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Review

European Cystic Fibrosis Society Standards of Care: Framework for the Cystic Fibrosis Centre

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Abstract

A significant increase in life expectancy in successive birth cohorts of people with cystic fibrosis (CF) is a result of more effective treatment for the disease. It is also now widely recognized that outcomes for patients cared for in specialist CF Centres are better than for those who are not. Key to the effectiveness of the specialist CF Centre is the multidisciplinary team (MDT), which should include consultants, clinical nurse specialist, microbiologist, physiotherapist, dietitian, pharmacist, clinical psychologist, social worker, clinical geneticist and allied healthcare professionals, all of whom should be experienced in CF care. Members of the MDT are also expected to keep up to date with developments in CF through continued professional development, attendance at conferences, auditing and involvement in research. Specialists CF Centres should also network with other Centres both nationally and internationally, and feed Centre data to registries in order to further the understanding of the disease. This paper provides a framework for the specialist CF Centre, including the organisation of the Centre and the individual roles of MDT members, as well as highlighting the value of CF organisations and disease registries.

Keywords: CF Centre; Multidisciplinary team; Continuing professional development

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1. Introduction

People with cystic fibrosis (CF) have complex care needs that demand specialist, medical and allied healthcare expertise. The life expectancy has increased significantly in successive patient birth cohorts [1] as a result of more effective treatments and crucially because most patients attend CF Centres in line with the demonstration that patients who attend CF Centres for their care have better well-being and lung function than those who do not [2,3]. Thus, the CF Centre has become the model of care for people with CF; patients should receive full care from the Centre or have local directed care supervised by the Centre [4–6].

The framework of the CF Centre is formed by a multi-disciplinary team (MDT), links with other medical and surgical specialties, the buildings and facilities, and the hardware and software that combined allow the MDT to provide a level of care that meets the complex medical challenges of this disease using effective diagnostics and holistic treatment programmes.

The MDT members are at the core of the CF Centre and should be supported with continuing professional development (CPD), audit and research. Each discipline should establish its own rigorous framework, to ensure that patients' needs related to their discipline are met. The CF Centre should have adequate resources (e.g. staffing, IT equipment) and an infrastructure (inpatient and outpatient facilities) that allow the MDT to provide a level of care that is in accordance with the European Cystic Fibrosis Society (ECFS) standards recommended in this document, ensuring a safe, cost-effective and high-quality service. It is recognized that this may not be immediately achievable throughout Europe, particularly in countries with a low gross domestic product. It is crucial that where these standards cannot be met, procedures are put in place to enable them to do so within the short- to mid-term future, and that the hospital management commits to supporting CF clinicians. Without proper resources, a Centre is at risk of providing uncoordinated and substandard care. A lack of homogeneity in CF care will impact on patient outcomes [7].

At present, access to specialist CF services across Europe is inconsistent. Qualifications, training and roles vary considerably. Clinical practice should where possible be evidence based and reflect current research findings, clinical guidelines and consensus views. CF specialist professionals should be appropriately trained, qualified and registered by the state/national health authorities and legally recognized to practice within that country. Specialists should practice within their professional code of conduct and competency. They have a responsibility to maintain, update and enhance their knowledge, skills, efficacy and expertise through a proactive approach to continuing professional development. Participation in uni- and multi-professional audit and research, benchmarking, external quality assessment schemes, service evaluation, development and improvement, both of the specialist service and its provision as a whole is essential. As a CF Centre may be the only facility in a city or region, national and international programmes to support CPD, benchmarking and service improvement are strongly encouraged.

The following sections describe the ECFS recommended standards for the individual specialties within the CF Centre.

2. Framework for the paediatric and adult Centres

Paediatric and adult CF Centres have many features in common, so that the requirements outlined below usually apply to both. As the health of children and adolescents continues to improve, the emphasis in paediatric care is on the prevention of disease progression. The morbidity and almost all of the mortality associated with CF have shifted to adults. Adult services should take into account the greater demands for inpatient provision and the higher prevalence of multisystem complications. Even before the transition from paediatric to adult care, it is most important that the two Centres work closely together. Regular meetings between the teams and shared protocols can smooth the transition process for the young adult and minimize the changes to their treatment. Effective communication between teams during this period is crucial to the success of the transition process.

Children with CF will have their care transferred to adult services around the time of their 17th/19th birthday. Children, and their families, should understand that they will transfer to an adult Centre at this age. The paediatrician is not trained or experienced to manage the emotional, social, or medical demands of the adult patient. In the adult population, the manifestations of this multisystem disease (e.g. CF-related diabetes, osteoporosis, renal and liver complications and atypical infection) are significantly more problematic. The adult physicians are also best placed to fully inform patients about the potential risks of pregnancy and are competent in the non-obstetric care of pregnant women with CF.

The young person with CF and his/her family must be involved in planning transfer at an early stage. The topic should be introduced when the diagnosis of CF is made and reinforced at appropriate intervals thereafter. Practical discussions should start at around 11 years of age in the context of educational, social and sexual conversations about growing up with a long-term condition. There must be close liaison with an agreed protocol for transition, and coordinators from both teams should be identified. The adolescent and his/her carers should have the opportunity to formally meet the adult team on more than one occasion. This is optimally achieved by having joint clinics with the adult team during the transition process. An opportunity to visit the adult facilities should be made available to patients and parents. Written information about each patient must be given to the adult team at the time of handover.

2.1. The Centre

Many of the required features are the same for adult and paediatric Centres. The CF Centre should have appropriate staff and facilities to provide comprehensive care and be capable of treating all CF-associated complications [4]. Patients should have direct access to the Centre 24 h a day.

In order to justify and maintain an appropriate level of expertise and experience, a specialist Centre should have a minimum of 100 adults or children with CF. In some circumstances, the geographical location of a specialist CF Centre, or a low disease frequency in certain populations, may mean that the number of patients seen by a Centre is less, but never below 50. Centres with fewer than 100 patients should be linked to a larger Centre until there are sufficient patients, experience and resources to run an independent service.

All patients with CF must have access to the Centre for routine and emergency care and advice. Patients should be reviewed regularly at a frequency appropriate to their individual needs. Routine appointments for people with stable disease should be every 2–3 months depending on the severity of their disease. Newly diagnosed infants should be seen more frequently (initially weekly).

All patients should have an annual assessment to ensure that, as a minimum, a full medical, dietetic, physiotherapy and psychosocial review is performed once a year and that all surveillance blood tests are requested. The report should be written by a consultant who should discuss the review findings with the patient/carers, and the treatment plan should be agreed by all.

2.2. The multidisciplinary team

A core MDT of trained and experienced CF specialist healthcare professionals should be responsible for patient care. The MDT should be of appropriate size for the clinic population and should include the following CF specialists and support staff:

- respiratory paediatrician/pulmonologist
- clinical microbiologist
- medical support from trainee(s)
- clinical nurse specialist
- specialist physiotherapist
- specialist dietitian
- clinical psychologist
- social worker
- pharmacist
- clinical geneticist
- secretarial support
- database coordinator.

There should be clear medical leadership of the MDT. The roles and responsibilities of all senior doctors in the team should be clearly defined.

The ECFS concurs with the staff numbers for paediatric and adult Centres recommended by the UK CF Trust (Tables 1 and 2) [6]. These numbers may vary according to health care organisation, geographical factors and local/regional variations in CF services. For example, the numbers of physiotherapists may vary according to the proportion of patients who self-administer intravenous antibiotics at home. Staffing numbers should reflect the model of shared care being used, taking into account time spent by staff from the specialist Centre in assessing and treating patients in a local hospital CF clinic. In addition, it is important that adequate cover is available for annual leave, study leave, and untoward events.

All members of the MDT must be registered with their relevant national health profession's council/body, and be a

Table 1
Whole-time equivalents per clinic size: full-time paediatric patients[a].

