

# **Services for People with Cystic Fibrosis in Ireland**

Conclusions of a Working Group established by the  
Health Service Executive

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## Executive Summary

In response to the Pollock Report which was published in 2005, the Health Service Executive established a Working Group with multi-disciplinary membership to undertake a wide-ranging review of the current infrastructure for CF in Ireland. The recommendations of the review endorse many aspects of the Pollock Report in identifying deficits in staffing levels and appropriate accommodation across the country which has not kept pace with the increase in the CF population.

Optimum care for people with CF as defined by the European Consensus document <sup>16</sup> is based upon multi-disciplinary care supervised by a specialist centre. It is clear that these requirements cannot be met in all of the hospitals currently providing CF care and that a new structure is required which coordinates:

- an appropriate level of care for patients at the most convenient location
- access to all of the specialist elements required for optimum outcomes
- appropriate transition between child and adult services
- information required to plan for a rapidly changing population which has ongoing service requirements
- access to care in an appropriate environment, e.g. facilities which enable infection control measures

The HSE Working Group concludes that the needs of the CF population would be best met by the following configuration of specialist cystic fibrosis centres offering either full care or supervision of structured shared care with satellite CF centres:

- Dublin North: Beaumont (adult) linked with Children's University Hospital, Temple Street (children)
- Dublin South: St Vincent's (adult) linked with a more closely integrated

OLCH, Crumlin / AMNCH, Tallaght service (children)  
*(pending the establishment of the new national children's hospital)*

- Cork: Cork University Hospital (children and adult)
- Limerick: Regional Hospital, Limerick (children and adult)
- Galway: Galway Regional Hospitals (children and adult)

The Working Group also recommends that Waterford Regional Hospital and Our Lady of Lourdes Hospital, Drogheda should provide shared paediatric care with a designated specialist centre (Our Lady's Children's Hospital, Crumlin and the Children's University Hospital, Temple Street respectively).

With the regard to adult patients, the group recommends that a Consultant Respiratory Physician with a special interest in Cystic Fibrosis be appointed to Waterford Regional Hospital with a view to the hospital developing as a CF specialist centre over time. Other units currently providing services may continue to do so on a shared care basis linked with a specialist unit.

The Working Group recommends that a tertiary clinical service linked with a national lung transplant programme should be formally designated for both adult and paediatric services. St. Vincent's University Hospital is suggested as the national referral centre for adult patients and Our Lady's Children's Hospital, Crumlin (pending the establishment of the new national children's hospital) as the national referral centre for paediatric patients due to the current availability of hepatobiliary, paediatric surgery and other specialist services required by a proportion of CF patients.

Other key recommendations of the report are as follows:

- The enhancement of staffing and accommodation to international guideline levels - proposed staffing requirements appropriate to the current number of patients are outlined in appendix 2.
- All services should be designed to minimise the risks of cross-infection by the adoption of a service control of infection policy.

- The establishment of a national CF reference laboratory should be formally designated as a priority.

# Chapter 1

## Introduction

### 1.1 Background

In February 2005, the Cystic Fibrosis Association of Ireland (CFAI) published a report entitled *Towards a Better Service*<sup>6</sup> (also known as the Pollock Report). This report summarised the findings of a study of hospital services in Ireland for people with cystic fibrosis and produced recommendations for service development involving the establishment of nine specialist cystic fibrosis centres across the country. This report was presented for consideration to the Health Service Executive (HSE).

### 1.2 The HSE Working Group

In April 2005, a Working Group (see Appendix 1 for membership) was established with the following terms of reference:

*To review the current configuration and delivery of services to CF patients in Ireland, across hospital and community, and to make recommendations for reconfiguration, improvement and development. The working group will consider the report prepared by Dr. Ronnie Pollock on behalf of the Cystic Fibrosis Association of Ireland and will encompass aspects of service alluded to but not covered in depth in that report including: CF services delivered in the community, the CF register, training and development of clinical staff, cost structures and cross border arrangements.*

*The output of the group will be an agreed, costed proposal for the development and reconfiguration of services for CF patients in Ireland with timescales. The proposal will form the basis of a bid for funding for implementation of the proposals through the Corporate Plan of the HSE.*

### 1.3 Information

Information was sought on the following areas to inform the work of the group:

- Review of literature with a focus on guideline / consensus documents on standards of care as the basis for development of a model of care suitable for the Irish health service context.
- Outcome and activity statistics for cystic fibrosis care
  - a. HIPE data
  - b. Central Statistics Office mortality data
  - c. CFAI CF Registry information
  - d. Census of CF service users
- Input from service providers on recommendations of the Pollock Report through the following channels:
  - a. Written submissions from current service providers and service managers through management structure of regional services.
  - b. Submissions from professional groups involved in cystic fibrosis care through the representatives on the Working Group:
    - i. Nurse Specialists in cystic fibrosis
    - ii. Dieticians
    - iii. Physiotherapists
    - iv. Medical and Scientific Council of Cystic Fibrosis Association
    - v. Microbiology Services (via questionnaire to laboratories)
- Views of voluntary organisations involved in advocacy for people with cystic fibrosis through representation on the Working Group, attending the annual conference of CFAI and meeting other groups not represented on the Working Group.
- Readiness and capacity of the current units delivering CF care to deliver the preferred model of care with current resources through structured telephone interviews with the senior clinician and/or nurse specialist in each unit.

## Chapter 2

### What is Cystic Fibrosis?

#### 2.1 Definition of Cystic Fibrosis

Cystic fibrosis (CF) is an inherited chronic disorder of the exocrine glands that affects the lungs and the digestion of food leading to frequent chest infections and under-nutrition. The disease causes progressive disability and early death and is characterised by a production of viscid mucous that obstructs the pancreatic ducts and bronchi, leading to infection and fibrosis. Difficulty breathing is the most common symptom and results from frequent lung infections.

#### 2.2 Epidemiology of Cystic Fibrosis

Cystic fibrosis is the most common life shortening, autosomal recessive disorder among people of European descent worldwide. CF is caused by a mutation in a gene called the cystic fibrosis transmembrane conductance regulator (CFTR). Only one working copy of the CFTR gene is required to prevent cystic fibrosis. Therefore affected people have an alteration in both copies of their CFTR gene; their carrier parents have an alteration in one copy. One in nineteen Irish people is a carrier, and approximately 35-40 children (one in 1,461) are born each year with cystic fibrosis (Cashman et al <sup>2</sup>). This frequency is higher than in other European populations (1 in 3,500 births) including Northern Ireland (1 in 1,850 births found by Hughes et al <sup>17</sup>) and England (1 in 2,500 births). A recent World Health Organisation (WHO) publication lists Ireland as the country with the highest known incidence of cystic fibrosis. Until recent decades CF was considered a disorder of childhood; improved survival rates now mean that almost 50% of people with CF (PWCF) on the CF register<sup>a</sup> are over 18 years and it is estimated that by 2009 the number of adults will exceed the number of children.

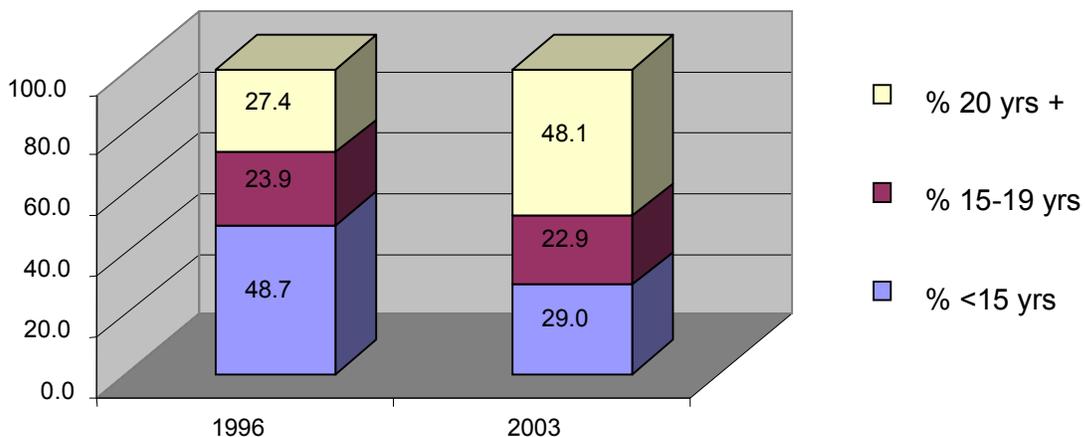
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<sup>a</sup> Cystic Fibrosis Registry of Ireland

### 2.3 Current Incidence of Cystic Fibrosis in Ireland

Prevalence of a disorder means the number of people of all ages in the population who have the condition at a given point in time in Ireland. At the end of 2005, there were a total of 1,094<sup>16</sup> cystic fibrosis patients in care; 520 of these were adults (47.5%) which is similar to other CF populations - in the US (41.8% in 2004) and Canada (47.6 % in 2002). The prevalence of the condition will increase as the rate of survival improves. The reason for this is that the number of children born with CF is likely to remain stable each year but the number of people who are living longer is increasing, resulting in an increase in the numbers of adult people needing services each year. This trend is borne out by the shift in hospital stay shown in Fig 2.1 towards an adult predominance over the 1996-2003 period. This increase in demand is particularly evident in the case of adult patients with an 11% increase in patient numbers in 2003 and 2004<sup>5</sup>. In addition, progression of the condition means that adults with CF are likely to develop more severe disease complications than children and to need a greater amount of in-patient intervention.

**Figure 2.1 - Change in the percentage of hospital bed-days by age-group 1996-2003**



Source: HIPE data, ESRI

## **2.4 Management of Cystic Fibrosis**

Improved survival rates depends upon a lifelong programme of:

- Preventing lung injury (with physiotherapy, oral, nebulised and intravenous antibiotics, anti-inflammatory and other medications, and effective infection control measures)
- Optimising nutrition (requiring intensive dietetic and psychological support)

## **2.5 Life Expectancy**

Survival rates for people with CF in this country will not be available until the national CF register is further developed. Although the median age of death (around the mid-twenties in 2003-4) is often used as a proxy for life expectancy, it is less reliable in small populations such as the population of Ireland. General trends however appear to be less favourable than our nearest neighbours in Northern Ireland where outcomes are better than the rest of the UK.

## **2.6 Cost and Funding of Care**

The cost of care for CF cannot readily be estimated for patients in Ireland which does not have a centralised cost and information structure such as exists in Northern Ireland. In countries where cost information is collected, it is evident that more than half of the costs relate to the use of specialist antibiotics. It can therefore be forecast that costs will rise steadily as the CF population ages and continues to increase and that these costs will constitute a significant cost pressure. In addition, many units currently depend on voluntary agency funding to support posts.

## Chapter 3

### Developing a Model of Care

Sound evidence exists from populations around the world (Nielson et al 1982<sup>23</sup>, Walters et al 1994<sup>26</sup>, Frederikson et al 1996<sup>12</sup>, Mahadeva et al 1998<sup>21</sup>) that centralised specialist care from a multi-disciplinary team markedly improves the quality and outcomes of care to cystic fibrosis patients.

A review of literature was undertaken with a focus on guideline or consensus documents relating to standards of care as the basis for developing a model of care suitable for the Irish health service context.

#### 3.1 International

The report *Services for Adults with Cystic Fibrosis - WHO/ICF(M)IACFA*<sup>30</sup> defined the range and extent of services to which an adult with cystic fibrosis should have access and emphasised the multi-disciplinary team approach at specialist centres.

#### 3.2 European guidelines

The European Consensus document by Kerem et al<sup>18</sup> is the most recent one to specify services to which a person with cystic fibrosis should have access (see Appendix 3).

The European Consensus accepts the need for agreed models of shared care in paediatric patients according to geographical need but specifies that a satellite CF unit in close liaison with a CF centre should have a minimum of 20 patients and input from a dietician, physiotherapist and a nurse, each with a special interest in CF who see patients at CF dedicated clinics. The centre team should perform the annual assessment and ultimate responsibility of care should rest with the CF centre Director. This is in contrast to the UK CF Trust guideline.

### 3.3 United Kingdom

UK guidance on the levels of professional expertise and the facilities required to provide CF care was updated in 2001 in the *Standards of Care* document<sup>8</sup> produced by the Clinical Standards and Accreditation Group of the Cystic Fibrosis Trust. This document recognizes two levels of CF care where patients receive their care:

- a) specialist CF centre, usually in a teaching hospital with over 100 patients
- b) CF clinic, in a local general hospital with smaller numbers of patients working with a specialist CF centre on a shared care basis.

The recommendations regarding CF staffing levels are included in Appendix 4 (a) of this report.

The CF Trust guidance acknowledges potential difficulties in the provision of shared care, particularly for adults where expertise in adult requirements does not exist in many general hospitals to the same extent as it does for children<sup>b</sup>. However, it provides a constructive framework (see Appendix 4 (b)) for suggested models of shared care as a means of providing specialist care for large numbers of patients whilst minimizing travel for patients and improving local expertise and community support.

### 3.4 United States

In the context of a country with a diversity of sources of healthcare funding which are mainly private, the Cystic Fibrosis Foundation (CFF) have sought to maintain standards of care through control of the accreditation process. They have produced *Clinical Practice Guidelines* (1997) and more recently the *Cystic Fibrosis Adult Care Consensus Conference Report* (2004) that strongly recommends a multi-disciplinary approach based on the strong association between the establishment of comprehensive CF care centres and improved patient outcomes.

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<sup>b</sup> Definition of paediatric for purposes of this report is under 18 - as per Children's Act 2001

### **3.5 Canada**

The Canadian Cystic Fibrosis Foundation (CCFF) adopts a different approach to ensuring standard compliance through its Clinic Incentive Grant Programme which provides supplementary funding based upon the number of patients served. Basic clinical care is the responsibility of each state government; the CCFF promotes quality assurance and consistency of care across Canada through their Clinic Site Visit Programme. By accepting a Clinic Incentive Grant each clinic agrees to participate in the Site Visit Programme that involves a full-day visit by two nominated site reviewers on a four to seven yearly cycle covering every aspect of care. Clinical data from each of the 38 CF clinics and 5 lung transplant programmes is also fed into the CCFF Registry facilitating audit.

### **3.6 Ireland**

#### **3.6.1 Newborn Cystic Fibrosis Screening**

In March 1999, the Chief Medical Officer at the Department of Health and Children, Dr. Jim Kiely, established a working group to “examine and make recommendations regarding routine newborn screening for cystic fibrosis. The group produced *The Interim Report of the Working Group on Newborn Cystic Fibrosis Screening* in April 2000. This report recommended the establishment of newborn screening for CF, and a group to plan implementation. The report noted: *“the successful implementation of this screening programme will be contingent on quality standards, treatment protocols, and further development of specialist centres for management of CF”*. As a result, the Programme of Action for Children established a Working Group on Newborn Cystic Fibrosis Screening which reported in March 2004<sup>22</sup> with recommendations for a two-tier programme integrated with the existing Newborn Screening Programme (“the heel prick”) and produced a detailed costed proposal (see Appendix 5). The report also recommended the development of a small number of specialist centres, adequately resourced to ensure optimal outcome. These recommendations were similar to those of the Pollock Report.

### 3.6.2 National Needs Assessment

The report by Dr. R. Pollock<sup>6</sup> commissioned by the Cystic Fibrosis Association of Ireland drew attention to the following issues:

- the large number of units in the context of the overall population
- the number of centres with very small patient numbers
- the imbalance between child and adult service provision
- staffing deficits
- variation in the suitability of physical accommodation with particular reference to infection control

The Pollock Report proposed:

- a) Centralised care at a smaller number of specialist units, five of which would be in Dublin (2 adult and 3 children's) and four outside Dublin providing adult and children's services.
- b) Increased provision of care (including transition) for the growing numbers of adults.
- c) Designation of tertiary service centres for adults and children.
- d) Facilitation of cross-border access to services in Belfast / Derry for patients.
- e) Establishment of a microbiology reference laboratory In Dublin.
- f) Introduction of neonatal screening for cystic fibrosis.

