INFECTION CONTROL GUIDELINES
FOR CYSTIC FIBROSIS
PATIENTS AND CARERS
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CFA

It is known that chronic infection with certain micro-organisms in people with cystic fibrosis (CF) is associated with increased respiratory symptoms and poorer outcomes. All people with CF can harbour important micro-organisms in their respiratory secretions (such as bacteria and viruses), some of which can potentially be transmitted to others with CF. The aim of these guidelines is to prevent transmission between people with CF and also their carers by reducing the possibility of contact with infected respiratory secretions. The information about the acquisition and transmission of organisms in CF is still incomplete. Consequently, these guidelines should be considered as “best practice” and based upon current information. The guidelines may change as new information regarding infection in CF becomes available.

These guidelines have been developed for people with CF, their families and carers.

Epidemiology of CF Pathogens

Micro-organisms capable of establishing infection and causing disease are termed ‘pathogens’. The origins of most pathogens in CF remains unknown, but several potential sources are possible. These include the natural environment (e.g. soil, water), the health care or hospital environment (e.g. sinks and surfaces), contaminated equipment (e.g. nebuliser, pulmonary function equipment), other contaminated objects or surfaces, and other people with CF who harbour potentially transmissible pathogens. People without CF, including household and family members, are not generally responsible for transmission of respiratory bacterial pathogens to people with CF. However, they can transmit viruses and other non-bacterial pathogens to someone who has CF, which can be equally important.

The most common bacterial pathogens found in CF secretions are Pseudomonas aeruginosa, Staphylococcus aureus and, less commonly, Haemophilus influenzae. Of these, P. aeruginosa is the most important. Other important pathogens in CF that require special precautions are Burkholderia cepacia complex, multi-resistant S. aureus (MRSA) and respiratory viruses. Of these, the B. cepacia complex has been the most extensively studied in the CF community and this group of pathogens serve as a prototype for infection control in people with CF.

As those with CF survive longer and have increasing exposure to powerful antibiotics, other micro-organisms are being found in CF sputum. These have been termed “late emerging pathogens” and include Stenotrophomonas maltophilia, Achromobacter xylosoxidans, Ralstonia picketti, Pandoraea apista, Inquilinus limosus, Aspergillus species, and non tuberculous mycobacteria (NTM). These micro-organisms are environmental bacteria, found in water, soil, and on plants, including fruit and vegetables. Whether all have a primary pathogenic role in CF lung disease and are capable of transmission between people with CF remains unclear.
Methods of Transmission

The following routes of transmission are considered important in the spread of microorganisms between people with CF.

Direct contact - Person to person spread occurs with actual physical contact. This includes intimate contact such as kissing or casual contact such as touching hands that are contaminated with secretions e.g. a handshake after covering the mouth during coughing.

Indirect contact - Involves contact with an object, which has been contaminated by secretions from an infected patient (e.g. sharing eating utensils, respiratory equipment, such as nebulisers, and toys) and contact with surfaces soiled by respiratory secretions.

Droplet route - This involves transmission of pathogens through the air in large particle droplets and can occur if a patient coughs or sneezes towards, or onto, the eyes, nose or mouth of a non colonised patient in close proximity (i.e. less than one metre or an arms length away). The droplets do not remain suspended in air for long, because of their large size. There is some evidence that droplet spread may occur with P. aeruginosa. Both contact, and to a lesser extent, droplet spread are important in the transmission of respiratory viruses.

Airborne - Infection is transmitted by inhalation of pathogens on tiny droplet nuclei, which have been expelled from the respiratory tract. These minute particles can remains suspended in air for a long time and carried substantial distances. Examples are TB, varicella (chickenpox), measles and, possibly, influenza amongst people kept in confined spaces. While this route is not believed currently to be important for transmitting respiratory pathogens associated with CF, this is still a subject of ongoing research.

