4.2.1 Physiotherapy

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1. INTRODUCTION

- Traditionally, the primary aim of physiotherapy for individuals with CF was clearance of excessive secretions from the airways. Modern physiotherapy is more than airway clearance techniques (ACT) and inhalation therapy. It involves treatments that are continually monitored and modified according to the individual's requirements in a long-term perspective.
- The proportion of CF patients who are middle-aged is increasing. Problems associated with increasing age should be recognized and addressed appropriately.

2. PHYSICAL THERAPY

- Physiotherapy is an integral part of adult CF care and airway clearance techniques (ACT) are a cornerstone. ACT aim to improve the clearance of secretions in order to:
  - reduce airway obstruction
  - reduce the risk of infection
  - maintain optimal respiratory function
  - improve exercise tolerance
- The assessment and treatment for cardiovascular fitness (exercise), musculoskeletal and postural disorders, and incontinence are further key points of physical therapy in CF **(Table 1).**

Table 1: Key points of physical therapy in CF

Physiotherapy should be performed at least once daily and more frequently with increased respiratory symptoms. Even in patients with mild chest involvement, regular daily physiotherapy helps to maintain respiratory function.
The choice of airway clearance technique (ACT) should be individualized. Currently, there are no randomized controlled trials showing superiority of one ACT over another.
Exercise benefits all patients. It should be encouraged from diagnosis so that it becomes a part of daily life.
The general principles of exercise training in CF individuals are no different from those for healthy individuals or even athletes.
Exercise should be individually tailored. According to patients' needs, an effective range of physiotherapy techniques can be used.
Annual assessment of exercise capacity and musculoskeletal status is indicated in all CF patients.
Pulmonary rehabilitation may improve respiratory symptoms, exercise tolerance and quality of life.

3. AIRWAY CLEARANCE TECHNIQUES (ACT)

- The CF-lung disease is characterized by abnormal and thick mucus production, resulting in airways obstruction (mucus plugging) and lung damage due to inflammation and infection **(see also Chapter "*Pathogenesis of CF*")**.
- **ACT are a cornerstone of CF care and should be performed throughout life.**
- The aim of ACT is to remove the sputum from the airways, maintain lung function, and improve quality of life.
- Various techniques are applied and they have in common that they
  - increase the expiratory flow rates in the airways
  - keep the airways opened
  - increase the overall ventilation
- The physiotherapist uses a wide range of assessment to determine the technique that suits the patient best. The patient should be encouraged to perform ACT independently.
- A Cochrane review has concluded that ACT have short-term beneficial effects on mucus transport in CF, however there was no evidence regarding long-term effects. To date there is little evidence regarding the long-term efficacy of ACT versus no treatment but as ACT is a well-established treatment, it is considered unlikely that such a trial can be conducted due to the ethical concerns.
- There are a number of evidence-based ACT. In order to maximize adherence and physiotherapist - patient cooperation it is recommended that
  - an appropriate **individualized physiotherapy program** is developed using sound clinical reasoning and input from the patient and his/her family.
  - factors such as age, adherence, independence, patient preferences and previous experience, cooperation, cognitive ability to understand and learn the techniques, family or social support and clinical status (i.e. lung function, sputum viscosity) should be taken into consideration.
- Common ACT techniques are presented in **Table 2.** A more detailed description of the most common ACT techniques recommended and taught by cf-physio.ch in courses for physiotherapists courses is provided in the following sections.

3.1. Autogenic Drainage (AD)

- AD developed by Jean Chevaillier in Belgium is an airway clearance technique which aims to
  - Move mucus in the airways through breathing in a controlled manner.
  - Increase the expiratory flow rate without causing compression or collapse of the airway.
- In pulmonary function tests, it has been demonstrated that, in contrast to forced expiratory maneuvers, correctly dosed expiratory movements improve the flow and volumes.
  - The expiratory flow rate is the active force that mobilizes and transports mucus.
  - The tidal volume, low-, mid- or high volume, is adjusted according to the localization of the mucus in the small, middle or more central airways.
- **The technique consists of**
  - cleaning the upper airways, nose and throat,
  - breathing in (whenever possible through the nose) while in a breath stimulating position, such as sitting upright or lying in a supine position,

Table 2: Summary of common airway clearance techniques (ACT)

