22. Cystic fibrosis

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Summary

The clinical picture of cystic fibrosis (CF) can vary considerably. The most common symptoms are malnutrition and rapidly progressive obstructive lung disease, which for most CF sufferers can entail respiratory insufficiency, secondary heart disease (corpulmonale) and the need for lung transplantation. At present, treatment is only symptomatic and is aimed at slowing the rapid advancement of the disease. With optimal treatment, individuals with CF can live well into their adult years. Half of all patients with CF in Sweden and Norway are now older than 18 years, meaning that CF is no longer only a childhood disease. Since the beginning of the 1980s, fitness, strength and flexibility training have become an important part of the basic treatment. The choice of exercises, intensity, duration and frequency must be adapted to the individual's requirements, conditions and current situation. For most patients, it is the continuing deterioration of lung function that gradually becomes the limiting factor for physical capacity, but all patients with CF can perform some kind of physical activity and training.

Definition

Cystic fibrosis (CF) is the most common hereditary (autosomal recessive) and potentially deadly disease in the white population (1–3), but occurs in all races. A recent estimate of the incidence in Sweden was approximately 1/5600 infants (4) and the incidence in Norway is estimated to be about the same. Cystic fibrosis is a disease that attacks several organs in the body and is due to disorders in salt transport across cell membranes. CF affects the body's exocrine glands (mucous and pancreatic glands), affecting the transport of sodium

and chloride through the cell membrane, which in turn leads to very thick, sticky mucus (1–3). Disorders in the transport system of salt also affect the patient's sweat, which contains high levels of salt (3). The diagnosis is made on the basis of clinical symptoms with the aid of a sweat test (1) and can now often be confirmed with gene analysis.

Symptoms

Symptoms present primarily in the lungs and gastrointestinal tract but may also occur in other parts of the body (5). The changed environment in the airways of the lungs leads to the mucociliary clearance system not working properly. Peripherally, that is, behind the "mucus plugs", this creates a deoxygenated environment, which serves as a breeding ground for the bacteria chronically found in colonised CF lungs. Studies have shown that seemingly asymptomatic infants have signs of infection and inflammation already at 4–6 weeks of age (6). Most of these children become chronic carriers of one or more types of bacteria found in our environment that do not affect healthy individuals. Stagnated secretions, inflammation and chronic bacterial infections of the pulmonary airways are the most common symptoms (1, 3).

Without treatment, the disease leads to malnutrition, chronic obstructive bronchitis, repeated cases of pneumonia and destruction of the lung tissue in the form of bronchiectasis, fibrosis and emphysema (1). This leads to escalating impairment of lung function, which in time can lead to respiratory insufficiency and cor pulmonale. At this point, lung transplantation is the only possible treatment option. The chronic obstruction can be caused by a number of different factors such as bronchial spasms, swelling of the mucous membrane, a collection of mucus and instability of the airways. In some patients there may also be an element of bronchial hyperresponsiveness or an asthmatic component (3). The risk of losing fitness, mobility and muscle strength increases as lung function deteriorates. Some patients also suffer from chronic infections and sinusitis. Spontaneous rib fractures can occur secondary to frequent coughing, as can problems with incontinence, especially in women, even in younger years. Herniation of the abdominal muscle wall or the groin can also occur. The obstructive respiratory pattern and pulmonary hyperinflation can lead to a stiff thorax, straining of the muscles used for inspiration and coughing, and rupturing of the intercostal muscles. Spontaneous pneumothorax can occur, as can haemoptysis, ranging from small harmless streaks of blood in the sputum to severe bleedings that require acute treatment.

In the gastrointestinal tract, the viscous secretion of the pancreas inhibits normal secretion of digestive enzymes, resulting in malabsorption of fat and fat-soluble vitamins (3), which also leads to vitamin- and mineral deficiencies. Left untreated, malnutrition in the childhood years leads to stunted growth and in adults to increasing weight loss. An obstructive respiratory pattern and increased respiratory exertion, chronically activated immune defenses and constant inflammation of the mucous membrane of the airways causes great expenditure of energy (7–9). The increased consumption of energy combined with malnutrition leads to increasing muscle atrophy (10). Osteopenia (diminished bone density) occurs as early as the late teens, with some individuals also developing osteoporosis (11). With age, CF-related diabetes may develop (3).

The clinical picture varies considerably. The disease is progressive in nature and treatment is symptomatic but primarily preventive. The rate of progression is also individual and varies between different periods of life in the same individual.

