National Steering Group for Specialist Children's Services

Report of the Complex Respiratory/Cystic Fibrosis Review

Content

Introduction

- As part of the National Delivery Plan for Specialist Children's Services in Scotland, work has been undertaken which aims to transform the current, fragmented approach to the provision of specialist paediatric services with an integrated service that improves access and quality of care. Many common respiratory illnesses can be very well managed in primary and secondary care. However, those children with severe or complicated presentation of common illnesses e.g. asthma or pneumonia, or with rare respiratory disease require the involvement of a specialist multidisciplinary team.

- Children with complex respiratory problems should be able to access high quality, effective and timely advice, assessment, diagnosis and treatment to enable them to live to their optimum health potential, and achieve independence as they grow and develop.

- An informal paediatric respiratory organisation(Scottish Paediatric Respiratory Interest Group - SPRING) comprising interested clinicians has evolved in Scotland over the last few years. This report reflects the views of clinicians working within that group and a small multidisciplinary working party convened to produce this report.

2. Background

- Respiratory disorders in children are common, diverse and largely managed in Primary and Secondary care. Tertiary respiratory services are required for the purposes of supporting the local clinicians in diagnosis and management of complex problems of common disease e.g. difficult asthma, as well as providing full or shared care for rare disease. In addition tertiary services have important links to many other paediatric specialties (in particular intensive care, neonatology cardiology and cardiothoracic surgery, oncology, ENT and neurology). The services are dependant not only on highly skilled consultant medical staff but also other essential disciplines include specialist nurses, dietetics, physiotherapy, physiological measurement technicians and psychology. The provision of ready access to specialist investigation facilities is also key to service delivery.

- Scotland has realigned services over recent years to focus specialist volume services (see Table A) in larger centres, to transport sick children to appropriate paediatric intensive care facilities and to increase the standard of healthcare for children dependent on technology. Tertiary Respiratory Medicine has expanded to enable these service redesigns and developments, but without any planning or co-ordination. There are large discrepancies in the services that can be provided at each of the tertiary centres.

Table A: Specialist respiratory services provided for large populations

- Complex Asthma
- Cystic Fibrosis
- Chronic lung disease requiring home oxygen an monitoring
- Interstitial lung disease
- Management of respiratory complications in neuromuscular disease
- Management of recurrent aspiration problems
- Investigation and treatment of recurrent lower respiratory tract infections
- Diagnosis and treatment of sleep disordered breathing
- Long term ventilation
- Bronchoscopy
- Diagnostic pulmonary function testing

3. Incidence/prevalence of Respiratory conditions

- Respiratory conditions are the commonest cause of paediatric hospital admission, accounting for 14% of UK hospital admissions[2] and over 50% of long-term illness in children[3]. Asthma is the commonest cause of school absence, Cystic Fibrosis the commonest life limiting inherited condition. Table B shows the incidence prevalence of respiratory conditions for a NHS Board area similar to Grampian or Tayside, with 100,000 children aged 0-16 years and approximately 6,000 births per year.

1. Table B. Incidence/prevalence of Respiratory conditions

Primary chronic conditions		
	Children affected	Comments
Asthma	20,000	2% with complex/severe disease
Cystic fibrosis	40	All require specialist care
Chronic lung disease of new-born	6 p.a.	Home oxygen required
Non-CF bronchiectasis	20-30	6 of whom have Primary Ciliary Dyskinesia
Sleep disordered breathing	100	Detailed assessment required
Long-term ventilation	2-4	Complex multidisciplinary care package needed
Rare serious lung disease	2-4	Requires specialist diagnostic services
Congenital lung and airway disease	6 p.a.	Requires specialist diagnostic services
Complications of other conditions		
Neuromuscular	25	Severe disease
Neurological	100	Cerebral palsy or degenerative conditions
Immune deficiency	4	Immunology input vital
Oncology	Unknown	Mostly "emergency" input for respiratory infection
Chest wall and scoliosis	Unknown	Liaison to plan and monitor around surgery
Acute admissions		
LRTI	300 p.a.	Often managed by non-specialist
Croup	100 p.a.	Often managed by non-specialist
Empyema	4 p.a.	Involves thoracic surgery

Figures base on Scottish single centre audits or published prevalence data

- The prevalence of asthma has increased over the last 20 years[4] but there is evidence that this may have peaked. However, the workload of seriously affected children, especially in adolescence, may have continued to increase. In these individuals the delicate balance of benefits to risks of steroid therapy requires specialist input.. The incidence of TB is already increasing with changes in ethnic minority groups in Scotland. Changing expectations from parents and professionals are increasing and the demand for more intensive respiratory intervention e.g. non-invasive ventilation for Duchenne Muscular Dystrophy. It is now recognised that several conditions, such as Primary Ciliary Dyskinesia, non-CF bronchiectasis and sleep disordered breathing (especially Obstructive Sleep Apnea in obese children) are currently under diagnosed, leading to preventable morbidity. A further influence on workload is the presence of

new cohorts of children who have survived previously fatal problems but with respiratory sequelae e.g. extreme prematurity.