The size and essential features of a specialist cystic fibrosis centre were defined using the staffing levels specified in the *Standards of Care* document produced by the UK CF Trust (see Appendix 4 (b)). The main differences were that it replaced the medical staff grade<sup>c</sup> sessions with consultant sessions and deferred consideration of a shared care model until the designated centres had been suitably resourced and were in a position to support shared care.

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<sup>c</sup> This grade does not exist at present in Ireland.

### **3.6.3 Regional Needs Assessment**

A review of service needs for adults with CF in the Mid-West conducted in 2000 highlighted the need for a regional adult CF service with a multi-disciplinary team and facilities based upon the UK CF Trust Guidelines due to:

- the increase in survival of this care group
- an international consensus that the major factor in increased survival is the coordinated management at specialised centres
- the availability of transplant as an option in management of end-stage disease

Five years after this report was produced, some elements have been implemented but the number of adults availing of services in the Mid-West has increased more rapidly than expected from 11 to 39.

The introduction in 1999 of a new model of physiotherapy provision for children with CF in the Mid-West, integrating hospital and community services according to individually assessed need, was positively evaluated. The model has more recently been implemented in Galway.

### **3.7 Summary of International Evidence**

Although the methods of ensuring compliance with optimal standards of care may differ according to the funding structure in each country, international consensus exists on a model of care encompassing the following elements:

- The care of people with cystic fibrosis should be overseen by a specialist centre with sufficient patients to ensure ready access to a multi-disciplinary team, appropriate accommodation and facilities and the full range of services that patients may need. Where geographical circumstances dictate shared care programmes may be developed in a structured format where overall responsibility for outcomes remains with the CF specialist centre.

- Infection control considerations are central to the planning of service infrastructure.
- The increasing number of adult patients requires the development of adult services as a priority, with appropriate arrangements for transition.
- The number of patients attending a centre is an essential consideration. The UK standards require at least 100 patients and the European model 50 at a minimum for a specialist centre and a minimum of 20 patients for a satellite unit.
- The Working Group agreed that the *European Consensus on standards of care for patients with Cystic Fibrosis*<sup>18</sup> best addresses the needs for a model of care in Ireland and would provide a basis for international comparison of the range of services to which a person with cystic fibrosis should have access (see appendix 3).

## Chapter 4

### Outcome and Activity Statistics for Cystic Fibrosis Care

#### 4.1 HIPE Data 1996 - 2003

Patterns of hospital usage by patients across Ireland with a diagnosis of cystic fibrosis were obtained through the Economic and Social Research Institute (ESRI) from Hospital In-Patient Enquiry (HIPE) statistics.

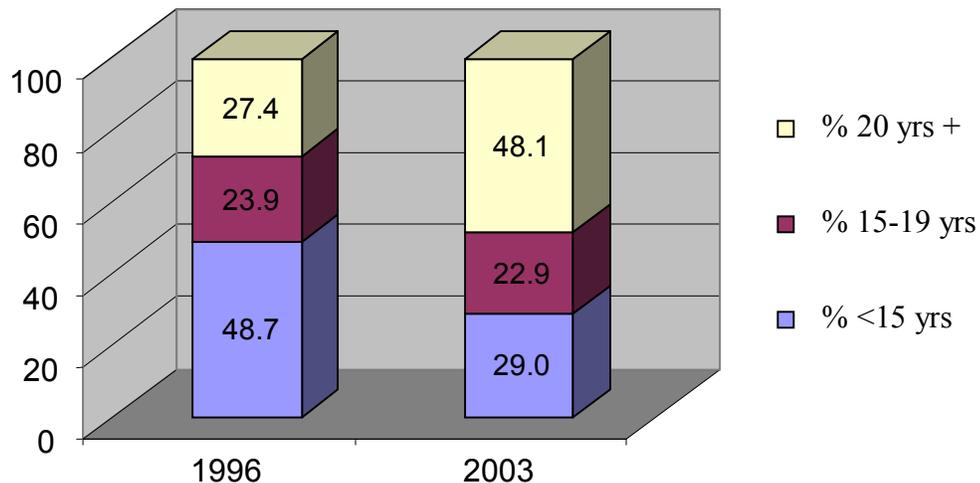
At a first glance this information does not indicate a significant change in hospital usage patterns over the eight year period ending on 31/12/2003: the number of in-patient episodes for each year was approximately 1,000 to 1,100 and the average length of stay was in the region of 10 days. However a four-fold increase in the number of day cases occurred between 1996 and 2003 leading to an increase of 45% in the total number of patient episodes. This reflects a shift in line with international trends to a more ambulatory form of care that remains hospital based.

**Table 4.1 - HIPE statistics for period 1996-2003**

| Ireland 1996-2003 |                     |           |                        |                        |          |
|-------------------|---------------------|-----------|------------------------|------------------------|----------|
| Year              | In-Patient Episodes | Day Cases | Total Patient Episodes | Average Length of Stay | Bed Days |
| 1996              | 1,036               | 157       | 1,193                  | 9.68147                | 10,030   |
| 1997              | 1,017               | 130       | 1,147                  | 9.49263                | 9,654    |
| 1998              | 1,021               | 119       | 1,140                  | 9.39373                | 9,591    |
| 1999              | 1,072               | 304       | 1,376                  | 9.70802                | 10,407   |
| 2000              | 1,162               | 531       | 1,693                  | 9.99139                | 11,610   |
| 2001              | 1,103               | 378       | 1,481                  | 10.3083                | 11,370   |
| 2002              | 1,043               | 425       | 1,468                  | 9.50623                | 9,915    |
| 2003              | 1,094               | 638       | 1,732                  | 10.6801                | 11,684   |

The age-spectrum of patients has also changed considerably over the 8 year time-span; in 1996 approximately half of all in-patient stays were by people under 15 but by 2003 this age-group accounted for only 29% as shown in Fig 4.1.

**Fig 4.1 - Change in percentage of hospital bed-days by age-group 1996-2003**



At the level of individual units, patient activity patterns varied widely as shown in Table 4.2 below. It is not possible for the ESRI to release figures for named hospitals without the consent of each hospital concerned but it is clear that out of the 30 units which provided in-patient services to patients with cystic fibrosis in 2003, three hospitals (one adult and two paediatric) were responsible for almost half of the admissions and almost two thirds of the bed days. Half of the hospitals (15) had less than 10 admissions per year.

Changing patterns of hospital usage were also evident in terms of location. Five units which accepted in-patients in 1996 had ceased to do so while 10 hospitals that had not done so in 1996 were now providing in-patient care. During 2003 75% of in-patient days were spent in hospitals in the Eastern Region although only 41% of patients were resident in the area.

**Table 4.2 - In-patient activity by unit in 2003**

| <b>Hospital</b> | <b>In-Patient</b> | <b>Day Cases</b> | <b>Total</b> | <b>Avg LOS</b> | <b>Bed Days</b> |
|-----------------|-------------------|------------------|--------------|----------------|-----------------|
| a               | 288               | 49               | 337          | 16.36          | 4,712           |
| b               | 157               | 25               | 182          | 9.87           | 1,549           |
| c               | 83                | 182              | 265          | 12.4           | 1,029           |
| d               | 78                | 107              | 185          | 7.44           | 580             |
| e               | 76                | 104              | 180          | 4.55           | 346             |
| f               | 73                | 4                | 77           | 6.66           | 486             |
| g               | 61                | 5                | 66           | 14.52          | 886             |
| h               | 55                | 2                | 57           | 9.27           | 510             |
| i               | 45                | 117              | 162          | 5.42           | 244             |
| j               | 38                | 4                | 42           | 6              | 228             |
| k               | 27                | 11               | 38           | 8.85           | 239             |
| l               | 18                | 5                | 23           | 5.44           | 98              |
| m               | 17                | 0                | 17           | 4.88           | 83              |
| n               | 15                | 3                | 18           | 6.2            | 93              |
| o               | 13                | 3                | 16           | 8.85           | 115             |
| p               | 9                 | 2                | 11           | 8.11           | 73              |
| q               | 8                 | 11               | 19           | 5.25           | 42              |
| r               | 7                 | 1                | 8            | 20.29          | 142             |
| s               | 4                 | 0                | 4            | 7.25           | 29              |
| t               | 4                 | 0                | 4            | 9.75           | 39              |
| u               | 3                 | 0                | 3            | 14.67          | 44              |
| v               | 3                 | 0                | 3            | 21.33          | 64              |
| w               | 3                 | 0                | 3            | 6.67           | 20              |
| x               | 2                 | 1                | 3            | 4              | 8               |
| y               | 2                 | 1                | 3            | 1              | 2               |
| z               | 2                 | 0                | 2            | 2.5            | 5               |
| aa              | 1                 | 0                | 1            | 1              | 1               |
| ab              | 1                 | 0                | 1            | 15             | 15              |
| ac              | 1                 | 0                | 1            | 2              | 2               |
| ad              | 0                 | 1                | 1            | -              | -               |
| <b>Total</b>    | <b>1,094</b>      | <b>638</b>       | <b>1,732</b> | <b>-</b>       | <b>11,684</b>   |

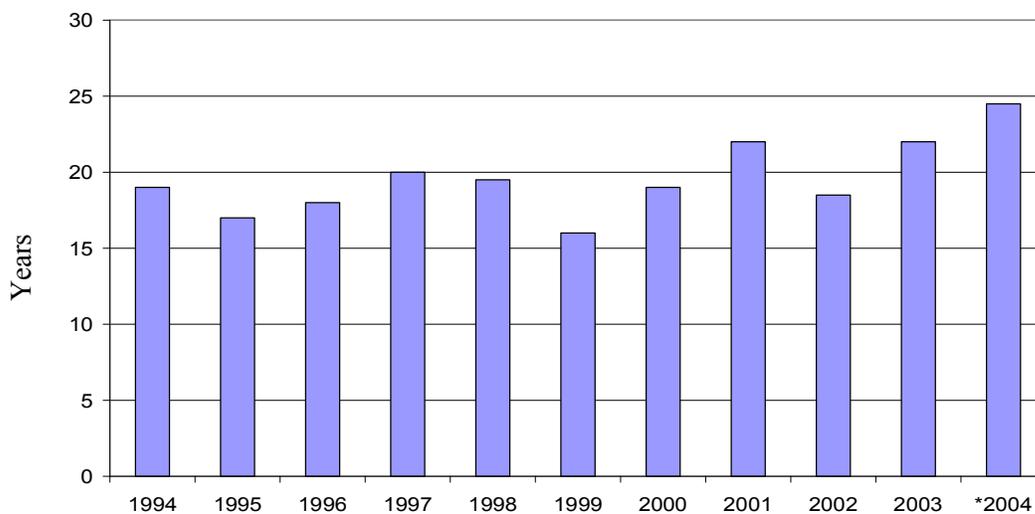
Source: HIPE, ESRI

## 4.2 Central Statistics Office Mortality Data

The prognosis for patients with cystic fibrosis has improved substantially over recent decades leading to an increase in survival across the world. It will not be possible to measure this in Ireland until the Cystic Fibrosis Register is further developed. However, the significant shift towards an increasing proportion of in-patient care for adults as shown in Fig 4.1 suggests this trend is also occurring in Ireland. In the interim, median age at death, which is readily accessible from mortality statistics, is commonly used as an indirect marker of current survival levels which can indicate trends over time. This information is illustrated in Fig 4.2; however, there are 2 reasons for caution in using these figures as follows:

- In smaller countries such as Ireland where the absolute number of deaths caused by cystic fibrosis each year is low, often in single figures, annual estimates of median age at death are unstable.
- In a disease such as cystic fibrosis for which life expectancy is constantly increasing, median age at death will invariably underestimate median survival.

**Fig 4.2 - Median age of death from CF (Ireland 1994-2004)**



\* Q 1-3 data only

### **4.3 Cystic Fibrosis Association of Ireland Information**

#### **4.3.1 Annual Census**

CF nurse specialists provide annual returns that are compiled to show the distribution and demographic characteristics of people with CF according to their attendance at CF centres for their annual assessment. The total of 1,182 at the end of 2005 composed of 543 adults and 639 children (<18 yrs): adult numbers had increased by 5% between 2004-2005 while paediatric numbers had a smaller increase of 2%.

#### **4.3.2 CF Registry**

The Cystic Fibrosis Registry was established by the Cystic Fibrosis Association of Ireland (CFAI) in 2001 with a grant from the Department of Health and Children. It began data collection in 2002 and by the end of 2005 had enrolled approx 50% of patients with CF attending centres in Ireland. The target enrolment figure is at least 75%. While this figure will not be reached for some time, the gender and age breakdown and geographical distribution of those enrolled to date suggests that it is representative of Irish patients and the information produced in recent annual reports is of value in assessing trends in levels of treatment and activity in Irish people with CF. The key findings from the 2004\* and 2005\*\* reports are as follows:

- Median age at diagnosis is approx 3 months \*\*
- 48% of those on the Registry also have had a sibling with CF \*
- 45% are aged 18+ and it is projected that this age group will make up half of the CF population by 2009 \*
- People aged 18+ are more likely to have an in-patient stay each year with an annual hospitalisation rate of 0.8 per PWCF compared to 0.5 for the paediatric group \*\*
- The rate of respiratory (chest) infections that required IV antibiotics was nearly three times higher in the adult group \*\* (1.4 per adult with CF compared to 0.5 for children)

- Adults have more complications than children with a rate of 4 per adult with CF for those assessed in 2005 in comparison to 2.2 per child under 18 \*\*
- Approx one in twenty five (4%) are without complications \*\*
- 10 categories account for over 90% of complications: pancreatic insufficiency, chronic pseudomonas infection, clubbing, osteopenia/osteoporosis, chronic staphylococcus infection, diabetes, abnormal liver function, DIOS (distal intestinal obstruction), nasal polyps, MRSA \*

We have seen from the HIPE data that in-patient hospitalisation rates have remained largely unchanged. Current health service information systems are not designed to reflect the substantial level of out-patient, ambulatory and home-based activity which has replaced this, however, the information gathered by the CF Registry can give some indication of the clinical service commitment which supports CF programmes.

- Culture analysis shows that the average number of microbiology specimens in 2005 was 5.3 for people with CF; approx 60% of these were sputum cultures
- Over 96% of children and 88% of adults with CF saw a dietician at annual assessment during 2005; although only 91% of children and 83% of adults are recorded as seeing a physiotherapist at the same assessment - data transfer may be an issue \*\*
- Most PWCF use more than one physiotherapy technique on a daily basis with “percussion” and “ PEP mask” used more frequently in childhood; “flutter” and “autogenic drainage” are used more commonly in adults \*\*
- The use of oral supplements, calorie supplements and supplemental feeding is more common in adults although almost all PWCF take both pancreatic enzymes and vitamins \*\*
- During 2005 a substantial proportion of children and adults received antibiotics orally, intravenously or by inhalation as shown in Table 4.3

**Table 4.3 - Route of Administration for Antibiotics for Patients on the CF Register in 2005**

|                 | <b>No. on CF Register with annual assessment</b> | <b>oral antibiotic</b> | <b>continuous oral antibiotics</b> | <b>hospital IV</b> | <b>home IV</b> | <b>inhaled antibiotics</b> |
|-----------------|--|------------------------|------------------------------------|--------------------|----------------|----------------------------|
| <b>Children</b> | 159  | 72 (45%)               | 16 (10%)                           | 21 (13%)           | 4 (3%)         | 64 (40%)                   |
| <b>Adults</b>   | 53   | 20 (38%)               | 9 (17%)                            | 10 (19%)           | 11 (21%)       | 35 (66%)                   |

Source: CF Register

Although the proportion of adults and children receiving both oral antibiotics and by the IV route in hospital is broadly similar, adult use of nebulised antibiotics and home IVs is much greater. The actual number of days per year of IV treatment is also considerably more for adults (16.2 days per adult) than for children (5.4 days per child). This information demonstrates the increased level of intervention required in the adult group and has implications for future service developments.

