NATIONAL CLINICAL PROGRAMME FOR CYSTIC FIBROSIS

CYSTIC FIBROSIS

A MODEL OF CARE FOR IRELAND

CYSTIC FIBROSIS

Feidhmeannacht na Seirbhíse Sláinte
Health Service Executive

ROYAL COLLEGE OF PHYSICIANS OF IRELAND

Developed by the National Clinical Programme for Cystic Fibrosis

CYSTIC FIBROSIS

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NCPCF Clinical Advisory Group Membership

All members appointed by the RCPI, following nomination by the Irish Thoracic Society.

Dr Desmond Cox	Consultant in Paediatric Respiratory Medicine, OLCHC
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Dr Margaret Hannan	Consultant Microbiologist, MMUH
Prof Edward McKone	Chair, Cystic Fibrosis Registry of Ireland
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Foreword by Cystic Fibrosis National Clinical Lead

This Model of Care for Cystic Fibrosis in Ireland was written by the Working Group of the National Clinical Programme for Cystic Fibrosis (NCPCF). It outlines how the care of People with Cystic Fibrosis should be organised and resourced now and in the future.

The NCPCF was set up by the Health Executive (HSE) in 2015 on the recommendation of the Royal College of Physicians in Ireland (RCPI) and others. The NCPCF Working Group was established in October 2015 and it is multidisciplinary. The Working Group includes nominees from different groups involved in the care of People with Cystic Fibrosis, including nominees from the Cystic Fibrosis Centres and Shared Care Centres, Cystic Fibrosis Team Members, Cystic Fibrosis Ireland, Cystic Fibrosis Registry of Ireland, HSE Acute Hospitals Division and others.

One of the first priorities of the NCPCF Working Group was to develop and define this inaugural Model of Care for Cystic Fibrosis in Ireland. Therefore, a Model of Care Subgroup was set up and the Subgroup reviewed different international and national documents and recommendations regarding Cystic Fibrosis care. The frequency, severity and complexity of Cystic Fibrosis are greater in Ireland than in most countries. Therefore, the Subgroup developed this Model of Care for Cystic Fibrosis for Ireland. Following several cycles of detailed review by the Working Group, the Model of Care was then sent out for external review and feedback. Written feedback was received from 44 individuals and organisations. This feedback was reviewed in detail by the Model of Care Subgroup and the Working Group and was incorporated into the Model document.

This final Model of Care document has been approved by the members of the NCPCF Working Group and the RCPI Cystic Fibrosis Clinical Advisory Group. The membership of the NCPCF Working Group, Model of Care Subgroup and the Cystic Fibrosis Clinical Advisory Group are given on Pages i, ii, iii.

I am very grateful to the members of the Model of Care Subgroup and the NCPCF Working Group who worked very hard to write, review and finalise this document; this is truly a team effort. I am very grateful to the Clinical Advisory Group for their careful review of and input into this document. I am very grateful to all who gave feedback. I want to acknowledge the very strong support, guidance and wisdom that I received from Dr Colm Henry, HSE Chief Clinical Officer and Dr Vida Hamilton, National Clinical Advisor and Group Lead, HSE Acute Hospitals Division. I am very grateful to Prof Mary Horgan, President RCPI; Prof Frank Murray, Past President RCPI; Dr Áine Carroll, previous National Director, HSE Clinical Strategy and Programmes Division for their strong support. I want to specially thank Mr Gary Killeen, NCPCF Programme Manager, for all his untiring work and dedication.

I am very grateful to the HSE Leadership, the RCPI, the Department of Health and the Irish Government for approving this Model. I am very grateful to Minister Harris for officially launching the Model of Care for Cystic Fibrosis today

This Model of Care is dedicated to People with Cystic Fibrosis, and their families, who bravely deal with CF every day of their lives. It is also dedicated to the People with Cystic Fibrosis who are no longer with us and their families; you will not be forgotten.

Professor Charles Gallagher

National Clinical Lead, National Clinical Programme for Cystic Fibrosis

Executive Summary

Life expectancy of People with Cystic Fibrosis (PWCF) and the complexity of Cystic Fibrosis (CF) care have both increased progressively in recent years. For each PWCF the complexity and severity of CF and the burden of CF care usually increase as they grow older, especially when they become adults. This increasing complexity and severity with age will not change with current disease-modifying treatments for CF because these drugs are disease-modifying, not disease-curing. Therefore, the healthcare needs of PWCF in Ireland will continue to increase and this will require ongoing patient-focused planning, involving all stakeholders, to ensure the highest quality of care is provided to PWCF.

Cystic Fibrosis care in Ireland must be delivered by a small number of designated Specialist CF Centres that are properly resourced and staffed.

All PWCF must receive their CF care from, and have regular follow-up by, a designated Specialist CF Centre. Adults with CF should receive all their CF care directly from a Specialist CF Centre. All children with CF should receive their CF care from a Specialist CF Centre; some children with CF who live far away from their Specialist CF Centre may receive part of their care from a designated Shared Care Centre closer to home, if that Shared care centre is in partnership with, and is following clinical guidelines from, the Specialist CF Centre.

Specialist CF Centres must:

- Have a specialist multidisciplinary team of trained experienced CF Specialist Medical, Nursing, Health and Social Care Professional and Administrative Staff whose time is dedicated to the care of PWCF. Staffing levels will be determined by the number of PWCF attending the centre; the required staffing ratios are outlined in this document. For CF Consultants and CF Team members, there must be a designated replacement to fill that post when they are on scheduled or unscheduled leave. For CF Team members who are working largely or solely in CF Care, their primary or sole reporting relationship should be with the CF Centre Director.
- Have dedicated single inpatient rooms with en-suite facilities. Adult Centres must have 5 inpatient rooms for each 50 patients attending the Centre. Paediatric Centres must have 3 inpatient rooms for each 50 patients attending the Centre. Inpatient rooms must always be available and access protocols must be in place to ensure that all PWCF who are acutely sick are admitted immediately and always within 24 hours. There must be at least 1 skilled nurse for each 3 CF inpatients during the day and 1 skilled nurse for each 4 inpatients during the night when all patients are stable because of the complexity and critical care nature of inpatient CF Care.

- Have dedicated day care rooms with en-suite facilities and outpatient rooms for assessment/treatment of PWCF as outpatients
- Have strict infection control facilities and protocols in place for inpatient, outpatient and day care to prevent cross infection from other PWCF and from other patients. PWCF can pick up dangerous organisms, especially from each other.
- CF Centres must have Consultants with a special interest in Cystic Fibrosis in the following specialties: Endocrinology, Gastroenterology, Hepatology, Nephrology, Oto-Rhino-Laryngology (ENT), Palliative Care, Psychiatry, Radiology including Interventional Radiology and Thoracic Surgery. Those CF Centres who do not yet have one or more of these on site should have clear referral pathways to them.
- Be designated as a Specialist CF Centre by the National Clinical Programme for Cystic Fibrosis (NCPCF) and must look after at least 50 PWCF

People with Cystic Fibrosis should have regular outpatient/day care review by the Specialist CF team every 3 months when medically stable and more often when they are unwell or unstable.

Follow up of PWCF Post-Lung Transplantation is very important; it is a complex issue internationally and it involves follow up for transplant related issues and also follow up for CF related issues. Because this is very important, the NCPCF will prepare a separate Model of Care document dedicated to the care of PWCF Post Transplantation; the recommendations of this present Model of Care and this Executive summary do not apply to PWCF Post-Lung Transplantation.

There must be a structured accreditation/peer review process by which CF Centres and Shared Care Centres are assessed and accredited/peer reviewed on a regular basis. The accreditation/peer review process needs to be properly resourced and it should be under the direction of the NCPCF or the National Cystic Fibrosis Office, when established.

The CF Registry of Ireland is an essential part of planning and monitoring Cystic Fibrosis Care. Its role and funding should be expanded so that, among other functions, it can provide the data needed to help plan CF resources in the medium and long term. It should also be resourced so that it can provide independent data on the effectiveness and side effects of new CF treatments.

The significant improvements in health and survival in CF internationally in recent years are due to the development and resourcing of designated Specialist CF Centres, aggressive treatment of respiratory and other manifestations of CF, the collection and organisation of data into registries, the development of accreditation/peer review and clinical standards, and the involvement of the whole CF community in research.