Whereas some micro-organisms are spread predominantly by contact (e.g. B. cepacia, MRSA and respiratory viruses) or by the droplet route (some respiratory viruses), it is likely most respiratory pathogens can be transmitted by a combination of pathways (i.e. both by contact and large droplets). Therefore attention to all areas of potential transmission is important, particularly for those pathogens, such as P. aeruginosa, where accurate information regarding transmission is incomplete.
Risk Factors Associated with Acquisition of B. Cepacia Complex

Although risk factors for transmission of respiratory pathogens between people with CF were developed by studying B. cepacia complex strains, they are relevant to all of those with CF, including their caregivers, who want to reduce the spread of any respiratory pathogen by the contact route.

Risk Factors

- Attendance at CF camps/attendance at an education program/participation in a group for adults with CF.

  Note: CF organisations in Australia no longer run any of the above activities. If people with CF choose to attend functions and events arranged by CF organisations they do so at their own risk.

- Close social contact
  - Kissing
  - Intimate contact
  - Prolonged car rides
  - Fitness class
  - Sharing eating and drinking utensils

- Sibling with B. cepacia complex

- Handshaking

- Inpatient exposures
  - Recent hospitalisation

- Sharing room or bathroom facilities with a B. cepacia complex infected patient
  - Sharing respiratory therapy equipment

Transmission of Specific Cystic Fibrosis Pathogens

B. cepacia complex

There is good epidemiologic evidence of person-to-person transfer in both health care and non-health care settings via contact and droplet routes with little current evidence of true airborne transmission. Transmission has been interrupted successfully by a combination of universal (standard) and “transmission based” precautions and these are applied to all patients with B. cepacia. Consequently, CF persons infected with B. cepacia are strictly segregated from each other and the general CF population.

MRSA (methicillin-resistant S. aureus)

This can be transmitted from person-to-person by contact transmission. Because of the potential risks to people with and without CF, strict contact isolation of those with MRSA as currently recommended by hospital infection control units is appropriate. People with MRSA should be managed with both standard and strict contact infection control measures as currently recommended by hospital infection control units.
**P. aeruginosa**

Available evidence suggests that in CF *P. aeruginosa* is acquired mainly from the environment and, other than amongst CF siblings, each person has their own unique strains. While people with CF in prolonged close contact situations (e.g. siblings, CF camp attendances) may share common strains, whether these are gained by cross-infection or from a common source exposure is still to be resolved. Most of the information regarding potential spread is derived from knowledge about *B. cepacia*, a much more highly transmissible micro-organism. However, frequent sharing of strains of *P. aeruginosa* that are indistinguishable from one another by modern molecular typing techniques (so-called ‘clonal’ strains) has been reported in some CF clinics. Whether this represents evidence of cross-infection in every instance is yet to be determined. Similarly, the clinical implications of acquiring these clonally-related strains is yet to be evaluated. Nevertheless, until more is learnt about these strains and their transmission pathways, it is reasonable for people with CF who are infected by *P. aeruginosa* to observe an awareness of general hygiene measures to reduce potential transmission between themselves and others with CF. Strict contact precautions are not usually required in this situation. However following recent reports of shared common *P. aeruginosa* strains within and between several Australian CF Clinics, it is recommended that, in order to avoid premature infection, those with *P. aeruginosa* should be separated from other people with CF who are still to acquire this pathogen. As an example, this might include having separate clinics and inpatient areas.

