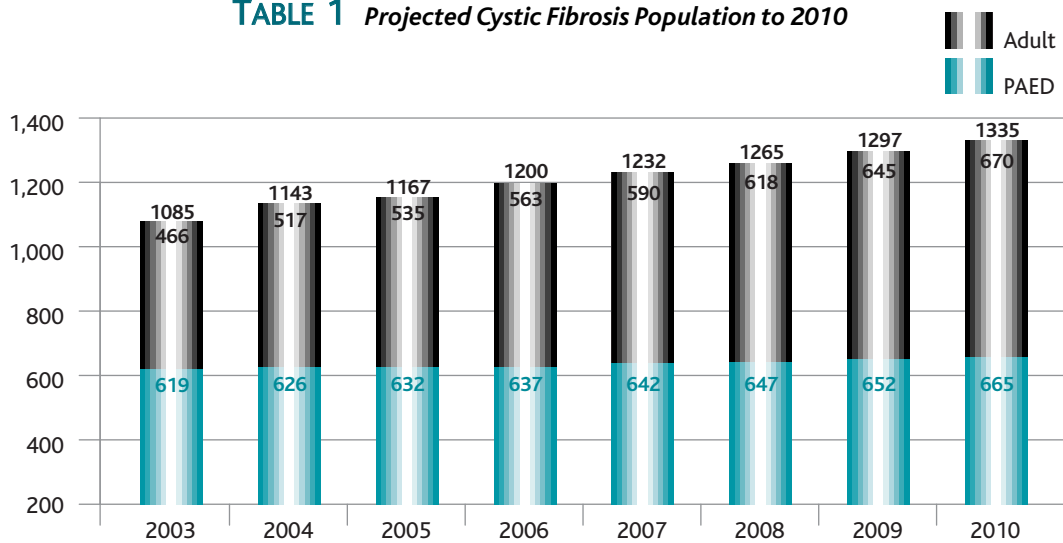
## PRESENT POSITION AND FUTURE GROWTH

A review of present services (2004) indicates that a total of 1,143 patients is under care in Ireland:-

- 517 are Adults (45%)
- 626 are Children (55%)

With increasingly successful care, survival is continuously improving. The pattern of growth showing an increasing adult population, but an almost constant paediatric population is shown in Table 1. It is thus anticipated that, in another ten years, the number of adults in care will come to equal, and then exceed, the number of children (Elborn, 1991)<sup>3</sup>; Bush (Personal Communication, 2003).

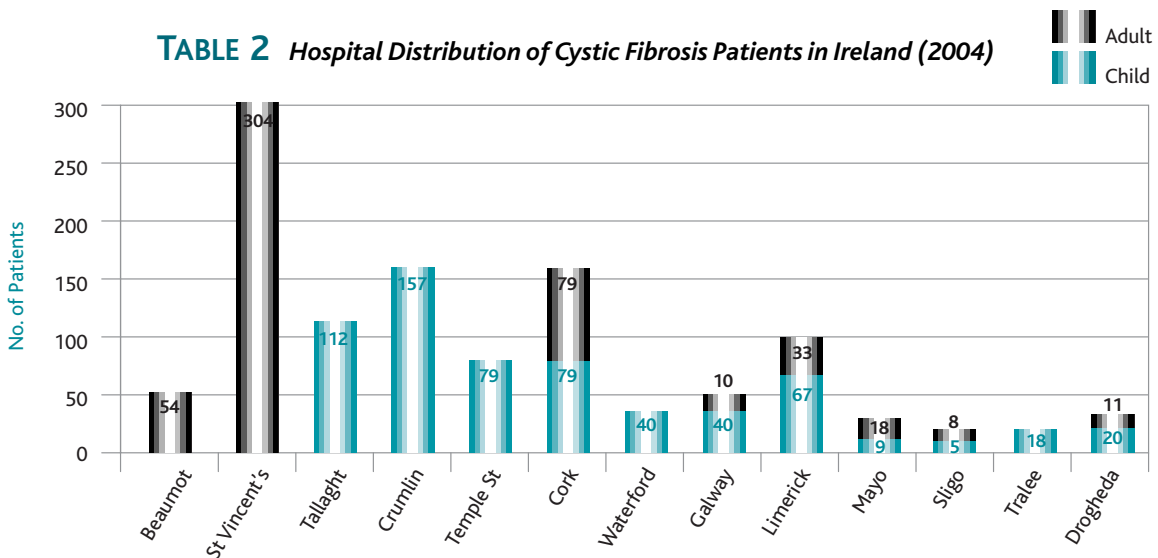
**TABLE 1** Projected Cystic Fibrosis Population to 2010



## WORKLOAD AND DISTRIBUTION OF SERVICES

An overview of the extent and distribution of Cystic Fibrosis services in Ireland reveals the current picture. Table 2 shows the distribution and workloads of the existing Cystic Fibrosis centres.

**TABLE 2** Hospital Distribution of Cystic Fibrosis Patients in Ireland (2004)





Of the 13 locations\* identified in the Cystic Fibrosis Association Registry.

- Two are purely Adult Centres: St. Vincent's University Hospital, Dublin (St Vincent's) and Beaumont Hospital, Dublin (Beaumont).
- Two have both Children's and Adult Services: Cork University Hospital (Cork) and Limerick Mid Western Regional Hospital (Limerick).
- Galway University Hospital (Galway), Mayo General Hospital (Mayo), Sligo General Hospital (Sligo) and Our Lady of Lourdes Hospital, Drogheda (Drogheda) have some Adult Services but are mainly Paediatric.
- The remaining five are purely Paediatric Centres.

*“...the amount of commitment to CF is everywhere seriously inadequate.”*

\* For the convenience of readers, in parts of this report hospital names have been abbreviated. The full hospital names are listed in Appendix 8.

Key issues arising from this are:-

1. The number of Centres which have very small patient numbers.
2. The large number of Centres overall, given the total size of the patient population.
3. The imbalance between Child and Adult Centres (of particular note when the expectation is for a 50/50 split within the next few years).

This adds complications to the already sensitive process of transition to Adult care; continuing care in a separate, usually distant unit imposes an additional travel and psychological burden.

Also, as a result of improving care, child patients will comprise a predominantly 'well' population, whereas, with the greater extension of life into adulthood, an increasing population of adult patients, with more complex manifestations and making greater demands on CF services, will come to exist.

## CURRENT STAFFING PROVISION

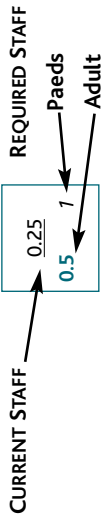
Table 3 shows the staffing position at each of the 13 locations.

The medical leadership of the team is clearly important yet the amount of commitment to CF is everywhere seriously inadequate, and usually the responsibility of a single individual, creating problems of cover during inevitable absences.

The survey also indicates a striking absence of Specialist Registrars, raising concerns about matching future consultant manpower needs.

*“...segregation from an infection control point of view is difficult, and the social mix of young CF patients and elderly – some incontinent – patients, is very unsatisfactory.”*

During this survey it has been evident that the lynch pins of the Cystic Fibrosis services are the Cystic Fibrosis Nurses, Physiotherapists, Dieticians, and Psychologists. However, their numbers are totally inadequate.



