

Cystic Fibrosis *our focus*

**National Consensus Standards for the
Nursing Management of cystic fibrosis**

May 2001

Fighting for a
Life Unlimited

UK Cystic Fibrosis Nurse Specialist Group Working Party

Thanks go to all members of the working party who helped compile this document. We are also grateful for the comments and suggestions from parents, patients and other professionals who reviewed this document.

Chairman

Susan Madge, MSc RGN RSCN, Great Ormond Street Hospital for Children, London

Members

Glenda Esmond, BSc (hons) RGN DipN, Barts and The London Adult CF Centre

Susan Saunders, RGN RSCN, Birmingham Children's Hospital

Janis Bloomer, RGN RSCN, Royal Victoria Infirmary, Newcastle upon Tyne

June Dyer, BSc (hons), RSCN Bristol Children's Hospital

Anne Morais, RGN, University Hospital of Wales, Cardiff

Carole Davies, BSc (hons) RGN, Llandough Hospital, Cardiff

Jenny Cottrell, RGN RSCN, Alder Hey Children's Hospital, Liverpool

These consensus standards were compiled by cystic fibrosis clinical nurse specialists (paediatrics and adults) working in the community and the hospital and based in regions around the UK. It is intended that by referring to these consensus standards all nurses involved in the care of cystic fibrosis will provide an optimum level of care for patients and their families.

National consensus standards for the nursing management of cystic fibrosis

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1. Introduction

Caring for people with cystic fibrosis (CF) can be complex and demanding and nurses finding themselves in this situation must ensure that they liaise closely with the nearest specialist CF centre. This document is offered as a guideline to nurses, both specialist and general, who care for people with CF, although throughout the text reference is made to the nurse specialist.

This document is not designed to be a textbook on CF or to be used as a detailed care plan or protocol, rather as a guide only. Nursing is a partnership between the patients, carers and nurses, if the suggestions given are used within the parameters of advocacy, clinical management, education, support, advice and research, a holistic plan of care can be made.

Although this document does not follow the strict criteria required for guidelines¹, it does offer examples of evidence with some recommendations made on the basis of these.

Scheme used for grading strength of evidence and recommendations

Levels of evidence
Level I - Randomised controlled trials, meta-analysis or systematic reviews
Level II - Cohort or case-control studies
Level III - Uncontrolled studies or consensus
Recommendations
*Based on level I evidence
**Based on level II or extrapolated recommendation from level I
*** Based on level II or extrapolated recommendation from level I or level II

1.1 Specialist care and the multi-disciplinary team

Cystic fibrosis is the most common, life-threatening inherited disease in the United Kingdom affecting approximately 1:2500 live births. Cystic fibrosis is a multi system disorder and although primarily the respiratory and digestive systems are involved there are often problems with the liver, joints, diabetes, fertility and reproduction².

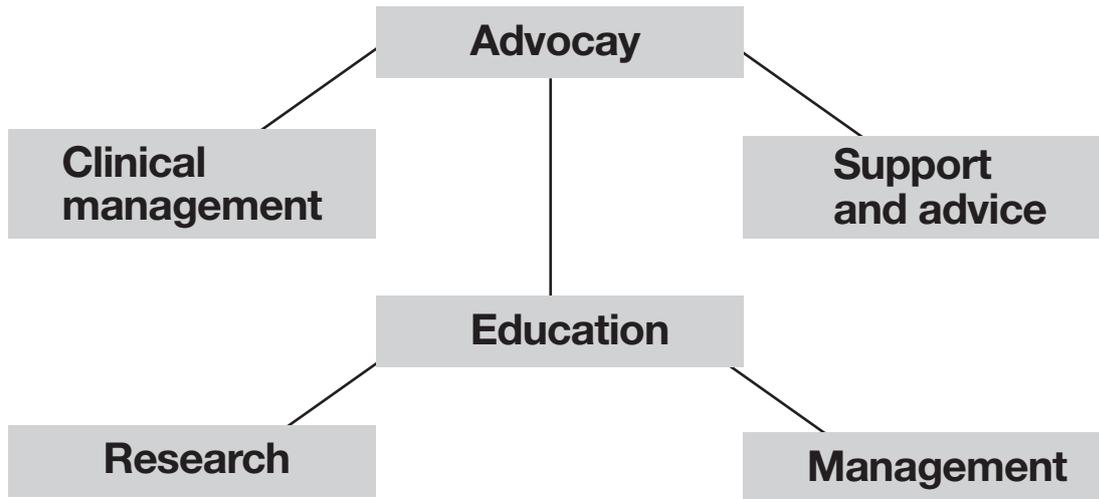
In 1989 the gene coding for cystic fibrosis was located on the long arm of chromosome seven and although 6F508 is the most common mutation found in the United Kingdom³, more than 900 mutations of the gene have been discovered to date (May 2001).

Advances in medical management over the last few years have improved both quality of life and longevity. However, due to the complexity of the disease it is widely recognised that people with CF should be cared for using a multi-disciplinary team approach. Cystic fibrosis teams in the UK include doctors, nurse specialists, dietitians, physiotherapists, psychologists and/or social workers. In addition it is recommended that these teams are supported by staff such as respiratory function technicians and ward nurses skilled in caring for people with cystic fibrosis⁴.

The multi-disciplinary team works very closely together to ensure a holistic approach to the care of their patients. Good communication is paramount and the team must allow time for regular meetings. Nurses working in areas where there is no opportunity to work within a team must communicate with the nearest specialist CF centre, both for their own support and to assure optimum care for their patients⁵.

1.2 The cystic fibrosis clinical nurse specialist

The roles of a nurse specialist often overlap and impact on each other



The cystic fibrosis nurse specialist has a five-fold responsibility to patients and their families and the staff that are involved in the care of those patients. These areas include: advocacy, clinical management, advice and support, education, research and management^{6, 7, 8}.

