



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 Cystic Fibrosis Western Australia (CFWA) or visit the website www.cysticfibrosis.org.au/wa



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What is CF?

CF is a chronic genetic illness which affects a number of organs in the body.

CF causes the mucus in the body to become thick and sticky, leading to chronic infections, 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, preventing the release of enzymes needed for the digestion of food. In healthy individuals, enzymes occur naturally to break down food, however in people with CF, mucus blocking the pancreas prevents their release. 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 fat from the diet.

There are more than 1,500 CF gene mutations and this causes significant variation in severity of the symptoms. When CF was first recognised in the 1930s the outlook was very poor. Although there is still no cure, earlier diagnosis, greater understanding of the condition and better treatments mean the majority of children live well into adulthood.

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).

With each pregnancy there is a:

- One in four (25%) chance that the child will have CF
- Two in four (50%) chance that the child will be a genetic carrier
- One in four (25%) chance that the child will not have CF and will not be a genetic carrier for CF.



Common symptoms of CF

As with many illnesses, CF has a broad spectrum of symptoms; some people will have a lot of symptoms while others will have very few. The main symptoms are respiratory related and will include a persistent cough (particularly after physical effort or with infection), mucus production and shortness of breath. Coughing can also lead to postural changes.

Problems with the digestive system and issues absorbing nutrients from food causes many people with CF to be small in stature and have a low body weight. This may cause other gut issues and result in having a bloated or distended belly.

Other medical complications typical in CF include gastro-oesophageal reflux, CF Related Diabetes (CFRD), infertility, osteoporosis and incontinence. These additional complications increase with age.

As with any chronic illness, someone affected by CF may also experience anxiety or depression. Regular exercise can be of benefit if people are experiencing these conditions.

CF primarily affects the lungs and digestive systems



How is CF treated?

Living with CF is a constant, complex and time consuming process. Treatments are designed to ease the symptoms and slow the progression of disease by maintaining optimal lung function and nutrition.

Treatments include a mixture of airway clearance, medications, diet and exercise. Lung transplantation is used as a last resort when lung function has declined despite traditional treatment methods.

AIRWAY CLEARANCE

Airway clearance is an integral part of the management of CF. The aims are to reduce airway obstruction and maintain optimal respiratory function by clearing mucus from the lungs. It is vital to clear the mucus from the airways to help prevent respiratory infections and lung damage. Airway clearance is required once or twice a day and may take anywhere from 15 minutes to more than 1 hour each session. 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.

The airway clearance technique used will be determined by the hospital physiotherapist.

MEDICATIONS

Antibiotics:

Antibiotics are often used as a way to fight infection in the lungs. These may be oral, nebulised or given intravenously. Contraction of specific bacteria can cause progressive airway damage and irreversible scarring on the lungs. An individual should still be able to carry out their exercise program while on antibiotics, however some modifications may be required if they are not feeling as well as normal.

Salt tablets:

People with CF are required to take salt tablets daily as they lose more salt in their sweat than healthy people. This can quickly lead to dehydration if not replenished. Consideration of this is very important during exercise, as it is vital they drink enough water and exercise in appropriate conditions (not too hot) to avoid dehydration.

HORDINA CONTRACTOR

DICES

STARCHES AND FATS

SUGAR AND STARCH FREE

Enzymes:

Enzyme capsules are taken every time carbohydrates, fats and proteins are eaten to aid in digestion. Enzyme dosage depends on how much fat is in the food and the strength of capsule being taken. Enzymes capsules are made from the pancreas of a pig and are not harmful to others if accidently taken.

NUTRITION

Due to mal-absorption 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 as a last option for end-stage lung disease. A lung transplant removes the CF lungs however it does not prevent or improve any problems that CF may be causing in other parts of the body, such as the pancreas and sweat glands.

Following transplant, individuals will be enrolled in the hospital's physiotherapy exercise program 2 to 3 times a week until they achieve a good level of fitness. They will then be provided with a program to follow at home and will be advised by the hospital physiotherapist when they are fit to return to their sport or exercise routine.

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 pulmonary 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.

Exercise can also:

- Improve daily functioning
- Slow the rate of decline in lung function
- Assist with airway clearance by helping to clear mucus from the lungs
- Improve the ability to perform activities of daily living (ADLs) such as cleaning and shopping etc.
- Preserve muscle strength and function
- Increase body mass
- Improve appetite
- Improve exercise tolerance

Increase bone density

Improve posture

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.