Treatment and its goals

There is at present no treatment that will cure CF, but symptomatic treatment is being developed continually (2). The goal of treatment is to prevent destruction of the lung tissue and to slow the disease's rate of progression by controlling symptoms and maintaining good physical function of the patient (12). Treatment includes both short- and long-term goals and involves active daily intervention. Achieving good compliance with treatment requires active support and ongoing education of patients and their families.

The physiotherapist must be able to define immediate and long-range problems and needs, and be able to present these in a positive manner. In order to maintain lung function and physical capacity in the long term, a practical and motivated treatment therapy must be the goal for every individual. To achieve good compliance, the agreed-upon treatment must be followed up, reviewed and evaluated frequently. The patient and physiotherapist always arrive at such agreements together, with both parties equal participants and willing to compromise. This is an important requirement to be able to achieve a high level of compliance with daily treatment (13–16).

The basic treatment aims at the following:

- **Nutritional status**
  The impaired ability to absorb nutrients (malabsorption) is treated by adding digestive enzymes, energy-rich food, vitamins and minerals. Active supervision of nutritional status is crucial, as are different types of nutritional supplements where needed (12).

- **Lung function**
  Inhalation of bronchodilators, mucolytic and anti-inflammatory drugs are often part of the treatment. Treatment to mobilise and clear the mucus from the airways helps to prevent stagnation of secreted mucus and mucus plugs, to keep all airways ventilated. The bacteria of chronically colonised airways cannot be eliminated, but the numbers can be minimised and the chronic inflammation caused by the infection held to a minimum. The bacteria growth is controlled in part by mucus mobilising treatment/physical exercise and in part with antibiotics. CF treatment incorporates a generous amount of antibiotics, given in tablet form, intravenously or via inhalation, as decided by using subjective and objective parameters (12).

The mucus mobilising portion of the treatment is very time-consuming. There are many different techniques today to loosen, transport and evacuate the viscous sputum from the airways (17). It is important to find a technique or combination of techniques that suits the particular individual. It is also important for people with CF to learn to control their cough, both to avoid urinary incontinence as well as for social purposes. In order to achieve optimal effect, the inhalation and mucus evacuation treatment for each individual should be planned strategically. The goal is for the treatment to be as gentle and effective as possible, from both a short- and long-term standpoint, in addition to encouraging the independence of the patient (13).

- **Fitness, mobility and strength**
  Physical training is carried out to maintain good functional status and counteract loss of fitness, poor posture, and to reduce the risk of a stiff chest (12, 13, 17). How the training is carried out varies according to the individual's age, symptoms, personality and interests.

Treatment outcomes and prognosis

Treatment concentrated to CF centres has shown good (2, 4, 12). Breathing exercises and physical training are considered the cornerstones of the treatment, along with medical treatment and nutritional supplements (5, 12, 17–23). Treatment outcomes have improved markedly in recent decades (2, 4). In Sweden, there are currently some 535 people between the ages of 0–65 years living with CF, half of whom are over 18 years. The corresponding figure for Norway is 260 people, where similarly more than half are over the age of 18 years. Recent estimates regarding the prognosis for children with CF born in 1991 or later is that 95 per cent will live to be more than 25 years old (4). Thus, CF is no longer only a childhood disease, but also a concern for adult medicine. With adequate treatment and good support, most people with CF can live a fulfilling life of a good quality well into their adult years. Many manage to maintain a good functional capacity and lung function. Despite poor lung function, others still have a good physical capacity. A survey study from 1998 showed that 75 per cent of adult CF patients who had finished school were working, and 39 (26 women and 13 men) had children (4).

Effects of physical activity

The objective of physical training for individuals with CF is to:

- Stimulate the respiratory apparatus and intervene with resting respiratory patterns to increase the ventilation volume and/or distribution of the ventilation, and to stimulate mucociliary clearance and mobilise the mucus.
- Maintain normal working capacity. A high level of fitness reduces the risk of worsening in connection with exacerbations (deterioration), and makes recovery easier. Despite poor lung function, fitness may be good.