4. Quality standards/outcome indicators:

- Standards of care for asthma are defined in the SIGN/BTS guidelines[7], and NICE guidance on inhalers[8]. The National Asthma Campaign has recently published a Patient's Charter[9].. Currently all Scottish centres participate in the UK asthma audit annually. The CF Trust has published on standards of care[6]. In addition the UK CF database enables outcomes to be assessed. There are guidelines for long-term ventilation[10] and the Scottish Ventilation Support database is at an advanced stage of development, which will enable annual reviews, and to aid the care. European standards exist for Primary Ciliary Dyskinesia[11], and for respiratory care in neuromuscular disorders. International standards have been published for the management of sleep-disordered breathing[12,13], and UK standards are being developed.

5. Mapping of current services:

- Tertiary respiratory services are currently provided in Aberdeen, Dundee, Edinburgh and Glasgow with varying levels or staff. The presence of Paediatric Intensive Care Unit (PICU) in Edinburgh and Glasgow is a significant influence on the service, particularly with respect to national services and ventilatory support. For some diseases, such as CF, these inequalities are being increasingly recognised. However, for other patient groups, who do not have strong support groups, their equity of access to high quality respiratory services remains unmet. Clinicians find it difficult to commission and fund these low volume, but complex and high cost services, which are often established on an ad hoc basis, without planning or co-ordination.

- As outlined above, Tertiary Respiratory services are dependent on many other specialties and high quality care requires a multidisciplinary team approach. The current national shortage of trained paediatric radiologists is of great concern as high quality imaging is an essential aspect of assessment and diagnosis.

- The possibility for shared care arrangements in Scotland is good, with informal links already in existence, with SPRING acting as a conduit. General paediatricians, with a significant respiratory interest, provide excellent levels of care in secondary centres. However, this provision is patchy and consideration should be given as to a method of providing a more even distribution of good quality respiratory care throughout Scotland (in terms of both medical and AHP care).

- No single centre in Scotland will be able to provide a comprehensive out-of hours service within European Working Time Directives.

- Whilst there are many conditions which can be placed under the banner of complex respiratory; for the purposes of this review it was agreed to focus on the areas listed, as improvements in these areas will have a positive effective on other areas of the complex respiratory service.

    - Cystic Fibrosis
    - Long-term Ventilation/Sleep disorder breathing

6. Cystic Fibrosis – History of service development to date

- Cystic Fibrosis is an inherited disease affecting children and adults. The main effect of the disease is on the lungs, with life expectancy being most closely related to the impairment in lung function. Until the last quarter of the 20th century Cystic Fibrosis (CF) was generally fatal during childhood and, therefore, seen as a purely paediatric problem. Affected children were largely cared for in general paediatric clinics by non-specialists. With the development of paediatric respiratory medicine came specialist CF centres and evidence (especially from Scandinavia[1] and Australia[2;3]) that survival was enhanced by a multi-disciplinary team of health professionals. There was also evidence that patients are more satisfied with the care they receive at specialised CF centres.[4]

- For many years paediatricians and the Cystic Fibrosis Trust recommended the introduction of a screening programme for CF. This was based on the assumption that if children are diagnosed before the onset of symptoms, particularly respiratory infections, then morbidity could be improved, and consequently improve quality of life and survival. However, evidence in the medical literature to support this view was only modest and a national screening programme was repeatedly rejected.

- In October 2001 the Scottish Executive Health Department (SEHD) published a Health Department Letter (HDL 73) proposing screening for CF. The Paediatric Subcommittee of the Scottish CF Group supported the introduction of screening, but strongly recommended that in order to achieve the maximum benefit to health, there would need to be a substantial improvement in clinical services. At this time it was felt that, - this could be most effectively accomplished by the development of a centrally funded Managed Clinical Network . The CF Trust and patient advocates supported this view and a proposal was submitted in December 2002. In parallel to this, the Scottish Executive commissioned a review of paediatric CF services [7] which, although coming to a similar conclusion; it recommended that networks should be regional and funded via local sources, as opposed to national.