The MDT	50 patients	150 patients	≥250 patients[b]
Consultant 1	0.5	1	1
Consultant 2	0.3	0.5	1
Consultant 3	–	–	0.5
Medical trainees	0.8	1.5	2
Specialist nurse	2	3	4
Physiotherapist	2	3	4
Dietitian	0.5	1	1.5
Clinical psychologist	0.5	1	1.5
Social worker	0.5	1	1
Pharmacist	0.5	1	1
Secretary	0.5	1	2
Database coordinator	0.4	0.8	1

[a] Patients with CFTR-related disorders should not be counted.
[b] When clinics care for significantly more than 250 patients, additional consultants should be added to the multidisciplinary team (MDT) at a rate of approximately one additional consultant per extra 100 patients. Additional allied health professionals and support staff will also be required. There is likely to be a limit to the number of patients who can be cared for effectively in a CF Centre. This number will vary according to the facilities available in the hospital housing the Centre and the capacity of that hospital to support adequate staffing for the Centre. The MDT in individual Centres should review patient numbers annually and appreciate when resources are becoming stretched beyond the limit allowing care to be delivered to the standards recommended in guidelines. Paediatric patient numbers are likely to remain relatively stable but adult numbers are increasing every year. The need to establish a new adult Centre in any region must be considered proactively. Supply must precede, or coincide with, need.

member of their national or international CF special interest group. They must have specialist knowledge and be experienced in the care of children and/or adults with CF. They must maintain CPD through attendance at local study days and national and international CF conferences.

2.3. Access to other specialists

It is essential that there is access to other medical and surgical specialists as needed. Essential supporting disciplines and services include: gastroenterology and hepatology (with expertise to perform emergency endoscopic ligation of oesophageal varices); diabetes and endocrinology; ear, nose and throat surgery; cardiothoracic and general surgery; specialist anaesthesia and pain control; rheumatology; nephrology; obstetrics and gynaecology; psychiatry; intensive care; and interventional radiology (with expertise to perform emergency bronchial arterial embolization, and elective percutaneous ultrasound-directed gastrostomy).

Table 2
Whole-time equivalents per clinic size: full-time adult patients[a].

The MDT	100 patients	150 patients	≥250 patients[b]
Consultant 1	0.5	1	1
Consultant 2	0.3	0.5	1
Consultant 3	–	–	0.5
Staff grade/fellow	0.5	1	1
Specialist registrar	0.4	0.8	1
Specialist nurse	2	3	5
Physiotherapist	2	4	6
Dietitian	0.5	1	2
Clinical psychologist	0.5	1	2
Social worker	0.5	1	2
Pharmacist	0.5	1	1
Secretary	0.5	1	2
Database coordinator	0.4	0.8	1

[a] Patients with CFTR-related disorders should not be counted.
[b] See footnote in Table 1.

2.4. Centre infrastructure

The facilities at the CF Centre must be appropriate for all age groups. There should be sufficient capacity within the Centre for outpatients to be seen urgently whether this is within a clinic session, in a day-case unit or during a ward visit. The number of inpatient beds should be sufficient to allow non-urgent patients to be admitted within 7 days and urgent patients to be admitted within 24 h. The hospital ward nursing staff who are in contact with patients should have sufficient knowledge and experience of CF care. Finally, there should be resources and staffing available for home intravenous antibiotic administration supervised by trained outreach nurses.

2.5. Segregation

All CF Centres must have a clear policy for infection prevention and control, and facilities must allow for adequate patient segregation to prevent cross-infection. Patients should not share rooms, bathrooms or toilets during a hospital stay and should not be in contact with each other in waiting areas, such as in CF clinics, wards, the pharmacy and radiology departments.

2.6. Access to specialist investigations

Within the specialists CF Centre, there should be easy access to specialist investigations. These include biochemistry and haematology laboratories for routine tests as well as analysis of sweat and measurement of fat-soluble vitamin levels, aminoglycoside levels, and measures of glucose metabolism (including continuous glucose monitoring systems). The microbiology services should have the ability to process samples from people with CF and to reliably detect *Burkholderia* spp., non-tuberculous mycobacteria, and fungal infection. Molecular pathogen typing and immunology for allergic bronchopulmonary aspergillosis monitoring should also be available.

Physiology must include lung function measures (both ward and outpatient spirometry), pulse oximetry including overnight O_2/CO_2 monitoring, exercise testing and fitness-to-fly testing.

The radiology and nuclear medicine service should include computed tomography (CT) scanning, liver ultrasound, and dual energy X-ray absorptiometry (DXA scan) bone scanning. High-frequency pure tone audiometry and flexible bronchoscopy should also be available.

2.7. Miscellaneous

Facilities must exist for a parent/carer to stay with their child in hospital, and for a child to receive suitable education within the hospital if they cannot attend school due to illness. Related to these facilities should be access to appropriate play and/or recreation, with facilities for study.

The CF Centre should be committed to active participation in clinical and translational research, and encourage patient participation in clinical trials. Each Centre should aim to be part of the expanding European Clinical Trials Network (www.ecfs.eu/ctn).

2.8. European policy aspects of CF Centre care

CF is a rare disease (i.e. <1 in 2000), and as such belongs to the domain of several European Union (EU) policy initiatives relevant to research and healthcare. CF Centres, in compliance with the European Committee of Rare Disease Experts guidelines for Centres of Expertise for rare diseases, could operate within the frame of the newly established European Reference networks for rare diseases. As a rare disease, CF has a special status (Art. 54) in the Cross-Border Directive that facilitates exchange of expertise and eventually patients, and could thus address disparities in care. The development of CF Centre care in EU member states is supported by the EU Council Recommendation on action in the field of rare diseases (EC 2009/C 151/02). The introduction of orphan medical products into CF care also falls into such policy initiatives.

3. Framework for the specialist doctor

3.1. The CF consultant

The Consultant who works in a specialist CF Centre should have received accredited training in paediatric or adult CF care, usually within the context of respiratory specialization. The knowledge and skills that the CF Consultant should have acquired are detailed below [8].

3.1.1. Knowledge

The CF Consultant should have knowledge of the epidemiology and pathophysiology of CF and the aetiology of respiratory and non-respiratory manifestations and complications of CF, including massive haemoptysis, pneumothorax, respiratory failure, gastrointestinal disease, diabetes, problems of fertility and pregnancy (adult care) and psychosocial problems. The relevant investigations required, including microbiological investigations, non-invasive imaging modalities such as chest X-ray, CT imaging scans, should be familiar to the Consultant. The CF Consultant also needs to be familiar with the pharmacology of inhaled, oral and systemic drugs that are prescribed to patients and with the varied interventions employed by physiotherapists. The nutritional requirements of individual patients should be monitored and enteral tube feeding initiated when appropriate.

Finally, the CF Consultant should know the indications for lung transplantation and be experienced in discussing this option with patients and carers.

3.1.2. Skills

CF Consultants should be able to apply the above knowledge to the management of respiratory and non-respiratory manifestations and complications. They should also be able to interpret the results from sputum microbiology tests and evaluate the functional status of patients. CF Consultants should also have good communication skills so that they can educate their patients and carers as the disease evolves.

The CF Consultant's job plan should include adequate time allocation for CF patients, both for clinical tasks as well as managerial duties. This must include the capacity to maintain his/her own CPD in CF, which should involve attendance at national or international respiratory/CF meetings. In order to keep up to date with advances in treatment and research, the CF Consultant should spend a minimum of 50% of his/her working time dedicated to CF issues.

3.2. The clinical lead

The Director of the specialist CF Centre is usually the Clinical Lead and will be expected to lead the CF MDT. He/she should act as a link between clinical experts and the hospital management. The Clinical Lead/Centre Director will be expected to head the team and to ensure that: the needs of its members are met in terms of professional development and adequate support; that opportunities for attendance at national and international CF meetings are available; and that research is encouraged. The Director should also ensure that a team approach is maintained and that all members of the CF MDT have the opportunity to have their observations and opinions considered in patient management.

It is essential that the Director understands the financial framework underpinning the country's healthcare system in order to develop and protect the financial support needed for the CF service. The Director should lead on staff recruitment, aiming to realize the human resource numbers as recommended in these Standards of Care. He/she should ensure that CF MDT meetings are held weekly, that Centre outcome measures are audited and that the results are reported back to the team so that standards of care are improved. In order to achieve the latter, the Director needs to oversee accurate data collection and documentation and that transfer of these data to the national and European registries is carried out.

In some Centres there may be co-leads/directors of the service. Clear definition of responsibility and communication is essential in this situation.

4. Framework for specialist nursing care

4.1. The role of the CF Clinical Nurse Specialist

The role of the CF Clinical Nurse Specialist should include [6]:

- education, advocacy and psychosocial support, particularly at important times such as:
  - notification of a screening result and diagnosis
  - first admission to hospital

- first course of intravenous antibiotics
- a second diagnosis (e.g. CF-related diabetes)
- transition from paediatric to adult care
- reproductive issues, pre- and postnatal care
- transplant and end-of-life issues

- provision of support and education at home, particularly for home intravenous antibiotic therapy, nebulizer therapy, enteral feeding and non-invasive ventilation
- provision of education to others about CF, including nurseries, schools, places of higher education and work places
- acting as a link between the patient and family, primary care, community services and hospital
- acting as a resource for training and education of other professionals involved in CF care.