**TABLE 3**  
Hospital Distribution of Cystic Fibrosis Patients in Ireland (2004): Current Staff Vs Staff Required

STAFF* REQUIRED PER 50 PATIENTS - ADULTS / PAEDS	BEAUMONT	ST. VINCENT'S	TALLAGHT	CRUMLIN	TEMPLE ST	CORK	WATERFORD	GALWAY	LIMERICK	MAYO	SILIGO	TRALEE	DROGHEDA	TOTAL
Consultant 1 0.8 / 0.7	0.5 0.9	0.6 5	0.3 1.6	0.3 2.2	0.3 1	0.1 1.3	0.25 0.6	0.1 0.2	0.25 0.5	Shared 0.3	Shared 0.1	0.2	0.2	0.3
Consultant 2 0.6 / 0.5	0.6	4	1	1.6	0.8	0.2 1	0.01 0.4	0.01 0.1	0.1 0.4	0.2 0.7	0.1 0.05	0.2	0.1	0.2
Staff Grade	0	0	0	0	0	0	0	0	0	0	0	0	0	0
CF Specialist Registrar	0.3 0.5	0.5 3	0.5 1	0.2 1.6	0.3 0.8	0.8 0.8	0.2 0.4	0.1 0.4	0.3 0.7	Shared 0.2	0.1 0.05	0.2	0.1	0.2
CF Nurse 1-1.5 / 1-1.5	1 1.3	3 8	1 3	2 4	1 2	1.5 2	1 1	1 0.3	1.5 0.8	0.5 0.2	0.2 0.1	0.5	0.3	0.5
Physiotherapist 2 / 2	0.4 2	1.25 12	0.4 4.5	0.9 6.3	0.5 3	0.5 3	0.5 (paeds+) 1.6	1 0.4	1.5 1.3	Shared 0.7	Shared 0.3	0.7	0.4	0.8
Dietician 0.4 / 0.4	0 0.4	0.8 2.4	0.5 1	0.9 1.3	0.5 0.6	0.2 0.6	0.5 (paeds+) 0.3	0.1 0.1	1 0.3	Shared 0.1	Shared 0.1	0.1	0.1	0.2
Social Worker 0.4 / 0.4	0.25 0.4	2.4	0.2 1	0.9 1.3	0.25 0.6	0.2 0.6	0.3 0.6	0.1 0.3	1 Shared 0.3	Shared 0.1	Shared 0.1	0.1	0.1	0.2
Psychologist 0.4 / 0.4	0.4	2.4	0.2 1	1.3	0.05 0.6	0.6 0.6	0.3 0.6	0.1 0.3	0.3 0.5	0.1 0.1	0.1 0.04	0.1	0.1	0.2
Secretary 1 / 1	0.4 1	1.2 6	1 2.2	0.4 3	0.30 7.6	1 1.6	0.8 1.6	0.05 0.2	0.7 1.3	Shared 0.4	Shared 0.2	0.4	0.2	0.4
Data Clerk 0.1 / 0.1	0.1	0.6	0.2	0.3	0.2	0.2	0.08	0.02	0.1	0.04	0.02	0.4	0.02	0.04
Pharmacist 0.3 / 0.3	0.3	2	0.2 0.7	0.9	0.5	0.5	0.2	0.06	0.2	0.1	0.05	0.1	0.07	0.1
<b>CURRENT STAFF</b>	2.85	8.85	4.3	5.6	3.20	3.70	2.46	2.35	5.35	0.5	Shared	Shared	0.5	40
<b>RECOMMENDED</b>	8.4	47	17	24	12	12	6	1.5	5	2.8	1.3	3	1.7	175

Adult	54	304	79	10	33	18	8	11	517
Child			112	157	79	40	40	67	626
<b>TOTAL PATIENTS</b>	54	304	112	157	79	158	40	50	1143

\* = From 'Standards of Care' Cystic Fibrosis Trust (UK) (Paeds +) = These cover all paediatric care not just cystic fibrosis. Shared = The exact allocation to cystic fibrosis is not quantifiable.



## PHYSICAL PROVISION

The standards of physical provision found during the survey varied quite widely.

Of the 13 units, ten were visited: Tralee General Hospital (Tralee), Mayo and Sligo General Hospital (Sligo) were not visited.

*“The major defects in the service however are the low numbers in the Paediatric Unit and the total lack of an Adult Service.”*

The visit to Beaumont involved only the interview with Professor McElvaney; the CF Nurse was absent on the day of the visit and the facilities were not visited. As Beaumont is a relatively new hospital it is assumed that the physical provision is adequate. However, it does not operate as a CF Centre, there being neither separate CF Clinics nor designated CF in-patient facilities. This pattern, it appeared, had been instigated at the behest of patients apparently in response to their concerns about cross infection risks when aggregating numbers of CF patients together. The staffing allocation is inadequate and there is no dietician allocated to the CF service.

At The National Children’s Hospital, Tallaght (Tallaght) the clinic facilities are good, as are the in-patient facilities, which provide well for single rooms; Pathology and Radiology support is impressive.

Elsewhere in Dublin the provision is very poor indeed.

The two dedicated children’s hospitals, Our Lady’s Hospital for Sick Children, Crumlin (Crumlin) and The Children’s University Hospital, Temple St (Temple St) present similar pictures: space standards are very poor both for in-patient and out-patients; segregation of patients and privacy are very difficult to achieve. In-patient provision at both hospitals is notable both for the limited space for clinical functions, and for the absence of facilities for parents.

At St. Vincent’s, with 304 patients the largest centre in Ireland, the position is bleak. The in-patient provision is seriously unsatisfactory, with CF patients and elderly respiratory patients cheek by jowl in tightly constrained space; segregation from an infection control point of view is difficult, and the social mix of young CF patients and elderly – some incontinent – patients, is very unsatisfactory. At times CF patients have to be admitted to other, general, wards where nursing staff do not have specific CF expertise.

The CF team members are dedicated and hard working, but their number and conditions are unsatisfactory. The sole consultant has a small office: the three CF nurses share a small room which acts as their office, the location for dealing with drop-in patients, and where patients undergoing testing or procedures are cared for. This is unrealistically, indeed dangerously, multi functional. The administration/secretarial space and resources are inadequate.

At each of the above locations there are some proposals for improvement.

At Crumlin a new CF Resource Centre, which has been funded by The Children’s Medical Research Foundation at the hospital, was completed in 2004. Following a critical report of the poor standards of provision in 2003 the Department of Health and Children commissioned a study for the preparation of an outline brief for the replacement of the hospital and this was completed during 2004.

*“The lack of segregation and isolation facilities, both for out patients and in patients is dangerous...”*



Temple St will be replaced by a purpose built 21st century paediatric hospital and will be co-located on The Mater Hospital site. It will incorporate a 20 bed intensive care unit, multiple isolation rooms and a new respiratory department including a purpose built respiratory function laboratory, and will provide a walk-in Cystic Fibrosis clinic service.

New development is currently under way at St. Vincent's but improvements in the situation for CF care, require the completion of the present building phase, decanting of other facilities, and refurbishment of the vacated space. This is unlikely before 2005.

Elsewhere in the country, physical provision is generally much better.

The exception to this statement is Drogheda where the CF service is located in a small general paediatric ward with limited segregation facilities. Patients are seen both here and at a clinic at Cavan. (At the time of the visit there was no CF physician at Drogheda, though a new appointment was expected. The CF Nurse is part time; physiotherapy and dietetic services are drawn from the general pool of services. The hospital has no Microbiology facility). (N.B. The figures in Table 2 relate to both Cavan and Drogheda attendance.)

