

**AUSTRALIAN & NEW ZEALAND NURSING  
STANDARDS OF CARE  
FOR  
CYSTIC FIBROSIS**



**The Thoracic Society**  
of Australia & New Zealand  
LEADERS IN LUNG HEALTH



**2015**

*A consensus statement prepared by the  
Australian and New Zealand Cystic Fibrosis Nurses Group*

## FOREWORD

The following Australasian Nursing Standards of Cystic Fibrosis Care document has been written by nurses with specialist knowledge of, and experience in, the physiological, psychological and functional impact of cystic fibrosis on affected individuals and families. The clinical practice standards are evidence-based where evidence has been found in the currently available literature and/or as agreed by the authors. Formal grading of the evidence has not been undertaken for this version in deference to the new grading system to be applied to the review of the Australasian Standards of Care for Cystic Fibrosis 2008, due for completion in 2017.

The creation of this document has been supported by Cystic Fibrosis Australia and has been endorsed by the Thoracic Society of Australia and New Zealand.

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We are grateful to Anna Montgomery (RN) who gave generously of her time to provide expert opinion and editing advice.

## INTRODUCTION

### **Background**

Nursing care of people with cystic fibrosis (CF) is multifactorial. The CF Nurse Specialist is involved in the overall management of the patient and support of the family. Areas of practice include advocacy, clinical management, support and advice, research and education of patients, families and staff involved in the management of patients with CF as well as education of external agencies such as schools and community organisations.

To date there has been no specific document to guide nursing care of CF in Australia and New Zealand. These guidelines are intended to outline the key nursing roles and responsibilities in the care of people with CF from diagnosis through the lifespan.

This document will refer to the Nurse Specialist. This can be translated to any suitably experienced nurse whose role is focused on the day to day management and nursing care of people with CF regardless of the title he or she holds.

### **Purpose and Scope**

The goal of this document is to optimise provision of nursing care for all people with CF in Australia and New Zealand.

The purpose of these Standards of Care is:

- To provide recommendations supporting best practice in the identified key areas of nursing care for infants, children and adults with CF.
- To standardise the nursing care provided to people with CF across Australia.
- To promote nursing care to people with CF that is based on the best available evidence and expert knowledge
- This document can be used a reference tool to support the training of nurses in the care of people with CF as well as provide direction to nurses who care for people with CF in rural and remote settings.

## INTRODUCTION

### Consultation Process

The value of having a document outlining nursing standards of care has been recognised now for several years. Following the development of the National Standards of Care document and similar documents from the Cystic Fibrosis Physiotherapy and Nutrition groups, it is timely that nursing formalised their practice through their own standards of care document.

### Timeline

August 2009 - State representatives were identified to coordinate the writing of the document, incorporating feedback from their states. A steering committee was also formed made up of members from the state representative group.

November 2009 - Steering committee met and identified key areas for the standards of care document.

Nov 2009 – June 2010 - Key areas that had been identified were allocated to each state to begin developing.

June 2010 - Steering committee met, collated and formatted the key areas that had been written by the individual states.

August 2010 - Work to date was presented at the Biannual Australian and New Zealand CF Nurses Conference in Perth. Comments and suggestions were sought from the group.

September 2010 – March 2011 - First draft was finalised and formatted incorporating feedback provided to date.

August 2011 - Draft document presented at the 9<sup>th</sup> Australian New Zealand Cystic Fibrosis Conference (Melbourne).

February 2012 – August 2014 - Draft document submitted to key stakeholders for final review and changes.

September 2014 – Final draft submitted to Cystic Fibrosis Australia and The Thoracic Society of Australia and New Zealand for ratification.

October 2014 – June 2015 – Inclusion of additional evidence

August 2015 – Launch

## INTRODUCTION

### **Methods**

A combination of review of current literature and expert opinion has been used to guide the development of these standards. The recommendations have been written by specialist nurses with experience in the area of CF and are based on the evidence that is available. They were formulated by general agreement amongst the writing group.

### **Review Process**

This document will be reviewed every 4 years. The review will include updating new evidence and incorporate changes based on the evaluation of the evidence.

### **Disclosure Statement**

The development of this document has been funded in part by unrestricted education grants from Roche pharmaceuticals and Cystic Fibrosis Australia. The funding was used to support travel for the steering committee members to meet and to facilitate editing of the document. There has been no involvement in the formulation of the standards document by any parties who provided funding support.

The following authors have declared Conflicts of interest having received reimbursement of expenses incurred as a result of participation on Advisory Panels or attendance at professional development meetings.

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## NURSING STANDARD of CARE 1: THE ROLE OF THE CF NURSE SPECIALIST

*“Nursing is a partnership between the patients, carers and nurses”.*

National Consensus Standards for the Nursing Management of Cystic Fibrosis. UK Trust 2001.

The CF Nurse Specialist is involved in all aspects of the overall management of the patient. These include the domains of clinical management and leadership, advocacy, education and research. These five domains are essential to inform a holistic plan of care for the patient and family.

The CF Nurse Specialist provides an important link for the patients and families to access both hospital and community based services.

This document acknowledges that there may be variation within the nursing role between Centres. For the purpose of this document, the term CF Nurse Specialist applies equally to the following; the Clinical Care Coordinator, Clinical Nurse Consultant and Clinical Nurse Specialist, Clinical Nurse and Nurse Practitioner.

### Clinical Management

- Responsible and accountable for assessing, recommending and coordinating the multi-disciplinary plans of care for people with cystic fibrosis.
- Triage and develop an interim management plan with referral to the appropriate members of the specialist medical and allied health team.
- Ensure continuity of care between the acute and ambulatory settings.
- Facilitate the transition from paediatric to adult care.
- Ensure infection control best practice is maintained.

### Advocacy

The CF Nurse Specialist will:

- Ensure patients and families have access to information and resources to enable them to make informed choices about their health care.

## NURSING STANDARD of CARE 1: THE ROLE OF THE CF NURSE SPECIALIST

- Facilitate communication between people with CF and health care providers.
- Respond to issues raised that have the potential to increase risk or reduce quality of patient care.
- Liaise with other specialist care providers, hospitals, health care and community agencies to engage services on behalf of people with CF and families as required.
- Prepare supporting documentation to ensure adequate resources for CF care.

### Leadership

The CF Nurse Specialist will:

- Establish collaborative relationships with the cystic fibrosis specialist team and other cystic fibrosis care providers.
- Mentor nurses with a special interest in cystic fibrosis to enable succession planning to ensure continuity of service provision.
- Foster links with colleagues in specialist centres nationally and internationally.
- Maintain membership of relevant professional bodies.
- Develop and support evidence-based clinical practice.
- Contribute to policy development as it relates to quality and safety of delivery of nursing care.

### Education

The CF Nurse Specialist will:

- Develop and provide education resources for people with CF, families, carers and the community.
- Develop and provide education resources for healthcare professionals caring for people with CF.
- Participate in ongoing professional development.

**NURSING STANDARD of CARE 1: THE ROLE OF THE CF NURSE SPECIALIST**

**Research**

The CF Nurse Specialist will:

- Monitor health outcomes according to key performance indicators.
- Develop and evaluate evidence-based programs to improve outcomes.
- Actively conduct, participate in and promote nursing research.
- Present and/or attend at local, national and international scientific meetings.
- Support and encourage junior researchers.

**NURSING STANDARD of CARE 2: PAEDIATRIC DIAGNOSIS**

**CF Standards of Care, Australia (2008) Section 2: Newly diagnosed children with cystic fibrosis (P22-23)**

**Standard 1**

Infants diagnosed through newborn screening should have prompt access to experienced medical and allied health CF personnel.

**Standard 2**

Following diagnosis, infants should be assessed for pancreatic insufficiency and enzyme therapy commenced when indicated.

**Standard 3**

Families should have access to up to date educational material regarding CF from the time of diagnosis.

**Standard 4**

Families of infants diagnosed with CF by newborn screening should have access to genetic counselling services.

**Standard 5**

Salt, electrolyte and vitamin replacement therapy should be considered in all infants at diagnosis; particularly in those geographical areas where hot and humid conditions prevail.

## NURSING STANDARD of CARE 2: PAEDIATRIC DIAGNOSIS

*Giving the diagnosis of cystic fibrosis needs to be handled confidently and sensitively. Families describe the way they were told about the diagnosis of cystic fibrosis for many years to come and, if done poorly, it can be a long term source of sadness and anxiety. (National Consensus Standards for the Nursing Management of Cystic Fibrosis, UK Cystic Fibrosis Nurses Specialist Group, CF Trust, May 2001, p4)*

**Nursing Standards of Care** as per Cystic Fibrosis Standards of Care, Australia 2008 (p22-23)

**Newly diagnosed patients should ideally be transferred to a paediatric tertiary CF treatment centre for initial assessment and parental education.**

- If the diagnosis is given by a local paediatrician or GP, the CF Nurse Specialist should make contact with the parents/carers the same day, or as soon as possible, after the parents receive notification of the diagnosis.
- If the diagnosis is to be given by the CF team, the CF Nurse Specialist should contact the family and arrange an appointment at the parents/carers earliest convenience. The diagnosis should be given in a face to face meeting, rather than over the phone whenever possible.
- The CF Nurse Specialist should ensure appropriate tests have been ordered, and carried out, to confirm diagnosis. This should include but is not limited to the following; repeat IRT, bloods for genetic typing and sweat testing. Sweat testing should be carried out in an accredited laboratory.
- During the initial discussion on the phone with the family, the information to be given should include, but is not limited to;
  - what is factually known from the tests carried out,
  - explanation of what tests (if any) still need to be conducted,
  - who makes up a CF team and who they will see at the hospital,
  - the approximate length of time they will need to be present to complete the post diagnosis education,

## NURSING STANDARD of CARE 2: PAEDIATRIC DIAGNOSIS

- suggest some suitable web sites if the family wish to do their own search about CF prior to meeting with the CF team.
- The CF Nurse Specialist will ensure the parents/carers are aware of the availability and the role of both a genetic counsellor and a social worker post diagnosis.
- Arrangements for the patient, with their parent/carer to be reviewed at a recognised CF centre should be made as soon as is suitable for the family. Minimising the time between diagnosis and initial contact with the CF team is important. (Sawyer and Glazner 2004)
- New patients should be reviewed in an outpatient or “care by parent” environment when possible, and medically suitable, to avoid unnecessary mixing with children who are unwell in a ward environment.

