

EXERCISE AND CYSTIC FIBROSIS

A GUIDE FOR PERSONAL TRAINERS



About this guide

This booklet is designed to provide information about cystic fibrosis (CF) and how it may affect an individual in an exercise or sporting environment.

We have covered most aspects of CF in this booklet but please be aware that some sections may not be relevant to your client. Individuals are affected very differently by this condition, so it is best to speak with the individual about their specific symptoms and limitations.

Training or coaching someone with CF means that you will be interacting with someone who has a complex chronic illness which requires input from a multidisciplinary team of health professionals on a regular basis. An individual's health can change considerably from month to month. Many people who have CF look like every other person, which can be misleading.

If you would like further information about CF, you can contact your local CF state organisation (details on page 16)



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1. What Is CF?

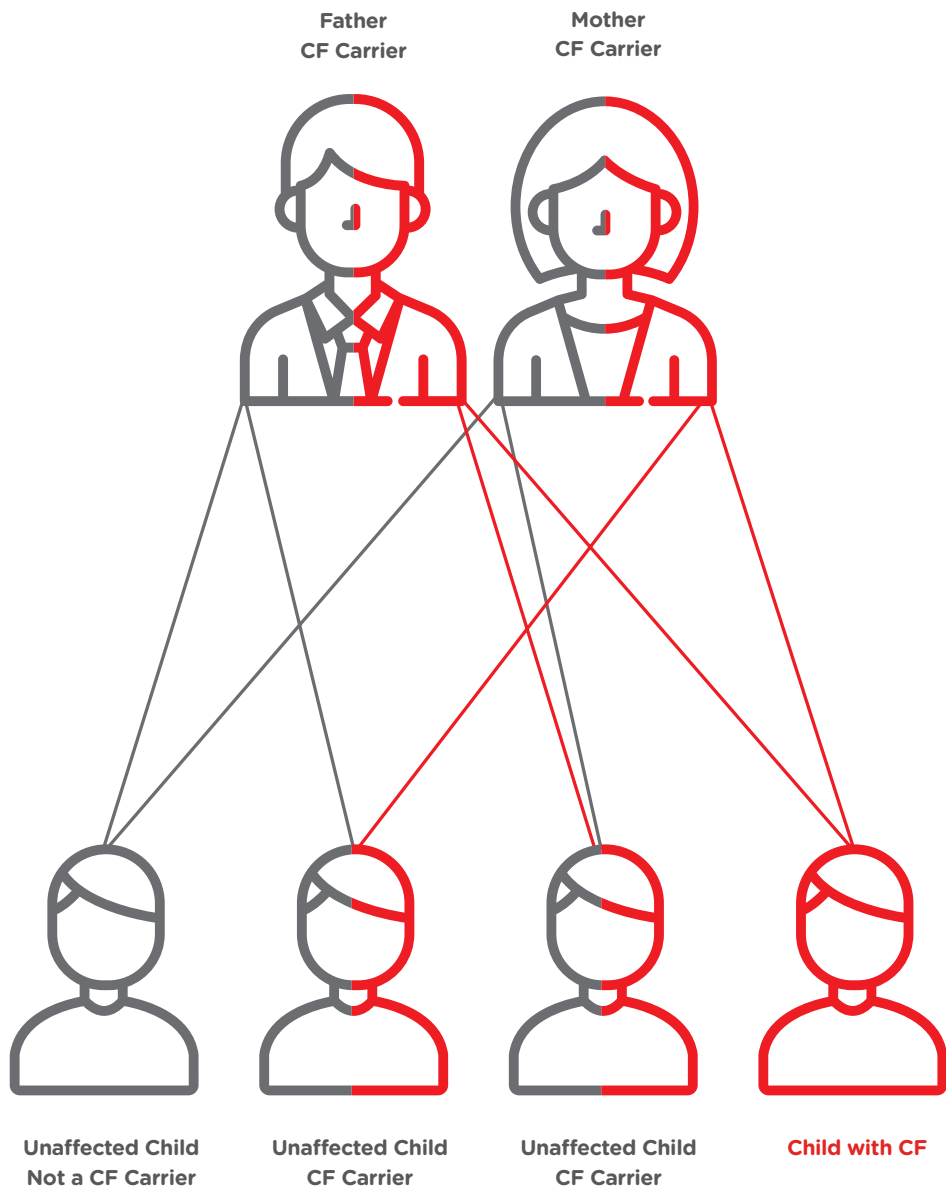
CF is a genetic illness which affects a number of organs in the body. It causes mucus to become thick and sticky leading to infection, inflammation and obstruction of the lungs. Daily and ongoing treatment is required to manage the illness and prolong the life of the person affected.

