Respiratory Bugs
Common in People with Cystic Fibrosis

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

Staphylococcus aureus

What is it?
Staphylococcus aureus is often referred to as ‘Staph aureus’ or just ‘Staph’. Meticillin resistant Staphylococcus aureus (MRSA) are Staphylococcus aureus bacteria that are resistant to meticillin and frequently other antibiotics.

Staphylococcus aureus (Staph) 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 Staph in their nose but it can also found be on the skin and other body sites.

Where do you get it?
Staph can be spread by skin-on-skin contact, by touching contaminated 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. Staph bacteria frequently affects the lungs of people with CF. Approximately 30% of people with CF are permanently colonised with Staph in the lungs.

How do we treat it?
The growth of Staph in the sputum should be treated with appropriate antibiotics. It may be difficult to determine if the isolated staph 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 Staph is flucloxacillin but there are many other antibiotics that are also effective.

Other important notes about this bug
• Staph 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 Staph infection (prophylaxis) especially in the first year of life (flucloxacillin).

Pseudomonas aeruginosa

What is it?
Pseudomonas aeruginosa is 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 can not be cleared.

Where/how do you get it?
Pseudomonas is an environmental species found in water and soil. The bacteria can also be transmitted by droplets. Exposure to Pseudomonas 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 and infection is possible from sharing contaminated equipment.

How do we 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:

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

**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 bug**

- 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.

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**MRSA: Meticillin resistant Staphylococcus aureus**

**What is MRSA?**

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

Some strains of Staph have become resistant to the antibiotic meticillin (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 MRSA?**

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 MRSA 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 e.g., 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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Treatment
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 treat MRSA. Suitable antibiotics differ from person to person and will be determined by the medical team caring for each individual person. It must be remembered that antibiotic treatment for MRSA will not always be successful in eradicating this bacteria and the person may end up having it present long term (be colonised).

What is the difference between infection and colonisation?
Colonisation means the bacteria is present but it is not causing disease or damage to the lungs. Infection means that the bacteria is present and multiplying and is also causing a disease and inflammation.

Candida
What is it?
Candida are 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 inhibit 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 usually not required when Candida is present in the lung of a PWCF. Treatment is only given when symptoms are present that do not respond to other treatments. Treatment is dependent on the CF treating team but usually consists of administering an antifungal medication (itraconazole, fluconazole or amphotericin B).

Burkholderia Cepacia
What is it?
Burkholderia cepacia (Cepacia) has recently (in the 1980’s) been recognised as an important pathogen in patients with Cystic Fibrosis. Formerly known as Pseudomonas cepacia, it is a bacteria found in approximately 5% of people with CF. 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 Cepacia in their clinic. Cepacia rarely causes problems in healthy individuals.

There are several different genomovars (types) of Cepacia. Some of these genomovars cause few problems and there is no significant change in the persons level of health.

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

Where do you get it?
Cepacia is an environmental organism found everywhere. It is often associated with soil, plants and water (just like Pseudomonas aeruginosa) it is capable of utilising a wide variety of substances for growth. Cepacia survives 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 we treat it?
As previously mentioned most types of 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 Cepacia on a long term basis. Antibiotic treatment, and when to treat, is decided by the persons CF Consultant.

The treatment of acute flare up’s of infection (exacerbation) in patients with Cepacia infection is more problematic. Even though in the lab 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 bug
Since the 1980’s when Cepacia was recognised as a problem for people with CF. Segregation and strict hygiene has helped limit its spread. People with Cepacia should be seen separately to other people with CF in clinic. They
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should be separated from other people with CF during inpatient hospital admissions. Individual hospitals will have policies on management of people with Cepacia infections.

In the past Cepacia may have excluded a person from lung transplant at some centres but more recent experience has shown that Cepacia doesn’t always cause problems post transplant and it is not longer recognised as an exclusion criteria.

Aspergillus and ABPA

What is it?
Aspergillus species are mould fungi found all over the natural environment.
Commonly found in soil, plants and vegetables and decomposing organic matter such as compost heaps. It 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 Cystic Fibrosis 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. It 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 we 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 we 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 this 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 is a bacterial pathogen which, in small numbers, are part of the normal flora of the pharynx and the mouth (upper respiratory tract). It is 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?
It is part of the normal flora of the upper respiratory tract. It is an opportunistic bacteria in people with CF. It can be passed from person to person through droplets or from coming into contact with similar surfaces.

