What is the risk of COVID-19 to people with CF?

- It is unclear yet if people with CF have a higher risk of getting more severe COVID-19 infection as compared to peers of their own age who do not have CF. Early experience from elsewhere in the world suggests this is not the case but this is being closely monitored across the world.
- People with lower lung function (FEV1 < 60% predicted), diabetes, high blood pressure or had organ transplants should be regarded as higher risk.
- People with CF and household family members can acquire COVID-19 infection like anyone else. Importantly people with CF who have the virus may potentially spread the virus more easily through coughing, physiotherapy and nebuliser treatments (see Physio section below). Extra precaution is needed in such cases.

What precautions should people with CF be taking to protect themselves and family?

- There is currently no vaccine to prevent COVID-19. The best way to prevent illness is to avoid being exposed to this virus.
- The virus is spread mainly from person-to-person:
  - Between people who are in close contact with one another (within 1.5 m).
  - Through respiratory droplets produced when an infected person coughs, sneezes or talks.
  - These droplets can land in the mouths or noses of people who are nearby or possibly be inhaled into the lungs.
  - Some recent studies have suggested that COVID-19 is also spread by people who are not showing symptoms.
- Wash your hands with soap and water for at least 20 seconds especially after you have been in a public place, or after blowing your nose, coughing, or sneezing.
- If soap and water are not readily available, use a hand sanitizer that contains at least 60% alcohol. Cover all surfaces of your hands and rub them together until they feel dry.
- Avoid touching your eyes, nose, and mouth with unwashed hands.
• Avoid close contact with people who are sick, even inside your home. If possible, maintain 1.5 m between the person who is sick and other household members.
• Maintain social distancing and put distance between yourself and other people outside of your home.
• Remember that some people without symptoms may be able to spread virus.
• Do not gather in groups
• Stay away from crowded places and avoid mass gatherings and public transport.
• Cover your mouth and nose with a cloth mask cover when around others. You could spread COVID-19 to others even if you do not feel sick.
• Wear a cloth mask if you go out in public places. The cloth mask is meant to protect other people in case you are infected and vice versa.
• Continue to keep about 1.5 m between yourself and others. The cloth mask is not a substitute for social distancing.
• If you are in a private setting and do not have on your cloth mask, remember to always cover your mouth and nose with a tissue when you cough or sneeze or use the inside of your elbow.
• Throw used tissues in the trash. Immediately wash your hands with soap and water for at least 20 seconds. If soap and water are not readily available, clean your hands with a hand sanitizer that contains at least 60% alcohol.
• Clean AND disinfect frequently touched surfaces daily. This includes tables, doorknobs, light switches, countertops, handles, desks, phones, keyboards, toilets, faucets, and sinks. If surfaces are dirty, clean them. Use detergent or soap and water prior to disinfection.

Is it safe for people with CF to return to work and children to school?

• The decision to return to work, college, university or school is a complex and personal one. Each person and family must make an informed decision after consulting with their doctors and relevant employers or education authorities. Each person and family have unique circumstances and risk levels which must be individually assessed.
• In general, it is sensible for people with CF to take extra precautions and avoid contact with other people as much as possible. This applies especially to those with lower or rapidly declining lung function (FEV1< 60%); diabetes, very frequent chest infections and organ transplant recipients.
• It is strongly recommended that adults work from home where at all possible and maintain strict social distancing all the time.
• It is strongly recommended that university and college students should where possible use remote learning and avoid campus activities where possible.
• Children and school:
  o parents and families must consult with their doctors to determine your child’s individual risk which considers personal, school and family circumstances.
  o In general, children are LESS likely to get or spread COVID-19 infection. If they do get infected then the illness is likely to be mild, like a common cold.
  o Until more evidence emerges, it is recommended that children with CF who are frequently sick, are prone in frequent chest infections or have any signs of lung damage should avoid school or pre-school at this stage and distance learning be followed where possible.
o For healthy CF children with heathy lungs and no CF-related complications, it is probably safe to attend school, provided the school takes all reasonable measures to prevent spreading infection at school which include:
  ▪ learners and staff are screened for symptoms on daily basis and quarantine measures are enforced for children or staff members who display symptoms or have close COVID-19 contacts.
  ▪ Classroom size is reduced, and distancing measures are in place.
  ▪ Frequent hand sanitising and wearing face masks is widely practised.
  ▪ Sport and extracurricular activities involving groups or close contact with other children are avoided.