Advocacy: There are many demands made on both the patient and the CF team therefore patient and family advocacy is the most important role for the nurse specialist. Patient wellbeing and satisfaction with care are paramount and successful advocacy can ensure this⁸.

Clinical management: The nurse specialist must take part in decision making and monitoring of care. In addition to the practical, day to day care that the nurse specialist offers, their responsibility is to be aware of all treatment modalities used in the management of CF and to ensure that each patient receives optimum care for their individual needs. The role of the nurse specialist is also to act as a link between patient and family, community services and the hospital multi-disciplinary team.

Support and advice: Both patients and their families and professional colleagues will benefit from the support, liaison and advice available from the nurse specialist⁹. This can be as a consistent carer, counsellor or as a confidante. Problems are often resolved more easily if pathways of communication are well established for all parties¹⁰.

Education for patient, families and carers: There are many treatment regimens that have to be learnt throughout the patient's life. Successful teaching of the patient and all concerned (parent, carer, school, work colleagues) and their understanding of the disease process, will ensure that treatment is carried out safely and effectively and that issues surrounding adherence to treatment can be more successfully dealt with.

Education, research and management: Nurse specialists are responsible for developing their own professional practice through reflection, participation on post registration courses and attendance at conferences and meetings. This also ensures that they keep up to date with new advances in treatment and new developments in the world of CF research enhancing their own research based practice^{10,11}.

2. Recommendations for best practice

2.1 Diagnosis

Diagnosis of a life limiting disease can be likened to bereavement and needs to be handled with honesty and sensitivity by skilled personnel. A diagnosis of CF may be made at any age and although most patients are diagnosed within the first year of life it is not unusual for adults, especially males to be diagnosed later on in life. When clinical evidence leads to suspicion of CF diagnosis can be made using a number of methods¹²:

- prenatal diagnosis - chorionic villus sampling
 - sweat testing
 - genotyping
 - nasal potential differences
 - pancreatic function tests
 - stool elastase
-
- Diagnosis should be carried out in a Specialist CF Centre or CF Clinic experienced in the techniques being used and if CF is confirmed immediate referral must be made to a CF team¹³.
 - The nurse specialist will ensure that pre-diagnosis support and counselling is available¹⁴.
 - The nurse specialist should be present when the diagnosis is being given either to a child and family or to an adult¹⁵.
 - The nurse specialist will help to determine the appropriate timing (to avoid inappropriate referral) for the introduction of information regarding¹⁵:
 - Contact numbers and availability of the CF team.
 - Involvement and liaison with the primary health care team.
 - Support agencies available, both local and national.
 - Government support.
 - The specialist nurse will offer continued psychosocial advice, support and counselling to the patient and their family with particular reference to:
 - Continued education about the disease.
 - The genetic implications to the immediate and extended family.
 - Expectations of care and day-to-day treatment regimens.
 - The nurse specialist will be available to offer advice, education and support to the patient, their families/carers and all staff involved¹⁰.

2.2 Inpatient care

- Ward nurses will have access to a CF nurse specialist.
- Each patient will be cared for or supported by specialist nurses who have a knowledge and experience of CF¹⁶, the disease process and the clinical and psychological outcomes, with specialist knowledge of:
 - psychological issues - living with a life limiting disorder
 - issues surrounding diagnosis
 - nutritional requirements
 - enteral feeding
 - CF related diabetes

- intravenous therapy
- respiratory complications and support
- nebuliser therapy
- care of indwelling venous devices
- infection control
- osteoporosis
- liver disease
- terminal care and symptom management
- Nurses caring for patients with CF and supported by a nurse specialist must assess, plan, implement and monitor care according to the needs of each patient at every admission, taking into account the variability of the disease.
- The nurse specialist will ensure that each patient has access to all members of the multidisciplinary team as often as their needs dictate.
- The nurse specialist will ensure a partnership in care with the patient and their family/carers¹⁷.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/ carers and all staff involved¹⁸.
- The nurse specialist, in liaison with ward staff will ensure that discharge planning needs are met with special reference to the GP, the primary health care team, the shared care hospital and school (where appropriate).
- self administration of intravenous therapy
- enteral feeding
- oxygen therapy
- psychosocial and emotional support
- The nurse specialist will liaise with patient and family on issues surrounding cross-infection and infection control¹⁹.
- The nurse specialist will ensure that all patients receive a comprehensive Annual Review in a recognised Specialist CF Centre or CF Clinic²⁰.
- The nurse specialist will liaise with the CF team, the primary health care team, shared care hospital, work and school (where appropriate)²¹.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/ carers and all staff involved¹⁰.

2.3 Outpatient care

- The nurse specialist will ensure that at every routine appointment each patient has access to¹³:
 - a doctor
 - a physiotherapist
 - a dietitian
 - a nurse specialist
 - a social worker
 - a psychologist
- The nurse specialist will ensure that at every routine appointment each patient has measured (as indicated)¹³:
 - lung function
 - oxygen saturation
 - weight and height
 - sputum or cough swab for microbiology
- The nurse specialist, in conjunction with appropriate members of the CF team, will be responsible for specific aspects of clinical management such as¹³:
 - nutritional needs
 - nebuliser therapy
 - care of indwelling venous devices

2.4 Community care

The nurse specialist will continue to support the patient in the community to ensure an equality of care to that offered in hospital.