The NCPCF strongly recommends that a National Cystic Fibrosis Office be established to coordinate, manage and integrate all aspects of CF care in order to optimise the health and quality of life of People with Cystic Fibrosis in Ireland

1 Background

Cystic Fibrosis (CF) is one of the commonest lethal inherited conditions in the world. It causes premature death and this is due to CF lung disease in the vast majority of people. CF has a high prevalence in Ireland and Ireland also has a higher proportion of more severe CF gene mutations than most countries. It is a major burden to People with Cystic Fibrosis (PWCF) and their families; even when well, many PWCF spend at least 2 hours every day taking their treatments. Over the last 25 years, there has been a switch from ad hoc treatment to a coordinated system of linked specialist CF centres with national registries, national and international standards of care and clinical guidelines; this has been associated with significant improvement in outcomes for PWCF internationally. The improvement in outcomes is partly related to better organisation and delivery of multidisciplinary care, partly to monitoring and aggressive early treatment of lung infections/lung disease and partly to management of the complex multi-organ manifestations of CF. The improvement in outcomes is also due to the understanding that the clinical features of CF vary significantly between PWCF and show a major change with time in each individual PWCF. Therefore, optimal management of CF involves treating a moving target(s). Programmes in other developed countries aimed at establishing a model of care, instituting and maintaining standards of care and ensuring ongoing accreditation/peer review of CF centres have proved very successful in terms of local and national outcomes. Predicted survival figures for Ireland have lagged behind those of other developed countries in the past but this gap has narrowed in recent times (Jackson et al., 2011a, Jackson et al., 2011b). With a high prevalence and awareness of CF and a small network of well-connected centres, organisational reform and appropriate resourcing of CF care have the potential to make Ireland a leader in CF care and outcomes.

In recent years, several countries have produced both *standards of care* and *models of care* for PWCF. These include documents from Australia and the United Kingdom (UK), where the healthcare systems resemble Ireland's healthcare system to an extent. In 2008, CF Australia published its document entitled *Cystic Fibrosis Standards of Care, Australia*, and in December 2011, the UK CF Trust published *Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (Second Edition)*. These documents deal with, among other things, the basic standards of care that should be expected by PWCF and their families, the standards expected for individual CF centres, and the overall structure of the network of CF centres in the country. While some of the content of these documents can be extrapolated to Ireland, additional local considerations must be taken into account. While considerable overlap exists between a *model of care* and *standards of care*, particularly in the context of CF in Ireland, in this document we will attempt to deal with the model of care in isolation.

2 Rationale

The evolution of models of care on a local and regional basis for PWCF in Ireland has been largely based on regional factors and has not been directed by a central authority. This could result in differing approaches in different areas to the management of PWCF. Referral pathways were ad hoc in the past and there could be significant variability in how care for PWCF is delivered in different areas.

In light of the historic differences in the development of CF services across the country, our outcomes for PWCF (which appeared to be behind those of other developed countries in the past) and the international trend for better organised, more standardised CF care delivery, a clear and integrated Model of Care for CF is required in Ireland.

3 Evolution and function of the service

Following publication of the Pollock report in 2005 (and the later HSE report on CF services in Ireland in 2009) investments were planned for the staffing of specialist CF services in Ireland. The identification of additional staff needs to address the deficits in CF care, and the investments in CF posts on foot of this, were effective in driving forward multidisciplinary management of PWCF in designated centres; however much of the progress in staffing was eroded by the recruitment moratorium in 2008. Also, local funding shortfalls in hospitals providing CF care, coupled with the strong demand from all specialist areas for the dwindling pool of nursing and health and social care professional (HSCP) staff in these hospitals, further negatively impacted staffing. Most, if not all, CF centres are now understaffed in key areas. Furthermore, in light of the lack of ongoing review of staffing numbers in relation to numbers of PWCF and severity, resources have been stretched in many centres as staff levels have dropped or remained static and numbers of PWCF have increased.

With the advent of newborn screening for CF, all children born since July 2011 who are diagnosed with CF are now referred directly to a specialist CF centre where their care is directed and coordinated (HSE, 2011). Prior to this, an ad hoc arrangement existed in relation to where children were cared for following diagnosis. Most children in Ireland with CF are treated in a designated CF specialist centre. Several local centres continue to provide care to children with CF; some of this care is provided in a shared care arrangement with a specialist CF centre. Individual practices have evolved over time and there is no clear national pathway for the provision or supervision of shared care services currently. There is a need for a clear and coherent model of care for children with CF, especially as it relates to the provision of specialist, shared or local services.

Paediatric CF care has evolved very significantly over the last 20 years. The vast majority of paediatric CF care is outpatient care. The numbers of children with CF are sufficiently large to enable expertise to be developed and sustained in regional designated specialist CF centres that have appropriate staffing and expertise. Ireland has a very effective network of paediatric CF centres, which have easy access to tertiary subspecialty expertise (such as gastroenterology, paediatric surgery, thoracic surgery) in the national paediatric referral centre at OLCHC Crumlin, and there are good communication pathways between centres. The number of children in Ireland with CF is predicted to increase by 21% between 2010 and 2025 (Burgel et al., 2015).

The number of adults with CF continues to rise in Ireland and over 50% of all PWCF are adults (CFRI, 2016). Importantly, a recent EU report highlighted that this number will increase by 65% between 2010 and 2025 in Ireland (Burgel et al., 2015). Adult services for PWCF are predominantly based at the following designated specialist CF centres: St Vincent's University

Hospital, Dublin; Beaumont Hospital, Dublin; Cork University Hospital, Cork; University Hospital Galway, Galway and University Hospital Limerick, Limerick. Transition arrangements from paediatric to adult services are generally well developed across the country. However, some variation in practice remains and there is room for improvement in this regard.

A significant number of PWCF are receiving the majority of their care through CF day care centres, including home intravenous antibiotic regimens with associated admission avoidance and/or early discharge. With aging there are increasing numbers of adults requiring complex care as complex multi-organ disease is becoming more common in adults (Goss and Burns, 2007, Kerem et al., 2005, Plant et al., 2013, Tuchman et al., 2010). In keeping with international trends with aging, the adult CF services are dealing with increasingly challenging respiratory problems including antibiotic multidrug resistant organisms, respiratory infections that are difficult to treat, pneumothorax and large volume haemoptysis (Kerem et al., 2005, Döring et al., 2004) resulting in high admission rates and prolonged length of stay. There is also increasing prevalence of CF related diabetes (Moran et al., 2009), liver disease (Herrmann et al., 2010, Rowland et al., 2015), kidney disease (Schechter and Stecenko, 2011), occult malignancy (Gory et al., 2014, Pang et al., 2015), osteoporosis (Conwell and Chang, 2014), anxiety/depression (Quittner et al., 2014, Quittner et al., 2016), and pregnancy/reproductive health related issues (Patel et al., 2015, Schechter et al., 2013). All these factors put increasing demands on the CF multidisciplinary teams (MDT) and on CF resources.

As has been seen in other jurisdictions, two key developments in CF in Ireland have helped to create an environment that delivers quality care, namely the establishment of CF Ireland (formerly CFAI) and the CF registry of Ireland (CFRI). The interaction between the CF Consultants and CF teams, the PWCF representative groups, the CF registries and the Health Service System in developed countries has produced an environment espousing consistently high standards where outcomes have improved considerably, even in an era prior to disease modifying therapies. CF Ireland has also provided considerable charitable funding to improve capital infrastructure in many CF centres in Ireland.

A fully functioning lung transplant programme has evolved in Ireland over the last 15 years and now caters for all adults with CF requiring lung transplantation. With the developments and opportunity of lung transplantation in Ireland, a cohort of critically ill PWCF requiring aggressive and real-time minute-to-minute multidisciplinary inpatient and outpatient care can bridge to successful transplant and subsequently post-transplant management (Hirche et al., 2014).

The planning, organisation, ongoing review and resourcing of CF care through the National Clinical Programme for Cystic Fibrosis (NCPCF) is likely to be associated with further improvements in outcomes for PWCF in Ireland.

4 Current service overview

4.1 Current service pathway, delivery and challenges

4.1.1 Paediatrics

Thanks to the establishment of a network of CF centres in the last decade, specialist services can now be provided effectively in all the Specialist CF centres. Despite this network, a number of challenges still remain in terms of service delivery in different centres (HSE, 2009). These challenges relate mainly to lack of appropriate staffing in medical, nursing and HSCP areas. In many hospitals, HSCPs, such as dietitians, pharmacists, physiotherapists, psychologists, secretarial and administrative staff and social workers allocated to provide CF services come from a pool of HSCPs in that institution. Contrary to the experience of our international counterparts, in many cases, the HSCPs allocated to CF services have other clinical commitments in addition to their CF work (HSE, 2009). Many centres have noticed over the last number of years that, in the context of shrinking staff pools of HSCPs within hospitals, HSCP allocation to CF services is inadequate and this is affecting care of PWCF. A system that protects the required number of whole time CF staffing is necessary for safe care of PWCF and for team stability and morale.

As well as the major staffing issues, care of PWCF in some centres is impeded by the lack of adequate infrastructural facilities for segregation and high-quality care delivery in an inpatient, outpatient and day care setting.

4.1.2 Adults

Adult CF services are delivered primarily in 5 designated centres in 4 distinct geographic regions (Cork, Dublin (2), Galway and Limerick). Paediatric services remain the primary referral pathway for the majority of PWCF and the transition process is usually completed by 18 years old. There are increasing 'new' referral pathways of late diagnosis of PWCF from male fertility services, non-CF Bronchiectasis and difficult Asthma clinics. This cohort requires extensive complex medical and genetic work up and counselling.