#### **4.4 Census of HSE CF Service Users in December 2005**

Accurate timely information on the complete population of people with cystic fibrosis (PWCF) by age-group and geographical area was a challenge as persons resident in Ireland do not have a unique identifier and there is no local area address coding (e.g. postcodes). The Working Group decided to construct a unique database of currently diagnosed PWCF accessing CF services in Ireland using patient initials, date of birth, and address area as identifiers.

##### **4.4.1 Method**

Cystic fibrosis centres in Ireland currently treating patients were contacted and provided the following details for each client: patient initials, date of birth, address area, together with main unit for treatment, secondary unit for treatment, referral unit and unit for annual assessment. To aid profile analysis each person was assigned to a county of residence and their age as at December 2005 was calculated. The data was then sorted by initials, date of birth and residence and

possible duplicate entries (where two centres claimed they were the main unit for the same patient) were extracted. These details were referred back to the cystic fibrosis nurse specialist at the relevant centres for confirmation. All identified duplicates were removed and within the constraints of the method the unique census was derived. This data was then used to provide a client profile by age, county and by treatment unit.

#### 4.4.2 Results

The total number of PWCF accessing specialist services in Ireland was determined at 1,094; 574 (52.5%) were aged less than 18 years and 520 (47.5%) were aged 18+ years.

1,229 records were assembled from the input of each unit, however, of these 135 were cited by more than one unit. 1,094 individuals were therefore identified using initials, county and DOB as identifiers. This is the best estimate of PWCF in Ireland in the absence of a unique identifier. Of the 1,094 individuals, 1 had an address in N. Ireland. Prevalence rates by county varied from 0.10 / 1000 in Donegal to 0.38 / 1000 in Tipperary with a national rate of 0.26 / 1000.

**Table 4.4 - Prevalence of PWCF by county of residence - December 2005**

| County   | Total Population | PWCF | Rate / 1000 Population |
|----------|------------------|------|------------------------|
| Carlow   | 50,471           | 14   | 0.28                   |
| Cavan    | 63,961           | 23   | 0.36                   |
| Clare    | 110,800          | 36   | 0.32                   |
| Cork     | 480,909          | 134  | 0.28                   |
| Donegal  | 146,956          | 15   | 0.10                   |
| Dublin   | 1,186,159        | 308  | 0.26                   |
| Galway   | 231,035          | 50   | 0.22                   |
| Kerry    | 139,616          | 41   | 0.29                   |
| Kildare  | 186,075          | 54   | 0.29                   |
| Kilkenny | 87,394           | 16   | 0.18                   |
| Laois    | 67,012           | 19   | 0.28                   |
| Leitrim  | 28,837           | 10   | 0.35                   |
| Limerick | 183,863          | 57   | 0.31                   |
| Longford | 34,361           | 4    | 0.12                   |
| Louth    | 110,894          | 28   | 0.25                   |
| Mayo     | 123,648          | 29   | 0.23                   |
| Meath    | 162,621          | 26   | 0.16                   |
| Monaghan | 55,816           | 17   | 0.30                   |
| Offaly   | 70,604           | 20   | 0.28                   |

|              |                  |              |             |
|--------------|------------------|--------------|-------------|
| Roscommon    | 58,700           | 7            | 0.12        |
| Sligo        | 60,863           | 19           | 0.31        |
| Tipperary    | 149,040          | 56           | 0.38        |
| Waterford    | 107,942          | 25           | 0.23        |
| Westmeath    | 79,403           | 20           | 0.25        |
| Wexford      | 131,615          | 27           | 0.21        |
| Wicklow      | 126,330          | 38           | 0.30        |
| <b>State</b> | <b>4,234,925</b> | <b>1,093</b> | <b>0.26</b> |

Source: HSE Census of CF Users (Dec 2005) and 2006 Census Figures (preliminary)

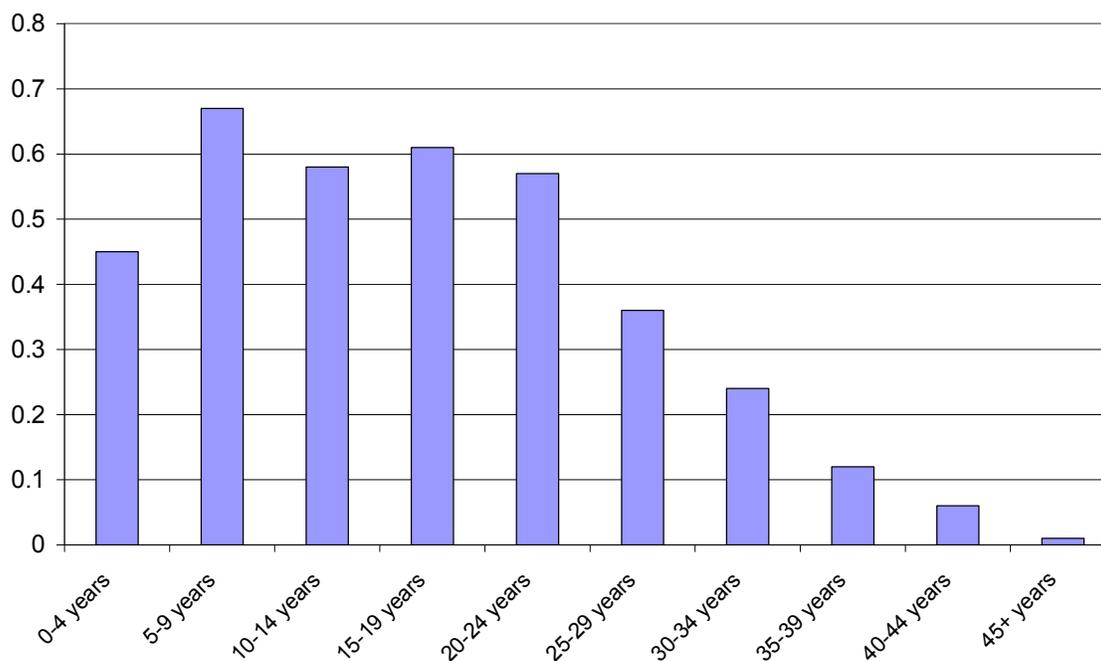
The age structure of the PWCF population using specialist services is central to future planning for CF services in Ireland; Table 4.5 illustrates that only 42.5% (465) are under 15 years of age. The Pollock Report <sup>6</sup> identified an urgent need to develop adult service provision, as it is primarily only available in Dublin at the present time.

**Table 4.5 - Age-structure of PWCF population**

| Age Group (years) | No. of Patients | % of PWCF Population |
|-------------------|-----------------|----------------------|
| 0-4               | 124             | 11.3                 |
| 5-9               | 176             | 16.1                 |
| 10-14             | 165             | 15.1                 |
| 15-19             | 192             | 17.6                 |
| 20-24             | 186             | 17.0                 |
| 25-29             | 112             | 10.2                 |
| 30-34             | 74              | 6.8                  |
| 35-39             | 36              | 3.3                  |
| 40-44             | 16              | 1.5                  |
| 45+               | 13              | 1.2                  |

Further analysis of age patterns as in Fig 4.3 shows that prevalence of CF increases by age-band during the first decade of life to a plateau that extends over the second and third decades. Delayed diagnosis patterns in a non-screened population may account for this picture, which may change with the proposed introduction of neonatal screening.

**Fig 4.3 – Prevalence of PWCF by 5-year age band in Ireland (rate per 1,000)**



Source: 2002 Census Figures (breakdown by age-group not yet available from 2006 Census)

#### **4.4.3 Shared care**

In addition to quantifying accurately the numbers of PWCF and their demographic profile, the census set out to quantify the proportion of CF patients whose care was shared by more than one unit; as shown in Fig 4.4. 13% (143) of PWCF have a major part of their care delivered by more than one unit with higher than average rates in the under 5 age group and in adolescence and early adult life. 8 individuals had a major input from 3 units. The reasons for this may include transitional arrangements from paediatric to adult care and delayed transfer because of absence of local adult services in some parts of the country; movement between centres during third level education may also be a factor. Referral for complex specialist services such as transplant assessment and management of complications influenced numbers attending more than one specialist unit.

**Fig 4.4 – Percentage of each age group managed by more than one unit**

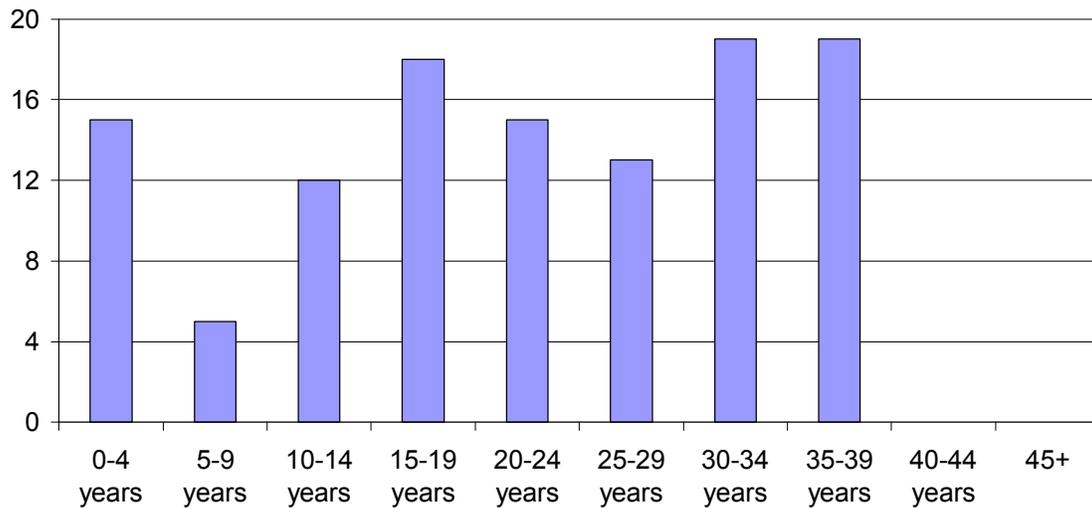


Table 4.6 lists the 10 centres in Ireland currently meeting the patient number requirements for CF specialist or satellite shared care centre as defined by the European Consensus report<sup>18</sup>. 93% (1,018) of PWCF obtain full care or a major part of their care (i.e. annual assessment) from one of these 10 units currently.

**Table 4.6 - Number of Patients Attending Centres**

| CF Treatment Centre | No. of patients on full care |          | No. of patients on shared care with major responsibility |          | Secondary Unit for PWCF | Total |
|---------------------|------------------------------|----------|--|----------|-------------------------|-------|
|                     | < 18 yrs                     | 18 yrs + | < 18 yrs   | 18 yrs + |                         |       |
| Beaumont            | 3                            | 44       | 0  | 10       | 6                       | 63    |
| Cork                | 76                           | 76       | 4  | 9        | 3                       | 168   |
| Crumlin             | 127                          | 3        | 5  | 1        | 24                      | 160   |
| Drogheda            | 17                           | 9        | 3  | 2        | 5                       | 36    |
| Galway              | 26                           | 20       | 1  | 1        | 3                       | 51    |
| Limerick            | 64                           | 25       | 6  | 6        | 2                       | 103   |
| SVUH                | 2                            | 235      | 1  | 45       | 30                      | 313   |
| Tallaght            | 74                           | 7        | 2  | 0        | 2                       | 85    |
| Temple St           | 79                           | 1        | 4  | 0        | 4                       | 88    |
| Waterford           | 28                           | 1        | 1  | 0        | 16                      | 46    |

## **4.5 Financial Information**

### **4.5.1 Staff**

Current staffing requirements can be ascertained from the Pollock Report <sup>6</sup> and consultation with hospitals. A number of posts are funded from sources other than the HSE (usually from charitable organisations) and often on a short-term basis. Many units without dedicated staff sessions rely on access to services which is more difficult to quantify precisely and thus cost.

The Working Group applied the staffing requirements as identified in the European model to the current numbers of CF patients at the various centres throughout the country. The result of this exercise indicates that there is an additional staffing requirement with an estimated cost of €10m to be implemented over three years (details attached at Appendix 2).

### **4.5.2 Drug Costs**

It was not possible to extract information on current medication costs incurred by treatment of cystic fibrosis in Ireland as no single cost-centre exists for CF unlike Northern Ireland where specialist drug costs representing one third of the total CF budget are held centrally by the Specialist Unit. In Ireland, drug costs are borne by each hospital for in-patients care, and a variety of systems cover out-patients costs including the long-term illness scheme, GMS cards, Drug Refund Scheme, health insurers and private prescription costs. Estimates produced by Dr. P. Murphy in AMNCH, Tallaght, Prof. G. Loftus in UCH, Galway and Prof. G. McElvanny in Beaumont Hospital have ranged between €25,000 - €30,000 annually for PWCF on maintenance antibiotics<sup>d</sup> who are likely to include the proportion of PWCF colonised with Pseudomonas estimated at approx. 50% in 2004 according to the CF Registry analysis.

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<sup>d</sup> These are drug acquisition costs only and do not include associated costs such as drug delivery systems e.g. IV giving sets, nebuliser costs, tobramycin drug assay monitoring costs, nursing / medical / pharmacist labour costs associated with drug delivery.

### 4.5.3 Capital costs

The need for infrastructural development is evident in many CF centres, particularly to enable appropriate management of infection control in both the inpatient and outpatient settings. The development of adequate single room, en-suite accommodation is a priority.

### 4.5.4 Funding Requirements

The HSE received €4.78m in the 2006 estimates process to commence implementation of interim recommendations of this report and an additional €2.0m in 2007.

**Table 4.7 - Strengths and Limitations of Current Data Sources for CF Services**

|                  | <b>Strengths</b>               | <b>Limitations</b>                       |
|------------------|--------------------------------|--|
| <b>Mortality</b> | Comparison over many years can | Limited value in any single year because |

|                               |   |   |
|-------------------------------|---|---|
| <b>Statistics</b>             | show trends   | of the small numbers involved in the Irish context  |
| <b>HIPE Data</b>              | Can provide trend-based information on patterns of attendance, balance between in-patient and day-case activity, changes in age-structure of care needs   | Does not reflect complexity of team based care, laboratory activity, increasing level of ambulatory care provided from a hospital base or activity in a community setting i.e. home IVs, community physiotherapy    |
| <b>CF Registry</b>            | Excellent database which has capacity to produce: <ul style="list-style-type: none"> <li>internationally comparable outcome measures including survival, pulmonary function, nutritional status</li> <li>access to treatment</li> <li>centre-based information which may be used for audit</li> </ul> | Not timely because of current system of manual collection of information from clinical records<br>Incomplete as database population not complete (50% at end of 2005)   |
| <b>CF Review Group Census</b> | Timely and complete<br>Accurate (omits duplicates)<br>Provides geographical distribution  | Snapshot only<br>Not derived from routine information system  |
| <b>Financial Information</b>  | Would be of use in planning service and monitoring trends as drug costs make up largest element of budget in Northern Ireland which has single cost-centre for CF   | No single cost-centre for CF – drug costs are borne by hospital for in-patients, and a variety of systems for out-patients incl. Long-Term illness Scheme, General Medical Services, High-Technology Medicines etc. |

## Chapter 5

### Comments on the Pollock Report

#### 5.1 Introduction to the Pollock Report

The Working Group agreed that key stakeholders - in particular the HSE Administrative Areas (former Health Boards and ERHA) and the constituent groups represented by the membership of the Working Group - should be invited to comment on the content and recommendations of *The Treatment of Cystic Fibrosis in Ireland: Problems and Solutions 2005*<sup>6</sup> more commonly referred to as the Pollock Report.

The Working Group agreed that each of those consulted would be asked to comment on how services in their areas were reflected in the report and to respond in line with the following framework:

- a) enhancement of adult cystic fibrosis services
- b) realignment of paediatric services
- c) screening
- d) reference laboratory
- e) information and registry

Responses were received between April and late July 2005. Respondents included Chief Officers of HSE Regions, HSE Hospital Network Managers, CEOs of voluntary hospitals, individual consultants and other parties. A synopsis of the responses are attached at Appendix 8.