4.2. Access, availability and facilities

There should be an adequate number of Clinical Nurse Specialists with expert knowledge of CF in the MDT [9]. The CF Clinical Nurse Specialist should deliver skilled support, advice and care directly to the patient and family wherever it is needed, both when attending hospital and at home. The service will vary according to differing patient populations, their needs and requirements. The role of the CF Clinical Nurse Specialist should continuously develop to meet the needs of the local CF population [4].

CF Clinical Nurse Specialists need sufficient time, office space, computer/printer and financial support in order to be able to provide a reliable service. They should stay in regular contact with patients and families in between clinic visits and therefore need access to technology such as email, phone and SMS texting.

4.3. Key stages for delivery of care

4.3.1. Diagnosis

Diagnosis through newborn screening is now common in many countries. The CF Clinical Nurse Specialist plays an active role in talking to parents at diagnosis and providing ongoing support and continuing education following the initial discussion. Where screening is not available, the CF Clinical Nurse Specialist plays a similar role offering support, advice and education, which has to be individualized at a level and frequency to meet differing needs, whether diagnosis is within the first year of life, in older children or during adulthood. Contact between the CF Clinical Nurse Specialist and the patient/parent is therefore essential, whether this is in hospital, through home visiting, or via email or telephone.

4.3.2. Pre-school

For many, after coming to terms with the diagnosis and learning how to carry out treatment regimens while adjusting back to family life, the early years can seem almost normal. However, there are a few areas where the CF Clinical Nurse Specialist can provide education, practical advice and psychosocial support [10], such as: administering medication; nutrition; adjusting pancreatic enzyme replacement therapy (judging the correct amount or offering advice when the child refuses to take the enzymes) in conjunction with the CF Dietitian; recognizing chest infections and making decisions about when to ask for advice or to start treatment; managing airway clearance and exercise in conjunction with the CF Physiotherapist; starting nursery; dealing with siblings; planning further children.

4.3.3. School age

When a child starts school it can be a traumatic experience for any parent. When the child has CF, parental anxiety about loss of control is likely. Many CF Clinical Nurse Specialists will visit the school (with parental permission) to educate and prepare teachers for managing CF in areas such as: maintaining good nutrition at school; administration of pancreatic enzymes and other medication (e.g. nebulizers/inhalers/oral); liaising with the school nurse; facilitating time off for hospital visits/admissions; dealing with the child's growing independence; advising on issues surrounding non-adherence, especially eating and airway clearance. The CF Clinical Nurse Specialist can help parents at this time, particularly with outreach contact as it gives parents time away from the clinical setting and allows them to discuss their anxieties in a safe and familiar environment.

Most school-age children with CF are relatively well and take part in all academic, sporting and social activities provided by the school. Occasionally, extra treatment is necessary. Supporting treatments such as intravenous therapy or enteral feeding in the home often allows children to continue attending school. The provision of an outreach service can help, as routine checks (such as spirometry) can be performed by the CF Clinical Nurse Specialist, and problems can be identified early [11,12].

4.3.4. Adolescence

Adolescents with CF go through the same physical and emotional changes and have the same expectations as their healthy peers, irrespective of the severity of lung disease [13]. The CF Clinical Nurse Specialist should be able to have open and honest discussions about issues such as: recreational drug use and the effects on CF; sexuality, safe sex and contraception; fertility and pregnancy; further education and employment; body image and self-esteem; adherence to treatment regimens; relationships with parents; promotion of self-care, adherence and responsibility; accurate information about their disease and treatment.

CF Clinical Nurse Specialists need to be sensitive and honest when giving information to young people with CF [14–16]. Much of the information they (and their families) receive is from peers, the media and the Internet. Information given by the CF Clinical Nurse Specialist must therefore be correct and up to date.

4.3.5. Transition from paediatric to adult care

All children with CF should move from paediatric to adult care. The importance of getting this transition process right is widely recognized [17,18]. Transition from paediatric to adult care happens at a time when the young person with CF is moving into adulthood in other areas of their life, such as further education or employment, forming relationships and

taking more responsibility for their own lifestyle. Transition can therefore be difficult for many reasons. CF Clinical Nurse Specialists involved with the transition process need to be aware of the many barriers that can prevent this process being successful [19,20]. Both the paediatric and adult CF Clinical Nurse Specialists play an important role in ensuring a successful transition and will manage details such as: patient and parent involvement in decision making; clear communication between paediatric and adult CF MDTs; appropriate transition clinics involving the MDT; ensuring attendance at the adult clinic with appropriate follow-up.

Young people may find that their first admission to the adult Centre is to an unfamiliar ward where they do not know the staff. For these individuals and their families, the first inpatient admission requires an increase in awareness and sensitivity from the ward staff and further CF Clinical Nurse Specialist support to both the patient and the family. The CF Clinical Nurse Specialist should liaise between the ward and the CF MDT.

4.4. Adult issues

CF Clinical Nurse Specialists play a vital role in helping adults maintain a balance between adhering to treatment and their lifestyle, and recognize the need to help individuals adapt treatment regimens to suit them. This will include: educating employers and work colleagues; liaison with government agencies and the work place to ensure maximum support (financial and practical) to enable patients to stay employed or to re-train; advocacy on a patient's behalf with local social services; educating and liaising with family doctors and local pharmacists; negotiating easier access to classes at school or university; increasingly, CF Clinical Nurse Specialists will work in collaboration with the family doctor, social services and the CF team to support patients caring for their ageing parents; providing education, counselling and support around reproductive issues for both men and women with CF; practical and emotional support throughout the neonatal and postnatal periods.

Complications occur more commonly in older patients with CF [21–23]. An outreach service led by a CF Clinical Nurse Specialist may have to manage complex medication regimens and organise care to help maintain a lifestyle/treatment balance.

4.5. Transplantation and end-of-life issues

When admissions become more frequent, longer in duration and the burden of treatment increases, patients or their families may wish to raise the issue of lung transplantation. Early discussion with the team raises questions and concerns for both patients and their families. The CF Clinical Nurse Specialist's role as advocate and educator for the patient is vital in this decision process.

CF continues to be life limiting. Death in childhood, although uncommon, does occur. Unlike other chronic diseases, the end stages of CF can be difficult to recognize. Patients often need opportunities to discuss their fears and anxieties but may feel uncomfortable or protective talking about these issues with their family for fear of upsetting them or 'letting them down'. Advocacy allows the CF Clinical Nurse Specialist to facilitate discussion between the patient and family. Early discussion about an individual's wishes for the terminal stage of their disease is essential to aid appropriate care planning. Issues that may be raised include transplantation, wills, funeral arrangements, writing letters or diaries to the family and where they would like to be when they die [24,25].

The CF Clinical Nurse Specialist plays a key role in providing individual emotional support for parents/partners. Although some families are willing to return to the hospital, many find this difficult. Visiting the family at home allows bereavement support to be offered in a safe and comfortable environment. Home visiting also allows other family members, siblings or grandparents for example, to receive support.

4.6. Core competencies, qualifications and professional development

4.6.1. Core competencies

The CF Clinical Nurse Specialist should be competent in the following key areas.

- Clinical practice
  - Diagnostic and assessment skills
  - Treatment skills
  - Recognizing and monitoring change
  - Facilitating programmes of care
  - Clinical research and audit
- Education
  - Knowledge of CF and associated issues
  - Evidence-based practice
  - Teaching and training: patients, carers, other healthcare professionals
- Communication
  - Patients and carers
  - MDTs
  - Liaison with clinical, social, educational, employment and other lay agencies
- Support and advocacy
  - Social care
  - Advocacy
  - Counselling skills
  - Legal and ethical issues.

4.6.2. Qualifications and professional development

The CF Clinical Nurse Specialists must be registered as licensed practitioners in their country. They should also have specialist knowledge and be experienced in the care of children (including specific paediatric training) and/or adults with CF. The CF Clinical Nurse Specialist must contribute to research in all areas of CF, either through developing individual projects or participating in research carried out by the CF MDT, and maintain CPD through attendance at courses and conferences.

CF is a demanding disease to manage for the patient, family and the CF MDT. The CF Clinical Nurse Specialist must act as a link between the patient and family, primary and community services, and the hospital. The CF Clinical Nurse Specialist has a responsibility to ensure that every patient receives appropriate

care for their individual needs. Patients should receive lifelong support and good-quality treatment through the coordination of care between patient and family, community services and hospital, both practically and through support and advice.

