CYSTIC FIBROSIS: GUIDELINES FOR MANAGEMENT - SCH

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Children with cystic fibrosis (CF) are at risk of respiratory tract colonisation and infection because of difficult to clear thick airway secretions.
- Acquisition of some organisms, such as *B. cepacia* and *P. aeruginosa* are known to cause respiratory morbidity and accelerate respiratory decline.
- Some organisms, including *B. cepacia*, some strains of *P. aeruginosa* and methicillin resistant *Staphylococcus aureus* (MRSA), may be transferred from one patient with CF to another.
- Respiratory pathogens associated with CF are spread by the contact and droplet routes.
- Children with CF requiring admission must be placed in separate bays to other children with CF.
- CF patients are cohorted into groups. Physical contact between CF patients should be limited to children within the same cohorted group.
- Children with CF with signs and symptoms of chest infections should be separated from immunocompromised children and other at risk patients

CHANGE SUMMARY

- Document due for mandatory review; No change in practice.
- Replaces SCH document: i.2.C.2 ***Cystic Fibrosis: Guidelines for Management - SCH***

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	
Date Effective:	1st October 2015	**Review Period:** 3 years
Team Leader:	Clinical Nurse Consultant Cystic Fibrosis	**Area/Dept:** Respiratory SCH

READ ACKNOWLEDGEMENT

- Ward nursing staff, members of respiratory medicine team, outpatient nursing staff, Hospital in the Home [HiTH] staff and physiotherapists should read and acknowledge they understand the contents of this document.

TABLE OF CONTENTS

Minimising Transmission of Respiratory Pathogens in Children

Policy Statement

Children with cystic fibrosis (CF) are at risk of respiratory tract colonisation and infection because they have thick airway secretions that are difficult to clear. Acquisition of some organisms, such as *B. cepacia*, *M abcessus* and *P. aeruginosa* are known to cause respiratory morbidity and accelerate respiratory decline. Some organisms, including *B. cepacia*, some strains of *P. aeruginosa* and methicillin resistant *Staphylococcus aureus* (MRSA), may be transferred from one patient with CF to another. Respiratory pathogens associated with cystic fibrosis are spread by the contact and droplet routes.[1,2]

Purpose and Scope

The aim of this guideline is to minimise the risk of hospital acquired transmission of organisms causing chronic colonisation and infection, in children with cystic fibrosis attending the Sydney Children's Hospital.

General Principles

1. Hand hygiene is one of the most important practices for minimising the transmission of infective agents. Adherence to hand hygiene is mandatory.
2. Use a liquid soap or a waterless, alcoholic based antiseptic hand rub before and after touching a patient, before or after a procedure and after touching a patient's surrounding. ("The 5 Moments in Hand Hygiene"). [5]
3. Staff must observe "Standard" and "Contact" precautions when in close contact with patients who are coughing. Appropriate personal protective equipment (PPE) must be donned to protect the mucous membranes prior to procedures that may induce cough in patients.
4. Physical contact between patients with cystic fibrosis should be discouraged.
5. Children with cystic fibrosis with signs and symptoms of chest infections should be separated from immunocompromised children and other patients at risk of severe lower respiratory tract infections.
6. Children with cystic fibrosis and their carers must be educated and encouraged to observe good hygiene practices such as covering their mouth when coughing, coughing into disposable tissues and hand hygiene after properly disposing of tissues they have coughed into.
7. Attendance at the physiotherapy department and gym is limited to one child with cystic fibrosis at any given time. Any equipment used by the individual will be cleaned with a disposable disinfectant wipe after use. Staff and patients apply alcohol based hand rub when entering and leaving the gym.
8. Attendance in the Respiratory Laboratory is limited to one child with Cystic Fibrosis at any given time. Children must attend the respiratory lab in order according to their clinic-cohort, such that a child with *P. aeruginosa* attends after a child without. Staff and

patients apply alcohol hand rub when entering and leaving the laboratory. Single-use bacterial filters and nose clips are used for every patient.