### Initial Education

- The CF Nurse Specialist should co-ordinate the education process for the family ensuring they have access to all members of the CF team. They should ensure there is ample time for each team member.
- The patient’s allocated physician would normally be the first person to see the family/patient and start the education process. The CF Nurse Specialist should be present during this education session whenever possible to explain anything the family need further clarification on.
- The CF Nurse Specialist should make sure the education and testing is spread out over a suitable period of time to ensure the family do not become too overwhelmed with the amount of information they are receiving.
- The CF Nurse Specialist should ensure the family/carer are aware of the following:
  - The role of the CF Nurse Specialist.
  - How and when to contact the CF Nurse Specialist.

NURSING STANDARD of CARE 2: PAEDIATRIC DIAGNOSIS

- Management of changes in respiratory symptoms.
- Initial medication requirements including dosage and role in the care of CF.
- How to obtain ongoing medications for the patient.
- What happens at CF clinic including how to make appointments and its location.
- Written and/or visual material should be provided to supplement the verbal education. This should include but not be limited to:
  - Daily management plan
  - Plan for changes in symptoms
  - Contact numbers
  - Informative websites
  - Basic written CF information to share with their family and friends

#### Follow-up

- The CF Nurse Specialist will ensure the patient has a follow up clinic appointment in a timely manner.
- It would be suitable for the CF Nurse Specialist to make regular phone contact with the family in the days immediately following education to ensure no new questions had arisen and the family was coping well with trying to instigate new treatments.
- The CF Nurse Specialist will ensure information is sent to local supports as identified by the parents/carer. This may include but is not limited to general practitioner, paediatrician, baby health nurse, local Cystic Fibrosis Association.
- The CF Nurse Specialist will explain the concept and importance of an annual review.

#### References

Cystic Fibrosis Standards of Care, Australia. 2008

Sawyer, S.M and Glazner, J.A “What follows newborn screening? An evaluation of a residential education program for parents of infants with newly diagnosed Cystic Fibrosis.” *Pediatrics* 2004;114(2):411-6

**CF Standards of Care, Australia (2008) Section 2: Newly diagnosed children with cystic fibrosis (P22-23, 26-28)**

**Standard 1**

Investigations such as a sweat test (sweat chloride >60mmol/L) and standard CFTR mutation testing may not be confirmatory in many cases of “atypical” CF.

**Standard 2**

For those patients with sinopulmonary symptoms, careful clinical assessment for extra-pulmonary symptoms and medical imaging of the sinuses and lungs is required (CT scan of the sinuses and high resolution CT chest).

**Standard 3**

After a diagnosis of “atypical” CF has been made, it is recommended that the patient receive medical follow-up by a physician expert in the management of patients with CF.

### NURSING STANDARD of CARE 3: ADOLESCENT AND ADULT DIAGNOSIS

*Giving the diagnosis of Cystic Fibrosis needs to be handled confidently and sensitively. A late diagnosis when an individual is an adolescent or adult can have a profound psychological impact on them and their families. This will also have implications for the adult or adolescent's family in relation to cascade genetic screening. . (National Consensus Standards for the Nursing Management of Cystic Fibrosis, UK Cystic Fibrosis Nurses Specialist Group, CF Trust, May 2001, p4)*

**Nursing Standards of Care** as per Cystic Fibrosis Standards of Care, Australia 2008 (pp 22- 23 & 26-28)

**Newly diagnosed patients should ideally be transferred to a tertiary CF treatment centre for initial assessment and education.**

- The CF Specialist Respiratory Physician involved in the diagnostic investigations is the appropriate person to give the diagnosis to the individual. The CF Nurse Specialist will contact the family and arrange an appointment at the earliest mutual convenience where the diagnosis can be given face to face. Minimising the time between diagnosis and initial contact with the CF team is important. (Sawyer and Glazner 2004)
- New patients should be initially reviewed in an outpatient area that is not the CF Clinic. This is to minimise the impact the diagnosis may have on the individual, as mixing with adults with CF may have a negative psychological impact when newly diagnosed. It also reduces the chance of cross infection.
- The CF Nurse Specialist may be required to provide initial support and counselling. They will provide the contact details and explain the relevance of the role of both a genetic counsellor and a social worker or clinical psychologist post diagnosis.
- The CF Nurse Specialist will be involved in the planning the most appropriate follow up for the individual including education, review of their health status and/or further diagnostic testing.

**NURSING STANDARD of CARE 3: ADOLESCENT AND ADULT DIAGNOSIS**

**Initial Education**

- The CF Nurse Specialist will co-ordinate the education process for the individual and their families ensuring they have access to and ample time with all appropriate members of the CF team.
- The allocated CF Physician would normally be the first person to see the individual and their families to start the education process. The CF Nurse Specialist should be present during this education session, whenever possible; to be able to explain anything the family may need further clarification about.
- The CF Nurse Specialist will, wherever possible, ensure the education and health status assessment is spread out over a suitable period of time and is individually tailored. This is to reduce the chance of the individual and their families becoming overwhelmed with the amount and type of information they are receiving.
- The CF Nurse Specialist should ensure the individual and their families are aware of the following:
  - The role of the CF Nurse Specialist and the other CF team members.
  - How and when to contact the CF team.
  - Recognition and management of changes in symptoms.
  - Initial medication requirements including dosage, frequency, delivery route and relevance in the treatment of their condition.
  - How to obtain ongoing medications.
  - How to obtain, use and care for new equipment.
  - How to make future appointments.
  - The location of appropriate areas of the hospital.

### NURSING STANDARD of CARE 3: ADOLESCENT AND ADULT DIAGNOSIS

- Written and/or visual material should be provided to supplement the verbal education. This should include but not be limited to:
  - A management plan for each co-morbidity - including a medication list.
  - A plan for management of changes in symptoms.
  - Contact numbers/email addresses.
  - Suitable websites.

#### Follow-up

- The CF Nurse Specialist will ensure the patient has a follow up appointment in a timely manner, with the appropriate team members and in an appropriate venue.
- The CF Nurse Specialist will have ongoing phone and/or email contact with the individual and/or family after initial diagnosis and review, providing support and resolving any problems with treatments and answering questions.
- The CF Nurse Specialist will ensure information is sent to appropriate contacts such as the general practitioner and/or specialist health care professionals.
- The CF Nurse Specialist will explain the concept and relevance of an annual review and arrange this on an ongoing basis in consultation with their treating CF physician.

#### References

Cystic Fibrosis Standards of Care, Australia. 2008

Sawyer, S.M and Glazner, J.A. "What follows newborn screening? An evaluation of a residential education program for parents of infants with newly diagnosed Cystic Fibrosis." *Pediatrics*. 2004;114(2):411-6.

National Consensus Standards for the Nursing Management of Cystic Fibrosis: UK Cystic Fibrosis Nurse Specialist Group, Cystic Fibrosis Trust. May 2001.

**NURSING STANDARD of CARE 4: INPATIENT CARE**

**CF Standards of Care, Australia (2008): Inpatient Care**

**Standard 1**

All hospitals should recognise the characteristics and needs of people with CF.

**Standard 2**

All hospitals should have available facilities, staff, and services to manage all complications of CF or the ability to arrange early transfer to an appropriate hospital.

**Standard 3**

All hospitals should have ward accommodation that meets the needs of people with CF, particularly with regard to optimal infection prevention.

**Standard 4**

All should have plans for emergency assessment and admission.

**Standard 5**

When people with CF are admitted to hospital and not under the direct care of the CF team, there should be prompt communication with the CF centre staff to ensure patients have access to the services, staff and facilities needed for optimal care.

**Standard 6**

There should be protocols for management of all common complications of CF in hospitals to which people with CF are admitted/treated.

## NURSING STANDARD of CARE 4: INPATIENT CARE

*People with CF frequently require admission to hospital. The most common reason for admission is a respiratory exacerbation. Less frequent reasons include complications of respiratory disease, gastrointestinal disease, liver disease, unstable diabetes, psychological problems or peri-operative care.*

*Nursing staff should be aware that patients with CF will have a longer average length of stay than most patients (10-14 days on average). They also need to understand patients will usually not be unwell for most of their admission and should be given leave for school, work, study or recreation.*

### Admission

CF admissions can either be planned or emergent. In each situation, the CF Nurse Specialist will be required to advocate for the patients to ensure their needs are met appropriately during the initial triage and admission process and then throughout the admission to discharge.

- The CF Nurse Specialist will facilitate admission to an appropriate ward area dependent upon patient's potentially infectious microbiological status, age, clinical status and familiar ward areas.
- Patients should be reviewed by the CF Nurse Specialist as soon as practical after admission and as needed throughout the admission.
- The CF Nurse Specialist is mindful of the importance of parent/carer support for the patient and will liaise with local supports to coordinate the availability of supports e.g. accommodation, car parking, transport.

### Venous Access

The CF Nurse Specialist should ensure that venous access is planned either prior to, or on admission.