5. Framework for physiotherapy care

The specialist CF Physiotherapist should take the lead in providing high-quality treatment of airway clearance, physical exercise and inhalation therapy. Physiotherapy programmes in CF care are primarily preventive, and regular input is required from the time of diagnosis. The aims of therapy are to maintain ventilation in all parts of the lungs, to postpone progression of pulmonary disease, to stimulate establishment and retention of normal physical capacity and to avoid pain and musculoskeletal complications due to pulmonary or bone disease [26]. The CF Physiotherapist should also develop strategies for the management of complications or co-morbidities experienced by the ageing patient and should optimize the respiratory physiotherapy programme, which includes highly technical equipment, non-invasive ventilation and physical exercise with oxygen supplementation. Physical rehabilitation is essential for patients on a transplant waiting list.

5.1. The role of the CF physiotherapist

The CF Physiotherapist should be available for regular contact and assessment of the patient for treatment, lung function testing, physical surveillance and therapy evaluation. The frequency of this will vary according to the patient's age and clinical status but as a minimum should happen at every routine outpatient clinic and daily during each hospitalization (including when patients are admitted under the care of other specialists and to intensive care). A more extensive assessment should take place annually.

5.2. Regular assessment and therapy

Regular lung assessments by the CF Physiotherapist should include lung function test data, respiratory signs, degree of dyspnoea, oxygenation cough characteristics and questioning about activity of everyday life. All interventions should be tailored to the individual, with consideration of their age, severity of disease, physical side-effects or complications, and social and domestic circumstances.

5.2.1. Inhalation therapy

As the CF Physiotherapists are responsible for inhalation therapy they should be familiar with techniques, equipment provision and appropriate maintenance of devices. There is a need for consideration of timing of inhalations in relation to airway clearance as there may be a positive interdependence between the two. Education of patients in appropriate inhalation techniques is essential for optimal deposition of inhaled drugs. The CF Physiotherapist should be familiar with the appropriate nebulizer systems proven to be safe and effective in the delivery of the medications prescribed. Cleaning and maintenance of the whole nebulizer system are essential to ensure that medications are delivered optimally and safely [26,27].

5.2.2. Airway clearance therapy

Physiotherapists are responsible for airway clearance therapy. This involves knowledge and experience of the full range of techniques available and immediate evaluation of therapy, for example by expiratory sounds, sputum volume and characteristics and by ability to control cough. Alternative physiotherapy techniques should be recognized and considered for individual patients. There are a variety of effective airway clearance techniques that allow patient independence. These are based on sound physiological concepts and allow the CF Physiotherapist to individualize treatment programmes [28]. There is no standard airway clearance regimen or conclusive evidence to promote one technique over another [6,26,29–37].

5.2.3. Postural and musculoskeletal assessment

Assessment of postural and musculoskeletal function is carried out to evaluate therapy. Physical exercises aimed at the maintenance of good posture and chest mobility should be included in the treatment from the beginning. As with all physiotherapy interventions the exercises should be individually tailored to each patient [26,38,39].

5.2.4. Exercise capacity

Exercise capacity and the opportunities for exercise prescription for the person with CF include any pre-transplantation preparation. Reduction in exercise capacity is associated with a decline in respiratory function and survival [40,41]. Physical exercise has been reported to improve lung function and decrease habitual inactivity in children with CF [42]. The CF Physiotherapist should perform regular exercise testing with a frequency dictated by disease progression, and in cases of specific needs like transplantation assessment or evaluation of a treatment. Care should be taken when prescribing exercise activities for patients with advanced disease, particularly when they may also experience haemoptysis, exercise-induced desaturation requiring supplementary oxygenation, pulmonary hypertension, cor pulmonale, joint arthropathies and other co-morbidities [26]. The CF Physiotherapist should also assess supplementary oxygen needs, for exercise or ambulation [26,43].

5.2.5. Non-invasive ventilation

It is recognized that non-invasive ventilation is a useful therapeutic adjunct to support airway clearance therapy and reduce the work of breathing and fatigue experienced by the severely ill patients during treatment. Non-invasive ventilation may also be useful during exercise management to decrease breathlessness, improve oxygenation and, consequently, to maintain or improve exercise tolerance [44–46]. Additionally, non-invasive ventilation is implemented to facilitate optimal function in patients with end-stage disease and possibly as a bridge to transplantation [45].

5.2.6. Other considerations and assessments

Surveillance regarding the incidence of urinary and faecal incontinence should also be the responsibility of the CF Physiotherapist. A sensitive and open approach with early recognition of symptoms should be adopted; questioning can occur as early as 10 years of age [47,48].

The CF Physiotherapist should also be responsible for:

- management of associated complications and issues with adherence while continuing to promote independence that is age appropriate
- appropriate inhalation and airway clearance therapy and physical exercise programmes during pregnancy [49]
- ensuring the appropriate maintenance and function of equipment provided for therapy and nebulization
- the education of patients, carers, teachers and local physiotherapists; the physiotherapists should work closely with the other professionals for the benefit of the patients' holistic care
- palliative care, especially in relation to relieving dyspnoea in the terminally ill, and advising on when to withdraw non-invasive ventilation.

5.3. Service provision

When patients are resident in hospital for the treatment of an exacerbation or for routine management they should be reviewed by the CF Physiotherapist within 24 h of admission, and a treatment plan focusing on airway clearance, inhalation therapy and exercise tolerance should be implemented. The CF Physiotherapist should have a comprehensive knowledge of all techniques, respiratory pathophysiology, the rationale for alternative approaches and any associated contraindications to the treatment techniques available [28]. CF physiotherapy services should be available 7 days a week, with an out-of-hours physiotherapy service available for those patients who may deteriorate overnight.

5.4. Professional development, research and availability

Education, clinical audit, research and contribution to a CF registry should be pursued. CPD is integral to the work of the CF Physiotherapist who should maintain and increase specialist knowledge by attendance at relevant postgraduate courses, lectures, and national and international conferences. They should preferably be an active member of their national CF physiotherapy group and be available to represent physiotherapy interests for their country at meetings of the International Physiotherapy Group for CF (IPG/CF) [50]. The CF Physiotherapist should contribute to research, development and evaluation by performing audits, participating in multicentre studies and contributing data to registries. They should collect annual data in order to evaluate their care [50,51].

6. Framework for dietetic care

A normal nutritional status is positively associated with better lung function [52,53]. Healthy body weight, height and BMI are positively associated with survival [53–55]. Ensuring normal growth in children and adolescents and maintaining a normal BMI in adults is essential.

Specialist CF Dietitians have an integral role to play in patient management and have overall responsibility for the delivery of expert nutritional care. They should be actively involved in the nutritional training, education, development and support of other healthcare professionals involved in CF care. Dietetic intervention should be both proactive and reactive, evolving in response to the needs of each individual patient. It is essential that the specialist CF Dietitian has expertise in managing the complex nutritional challenges and rare complications of the disease.

6.1. The role of the specialist CF dietitian

The specialist CF Dietitian should take the lead in providing high-quality treatment and care to ensure optimal nutritional status, including nutritional screening and surveillance, and regular patient assessment with review of all aspects of nutrition and gastrointestinal status. The frequency and type of assessment will vary with age and clinical status.

The specialist CF Dietitian should advise and educate patients and carers about the principles of nutritional management in CF to enable them to meet their nutritional needs and achieve optimal growth, weight and body composition. Advice may be required on the management of pancreatic insufficiency, fat-soluble vitamin deficiency, altered gastric motility, gastro-oesophageal reflux, impaired glucose tolerance/diabetes, reduced bone mineral density, renal disease and liver disease.

Age-specific individualized advice should be offered. This advice should consider psychosocial barriers (especially during adolescence) and be supported by written literature, visual, audio and/or audiovisual aids, computerized learning packages and 'apps'. This is an ongoing and evolving process and must take into account the key times that may require more intensive dietetic intervention and support, such as diagnosis, early infancy, initiation of pancreatic enzyme replacement therapy, weaning, adolescence and self-management, pregnancy, initiation of enteral tube feeding, diagnosis of CF-related diabetes, transplantation and end-of-life care.

It is important to remember that patients diagnosed later in life tend to present atypically and have unique educational requirements. Improving adherence to the many prescribed nutritional therapies is a key challenge. The specialist CF Dietitian should provide a collaborative approach to increase motivation to change and support patients' efforts to change. This is based on providing information and facilitating open discussion. It is important to address emotional and perceptual barriers to adherence, e.g. a reluctance for females to gain weight during adolescence.

