



PAEDIATRICS

A NATIONAL MODEL OF CARE FOR PAEDIATRIC HEALTHCARE SERVICES IN IRELAND

CHAPTER 42: PAEDIATRIC RESPIRATORY MEDICINE



Féidhmeannacht na Seirbhíse Sláinte
Health Service Executive

Clinical Strategy and Programmes Division



**ROYAL
COLLEGE OF
PHYSICIANS
OF IRELAND**

TABLE OF CONTENTS

42.0	Introduction	2
42.1	Evolution of Services	4
42.2	Current Service Provision	9
42.3	Proposed Model of Care	22
42.4	Requirements for Successful Implementation of Model of Care	39
42.5	Programme Metrics and Evaluation	51
42.6	Key recommendations	55
42.7	Abbreviations and Acronyms	55
42.8	References	56
42.9	Appendices	58

42.0 INTRODUCTION

Ireland has a strong network of paediatric respiratory subspecialists in tertiary and regional centres built around providing a service to children with cystic fibrosis. This lends itself very well to developing a coherent national network of paediatric respiratory centres capable of providing a range of subspecialty services at different levels of complexity. Paediatric respiratory medicine is a subspecialty that is seeing significant change internationally. The areas of sleep and long-term invasive and non-invasive ventilation are growing at a rapid rate internationally and becoming a large part of the service provided by specialists in paediatric respiratory medicine. There is sufficient volume and complexity in the areas of cystic fibrosis, paediatric sleep medicine, long-term ventilation and poorly controlled asthma to warrant the development of regional centres of expertise in paediatric respiratory medicine, as part of a national hub-and-spoke model, which are appropriately staffed and resourced.

The current model of practice in the tertiary subspecialty centres in Dublin, which includes the provision of general paediatric care, involves a variable scope of practice across three sites. This needs to change, in order to reflect both the change in make-up of the subspecialty internationally and the development of the new children's hospital in Dublin. Significant investment and development is required, particularly in the areas of paediatric sleep medicine and long-term ventilation (invasive and non-invasive). A modernisation of the provision of subspecialty services in the units in Dublin, and the development of regional centres of expertise in paediatric respiratory medicine, will provide a fantastic opportunity to provide a world-class paediatric respiratory medicine and sleep service to the children of Ireland. A strong national network with a coordinated approach to quality, education, audit, research, and service development in paediatric respiratory medicine is within our grasp.

International and National Models

Cystic fibrosis (CF)

Several countries around the world have drafted standards of care and models of care for children with cystic fibrosis (CF). Recent key documents originate from Australia and the United Kingdom (UK), and these healthcare systems more closely resemble Ireland's healthcare system compared with North American models. 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 standard of care that should be expected by children and their families, the minimum acceptable standards of care from centres, and the overall structure of the network of CF centres in the country. While much of the content of these documents can be extrapolated to Ireland, additional local considerations must be taken into account. A switch from ad hoc, centre-based treatment to a coordinated system of linked specialist centres with national patient registries, national and international standards of care and clinical guidelines in CF has occurred over the last 10 to 20 years.

Note should be made of the formation in Ireland of a National Clinical Programme dealing specifically with CF. This Programme is charged with the development of both a model of care and standards of care for children and adults with CF in Ireland. Official recommendations on the appropriate national distribution of services for children with CF must take into account both local paediatric medicine services and adult services for people with CF. In light of the remit of the National Clinical Programme for CF, this document will not make specific recommendations in relation to the model of care to ensure that a more balanced and complete picture is considered prior to any official recommendations being made.

Sleep

Similar to CF, the key documents relating to standards of care in paediatric sleep originate from the UK and Australia rather than North America, as their healthcare services are more comparable. In 2009, the Royal College of Paediatrics and Child Health published standards for services for children with disorders of sleep physiology. This document was developed by the working party on sleep physiology and respiratory control disorders in childhood. In June 2012, the Australasian Sleep Association published Standards for Sleep Disorders Services, which addresses both adult and paediatric sleep requirements. These documents highlight the patient groups that require sleep services, the types of assessments required, the management and technical requirements and the minimum acceptable standards of care that children should receive. The Irish model needs to take into account the difference in geography when extrapolating from these documents, but otherwise much of the content can be applied to Ireland. A proposal for a National Clinical Programme for Sleep Medicine, which includes paediatrics, was submitted to the Health Service Executive (HSE) in 2013. A final decision regarding this proposal is still awaited.

Neuromuscular Disease (NMD)

Children with neuromuscular disease (NMD) are initially diagnosed and cared for by neurologists. NMD is a life-limiting disorder, and approximately 90% of patients die of respiratory failure. Therefore, the respiratory consultant and team have significant involvement in the care of the patient; such involvement tends to increase as the child gets older. While the majority of these children die in their teens or early twenties, a significant proportion die much earlier, i.e. within the first few years of life. Units taking care of children with NMD require strong sleep-medicine support and non-invasive ventilation because almost all of these patients require regular sleep assessment and non-invasive ventilation as their health deteriorates. The neurology literature contains national and international guidance on the provision of care for children with NMD. The core component of service delivery to this group of children is a multidisciplinary team in a specialist paediatric hospital. As part of the multidisciplinary team, consultants in paediatric respiratory medicine, respiratory nurse specialists and respiratory physiotherapists see all children with NMD, and undertake assessment and treatment of neuromuscular respiratory compromise. Given the degree of expertise required across many different disciplines, NMD clinics are usually based at specialist paediatric hospitals. Specific, up-to-date recommendations in relation to the model of care delivery and standards of care can be found in the British Thoracic Society document Respiratory Management of Children with Neuromuscular Weakness (July 2012).

Long-term Ventilation

Long-term mechanical ventilation in children is a specialised and complex practice that can only be carried out in a small number of highly specialised centres. Like other highly specialised low-frequency conditions, the geographical dispersion of the patients and lack of expertise locally means that the delivery of care has to be concentrated in, and coordinated by, the specialist centre with local liaison. As is the case in many international centres, this model of delivery of care requires the involvement of a paediatric respiratory medicine consultant (with expertise in long-term mechanical ventilation) and a nurse specialist service. A multidisciplinary team is required in the specialist centre. This team, which is based in a specialised inpatient facility catering for children on long-term ventilation (LTV), will liaise closely with the child's family while the child is in hospital; when the child is at home, the team will liaise with the family's local community services.

The area of LTV is a rapidly expanding subspecialty of paediatric respiratory medicine. Advances in ventilator technology now provide us with the opportunity to care for many of these children in their home environment. A number of national guidelines on the care of children on LTV have been developed in the USA, UK and Australia. Different groups in the UK have published a number of documents pertaining to the care of children on LTV. The five most prominent of these documents are:

- Core guidelines for the discharge home of the child on long-term assisted ventilation in the United Kingdom (Thorax, 1998)
- Breathing Space: Community Support for Children on Long-term Ventilation (Ludvigsen and Morrison, 2003)
- From Hospital to Home: Guidance on discharge management and community support for children using long-term ventilation (Noyes and Lewis, 2005)
- The Thoracic Society of Australia and New Zealand (TSANZ) (2008) Ventilatory Support at Home for Children. A Consensus Statement from the Australasian Paediatric Respiratory Group
- The American College of Chest Physicians published a consensus statement in 1998 titled Mechanical ventilation beyond the intensive care unit

Each of these documents provides a framework for the management of children with complex conditions as they transition from the paediatric intensive care unit (PICU) to home. A well-structured and funded paediatric LTV service in Ireland, in which the new children's hospital would take the lead in overseeing the initial management with local expertise involved from an early stage, would fit well into the hub-and-spoke model proposed for the national paediatric respiratory service.

42.1 EVOLUTION OF SERVICES

General Respiratory Services

Twenty years ago, paediatric respiratory clinical practice mainly comprised CF, asthma, bronchopulmonary dysplasia, respiratory infection and a constellation of rare disorders. Over the last 10 years or so there has been a very rapid growth in paediatric sleep medicine, long-term invasive ventilation and long-term non-invasive ventilation (incorporating children with NMD). Today, these three areas comprise up to 50% of day-to-day activity in a busy tertiary centre. Bronchopulmonary dysplasia is now much less severe and less prevalent than in the past, mainly as a result of improvements in neonatal intensive care. As general paediatric expertise in asthma improves, tertiary subspecialists in paediatric respiratory medicine are increasingly treating fewer mild and moderate asthma patients, and are focusing solely on severe asthma patients. The respiratory subspecialist is tasked with the increasing volume of more complex subspecialty work. Severe asthma clinics are now commonplace in large tertiary paediatric respiratory centres.

As the scope and complexity of general paediatric medicine (and other subspecialties) broadens, subspecialist practice often becomes more specialist and complex, and less general. A number of examples of this can be seen in subspecialty practice in both Our Lady's Children's Hospital, Crumlin (Crumlin) and the Children's University Hospital, Temple Street (Temple Street). In the last number of years, the number of consultations referred from cardiology specialists has increased significantly. With the growing realisation internationally that obstructive sleep apnoea and pulmonary aspiration in infants with congenital heart disease (particularly those with trisomy 21) can have a very significant haemodynamic effect on pulmonary circulation and the outcome from cardiac surgery, these conditions are now considered in all children with congenital heart disease with any risk factors. Similarly, as information becomes available on the pulmonary abnormalities of children with sickle cell disease, the demand on paediatric respiratory and sleep services from the sickle cell clinic has risen very significantly in the last five years.

In Temple Street, a significant increase in respiratory workload has been identified from the National Centre for Inherited Metabolic Disorders. Metabolic diseases are complex medical conditions requiring specialised diet and care. In many patients, the prevalence of sleep-disordered breathing is up to 90-100%. Additionally, the national

paediatric neurosurgical centre has been moved to Temple Street, and these complex cases are often challenging and difficult from a respiratory viewpoint. The significant increase in NMD referrals over recent years from the Central Remedial Clinic, coupled with referrals from the national renal transplant centre, have also significantly added to the workload at Temple Street. With the increased understanding of the detrimental effects of obstructive sleep apnoea, especially in infants, and the increasing availability of sleep services internationally, there has been a marked increase in requests for consultation and sleep investigations in children with trisomy 21, craniofacial abnormalities and complex neurological conditions.

Overall, the complexity of the inpatient, consultation and outpatient service has increased over the last 10 years. The volume of inpatient and consultation work has also increased in parallel. In the tertiary centres in Dublin, outpatient activity has generally changed from a predominantly high-volume low-complexity picture to a predominantly low-volume high-complexity picture, where patients require more specialist input and therefore fewer patients can be scheduled. With the appointment of paediatric respiratory specialists in regional centres to provide services for children with CF, children in those centres with respiratory problems other than CF are increasingly being referred there. These subspecialists are also providing a regional non-CF subspecialty respiratory service, but are restricted by both the large volume of general paediatric workload and the lack of availability of specialised nursing, health and social care staff and appropriate facilities in this area. As well as managing children with CF, paediatric respiratory specialists in regional centres see larger numbers of lower complexity respiratory patients as well as complex respiratory patients in clinics. A limited sleep service is being provided in many regional centres which cannot meet current demand without significant investment in personnel and equipment. Given the significant volume of children with highly prevalent conditions, for example, troublesome asthma or obstructive sleep apnoea, regional subspecialty services need further investment to allow them to effectively manage this workload, the volume of which far exceeds the capacity of tertiary centres.

Cystic fibrosis (CF)

Following publication of the Pollock report, and the subsequent HSE report on CF services in Ireland, significant investments were made in the staffing of tertiary and regional paediatric CF services in Ireland in the first decade of the 21st century. These investments have been very effective in providing, in the six designated specialist paediatric CF centres in Ireland, high-quality uniform standards of CF care for children. Unfortunately, much of the progress in staffing levels up to 2008 has been eroded due to local funding shortfalls in hospitals providing CF care, coupled with the strong demand from all specialist areas for the dwindling pool of nursing and allied health professional staff in these institutions. Most paediatric CF centres in Ireland are now significantly understaffed.

Paediatric CF care has evolved very significantly over the last 20 years. Most care provided for children with CF can be delivered through regional centres that have appropriate staffing and expertise. For children who do not have complex disease, the same level of care can be as readily provided in a regional centre as in a tertiary specialist centre. The vast majority of care is ambulatory, and the numbers of children with CF are sufficiently large to enable expertise to be developed and sustained locally. Ireland has a very effective network of paediatric CF centres, which have easy access to tertiary subspecialty expertise (such as gastroenterology, paediatric surgery, cardiothoracic surgery) in the national referral centre, and good communication pathways between centres.

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 from where their care is directed and coordinated. 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 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 centre. Individual practices have evolved over time and there is no clear 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.

Adult services for patients with CF are predominantly based at St Vincent's University Hospital and Beaumont Hospital in Dublin, and the University Hospitals in Cork, Limerick and Galway. 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. This will be addressed specifically by the National Clinical Programme for Cystic Fibrosis.

Sleep

Paediatric sleep services are in their infancy in Ireland. Internationally, North America and Australia have recognised the need for paediatric sleep services, and now have well-established training programmes and services in this area. Other countries, including the UK and Ireland, have been slow to recognise the high prevalence of sleep disorders in children, and the need for structured services. Currently, there is no established service in place in Ireland for children with behavioural sleep disorders. A number of paediatric respiratory centres have purchased a limited amount of sleep equipment, but most are unable to cover the cost of providing the requisite staffing levels to deliver a paediatric sleep service. With increased recognition of the prevalence of sleep disorders in children, the demand for services has increased dramatically over the last four years. A national audit of the six paediatric respiratory centres carried out in 2011 showed a greater than 400% increase in diagnostic sleep tests and a greater than 600% increase in the number of children requiring non-invasive ventilation. This area is the fastest growing branch of paediatric respiratory medicine and is continuing to expand. A national business case for paediatric sleep services has been developed and was presented to the Paediatric Integration Group in 2014. This demand for service highlights the need for an integrated national paediatric sleep service based on a hub-and-spoke model involving local, regional and supra-regional units.

Respiratory management of neuromuscular disease (NMD)

Standard international practice for children with NMD is that they attend a specialist multidisciplinary team (MDT) in a recognised tertiary centre. This MDT is led by a neurologist, and incorporates physiotherapy, occupational therapy, dietetics, speech and language therapy, orthopaedics, respiratory medicine, cardiology, psychology and social work among others. The key involvement from a respiratory medicine perspective includes management of sleep-related breathing disorders, impaired airway clearance and evolving respiratory failure. Additional factors, such as progressive bulbar weakness leading to recurrent aspiration pneumonia, and evolving kyphoscoliosis causing restrictive lung disease, further contribute to respiratory insufficiency. As respiratory function worsens in individuals with NMD, respiratory infection becomes more common and more difficult to treat. In addition, the requirement for non-invasive ventilation at night and during the day increases demand on respiratory services significantly. If managed by a well-resourced respiratory team, frequent and extended hospital inpatient stays can be avoided by attention to detail with regard to airway clearance and adequacy of ventilation. Technological advances, particularly in the area of long-term non-invasive ventilation, mean that we can now achieve a much better quality of life for children at home than before. This preventive approach reduces the number of PICU admissions and the need for acute invasive ventilation in children with NMD, particularly Duchenne Muscular Dystrophy. However, this is staffing resource intensive.

Currently, patients with NMD and respiratory involvement are seen in Cork University Hospital, Our Lady's Children's Hospital, Crumlin and Tallaght Hospital. Cork University Hospital has a paediatric neurology service that runs a neuromuscular clinic. The consultants in paediatric respiratory medicine at Cork University Hospital take care of the respiratory needs of these patients. The greatest numbers of patients are seen between Temple Street and the Central Remedial Clinic, where an established neuromuscular programme is in place. MDT staffing here is poor. There is a pressing need to adequately resource MDTs looking after children with NMD, and to establish very clearly the clinical governance of this population. In some centres, neurologists lead the MDT; in other centres the team is led by general paediatricians or respiratory physicians. It is essential to ensure that whichever service is charged with holistic management of this group is adequately resourced. For the purposes of this model-of-care document, resources reflect management of the respiratory component only by the respiratory team.

Inadequate physiotherapy staffing levels (dedicated to children with NMD) in tertiary and regional centres results in a significant gap in service provision. Currently, there is no respiratory physiotherapy presence dedicated to any NMD respiratory clinic. The lack of dedicated physiotherapy staffing in these clinics leads to a fragmented respiratory service for children and families. These children and families require respiratory physiotherapy input at clinics, so as to provide education and support in relation to physiotherapy techniques and respiratory equipment, e.g. cough assist, and provide a clear link to local community disability team physiotherapists. Integrated care pathways need to be developed for children who require specialist respiratory physiotherapy support when attending an NMD clinic, in addition to physiotherapy support provided by their local physiotherapist. Physiotherapy staff on local disability teams are unlikely to have the specialist respiratory knowledge and linkages with the NMD clinics that are required in order to provide a high-quality integrated respiratory physiotherapy service.

Across all centres, there are poor resources for the diagnostic evaluation of sleep-disordered breathing (poorly resourced sleep laboratory at Crumlin, poor resources for ventilation assessment at Temple Street, lack of clinical nurse specialist (CNS) posts in sleep medicine, difficulty admitting patients for overnight assessment), management of impaired airway clearance (poor physiotherapy staffing particularly at Crumlin), management of respiratory failure (high demand on scarce resources in the last number of years, lack of nurse specialist services). In the Dublin centres, the service would benefit from centralisation and pooling of resources. However, with the current arrangement of paediatric services across the city, and given the complexity and multidisciplinary nature of NMD, such centralisation and pooling does not appear possible in the current climate.

Long-term Ventilation

'Ventilation' is the term used to describe the movement of air in and out of the lungs, so that gas exchange can occur at the alveoli. Air is 'pumped' in and out of the lung in healthy individuals with normal airways and lungs by the muscles that form the respiratory pump. Ventilation, therefore, occurs in all of us. Supplementary, or mechanical, ventilation is required in some individuals if their pump, their airways, or their lungs, are incapable of ventilating effectively unaided.

Non-invasive ventilation (NIV) is a method of assisting a person's own ventilation efforts by applying positive pressure to the airway, typically via a mask covering the nose, or nose and mouth. The technique can be applied day and night in the acute setting, but over the longer term it is most commonly done when the patient is sleeping. Invasive ventilation refers to mechanical ventilation of a patient through a tube placed directly in the trachea, the main windpipe. This occurs most commonly on a temporary basis, such as in an operating theatre or intensive care

unit. For temporary indications, patients are heavily sedated and the tube is inserted directly into the trachea via the nose or mouth. In certain situations, some children are unable to wean successfully off mechanical ventilation, due to dynamic airway instability and/or other chronic conditions affecting the child's ability to breathe on their own, such as chronic lung disease of prematurity and NMD. For medium- or long-term ventilation, tubes are more effectively inserted directly through the neck into the trachea (tracheostomy tubes).