Day-to-day care is provided through daycentre facilities and these should be supported by inpatient designated protected beds (recommended figure of 5 inpatient beds per 50 adults attending a service). The ongoing development and maintenance of infrastructural day care and inpatient care facilities is critical to cater for increasing numbers as a 65% increase in adult numbers in Ireland is predicted between 2010 and 2025 (Burgel et al., 2015, CFRI, 2016). This will result in clear capacity issues and associated clinical risk, if not planned for and project managed in time. Equally importantly, and not catered for in the recent projections, are the emergence of new disease-modifying treatments for the majority of these PWCF which will potentially further increase the number of Adults with CF significantly through improved survival

and reduced rates of decline in pulmonary function (Davies et al., 2013, Elborn et al., 2016, McKone et al., 2014, Van Goor et al., 2014, Wainwright et al., 2015).

Disease complexity in adults with CF is now a significant challenge (Plant et al., 2013). Extrapulmonary manifestations of the disease are increasing as co-morbidities and multi-morbidities continue to change the dynamic of care in adults with CF ; for example, 30% of 30 year olds will have CF related diabetes (Moran et al., 2009), anxiety and depression is double that of the healthy population (Quittner et al., 2014) and renal replacement therapy for CF kidney disease is an increasing reality (Plant et al., 2013).

The inadequate numbers of appropriate staffing in medical, nursing and HSCP areas is a problem in delivering appropriate multidisciplinary care in keeping with best medical practice (Pollock, 2005). In most hospitals, the numbers of HSCPs, such as dietitians, nurses, pharmacists, physiotherapists, psychologists, secretarial and administrative staff and social workers are clearly inadequate. As with paediatric care, a system that protects whole time equivalent staffing for CF is necessary for safe care of PWCF and team stability.

4.2 Specialist CF centres

Specialist centres have been in operation in Ireland for many years and are recognised by the HSE as such (HSE, 2009). There are currently 6 designated paediatric centres and 5 designated adult centres.

4.2.1 Designated specialist CF centres

The HSE Report into CF services in 2009 has designated the following as specialist CF centres:

- Cork University Hospital (adult and paediatric CF Centres)
- Dublin North - Beaumont Hospital (adults) linked with Temple Street Children's University Hospital
- Dublin South – St. Vincent's University Hospital (adults) linked with OLCHC Crumlin (paediatric) and Tallaght Hospital (paediatric)
- University Hospital Galway (adult and paediatric CF Centres)
- University Hospital Limerick (adult and paediatric CF Centres)

The 3 Dublin Paediatric CF Centres will be amalgamated into a single Paediatric CF Centre in the new Children's Hospital.

Transition arrangements are in place between the adult and paediatric centres and these need to be strengthened and standardised in some cases.

4.2.2 Hospitals other than specialist CF centres currently providing services to PWCF

The most recently available data from the CFRI indicate that, in addition to the Specialist CF centres, the following hospitals also provided care for PWCF in 2017; some of these had shared care arrangements with Specialist CF centres:

- Cavan General Hospital
- Letterkenny University Hospital
- Mater Misericordiae University Hospital (Post Transplant)
- Mayo University Hospital
- Midland Regional Hospital at Tullamore
- Our Lady of Lourdes Hospital, Drogheda
- Portiuncula University Hospital
- Regional Hospital Mullingar
- Sligo University Hospital
- St. Luke's General Hospital Carlow/Kilkenny
- University Hospital Waterford

4.3 Shared care

No overarching guidance is currently provided in terms of the scope of CF shared care arrangements in Ireland.

In paediatrics, historic shared care arrangements exist between a number of local hospitals and specialist centres, although the degree to which this occurs, the centres providing shared care and the operational models of shared care are ad hoc and variable (HSE, 2009).

Adult services do not generally adopt a Shared Care approach, although some historic local ad-hoc arrangements are in place. The complexity of medical issues are generally best served through direct access to the designated CF centres and teams. Shared care is not recommended in the UK standards of care for adults with CF.

4.4 Newborn screening

A two tier newborn screening programme based on the heel prick test has been in operation in Ireland since July 2011. Infants diagnosed with CF through the CF newborn screening programme are referred to the specialist CF centre in the catchment area where the child was born (HSE, 2011). Parents of all children screened positive through the programme are referred for genetic counselling. As of now therefore, the vast majority of newly diagnosed children with CF are referred directly to a specialist CF centre.

4.5 Primary care

With the modernisation and evolution of specialist CF services concentrating on direct access to specialist treatment for CF health related issues, including those typically associated with primary care such as intercurrent infections, the role of primary care in managing day to day CF related issues has reduced. This includes paediatric care where complexity and comorbidity have reduced with improving outcomes and many children have all their care provided in an ambulatory setting. Primary care involvement is however vitally important because PWCF and their families will continue to require a relationship with their General Practitioner for some CF related issues (in partnership with the CF centre) and for non-CF related primary care issues including immunisation. Ensuring clear and consistent two-way communication between CF centres and General Practitioners/Primary care teams is vital. This should be aided by the introduction of electronic healthcare records and unique identifiers in the future.

5 Model of care 2019 onwards

A significant body of work has been involved in the development of models of care in other jurisdictions, much of which is similar between models, and is internationally applicable. Recent comprehensive recommendations on standards and models of care are relevant to the Irish context and form the basis of many of our recommendations. There are similarities between our Model of Care recommendations and those of other countries, but there are national and regional considerations in Ireland which will impact on some of our recommendations.

5.1 Principles of CF care

- All children born in Ireland should be screened for CF through the CF Newborn Screening Programme. The diagnosis of CF should not be delayed, must be handled sensitively and followed by education of the parents/carers and PWCF. All individuals (or their parents if minors) diagnosed with CF should receive counselling on the genetics of CF.
- All PWCF must be under the direct supervision of, and receive regular follow-up from, an adequately resourced, designated specialist CF centre (sometimes in partnership with a designated local shared care CF centre for children).
- Specialist multidisciplinary care must be delivered by a team of trained and experienced CF specialist health professionals, with staffing levels determined by the size of the PWCF population at a given centre. This should be regularly monitored, reviewed and where applicable rectified to ensure uninterrupted staffing levels and parity for all designated centres and their cohorts of PWCF. For CF Team members who are working largely or completely in CF Care, their primary reporting relationship should be with the CF Centre Director.
- For CF Consultants and CF Team members, there must be a designated replacement to fill their post when they are on scheduled or unscheduled leave. All CF Centres must have more than 1 CF Consultant.
- Treatment should be safe with a focus on PWCF health and wellbeing and should be of consistently high quality in line with national standards of care, independent of where it is delivered.
- Research, audit and quality improvement are key aspects of the CF care landscape and have led to significant improvements in outcomes. Research, audit and quality improvement should be seamlessly integrated with the clinical care of PWCF.
- The facilities available to specialist CF centres for inpatient, day care and outpatient care should be to a high standard and must be sufficient to prevent cross-infection between PWCF and from other patients.
- Treatment of exacerbations of CF is critical, and antibiotics are a key part, but not the only part, of treatment of exacerbations. Access to treatment in hospital should be direct

(avoiding the emergency department) and swift when required (within 24 hours for acute care and within 7 days for non-acute care).

- Respiratory physiotherapy with airway clearance and physical exercise are key lifelong parts of treatment and the most effective types of treatment usually change with time in the same PWCF. There is a need to ensure continued uninterrupted designated specialist support of this as part of daily outpatient and inpatient care.
- Nutritional support is crucial for all PWCF regardless of age. With the emergence of new therapies including disease modifying treatments, this is a dynamic evolving area where real time adjustments are critical. There is a need to ensure continued uninterrupted designated specialist support of this as part of on-going outpatient/day care and inpatient care.
- Psychosocial support is of great importance, clearly improves outcomes, and should be routinely available to all PWCF and, where appropriate, their families.
- Access to treatment through CF daycentres should be swift when required (within 24 hours for urgent care and within 7 days for non-urgent care).
- Regular measurement of Pulmonary Function Tests is crucially important for PWCF to monitor lung disease and to assess responses to treatment. Respiratory Muscle Strength, Cardiopulmonary Exercise Testing and Lung Clearance Index also need to be measured in some cases. Respiratory Physiologists have an essential role in this.
- Extrapulmonary manifestations of CF, as well as complications (e.g. liver disease, CF related diabetes, kidney disease, and occult malignancy as well as vascular access anomalies and antibiotic resistance) must be screened for as appropriate, recognised and dealt with through the CF MDT. With improved survival/aging this area is increasing and poses a considerable disease burden, with a need for significant additional resources, particularly for Adult CF centres.
- Transition to adult care should be planned and managed appropriately through a uniform national approach.
- Palliative and end-of-life care must be planned and managed appropriately, recognising the disease specific challenges that CF adds to this area (Cystic Fibrosis Trust, 2011). Palliative care is a very important ongoing part of treatment for many PWCF throughout their life; end-of-life care is an important part, but only a part, of Palliative care for PWCF.
- Post bereavement psychosocial support should be offered to families and partners of PWCF who have died
- Lung transplantation and, in some cases, liver transplantation and kidney transplantation are important parts of treatment for some PWCF. Transplantation services (including expedited transport to transplant centres) should be readily available to both children and adults with CF.

