CYSTIC FIBROSIS: CAPAC PATIENT MANAGEMENT

PRACTICE GUIDELINE©

DOCUMENT SUMMARY/KEY POINTS

- Children with Cystic Fibrosis who require intravenous (IV) antibiotic therapy and increased physiotherapy for an exacerbation in their lung disease can safely and effectively be treated at home in a Community Acute/Post-Acute Care service (CAPAC) model of care.
- The following Practice Guideline identifies:
  - Patient Flow to the CAPAC service
  - CAPAC Treatment Plan
  - How to complete an accurate assessment

Patients excluded from these Guidelines: Refer to the **CAPAC admission criteria**: http://chw.schn.health.nsw.gov.au/ou/kols/resources/forms/CAPAC_criteria.pdf

CHANGE SUMMARY

- Due for mandatory review – no changes made.

READ ACKNOWLEDGEMENT

- CAPAC staff & Respiratory team should read and acknowledge they understand the contents of this document.

This document reflects what is currently regarded as safe practice. However, as in any clinical situation, there may be factors which cannot be covered by a single set of guidelines. This document does not replace the need for the application of clinical judgement to each individual presentation.

Approved by:	SCHN Policy, Procedure and Guideline Committee	CAPAC Management
Date Effective:	1st February 2014	**Review Period**: 3 years
Team Leader:	Clinical Nurse Specialist	**Area/Dept**: CAPAC

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1 Preamble

Community Acute/Post-Acute Care service (CAPAC) provides acute hospital substitution and/or hospital avoidance services for children who present to SCHN Westmead and Randwick.

Children with Cystic Fibrosis who require intravenous (IV) antibiotic therapy and increased physiotherapy for an exacerbation in their lung disease can safely and effectively be treated at home within the CAPAC model of care.

2 CAPAC Admission Criteria

Acceptance of referrals depends on

- Geographical location (within one hour drive of either Randwick or Westmead site)
- Patient is clinically/medically stable as determined by the treating team
- AMO consent for transfer
- Consent from parents/carers for CAPAC
- Carer presence during home visits
- Agreement with CF team that patient care needs can be met with home management
- Phone access
- Mutual recognition of identified goals of care
- Medicare eligibility
- Family have access to transport
- No issues identified that can compromise staff safety during home visits

3 Patient Flow

3.1 Booked admission

- Patients are referred to CAPAC by either the CF Clinicians or Clinical Nurse Consultants.
- The patient presents to the hospital for insertion of a Central Venous Access Device (CVAD), or for Implantable Vascular Access Device (IVAD, otherwise known as ports) to be accessed, Pre-treatment Pulmonary Function Test and any other required tests as determined by the treating team.
- Depending on the child's clinical status they may remain in hospital for 1 day to 7 days of inpatient treatment before being transferred to CAPAC for continuation of treatment.
- Usually a CF tune up lasts 14 days

3.2 Inpatient referral

- Patients who have been admitted to an inpatient ward and meet the CAPAC criteria are eligible for transfer to CAPAC if clinically stable.
- Patient referred to CAPAC using Power Chart referral form at Westmead site and Paper referral at Randwick site.
- Referrals can only be accepted between 0730 - 1600hrs after discussion with the NUM on Westmead page 7115 or mobile 0409830213.
- Patient has medication ordered on hospital drug chart MR73 after the ID team have approved treatment.
- Antibiotics that require 23hr infusions need to be ordered from Baxter via pharmacy. This requires 48hrs notice to guarantee delivery.
- Patient has antibiotic assays attended and appropriate antibiotic dose prescribed.
- CAPAC team will liaise with the family regarding first visit time, complete Home Visiting risk assessment and consent for treatment.
- Patient transferred from inpatient ward to CAPAC ward on inpatient Patient Management system.

4 Treatment

4.1 Aim of treatment

The aim of antibiotic therapy for an acute exacerbation in a Cystic Fibrosis patient is to decrease the bacteria and inflammation present in the airways.

Length of treatment is determined by improved Pulmonary Function Test's (PFT's) and a return to the patient's baseline pulmonary status.

4.2 CAPAC Treatment Plan

Admit patient to the hospital for insertion of CVAD, or IVAD access, order medications and obtain ID approval. Organise physiotherapy, dietician, social worker, medical and CNC review. When the decision is made to transfer to CAPAC, order required medication from Baxter via pharmacy and complete a CAPAC referral.

- Day 1 in CAPAC transfer completed.
- Daily nursing visit for a total of 14 days or until treatment completed.
- Nursing care includes:
  - Daily assessment and observations, including temperature, pulse, respiration and oxygen saturation.
  - Weigh twice each week on CAPAC scales.
  - Daily visits for administration of Intravenous Antibiotics

- Care of CVAD or IVAD
- If BD or TDS Antibiotics are prescribed, arrange for parent education

- Daily or twice daily physiotherapist visit when available.
- Medical review and lung function test on day 7 and 14 of treatment.