- The introduction of screening was delayed until February 2003. Since then there has been evidence published which shows that infants diagnosed before the onset of symptoms, who subsequently get specialist care, are deriving some health benefit. A lack of investment in paediatric services

means that the potential improvement to the health of children diagnosed by newborn screening, remains unmet.

- Children and young people with CF are seen in either a general or teaching hospital setting with variable support services available. The 2002 SE review of paediatric CF services demonstrated that no hospital met the minimum standards of care, in terms of personnel (particularly for Allied Health professionals), as recommended by the CF Trust or European Cystic Fibrosis Society consensus statements[9]. This is still the case and, therefore, there exists a significant inequality in the care that children receive depending on their area of residence.

- Delivering a Healthy Future (An Action Framework for Children and Young People's Health in Scotland) again proposes that the CF service would benefit from the development of a national managed clinical network.

- In Scotland services for adults with Cystic Fibrosis have been commissioned and funded nationally since September 1992. The service is located in three centres – at Gartnavel General Hospital in Glasgow, Aberdeen Royal Infirmary and the Western General Hospital in Edinburgh. Each of these centres aims to meet the specification of a level II centre as described in the clinical guidelines for cystic fibrosis care published by the Cystic Fibrosis Trust, the British Paediatric Association and the British Thoracic Society.

7. Incidence and Prevalence

- CF is a relatively rare disease with an incidence of ~1 in 2500 live births. Following the introduction of screening in Feb 2003 , 85 infants have been diagnosed by screening.

- The latest audit shows that there are 395 children and young people, (under the age of 16) with cystic fibrosis living in Scotland.. Based on previous modelling Scotland is approaching the anticipated rise in the total number of children with CF to ~400 (a 17% increase from 2001 data).

8. Current Situation

- In Scotland children and young people with CF attend 10 different hospitals. Not all of these hospitals have designated CF clinics, but each has a consultant with training and/or a special interest in CF. Tertiary CF services are currently provided in Aberdeen, Dundee, Edinburgh and Glasgow with varying levels of staffing. Following a review of paediatric CF services in 2002 which was commissioned by the SEHD, demonstrated that all of these clinics have inadequate provision of medical, nursing and AHP staffing, as defined by "Standards for the clinical care of children and adults with CF in the UK in 2001".

- This review was revisited as part of development process for National Delivery Plan for Specialist Children's Services in Scotland and the situation regarding staffing has not improved.

- The challenges facing the paediatric CF service are considerable and include:

    - Inequality, in the access to the specialist CF multidisciplinary team, dependent on area of residence, annual reviews and diagnostic services, in order to provide appropriate care to meet a child's needs.

    - Inequalities, in the local care depending on area of residence that can be provided for the ongoing care appropriate to a child's needs.

    - Sharing of clinical experience, good practice and discussion of different approaches is patchy and inadequate.

    - There are insufficient good practice guidelines in operation, which are measurable and auditable.

    - Audit and Research is poor even when guidelines exist.

    - Training opportunities for nursing and allied health professionals are not adequate. Edinburgh RHSC runs a Degree Nursing Module in Paediatric Respiratory Medicine which includes CF, but there are no known courses for AHPs.

    - Lack of national or regional service co-ordination makes it difficult to identify service gaps/pressures in a planned approach.

    - Peer support, especially for staff in district units, is variable for those working with severe and complex cases.

    - Anecdotal evidence suggests that in several Health Board Areas, GPs cannot prescribe Alfa Dornase. It has also been suggested that even simple appliances and equipment for physiotherapy have to be purchased from charitable donations because of inadequate funding of physiotherapy budgets. Not surprisingly, as treatment becomes ever more complex and aggressive, and ever more expensive, there is real anxiety that funding will not keep pace with developments.

9. Models of care

- The Cystic Fibrosis Trust has produced standards which are recognised nationally and internationally. These standards provided a strong basis to model the development of a future service in Scotland and the development of any future model of care must achieve the standards highlighted with this document.

- In previous reports it has been proposed that CF would benefit from the establishment of either regional or national MCN's. However an MCN, without any investment in service provision will not reach the standards or address the inequalities within the current service.

- It is therefore proposed that National Service Network should be established. This would enhance support for local clinics and reduce the current inequalities, by augmenting the three regional specialist centre multidisciplinary teams and developing outreach clinics. In addition a NSN would provide the infrastructure to develop national standards of care, education, audit and research throughout Scotland.