6.2. Clinical governance, research and quality framework

The specialist CF Dietitian should be a member of, and an active participant in, specialist interest groups locally, nationally and internationally, (e.g. European Cystic Fibrosis Nutrition Group) in order to support their practice. They should be

encouraged to be an Allied Healthcare Professional Member of the European Cystic Fibrosis Society.

6.3. Dietary assessment

6.3.1. The annual assessment

Dietetic staffing should allow for a structured annual assessment of dietary intake and nutrition. Formal assessment of dietary intake using a written diet and enzyme diary should be targeted at selected individuals only; in large CF Centres, such an exercise is unsustainable if applied to all patients and is unlikely to provide additional information for patients with stable nutritional status. The annual evaluation should address all aspects of nutritional status assessment, nutritional intake, pancreatic enzyme replacement therapy, and the management of nutritional and metabolic complications. The annual assessment will help to provide the framework for future care planning and anticipatory guidance.

The reader is referred to the document "European Cystic Fibrosis Society Standards of Care: Best Practice guidelines – Optimal Nutrition and Management of Metabolic Complications of Cystic Fibrosis" for details on the assessment of the following:

- pancreatic status and absorption
- growth and nutritional status
- bone mineral density, and
- glycaemic status.

6.4. Service provision framework

Traditionally the framework for service provision is divided into:

- inpatient care
- outpatient care
- home care
- shared care
- transitional care
- annual review.

All patients should have access to a specialist CF Dietitian at all of these times. The same dietitians should provide inpatient and outpatient advice to ensure continuity of care and to prevent the important minutiae of care being overlooked. Advances in telecommunications and technology allow opportunities to re-evaluate service delivery.

6.5. Key considerations of service provision

6.5.1. Inpatients

A clear discharge plan and follow-up arrangements should be provided for patients treated in hospital, especially for those requiring ongoing nutritional support.

6.5.2. Home treatment

For those being treated with intravenous antibiotic therapy at home, there should be access to a specialist CF Dietitian at the start and end of this treatment, with ongoing nutritional support provided remotely (e.g. by telephone, telemedicine) or via the CF Clinical Nurse Specialist. There should be clear channels of communication between the CF Clinical Nurse Specialist and the specialist CF Dietitian.

6.5.3. Outpatients with CF-related diabetes

Outpatients with CF-related diabetes should have access to a specialist CF Dietitian with experience in the management of this CF complication.

6.5.4. Shared care

In general, due to the complexity of the dietetic needs of adults with CF, shared care is not appropriate. In paediatric Centres there should be:

- protocols for the delivery of care and lines of responsibility for nutritional management
- an identified dietitian within the shared care hospital who will liaise with the specialist CF Dietitian at the Centre
- review of all patients by the Centre's specialist CF Dietitian at least twice a year.

6.5.5. Transition

The paediatric and adult specialist CF Dietitians should work together to promote autonomy, facilitate self-management and ensure a smooth transition. At the time of transfer, the paediatric specialist CF Dietitian should provide a clear and concise summary of the nutritional management and challenges for each patient. Where possible the paediatric specialist CF Dietitian should provide a written treatment plan.

7. Framework for microbiology

A Clinical Microbiologist with specialist knowledge of CF infection should be part of the CF MDT. This individual may be a medically trained clinical microbiologist/infectious disease specialist; alternatively, a clinical scientist with relevant knowledge and experience may be able to undertake this role. The CF Clinical Microbiologist should work closely with the microbiology laboratory providing diagnostic services for the CF MDT and also with the local infection control and prevention team.

In order to provide support to the CF MDT for the diagnosis and treatment of infection, the CF Clinical Microbiologist needs to know about the range of infections in CF. In particular, they need to be aware of the role of unusual micro-organisms, the risk of cross infection and the impact of long-term chronic infection on microbiological laboratory testing and treatment. In addition to a good basic knowledge, the CF Clinical Microbiologist should have evidence of CPD in CF microbiology and attend specialist CF meetings and conferences.

7.1. The role of the CF Clinical Microbiologist

The CF Clinical Microbiologist should ensure that appropriate laboratory microbiology provision is in place. The individual may be part of the management of the laboratory. Alternatively,

these services may be provided through an external contract, in which case the CF Clinical Microbiologist should be involved with setting the terms of the contract and act as an advocate for the CF Centre.

The CF Clinical Microbiologist should advise on the diagnosis and treatment of infection including the monitoring of antibiotics. This may be achieved by attendance at the CF MDT meetings. The CF Clinical Microbiologist should also act as an advisor on infection prevention and control in the CF Centre. This may be delegated to the designated infection control doctor if such a position exists.

7.2. Overview of laboratory services

The CF Clinical Microbiologist should ensure that the full range of microbiology laboratory tests needed for the CF Centre is available and that the laboratory service provided is based on published guidelines [56–58]. The laboratory should be fully accredited by a recognized national scheme for clinical microbiology and should participate in external quality assurance, which includes CF-associated pathogens. There should be provision to send relevant samples to a reference laboratory specializing in CF microbiology when required.

The laboratory should provide accurate and timely results to the CF Centre with an agreed system for notifying urgent and important results. The technical staff in the laboratory should have sufficient expertise and knowledge to deal with the complex microbiology of CF infections.

There should be a framework for recording and investigating errors and other incidents, with evidence of how the lessons learned are used to inform a programme of service improvement.

The service should be regularly audited. Examples of audits are the turn-around time (i.e. the time between the receipt of the sample in the laboratory and the time when the result is available to the CF MDT), the accuracy of identification and susceptibility testing, and the appropriate and prompt communication of urgent results to the CF MDT.

7.3. Clinical microbiology services and the CF MDT

The following items should be agreed between the CF MDT and the clinical microbiology service.

- Which respiratory samples should be taken and how should they be processed (e.g. sputum, bronchoalveolar lavage, cough swab or a pharyngeal swab).
- Which samples should be taken for the diagnosis of an infected intravascular line.
- Diagnosis of other infections including infections of the gut (e.g. enteric viruses, when and how to test for toxigenic *Clostridium difficile*)
- The level of identification of micro-organisms (e.g. genus, species, subtype) required in individual cases. This may include a discussion on the tests that can be performed in a local laboratory and what may need to be referred to a specialist laboratory with more advanced testing methodology (e.g. confirmation of first infection with *Burkholderia* spp. with accurate species identification).
- Typing methods and frequency of typing (i.e. how often the CF MDT should send samples for routine surveillance and when additional typing should be done due to suspicion of cross infection)
- Measurement of antipseudomonal antibodies where appropriate
- Provision of diagnostic testing for fungal and mycobacterial infection together with level of identification and role of typing
- Susceptibility testing — agreement on the antibiotics to be tested and when susceptibility testing is helpful
- Virology services should include rapid identification of highly pathogenic viruses that may spread between patients — both familiar (e.g. influenza virus) and emerging viral pathogens (e.g. SARS, MERS coronavirus).
- Which results need to be phoned urgently to the CF MDT (e.g. first growth of *Pseudomonas aeruginosa*, new isolation of *Burkholderia cepacia complex* and other *Burkholderia* species, MRSA, possible *Mycobacteria* seen in sputum).
- Advice on infection prevention and control.

In addition, a robust framework for communication between the microbiology services and the CF MDT should be agreed (e.g. telephone contact, ward rounds to review patients, participation in MDT meetings).

7.4. Clinical advice on treatment of infection

The CF Clinical Microbiologists should work with the CF MDT to draw up guidelines for the use of antimicrobials, including the selection of treatment for clearance of new infections, therapy for acute exacerbation and long-term suppressive antibiotics. The aim is to reduce morbidity and hospital admissions and to use antibiotics responsibly in order to limit the development of resistance.

There must be provision of therapeutic drug monitoring of antibiotics. The CF Clinical Microbiologist should ensure that guidelines and advice are available on the maintenance of optimum antibiotic levels in the patient in order to promote effective treatment while minimizing side-effects.

7.5. Infection prevention and control

The CF Clinical Microbiologist should work with the CF MDT and the local infection control team to develop a local infection control and prevention policy and procedures in line with expert national and international guidelines [59–63]. This policy should include:

- how patients with transmissible infections are managed, both in the community and in hospital, in order to prevent the spread of infection
- surveillance for transmissible infection (e.g. how often to screen and which samples to send to the laboratory)

- antimicrobial treatment to clear carriage of potentially transmissible micro-organisms
- guidelines for staff with infections
- the investigation of outbreaks
- the provision of facilities for the CF Centre and the outpatient department — this should include the cleaning and maintenance of equipment and involvement in any plans for refurbishment or re-build of the department.