- Each patient will have access to a CF nurse specialist¹³.
- Nurses caring for patients with CF in the community will be supported by a nurse specialist who has knowledge and experience of CF. This will include the disease process, the clinical and psychological outcomes of the treatment modalities to ensure safe and effective care meeting the minimum nursing standards of care²².
- The nurse specialist will ensure that there is close liaison between the CF team, the GP, the primary health care team, the shared care hospital and work or school²².
- The nurse specialist will ensure that advice and support is available for the management of complications associated with CF such as¹³:
 - CF related diabetes
 - arthropathy
 - haemoptysis
 - pneumothorax
 - breathlessness
 - liver disease
 - pain
 - osteoporosis
- The nurse specialist, in liaison with colleagues, will support and co-ordinate:
 - home supplementary nutrition
 - home intravenous therapy
 - respiratory support

- The nurse specialist will assess and monitor the response to treatments carried out in the community²².
- The nurse specialist, in liaison with colleagues will offer psychosocial and emotional support surrounding issues of living with a genetic, life limiting disease to families and patients of all ages.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/ carers and all staff involved¹⁰.

2.5 Issues surrounding adolescence and adulthood

Adolescence and young adulthood is a particularly difficult time for those having to deal with both the psychological and practical burden of CF. Many treatment related issues can become a problem and the nurse specialist must be alert to problems, especially those surrounding adherence and conflict between teenager and parents²³.

- The nurse specialist will be instrumental in promoting self care and responsibility in the young adult and offering support and advice to the parents²⁴.
- The nurse specialist will liaise with schools and colleges to support continuing education⁹.
- The nurse specialist, in liaison with colleagues, will ensure that adolescents receive appropriate knowledge regarding issues such as²⁵:
 - fertility
 - pregnancy
 - contraception/safe sex
 - cross-infection
 - further education/employment
 - smoking/substance abuse
- The nurse specialist should use experience and knowledge to advise on the appropriate time (however long) for transition and transfer to adult care for each patient. This will include²⁶:
 - Liaison and communication between the paediatric and adult specialist CF centres about all aspects of care, e.g. level of knowledge of CF at time of transfer.
 - The co-ordination of joint transition clinics, parallel care and visits to the adult specialist CF centre where appropriate.
 - Provision of adequate information and ongoing support for patients and parents during the transition period.
- The nurse specialist will allow opportunity for discussion between the patient, parents and CF team members when problems arise²⁷.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/ carers and all staff involved¹⁰.

2.6 Issues surrounding the care of advanced cystic fibrosis

Although children still die, CF is becoming a disease of adulthood. However, pre-terminal grief at diagnosis onwards must be acknowledged throughout the patient's life. Treatment choices - the dilemma of aggressive management versus palliative care, is difficult for both the families and staff. The pre-terminal and terminal stages must be handled with sensitivity and compassion together with sound clinical judgement and involve the patient, their family and staff.

- The specialist nurse will recognise the patient's complexity of care and changing needs and will be able to offer support to the patient and their family to help them come to terms with and adapt to the changes²⁸.

The nurse specialist must be aware of the concerns involved with heart/lung transplantation and liver transplantation as a treatment option, these include²⁹:

- the stress of waiting
- the complications of transplantation
- the loss of a recognisable end point
- death with dignity
- The nurse specialist will advocate on the patient's and family's behalf. This will involve accepting new ways of coping, recognising denial, respecting their wishes and decisions about treatment, and allowing discussion around issues of dying³⁰.
- The nurse specialist will ensure that the patient and family receive sufficient knowledge to make informed decisions about treatment and where possible allow flexibility of choice.

These treatment decisions will include^{28,31}.

- terminal care – hospital or home (within available resources)
- continuation of enteral feeding and intravenous therapy
- the options for respiratory support
- pain management and symptom control
- dealing with complications such as haemoptysis, liver disease, pneumothorax, trans-plantation and management of cardio-respiratory failure
- bereavement management
- The nurse specialist will be able to recognise and support the healthcare team – both hospital and community based – in accepting the outcome²⁸.
- The nurse specialist will involve other support and specialist agencies where appropriate.
- The nurse specialist will maintain ongoing liaison and communication with the primary healthcare team.
- The nurse specialist, in liaison with colleagues will ensure that appropriate bereavement support is offered to the family both in the terminal stages and after death.

Caring for the dying patient is complex and stressful, however, the nurse specialist should recognise that with support from the CF team and the available support network it can become a rewarding and challenging experience.

3. Consensus standards of care for nursing practice

These are minimum standards of nursing care and should be adapted to meet local needs.

Standards of care for

3.1 Diagnosis

3.2 Inpatient care

3.3 Outpatient care

3.4 Community care

3.5 Nebuliser therapy

3.6 Care of central venous access devices

3.7 Self-administration of intravenous therapy

3.8 Enteral feeding

3.9 Oxygen therapy

3.10 Cystic fibrosis related diabetes
(insulin dependant)

3.11 Transfer from paediatric to adult care

3.12 Terminal care and symptom management

Consensus standards for the nursing management of cystic fibrosis

3.1 Diagnosis

Standard statement

Once the diagnosis has been confirmed the nurse specialist will provide a full explanation of the condition and provide individual clinical and psychological care for each patient and their family.

Structure

- 1. Notification will be made to a recognised Specialist CF Centre of all newly diagnosed patients.
- 2. A CF team consisting of doctors, nurse specialist, physiotherapist, dietitian, psychologist and social worker will be available.
- 3. CF literature will be available.

Process

- 1. The nurse specialist will assess and document the physical and emotional needs of the patient/family.
- 2. The nurse specialist will support the family to enable them to cope with the emotional impact of diagnosis and allow time for discussion.
- 3. The nurse specialist will provide relevant literature to support the information being given to the patient/family at appropriate times.
- 4. The nurse specialist will formulate a treatment plan with explanation of symptoms, implications of CF and reasons for treatment.
- 5. The patient/family will be provided with contact numbers and information about the CF team.
- 6. The patient/family will be informed of follow up arrangements and ongoing support available.