Airway clearance technique	Indications / Description	Contraindication / Precaution
Coughing / Huffing	Coughing is the most basic ACT. The reflex clears mucus from the central airways with high-speed airflow. Huffing / Huff is a forced expiration with an open glottis. It involves taking a breath in and actively exhaling. It is not as forceful as cough and therefore is less tiring. Depending on the position of the sputum the appropriate inspiratory volume (at low volume for more peripheral airways to medium or high volumes for central airways) is applied. Coughing and huffing clears mucus mainly from the central airways	• Important gastroesophagal reflux may be exaggerated by forced cough
Autogenic Drainage	It aims to move mucus in the airways through breathing, in a controlled manner (i.e. increase of the expiratory flow rate without causing compression or collapse of the airway). *Active patient participation is required*	
Positive expiratory pressure (PEP) devices		
Constant PEP devices	These are devices that the patient exhales into (added resistance), using a mouthpiece or a mask → the created pressure in the lungs helps to keep the airways from closing → the air flowing through the PEP device moves the mucus into the larger airways → a huff/ cough will help to move the mucus out of the airways.	• Patient with increased work of breathing may not tolerate PEP • Intracranial pressure >20mmHg • Haemodynamic instability • Recent facial, oral, or skull surgery or trauma • Acute sinusitis

(continued)

Oscillating PEP devices	Breathing with these devices accelerates the airflow and induces vibrations of the large and small airways → through these vibrations the mucus is thinned, dislodged and moved → the person coughs or huffs. *The cycle is repeated several times. Active patient participation is required*	• Epistaxis • Oesophageal surgery • Active haemoptysis • Undrained pneumothorax • Nausea • Known or suspected tympanic membrane rupture or other middle ear pathology • Claustrophobia
Passive oscillatory devices	Passive oscillatory devices do not require the active participation of the patient. For an effective therapy, huff/cough and a regular coughing must be done during and after the treatments.	
High frequency clearance techniques (commonly referred as the 'vest')	The "vest" is a machine with an air-pulse generator and an inflatable vest that wraps completely around the chest → the created rapid bursts of air make the vest inflate and deflate against the chest wall → the applied chest vibrations help to loosen mucus. Debated effectiveness. High cost.	• Fractured ribs or unstable chest
Intrapulmonary percussive ventilation (IPV) - Percussionaire	This device delivers intrapulmonary percussive ventilation. It combines internal thoracic percussion through rapid minibursts of inhaled air and continuous therapeutic aerosol delivered through a nebulizer. Evidence of effectiveness is limited evidence. High cost. The IPV is a therapeutic concept for the acute or chronic care in patients displaying the typical symptoms of obstructive pulmonary diseases.	• Undrained pneumothorax • Fractured ribs or unstable chest • Inefficient cough (≤180L/min) • Lack of adequate, skilled supervision • Lack of patient cooperation • Cardiovascular insufficiency • Hemoptysis
Non-invasive ventilation (NIV)*	Ventilatory support provided via a mask or mouthpiece may be used – in hypercapnic respiratory failure – in nocturnal hypoventilation – as a bridge to lung transplantation – as an adjunct to airway clearance – to facilitate exercise	• Undrained pneumothorax • Facial trauma • Severe bronchial obstruction • Non-collaboration • Reduced consciousness (risk of bronchoaspiration)

***see also Chapter "*Oxygen therapy and non-invasive ventilation (NIV)*"**

- breathing in by using the diaphragm,
- a post-inspiratory hold of 3-4 seconds and
- breathing out with an increased expiratory flow.

- This cycle is repeated until the mucus reaches the central airway and can be expectorated by a cough or huff.
- As AD is not an easy technique to learn for both patient and therapist, considerable patience is required. Moreover, a physiotherapist should spend time with a physiotherapist experienced in teaching this technique.

3.2. Positive expiratory pressure (PEP) devices

- The PEP techniques consist of expiration into an added resistance by a mouthpiece or mask.
- From pulmonary function tests we know that, in obstructive patients, forced expiratory maneuvers result in a rapid decline in the flow/volume curve as a result of
  - the compressed or collapsed airways,
  - the increase of the residual volume (RV) and hyperinflation.
- By breathing out through a resistance
  - the intrabronchial pressure increases,
  - the airways remain open,
  - the length, the flow rate and the volume of the expiration is increased in comparison to forced expiratory maneuvers.
- Favorable effects have been demonstrated using pulse frequencies of approximately 15Hz (pressure 10-20 cmH_20).

3.2.1. Constant PEP devices

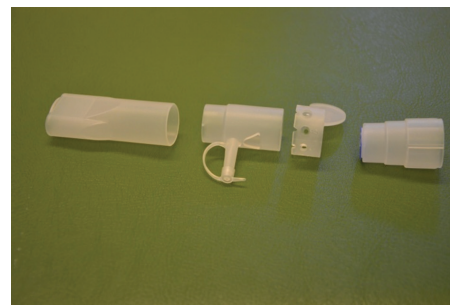
- They consist of a **mouthpiece** or a **facemask** added to a **middle piece** (optional to connect a manometer) and a **lid with holes** of 1.5 – 4.0mm diameter that functions as a resistor and can therefore be adapted to patients as needed. There is a possibility of combining some PEP devices with a nebulizer. **Figure 1** presents examples of constant PEP devices.
- By breathing out through the device → mucus is transported from the small to the large airways → it can be evacuated by coughing or huffing.