In most people, the digestive system is also affected. This can cause difficulties in digesting food and nutrients, leading to malnutrition and poor weight gain. This requires people with CF to consume extra calories in their diet and also to take enzyme tablets with most foods to enable adequate absorption of nutrients from the diet.

DIAGNOSIS

The majority of people with CF are diagnosed in the first few weeks of life via the Newborn Screening Test. Some people, although rare, are not diagnosed until symptoms present in later infancy or early childhood. Diagnosis can be confirmed with a sweat test. Treatment begins from the time of diagnosis and continues every day for the rest of their lives.

CF is not contagious. For someone to be born with CF, the CF gene must be passed on by both parents (as demonstrated in the chart to the right).



2. Common symptoms of CF

As with many illnesses, CF has a broad spectrum of symptoms; some people will have a lot while others will have very few.

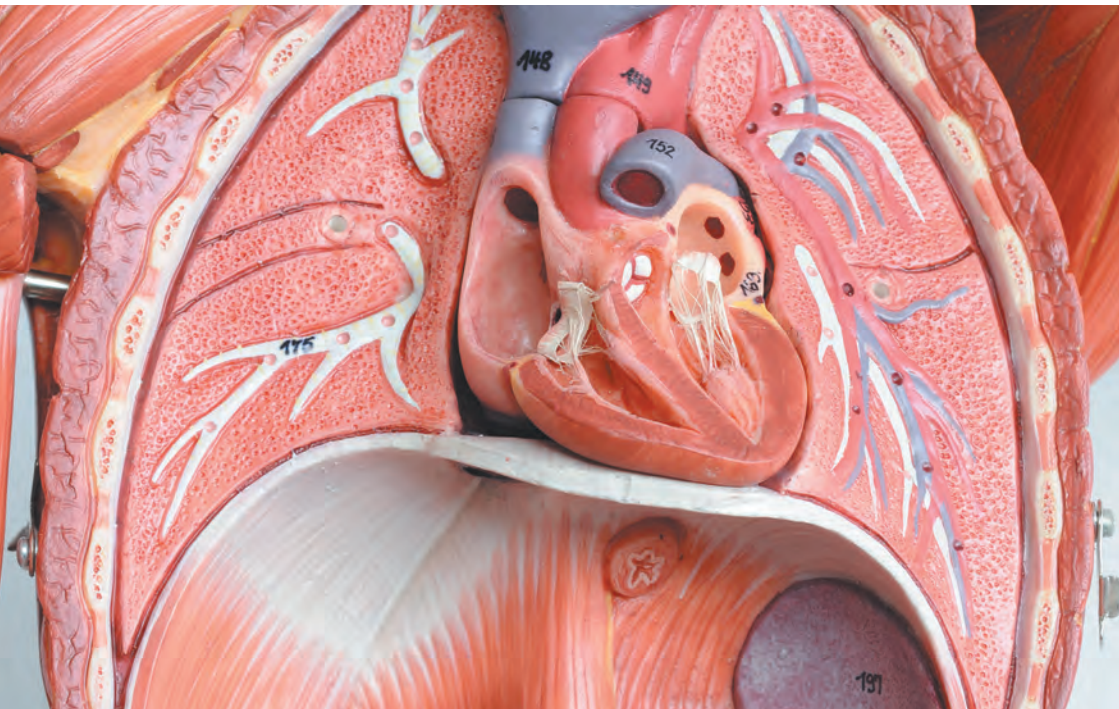
Common symptoms include:

- A persistent cough (particularly after physical effort or with infection)
- Mucus production
- Shortness of breath
- Postural changes
- Low body weight
- Small stature
- Gut issues, including bloated/distended belly

Other medical complications typical in CF include:

- Weakened pelvic floor muscles leading to incontinence
- CF Related Diabetes (CFRD)
- Osteoporosis
- Reflux
- Anxiety and/or depression

The incidence of additional complications increases with age.