Cork, which cares for 158 patients, is an excellent University Hospital, with good physical provision. The configuration of the unit is unique in that Adult and Children's services are located close to each other on the same Out-Patient Department corridor. The teams for both adults and children comprise the same personnel – only the consultants are different. The 'problems' of transition are as a result, minimised. The atmosphere among the team was positive and dynamic, though – as elsewhere – the staffing level is too low.

*“It is essential that there is a Consultant Microbiology presence at every CF Centre.”*

Limerick has an excellent Paediatric Unit, within a modern hospital. It cares for 67 Child CF patients (there are 33 Adults). Fifty percent of the beds are in single rooms with sanitary facilities and good facilities for mothers to 'room in'. The staff are dynamic and enthusiastic and staffing levels generally are among the best in the country, though input from the consultants is below a desirable level. Unfortunately the present links for Adult Services are less than perfect and care is provided by two respiratory physicians, one who provides out-patient care, and a different physician who provides in-patient care.

The Galway unit is in a high calibre University Hospital. The unit however cares for only 40 Children and 10 Adults. The staffing provision is one of the best in the country though below desirable levels in dietetics and psychology. The staff are dynamic and committed. The principal problem is that Adult care is not well integrated with Paediatric care, and is fragmented, being dealt with by two different consultants on two sites.

The Waterford Regional Hospital (Waterford) is pleasant, and fairly new, with good facilities for clinics and in-patients. Parent facilities too are good. Though there is an excellent full time CF Nurse, other staff are not specifically dedicated to the unit, physiotherapy and dietetics being served by general paediatric personnel. The major defects in the service however are the low numbers in the Paediatric Unit and the total lack of an Adult Service.



## SUMMARY OF DEFICIENCIES IN CURRENT SERVICE

- The current staffing for Cystic Fibrosis is seriously inadequate. It is very unbalanced, and it is too thinly distributed over too many, too small, units.
- In many locations physical resources, particularly in Dublin, fall well below accepted standards. St Vincent's is particularly poor. The lack of segregation and isolation facilities, both for out-patients and in-patients is dangerous, creating significant risks of cross infection with virulent organisms (such as MRSA, Burkholderia Cepacia, and resistant Pseudomonas), additionally carrying risks of litigation (as with Hepatitis C) if an outbreak were to arise.
- There are huge deficiencies in the provision of adult services, and the availability of consultants is further diminished by their heavy General Medical 'on-call' responsibilities.
- This is of special concern because of the steadily increasing proportion of adults, and the necessary complexity and severity of their condition.
- Funding is unstable and at risk from other pressures.
- There is no system of accreditation of CF Centres.

*“There must be a high standard of hygiene practiced by all concerned, particularly washing or disinfection of the hands and regular expert microbiological surveillance.”*

## DESIRABLE PATTERN OF CARE

### a) Centralisation

The case that Centralised Specialist Care markedly improves the quality and outcome of care to Cystic Fibrosis patients is overwhelming.

It has been demonstrated that it provides better symptom control, more concentrated expertise, more intensive care and greater patient satisfaction (Nielson et al 1982)<sup>4</sup> (Walters et al 1994)<sup>5</sup>.

Frederikson et al (1996)<sup>6</sup> demonstrated improved nutritional and pulmonary status, and improved survival in The Danish "Centre-treated" model.

Mahadiva et al (1998)<sup>7</sup> have demonstrated conclusively that the most effective care for both Children and Adults is delivered through a team of experienced professionals operating in a Specialist CF Centre.

### b) Size and Essential Features of a Specialist Cystic Fibrosis Centre

The essential component of a Specialist CF Centre is people – both sufficient patients with CF (at least 50/100 children for a paediatric centre and 50/100 adults for an adult centre) and adequate numbers of suitably experienced staff to advise them on treatment. Also there are numerous other people in the hospital, some of whom may never come into direct contact with patients and families, who nevertheless are essential and contribute to the smooth running and expertise of the Specialist CF Centre's service.

#### A Functions of a Specialist CF Centre

- i) Care and Annual Review of patients including the early assessment and management of infants detected by antenatal and neonatal screening. The availability and offer of open access service gives necessary support to patients and their families as well as to general practitioners.
- ii) Provision and co-ordination of a wide range of treatments and services which require special expertise in patients with CF. These would include treatment of newborn infants with meconium ileus, massive



haemoptysis, pneumothorax, unusual and complex respiratory and gastrointestinal problems, liver and biliary complications, enterostomy feeding, thoracic surgery, ENT surgery, major elective surgery, cystic fibrosis related diabetes mellitus, liver disease, obstetric management, rheumatology and psychology.

Counselling about fertility and pregnancy. Critically this requires the availability of consultant Gastroenterology and Endocrinology staff with a special interest in Cystic Fibrosis.

- iii) Provision of expertise in specialised procedures is often required by people with CF. For example, inserting totally implantable venous access devices, upper gastrointestinal endoscopy, gastrostomy, non-invasive ventilation, bronchial artery embolisation and fibreoptic bronchoscopy on infants and children.
- iv) Access to diagnostic and specialised laboratory facilities including genetic investigations and counselling, antenatal diagnosis, neonatal screening investigations, sweat testing, CT and V/Q scanning, specialised cardiovascular investigations, and specialist investigation and evaluation of pancreatic function, gastrointestinal and liver disease. Detailed respiratory function tests. It is essential that there is a Consultant Microbiology presence at every CF Centre. It is also critical that a Microbiology Reference Laboratory is established which would have available the molecular methods which are necessary in the speciation of BCepacia.
- v) Psychosocial support for problems specific to CF. Education, employment, financial benefits, personal and family support particularly around the time of diagnosis, problems with burden of treatment, adapting to progressive disease, pre-transplant psychosocial assessment and counselling, terminal care and bereavement counselling. Treatment of depression, anxiety and a range of emotional difficulties, disordered eating and non-adherence problems.
- vi) Ensuring the smooth transition of patients from paediatric to adult care, usually by running a regular adolescent/transition clinic with the appropriate paediatric and adult CF unit staff.
- vii) Liaison with transplant centres and assessment of patients.
- viii) Providing opportunities for patient involvement in the monitoring and development of the service and support for patient advocacy.
- ix) Responsibilities for entering data of all patients into the CF Registry of Ireland.
- x) Performing clinical research aimed at increasing the understanding of CF and introducing and evaluating new treatments. Collaboration with multi-centre trials.
- xi) Attendance of the staff at national and international CF meetings. Presentation and publication of the results of research.
- xii) Additional specialised investigations should be available at designated Tertiary CF centres, e.g. complex lung function tests particularly for young children and infants. Nasal potential difference measurements.

*“With improving survival, there will be more adults in need of complex care.”*

*“Arrangements for direct admission to the ward should be in place to avoid long delays in the Accident and Emergency...”*

## **B Facilities Necessary for the Care of People with Cystic Fibrosis**

### *B1 Out-patient facilities which should be available at all Specialist CF Centres*

Sufficient clinic sessions designated for patients with CF are required to allow adequate time for discussion and to avoid overcrowding in the waiting areas.



Ideally, there will be an area in each Specialist CF Centre where patients and families attending for the day ('day cases') can be seen and treated on an ad hoc basis. This area can also be used for patients coming from other hospitals for consultations, annual reviews, specialist investigations, and for the provision of Home Intravenous Antibiotic Therapy which is now used to treat many patients with acute exacerbations of CF. Relatives and patients require some accommodation, other than the waiting area, as they will often be spending many hours at the unit with young children and need a base for the day.