When invasive ventilation is required indefinitely it is typically referred to as 'long-term ventilation (LTV)'. It is typically applied 24 hours a day, for seven days a week initially, with planned weaning over an extended period, depending on the child's condition. Due to technological advances in the areas of neonatal and paediatric intensive care and respiratory medicine, the number of children requiring respiratory support by LTV has increased significantly worldwide over the past 10 years, and this trend has been reflected in Ireland. LTV is probably unique, in its aspiration to provide intensive-care-level support in the home environment, long term. This is hugely challenging for families, GPs, community care services, and regional paediatric services.

When clinically stable, children on LTV can be cared for very effectively in the home environment, conferring benefits on the child, the family, the community and the hospital. The improved morbidity and mortality that has resulted from the ability to provide LTV to children with complex conditions has not been coupled with a sufficient increase in resources and staffing in LTV care. Children stay in hospital until a sufficient homecare package (HCP) to enable the family to care for their children in the home environment has been agreed/provided. Once home, most children can be weaned off LTV in a staged process.

A respiratory paediatrician coordinates respiratory care throughout this journey, optimising ventilation needs, planning the weaning process, and any other general respiratory issues. As initially these patients all are cared for in a tertiary PICU, the burden of respiratory care of these children has historically fallen on the Temple Street and Crumlin hospitals. The same level of training, but less ongoing exposure, is present at consultant level in each of the regional centres to provide this supervision, but expertise in nursing and allied health professionals is lacking in these centres.

Severe Asthma

Asthma is one of the most common diseases of childhood. The vast majority of children with asthma have very mild disease, and can be treated in primary care. In Ireland, all hospitals providing care for children, including regional and specialist centres, will treat children with asthma at a secondary care level. Children with poor asthma control, despite intervention at a secondary care level, are currently referred to either regional or tertiary centres. No guidance exists to help direct referrals. Consultants in paediatric respiratory medicine at all the regional and tertiary centres are well trained to care for the vast majority of children who suffer from poor asthma control despite high doses of asthma medication. There is a small and particularly complicated group of patients who suffer from extremely difficult-to-control asthma, and these patients may require systemic anti-inflammatory therapy or complex investigations, in order to rule out other diagnoses. Other countries have 'super-specialised' centres that concentrate on this very difficult group of patients, and build a level of expertise in evaluation and treatment of these children. No such referral pathway exists in Ireland. It is likely that there are sufficient patients to merit a single national centre for complex therapy-resistant asthma, where expertise can be further developed in this area. There is a very small number of patients in each individual centre who might require this type of service, but the number is sufficiently large to merit the establishment of a 'Severe Asthma Clinic' in a single centre.

Primary Ciliary Dyskinesia (PCD)

The prevalence of PCD is noted to be approximately 1 in 16,000 live births in developed countries. Given the increased frequency in the consanguineous Traveller population in Ireland, it is likely that this figure is higher here. However, accurate data on the number of patients with PCD in Ireland are lacking. On the basis of the birth rate in Ireland, and the noted international prevalence of PCD, it is likely that there are between 80 and 100 children with PCD in Ireland at any one time. The international norm for children with PCD is to establish a specialist PCD service in a tertiary children's hospital that develops expertise in the condition and sees children in a multidisciplinary setting, similar to what is practised for children with CF. The UK, which has a population of 63 million, has six specialised centres for children with PCD.

Children with PCD are cared for in all of the regional and specialist centres in Ireland. In line with the international experience, it is likely that there are many children with undiagnosed PCD in Ireland being treated in a primary or secondary care setting. There are no dedicated services for children with PCD in Ireland. The Irish Paediatric Surveillance Unit is currently conducting a study into the prevalence of PCD in Ireland. A specialist service, based in the New Children's Hospital, and focused on the evaluation and treatment of children with PCD, should be established. This specialist service would include a shared care programme with regional centres.

42.2 CURRENT SERVICE PROVISION

Specialist Centres

In Dublin, three centres provide subspecialist respiratory services. These centres are: Crumlin, Temple Street and Tallaght. Crumlin is the largest of these, and provides a wide range of subspecialty services including tertiary sleep, long-term invasive and non-invasive ventilation, and a full inpatient and consultation service. Crumlin has an eight-bed transitional care unit for children on long-term mechanical ventilation. All major subspecialties, with the exception of metabolic medicine and neurosurgery, are represented in Crumlin. A small cohort of children with NMD is seen at the respiratory clinic in Crumlin, with much larger numbers seen in the other hospitals.

Temple Street is the next largest centre providing a wide range of subspecialty respiratory services, including care of patients with CF, long-term invasive and non-invasive ventilation, the largest cohort nationally of NMD patients and a full inpatient, respiratory consultation service and tertiary referral outpatients service. Temple Street has the national centres for inherited metabolic disorders, paediatric neurosurgery and spina bifida, to which the respiratory service provides consultation. Temple Street has a wide array of additional subspecialty areas including craniofacial surgery, PICU, neonatology and ear, nose and throat (ENT). The hospital also provides a limited sleep service and does not provide polysomnography. Temple Street provides long-term invasive ventilation, but lacks a transitional care unit.

The National Children's Hospital is attached to Tallaght Hospital, an adult academic teaching hospital. Tallaght has specialist services for respiratory paediatrics, endocrinology, paediatric surgery and paediatric ENT, as well as consultation services for gastroenterology, cardiology, neurology, allergy, infectious diseases and other services, most of which are shared with Crumlin. The respiratory team at Tallaght provides services for children with CF, severe asthma, paediatric sleep medicine and general respiratory problems. Tallaght provides a polysomnography service, but the hospital does not have the facilities to provide an invasive ventilation service.

Regional Centres

The regional paediatric centres in Cork, Limerick and Galway are staffed to provide a full range of services (with the exception of complex tertiary care) to children with CF. Each centre should be staffed with a full CF MDT. Each unit provides a neonatology service. A limited polysomnography service is provided in Cork, but without any technical or administrative back-up. Basic diagnostic sleep services are provided in all regional centres, but the services are very limited both in terms of scope and volume due to the lack of available dedicated staff in this area. Subspecialists in the regional centres manage a wide variety of paediatric respiratory problems apart from CF. The ability to provide the respiratory service is limited by the demands on subspecialists from general paediatrics and the lack of appropriate staffing or expertise in the regional centres.

Local Services

Local paediatric centres around Ireland provide secondary care services for all children including those with asthma and recurrent respiratory infections. In the last number of years, some local centres have started to provide diagnostic overnight oximetry, but this occurs on an ad hoc basis and no formal pathway exists for onward referral or service development. Increasingly, local centres, in some instances, refer children with respiratory concerns to regional as opposed to tertiary subspecialist centres. There is currently no dedicated paediatric physiotherapy service in any of the local paediatric units. When children are admitted, they are seen by adult respiratory physiotherapists on an ad hoc basis, and without the appropriate linkages to the respiratory physiotherapist in the relevant tertiary or regional centre. There is no paediatric physiotherapy respiratory outreach service for children available from the local paediatric unit. A dedicated paediatric physiotherapist in each paediatric unit, with 0.5 physiotherapists dedicated to outreach, would bridge the significant gap in local service provision for this vulnerable group of children and families; these physiotherapists would provide support, education and local respiratory management as dictated by a defined, integrated care pathway with the relevant regional or tertiary centre paediatric respiratory physiotherapist.

Main Clinical Caseload Areas

<p>Cystic fibrosis (CF)</p>	<p>Cystic fibrosis (CF) services are delivered at all regional and specialist centres. These centres have been designated as specialist CF centres as part of the national newborn screening programme. Some local centres provide CF care with limited staffing. Shared care arrangements between local, regional and specialist hospitals are ad hoc. Crumlin is the national referral centre for children with CF, due to the availability of hepatology, general paediatric surgery, cardiothoracic surgery and endocrinology in the one institution. Children are referred from both local and designated CF centres for services which are not available at the referral source.</p>
<p>Obstructive sleep apnoea (OSA)</p>	<p>Obstructive sleep apnoea has a prevalence of 3% in the general population, making it one of the most common conditions of childhood. The prevalence is significantly higher in conditions such as trisomy 21, craniofacial disorders and NMD. Prior to the advent of diagnostic sleep services for the diagnosis of OSA, children with symptoms suggestive of OSA, such as snoring, difficulty breathing at night, fatigue and behavioural problems during the day, were referred to ENT surgeons. This occurred at all local regional and tertiary centres. Historically, the decision to perform adenotonsillectomy was subjective, and based on the history and the degree of adenotonsillar hypertrophy. As international practice has evolved, diagnostic sleep tests are increasingly sought prior to consideration of treatment for obstructive sleep apnoea. This has been most evident in tertiary centres that provide polysomnography, where the demand has been very significant due to the high prevalence of children with complex conditions at high risk</p>

	<p>of sleep-disordered breathing. Regional centres that now provide an under-resourced, limited sleep service have also seen a dramatic increase in referrals for sleep assessment, mainly from local ENT services. It is likely that historic practice continues at local and many regional centres. The majority of tertiary sleep services are provided in Crumlin, with a small smaller volume provided in Tallaght. Temple Street provides a limited service. Regionally, Cork, which has poor staffing levels, provides secondary-level sleep studies, whereas both Limerick and Galway provide primary-level studies. All areas are experiencing increasing demands on their very limited services.</p>
Sleep-related hypoventilation	<p>Children with sleep-related hypoventilation are seen at all centres. Initial diagnosis and treatment is often carried out in tertiary subspecialty centres, with some follow-up and titration increasingly occurring at regional centres. Initial diagnosis and treatment is more frequently being provided at some regional centres, such as Cork and Limerick. With appropriate staffing this level of service could be expanded in the future, thus significantly alleviating pressure on the supra-regional unit.</p>
Narcolepsy	<p>Narcolepsy is an extremely rare disorder with what appears to be a once-off transient increase in prevalence related to H1N1 vaccination. Services for children with narcolepsy have been concentrated in the Temple Street neurology department, with diagnostic testing (multiple sleep latency testing) taking place in the Mater Private Hospital.</p>
Behavioural sleep problems	<p>In Ireland, no services exist in the public health system for children with behavioural sleep problems. There are no individuals in the public system with sufficient training and expertise in this area. The Mater Private Hospital does, however, provide a service for children with behavioural sleep problems.</p>
Respiratory management of neuromuscular disease (NMD)	<p>Children with NMD have very complex respiratory needs, particularly as they get older. Neuromuscular weakness affects the respiratory system in two main ways:</p> <ul style="list-style-type: none"> • Respiratory muscle weakness severely affects the efficacy of coughing, and puts children at high risk of recurrent respiratory infections. Children often struggle to overcome respiratory infections with standard treatment, due to respiratory muscle weakness. • Children with respiratory muscle weakness often have sleep-related hypoventilation. This can be very difficult to manage in such children. Many children in the late stages of NMD go on to develop diurnal hypoventilation, and require mechanical ventilation at night and during the day. <p>From a respiratory point of view, the treatment of children with neuromuscular weakness is a highly complex area; such children have very poor reserves, can get very sick very quickly, and are often poorly understood by non-respiratory specialists. There is a very high rate of morbidity and mortality in this group. Most of the burden of this morbidity is borne by the respiratory services, as 90% of mortality is due to respiratory failure. This is a very resource-intensive area of respiratory practice, and requires significant development.</p>

	<p>Multidisciplinary clinics for children with NMD are held in Temple Street and Tallaght. The neuromuscular clinic in Temple Street sees larger volumes of patients. Smaller numbers of children with non-specific neuromuscular weakness, unrelated to specific neuromuscular conditions, are treated in all centres. Crumlin looks after the respiratory component of NMD for a small number of patients. Some regional centres, such as Cork, which has two paediatric neurologists, manage the respiratory needs of children with neuromuscular weakness locally (including initiation of non-invasive ventilation).</p>
Long-term ventilation (LTV)	<p>Long-term ventilation (LTV) is provided in Crumlin and Temple Street. Crumlin has a dedicated transitional care unit and a discharge coordinator for children on LTV. Greater numbers of children requiring LTV are seen in Crumlin; this is mainly due to the hospital's wider range of specialties and larger intensive care unit. Children on LTV are initially cared for in hospital and subsequently discharged to the community with a homecare package, and are under the supervision of a respiratory consultant. The most common reason for LTV is dynamic airway instability. A number of children with chronic lung disease of prematurity, complex cardiorespiratory disease and NMD also require LTV. In some instances, children on long-term invasive ventilation have been transferred to an inpatient setting in regional units. Access to this service is limited by lack of local expertise, staffing and back-up.</p>
Severe asthma	<p>Among specialist centres, the majority of children with severe asthma are cared for in Temple Street and Tallaght. Crumlin does not have an asthma nurse. A well-developed asthma/allergy service is in place in Tallaght. All of the regional centres care for children with severe asthma and refer them on an ad hoc basis to specialist centres, if required. There is no recognised national referral pathway for children with severe asthma. There are no data available on the number of children with severe asthma in local, regional or specialist paediatric centres.</p>
Primary ciliary dyskinesia	<p>Primary ciliary dyskinesia (PCD) is a lifelong, genetically inherited disorder characterised by recurrent respiratory, sino-nasal and ear infections. This lung disease is analogous to CF. Children require long-term chest physiotherapy and intermittent treatment for exacerbations either at home or in hospital. The diagnosis of PCD is extremely complex. There is no cure for PCD, and individuals with PCD will develop progressive lung disease characterised by bronchiectasis and, ultimately, respiratory failure. There is no single centre specialising in the provision of care for children with PCD in Ireland. Patients with PCD attending Temple Street are seen for diagnostic testing, and annually thereafter, at the Royal Brompton Hospital in London in a shared care arrangement.</p> <p>It is estimated that there are between 80 and 100 children with PCD in Ireland at any one time. The international norm for children with PCD is to establish a specialist PCD service in the tertiary children's hospital (which would include respiratory physiotherapy) that develops expertise in the condition and sees children in a multidisciplinary setting, similar to what is practised for children with CF. There is currently a significant unmet need for treating patients with PCD in Ireland.</p>

	<p>In the tertiary units, inpatients are seen by a respiratory physiotherapist, as referred, but there is currently no physiotherapy cover at outpatient clinics, and there is a lack of services available for local follow-up both in tertiary units and, locally, in the community. No local outreach service is available for children and families with a diagnosis of PCD and, as diagnostic rates increase, this places a significant demand on paediatric respiratory physiotherapy services, which cannot be addressed with current resources.</p>
General respiratory	<p>The workload of the respiratory subspecialist, other than that related to the conditions referred to above, can broadly be broken down into generic respiratory problems and rare disorders:</p> <ul style="list-style-type: none"> • Thoracic insufficiency syndrome – conditions such as scoliosis, arthrogryposis, skeletal dysplasia. Children with thoracic insufficiency syndrome often require non-invasive ventilation, and some diurnal ventilation. • Pulmonary aspiration – pulmonary aspiration is very common in children with co-morbidities, and is much more common than has been recognised in the past. Diagnosis and treatment are very complex and time-consuming. Pulmonary aspiration is poorly understood by the non-specialist. • Recurrent respiratory tract infections – conditions such as non-CF bronchiectasis, immune deficiency, structural airway problems. The number of patients with non-CF bronchiectasis is unknown. • Airway disorders – conditions such as congenital airway obstruction, acquired airway obstruction, tracheobronchomalacia. These are very complex and time-consuming disorders. • Complicated respiratory infection – conditions such as empyema. Empyema requires a very significant time input for the inpatient service. • Congenital thoracic malformations – conditions such as congenital cystic adenomatoid malformation/congenital pulmonary airway malformations, sequestration and congenital lobar emphysema. These lesions are complex and require multidisciplinary input. • Suspected asthma – diagnosis unclear. Increasingly, subspecialists are seeing children with significant morbidity from a respiratory perspective who do not have a clear diagnosis of asthma. These children must be investigated and managed carefully. Communication is vital here. This is a time-consuming patient population. • Children with lower respiratory tract infection with and without other co-morbidities, e.g. cerebral palsy.
Rare disorders	<p>Interstitial lung diseases – conditions such as surfactant protein disorders, alveolar proteinosis, pulmonary haemosiderosis, interstitial glycogenosis.</p> <p>There is a high volume of extremely rare conditions that are rarely seen in individual centres and are too numerous to list.</p>

Paediatric Respiratory Services – Key Clinical Relationships

Health and social care professionals

- Respiratory scientists are involved in almost all areas of clinical respiratory practice encountered by the respiratory team. Respiratory scientists run the pulmonary function and sleep laboratory, and work very closely with the respiratory team and clinical engineers in terms of clinical diagnostics and therapeutics, both from a general respiratory and sleep perspective.
- There is close collaboration with physiotherapy colleagues. In children with CF, airway clearance is one of the core components of their treatment. Assessment and management of effective airway clearance, musculoskeletal function and cardiopulmonary exercise capacity are core elements of CF respiratory management. Physiotherapists are vital members of the multidisciplinary team. They provide:
 - Individual treatment focused on airway clearance to prevent airway damage, and also focused on maintaining exercise tolerance by actively encouraging people with CF to participate in regular physical activity and exercise.
 - Continuous/ongoing education and training for patients and their families to enable them to manage their condition through essential, daily home physiotherapy.
 - Assessment and advice for secondary complications such as musculoskeletal and postural problems, bone health, and continence issues.
 - An inpatient service including assessment, airway clearance and exercise.
 - An outpatient service which involves attending clinics and annual assessments, and providing outpatient appointments when/if needed.

In addition to providing treatments for children with CF, we care for a large number of children who have difficulties with airway clearance, including those with neuromuscular disease, non-CF bronchiectasis, pulmonary aspiration, complex respiratory infection, neurological disability and spinal trauma, as well as children on LTV. Physiotherapy plays a significant role in the management of children in transitional care in hospital, and in both pre- and post-operative multidisciplinary assessment.

Physiotherapy provides a weekend, emergency weekend and on-call service for all respiratory patients in tertiary, regional and local units who meet the treatment criteria. The respiratory physiotherapy service facilitates the MDT management of a broad range of clinical presentations which are likely to deteriorate significantly where the patient does not have access to respiratory physiotherapy techniques.

The vital role of this out-of-hours and weekend respiratory physiotherapy service on the ward may prevent a patient requiring intubation if secretions can be cleared and respiratory function improves. In order to ensure the smooth running of these services, all physiotherapists are expected to partake in on-call training and maintain their competencies. A key responsibility of the respiratory physiotherapy service is to educate and provide training for all physiotherapy staff who are contracted to work these rotas. Adequate staff is imperative to ensure that these emergency out-of-hours and weekend services are continued.