- Maintain good mobility, primarily of the thorax (24). Mobility of the thorax, back and shoulders must be maintained in order to perform effective mucus evacuation therapy (16). Stretching tense structures is time-consuming, painful and often unpleasant – preventing stiffness is easier and much more pleasant.
- Maintain good muscle strength. Strength training for the postural muscles helps to preserve mobility and avoid thoracic kyphosis. Good posture also helps patients to maintain the image of looking like everyone else, despite their advanced lung disease.
- Avoid osteopenia and osteoporosis.
- Improve/maintain good body awareness.
- Learn to coordinate muscle contractions to avoid urinary incontinence in connection with coughing or other physical exertion.
- Learn to distinguish between acceptable shortness of breath and abnormal dyspnoea and be able to manage these conditions.
- Increase self-confidence (25).

Strength and endurance of the peripheral skeletal muscles can be impaired in patients with lung disease (10). Both oxygen transport and energy metabolism in the muscle cells are worse than in healthy individuals for many reasons, including a change in the distribution of different types of muscle cells, a low capillary density, and biomechanical changes. Possible causes are the effects of chronic inflammation, malnutrition, hypoxia (decreased concentration of oxygen in the body's tissues), hypercapnia (increased concentration of carbon dioxide in the blood), use of corticosteroids and low level of physical activity (10, 26). Strength training that focuses on peripheral skeletal muscles has, however, shown to be effective (27, 28). Improved oxidative capacity reduces the production of carbon dioxide, which in turn reduces respiratory need, dyspnoea and muscular fatigability (27).

Physical activity affects both circulation and ventilation (28). Many individuals experience a mucus-mobilising effect in connection with activity. This effect can likely be attributed to the increased ventilation, both general and regional, increased tidal volume, increased rate of air flow and a temporary elevation of functional residual capacity (FRC) during physical exertion in individuals with obstructive pulmonary disease (28). Blocked airways are thus opened, and mucus dislodged and transported to larger airways. An increase in mucociliary clearance and positive biochemical factors such as less viscous mucus also likely play a role (29). During regular breaks in the physical activity, for example, in interval or circuit training, or after an exercise session, the loosened mucus may be evacuated. The combination must be stressed, however, in order to achieve mucus evacuation (13). This method of managing the mucus-mobilising part of treatment has been shown to be equally effective (18), and in certain cases more effective than other respiratory exercises, and is associated with the following advantages:

- It is effective from a time standpoint as well, also providing fitness training, mobility training and training of muscle strength.
- Anyone can take part as long as the objectives are maintained – not only CF patients benefit from physical exercise, which can improve compliance with the treatment.

- It can easily be changed and adapted according to the severity of the disease, the individual's interests and moods, location, weather, etc.
- It is easy to "take with you" to school, work, on holidays, etc.
- It can be done on one's own and thereby gives independence.
- It is, for the most part, stimulating and fun.

A high level of fitness impacts both survival and quality of life, helps individuals with CF to function like others, and enables them to function at work and have a family (21–23, 30, 31). Patients with a well-functioning basic therapy can, however, not expect to see further improvement in lung function from the increase in physical exercise. For these individuals, unchanged lung function values in the long term are seen as a positive outcome. However, if the current "treatment package" is insufficient, improvements in lung function can be achieved when treatment is optimised. Improved work capacity thus depends on the frequency, intensity and duration of the exercise training, similarly as in healthy individuals.

Prescription

Physical activity and training is an established and important part of the daily treatment of CF today. Physical activity/training should be carried out during antibiotic treatment despite the presence of chronic infection. Physical activity/training can serve as a part in mucus-mobilising treatment to increase ventilation and loosen secretions (13, 18) and/or as a supplement to other therapies (13, 16, 31). Treatment plans are holistic and include different types of strength training, for the core muscles as well as large and small muscle groups in both the upper and lower extremities, and exercises for the pelvic floor.

Individual adaptation and dosage

Physical activity/exercise must be adapted to the individual. Factors of importance for the type and dosage relate firstly to age, nutritional and functional status, lung condition, with special regard to the degree of obstruction, amount of secretion, and presence of hyperresponsiveness or instability of the airways. Exercise training can have an impact on the acceptable intensity level and perceived dyspnoea, while these are also dependent on daily condition and personality. Finding an exercise regime that can be tolerated in the patient's current state and is perceived as positive is essential to achieve a high level of compliance (16, 31). The need for pre-medicating with inhaled bronchodilation therapy should be evaluated, as well as the warm-up before exercise sessions, whenever treatment or the requirements and conditions for treatment change. For patients who desaturate (oxygen saturation decreases) during physical exercise, the need for providing oxygen during training should be evaluated in order to maintain a saturation of more than 90 per cent in the blood. This helps to reduce ventilatory and cardiovascular demands during training. An alternative can be to control the exercise intensity to maintain oxygen saturation over

90 per cent (32). Many patients benefit from "pursed-lip" breathing to lower the respiratory level, increase the size of each breath, and thereby improve gas exchange in the lungs. Constant optimisation of the treatment in cooperation with the patient strengthens the daily routines. Close follow-up and evaluation is required to motivate the patient to comply with the treatment.