*“These figures should therefore be regarded as representing the minimum acceptable standards.”*

There should be separate rooms available for medical staff, physiotherapists, dieticians and psychologists/social workers and the nurse specialist along with consulting and treatment rooms, where patients can be seen and minor procedures performed.

There must be a high standard of hygiene practiced by all concerned, particularly washing or disinfection of the hands and regular expert microbiological surveillance. This further emphasises the importance of on site Consultant Microbiology Services.

*“...it is vital to recognise that regular contact with a Specialist CF Centre is an essential component of adequate care.”*

Respiratory function equipment should be available, appropriately calibrated, and adequate to cater for patients with transmissible infections.

#### **B2 In-patient facilities which should be available at all Specialist CF Centres**

Specialist CF Centres should have a clear infection control policy. Patients with cystic fibrosis are at particular risk of harbouring multi-resistant organisms, e.g. methicillin resistant S.aureus (MRSA), P.aeruginosa, and Burkholderia Cepacia; (Jones et al (2000)<sup>8</sup>, Armstrong et al (2000)<sup>9</sup>). This poses a potentially fatal risk to themselves, to other patients with CF and also to susceptible patients with other illnesses (McCallum et al (2000)<sup>10</sup>).

The beds should be in single rooms (the main reason being to reduce the likelihood of cross-infection), with en-suite bathrooms. Children should be in single room accommodation, which has adequate observation and appropriate facilities for parental sleeping.

The equivalent of three to five beds is required for every 50 patients who are receiving full care at the Specialist CF Centre. The number of beds required would vary according to whether the patient population is mainly composed of relatively well children or adults at a Specialist CF Centre, a significant proportion of whom may be severely affected. With improving survival, there will be more adults in need of complex care.

Facilities should be available for education and recreation taking account of the potential risks of cross-infection.

*“...the skills and experience of the existing clinical staff at the smaller units must not be lost...”*

Equipment for measurement of height and weight should be available.

It is inappropriate for patients who have an exacerbation of their chest infection to join a waiting list. They should be admitted the same day if the doctor considers this is necessary. Arrangements for direct admission to the ward should be in place to avoid long delays in the Accident and Emergency Department, and prevent cross infection.

#### **C Staffing of a CF Centre**

These staffing requirements have been accepted as representing an overview of expert opinion. The requirements vary depending on the age of the patients, the severity of their condition and the amount of



community support that a Specialist CF Centre provides. The numbers may have to be increased where there is a constant higher proportion of very unwell patients with more highly complex needs. It is suggested that an individual will require approximately half or more of their time working in CF patient care to maintain their necessary expertise.

The suggested number of whole time equivalent staff (WTE) required for every 50 patients on full care:-

STAFF MEMBER	SPECIALIST PAEDIATRIC CENTRE	SPECIALIST ADULT CENTRE
Consultant 1	0.7	0.8
Consultant 2	0.4-0.5	0.5-0.6
Staff Grade*	-	-
CF Specialist Registrar	0.5	0.5
CF Nurse	1.0-1.5	1.0-1.5
Physiotherapist	2.0	2.0
Dietician	0.4	0.4
Social Worker	0.4	0.4
Psychologist	0.4	0.4
Secretary	1.0	1.0
Data Clerk	0.1	0.1
Pharmacist	0.3	0.3

Consultant 1 has a major commitment to CF care (Clinic Director or Lead Clinician in Cystic Fibrosis); Consultant 2 is involved with routine CF care but to a lesser extent. At Specialist CF Centres, there is a Specialist Registrar (CF Sp. R) with full commitment to CF, in addition to the usual junior staffing of the paediatric/medical team. \*(As a Staff Grade does not exist in Ireland, the ratio shown in the CF Trust proposals for that grade has been included in the Consultant grade, though in Adult Care the option may be taken to allocate a proportion to the Specialist Registrar Grade). It should be noted that Centres of Excellence elsewhere – notably in North America – not only attain but exceed these standards. These figures should therefore be regarded as representing the **minimum** acceptable standards.

## LOCATION OF CF CENTRES

The foregoing considerations of the benefits of centralisation; the appropriate size and features of a CF Centre, and its staffing, dictate that in future it will be essential, to base care on many fewer units, which are each of significant size, and so able to sustain viable staffing levels.

The determination of the precise number of Centres which would most effectively service Ireland, will have to take account of accessibility on the one hand and the necessity to create centres of such mass as to ensure international class care on the other. There will inevitably be conflict between these two forces: in resolving it, it is vital to recognise that regular contact with a Specialist CF Centre is an essential component of adequate care. It is not a service to patients and families to provide ease of access at the cost of less than maximally effective services.



Clearly the locations at Tralee, Mayo, Sligo and Drogheda have insufficient workload to justify the clinical establishments needed to ensure first class service.

The inclusion of the Mayo and part of the Sligo workload at Galway, would boost the patient load there to the desirable level for a CF Centre and so ensure an improved standard of care.

The population around Sligo and to the North, particularly in Donegal currently presents an unsatisfactory picture with no easily accessible major centre. However, if the problem is viewed without geographical constraints, a solution at a centre in Northern Ireland seems logical (particularly if proposals for a new centre at Derry are consummated in the reasonably near future).

Waterford is the largest of the small units, but has only 40 child patients, and no adult service at all, so that in terms of workload it appears to fall below the 'absolute minimum' advised in the CF Trust 'Standards' document. However some 20 further children who are Waterford residents have been identified as being treated elsewhere; with a properly staffed unit, and the development of Adult Services they could be cared for without having to travel. These considerations have led to the conclusion that the unit should be developed as a Centre, and that full adult services should be developed.

*“...that early diagnosis of Cystic Fibrosis through neonatal screening (combined with aggressive nutritional therapy) can result in significantly enhanced long term nutritional status....”*

In the refocusing of care to CF Centres which are of sufficient size as to justify the recommended staffing levels, the skills and experience of the existing clinical staff at the smaller units must not be lost, and attention should be given to harnessing them (e.g. through variations of contract to allow them to participate in CF Centre care).

Table 4 identifies possible future centres and their indicative workload. Table 5 shows proposed future staffing levels for each centre.

**TABLE 4** Proposed Centre based cystic fibrosis care and patient distribution using 2010 projections

LOCATION	CHILDREN	ADULTS
North Dublin and the North East	Temple St. 105	Beaumont 110
South Dublin	Crumlin } Tallaght } 210	St. Vincent's 240
South Ireland	Cork 80	75
	Waterford 55	55
South West Ireland	Limerick 75	65
West Ireland	Galway 70	60
Donegal and the North West	Northern Ireland (Belfast or possibly Derry) 70	65
<b>TOTAL</b>	<b>665</b>	<b>670</b>