## 5.2 Common Issues Raised

A number of common themes emerged in the responses. These are, *inter alia*:

- Agreement with the conclusions of the Pollock Report regarding existing deficits in service provision, staffing and infrastructure.
- Concern that the HSE CF Working Group move rapidly to address the deficits identified by the Pollock Report.
- An increase in patient awareness of deficits in the quality of service and related patient concern regarding their treatment in existing locations.
- Broad agreement with the recommendations of the Pollock Report regarding the need to establish specialist CF centres.
- Broad support for a variation on the service configuration to that outlined in the Pollock Report. Many of the submissions advocate the establishment of specialist CF centres which would provide regular outreach, support and diagnostic services to local CF centres.
- Some centres provided patient figures that were sufficiently different from those cited by Pollock to merit a re-examination of the report's recommendations regarding the future configuration of CF services.
- A minority of submissions queried the rationale behind some of the staffing recommendations in the Pollock Report. A number of centres also provided revised staffing figures and other amendments to the report.
- A number of centres highlighted the need for additional dedicated staffing, enhanced provision of existing associated services such as microbiology, pulmonary function, chemical pathology and radiology.
- Concern that the consultation process supporting the Pollock Report was inadequate and failed to reflect the viewpoints of a number of smaller, more geographically isolated centres and also that the membership of the HSE CF Working Group may not have addressed this issue either.
- Significant concern regarding the potential increase in patient travel time and rapid access to services associated with establishment of specialised centres and the associated transfer of services from smaller centres.

## Chapter 6

### Current Cystic Fibrosis Service Provision

#### 6.1 Model of Care

The Working Group agreed that the *European Consensus on standards of care for patients with Cystic Fibrosis*<sup>18</sup> best addressed the needs for a model of care in Ireland identified in the Pollock Report and would provide a basis for international comparison of the range of services to which a person with cystic fibrosis should have access.

#### 6.2 Staffing

The number of units currently offering services to patients was unchanged from the Pollock Report. The information on staffing levels presented in the Pollock Report indicated significant deficits in units across the country. The Working Group undertook to update this information (shown in Appendix 2) for the following reasons:

- Staff changes had occurred in the interim, including new consultant appointments in Sligo, Drogheda and Tallaght and retirements in Waterford and Cork.
- Voluntary funding of some additional posts through the Cystic Fibrosis Association of Ireland and the Hope Source Foundation for Cystic Fibrosis.
- Indications from some units that sessional commitments had reflected generic posts rather than dedicated CF sessions.

#### 6.3 Patterns of Shared Care

- Paediatricians in smaller units opt to share care with larger units that have expertise in respiratory medicine and access to paediatric surgery.

- Few patients in Cork, Limerick or Galway (children) are involved in shared care with another unit; where this occurs it is usually for transplant assessment, or a specific clinical problem.
- None of the Dublin units has exclusive shared care arrangements with any of the units outside Dublin which leads to a complicated matrix of linkages.
- Shared care arrangements are not formalized – they range from an annual assessment visit to a virtual transfer of care where a local service is in the process of change.
- In some instances students with a home base and a Dublin base e.g. Castlebar and Cavan may arrange IV treatment in their home hospital over weekends to avoid missing weekday commitments.
- No outreach services are provided by the Dublin units to the other units with which care is shared.
- Patterns of shared care are based upon professional rather than geographical linkages in response to availability of expertise.
- Tertiary referrals for specialist procedures are generally made to St. Vincent's for adults and Crumlin for children.
- Paediatric services outside Dublin are more willing to take on patients for either full or shared care because of the reduction in complication levels in this age group in recent years.
- Adult services have not developed in many locations outside Dublin due to the complexity of adult service provision.

#### **6.4 Working Arrangements**

Additional information on shared care, cover arrangements for staff absence and community supports was received from either the senior clinician or CF nurse specialist in each unit. The following facts were established:

- Cover arrangements for specialist staff are internal because of the specialised nature of the service and in many cases involve significant

- flexibility from the multi-disciplinary team in responding to unplanned patient queries where the relevant professional is away.
- Larger units where an individual physiotherapist, dietician or CF nurse specialist provides specialist team input are particularly vulnerable to gaps in service and in some instances funded sessions are used in providing cover rather than in additional service provision.
  - Pharmacy team commitment is at a low level; six units reported no pharmacy team input.
  - Psychology and social work input to multi-disciplinary teams is limited with only three units reporting dedicated psychology sessions and four reporting no social work input. (The CFAI funds emergency private counselling sessions when none is available).
  - In some instances specialist CF nurses may not be present at weekends.
  - No CF unit has a data clerk for database maintenance and audit leading to a limited capacity to audit local services.
  - Provision of home-based services is undertaken mainly by CF nurse specialists in supporting home IV treatment: in 2 areas (Mid-West region and Galway, domiciliary physiotherapy is organised through an integrated service supervised by the specialist centre. The CFAI funds and provides domiciliary physiotherapy services upon recommendation by the hospital consultant. (The CFAI reports that demand for and costs of this has doubled over the last year).
  - Consultant microbiology services are unevenly distributed across the country; the Limerick, Drogheda, and Mayo services have no access to consultant microbiology advice (a full-time consultant microbiologist took up post in Limerick in September 2005).
  - Although the numbers of consultant paediatricians who have specialist training in respiratory medicine are small (6), all of these are centrally involved in CF units. In comparison, there are many more adult respiratory physicians in post throughout the country but few are involved in CF care.

- This may reflect a training deficit and the competing needs of sarcoidosis, malignant, auto-immune, infection and tobacco-related disease in adults.
- Only two units have two consultants with a specialist level training in CF to provide internal cover.
  - Although five of the most specialized CF units are based in Dublin (two adult and three paediatric) no formal cross cover arrangements are in place between units.
  - No additional sessions are designated for tertiary unit functions in either adult or paediatric services.

## **6.5 European Consensus Document**

Significant elements of the European Consensus document <sup>18</sup> are listed in the boxes below and their application in the Irish context is discussed following each box.

### **6.5.1 Professional Linkages**

The model of care proposed by the European Consensus document describes the CF centre as having close links with consultants within the hospital or in nearby hospitals specialising in gastroenterology, hepatology, endocrinology, ear, nose, and throat (ENT) surgery, general, hepatobiliary and paediatric surgery, radiology, obstetrics and gynaecology (including assisted conception), infectious diseases and infection control, rheumatology, ophthalmology and nephrology.

- Only St. Vincent's, Beaumont and Crumlin are currently self-sufficient in the above disciplines other than for obstetrics and gynaecology which are provided in linked maternity units. The majority of recommended linkages are also in place in Galway and Cork with the exception of paediatric surgery; in Limerick a number of pending appointments will bring the service linkages to a similar level.

- All units have ready access to radiology, ENT surgery, ophthalmology, and nephrology. Approximately one in six people with CF is diagnosed with meconium ileus as an infant, a complication which requires transfer to a Dublin paediatric hospital where neonatal surgery is available. Most inter-hospital referrals for children are sent to Our Lady's Children's Hospital, Crumlin because of the availability of paediatric surgery, gastroenterology and hepatology. In the case of adults, most surgical procedures take place at a local level but the majority of referrals are to St. Vincent's University Hospital for hepatology and transplant assessment.

### **6.5.2 Protocol-based Management**

The CF centre will have effective referral and assessment protocols with a national transplant centre, written guidelines and facilities for the treatment of all complications of CF and direct access to the CF centre for telephone advice or emergencies.

- Written guidelines are used in all CF units although the source varies according to the location of training of the respiratory physician or paediatrician. Those in current use include the UK CF Trust guidelines, North American CF Foundation guidelines, Leeds guidelines and the European Consensus guidelines.
- All centres other than in Tralee report effective links with the national transplant centre.
- Direct access to the CF centre on a 24-hour basis without going through A&E is more likely in smaller units. In the Dublin-based units (other than Temple Street) and in Cork, and Limerick admissions go through the Day Ward from 9-5 on weekdays but through A&E out-of hours.

### 6.5.3 Accommodation

Appropriate in-patient accommodation will be provided in single rooms with ensuite facilities with out-patient visits taking place in a designated clinic allowing patient segregation according to infection category and sufficient rooms for members of the multi-disciplinary teams.

- Access to single rooms with ensuite facilities varies across the country.
- Hospitals providing CF care should incorporate appropriate accommodation into their capital plans.
- The HSE will examine the need for segregation / isolation capacity in the bed capacity review.

### 6.5.4 Facilities

The following facilities will be made available:

- a radiology department with CT scanning facilities
- expertise in bronchial artery embolisation for pulmonary haemorrhage
- a pulmonary function laboratory
- expertise in the placement of
  - totally implantable venous access devices
  - nasogastric and gastrostomy tubes
- a microbiology service expert in examining specimens from PWCF with established contacts with a CF microbiology reference laboratory
- a full diagnostic capability including reliable sweat testing and CFTR gene mutation analysis

- All units have access to a radiology department with CT scanning facilities.
- St. Vincent's, Beaumont and Galway for adults and Crumlin and Tallaght in the case of children provide access to bronchial artery embolisation.

- All units have access to pulmonary function testing although this varies between portable equipment operated by members of core team and fully equipped pulmonary function laboratories supporting research.
- Implantable venous access devices are inserted mainly by vascular surgeons or radiologists – for adults and older children this is usually carried out at a local level while younger children outside the Dublin area are usually sent to Crumlin for this procedure. The exceptions to this were Limerick (until the appointment of another vascular surgeon who took up post September 2005), Waterford, Mayo, Sligo and Drogheda where children of all ages are referred to Crumlin.
- Outside of Dublin, where all units have expertise in implanting gastrostomy tubes, Cork and Limerick undertake this procedure for adults and adolescents but refer younger children to Crumlin.
- St. Vincent's and Crumlin are self-sufficient for microbiology with exception of Cepacia speciation which is sent to Tallaght. The other hospitals refer generally to Tallaght with the exception of Drogheda which refers to Crumlin.
- Sweat testing is carried out in all units. Since the close of the facility in Galway, gene mutation analysis is undertaken only in Crumlin with the shortfall in service to some units being addressed by sending tests abroad.

## Chapter 7

### Microbiology Services for Cystic Fibrosis

#### 7.1 Requirements

Respiratory infection is the most common complication in people with cystic fibrosis. Microbiology support is therefore central to successful clinical management and is critical in making decisions about patient segregation and antibiotic management in the case of multi-resistant bacteria. Most routine microbiology laboratories are unable to provide the extensive range of reference procedures required as described in Table 7.1.

**Table 7.1 - Laboratory workload generated by CF service**

|  |
|--|
| <ul style="list-style-type: none"><li>• Culture and conventional susceptibilities</li><li>• Special susceptibilities<ul style="list-style-type: none"><li>- Inhalation susceptibilities</li><li>- Synergy FICs</li><li>- Bio film susceptibilities</li></ul></li><li>• Pseudomonas aeruginosa serology</li><li>• Aspergillus serology</li><li>• Mycobacteria culture<ul style="list-style-type: none"><li>- Routine</li><li>- Special CF Mycobacteria culture</li></ul></li><li>• B. Cepacia complex identification</li><li>• Non-cepacia Burkholderia identification</li><li>• Clonal analysis for CF centre epidemiology</li></ul> |
|--|

In recent years the trend in developed countries has been to refer a varying amount of work to other more specialised laboratories, usually at a national level. In some cases these reference laboratories have been funded by voluntary organisations such as the CFF funded laboratory service in the U.S. In other instances, the reference function forms part of a state-funded national laboratory service such as the Laboratory of Healthcare Associated Infection at Colindale,

London under the auspices of the Health Protection Agency for England and Wales.

The Pollock Report recommended that a microbiology reference laboratory should be established in Dublin to support and inform the centres and to champion advances in knowledge and treatment. A consultant microbiology service was also required at each centre.

## **7.2 Current Microbiology Services**

A questionnaire was issued under the auspices of the HSE Working Group to Irish laboratories supporting current CF centres. The questionnaire sought information on current workload, staffing, level of service locally and current need for reference work. The survey showed a spectrum of practice that is summarised in Appendix 6.

Although some laboratories also refer specimens to the Colindale, Central Public Health Laboratory in London (which has a turnaround time of up to 6 weeks and a cost per test of €250 per specimen) most of the laboratories supporting current CF centres refer the molecular speciation of *B. Cepacia* to the microbiology laboratory at Tallaght which has a dedicated interest in CF microbiology. In the absence of a designated national state-funded CF reference laboratory facility, the CFAI has provided short term funding to supplement existing staffing and the Hope Source Foundation has also provided additional resources for specialist equipment. This informal reference service is highly valued by the referring CF centres but is not a long-term solution due to the following factors:

- time-limited voluntary funding
- projected increases in workload because of enhanced survival of people with CF and development of the lung transplant programme will place additional pressures on existing staffing and result in overflow referrals to UK laboratories who are likely to prioritise their own increasing workload
- the WTE ceiling on posts will cause staffing difficulties in HSE services without a formal designation established with voluntary agency support

It is therefore recommended that:

- The Working Group on Reference Laboratory Facilities which has been established by the HSE, should consider the need for a designated reference facility service for cystic fibrosis as a priority.
- The existing expertise built up at the microbiology laboratory in Tallaght should be supported in the interim.
- Every CF centre should have a consultant microbiology service within the hospital with appropriate access to the national referral service.

### **7.3 Infection Control**

Infection control is especially vital to good CF care and must be carefully implemented at all levels both inside and outside the hospital. Consideration should be given to the standard risk groups and to the intermittent stage before chronic colonisation is recognised.

#### **a) inpatients:**

All hospitalised PWCF should be accommodated in single rooms of large air volume with en-suite facilities where possible and further cohorting of rooms if infrastructure allows. New buildings being commissioned should meet these standards. Where older centre infrastructure does not allow these standards, infection control should be optimised as far as is possible.

#### **b) out-patient attendances:**

Attendances at the clinic should be arranged to allow cohorting between the alert pathogen groups, (*Burkholderia cepacia* complex, *Pseudomonas aeruginosa* and *Staphylococcus aureus* particularly those of multidrug resistance) and as far as possible the more resistant pathogens at the end of the clinic. Cleaning and hygiene at the end of the clinic is also important especially if two consecutive clinics are held on the same day.

**c) laboratory:**

The accurate diagnosis and speciation of alert pathogens is especially essential in CF care. The use of the reference laboratory is essential in the speciation of *Burkholderia cepacia* complex bacteria, *Pseudomonas aeruginosa* serology for chronic colonisation status and clonal analysis within pathogen groups which should be performed at intervals decided by the team to identify the bacteria of high endemicity within a centre. A member of the microbiology staff should be an integral part of the CF team to advise on infection status and antibiotic susceptibility.

**d) community:**

Guidelines for out of hospital infection control should be promoted and adopted in the home, the workplace or school, for social activities and for PWCF group interaction. Appropriate guidelines can be found from the UK CF Trust ([www.cftrust.org.uk](http://www.cftrust.org.uk)), North American CF Foundation ([www.cff.org](http://www.cff.org)), and the CF Association of Ireland ([www.cfireland.ie](http://www.cfireland.ie)).

## Chapter 8

### Recommendations

#### 8.1 Overview

Although a consensus exists around the world about the model of appropriate care for people with CF, the configuration of service development varies according to the incidence levels, geography, training and health funding structures. The challenge for each country is to find a service framework which best meets the needs of service users within these constraints. The model of care which the Working Group recommends is the one adopted in the European Consensus document (see Appendix 3).