There is seasonal fluctuation in the workload of the respiratory physiotherapy service, with referrals for inpatient and weekend/on-call services peaking annually in the winter months. A significant number of children with NMD/development delay/metabolic conditions require intensive chest physiotherapy input during this time, and this impacts enormously on both inpatient and weekend/out-of-hours services.

- Pulmonary aspiration is extremely common, especially in the complex group of patients who are cared for in tertiary centres. There is close liaison with speech and language therapy colleagues in terms of assessing swallow function and the risk for pulmonary aspiration. In the transitional care unit there is a reliance on the speech and language therapist in terms of assessment and prevention of pulmonary aspiration and optimisation of vocal function in children on LTV.
- Nutritional management of children with CF is one of the core aspects of their long-term care. Physiotherapists work very closely with colleagues in nutrition and dietetics in managing the long-term needs of patients with CF. Dietetic input is essential for children with NMD in terms of obesity management, bone mineral density and enteral feeding.
- Occupational therapists play an integral role in the assessment and treatment of children with respiratory conditions, in particular LTV patients and patients with NMDs. This cohort of patients may require 24-hour postural management systems (sleep systems, specialised seating, bathing equipment), specialist equipment for participation in activities of daily living (ADLs), assistive technology and splinting. The occupational therapist also assesses a child's development in terms of the skills needed to participate in ADLs. The acute hospital occupational therapist will refer to community therapists for ongoing developmental intervention, rehabilitation, postural management (specialised seating) or environmental adaptations. Children who have had respiratory infections that have rendered them severely de-conditioned are also seen by occupational therapists for rehabilitation, energy conservation education, splinting, transfer practice, equipment provision and onward referral to the community.
- All medical and surgical teams need to work closely with social work colleagues in order to optimise outcomes from a social perspective and ensure that the appropriate entitlements for patients and families are provided. The psychosocial aspects of the management of children with CF and NMD cannot be underestimated. Maintenance of a strong relationship between the family and the centre is of key importance. This practice is strongly supported by social work.
- The psychologist is a key part of the CF MDT. Chronically incapacitated children, with life-shortening illness, such as the NMD cohort and severe asthmatics, require psychology input. In addition to providing treatment for CF and NMD patients, CF services have the same requirement for the services of a psychologist as the vast majority of other teams, and such services are provided on a case-by-case basis. In CF, early intervention and effective psychological management can improve patient compliance and patient comprehension, and can also lead to better medical outcomes.

Diagnostics

- Pulmonary imaging is a core part of the assessment and management of fire patients. There is a heavy reliance on the radiology department in this regard. For children with airway compromise, parenchymal abnormalities, cardiopulmonary abnormalities and thoracic insufficiency, physiotherapists could not practice as they do without the clinical input of the Department of Radiology. In Crumlin, a monthly airway/cardi thoracic/respiratory radiology meeting takes place where paediatric patients with complex airway/thoracic/pulmonary problems drawn from all three hospitals can be discussed in a multidisciplinary setting. Weekly radiology meetings are held in Temple Street with the respiratory teams.
- The department of Clinical Engineering provides key support in terms of invasive and non-invasive ventilation in children, particularly in terms of ventilators and interfaces for children, but also in the context of the diagnostic sleep service.

- Short- and medium-term vascular access is a very significant issue for children with CF; they will require repeated courses of IV antibiotics. The approach to short- and medium-term vascular access differs across the city. In the optimum scenario short- and medium-term access will be provided through a peripherally inserted central catheter (PICC) line service provided by the Department of Interventional Radiology.
- Paediatric respiratory services rely heavily on the departments of Microbiology and Infectious Diseases in terms of the management of children with CF. Assessment and management of airway infection is one of the key aspects of CF management. There is close liaison with microbiology colleagues in terms of optimising diagnostic practices, interpretation of tests and management of findings. Infection control is also a significant issue with patients, who tend to spend significant periods of time in hospital.
- Like many other medical teams, paediatric respiratory services rely, on a regular basis, on haematology and biochemistry laboratories. Particularly in the case of children with CF. There is close liaison with the biochemistry laboratory to optimise the sweat test service available to children with CF at the time of newborn screening and diagnosis. Paediatric respiratory services also have a close working relationship with the pathology laboratory in terms of diagnostic lung biopsies, but particularly cytology findings related to bronchoalveolar lavage (BAL).

Medical

For a number of reasons paediatric respiratory services have a close working relationship with many medical teams. With several teams, this is based on the range of pathologies encountered; with others it is based on the work of our two core diagnostic laboratories of pulmonary function and sleep.

The following list summarises most of the key medical interdependencies:

- Gastroenterology: children with CF-associated liver disease, children with oesophageal dysmotility and pulmonary aspiration
- Cardiology: children with pulmonary hypertension, obstructive sleep apnoea and pulmonary disease causing pulmonary hypertension, children with trisomy 21 and congenital heart disease
- Metabolic: children with metabolic disease have significant sleep-disordered breathing problems – up to 90-100% in certain common metabolic disorders
- Neurology: children with NMD resulting in pneumonia or hypoventilation, and who die of respiratory failure in the first years of life or in their teens/twenties
- Endocrinology: children with CF-associated diabetes, children with Prader Willi syndrome or with suspected obstructive/central sleep apnoea, children on growth hormone with suspected obstructive sleep apnoea, obese children with suspected obstructive sleep apnoea
- Oncology: children with pulmonary insufficiency as a result of thoracic tumours, spinal or thoracic radiation or chemotherapy-induced pulmonary damage
- Renal: children with complex pulmonary renal syndromes and those who are immune suppressed after a kidney transplant
- Developmental paediatrics: children who have neurodevelopmental disorders with suspected pulmonary aspiration and/or obstructive sleep apnoea
- Neurology: children with NMD affecting airway clearance and ventilation
- Haematology: children with sickle cell disease with pulmonary involvement and/or obstructive sleep apnoea
- Neonatology: infants with chronic lung disease secondary to prematurity, newborn infants with complex problems including pulmonary aspiration and/or obstructive sleep apnoea
- Immunology: children with an immune deficiency that is affecting their mucosal immunity, including recurrent respiratory infection

- Allergy: children with severe allergy incorporating poorly controlled asthma; suspected allergic reactions to intravenous medications in children with CF
- Palliative care: respiratory management of children with end-stage respiratory failure, and end-of-life issues in children with CF

Surgical

Paediatric respiratory services also have a very close relationship with many surgical colleagues.

- ENT: joint management of children with complex congenital airway anomalies, joint management of children with tracheostomy tubes, assessment of surgical reversibility of obstructive sleep apnoea
- Cardiothoracic surgery: joint management of children with complex vascular thoracic anomalies, management of pleural disease including empyema, lung biopsies in children with established lung disease
- Orthopaedics: pre-and post-operative assessment and management of children with scoliosis and other thoracic insufficiencies
- Plastic surgery: assessment and management of children with complex craniofacial abnormalities with a risk of pulmonary aspiration and/or obstructive sleep apnoea
- General and abdominal surgery: management of enteral feeding in children at risk of aspiration, long-term implantable venous access devices and enteral feeding devices for children with CF. Most general surgeons also perform thoracic surgical procedures.
- Renal transplant surgery: This service requires access to respiratory care and rehabilitation.
- Neurosurgical: Including brain tumour, spinal cord injury and spinal surgery which all require acute PICU admission post-operatively, and multidisciplinary management.

Current Challenges in Service Provision

<p>Cystic fibrosis (CF)</p>	<p>Thanks to the establishment of a network of CF centres in the last decade, specialist services can now be provided effectively in all of the six main CF centres. Despite this well-thought-out network, a number of challenges still remain in terms of service delivery in different centres. These challenges relate mainly to lack of appropriate staffing in medical and allied health professional areas. In most hospitals, health and social care professionals (HSCPs), such as physiotherapists, dietitians, occupational therapists, social workers and psychologists, allocated to provide CF services come from a pool of HSCPs in that institution. In the majority of cases the HSCP allocated to CF services has other clinical commitments in addition to his/her CF work. All centres have noticed over the last four years that, in the context of shrinking staff pools of HSCPs within hospitals, allocation to CF services has reduced significantly to a point where it is affecting service delivery. Outside of the staffing issues, the centres in Cork and Galway are impeded in terms of seamless service delivery by the absence of adequate infrastructural facilities for segregation and high-quality care delivery in an inpatient and outpatient setting.</p>
<p>Sleep</p>	<p>Paediatric respiratory medicine in Ireland is very fortunate to have a good network of respiratory centres that already work cohesively, particularly in the area of CF. This allows for a natural sleep network to follow this path with a supra-regional New Children's Hospital and three main regional units – Cork, Limerick and Galway. The biggest challenge currently is manpower. The need for sleep scientists and non-invasive specialist nurses is the most pressing staffing issue. These posts are essential for the development of a functioning paediatric sleep service. Currently, demand for sleep studies far outstrips supply, resulting in long waiting lists, and this situation continues to deteriorate. Children diagnosed with sleep-disordered breathing requiring non-invasive ventilation have poor support in establishing and maintaining this treatment.</p>

Respiratory management of NMD	<p>Children with NMD require input from respiratory consultants, respiratory scientists, respiratory nurse specialists, respiratory paediatric physiotherapists, as well as the MDT working on the neurological aspects of the patient's condition. A close working relationship is required with local sleep services. The current difficulties with provision of sleep and pulmonary function services for children in Ireland are having a significant negative impact on the quality of the respiratory care delivered to this group of children with complex conditions. The absence of structured respiratory nurse specialist posts in all centres represents a real challenge in terms of providing the quality of care that is required for these children. The physiotherapist should link directly with local services on discharge to ensure that respiratory physiotherapy treatment programmes are overseen by the local physiotherapy team. Occupational therapists work with children with NMD under the direction of the neurology team primarily. However, currently, Crumlin does not have an occupational therapy presence at clinic appointments or at transition clinics, due to demands on the service.</p> <p>Even in Temple Street, which provides the majority of respiratory care to children with NMD, access to the core requirements of sleep testing, respiratory physiotherapy and nurse specialist follow-up is an obvious area of significant shortfall currently. This needs to be comprehensively addressed in the short term.</p>
LTV	<p>Children requiring LTV may come from any part of Ireland, and so a key aspect of a programme to manage children on LTV requires the concentration of expertise in one centre and the maintenance of a functioning network incorporating both regional centres and community services across the country. Inpatient services for children on LTV are almost exclusively provided in Crumlin and Temple Street. Some children from regional centres are sent there prior to discharge home. The frequency of such referrals has increased in the last two years. The key issues with inpatient service delivery relate to insufficient inpatient resources in most centres. In Crumlin, the purpose-built Transitional Care Unit is well equipped to deal with this patient population, but at times of high demand there are insufficient numbers of beds. With the exception of Crumlin, inpatient resources are poor. In Temple Street, this group of children with complex needs is managed either in intensive care beds or on key inpatient wards with relevant staffing experience. Regional centres use either regular inpatient beds or beds designed for high-dependency use. There is no designated LTV nurse in either Crumlin or Temple Street to act as a key worker overseeing the journey of these patients from tracheostomy insertion to discharge home. This person would act as an invaluable liaison between the LTV centre, the peripheral hospitals and the community. Crumlin does not have a discharge coordinator who is responsible for the facilitation of discharge of, among other things, children on LTV. An ideal system would involve the provision of both an LTV specialist and a dedicated discharge coordinator for children on LTV. These two individuals would need to have a close working relationship.</p> <p>The main issue for children on long-term mechanical ventilation is the very slow process surrounding discharge to the community. This is a disparate arrangement with variable practice all over Ireland in local community areas, and unacceptable delays in the sanctioning and provision of funding and in the allocation and recruitment of staff. This</p>

causes undue backlog within acute hospitals where children on long-term mechanical ventilation must remain as inpatients; it also has a further negative impact on ICU beds. In Crumlin, over the last number of years, the use of ICU beds by children on long-term mechanical ventilation has forced the cancellation of many ICU admissions and surgical procedures. The current application process for a home care package (HCP) is not fit for purpose. There is no centralised body identified in the HSE to oversee the approval of HCPs, thus leading to long delays in the discharge process. The process of discharge to the community should start immediately the decision to initiate LTV is taken. A pathway has been established in Crumlin in this regard. Work is currently underway in the HSE to establish a national framework of the discharge of medically fragile children from tertiary centres to the community. Given the low numbers and high complexity of these children's conditions, and also given the wide range of local community areas with their own funding, it can be extremely challenging for services in a local area to take on such patients when they may have no funding and no experience in this branch of medicine. It will, therefore, be vital to have a national funding stream for these children, and also to have a single national centre of expertise, so as to ensure the rapid and appropriate funding of HCPs when such is required. Ongoing surveillance of these HCPs will be a core part of any undertaking.

Occupational therapists have a key role in the facilitation of discharge planning. This process begins at the moment of referral. If a child presents with severe postural management requirements beyond those that can be addressed by acute hospital equipment (specialised buggies) liaison with outside specialist services is required (for example CRC, SeatTech Enable Ireland). Joint assessment and prescription for specialised moulded seating systems, custom hoist slings and sleep systems take place. Consideration is given to the child's and the family's requirements in terms of maximising involvement in daily activities (for example a chair that lowers to the floor for circle time in school, playing with siblings). Adaptations to the buggy or wheelchair to accommodate oxygen tanks, suction equipment and feeding poles are also extremely important in terms of mobility. Early referral to community colleagues is essential in order to support an easy transition to home.

The increase in the number of children requiring and obtaining tracheostomies in Temple Street has added to the increased training needs required by physiotherapists. These patients are often LTV dependent and can be difficult to rehabilitate from a manpower point of view (two people trained in tracheostomy changes and care are required to leave the ward and attend rehabilitation gym).

Another significant challenge currently, and a cause for much delay in the successful operation of home care for children on LTV, is the recruitment and training of healthcare professionals to care for children on LTV at home. A national system to coordinate and fund care in the community for these children should involve the use of a single nursing

	<p>agency, as opposed to the current practice which involves the use of several different agencies in different areas. Maintenance of expertise and concentration of liaison points between nursing agencies and specialist services for children on LTV is vital. Many staff assigned to HCP patients are agency-based nurses. The HSE currently has no community paediatric nursing staff designated to caring for children with tracheostomies and LTV. The absence of adequately trained nurses, coupled with the problems of staff retention and turnover, creates significant clinical governance and clinical risk challenges. Once a child on LTV is discharged to the community, there is a governance issue as to who is responsible for the overall care of that child.</p> <p>The strengthening of regional centres with specialist nurses and respiratory scientists will assist significantly in the liaison between discharge coordinator and LTV nurse specialist in the tertiary centres, in order to enable local support after discharge, or to enable bridging to local discharge via regional centres.</p>
Severe asthma	<p>Successful and effective management of these children requires a multidisciplinary team approach. Services should be centralised to a single national centre with significant expertise and appropriate staffing (consultant in paediatric respiratory medicine, respiratory scientists, asthma nurse specialists, paediatric pharmacists, psychology and social work). Services are provided most effectively if all children with complex and severe asthma are managed in one centre. Currently, any of the six paediatric respiratory centres could manage children with asthma with high levels of complexity; however, most of these centres have inadequate staffing and resources to do so. There is no allocated national referral centre for children with complex/severe asthma and, currently, referral pathways are ad hoc.</p>
PCD	<p>Currently, all diagnostic testing for Ireland is provided by specialist paediatric PCD centres in the UK. Ireland is likely to have a sufficient number of patients with PCD to set up a single specialist PCD centre that may be capable of providing a complete, or almost complete, diagnostic and therapeutic service. There are currently no clinical nurse specialists (CNSs) for children with PCD. Most centres have difficulty accessing standard aspects of care such as nurse specialist access, regular physiotherapy and specialist ENT opinion.</p> <p>There is also a significant lack of appropriate physiotherapy respiratory services. Inpatients are seen in tertiary units, as referred. However, no physiotherapy cover is provided at outpatient clinics. There is a lack of physiotherapy outpatient follow-up service/absence of community physiotherapy respiratory services for patients with PCD, which further restricts access for these children.</p> <p>Currently, patients with PCD in Ireland have a huge number of unmet needs. As PCD is likely to be under-diagnosed in Ireland, and as related services evolve and become more established, the diagnosis and referral rate is likely to increase pro rata. Currently, there is no physiotherapy staff provision for this service in tertiary and regional units, and this issue should be addressed.</p>

General respiratory services	In addition to the above challenges relating to specific aspects of respiratory medicine, the provision of general respiratory services also faces a number of challenges in many paediatric units. Most units have long waiting lists for outpatient appointments for respiratory clinics. The primary reason for this is the inappropriate distribution of patients to these clinics. A significant proportion of patients seen in all respiratory clinics could easily be dealt with at a primary or secondary care level by GPs and general paediatricians. Many children seen in the tertiary units in Dublin could be dealt with at a local level if appropriate respiratory scientist and nurse specialist resources were in place at that level. In order to meet current service demands, there would be a sufficient number of specialists available to see children in a timely fashion were it not for requirements for specialists to see children who could be effectively dealt with by other professionals. The distribution of clinical caseload, complexity and staffing is uneven across Dublin.
-------------------------------------	---

Challenges in Regional Service Provision

	Cork	Limerick	Galway
Access to paediatric pulmonary function testing	No laboratory for children. No paediatric staff. Very limited access to adult pulmonary function testing (PFT) laboratory. New paediatric respiratory laboratory needed for pulmonary function/sleep.	No laboratory for children. No staff. New respiratory laboratory needed for pulmonary function/sleep.	No laboratory for children. No staff. New respiratory laboratory needed for pulmonary function/sleep.
Initiation of non-invasive ventilation (NIV)	No CNS, no clinical engineering support.	No CNS, no clinical engineering support.	No CNS, no clinical engineering support.
Non-CF respiratory clinical nurse specialist (CNS)	None.		
(Asthma nurse specialist part funded via fundraising for six hours a week)	None	None	
Access to non-CF respiratory physiotherapy	Very limited	Very limited	Very limited
Sleep diagnostics	CR-PSG equipment but no staff or other resources. TCO ₂ , overnight oximetry.	TCO ₂ , home overnight oximetry. No staff or other resources.	TCO ₂ only. No staff or other resources.
Beds for children on LTV	Two HDU beds available, usually full in winter with respiratory patients.	Two HDU beds available, usually full in winter with respiratory patients	No dedicated or supported beds

Barriers to taking children on LTV	Lack of training, awareness among non-consultant hospital doctors (NCHDs), nurses. Limited HDU beds at times.	Limited HDU beds (at times). Delays in establishing home care package – funding, staffing, training.	Lack of training, awareness among NCHDs, nurses. No HDU beds.
Access to VFL and SALT	SALT and videofluoroscopy available on site but SALT staffing levels suboptimal.	None	Limited access
Access to chest CT under sedation/general anaesthesia (GA)	Limited access to CT chest under GA. Sedated CT chest possible.	No access to GA, sedated children over one year possible.	MRI/CT available under GA on case by case basis.
Access to bronchoscopy	Regular list – Limited expertise in infants < one year old.	Regular list (four weekly). Limited anaesthetic support for young infants and toddlers – solution being sought.	None

Flow of Complex Patients

A number of children with complex respiratory needs (such as pulmonary aspiration, obstructive sleep apnoea, requirement for non-invasive ventilation or airway adjuncts) are transferred from regional centres to tertiary centres in Dublin for comprehensive multidisciplinary evaluation and treatment. It is often extremely challenging to repatriate these children to their regional centres as a bridge to their discharge home. This relates mostly to a lack of resources, training and expertise at a ward level in the regional centres. The development in staffing of regional centres with localised respiratory expertise, working within a national network, would significantly facilitate this and assist in the effective management of patient flow in and out of the tertiary centres (and ultimately the new children's hospital). This would have clear benefits in terms of bed utilisation.