**TABLE 5 Proposed Future (2010) Cystic Fibrosis Staffing (in whole time equivalents)**

STAFF PER 50 PATIENTS IN IRELAND*	BEAUMONT		TEMPLE ST		ST VINCENT'S		CRUMLIN/TALLAGHT		CORK		WATERFORD		LIMERICK		GALWAY		N. I. PATIENTS		TOTAL STAFF
	Paed	Adult	Paed	Adult	Paed	Adult	Paed	Adult	Paed	Adult	Paed	Adult	Paed	Adult	Paed	Adult	Paed	Adult	
0.7	0.8	Consultant 1	1.60	3.60	3.10	1.10	1.10	1.10	0.80	0.70	1.10	1.00	1.10	1.00	1.10	1.10	1.10	1.10	20.00
0.5	0.6	Consultant 2	1.20	2.25	2.20	0.60	0.60	0.60	0.60	0.60	0.75	0.70	0.80	0.70	0.80	0.70	0.80	0.70	13.80
-	-	Staff Grade	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
0.5	0.5	CF Special Registrar	1.00	2.25	2.20	0.80	0.70	0.60	0.60	0.50	0.80	0.60	0.70	0.50	0.70	0.50	0.60	0.60	13.05
1-1.5	1-1.5	CF Nurse+ (Average)	2.0-3.0 2.50	4.5-6.75 5.63	4.4-6.6 5.50	1.6-2.4 2.00	1.4-2.1 1.75	1.1-1.7 1.40	1-1.5 1.25	1.1-1.7 1.40	1.5-2.2 1.85	1.2-1.8 1.50	1.4-2.1 1.75	1.1-1.7 1.40	1.4-2.1 1.75	1.4-2.1 1.75	1.4-2.1 1.75	1.2-1.8 1.50	32.53
2.0	2.0	Physiotherapist	4.00	9.00	8.80	3.20	2.80	2.20	2.00	2.00	3.00	2.40	2.80	2.20	3.00	2.40	3.00	2.40	52.20
0.4	0.4	Dietician	0.80	1.80	1.80	0.60	0.60	0.60	0.40	0.40	0.60	0.50	0.60	0.50	0.60	0.50	0.60	0.50	10.80
0.4	0.4	Social Worker	0.80	1.80	1.80	0.60	0.60	0.60	0.40	0.40	0.60	0.50	0.60	0.50	0.60	0.50	0.60	0.50	10.80
0.4	0.4	Psychologist	0.80	1.80	1.80	0.60	0.60	0.60	0.40	0.40	0.60	0.50	0.60	0.50	0.60	0.50	0.60	0.50	10.80
1.0	1.0	Secretary	2.00	4.50	4.40	1.60	1.40	1.10	1.00	1.00	1.50	1.20	1.50	1.10	1.50	1.10	1.50	1.10	26.10
0.1	0.1	Data Clerk	0.20	0.45	0.45	0.20	0.20	0.10	0.10	0.10	0.20	0.10	0.20	0.10	0.20	0.10	0.20	0.10	2.80
0.3	0.3	Pharmacist	0.60	1.35	0.90	0.50	0.40	0.40	0.30	0.30	0.50	0.40	0.50	0.33	0.50	0.36	0.50	0.36	7.74
<b>201</b>																			

\* From 'Standards of Care' Cystic Fibrosis Trust (UK)

+ Median Figure used for calculating totals

NB: Those Centres in Dublin which are designated 'Tertiary Centres' will require a level of staffing above the countrywide norm, because of their high proportion of patients with highly complex needs



## FUNDING

Critical to the full development of the desirable reformation of the Cystic Fibrosis service, is an adequate financial and funding framework.

Cystic Fibrosis is a small discipline; as such it is particularly vulnerable to vagaries of funding.

Also, because of the constantly changing pattern of patient care within the system (as the patient moves from childhood to adulthood), and from relatively uncomplicated to immensely complicated stages of treatment, constant responsive adjustments to its budget are required.

This problem has been successfully addressed within the UK, by the development by The Department of Health of a banding system, embracing five bands of increasing severity – and thus increasing costs.

This system has the great advantage that, by its use, the budget for each year is set at the beginning of the year with direct reference to specific patient needs, thus making absolutely clear, to purchasers and providers, the responsibilities and requirements throughout the year.

The system was introduced in the UK ten years ago. It has been operational in Northern Ireland for the last nine years with very considerable success. A paper describing the operation of the system is attached as Appendix 5.

## ESTABLISHMENT AND MAINTENANCE OF STANDARDS

The proposals in this paper for the definition of a new shape for CF services based on a smaller number of appropriately geographically distributed Centres, giving equal provision to Adults and Children, and staffed in accordance with accepted best practice, create the climate for the development and delivery of an international class service for Ireland.

The framework, however, will not, of itself, generate and maintain the standards of excellence, and the improvements in survival, which are the goals. To achieve this it will be necessary to establish a system of accreditation for the centres.

All centres should be subject to regular accreditation through a structured review process as happens in North America. Each Centre should be visited and accredited on a regular basis (e.g. every six years) by an Accreditation Committee comprising senior clinician, Department of Health and Children officials and representatives of the CF Association.

For this process to be effective, and for the proper monitoring of progress and standards throughout the country, it is essential that the work of The CF Association Registry of Ireland be continued and developed.



## FURTHER FUTURE ACTIONS TO IMPROVE OUTCOMES

Having considered the logistics of developing a pattern of centre-based care, we now turn to the issue of neonatal screening.

The debate on this has been lengthy and it has been extensively summarised in Littlewoods monograph (1997) for the Cystic Fibrosis Trust (UK).

The position now is dominated by the output from the randomised controlled trial at the University of Wisconsin (Farrell et al 2001)<sup>11</sup> – the evidence from which becomes stronger with each succeeding year – which has demonstrated that early diagnosis of Cystic Fibrosis through neonatal screening (combined with aggressive nutritional therapy) can result in significantly enhanced long term nutritional status, improved long term growth (and, by implication, improved survival).

Neonatal screening has the potential to improve outcomes by avoiding the period of decline in health status which accompanies delayed diagnosis. Early diagnosis, by contrast allows vigorous physiotherapy and dietetic measures to be introduced with measurably improved scores in body indices and pulmonary function.

The not unreasonable deduction is that these benefits will in time be marked by improved survival (as suggested by the Boston/Toronto Study) (Corey et al 1988)<sup>12</sup>.

It is critical, however, that there is an appreciation of the fact that screening per se cannot achieve these benefits.

For success, it is **essential** that the child, identified through the screening process, should be enrolled in a care programme at an identified CF Centre and that the Centre be staffed so that every patient has the benefit of a full team appraisal in which all the identified and important disciplines (Medicine, Nursing, Physiotherapy, Dietetics and Social Work) are fully and properly represented and where there is ease of access to other required disciplines, e.g. Psychology.

A further development, essential for a fully comprehensive CF service, is the provision at each CF Centre, of a full Consultant Microbiology Service, which should be backed up by the provision of a Microbiology Reference Laboratory.

The benefits of creating a Reference Laboratory and the case for its development were set out in a submission to The Department of Health and Children by Dr. Philip Murphy of Tallaght Hospital in 1999. The beginnings of such a facility were facilitated at Tallaght by the Cystic Fibrosis Association but further development is now essential.

Extracts from Dr. Murphy's paper (The Background and Executive Summary) present a lucid and self explanatory case. They are attached as Appendix 6.