- The establishment of a NSN, requires additional investment for staffing and resources. It is proposed that this be phased over a two year period, with the regional centres being develop in year 1 and local centres in year 2. As with the adult , the paediatric regional centres will meet the specifications of a level II centre as described in the Standards of Care

- At present, informal networks exist to support professionals working with CF Children and young people. A NSN would formalise these networks, ensuring the care of children through facilitating and enhancing local service quality, and also facilitating planning and governance of paediatric CF services. It will be developed on a regional basis, similar to the adult service, working from the three children's hospitals, providing a substantial outreach service. The following describes some of the advantages that could be brought by a combined approach.

- **Sharing Good Practice and Supporting Local Services**

  A key benefit of a network will be the provision of support to local services, ensuring where possible that children and families are provided with appropriate care, close to home. In addition to telephone advice provide by the team, it is proposed that the specialist multi-disciplinary teams undertake regular visits to the local clinics in their region. This will give all children and their parents access to the specialist health professionals, and will aid the delivery of the other benefits from training and education, etc. as delineated below.

- **Coordination of appropriate care**

  For a safe level of care, it is essential that the most appropriate resources available at any one time are utilised to meet a child's needs. Inadequate knowledge about potential resources, inflexible use of potentially valuable resources and inadequate understanding of a child's needs can lead to suboptimal treatment. An important function of a NSN would be a sharing of knowledge regarding different resources and treatment approaches to improve clinical care.

- **Training and Education**

  Whilst shared experience in the management of single cases provides one form of training, knowledge and experience can be shared in other ways. This may involve exchanges between units of training materials or other resources. The combined approach will also facilitate educational programmes that include travelling speakers, national meetings, sabbatical and exchanges as staff temporarily move between placements in different centres.

- **Partnership with children, young people and families**

  It can be difficult to ensure parents, children and young people have a voice in the planning of specialist services. The network will provide a conduit of communication between specialist units and parent representatives. This will be important both for the units to get input from parents, and for the dissemination of information by the units. The NSN will facilitate the development of appropriate information about CF services, where advice is to be sought locally, and actively encourage patients/carers to participate in the network directly. This would inform the planning of services from a user perceptive.

- **Standardised practice.**

  It is important that all children have access to an appropriate level of care wherever they live in Scotland. This requires uniformity of service delivery across Scotland. The network will support the development and implementation of national standards on the management of children and young people with CF and the resources required to deliver those standards.

- **Audit**

  The network will facilitate audit programmes to examine the clinical effectiveness of services. A national network would also make it possible to audit practice across Scotland

- **Research**

  Advances in CF care have been hampered by studies from single centres that are inadequately powered to answer research hypotheses. By harmonising practice and developing a NSN will develop a Scottish multi-centre approach to cystic fibrosis research. In addition established (but poorly researched) and new therapeutic interventions could be investigated in Scotland through the Medicines for Children initiative.

- **Communication**

  The enhancement of communication between CF clinicians, through the work of the network, would also help to reduce the isolation experienced by clinicians and the resulting stress that isolation can bring. It is also hoped this would improve staff retention as staff often site stress and isolation as a reason for leaving their post.

10. Resources Issues

- As part of the SEHD CF review in 2002 a range of staffing short falls were identified .This work was revisited as part of the review process, and identified significant gaps in the provision of AHP, nursing and psychological services. To bring the service in line with the CF Trust standards would require a staffing investment. Staffing information is attached as appendix 1

- As part of the planning and commissioning process in England work has been undertaken to cost the level of care required based on the severity of an individuals condition. This banding approach costs the level of hospital support and cost of drug therapy required. This would support early intervention approaches as conditions become more severe they also become more costly. Appendix 2 provide more detailed information, based on work undertaken by the Department of Health.

11. Establishing a NSN for CF

During it's first year a NSN would -

National

- Establish a national steering group and its terms of reference
- Agree roles and responsibilities of key network stakeholders
- Agree network membership
- Within three months develop a NSN development plan detailing specific timescales and priorities for year one
- Establish working groups and/or individuals responsibility for taking forward the elements of the development plan

Regional

- Recruit staffing to support implementation of NSN via three regional centres
- Support implementation of NSN

Locally

- Recruit staffing to support implementation of NSN with District General Hospitals
- Support implementation of NSN

12. Long –term Ventilation/ Sleep disordered Breathing

Background

- Long-term ventilation is a mode of respiratory therapy which can be delivered in a variety of settings including the child or young person's home. It is a relatively new development which has had a significant impact for a number of children who would previously have died of respiratory failure and there is evidence that many of these children experience a significantly improved quality of life.