**Clonal P. aeruginosa strains**

Direct and indirect person-to-person spread of clonal *P. aeruginosa* strains within and between CF clinics has been demonstrated in Europe, the United Kingdom, Canada and Australia. Some of these “clonal” strains (identified by molecular finger printing typing techniques) may be more virulent than other *P. aeruginosa* strains and, as they are typically multi-resistant, they can also be more difficult to treat. Fortunately, not all clonal strains are highly transmissible and their common presence may simply reflect a high prevalence in the environment. Only a few centres have the ability to identify these micro-organisms by using sophisticated molecular typing techniques. Clinical suspicion should be raised, however, if a newly acquired strain of *P. aeruginosa* is mucoid and has a multi-antibiotic resistance pattern. These properties contrast with the usual non-mucoid appearance of these initial infecting strains and their sensitivity to most anti-pseudomonal antibiotics. Similarly, newly referred patients with *P. aeruginosa* who come from CF clinics known to have clonal strains should also be under suspicion and arrangements made for molecular typing of their strains. If clonal *P. aeruginosa* strains are identified, a combination of standard and contact based precautions, including segregation from other CF patients should be instituted. Some authorities recommend additional transmission based precautions for all people with CF harbouring *P. aeruginosa* isolates that demonstrate multi-resistance (i.e. resistance to all members of 2 or more classes of anti-pseudomonas antibiotics). However, there is little evidence to support this recommendation. Indeed, a general cohort isolation policy based upon antibiotic resistance makes little sense. If there is no epidemic strain, the evidence for cross-infection of *P. aeruginosa* is likely to be negligible. Conversely, if an epidemic strain is present, placing people with CF together promotes the cross-infection that such a strategy seeks to avoid. A cohort isolation policy must be strain-specific and based upon molecular typing, not antibiograms or other equally unreliable phenotypic methods.
Respiratory viruses (RSV, parainfluenza, influenza)

Respiratory viruses are highly contagious and are spread predominantly by contact with infected secretions and to a lesser extent by large particle droplets. Consequently, additional contact and droplet precautions are recommended for people with CF who have symptoms of a respiratory viral infection. This should include isolation in a separate room or attendance at a separate clinic or day centre.

“Late emerging” pathogens

As people with CF live longer and are exposed increasingly to more antibiotic courses, S. maltophilia, A. xylosoxidans, R. picketti, Pandoraea species, Aspergillus and NTM are being detected more frequently. These micro-organisms appear to be less virulent than other bacteria found in CF respiratory secretions and their pathogenic role in CF lung disease is unclear. Person-to-person spread has only occasionally been documented and is controversial. Routine standard and contact precautions are recommended. However, patients colonised with these micro-organisms should not be nursed alongside those with impaired immunity.

General Guidelines

The guiding principle is to decrease the spread of respiratory pathogens between people by reducing the opportunity for contact with their respiratory secretions either directly or indirectly. The risk of transfer through secretions increases with the time spent together and closeness of contact. Respiratory viruses (colds and flu) are highly contagious in these situations. The following precautions should be applied by all people at home, in hospital and in outpatient clinics irrespective of the microorganisms in their sputum.

General hygiene measures for children and adults with CF

- Wash your hands using soap and water or waterless antiseptic hand wash (e.g. alcohol based hand rub) frequently before eating, after coughing or handling sputum or tissues, and when using bathroom facilities. Waterless antiseptic is provided for use in hospitals and is also effective. It is very important to dry your hands thoroughly after washing with soap and water.
- Always cover your mouth and nose with your hand or tissue when you sneeze or cough. Throw away tissues immediately after use and wash your hands afterwards.
- Sit or stand a reasonable distance (one metre) from each other to prevent transmission from coughing or sneezing.
- Try not to shake hands with each other, a light tap on shoulder for example is better.
- Throw away tissues immediately after you use them.
- Do not leave sputum pots uncovered.
- Do not share eating or drinking utensils with each other.
- Do not share drink cans, cup or bottles.
- Do not share toothbrushes or towels.
• Do not share rooms with other people with CF if staying overnight.
• Avoid long car journeys with CF friends.
• Do not share respiratory equipment (nebuliser, inhalers, spacers etc).
• If possible infants and children with CF should avoid sharing toys.
• If you have symptoms of a viral respiratory infection (e.g. runny nose, cold or flu like illness) you should take particular care in following these general hygienic measures.

