Clinical guidelines for cystic fibrosis care

SUMMARY OF GUIDELINES PREPARED BY A WORKING GROUP OF THE CYSTIC FIBROSIS TRUST, THE BRITISH PAEDIATRIC ASSOCIATION AND THE BRITISH THORACIC SOCIETY

By the early 1980s the majority of patients with cystic fibrosis (CF) were surviving to adult life and in 1990 the Royal College of Physicians published a report [1] which set out the general principles of care for adults with CF. Three years later the Clinical Standards Advisory Group (CSAG), an independent source of expert advice to the government, reported on access to and availability of specialist services for patients with CF [2]. They concluded that a substantial number of patients were receiving a poor standard of care and that there was an urgent need for guidance in appropriate care. The government's response to the CSAG report [3] welcomed the recommendation that national guidelines concerned with service specifications and quality standards should be developed and the NHS Management Executive (NHSME) subsequently announced its intention to work with the professions to identify and develop clinical guidelines which would be useful in informing discussions between purchasers and providers [4]. In light of these events a working party of consultant physicians and paediatricians with a special interest in CF was convened under the aegis of the Cystic Fibrosis Trust to prepare a document setting out clinical guidelines for CF care based on research evidence and current best practice [5]. This article is based closely on that document.

Cystic fibrosis

In the UK the carrier rate for CF is one in 25 and the incidence one in 2,500. There were approximately 6,250 patients with CF in the UK in 1994, of whom 40% were over the age of 16. The median survival for children born with the disease in the 1990s is predicted to be more than 40 years [6]. Carrier screening is unlikely to affect overall numbers significantly but neonatal screening will lead to earlier diagnosis and a probable further increase in survival.

The disease results from the mutation of a gene located on chromosome 7 which encodes for a chloride channel (cystic fibrosis transmembrane conductance regulator—CFTR). Faulty regulation of the movement of salt and water across cell membranes results in thickened secretions in mucous glands in various organs such as the lungs, pancreas and intestines. Some cases present at birth with meconium ileus, a minority are diagnosed in adult life, but the majority of patients present in early childhood with persistent respiratory tract infection and intestinal malabsorption with failure to thrive. Most of the morbidity and more than 90% of the mortality from CF is due to chronic bronchial infection and its complications. Older patients suffer a variety of complications outside the respiratory tract, for example distal intestinal obstruction syndrome, liver disease and diabetes.

The need for special care

CF is a multisystem disease which requires a holistic approach to care from a wide range of disciplines. This type of care is best provided by a team of trained and experienced professionals, including paediatricians, adult physicians, nurse specialists, physiotherapists, dietitians, social workers and secretaries, working in a specialist centre. Specialist teams must be supported by staff from other specialties and full facilities for treatment, investigation and research. Specialist care must be complemented by local hospital and primary care support. Evidence from the UK and overseas indicates that patients receiving their care in specialist clinics have significantly longer survival [7–10], better clinical status and greater satisfaction with their care [11].

The CSAG defined four levels of hospital care which are summarised as follows:

Level I *National Resource Centre* (large numbers of patients; national education, training, research and resource centre; national tertiary referral centre)

Level II *Major Specialist Centre* (over 100 patients; major specialist interest by medical and paramedical staff; provide care for patients from a local region; local research and resource centre)

Level III *Local Specialist Centre* (over 35–40 patients, including some adults; special CF clinics; some specialist interest among staff)

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Level IV Local Hospital (about 10 patients seen in special or general clinics; providing day to day general care)

The guidelines working group recognised that in 1995 the CSAG definitions of levels of CF care needed to be modified. Standards of care were even more important than numbers of patients. Some specialist centres with less than 100 patients were able to provide the comprehensive specialist service required at Level II but a minimum of 50 patients was required to develop and maintain a specialist team with the necessary expertise. As a general principle all patients with CF should have access to Levels I/II as an ideal and to Level III as a minimum, although this access could be achieved by shared care.

Shared care

Shared care between specialist centres and the local hospital and primary care services has developed as a means of providing specialist care for larger numbers of patients with the minimum of travel and of improving local expertise. Since Level III centres do not have the full range of services available to Level I/II centres they will normally need to refer patients to the latter for tertiary care. In certain more remote parts of the country, however, it may be appropriate for Level III centres themselves to offer shared care to local hospitals.