3. How is CF Treated?

CF treatments are designed to ease the symptoms and slow the progression of disease. Treatments include a mixture of physiotherapy airway clearance, medication, diet and exercise. Lung transplantation is used when lung function has declined despite other treatment methods.

Physiotherapy Airway Clearance

Airway clearance is an important part of the management of CF. The aim is to clear mucus from the lungs to prevent infections and lung damage. Exercise enhances airway clearance. Individuals will complete their airway clearance at home either before or after exercise.

There are various airway clearance techniques used, and these will change over the individual's lifespan as their disease changes.

Medications

Some medications may have side effects that interfere with an individual's ability to exercise. E.g. nausea, increased sensitivity to sunlight. Please discuss all medications with your client.

Commonly used medications are:

- Antibiotics
- Salt supplements
- Digestive enzymes
- Vitamin supplements
- Anti-inflammatories
- Ventolin



Nutrition

Due to malabsorption of food and increased energy expenditure of people with CF, a high energy diet is required. Most people with CF require 20-50% more calories and three to four times more salt each day to maintain their energy levels, fight infections and promote growth. Eating the required calories can often be a struggle, especially if the person is feeling unwell. The excess salt loss also means they need to increase their salt intake through food. Some people may also need supplementation drinks like Sustagen or Ensure to try and reach their daily calorie requirements.

Just like anyone else, a person with CF may suffer from malnutrition if their body does not get the nutrients it needs.

People with CF follow a food cube diet, rather than the traditional pyramid. They require an even spread of all food groups including high amounts of fats, carbohydrates and proteins to allow their body to function.

Transplants

A double lung transplant may be considered for end-stage lung disease. A lung transplant replaces the CF lungs with healthy lungs from a donor, however it does not prevent or improve any problems that CF may be causing in other parts of the body, such as the digestive system and sweat glands.

Following transplant, individuals will be closely monitored by their transplant team. Before commencing any new exercise program, it is important that the individual discusses, and gets the all clear, from their transplant team.

People who have had a lung transplant may not be able to achieve the same level of fitness as their peers.

4. Exercise and CF

An active lifestyle is extremely important for the CF population, with benefits including increased lung function and overall quality of life. People with CF who keep fitter are shown to cope better with the impact of CF, have fewer infections and generally have healthier lives.

The benefits are not only physical, with improvements often seen in self-esteem and emotional wellbeing as well as decreased anxiety and depression, all of which affect adherence with treatments.

Exercise can also:

- Improve daily functioning
- Slow the rate of decline in lung function
- Assist with clearing mucus from the lungs
- Improve the ability to perform activities of daily living such as cleaning and shopping etc.
- Increase bone density
- Preserve muscle strength and function
- Increase body mass
- Improve appetite
- Improve exercise tolerance
- Improve posture

Aerobic Exercise

Aerobic training is particularly beneficial as the movements involved, such as vibrations in walking, often help to clear mucus from the airways. Aerobic training should never replace airway clearance.

The level of aerobic exercise your client can complete will depend on various factors including current physical fitness, lung function and nutritional status. Just as with any client, it is important to start with a moderate level intensity and build up slowly over time. Your client will be able to advise you if the exercise is too hard for them. Moderate intensity interval training, rather than high intensity work, has been proven to be just as beneficial to people with lung disease.

It is important to consider that weight loss is not usually an exercise goal for people with CF, therefore the calorie burning potential of aerobic exercise should be considered when prescribing exercise for this population. Aerobic training should focus more on building endurance and fitness rather than calorie burning. A well rounded exercise program coupled with an optimal diet is the best way to avoid weight loss.

