Respiratory Microbes
Common in People with Cystic Fibrosis

People with cystic fibrosis are vulnerable to microbes that can cause serious infections. These infections are often difficult to treat with antibiotics so it is important to keep these microbes from spreading. This information sheet outlines some of the most significant respiratory microbes in people with CF.

Staphylococcus aureus

What is it?

Staphylococcus aureus are bacteria that are often reffered to as "Staph". Meticillin resistant *Staphylococcus aureus* (MRSA) are *Staphylococcus aureus* bacteria that are resistant to meticillin and frequently other antibiotics.

Staphylococcus aureus are bacteria found on the skin and in the nose of healthy people. When bacteria are living on or in the human body, but are not causing infection, it is called "colonisation" and the person is said to be a "carrier". Humans are most often colonised with *Staphylococcus aureus* in their nose but it can also found be on the skin and other body sites.

How do you get it?

Staphylococcus aureus can be spread by skin-on-skin contact, by touching infected surfaces or droplet infection. It can cause a range of mild to severe infections. This bacteria is one of the most common causes of skin infections such as pimples and boils.

Staphylococcus aureus frequently affects the lungs of people with CF. Approximately 30% of people with CF are permanently colonised with *Staphylococcus aureus* in the lungs.

How do you treat it?

The growth of *Staphylococcus aureus* in the sputum should be treated with appropriate antibiotics. It may be difficult to determine if the isolated *Staphylococcus aureus* was from the sputum or from upper airway contamination (especially in young children requiring cough swabs).

For this reason all isolates are usually treated. The most commonly used medication for treating *Staphylococcus aureus* is flucloxacillin, but there are many other antibiotics that are also effective.

Other important notes about this microbe

- *Staphylococcus aureus* is more common in children with CF than in adults when other bacteria can become more of a problem.
- Some clinics give antibiotics to prevent *Staphylococcus aureus* infection (prophylaxis) especially in the first year of life (flucloxacillin).

MRSA: Meticillin resistant *Staphylococcus aureus*

What is it?

Staphylococcus aureus are bacteria that are found on the skin and in the nose of about one third of humans.

Some strains of *Staphylococcus aureus* have become resistant to the antibiotic methicillin (flucloxacillin) and other antibiotics used to treat Staph infections. These are referred to as MRSA and are more difficult to treat.

How do you get it?

You can be infected by MRSA in the community as well as in a hospital or other health-care setting. MRSA can cause infection by getting into the body through broken skin, into the blood stream or into the lungs. People with CF are prone to chest infections and can have MRSA in their sputum and lungs.

How is it spread?

MRSA is spread by direct contact with other people who are infected with MRSA. It can also be spread by using towels, clothes or bed sheets used by a person with MRSA. It can be spread through infected sputum.

How can you prevent the spread of MRSA?

- ***Strict hand washing*** is the best method of prevention.
- Dispose of tissues and sputum appropriately and wash hands.
- Don't share towels, clothes or bed linen.
- Don't share grooming items. For example: toothbrushes, scissors, tweezers or razors.
- Single room isolation is recommended when someone with MRSA is in hospital as well as in the CF outpatient clinic.

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How do you treat it?

Sometimes MRSA is isolated in the sputum of a person with CF but is not thought to be causing problems clinically, i.e., not contributing to lung disease. For this reason there will be some instances in which MRSA is not treated.

Specific antibiotics can be used to medically treat MRSA. Suitable antibiotics differ from person to person and will be determined by the medical team caring for each individual patient. Antibiotic treatment for MRSA will not always be successful in eradicating this bacteria and the patient may end up having it present long term (be colonised).