9. Any reusable instrument or respiratory equipment that comes into contact with the mucous membranes of a patient with cystic fibrosis must be cleaned and disinfected or sterilised prior to its use. Respiratory equipment must be cleaned after use according to manufacturers' instructions or hospital or NSW Health protocols. Any equipment in the vicinity e.g. computer key boards, should be wiped down with a disinfectant wipe.
10. Attendance at the Starlight Room must be restricted to one child with CF at any given time. Bookings for this attendance can be made with the Starlight Room directly. Parents are educated to check with the Starlight Room staff that no other children with CF are present in the Starlight Room before entering. Individuals with CF or their parents are responsible for wiping down any equipment they use within the Starlight Room with a disposable cloth impregnated with disinfectant wipe.

Inpatients

1. Children with CF requiring admission must be placed in separate bays to other children with CF. Where possible a nurse should only care for one patient with CF. If this is not possible then cohorting CF patients needs to occur.
2. Where possible, each patient with CF should have a designated toilet and shower that is not shared by other patients with CF. If there are too many patients with CF on the one ward to allow for this, then patients should be encouraged to follow good personal hygiene standards, especially the washing of hands upon entering and leaving the bathroom.
3. Attendance by CF patients at the Hospital School must be limited to a maximum of one child in primary and one child in high school at any given time. When there are multiple inpatients, other students are to do school work in the ward under the supervision of their parent(s), ward teacher or work independently. Children will be nominated to attend the hospital school at the discretion of the school in consultation with the appropriate medical staff.
4. Equipment such as computer keyboards should be cleaned with a disinfectant wipe after use. Other equipment may be washed with neutral detergent.
5. School staff must observe hand hygiene practice prior to and after physical contact with patients, their immediate environment or secretions. [5]
6. Contact between patients by non-physical means should be encouraged to prevent social isolation e.g. the use of the telephones, livewire or emails between patients in the wards.
7. Where possible, each patient with CF should have a designated toilet and shower that is not shared by other patients with CF. If there are too many patients with CF on the one ward to allow for this, then patients should be encouraged to follow good personal hygiene standards, especially the washing of hands upon entering and leaving the bathroom.

C1SW

- CF patients admitted to C1SW for their routine annual bronchoscopy should be placed in separate bays.

Multi resistant staphylococcus aureus (MRSA)

- CF inpatients infected or colonised with MRSA, must be isolated in a single room with en-suite. See Multi Resistant Staphylococcus Aureus (MRSA): Management – SCH Procedure.
- Lung Function testing will be performed on the portable spirometer which is brought to the patient to be tested in their room. The spirometer is cleaned after each use, and is stored in the Respiratory Laboratory.

Multi-Resistant Pseudomonas aeruginosa

- CF inpatients infected or colonised with multiresistant *P. aeruginosa* or "clonal strains" of *P. aeruginosa* must be admitted to separate wards from other CF patients. Isolation from the otherwise healthy patient population is not required.

Mycobacterium Abcessus

- CF inpatients infected or colonised with *M. abcessus* must be admitted to separate wards from other CF patients. Isolation from the otherwise healthy patient population is not required.

Burkholderia cepacia

i. Wherever possible, children with *B. cepacia* must be isolated in a single room with en-suite, and not in the same ward as another CF inpatient.

ii. Patients with *B. cepacia* must not attend the Hospital School, Starlight Room, Respiratory Lab, the gym or hospital functions e.g. children's concerts or parties.

iii. Patients with *B. cepacia* attending clinics are to remain in the nominated room in outpatients. Lung Function testing will be performed on the portable spirometer (Easy One Spirometer) which is brought to the patient to be tested in their room. The spirometer is cleaned after each use, and is stored in the Respiratory Laboratory.