## NURSING STANDARD of CARE 4: INPATIENT CARE

CF Nurse Specialists need to ensure that:

- Intravenous access for each person is individualised based on knowledge of their history of access.
- Options for intravenous access should be discussed with patient and family at each admission.
- Clinical staff are made aware of hospital guidelines around safe management of the patient's intravenous access.

NB: Needle/procedural phobias are important considerations; sedation or general anaesthesia may be indicated. Diagnosed phobias require referral to clinical psychology or other appropriate therapeutically relevant services.

### Clinical Care

Nurses caring for patients with CF on the ward must plan, assess, implement and monitor care according to the needs of each patient at every admission, taking into account the variability of the disease.

- The CF Nurse Specialist will ensure that each patient has access to allied health members of the CF team.
- The CF Nurse Specialist will advise on patient care according to local centre guidelines and best practice. This will include day to day care as well as management of complications such as haemoptysis, DIOS, pneumothorax, CFRD.
- The CF Nurse Specialist will provide education to local ward staff.

### Discharge Planning

Expected date of discharge should be determined on the day of admission to optimise discharge planning.

- Arrangements for follow-up are to be discussed with the patient and family.
- In consultation with the patient, develop an ongoing management plan, with referral to the appropriate members of the specialist medical and allied health team.
- Discharge treatments are outlined and education provided for any new or changed treatment.

**NURSING STANDARD of CARE 4: INPATIENT CARE**

- Patients and families should be made aware of community services that can assist them to manage their treatment at home and appropriate referral can be made.
- Community services that have long standing relationships with families are to be made aware that the patient is due to return home, e.g. for timing of TIVAD flushing etc.
- Ensure discharge summaries are completed and sent to the appropriate care providers.

**References**

Cystic Fibrosis Standards of Care, Australia. 2008

National Consensus Standards for the Nursing Management of Cystic Fibrosis: UK Cystic Fibrosis Nurse Specialist Group, Cystic Fibrosis Trust. May 2001.

**CF Standards of Care, Australia (2008) Section 4: Outpatient Care (p33-36)**

**Standard 1**

Treatment should be coordinated by a multidisciplinary team in specialised CF centres.

**Standard 2**

All patients should be seen at least four times per year (including at least twice by the CF specialist team).

**Standard 3**

Access to the specialist CF multidisciplinary team should be available at all clinics.

**Standard 4**

An annual review, including appropriate tests, should be undertaken followed by a written report to the general practitioner. This should include assessment of current status and progress.

**Standard 5**

Adequate policies, facilities and procedures should be in place to comply with, and promote infection control guidelines.

**NURSING STANDARD of CARE 5: OUTPATIENT CARE**

*The review of ambulatory patients in an outpatient setting should be undertaken by a multidisciplinary team that consists of a CF specialist physician, nurse, physiotherapist, dietitian and social worker, with access to other specialist services as clinically indicated.*

**Nursing Standard of Care** as per Cystic Fibrosis Standards of Care, Australia 2008 (p33-36)

**Recommendations for Clinic**

The CF Nurse specialist will:

- Ensure that patients and families are aware of the availability of CF outpatient services and how to contact the clinic, make and reschedule appointments.
- Facilitate access to the allied health multidisciplinary team as per individual patient assessment.
- Triage the patient and undertake a clinical assessment.
- Perform clinical procedures as required e.g. TIVAD flushing, gastrostomy care, sputum collection, vaccinations.
- Conduct an Annual Review as per best practice. (CF Standards of Care)
- Ensure that the patient and their families/carers have timely access to education, support and equipment at the completion of both routine and annual reviews.
- Have access to the most recent result of each individual's sputum microbiology test to ensure effective infection control is maintained.
- Ensure that at each clinic appointment, the patient has appropriate tests and investigations as clinically indicated.
- Promote efficient use of clinic time by guiding patient flow through the clinic.
- Establish strategies to identify and contact patients who fail to attend clinic appointments.
- Ensure follow up arrangements are in place according to local policy e.g. results, referrals to other services, and future clinic appointments.

Note: At first visit to clinic (not including neonatal diagnosis) the patient is to be isolated until confirmation of sputum microbiology results.

**NURSING STANDARD of CARE 5: OUTPATIENT CARE**

**Recommendations for Outreach/Shared Care Clinic**

The CF Nurse Specialist:

- Will be the point of contact for patient and families to facilitate access to appropriate health care services.
- Liaises with local services to facilitate continuity of care.
- Provides information to patient and families on how to access local services.
- Will provide a current plan for management of an acute exacerbation. This should preferably be developed with the patient and/or family and communicated to other health care teams involved.
- Will ensure that patients and families are aware of the availability of CF Outreach Clinics and how to contact the clinic, make and reschedule appointments.
- Will facilitate Outreach Clinic/s, with consideration given to;
  - individual's microbiology results to ensure effective infection control is maintained (at first visit to clinic, not including neonatal diagnosis, the patient is to be isolated until confirmation of sputum microbiology results),
  - appropriate tests and investigations as clinically indicated,
  - patient and their families/carer having timely access to education, support and equipment at the completion of both routine and annual reviews,
  - processes being in place for the documentation of all clinic visits (routine and annual review) and hospitals admissions being made available to the local health care providers in a timely fashion,
  - follow up arrangements according to outcome of clinical review, e.g. results, referrals to other services, and future clinic appointments.

## References

Cystic Fibrosis Standards of Care, Australia. 2008

National Consensus Standards for the Nursing Management of Cystic Fibrosis: UK Cystic Fibrosis Nurse Specialist Group, Cystic Fibrosis Trust. May 2001.

**NURSING STANDARD of CARE 6: SEXUAL AND REPRODUCTIVE HEALTH**

*Adolescents and young adults with CF deserve health care professionals who are knowledgeable about sexual and reproductive health issues with strong communication skills. (Sawyer, 1996)*

**Initial Education**

- The CF Nurse Specialist co-ordinates the new diagnosis education process for the family ensuring that this includes information specific to sexual and reproductive health issues, including probable male infertility.
- Age appropriate information (initiated no later than mid-adolescence)
  - The CF Nurse Specialist should ensure that each patient/parent/partner receives verbal and written information at age and illness appropriate stage.
  - The CF Nurse Specialist facilitates access to the appropriately qualified health care professional.

<b>Gender Specific Topics</b>	
Male	Female
Difference between impotence and infertility	Fertility status
Safe sex practices (condoms)	Safe sex practices (condoms)
The role of semen analysis	Contraceptive options
Small volume ejaculate	Impact of antibiotics on vaginal flora
Reproductive options	Preconception counselling – potential health implication for mother and foetus
	Pregnancy testing and decision making
Continence	Continence
Genetic counselling – partner screening	Genetic counselling – partner screening
Illness severity and sexual intimacy	Illness severity and sexual intimacy

(Sawyer and Glazner, 2003).

## NURSING STANDARD of CARE 6: SEXUAL AND REPRODUCTIVE HEALTH

### Pre, Peri and Post Natal Care Education

- The CF Nurse Specialist should ensure that each patient receives access to information, support and counselling from appropriately qualified health care professionals.

The framework should include, but not be limited to:

#### Prenatal

- Pre-conception counselling:
  - genetic counselling
  - partner carrier screening
  - discussion of potential risks to the mother, the risks to the foetus
  - short and long term issues surrounding parenthood in CF.
- A strong emphasis needs to be placed on achieving the best possible respiratory and nutritional status prior to conception.
- Education on the interactions of currently prescribed medications and treatments and pregnancy.
- Access to information and education on normal physiological adaptations during pregnancy.

#### Perinatal

- The CF Nurse Specialist facilitates:
  - Regular review with both the specialist CF and obstetric teams.
  - Access to antenatal education, including birthing options, analgesia and breast feeding.

#### Postnatal

- Ensure that maternal genotype is noted on the Neonatal Screening request.
- The CF Nurse Specialist facilitates prompt review with the specialist CF Team post birth.
- Ensure that referral has been facilitated to local maternal and child health services.

## References

Cystic Fibrosis Standards of Care, Australia (2008).

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**NURSING STANDARD of CARE 7: TRANSITION**

**CF Standards of Care, Australia (2008) Section 7: Transition from Paediatric to Adult Care (p48-49)**

**Standard 1**

Transition from paediatric to adult care involves a process of close cooperation between paediatric and adult specialist care teams.

**Standard 2**

All CF Centres should have a transition program incorporating active education on adult issues, i.e. fertility and the processes should engage the young person with CF and their family in a positive way.

**Standard 3**

The concept of transition should be raised soon after diagnosis with more active discussions commencing around secondary school entry (12 years) and the process finishing with transfer to adult care around school leaving age (18 years).

**Standard 4**

Paediatric and adult specialist care teams should meet regularly to discuss individual's transitions.

**Standard 5**

The adult co-ordinator should meet individuals during the year before transfer and the adolescent should have the opportunity to visit the adult CF Centre at this time.

**Standard 6**

A comprehensive summary of medical and social issues should be available to the adult team well in advance of transfer. The local CF Association can be involved to help facilitate the process.

## NURSING STANDARD of CARE 7: TRANSITION

*“Transition is defined as the process of preparing for the transfer from paediatric to adult care systems. The transfer must facilitate chronically ill children beginning a productive life and achieving social integration as independent adults in society (Schidlow 2002; Sawyer, Drew et al. 2007). The objective of an integrated CF service is to develop a local programme for transition that addresses the young person’s concerns and considers the factors affecting their health at the time of transition (Craig, Towns et al. 2007).” Cystic Fibrosis Standards of Care, Australia 2008 p47*

### Guiding Principles

- All CF centres should have a transition program that prepares children and adolescents with CF to transfer to an Adult CF centre.
- Transition involves a judgement on the optimal time for an individual to transfer to the adult centre and should include specific educational material to facilitate the process.

Transition is a four staged process – Early, Middle, Late and Completion.