4.3 Physiotherapy

Physiotherapy plays a key part in the overall management of Cystic Fibrosis and assists in the removal of secretions. Airway clearance therapy, exercise and inhalation therapy are the cornerstones of treatment and are associated with improved long term outcomes. Physiotherapists are also involved in musculoskeletal management, care of complex patients, infection control, education of patients and their families, promotion of self-management, transition to adult care, pre- and post- transplant care and end-of-life care.

Airway clearance techniques vary between patients. The most effective and appropriate method is determined by the treating therapist with guidance from the CF physiotherapist and in conjunction with the patient. In some cases (e.g. post sinus surgery, pneumothorax, haemoptysis) guidance from the medical team is sought in regards to the commencement and cessation of therapies. Airway clearance techniques include but are not limited to:

- Active cycle of breathing techniques (ACBT)
- Positive expiratory pressure therapy (PEP)
- PEP performed in combination with nebulised therapy (mask or mouthpiece)
- Oscillating PEP
- Autogenic drainage
- Modified postural drainage
- Percussion and Vibrations

Physiotherapy and Inhalation Therapy

Physiotherapy is often timed with nebuliser therapy to optimise the effectiveness of nebulised agents. Nebulisers are prescribed by the medical team before transfer to CAPAC. These commonly include:

- Hypertonic saline is used before or in combination with airway clearance to loosen secretions.
- Bronchodilators are usually taken before the administration of hypertonic saline to reduce associated bronchospasm. This is especially critical in patients who are identified as having hyper-reactive airways as determined by a challenge test in the pulmonary function laboratory.
- Dornase alpha (pulmozyme©) is a mucolytic agent used before or after airway clearance to decrease the viscosity of sputum. A minimum of 30 minutes should be allowed between nebulisation and the commencement of airway clearance.
- Nebulised antibiotics (although normally wouldn't be used during a CAPAC admission as the child would be receiving antibiotics intravenously) is done after airway clearance.

Physiotherapy Assessment

The CAPAC physiotherapist will carry out assessment and reassessment of the patient's respiratory system and exercise tolerance as indicated throughout the treatment period.

Physiotherapy Treatment

Typically, airway clearance therapy is performed twice daily during respiratory exacerbations and exercise is also utilised daily as part of management. As the physiotherapy service is currently only available in the morning, the CAPAC physiotherapist will endeavour to perform one session of airway clearance therapy and/or exercise session if time allows.

The family will be responsible for performing additional airway clearance therapy and exercise sessions with appropriate education from the CAPAC physiotherapist.

Communication and continuity of care

Where possible, the CAPAC physiotherapist will endeavour to treat potential CAPAC patients whilst in hospital, or receive handover from the treating ward physiotherapist. There is regular communication between the CAPAC physiotherapist and the CF physiotherapist to provide updates to the CF team on patient progress. Urgent matters or acute changes to the patient's condition will be directly communicated to the CF medical team.

Useful Resources

SCHN **Randwick CAPAC staff**: see hospital specific guidelines in the CF manual. (hard copy)

SCHN **Westmead CAPAC staff**: see Appendix 1.

All SCHN CAPAC staff:

- **Physiotherapy for Cystic Fibrosis in Australia: A Consensus statement:** http://www.thoracic.org.au/documents/papers/physiotherapyforcf.pdf

5 Assessment

5.1 Respiratory

Cystic Fibrosis is a complex disease primarily affecting the lungs as well as other organs leading to gastrointestinal, nutritional and pulmonary complications. It is characterised by the production of thick secretions leading to an alteration in lung environment causing recurrent lung infections, bronchial damage, bronchiectasis and eventual respiratory failure. Treatment is directed at identifying and attempting to eradicate or control bacterial infection in the airway with regular planned courses of intravenous antibiotics ("tune-up") to maintain or improve the patient's clinical condition.

Nursing treatment required for patients whilst in CAPAC:

- Daily visits for administration of Intravenous Antibiotics
- Care of CVAD or IVAD as per hospital policy

- If BD or TDS Antibiotics are prescribed, arrange for parent education
- Daily observations including temperature, pulse, respiration and O_2 saturation
- Daily Respiratory Assessment observing and recording respiratory rate and pattern, O_2 saturation, use of accessory muscles, any complaints of dyspnoea, haemoptysis, chest pain or wheezing, increased cough, increased secretions, use of supplemental O_2, exercise tolerance, current nebuliser therapy and compliance with physiotherapy.

5.2 Nutritional

Most patients with Cystic Fibrosis are pancreatic insufficient and require replacement enzyme supplementation with meals. All patients require added energy foods to promote optimal growth and normal nutritional status.