Extended Spectrum Beta Lactamase organisms (ESBL)

e.g. ESBL *Klebsiella spp*.

- CF inpatients infected or colonised with MRSA, must be isolated in a single room with en-suite. Lung Function testing will be performed on the portable spirometer (Easy One Spirometer) which is brought to the patient who is tested in his/her room. The spirometer is cleaned after each use, and is stored in the Respiratory Laboratory.

Outpatients

1. **Cohorting:**
   The outpatient cystic fibrosis clinics will be organised by cohorting children with similar organisms into the following groups: Red (*Pseudomonas* negative), Green (*Pseudomonas* positive), Yellow (*Stenotrophomonas*), and Purple (*B. cepacia*, multi-resistant *Pseudomonas*, ESBL, Atypical mycobacterium and MRSA).
2. Patient confidentiality must be maintained.
3. Sputum will be collected every 3 months where possible, or if a change in clinical state occurs. The treating physician and CF coordinator(s) will be responsible for checking results. All request forms must advise the laboratory that the patient has CF. If organisms other than the general respiratory pathogens and *P. aeruginosa*, *Staph. aureus* and *B. cepacia* are sought after, the request form must state this (e.g. *Stenotrophomonas*, *Achromobacter*, *Mycobacterium* or fungi).
4. Children who are too young to produce sputum should undergo annual bronchoscopy with bronchoalveolar lavage (BAL) taken from several lobes for microscopy, culture and sensitivity (MCS).
5. The “CF Cohort Groups” will be updated as sputum cultures become available. Parents will be notified of any changes for their child.
6. Children who attend the CF clinic are placed in a consultation room and remain in that room for the duration of their appointment. Staff needing to see that child enter and leave that room. After a child’s clinic visit is completed and prior to another child with CF entering that room, the room is cleaned by changing bed linen and wiping hard surfaces with a disinfectant wipe.
7. To minimise time spent in the waiting room, appointments at CF clinic are made in two timeslots (0915 and 1030) in order for children to be moved straight into an empty room without time spent waiting in common areas.
8. Children in the Purple group attend at 1100 in an attempt to avoid contact with other patients. Rooms used by children in the purple group are in a separate area within the outpatient department and are terminally cleaned following their appointment.
9. Outpatient appointments can only be made by contacting the outpatient department or CF clinic coordinators, who will use the “CF Outpatient Cohorted Roster”. The CF Clinical Coordinator can be contacted on ext. 2-1476. The CF CNC can be contacted on pager 44903.
10. Appointment dates and times are to be adhered to strictly. This means each patient in each designated cohort must observe their appointed day and time slot to minimise chances of contact with patients outside of their own cohort.
11. If children enter the clinic area on a day when another cohort of children with cystic fibrosis is being seen, they will be ushered immediately to a distant consultation room or to the emergency department.
12. All consultations should occur in consulting rooms not in the corridors, testing areas or waiting rooms.

13. Toys should be removed from outpatient rooms prior to seeing children with cystic fibrosis in the room. Children should be encouraged to bring their own toys to hospital.
14. Unscheduled appointments must be made through the clinic coordinators in order to be seen in the outpatient department. The CF Clinical Coordinator can be contacted on ext. 21476 and the CF CNC can be contacted on pager 44903
15. Urgent reviews should be arranged by contacting the Respiratory Unit by paging the Respiratory Fellow (pager 45415) or the CF CNC (pager 44903).

Emergency Department visits

- Cystic fibrosis patients seen in the Emergency Department shall ideally be segregated from other cystic fibrosis patients.

De-isolation

1. *P. aeruginosa* and *Stenotrophomonas*

A child with newly acquired *P. aeruginosa* can only be considered "negative" for the purpose of cohorting only after 3 sputum samples, one month apart in a 6 month period remain negative for *P. aeruginosa*, or after a BAL sample taken ≥ 6 months after the last *P. aeruginosa* positive sputum or BAL is negative for *P. aeruginosa*. Children chronically infected with *P. aeruginosa* will not be returned to the negative group.