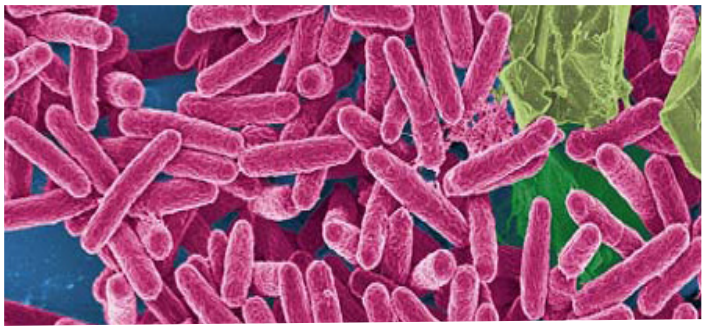
What is the difference between infection and colonisation?

Colonisation means the bacteria are present but are not causing disease or damage to the lungs. However, infection means that the bacteria are present and multiplying, which leads to disease and inflammation.

Pseudomonas aeruginosa

What is it?

Pseudomonas aeruginosa are the most important bacteria resulting in significant lung infection in people with CF. The majority of people with CF will develop a *Pseudomonas* infection at some point. US data quotes the following figures: 30% of children at age 5yrs will have had a Pseudomonas infection and 80% of people with CF over the age of 18yrs have a chronic *Pseudomonas* infection.



Reference is made to non-mucoid and mucoid *Pseudomonas*. For most people with CF their first *Pseudomonas* infections are with the non-mucoid strain. There is good evidence that with aggressive treatment non-mucoid *Pseudomonas* can be cleared.

Over time there is a transition to the mucoid form of the bacteria. Mucoid Pseudomonas has developed a "film" around it that makes it much more resistant to antibiotics. It almost always results in a chronic infection and most doctors agree that mucoid Pseudomonas cannot be cleared.

How do you get it?

Pseudomonas aeruginosa are environmental microorganisms found in water and soil. The bacteria can also be transmitted by droplets. Exposure to Pseudomonas aeruginosa in the community is unavoidable.

People with CF will develop *Pseudomonas* at different times in their lives. It is most commonly acquired from an environmental source but may be acquired via cross infection from another person already infected with the organism. Several studies have reported cross infection between patients is possible from sharing contaminated equipment.

How do you treat it?

Prevention

It is recognised that prevention of chronic colonisation with *Pseudomonas* is important. This is not as easily achieved because, as noted above, the most likely place of acquiring *Pseudomonas* is from the environment.

Some of the ways you can decrease the likelihood of acquiring *Pseudomonas* are:

- Careful handwashing at all times, but especially when in a hospital environment including lung function units,
- Maintaining a distance of approximately 1metre from other people with CF.
- Avoiding areas that are known to hold an increased risk, for example, spas and hot tubs.
- Attending CF clinics with strict policies of separating patients with *Pseudomonas* from those without Pseudomonas.

Antibiotic Treatment

There are few oral antibiotics suitable for treating *Pseudomonas*. One oral antibiotic, Ciprofloxacin, is useful and is often used in conjunction with nebulised antibiotic treatment.

Nebulised antibiotics are the best option for home treatment of Pseudomonas. Tobramycin is most commonly used but other nebulised antibiotics include Colistin, Aztreonam, Ceftazidime and Timentin.

Nebulised antibiotics are beneficial as they target the site of infection. A course of nebulised antibiotics can last anywhere from a couple of weeks to a couple of months, depending on the CF centre policy and the severity of the infection. Many people with mucoid *Pseudomonas* take nebulised antibiotics on a long term basis (often for years). This is to keep the levels (bacterial load) of *Pseudomonas* low and to minimise symptoms.

Intravenous Antibiotics (IVAB) are commonly used for *Pseudomonas* infection. Initially doctors are likely to use IVAB if nebulised antibiotics have failed to clear non-mucoid *Pseudomonas*. They may be used instead of nebulised antibiotics for *Pseudomonas* if the person is unwell or displaying other symptoms (weight loss, decreased lung function, etc).