- Follow up of PWCF Post Lung Transplantation is very important; it is a complex and evolving issue internationally and it involves follow up for transplant related issues and also follow up for CF related issues. The NCPCF has established a NCPCF Transplant Subgroup, which will prepare a separate Model of Care document dedicated to the care of PWCF Post Transplantation; the recommendations of this present Model of Care do not apply to PWCF Post Lung Transplantation.
- Pregnant women with CF should attend an Obstetric Hospital that has experience of looking after women with CF and that has formal links with a designated CF Centre. The NCPCF will make further recommendations on Pregnancy and Fertility in CF in the future.
- The NCPCF strongly recommends that a National Cystic Fibrosis Office be established to coordinate, manage and integrate all aspects of CF care in order to optimise the health and quality of life of People with Cystic Fibrosis in Ireland

5.2 Specialist CF centres

Both the 2011 UK *Standards* document and the 2008 Australian document *Cystic Fibrosis Standards of Care, Australia* underlined the importance of the development of specialist centres of expertise for the delivery of CF care. An ideal system would ensure that each PWCF has access to a high level of clinical expertise and excellence on each occasion that they interact with the healthcare system. The challenge of implementing a system such as this – as is often the case in healthcare – is ensuring an appropriate balance between concentration of expertise and geographical access to care. In most situations, and most particularly with serious conditions such as CF, concentration of expertise and delivery of a high-quality service takes precedence over issues of geographical access. This has been seen in relation to cancer services where concentration of expertise in specialist centres has led to improvements in outcome.

Concentrating care for PWCF in specialist CF centres ensures that the multidisciplinary team will see sufficient numbers of PWCF to enable them to maintain expertise, so that they treat PWCF effectively, recognise the more unusual manifestations and delay the onset of the complications that occur in CF. The importance of Specialist CF Centre care cannot be over-emphasised, and has been recognised by the HSE (HSE, 2009). The US Cystic Fibrosis Foundation, the European Cystic Fibrosis Society, the Royal College of Paediatrics and Child Health and the British Thoracic Society all strongly endorse the principle and the importance of Specialist CF Centre care (Farrell et al., 2008, Jackson, 1996, Kerem et al., 2005, Smyth et al., 2014). Outcomes for children and adults attending designated specialist CF centres with multidisciplinary teams are superior, compared with those attending non-specialist clinics. The ideal model of care, therefore, would involve all PWCF in Ireland attending specialist centres for all of their care. It should be noted, however, that some PWCF do not live within close

commuting distance of a specialist CF centre in Ireland, and access to services for them can be challenging. The concept of shared care will be discussed later in this document.

With the advent of the hospital group structure in Ireland and, in particular, the grouping of the children's hospitals in Dublin into a single group pending the establishment of the new children's hospital, the official designation of current specialist CF centres should be revised as follows:

Children's Hospital Group

- OLCHC, Tallaght Hospital (paediatric) and CUH Temple St pending the establishment of the new children's hospital

Ireland East Hospital Group

- Dublin South - St Vincent's University Hospital (Adults)

RCSI Hospital Group

- Dublin North - Beaumont Hospital (Adults)

Saolta University Health Care Group

- University Hospital Galway adult centre
- University Hospital Galway paediatric centre

South/South West Hospital Group

- Cork University Hospital adult centre
- Cork University Hospital paediatric centre

University Limerick Hospitals Group

- University Hospital Limerick adult centre
- University Hospital Limerick paediatric centre

It is vitally important to maintain a high level of multidisciplinary expertise in both paediatric and adult specialist CF centres to ensure outcomes are optimised. Maintenance of expertise requires that MDT members see and treat a critical mass of PWCF, and so the number of PWCF per centre is important. The following Criteria must be met for the centres to be regarded as specialist centres:

- Designation as a Specialist CF Centre by the accreditation/peer review committee of the NCPCF (on the basis of the following).
- Each Specialist CF centre should care for at least 50 PWCF.
- Each specialist CF centre must have a core group of adequately trained and experienced healthcare professionals who are dedicated to CF care. For CF Team members who are working largely or completely in CF Care, their primary or sole reporting relationship should be with the CF Centre Director. For CF Consultants and CF Team members, there must be a designated replacement to fill that post when they are on scheduled or unscheduled leave

- The number of staff in each discipline should be appropriate for the number of PWCF attending that centre (see below):
    - Consultants in adult or paediatric respiratory medicine with a special interest in CF
    - Non-consultant hospital doctors (including Specialist Registrars)
    - CF Specialist Dietitians*
    - CF Nurse Specialists
    - CF Specialist Pharmacists*
    - CF Specialist Physiotherapists*
    - CF Specialist Psychologists * [Clinical or Counselling]
    - CF Secretarial and administrative staff
    - CF Specialist Social workers *
    - Data managers
    - Microbiology Laboratory scientists
    - Respiratory Physiologists (formerly Pulmonary Function Scientists)

*In these disciplines we strongly recommend that posts must be Clinical Specialist and Senior, because of the complex and changing health care needs of PWCF, with possibly a small number of rotating staff grade posts, where applicable, for training.

- All CF Centres must provide data to and assist in the collection of data by the CFRI, which collects data on behalf of the NCPCF
- Specialist centres must have direct access on site to the following resources:
    - Have dedicated single inpatient isolation rooms with en-suite facilities. Adult units need to have 5 inpatient rooms for each 50 PWCF attending the unit. Paediatric units need to have 3 inpatient rooms for each 50 PWCF attending the unit. Inpatient rooms must be available, and protocols must be in place to ensure that PWCF who are acutely sick are admitted immediately and always within 24 hours.
    - It is essential that there be an appropriate number of nurses and other staff for inpatient care. Because of the complexity and critical nature of inpatient CF Care, there should be at least 1 skilled nurse for each 3 CF inpatients during the day and 1 skilled nurse for each 4 inpatients during the night when all patients are stable. The ratio of nurses/patients must be greater when patients are unstable or critically ill
    - Sufficient dedicated day care rooms with en-suite facilities and outpatient rooms for assessment/treatment of PWCF as outpatients to facilitate the highest standards of assessment and treatment

- PWCF are at risk of picking up severe infections from other PWCF and from other patients. Cystic Fibrosis Centres must have strict infection control facilities and protocols in place for inpatient, outpatient and day care to prevent cross infection from other PWCF and from other patients. PWCF should not meet or mix with other PWCF in hospital or elsewhere
- Full pulmonary function laboratory capacity staffed by Respiratory Physiologists. The Pulmonary Function laboratory will measure pulmonary function tests routinely and also Respiratory Muscle Strength, Cardiopulmonary Exercise Testing and Lung Clearance Index in some cases
- A Microbiology laboratory skilled in the specific evaluation of CF specimens and following International CF specific guidelines
- Chest CT (including under sedation or anaesthesia in children)
- Same day processing of therapeutic drug levels
- CF Centres must have Consultants with a special interest in Cystic Fibrosis in the following specialties: Endocrinology, Gastroenterology, Hepatology, Nephrology, Oto-Rhino-Laryngology (ENT), Palliative Care, Psychiatry, Radiology including Interventional Radiology and Thoracic Surgery. Those CF Centres who do not yet have one or more of these on site should have clear referral pathways to them e.g. through the national paediatric hospital for paediatric centres.
- Access to occupational therapy services, either through specialist centres or the community is important for a subset of children and adults with CF, particularly adults with more advanced disease.

It is of fundamental importance that appropriate staff numbers are maintained to ensure safe, efficient and effective care of PWCF in all CF Centres. Table 1 outlines the staffing of CF centres per 50 PWCF that are necessary for safe patient care. These were developed and benchmarked with reference to international recommendations. It is crucial to emphasise that these figures relate to whole time equivalent spent in the direct delivery of CF care; if, for example, a fulltime CF team member spends 60% of their time delivering CF care and 40% delivering other care, they contribute 0.6 whole time equivalent to CF.

It is vital that, in every discipline, ongoing staff training and continuing professional development are available to ensure ongoing high standards. Funding should be made available routinely for all CF team members for this.

These staffing recommendations are appropriate in 2019. They should be reviewed and may need to be increased in the future as the complexity of CF and the complexity of CF care are expected to increase even more in the future.

Of vital importance in the future will be the need for ongoing assessment of numbers of PWCF attending and the staffing levels and resourcing at each centre to ensure that centres continue to meet the criteria laid out in the model of care for CF in Ireland.

To do this, it is crucially important that the accreditation/peer review programme recommended by the NCPCF be properly resourced. Given the specialist nature of CF services and the limited overlap with other disciplines, funding for CF staffing should be ring-fenced. This will ensure ongoing maintenance of appropriate staffing levels and facilitate accreditation/peer review and clinical governance.