42.3 PROPOSED MODEL OF CARE

Cystic fibrosis

A comprehensive review of the model of care and standards of care for adults and children with CF is outside the scope of this document. This will be dealt with in greater detail by the National Clinical Programme for Cystic Fibrosis.

In December 2011 the UK Cystic Fibrosis Trust published Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (Second Edition). This document provides recommendations across a range of areas as follows:

- The diagnosis of CF should not be delayed, but must be handled sensitively and followed by education of the parents/carers and patient.
- All patients must be under the direct supervision of, and with regular follow-up from, an adequately resourced, designated specialist CF centre (sometimes in partnership with a local CF centre).

- Specialist multidisciplinary care must be delivered by a team of trained and experienced CF specialist health professionals, with staffing levels appropriate to the size of the patient population.
- Measures must be in place to prevent cross-infection from other patients.
- Treatment of airway infections is critical, and antibiotics are a key part of CF therapy.
- Chest physiotherapy with airway clearance techniques is a lifelong mainstay part of treatment.
- Nutrition support is crucial for all patients.
- Other manifestations of CF, as well as complications – particularly liver disease and impaired glucose metabolism – must be recognised promptly and, in some cases, should be screened for.
- Psychosocial support is often required and should be available.
- Transition to adult care should be planned and managed appropriately.
- Transplantation, palliative and end-of-life care must be planned and managed appropriately.

Both the recent 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 child 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, concentration of expertise and delivery of a high-quality service takes precedence over issues of geographical access. Concentrating care for CF patients in specialist paediatric CF centres ensures that the multidisciplinary team will see sufficient numbers of patients to enable them to maintain expertise, so that they treat CF patients effectively, recognise the more unusual manifestations and delay the onset of the multi-system complications associated with the condition. The importance of Specialist CF Centre care cannot be over-emphasised, and has been recognised by the 2009 HSE report on CF Services in Ireland: <http://www.hse.ie/eng/services/publications/topics/reptcysticfibrosis.pdf>

The Royal College of Paediatrics and Child Health, the Royal College of Physicians of London, the British Thoracic Society, and the British Paediatric Respiratory Society all agree on the need for specialist CF centres. The US Cystic Fibrosis Foundation and the European Cystic Fibrosis Society also strongly endorse the principle and importance of Specialist CF Centre care. 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 children in Ireland attending specialist centres for all of their care. It should be noted, however, that many children do not live within close commuting distance of one of the six specialist CF centres in Ireland, and access to services for these children could be challenging.

In other countries, including the UK, specialist CF centres work with local hospitals in an attempt to provide specialist care closer to home while maintaining a high standard of care. This has worked well in Wales, in particular, where outcomes in an unsupervised, locally based system have been shown to be poor. In contrast, outcomes approach specialist care levels where a local hospital works in a partnership under the supervision of a specialist centre (Doull and Evans, 2012). 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, and there are no clear guidelines or standards for the operation of shared care in these centres.

Basic Principles of CF Care Delivery

- All patients with CF must have their care delivered under the direct supervision of a recognised specialist CF centre for treatment throughout their lives.
- The child, family and local clinic should be aware that they are a patient of the specialist centre.
- Children will either receive full care from a specialist CF centre, or shared care within a designated local clinic within the specialist centre's network.
- Parents should be aware of their options and know that they can choose full care from a specialist CF centre if they wish.
- Care delivered by a combination of a designated local hospital 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 and standard operating procedures as laid down by the specialist CF centre. There must be regular communication between the consultants and the multidisciplinary team in the specialist and designated local CF centres.
- The responsibility for the management of the quality of the shared care arrangement, and the quality of the patient care, should rest with the specialist CF centre. If the quality of local hospital services cannot be safely maintained, e.g. loss of staff or facilities, patient care should be delivered exclusively at the specialist CF centre.

Specific recommendations in relation to designated shared care centres, geographical referral patterns and guidance on service-level agreements for shared care will be produced by the National Clinical Programme for Cystic Fibrosis. The current model of CF care delivery within the six specialist cystic fibrosis centres should continue. It is vital that appropriate staff resources are put in place to ensure efficient and effective running of each of these services and to avoid any variation in service quality or availability.

HSE recommendations made in 2009 for staffing of CF centres are outlined in Table 42.1. Current staffing levels in all paediatric CF centres fall below recommended levels.

Staff member	Paediatric	Adult
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
Dietitian		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

Table 42.1: Recommended staffing levels for specialist paediatric and adult CF centres per 50 patients. Source: Services for People with Cystic Fibrosis in Ireland. HSE 2009.

Sleep

In light of the very underdeveloped services in Ireland for sleep disorders in children, the high prevalence of these disorders, and the established network of paediatric respiratory centres in Ireland (based on specialist CF centres), we have an exciting opportunity to develop an integrated national service spanning primary care to tertiary subspecialty services.

Primary Care

Information on relevant symptomatology and possible consequences of disorders of childhood sleep physiology should be incorporated into training for public health nurses, school nurses and those involved in developmental screening in childhood. The development and incorporation of appropriate questionnaires on sleep into routine developmental screening, coupled with more widespread recognition of the potential contribution of sleep disorders to poor school performance and behavioural problems, is likely to increase the appropriate and early recognition and referral of affected children. General practitioners should be aware of sleep disorders in children, and have ready access to local diagnostic services through the local hospital.

Local Paediatric Units

The high prevalence of sleep disorders in children means that the majority with suggestive symptoms will be most appropriately seen, investigated and treated by the local paediatric secondary care service. In addition to the appropriate level of training for consultant paediatricians, this will require the availability of the necessary equipment (with robust artefact detection or signal extraction facilities) to carry out overnight recordings of pulse oximetry on children at home. For a local paediatric service serving a population of between 50,000 and 60,000 children, with 3,000 births per year, a single-recording oximetry system is likely to be sufficient for this purpose. Some provision for this service must be made in job plans for medical and support staff, even though the time commitment is likely to be small (approximately six hours of consultant time per month). It is essential that the clinicians involved in this service work in liaison with the regional centre to ensure a smooth patient journey.

Regional Paediatric Units – Cork, Limerick and Galway

Regional units would all have the capacity to perform home oximetry studies, inpatient TCOM studies and cardiorespiratory polysomnography (either at home or in inpatient beds). In order to perform these types of studies, personnel trained in the technical aspects of equipment maintenance, scoring, basic analysis and quality control will need to be in place. The regional centres will also be required to initiate children on NIV, perform titration studies, downloads and compliance checks, and provide back-up and support to the patients at home. Based on the workload of such centres currently undertaking this level of service provision in the UK, and covering a population of approximately 1 million, the equivalent figure for Ireland is likely to be in the range of 0.5 whole time equivalent (WTE) consultant's time, plus one WTE nurse specialist (or respiratory scientist) per regional unit as a minimum.

Supra-regional Units – Crumlin/ Temple Street/ Tallaght (pending development of New Children's Hospital)

On the basis of the 2009 Royal College of Paediatrics and Child Health (RCPCH) report, tertiary-level investigational and treatment services for children with sleep disorders should be available in at least one unit nationally. Facilities should also include the full range of "second line" investigations, together with appropriate staff and resources to conduct such investigations. The supra-regional unit will also be required to initiate children on NIV, perform titration studies, downloads and compliance checks, and provide back-up and support to the patients at home. It is envisaged that the supra-regional centre will have significant numbers of children on NIV, many of whom may have complex needs and co-morbidities.

The unit will also need to provide more complex investigational facilities and expertise, e.g. quantitative recordings of minute ventilation, and combined neurophysiological and respiratory recordings for children with complex neurological disorders affecting sleep physiology, and those with disorders of respiratory control, e.g. congenital central hypoventilation syndrome (CCHS). For a centre providing detailed investigational and treatment facilities for children with complex neurological and respiratory control disorders, and covering a population of 5 million people, the additional staffing required (based on the current workload in a similar system in Sheffield at present) will require at least two WTE consultants, two nurse specialists and three basic respiratory scientists or equivalent, one WTE senior respiratory scientist and one WTE chief respiratory scientist as a minimum.

The supra-regional sleep laboratory will require overnight staffing by technical personnel, in order to ensure the appropriate quality of the output. Efficiency will need to be maximised in terms of both staffing and turnover. What is envisaged is a four-bed unit requiring two respiratory scientists overnight to perform a combination of full polysomnography (PSG), cardiorespiratory PSG and TCOM on any one night (Standard for Sleep Disorders Services ASA 2012). This service could run a minimum of three nights per week, and subsequently be increased as necessary on the basis of service needs and staffing. This allows for a more cost-effective service than a single or two-bed unit. This has been well demonstrated in the UK (Transforming Respiratory and Sleep Diagnostic Services DOH NHS 2009). Staffing will be required during daytime hours for scoring (very labour intensive), maintenance, laboratory management, data downloads and nap studies in infants.

The key organisational idea in developing a tertiary/quaternary sleep service is to keep children requiring outpatient tests out of inpatient beds. This is not happening at the moment, and is incurring significant costs, inefficiencies and an increase in workload for all concerned. An efficient system should run completely in parallel with the inpatient service. For this reason, sleep laboratories need to be observed by trained respiratory scientists. Sleep laboratories do not need nursing support, as sleep testing is seen as a purely outpatient diagnostic service such as EEG, or pulmonary function. Children come directly from home for their sleep diagnostic test and return home afterwards. Children who are too sick or unstable to have sleep tests safely performed as an outpatient should, by definition, be in an inpatient bed until they are well enough to be cared for safely at home.

The model of care for Ireland should be based on a hub-and-spoke model, with the supra-regional centre as the hub for regional centres, and the regional centres as the hub for the general paediatricians in the local centres. Each centre needs to have a clearly identified consultant and nurse/technician who take responsibility for their service. There should be clear referral pathways, clinical guidelines and procedures between primary, secondary and tertiary-level services, with clinical leadership and support provided by the paediatric sleep consultants to the respiratory paediatricians and general paediatricians involved in the diagnosis and assessment of sleep disorders. Diagnostic sleep equipment and software should be standardised across all centres, with the potential for a small national network where studies can be seen in all centres (akin to the National Integrated Medical Imaging System(NIMIS)), helping with quality control, standards, education and training.

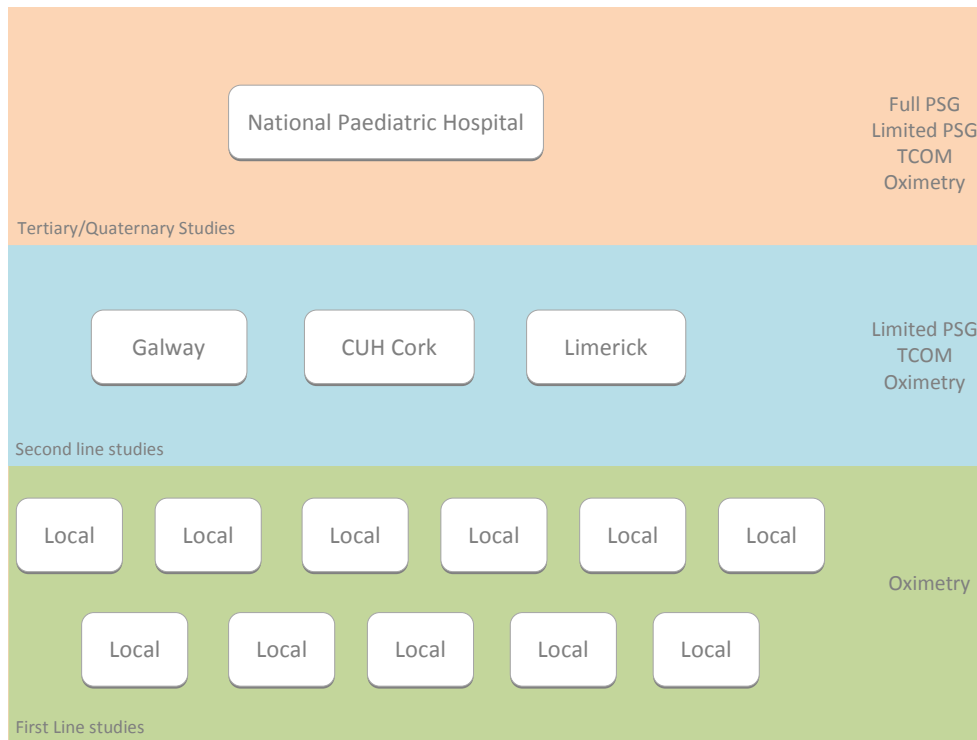


Figure 42.1: Proposed national paediatric sleep service structure

As part of the plan to develop a national paediatric sleep service, as outlined in Figure 42.1, the following minimum staff will be required per unit (for sleep services specifically):

Local	Regional	Supra-regional
One session per month consultant time	0.5 WTE consultant	Two WTE consultants
0.2 WTE nursing/technician support	One WTE nurse/respiratory scientist	Two WTE respiratory nurses One WTE chief respiratory scientist One WTE senior respiratory scientist <ul style="list-style-type: none"> • Four WTE basic respiratory scientists • 0.5 WTE clinical engineers • 0.5 WTE psychologist
Physiotherapy support	Physiotherapy support	Physiotherapy support
Administrative support	0.2 WTE Administrative support	One WTE Administrative support

<p>Rationale: Local paediatric units will be dealing mainly with low-complexity, moderate- volume caseloads. The majority of the workload here will be in the form of overnight oximetry in children with suspected obstructive sleep apnea (OSA) referred from GPs and ENT surgeons. The administration and technical aspects of this workload could be carried out by nurses, a proportion of whose time could be allocated to diagnostic sleep testing.</p>	<p>Rationale: Given the local catchment workload, acceptance of children from linked local centres, provision of first- and second-line studies, an NIV service and liaison with the tertiary centre, and full-time technical/nursing support will be required.</p>	<p>Rationale: The supra-regional unit would act as the hub of the national paediatric sleep service. It would deal with tertiary sleep referrals which require a fully operational, four-bed sleep laboratory. It would also undertake provision of local first- and second-line studies and the running of an NIV service. The supra-regional centre would create national guidelines and policies on paediatric sleep and NIV, and would ensure adequate support and education provision for regional units.</p>
--	---	--

Neuromuscular Disease (NMD)

The overall care of children with neuromuscular disease (NMD) should be under the supervision of a neurology team. As children get older and develop respiratory issues, the requirement for respiratory input increases significantly. Multidisciplinary input is essential from the following services: respiratory, cardiology, orthopaedic surgery, physiotherapy, occupational therapy, speech and language therapy, dietetics, specialist nursing, psychology and social work. For this reason, and based on concentration of specialist care, a multidisciplinary neuromuscular clinic is, therefore, best suited to a tertiary referral hospital. From a respiratory perspective – and certainly prior to the advent of diurnal hypoventilation – children with NMD can be effectively cared for in a regional centre of expertise. With the evolution of more advanced respiratory failure and diurnal hypoventilation, the need to treat children with NMD from a respiratory perspective becomes more specialised, and is likely to benefit from the input of tertiary physiotherapy, occupational therapy, orthopaedic and respiratory input. A single national referral centre for children with NMD is likely to be established at the new children’s hospital. From a respiratory perspective, all such children should be seen in this hospital at least once a year, but more frequently if they are based in the Dublin area or have more advanced lung disease. The respiratory and physiotherapy teams at the new children’s hospital should liaise closely with regional centres that may be reviewing these patients between hospital visits.

The new children’s hospital should have a clinical lead for respiratory complications associated with NMD. This person would be a consultant in paediatric respiratory medicine who predominantly attends neuromuscular multidisciplinary team clinics, and leads on education and service development in the area of respiratory complications associated with NMD. A respiratory nurse specialist with responsibility for long-term invasive and non-invasive ventilation should work alongside both the consultant in paediatric respiratory medicine and an NMD nurse specialist (allied to the neurology team) in terms of diagnostic and therapeutic respiratory interventions for this group of patients. Within the respiratory section of the Department of Physiotherapy one or two physiotherapists should lead in the area of NMD in terms of education and service development. Respiratory physiotherapy for patients with NMD is vital both in the home setting (for out-of-hospital maintenance) and during the acute in-hospital stay period.

Long-term Ventilation (LTV)

Children on long-term mechanical ventilation (LTV) are referred directly as inpatients, (usually while in a critical care setting), prior to the insertion of a tracheostomy tube and the initiation of LTV. The expertise for LTV should be centred at Crumlin/ Temple Street, pending the establishment of the new children’s hospital. The centre for LTV in Crumlin is already established, and comprises a multidisciplinary team of nurses, occupational therapists, physiotherapists, dietitians, speech and language therapists, social workers and respiratory consultants. While it has ring-fenced staffing for the provision of physiotherapy and speech therapy, it does not have dedicated staff for the provision of occupational therapy. Similarly, in Temple Street, children are seen by the general staff pool. For the LTV patient, physiotherapy is of significant importance. The physiotherapist works closely with the patient during periods of respiratory compromise, in addition to providing regular neurodevelopmental therapy aimed at maximising physical independence. Achieving motor milestones is crucial for the LTV child, as mobility and general activity will optimise respiratory health. Occupational therapists working with LTV patients will provide assessment, advice, education, training, intervention, equipment provision, rehabilitation and discharge planning and facilitation. Currently, neither Crumlin nor Temple Street has an LTV nurse specialist who could act as a key worker for the management of these patients. A dedicated discharge coordinator for LTV patients should be identified in each centre. There needs to be clear communication between both a liaison nurse based in the community, the LTV nurse specialist and discharge coordinator, and specialist nurses in regional centres, in addition to a liaison person who will communicate with consultants in specialist centres as well as consultants in regional centres. In addition to the provision of key workers in the LTV area, there should be a system-wide approach to the funding and delivery of home care packages.