The Working Group has also made a number of recommendations based on the following principles:

- a) The clinical care of people with cystic fibrosis should be overseen by a specialist centre with sufficient patients to ensure provision of the range of services patients need. Where local geography requires some elements of care to be delivered by smaller general units or through community services this should happen under the supervision of the specialist team according to a structured shared care arrangement.
- b) At diagnosis, all patients should be referred to the most appropriate specialist centre. At this point they should be fully informed of all options available to them including:
  - the range of services available (to include voluntary, primary/community, local unit, specialist centre and tertiary centre)
  - the options for provision of full care through the specialist unit or shared care
  - other supports including genetic services
  - transition arrangements between paediatric and adult services
- c) Cross-border arrangements should be facilitated if this provides closer access to specialist care and is the preference of the person with CF. These would be to the Belfast City Hospital at present and possibly to Altnagelvin Hospital in Derry where a service is currently being developed.
- d) Tertiary centres for adults and paediatric patients should be identified due to the necessity for availability of hepatobiliary, paediatric surgery and other specialist services required by a proportion of CF patients.

## 8.2 Levels of Care

The group recommends three designated levels of care:

- National Referral Centre
- Specialist Centre
- Shared Care

### 8.2.1 National Referral Centres

The Working Group recommends that the national referral centres should be as follows:

**Paediatric:** Our Lady's Children's Hospital, Crumlin should be designated as the national referral centre for paediatric patients (pending the establishment of the new national children's hospital).

**Adult:** St. Vincent's University Hospital should be designated as the national referral centre for adult patients.

### 8.2.2 Specialist Centres

The Working Group recommends that specialist centres should be located as follows:

**Dublin North:** Beaumont (adult) linked with Temple Street (children) (pending the establishment of the new national children's hospital)

**Dublin South:** St Vincent's (adult) linked with OLCH, Crumlin (children) integrated with AMNCH, Tallaght (pending the establishment of the new national children's hospital)

**Cork:** (children and adult)

**Limerick:** (children and adult)

**Galway:** (children and adult)

The recommended configuration of Dublin centres takes account of the need for a clear structure of transition from paediatric to adult services.

### 8.2.3 Shared Care

**Waterford:** The Pollock Report recommendations acknowledged that Waterford Regional Hospital was a borderline situation but anticipated an increase in patient numbers. Because the number of paediatric patients has reduced, it is recommended at present that Waterford should provide

shared paediatric care with a designated specialist centre (Our Lady's Children's Hospital, Crumlin).

With the regard to adult patients, the group recommends that a Consultant Respiratory Physician with a special interest in Cystic Fibrosis be appointed to Waterford Regional Hospital with a view to the hospital developing as a CF specialist centre over time. The job description of the individual will provide for 5.5 sessions for CF to be reviewed as patient numbers increase. A mentoring programme with St. Vincent's University Hospital (nominally two sessions) will be initiated for the postholder. The 50:50 sessional split will also apply to on call arrangements.

**Drogheda:** It is recommended that Our Lady of Lourdes Hospital, Drogheda should provide shared paediatric care with a designated specialist centre (The Children's University Hospital, Temple Street).

**Other Units:** Other units currently providing services may continue to do so on a shared care basis linked with a specialist unit. In the case of patients opting for shared care, a structured package should be agreed between the participating specialist centre, the local unit and the person with CF. Shared care arrangements are discussed in more detail in Appendix 7.

### **8.3 Staffing**

Appendix 2 details the proposed staffing requirements appropriate to the current number of patients. It is recommended that these staffing levels be developed over a three-year period.

### **8.4 Physiotherapy**

Physiotherapy services should be hospital based within the specialist multi-disciplinary team with an integrated outreach service in the community.

## **8.5 Infection Control**

All services should be designed to minimise the risks of cross-infection by the adoption of a service control of infection policy to include:

- provision of home care for suitable patients (e.g. home IVs, domiciliary physiotherapy)
- stratification of out-patients by infection grouping
- availability of en-suite single rooms for inpatients
- 24-hour access to a CF centre with direct admission to CF unit (i.e. without having to be admitted through A&E)

A national reference laboratory should be formally designated as a priority. It is recommended that the service currently provided at AMNCH, Tallaght should be supported until the HSE Working Group on Reference Laboratory Facilities rules on the location of a designated reference facility service for cystic fibrosis.

The infection control policy should be overseen by the consultant microbiology service in each specialist centre with access to a national microbiology reference laboratory.

## **8.6 Evaluation of Care**

There is scope for development of a more sophisticated financial model which would provide improved accuracy of the cost of CF care for planning purposes in a 'band of care' model (as illustrated in the Pollock Report).

An approach to quality assurance/accreditation of all specialist units should be developed based on either accreditation or incentive programmes. Hospitals providing shared care should also be part of the accreditation process.

Information systems should be integrated to reflect the need for timely clinical, activity, outcome and cost information. The CFAI Registry should continue to be supported by the HSE and integrated with a computerised CF information system to be developed by the HSE to address the following needs:

- Clinical information for each individual unit to support local service audit and accreditation.
- Timely activity data for planning and service monitoring purposes.
- Costs of treatment to allow for banding packages of care.
- Information downloads to the CFAI Register capable of providing outcome information on survival, lung function (FEV 1), nutritional status, number of infections per year, quality of life measures (e.g. fertility in adults, employment) and symptom control.

## 8.7 Transplantation

Transplantation is an important element of CF care referred to but not covered in this report.

## 8.8 Training

The Working Group recommends that a training plan be developed which addresses:

- Future expansion in the client group due to increased survival.
- The need to provide training for all professionals who will be required to provide adult services in new locations.
- The need for training in specific procedures including short, medium and long term venous access and peg feeding.
- The potential for inclusion of CF modules on existing training programmes / rotations in adult and paediatric medicine, physiotherapy, dietetics etc.
- The dedicated funding of research fellowships in CF at MSc, MD and PhD level for doctors, nurses and allied health professionals through the Health Research Board

## 8.9 Neonatal Screening

A neonatal screening programme as described in the *Report of the Newborn Cystic Fibrosis Screening Working Group*<sup>22</sup> should be established as soon as services and infrastructure are in place to meet newly identified need.

## **8.10 Infrastructure**

The Working Group acknowledges the deficiencies in infrastructure in some centres providing CF services and recommends that the hospitals concerned prepare capital proposals that will enable the enhancement and expansion of services.

## **8.11 Financial Hardship**

The Cystic Fibrosis Association of Ireland reports that many CF patients and their families experience financial hardship in terms of:

- inconsistent interpretation of rules of allocation of medical cards in different parts of the country
- payment for GP visits (in the case of patients not eligible for medical card)
- hospital in-patient charges (CF patients frequently require in-patient treatment)

The Working Group recommends that the HSE should review the issue of variability in the allocation of medical cards and practices relating to consistency of application of hospital in-patient charges. The work of the Department of Health & Children on eligibility issues which is currently underway should update eligibility provisions and assist in providing clarification so that people will be clear about their entitlements.

## **Appendix 1**

### **Membership of the Working Group**

Ms. Louise McMahon, HSE, National Hospitals Office (Chair)

Mr. Denis O’Sullivan, Principal Officer, Secondary Care, Department of Health and Children

Dr. Tessa Grealley, Specialist in Public Health, HSE Mid-Western Region

Prof. Gerry Loftus, Paediatrician, University College Hospital, Galway

Professor McElvaney, Consultant Respiratory Physician, Beaumont Hospital

Dr Philip Murphy, Consultant Microbiologist, AMNCH, Tallaght

Ms Mary Hanratty-Woods, Hon. Sec. of the Cystic Nurse Specialist Group

Ms Hilary Colgan, Senior Dietician, Our Lady’s Children’s Hospital, Crumlin (Representing Allied Health Professionals)

Ms Fionnuala Duffy, Senior Commissioner, HSE Eastern Region

Ms Prya Prendergast Primary, Community and Continuing Care Directorate of HSE

Dr. Charles Gallagher, Consultant Respiratory Physician, St. Vincent’s University Hospital, and Chairperson of the Medical and Scientific Council of the Cystic Fibrosis Association

Mr. Godfrey Fletcher, Chief Executive, Cystic Fibrosis Association of Ireland

Mr Carl Rainey, Vice Chairperson, Cystic Fibrosis Association of Ireland

Ms. Mary McCarthy, Business Manager, Division of Internal Medicine, Cork University Hospital

Ms Suzanne O’Reilly, HSE, National Hospitals Office

## **Appendix 2**

### **Staffing Recommendations of the Working Group**

The recommendations below are based on the report *Standards of Care* document produced by the UK CF Trust. The report included recommendations for 2 types of Consultant depending on their level of commitment to CF care. The following recommendations do not make this distinction. It is envisaged that the Consultant Specialists in CF would be supported by other Consultant Respiratory Physicians within the hospital and that in the future, all Consultants dealing with CF patients would have previous specialist training in CF. As a Staff Grade does not exist in Ireland, the recommendations for this grade have been included in the Consultant Grade.

The recommendations of the UK CF Trust report provided a small range of staffing levels for Consultant 2 type posts and CF Nurses. The Working Group recommends 0.2 WTE Consultant post to represent the Consultant 2 type post and 1.5 WTE CF Nurses.

**Table (i) - Recommended staffing levels for specialist CF centres per 50 patients with CF**

| <b>Staff Member</b>           | <b>Paediatric</b> | <b>Adult</b> |
|-------------------------------|-------------------|--------------|
| Consultant 1                  | 0.7               | 0.8          |
| Consultant 2                  | 0.3               | 0.3          |
| CF SpR                        | 0.5               | 0.5          |
| CF Nurse                      | 1.5               |              |
| Physiotherapist               | 2.0               |              |
| Dietician                     | 0.4               |              |
| Social Worker                 | 0.4               |              |
| Psychologist                  | 0.4               |              |
| Secretary                     | 1.0               |              |
| Data Clerk                    | 0.1               |              |
| Pharmacist                    | 0.3               |              |
| Pulmonary Function Technician | 0.7               |              |
| Medical Scientist             | 0.7               |              |

**Note:** patients on shared care should be assigned a 50% allocation of those on full-care.  
Based on the number of paediatric, adult and shared care patients in the service at December 2005, the following table outlines the Working Group's recommended staffing levels for specialist cystic fibrosis centres:

**Table (ii) – Recommended Staffing Levels for each of the recommended specialist CF units**

| Staffing                              | Dublin / North East |                 | Dublin / Mid-Leinster |                                   |
|---------------------------------------|---------------------|-----------------|-----------------------|-----------------------------------|
|                                       | Beaumont            | Temple Street   | St. Vincent's         | Crumlin<br>incorporating Tallaght |
|                                       | adult only          | paediatric only | adult only            | paediatric only                   |
| Consultant1                           | 0.8                 | 1.2             | 4.1                   | 2.6                               |
| Consultant 2                          | 0.3                 | 0.5             | 1.6                   | 0.8                               |
| CF SpR                                | 0.8                 | 0.8             | 2.6                   | 1.3                               |
| CF Nurse                              | 1.5                 | 2.5             | 7.7                   | 5.3                               |
| Physiotherapist                       | 2.0                 | 3.3             | 10.0                  | 7.4                               |
| Dietician                             | 0.4                 | 0.7             | 2.0                   | 1.6                               |
| Social Worker                         | 0.4                 | 0.7             | 2.0                   | 1.6                               |
| Psychologist                          | 0.4                 | 0.7             | 2.0                   | 1.6                               |
| Secretary                             | 1.0                 | 1.7             | 5.0                   | 3.4                               |
| Data Clerk                            | 0.1                 | 0.2             | 0.5                   | 0.3                               |
| Pharmacist                            | 0.3                 | 0.5             | 1.0                   | 1.0                               |
| Pulm. Function<br>Technician          | 0.7                 | 1.2             | 3.6                   | 2.0                               |
| Medical Scientist                     | 0.7                 | 1.2             | 3.6                   | 2.0                               |
| <b>Recommended<br/>Staffing Total</b> | <b>14.3</b>         | <b>18.6</b>     | <b>46.4</b>           | <b>30.9</b>                       |

| Staffing                              | South       |      | West        |      |             |      | Total        |      |
|---------------------------------------|-------------|------|-------------|------|-------------|------|--------------|------|
|                                       | C.U.H.      |      | Galway      |      | Limerick    |      | adult        | paed |
|                                       | Adult       | paed | Adult       | paed | adult       | paed |              |      |
| Consultant 1                          | 1.3         | 1.0  | 1.0         | 0.5  | 0.5         | 1.0  | 7.7          | 6.3  |
| Consultant 2                          | 0.5         | 0.5  | 0.2         | 0.0  | 0.2         | 0.7  | 2.8          | 2.5  |
| CF SpR                                | 0.8         | 0.8  | 0.5         | 0.5  | 0.3         | 0.7  | 5.0          | 4.1  |
| CF Nurse                              | 4.7         |      | 1.35        |      | 3.0         |      | 26.05        |      |
| Physiotherapist                       | 6.3         |      | 1.8         |      | 4.0         |      | 34.8         |      |
| Dietician                             | 1.3         |      | 0.4         |      | 0.8         |      | 7.2          |      |
| Social Worker                         | 1.3         |      | 0.4         |      | 0.8         |      | 7.2          |      |
| Psychologist                          | 1.3         |      | 0.4         |      | 0.8         |      | 7.2          |      |
| Secretary                             | 3.0         |      | 1.0         |      | 2.0         |      | 17.7         |      |
| Data Clerk                            | 0.3         |      | 0.1         |      | 0.2         |      | 1.7          |      |
| Pharmacist                            | 1.0         |      | 0.3         |      | 0.6         |      | 4.7          |      |
| Pulm. Function<br>Technician          | 2.0         |      | 1.0         |      | 0.7         |      | 11.2         |      |
| Medical Scientist                     | 2.0         |      | 1.0         |      | 0.7         |      | 11.2         |      |
| <b>Recommended<br/>Staffing Total</b> | <b>28.0</b> |      | <b>10.5</b> |      | <b>17.0</b> |      | <b>128.4</b> |      |

**Table (iii) – Recommended Staffing Levels for each of the other recommended CF units**

| Staffing                          | South           | Dublin / North East | Overall Total<br>(specialist and CF units) |      |
|-----------------------------------|-----------------|---------------------|--|------|
|                                   | Waterford       | Drogheda            | adult                                      | paed |
|                                   | paediatric only | paediatric only     |  |      |
| Consultant 1                      | 0.3             | 0.2                 | 7.7  | 6.8  |
| Consultant 2                      | 0.0             | 0.0                 | 2.8  | 2.5  |
| CF SpR                            | 0.0             | 0.6                 | 5.0  | 4.1  |
| CF Nurse                          | 0.6             | 0.4                 | 27.25                                      |      |
| Physiotherapist                   | 1.2             | 1.0                 | 37.0                                       |      |
| Dietician                         | 0.2             | 0.2                 | 7.6  |      |
| Social Worker                     | 0.2             | 0.2                 | 7.6  |      |
| Psychologist                      | 0.2             | 0.2                 | 7.6  |      |
| Secretary                         | 0.3             | 0.3                 | 17.7                                       |      |
| Data Clerk                        | 0.0             | 0.0                 | 1.7  |      |
| Pharmacist                        | 0.1             | 0.1                 | 4.9  |      |
| Pulm. Function Technician         | 0.4             | 0.0                 | 11.6                                       |      |
| Medical Scientist                 | 0.4             | 0.                  | 11.6                                       |      |
| <b>Recommended Staffing Total</b> | <b>3.9</b>      | <b>2.8</b>          | <b>134.6</b>                               |      |

**Note:** these CF units will provide shared paediatric care with a designated specialist centre

The Working Group also recommends the appointment of Consultant Respiratory Physician with a special interest in Cystic Fibrosis to be appointed to Waterford Regional Hospital with a nominal 2 sessions per week at St. Vincent’s University Hospital to initiate a mentoring programme.