7.6. Role in clinical research and data collection

CF Clinical Microbiologists can have an active role in CF clinical research. They may be involved with the design of innovative research but also have a role in the provision of reliable and accurate laboratory support for clinical studies. They can also help to ensure that accurate microbiological results are available for national and international database collection.

8. Framework for medicines management

Optimal care of people with CF requires complex multidrug treatment plans. These drugs may be administered by oral, intravenous and inhaled routes. Adverse effects and drug interactions are common.

Many of the drugs are expensive and require specialist assessment and instruction on optimal administration. Adherence is a major challenge for patients and parents/carers. Non-adherence is associated with poor outcomes. CF Centres must have an effective medicines management programme to support patients in optimizing their therapy. The CF Clinical Pharmacist is pivotal in this process [64]. In European countries, decentralized clinical services, with a pharmacist working in the ward at least 50% of his/her time, or with pharmacists visiting wards daily, are not very common [65]. Only two countries, UK and Ireland, have developed these clinical services to a significant extent [65].

8.1. The role of the CF Clinical Pharmacist

CF Clinical Pharmacists have a central role in managing medicines effectively [66]. The overall goal of clinical pharmacy activities is to promote the correct and appropriate use of medicinal products and devices [67]. These activities aim to:

- maximize the clinical effect of medicines (i.e. using the most effective treatment for each patient)
- minimize the risk of treatment-induced adverse events (i.e. monitoring the therapy course and the patient's compliance with therapy)
- optimize the expenditures for pharmacological treatments borne by the national healthcare systems and by the patients.

The principle objective of the service provided by the CF Clinical Pharmacist is to provide patient-focused pharmaceutical care, defined as the responsible provision of medication to achieve definite outcomes that improve patients' quality of life and long-term survival. The service is the process through which the pharmacist cooperates with a patient and other healthcare professional in designing, implementing and monitoring a therapeutic plan to produce these specific health outcomes [68].

Effective provision of a clinical pharmacy service to the CF Centre relies on the knowledge and skills of a CF Clinical Pharmacist and the quality of various support services, such as a medicine information service with experience in the problems of CF and paediatrics (if applicable), and the necessary procurement and distribution services that can provide an efficient medicine supply service for inpatients. A dispensing service should also be provided as required. Access to an on-call service for the supply of urgent medication, information and advice for inpatient care, and an aseptic dispensing service for the preparation of intravenous antibiotics including complex desensitization regimens should also be available.

The CF Clinical Pharmacist should:

- dispense medications to inpatients or outpatients as required in their institution
- attend CF wards rounds and CF MDT meetings
- support and provide information to other pharmacists in the department who may not be familiar with CF
- liaise with paediatric and adult Centres during transition of care and transfer of patients
- support and provide information to pharmacists working in primary care and other hospitals
- maintain CPD through appropriate study and attendance at relevant study days, and at national and international conferences
- network with other CF Pharmacists for advice and CPD.

8.2. Pharmaceutical care practice for CF Clinical Pharmacists

8.2.1. Managing formularies, clinical guidelines and treatment protocols

The CF Clinical Pharmacist should assist in the completion of formulary applications to ensure that the appropriate medicines are introduced into clinical practice. They should also assist in the development and support of homecare services, such as home intravenous antibiotics, and manage and monitor the delivery of medication in this setting.

Effective communication should exist between the CF Clinical Pharmacist and other members of the CF MDT. As for all clinical professionals in the CF MDT, the Clinical Pharmacist should participate in CPD and attend CF conferences and relevant study days. They should also contribute to education and training of other healthcare professionals, including those working in primary care, as appropriate. The CF Clinical Pharmacist should act as an advisor on the legal and ethical responsibilities of using medicines, including sourcing and administration of unlicensed and off-label medicines. Any problems with medication supply should be resolved by the CF Clinical Pharmacist and communicated to the CF MDT.

The CF Clinical Pharmacist may be required to collaborate with CF research and development and assist in the completion of individual funding requests or exceptional case requests for

the supply of specific medications for individual patients where no such mechanism exists to currently fund that treatment.

8.2.2. Medication reconciliation/history taking

The CF Clinical Pharmacists is responsible for medicines reconciliation at admission/transfer from other institutions and on discharge, including alternative over-the-counter medications, trial medications and medications used for other conditions. They should ensure that an accurate history is recorded, including previous allergic reactions/adverse drug reactions.

8.2.3. Prescription monitoring and medication review service

In the monitoring and review of patient medication, the CF Clinical Pharmacists should ensure that medication and the formulation are appropriate for the patient, oversee extended prescribing for allied healthcare professionals including other pharmacists, and check for drug interactions. The CF Clinical Pharmacist is also responsible for ensuring that prescriptions are complete, unambiguous and legal, and for detecting potential medication errors.

8.2.4. Identifying patient and medication risk factors

It is the CF Clinical Pharmacist's responsibility to ensure that patient characteristics, including age, pregnancy or breast feeding, and organ dysfunction are taken into account when medicines are prescribed, and to check the response to previous and current medication. The use of non-drug and complementary therapies should also be taken into account when managing the patient's medication.

8.2.5. Preventing, detecting and reporting adverse drug reactions

The CF Clinical Pharmacist needs to document and report all reactions to newer medications and serious reactions to established medications to the appropriate national body. This includes documentation of individual toxicity/allergies/hypersensitivity reactions and contraindications, and monitoring for any adverse drug reactions. The appropriate use, storage and disposal of medicines should be ensured in order to minimize adverse events.

8.2.6. Individualizing drug and dosage requirements

The CF Clinical Pharmacist should aim, whenever possible, to maximize the therapeutic potential and minimize the adverse effects of medicines. Therapeutic drug monitoring of specific medicines (e.g. aminoglycosides, azoles) according to an individual's pharmacokinetic variables and monitoring and reviewing the outcome of an individual's need for medication are also required. While optimizing the use of medicines the CF Clinical Pharmacist also needs to take into account the patient's wishes and lifestyle.

The CF Clinical Pharmacist needs to keep up to date with newly available medications and therapies (e.g. new nebulized antimicrobials), and find a place for them in the treatment regimen.

8.2.7. Educating and counselling patients and carers

The CF Clinical Pharmacist has an important role to play in providing appropriate patient education and counselling to ensure the safe and effective use of medicines. This may include patient information leaflets about medicines and other appropriate methods of improving adherence to treatment. Pharmacists should also agree an informed plan with a patient/carer to achieve the best possible concordance with medication.

8.2.8. Evaluating medicine use

The CF Clinical Pharmacist's non-clinical responsibilities will include financial reporting to the CF MDT, hospital management, and other authorities, as appropriate, on CF medication usage. They should audit treatment guidelines, new therapies and homecare services.

9. Framework for psychosocial care

Living with CF provides many challenges for patients and their families. The CF Centre should provide adequate psychosocial care and support to help the person with CF and the families meet these challenges. In order to deliver optimal care CF Centres need a multidisciplinary framework to include access to psychosocial professionals throughout the patient's life. The core psychosocial professionals available should be a Clinical Psychologist and a Social Worker, though variations on these professions are acceptable provided core competencies are met (see below). Psychosocial professionals should be proficient in the following areas: working with children, families and adults according to the needs of the specific CF Centre; working with patients presenting with a range of clinical severity; and delivering care in all CF settings including outpatients, inpatients, community and residential care.

The variability of patient needs and availability of expertise in CF Centres prevent the formulation of a single programme. Psychosocial care should be provided within the context of the patients' development: from infancy to toddler, childhood, adolescence, young adulthood, adult life and old age. In each stage there are age-related themes as well as CF themes [69,70] (Table 3). Key stages include the time around diagnosis [71], the transition to adult care [19,72–76] and the transition to end-of-life/transplantation care [77,78]. The disease trajectory itself also necessitates times of increased psychosocial support, for example, at diagnosis, first *P. aeruginosa* infection, first inpatient admission, diagnosis of CF-related diabetes, the need for gastrostomy tube feeding, supplementary oxygen, non-invasive ventilation, and assessment for lung transplantation. Psychosocial professionals should be included in the multidisciplinary care at all of these stages (Table 3).

9.1. The CF social worker

9.1.1. Role

The CF Social Worker provides expertise in helping patients and families with their emotional and practical needs and supports patients and families in coping with CF in the different developmental and disease stages. The CF Social Worker should bridge the gap between hospital life and home life and liaise with locally available support so that local services can be accessed. The CF Social Worker is actively involved in the different

Table 3
Examples of important life events where psychosocial support is crucial in people with cystic fibrosis.