- There are several groups of children who may require LTV –
    1. Children surviving a period of neonatal or paediatric intensive care with significant ongoing respiratory insufficiency
    2. Children with some chronic and degenerative conditions such as – neuromuscular disorders and skeletal disorders.
    3. Children with congenital respiratory control disorders
    4. Children with obstructive airway problems including Obstructive sleep apnoea

- Whilst this service needs to be led by the NHS, it requires to work closely with social work and education services to provide high quality integrated care of children and young people.

Sleep disordered breathing

- The demand for diagnostic and treatment services is increasing rapidly due to increasing knowledge, differing expectations of what can be achieved ad availability of expertise. In addition, the increasing prevalence of obesity is leading to increased numbers of children with obstructive apnea and sleep hypoventilation.

Current Situation

- Whilst there are examples of good service there is not equality of provision or access provided across Scotland in the areas of sleep breathing diagnosis and management. Consequently it is felt that there are significant improvements which could be made. But to clearly identify the currently level of the service and challenges it may face, there is a need to undertake a multi-agency review, as well as a needs assessment to clearly identify future demands.

13. Other aspects of respiratory Care which require to be considered –

- **Transport** - Scotland covers a wide geographical area, and it is by no means easy to provide a satisfactory service for patients in remote areas – for parents of young children, a distance of only 40 miles can be a major barrier to regular clinic attendance.

- **Inadequate funding of paediatric clinics** – in most Health Boards paediatric CF/respiratory services are considered to be part of the general provision for paediatrics, and no specific funding is identified. Based on previous work all clinics have inadequate provision of medical and in particular AHP staff.

- **Respiratory care of children with neurological disorders.** - Changing expectations and demonstration of gains in quality and quantity of life have led to an increase in the intensity of respiratory support provided. Children with chronic severe disability are living longer; occupying more intensive care beds, and are becoming more technology-dependent. Respiratory paediatricians are asked to lead the provision of care for these patients, but without adequate resources to do so.

- **Diagnostic services.** - Appropriate, standardised, equipment with fully trained technical staff is crucial to any tertiary service. Investment in IT would allow investigations to take place locally, thereby reducing unnecessary travel for patients. Flexible bronchoscopy is established in 3 centres but large amounts of time/effort are wasted as no centre has adequate access to dedicated theatre sessions or supporting staff.

- **Transition Issues** -The transition into the respiratory service for those with neonatal conditions such as chronic lung disease is inconsistent, with some home oxygen services provided by neonatologists and some by respiratory paediatricians. Hospital readmission is common and it is imperative that a seamless service is provided for these young people. The transition between paediatric and adult respiratory services is also very variable. CF generally has good transitional arrangements, probably because of the existence of dedicated relatively well funded adult services. In other conditions e.g. asthma, immune deficiency, congenital lung disease, etc. good transition arrangements are not established.

- **Oxygen Therapy -** It is disappointing that recent NHS Scotland pronouncements on Oxygen therapy appear to have overlooked paediatric aspects of care. Children may need this service in fewer numbers but the health gain is relatively large.

14. Recommendations

- Similar to other services the development of Managed Clinical Networks is seen as the best way of delivering Tertiary Respiratory Medicine across Scotland, while ensuring equity of access. Without substantial investment in resources in Tertiary Paediatric Respiratory Medicine it will not be possible to meet these aspirations or even maintain the current level of service with the impending changes to working patterns set by the European Working Time Directives.

- Specific recommendation for Cystic Fibrosis and Long-term Ventilation and Sleep disorder breathing are outlined below

Cystic Fibrosis

Recommendation	**Time-scale**	**Lead**
In year 1 establish a National Service Network, provided via three regional centres, incorporating outreach services to DGH's	August 2008	
In year 1 each Region to produce an implementation plan for CF services, ensuring that the Cystic Fibrosis Trust Standards are met.	March 2009	
In year 2 appoint additional staff for 3 Regional Centres to support service network and ensure minimum staffing levels recommended by the Cystic Fibrosis Trust are achieved	March 2009	
In year 3 appoint staff for DGH's support service network and ensure minimum staffing levels recommended by Cystic Fibrosis Trust are achieved,	March 2010	
Develop pathways of care, ensuring effective transition to Adult service	March 2009	
Undertake a review of services to ensure Cystic Fibrosis Trust Standards are being achieved in all centres providing CF care	March 2010	

Long - term Ventilation

Recommendations	Time-scale	Lead
Undertake a national multi-agency review of long-term ventilation provision for children and young people, including invasive and non-invasive ventilation.	March 2009	
Complete a needs assessment on children's and young peoples future needs/demands for long-term ventilation	December 2009	
Identify resource requirements for current and future provision of long-term ventilation	March 2009	
Develop quality standards to ensure equity and sustainability of long-term ventilation services	March 2010	