Table 1. Recommended whole time equivalent staffing levels for specialist paediatric and adult CF centres per 50 PWCF

Staff member	Paediatric	Adult
Consultant	0.7	0.9
CF Specialist Registrar	0.5	0.5
CF Nurse	1.5	1.5
Physiotherapist	2.0	2.0
Dietitian	0.5	0.6
Social worker	0.4	0.5
Psychologist	0.5	0.6
Secretary	0.8	0.8
Data clerk	0.25	0.25
Pharmacist	0.4	0.5
Respiratory Physiologist	0.5	0.6
Medical Laboratory Scientist	0.7	0.7

*see appendix 3

5.2.1 Challenges in specialist CF centre care

A recent EU report (Burgel et al., 2015) based on existing CF registry data (including Irish CF registry data) reported that, with population growth and increased survival rates, overall the number of PWCF will rise by 50% between 2010 and 2025. This represents an increase of 21% and 65% respectively of children and adults with CF accessing services. These projections predate the emergence of new disease modifying drugs and therefore may under-represent growth (through greater survival). The expected increase in numbers will likely lead to higher numbers of PWCF in many centres and this should be taken into account in future planning.

Lung transplantation is now an important opportunity for many PWCF with end stage CF lung disease. The care of patients post lung transplantation is still in evolution in Ireland. Currently many PWCF who had lung transplantation are prospectively followed up for all their care under the transplant programme at the Mater Hospital. Other PWCF, often from centres outside Dublin, receive care from a combination of the transplant centre and their local Specialist CF centre. The NCPCF has established a NCPCF Transplant Subgroup, which will prepare a separate Model of Care document dedicated to the care of PWCF Post Transplantation; the recommendations of this present Model of Care do not apply to PWCF Post Lung Transplantation.

Co-morbidity in CF is increasing with improved survival, particularly in adult patients. The CF MDT is heavily reliant on other medical teams to deliver a high standard of care to our patients. Conditions such as liver disease, diabetes, bowel disease, renal disease and malignancy can impose a significant burden on other services, and this should be kept in mind in terms of resources. As highlighted previously, CF Centres should have Consultants with a special interest in Cystic Fibrosis in the following specialties: Endocrinology, Gastroenterology, Hepatology, Nephrology, Oto-Rhino-Laryngology (ENT), Palliative Care, Psychiatry, Radiology including Interventional Radiology and Thoracic Surgery.

The new children's hospital in Dublin will incorporate the 3 existing paediatric hospitals. Based on current numbers and predicted growth in the future it is expected that at the time of opening there will be approximately 350 PWCF in the CF centre. Despite the high collective numbers in the new children's hospital, an alternate centre does not exist for the care of children in the greater Dublin area. Management of the operation of the centre in the context of these high numbers will need to be carefully planned.

A national microbiology reference laboratory for CF is not currently designated. The 2009 HSE report suggested that existing services available in Tallaght Hospital be supported, pending the establishment of a national microbiology reference laboratory. The CF microbiology lead in Tallaght has retired and the CF centre in Tallaght will close in due course with the opening of the new children's hospital. It is recommended that a National Reference Laboratory for CF Microbiology be designated via a formal consultative process through NCPCF in conjunction with the CF subgroup of the Irish Society of Clinical Microbiologists. In the interim the microbiology laboratory at St Vincent's University Hospital (the current largest CF centre) should serve as the microbiology reference laboratory.

5.3 Shared CF care for paediatrics

There have been impressive improvements in outcomes in the last 30 years in CF, and this predates the development of novel disease modifying treatments. This is largely due to attention to detail by CF Teams and improvements in the organisation, delivery and audit of

care at centre and national levels. CF care has become more subspecialised, treatment thresholds are reducing, and the understanding of early CF lung disease is increasing. It is the access to experience and expertise at every interaction with the service that is the key difference between local and specialist services. Care in children with CF has become more about health maintenance and prevention of illness as opposed to treatment of complications alone. Much care is therefore delivered in an ambulatory setting in the paediatric age group and is planned. Access to expert opinion at all times is key to maintaining high standards. Specialist services with larger numbers and greater staff pools will be in a position to reliably provide a higher quality of care on a more consistent basis. It is vital that future services for children with CF are planned on the basis of having a robust system in place that can ensure quality outcomes as opposed to depending on an historic ad hoc model that relies upon individuals in individual centres without robust backup. With a view to maximising quality of outcomes and ensuring equity of access to services, all children should have their care delivered by a specialist CF centre. Local centres may also be involved in the care of children with CF under the umbrella of the specialist centre, and in an arrangement that optimises the balance between excellence and access.

The concept of a 'shared care' approach to CF features more in the paediatric as opposed to the adult literature. In other countries, including the UK, specialist Paediatric CF centres work with local hospitals in an attempt to provide specialist care closer to home while maintaining a high standard of care. Outcomes in unsupervised, locally based systems working in isolation have been shown to be worse but approach specialist care levels where a local hospital works in a partnership under the supervision of a specialist CF centre (Doull et al., 2012, Doull, 2012). This type of system is generally more resource intensive but can be particularly useful where distances are great and access to specialist services challenging. There has been a shared care system in some CF centres in Ireland for many years, but it is not arranged in a geographical distribution, does not operate under any governance framework and there are no clear guidelines or standards for the operation of shared care in these centres.

A shared care model can work well in terms of allowing access to expertise at all times and, where possible, minimising unnecessary travel for families. The degree of care shared locally will depend on the facilities and staffing available to the local centre, their ability to provide the required level of expertise to ensure high standards of care, and the need for local services. Shared care centres can provide anything from minimal commitments on an infrequent basis to more significant and regular commitments, as long as clear communication is in place and as long as high clinical standards are maintained.

The following guidance is general and designed to ensure a satisfactory minimum standard. Specific further arrangements may be necessary or desirable in certain settings of shared care between local hospitals and specialist centres. The specialist CF centre has the responsibility

to ensure that shared care services between the two hospitals are consistently monitored to ensure that a high standard of services is being maintained. If a satisfactory standard cannot be maintained, a shared care model should not be used.

For the system to work well there must be rapid and effective communication between the Shared Care local hospital and the specialist CF centre at all times. The requirement for effective communication cannot be overstated. It is vital that all records relating to the care and management of children with CF, and all of their interactions with healthcare professionals, are maintained in a central location at the specialist CF centre. Clear and unambiguous written guidance should be provided to the local hospital to aid them in providing services to children with CF at each clinical interaction, consistent with the practices of the specialist centre.

5.4 Model of Shared CF Care

- All infants diagnosed with CF through newborn screening or otherwise must be referred to a specialist CF centre.
- All PWCF must have their care delivered by a recognised specialist CF centre throughout their lives.
- Given the increasing complexity of CF in the adult population, adults should receive all their CF care directly from a specialist CF centre.
- Children will either receive full care from a specialist CF centre, or in some cases shared care with a designated Shared Care Centre at a local hospital.
- In shared care arrangements, the child, family and local hospital should be aware that they are a patient of the specialist centre.
- Care delivered by a combination of a designated Shared Care Centre and a specialist CF centre should be to the same standard as that delivered exclusively by the specialist CF centre.
- Shared care should be delivered as part of an agreed service plan with a service-level agreement, standard operating procedures and clinical guidelines *as laid down by the specialist CF centre*.
- Clear ongoing communication between the multidisciplinary team in the specialist CF centre and staff at local hospitals is essential for patient safety and for the maintenance of high standards of care.
- Shared care of children with CF should be seen as fluid. Depending on the resources available at local hospitals, in primary care or within hospital groups, the specialist centre can work with other centres to ensure the appropriate delivery of high-quality services to their PWCF. The degree of shared care may differ between hospital groups based on availability of resources and staffing.

- The responsibility for the management of the quality of the shared care arrangement, and the quality of the care for PWCF, rests with the specialist CF centre. If the quality of local hospital services cannot be safely maintained (e.g. due to loss of staff or facilities), care for PWCF should be delivered exclusively at the specialist CF centre.

5.4.1 Delivery of care to children with CF in a shared care model - CF clinic attendances every 3 months

All clinic attendances every 3 months should be at the specialist centre or by the team from the specialist centre at an outreach clinic. At clinic visits, the child should see the CF dietitian, CF nurse, CF physiotherapist and CF consultant/CF Registrar at a minimum. This will allow consistent delivery of a high standard of CF care to the child with minimal variability and clear communication with the child and family.

5.4.2 Scheduled diagnostics – radiology, bloods, others

Ideally all scheduled diagnostics should occur in the specialist centre and should be arranged to coincide with scheduled clinic appointments to prevent unnecessary travel. This will ensure availability of results in the laboratory system at the specialist centre when required. Certain investigations including oral glucose tolerance tests may be performed at local centres. In such situations results should always be forwarded to the specialist centre, ideally between laboratories to ensure continuity of results within the specialist centre's laboratory system. The availability in the future of the MedLIS system should ensure a single laboratory record for each patient and will allow scope for some diagnostic testing to be performed closer to home.

5.4.3 Respiratory diagnostics – pulmonary function testing and airway microbiology

Microbiology samples are a key aspect of CF care and considerable expertise is required to perform microbiology testing on CF samples to an acceptable international standard. Laboratories in specialist centres processing large volumes of samples are in the best position to process these samples to the required standards. It is also of key importance that results are available in the laboratory system of the specialist centre when required. Therefore, it is recommended that airway microbiology samples should always be sent to the specialist centre laboratory. For any samples sent to laboratories other than the specialist centre, reports should always be forwarded to the specialist centre, ideally between laboratories to ensure continuity of results within the specialist centre's laboratory system.