Other important notes about this microbe

- The majority of CF clinics now separate patients with known *Pseudomonas* infections from patients without *Pseudomonas*.
- Be aware of places where water lies stagnate as a possible high risk of infection. *Pseudomonas* has been isolated from children's bath toys (the kind that have holes and hold water).
- Regular sputum sampling is important to detect *Pseudomonas* early.

Candida

What is it?

Candida is a type of yeast fungi that normally inhabit our digestive system: mouth, throat and intestines.

Candida is a normal part of the bowel flora (the organisms that naturally live inside our intestines). It has many functions inside our digestive tract, one of them is to inhbit growth of harmful bacteria. Without Candida in our intestines we would be defenceless against many pathogenic bacteria.

Even though there are more than 150 species of *Candida*, no more than ten cause disease in humans. *Candida* is grown frequently from the sputum of individuals receiving antibiotics for chest infections.

Where do you get it?

Candida can be found in soil, inanimate objects, food and hospital environments.

How do you treat it?

Treatment is not always required when *Candida* is present in the lung of a CF patient. Treatment is usually only given when symptoms are present. Treatment is also dependent on the CF treating team but usually by administering an antifungal medication (intraconazole, fluconazole or amphotericin B).

Burkholderia cepacia

What is it?

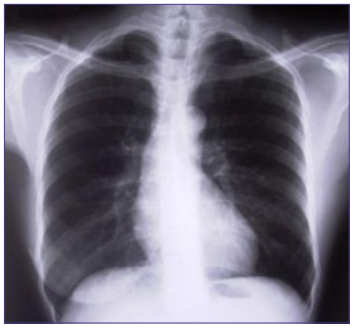
Burkholderia cepacia have recently (in the 1980's) been recognised as important pathogens in patients with CF. Formerly known as *Pseudomonas cepacia,* they are found in approximately 5% of people with CF. *Burkholderia cepacia* has generated considerable anxiety amongst people with CF because of its resistance to many available antibiotics and its ability to be passed from person to person. Increased knowledge about this bacteria has changed the way in which CF care teams manage people with *Burkholderia cepacia* in their clinic. *Burkholderia cepacia* rarely causes problems in healthy individuals.

There are several different genomovars (types) of *Burkholderia cepacia.* Some of these genomovars cause few problems and there is no significant change in the patient's level of health.

Other genomovars, however, have been associated with a significant change in the patient's lung function and overall health status *Burkholderia cepacia* Syndrome is when someone infected with the bacteria experiences a rapid decline in health. A patient's sputum can be tested to see what type of *Burkholderia cepacia* (genomovar) is present.

Where do you get it?

Burkholderia cepacia are environmental microorganisms found everywhere. They are often associated with soil, plants and water (just like *Pseudomonas aeruginosa*) and are capable of utilising a wide variety of substances for growth. *Burkholderia cepacia* survive on the skin for up to 60 minutes, on a moist surface for up to one week, and in water for years. This makes indirect spread possible.



How do you treat it?

As previously mentioned, most types of *Burkholderia cepacia* are very resistant to antibiotics. Once someone has acquired *Burkholderia cepacia*, it is very difficult to clear it from the sputum and most people are said to be colonised from that point on. People who have few symptoms and who have not shown significant decline in health may not be 'actively treated' (given antibiotics) specifically for *Burkholderia cepacia* on a long term basis. Antibiotic treatment, and when to treat, is decided by the patient's CF consultant.

The treatment of acute flare ups of infection (exacerbation) in patients with *Burkholderia cepacia* is more problematic. Even though in the lab *Burkholderia cepacia* appears to be resistant to most antibiotics, a number of studies have demonstrated that there is frequently a reasonable response (improvement in health status) to some antibiotics used in combination. Each individual can have their sputum tested to identify the best combination of antibiotics for them.

Other important notes about this microbe

Since the 1980's, when *Burkholderia cepacia* was recognised as a problem for people with CF, segregation and strict hygiene has helped limit the spread of the microbe.