Options for using physical activity/exercise as a part of mucus-mobilising therapy

There are four main ways of using physical activity/exercise for patients with CF to mobilise mucus with a loose delineation between them (13, 14). The factors that determine the option chosen for a particular individual are mainly age, amount of mucus in the airways, lung function, possible complications, and what subsequently proves to be the most effective (15).

The choices are:

- *Alternate dislodging, moving and evacuating of mucus with physical activity/exercise*
  This option involves short intervals of physical activity/exercise to loosen the mucus and breaks between the intervals to assess the amount of secretion/expectorate the mucus. The intensity of the intervals should be tailored to the individual, with high intensity activities having proven to be effective. The breaks can include careful chest compression and manual coughing support for the very young, followed by specific coughing technique, huffing (17) and coughing.
- *Dislodge the mucus during physical/exercise and move and evacuate it afterwards*
  This option involves 30 minutes of individually tailored physical activity/exercise to loosen the mucus, followed by cycles of individually tested mucus-mobilising techniques to evacuate the mucus using specific coughing technique, huffing and coughing.
- *Dislodge, move and evacuate the mucus before physical activity/exercise*
  This option is for patients with large amounts of mucus who have a need for individually tested mucus-mobilising treatment before physical activity/exercise.
- *Dislodge, move and expectorate the mucus while conducting endurance training*
  This option involves patients with small amounts of mucus and slightly reduced lung function being able to take short breaks to assess and expectorate possible mucus. The short breaks need not necessarily affect the intensity.

Physical activity/exercise can affect mucus-mobilisation by, for example, opening blocked airways and getting air in "behind" the mucus as well as increasing the breathing movements (respiratory pump) of the thorax. This helps to loosen and transport the mucus from the small airways into the larger ones. Physical activity/exercise combined with a specific coughing technique, huffing and coughing, is then used as a mucus-mobilising treatment option. This treatment option is often the first choice for children since it can be perceived as a natural approach when it comes to treatment.

One or more test treatments should be carried out to evaluate the individual effect of the physical activity/exercise. Evaluation of the response and effect determines whether physical activity/exercise can be used as part of the mucus-mobilising treatment for that individual. The trial treatment should provide an answer regarding the level and type of physical activity/exercise that will contribute to the treatment and, based on this, needs, possibilities/limitations and dosage can be determined (13).

Patients with CF perform inhalation and mucus-mobilising therapy 1–3 times per day according to their individual needs. Seemingly symptom-free patients are generally treated once a day. Physical activity/exercise is part of the main therapy. For patients with more pronounced symptoms, additional treatment sequences on the same day can comprise inhalation combined with other mucus-mobilising techniques.

Age-related treatment plans

Physical activity for very young children, age 0 to 1 year, comprise motor stimulation according to the child's motor development and activation of motor reflexes. Positive stimulation and activation of reflexes is done in different body positions with the aim of influencing the breathing pattern, increasing the amount of inspired air, affecting the ventilation distribution, and increasing the demands on the respiratory apparatus. The flow of exhalation can be increased with careful chest compressions to loosen and transport the mucus to the central airways. The compressions must be carried out with appropriate force during exhalation with the aim of increasing the expiratory flow and enabling the child to prolong exhalation. The compressions must also follow the breathing pattern, frequency and exhalation movement. Mobilised mucus induces a coughing reflex and the force of the cough can be enhanced manually. All of these techniques require education and training as the dosage of force must be such that it does not give the opposite effect (13, 16, 33).

From the age of 1 to about 4 years, the physical activity/training comprises chasing games and other active play. These games should also include fun "exercises" for strength and mobility. Those conducting the physical activity and exercise training with the children must learn what games are suitable. At 2–3 years of age many children can begin to lengthen exhalation and hold obstructed airways open by playing "blowing" games. The children are made aware of coughing and coughing technique. "Steaming up the mirror" can be used as a starting point for later learning the huffing technique. The chest compressions can then be replaced by specific coughing technique, huffing and coughing (14). In time, most 4- and 5-year-olds will be able to control their breathing technique, huff effectively, control the strength of their cough and achieve peak expiratory flow (PEF).