Childhood years	Adolescent years	Adult years
Starting kindergarten/day care/pre-school	Starting secondary school	Starting higher education
Starting school	Being a teenager with CF	Starting to work
First awareness of being different	First relationship	Starting a long-term relationship
Eating problems	First sexual experience	Parenthood
Sleeping problems	Death of a CF friend	Death of a CF friend
Behavioural problems (e.g. non-adherence)	Behavioural problems (e.g. non-adherence)	Behavioural problems (e.g. non-adherence)
Examples of key medical stages		
CF diagnosis	Diagnosis of infertility	
First *Pseudomonas* infection	Treatment of infertility	
First episode of allergic bronchopulmonary aspergillosis	Awareness of deteriorating disease	
Gastrostomy placement	Transition to the adult clinic	
Diagnosis CF-related diabetes	Transition to transplantation	
First haemoptysis and other complications	Transition to end-of-life care	
Supplementary oxygen dependence	Transplantation	

CF, cystic fibrosis.

transition stages, including transition to adult care and transition to transplantation care (Table 3). The CF Social Worker has an expertise that complements that of the CF Clinical Psychologist.

The CF Social Worker must have skills in the assessment of practical needs and provide a range of services that are available in the patient's country. Up-to-date knowledge of the country's system of benefits and allowances is essential. The CF Social Worker has to be able to liaise with other agencies (e.g. health insurance companies, child benefit agencies, social welfare agencies, hospital administration, school, CF patient associations) and serve as an advocate for patients and families. The CF Social Worker must have expertise in educational and career issues of the country where the patient resides. They should be skilled in the implementation of child protection procedures, and ensure effective information sharing, referral and liaison to home authority teams, where appropriate. Home visits can greatly contribute to best care and should be available where needed.

9.1.2. Professional development

CF Social Workers need to keep up to date with changes in healthcare systems, financial or social security matters, educational and work aspects, and patient and family welfare concerns. CF Social Workers need continuous education about CF issues. They should attend national and international conferences regularly to maintain up-to-date knowledge about CF and new scientific developments.

9.2. The CF Clinical Psychologist

During their life people with CF have to acquire specific CF-related healthcare behaviours in conjunction with acquiring normal developmental tasks. The CF Clinical Psychologist can help patients and families with these challenges and support them in coping with CF and its treatment throughout life.

9.2.1. Role

In relation to the patient and the family, the key responsibilities of the CF Clinical Psychologist are the assessment of, and intervention in, emotional, behavioural and psychological difficulties, using evidence-based treatments where indicated and making onward referrals where appropriate. The CF Clinical Psychologist is responsible for all the psychological work in the CF Centre. They should offer outpatient clinics as well as caring for hospitalized patients. Adherence, eating behavioural problems, anxiety and depression, demoralization, pain, phobias and sleep are day-to-day themes that need psychological care.

The CF Clinical Psychologist may use mediation techniques in working with other CF MDT members. In addition, the CF Clinical Psychologist should provide a consultation and supervision service to other members of the CF MDT in their work with patients, and provide staff support in coping with 'working in CF'.

The CF Clinical Psychologist must be registered with their national governing body. They should have expertise in child/adolescent and/or adult clinical psychology and also in systemic psychology and the psychology of grief and bereavement. The CF Clinical Psychologist must be skilled in applying therapeutic techniques that have proven efficacy in patients and families with CF. These include, for example (cognitive) behavioural techniques [79,80] and motivational interviewing [81]. Finally, the CF Clinical Psychologist has to be up to date with research on psychological issues in CF, including adherence, self-management and self-care, impact of chronic illness on human development, impact of chronic illness on family and social life, end-of-life issues and palliative care. The CF Clinical Psychologist should have or should develop, skills and experience in conducting psychological research with the aim of improving the care of patients and the understanding of psychosocial issues in CF.

9.2.2. Professional development

CF Clinical Psychologists have a responsibility to engage in CPD and in some countries this will be assessed as part of annual registration. Clinical Psychologists working in CF have a responsibility to update their knowledge of medical aspects of CF as well as mental health. Some countries have a national body for psychosocial professionals, such as in the UK, and membership is a requirement of practising in a CF team. CF Clinical Psychologists should attend national and international

conferences regularly to maintain CF knowledge and to be aware of scientific developments.

9.3. Facilities and requirements for psychosocial care

The CF Clinical Psychologist and the CF Social Worker need sufficient time, office space, and facilities, and the support and respect of the CF MDT. The CF Social Worker and the CF Clinical Psychologist often need to stay in regular contact with patients and families in between clinic visits and therefore need access to modern media (e.g. email, phone, texting). They both need an up-to-date interactive referral system to external psychosocial professionals and institutions in order to provide problem-specific psychosocial/mental healthcare (e.g. for attention deficit hyperactivity disorder or autism) within the vicinity of a patient's home. The CF Clinical Psychologist and CF Social Worker must participate and contribute to the MDT meetings, decision making and patient consultation.

10. Framework for clinical genetics

Clinical Geneticists, Medical Molecular Genetic Laboratory Specialists and Genetic Counsellors have an increasingly important role in the complex diagnosis and disease management of CF, particularly in the areas of disease diagnosis, informed reproductive choices and the assessment of disease liability of *CFTR* variants detected by DNA sequencing. Furthermore, the Clinical Geneticist assesses the linkage phase through family segregation analysis, and ensures that complex alleles (which may be associated with variable CF phenotype) are not under- or misdiagnosed.

The Genetic Counsellor or the Clinical Geneticist provides counselling on reproductive options to the families of newly diagnosed children and to adult patients, and facilitates identification of at-risk family members who are genetically related to the patient.

Clinical Geneticists, working within the CF Centre, also coordinate data sharing with specialized registries (e.g. the ECFS Registry) and submission of detected *CFTR* mutations to both the Cystic Fibrosis Mutation Database (CFTR1), which serves as a locus-specific database for mutations and variants identified on a world-wide scale, and the Clinical and Functional Translation of CFTR online interactive resource (CFTR2), in order to objectively substantiate the disease liability of identified *CFTR* gene variants.

If administration of CFTR modulating therapies is considered, the CF Clinical Geneticist is responsible for the laboratory verification of the *CFTR* genotype in eligible patients (optimally by independent sampling and DNA sequencing) and that the laboratory examination is performed in an ISO 15189-accredited laboratory that assures appropriate turn-around time.

11. Framework for data collection

CF is a multisystem clinically heterogeneous disease with variable outcomes despite its monogenic origins. Although phenotypic variation is influenced by genotype, siblings with the same genotype differ in outcome suggesting the influence of other factors such as modifier genes, the environment, airway microbiota, social class, sex, access to healthcare and adherence to treatment [82]. Collecting data at a national and international level remains a key process to aid the understanding of the epidemiology and outcomes of the disease. It is only through accurate data collection that disease progression, outcomes, health economics and the need for change can be identified [7]. High-quality data can also be used by policy makers to focus on and prioritize future strategies and interventions.

CF is a relatively rare disease with small patient numbers. Collecting data from a single institution limits the level at which clinical and translational research can be undertaken, and does not capture the significant variability in geographical outcome [83]. It is therefore essential that both small and large CF units submit data at least annually to national and/or European CF registries in order to ensure that appropriate longitudinal data are collected. The registry also acts as a monitor for an individual Centre's outcomes, providing an additional tool for ensuring standards of care and appropriate clinical governance.

Healthcare professionals can find collecting and submitting data to national registries laborious and time consuming. Every effort should be made to ensure that data sets are limited to those of predictive value and that individuals can upload data through a secure intuitive interface. Funders must ensure that larger CF Centres have the resource to employ either a data clerk or alternative individual whose ring-fenced responsibilities include national data submission. To have value, such a person must have meaningful access to the management structure so that their voice counts and they can submit gold standard data.

There is a need for the international CF community to adopt a standard coding structure. Creating uniform clinical terminologies and classifications of disease through a primary coding structure would allow clinical data to be mapped and shared between registries as well as other data sets. The changes would result in a common digital language allowing effective international collaboration and would remove key barriers to electronic connectivity.

Addressing this issue is a matter of urgency, as a new era of medical informatics and electronic health records is upon us. Health services have started to successfully deploy electronic patient records, which automate the capture of data and have the potential to feed large continuous data sets directly into national and international registries. In Europe, a standard called XML has been adopted as a first step to harmonizing data from national registries. Almost all national organisations follow this initial standard, but this is only a first step in order to standardise the uploading; if the data collected do not adhere to common unequivocal definitions, the XML format cannot correct it. Therefore, common definitions and coding in both national registries and directly reporting individual centres are crucial.