**Table (iv) – Current Staffing Levels in CF Specialist Centres**

| <b>Hospital</b>               | <b>Beaumont</b> | <b>Temple Street</b> | <b>St. Vincent's</b> | <b>Crumlin</b> | <b>Tallaght</b> | <b>CUH</b>       | <b>Galway</b> | <b>Limerick</b> |
|-------------------------------|-----------------|----------------------|----------------------|----------------|-----------------|------------------|---------------|-----------------|
| Consultant 1                  | 0.30            | 0.30                 | 0.60                 | 0.30           | 0.30            | 0.10             | 0.10          | 0.25            |
| Consultant 2                  | 0               | 0                    | 0                    | 0              | 0.30            | 0.20             | 0.01          | 0.10            |
| CF SpR / SHO                  | 0.30            | 0.20                 | 0.50                 | 0.40           | 1.00            | Rotational input | 0.10          | 0               |
| CF Nurse                      | 1.00            | 1.00                 | 3.00                 | 2.00           | 1.20            | 1.50             | 1.00          | 1.50            |
| Physiotherapist               | 0.40            | 1.00                 | 1.50                 | 0.90           | 0.50            | 0.50             | 1.00          | 1.50            |
| Dietician                     | 1.00            | 0.75                 | 1.00                 | 0.70           | 0.40            | 0                | 0.10          | 0.75            |
| Social Worker                 | 0.25            | 0.25                 | 0                    | 0.90           | 0.20            | 0.20             | 0             | 0               |
| Psychologist                  | 0.00            | 0.05                 | 1.00                 | 0.30           | 0.20            | 0                | 0             | 0               |
| Secretary                     | 0.40            | 0.20                 | 1.20                 | 0.40           | 1.00            | 1.00             | 0.05          | 0.50            |
| Data Clerk                    | 0               | 0                    | 0                    | 0              | 0               | 0                | 0             | 0               |
| Pharmacist                    | 0               | 0                    | 0.50                 | 0              | 0.20            | 0                | Access        | Access          |
| Pulmonary Function Technician | 0.30            | 0                    | 0                    | 0              | 0               | 0                | 0             | 0               |
| <b>Total</b>                  | <b>3.95</b>     | <b>3.75</b>          | <b>9.30</b>          | <b>5.90</b>    | <b>5.30</b>     | <b>3.50</b>      | <b>2.36</b>   | <b>4.60</b>     |

**In addition to the above, the Cystic Fibrosis of Ireland currently funds the following posts:**

**1 year – laboratory scientist – Crumlin – started January 2005**

**2 year – CF dietician – C.U.H. – started June / July 2005**

**2 year – CF Physiotherapist – C.U.H. – started August 2005**

**Table (v) - Number of CF patients being treated in CF units at December 2005**

| Hospital      | Total Patients on full care or on shared care with major responsibility |            |            | No of Patients on shared care with major responsibility |           |           | Secondary Unit for PWFC |           |           |
|---------------|---|------------|------------|---|-----------|-----------|-------------------------|-----------|-----------|
|               | Total   | Adult      | Paed       | Total   | Adult     | Paed      | Total                   | Adult     | Paed      |
| Beaumont      | 57  | 54         | 3          | 10  | 10        | -         | 6                       | 6         | -         |
| Bons Tralee   | 2   | -          | 2          | 1   | -         | 1         | -                       | -         | -         |
| Cavan         | 12  | 7          | 5          | 4   | 1         | 3         | 8                       | 5         | 3         |
| Crumlin       | 136   | 4          | 132        | 6   | 1         | 5         | 24                      | 9         | 15        |
| C.U.H.        | 165   | 85         | 80         | 13  | 9         | 4         | 3                       | 3         | -         |
| Drogheda      | 31  | 11         | 20         | 5   | 2         | 3         | 5                       | 2         | 3         |
| Galway        | 48  | 21         | 27         | 2   | 1         | 1         | 3                       | 3         | -         |
| Kerry         | 8   | -          | 8          | -   | -         | -         | 3                       | 3         | -         |
| Limerick      | 101   | 31         | 70         | 12  | 6         | 6         | 2                       | 2         | -         |
| Mater         | 2   | 2          | -          | -   | -         | -         | 1                       | 1         | -         |
| Mayo          | 22  | 10         | 12         | 2   | 1         | 1         | 7                       | 7         | -         |
| Mullingar     | 12  | 1          | 11         | -   | -         | -         | -                       | -         | -         |
| Portiuncula   | 1   | -          | 1          | -   | -         | -         | -                       | -         | -         |
| Portlaoise    | 1   | -          | 1          | -   | -         | -         | -                       | -         | -         |
| Sligo         | 13  | 4          | 9          | 4   | 3         | 1         | 4                       | 2         | 2         |
| St. Vincent's | 283   | 280        | 3          | 46  | 45        | 1         | 30                      | 26        | 4         |
| Tallaght      | 83  | 7          | 76         | 2   | -         | 2         | 2                       | -         | 2         |
| Temple St.    | 84  | 1          | 83         | 4   | -         | 4         | 4                       | 2         | 2         |
| Waterford     | 30  | 1          | 29         | 1   | -         | 1         | 16                      | 13        | 3         |
| Wexford       | 2   | -          | 2          | -   | -         | -         | -                       | -         | -         |
| No Main       | 1   | 1          | -          | -   | -         | -         | -                       | -         | -         |
| <b>Total</b>  | <b>1,094</b>  | <b>520</b> | <b>574</b> | <b>112</b>  | <b>79</b> | <b>33</b> | <b>118</b>              | <b>84</b> | <b>34</b> |

## **Appendix 3**

### **Recommendations of *Standards of Care for Patients with Cystic Fibrosis: a European Consensus* (Kerem et al, 2005) <sup>18</sup>**

A person with cystic fibrosis should have access to:

- A CF Centre caring for a minimum of 50 patients attached to a teaching hospital with a centre director who is an experienced CF physician working in close collaboration with at least one other physician knowledgeable in CF medicine and the following staff according to patient numbers quoting on the UK CF Trust recommendations:
  - specialist CF nurses
  - dieticians
  - physiotherapists
  - social workers
  - psychologists;
  - pharmacists
  - microbiologists
  
- A CF Centre which has close links with consultants within the hospital or in nearby hospitals specialising in gastroenterology, hepatology, endocrinology, surgery, ear, nose and throat (ENT) surgery, general, hepatobiliary and paediatric surgery, radiology, obstetrics and gynaecology (including assisted conception), infectious diseases and infection control, rheumatology, ophthalmology and nephrology.
  
- A CF Centre which has effective referral and assessment protocols with a national transplant centre, written guidelines and facilities for the treatment of all complications of CF and direct access to the CF Centre for telephone advice or for emergencies or other consultations.
  
- Appropriate in-patient accommodation provided in single rooms with en-suite facilities with out-patient visits taking place in a designated clinic allowing patient segregation according to infection category and sufficient rooms for members of the multi-disciplinary teams.

- A CF Centre with the following facilities:
  - a radiology department with CT scanning facilities
  - expertise in bronchial artery embolization for pulmonary haemorrhage
  - a pulmonary function laboratory
  - expertise in the placement of
    - totally implantable venous access devices,
    - nasogastric and gastrostomy tubes
  
- A microbiology service expert in examining specimens from people with CF with established contacts with a CF microbiology reference laboratory
  
- A full diagnostic capability including reliable sweat testing and CFTR gene mutation analysis.

The European Consensus accepts the need for agreed models of shared care according to geographical need but specifies that a satellite CF unit in close liaison with a CF centre should have a minimum of 20 patients and input from a dietician, physiotherapist and a nurse, each with a special interest in CF who see patients at CF dedicated clinics; the centre team should perform the Annual Assessment and ultimate responsibility of care should rest with the CF Centre Director.

## **Appendix 4**

### **Recommendations of *Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK* (UK Cystic Fibrosis Trust, 2001) <sup>7</sup>**

#### **(a) Staffing Recommendations (per 50 patients)**

| <b>Staff Member</b> | <b>Local Centre<br/>(&lt;50 patients)</b> | <b>Specialist Centre<br/>(paediatric)</b> | <b>Specialist Centre<br/>(adult)</b> |
|---------------------|---|---|--------------------------------------|
| Consultant 1        | 0.5                                       | 0.5                                       | 0.5                                  |
| Consultant 2        | Nil                                       | 0.2 – 0.3                                 | 0.2 – 0.3                            |
| Staff Grade         | Nil                                       | 0.4                                       | 0.6                                  |
| CF SpR              | Nil                                       | 0.5                                       | 0.5                                  |
| CF Nurse            | 1   | 1 – 1.5                                   | 1 – 1.5                              |
| Physiotherapist     | 1.0 – 2.0                                 | 2.0                                       | 2.0                                  |
| Dietician           | 0.4                                       | 0.4                                       | 0.4                                  |
| Social Worker       | 0.4                                       | 0.4                                       | 0.4                                  |
| Psychologist        | 0.4                                       | 0.4                                       | 0.4                                  |
| Secretary           | 0.5                                       | 1.0                                       | 1.0                                  |
| Data Clerk          | Nil                                       | 0.1                                       | 0.1                                  |
| Pharmacist          | 0.2                                       | 0.3                                       | 0.3                                  |

The above recommendations refer to the number of whole-time equivalent (WTE) staff required for every 50 patients on full care with a 50% allocation for those on shared care.

**(b) Arrangements for Shared Care**

It is recommended that shared care arrangements between a CF Clinic and a Specialist CF Centre are covered by an agreement setting out the arrangements and responsibilities along the following lines: -

- The frequency with which the CF Consultant and members of the multi-disciplinary CF team see the patient e.g. at least once and ideally twice a year.
- An Annual Review is performed (Section 3.5). It should be specified where the Annual Review would be done i.e. at the Specialist CF Centre or the local CF Clinic.
- The CF Consultant from the Specialist CF Centre sees the results of the Annual Review and is responsible for reporting the results to the UK Database.
- It is desirable that the CF Consultant should write to the patient/parents summarising the results of the Annual Review and detailing any recommendations arising from the results.
- Arrangements for telephone contact between the local CF Clinic Consultant and the CF Consultant are agreed.
- Communication is improved if copies of the clinic data sheets and letters from the local CF Clinic are sent to the CF Consultant and reviewed by him/her before filing at the Specialist CF Centre.
- Events requiring notification to, discussion with or referral to the Specialist CF Centre are agreed (suggestions are listed in this document, Section 3.10).
- Prescribing arrangements for drugs are agreed as to whether they are obtained from the Specialist CF Centre, the local CF Clinic or the General Practitioner.
- The most appropriate method of contact for the patient/parents to obtain advice should be specified. This would usually be the Consultant at the local CF Clinic but may be the CF Consultant when the local Consultant is on leave or otherwise absent, particularly if there is no local senior medical cover.
- Arrangements for regular clinical audit utilising data stored in the UK CF Database.
- The ultimate responsibility for the care of patients receiving shared care usually lies with the Consultant at their local CF Clinic (N.B.) but must be clearly defined in the agreement and made known to the patient/parents.

*(Note: numbering refers to CF Trust document)*

## **Appendix 5**

### **Executive Summary of *Implementation of Newborn Cystic Fibrosis Screening in the Republic of Ireland (March 2004)* <sup>22</sup>**

1. A two-tier IRT/DNA analysis screening programme for Cystic Fibrosis is recommended. It is estimated that approximately 250 infants would screen positive on the initial IRT test. Of these, 33 would have CF and 40 would be carriers for the disorder.
  
2. A steering group should be appointed in the initial phase to oversee and coordinate the implementation of the screening programme and the appropriate developments in clinical services, including treatment protocols. Membership of this group should include; representation from;  
  
The Department of Health and Children  
Each designated CF clinic (individuals from different professional backgrounds should be nominated e.g. Paediatrician, Neonatologist, Paediatric Gastroenterologist, CF Nurse, Dietician, Physiotherapist)  
The National Newborn Screening Laboratory, The Children's University Hospital, Temple Street  
The National Centre for Medical Genetics, Our Lady's Hospital for Sick Children, Crumlin  
The Programme of Action for Children  
Director of Nursing, Maternity unit  
Director of Public Health Nursing  
Irish College of General Practitioners  
Faculty of Public Health Medicine  
CF Registry  
CF Association of Ireland
  
3. Since CF screening will be integrated into the existing Newborn Screening Programme, overall responsibility for the CF screening programme would rest with the Steering Group with delegated responsibility to the Director of the NNSL.
  
4. A clinical Liaison officer, with responsibility for liaison regarding the Newborn Screening Programme, including CF screening should be appointed at the NNSL to support the programme.

5. Health promotion material for both parents and sample takers will need to be developed and circulated to all staff involved in supporting parents during the ante-natal and post-natal period. Culturally appropriate materials will be developed in consultation with support groups for Irish Travellers and other ethnic groups.
6. Written parental consent to screening of the infant for CF, will be requested, ideally during the ante-natal period. Since the heel prick blood spot sample will also be used for testing for all conditions currently included in the National Newborn Screening Programme, a new consent form will be designed to reflect this. It will state clearly that DNA testing may be carried as part of the CF screening process. The existing programme has virtually universal coverage and it is essential to ensure that introduction of any new screening procedure will not in any way undermine its success.
7. Samples for IRT testing will be collected, as part of newborn screening samples for the National Newborn Screening Programme (NNSP). These are collected when the infant is aged between 72-120 hours of age. Sample collectors will need to collect no additional blood, but should endeavour, as at present, to collect an adequate sample in the manner prescribed in the guidelines produced by the National Newborn Screening Laboratory (NNSL).
8. The existing newborn screening card (NSC), which may also be used for sample collection for IRT, is currently being redesigned to comply with best international practice and to accommodate space for recording of additional information.
9. A review of the National Newborn Screening Programme (NNSP) has recently been conducted. It is essential that the recommendations made in that review are adopted and implemented before the newborn CF screening programme is started. The review recommends provision of additional resources to support the NNSP. Once in place, these resources will facilitate delivery of the CF screening programme in the NNSL.
10. IRT testing will be carried out at the National Newborn Screening Laboratory (NNSL), Children's University Hospital, Temple Street.
11. DNA testing for CF will be carried out at the National Centre for Medical Genetics (NCMG) at Our Lady's Hospital for Sick Children, Crumlin. The original blood sample, which will have tested positive on IRT testing, will be passed on from the NNSL, Children's University Hospital, Temple Street.

12. The NNSL will be responsible for liaising with Paediatricians, General Practitioners and Community Care Services regarding results from the CF Screening Programme.
13. The Molecular Genetics Laboratory at the NCMG will evaluate the various test kits commercially available in the initial stages of the screening project, with a view to identifying the most suitable kit for the programme.
14. A mechanism needs to be put in place to deal with the anticipated increased demand for CF carrier testing. Due to the large number of carrier tests which the programme is expected to generate, a full-time Genetic Counsellor, based at the NCMG, will be required to support the programme.
15. Diagnostic sweat testing is currently being carried out at a number of centres throughout the country. In order to ensure high quality testing, sweat tests performed as part of the newborn CF screening programme should be performed in laboratories associated with designated or recognised CF centres.
16. A standardised pilocarpine sweat chloride test with measurement of sweat chloride should be adopted, as the conductivity method has yet to be recommended and adopted by the CF associations in the US and UK.
17. Additional resources, as outlined in detail in the text, will be required at the National Newborn Screening Laboratory and the National Centre for Medical Genetics at Our Lady's Hospital for Sick Children, Crumlin to support delivery of the programme.

**Estimated costs (at January 2004 prices) are:**

**Total Year 1 €654,000**

**Total Year 2 and recurring €392,000**

18. Additional 0.5 sweat test technicians will need to be appointed at the five designated sweat testing centres at Our Lady's Hospital for Sick Children, Crumlin, the Children's University Hospital, Temple Street, National Children's Hospital, Tallaght, University College Hospital, Galway and Waterford Regional Hospital at a **total cost of €87,500 for year one and recurring.**
19. Estimated cost of production of health information leaflets regarding the CF screening programme is **€70,000 for one million leaflets.**

20. Having considered various options for optimal service delivery, the working group recommends that seven centres as outlined on page 17 should be designated as specialist referral centres. Each designated centre would have a minimum throughput of at least 40 CF clients, and it is anticipated in the future that this number would increase to 100. This smaller number of dedicated centres should be fully resourced to provide outreach and shared care for the hospitals within their local network.
21. Since the Cystic Fibrosis Association of Ireland is currently carrying out a review of clinical CF services, including a comparison between existing and recommended staffing levels, the working group did not consider it appropriate to address staffing levels in this report.
22. The screening programme should be audited annually to facilitate quality assurance and comparison with other international screening programmes.
23. Uniform data collection procedures should be developed at all sites involved in the screening and treatment service to facilitate this audit.
24. Appropriate clerical and IT support will be required to ensure high quality data entry and data management.