People with *Burkholderia cepacia* should be seen separately to other people with CF in a clinic and during inpatient

hospital admissions. Individual hospitals will have policies on management of people with *Burkholderia cepacia* infections.

In the past, *Burkholderia cepacia* may have excluded a person from lung transplant at some centres, but more recent experience has shown that B*urkholderia cepacia* doesn't always cause problems post transplant and is no longer recognised as an exclusion criteria.

Aspergillus and ABPA

What is it?

Aspergillus species are mould found in the natural environment. Commonly found in soil, plants, vegetables and decomposing organic matter, such as: compost heaps. They can also be found in air conditioning systems.

How do you get it?

The *Aspergillus* spores are a perfect size to be breathed in and can reach far into the airways and airsacs (alveoli) where they can grow at human body temperature.

Growth of *Aspergillus* from sputum in CF is relatively common. The wide range of reported prevalence is related to the degree of exposure to the spores. In general, people living in rural areas or poorly ventilated houses have higher rates of colonisation.

In most people, *Aspergillus* will not usually cause disease. However, some people, particularly those with CF and Asthma, may be allergic to *Aspergillus* and this allergy, or hypersensitivity, can cause a disease called Allergic Bronchopulmonary Aspergillosis (ABPA).

What is ABPA?

ABPA is an allergic reaction that occurs when the airways become colonised with Aspergillus. ABPA has been reported in 6-25% of people with CF and is less common in children. Whilst ABPA is uncommon, it is a serious respiratory condition characterised by chronic airway inflammation and airway damage.

The symptoms of ABPA may include wheezing and breathlessness, coughing up of brown mucous plugs or haemoptysis (blood in your sputum), loss of appetite, and general malaise / lethargy. Symptoms of ABPA are similar to many other symptoms that occur in CF lung disease. ABPA should be considered in people with worsening chest symptoms or falling lung function despite treatment.

How do you diagnosis ABPA?

It is often difficult to confirm a diagnosis of ABPA in people with CF. If ABPA is suspected a range of diagnostic tests will be performed. These include chest X-ray, skin Prick Tests to look for allergies to the fungus aspergillus, blood tests and sometimes sputum tests or CT scans.

In order to diagnose ABPA the person must show a positive skin test and evidence of allergic inflammation with elevated IgE (an immunoglobulin level) in the blood.

How do you treat it?

The treatment of ABPA aims to control episodes of inflammation and prevent damage to the lung. Corticosteroids, such as Prednisolone, are very effective at reducing this inflammation, and is the most common treatment. There is also more recent evidence to support the use of anti-fungal agents, such as Itraconazole, in the treatment of ABPA, and this may be used in conjunction with Prednisolone. Both of these treatments are tablets taken on a daily basis over a period of time. They are not usually long term treatments.

Haemophilus Influenzae

What is it?

Haemophilus influenzae are bacterial pathogens which, in small numbers, are part of the normal flora of the upper resperatory tract (pharynx and the mouth).*Haemophilus influenzae* are the third most commonly found bacteria in the respiratory tract of young people with CF, affecting approximately 5 - 10% of patients in the first five years. It is infrequently seen in adult patients with CF.

Where do you get it?

Haemophilus influenzae are part of the normal flora of the upper respiratory tract. They are opportunistic bacteria in people with CF and can be transmitted from person to person through droplets or from coming into contact with infected area.

How do you treat it?

Haemophilus influenzae can be treated with a wide variety of antibiotics, examples include; amoxicillin, erythromycin, co-trimoxazole (Bactrim), augmentin duo.

Further Information:

- Infection Control Guidelines for People with Cystic Fibrosis (PWCF) outside of hospital – CF Ireland

Note: this leaflet was reproduced with permission from the New South Wales CF Nurses Working Group - May 2006; www.cysticfibrosis.org.au. It was adapted by the Irish Cross-Infection Working Group in 2014.

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