There are three patterns of shared care:

- *Annual assessments*—Patients receive most of their care from the local hospital and attend the specialist centre for annual assessment which includes specialist investigations, dietary and physiotherapy assessment and a full report giving plans for treatment over the following year.
- *Alternating visits*—Patients attend the local hospital and the specialist centre alternately. Good communication is essential if patients are not to be given conflicting advice.
- *Joint clinics locally*—Patients attend the local hospital which one or more members of the specialist centre team visit at regular intervals to run a joint clinic with the local health professionals. Occasional visits to the centre may be needed for certain complex investigations.

Functions of the specialist centre (Levels I/II)

The functions of the specialist centre (Levels I/II) are:

- Comprehensive assessment and care, including open access, for local patients and those referred for shared care.
- Provision of a wide range of treatments and services not readily available at local hospitals or when special expertise is required, for example, management of unusual and complex respiratory and gastroenterological problems or pre-transplant care.
- Provision of diagnostic and specialised laboratory facilities, including genetic investigations and counselling, sweat tests and complex lung function tests.
- Psychosocial support for problems specific to CF such as education, employment, transplantation and bereavement counselling.
- Provision of education for regional colleagues and research, both clinical and operational.

Facilities and staffing of the specialist centre (Levels I/II)

There should be 3–5 beds in single rooms for every 50 patients with CF. Outpatient rooms should be available where medical staff, physiotherapists, dietitians, nurse specialists and social workers can interview patients. Separate arrangements must be available for patients who pose the risk of cross infection.

The following recommended staffing levels are based on about 50 patients; for larger clinics more staff will be needed in proportion:

- One consultant with a major commitment of 3–4 sessions with further sessions from another senior doctor, making a total of 6 sessions weekly.
- Intermediate grade medical staff to provide at least 5 sessions weekly in addition to the usual junior staffing of the ward team.
- One whole-time clinical nurse specialist
- Two whole-time physiotherapists
- One whole-time secretary/clerk
- Part-time input from a psychiatrist and/or a psychologist

Standards of care

The guidelines set out the desirable standards of care for all patients with CF under the following headings:

Diagnosis

Diagnosis is confirmed by demonstrating high levels of sodium and chloride in the sweat. Two sweat tests should be performed in a laboratory experienced in the technique, ideally at least one at a specialist centre (Levels I/II). All patients should be genotyped. As soon as the diagnosis is confirmed full explanation of the disease and its effects on the patient and family should be given and the arrangements for long-term care outlined. All newly diagnosed patients should be notified to the regional (Levels I/II) specialist centre.

Routine outpatient reviews

Reviews should be carried out at intervals of not less than three months, more often in children and

patients with complications. They should include measurement of height and weight in children and weight in adults, clinical assessment by the doctor, physiotherapist and dietitian and referral to the nurse specialist and social worker if necessary. Investigations should include sputum culture (or cough swabs in children) using appropriate media and, when indicated, oximetry, spirometry and urinalysis for glucose. A full report of the review should be sent to all relevant colleagues within 10 days.

Annual assessments at a specialist centre (Levels I/II)

Annual detailed assessments should be made by the CF specialist team. In addition to the investigations at routine reviews the following are carried out: chest x-ray, more detailed lung function tests, liver function tests (which may include abdominal ultrasound), blood glucose, assessment of intestinal absorption and various other tests. If the patient is attending the centre for the first time the reliability of the diagnosis and genotyping are checked, long-term treatment plans are discussed, all members of the specialist team are introduced and follow-up arrangements made.

Inpatient services

A wide range of inpatient services is essential. These include all aspects of respiratory care including management of ventilatory support and cardiorespiratory failure, total parenteral nutrition and medical, surgical and diagnostic services to deal with such events as meconium ileus, distal intestinal obstruction syndrome, complications of liver disease, and severe haemoptysis. Appropriate management must be provided for patients awaiting a transplant and for all aspects of the care of the dying.

Care in the community

Carrying specialist CF care into the community is cost-effective and is favoured by patients. This form of shared care should be supervised by the multidisciplinary team from the centre but close cooperation with the local primary and community care services is essential. It includes physiotherapy, intravenous feeding and drugs, oxygen therapy, management of nasogastric or gastrostomy feeding and the instruction and support of patient and family, especially for those with terminal disease.