Figure 42.2 outlines the proposed model of care for LTV services across Ireland:

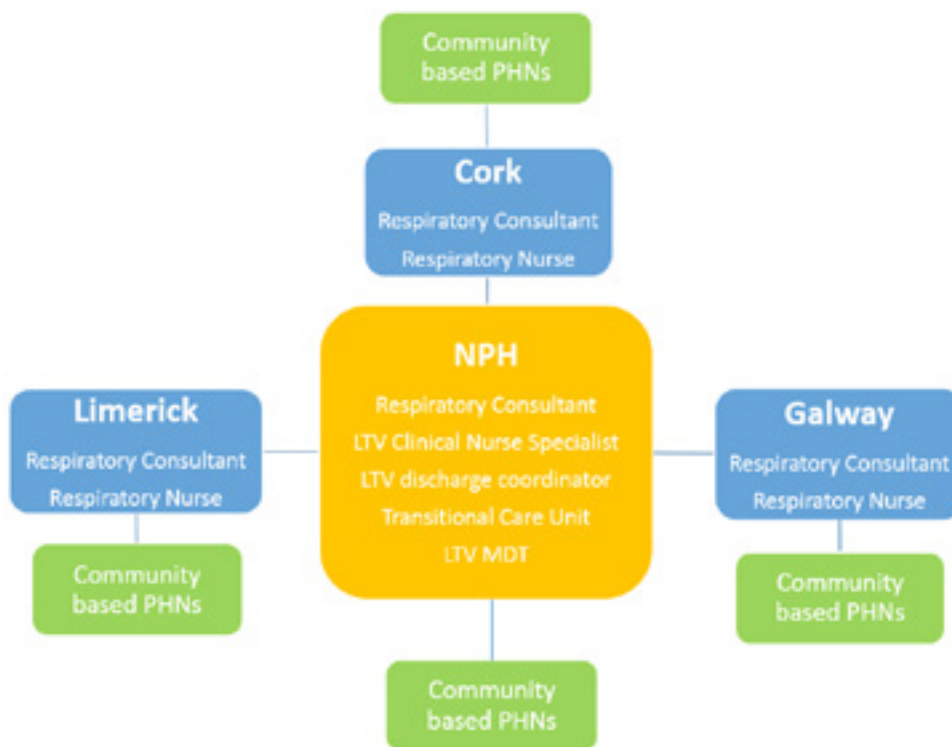


Figure 42.2: Proposed model of care for LTV services across Ireland

Severe Asthma

Asthma is an extremely common condition, and the vast majority of children with asthma have mild-to-moderate disease. Most children with asthma can be effectively cared for in a primary and secondary care setting. Children whose asthma control is poor, despite attending a secondary care paediatrician, should be referred to a consultant in paediatric respiratory medicine. The vast majority of children who are referred to a paediatric respiratory physician can be effectively managed without the requirement for advanced diagnostic testing or further onward referral. This document will not deal in detail with non-severe asthma, as this is addressed in detail by the National Clinical Programme for Asthma.

In order to plan a model of care for delivery of severe asthma services, it is essential to agree a meaningful definition of severe asthma. In 2009, an expert panel met to propose the World Health Organization (WHO) definition of asthma severity and control. This group suggested that severe asthma includes three distinct groups:

- Untreated severe asthma, due to undiagnosed asthma or unavailability of therapy
- Difficult-to-treat severe asthma (due to adherence issues, inappropriate or incorrect use of medicines, environmental triggers or co-morbidity)
- Treatment-resistant severe asthma, including asthma for which control is not achieved despite using the highest level of recommended treatment, or asthma which is controlled only with the highest level of recommended treatment.

Children in the first two groups above can effectively be managed at any of the tertiary or regional centres. It is suggested that a single severe asthma clinic be established at the New Children's Hospital. Referrals to this clinic should be made only by other consultants in paediatric respiratory medicine.

Primary ciliary dyskinesia

Primary ciliary dyskinesia (PCD) is a disorder with a respiratory component that is not dissimilar to CF. Children with PCD have chronic and recurrent respiratory infection and require ongoing management of infection and airway clearance. A PCD service would have close ties, and would overlap, with a CF service. While PCD is a rare disorder, it is likely that there is a sufficient number of patients with this condition in Ireland to merit developing a specialist national centre for children with PCD, based at the new children's hospital. The specialist centre should be staffed by a respiratory consultant (PCD centre director) and have access to a nurse specialist and physiotherapy services. Patients should have access to regular assessment by a physiotherapist with expertise in respiratory medicine. Daily physiotherapy, including airway clearance and exercise, is essential for augmenting airway clearance, in order to delay the onset and progression of obstructive airway disease. Specialist paediatric ENT services should be available, with expertise concentrated in one or two ENT surgeons. Given the geographical dispersion of patients with PCD within the respiratory subspecialty network, a shared care arrangement for these patients should be in place; such children should be seen annually at the specialist centre and at all other times at the local regional centre. Regionally, care for children with PCD should be provided in parallel with CF services. There are insufficient numbers of PCD children in any of the regional centres to necessitate PCD-specific nurse specialist services. However, given the small numbers of children with PCD, amalgamation of PCD services with local CF services (if appropriately staffed) would seem logical. At the new children's hospital, a designated nurse specialist for PCD should be employed with cross-cover in place between the PCD nurse specialist and CF nurse specialists. Physiotherapy services for children with PCD should be included in the calculation of physiotherapy requirements for children attending the new children's hospital. There should be protected numbers of physiotherapists for each of the individual specialties of PCD, CF and NMD. However, there should be the ability to cross-cover between these services when required.

Much of the complexity involved in the management of children with PCD involves establishing a diagnosis.

The key diagnostic tools are as follows:

- bronchoscopy with ciliary brushing
- electron microscopy of epithelial cells
- live video ciliary analysis
- nasal nitric oxide measurement
- primary epithelial culture.

There are six PCD diagnostic centres in the UK, covering a population of 63 million. Currently, in Ireland, there is no expertise in electron microscopy of epithelial cells from children with suspected PCD. Nasal nitric oxide and video ciliary analysis are intermittently available, but only for a small number of patients. Primary culture of epithelial cells is not available in a clinical setting, but is available in a research setting. Overall, there is insufficient infrastructure and expertise in Ireland to establish a completely independent PCD diagnostic service. Currently, paediatric centres in Ireland send samples to the UK for diagnosis. The need for Irish children to travel to the UK for diagnosis occurs only infrequently. Formal transition links between specialist paediatric services and specialist adult services should be fostered and actively managed.

Respiratory Services at the New Children's Hospital

The new children's hospital will house a large and complex Department of Respiratory Medicine. The Department will have a director (a consultant in paediatric respiratory medicine) and should have a lead administrator (a senior secretary/administrator). There will be a requirement to have a number of virtual centres within the Department:

- The Sleep Centre
- The Cystic Fibrosis Centre
- The Centre for Long-Term Ventilation

Each of these centres will have some of its own dedicated staff (although there will be some overlap) and a director (a consultant in paediatric respiratory medicine). Some departmental staff will be shared across all centres.

In addition to these centres, the hospital will run a number of complex clinics:

- The severe asthma clinic
- The primary ciliary dyskinesia clinic
- A multidisciplinary clinic for children with neuromuscular disease (run by the neurology service)
- The sleep clinic

The Department will have two diagnostic laboratories:

- The pulmonary function laboratory
- The sleep laboratory

These diagnostic laboratories will operate separately and have separate staff. Each will have a director (a consultant in paediatric respiratory medicine).

The department will have several different broad groups of inpatients accounting for most of the inpatient activity:

- Children with cystic fibrosis
- Infants with suspected pulmonary aspiration, obstructive sleep apnoea, respiratory failure
- Children with neuromuscular disease and respiratory compromise
- Children with empyema
- Children on long-term mechanical ventilation in the transitional care unit (TCU)
- Children with complex airway disease
- Children with pre-existing respiratory conditions with acute infection.

From a respiratory perspective, management of these children would be best facilitated if children could be cohorted to a single ward or location. The core competencies required from inpatient staff on a respiratory ward include management of children with cystic fibrosis, an understanding of non-invasive and long-term mechanical ventilation, an understanding of basic and assisted airway clearance, and familiarity with children with tracheostomies.

Currently, in Crumlin, governance and medical supervision of TCU is unclear. TCU originally started as an offshoot of PICU, and therefore PICU has always been involved in the management of TCU patients to some degree. Due to increasing familiarity with stable children on LTV, and an increasing realisation that the primary focus of the inpatient stay is centred around developmental progress, with mechanical ventilation merely as an adjunct to achieve this, it is clear that TCU should be viewed more as a regular inpatient ward than an intensive care area. The governance of TCU from a medical point of view should be shared between respiratory and general paediatric services. From a nursing point of view, given that clinical practice in TCU is more similar to respiratory inpatient management than intensive care, governance of TCU should be grouped with the respiratory inpatient ward rather than with ICU. A respiratory inpatient ward should be capable of looking after children with CF, children with suspected pulmonary aspiration, children with obstructive sleep apnoea, children with empyema, children on long-term mechanical ventilation, children on non-invasive ventilation, and children with neuromuscular weakness affecting the respiratory system. The respiratory inpatient ward should include an area designated for TCU patients. Some plasticity will be necessary here, given the natural variation in numbers of patients on long-term mechanical ventilation over time.

The Department will liaise closely with a number of health and social care professional groups. Key among these will be physiotherapy. Physiotherapy is of central importance for CF, LTV, NMD, PCD, non-CF bronchiectasis and several other respiratory diseases. Given the very close relationship between physiotherapy and respiratory medicine, and the overlap between the physiotherapy requirements of the different disorders, a finite 'respiratory physiotherapy' group within the Physiotherapy Department should be developed. Within this group, specific and unique expertise will be required in the areas of CF and NMD, in which there should be natural specialisation. Occupational therapists work closely with LTV and NMD patients. There is an opportunity to establish and develop a role for occupational therapists to work with other patients who have respiratory conditions, and are being treated in the new children's hospital.

As the centre of the national hub-and-spoke model for paediatric respiratory medicine, the Department of Respiratory Medicine in the new children's hospital will need to maintain close clinical and administrative liaison with regional specialist centres in terms of education, audit, quality assurance, research and service development. In this regard, a chief administrator and full-time data manager will be required. An educational network will also need to be maintained; it should be directed by the new children's hospital, and be under the auspices of the Irish Thoracic Society, for the maintenance of skills and professional development in both the supra-regional centre and regional specialist centres in the following areas:

- Medical
- Nurse specialists
- Respiratory physiotherapy
- Respiratory scientists

A key aspect of this vertical relationship with the regional centres should involve some element of joint working or joint appointments. It should be possible for specialists in regional centres, if desired, to have a joint appointment with the new children's hospital and attend on a scheduled basis to participate in clinics, reporting and inpatient care. In a similar vein, satellite clinics for patients with complex conditions should be held on a scheduled basis in each of the regional centres; tertiary specialists can also run clinics in these centres jointly with regional specialists. This arrangement would foster improved communication, quality assurance and education, and strengthen the specialist network.

Specialist Regional Units

Specialist regional units will need to provide specialist care in the following areas:

- Cystic fibrosis
- Paediatric sleep medicine
- Non-invasive ventilation
- Asthma
- Neuromuscular disorders
- General respiratory services for local children (in some instances in a shared care arrangement)

In order to provide a high standard of service across all of these areas (between which there is significant overlap), each regional specialist centre will need access to the following staff:

- A full-time respiratory scientist (pulmonary function testing – CF, NMDs, general respiratory and diagnostic sleep testing)
- A full-time respiratory physiotherapist (CF, NMD, non-CF bronchiectasis, general respiratory services, local inpatient respiratory physiotherapy)
- Full-time administrative support (CF, paediatric sleep medicine, NMD, NIV, liaison with the new children's hospital administration and data management)
- Full-time respiratory nurse specialist (NMD, asthma, sleep, NIV, general respiratory services)

Each regional specialist centre should have the following staff:

- Consultant (covering respiratory, sleep, NMD and CF)
- Administrator (covering respiratory, sleep and CF)
- CF nurse specialists
- CF dietitian
- CF psychologist
- CF social worker
- CF pharmacist
- Respiratory nurse specialist (covering respiratory and sleep)
- Respiratory physiotherapist (covering respiratory and CF)
- Respiratory scientist (covering respiratory, sleep and CF)
- Access to a paediatric occupational therapist

There will be a requirement, at regional level, for the respiratory scientist, respiratory physiotherapist and respiratory nurse specialist to work closely together for a number of reasons. Firstly, there is significant overlap in the roles of each of these professionals in terms of service delivery. Secondly, in order to optimise efficiency and reduce service gaps during leave, some element of cross-cover in certain aspects of practice (generic skills) will

need to be provided. There is unlikely to be sufficient need for two WTE posts in each of these areas at the outset, and so cross-cover will ensure some degree of ongoing service delivery. Thirdly, this will help significantly in terms of development of overall staff education and competence. All staff in the subspecialty respiratory service will be required to take part in continuous professional development and education within the national network for paediatric respiratory medicine.

Specialist regional centres should have access to the following diagnostic modalities locally:

- Spirometry, lung volumes
- Overnight oximetry
- Overnight oximetry/capnography
- Cardiorespiratory polysomnography
- Data download from non-invasive ventilators
- Speech and language therapy with videofluoroscopy
- Specialist regional centres should have access to the following diagnostic modalities either locally (preferable) or by direct referral to another centre:
 - Paediatric bronchoscopy
 - CT scanning under sedation/anaesthesia

It is expected that regional specialist centres will be able to effectively manage the following conditions:

- Cystic fibrosis
- Obstructive sleep apnoea
- Neuromuscular disorders with mild/moderate respiratory involvement
- Moderately severe asthma
- Sleep-related hypoventilation
- Chronic non-invasive ventilation
- Patients with complex respiratory conditions. Such patients would be managed in conjunction with the new children's hospital.

Regional centres should act as first-line referral centres for respiratory problems in the region, except in situations where a clear requirement exists for direct tertiary referral. In addition, regional centres will be required to work with the supra-regional centres to allow some element of shared care services to be delivered closer to the homes of some children with complex respiratory disorders.

Patient Journey

This section of the document details some current issues with referral pathways, and outlines what we feel should apply in the future.

Given the array of different services provided by specialists in paediatric respiratory medicine, referral pathways differ for many respiratory conditions. Some pathways are outlined below.

- *Cystic fibrosis (CF)*

Today, the vast majority of children diagnosed with cystic fibrosis (CF) will be diagnosed through the newborn screening programme. These children are automatically referred to one of the six specialist CF centres. Most of their care will be provided locally in specialist CF centres. There may be some requirements for referral to a tertiary centre or to the national referral centre, which can cater for more complex care requirements.

Given the excellent network of paediatric CF centres that already exists in Ireland, access to services for children with CF is very good overall. Currently, the exception to such access occurs whenever CF centres are experiencing temporary staff shortages. In the past few years, a number of paediatric CF centres have experienced ongoing staffing problems, which resulted in access to services being suboptimal. CF children and their families should have access to a nurse specialist, dietitian, physiotherapist, and social worker on demand during normal office hours. Moreover, there should be ready access to CF consultant and psychologist support and advice on a day-to-day basis.

- *Obstructive sleep apnoea (OSA)*

Children referred for evaluation of suspected obstructive sleep apnoea (OSA) (the most common sleep disorder) can be broadly categorised into two groups. As follows:

1. Otherwise well children, with OSA as an isolated problem

Currently, many children who are suspected of having OSA are referred directly to an ENT surgeon. In many situations, management of the patient proceeds without any sleep investigations. Currently, if an ENT surgeon requires a paediatric patient diagnosis, he/she must refer the patient to a paediatric sleep specialist, and then wait for a report, prior to reviewing the child again. In terms of efficiency, clinical excellence, avoidance of morbidity and communication effectiveness, it is preferable for a child to see a paediatric sleep specialist in advance of seeing an ENT surgeon if the medical team are concerned that the child may have OSA.

The appropriate referral pathway for otherwise well children with suspected OSA should be from their general practitioner to a local/regional paediatrician with training in sleep medicine. The paediatrician should counsel the family about OSA, the need for investigations, various treatment modalities, and the natural history of the condition. The paediatrician should then make the appropriate referral, depending on the diagnosis. If a diagnostic test is not required, appropriate advice should be given and the child should be observed. If a diagnostic test is required, the child should be referred to the local/regional paediatric sleep service. If OSA is excluded, the child should be observed. If OSA is diagnosed, an assessment should be made as to whether there may be a surgically reversible component to the obstruction. In most circumstances this should be determined by an ENT surgeon. If the condition is felt not to be reversible, or cannot be cured with surgical intervention, referral to a regional/tertiary sleep specialist for initiation of non-invasive ventilation may be necessary.

2. Children with complex medical needs/co-morbidities who are suspected of having OSA

Children with complex medical needs usually attend a consultant paediatrician. As part of a system review – and if the consultant paediatrician is concerned about potential OSA – the child should be referred for investigation and management of this condition. Currently, most paediatricians will refer directly to an ENT surgeon. In most circumstances, an ENT surgeon will request a formal diagnostic sleep test. In some instances a surgical procedure will be carried out, but for many children with complex medical needs OSA is not surgically reversible, and medical treatment will be required under the direction of a paediatric sleep specialist.

Children with complex medical needs who are suspected of having OSA should be referred to a paediatric sleep specialist in the first instance. The paediatric sleep specialist must perform the appropriate diagnostic tests and, if necessary, refer the child to an ENT surgeon. Many children will require treatment with non-invasive ventilation or other nonsurgical approaches. This can often be commenced immediately after they have been seen by a paediatric sleep specialist, and without having to wait for an ENT appointment.

Given the underdeveloped nature of paediatric sleep services in Ireland, access to services is currently poor in most parts of the country. Access to expertise, diagnostic tests and treatment modalities is inequitable, and is dictated by where the child lives.

While tertiary paediatric sleep services are provided in the Dublin centres, waiting times for diagnostic testing and treatment are prolonged. The provision of sleep services outside of Dublin is patchy. Cork and Limerick provide a limited paediatric sleep service without the aid of technicians or nurse specialists. Some regional and local centres provide oximetry, which can be used as a screening tool for OSA but, due to a lack of local expertise and equipment, many of these cases subsequently require referral to tertiary centres. Much of the sleep services provided by tertiary centres could be provided in regional and local centres, if they were adequately resourced. The inappropriate overloading of tertiary centres with non-tertiary-level sleep disorder cases has resulted in excessively long waiting lists.