Nursing treatment which may be required for CF patients whilst in CAPAC:

- Record weight twice weekly on CAPAC scales.
- Discuss and record oral intake.
- Trial of nutritional supplements as prescribed by the dietician
- Support and guidance with nasogastric or gastrostomy feeds and use of feeding pumps if required.
- Assess appropriate use of pancreatic enzyme replacement therapy.
- Promotion of self-care and independence

Useful resources:

- Australasian Clinical Practice Guidelines for Nutrition in CF 2006

5.3 Gastrointestinal

Cystic Fibrosis affects the pancreas and liver in a high proportion of patients. Patients can suffer from Gastro -oesophageal reflux, distal intestinal obstruction syndrome (DIOS), gallstones, chronic liver disease, portal hypertension, and colonic strictures.

- Document past history of oesophageal bleeds/ haemoptysis
- Document and report to CF team any gastrointestinal symptoms such as abdominal pain, change in bowel habits or loss of appetite.

5.4 Endocrinology

Cystic Fibrosis related Diabetes (CFRD) is a common complication in people who have CF. The person does not produce enough Insulin due to scarring of the pancreas. Insulin resistance, or the body not using Insulin normally, is another reason people develop CFRD. Prevalence increases with age.

Symptoms include increased thirst and urination due to increased blood sugar levels, extreme tiredness, weight loss and unexplained decreased lung function.

Nursing treatment which may be required for patients whilst in CAPAC include:

- Record weight twice weekly
- Monitor BSL and liaise with CNC, Endocrinology or CF team
- Observe Insulin administration
- Encourage food diary
- Trouble shoot family concerns

6 Parent Education

Education begins at admission and continues throughout duration of treatment. Negotiation of care is necessary to provide family centred care. Negotiation of physiotherapy is required and physiotherapist to instruct parent on physiotherapy techniques.

Useful resources

- **CVC Parent/Carer Handout**
- **Central Venous Catheter Home Care – Parent Training & Information**(to be given to parents caring for a CVC at home in conjunction with appropriate face-to-face education and training):
- **Heparin Locking a Central Venous Catheter at Home – Parent Training & Information** (to be given to parents heparin locking a CVC at home in conjunction with appropriate face-to-face education and training):
- **IVAD Parent/Carer Handout** (to be given to parents of children receiving an IVAD for the first time):
- **Implantable Vascular Device (IVAD) Home Care – Parent Training & Information** (to be given to parents caring for an IVAD at home in conjunction with appropriate face-to-face education and training):

7 Discharge

- Patient attends hospital for medical review and end of treatment lung function
- Discharge exercise tolerance test is performed and recorded by a physiotherapist.
- Remove CVAD or hep-lock IVAD as per hospital policy
- Complete documentation
- Record weight on discharge
- Discharge from computer

8 References

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3. Huot L, Durieu I, Bourdy S et al. Evolution of costs of care for Cystic Fibrosis patients after clinical guidelines implementation in a French network. Journal of Cystic Fibrosis 2008.
4. Proesmans, Heyns, Moons, Havermans, De Boeck. Real life evaluation of intravenous antibiotic treatment in a Cystic Fibrosis Centre: Outcome of home therapy is not inferior. Respiratory medicine 2009.
5. Termoz,Touzet,Bourd, Decullier et al. Effectiveness of home treatment for patients with Cystic Fibrosis: the intravenous administration of antibiotics to treat respiratory infection. Paediatric Pulmonology 2008.
6. Cystic Fibrosis Trust 2001. Standards of clinical care of children and adults with Cystic Fibrosis in the UK.
7. Michel S.H., Mueller D.H. Are eating disorders present in Cystic Fibrosis. The CF Supplement, Volume 8, Issue 1, Feb 2010.
8. Dietitians Association of Australia: National Cystic Fibrosis Interest group (2006). Guidelines for Nutrition in Cystic Fibrosis.(p7-83)

Appendix 1: SCHN Randwick and Westmead useful resources

SCHN Westmead

- CVAD Practice Guidelines:

  http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2013-9037.pdf
- CVC Home Care:
  http://chw.schn.health.nsw.gov.au/o/documents/policies/homecare/2006-8125.pdf
- IVAD Home Care:
  http://chw.schn.health.nsw.gov.au/o/documents/policies/homecare/2013-8001.pdf
- Home Intravenous Medication: Parent/Carer Administration:
  http://chw.schn.health.nsw.gov.au/o/documents/policies/policies/2006-8301.pdf
- Medication Management CHW:
  http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2006-8232.pdf
- Home IVAB Administration for CAPAC:
  http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2010-0005.pdf
- CF Manual - CHW: (currently under review)

SCHN Randwick

- CF Management Guidelines: SCH
  http://sch.sesahs.nsw.gov.au/policy/manuals/infection/cystic%20fibrosis,%20guidelines%20for%20management.pdf