The European Commission has decided to start funding a European Platform on Rare Diseases Registration, which will provide services and tools for the existing and future rare diseases registries, in accordance with the Council Recommendation on action in the field of rare diseases (2009/C 151/02).

The interconnectivity will maximize patient benefit for minimum outlay, which is a key given the difficult financial situation in healthcare. Adopting a detailed coded registry structure can have the added advantage of reducing costs and improving productivity [84,85]. All such European and/or international data aggregations will have to make the best possible use of local/regional/national data acquisition and storage and intelligent ways of data sharing or retrieval will have to be developed.

Registries are here to stay and should be seen as a key part of any chronic disease management.

12. Challenges relating to developing health services in low income countries

The aim of the ECFS Standards of Care Guidelines is to improve the quality of care for patients with CF and to establish best practice across the whole of Europe. Immediate implementation of these guidelines may prove difficult for less economically advantaged countries where CF services are absent or inadequate. The EU EuroCareCF European Commission 6th Framework Coordination Action project identified a persisting wide difference in the standards of care across Europe, with some Eastern European countries having very basic or no recognizable CF services [7]. The likely reason for such dramatic inequalities has been the absence of appropriate funding, a lack of staff recruitment and training, and also a lack of political prioritization.

The current situation in Eastern Europe was assessed from the responses to a questionnaire distributed to most Eastern European countries by the ECFS Standards of Care Committee. The aims were to evaluate:

- a minimum number of patients who attend CF Centres in Eastern Europe
- national recognition of CF Centre networks, and
- the composition of CF MDTs and the cooperation between paediatric and adult Centres.

Each question was answered on the basis of the current situation in the country and in relation to the ECFS Standards of Care [4]. The response rate was 44% (7/16: Czech Republic, Hungary, Latvia, Poland, Serbia, Slovakia, Ukraine).

The major observations were as follows. While the number of patients per CF Centre is currently below 50 in some Eastern European countries, a requirement for a minimum number of 50 was seen to be an achievable goal. Centralized CF care has been endorsed by government authorities in only three of seven Eastern European countries. A paediatrician/pulmonologist, a physiotherapist and a CF nurse specialist were agreed to be essential team members. Many CF Centres lack a full-time CF nurse specialist, a dietitian, a microbiologist, a psychologist, a social worker or secretarial support. Collaboration between paediatric Centres and those for adults with CF had not been established in two of the countries that replied to the survey.

It is imperative that all European countries should strive to implement best practice in accordance with the ECFS recommendations. An initial stepwise approach may be needed in some low income countries where there are no established services and CF MDTs virtually do not exist. For example, initial recruitment of core medical, nursing and physiotherapy staff may be the most appropriate initial investment on the way to establishing a service that meets all ECFS standards. It is no longer acceptable to have such dramatic variation in the survival of people with CF across European nations, and every effort must be made to deliver equality and high standards of care.

CF care in low income countries should be centralized in well-established CF Centres that can guarantee a reasonable standard of complex care for both paediatric and adult patients. The Centre should care for at least 100 patients, although a minimum of 50 may be temporarily considered acceptable. Because of the financial and staffing limitations in Eastern Europe, shared care with local hospitals is not a preferred model of care. Resources should be directed at establishing state-of-the-art CF care at a national level by the development of specialized CF Centres at major hospitals, and where possible at the level of University Hospitals. The minimum staff requirements for a specialist CF team include a physician and a specialist nurse (one each for children and adults in Centres that care for all age groups), and a CF specialist physiotherapist. The goal is to develop teams that include a microbiologist, dietitian, psychologist, social worker and clinical geneticist. Meanwhile the absence of these specialists should not delay the setting up of regional CF Centres. Their roles may be temporarily performed by specialist consultants of the hospital who can be accessed by the CF service, even though not primarily allocated to a CF team and therefore not able, for instance, to participate in regular CF MDT meetings.

13. Perspective from European CF associations

13.1. The function and role of national CF patient organisations in Europe

Most European countries have their own national CF patient organisation. These vary in size and methodology but have one thing in common: they fight for the interests of people with CF, in the broadest sense of the word. They work together with volunteers, who are often experts in CF but who may be non-medical. In Europe patient organisations are developing their own in-house expertise and employing professional staff, for example in the area of healthcare quality, scientific research, communication, fund raising, information provision, and the legal and psychosocial aspects of CF. The organisations aim to support patients and their parents, both individually and collectively, and to define research agenda, to finance scientific research and to test healthcare quality. They are often closely involved in the setting up and maintenance of guidelines for the diagnosis and treatment of CF.

The national CF organisation can play an important role in the national CF registry. Many have helped found these registries and continue to support and finance them. In a number of countries, the CF organisation plays a role in the set-up and organisation of research networks and healthcare quality improvement programmes for people with CF.

Fund raising is an important prerequisite to enable the organisations to successfully achieve the above aims. Over the past few years, CF organisations in countries such as the UK, Germany, Belgium, France, Italy and The Netherlands have invested millions of Euros in scientific research and healthcare quality. As a result, they have made important contributions to the progress that has been made in many healthcare areas of CF.

Patient participation is an important part of the work achieved by patient organisations, as it is exactly the patients' perspective that can and should be expressed by the national CF organisations.

13.2. National organisations

The national CF organisations have responsibilities to provide information. This material, which is made available to people with CF and their parents/carers, is often developed in collaboration with the CF Centres and covers all aspects of life with CF: diagnosis, treatment, growing up with CF, raising a child with CF, going to school with CF and building a life with CF. National CF groups should work closely with CF Centres in the organisation of healthcare, a cooperation that may vary in its infrastructure in different countries. However, in all cases, representatives of those Centres should be present on medical advisory boards and scientific councils.

CF organisations should organise meetings for parents and, with the use of information technology to safeguard against cross-infection risks, for the patients themselves. E-health has many advantages and is a subject that is being researched and developed.

CF organisations are the obvious parties to represent and protect the interests of patients and their parents/carers, for example by supporting ready availability of new medications, securing reimbursements for medication and access to high-quality healthcare. CF organisations should lobby and exert pressure on the authorities, government and insurance companies in this regard. They should also organise congresses, symposia and other meetings (in collaboration with or under the supervision of the healthcare organisations), in which the specific (scientific) aspects of CF are discussed.

CF patient organisations are non-medical yet through close cooperation with CF Centres have developed a lot of CF expertise. It is important that the Centres and the organisation exchange information on a regular basis in order to facilitate a proactive approach to developments in the healthcare industry and scientific research. This collaboration will help to improve communication with patients, facilitating their inclusion in scientific studies and solving problems on a national level more swiftly.

13.3. European organisations

The European CF patient organisations are united in a single European society — CF Europe (CFE). The importance of collaboration in Europe is growing, especially with regard to healthcare access, which is not uniform, or even available, in all European countries. As a result, collaboration in the fields of research, research financing and fund raising is continually increasing.

The European collaboration should lead to the CF organisations in countries where CF healthcare is already at a high level accepting their responsibility and offering their expertise to countries in which adequate healthcare, access to healthcare and the ready availability of medication are not universal. This should take place in close collaboration with the CF Centres. Collaboration through CF Europe has made it easier to lobby effectively on a European level with regard to subjects such as organ donation, accessibility, quality and affordability in healthcare.

Another level of European collaboration is partnering with the European overarching patient support organisation (Eurordis), which is representing the majority of rare disease national and regional organisations (including CF) in e.g. terms of awareness building, access to care, reimbursement policies, development of European guidelines for centre care fundraising, introduction of orphan medicinal products into therapy, including training courses on various aspects of patient advocacy.

CF patient organisations are working increasingly with the ECFS, for example by participating in the executive boards of the EFCS Patient Registry and the EFCS Clinical Trials Network. The latter is financially supported by a number of patient organisations. Through the CF patient organisations, patients and parents have become more involved recently in the assessment of the ECFS Clinical Trials Network research protocols.

Conflict of interest

S. Conway, K. De Rijcke, P. Drevinek, J. Foweraker, T. Havermans, H. Heijerman, L. Lannefors, A. Lindblad, M. Macek, S. Madge, M. Moran, L. Morrison, A. Morton, J. Noordhoek, D. Sands, A. Vertommen, and D. Peckham have no conflicts of interest to report. I. M. Balfour-Lynn declares personal fees from Vertex, outside the submitted work.

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