At 5–10 years old, the physical activity/training can be scheduled as various gym games or as relays and obstacle courses. The training should include fun exercises for fitness, strength and mobility. Breaks in the training are used for cycles of specific coughing technique, huffing and coughing to move and evacuate the loosened mucus. Those who began physical activity early are now well-developed from a motor standpoint and win over their peers, siblings, parents, the physiotherapist and physician, which as a rule creates self-confidence and is a good investment for future treatment.

After the age of 10 years, the physical training can be planned as circuit training with various content. A combination of low and high intensity exercises is recommended, often in the form of interval training. This training includes exercises to maintain mobility and strengthen the muscles of the thorax. Breaks in the training are used for cycles of specific coughing technique, huffing and coughing to move and evacuate the loosened mucus. This type of exercise can be alternated with running with an adult. Running gradually becomes popular with some people since it is perceived as the most time-efficient and "normal". Running can be complemented with simple mobility and strength exercises. Specific coughing, huffing and coughing are done at the end when the exercising is finished.

Physical training as a complement to mucus-mobilising therapy

All individuals with CF can perform physical training of some type regardless of their symptoms. For those with normal or slightly reduced lung function, training schedules, including intensity, are the same as for healthy individuals. In order to achieve as wide an effect as possible, a combination of different types of training should be used. Both high and low intensity training should be used, see Table 1. An effective way to exercise oxidative capacity is to perform high intensity training in intervals of 30 seconds at maximal exertion and 30 seconds at rest, for 30 minutes, or perhaps 3 minutes of intensive exertion and 3 minutes of rest, for 3–5 repetitions (34–36).

All-round strength training and mobility training should also be included. A good starting point for many people is to find, early on, a type of physical training that is also socially stimulating and that can be done with friends, such as playing football, field or ice hockey, bandy, horseback riding, jogging, Nordic walking, swimming, spinning, etc. This activity can then be complemented with strength and mobility exercises. Many patients choose to go with their friends, spouse or partner to fitness, aerobics or other exercise classes that offer aerobic, strength and flexibility training. For others, in-home exercise programmes using simple aids such as an exercise ball, Bobath ball, trampoline, exercise bike, weights, Thera-bands, wall bars, etc., may be a better option. The programme is planned by the physiotherapist in cooperation with the patient/parents.

Table 1. Specialised training/physical activity for different stages of cystic fibrosis.

Status	Training
Normal lung function/strength/flexibility.	No restrictions. Regular aerobic fitness and strength training principles. Enjoyable sports activities. CF-specific mobility and strength training.
Normal or slightly reduced lung function – FEV_1* > 70% of expected value – oxygen saturation does not decrease during exertion.	As above. Close follow-up.
Moderately reduced lung function – FEV_1* 40–70% of expected value – risk for desaturation at night and during exertion – possibly dependent on supplemental O_2 during sleep	High intensity interval training with long breaks, and low intensity training. Flexibility training, above all for back, chest and shoulders. Strength training, above all for postural muscles and pelvic floor. Evaluate need for supplemental O_2 during training.
Severely reduced lung function – FEV_1* < 40% of expected value – high risk for desaturation at rest – evaluate 24-hour dependency on supplemental O_2.	High intensity interval training with shorter training intervals and longer breaks, and low intensity training. Flexibility training, above all for back, chest and shoulders. Strength training, above all for postural muscles and pelvic floor. Need for supplemental O_2 during training.
Respiratory insufficiency while awaiting lung transplantation.	Light physical exercise. Flexibility training, above all for back, chest and shoulders. Adequate strength training, above all for postural muscles and pelvic floor. Requires supplemental O_2 during training.

* FEV_1 = Forced Expiratory Volume in one second.

There are many examples of adults with CF who have been able to take part in sports at a high level. It has also been shown that patients with CF can run a marathon with normal biochemical, metabolic and endocrinological response (36, 37).

Special considerations

Pronounced dyspnoea

Patients must be trained to distinguish between acceptable shortness of breath and abnormal dyspnoea, and to manage their shortness of breath and to recognise dyspnoea that can lead to panic and anxiety early. Training intensity, equipment and aids should be adapted to the individual's level of function and ability.