- *Neuromuscular disease (NMD)*

In order to provide the most effective care for children with NMD, a very significant number of staff is required. Children with generalised neuromuscular disease (NMD) will have first been seen by a paediatrician or paediatric neurologist. As outlined above, children with generalised NMD should be seen in a multidisciplinary clinic. In addition, as part of the service, all children should be seen by a respiratory physician on a regular basis. Children with NMD require regular pulmonary function testing and need to undergo sleep studies. In order to manage these children's health needs, a very significant input is required from respiratory physiotherapy services. The ideal delivery of services to children with NMD should be coordinated by a central team, and should involve input from all of the subspecialty teams. In centres where this works well, the supervision of care is assumed by general paediatricians, neurologists or specialists in physical medicine and rehabilitation. It is vital to establish the clinical governance for children with NMD. Prior to the establishment of the new children's hospital the governance and responsibility for this group will need to be clearly agreed. Currently, there are disparate arrangements in place in the various children's hospitals for children with NMD. Overall, however, the numbers of staff dedicated to caring for children with NMD are insufficient.

Access to respiratory services for children with NMD are non-uniform across the country. Children with respiratory complications of neuromuscular disease should ideally be referred directly through multidisciplinary neuromuscular clinics to a consultant in respiratory medicine. Children with NMD can suffer acute and rapid decline, and their respiratory management is not well understood outside of respiratory medicine. Children should have immediate access on a day-to-day basis to respiratory nurse specialists, respiratory physiotherapy and the respiratory consultant and NCHDs.

- *Long-term ventilation (LTV)*

The optimal patient journey for children requiring long-term ventilation (LTV) is detailed here.

Typically, children who require LTV are emergency transfers from peripheral centres, as neonates or in early infancy, to a tertiary neonatal or paediatric centre. In the majority of cases, they are initially unstable, with high mortality and morbidity associated with their underlying condition and requiring intensive care treatment, including invasive ventilation. If, over a period of time, their condition stabilises, the need for medium to long-term ventilation becomes apparent, and a tracheostomy tube is inserted as a conduit to providing this. Shortly thereafter, it may be possible to transfer the child to the TCU, where discharge planning can commence while intensive care is provided in a less critical setting. The focus at this stage is very much on "transitioning" the

child and family to their home. This involves a process of putting in place services, equipment and staffing as part of a HCP for the family. When the HCP is in place – and barring exceptional circumstances – patients should make the final “step down” with a transfer to the regional paediatric centre for a short period, prior to final discharge home.

Physiotherapists are heavily involved with LTV patients both in the ICU and TCU setting.

The physiotherapist will liaise closely with community therapy services to ensure that physiotherapy input will continue on discharge. Such arrangements are often put in place in conjunction with other health professionals, e.g. speech and language therapists. The physiotherapist also provides education to parents regarding ongoing respiratory physiotherapy if it is required on discharge home. Currently, there is no local outreach paediatric respiratory physiotherapy service available to these children (and which could link easily with the children’s local paediatrician). Consideration should be given to developing this service, as it could address a significant unmet need.

Children on LTV require a complex care package as they journey from the hospital inpatient setting to the community. At each stage, the correct support structures, provided by a designated multidisciplinary team, need to be in place in order for the child to always be cared for in a safe and stable environment. Once the child is medically stable, it is inappropriate to care for them in a high-level intensive care unit. In order to promote normal growth and development, children on LTV are much better off in their home environment. A structured pathway is essential in order to ensure that discharge to their home environment is facilitated as soon as the child is deemed medically stable for such discharge.

From the outset, once it is decided that a patient is suitable for tracheostomy and LTV, a key worker, ideally a nurse specialist for LTV, should be identified to work with and support LTV patients and their families, as well as carers and health and social care professionals in the hospital and in the community. Discussions with the community care and local paediatric hospital services should take place, so as to involve these services in early discharge planning, and also to nominate a key worker from both services. Once the child is medically stable, a home care package should be requested; this should be based on the child’s needs and the child should be transferred to a transitional care unit, where more of their development and growth needs can be met.

The decision on the level of care required from the home care package lies with the respiratory consultant, LTV clinical nurse specialist and multidisciplinary team. While awaiting approval of a HCP, the parents should undergo intensive training in the care involved with tracheostomy and LTV. This training should be coordinated by both the airway and LTV nurse specialists in the tertiary hospital. Throughout this process, a designated respiratory consultant should oversee the management of the child’s ventilation requirements, and other members of the multidisciplinary team should ensure that the child’s developmental and nutritional needs are met. For children with complex, multi-system disorders, a general paediatrician should coordinate the patient’s overall care. This process should take approximately eight weeks.

Once the child is medically stable, a home care package has been approved, and the parents are fully competent in all aspects of care associated with tracheostomy and LTV, the community services key worker should be notified and the child should be discharged home. In certain circumstances it may be prudent to discharge a child on LTV to their peripheral hospital for a short period prior to moving to their home environment. In this case, the key worker from the peripheral hospital (preferably a respiratory nurse specialist) should assist

in coordinating the transition process. Once home, the support staff should comprise a mix of both nurses and healthcare assistants (HCAs) fully trained in managing a child with a tracheostomy and who is on LTV. Staffing should be appropriate for the level of care that the child requires. (See Appendix 1 for proposed LTV discharge pathway.)

- Primary ciliary dyskinesia (PCD)

In general, children with PCD present either in the neonatal period or in early childhood. The most common neonatal presentation is of persistent tachypnoea and oxygen requirement. Other children may have a diagnosis established by the discovery of dextrocardia or situs inversus in the neonatal period. These children are usually referred directly to a consultant in paediatric respiratory medicine. Children who do not present in this manner usually only get diagnosed with PCD if they manifest with a marked increase in the frequency of chronic nasal, ear and respiratory symptoms. Children presenting in this way usually have a delayed diagnosis because of the commonness of recurrent upper respiratory infection in the general population. A greater awareness of PCD among GPs and paediatricians would improve this situation. Indeed, GPs and paediatricians should actively consider the possibility of a PCD diagnosis, and any child with recurrent nasal and respiratory symptoms should be examined with the possibility of a PCD diagnosis in mind, particularly if recurrent ear infections are an issue. Children with this pattern of presentation should be referred to a consultant in paediatric respiratory medicine.

In the future, it is recommended that a national specialist centre be established at the new children's hospital. In the greater Dublin area, children with PCD should attend the national specialist centre on a regular basis. Children living in the catchment area of regional specialist centres should attend those centres, and they should also attend the national centre on an annual basis for review. Shared care arrangements should be established between the national centre and the regional centres.

Children with PCD have variable access (usually poor) to respiratory physiotherapy, and have no access to PCD nurse specialists. Children have ad hoc access to an inpatient treatment service, depending on where they live. Given the lack of a designated service for children with PCD, and also given the very poor awareness of PCD among general paediatricians, it is likely that there are many undiagnosed children with PCD in the community. The establishment of a designated national service, coupled with the appointment of a nurse specialist in PCD, is of vital importance both in terms of providing education and awareness of PCD and also in terms of improving access to care for children who have PCD. Based on the fact that there are between 80 and 100 children with PCD in Ireland at any one time, it is estimated that one WTE physiotherapists for patients with PCD would be required in order to provide a national service. Children with PCD should have direct access to a multidisciplinary team comprising a clinical nurse specialist, respiratory physiotherapist, and social worker. Via this multidisciplinary team network, the child and their family should have access to a consultant in paediatric respiratory medicine and an ENT consultant. Children with PCD will require intravenous antibiotics and should have ready access to an inpatient hospital bed and a PICC line service.

- Severe asthma

Currently, children with severe asthma usually attend a consultant in paediatric respiratory medicine. The quality of, and access to, this service vary throughout Ireland and are based on the limited availability of medical and nurse specialist staffing in different units. All tertiary and regional units should have full-time staff, which should include respiratory nurse specialists. Specialist asthma nurses should be in place in the tertiary centres and, ultimately, in the new children's hospital. In the regional centres, some overlap is

expected in the nurse specialist role between the conditions of asthma, sleep disorders and long-term non-invasive ventilation. All children with difficult-to-treat asthma should have immediate access to an asthma nurse specialist locally. Children with severe asthma who are referred to the severe asthma clinic should have immediate access to a nurse specialist who is responsible for the clinic, and is also responsible for ensuring urgent access for the patient, if required, to a consultant in paediatric respiratory medicine. Children with severe asthma requiring review at the National Severe Asthma Clinic will have initially been seen by a specialist in paediatric respiratory medicine. These children should be referred directly to the severe asthma clinic. It is not anticipated that children will be referred from a secondary care general paediatrician directly to the severe asthma clinic.

- General respiratory problems

Children may be referred to tertiary respiratory subspecialists from a number of sources:

- Other tertiary subspecialists
- Secondary care consultant paediatricians
- Emergency department.

As is the case with many other paediatric subspecialties, the experience in paediatric respiratory medicine is that many GPs refer a high volume of non-complex cases directly to subspecialists rather than to general paediatricians. (This is partly due to historic practice and partly due to the fact that there are so few general paediatricians.) Such practices are evident in tertiary units in the Dublin paediatric hospitals. This is an inefficient system, which reduces access and increases waiting times for children who have already been seen and assessed by a paediatrician, and require urgent assessment by a subspecialist. GPs should not refer children directly to tertiary paediatric respiratory subspecialists for evaluation of suspected asthma or for recurrent respiratory infection. These referrals should be directed to general paediatricians who would be in a position to screen and manage straightforward cases and refer on cases requiring subspecialty opinion.

Waiting times for tertiary subspecialty respiratory opinion vary from immediate to more than one year. Children with respiratory problems requiring tertiary subspecialty review should, depending on the severity/acuity of the problem, be seen within three months of referral. Children with general respiratory problems should have immediate direct access from the respiratory clinic to the appropriate clinical nurse specialist, respiratory physiotherapy and pulmonary function testing, and diagnostic sleep testing as an outpatient, within three months or sooner.

42.4 REQUIREMENTS FOR SUCCESSFUL IMPLEMENTATION OF MODEL OF CARE

Infrastructure

Regional centres

The major deficiency in regional centres, in terms of provision of adequate respiratory services for both children with CF and other respiratory conditions, is the lack of any significant paediatric pulmonary function laboratory capacity. In order to provide a satisfactory service for subspecialty respiratory patients, a paediatric pulmonary function laboratory is an absolute necessity.

	Cork	Limerick	Galway
CF inpatient facilities	Inpatient facilities inadequate – no en-suites, no isolation rooms. Suboptimal for infection control	Inpatient facilities adequate	Inpatient facilities adequate with en-suite rooms available
Plans for development	Application for funding for new inpatient unit, including isolation space for children with CF, in progress	None	Standalone outpatient department (OPD) building nearing completion for day cases/clinics/reviews/procedures
CF outpatient facilities	Inadequate currently. Takes place in general outpatient unit shared with adult clinics (non-CF), no en-suite rooms. Inadequate infection control.		
Poor access to day beds.	Outpatient facilities adequate	As above	
Plans for development	Limited CF day unit/ outpatient unit currently in planning. Funding from local CF charity Build4Life – not HSE funded. Four single en-suite rooms with office space for MDT and gym required. Four rooms with only two en-suites with no MDT office space planned. Restricted by limited space and funding available.	None	No further plans
Clinical space needed (to run full regional service)	Respiratory pulmonary function laboratory. Four day unit isolation CF rooms. Four inpatient en-suite isolation CF rooms. Two treatment rooms for day unit reviews of non-CF patients – sleep, asthma, TB, non-CF bronchiectasis, PCD	Respiratory laboratory, dedicated inpatient sleep beds. Three treatment rooms.	Respiratory laboratory
Office space needed (for respiratory nurse, scientist, physiotherapist, dietitian, SLT)	One large office for CF MDT. Three CF nurse specialists (not in post). CF physiotherapist. CF dietitian. One other office for asthma nurse specialist and respiratory nurse specialist (not in post)	Office space needed for respiratory nurse(s), respiratory physiotherapist, speech and language therapist, respiratory scientist, psychologist	Office for respiratory nurse and scientist

New Children’s Hospital

As the planning of the new children’s hospital on the St James’s Hospital site is already underway, significant infrastructural change in the three existing paediatric hospitals would not seem justified at this time.

Ambulatory services for patients with CF are reasonably well catered for in each of the three centres in Dublin. From an infrastructural point of view, the sleep service in Crumlin is specifically designed to cater for significant increase in demand as it arises. However, increased demand would require additional staffing resources. In both Temple Street and Tallaght, with the exception of providing inpatient beds, facilities for sleep diagnostics do not exist. Prior to the establishment of the new children’s hospital, the service across the three paediatric hospitals will concentrate on maximising the utility of the day-case beds provided for sleep diagnostics at Crumlin.

Staffing

National and international recommendations

With regard to caring for children with CF, clear staffing recommendations have already been agreed, both from an Irish perspective (HSE CF Report, 2009) and from a UK perspective (UK CF trust). The RCPCH report on sleep disorders in children provides some guidance in relation to suggested staffing numbers per region, but this guidance is not as specific as that contained in the CF reports. Apart from these documents, there is little by way of structured recommendations in relation to staffing provision within the specialty. Table 42.2 was published in the HSE report on CF services in 2009:

Staff member	Paediatric	Adult
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	
Dietitian	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	

Table 42.2: Recommended staffing levels for specialist CF centres per 50 CF patients

Some modernisation of the infrastructure for the provision of care to children with CF is anticipated. This will form the remit of the National Clinical Programme for Cystic Fibrosis. It is expected that shared care and referral pathways will be consolidated, in order to reflect the emergence of six specialist CF centres, as outlined in the national newborn screening strategy. A quick review of data from the CF Registry of Ireland would suggest the following total patient numbers in the future: New Children’s Hospital – 300; Cork – 110; Limerick – 90; Galway – 70.

Consultants

The most urgent requirement for specialists in paediatric respiratory medicine, in terms of consultant workload, is to be able to concentrate on core subspecialist work and have the burden of general paediatric, as well as primary and secondary respiratory care, eliminated. Respiratory consultants should be on a respiratory rota, and not a general paediatric rota. This inappropriate overloading with non-specialist clinical work is currently the case both in tertiary centres in Dublin (where specialists in paediatric respiratory medicine are seeing primary and secondary care respiratory problems in local children) and also in regional centres (where specialists must provide a high volume of general paediatric and/or neonatology cover). The alleviation of this workload is the priority for the subspecialty in terms of consultant staff. A consultant in paediatric sleep medicine needs to be appointed as a matter of urgency. The rapidly growing area of paediatric respiratory medicine, which is still a relatively new specialty, encompasses conditions with a very high level of prevalence and complexity. A consultant in paediatric sleep medicine would be responsible for the guidance of the sleep service nationally, coupled with maintenance of clinical standards, leadership in the area of sleep medicine, and maintenance of quality and effectiveness from a technical point of view in diagnostic sleep services. The sleep service at the New Children's Hospital will require a number of consultants whose primary interest is in paediatric sleep medicine.

The new children's hospital will care for approximately 300 CF patients. Based on the HSE report into CF services (2009), caring for this number of patients will require six WTEs, i.e. four 'consultant 1' and two 'consultant 2' consultants for patients with CF alone. Apart from guidance on the number of consultants that Ireland needs in order to care for CF patients, specific international or national guidance on consultant numbers is not available. Cork is appropriately staffed from a consultant point of view, with 2.0 WTEs, which equates well with regard to the current and expected CF patient numbers in the future.

In order for the sleep service to function effectively and develop in Cork, an additional 0.5 WTE consultant post is required. The key requirement here is alleviation of general paediatric and secondary care work currently being provided in addition to the subspecialty component. The centre in Limerick remains understaffed from a consultant point of view. The HSE report would suggest that the requirement is for 1.8 WTE consultants for CF care. There is currently one full-time consultant in place, and some provision of WTE (approximately 0.2) by a secondary consultant. The number of consultant staff in Limerick needs to be increased, with the addition of a second consultant with specific training in paediatric respiratory medicine and CF committing 80% of their time to respiratory medicine and 20% of their time to general paediatrics. Of key importance in Limerick will be the alleviation of the respiratory team's general paediatrics and neonatology workload.

Ultimately, in Galway there will be a requirement for 1.4 WTE consultants. The geographical area covered by the Saolta Hospital Group is very large. Travel distances from some areas within the Saolta Hospital Group catchment area can exceed the travel distance from place of origin to Dublin. It is likely that a substantial outreach or shared service will be required. This service could be located in either Sligo or Castlebar, with Sligo seeming more sensible, given the distance from Galway to Sligo. A shared consultant appointment between Galway and the new centre (50% respiratory, 50% general paediatrics) will be required. This appointee should have training in paediatric respiratory medicine and CF, and concentrate on forming a strong link between the two centres, coordinating shared care and outreach. The priority in the short term will involve removal of secondary care, neonatology and general paediatrics from the respiratory consultant remit in Galway.

With the very significant growth in numbers in the area of LTV, NIV and sleep, this area of practice has now taken over from CF in terms of the greatest time commitment within consultants' individual practices. Between 2007

and 2011 in children's units across Ireland, consultants have seen a more than threefold increase in the number of sleep studies performed and the number of children on NIV. Currently, the number of children on NIV, divided between the three paediatric hospitals, totals more than 300. Given the very rapid increase in numbers in the past few years, it is likely that this will more than double prior to the commencement of services at the New Children's Hospital.

A review of current clinical practice in Crumlin and Temple Street (which, combined, encompass all the subspecialty services that will be provided in the new children's hospital) gives an approximate breakdown of the workload of the respiratory service as follows:

- Cystic fibrosis 30%
- Sleep 25%
- Neuromuscular 15%
- General respiratory 15%
- Long-term ventilation 10%
- Other disorders 5%

Internationally, the management of children on long-term mechanical ventilation is increasingly becoming the role of respiratory physicians, as opposed to intensive care doctors. It is envisaged that, in the new children's hospital, the transitional care unit will form part of the Department of Respiratory Medicine. Management of this complex group as inpatients, and also as outpatients, is time-consuming from both a clinical and administrative point of view, and will involve an additional workload in the future. A modern Department of Respiratory Medicine in the New Children's Hospital will require at least 12 WTE consultants, of whom two should hold academic posts.