Outcome

- 1. The patient/family will have an assessment of their physical and emotional needs that will be documented.
- 2. The patient/family will have a treatment plan which includes explanation of symptoms, the implications of CF and reasons for treatment.
- 3. The patient/family can state the names, roles and contact numbers of the CF team.
- 4. The patient/family can state when the CF team and nurse specialist will review them.

Audit

Standard Statement (Diagnosis)

Once the diagnosis has been made the nurse specialist will provide a full explanation of the condition and provide individual clinical and psychological care for each patient and their family.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	S1	State which recognised specialist CF centre patient has been referred to?
	Check	P3	Is there literature available for newly diagnosed CF patient/family?
	Check	P5	Is there written information stating CF team members and contact numbers?
Records	Check	O1	Is there a documented assessment of physical and emotional needs?
	Check	O2	Is there a written plan of care?
Patient/Family	Ask	O3	Can the patient/family state the names, roles and contact numbers of the CF team?
	Ask	O4	Can the patient/family state what follow up care has been arranged?
	Ask	O2	Did the patient/family have the opportunity to discuss the impact the diagnosis has made on them?

Consensus standards for the nursing management of cystic fibrosis

3.2 Inpatient care

Standard statement

At each admission staff experienced in the management of CF and able to meet the individual's physiological, psychological and social needs will provide care.

Structure

- 1. A recognised specialist CF centre or CF clinic will be available.
- 2. Facilities and equipment to perform lung function testing, oxygen saturation and microbiological analysis of sputum will be available.
- 3. A CF team consisting of a doctor, nurse, physiotherapist, dietitian, psychologist and social worker will be available.
- 4. Ward staff will be experienced in the care of CF.

Process

- 1. The CF patient/family will have access to a recognised specialist CF centre.
- 2. On admission the patient will be fully assessed, this may include lung function, height, weight, urinalysis, oxygen saturation, chest x-ray and sputum analysis where appropriate.
- 3. The patient/family will have the opportunity to discuss treatment issues with members of the CF team.
- 4. The option of home therapy will be discussed with the patient/family where indicated.
- 5. Treatment plans will be monitored and changed where indicated.
- 6. Appropriate follow up will be arranged at discharge.

Outcome

- 1. The patient is assessed and a plan of care is available.
- 2. The patient/family have had the opportunity to discuss treatment options.
- 3. Treatment changes are monitored and documented.
- 4. The patient will have the correct medication and equipment on discharge.
- 5. The patient/family can state when the CF team will review them.

Audit

Standard Statement (Inpatient care)

At each admission care will be provided by staff experienced in the management of CF and able to meet the individual's physiological, psychological and social needs.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	P1	State which recognised specialist CF centre the patient is registered with?
	Ask	S4	Do ward staff have experience of caring for CF patients?
Records	Check	P2	Is there evidence that the patient has been assessed on admission and that results of investigations are documented?
	Check	O1	Is there a plan of care?
	Check	O4	Is there evidence of discharge planning?
Patient/Family	Ask	O2	Did the patient/family have the opportunity to discuss care with the CF team?
	Ask	P4	Did the patient/family have the opportunity to discuss the option of home therapy?
	Ask	O4	Did the patient have the correct medication and equipment on discharge?
	Ask	O5	Can the patient/family state what follow up care has been arranged?

Consensus standards for the nursing management of cystic fibrosis

3.3 Outpatient care

Standard statement

At each routine outpatient visit a multi-disciplinary team experienced in the management of CF and able to meet the individual's physiological, psychological and social needs will provide care.

Structure

- 1. Facilities and equipment to perform lung function, oxygen saturation, weight, height and sputum/cough swab microbiology will be available.
- 2. Growth parameters will be recorded where appropriate.
- 3. A multi-disciplinary team, specialising in CF and consisting of a doctor, nurse specialist, physiotherapist, dietitian, social worker and psychologist will be available.

Process

- 1. The patient/family will have access to a recognised specialist CF centre.
- 2. Written information regarding the roles, names and contact numbers of the CF team members are given to the patient/family.
- 3. At each outpatient visit the patient will have their FEV₁, FVC, SaO₂, height, weight and a sputum/cough swab specimen taken for analysis where appropriate.
- 4. At each outpatient visit the patient/family will have the opportunity to discuss their care with a doctor, nurse specialist, physiotherapist, dietitian, social worker and psychologist.
- 5. The patient will have an Annual Review performed.

Outcomes

- 1. The patient/family can name to which recognised specialist CF centre that they have access.
- 2. The patient/family has the names and contact numbers of the CF team.
- 3. The patient has their FEV₁, FVC, SaO₂, height, weight and a sputum/cough swab specimen taken for analysis at each visit.
- 4. The patient has a documented Annual Review.
- 5. A doctor, nurse specialist, physiotherapist, dietitian, social worker and psychologist with knowledge of CF are available to see the patient/family during their visit where appropriate.

Audit

Standard Statement (Outpatient care)

At each routine outpatient visit care will be provided by a multi-disciplinary team experienced in the management CF and able to meet the individual's physiological, psychological and social needs.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	P1	Can staff state where the patient's recognised specialist CF care takes place?
	Ask	O5	Can staff state the names of CF team that are involved in the patients care?
	Check	P2	Is there any written information stating CF team members and contact numbers?
Records	Check	O3	Is there evidence that the patients FEV ₁ , FVC, SaO ₂ , height, weight and sputum/cough swab specimen were performed and recorded?
	Check	O4	Is there evidence that the patient has had an Annual Review performed?
Patient/Family	Ask	O2	Can the patient/family state the names of the individual members of the CF team and how to contact them?
	Ask	O5	Did the patient/family have an opportunity to discuss their care with a doctor, nurse specialist, physiotherapist, dietitian, social worker and psychologist during their visit?