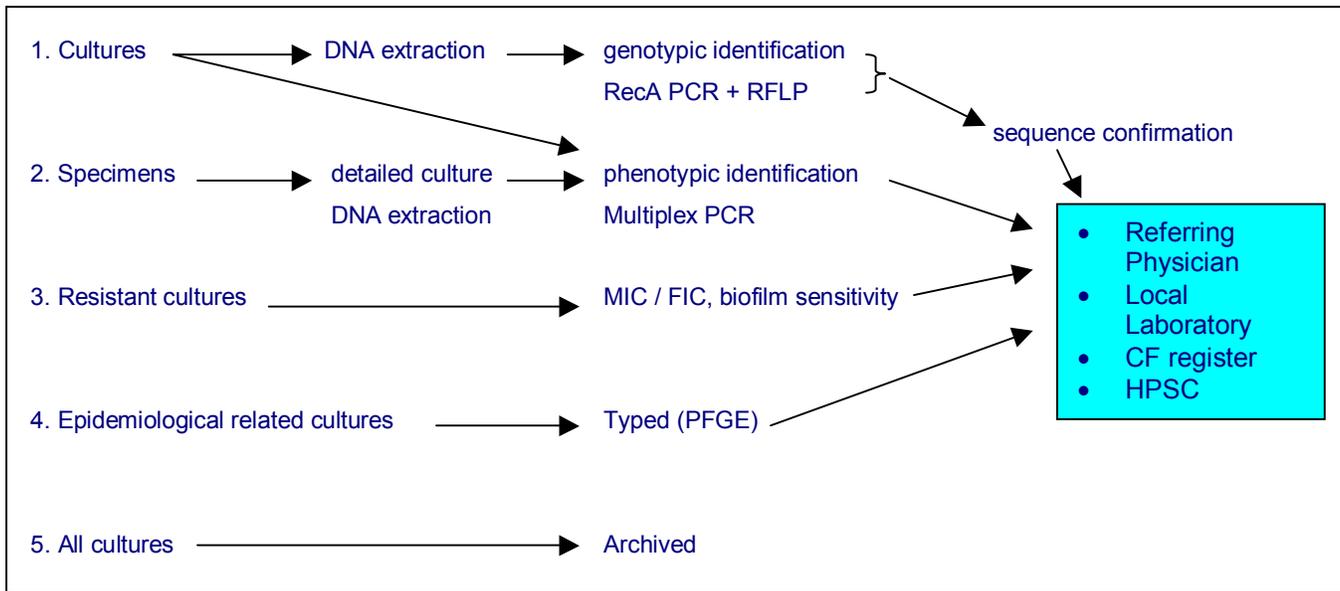
## Appendix 6

### Appropriate Management of Specimens and Infection Control

#### (a) Appropriate management of specimens between routine and reference laboratories

|  | Routine Laboratory | Reference Laboratory |
|--|--------------------|----------------------|
| <b><i>Conventional respiratory Pathogens</i></b> | +                  | +                    |
| <i>Staphylococcus aureus</i>                     | +                  | +                    |
| <i>Pseudomonas spp.</i>                          |                    |                      |
| <i>Pseudomonas aeruginosa</i>                    | +                  | + PCR epidem.strains |
| Non-aeruginosa <i>Pseudomonas</i>                | +                  | + PCR confirmation   |
| Polyclonal analysis                              | minimal            | detailed PCR         |
| Related Gram neg. nonferm. genera                | +                  | + PCR confirmation   |
| <i>Burkholderia cepacia</i>                      | +                  | + PCR confirmation   |
| <i>B.cepacia</i> genomovar I-IX                  | -                  | + gene sequencing    |
| <i>B.cepacia</i> virulence marker                | -                  | + by PCR             |
| <i>B. cepacia</i> transmissibility marker        | -                  | + by PCR             |
| Non-cepacia <i>Burkholderia</i>                  | -                  | +                    |
| Mycobacteria                                     |                    |                      |
| and others                                       | +                  | + transplant work-up |
| Susceptibility testing                           | +                  | + with MIC           |
| Synergy testing/biofilm MIC/Ih tests             | -                  | + with FIC           |
| Genotyping                                       | -                  | +                    |
| Serology   | -                  | +                    |

**(b) The Processing Scheme:**



**Source:** Dr Philip Murphy, Consultant Microbiologist, AMNCH, Tallaght, 2005

## **Appendix 7**

### **Shared Care Provision**

The Working Group fully endorses the recommendations and comments of the European Consensus document <sup>18</sup>, which states that:

In countries where there is shared care between the CF centre and smaller hospitals that are closer to the patient's home, the centre should coordinate care and hold ultimate responsibility for the patients' treatment and outcome. Shared care cannot be guaranteed to be equivalent to centre care and should be reserved for patients who live far from the CF centre or for whom social difficulties make regular attendance at the centre impossible. Shared care clinics must meet the same standards as at the main centre, allowing that this may require help from the multidisciplinary centre team and subspecialty consultations from the centre.

*“The practice of the CF centre sharing the care of patients with the staff at their local hospital has become established because some families and patients cannot, and others will not, travel long distances for their routine treatments.”*

*“Agreed models of shared care are needed as a response to patient/parent demand but they should not be allowed to result in suboptimal care. There is no place for doctors working in isolation and caring for small numbers of people with CF. A satellite CF unit in close liaison with the CF centre should have a minimum of 20 patients and input from a dietician, physiotherapist, and nurse, each with a special interest in CF. Patients with CF at the satellite unit should attend the CF dedicated clinics and should not be included in general paediatric or adult respiratory outpatient sessions. Basic care at the satellite unit should be of an equivalent standard to that delivered at the centre. Only in exceptional circumstances should the CF centre agree to share management with a doctor who cares for very few patients. Contact with the centre may be by the centre team visiting the satellite unit or by the patient periodically attending CF centre or both, at least once and ideally twice a year. The centre team should perform the annual assessment and ultimate responsibility of care should rest with the CF centre director.”*

It is the view of this group that the annual assessment should be conducted in the specialist centre that has all of the necessary infrastructure.

*“Shared care is more appropriate for children with CF than for adults. The latter are likely to have more complex disease requiring input from other specialists who have acquired an expertise for CF associate problems in their own discipline, e.g. obstetrics, gynaecology, diabetes and are therefore best cared for by the CF centre staff.”*

In order for shared care to be effective it was agreed that:

- a) the records for the patient would be held at both the specialist centre and the outreach hospital
- b) consultants at both hospitals are responsible for maintaining and communicating patient information within and between the two hospitals and General Practitioners as appropriate
- c) the specialist centre would be responsible for updating of the CF Registry

**Note: It is important that it is clearly explained at the first clinic attended by the patient and parents the advantages and disadvantages of specialist centre care versus shared care and that the patients or parents have the option of changing from one to the other at any time.**

## **Appendix 8**

### **Comments from Service Providers in Ireland on the ‘Pollock Report’**

#### **“The Treatment of Cystic Fibrosis in Ireland: Problems and Solutions”**

Key elements of each response are summarised below:

- 1) **St Vincent’s Health Care Group (including St Vincent’s University Hospital, St Vincent’s Private Hospital and St Michael’s Hospital) - Chief Executive**
  - Numbers of patients shown for the hospital all receiving hospital care simultaneously - extent to which each patient required hospital care varied depending on disease profile
  - Treatment of a number of older patients puts exceptional pressure on facilities and staff
  
- 2) **Cystic Fibrosis Registry of Ireland - Director**
  - Key objective is to provide utilisation details for services and resources.
  - 39% (445) of the current estimated CF population (1,143) on the CFRI database.
  - Further and more useful analysis is dependent on a larger data set (closer to 100%).
  - A key limiting step is patient consent to data recording and entry.
  - Goal of 100% patient enrolment would require additional resources.
  - Internationally, large CF registries cover approximately 66% of their target populations.
  - Ideally, registry staff would be located at a CF centre.
  - A long-term guaranteed funding stream is required.
  
- 3) **Cystic Fibrosis Nurses - Secretary, Cystic Fibrosis Nurses Association**
  - Staffing levels in the major centres should be brought in line with the recommended staffing levels prior to phasing out the smaller centres in order for them to be able to absorb the increased workload.
  - Inpatient facilities should be a single room and bed allocation should be addressed at management level in each hospital as to how they would meet the requirement of 3 to 5 beds per 50 patients.
  - Transition programmes should be further developed and resourced. The ideal is that transition begins in early teens with several meetings between adult and paediatric services. This is not currently in place due to work commitments on both sides at present – exacerbated when not on same site.
  - There needs to be an examination of what community services are currently available and what scope or need for these to be further developed such as homecare nurses for patients receiving intravenous antibiotics.
  - Up-skilling GPs and the development of a community cystic fibrosis, Clinical Nurse Specialist.
  - Training of existing staff should be examined and possible course established for all CF staff i.e. in counselling and bereavement skills and research skills.
  - Development of nursing research in CF with adequate staffing levels and this would provide an opportunity for nursing research to be done in this area and also the development of CF nurses in major CF centres to co-ordinate all nursing research around the country.

*(It was also noted that in the majority of cases CF nurses would also have contributed to their hospital’s submission).*
  
- 4) **Physiotherapists working with Cystic Fibrosis patients - Acting Senior Physiotherapist in cystic fibrosis, Our Lady’s Children’s Hospital, Crumlin.**
  - Physiotherapists working in CF centres agreed the Pollock Report, were aware of flaws in the system and acknowledged the need for more specialised CF centres.
  - Emphasised the lack of adequate treatment space and facilities, the lack of scheduled time for the treatment of CF patients, the lack of physiotherapy and wider CF staffing.

- Were the proposed staffing levels were to take effect there would be no need for the involvement of community physiotherapy. Instead, physiotherapists based in specialised CF centres would carry out home visits
- Key concern is the level of training and service provided by community physiotherapists to those patients whose physiotherapy was initiated in a CF centre.

**5) Cystic Fibrosis Interest Group of The Irish Nutrition and Dietetic Institute (INDI) - Institute Secretary**

- Professional dietetics input is an essential element in the treatment of CF
- Dietetic staffing guidelines proposed by Dr. Pollock insufficient to meet the need
- Clinical Specialist positions should be created in both adult and paediatric services
- Dieticians working fulltime in CF care should be at senior grade
- Should be access to a gastroenterology and endocrinology service which has specialist knowledge of CF
- Care should be provided in a specialist centre, with teaching, by experienced staff
- While a community dietetic service would be beneficial, neither the staffing nor expertise or currently available in the community to support such a service
- Need more staff for OLCH, Crumlin than Pollock states (details provided)
- OLCH, Crumlin submission notes the need for a newborn CF screening programme and associated paediatric dietetic staffing with expertise in CF to support such a service
- Temple Street Hospital submission notes that the hospital exceeds the Pollock Report's recommendations. Pollock recommended 0.8 wte per 100 patients. Temple Street currently has 0.9 wte for 82 patients.

**6) Psychologists working with CF patients**

- Pollock report does not give appropriate weight to the role of the psychologist in the multi-disciplinary team for CF patients.
- In the CF population the physical disease can bring an increase in psychological disease - both patient and family may require psychological support
- Psychologist must be full-time in CF context – sessional work is inadequate for professional support to other members of the team and to build appropriate relationships with patients and families for ongoing treatment and care across the full range of CF management including consideration of lung transplantation
- Adequate staffing: 1:80 patients, 14-25 patients – 80% time spent with in-patients, 15% outpatients, 5% non-clinical work; patient education and continuous professional education.

**7) The Children's University Hospital, Temple Street – Consultant Respiratory Physician with a special interest in cystic fibrosis**

**Enhancement of Adult Services**

- Extra funding is required to improve staffing.
- Further isolation rooms need to be created and a treatment room provided for drop-in purposes.
- While international evidence points to 50-100 patients per CF centre, in excess of 200 patients is not necessarily beneficial - very high numbers of patients could lead to increased cross-infection.
- The recently developed adult cystic fibrosis centre in Beaumont is increasing its numbers and will help ease pressure on St Vincent's Hospital.
- The majority of patients from Temple Street are currently transitioned to Beaumont Hospital for practical purposes.

**Realignment of paediatric services**

Notes the differences in the NCHD staffing of adult and paediatric consultant-led teams. Adult teams have more NCHDs, all of whom, generally speaking, participate in the general medical on-call rota. It is not clear how increasing the number of NCHDs on paediatric teams would correspond to training requirements.

## **Staffing**

Reasons for a rapid increase in patient numbers in Temple Street include the appointment of a Consultant Respiratory Physician with a special interest in cystic fibrosis; regular referrals of neonates from the Rotunda and Holles Street – many of whom require neonatal surgery; and Temple Street's participation in 'shared care' arrangements with other hospitals.

Temple Street provides a wide range of services to CF including annual review, neonatal care and treatment, ENT surgery, genetic counselling, specialised procedures specific to CF, access to diagnostic and laboratory services – including national neonatal screening, and psychological support, clinical research and provision of opportunities for patient and familial advocacy.

## **Reference Laboratory**

Emphasises the need for a National Reference Laboratory, based – due to existing expertise (in particular regarding B. Cepacia) in Tallaght. It should be noted that Dr Slattery notes that the patient population in Tallaght is likely to fall due to the absence of a 'neonatal feeding hospital' (although it is not clear where neonates from The Coombe Women's Hospital are referred to) and that increasingly, patients will be transitioned to St Vincent's.

## **Development of Screening Programmes**

Funding for cystic fibrosis centres should be allocated to improve staffing, inpatient and outpatient services before a neonatal screening programme is introduced. This is to address the potential for neonatal screening to identify a large number of new patients simultaneously rather than them presenting over a number of years as is the case at the current time.

## **Information Registry**

The submission notes that the Cystic Fibrosis Registry should be maintained and developed and that additional funding is needed.

Ireland should seek to attain a level of CF service provision consistent with that offered in the United States or Canada. The level of services recommended in the UK should form a 'minimum basic' or 'first step'.

## **8) Our Lady's Children's Hospital, Crumlin**

### **CF services at OLCH, Crumlin; Strengths, Deficiencies and Realignment of Paediatric Services**

- OLCH is the largest paediatric CF centre in the state -160 CF patients. 31 patients are on a shared care programme with regional CF centres. The retirement of a Consultant Respiratory Physician in Waterford has resulted in an increase in patient numbers in recent months.
- Emphasises the need for single rooms for all CF patients and associated isolation facilities. A Development Control Plan has been submitted to the Department of Health and Children and is being discussed in the context of the future development and location of paediatric services in Dublin. In the interim period, OLCH state that 3-5 beds per 50 CF patients are required – equating to 9-15 beds in OLCH.
- In relation to outpatient facilities, the submission notes the development of a purpose-built, privately-funded facility on-site. OLCH hope to develop combined respiratory / gastroenterology and respiratory / diabetic clinics in this facility.
- Highlights a need for the appointment of a locum consultant paediatrician as an emergency measure and states that the effectiveness of a new post of consultant in respiratory medicine (for which approval is awaited from the HSE) is dependent on the appointment of approximately 11.5 additional support staff of various types.
- Describes current CF research in Crumlin, including that on CF related Liver Disease, CF related diabetes and anti-inflammatory agent in CF lung disease.
- Supports the establishment of an All-Ireland CF Research Consortium at the Dublin Molecular Medicine Centre to facilitate basic training and research and foster links with the National Institutes of Health in the US.
- OLCH, as a national centre of excellence and tertiary referral centre, is 'ideally suited' to become the National Tertiary Centre for CF patients.
- The Pollock Report's recommendations with regard to discontinuing service delivery at some smaller units outside Dublin would result in increased travel times for a number of patients and recommends the development of cross-border links.