### Early Stage (13 - 14 yrs)

The young person is supported to practice skills directed at self care.

- The Paediatric CF Nurse Specialist introduces the concept of transition when the patient is 13 years old, to both patient and family.

### Middle Stage (15- 16yrs)

The young person practices skills, gathers information and actively participates in his/her own care.

- The Paediatric CF Nurse Specialist facilitates a series of multidisciplinary joint clinics (e.g. Adolescent Clinics) where the patient and family meet and interact with the Adult Team.

## NURSING STANDARD of CARE 7: TRANSITION

### Late Stage (17 -18yrs)

The young person uses independent behaviours to move into adult care.

- The Paediatric CF Nurse Specialist facilitates the final handover to the adult team, including multidisciplinary case conference and comprehensive summary.

### Completion Stage

The Adult CF Nurse Specialist facilitates follow-up contact and access to CF Services within 2 weeks of the final transition clinic.

### Resource Tool

The framework for the resource tool to support the transition process should include, but not be limited to:

1. What is Transition?
  - When Does Transition Start
  - Continuing the Journey
2. Adolescent Clinics
3. The Adult CF Service
  - What Can I Expect When I Get There?
  - Differences between paediatric and adult services
  - How do I contact the Adult CF Team?
  - Where are CF Clinics held?
  - What happens at the CF Clinic?
- What happens when I am admitted to the adult hospital?
- Prescriptions
- General Practitioner

## NURSING STANDARD of CARE 7: TRANSITION

4. Confidentiality
5. Meet the Team
6. References

### Process

The Paediatric and Adult CF centre staff should facilitate transfer to adult care by ensuring regular communication and documentation.

The role of the Paediatric and Adult CF nurse specialist in preparing the patient and family is to:

- develop transition protocol with accompanying documentation, including:
  - checklist, patient information
  - parent/care provider information
- Facilitate joint clinic with Paediatric and Adult Teams
- Accept that it may take several years for patients to develop therapeutic relationships with the new care team and for families to trust also.

### Suggested Resources

1. "MOVING ON TO CHARLIES" Sir Charles Gardiner CF Services (Western Australia)
2. "Heading North" RCH/TPCH CF Services (Queensland)
3. [www.health.nsw.gov.au/gmct/transition](http://www.health.nsw.gov.au/gmct/transition)

### References

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**NURSING STANDARD of CARE 8: TRANSPLANT**

**CF Standards of Care, Australia (2008) Section 9**

**Standard 1**

End of life care discussions, including the role of lung and/or other organ transplantation, should occur before predicted life expectancy is less than two years.

**Standard 2**

Clinical management of the patient during end of life should include a combination of active treatments and therapies to enhance symptom control.

**Standard 3**

Patients with end stage lung and/or other solid organ disease should receive timely referral for single, multi-organ or multi-organ sequential transplantation assessment.

**Standard 4**

Equitable access to transplantation services should be available for all patients.

**Standard 5**

There should be optimal post-transplantation care of all CF related complications.

**Standard 6**

Prior to and during the terminal period effective communication with the patient and their family/carers must be provided.

**Standard 7**

Patients with end stage organ failure should have access to palliative care services when available and desired by the patient.

**Standard 8** Following the death of a patient with CF, ongoing support should be available for the family / carers.

## NURSING STANDARD of CARE 8: TRANSPLANT

### Definitions

End of life is a period of time when the clinical status of the patient is inexorably declining and death is likely within a limited time frame (usually two years).

Transplantation and palliative care are the treatment options that should be considered at this time.

Palliative care is defined as the provision of co-ordinated nursing, medical and allied health services for people facing a life-limited illness. Palliative care provides physical, psychological, social, emotional and spiritual support for the patient and their family and friends, ideally in the environment of choice of the patient (World Health Organisation).

### Guidelines for Referral to Lung Transplant Services (Appendix 1)

- FEV1 below 30% predicted and an accelerating rate of decline in FEV1.
- Anyone who has had an exacerbation of pulmonary disease requiring admission to intensive care unit.
- Increasing frequency of pulmonary exacerbations requiring prolonged periods of intravenous antibiotic therapy.
- Refractory and/or recurrent pneumothorax.
- Recurrent haemoptysis not controlled by bronchial artery embolisation.
- Other specific criteria may include:
  - Reduced six minute walk (less than 400m).
  - PA systolic pressure >35mmHg.

## NURSING STANDARD of CARE 8: TRANSPLANT

### The CF Nurse Specialist should:

- Be aware of the clinical criteria for transplantation.
- Be aware of with the current clinical practice guidelines relating to communicating prognosis and discussing end of life issues.
- Orientate the patient and family to the need for consideration of end of life planning including transplantation.
- Provide information and access to supportive therapies as clinically indicated.
- Facilitate and advocate for multidisciplinary referrals to the transplantation services.
- Establish a process to facilitate feedback to the multidisciplinary CF health care team on patient's transplant status (i.e. pre assessment, listed).
- Facilitate and advocate for regular review by multidisciplinary cystic fibrosis health care team for ongoing management of cystic fibrosis.
- Ensure that the patient and family have access to spiritual/pastoral support as desired.

Note: Contact relevant service provider for specific referral criteria for other organ transplants.

### Resources

- Patient Information Guide to Lung Transplantation
  - The Alfred (Vic)
  - The Prince Charles Hospital (Qld)
- Patient Information Guide to Liver Transplantation
  - The Austin (Vic)
- Patient Information Guide to Renal Transplantation
  - The Alfred (Vic)
  - The Royal Melbourne

**NURSING STANDARD of CARE 8: TRANSPLANT**

**References**

Cystic Fibrosis Standards of Care, Australia. 2008

2014 ISHLT Recipient Selection Criteria for Lung Transplantation (personal communication, Prof Greg Snell.

National Consensus Standards for the Nursing Management of Cystic Fibrosis: UK Cystic Fibrosis Nurse Specialist Group, Cystic Fibrosis Trust. May 2001.

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## NURSING CLINICAL PRACTICE STANDARD 1: CYSTIC FIBROSIS BONE DISEASE (CFBD)

### Definition

Reduced bone accretion and accelerated bone loss are thought to contribute to reduce bone mineral density (BMD) in people with CF.

The definition used to indicate reduced BMD is:

- BMD Z-score#  $< -2$  i.e. less than 2 standard deviations from the population average, at one or more site, or
- BMD T-Score<sup>^</sup> of  $-2.5$  at one or more sites.

Cystic Fibrosis Bone Disease (CFBD) is associated with many factors, including:

- Malnutrition
- Vitamin D deficiency
- Sex hormone deficiency and delayed puberty
- Lung disease and infection
- Prolonged corticosteroid exposure
- Reduced physical activity
- Delta F508 genotype

### Assessment

DEXA scanning is used to assess BMD.

- Monitoring and management decisions are usually based on the site with the lowest BMD level.
- Not indicated in children  $<$  eight years due to:
  - Low prevalence of subnormal BMD prior to adolescence
- The frequency of follow up is determined by:
  - Baseline BMD result.
  - Type of pharmacological agent, frequency of, adherence and response, to any treatment initiated.

## NURSING CLINICAL PRACTICE STANDARD 1: CYSTIC FIBROSIS BONE DISEASE (CFBD)

- Emergence of further risk factors, e.g. corticosteroid treatment, a significant decline in lung function, poor nutritional status, delayed puberty, CFRD, fractures, prolonged period of immobility and readiness for lung transplantation.

### Management:

Full allied health team review:

- Optimisation of nutritional status.
- Vitamin D and calcium supplementation +/- a bone resorption antagonist\*.
- Modification of any occupational or habitual exercise risk factors.
- Adopt an active breathing technique (non-percussion) for effective airway clearance.

# Z-score is based on comparison with the mean BMD of an age-matched population and is therefore the parameter of choice when assessing BMD of children but can be used in adults as well.

^ T-score is based on the mean population data for a group of young adults and so is less reliable in assessing BMD in children.

\*Administration of intravenous bisphosphonates is usually undertaken in consultation with an Endocrinologist following assessment and treatment of any dental and/or oral health issues. Early consideration of removal of wisdom teeth is usually given if any are impacted and the young person is approaching lung transplantation (FEV1~40%predicted) and requiring bisphosphonates.

## References:

Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, 2006. Chapter 8.

Osteoporosis.

Cystic Fibrosis Standards of Care, Australia. 2008.

## NURSING CLINICAL PRACTICE STANDARD 2: CYSTIC FIBROSIS RELATED DIABETES (CFRD)

### Definition

Cystic Fibrosis Related Diabetes (CFRD) is considered to be a distinct form of diabetes. The primary cause of CFRD is insulin deficiency resulting from cumulative loss of endocrine pancreatic tissue. The incidence of CFRD increases with age. The median age of diagnosis is 18-21 years. Up to 50% of individuals with CF will have CFRD.

### Clinical signs

- Polyuria
- Polydipsia
- Delayed progression of puberty and/or poor growth velocity
- Inability to gain weight / weight loss
- Unexplained accelerated decline in pulmonary function

### Categories of CFRD

	Fasting Blood Glucose	OGTT: BGL @ 2hours
Normal Glucose Tolerance	< 5.6 mmol/L	< 7.8 mmol /L
Impaired Glucose Tolerance	5.6 – 6.9 mmol/L	7.8 – 11.1 mmol/L
CFRD	> 7.0 mmol/L*	> 11.1 mmol/L**

\*On two consecutive occasions – no OGTT required

\*\* On two separate occasions

### Screening

#### Oral glucose tolerance test (OGTT):

- Recommended annually in those over the age of 10 years.

#### Routine screening should include:

- Opportunistic fasting, random and post-prandial blood sugar testing & HbA1c.