2. MRSA

CF patients with known MRSA can be de-colonised and subsequently de-isolated if they satisfy the following criteria.

- They have 3 negative MRSA screen tests ≥ 6 months after the FIRST negative MRSA screen (taken under the conditions as outlined below).
- And only if they do not have in-dwelling devices (e.g. gastrostomy tubes)

a). Decolonisation regimen *(oral + topical concurrently)*

- Oral regimen of:
  Rifampin for 5 days.
  Dose: 10 mg/kg/day, once daily or in two divided doses
  (max 450mg less than 50 kg and 600mg greater than or equal to 50 kg)

Plus

Sodium fusidate* for 5 days.
Dosing: Only available as 250mg tablets -

- Infants – 1a mg/kg/dose, TDS
  (Dissolve ½ or whole tablet in water or juice and round up)
- Child greater than 5 years – 250mg, TDS.

* *Nb: Fusidic acid (paediatric syrup) – not available in Australia*

- Plus a concurrent topical regimen of:
  - Nasal mupirocin
    - Wash hands before use.

- Dispense a small amount (size of match head) of 2% nasal mupirocin (Bactroban) onto a clean cotton bud tip, apply to nostril and massage gently around the inside of nostril. Do no insert too deeply. Use a new cotton bud for each nostril
- Apply three times a day for 5 days.
- After applying the ointment, press finger against the nose next to the nostril opening and use a circular motion to spread the ointment within the nose.
- Wash hands after application.

AND

- Antiseptic body wash
  - Use a chlorhexidine-based wash (e.g. *Microshield*™[1][1] or *Microshield*™[2]). If the child has eczema, then Oilatum[2] Plus (for children greater than or equal to 6 months of age) should be used.

    Note: Bleach baths and may be an alternative for patients with eczema. Protocol available from ID service.
  - Apply the antiseptic body wash in the bath or shower daily for the same 5 days as nasal mupirocin.
  - Take care to wash hair, under the arms, inguinal region and in any skin folds.
  - Allow the antiseptic to remain on the skin for at least 5 minutes before washing off.

b). Screening samples for deisolation

- Samples: Nose + throat +/- any other relevant clinical samples – e.g. sputum
- First post-decolonisation screening sample should be taken **greater than or equal to 2 weeks** after the topical regimen for decolonisation and **greater than or equal to 3 months** after the oral decolonisation is complete
- Subsequent screening samples are to be taken on a separate occasions, at least 72 hours apart
- Samples must not be taken within a months of an MRSA active antibiotic e.g. vancomycin, rifampin and fusidic acid, ciprofloxacin and other quinolones, linezolid, with an additional list to include bactrim, clindamycin, lincomycin or erythromycin if the original isolate was a "non-multiply MRSA resistant" strain.

c). Deisolation

- Deisolation can occur **greater than or equal to 6 months after last negative** MRSA sample if there are 3 documented screening samples taken under the above conditions <u>after</u> the first negative MRSA sample.

d). Follow-up screen

- Screening for MRSA should continue however till a year after first negative screen (same conditions for interpreting the sample(s) apply).

[1] *Contains 2% or 4% chlorhexidine gluconate respectively. Not recommended for children < 2 months old*
[2] *Benzalkonium chloride 6% w/w, Triclosan 2% w/w, light liquid paraffin 52.5% w/w – adhere to 'Instructions for Use'*

Related SCH Infection Control Documents

- Personal Protective Equipment Guidelines - SCH
- Acute Respiratory Infections: Transmission and Prevention – SCH: http://chw.schn.health.nsw.gov.au/o/documents/policies/policies/2013-7050.pdf
- Multi Resistant Staphylococcus Aureus (MRSA): Management – SCH: http://chw.schn.health.nsw.gov.au/o/documents/policies/procedures/2015-7017.pdf

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