Pulmonary function testing is a core component of diagnostics in CF and is both operator dependant and dependant also on equipment quality and calibration. One of the key outcomes from pulmonary function testing is the assessment of change in values over time. This requires having many measurements, preferably in the same centre over time with a consistent measurement approach provided by highly trained respiratory physiology scientists.

For best results and to ensure adequate pulmonary surveillance, Pulmonary function testing should always be performed in the specialist centre.

In some instances, where distances are great and frequent measurements may be needed, pulmonary function testing can be performed in the local centre, however this should be co-ordinated with the pulmonary function laboratory in the specialist centre to ensure the appropriate equipment, techniques and standards are used so that the collected data is clinically relevant.

5.4.4 Scheduled Day-case procedures – port flushes, PEG care, dressings etc.

Many day case procedures, particularly ones required on a regular basis like monthly port flushes can be performed at the local centre, or indeed in a primary care setting, as applicable, as long as the required expertise and experience is available.

5.4.5 Scheduled clinical reviews

Scheduled clinical reviews for example post discharge from hospital, during home IV antibiotics or to monitor the progress of acute illnesses should be performed by the specialist team to ensure consistency of approach for PWCF and within defined illnesses.

5.4.6 Unscheduled reviews

Unscheduled reviews will usually arise when PWCF are unwell. Communications from PWCF or from families should go directly to the CF nurse specialists at the specialist CF centre. In some instances, MDT advice can be given over the phone and prescriptions emailed or faxed, reducing the need for face to face review; only the CF team can decide if this is appropriate. If clinical review is required, there may be a requirement for point of care diagnostics as well as clinical assessment by the MDT and, of key importance, access to specialist opinion. Unscheduled reviews therefore should always be conducted by the specialist CF team. This is one of the most important times that the PWCF will require input from the MDT at the specialist centre. In children, as in adults, much of the clinical assessment involves experience and clinical judgement. Clinical judgement in these circumstances is informed by subspecialty expertise, experience, volume of similar PWCF and knowledge of potential pitfalls. Given this requirement there is a danger of significant variability in approach if clinical opinion is being sought from individuals at different locations, particularly those not experienced in the management of large numbers of children with CF.

5.4.7 Admission to hospital for inpatient treatment

The acuity and complexity of inpatient treatment for CF is variable in children. In situations where disease is complex, where the PWCF's condition is changeable or where daily specialist centre MDT input is required, the PWCF should be an inpatient at the specialist centre. In some situations, such as where children are on an elective IV schedule, where disease is stable but

prolonged IV therapy is necessary or where completion of a course of IV therapy at lower acuity is envisaged, IV therapy may be delivered at local centres as long as the following resources are in place to facilitate this. The following are considered minimum requirements to be able to deliver IV antibiotic therapy to children with CF:

- Isolation rooms (with en-suite facilities)
- Access to paediatric physiotherapy
- Access to same day therapeutic drug levels (aminoglycosides)

Inpatient treatment supervision should be provided by the specialist centre with key treatment decisions made in conjunction with the specialist centre. Availability of paediatric dietitian and pharmacy staff is important. These staff members would be expected to liaise with their counterparts in the specialist centre. It is recommended that PWCF are seen at least at the start and the end of IV treatments in the specialist centre for clinical review and diagnostic testing. Decisions regarding suitability for shared care inpatient treatment should be continuously reviewed and are the responsibility of the specialist centre.

5.4.8 Clinical decision making

It is vitally important to clarify this in a shared care model, as otherwise this area is ripe for error and inconsistency. In general, all clinical decisions regarding treatment choices in CF should be made by the specialist centre team to ensure a consistent approach, even in what would appear to be straightforward situations such as whether or not to use antibiotics for respiratory symptoms, what type and what dose. This might seem to be a straightforward question for many non-CF clinicians including GPs; however, decision making is very different in the case of CF compared to otherwise healthy children or children with other disorders.

5.5 Shared care centres - Requirements

5.5.1 Staff

The following team members are essential to ensure a high standard of clinical care at the local Shared Care hospital:

- Consultant paediatrician nominated as Local CF lead. This person should have an interest in CF and will be responsible for the organisation and provision of services for children with CF in the local hospital and for liaison with the Centre Director in the CF centre. There must also be another Consultant paediatrician nominated to fill this role when the Local CF lead is on leave
- Liaison Nurse – the Liaison Nurse, at a nurse specialist level, should be familiar with aspects of CF care such as port flushes, PEG care, OGTT. Liaison nurses will have a close working relationship with the CF CNSs at the specialist centre.

- Paediatric Physiotherapist – with training in respiratory physiotherapy who can provide predominantly inpatient physiotherapy services as required and, in some cases, outpatient resources, under the supervision of the specialist CF physiotherapist at the specialist CF centre.
- Paediatric Dietitian – who can provide predominantly inpatient dietetic advice and supervision when required and, in some cases, outpatient resources, under the supervision of the specialist dietitian at the specialist CF centre.

It is envisaged that all of the above staff will be part of the general paediatric team in the local paediatric centre and provide services to children with CF as part of a wider general paediatric remit. For each post, there must be a replacement to fill that post when they are on scheduled or unscheduled leave.

5.5.2 Facilities

For inpatient care the Shared Care Centres should provide single isolation rooms with en-suite facilities and preferably with parent accommodation. For day-case visits Shared Care Centres are expected to provide single rooms, with a facility to ensure adequate infection control precautions during the PWCF's stay. Shared care centres are expected to be capable of performing basic laboratory tests and therapeutic drug levels (aminoglycosides).

5.5.3 Communication

Parents of children with CF should always contact the CF nurse specialist at the specialist CF centre as the first port of call. The CF nurse specialist will act as the central communication point to facilitate working with the local Shared care centre and specialist centre to ensure the appropriate balance between convenience and access to appropriate services at each interaction of the child with CF with medical services.

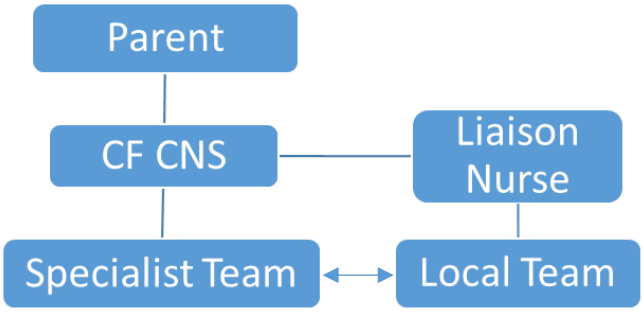

Figure 1. Communication pathway for children with CF

5.5.4 Specialist Centre Care, Shared Care, Outreach

The majority of CF services can be delivered efficiently at specialist CF centres as long as families live within approximately two hours travel time from the nearest CF centre. Even in this context, situations may arise where some aspects of care can be seamlessly provided at a local centre closer to the child's home as laid out above. In situations where children may live

more than approximately 2 hours from their nearest specialist centre, consideration should be given to providing outreach services at local hospitals where the number of PWCF merits this approach. In these situations, the CF team from the specialist centre would travel together to run scheduled clinics at local hospitals. This approach is best suited to a situation where all centres are within one hospital group. The northwest of Ireland (Saolta University Healthcare Group) in particular may benefit from this approach. Here, the specialist centre is located at one end of the geographic area, and distances can be great with up to 5 hours of travel time. There are 3 local hospitals in this area with paediatric units, all of which currently offer some level of CF service, but not in a uniform co-ordinated fashion. The resourcing of CF services in this area in particular should take account of the unique population needs and the desire to provide an outreach service. Outreach services should be agreed as part of the service level agreement between the centres. Outreach services require an adequate number of CF Team members so that services continue to be provided at the CF Centre while some team members are at the Shared Care Centre; if this is not the case, outreach services cannot be provided.

5.5.5 Arrangement of shared care

Where possible, shared care is best served by linking specialist CF centres and local hospitals within hospital groups. This will help significantly in terms of development of outreach, funding models and service level agreements. This is not possible in Dublin as the children's hospitals themselves form a group. The following shared care arrangements should be put in place for newly diagnosed children where shared care is appropriate, and existing arrangements amended over time to reflect this organisation. Some children's hospitals in the following list are not currently providing services to children with CF and this guideline does not oblige them to do so; rather it outlines referral patterns to guide future arrangements that might be put in place.

Cork University Hospital

- South Tipperary General Hospital
- University Hospital Kerry
- University Hospital Waterford

New Children's Hospital (Currently OLCHC, CUHTS, Tallaght)

- Cavan General Hospital
- Midland Regional Hospital, Mullingar
- Midland Regional Hospital, Portlaoise
- Our Lady of Lourdes, Drogheda
- St. Luke's General Hospital Carlow/Kilkenny
- Wexford General Hospital

University Hospital Galway

- Letterkenny University Hospital
- Mayo University Hospital
- Sligo University Hospital
- Portiuncula University Hospital

University Hospital Limerick

- No other paediatric hospitals in hospital group

5.6 Governance

Clear governance arrangements should be in place both at specialist CF centres and at a national level through the NCPCF programme.