Sleep breathing Disorder

Recommendations	Time-scale	Lead
Complete review of current service provision	March 2009	
Complete a needs assessment on children's and young peoples future needs/demands for sleep breathing disorder	March 2009	
Develop quality standards to ensure equity and sustainability of sleep disorder services	March 2010	

References:

1. Specialised Services National Definitions Set (2nd Edition), Department of Health
2. Department of Health (England). Hospital Episode Statistics
3. Joint Health Surveys Unit 1999. Health Survey for England: The Health of Young People 1995-97. The Stationery Office, London
4. Omran M, Russell G. Continuing increases in respiratory symptoms and atopy in Aberdeen school children. BMJ 1996; 312 (7022): 34
5. Standards of Clinical Care of Children and Adults with CF in the UK 2001, CF Trust
6. BTS/SIGN Guidelines on asthma. Thorax 2003;58(suppl 1):i1-i92
7. http://www.nice.org.uk/pdf/Niceinhalers 1dC38GUIDA.pdf and http:///www.nice.org.uk/pdf/NiceINHALERguidance.pdf
8. http://www.asthma.org.uk/images/charter.pdf
9. Jardine E, Wallis C. Core guidelines for the discharge home of a child on long-term assisted ventilation in the UK. UK Working Party on Paediatric Long-term Ventilation. Thorax 1998;53 (9): 762-767
10. Bush A, Cole P, Hariri M, MacKay I, Phillips G, O'Callaghan C et al. Primary Ciliary Dyskinesia: diagnosis and standards of care. European Respiratory Journal 1998; 12: 982-988
11. Thoracic Society of Australia and New Zealand and Australasian Sleep Association. Accreditation of sleep disorder services, including standards of paediatric laboratories. 2002
12. American Academy of Pediatrics. Clinical Practice Guideline: Diagnosis and management of Childhood Obstructive Sleep Apnea Syndrome. Pediatrics 2002; 109: 704-12
13. Frederiksen B, Lanng S, Koch C, Hoiby N. Improved survival in the Danish center-treated cystic fibrosis patients: results of aggressive treatment. *Pediatr Pulmonol* 1996;**21**:153-8.
14. Phelan PD, Allan JL, Landau LI, Barnes GL. Improved survival of patients with cystic fibrosis. *Med J Aust.* 1979;**1**:261-3.
15. Hill DJ, Martin AJ, Davidson GP, Smith GS. Survival of cystic fibrosis patients in South Australia. Evidence that cystic fibrosis centre care leads to better survival. *Med J Aust.* 1985;**143**:230-2.
16. Walters S, Britton J, Hodson ME. Hospital care for adults with cystic fibrosis: an overview and comparison between special cystic fibrosis clinics and general clinics using a patient questionnaire. *Thorax* 1994;**49**:300-6.
17. http://www.show.scot.nhs.uk/sehd/publications/DC20030221CysticReport.pdf
18. Kerem E, Conway S, Elborn S, Heijerman H. Standards of care for patients with cystic fibrosis: a European consensus. *J Cystic Fibrosis* 2005; 4: 7-26.

Appendix 1

Current Level of Staffing

Currently Levels of Staffing

		Consultant 1	Consultant 2	Staff Grade	CF Sp R	CF Nurse	Physio	Diet	Psychologist	data clerk	Pharmacist
Grampian		0.2	0.2	0.3	0.2	1	0.1	0.1	0	0.1	0
Glasgow/Clyde		0.5	0.4	0.5	0.5	2	2.5	0.5	0	0.2	0.3
Lothain		0.3	0	0	0	2	0.4	0.3	0.2	0.6	0.05
Highland		0.1	0	0	0	1	1	0.1	0	0.2	0
Forth Valley		0.2	0	0	0	0.2	0.1	0.2	0	0	0.1
Dumfries		0.1	0	0	0	0	0	0	0	0	0
Borders		0.1	0	0	0	0	0	0	0	0	0
Tayside		0.2	0.2	0.2	0.2	1	0.5	0.2	0	0.5	0.1
Lanarkshire		0.1	0	0	0	0	0	0	0	0	0
Total		**1.8**	**0.8**	**1**	**0.9**	**7.2**	**4.6**	**1.4**	**0.2**	**1.6**	**0.55**
Required Levels		3.2	1.3	2.2	2.6	6.3	13.9	3	4.2	0	1.95