- Designation of OLCH as the national centre should be contingent on the full implementation of the Pollock Report.

#### **Enhancement of adult CF Services**

- OLCH note that while it currently caters for patients up to 16-18 years of age, care often continues beyond this period
- States that the hospital is already exploring the area of services to adolescents and that there is a need for a purpose-built adolescent unit. Makes reference to patient anxiety about leaving OLCH as a result of adverse reports of services available in adult hospitals as outlined in the Pollock Report
- Proposes a two-year lead-in prior to transfer of patients from paediatric to adult services.

#### **Reference Laboratory and Microbiology Services**

- The submission describes the wide range of specialties and services provided in OLCH with activity details.
- The National Centre for Medical Genetics is based in OLCH and provides a service to patients and families affected by genetic disorders in Ireland. The Centre offers a variety of tests for CF and seeks to expand CF genetic counselling services.
- OLCH recently underwent the first stage of IHSAB accreditation. The laboratory has also recently undergone CPA accreditation.

#### **Development of Screening Programmes**

- Welcomes the proposed introduction of a newborn screening programme for CF patients but would require additional resources. In the event of a newborn screening programme OLCH would wish to develop an Infant Lung Function laboratory.

#### **Information / Registry**

- OLCH is closely involved with the Cystic Fibrosis Registry
- OLCH concludes by noting that the 'packages of care' funding mechanism proposed by Pollock needs further examination. In addition, OLCH note that the example of a banding system used in the Report seems to apply to an adult rather than a paediatric population.

#### **9) Health Service Executive – South Western Area, Assistant Chief Executive Officer**

- Children with CF in the HSE South Western area receive a long-term illness card and may benefit from Domiciliary Care Allowance and an Annual Respite Care Grant.
- The National Physical and Sensory Disability Database is the planning tool used for the development of disability services. At the current time, 19 adults and 49 children with CF are registered on the database. This would appear to be a significant underestimate of the numbers and thus is of limited value for planning purposes.

#### **10) Health Service Executive – Southern Area**

##### **a) Community Physical and Sensory Disability Services: Development Manager:**

- The Pollock Report does not address services for people with CF in the community or in the interface between Acute and Community Services.
- Model for the delivery of community nursing services which links both Acute and Community Services currently in development.

##### **b) Cork University Hospital, Divisions of Internal Medicine / Paediatrics:**

- The submission welcomes the Pollock Report and notes the 'unstructured' manner by which services for cystic fibrosis patients have evolved.
- Need for standards to underpin any proposed planning for service development define the need for multidisciplinary staffing and set out defined care pathways for patients.

##### **c) Cork University Hospital, Physiotherapy Services, Physiotherapy Manager:**

- Notes and specifies gaps and deficiencies in the CF physiotherapy service.
- A physiotherapist should assess most 'walk ins' to the day unit,
- There should be physiotherapy follow-up of patients on home I.V. treatments

- Suggests that CUH be developed as the Southern Region Adult Centre in addition to existing paediatric services.
  - Support for the recommendations of Report regarding screening, reference laboratories and information.
  - Development of the Cork University Hospital Neonatal Service may allow for the establishment of neonatal screening within the CUH Group.
  - Advocates the use of common physiotherapy assessment tools and exercise testing to support standardisation and quality improvement.
- d) Cork University Hospital, Nursing Services, Nurse Service Manager and Clinical Nurse Specialist, cystic fibrosis
- Suggests all CF nurses should use the current model in use in Cork where nurses provide and maintain skills in both paediatric and adult CF care with no division between the two.
  - Increases in staffing would need to be matched to infrastructural improvements.
  - A CF nursing postgraduate diploma qualification be introduced.
- e) Cork University Hospital, Microbiology Services, Consultant Microbiologist
- Recommends the creation of specific CF laboratory facilities staffed by a senior laboratory scientist and two basic grade scientists with appropriate molecular equipment. The submission notes that over 50% of the 158 CF patients in the Southern region are *Pseudomonas aeruginosa* carriers. Potential *Burkholderia* samples are sent to the UK for testing and there is currently a 6-week turnaround time. (Since this information was originally gathered these tests are now sent to AMNCH).
- f) Cork University Hospital, Social Work Department, Social Work Manager,
- Inadequate counselling and social work service in place for CF patients
  - Staffing requirements additional to those stated in Pollock report
- g) Cork University Hospital, Nutrition and Dietetics Department, Dietician Manager
- Pollock Report described physical facilities as good – this is not the case.
  - Welcomes the recommendation that both adult and paediatric CF patients be treated by the same personnel.
  - Emphasises the need for prompt and thorough follow-up of children identified as having CF in an adequately-staffed specialist centre.
  - Notes that the list of people interviewed during the preparation of the Pollock Report does not appear to include dietitians, physiotherapists or social workers.

## 11) Health Service Executive – North Western Area

- a) Consultant Paediatrician, Sligo General Hospital
- Number of CF patients attending Sligo General Hospital cited in the Pollock Report should be amended from 13 to 21.
  - Pollock team did not consult staff in Sligo or visit the Sligo service;
  - A key factor in travel to Dublin, Galway or Derry is distance – most patients would not seek to travel to this extent on a regular basis; the distance involved could be problematic in emergency cases.
  - In the UK, CF services are provided locally even where numbers are small. A team from the specialist CF centre visits the local centre on a regular basis in place of patients travelling to the specialist centre. In this context, suggests improving the CF services in Sligo and supporting service provision with regular input from a specialist CF centre.
- b) Consultant Paediatrician, Sligo General Hospital
- Disagrees with the Report's conclusions regarding the North West.
  - Dr Pollock did not contact staff in the North West
  - Model operating in the UK includes outreach from specialist centres to smaller CF units

- Most patients in the North West have closer links to Dublin than Galway and that it cannot be assumed that they would transfer to Galway for CF services.
- Emphasises the need to examine the benefits of shared care between satellite centres and a national centre and the need to avoid the disruption associated with a regular travel to a geographically distant national centre.

## 12) Health Service Executive – South Eastern Area

- a) Regional Manager, Acute Services and Consultant Paediatrician, Waterford Regional Hospital (on behalf of the multidisciplinary team caring for CF patients)
- Welcomes the recommendation of the Pollock Report that Waterford Regional Hospital be developed as a CF centre.
  - Suggests that a forthcoming post of Consultant Respiratory Physician be structured to include a number of CF sessions.
  - Notes that while there are single rooms, none of these have en-suite facilities.
  - Notes the need for improvements in physical infrastructure, physiotherapy, social work and psychologist staffing.
  - Need for dedicated staff to support the operation of the CF Registry in Waterford.

### Dietetics:

- Currently 0.15 wte dietician involved in CF services – Pollock Report states 0.5 including general paediatric dietetics.
- Pollock recommendation of 0.4 wte per 50 CF patients has not been ratified in the Irish context of sicker patients with a historically lower level of service. The submission proposes a figure of 0.8 wte.
- Time is evenly split between inpatient and outpatient services. Paediatric and adult dieticians work separately – this gap needs to be bridged

## 13) Health Service Executive – North Eastern Area

- a) Consultant Paediatrician, Our Lady of Lourdes Hospital, Drogheda
- Welcomes the principle behind the Pollock Report but states that the model of care proposed by the Report does not optimise care to achieve optimal outcomes while achieving an optimal level of consumer satisfaction
  - Either a 'supra-regionalised' approach or a 'hub and spoke' model recommended
  - May be difficult to achieve required medical staffing levels in the number of centres proposed by Pollock. Excessive on-call commitments may remain as medical staff are spread too thinly
  - Table 2 of the Pollock report refers to patients attending both Drogheda and Cavan. However, the figure used in the report refers solely to patients attending Drogheda.
- b) Assistant Chief Officer, Acute Hospital Services included comments from:  
Consultant Paediatrician, Our Lady of Lourdes Hospital, Drogheda
- Makes many of the same points in this submission as in his own, separate submission described above.
  - Essentially 1 wte consultant respiratory physician and 1 wte CF nurse involved in the care and treatment of CF patients in Drogheda. Patient figures in the Pollock report do not include Cavan and should be amended from 31 to 55.
  - Recommends the provision of clinical and office space for outpatient reviews, the termination of CF nursing commitment to Cavan, the development of outreach CF services and the provision of consultant microbiologist and nutritionist services in Drogheda.
  - Would be more logical to develop a regional centre in Drogheda than in Waterford and that such a centre would better serve the needs of patients from the North West than a centre in Derry would.
- c) Consultant Respiratory Physician,

- While benefits of greater centralisation of services are clear, that there are two problems.
  - Firstly, there is no dedicated CF centre north of a line between Dublin and Galway; and
  - Secondly, patients living some distance from CF centres may have difficulties accessing services due to problems with bed availability.

c) Consultant Paediatrician,

- Notes that there is no CF centre in Ireland comparable to those in continental Europe or the United States then describes a series of deficits in CF service provision nationally and in the north east.

d) Acting Clinical Nurse Manager 3 and Cystic Fibrosis Clinical Nurse Specialist

- Notes the inaccuracies in the Pollock Report regarding patient numbers in the North East described above.
- Describes existing deficits in CF services in the region and identifies a series of infrastructural and staffing needs, including a need for specialist surgical expertise.

e) Consultant in Respiratory and General Internal Medicine,

- Describes existing deficits in CF services
- Would recommend that adult CF patients 'with maternity issues' attend a specialised unit in Dublin for safety reasons.
- Given the deficits in service provision and infrastructural problems, without significant investment, CF care in Drogheda is unsustainable. Will not be in a position to take on new referrals from paediatric services.
- Currently no formal process in place to support the transition of such patients to CF services in other hospitals
- Issues raised by the Pollock Report have implications not only for CF services but for Respiratory Medicine Services in General.

**14) Health Service Executive – Midland Area, Assistant Chief Officer**

- General concurrence in the Midland Area to the broad thrust of the Pollock Report's recommendations.
- Recommends that there should be outreach clinics in each hospital network about three times a year.
- This model is in operation in the UK and ensures optimal patient care locally without unnecessary travel; maintains skill levels regarding the treatment of CF amongst the local paediatric team and supports links between secondary and tertiary CF treatment services.

**15) Health Service Executive – Northern Area, Assistant Chief Officer**

- Notes the recommendations of the Pollock Report
- Refers to the significant deficits in current CF services
- Identifies the number of children moving into the adult service as key area
- Notes that planning should address the needs of an older population with more complex problems.

**16) Health Service Executive – Western Area, Regional Coordinator, Acute Services**

- Broad agreement to the recommendations of the Report relating to screening, reference laboratory, information and registry.
- In relation to Mayo General Hospital notes that Dr Pollock did not visit the hospital and that concerns have been expressed that the position of patients in this area had not been adequately represented.
- Includes a detailed paper on the development of CF services in Galway prepared in early 2004 and notes that this paper is broadly in line with the Pollock recommendations regarding Galway.

## 17) Health Service Executive – Mid-Western Area

- a) Consultant Paediatrician, Mid-Western Regional Hospital, Limerick
- Describes a series of deficits in the CF services provided in the Mid-Western area while detailing the range and nature of CF services provided.
  - There has been difficulty relating to the position of CF Nurse Specialists since 1998.
  - Each of the 3.3 wte CF nurses recommended by the Pollock Report should be Nurse Specialists.
  - This will involve the appointment of Clinical Nurse Specialists.
  - Existing CF nurse staffing in Limerick is provided by nurses in temporary positions and the regular retraining is required.
  - Neonatal CF screening should be a priority.
  - There is a need for Clinical Psychology service.
- b) Regional Manager, Acute Hospital Services
- The two Consultant Respiratory Physicians collaborate closely in relation to CF patients and that links between the paediatric and adult services work well
  - The figures in the Pollock Report concerning staffing figures in Limerick are slightly incorrect (details provided)
  - The hospital Group would not view the establishment of neonatal screening as the highest priority
  - Lists a series of staffing and requirements and makes reference to the need for additional infrastructural investment and support – particularly if patients transfer from the Tralee CF service
  - Key priority is the development of an Adult Day Unit for the treatment of CF patients

## 18) Adelaide and Meath Hospitals incorporating the National Children's Hospital (Tallaght)

- The hospitals would seek to develop the current respiratory services to manage transition, adolescent and adult CF patients creating seamless progress for the current paediatric patients and easing pressure on the adult service at St.Vincent's.
- The hospitals currently have a strategy of strengthening links with OLHSC and the Coombe Women's Hospital and seek to develop maternity and neonatal services on site.
- Consider OLHSC as natural partner in relation to future paediatric arrangements
- AMNCH has well-developed adult respiratory services on site and is developing non-acute respiratory services with Peamount hospital.
- Hospitals' strategy includes development of chair in clinical genetics with TCD.

### Reference Laboratory

- AMNCH previously submitted proposal for designation of CF reference laboratory at Tallaght in 1999. A service has been established using charitable grants from the Cystic Fibrosis Association of Ireland.

### Screening

- Supports Pollock report's recommendation on neonatal screening but emphasises need to have required services to meet screening outcomes.
- Submission notes that maternity hospitals tend to refer to paediatric hospital with which they have sessional commitments rather than the paediatric CF centre in the patient's catchment area – this may lead to distortion of patient distribution among centres with under-referral to some CF centres and over-referral to others. The National Children's Hospital would recommend that neonatal screening programme would address referral patterns of children based on patient choice and catchment area.
- The hospital fully supports and collaborates with the CF Registry.

### 18.1) Reference Laboratory

- The hospitals made a separate, comprehensive submission seeking the designation of the current referral service as the national reference laboratory for CF.

- The document reflects an updated proposal previously submitted to the Department of Health and Children in 1999. The service currently provided is used by all hospitals currently providing CF treatment.
- The current service is provided through use of existing equipment and staff supplemented by funding from charitable sources.

**19) Beaumont Hospital, Clinical Services Co-ordinator**

- Currently adult 68 patients (no paediatrics), referred by Temple St.
- Adult CF would be the intended core patient group for future
- Patients largely from north city and county Dublin and HSE North Eastern Region.
- Aim to develop comprehensive multi-disciplinary outpatient service thus keeping admissions to a minimum.
- Disagree with assumption in Pollock report that physical infrastructure is adequate - significant problem is lack of single rooms (vital for prevention of cross-infection) – two to three single rooms required.
- Outpatient department of the hospital currently preparing to move to three sessions per day – this would provide greater flexibility for patients but physiotherapy space required.
- Service currently provided by combination of general medical services staff, CF specialist staff and post(s) (current and planned) funded by the Cystic Fibrosis Hopesource Foundation as well as small number of dedicated CF staff or sessions.
- Submission also highlights the need to increase adjunct diagnostic supports for CF (not explored in detail in the Pollock report), e.g., additional post required in pulmonary laboratories as well as increased demands on immunology, microbiology in the pathology division and on radiology.
- Estimated will have 110 patients by 2010 (currently 68)
- Identifies substantial amount of equipment required for comprehensive service – for immunology, pulmonary laboratory, physiotherapy, social work and need to develop services to accreditation standard.
- Detailed description of current service (where provided) and future plans for CF nursing, dietetics, medical social work, physiotherapy, psychology, laboratories and radiology.

## **Appendix 9**

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- Hope Source Foundation for CF
- ESRI
- CSO
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- All of the staff linked to the following units caring for CF patients who provided information on current staffing and facilities:
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  - St Vincent's University Hospital
  - The Children's University Hospital, Temple Street
  - Our Lady's Hospital for Sick Children, Crumlin
  - Adelaide, Meath and National Children's Hospital, Tallaght
  - Waterford Regional Hospital
  - Cork University Hospital
  - Tralee General Hospital
  - Limerick Regional Hospital
  - Galway University Hospital
  - Mayo General Hospital
  - Sligo General Hospital
  - Letterkenny General Hospital
  - Cavan General Hospital
  - Our Lady of Lourdes Hospital, Drogheda
- Members of the Working Group
- Patients with CF

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