## **RECOMMENDATIONS FOR FUTURE DEVELOPMENT**

1. *Urgent action should be initiated to correct the dangerously inadequate staffing position in cystic fibrosis.*
2. *A small number of highly staffed, fully supported CF Centres should be designated to combine:-*
  - a) *Reasonable geographic access.*
  - b) *The expansion and consolidation of team expertise. This must include the availability of consultant Gastroenterology, Endocrinology and ENT staff with a special interest in cystic fibrosis.*
3. *The existing Dublin Centres should remain but the links between Adult and Children's Units should be strengthened and developed along geographical lines (e.g. Temple Street linking with Beaumont and Crumlin and Tallaght linking with St. Vincent's).*
4. *Tertiary Paediatric services should be designated.*
5. *Adult provision outside Dublin should be developed urgently to create a balanced service with broad geographical coverage. This would reduce the excessive burden on St. Vincent's.*

*St. Vincent's should retain its designation as the Adult Tertiary Centre.*
6. *All beds for cystic fibrosis patients should be in single rooms with ensuite toilet facilities to prevent the transmission of dangerous organisms.*
7. *A Microbiology Reference Laboratory should be established in Dublin to support and inform the Centres and to champion advances in knowledge and treatment. There should be a consultant microbiology service at each Centre.*
8. *A Neonatal Screening Programme should be established following the establishment of a logical pattern of joint child/adult CF Centres (so as to maximise the potential benefits of screening).*
9. *A structured, regular Accreditation Process should be developed, and form part of the funding methodology.*
10. *A new funding methodology (possibly based on banded 'packages of care') should be developed to create the stability required in the system.*
11. *The CF Registry of Ireland should be maintained and developed.*



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## ***APPENDICES***

1. Units Visited and People Interviewed
2. Present Distribution of CF Units
3. Proposed Distribution of CF Centres
4. Content of Routine Review and Annual Review at CF Centres
5. Practical aspects of providing banded packages of care for Adults with Cystic Fibrosis
6. Proposals for Reference Microbiology Facilities
7. Transition Documentation
8. Full names of the 13 locations identified in the Cystic Fibrosis Association Registry



## APPENDIX 1

UNITS VISITED	PEOPLE INTERVIEWED
<b>UK</b>	
Royal Brompton Hospital	Dr. A Bush Prof. M. Hodson Jackie Francis (CF Nurse)
Great Ormond Street	Dr. A. Jaffe Denise Sheehan (CF Nurse)  Dr. J. Littlewood } Sandra Kennedy } CF Trust (UK) Rosie Barnes }  Prof. S. Elborn (City Hospital Belfast)  Dr. S. Walters (Birmingham University)
<b>IRELAND</b>	
Crumlin Hospital	Dr. G. Canny
Beaumont Hospital	Prof. G. McElveney
Temple Street	Dr. D. Slattery Mary Morgan (CF Nurse)
St Vincent's Hospital	Dr. C. Gallagher Catherine Carroll } Josephine Doyle } CF Nurses Tara McMahon }
Tallaght Hospital	Dr. P. Greally Dr. P. Murphy
Cork University Hospital	Dr. C.P. Bredin Cathy Shortt } Clare Hayes } CF Nurses
University College Hospital, Galway	Prof. B. Loftus
Limerick	Dr. M. Mahoney Helen Caffey (CF Nurse)
Waterford	Dr. J. Gosgrove Anne Groggin (CF Nurse)
Drogheda	Mary Hanratty (CF Nurse)



## APPENDIX 2 & 3



### PRESENT DISTRIBUTION OF CF CENTRES

### PROPOSED DISTRIBUTION OF CF CENTRES





## APPENDIX 4

*(The material in this Appendix is drawn from the UK CF Trust's 'Standards of Care' document)*

### ROUTINE REVIEW AT THE SPECIALIST CF CENTRE

Regular routine reviews should take place ideally every six to eight weeks, and at least every three months.

They should comprise:-

- Measurement of weight and height (for children only).
- Culture of Sputum.
- Consultation and physical examination with an experienced CF specialist reviewing drug treatment and making changes as necessary.
- Spirometry.
- Chest X-ray (if there is a fall in lung function).
- Review by Physio of respiratory status in relation to current physical therapy including reviewing:-
  - Airway clearance techniques.
  - Inhaler and nebuliser techniques.
  - Review of exercise programmes.
- Review by dietician of nutritional status including:-
  - Assessment of dietary intake.
  - Dietary sufficiency/enzyme use.Also review of diabetic treatment and review of vitamin supplementation.
- Access to Social Worker, if required.

### ANNUAL REVIEW AT THE SPECIALIST CF CENTRE

This is a detailed assessment of every aspect of the patient's treatment and condition with a view to identifying sub-optimal treatment, and reformulating the patient's individual treatment plan.

The Review should include:-

- i) Detailed assessment of progress and review of the patient's and family's knowledge of the condition by an experienced doctor. If appropriate, fertility issues could be discussed.
- ii) Detailed assessment by an experienced Physiotherapist.
- iii) Review by the Psychologist or Social Worker.
- iv) Review by the CF Nurse Specialist.
- v) Full assessment by the Dietician. The need for further nutritional intervention should be considered, e.g. dietary supplements, enteral feeding by nasogastric tube or gastrostomy.



- vi) Clinical measurements. In addition to those done as part of a routine out-patient review (i.e. weight and height in children, weight in adults also height until growth has ceased, sputum culture, oximetry, spirometry) the following are usually included as part of the Annual Review:-
- Sputum culture for non-tuberculosis mycobacteria.
  - Chest X-ray score using a recognised scoring system, e.g. Northern CF X-ray Score or Chrispin Norman score.
  - Reversibility test to bronchodilator using either a nebuliser or inhaler and spacer device when appropriate. Changes are measured using the usual clinic spirometer.
  - When appropriate, more detailed lung function tests, depending on local availability, including lung volumes, CO transfer, exercise testing and challenge testing.
  - Blood gases in adults, if indicated, and only if the SaO<sub>2</sub> is less than 92% in air.
  - Full blood count, urea and electrolytes, liver function tests including serum albumin, clotting studies, fat soluble vitamin levels A, D and E also K if available. Aspergillus species RAST and precipitins, IgA, IgG, IgM and IgE. Venous blood gas, glucose and oral glucose tolerance test if >10 years of age.
  - Pseudomonas antibody levels, as an indication of chronic infection.
  - Assessment of the adequacy of intestinal absorption by a combination of clinical and laboratory methods. The latter include a semi-quantitative estimation of faecal fat, e.g. by microscopy or acid steatocrit. Estimation of 3-day faecal fat is useful, in addition to other relevant gastrointestinal investigations, if there are significant gastrointestinal problems.
  - Random or fasting blood glucose in all patients and some assessment of glucose metabolism in patients over 12 years or if clinically indicated. This is more reliably achieved by an oral glucose tolerance test. Patients with established diabetes should have the adequacy of their control measured by serum HbA<sub>1c</sub>.
  - Electrocardiogram if clinically indicated.
  - An assessment of liver status using a combination of one or more clinical, biochemical or imaging methods. Annual ultrasound of the liver and biliary system is recommended by some paediatric hepatologists and is now routine in many Specialist CF Centres.
  - Urinalysis for glucose, protein and blood.
- vii) Review of results and planning of future therapy and immunisations including annual influenza immunisation. Discussion with other members of the CF multi-disciplinary team.
- viii) Collection of data for audit and research. The results are entered into the Specialist CF Centre's database and also reported to the CF Association of Ireland Registry.

In addition, if it is the first visit or contact with the Specialist CF Centre:-

- The diagnosis is reviewed. It is advisable to repeat the sweat test if two CF mutations have not been identified.
- Genotype if not already done.
- Confirm pancreatic status by faecal pancreatic elastase 1; there is no need to stop the patient's pancreatic enzyme treatment.
- Introduction to CF team members; it is an opportunity to see facilities at the Specialist CF Centre.
- Provision of the CF management information from the Specialist CF Centre.
- Ensure patient knows of the Cystic Fibrosis Association.