• In households where vulnerable or unwell individuals with CF live, the risk of family members/siblings bringing home COVID-19 from work school must be considered. In some high-risk instances, it may be necessary for family members to take extra precaution with social distancing and self-quarantine to protect their loved ones with CF at home.

Should I attend regular check-ups at my doctor or CF clinic for scheduled appointments?

• Check with your doctor or clinic for updates about routine check-ups and appointments. It is generally recommended that routine in-person visits are deferred until further notice if you or your child are WELL. Tele-consultations should be attempted as far as possible.
• It is important to make sure your supply of chronic medications is not disrupted, and prescriptions are regular renewed.
• Discuss with your doctor the need for routine blood tests, lung function, x-rays or other tests. Some of these can be deferred but others not. Regular sputum tests should be continued to screen for infections that need prompt treatment. See section below on the best way of collecting sputum samples.
• If you do attend clinic, wear a cloth mask and practice good hand and cough hygiene. Try avoiding public transport or crowded public areas if possible.

When is it necessary to do lung function testing and is it safe?

• Lung function testing should generally be avoided because it requires the individual to forcibly blow into a mouthlike which will release a lot of droplets into the air. This poses a risk to health care workers and other people who enter the room. Fomite spread onto surfaces may also pose a risk to patients that follow.
• Lung function testing at your doctor, clinic or hospital should only be performed if it will influence clinical management or treatment.
• Ideally, lung function testing should be done only after a negative COVID-19 PCR result is obtained within 48 hours before the time, or the person has been in an isolation room or in self quarantine since performance of the last negative COVID-19 PCR test. If a COVID-19 test is not done or result not available, and lung function testing deemed urgent by your doctor, then:
  o the technician/doctor must use full PPE – as directed by your respective hospital/clinic for anyone performing an aerosol generating procedure.
  o the room/space in which the lung function is performed has adequate ventilation.
  o the room/space in which the lung function is performed is deep cleaned following the performance of the lung function i.e. 3 cleans with or hypochlorite solution, followed
by “fogging” with hydrogen peroxide or “zapping” with high frequency ultra-violet light, before staff or another patient use the room or lung function equipment again

- The room/space should not be used by anyone else for at least 30 minutes after a lung function test is done.
- In some instances, your doctor will recommend lung function self-monitoring at home on handheld devices linked to a smartphone app or computer. It is strongly recommended that health care funders fund these from an equipment or special COVID-19 budget if possible. Check with your doctor if home lung function monitoring is advisable.

**What if I or my child gets sick and I need to see my doctor or CF clinic?**

- Unless it is an emergency, please DO NOT visit your doctor or any emergency department without a prior appointment or making contact for advice ahead of time.
- It is important that you contact your doctor or CF clinic for advice and a teleconsultation if possible. Your doctor will decide if you will need to be seen or what treatment or tests are needed.
- If you do travel to the visit doctor or hospital, always wear a cloth mask and practice good hand and cough hygiene. Try avoiding public transport or crowded public areas where at all possible.

**When is it necessary to do sputum tests and what is the safest way to do sputum tests?**

- If you/your child are well: It is important to continue to take regular “surveillance” sputum tests, so that any new infections are picked up and treated quickly. These should be taken at least 4 times a year.
- In young children or infants who cannot cough and spit secretions, discuss with your CF doctor about when it is necessary to take a routine “cough swab” If a routine cough swab is obtained by a health-care professional, this should be considered an aerosol generating procedure, and appropriate PPE should be worn for airborne transmission.
- If you/your child become sick with a chest infection, you should try collect and send off a sputum specimen, after discussing with your CF doctor.

*For outpatients:* If possible, a sputum specimen should ideally be taken at home, for older children and adults able to cough and spit. The specimen should be sealed and brought to the laboratory or hospital (discuss with your CF doctor). When taking a specimen at home, the person with CF should either be outside (ideally); or in a separate room from other members of the household, with the windows wide open to improve ventilation.
- Wash hands before taking the specimen, and then loosen the specimen bottle lid.
- Whilst coughing, a tissue or the crook of the elbow should be in front of the mouth, to limit aerosol spread.
- After sealing the specimen bottle, wash hands again and disinfect the outside of the bottle.
- Place the bottle in a zip-seal plastic bag.
Younger children and infants may need to come to the hospital or laboratory service to obtain a sputum specimen and/or for clinical examination and/or treatment.