Specific problems

Specific problems which are indications for referral to or discussion with the Levels I/II centre are:

- Respiratory infections which are prolonged for more than 2–3 weeks, especially those due to *Burkholderia cepacia*, multiresistant *Pseudomonas aeruginosa* or methicillin resistant *Staphylococcus aureus*
- Deterioration in pulmonary function not responding to treatment
- Pneumothorax
- Severe haemoptysis (>100 ml)
- Allergic bronchopulmonary aspergillosis
- Significant or persistent atelectasis
- Respiratory or cardiac failure
- Gastrointestinal bleeding (usually from varices)
- Acute or subacute intestinal obstruction or severe abdominal pain
- Jaundice
- Significant or unexplained weight loss or failure to thrive
- Onset of glucose intolerance
- Vasculitis
- Arthropathy
- Pregnancy
- Serious psychosocial problems
- Need for transplantation

Contracts and the delivery of specialist CF care

The essential services to be covered by contracts are therefore:

- Appropriate numbers of medical and paramedical staff.
- Designated outpatient facilities and sessions which allow sufficient time for the necessary consultations with patients and families.
- Inpatient facilities which should ideally be in designated single rooms.
- An investigation service relevant to the care of CF
- Other supporting services with experience in the management of CF.
- Facilities for assessment and referral of cases for organ transplantation.
- A community treatment service and the provision of equipment for home treatment, the financial responsibility for which should be clearly defined.

Contracts with Level I/II centres should include all these essential services and all the functions mentioned above, providing for direct care for all local patients and for other patients who require or desire it. The centre should also provide tertiary and shared care for Level III centres and local hospitals.

Contracts with Level III centres should provide most of the above essential services and functions for local patients and, in certain parts of the country, shared care for patients from neighbouring districts without a Level III centre. All newly diagnosed patients should be notified to the regional Level I/II centre for registration purposes.

General principles for commissioning and contracting

- Commissioning authorities should ensure that the contracts they place reflect the total clinical needs of all their resident adult and child patients with CF.
- All patients should ideally have a right of access to Level I/II care, at least through shared care, or to Level III care as a minimum. Some patients, particularly those living in isolated areas, may prefer to be cared for in a local hospital but they must be offered shared care with a Level I/II centre (or, depending on local circumstances, Level III).
- Contracts should cover shared care and referrals for tertiary care. They should specify the different standards of care expected in Levels, I, II and III centres and local hospitals, their roles in education, training and research and their responsibility for audit and the maintenance of a patient register.
- Contracts should define which treatments and procedures are considered to be undergoing research and evaluation and those which are funded from contract monies. This will require annual updating.
- Patients should not be denied treatments of demonstrated clinical benefit on financial grounds. Contracts should clearly define the financial responsibility for providing drugs, nutritional support and the provision and servicing of equipment.
- Contracts should include resources for community services provided by centres and for the education and training of other staff in the region.

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References

1 Royal College of Physicians. *Cystic fibrosis in adults.* Recommendations for the care of patients in the UK. Report of a working party. London: RCP, 1990.

2 Clinical Standards Advisory Group. *Cystic fibrosis: access to and availability of specialist services.* London: HMSO, 1993.

3 Government Response to the Reports by the Clinical Standards Advisory Group on Access to and Availability of Specialist Services. London: Department of Health, 1993.

4 NHS Management Executive. *Improving clinical effectiveness.* EL(93)115. Leeds: NHSME, 1993.

5 Cystic Fibrosis Trust. *Clinical guidelines for cystic fibrosis care.* Recommendations of a working party. London: CFT and RCP, 1996.

6 Elborn JS, Shale DJ, Britton JR. Cystic fibrosis: current survival and population estimates to year 2000. *Thorax* 1991;**46**:881–5.

7 British Paediatric Association Working party. Cystic fibrosis in the United Kingdom 1977–85; an improving picture. *Br Med J* 1988;**297**:1599–602.

8 Nielsen OH, Schiotz PO. Cystic fibrosis in Denmark in the period 1945–1981: evaluation of centralised treatment. *Acta Paed Scand (Suppl)* 1982;**301**:107–19.

9 Phelan P, Hey E. Cystic fibrosis mortality in England and Wales and in Victoria, Australia 1976–80. *Arch Dis Child* 1984;**59**:71–83.

10 Corey M, McLaughlin FJ, Williams M, Levison H. A comparison of survival, growth and pulmonary function in patients with cystic fibrosis in Boston and Toronto. *J Clin Epidemiol* 1988;**41**:583–91.

11 Walters S, Hodson ME, Britton J. 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.

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A full version of these guidelines will be available from the College in September.