#### Continuous Glucose Monitoring:

Continuous Glucose Monitoring (CGM) has been proven to offer greater insight into glucose excursions. Guidelines have not yet been established to enable CGM as a diagnostic tool.

#### Increased surveillance required during:

- Infective exacerbation.
- Corticosteroid therapy.
- Pregnancy.
- Malnutrition.
- Enteral feeding.
- Other organ dysfunction, e.g. CF related

## NURSING CLINICAL PRACTICE STANDARD 2: CYSTIC FIBROSIS RELATED DIABETES (CFRD)

Individuals with CF and their families should be educated at an early age around the increased risk of developing diabetes, including the signs & symptoms of diabetes.

### Treatment

#### Patients with CFRD:

- Usually require a combination of insulin treatments. Oral agents rarely used.
- Treatment needs to be individually tailored.
- Where appropriate, HEHP diet is maintained, and insulin adjusted accordingly.
- Insulin requirements may fluctuate as health status changes.

### Management

#### CFRD should be managed by a multidisciplinary team including

- CF Physician, CF Nurse, Endocrinologist, Diabetes Nurse Educator (DNE), Dietitian and Physiotherapist.

#### Blood glucose aim

- Fasting: 4-8mmol/L. Post-prandial: <10mmol/L. HbA1c: <7.0%.

#### Newly Diagnosed

- Manage hyperglycaemia (insulin, hydration, monitor biochemistry).
- Refer to multidisciplinary team for review / education.

#### Established CFRD

- Annual diabetes risk assessment as per Endocrinologist (HbA1c, urinary ACR, eye check).
- Consider admission when: fasting required, commencing enteral feeding or oral corticosteroids.
- Ongoing support on a regular basis from multidisciplinary team.

#### CFRD and pregnancy

- CF nurse / DNE closely monitor, pre, during and post pregnancy.
- Regular monitoring required by pregnant individual at home.
- Regular medical review (CF & ENDO).

#### Pregnancy with no previous diagnosis of diabetes:

- Screening is recommended at both 12-16 weeks and 24-28 weeks
- Diagnosis of Gestational Diabetes differs to traditional OGTT  
Fasting  $\geq 5.1$ mmol/L, 1 hour  $\geq 10$ mmol/L, 2 hour  $\geq 8.5$ mmol/L.

#### Travel:

- Counsel individual around correct storage of medications and sick day management.
- Ensure detailed medical letter and medication list is completed.

**NURSING CLINICAL PRACTICE STANDARD 2: CYSTIC FIBROSIS RELATED DIABETES (CFRD)**

Together with the DNE, the CF Nurse will endeavour to give the individual with CFRD the necessary tools to successfully manage their diabetes, and therefore minimise complications.

**References:**

Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, 2006

Brennan, A et al. Airway glucose concentrations and effect on growth of respiratory pathogens in cystic fibrosis. *Journal of Cystic Fibrosis*. 2007; 6: 2: 101-109

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**NURSING CLINICAL PRACTICE STANDARD 2: CYSTIC FIBROSIS RELATED DIABETES (CFRD)**

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## NURSING CLINICAL PRACTICE STANDARD 3: DISTAL INTESTINAL OBSTRUCTION SYNDROME (DIOS)

**Definition:** (As per the ESPGHAN Cystic Fibrosis Working Group, 2010).

Constipation/ Faecal loading -Abdominal pain and/or distension or a decline in the frequency of bowel movements in the last few days to weeks or increased density of stools in the last few weeks or months, where the symptoms are relieved by the use of laxatives.

Partial DIOS - Incomplete or impending DIOS defined as a short history (days) of abdominal pain or distension or both and a palpable mass in ileo-caecal junction without signs of complete obstruction (i.e. presence of bowel sounds and passing of flatus).

Complete DIOS - Complete intestinal obstruction, as evidenced by vomiting of bilious material and/or fluid levels in small intestine on an abdominal radiography with a faecal mass in ileo-caecal junction and abdominal pain or distension or both.

Note: These definitions also make a distinction between the fairly acute onset of symptoms as seen in complete and partial DIOS, and the more gradual onset of symptoms as usually seen in constipation.

### Clinical Assessment

- Assessment should include, but is not limited to:
- Clinical history:
  - abdominal pain/distension, length of time of onset
  - frequency and consistency of bowel movements recently
  - vomiting/nausea
  - absence or presence of flatus
  - decrease in appetite
  - history of previous episodes of DIOS, constipation and/or abdominal surgery
- Abdominal examination - hard immobile mass is often palpable in right iliac fossa.
- Dietary intake – specifically pancreatic enzyme replacement therapy (PERT), fluid and salt intake.
- Sputum volume/tenacity and is the patient prone to sputum swallowing
- Medication review specifically recent use of PPIs, aperients and osmotic agents e.g. Movicol™, Lactulose™, Gastrograffin™, Colonlytely™.
- Psychosocial review.

**NURSING CLINICAL PRACTICE STANDARD 3: DISTAL INTESTINAL OBSTRUCTION SYNDROME (DIOS)**

**Investigations may include**

- Plain erect abdominal X-ray.  
DIOS typically shows faecal loading in the right iliac fossa (often granular or bubbly in appearance), dilatation of the ileum and an empty or gas-filled distal colon. Air-fluid levels and a variable degree of small bowel dilatation may be seen during acute episodes.
- Computed tomography (CT)
  - CT may help to clarify the diagnosis. Features seen include dilated small bowel and proximal colon with or without intestinal wall swelling. The intestines are filled with homogenous masses (faecolith) and there are usually varying degrees of fluid in the small bowel.
- Abdominal ultrasound (Abdo US)
  - Abdo US may be helpful in identifying the obstructing mass, but cannot be relied upon to exclude other serious causes of pain and obstruction such as intussusception
- Serum amylase and electrolytes should be checked to exclude pancreatitis.
- Contrast enema or colonoscopy should be performed if the condition is recurrent or unresponsive to medical treatment.

**Suggested Treatments:**

Medical management is first line treatment. However early and regular review by an experienced Upper GI Surgeon and Anaesthetist is recommended.

**NURSING CLINICAL PRACTICE STANDARD 3: DISTAL INTESTINAL OBSTRUCTION SYNDROME (DIOS)**

	Fluid/Salt	Aperients	Laxative	Intestinal lavage	Decompression	Analgesia	Anti emetic
Constipation	Oral e.g. Glucolyte™	Stool Softener, e.g. Lactulose™; Microlax enema, Pear Juice	Osmotic agent, e.g. Movicol™				
Partial DIOS	Oral/IV e.g. Glucolyte™	Stool Softener, e.g. Lactulose™; Microlax enema, Pear Juice	Osmotic agent, e.g. Movicol™, Hyper Osmotic e.g. Gastrograffin™	*Bowel prep, e.g. Golytely™, Colonlytely ™, Picolax™		Opioid	Yes
Complete DIOS  (Surgical Review required)	NBM - IV		Osmotic agent, eg Movicol™, Hyper Osmotic e.g. Gastrograffin™	*Bowel prep, e.g. Golytely™, Colonlytely ™, Picolax™	Via NG or Gastrostomy – free drainage	Opioid	Yes

\*Administer via gastrostomy, if in situ, or the patient may require nasogastric placement if unable to tolerate oral administration of therapy.

### Nursing Considerations

- Frequent and regular observations including pain and volume of nasogastric drainage, bowel sounds, bowel motions.
- Encourage gentle ambulation
- Facilitate regular airway clearance, coordinate treatments with physiotherapist.
- Review all prescribed medications with treating medical team.
- Facilitate review by specialist CF Dietitian e.g. contact nearest tertiary centre.
- Liaise with family/social supports.

### Maintenance / Prevention

An individualised education program for the patient is developed after dietetic and medical review. Treatment plan should include the following:

- **GI aperients** - consider an individualised regimen of laxative intake: Movicol™/Lactulose™/Gastrograffin™ to assist regular GI motility
- **Hydration** - adequate fluid and sodium intake including Glucolyte™ based on 30-40mls/kg/BW and 4000-6000mg sodium/day to prevent risk of dehydration

### NURSING CLINICAL PRACTICE STANDARD 3: DISTAL INTESTINAL OBSTRUCTION SYNDROME (DIOS)

- **Pancreatic Enzyme Replacement Therapy** - review adherence and dosing and provide education as appropriate to prevent over or under dosing of enzymes which may contribute to DIOS and/or ongoing constipation.
- **Pear juice** up to 2 glasses (500mLs) per day may assist GI hydration

#### Resources

Guideline for the diagnosis, treatment and prevention of Distal Intestinal Obstruction Syndrome (DIOS) Adult Cystic Fibrosis Centre, The Prince Charles Hospital – Brisbane

FACT SHEET - Distal Intestinal Obstruction Syndrome - Cystic Fibrosis Centre, Royal Children's Hospital - Brisbane

#### References

Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, 2006

Cystic Fibrosis Standards of Care, Australia. 2008

Guideline for the diagnosis, treatment and prevention of Distal Intestinal Obstruction Syndrome (DIOS) Adult Cystic Fibrosis Centre, TPCH – Brisbane, 2011

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## NURSING CLINICAL PRACTICE STANDARD 4: GASTROINTESTINAL COMPLICATIONS

### Gastro-oesophageal Reflux Disease (GORD)

GORD is defined as an abnormal increased frequency or duration of regurgitation of gastric contents into the oesophagus and may cause the following symptoms; “heartburn”, dyspepsia, acid taste in the mouth and cough (especially if worse at night), but can extend to dysphasia, anorexia, nausea and vomiting. Some patients report no symptoms, however limit their diet. Detailed patient history is important in determining diagnosis. Referral to a Gastroenterologist is required for full assessment.