Resistance/Strength Training

Strength training, just as in the general population, is very beneficial for people with CF. Muscle atrophy is common in CF patients, which is often linked to nutritional deficiencies and cardiorespiratory limitations of the disease. Improvements in muscular strength can be seen through weight training, although progress may be slower than for the general population. To build muscle, increased protein intake is required as CF patients have higher protein requirements, however it is best for their hospital dietitian to advise any changes in diet.



Many people with CF have low bone mineral density (BMD). Weight training is particularly beneficial in increasing bone strength and preventing fractures and osteoporosis.

UPPER BODY TRAINING

Upper body strength training is particularly useful for increasing chest mobility and strength, which aids in the removal of mucus from the airways. People with CF often experience postural changes and can sometimes appear slightly hunched or barrel chested. Upper body strength training and stretching is helpful for minimising such symptoms and improving posture.

LOWER BODY TRAINING

Lower body strength training has the ability to increase leg strength making Activities of Daily Living (ADLs), such as walking, shopping and cleaning easier.

Flexibility

Many people with CF will have postural changes due to coughing and increased work of breathing, including barrel shaped chest and forward rotation of the shoulders.

These changes can result in back pain, joint problems and have a detrimental effect on lung capacity

Flexibility and core strengthening exercises can be particularly beneficial for improving posture and breathing. Keeping the spine, ribcage and shoulders flexible, assists in maintaining good posture and preserving full movement of the joints and muscles around this area.

5. Exercise considerations for people with CF

Hydration

Due to the high level of salt lost in sweat, people with CF are placed at a greater risk of complications from dehydration, so it is important to allow plenty of drink breaks, particularly in hot weather. If your client is exercising in summer, they will need to replace the salt and electrolytes lost in sweat by drinking extra water or sports drinks, taking salt tablets and eating salty foods.

Early signs of dehydration

- Headaches
- Dizziness
- Poor concentration
- Feeling tired/fatigue
- Dry mouth
- Salt crystals on the skin
- Thirst
- Dark urine colour

Late signs of dehydration

- Loss of appetite
- Nausea/vomiting
- Muscle cramps
- Thickened mucus
- Constipation

If your client experiences these symptoms, cease exercise and encourage them to drink more water or a sports drink to help balance out electrolytes. A gym or indoor area with controlled temperature is best during the summer months.

Cough

During aerobic exercise, your client may experience coughing, wheezing or breathlessness. This may be normal, and the coughing should not be suppressed as it is important for clearing the airways. They may need to spit the mucus out or they may just swallow it. Being understanding of

this may help minimise any feelings of embarrassment or discomfort for the client. Any change in normal CF symptoms should be reviewed by the CF team.

Sometimes there may be blood in the mucus which is known as haemoptysis. This can be common, however only in small quantities. If larger quantities are being coughed up, cease exercise immediately. The person should be encouraged to seek medical advice ASAP.

Weight and Food

Individuals should be speaking with their CF care team about their nutrition and should have a good understanding of their own dietary needs.

Exercise will cause the already high energy needs of people with CF to increase even more, so it is important they are eating enough calories to replace those lost in exercise.

A diet high in protein and energy, as well as eating regular meals and snacks, is important to help maintain weight and a higher energy intake. Food should include plenty of meat, fish and eggs, and several high-calorie snacks in between. Encourage your client to speak to their dietitian when starting a new exercise program.

Exacerbation

An exacerbation is an increase in the severity of normal CF symptoms. It may include any or all of the following symptoms:

- Increased cough
- Increased mucus production or change in the colour of mucus
- Shortness of breath (dyspnea)
- Fatigue
- Described sensation of tightness in the chest
- Wheezing
- Decreased exercise tolerance
- Fever

Exercise is not likely to be the cause of an exacerbation, however if your client experiences such symptoms (worse than normal) you should stop exercise and encourage them to contact their CF medical team