Consensus standards for the nursing management of cystic fibrosis

3.4 Community care

Standard Statement

The patient/family will have access to specialist advice, support and direct supervision as required, enabling the individual to meet their health needs in the community setting.

Structure

- 1. Availability and access to a specialist CF centre and multi-disciplinary team for direct care or as a resource to local services.
- 2. Resources and equipment to facilitate safe and effective home care will be available.
- 3. Information regarding local government guidelines/resources for community care are available.

Process

- 1. The CF patient/family will have access to care and advice from a specialist CF centre.
- 2. The patient/family are given the opportunity to discuss community care and together with the specialist nurse a plan of care is formulated and re-assessed on a regular basis.

- 3. Relevant teaching programmes and instruction will be implemented prior to commencement of home care.
- 4. The patient/family are given contact numbers to cover the 24 hour period for support and advice.
- 5. Drugs, equipment and written information are available for the patient at the commencement of home care and throughout its course.
- 6. Continued monitoring and supervision is carried out to ensure the safe delivery of and effective treatment/care in the community.

Outcomes

- 1. The patient/family can name which specialist CF centre they attend or have shared care with.
- 2. The patient/family have a plan of care.
- 3. The patient/family feel competent and confident to continue with community care/treatment as outlined in the plan of care.
- 4. The patient/family can state who to contact for help and advice covering the 24 hour period.
- 5. The patient/family have the equipment, drugs and written information to enable plan of care to be implemented.
- 6. The patient/family acknowledge that episodes of continuing care are safe, effective and appropriately supported.

Audit

Standard Statement (Community care)

The patient/family will have access to specialist advice, support and direct supervision as required, enabling the individual to meet their health needs in the community.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Check	S3	Is there information available on local government guidelines for community care?
Records	Check	O2	Is there a plan of care
Patient	Ask	O1	Can the patient/family state whether specialist CF centre care or CF clinic care is received?
	Ask	O3	Does the patient/family feel competent and confident to undertake treatment plan at home?
	Ask	O4	Can the patient/family state the names and contact numbers covering 24 hour period for support and advice?
	Ask	P3	Did the patient/family have instruction/education prior to commencement of home care?
	Ask	O5	Did the patient/family have the correct equipment, drugs and written information to enable to undertake treatment at home?
	Ask	O6	Did the patient/family feel supported to undertake care effectively and safely at home?

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3.5 Nebuliser therapy

Standard statement

The patient receiving nebulised medication will understand the importance of the therapy and how to use and care for the equipment.

Structure

- 1. A CF nurse specialist and/or physiotherapist will be available.

- 2. The nurse specialist will have knowledge of the medication.
- 3. The following resources will be available:
 - compressor
 - appropriate nebuliser
- 4. The patient/family is provided with appropriate knowledge in the therapy and in the use and storage of the equipment.

Process

- 1. Together with the patient/family, the nurse specialist and/or physiotherapist assesses and formulates an appropriate plan of action to ensure safe practice.
- 2. The patient/family will be provided with the appropriate nebuliser and compressor delivery system and ancillaries.
- 3. The patient/family will be provided with contact names and numbers for advice and for problems associated with nebuliser and compressor system.
- 4. The patient/family will be advised on frequency and method for maintaining compressor.

Outcomes

- 1. The patient/family has a plan of care to meet their individual needs and can state the problems associated with nebuliser therapy.
- 2. The patient/family are confident at administering nebuliser therapy.
- 3. The patient/family can state who to contact for advice or problems.
- 4. The patient has the appropriate delivery system and ancillaries for their needs.
- 5. The patient/family can state frequency and method of maintaining the compressor.

Audit

Standard Statement (Nebuliser therapy)

The patient receiving nebuliser therapy and their family will be taught how to administer medication safely and effectively.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	S1	Is there a nurse specialist available with experience of nebuliser therapy?
Records	Check	O1	Is there a recorded plan of care?
Patient	Ask	O2	Can the patient/family demonstrate how to use nebuliser equipment correctly and safely?
	Ask	O3	Can the patient/family state names and contact numbers with regard to nebuliser therapy advice?
	Ask	O4	Has the patient/family the correct supplies to administer nebuliser therapy at home?
	Ask	O5	Can the patient/family state frequency and method of maintaining the compressor?

Consensus standards for the nursing management of cystic fibrosis

3.6 Care of a central venous access devices

Standard statement

The care of a central venous access device (CVAD) is managed safely by a suitably trained person to prevent complications.

Structure

- 1. An experienced CF nurse specialist is available for advice.
- 2. Training programmes and assessment processes for all persons caring for the CVAD will be available.
- 3. Supporting literature will be available.

Process

- 1. A CF nurse specialist with knowledge of CVAD will be available to support and advise on management.
- 2. Teaching will be available to all the patient/family and relevant staff.
- 3. A person/persons will be identified and trained to undertake routine flushing of the CVAD.
- 4. Appropriate literature will be readily available to support teaching.
- 5. Action will be taken on all reported complications and problems relating to the CVAD.

Outcome

- 1. The patient/family can state who to contact with regard to the CVAD.
- 2. Competency of accessing and managing the CVAD will be recorded on all persons.
- 3. All those trained in CVAD management will state complications and be able to problem solve.
- 4. The patient/family will be able to state who undertakes routine flushing of the CVAD.
- 5. The patient/family will have literature relating to the management of CVAD.

Audit

Standard Statement (CVAD)

The care of a central venous access device (CVAD) is managed safely by a suitably trained person to prevent complications

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	S1	Is there a nurse specialist available with experience of nebuliser therapy?
	Check	S2	Is there a training and assessment process available for caring for CVAD?
	Ask	O3	Can staff state complications of CVAD and problem solve?
	Check	P4	Is there literature to support teaching available?
Records	Check	O2	There is a record of all those trained to access and manage CVAD?
Patient	Ask	O1	Can the patient/family state who to contact for advice with regard to the CVAD?
	Ask	O3	Can the patient/family state complications of CVAD?
	Ask	O4	Can the patient/family state who performs routine flushing of CVAD?
	Ask	O5	Did the patient/family have literature relating to the management of the CVAD?