Clinical investigations may include, but not restricted to:

- 24 hour oesophageal pH monitoring and manometry to assess oesophageal motility and sphincter integrity.
- Endoscopy (gastroscopy) to visualise the upper gastrointestinal (GI) tract for any abnormalities.
- Barium swallow and follow through to assess gastric emptying transit time i.e. bowel motility.

Management:

Full medical, nursing and allied health review to assess, treat and educate, as appropriate, on:

- Lifestyle modification, e.g. dietary, sleep position, airway clearance options.
- Pharmacological agents including proton pump inhibitors, gastric acid antagonists, GI motility agents.
- Surgical e.g. Fundoplication to reinforce the oesophageal sphincter and reduce regurgitation of stomach contents.

### Pancreatitis

Pancreatitis is inflammation of the pancreas, which is characterised by central abdominal pain radiating to the back, vomiting and raised serum amylase. This most commonly occurs in patients who are pancreatic sufficient (PS).

Clinical investigations may include, but not restricted to:

- Blood tests including an assessment of the level of serum amylase and inflammation.
- Abdominal ultrasound (to exclude other causes).

## NURSING CLINICAL PRACTICE STANDARD 4: GASTROINTESTINAL COMPLICATIONS

### Management:

- Treatment of acute episodes is similar to that of other aetiologies, which is based on symptom management:
  - Nil by mouth – with careful reintroduction of diet.
  - Electrolyte and fluid replacement and monitoring.
  - Analgesia
  - Note: Recurrent pancreatitis may respond to low dose Pancreatic Enzyme Replacement Therapy (PERT) in PS individuals.

### Cystic Fibrosis Related Liver Disease (CFLD)

CFLD usually presents as an early clinical manifestation i.e. in childhood or adolescence, in pancreatic enzyme insufficient patients with CF. Many people with CF liver disease may be asymptomatic but have elevated liver enzymes on blood testing. Cirrhosis with portal hypertension and oesophageal varices are late complications; however regular monitoring and prompt referral to a Gastroenterologist at the earliest signs of liver abnormalities may slow progression in patients thought to be at risk.

### Clinical investigations may include, but not restricted to:

- Clinical examination.
- Abdominal ultrasound.
- Blood test – full blood count, serum liver function tests, coagulation profile and vitamin levels.
- Endoscopy.

### Management may include:

- Pharmacological agents– e.g. Ursodeoxycholic acid (URSO).
- Fat soluble vitamin supplementation.
- Nutritional support.
- Oesophageal banding.

In patients with advanced liver disease, referral for liver transplantation may be considered in the absence of significant other organ compromise. The criteria and information required for consideration for transplant by designated State centres can be found on the Transplantation Society of Australia and New Zealand website.

## NURSING CLINICAL PRACTICE STANDARD 4: GASTROINTESTINAL COMPLICATIONS

### Bowel Cancer

Bowel cancer is a well recognised health problem in the general population. There is a national screening program for everyone over 50 years in Australia. The incidence of bowel cancer in adults with CF is higher than the general population (SIR 3.1 95%CI 2.7-4.2), however, consensus recommendations on how screening for bowel cancer in CF should be implemented, are yet to be formulated.

Studies are underway to shed light on the cause of this emerging problem and to guide clinicians in implementation of best practice screening and management.

#### Clinical assessment and management:

- Regular review by full CF medical, nursing and allied health team and a Gastroenterologist.
  - Optimisation of nutritional status with regular assessment of anaemia, bowel habit in conjunction with PERT, aperients use, salt supplementation and hydration.
  - Physicians will have a low threshold to refer for colonoscopy.
  - Education and planning to maximise the effectiveness and minimise the complications of the bowel preparation regime for colonoscopy, as per local protocol. May require CF Dietitian and Endocrinology input.
  - Ensure repeat screening occurs at the recommended interval or prior to this if there is any clinical suspicion.

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Maisonneuve P, Marshall BC, Knapp EA, Lowenfais AB. Cancer risk in cystic fibrosis: a 20 year nationwide study from the United States. L Nati Cancer Inst 2013 Jan 16;105(2):122-9.

## NURSING CLINICAL PRACTICE STANDARD 5: HAEMOPTYSIS

### Definition

Haemoptysis is defined as the expectoration of blood from the lungs as a result of pulmonary or bronchial vascular haemorrhage.

### Cause

The most common reason for blood vessels in the lungs to leak is the presence of inflammation and infection of the airways. The architecture of the lung parenchyma and the blood supplies are abnormal due to constant remodelling that occurs as a consequence of persistent infection. Sudden increases in the amount of blood flow and pressure through the vessels due to exertion or postural changes can also cause thin-walled vessels to leak. Vessels are usually very efficient at forming a clot and for this reason bleeding normally slows and then stops within a 5-15 minute period. The appearance of blood streaking through the sputum should reduce over the next 24 – 48 hours. The timeframe vary widely between individuals.

### Management

Medical management may consist of:

- Full medical, nursing and allied health team assessment and intervention.
- Regular/frequent observation of blood pressure, heart rate and oximetry for 24-48 hrs post episode
- Appropriate antibiotic therapy to treat any underlying infection.
- Initiation of anti-thrombolytic therapy e.g. tranexamic acid.
- Review of Vitamin K prescription
- Major haemoptysis or persistent small to moderate volume haemoptysis unresponsive to conservative management may require radiological imaging and occlusion of any abnormal or leaking vessels using bronchial artery embolisation (BAE).

**NURSING CLINICAL PRACTICE STANDARD 5: HAEMOPTYSIS**

	Airway Clearance/ Exercise	Aerosol Treatment	Clinical Review Advised	Investigations	Treatment
MINOR* Blood Streaked Sputum	<ul style="list-style-type: none"> <li>Reduce the force of coughing</li> <li>Cease high PEP</li> <li>Minimise head down tilt positions</li> </ul>		If persistent		<i>Consider oral antimicrobial treatments</i>
MODERATE <250mL in 24 hour period	<ul style="list-style-type: none"> <li>Maximise controlled airway clearance and gentle coughing/huffing</li> <li>Cease PEP, percussion, vibrations and head down tilt positions</li> <li>Adequate humidification</li> <li>Cease vigorous exercise</li> </ul>	Cease Mucolytic e.g. dornase alpha, hypertonic saline	Required	Coagulation profile CT –Pulmonary Angiogram	<u>Symptomatic Treatment</u> <ul style="list-style-type: none"> <li>Tranexamic Acid</li> <li>Review Vitamin K</li> </ul> <i>Consider Intravenous antimicrobial treatments</i>
MAJOR >250mL in 24 hour period	<ul style="list-style-type: none"> <li>Position into high side lying with bleeding side down</li> <li>Airway clearance as per “Moderate”</li> </ul>	Cease Mucolytic e.g. dornase alpha, hypertonic saline	<b>URGENT</b>	Coagulation profile; FBC and Group & X- match CT –Pulmonary Angiogram	<ul style="list-style-type: none"> <li>As for” Moderate”</li> <li>Bronchial Artery Embolisation may be indicated</li> <li>Establish venous access (e.g. large bore intravenous catheter)</li> </ul>

- **These criteria may vary between institutions and will be dependent on access to, and the professional preference/expertise of, the Interventional Radiologist/s concerned.**

## References

Physiotherapy for Cystic Fibrosis in Australia: A Consensus Statement, The Thoracic Society of Australia and New Zealand. 2008.

Cystic Fibrosis Standards of Care, Australia. 2008.

Moen C, Burrell A, Dunning J. Does tranexamic acid stop haemoptysis? *Interact Cardiovasc Thorac Surg.*, 2013 Dec; 17(6):991-4. Epub 2013 Aug 21.

Sidhu M, Weisler K, Burdick TR, Shaw DW. Bronchial artery embolisation for hemoptysis. *Semin Intervent Radiol.* 20-08 Sep; 25(3):310-18.

## NURSING CLINICAL PRACTICE STANDARD 6: ENTERAL NUTRITION

### PERCUTANEOUS ENDOSCOPIC GASTROSTOMY OR JEJUNOSTOMY

#### Definition

A percutaneous endoscopic gastrostomy (PEG) is a device that enters the stomach directly through the abdominal wall. In most instances it is a small device called a “button” that can be seen externally. Gastrostomy tubes are of benefit to people with CF that require long term nutritional supplementation via enteral feeding. In people who have significant gastro-oesophageal reflux disease and may not be able to undergo fundoplication prior to, or in conjunction with, the stoma formation, a longer tube may be placed. To minimise the risk of aspiration, the end of the tube may be passed through the pyloric sphincter into the small bowel and is therefore called a percutaneous endoscopic jejunostomy (PEJ) tube.

#### Assessment

Prior to surgical placement of either enteral feeding tube, patients will require assessment from several members of the team to ensure that it is a suitable option for them.

- **Nursing Assessment:** To ensure the patient is prepared psychologically, practically and has appropriate home support to enable insertion of the tube and provide assistance if necessary for ongoing maintenance of the same. To ensure the patient and family have the means to obtain prescribed formulas, pumps and consumables required to deliver feeds.
- **Gastroenterological assessment:** To assess, treat and monitor for gastro oesophageal reflux and any other potential GI risk factors. Regular review of the integrity of the gastrostomy/jejunostomy site and device.
- **Dietetic Assessment:** To assess and advise on the nutritional requirements of the patient and the volume of feed required to achieve optimal growth and nutrition and optimise the PERT required depending on the type of feed prescribed.
- **Diabetes education** may be required if the patient has glucose intolerance or CFRD and is intermittently or permanently insulin requiring.

#### Management

Patients should be educated in how to care for their enteral feeding device in line with local hospital policy.

General principles include:

- Cleaning of entry site daily or more frequently as required.
- Turning of the “button” each day to prevent the device adhering to the skin around the tract.
- Flushing the button at the completion of each use (feed /medication) or daily if not being used.