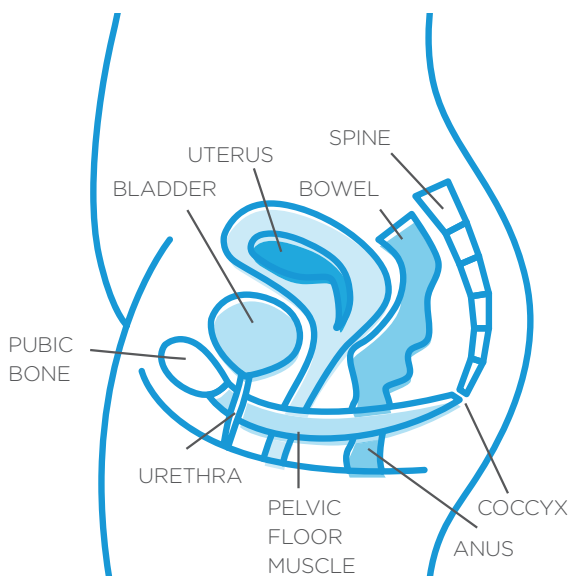
for review. Respiratory exacerbations often require hospitalisation and antibiotic treatment. Frequent exacerbations negatively influence quality of life, leading to absence from work or school, decreased exercise tolerance, decreased appetite and can accelerate lung function decline.

Continence

Many people with CF have weakened pelvic floor muscles which can lead to incontinence of the bladder and bowel. Urinary incontinence (UI) frequently occurs with exercise.

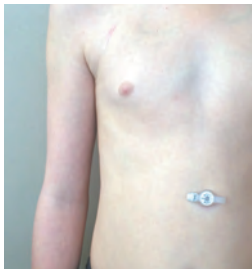
Both men and women with CF are at greater risk of experiencing UI than the general population. This is even common in young people with CF. Symptoms may be worse with high impact exercises. So, as a general rule, try to avoid prescribing high impact or high intensity exercises that place downward pressure on the pelvic floor such as jumping, skipping and running.

Be aware that your client may need more frequent toilet breaks and may feel embarrassed about leakage.



The Pelvic Floor

PEGS, PICCS and PORTS

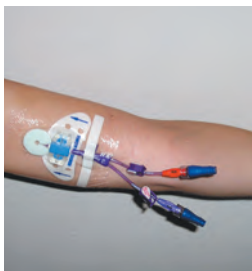


PEG

A PEG (Percutaneous Endoscopic Gastrostomy) is a tube which goes through the abdominal wall into the stomach to provide a means of supplemental feeding when oral intake is not adequate.

As seen in the image, PEGs are quite discreet and are likely to go unnoticed under an individual's t-shirt. A PEG should not impact on an individual's ability to continue their exercise program, however they may occasionally suffer side effects such as nausea, abdominal bloating or diarrhoea. If any complications occur with the PEG, such as infection of the wound, dislodgement, blockage/fracture or leakage of the tube, your client should seek medical attention immediately.

Accidental removal requires urgent action as the tract begins to close immediately and may close completely within hours.



PICC

A PICC line (peripherally inserted central catheter) is a form of intravenous (IV) access that can be used for a prolonged period of time, for antibiotic therapy. For some, lengthy hospitalisations will interrupt day to day life, including their usual exercise routine. Some people may be discharged

from hospital with a PICC line. This allows them to continue to have antibiotics intravenously at home, and they may even return to exercise if they are feeling up to it physically.

As a general rule you should follow these guidelines:

- Do not lift heavy objects with the PICC arm.
- Do not play contact sports.
- Do not swim.
- Do not do repetitive motions such as vacuuming, raking or golfing.



PORT

A port (portacath) is inserted under the skin and connects to a vein to allow direct access to the blood stream. Medications can be injected into the port and blood samples can be drawn many times.

If there are signs of inflammation, swelling, tenderness, discharge or chest pain, the patient should seek medical advice.

If the client has a port, they are still able to exercise as normal. If the client is receiving regular treatment, such as IV antibiotic therapy, the port will be needed. During this time, they should avoid swimming and should use lighter weights for upper body exercises.