Non-consultant hospital doctors (NCHDs)

The following areas of practice in the new children's hospital will require NCHD staffing:

- CF: inpatients, drop-ins, outpatient clinics Three WTEs
- Inpatient service Three WTEs
- Consultations 1.5 WTEs
- Outpatient clinics, non-CF drop-ins Five WTEs
- Transitional care unit One WTE
- Day case: bronchoscopy, nasal brushings, sedation/GA scans 0.5 WTE

The HSE report into CF services recommends that for each 50 patients with CF, 0.5 WTE CF-specific specialist registrars (SpRs) are required. For the new children's hospital, therefore, purely in terms of CF, three WTE SpRs are required. On the basis of the specific requirement of subspecialty workload – both in the inpatient and outpatient setting – the component represented by CF, the overall current number of NCHDs currently in the system, and an appropriate NCHD to consultant ratio, the number of NCHD staff required for the new children's hospital would be:

- SpR Six WTEs
- Registrar Two WTEs
- SHO Six WTEs

Each regional specialist unit, if it is to operate as such, will require dedicated respiratory NCHD staff rather than sharing a pool of general NCHD staff. Familiarity with the specialty and consistency are key here if the unit is to operate satisfactorily. Based on putative projected numbers of CF patients and further regional specialisation of

centres, one might expect the following patient distribution: Cork – 110; Limerick – 90; Galway – 70. With this in mind, in terms of CF-specific NCHDs, Cork and Limerick would require one full-time CF SpR, and Galway would require a respiratory SpR covering all aspects of respiratory medicine including CF. The overall number of NCHDs required in the three units is Cork – 3.0 WTEs; Limerick – 2.5 WTEs; Galway – 2.0 WTEs.

Nationally, therefore, the following number of NCHDs in respiratory medicine are required.

- SpR 9 WTEs
- Registrar 3 WTEs
- SHO 9.5 WTEs

Clinical nurse specialists

Outside of the area of CF, which has a well-established staffing requirement, as laid down by international bodies including the HSE, staffing for nurse specialists in paediatric respiratory medicine is poor. There are no non-CF nurse specialists in Crumlin; there is one in Temple Street and there are two in Tallaght. Currently, these nurse specialists are seeing a combination of patients with asthma, allergy, NMD and NIV.

Asthma services within the Department of Respiratory Medicine at the New Children's Hospital are likely to concentrate on difficult-to-control asthma, and the majority of children with asthma will be cared for by general paediatricians. Given the prevalence of asthma, a number of asthma nurse specialists will be required by the new children's hospital. Access to asthma nurse specialist services should be available to all outpatients with asthma who are attending the hospital; access should also be available to inpatients admitted with asthma exacerbations, regardless of which medical team is caring for these patients. The time commitment of asthma nurses must be split between the difficult-to-control asthma service based in the Department of Respiratory Medicine, and the majority of asthma patients in the hospital who are being cared for by general paediatricians.

Apart from caring for asthma and CF patients, respiratory nurse specialists' main areas of practice will include:

- long-term mechanical ventilation
- long-term nocturnal non-invasive ventilation
- sleep medicine
- thoracic insufficiency
- respiratory complications associated with neuromuscular disease.

Each of these areas of practice is large, with distinct differences in terms of approach and practice. There is, however, a common thread between them, as well as a degree of natural overlap. In terms of staffing plasticity, cover for leave, education and training it would seem logical to have cross-cover between the specialists in these different fields. Respiratory nurse specialists from the regional centres will deal with some aspects of each of these areas. Therefore, it will be of benefit for respiratory specialists at the New Children's Hospital to have a similar level of exposure across all of these areas. It is envisaged that certain nurse specialists will specialise in each of these areas, but will also have a good basic understanding of all areas of practice. Based on the very significant numbers of patients in each of these areas of practice, seven full-time non-CF respiratory nurse specialists (and one or more advanced nurse practitioners (ANPs)) will be required (four for asthma; one for long-term mechanical ventilation; two for long-term nocturnal non-invasive ventilation and sleep; one for thoracic insufficiency and NMD).

Advanced nurse practitioners (ANPs)

One CF nurse specialist in Tallaght is currently enrolled in an ANP training programme. Another respiratory nurse in Tallaght has trained, and is now operating, as an ANP. ANPs would be very valuable for the service in the areas of CF, LTV, NMD, sleep and severe asthma. Depending on the clinical service requirement and institutional nursing policy in each of the centres, clinical nurse specialist posts could be replaced with ANP posts.

Physiotherapy

Physiotherapy is central to many of the different subspecialty areas of paediatric respiratory medicine. Respiratory physiotherapy input is essential for CF, NMD, long-term mechanical ventilation, and general respiratory medicine. Within respiratory medicine, CF accounts for physiotherapists' core workload. However, a large volume of patients are also referred for respiratory physiotherapy by other teams, as well as the post-operative patients. For example, 47.6% of all PICU admissions in Crumlin from 2011 to 2013 were categorised as cardiac/cardiothoracic (see Crumlin, Paediatric Intensive Care Unit, 4th Annual Report). Given the very significant requirement for a physiotherapy input in caring for CF patients, as outlined in the HSE report, each centre should have a number of physiotherapists dedicated to respiratory medicine. Given the volume of non-CF respiratory issues, the intermittent nature of patient flow and the mix of inpatient and outpatient workload, and also given the overlap between CF and non-CF respiratory physiotherapy, cross-cover between the areas within the recommended number of therapists for CF is envisaged – particularly in regional centres.

A dedicated paediatric physiotherapist in each paediatric unit – with 0.5 dedicated to outreach – would bridge the significant gap in local service provision for this vulnerable group of children and families. Moreover, this physiotherapist would provide support, education and local respiratory management as dictated by a defined integrated care pathway with the relevant regional or tertiary centre paediatric respiratory physiotherapist. This is one of the most significant challenges faced by the physiotherapy profession to date, as the numbers of children in the community with complex respiratory conditions continues to rise. The increase in the numbers of such children puts further pressure on the tertiary respiratory physiotherapists who try to provide support and education to local physiotherapy hospital and disability staff. Access to local paediatric respiratory physiotherapy is critical in order to facilitate discharge, and also in order prevent re-admission with an acute episode, in so far as possible, by providing a continuum of respiratory physiotherapy support as part of the patient's package of care.

Figure 42.3 provides an overview of respiratory physiotherapy requirements.

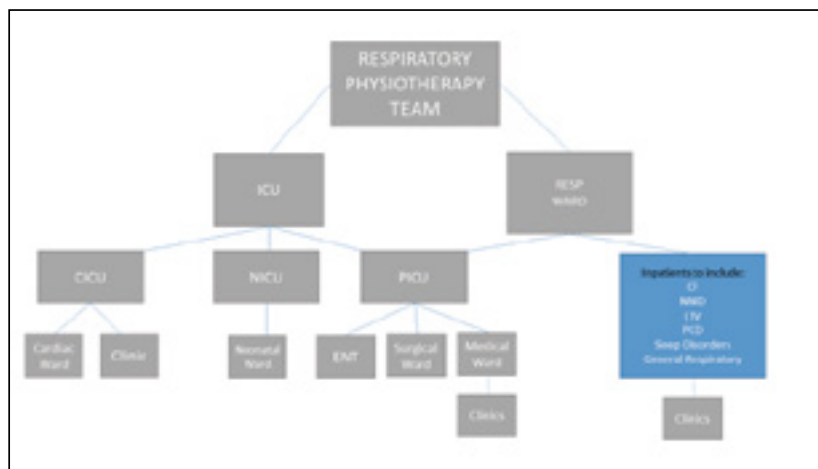


Figure 42.3 Overview of range of physiotherapy respiratory services

Other health and social care professionals

The HSE report into CF services provides very clear recommendations on staffing levels for a range of health and social care professionals. Some, such as physiotherapy, psychology, social work, and respiratory scientists, which are listed here, are required both for CF and for non-CF respiratory areas. Numbers in the recommended total national staffing component reflect this. For areas such as nutrition and dietetics, pharmacy and medical scientists, non-CF respiratory needs are in line with those of all patients in the hospital, and no structured allocation to respiratory services is required over and above this. This is contingent on a clear plan of clinical governance for children with NMD. The lead team supervising their care needs to be well resourced in terms of relevant health and social care professionals.

Speech and language therapists are of increasing importance to the provision of respiratory services. Pulmonary aspiration occurring as a result of disordered swallowing is very common in paediatric respiratory practice, significantly more common than previously suspected. Swallowing disorders are common in children with neurodevelopmental delay, neuromuscular disorders, neurosurgical patients, trisomy 21, craniofacial disorders and many other children with complex medical needs. The volume of children with these types of issues far exceeds the potential capacity of specialist centres to address them. Full-time speech and language therapy service provision, including the provision of video fluoroscopy, should be available in each of the specialist and regional centres.

Table 42.3 outlines the current and proposed position in relation to CF alone. All proposed numbers in this table are based on the HSE report on CF services 2009:

Staff category	Current (WTE)	Proposed (WTE)
Consultant	11 (8.0 respiratory)	11 (purely CF)
Specialist registrar	4.5 (3.2 respiratory)	5.5 (purely CF)
Registrar	3 (2.5 respiratory)	0
SHO	5 (4.0 respiratory)	0
Specialty trained nurses	0	0
Advanced nurse practitioner	0	0-3
CF clinical nurse specialist	12.5	16.5
Dietitian	3.5	4.5
Physiotherapist	6.5	22.8
Pharmacist	1.2	3.3
Occupational therapist	0	0
Psychologist	1.7	4.5
Social worker	2.2	4.5
Data clerk	0	1.1
Respiratory scientist (PFTs)	6.5	7.7
Medical scientist	*	7.7
Secretary/Administrator	5	11

*Data not available

Incorporating the CF and non-CF figures, Table 42.4 outlines the national position for respiratory medicine:

Staff category	Current (WTE)	Proposed (WTE)
Consultant	11** (8.0)	18.5
Specialist registrar	4.5** (3.2)	9
Registrar	3** (2.5)	3
SHO	5** (4.0)	10
Specialty trained nurses	0	0
Advanced nurse practitioner	1	1-3 (taken from CNS numbers)
Clinical nurse specialist	12.5CF*/2.6Non-CF	28 (17CF*/10 Resp)
Dietician	3.5	4.5
Physiotherapist	6.5	37.8
Pharmacist	1.2	3.3
Occupational therapist	0	1
Psychologist	1.7	6 (5 CF/1 sleep)
Social worker	2.2	6 (5 CF/1 resp)
Data clerk	0	2 (1.1 CF/0.9resp)
Respiratory scientist	6.5 (2 sleep/4.5 PFT)	15.5 (8Sleep/7.5PFT)
Medical scientist	***	7.7
Secretary/Administrator	5	11.5

* CF numbers based on a national population of 550 children with CF.

** Currently, medical workload for respiratory teams in Dublin hospitals comprises approximately 20% general paediatrics and 80% respiratory. In regional centres, these figures are approximately 30% general and 70% respiratory. (WTE here indicates the time committed to respiratory medicine.) Posts proposed should be 100% respiratory.

*** Data not available

Table 42.5 outlines the proposed staffing based on service need. Numbers refer to dedicated respiratory staff, except where specifically indicated otherwise.

Staff category	New children's hospital	Cork	Limerick	Galway
Consultant	12	2.5	2 (0.2 shared)	2 (0.5 shared)
Specialist registrar	4.5	2	1	1
Registrar	3	0	0	0
SHO	6	1	2	1
Specialty trained nurses	0	0	0	0
Advanced nurse practitioner	1-3(v. CNS)	0	0	0
Clinical nurse specialist	9 CF/7 Resp	3.5 CF/1 Resp	2.5 CF/1 Resp	2 CF/1 Resp
Dietitian	2.3	1.0	0.7	0.5
Physiotherapist	24	5.4	4.6	3.8
Pharmacist	1.7	0.7	0.5	0.4

Occupational therapist	1	0	0	0
Psychologist	3.5 (2.5 CF)	1	1	0.5
Social worker	3.5 (2.5 CF)	1	1	0.5
Data clerk	1	0.4	0.3	0.2
Respiratory scientist	12 (8 sleep)	2	1.5	1
Medical scientist	4.1	1.6	1.1	1
Secretary/Administrator	7	2	1.5	1

Education, Training and Continuous Professional Development

An absolute prerequisite for the establishment of a national network is a strong education/training/continuous professional development agenda. To some degree, this is already present in terms of the structure around the provision of CF care in specialist centres. A national CF meeting is held on an annual basis, and this meeting has proved to be extremely successful in terms of the interaction of the teams in different centres across all disciplines. The Irish Thoracic Society meeting, which is held annually, has a paediatric subgroup which provides an effective education and research outlet, mainly for physicians, and to a lesser degree for nurses and health and social care professionals (HSCPs). Given the limited number of nurses and HSCPs outside of CF in the network currently, little progress is possible in this regard.

The plan to ensure regional distribution of expertise, outside of the CF field there will be respiratory scientists and respiratory nurse specialists in all centres. In addition, there is considerable potential to broaden the scope of the education and training agenda of the paediatric subgroup of the Irish Thoracic Society. Such a move would provide an ideal opportunity for respiratory scientists, respiratory physiotherapists and respiratory nurse specialists to focus on the issue of education, training and continuous professional development (CPD).

One of the key drivers of the urgent need to increase the numbers of respiratory scientists, respiratory physiotherapists and nurse specialists across the country is the necessity to incorporate education, training and CPD into current day-to-day practice. Currently, the level of service requirement is overwhelming, stifling the ability of staff members to conduct audits, develop services, improve quality and focus on education and training.

One of the newer areas of paediatric respiratory medicine where significant change has occurred in the past five years is the area of sleep medicine. With this in mind, a multidisciplinary study day was recently held revolving around the diagnosis and management of children with sleep disorders. This is the first of what will be an annual education and training day in relation to sleep medicine. A second annual meeting is proposed (in order to have an education/training meeting every six months) revolving around long-term invasive and non-invasive ventilation. In the future, a schedule as follows is envisaged which will maximise the education/training component of paediatric respiratory medicine in a meaningful way without unnecessarily diluting the content or producing meeting fatigue:

- | |
|--|
| <p>The national CF meeting</p> <ul style="list-style-type: none"> o Medical o Nursing o Allied health professionals |
|--|

The Irish Thoracic Society Meeting
o Medical
o Nursing
o Respiratory physiotherapy
o Respiratory dietitians
o Respiratory scientists
An annual study day for paediatric sleep medicine
o Medical
o Nursing
o Respiratory scientists
An annual study day for long-term ventilation
o Medical
o Nursing
o Respiratory physiotherapy
o Respiratory scientists

The generation of new national clinical guidelines, protocols and standards, in the context of a national network of paediatric respiratory medicine, will be a multidisciplinary process and will involve staff from all centres together.

Governance

The collective goal is to establish a high-quality national collaborative service for children with respiratory disorders. A key aspect of this national collaborative approach is a strong working relationship, both vertically and horizontally, between professionals in the network. Services are organised locally, regionally and nationally. Paediatric services at a local, regional and national level have complex governance structures in place; these structures govern clinical practice and management. National regional and local services in paediatric respiratory medicine come under the overall governance structures in place in the relevant institutions.

From a medical point of view, all consultants in paediatric respiratory medicine are members of the Irish Thoracic Society, a professional body. The Irish Thoracic Society has a paediatric subgroup with a permanent chair in place. The chair of the paediatric subgroup of the Irish Thoracic Society is responsible for representation on behalf of consultants in paediatric respiratory medicine; for distribution of relevant information from the professional body to its members; for the organisation and management of annual scientific meetings; it also acts as an important liaison between paediatric and adult respiratory groups. The chair is an important role in the promotion of research and education relating to paediatric respiratory medicine. The chair will drive the educational agenda of the national group of practitioners in paediatric respiratory medicine and will act as a link between the new children's hospital and the regional centres. The chair will be responsible for fostering development of national guidelines and ensuring that centres work as closely as possible.

In 2014, as part of the rearrangement of the clinical directorate of paediatrics in Dublin, and the amalgamation of three children's hospitals, subspecialty leads across the three children's hospitals were appointed. The subspecialty leads are charged with supporting high quality healthcare across the specialty and across hospitals ensuring continuous quality improvement, development of key performance indices, audit and implementation of standardised clinical guidelines. The Department of Respiratory Medicine in the new children's hospital will have an administrative head who will operate as head of department under the directorate of which the department is part. The administrative head will be responsible for clinical leadership within the department, administration, co-ordination of departmental staff and financial aspects of the running of the department.

Standards of Care

Documents outlining standards of care for children with respiratory and sleep problems are available in a number of the above areas. Most standards of care documents are published by UK, European and US bodies. Further details on policies, procedures and guidelines are detailed in Appendix 2.

CF

As outlined previously, the most recent document detailing standards of care for children with CF was published in the UK. The National Clinical Programme for Cystic Fibrosis in Ireland is charged with publishing similar standards of care for Irish children; these standards of care are likely to be similar to those contained in the UK document.

Sleep

In 2009, the Royal College of Paediatrics and Child Health published standards for services for children with disorders of sleep physiology. This document was developed by the working party on sleep physiology and respiratory control disorders in childhood. In June 2012, the Australasian Sleep Association published Standards for Sleep Disorders Services, which addresses both adult and paediatric sleep requirements.

A national service plan and model of care for paediatric sleep medicine in Ireland has recently been drafted and submitted to the Paediatric Integration Group.

Respiratory management of neuromuscular disease (NMD)

Standards and clinical guidelines for the respiratory management of children with neuromuscular disease (NMD) have been produced by the British Thoracic Society in 2012. See: Respiratory management of children with neuromuscular weakness.

Long-term ventilation (LTV)

The following key documents outline best practice and clinical guidelines in the area of long-term ventilation:

- Core guidelines for the discharge home of the child on long-term assisted ventilation in the United Kingdom: Jardine E, Wallis C. Thorax 1998
- Breathing Space: Community Support for Children on Long-term Ventilation Ludvigsen and Morrison 2003. Barnardos
- From Hospital to Home: Guidance on discharge management and community support for children using long-term ventilation. Noyes and Lewis 2005. Barnardos
- The Australasian paediatric respiratory group has produced a comprehensive document relating to children on invasive and non-invasive ventilatory support at home: Ventilatory Support at Home for Children – A Consensus Statement from the Australasian Paediatric Respiratory Group February 2008

Severe asthma

The National Clinical Programme for Asthma has produced recommendations on standards of care for children with asthma.

Primary ciliary dyskinesia (PCD)

Standards of care for children with PCD in the UK were published in 1998. These recommendations remain largely unchanged. This document outlines the requirement for all children with PCD to attend a specialist centre and to have regular contact from a nurse specialist and direct and frequent access to respiratory physiotherapy

services. The European Respiratory Society produced a comprehensive document on the diagnosis and treatment of children with PCD in 2009. This provides up-to-date guidance on appropriate investigations and treatment of PCD.

42.5 PROGRAMME METRICS AND EVALUATION

Cystic fibrosis (CF)

Extensive details on metrics and evaluation have been published in the UK and Australia in particular. These metrics will be introduced in conjunction with the introduction of the other elements of the National Clinical Programme for Cystic Fibrosis in Ireland. There is much international agreement on key performance indicators for CF centres and metrics for delivery of effective CF care. The National Clinical Programme for Cystic Fibrosis will ensure that sufficient staff and physical resources are in place to enable delivery of programme metrics and ongoing programme evaluation.