5.6.1 National Clinical Programme for Cystic Fibrosis

The NCPCF is the body that will plan and direct the delivery of care to PWCF. The membership of the NCPCF Working Group is multidisciplinary including nominees from different groups involved in the care of PWCF, including nominees from the Cystic Fibrosis Centres and Shared Care Centres, Cystic Fibrosis Team Members, Cystic Fibrosis Ireland, Cystic Fibrosis Registry of Ireland, HSE Acute Hospitals Division and others.

The NCPCF is governed by a dual governance structure comprising:

- A Clinical Advisory Group the chair of which reports to the President of the RCPI
- The National Clinical Advisor and Group Lead for Acute Hospitals

This governance structure is illustrated in Appendix 2.

The NCPCF includes a number of workstream subgroups chaired by subject matter experts reporting to the NCPCF Clinical Lead and the Working Group. All documentation, policies, etc. produced by each subgroup must be first signed off by the subgroup itself, followed by approval by the wider NCPCF working group and then by the Clinical Advisory group and the CSPD Senior Management Team prior to submission to HSE Leadership.

The Patient Data/Information Systems Subgroup is developing a system to collect, manage and collate PWCF information on a national basis. The CFRI has a crucial role in this collection of accurate data. This is essential for the planning of CF care in the short and long term.

The recent initiation of a regular NCPCF national audit/quality assurance assessment involving all Specialist CF Centres and Shared Care Centres and relevant stakeholders would provide a unique opportunity for those involved in CF care to continue to improve the quality of care for all PWCF. The NCPCF has established a Data Committee which will lead the data and quality agenda in conjunction with the National Clinical Lead and Programme Manager.

The NCPCF strongly recommends that a National Cystic Fibrosis Office be established to coordinate, manage and integrate all aspects of CF care in order to optimise the health and quality of life of People with Cystic Fibrosis in Ireland

5.6.2 Service

Each specialist CF centre should have a CF centre director. The centre director should be a senior CF consultant with experience in management and leadership. The centre director will be responsible for all aspects of the delivery and audit of CF care in the centre. The CF centre director will report through the local clinical directorate structure in terms of clinical operations and local governance. The CF centre director will report to the national clinical lead in CF in relation to issues concerning the delivery of CF care, standards and accreditation/peer review. For CF Team members who are working largely or solely in CF Care, their primary reporting relationship should be with the CF Centre Director.

Each centre should have a PWCF (or parent/guardian in Paediatrics) representative and a nominated staff member (client liaison) charged with liaison with the PWCF representative. Regular scheduled communication between the centre staff and PWCF representative is encouraged.

5.7 Transfer of care of PWCF between CF centres

Because of the life-long and changing nature of CF, continuity of care is very important. Many PWCF will attend one Paediatric CF Centre during childhood and adolescence and, after transition, they will attend one Adult CF Centre. Apart from transition from Paediatric to Adult care, there are a number of reasons why the care of a PWCF may be transferred to another CF Centre.

5.7.1 Transfer for geographical reasons

If a PWCF moves to a different place that is nearer another CF Centre, their care may be transferred to the other CF Centre if they wish. The PWCF or their parent/guardian should inform their current CF Centre that they want to move Centre; if possible, that should happen at least 3 months before the move so that the transfer of care can be planned. The current CF Centre will then send a Transfer Letter and relevant documents to the new CF Centre

5.7.2 Transfer for health reasons

In rare situations, transfer to another CF Centre may be appropriate if the PWCF needs specialised health care resources that are available in the other CF Centre and are not available in the current CF Centre. In some cases, the Transfer of Care will be permanent and in others it may be temporary. In other cases, a Consultation may be obtained from the other CF Centre without a Transfer of Care

5.7.3 Transfer because of communication/relationship issues between the PWCF and the CF Team

On rare occasions, a Transfer of Care may be necessary because of communication/ relationship issues between the PWCF and the CF Team.

If a PWCF or their parent/guardian are concerned at communication/relationship issues with the CF Team, they should inform the CF team of the concerns and should request a meeting with the CF team to discuss these. If they are still concerned at the issues after the meeting and they want a Transfer of Care to another CF Centre, they should indicate that in writing to the CF Team and specify what CF Centre they want to transfer to. The CF Team will then send a Transfer Letter and relevant documents to the new CF Centre.

If a CF Team are concerned at communication/relationship issues with a PWCF or their parent/guardian/family, they should inform them of the concerns and should arrange a meeting with them to discuss these. If the CF Team are still concerned at the issues after the meeting and they feel that the issues are a barrier to the health or safety or quality of Life of either the PWCF or CF Team members, they should arrange a transfer of care to another CF Centre. The CF Team should indicate in writing to the PWCF or their parent/guardian that they are arranging a Transfer of Care. The CF Team will then send a Transfer Letter and relevant documents to the new CF Centre. If a PWCF or their parent/guardian are unhappy with this Transfer of Care, they may appeal the decision by writing to the NCPCF. The NCPCF will appoint a committee to assess this and the committee will have no members from either the transferring Centre or the Centre to which they are being transferred.

5.8 Future developments in CF care

5.8.1 CF Registry of Ireland

The development and maintenance of disease registries is key to understanding changes in patterns of disease and outcomes, particularly as they relate to cause and effect. The CFRI has been in operation since 2001 and is part of the European CF registry. CFRI operates under a service level agreement with the HSE. Historically registries relied upon retrospective data entry and so entries were at times delayed and incomplete. International registries, including the Irish and European CF registries, want to transition towards live data entry at the point of assessment and treatment thus allowing rapid access to live registry data and rapid access for the clinician and PWCF to trends. This exciting development in data collection and use, when it is set up, will improve the experience of the PWCF and will be a very useful tool in audit, benchmarking and accreditation/peer review. It will also be of significant value to CF researchers.

5.8.2 Recruitment and retention of CF team members and nursing staff on inpatient CF wards

Recruitment and retention of health care professionals who look after PWCF is a major issue internationally for several reasons. Firstly, treating people with a severe chronic illness over a number of years has always been stressful, especially when CF Teams treat a number of PWCF who deteriorate and die. Secondly, the expectations of the public for health improvement nowadays exceed the reality of day to day CF care; despite new "disease modifying treatments", CF is still a chronic condition which causes major health issues and is an ongoing burden for PWCF. This "expectations gap" is stressful for PWCF and their families and also for CF health care professionals. The rare cases of abuse (personal and on social media) of CF Team members and Nurses on CF wards has also made it harder to recruit and retain CF team members.

Recruitment and retention of CF team members and also nurses on inpatient CF Wards is and will be a very important issue for care of PWCF in Ireland, especially with the expected increase in the number of PWCF. It is essential that CF team members and also nurses on inpatient CF Wards have time and funding available for regular continuing professional education and are provided with the appropriate supports to enable them to deal effectively with this high stress environment.

5.8.3 Incorporation of new national eHealth initiatives

New national initiatives such as the national medical laboratory system (MedLIS), individual health identifier (IHI), electronic health record (EHR) are likely to have significant positive impacts on management of PWCF particularly when it comes to promotion of efficiency and reduction in duplication, medication reconciliation, referral of PWCF between centres, communications between clinicians, service access for users and safety of PWCF. Given the level of interaction with healthcare professionals involved in the management of their CF, this is likely to be particularly beneficial for PWCF.

5.8.4 Telemedicine for outreach, virtual clinics

In areas with challenging geographical access, where outreach and virtual clinics could be of benefit, rollout of high-speed broadband to remote areas may facilitate more regular and useful staff/PWCF interactions. This would complement, not replace, the need for regular face to face meetings with the CF team. The development of home testing solutions might further add to this.

5.8.5 Research

Support for basic science, clinical and translational research in CF is one of the reasons that outcomes for our patients have improved so dramatically in the past 30 years. Ireland has a

strong track record in CF research, but this has happened in an ad hoc and often fortuitous way in the past and has been very dependent on individuals. It is vitally important for the future of CF care and outcomes in Ireland that research is strongly integrated with clinical care. The NCPCF recognises the importance of ensuring that the organisation of CF care in Ireland lends itself to the conduct of high quality, clinically relevant research.