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3.7 Self-administration of intravenous therapy

Standard Statement

The patient/family participating in self-administration of intravenous (IV) drugs at home will be able to administer their own drugs safely, ensuring minimal disruption to the individual's life style.

Structure

- 1. The registered nurse has accreditation in IV therapy and has access to IV policy and United Kingdom Central Council (UKCC) Scope of Practice.
- 2. The patient/family is provided with the following resources in the administration of IV therapy:
 - assessment criteria
 - medication as prescribed
 - ancillaries
 - appropriate sized Sharps bin
 - information booklet with drug regime and contact numbers

Process

- 1. Together with the patient/family (and the community team when available), the specialist nurse assesses and formulates an appropriate individual plan of action to ensure safe practice.
- 2. The patient/family are given the opportunity to discuss the option of self-administration of IV drugs at home, so that they can make an informed decision that is reversible.
- 3. a) A teaching plan is formulated to meet the individual's needs and requirements.
b) A period of supervision/instruction will be carried out to meet the needs of the individual.
c) Patient assessment is carried out according to set criteria.
- 4. The patient/family are given contact numbers to cover 24 hour period. The patient/family are contacted during IV course or within 72 hours of discharge when carrying out self- administration for the first time.
- 5. Medication, equipment and information booklet are available for the patient when commencing each course of self-administered IVs at home.

Outcome

- 1. The patient/family has a plan of care to meet individual needs.
- 2. The patient/family are competent at self-administration of IV therapy.
- 3. The patient/family can state who to contact during 24 hour period.
- 4. The patient/family self-administering IV drugs were contacted during home IV course or within 72 hours of discharge if self administering home IVs for the first time.
- 5. Response and compliance with treatment and any problems occurred are recorded.
- 6. (a) The patient received the correct supplies on discharge.
(b) Patient has information booklet.
(c) Patient correctly disposes of the sealed Sharps bins.
- 7. The patient did not acquire IV related sepsis.

Audit

Standard Statement (Self-administration of intravenous therapy)

The patient or family participating in self-administration of intravenous drugs at home have the skills and support to administer their own therapy.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Records	Check	O1	Is there a written plan of care?
	Check	P3	Is the assessment criteria available?
	Check	O2	Is there evidence that the patient/family have been formally assessed in the administration of intravenous medication?
	Check	O5	Is there evidence that the patient's response, compliance to treatment and any problems that may have occurred during treatment been evaluated?
Patient	Ask	P2	Was the patient/family given the opportunity to discuss the option of self-administration of intravenous medication at home prior to discharge?
	Ask	O3	Does the patient/family know who to contact during a 24 hour period if there are any problems when administering the medication at home?
	Ask	O4	Was the patient/family contacted (within 72 hours if it was a first home IV course) during their course of treatment?
	Ask	O6a	Did the patient/family receive the correct supply of medication and equipment to complete the IV course?
	Ask	O6b	Did the patient/family have written instructions regarding the medication regimen?
	Ask	O6c	Does the patient/family know how to dispose of the sealed Sharps bin?

Consensus standards for the nursing management of cystic fibrosis

3.8 Enteral feeding

Standard Statement

The patient/family administering enteral feeding at home will be able to manage the gastrostomy or naso-gastric tube and administer their feed safely ensuring minimal disruption to the individual's life style.

Structure

- 1. A nurse specialist and dietitian will be available for all patients requiring enteral feeding.
- 2. The following resources will be available:
 - replacement tubes (where appropriate)
 - ancillaries
 - feed
- 3. Information regarding feeding regime and contact numbers.

Process

- 1. The nurse specialist will ensure that the patient is assessed for an appropriate education plan.
- 2. The patient/family are given the opportunity to discuss enteral feeding at home.

- 3. A period of supervision/instruction will be carried out to meet the needs of the individuals concerned.
- 4. The patient/family will be provided with contact names and numbers.
- 5. Assessment (height, weight, body mass index where appropriate) of the patient's nutritional status will be carried out regularly.
- 6. Feed and equipment will be available for the patient.
- 7. The nurse will ensure that the patient does not acquire any complications associated with enteral feeding.

Outcome

- 1. An assessment (height, weight, body mass index where appropriate) and record of progress is documented.
- 2. The patient/family has a plan of care to meet their individual needs.
- 3. The patient/family are confident at administering the feed.
- 4. The patient/family can state who to contact.
- 5. The patient has the correct supplies and feed.

Audit

Standard Statement (Enteral feeding)

The patient/family administering enteral feeding at home will be able to manage the gastrostomy or naso-gastric tube and administer their feed safely ensuring minimal disruption to the individual's life style.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	S1	Is there a nurse specialist and dietitian experienced in enteral nutrition?
Records	Check	O1	Is there a recorded assessment that includes height and weight?
	Check	O2	Is there a recorded plan of care?
Patient	Ask	P2	Did the patient/family have the opportunity to discuss enteral feeding at home?
	Check	O3	Can the patient/family demonstrate how to administer feed confidently?
	Ask	O5	Have the patient/family been given the correct supplies to administer feed at home?
	Ask	O4	Can the patient/family state a name and contact number with regard to enteral feeding?
	Check	P7	Did the patient remain free of any complications associated with enteral feeding?

Consensus standards for the nursing management of cystic fibrosis

3.9 Oxygen therapy

Standard statement

The patient receiving long term oxygen therapy (LTOT) and their family will be taught how to administer oxygen safely and effectively.