The key service metrics outlined in the UK CF report are as follows:

General

- Is there a multidisciplinary team of trained and experienced CF specialist health professionals in the specialist centre and other staffing levels appropriate for the clinic size?
- Do all patients have a named consultant, and how often are the patients seen by them?
- What is the median lung function of the whole clinic and patients at transition to adult services?
- What proportion of patients have had an annual review in the last year?
- Is there a formal pathway in place for transition to adult care?
- Is there access to a paediatric palliative care service?
- Is there evidence that the staff maintain their CPD relative to CF?

Microbiology

- What proportion of patients are infected with pseudomonas aeruginosa, burkholderia cepacia complex, MRSA and what is the annual rate of new acquisition of these organisms?
- What arrangements are in place to minimise the risk of cross-infection in clinics and inpatient facilities?
- Is there evidence of cross-infection in the unit?
- What is the process for checking the result of clinical respiratory microbiology samples?
- What proportion of first isolates of pseudomonas aeruginosa received antimicrobial therapy?
- What is the mean duration between isolation of first (or first after a prolonged period of absence) pseudomonas aeruginosa and initiation of treatment?
- What proportion of patients with chronic pseudomonas aeruginosa infection are on long-term inhaled antibiotics?
- Are tobramycin levels available on inpatients and those on home IV antibiotics within 24 hours?
- Does the microbiology laboratory at the specialist CF centre comply with international criteria for processing CF specimens?

Newborn screening

- What proportion of children in the newborn screening programme receive a diagnosis of cystic fibrosis before the age of four weeks?
- What proportion of sweat tests in children aged 3-4 weeks failed for technical reasons?
- How many sweat tests were performed in the centre in the last year?
- What proportion of siblings of newly diagnosed patients with cystic fibrosis have had a sweat test performed?
- What proportion of patients diagnosed through newborn screening were invited to enrol in the CF registry?

Exacerbations

- What was the median waiting time for admission for a chest exacerbation?
- How often are inpatients seen by a physiotherapist?
- What proportion of patients receive home IV antibiotics?
- Have all patients/carers administering home IV antibiotics undergone competency assessment?

Clinics

- What was the average number of clinic visits for patients attending the centre per year?
- What arrangements are in place for segregation in the clinic?
- Are all MDT members present at every clinic?

Sleep

Internationally, there is little information available on which to base metrics and prospective evaluation for a national paediatric sleep service. On the basis of proposals for a new national paediatric sleep service, the following metrics will be useful for prospective evaluation of the suitability of the service delivered:

Regional specialist centres

- Is domiciliary overnight oximetry routinely available?
- What is the waiting time for domiciliary overnight oximetry?
- Have a representative sample of overnight oximetry studies had inter-scorer and inter-reporter reliability performed in the last year?
- Is overnight oximetry/capnography routinely available?
- What is the waiting time for overnight oximetry/capnography?
- Have a representative sample of overnight oximetry/capnography studies had inter-scorer and inter-reporter reliability performed in the last year?
- Is overnight cardiorespiratory polysomnography routinely available?
- What is the waiting time for cardiorespiratory polysomnography?
- Have a representative sample of cardiorespiratory polysomnography studies had inter-scorer and inter-reporter reliability performed in the last year?
- Is a respiratory nurse specialist/respiratory scientist available within 24 hours during the week and 48 hours at the weekend for urgent issues for children on non-invasive ventilation?

Supra-regional centres

- What is the waiting time for domiciliary overnight oximetry?
- Have a representative sample of overnight oximetry studies had inter-scorer and inter-reporter reliability performed in the last year (with a member of the UK paediatric sleep group)?

- What is the waiting time for overnight oximetry/capnography?
- Have a representative sample of overnight oximetry/capnography studies had inter-scorer and inter-reporter reliability performed in the last year (with a member of the UK paediatric sleep group)?
- What is the waiting time for cardiorespiratory polysomnography?
- Have a representative sample of cardiorespiratory polysomnography studies had inter-scorer and inter-reporter reliability performed in the last year (with a member of the UK paediatric sleep group)?
- What is the waiting time for full polysomnography?
- Have a representative sample of full polysomnography studies had inter-scorer and inter-reporter reliability performed in the last year (with a member of the UK paediatric sleep group)?
- What proportion of full polysomnography studies are directly observed overnight by a respiratory scientist?
- Is a respiratory nurse specialist available within 24 hours during the week and 48 hours at the weekend for urgent issues for children on non-invasive ventilation?
- What percentage of children requiring NIV achieve adherence?
- What percentage of children initiating NIV receive weekly support during the initiation phase from a respiratory nurse specialist?

Respiratory management of neuromuscular disease (NMD)

- What proportion of patients have been seen twice in the last year by a neuromuscular MDT?
- What proportion of patients greater than six years of age have had full pulmonary function testing, including sniff nasal and respiratory pressure, in the last year?
- What proportion of patients have had domiciliary overnight oximetry in the last year?
- What proportion of patients have had a respiratory physiotherapy assessment in the previous year?
- What proportion of patients on nocturnal non-invasive ventilation have had daytime oximetry capnography in the last year?
- What proportion of patients on non-invasive ventilation have a fully functioning back-up ventilator?
- What proportion of patients on cough assist device have had advice from the service in the last year?

Long-term ventilation

- What proportion of patients had an MDT meeting organised prior to the decision to perform a tracheostomy?
- What was the average duration between the organisation of the MDT meeting and initiation of the discharge process?
- What was the average duration between the decision to discharge and the granting of the homecare package?
- What was the average duration between the granting of the homecare package and discharge from hospital?
- What proportion of patients had an MDT meeting organised immediately prior to discharge?
- What proportion of patients had a written respiratory management plan on discharge?

Severe asthma

- What proportion of patients had their inhaler technique checked by an asthma nurse specialist on the day of the clinic?
- What proportion of patients have a written asthma action plan?
- What proportion of patients on oral steroid therapy have had a DEXA scan in the last year?
- What proportion of patients on oral steroid therapy have been seen by an ophthalmologist in the last year?
- What proportion of patients on oral steroid therapy have had their blood pressure checked in the last year?
- What proportion of patients on high-dose inhaled corticosteroids have had an assessment of their adrenocortical axis in the last year?

Primary ciliary dyskinesia (PCD)

- What proportion of patients are under regular review by an ENT consultant?
- What proportion of patients have seen a respiratory physiotherapist at least twice in the last year?
- What proportion of patients have been seen at the specialist PCD centre for annual review?

General respiratory services

- What is the average waiting time for a respiratory outpatient appointment?
- What was the longest waiting time for a respiratory outpatient appointment?
- What proportion of referral letters were triaged by the respiratory consultant?
- What is the average waiting time for a respiratory physiotherapy outpatient appointment after a referral from the respiratory clinic?
- What is the average waiting time for full pulmonary function testing?
- What is the average waiting time for a diagnostic CT thorax under general anaesthetic?
- What is the average waiting time for bronchoscopy under general anaesthetic?

Patient and family experience of the service

In the Irish health service, where subspecialists have high volumes of patients with complex conditions and very limited availability of appropriate staffing or infrastructural resources, it is an unfortunate fact that pressing clinical concerns rarely allow patient or family experiences to reach a level of priority that would allow them to be appropriately addressed. If excellent services are to be developed in line with those in developed Western countries, our clinical services must be staffed appropriately to allow us to address not only the urgent and routine clinical workload, but vitally, to begin to seriously address quality, patient safety, patient outcomes and patient/family experiences of the service.

The cystic fibrosis service in Crumlin is currently embarking on a coordinated programme of education and feedback with families using the service. This will comprise a quarterly newsletter, annual or biannual information nights and question-and-answer forums for parents, as well as a mechanism for patient and family feedback on the service. There is the potential to expand this in the future on a national basis, both in the area of CF and non-CF respiratory medicine. This will require time, effort and resources to ensure that it is successful.

Temple Street has organised parents' nights, with speakers from the team covering different topics, and parent group sessions in the evenings run by members of the multidisciplinary CF team. The group nights for parents of newly diagnosed children with CF have been particularly well received. Camps for siblings of children with CF have been run by the CF/respiratory psychologist and social worker. A comprehensive information folder has been produced for parents, containing information on CF and details of related websites. The folder has been published in conjunction with CHIC representatives, thus ensuring that the language is parent friendly. A number of our children with CF agreed to allow use of their art work on the folder cover. Temple Street has produced both parents' and children's booklets containing information on transition of care to adult services. The feedback on all of the above initiatives has been very positive.

Most of these initiatives have been designed with the input of children and parents.

42.6 KEY RECOMMENDATIONS

- Relieve respiratory subspecialists in the national tertiary centre of general paediatric workload, in order to enable them to concentrate on delivering and developing the subspecialty service.
- Increase multidisciplinary staffing levels in tertiary and regional units, in order to provide safe, accessible and effective services for children with complex respiratory diseases, including CF, PCD, NMD and sleep-disordered breathing.
- Paediatric pulmonary function laboratory capacity (incorporating basic sleep laboratory services) is urgently required in regional centres.
- Appropriate infrastructural facilities are required for patients with CF in the (currently six) specialist CF centres.
- Develop a standardised process for children requiring long-term mechanical ventilation, in order to enable discharge to the community and ongoing review of appropriate resource allocation.

Urgent Priorities

- Full implementation of the business case for a national paediatric sleep service. This is well developed and has been partially funded to date.
- Paediatric respiratory laboratories with 1xWTE respiratory physiologist (incorporating sleep and pulmonary function services) are urgently required in each of the regional centres.
- Respiratory nurses (initially 1xWTE each) are urgently required in each regional and tertiary centre
- Respiratory subspecialists in the national tertiary centres must cease general paediatric commitments in order to enable them to concentrate on delivering, developing and streamlining (the national picture and clinical service integration for the new children's hospital) the subspecialty service.

42.7 ABBREVIATIONS AND ACRONYMS

ANP	advanced nurse practitioner
BAL	bronchoalveolar lavage
CCHS	congenital central hypoventilation syndrome
CF	cystic fibrosis
CNS	clinical nurse specialist
CPD	continuous professional development
CT	computer-aided tomography
ENT	ear, nose and throat
GA	general anaesthetic
HCA	healthcare assistant
HCP	Homecare package
HDU	high dependency unit
HSCP	health and social care professionals
HSE	Health Service Executive
LTV	long-term ventilation
MDT	multidisciplinary team
MRI	magnetic resonance imaging

NCHD	non-consultant hospital doctor
NIV	non-invasive ventilation
NMD	neuromuscular disease
OSA	obstructive sleep apnoea
PCD	primary ciliary dyskinesia
PICU	paediatric intensive care unit
PSG	polysomnography
RCPCH	Royal College of Paediatrics and Child Health
SALT	speech and language therapy
TCOM	transcutaneous overnight oxycapnography monitoring
TSANZ	Thoracic Society of Australia and New Zealand
UK	United Kingdom
USA	United States of America
WTE	whole time equivalent

42.8 REFERENCES

Cystic Fibrosis Standards of Care, Australia: 2008 – Cystic Fibrosis Australia

Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (Second Edition): 2011 – UK Cystic Fibrosis Trust

Towards a better service: The treatment of Cystic Fibrosis in Ireland 2005, Problems and Solutions. CF Ireland 2005. www.cfireland.ie/index.php/cfservicesreports

Report on Services for People with Cystic Fibrosis. HSE. 2009 www.hse.ie/eng/services/news/newsarchive

'Full, shared and hybrid paediatric care for cystic fibrosis in south and mid Wales' Doull L. and Evans H. Arch Dis Child 2012 Jan;97(1):17-20

Standards for Sleep Disorders Services 2012 – Australasian Sleep Association

Transforming Respiratory and Sleep Diagnostic Services. DOH (UK) NHS 2009

Standards for Services for Children with Disorders of Sleep Physiology. 2009 Royal College of paediatrics and Child health

Guidelines for respiratory management of children with neuromuscular weakness. British Thoracic Society July 2012

'Core guidelines for the discharge home of the child on long-term assisted ventilation in the United Kingdom' Jardine E. and Wallis C. Thorax 1998

'Breathing Space: Community Support for Children on Long-term Ventilation'. Ludvigsen and Morrison 2003. Barnardos

'From Hospital to Home: Guidance on discharge management and community support for children using long-term ventilation'. Noyes and Lewis 2005. Barnardos

Ventilatory Support at Home for Children – A Consensus Statement from the Australasian Paediatric Respiratory Group February 2008

Primary Ciliary Dyskinesia: Diagnosis and Standards of Care. Bush A et al. Eur Respir J 1998 Oct;12(4):982-8

'Diagnosis and Management of Primary Ciliary Dyskinesia' Lucas J. et al. Arch Dis Child 2014 Apr 25 epub.

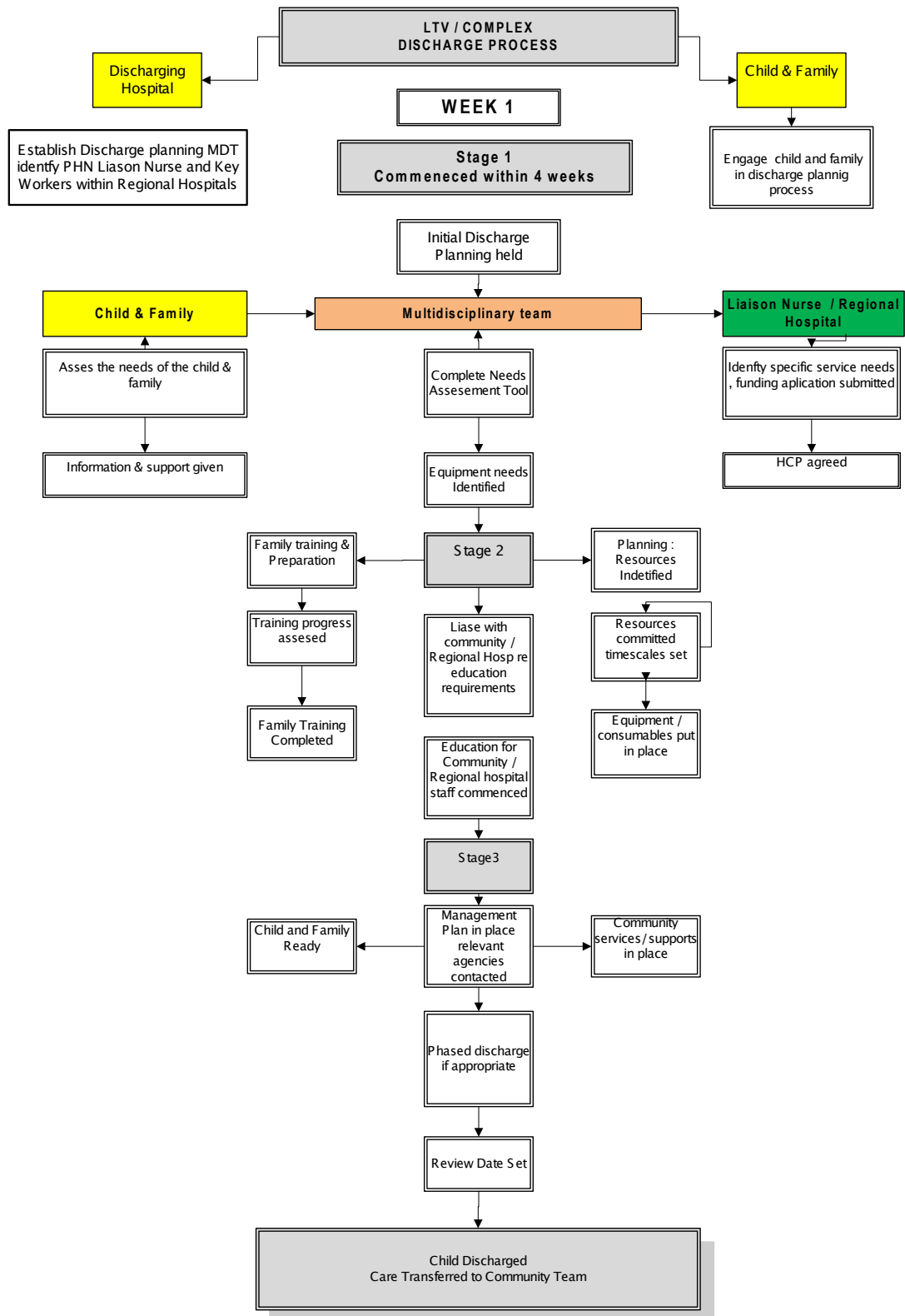
Primary Ciliary Dyskinesia: A consensus statement on diagnostic and treatment approaches in children. Barbato A. et al. Eur Resp J 2009 Dec;34(6):1264-76

'WHO universal definition of severe asthma' Bush A. and Zar H. Curr Opin Allergy Clin Immunol 2011 Apr;11(2):115-21

Report of the National Clinical Programme for Asthma – Draft. 2012

42.9 APPENDICES

Appendix 1 Hospital discharge pathway for long-term ventilation



Appendix 2 Policies, procedures and guidelines

Due to the small size of Ireland's population, clinicians treating less common conditions will often use guidelines from bigger jurisdictions such as the UK, Europe or the US. In most cases, it is unnecessary to duplicate the work involved in creating new guidelines, not least because greater expertise usually exists in these bigger jurisdictions. However, given the various different publications that may already exist, coupled with the disparate approaches used in different centres in Ireland that are currently dependent on local teams, there is a need to streamline approaches and ensure that all centres are operating from a similar set of guidelines.

Cystic fibrosis

Work is underway on streamlining various different recommendations from international centres and bodies, and collating them into a national paediatric set of guidelines for clinical practice in CF. Although much of our clinical practice in the six centres is similar, some variations in practice still remain.

Sleep

Paediatric sleep medicine is still a reasonably new specialty. The most common sleep-disordered condition, obstructive sleep apnoea (OSA) is frequently encountered in clinical practice and, currently, a wide array of different practices exist in terms of diagnosis, management and follow-up of this condition. The clinical guideline for paediatric OSA has been drafted and reviewed by the Irish paediatric respiratory consultants. This is currently under review by subspecialty stakeholders. There is very little guidance in relation to sleep-related hypoventilation in children, and no universal guidelines for diagnosis and management are available.

Neuromuscular disease (respiratory management)

Standards and clinical guidelines for the respiratory management of children with neuromuscular disease (NMD) were produced by the British Thoracic Society in 2012: Respiratory management of children with neuromuscular weakness.

Difficult asthma

The National Clinical Programme for Asthma includes comprehensive information on policies and clinical guidelines relating to asthma management in children.

Primary ciliary dyskinesia (PCD)

The European Respiratory Society produced a comprehensive document on the diagnosis and treatment of children with PCD in 2009. This provides guidance on appropriate investigations and treatment of PCD.