Cystic Fibrosis Related Diabetes

Diabetes is a common complication that develops over time in many people with CF. Cystic fibrosis related diabetes (CFRD) is a unique type of diabetes that only people with CF can get. Consequently, treatment of CFRD is not the same as treatment of other types of diabetes.

Exercise is great for people with CFRD as it can help with diabetes control. Any clients with CFRD should have spoken with their CF and diabetes teams before commencing any new exercise program, as exercise can affect Blood Glucose Levels (BGL). Your client and their hospital care team should have the diabetes under control, and it should not impact them in an exercise environment.

People with CF should always carry a fast-acting (jelly beans) and a slow-acting (banana, muesli bar) carbohydrate snack when exercising in case of a drop their BGL (hypo).

Signs of a hypo may include:

- Feeling shaky and weak
- Hunger
- Sweating
- Lightheadedness and dizziness
- Headache
- Pins and needles around the mouth
- Mood change, confusion, irritability

If your client experiences these symptoms, they should stop exercise immediately and check their BGL. If their BGL is low they should have a fast-acting carbohydrate snack followed by a slow-acting carbohydrate snack, monitor their BGL and not return to exercise until they have returned to safe BGL (5.5mmol/L-15mmol/L).

Infection Prevention And Control

A part of everyday life involves reducing the risk of catching and spreading infections. This is true for everyone, not just people with CF. However, common colds and viruses can have more serious consequences for people with CF and can even result in hospitalisations and irreversible damage to the lungs.

Practicing good hand hygiene and wiping down gym equipment is the best way to minimise the spreading of germs. Your client may wish to clean equipment thoroughly before use and may even bring along their own antibacterial wipes.

If you are sick with flu, colds or gastro etc. you should stay at home or inform your client that you have a cold so that they may choose to stay at home.

People with CF are prone to certain lung infections which can prove harmful to others with the condition. To avoid the risk of cross-infection, people with CF are advised not to mix with each other and special care needs to be taken to clean equipment between use if you have more than one client with CF.

6. Useful Resources

CFFIT

Exercise and Cystic Fibrosis: My Exercise Diary

Exercise and Cystic Fibrosis: A Guide for People Living with CF

CFFACT

A range of quick, easy to read fact sheets on various CF-related topics, including

- Airway clearance techniques
- Bone health
- Continence
- Cystic Fibrosis Related Diabetes
- Dehydration
- Germs and cross-infection
- Lung transplants
- Medications
- PEGs
- PICCs
- Ports

CFFOOD

Cystic Fibrosis Nutrition Program: A Guide for Adults

CFBites: Snacks and Meals for those with Cystic Fibrosis

CFCOOKING

A series of cooking demonstration videos for people with CF

All our resources available from our website www.cfwa.org.au

OTHER WEBSITES

Cystic Fibrosis Australia www.cysticfibrosis.org.au

Pelvic Floor First www.pelvicfloorfirst.org.au

Continence Foundation of Australia www.continence.org.au

7. Useful Contact Details

ACT

Cystic Fibrosis Association ACT Inc

☎ (02) 6292 9866
✉ info@cfact.org.au
🌐 www.cysticfibrosis.org.au

NSW

Cystic Fibrosis Community Care (NSW)

☎ (02) 8732 5700
✉ admin@cfnsw.org.au
🌐 www.cysticfibrosis.org.au

QUEENSLAND

Cystic Fibrosis Queensland

☎ (07) 3359 8000
✉ admin@cfqld.org.au
🌐 www.cysticfibrosis.org.au

SOUTH AUSTRALIA

Cystic Fibrosis SA

☎ (08) 8221 5595
✉ cfsa@cfsa.org.au
🌐 www.cysticfibrosis.org.au

TASMANIA

Cystic Fibrosis Tasmania

☎ (03) 6234 6085
✉ general@cftas.org.au
🌐 www.cysticfibrosis.org.au

VICTORIA

Cystic Fibrosis Community Care (CFV)

☎ (03) 9686 1811
✉ admin@cfv.org.au
🌐 www.cysticfibrosis.org.au

WESTERN AUSTRALIA

Cystic Fibrosis Western Australia

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www.cfwa.org.au