Required numbers of staff to meet CF standards – based on numbers of Children & Young people being treated

Unit											
		Consultant 1	**Consultant 2**	**Staff Grade**	**CF Sp R**	**CF Nurse**	**Physio**	**Diet**	**Psychologist**	**data clerk**	**Pharmacist**
Grampian		0.5	0.3	0.4	0.5	1.5	2	0.4	0.4	0.1	0.3
Glasgow/Clyde		1	0.6	1	1	3	4	1	1	0.2	0.5
Lothain		1	0.6	1	1	3	4	1	1	0.2	0.5
Highland		0.5	0	0	0	1	1.5	0.4	0.4	0	0.2
Forth Valley		0.5	0	0	0	1	1.5	0.4	0.4	0	0.2
Tayside		0.5	0.3	0.4	0.5	1.5	2	0.4	0.4	0.1	0.3
Lanarkshire		0.5	0.3	0.4	0.5	1.5	2	0.4	0.4	0.1	0.3
Totals		**5**	**2.1**	**3.2**	**3.5**	**13.5**	**18.5**	**4.4**	**4.4**	**0.7**	**2.5**

Appendix2

THE CLINICAL CARE OF CHILDREN AND ADULTS
WITH CYSTIC FIBROSIS
BANDINGS AND ASSOCIATED COSTINGS

THE CLINICAL CARE OF CHILDREN AND ADULTS
WITH CYSTIC FIBROSIS
BANDINGS AND ASSOCIATED COSTINGS

2.

3. THE IMPORTANCE OF BANDING

3.1

3.2 The banding of patients with CF according to the severity of their disease and ensuring appropriate levels of care are properly funded is of vital importance for the following reasons:

1 Hospital Trusts and CF Specialist Centres are unable to fund proper CF care from 'block contracts', including paediatric 'block contracts' and respiratory 'block contracts'.

2 The cost of proper CF care should be broadly similar regardless of geographical location. There is no reason or excuse for specialist CF care to be funded at entirely different levels in different parts of the country.

3 It would be completely impractical to try to cost CF care on an 'episode' basis, including outpatient appointments, inpatient care and hospital support for home treatment, as is the plan of the NHS 'National Tariff' initiative, which is currently being implemented for other conditions.

4 Further concern has arisen because early indications from the 'National Tariff' focus on inpatient episodes and tend to discount outpatient and home care provision, which would make current CF care patterns impossible to fund and therefore to deliver. If this was implemented, it would act as a total disincentive for any hospital Trust to provide outpatient or home care for CF. The laudable efforts of clinicians to manage even the most severe patients from time to time within an outpatient setting, would therefore be jeopardised, and in order to cover the cost of IV drugs, patients would need to be admitted rather than administering IV's themselves at home. The current proposal would also make it impossible for patients in Band 2 to receive intense therapy at home in the form of IV's, which many of them currently receive at present, either to tackle an episodic infection or as a preventative measure. The only way to continue this established good practice in the context of the present proposals would be to admit these patients on a regular basis for intravenous antibiotics. This would clearly be contrary to current Department of Health guidance as there is an obvious intention to encourage as many patients as possible to be seen as outpatients rather than to be admitted.

5 Similarly, elective as opposed to non-elective treatment is not a relevant consideration in assessing, delivering and costing CF care.

6 It is important that it is understood that bandings will include some inpatient and outpatient care, and that a patient may stay in or even revert to a lower banding of care than would be the case if they were treated less aggressively. Higher costs do not therefore always indicate severity of disease or deterioration, but reflect the need for aggressive treatment to keep infections under control and prophylactic treatment to protect CF lungs from infection.

7 At present, there are no realistic proposals whereby CF care will be delivered to a high and equal standard across the UK. PCTs in England do not have the commitment or resources to address such a complicated by relatively rare condition.

It is, therefore, extremely important that bandings which represent 'Packages of Care' and associated costs for CF are accepted by the NHS and particularly by the Healthcare Resource Group, as was the case in relation to the National Specialised Services Definition (10) for Cystic Fibrosis.

The bandings which have been developed are as follows:

Diagnostic Year

Some patients may be admitted to hospital after they have been diagnosed whilst most will be cared for as outpatients. The outpatient input will be intense and they will spend a lot of time with doctors, CF nurses, physiotherapists and dieticians learning how to manage their CF.