Acute infection and fever

Temporarily stop physical exercise and strength training that give rise to an increased heart rate. Flexibility training can still be carried out.

Nutritional status and energy balance

In the case of malnutrition, physical activity/training contributes to further weight loss and muscle atrophy. The need for proper nutritional support combined with dosage of physical activity/training is assessed in cooperation with a dietitian/nutritional physiologist and physician, in order to build up muscle mass and muscle function (38).

Asthma or bronchial hyperresponsiveness

The need for pre-medication is assessed with a reversibility test, both at work and in connection with exertion. The test should be repeated when the symptom picture changes.

Diabetes

Patients with CF-related diabetes can experience a substantial drop in blood sugar during physical activity/training, which they must learn to manage in cooperation with the dietitian/nutritional physiologist and physician.

Over-exertion

All-round training is recommended to avoid over-exertion and to enable optimal function in day-to-day life.

Decrease in oxygen saturation of the blood

The need for supplementation is determined with the help of an oxygen saturation meter (SpO_2).
Oxygen saturation < 90 per cent, measured as SpO_2, should be avoided.
Training intensity and/or oxygen supplementation during training is determined in relation to SpO_2.

Joint problems and arthritis (joint inflammation)

The need for alternative forms of training and relief is assessed.

Reduced spleen or liver function

Avoid physical activity/training that can lead to trauma to the abdomen/back.

Salt and mineral deficiencies

Excess sweating can result in symptoms of extensive loss of fluids and salts (39). Ample fluids and salt tablets should be administered for long sessions of high-intensity physical training.

Haemoptysis

In the case of minor symptoms (streaks of blood in the sputum or small bloody expectorations), stop the training session. In the case of massive haemoptysis (large amounts of coughed up blood), seek emergency medical attention.

Pneumothorax
In the case of sudden, increased dyspnoea and chest pain, pneumothorax may be suspected. Stop the training session and seek medical attention immediately.

Functional tests

Patients usually visit the clinic every six weeks, and every visit includes contact with the physiotherapist. In Sweden, meeting with the physiotherapist always includes at least one treatment session, where evaluation of the prescribed inhalation therapy, mucus-expectoration treatment and compliance occurs. A spirometric examination and functional tests are also conducted, in which chest flexibility, muscle strength and work capacity are followed up. Many also have an out-patient visit to the physiotherapist in between. Once a year an extensive lung function test is carried out, at a clinical physiology laboratory, which includes both static and dynamic volumes, as well as maximal exercise test (12, 40, 41). The treatment is continually adjusted to the measured outcomes and compliance.

The testing programme in Norway includes spirometry at every visit to the clinic. The physiotherapist evaluates and follows up the different parts of the pulmonary physiotherapy, that is, mucus evacuation therapy and physical function, posture, work capacity and work tolerance. When necessary, the patient is referred to a specialist in manual therapy. Every or every second year, an extensive 3-day cross-disciplinary review is conducted, covering lung function exams and maximal exercise tests.

Interactions with drug therapy

Many patients use inhaled beta-2 agonists, which have a heart rate-increasing effect. This seldom has significance for the planning of physical training or its outcomes, but be known for the evaluations. Insulin has a blood glucose-lowering effect as does physical training. Consideration should be given to the balance between blood glucose-lowering effect and food intake, especially in intensive and/or extended training.

In connection with lung transplantation

CF is a chronic destructive disease whose progression cannot always be slowed despite intensive treatment. Lung transplantation may ultimately be the only remaining treatment option. In this case, physical training is of utmost importance so that the patient will be in optimal physical condition before this big operation. The training does not differ from that described earlier however (see Table 1). Even patients being treated with non-invasive ventilation should engage in physical exercise.

For the period immediately post-lung transplantation, the physical training is different than for other intensive care patients. Even patients who need extended assisted ventilation

should perform physical training. The goal is to successively regain normal physical function. The physical training can then be carried out according to the usual principles. Maximal oxygen uptake (> 30 ml/kg/min) is seldom attained, however, despite normal lung function. Many patients are limited by accumulation of lactic acid, experienced as tiredness in the legs, due to changes in muscle metabolism. A few individuals have taken part in a marathon race (37). The lungs are large organs and therefore require large doses of immune-suppressing drugs. Despite the lungs being extremely vulnerable to the environment, the immune defense against bacteria remain intact. Patients may, however,be more susceptible to occasional infections.

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