AEROBIC EXERCISE

Aerobic training is particularly beneficial as the movements involved, such as vibrations in running, often help to clear secretions from the airways. Aerobic training should always be used in conjunction with airway clearance. Regular training helps make everyday life easier, by improving overall fitness and endurance for carrying out day to day tasks.

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, if not more beneficial, to people with lung disease.

During aerobic exercise, your client may experience coughing, wheezing or breathlessness. This is normal and the coughing should not be suppressed as it is

important for clearing the airways. They may need to spit the secretions out or they may just swallow it. Being understanding of this may help minimise any feelings of embarrassment or discomfort for the client.

Individuals will often complete airway clearance at home following their workout.

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. For muscle hypertrophy to occur, increased protein intake is required as CF patients have higher protein requirements, however it is best for their hospital dietician to advise any changes in diet.

Many people with CF have low bone mineral density (BMD), predisposing them to fractures. 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. Due to coughing, people with CF often experience postural changes and can sometimes appear slightly hunched. 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 shopping and cleaning, easier.

CFfit

FLEXIBILITY

The muscles in the trunk are needed for both posture and breathing, therefore when the body has to use these muscles excessively for breathing and coughing (often in a bent over position), posture will start to be adversely affected. Increased pressure in the chest, due to lung disease as well as excessive coughing can push outwards on the skeleton, leading to curvature of the spine, "barrel" shaping of the chest and rotation of the shoulders forward, giving a hunched and rounded shoulder appearance. 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.



Exercise considerations for people with CF

HAEMOPTYSIS

As previously discussed, exercise may cause people with CF to cough up mucus, which they may either spit out or swallow. Sometimes there may be blood in the mucus which is known as haemoptysis. This is 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.

EXACERBATIONS

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

 Increased cough 	Fever
 Increased sputum production 	 Shortness of breath (dyspnea)
 Change in the colour of sputum 	 Described sensation of tightness in the chest
 Wheezing 	 Decreased exercise tolerance

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.



HYDRATION

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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.

Symptoms of dehydration are:

 Headache 	 Dizziness
 Fatigue 	 Vomiting
 Dry mouth 	 Excessive thirst

Flushed skin

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.

INCONTINENCE

Due to coughing associated with CF, individuals often have weakened pelvic floor muscles leading to incontinence.

This may be exacerbated during coughing and high impact exercises such as jumping, skipping and running. This is even common in young people with CF. Be aware that your client may need more frequent toilet breaks or may feel uncomfortable performing such exercises.

WEIGHT AND FOOD

Individuals should be speaking with their hospital 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. 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 dietician when starting a new exercise program.

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. PEGs are used in CF patients when they

are dealing with chronic malnutrition. Individualised feeding regimes are developed to ensure nutritional needs are met. Feeding occurs at home, usually during the night.

As seen in the image, PEGs are quite discrete 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 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 swim.

- Do not play contact sports.
- Do not do repetitive motions such as vacuuming, raking or golfing.





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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 antibiotics, the port will be needled. It may be like this for up to 3 weeks until treatment is finished. During this time they should avoid swimming and should use lighter weights for upper body exercises.

CYSTIC FIBROSIS RELATED DIABETES (CFRD)

Diabetes is a very common complication that develops over time in many people with CF. In fact, many adults living with CF have some degree of diabetes or glucose intolerance. 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.

CFRD is usually treated with a combination of insulin, exercise and diet. The diet for people with CFRD is different than the calorie-restricted diet typically prescribed for people with other types of diabetes. Despite their diabetes, people with CF must maintain high-calorie, high-fat diets and compensate by adjusting insulin doses. 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 glucose snack when exercising in case of a drop in blood glucose levels.

ASTHMA

Some people with CF also suffer from asthma. This occurs when the airways in the lungs contract and narrow. Bronchodilators and steroids used for people who have asthma also work well in people with CF who are experiencing wheezing.

INFECTION CONTROL AND CROSS INFECTION

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.

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.

Useful Contact Details

If you have any questions about any information contained in this booklet please contact your health care team or Cystic Fibrosis WA.

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