Specific pathogens
• You or your child should know your own micro-organisms and understand the precautions you need to take. Some pathogens (e.g. *B. cepacia*, MRSA, clonal *P. aeruginosa* strains) are more transmissible or infectious and can be difficult to treat. If you have these particular micro-organisms you will be asked to undertake special precautions in the hospital and at clinic, for example, coming early or late to clinic, staying in a single room, or attending a separate clinic or day centre, so as to reduce contact with other people with CF.
• Your doctors will check your respiratory secretions (e.g. sputum) regularly so that they are aware of the usual pathogens in your lungs when you come to clinic.

Hospital Outpatient
• Please wash your hands at the beginning and end of clinic, and after contact with respiratory secretions (e.g. before and after lung function testing, coughing into hands, sputum pots and tissues containing sputum). Waterless antiseptic hand wash (e.g. alcohol based hand rub) will be provided in all patient rooms, clinic waiting areas and the lung function laboratory/testing area.
• Avoid sharing toys in common waiting area. Families should bring their own toys for children.
• Use waste bins provided for tissues. These should be easily available.
• People with CF should maintain a distance of at least one metre from other people with CF in the clinic waiting area.
• Avoid hand shaking and general physical contact.

Specific Pathogens
People not yet infected with *P. aeruginosa* will be segregated from other people who harbour this bacterial pathogen and will be placed in a separate clinic, if available.

*B. cepacia* complex, MRSA and clonal *P. aeruginosa* strains
If you have these bacterial pathogens you will be segregated from other unaffected people with CF and in the case of *B. cepacia* from each other. Separate clinic rooms will be allocated, where you will be placed immediately and you will be visited by the CF team in your room. You may be asked to come
You will be asked to follow good personal hygiene measures whilst in hospital.

### Room sharing
- Whenever possible all people with CF will have single rooms with their own washing facilities.
- If this is not possible then you will be placed in a shared room with other people with CF who have the same pathogens as you do (except those with B. cepacia complex) or people who do not have CF.
- You should not sit on the bed or visit rooms of other people with CF on the ward, particularly if they are having physiotherapy or nebuliser treatment.
- If group activities are allowed you should sit at least one metre apart. Take care when coughing and avoid touching so that you do not pass on to others any respiratory pathogens that you may carry.

### Specific pathogens
- If you have B. cepacia complex you will be placed in a single room and treated with isolation precautions. There will be no mixing with other people with CF or school visits.
- If you have MRSA, a clonal P. aeruginosa strain or a virus causing a respiratory infection (eg. cold or the flu) you will be placed in a single room and treated with isolation precautions. No mixing with other people with CF or school visits is allowed. If a single room is unavailable you will share a room with another person with the same micro-organism as your own.

### Hospital school
- People with B. cepacia complex, MRSA, clonal strains of P. aeruginosa or a respiratory virus (eg influenza) should not attend school while they are in hospital, but have schooling in their own rooms.
- Other people with CF can attend school, but should sit at least one metre apart from others with CF.
- If you use a computer it should be wiped down after use.

### Colds and flu
People with viral infections, such as colds and flu like illnesses should ideally be seen away from the clinic, either in separate rooms distant from the main clinic area or in a CF day centre. If you have symptoms to suggest a cold or flu please inform the CF clinic in advance.
Outside rooms or off the ward

- Observe good general hygiene measures, sit in well ventilated areas at least one metre apart from others with CF. In certain circumstances you may be asked to wear a mask, for example if you have symptoms of the flu.

Equipment

- Do not share your nebulisers or physio equipment

Eating and drinking

- Observe the general hygiene rules for sharing eating and drinking utensils.

In the community

- Be aware of infection control hygiene measures.
- People with CF and their carers should be aware of their infectious status.
- Spas should be avoided.
- In the workplace people should be aware of their infection status and discuss infection risks.
- Siblings at home should have separate bedrooms.
- No communal CF camps or holidays.

Schooling

- People with CF may attend the same school.
- It is preferable for people with CF to attend different classes.
- Non-classroom contact should be minimised e.g. indoor gym session.
- Personal hygiene measure should be followed at all times.
- Involvement in school activities with non-CF people is encouraged.