**NURSING CLINICAL PRACTICE STANDARD 6: ENTERAL NUTRITION**

**Nursing Considerations**

There are several types of enteral feeding devices used and each has their own merits and requires slightly different long term management. Refer to product information and local policy to ensure patients adequately educated in the use of the type of device they have.

Stomal Therapists and PEG Accredited Dietitians are valuable resource for patients and can provide education and assist with trouble shooting any problems and should be involved in patient education pre-insertion if appropriate.

**References**

Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, 2006.

Gastrostomy Management Procedure: The Children's Hospital at Westmead, 2009.

Gastroenterological Society of Australia (GESA): PEG Patient Information Sheet.  
[http://www.gesa.org.au/files/editor\\_upload/File/GESA%20PEG.pdf](http://www.gesa.org.au/files/editor_upload/File/GESA%20PEG.pdf)

## NURSING CLINICAL PRACTICE STANDARD 6: ENTERAL NUTRITION

### NASOGASTRIC TUBE

#### Definition

A nasogastric (NG) tube is a flexible tube passed through the nose and into the gastric pouch. In cystic fibrosis the primary use for NG tubes is short term nutritional support during respiratory exacerbations or as a trial prior to commencing gastrostomy feeding. NG tubes are not a good long term option for most people as they need to be replaced regularly, can become dislodged easily (especially with coughing) and in general they are less aesthetically suitable.

#### Assessment

Prior to placement of a NG tube patients will require assessment from several members of the team to ensure that it is a suitable option for them.

- **Nursing Assessment:** To ensure the patient is prepared psychologically and practically for insertion of the tube and the ongoing maintenance of the same.
- **Gastroenterological assessment:** To assess, prescribe treatment and monitor for gastro-oesophageal reflux or other potential GI risk factors.
- **Dietetic Assessment:** To measure the nutritional requirements of the patient and the volume of feed required to achieve optimal growth and nutrition and optimise Pancreatic Enzyme Replacement Therapy.

Note: If patients are to be discharged home with NG feeds the CF Nurse Specialist should ensure they have the means to obtain prescribed formulas, pumps and consumables required to deliver feeds.

#### Management

NG tubes should be managed in line with local hospital policy.

General Principles include:

- Ensuring feeding tube is checked for correct placement prior to every use
- Flushing the tube after each use.
- Ensuring pressure area care for the nares and face.
- Ensure the tube is taped into position appropriately.

## NURSING CLINICAL PRACTICE STANDARD 6: ENTERAL NUTRITION

### Nursing Considerations

Whilst NG tubes are primarily used during inpatient stays, if a patient is to be discharged home having enteral feeds via an NG tube, ensure the patient has adequate information about:

- Care and management of the tube.
- Obtaining equipment required to carry out this care as well as to deliver feeds appropriately.
- Contact details for relevant staff members in case of adverse effects.

Most local hospital will have policies and procedures incorporating the delivery of enteral feeds via NG tubes in both the home and hospital environment.

### References

Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, 2006

Enteral Feeding Tubes and the Administration of Enteral Nutrition Practice Guideline, 2009. The Children's Hospital at Westmead.

## NURSING CLINICAL PRACTICE STANDARD 6: ENTERAL NUTRITION

### PARENTERAL NUTRITION

#### Definition

Parenteral nutrition is provision of elemental feeding via an intravenous line. It is only used as a short term intervention in severely malnourished individuals who are unable to maintain or gain weight as a result of acute, severe illness, a non-functional gut, anorexia associated with overwhelming sepsis and in whom suitable intravenous access is possible. The risks include line sepsis and /or metabolic imbalances.

#### Assessment

Prior to the initiation of parenteral feeding, patients will require assessment from several members of the team to ensure that the benefits outweigh the risks.

- Medical assessment: To diagnose and treat the underlying cause of malnutrition and determine the likely duration of enteral nutritional supplementation.
- Nursing Assessment: To ensure the patient has suitable intravenous access i.e. a separate intravenous line to that being used for other iv medications that is clearly marked as an enteral nutrition line only. To ensure the patient understands the reasons for this type of intervention and the possible risks associated with this therapy including the requirement for regular monitoring of electrolytes and blood sugar levels and possibly arterial blood gases.
- Dietetic Assessment: To assess the nutritional requirements of the patient, advise on the type of elemental preparation to be infused and the regimen that best meets the needs of that individual.

#### Management

The initiation, frequency of monitoring and duration of therapy should be managed in line with local hospital policy including:

Regular monitoring of weight, fluid intake, electrolytes and blood sugar levels.

Regular nutritional assessment of adequacy of oral intake and ongoing need for enteral feeding.

**NURSING CLINICAL PRACTICE STANDARD 6: ENTERAL NUTRITION**

**Nursing Considerations**

Administration of the enteral feeds as prescribed.

Monitoring and regular dressing of the iv insertion site to prevent infection.

Assessment of blood sugar levels.

Monitoring for any signs of adverse effects.

**References**

Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, 2006. Section 5.8.

## NURSING CLINICAL PRACTICE STANDARD 7: NEBULISED MEDICATION ADMINISTRATION

### Definition

Nebulisers provide a means of administering aerosolized drugs to the airways for local effect or systemic absorption. To effectively achieve this, the nebuliser must produce very small particles (<5 micron) which are breathed deeply into the lungs. This procedure describes the appropriate equipment & process to achieve effective administration of the listed medications by this route.

### Considerations for optimal medication delivery

- Nebuliser selection and use
  - Conventional e.g. sidestream
  - High efficiency e.g. Pari LC plus™, Pari Star™, Pari Sprint™, AeroNeb™
- Patient interface (must match nebuliser type)
  - mouthpiece
  - mask
- Gas flow and type
- Storage and care of all equipment

### Nebuliser Selection and Use

Medication Type	Nebuliser Type	Patient Interface	Filter Needed
Bronchodilator	Conventional and High efficiency	Mouthpiece*	No
Mucolytic, e.g. dornase alpha, hypertonic saline †	High efficiency	Mouthpiece Mask <4yrs of age	No
Antimicrobials†	High efficiency	Mouthpiece Mask <4yrs of age	Yes

\* Mask may be used as per patient assessment (except Atrovent® if patient has glaucoma) or if otherwise clinically indicated

† Test dose with pre & post spirometry is usually given to check for bronchospasm (as per local facility clinical guidelines)

#### Note:

- Filters are used to prevent environmental deposition of nebulised antibiotics in hospital and the risk of developing antibiotic resistant organisms.
- Filter Pad to be replaced after use

## NURSING CLINICAL PRACTICE STANDARD 7: NEBULISED MEDICATION ADMINISTRATION

- If the patient is to nebulise antibiotics at home they do not require a nebuliser with filter. Consideration, however, should be given to the home environment when making this decision. The usual nebuliser is used and they are instructed to use it in a well ventilated area, e.g. outside, near an open window or vent the nebuliser outside with a 75-100 cm length of tubing.

### Patient Interface

Mouthpieces are the recommended interface for nebuliser therapy as they:

- promote mouth breathing & reduce upper airway deposition
- provide better lung deposition than masks
- reduce facial/eye irritation
- increase user comfort (less claustrophobic)

Note: Mouthpiece must be used for antimicrobials, steroids, dornase alpha and ipratropium bromide.

Mask interface is indicated for children under five years, and for patients who are confused, unconscious or have impaired coordination.

For optimal effect, masks must:

- have an expiratory valve
- be firmly fitted
- be used with mouth breathing

### Gas Flow and Type

- Nebulisation **by air** to be used for patients **requiring  $\leq 4\text{L/min}$** , especially for known or suspected CO<sub>2</sub> retainers (chronic hypercapnia)
- O<sub>2</sub> via Nasal Prongs should be continued during nebulisation on air
- Nebulisation **by O<sub>2</sub>** may be used for those **requiring  $\geq 5\text{L/min}$**

## Storage and Care of Equipment

As recommended by the manufacturer and local facility Infection Prevention Unit and CF Physiotherapy Guidelines.

## References

Infection control guidelines for CF patients and carers (2008)

Metro North Health Service District – The Prince Charles Hospital Primary & Community Health Services – Nebuliser Administration Procedure (2010)

Practice Guideline: Administering nebulised antibiotics in Hospital, Children's Hospital Westmead (2013).

The Alfred: Care of Airway Clearance Equipment for Inpatients. 2010.

## NURSING CLINICAL PRACTICE STANDARD 8: NON INVASIVE VENTILATION (NIV)

### Definition

Non Invasive ventilation (NIV) is the application of positive pressure via the upper respiratory tract using a nasal or facemask for the purpose of augmenting alveolar ventilation.

NIV has an expanding role in the management of:

- Acute respiratory failure
- Chronic respiratory failure
- Bridge to transplant
- Airway clearance and exercise

“The physiological rationale for use of NIV is that the unloading of respiratory muscles with positive pressure results in improved respiratory muscle performance, increased alveolar ventilation and improvement in gas exchange” (Physiotherapy for Cystic Fibrosis in Australia: A Consensus Statement, The Thoracic Society of Australia and New Zealand)

### Bi-Level Positive Airway Pressure (BIPAP)

BIPAP alternates between an inspired positive airway pressure (IPAP) and expired positive airway pressure (EPAP). It provides a higher level of positive pressure during inspiration than expiration.

- IPAP assists with PaCO<sub>2</sub> clearance
- EPAP and oxygen assist with the PaO<sub>2</sub>

Nurses and physiotherapists have a joint role in establishing and maintaining patients on NIV therapies.