- Induced sputum specimens with nebulisation of hypertonic saline are not recommended.
- Sputum specimens obtained by naso- or oropharyngeal suctioning should be considered high-risk procedures. If clinically necessary, these should be done in an isolation facility/room without other patients or staff present, and full airborne infection control precautions and PPE.

Inpatients admitted to hospital should provide a sputum specimen. They should be in an isolation facility/room (ideally with negative pressure ventilation) during the procedure.

Older children and adults should ideally produce a specimen without anyone in attendance.

Where a physiotherapist or other healthcare worker is needed to help the patient produce the specimen, full PPE and infection control measures for airborne transmission should be adhered to.

**Should I continue chest physiotherapy on nebuliser treatments at home or in hospital and when is it not safe to do this?**

**Airway clearance/physiotherapy at home**

- It is very important that you/or your child continue doing your usual regular chest physiotherapy/airway clearance techniques at home during the COVID-19 pandemic.
- If you used a nebuliser before the COVID-19 outbreak, you should carry on using the nebuliser at home (e.g. for hypertonic saline/pulmozyme/inhaled antibiotics).
- People with CF should try and continue with regular exercise as far as possible.
- If a person with CF becomes infected with COVID-19, they should ideally do their airway clearance treatment in a separate room from other household members, or outdoors (if possible).
- “Cough etiquette” should be adhered to, to prevent spread of possible infection:
  - turn head away from others during cough and expectoration (spitting)
  - if possible, you should “catch your cough” with a tissue, dispose of tissue and perform hand hygiene.
  - If no tissue, rather cough into the crook of your elbow sleeve
- It is very important to keep all equipment as clean as possible. All airway clearance devices (e.g. flutter/bronch-U-vibe/PEP/Acapella) and nebuliser consumables must be washed with warm soapy water after every use, and sterilised once a week (e.g. using a microwave steriliser or Miltons)
- If nebuliser filters need replacing, preferably do this outside and discard the filter immediately. Wash hands thoroughly afterwards. It is best if someone in the household without CF changes the nebuliser filter.
- Wash hands well after nebulising and after every physio session.

**Airway clearance/physiotherapy and nebulisation in hospital**

- If a person with CF is admitted to hospital, they should receive regular airway clearance therapy by a trained physiotherapist, wherever possible and when indicated.
o Where medically necessary, airway clearance therapy should be done in a negative-pressure room, if available, or in a single room with the door closed.

o Only the minimum number of required staff should be present, and they must all wear PPE as appropriate for airborne transmission.

o Entry and exit from the room should be minimised during the treatment.

o If possible, when performing techniques that are likely to generate a cough, physiotherapist should position themselves ≥ 2m from the patient and out of the “blast zone” or line of cough.

o Bubble-PEP is not recommended.

o Where airway clearance devices (e.g. flutter/bronch-U-vibe/PEP/Acapella) are used, these should be single-patient use, and must be cleaned after each use.

- Where medically necessary, nebulisation of essential medication (e.g. pulmozyme or antibiotics) may continue in hospital, but ideally these should be given in a negative pressure/single room with the door closed and no staff in direct attendance.

- Because people with CF potentially aerosolise more droplets into the air with coughing and physiotherapy, it is advisable that the all healthcare workers entering the hospital room wear PPE (N95 mask, face shield/goggles and gloves/apron) at all times, even if the COVID-19 test is negative. This is because the COVID-19 test can sometimes be inaccurate i.e. may be negative at first and then become positive later.

What if I or my child needs hospital admission, IV treatment or other urgent medical treatment or tests?

- Admission to hospital for IVs or other reasons should proceed as usual if required. The choice of hospital vs. home-based IV therapy should be guided by your doctors’ recommendations and opinion, including re-admission to hospital if home IV therapy is not working.

- a COVID-19 test (with result) prior to admission must be done for all planned or non-urgent admissions to hospital. This is important to exclude COVID-19 infection and avoid inadvertent exposure of COVID-19 to healthcare workers and people around you/your child.

- With exception of lung function, all relevant laboratory and other tests deemed necessary by your doctor must be done as usual.

- Each hospital has instated COVID-19 admission policies and rules which will need to be followed in case of emergency and any admissions. Please discuss with your doctor.

- General CF care should not be neglected nor set aside during admission. Whilst in the ward with emergency admission, all CF treatments must continue as normal with appropriate distancing and PPE measures in place.