3.2.1 Band 1

Patients who come only to outpatients, receive outpatient care in terms of input from physiotherapist, doctors, social workers, dieticians etc. They may receive nebulised antibiotics and courses of oral antibiotics from time to time and they receive regular pancreatic enzyme supplements and vitamin supplements as do 95% of CF patients

3.2.2 Band 2

Patients who receive the above and in addition receive outpatient intravenous antibiotics up to 3-4 times a year. They may occasionally be admitted. The input as outpatients may be more intense. Some of them will only be kept in Band 2 level by intensive and sometimes expensive drug treatment to keep Pseudomonas aeruginosa at bay.

3.2.3

3.2.4 Band 3

Similar to 1 and 2 but essentially intravenous antibiotics are received as an inpatient 3-4 times a year. They may also have Diabetes, require feeding gastrostomies, and require a higher input overall.

3.2.5

3.2.6 Band 4

Patients with severe disease, who will come into hospital at least 3-4 times a year for intravenous antibiotics, and have increasingly disease severity. They may have Diabetes and more resistant organisms. They may be under consideration for transplantation.

3.2.7

3.2.8 Band 5

These patients have usually been in Band 4 for at least a year and need to stay in hospital for 4-6 months throughout the year whilst awaiting transplantation or receiving palliative care. They are unable to go home because of oxygen dependence, nocturnal ventilation and feeding gastrostomies and need intravenous antibiotics every day, sometimes for 2-3 years, although on average, these patient's life expectancy is usually no more than a year to 18 months.

3.2.9 ASSOCIATED COSTS

To translate this into costs has been done by taking the average annual plan of hospital care for a patient in each band, including outpatient and inpatient care. Outpatient care will either be routine CF care or intensive care, including IV or other intense drugs. Inpatient care is divided into those requiring an admission of 6 nights or less which is categorised as a Routine Admission, and those requiring an admission of 7 nights or more, which is categorised as an Intensive Admission.

It would be relatively easy, given the UK CF Database, to band each patient within each CF Centre according to this criteria and to arrive at a proper costing for each patient. This information could be given to each PCT, thus enabling the money to follow the patient. This would reflect an estimated level of care based on a yearly package for each CF patient. Commissioners and providers will then be able to monitor actual activity and adjust funding accordingly. There is likely to be relatively little unexpected change of a significant nature, which is an advantage in H.R.G. terms, where there is particular concern about volatility and unexpected high costs.

An alternative would be for a specialised PCT to be established, representing a community of those with the same condition, rather than a geographic community.

The following costs have been fully worked out and are based on the care pattern, which shows how the annual cost relates to the outlined episodes costs. Hospital costs exclude drugs routinely prescribed by GP's. (pancreatic enzyme supplements, vitamin supplements, courses of oral and nebulised antibiotics), but practice in respect of additional CF medication varies considerably throughout the country, with some GP's prescribing drugs which are elsewhere prescribed by hospitals, either reflecting hospital practice or a GP's refusal to prescribe particular drugs.

THE OVERALL COST TO THE NHS OF CARE FOR THOSE WITH CYSTIC FIBROSIS

← HOSPITAL COSTS →← DRUG COSTS* maximum likely →

	4. % 5. patients	*Total*	*Variable*	*Fixed*	*Total*	*Centre*	*GP*	**TOTAL COST**
6. Diagnostic Year	**3**	**8,103**	**3,715**	**4,388**	**2,000**	-	**2,000**	**10,103**
7. Band 1 8. Mild	**37**	**1,850**	**724**	**1,126**	**2,000**	-	**2,000**	**3,850**
9. *Band 2* 10. Moderate	**17**	**12,388**	**8,000**	**4,388**	**23,500**	**8,000**	**15,500**	**35,888**
Band 3 **Severe**	**24**	**24,877**	**11,398**	**13,479**	**36,900**	**12,000**	**24,900**	**61,777**
Band 4 **Extra Severe**	**16**	**53,035**	**22,730**	**30,305**	**41,900**	**16,000**	**25,900**	**94,935**
Band 5 **Awaiting Transplant**	**3**	**Average 83,548**	**34,982**	**48,566**	**48 – 100,000****	**48,000 – 100,000**	**-**	**-**

Most children will be in the lower bands.

NOTE: The drugs cost is normally cumulative, although in some cases substitutions may occur. As patients move up the banding system, more drugs will be necessary. Expensive drugs such as DNase and TOBI will normally be given on a shared basis, with the CF Specialist Hospital Centre bearing the cost of the trial period, after which the GP will be asked to prescribe the drug if it is recommended to be continued.

*** Not all patients will need all of these drugs.**

**** At this stage, patients spend most of their time in hospital, with spells of being cared for intensively by the CF team at home, so their drug costs will be borne by the hospital.**

26