### Patient Interface

- Full Face Mask
- Nasal Mask
- Mouthpiece

## NURSING CLINICAL PRACTICE STANDARD 8: NON INVASIVE VENTILATION (NIV)

### Nursing Care Considerations

Monitoring – the monitoring required will vary depending on the underlying indication for use (e.g. acute respiratory failure or augmentation of airway clearance). Monitoring heart rate, respiratory rate and effort is essential in assessing the response to treatment. When monitoring the patient, patient acuity should be considered, the level of monitoring required and the nursing staff level required to safely care for the patient.

Hygiene – the patient should have regular periods with mask off to protect skin integrity and to facilitate oral/eye care.

#### Interface (full face/nasal/mouthpiece)

- Check appropriate fit
  - Placement
  - Pressure area care to high risk areas, e.g. nasal bridge, ears, back of head.
- Clean mask cushion daily

#### Equipment maintenance

- Circuit:
  - Check temperature
  - Ensure no leaks
  - Change as per local policy (Refer to local tertiary centre)
- Humidification:
  - All NIV should be delivered with humidified circuit
  - Ensure humidification is maintained at all times
  - Change as per local policy (Refer to local tertiary centre)

### References

Physiotherapy for Cystic Fibrosis in Australia: A Consensus Statement, The Thoracic Society of Australia and New Zealand

Cystic Fibrosis Standards of Care, Australia. 2008

Guideline: Non Invasive Ventilation (NIV) Management Bayside Health (2011)

## NURSING CLINICAL PRACTICE STANDARD 9: OXYGEN THERAPY

### Definition

Oxygen therapy is indicated in CF for the management of acute and chronic respiratory failure and may also be required intermittently during exercise or in hypoxic environments (e.g. High altitude or air travel).

### Considerations for the safe administration of prescribed supplemental oxygen therapy

- When is oxygen therapy indicated and at what dose?
- What flow is required to maintain the predetermined PaO<sub>2</sub> and/or saturation? (Note: A saturation of 90% is required to prevent tissue hypoxia)
- How oxygen is best delivered?
- Is humidification required?
- What are the dangers of oxygen therapy for this patient?
- What assessment and monitoring is required?
- When should it be stopped?

### Assessment

#### Acute

At times of acute exacerbation patients may have an increase oxygen requirement. It is important that this is monitored. Nursing observations may include, but not restricted to:

- Heart rate.
- Respiration rate and effort (BORG's Scale).
- Oxygen Saturation – at rest and post exertion.
- Skin colour – peripheral or central cyanosis.
- Use of accessory respiratory muscles.
- Temperature.

Note:

- Medical staff prescribe oxygen therapy and advise the required flow rate and delivery device or the SaO<sub>2</sub> range to aim for if flow is to be titrated.
- Regular review of oxygen prescription is essential.

## NURSING CLINICAL PRACTICE STANDARD 9: OXYGEN THERAPY

Clinical measurements may include, but not restricted to:

- Overnight oximetry
- Sleep study
- Arterial blood gas

### Chronic

Indications for chronic therapy at home are based on hypoxia at rest. Clinical measurements requirements may include, but not restricted to:

- Overnight oximetry
- Sleep study
- Arterial blood gas test
- Six minute exercise test

Refer to local health area guidelines for the criteria to qualify for home O<sub>2</sub> therapy.

### Intermittent

- Exercise - Oxygen saturation is monitored as the patient ambulates with and without oxygen. Saturations below 88% whilst ambulating on room air may indicate that patient would benefit from supplemental oxygen with exercise and would warrant formal assessment.
- High Altitude - High Altitude Simulation Testing (HAST) is performed on patients who wish to travel by aircraft or who intend to be staying at high altitude such as skiing holidays. The test involves simulation of oxygen levels that will be experienced during flight. (FiO<sub>2</sub> = 15% during flight). If SpO<sub>2</sub> drops below 88% during simulation of altitude then additional oxygen is given via nasal prongs to maintain SpO<sub>2</sub> > 88%.

## Delivery Systems

Refer to your local State/ Territory and Health Authority Oxygen Therapy Administration Guidelines

## References

Adult Domiciliary Oxygen Therapy Clinical Practice Guideline. The Thoracic Society of Australia and New Zealand. March 2014.

Physiotherapy for Cystic Fibrosis in Australia: A Consensus Statement. The Thoracic Society of Australia and New Zealand. 2010

Cystic Fibrosis Standards of Care, Australia. 2008

## NURSING CLINICAL PRACTICE STANDARD 10: PNEUMOTHORAX

### Definition

A pneumothorax is defined as the presence of air within the pleural space. In CF, it is generally termed a secondary pneumothorax as it occurs as a result of underlying lung disease. A pneumothorax may occur in a patient with CF as a result of rupture of sub-pleural blebs on the visceral pleura.

### Clinical Presentation

Symptoms vary, but may include the following:

- Sudden or gradual onset of chest pain – sometimes localised to shoulder tip, may be described as a “stitch”.
- Sudden or gradual onset of dyspnoea.

Assessment:

- Normal observations including site and severity of chest pain x/10.
- Auscultation to assess extent and side.
- Radiological imaging.

### Management

The aim of management is to create an environment that facilitates healing of the breach in the lung surface by removing the air from the space and encouraging the lung pleural interface to adhere and prevent the development of an empyema.

For a small pneumothorax in an asymptomatic patient, medical management usually involves observation and/or aspiration and low flow supplemental oxygen therapy. A large pneumothorax requires intercostal drainage with or without suction. Intravenous antimicrobial treatment should be commenced at the same time to prevent infection and resultant sputum retention, which may delay re-expansion of the collapsed lung. A recurrent pneumothorax may require surgical intervention i.e. a pleurodesis.

In the case of potential need for surgery, usually considered if the lung fails to come up on suction or goes down when suction is removed after 48+ hrs, early referral to an experienced cardiothoracic surgeon is advisable.

NURSING CLINICAL PRACTICE STANDARD 10: PNEUMOTHORAX

	Airway Clearance/ Exercise	Investigations	Treatment
Small pneumothorax	<ul style="list-style-type: none"> <li>• Maximise controlled airway clearance and gentle coughing/huffing</li> <li>• Cease PEP, percussion and vibrations, and head down tilt positions</li> <li>• Adequate humidification</li> <li>• Avoid upper limb resistance exercises</li> </ul>	CXR – PA + Lateral Chest CT	<ul style="list-style-type: none"> <li>• Consider intravenous antimicrobial treatments.</li> <li>• Low flow supplemental oxygen.</li> </ul>
Large Pneumothorax	<ul style="list-style-type: none"> <li>• If pneumothorax is <b>undrained</b>, cease treatment and liaise with medical team.</li> <li>• If pneumothorax is <b>drained</b>: <ul style="list-style-type: none"> <li>▪ Gentle Coughing</li> <li>▪ Review PEP and non-invasive ventilation</li> <li>▪ Avoid PEP 1-2 weeks post resolution of pneumothorax</li> </ul> </li> </ul>	CXR – PA + Lateral Chest CT	<ul style="list-style-type: none"> <li>• Intravenous antimicrobial treatments</li> <li>• Intercostal catheter placement</li> <li>• Ensure adequate analgesia and humidification</li> </ul>

**Note: Lung function testing, air travel and diving (or any other breath-holding activity) should be avoided for several weeks post resolution, as medically deemed appropriate.**

## References

Physiotherapy for Cystic Fibrosis in Australia: A Consensus Statement, The Thoracic Society of Australia and New Zealand.

Cystic Fibrosis Standards of Care, Australia. 2008. Revised 2013

## NURSING CLINICAL PRACTICE STANDARD 11: SALT REPLACEMENT THERAPY

### Definition

People with CF are at risk of increased sweat losses of sodium and chloride. Infants with CF are particularly at risk of salt depletion due to low levels of sodium in breast milk and substitutes. Sodium chloride deficiency is characterised by:

- Hyponatraemia
- Decreased serum osmolarity
- Decreased appetite
- Nausea and vomiting
- Muscle cramps
- Deposit of sodium chloride crystals on the epithelial surface
- Fatigue
- Poor concentration

Salt depletion has clinical implications by contributing to increase sputum viscosity which may lead to sputum retention and also to distal intestinal obstruction syndrome.

The recommended dose of salt replacement for people with CF is based on an individual's symptoms, dietary intake, climate conditions, and exercise/activity level.

General Recommended doses of sodium	
Infant	500mg/day
Child	Up to 4000mg/day
Adolescent/Adult	6000mg/day

### Considerations

- Salt requirements may increase:
  - During rapid growth
  - During periods of illness
  - Changes in dietary intake
  - With additional strenuous work or physical activity
  - With change to hot/humid climate
- Facilitate review by specialist CF Dietitian e.g. contact nearest tertiary centre, minimum annually or at time when changes in requirements occur.

## Resources

“Salt, just do it” Adult Cystic Fibrosis Centre, TPCH – Brisbane

FACT SHEET – Salt Replacement Therapy, Paediatric Cystic Fibrosis Centre, Royal Children’s Hospital – Brisbane

“Don’t dry out” – CF Service, The Alfred, Melbourne

## References

Australasian Clinical Practice Guidelines for Nutrition, 2006

Cystic Fibrosis Standards of Care, Australia. 2008

**GLOSSARY OF TERMS**

- CF - Cystic Fibrosis
- CFRD - Cystic Fibrosis Related Diabetes
- CFTR - Cystic Fibrosis Transmembrane Regulator
- CO<sub>2</sub> - Carbon Dioxide
- CT - Computerised Tomography
- DIOS - Distal Intestinal Obstruction Syndrome
- DNE - Diabetes Nurse Educator
- ENDO - Endocrine
- FEV1 - Forced Expiratory Volume in 1 second
- GI - Gastrointestinal
- HbA1c - Haemoglobin A1c
- HEHP - High Energy High Protein
- L/min - Litres per minute
- O<sub>2</sub> - Oxygen
- TIVAD - Totally Implanted Venous Access Device