Structure

- 1. The CF nurse specialist and/or physiotherapist will be available.
- 2. The following resources will be available:
 - oxygen (cylinders, concentrator or liquid oxygen)

- nasal cannulae or mask
- 3. Information regarding flow rate, hours of usage and contact numbers.

Process

- 1. The patient will be assessed to determine flow rate and hours of usage (arterial blood gases and oxygen saturation measurements³¹).
- 2. Together with the patient/family, the nurse specialist and/or physiotherapist assesses and formulates an appropriate plan of action to ensure safe practice.
- 3. The patient/family will be provided with the appropriate oxygen delivery system and ancillaries.
- 4. The patient/family will be provided with contact names and numbers for advice and for problems associated with oxygen delivery system.

Outcomes

- 1. The patient has an assessment documented (arterial blood gases and oxygen saturation).
- 2. The patient/family has a plan of care to meet their individual needs and can state the complications associated with oxygen delivery.
- 3. The patient/family are confident at administering oxygen therapy.
- 4. The patient/family can state who to contact for advice or problems.
- 5. The patient has the appropriate delivery system and ancillaries for their needs.

Audit

Standard Statement (Oxygen therapy)

The patient receiving long term oxygen therapy (LTOT) and their family will be taught how to administer oxygen safely and effectively.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	S1	Is there a nurse specialist available with experience of oxygen therapy?
Records	Check	O1	Is there a documented assessment that includes arterial blood gases and oxygen saturation?
	Check	O2	Is there a recorded plan of care?
Patient	Check	O2	Can the patient/family demonstrate how to use oxygen therapy correctly and safely?
	Ask	S3	Can the patient/family state oxygen flow rate and hours of usage?
	Ask	O4	Can the patient/family state names and contact numbers with regard to oxygen therapy advice?
	Ask	O5	Have the patient/family been given the correct supplies to administer oxygen at home?

Consensus standards for the nursing management of cystic fibrosis

3.10 Cystic fibrosis related diabetes (insulin dependant) Standard statement

The patient with cystic fibrosis related diabetes (CFRD) will be taught to manage their condition, how to obtain diabetic control and how to prevent associated complications.

Structure

- 1. A diabetologist, diabetic nurse specialist and dietitian will be available for all patients newly diagnosed with CFRD.

- 2. The following resources will be available:
 - blood glucose monitoring equipment
 - insulin and pens
 - CFRD plus other relevant literature on diabetes will be available

Process

- 1. The nurse specialist will ensure that the patient is assessed for an appropriate education plan.
- 2. A diabetologist and diabetic nurse specialist will liaise with the CF team.
- 3. The patient will be supervised and instructed regarding:
 - blood glucose monitoring
 - injection sites and techniques
 - disposal of equipment
 - prescribed insulin regimens
 - hypo/hyperglycaemic management
- 4. The patient/family will be provided with contact names and numbers of the CF and diabetic teams.
- 5. The patient will have a diabetic annual assessment.

Outcomes

- 1. The patient/family have access to a diabetologist, diabetic nurse specialist and dietitian with knowledge of CFRD.
- 2. The patient with CFRD has a documented diabetic annual assessment.
- 3. The patient can demonstrate how to monitor their blood glucose level, inject insulin and manage hypo/hyperglycaemia.
- 4. The patient/family has literature relating to CFRD.

Audit

Standard Statement (Cystic fibrosis related diabetes (CFRD))

The patient with CFRD will be taught to manage their condition, how to obtain diabetic control and how to prevent associated complications

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	S1	Is there a diabetologist, diabetic nurse specialist and dietitian available?
	Ask	P2	Can staff members state how the diabetic team and CF team liaise?
Records	Check	P1	Is there a documented education assessment?
	Check	P1	Is there a recorded plan of care?
	Check	O2	Is there a documented diabetic annual assessment?
Patient	Check	O3	Can the patient/family demonstrate how to monitor blood glucose level, inject insulin and manage hypo/hyperglycaemia?
	Ask	P4	Can the patient/family state names and contact numbers with regard to CFRD?
	Ask	O4	Does the patient/family have literature relating to the management of CFRD?
	Ask	S2	Does the patient/family have the correct equipment to control and monitor CFRD?

Consensus standards for the nursing management of cystic fibrosis

3.11 Transfer from paediatric to adult care

Standard statement

Each patient/family will receive continued support from the multi-disciplinary CF team appropriate to their individual needs during transition from paediatric to adult care.

Structure

- 1. Information regarding recognised adult specialist CF centres will be available.
- 2. The CF team consisting of a doctor, nurse, physiotherapist, dietitian, psychologist and social worker will be available at the recognised adult specialist CF centre.

Process

- 1. The patient will be transferred from the paediatric specialist CF centre to a recognised adult specialist CF centre.
- 2. A flexible approach to the age at transfer will be adopted according to individual needs.
- 3. Transition to adult care will be discussed and planned with the patient/family at the appropriate time and a date for transfer agreed.
- 4. The CF nurse specialist will act as a key person to liaise between the paediatric and adult specialist CF centres during the transition period.
- 5. The patient/family will have the opportunity to visit the adult specialist CF centre on a social basis.
- 6. Contact names and numbers of the adult CF team will be provided.
- 7. A clinic appointment with the adult CF team will be offered prior to the final paediatric appointment.
- 8. The paediatric CF team will promote autonomy of care prior to transfer.

Outcome

- 1. A date for transfer is agreed between the patient/family, the paediatric CF team and the adult specialist CF centre.
- 2. The patient/family are able to outline the plan for the process of transition.
- 3. The patient/family will have been offered the opportunity to visit the adult specialist CF centre prior to transfer.
- 4. The patient/family has the names and contact numbers of the adult CF team.
- 5. The patient will have an outpatient appointment with the adult specialist CF centre.