## APPENDIX 5

# PRACTICAL ASPECTS OF PROVIDING BANDED PACKAGES FOR ADULTS WITH CYSTIC FIBROSIS

## Introduction

Care for adults with cystic fibrosis is very expensive. The disease is life-long and associated with new expensive treatments. The budget for a CF centre can be divided into the provision of intravenous antibiotics, staffing, stock and non-stock costs. The provision of intravenous antibiotics by the centre for home and domiciliary care usually comprises 50% of the budget.

Ten years ago a system was devised using a bottom-up approach to costing and is used in several units across the country. The work is published 'A cost description of an Adult Cystic Fibrosis Unit and Cost Analysis of the different categories of patients'. (Thorax 1992; pages 684-669.)

The bands are as follows:-

- Band 1:** Patients who come to outpatients, receive only outpatient care in terms of input from physiotherapists, doctors, social workers, dieticians etc.
- Band 2:** Patients who receive the above and in addition require outpatient intravenous antibiotics 3-4 times a year at home. They may be occasionally admitted to hospital. The input as an outpatient is greater than Band 1.
- Band 3:** Similar to 1 and 2 but essentially intravenous antibiotics are received as an inpatient 3-4 times a year. Outpatients require multi-disciplinary input.
- Band 4:** These patients have severe disease, come to hospital 3-4 times a year for intravenous antibiotics, have increasing disease severity. They may have diabetes and more resistant organisms. They may be under consideration for feeding gastronomies, transplantation.
- Band 5:** These patients have usually been in Band 4 for at least a year and need to stay in hospital for 4-6 months awaiting transplantation care or palliative care. They are unable to go home because of oxygen dependence, nocturnal ventilation, and feeding gastronomies and need intravenous antibiotics every day sometimes for 2-3 years. Patients life expectancy is usually no more than a year to 18 months.



## ILLUSTRATION OF THE BANDING SYSTEM

The way in which this banding system is operated within one NHS Trust is described below:

Cystic fibrosis is a disease that progresses slowly. Deterioration is slow and rapid change from one band to another occurs usually over years rather than within a year. On this basis the bands are revised at the beginning of each financial year by a small group of staff who know the patients best. This will usually comprise 2-3 doctors and a physiotherapist. Records are reviewed for the previous year, which are held on a database. Each patient is allocated a band from 1-5 at the beginning of each financial year. These numbers are then provided to the finance department and provider group. The increase to the costing each year is usually based on inflation. These costs are provided to the respective local health authority at the beginning of each financial year. New patients referred within the year are then placed on the CF database for the remainder of the year. Similarly, if a patient dies, the Health Authority is informed and the charge removed.

Commissioners on the whole like this system of costing they are aware of the volume and banding of patients, and they will be aware of the costs from last year and can have some idea of the projected costs for the next few years. Correspondingly the CF center can plan its care for the forthcoming year according to income received.

Each health authority will not have many patients. If they are uncertain about the revision of the banding then they can ask the CF Centre for the reasons why these changes have been made.

Bands 1-4 have been in existence for over ten years. Band 5 has only recently been added. It is clearly much more expensive but if the patients are assigned to this correctly it is highly unlikely that the purchasers will continue to pay large sums of money for a long period of time. Usually a patient in Band 5 will have received a transplant within the year or have died with that year.



## APPENDIX 6

# PROPOSALS FOR REFERENCE MICROBIOLOGY FACILITIES FOR PEOPLE WITH CYSTIC FIBROSIS – NOV 1999

## 1. Executive Summary

### Background

- Cystic Fibrosis, (CF) is the commonest lethal genetic condition in Caucasian populations and Ireland has the highest birth incidence of Cystic Fibrosis in the world. Resources comparable with other developed countries are therefore appropriate to support our CF population.
- As respiratory infection is the main clinical problem and leading cause of death in cystic fibrosis, the Microbiology support is particularly crucial to management.
- Routine microbiology laboratories usually struggle in both the detection and speciation of the bacteria responsible, and in the case of *B. cepacia* complex can not speciate the seven new species. The bacteria are commonly multiresistant, and often resistant to all available antibiotics, requiring multidrug antibiotic combination therapy.
- Most developed countries have a centralised reference laboratory service to confirm the identification of difficult organisms, molecular speciation of the *B. cepacia* genomovars and perform antibiotic combination sensitivity testing to these highly resistant bacteria. Routine microbiology laboratories do not perform these tests.
- The Faculty of Pathology of the Royal College of Physicians in Ireland has submitted a document to the DOH&C with respect to the establishment of a network of a Microbiology reference laboratories for Ireland. Three of these are now established but the proposal pre-dated the opening of the Tallaght laboratories.

### Health Gain

- Optimal detection of the *B. cepacia* complex and other multi-resistant bacteria will reduce the prevalence of these problem organisms in CF by facilitating proper infection control segregation practice. This is currently not possible in Ireland.
- Overall, it should be possible to reduce pulmonary exacerbations, multi-drug antibiotic resistance, hospital stay and add to the increasing life expectancy of patients with Cystic Fibrosis.
- Antibiotic synergy testing will facilitate optimal antibiotic combination therapy. High MIC sensitivity testing will direct nebulised antibiotic delivery.
- The laboratory could properly support CF patient assessments for the Lung Transplant Programme (Newcastle/Mater) and contribute to its efficiency.
- Quality epidemiological data integration with the CF register and antibiotic resistance data integration with NDSC for these highly resistant antibiotic bacteria.

## 2. Background

### 2.1 Clinical Background

Recent medical progress in the management of cystic fibrosis has been innovative, full of potential and life expectancies are improving. However, patients continue to suffer from recurrent and chronic respiratory infection and most of their problematic morbidity and mortality is due to pulmonary infection for much of their lives.



Pulmonary infection is a major complication in the management of patients with cystic fibrosis and a pulmonary trophism exists for particular bacterial species which have the ability to establish an environmental niche in the CF lung. Some species seem to have the ability to colonise only, with little contribution to pathology, while others are equipped with necessary virulence factors to cause very substantial pathology and in some cases a rapid and fulminant necrotic pneumonic process leading to terminal respiratory failure.

The acquisition of *Pseudomonas aeruginosa* or *Burkholderia cepacia* significantly affects pulmonary function and correlates with a poorer outcome.

Clinically, sputum microbiology can present several dilemmas:-

- Infection Vs colonisation – when to institute antibacterial therapy.
- Speciation of isolates – molecular speciation required for *B. cepacia* complex and related bacteria helpful in determining clinical significance of cultures.
- Polyclonal infection – co-existence of several strains of a single species.
- Differential antibiotic targeting of species or several clones of a species.
- Treatment Vs prophylaxis.
- Conventional sensitivity testing Vs high MIC breakpoint testing for nebulised antibiotic therapy.
- Synergistic combinations – when to use one, two or three potentially synergistic combinations.
- Dosage and route of administration – when to treat more aggressively with higher dosages of antibiotics or by inhalation.
- Infection control issued based on risk assessment – hospital and psychosocial implications of multi-drug resistant *P. aeruginosa* and *B. cepacia*.

## 2.2 Laboratory Background

There is a common assumption among many CF physicians of optimal quality from the analysis of respiratory secretion microbiology. However, a routine hospital Microbiology service can not meet all the clinical demands required for optimal care. Many non-routine technical issues can affect laboratory performance from selective liquid broth enrichment to PCR amplification of target pathogens and from conventional sensitivity testing to high MIC breakpoint testing for inhalation antibiotics and synergy testing. It is axiomatic in clinical microbiology that the harder a pathogen is looked for, the more it is found. The detection of CF pathogens is no exception.