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Appendix 1

Abbreviations

AHD – Acute Hospitals Division

BH – Beaumont Hospital

BCH – Belfast City Hospital

CF – Cystic Fibrosis

CFI – Cystic Fibrosis Ireland

CFRI – Cystic Fibrosis Registry of Ireland

CFTR – Cystic Fibrosis Transmembrane Conductance Regulator

CGH – Cavan General Hospital

CUH – Cork University Hospital

CUHTS – Children's university Hospital Temple Street

HSE – Health Service Executive

MDT – Multi-Disciplinary Team

MMUH – Mater Misericordiae University Hospital

NCP – National Clinical Programme

NCPCF – National Clinical Programme for Cystic Fibrosis

ODTI – Organ Donation and Transplant Ireland

OLCHC – Our Lady's Children's Hospital Crumlin

PWCF – Person/People with Cystic Fibrosis

RCPI – Royal College of Physicians of Ireland

RCSI – Royal College of Surgeons in Ireland

SVUH – St. Vincent's University Hospital

TH – Tallaght Hospital

UHG – University Hospital Galway

UK – United Kingdom

UHL – University Hospital Limerick

UHW – University Hospital Waterford

Appendix 2

Structure and Governance of the RCPI National Clinical Programmes

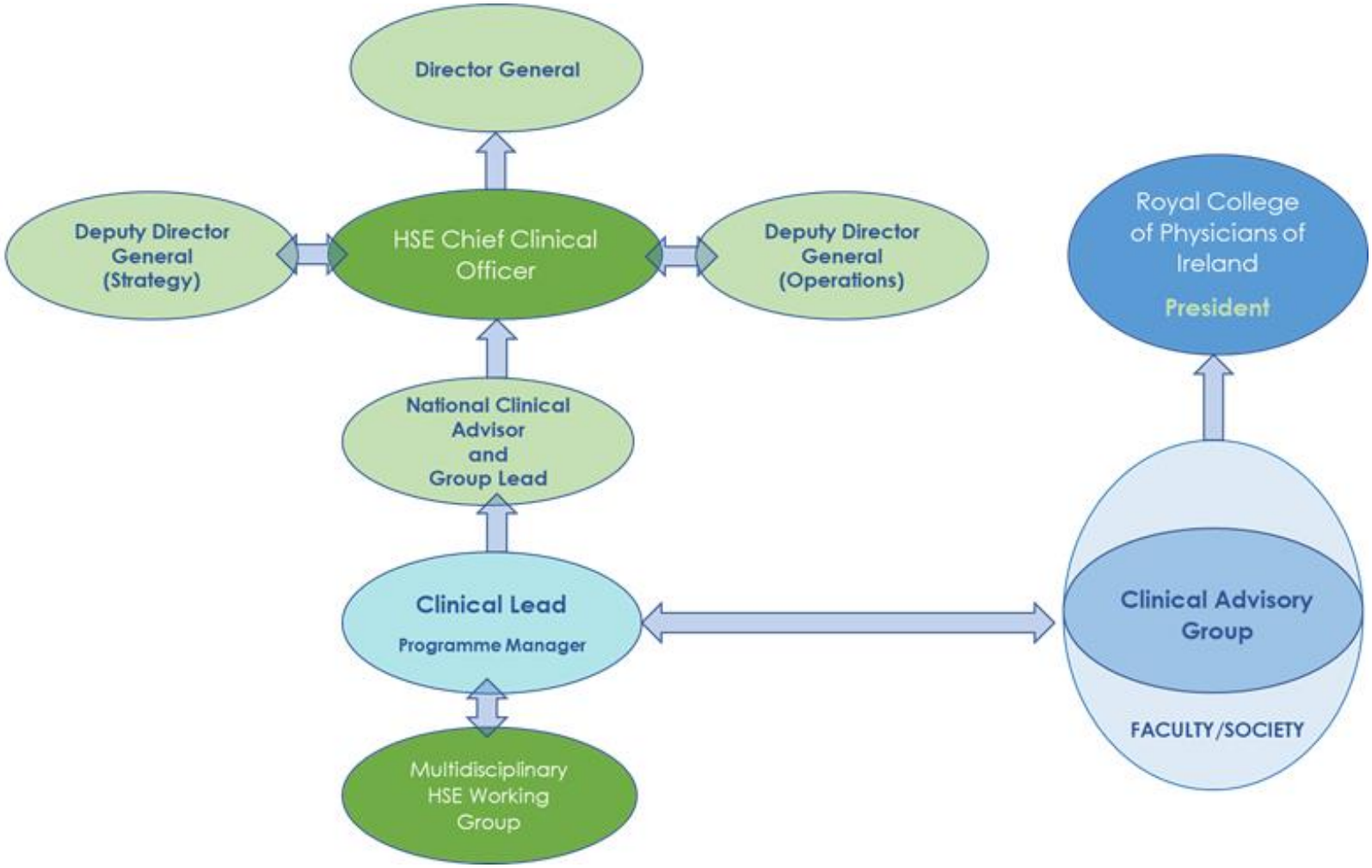

Figure 2. Structure and Governance of the RCPI National Clinical Programmes

Appendix 3

Notes on staffing ratios contained in Table 1

Consultant posts in the HSE document from 2009 were allocated as consultant 1 and consultant 2. In practice this has never been applicable in an Irish context. On the basis that consultant 1 would spend the vast majority of their time on CF and consultant 2 a lesser proportion of their time we have recalculated to a total WTE figure for consultant time spent in the delivery of CF care. It is crucial to emphasise that, as for all CF team members in Table 1, these figures relate to whole time equivalent spent in the direct delivery of CF care.

We recommend an increase in WTE allocation for dietitians on the basis of the increasing complexity of CF patient workload with both the introduction of CF newborn screening and the prolonged survival and complexity in adult nutritional CF care.

We recommend increasing WTE for adult and paediatric pharmacists from 0.3 to 0.5 and 0.4 respectively. This is due to the increase in complexity and workload with the introduction of CFTR modulators, particularly in adults where medication interactions are common.

We have amended the recommendations for secretaries downward from 1.0 to 0.8 and increased the data clerk WTE from 0.1 to 0.25. Increasingly in CF the accurate and timely use of data is vital in terms of service planning, audit, quality and importantly, working with the CF registry to ensure outcomes are captured in individuals on CFTR modulator compounds.

Psychology allocation was increased in children from 0.4 to 0.5 and in adults from 0.4 to 0.6 on the basis of the increase in psychological morbidity and mental health issues with prolonged survival and disease comorbidity particularly in adult patients.

Appendix 4

Document approval and consultation methodology

The draft model of care document was approved by the model of care subgroup before being circulated to the wider National Clinical Programme for Cystic Fibrosis Working Group for a formal internal consultation process. Membership of both these groups can be found on pages i and ii.

Following this process, amendments and inclusions were made to the draft document prior to submission to a formal consultation process with the National Clinical Programme for Cystic Fibrosis Clinical Advisory Group. Membership of this group can be found on page iii. The document was again revised based on feedback received and approved.

Once approved by the National Clinical Programme for Cystic Fibrosis Working Group and Clinical Advisory Group, the document was submitted to a national open external consultation process. The document was circulated to all relevant organisations and individuals for comment. Each organisation and individual were also given the option to forward the document on to anyone they felt appropriate.

Forty-four (44) individual submissions were received during this process. These submissions were logged individually in their entirety and also separated chronologically relating to the relevant pages and sections of the draft document. Following this process, amendments and inclusions were made to the draft document prior to submission to the HSE National Clinical Advisor and Group Lead for Acute Hospitals for final HSE approval before publication.

Figure 3 illustrates the above process.

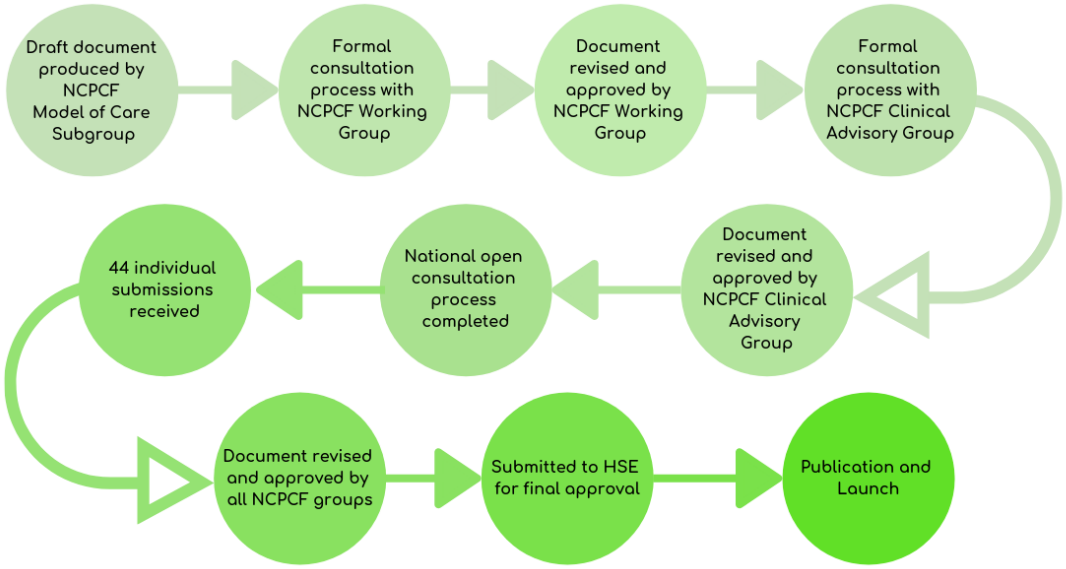

Figure 3. Model of care approval methodology

CYSTIC FIBROSIS

Feidhmeannacht na Seirbhíse Sláinte
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