How do we treat it?
Haemophilus can be treated with a wide variety of antibiotics, examples include; amoxicillin, erythromycin, cotrimoxazole (Bactrim), augmentin duo.
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E.coli (Escherichia coli)

What is E. coli?
E. coli is a gram negative bacteria in the large family of Enterobacteriaceae. It is a common type of bacteria which normally lives in your intestines where it helps your body break down and digest food; however if E. coli finds its way into other parts of your body such as your urinary tract or lungs it can cause you to become sick. E. coli can be present without causing any symptoms.

As CF lungs are prone to bacterial infections, it is not uncommon for E. coli to be present in the lungs of young CF babies, who have a reduced ability to fight infection. It will often not cause any symptoms and will clear without treatment.

Where/How do you get E. coli?
E. coli bacteria are everywhere in the environment. Our hands can become contaminated with our own E. coli if we don’t wash them properly after using the bathroom. They are a common bacteria of all animals. Therefore anytime we eat or drink something or touch something that has either been a part of or near where animals are, there is an increased potential for coming into contact with this bacteria.

To reduce the chances of E. coli infection:
• Good hand hygiene is recommended, especially after using the bathroom or changing nappies because there are large amount of E. coli in faecal matter.
• Avoid undercooked meat and ensure kitchen benches are cleaned thoroughly after preparing raw meat.

How do we treat E. coli?
E. coli is often not treated if the person is not symptomatic.

Klebsiella Pneumoniae

What is it?
Klebsiella is one of the most common gram negative bacteria, classified in the large family of Enterobacteriaceae. It is part of the normal flora in the intestine. Klebsiella is everywhere and may colonise the skin, throat or gastrointestinal tract in humans.

Where/How do you get it?
Klebsiella is an opportunistic pathogen found in the environment and on mucosal surfaces. It is commonly passed by direct contact and spread may be the result of poor hand hygiene.

How do we treat it?
Treatment is often not required if the person is not symptomatic.

Serratia Marcescens

What is it?
Serratia Marcescens is an opportunistic gram-negative bacteria. It belongs to the large family of Enterobacteriaceae.

Where/How do you get it?
It is found naturally in water and soil and may opportunistically cause respiratory infections. It thrives in moisture, dust and phosphates and needs almost nothing to survive.

How do we treat it?
Treatment is not required if the person is not symptomatic.

Further Information:
• Infection Control Guidelines for People with Cystic Fibrosis (PWCF) outside of hospital – CF Ireland

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Definition of Terms and Glossary

Antimicrobial
A drug used to treat infections caused by bacteria and other microorganisms.

A chemical substance that has the capacity to kill or inhibit the growth of microorganisms.

Bacteria
Single-celled microorganisms which can exist either as independent (free living) organisms or as parasites (dependent on another organism for life).

Colonisation
Colonisation means the bacteria/fungus is present but it is not causing disease or damage to the lungs.

Cross Infection
Spread of infection from one person to another.

Environmental
Pertaining to the environment; to exist in the natural community, in air, dust, sediment, soil and water.

The complex of physical, chemical and biotic factors (as climate, soil and living things) that act upon an organism.

Fungi
Fungi can be true pathogens that cause infections in healthy people or they can be opportunistic pathogens such as aspergillus and candida that cause infections in immunocompromised people. An example of a common fungus is the yeast organism candida that causes thrush.

Fungi are present in the soil, air and water but only a few species can cause disease.

Infection
A process of infecting or being infected by bacteria/fungi/viruses. Infection means that the pathogen is present and multiplying and is also causing disease and inflammation.

Intravenous
Administration of fluids or medications into a vein.

Normal Flora
The growth of the usual bacteria that live in the intestinal tract and the mouth.

Opportunistic
An infection that occurs because of a weakened immune system.

A bacteria, virus or fungus that takes advantage of certain opportunities to cause disease. Those opportunities are called opportunistic conditions.

Pathogen
Agent causing disease.

A disease producer. The term pathogen is most commonly used to refer to infectious organisms. These include bacteria, viruses and fungi.

Prevalence
The proportion of individuals in a population having a disease. Prevalence is a statistical concept referring to the number of cases of a disease that are present in a particular population at a given time.

Resistant
The natural ability of a normal organism to remain unaffected by noxious agents in its environment.

The ability of bacteria and other microorganisms to withstand an antimicrobial to which they were once sensitive.

Symptomatic
Exhibiting the symptoms of a particular disease.