The pathogens in the early stages of disease such as *Staphylococcus aureus*, *Haemophilus influenzae* and *Streptococcus pneumoniae* are easily managed in the routine microbiology laboratory. Confirmation of an MRSA can be problematic and certainly thymidine dependent *Staphylococcus aureus* (TDSA) will not be detected without special media and extra attention to routine protocols.

*Staphylococcus aureus* is the major pathogen of this group of early colonisers in the CF lung and surprisingly the conventional respiratory pathogens such as *Streptococcus pneumoniae* rarely cause chronic infection in CF. On the other hand the Gram negative non-fermenters such as *Pseudomonas aeruginosa*, and *Burkholderia cepacia* complex can present major difficulties in both isolation and identification and the clinical implications can be serious. The former *B. cepacia* is now recognised as seven new species which can only be differentiated using molecular methods not available in routine laboratories.

Antibiotic susceptibility testing for these pathogens is also particularly demanding as most patients are treated with antibiotic combinations in the hope that synergy may exist. However, this is rarely supported with laboratory evidence and the potential for antagonism between the chosen antibiotics is also real. Synergy MIC testing is not available in routine laboratories.



## APPENDIX 7

### *Transition Documentation*

#### **MOVING TO ADULT CARE**

Next year will be your transition year – a move from the Children's CF clinic to the Adult CF clinic.

We would like to try and answer some of your concerns about this move. Some of the questions asked by other teenagers and their parents who have made this move in the past include:

#### **Do I get a choice of where I want to go when I leave the Children's CF Clinic?**

Yes, the choice is entirely up to you – it usually depends on where you live. Many people choose the Adult CF Centre nearest to their home. The most popular choices are: The Brompton Adult CF Centre, Popworth Adult CF Centre in Cambridge, and Barts and the London CF Centre in east London. All these CF clinics have the same full CF teams as you are used to here, all of them carry out annual reviews similar to the ones you have had here and all have homecare nurses who will visit you at home if you need them. If you would like help making this decision please talk to Jackie Francis or Su Madge. We can contact the centre of your choice and arrange a visit for you. If you choose Popworth or Barts or London, and have spoken to Jackie or Su, you do not need to read the rest of this information sheet.

#### **Where is the Adult CF Clinic at the Brompton held?**

There are two consultants in the adult clinic, Professor Geddes has his clinic on a Monday afternoon in the outpatients department below the children's clinic and Professor Hodson has her clinics on a Tuesday afternoon and a Friday afternoon in outpatients west (just along the corridor).

#### **Do they do annual reviews?**

All annual reviews are carried out as a day case admission to Lind Ward (4th floor of the Fulham Wing) because, as you get older, there are a few extra tests that need to be carried out. If you would like more information about the annual reviews please ask Jackie or Su.

#### **Are my mum or dad allowed to come to clinic with me?**

Mums and Dads are allowed to come to clinic with you if that is what you and they want. No one will be asked to leave, but you – as an adult – will be asked what you want.



### Who are in the Adult CF team and how will we contact them?

Professor Duncan Geddes	Consultant	ext 8182
Professor Margaret Hodson	Consultant	ext 8041
Dr Khin Ma Gyi	Associate Specialist	ext 8009
Su Madge	Nurse Consultant	bleep 7032
Liz Bramwell	Homecare Nurse Specialist	07860 391642
Christine Hockings	Homecare Nurse Specialist	07801 745249
Penny Agent and Fran Alton	Physiotherapists	bleep 7302
Sarah Collins and Caryn Straker	Dieticians	bleep 7102
Steve Barton	Ward Manager	ext 8069
Sandra Scott	Research Nurse	bleep 0021
Juliana Burgess	Annual Review Coordinator	ext 4935
Alison Higgs	Social Worker	ext 4739
Carol Wingett	Welfare Rights Advisor	ext 4736
Nick Savage	Patient Advocate	cfadvocate@onetel.net.uk

### Where is the ward and what does it look like?

The ward is called Foulis Ward and is on the 3rd floor of the Fulham Wing. It has 25 single ensuite rooms and everyone with CF has priority for using these rooms.

### Will my treatment change?

The doctors will not change your treatment until they get to know you and find out what your needs are. However, as you get older you may need some things changed, some things stopped and some things added – all this will be in discussion with you.

We hope that we have answered any questions you may have had. If you have other worries please contact Jackie – bleep 1213 or Su – bleep 7032 and we will try to help you.

### Your transition checklist to keep and work through

Have you had a discussion about moving?

Have you had time to talk and ask questions fully?

Have you felt fully involved in the plans?

Have you met the adult team?

Have you seen the adult clinic?

Have you been to a transition clinic?

Do you know who the two key people for your transition are?

Have you been given any information booklets?





## APPENDIX 8

Full names of the 13 locations identified in the Cystic Fibrosis Association Registry

St Vincent's:	St Vincent's University Hospital, Dublin
Beaumont:	Beaumont Hospital, Dublin
Temple St:	The Children's University Hospital, Temple St, Dublin
Tallaght:	The National Children's Hospital, Tallaght, Dublin
Crumlin:	Our Lady's Hospital for Sick Children, Crumlin, Dublin
Waterford:	Waterford Regional Hospital
Cork:	Cork University Hospital
Tralee:	Tralee General Hospital
Limerick:	Limerick Mid Western Regional Hospital
Galway:	Galway University Hospital
Mayo:	Mayo General Hospital
Sligo:	Sligo General Hospital
Drogheda:	Our Lady of Lourdes Hospital, Drogheda



## CYSTIC FIBROSIS IN IRELAND SOME FACTS

*Ireland has the highest incidence of cystic fibrosis in the world. There are more than 1,100 cystic fibrosis patients (45% adults & 55% children) in the Republic of Ireland.*

*Cystic fibrosis is also Ireland's most common life-threatening genetically inherited disease affecting one in every 1,600 births.*

*Ireland displays a very much higher death rate than does England, Wales and Northern Ireland.*

*There is no cure for cystic fibrosis so patients rely heavily on the health system to manage their often severely debilitating symptoms.*

*Cystic fibrosis primarily affects the lungs and digestive system but can also damage other organs including the pancreas, liver and reproductive system.*

*Respiratory infections are the main clinical problems and leading cause of death.*

*Patients are prone to chest infections and malnutrition, and the absence of access to qualified staff and appropriate facilities leads to unnecessary visits to A&E units, for emergency relief, rather than specialists CF units.*

*Cystic fibrosis services are provided in 13 locations around the country.*

*Cystic fibrosis causes a build up of mucus in the lungs which leads to a cycle of lung infections and inflammation. The build up of mucus in the pancreas can also make it difficult to digest and absorb food.*

*The severity of the illness can vary and generally gets progressively worse with age and becomes increasingly more expensive to manage as symptoms become more acute.*

*At one end of the scale patients can be treated as out-patients with input from physiotherapists, doctors, dieticians, etc. while others, with more severe symptoms, must remain in hospital and require organ transplantation or palliative care.*

*Treatment includes consumption of pancreatic enzymes with food, daily chest physiotherapy, nutritional supplements and tube feeding, antibiotic treatment (including intravenous) and organ transplant.*

*The Cystic Fibrosis Association was founded in 1963, has 22 branches and 1,500 members. It provides assistance and support for people with cystic fibrosis and their families.*