Audit

Standard Statement (Transfer from paediatric to adult care)

Each patient/family will receive continued support from the multidisciplinary CF team appropriate to their individual needs during transition from paediatric to adult care.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	P1	Can staff state the adult specialist CF centre where the adolescent's care will be transferred?
	Ask	P4	Can staff state the key person who will liaise between the paediatric and adult specialist CF centres?
Patient	Ask	O1	Does the patient and their family know when their agreed date of transfer is?
	Ask	O2	Has the transition to adult care been discussed with the patient and their family?

Patient	Ask	O3	Did the patient and family have the opportunity to visit the adult specialist CF centre on a social basis?
	Ask	O4	Have the patient and family been given names and contact numbers of the CF adult team?
	Ask	P7	Has a clinic appointment been made with the adult CF team prior to the patient's final paediatric appointment?

Consensus standards for the nursing management of cystic fibrosis

3.12 Terminal care and symptom management

Standard statement

Patients in the terminal stages of their disease will receive support, symptom control and care appropriate to their individual needs to allow them to die with dignity.

Structure

- 1. A CF nurse specialist will be available.
- 2. Appropriate support services will be available.

Process

- 1. Together with the patient/family the CF nurse specialist assesses and formulates an appropriate plan of care.
- 2. The patient/family will have the opportunity to choose home or hospital based terminal care. This arrangement will be flexible with changing needs.
- 3. Regular assessment and monitoring of symptoms will be carried out and management planned accordingly.
- 4. The patient/family will be kept informed of changes and have the opportunity to express their feelings and needs.
- 5. The nurse, in conjunction with other experienced professionals, will provide:
 - management of pain and symptoms (table 1)
 - a support structure to the patient/family during end stage and the period following death
 - education and support for colleagues involved in the care.
- 6. Bereavement support will be identified and offered.

Outcome

- 1. The patient/family have a plan of care that meets their needs.
- 2. The patient/family's decision for hospital or home care is recorded along with any management changes.
- 3. The patient/family can state who to contact for support and advice.
- 4. The family will state that all relevant equipment and medication is available to provide symptom control.
- 5. A support service for staff is available.
- 6. Bereavement support is organised according to the wishes of the family.

Symptom control

Suggested approaches

Problem	Pharmaceutical management	Other measures
Dyspnoea	<ul style="list-style-type: none"> ▪ oxygen ▪ morphine syrup (titrate and breakthrough) ▪ MST (maintenance) ▪ IV/SC diamorphine infusion (if no oral route) ▪ nebulised morphine ▪ fentanyl patches (slow release) ▪ bronchodilators salbutamol terbutaline 	<ul style="list-style-type: none"> ▪ fan positioning ▪ sitting over pillows ▪ high side lying
Viscous lung secretions	<ul style="list-style-type: none"> ▪ mucolytic agents ▪ DNase ▪ N-acetylcysteine ▪ Nebulised saline (hypertonic) 	<ul style="list-style-type: none"> ▪ adequate hydration (oral/enteral/IV) ▪ humidification ▪ modified physiotherapy
Copious secretions	<ul style="list-style-type: none"> ▪ review mucolytics ▪ anticholinergic agents if distressed: <ul style="list-style-type: none"> ▪ atropine ▪ hyoscine 	<ul style="list-style-type: none"> • oropharyngeal suction if necessary
Oedema	<ul style="list-style-type: none"> ▪ diuretics if symptoms are troublesome 	
Anxiety	<ul style="list-style-type: none"> ▪ diazepam (oral, PR, IV or infusion) ▪ chlorpromazine ▪ midazolam infusion 	<ul style="list-style-type: none"> ▪ reassurance and security ▪ information and involvement distraction ▪ complementary therapies ▪ counselling
Mobility		<ul style="list-style-type: none"> ▪ wheelchair ▪ commode/urinal/bedpan ▪ pressure relieving mattress ▪ attention to pressure areas and personal hygiene
Loss of appetite	<ul style="list-style-type: none"> ▪ metoclopramide ▪ cyclizine ▪ prochlorperazine ▪ domperidone 	<ul style="list-style-type: none"> ▪ avoid strong smells ▪ small amounts of food and drink
Constipation	<ul style="list-style-type: none"> ▪ laxatives and stool softeners ▪ senna, lactulose (consider starting with opiates) 	<ul style="list-style-type: none"> ▪ hydration ▪ diet
Pain	<ul style="list-style-type: none"> ▪ identify and treat source with appropriate analgesia (eg non-steroidals) 	<ul style="list-style-type: none"> ▪ involve symptom control/pain team ▪ patient controlled analgesia ▪ TENS

Audit

Standard Statement (Terminal care)

Patients in the terminal stages of their disease will receive support, symptom control and care appropriate to their individual needs to allow them to die with dignity.

Key to table: S = structure, P = process, O = outcome			
Target Group	Method	Code	Audit Criteria
Staff	Ask	S1	Is there a CF nurse specialist available to advise on terminal care?
	Ask	O5	Can staff identify what support is available?
	Check	O3	Is there written information stating CF team members and contact numbers?
	Ask	O4	Can staff state what equipment and medication is available for symptom control?
Records	Check	P1	Is there an assessment of symptoms and needs recorded?
	Check	O1	Is there a plan of care recorded?
	Check	O2	Has the option of home or hospital care been discussed with the patient/family and documented?
	Check	O6	Have the bereavement needs of the family been assessed and documented?

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Cystic Fibrosis Trust
2nd Floor
One Aldgate
London EC3N 1RE

020 3795 1555
enquiries@cysticfibrosis.org.uk
cysticfibrosis.org.uk

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At the Trust we are:

- Investing in cutting-edge research
- Driving up standards of clinical care
- Providing support and advice to people with CF and their families
- Campaigning hard for the issues that really matter

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