3.2.2. Oscillatory PEP devices

- The oscillating PEP combines the PEP with an oscillating airflow. The oscillation of the airflow and subsequently of the bronchial wall seems to loosen the sputum and helps to transport it.
  - The oscillation is created by the rising and falling of the expiratory pressure, which opens the exit – hole (see details below).
- Commonly used PEP devices include but are not limited to the following
  - **The Flutter valve device (VRP1®):**
    - It is the original oscillating PEP, developed in Switzerland in the late 1980s. The Flutter VRP1® is pipe-shaped, consists of a **mouthpiece** and a **steel ball**, which lies in a **plastic cone (Figure 2a)**.

Figure 1: Examples of constant PEPs



- While exhaling into the device → the expiratory pressure rises → it pushes the ball to open the exit-hole → then the pressure drops → the ball falls back (this happens many times a second throughout the exhalation) → the Flutter VRP1® oscillations (frequency range 2 - 32 Hertz) are transmitted through the airways → this helps loosen the mucus and move it to the upper airways, supported by the generated positive pressure of 10-25 cm of H_2O.
- **Technique:**
  - The patient is in a breath stimulating position such as sitting upright or lying supine.
  - Inspiration takes place through the nose, followed by a post-inspiratory hold of 3-5 seconds in order to allow the air to get behind the mucus and to promote movement of the secretions.
  - Subsequently the patient exhales into the Flutter VRP1®.
  - Followed by huffing or coughing
  - One cycle consists of 10-15 breaths and can be repeated several times, untill the patients feel relieved from the secretions.

◦ **Acapella® choice:**
- Acapella® choice is **pear-shaped** and has a **mouthpiece (Figure 2b)**.
- The oscillation is created by a magnetic mechanism: The rising pressure opens the valve → the magnetic force closes it again when pressure drops.
- It is used in the same way as Flutter VRP1®.

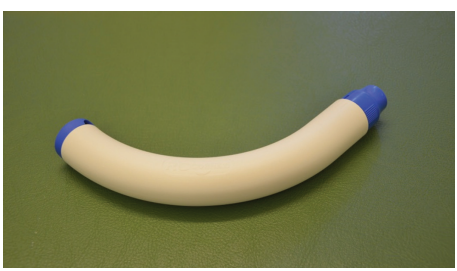
- **RC Cornet®:**
  - The cornet consists of a **mouthpiece** and a **curved plastic hose** with an elastic rubber inside **(Figure 2c).**
  - While exhalation the air has to move through the elastic rubber, which creates the PEP. The cornet is used in the same way as other oscillating PEP devices.
- Newer devices such as the Aerobika® Oscillating Positive Expiratory Pressure Therapy System need further studies to establish their efficacy.

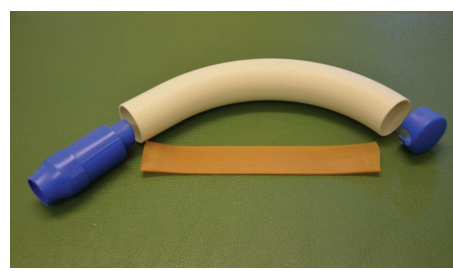
Figure 2: Examples of **Oscillatory PEP devices** a) Flutter VRP1®, b) Acapella® choice and c) RC Cornet®

a)

b)

c)





3.3. Passive oscillatory devices

3.3.1. High frequency chest wall oscillation (HFCWO) – Commonly referred as 'Vest'

- It consists of an **inflatable vest** and an **air-pulse generator**.
- The patient wears the vest on his chest → the vest gets inflated and creates a constant pressure on the chest → superimposed is an oscillation of various frequencies by the air-pulse generator throughout inspiration and expiration. The patient breathes normally and as in all other ACT the patient can use the vest by himself.
- There is an ongoing debate about the effectiveness of the vest. Latest research in a long term clinical trial comparing HFCWO versus PEP showed no difference in lung function, quality of life and patient satisfaction. Patients using the vest had a significant higher hospitalization rate than the control group using PEP.
- The HFCWO is home based, it needs an electric power source and it is by far the most expensive device.
- Its usage should be restricted to selected situations.

3.3.2. Intrapulmonary percussive ventilation (IPV) - Percussionnaire

- IPV was developed by Dr. Forrest Morton Bird in 1980s.
- It was designed to
  - promote airway clearance
  - recruit areas of lung
  - improve pulmonary gas exchange
- Its principle is to administer **bursts of small tidal volume at high frequency**. Therefore, the IPV is used to treat peripheral airway obstruction as well as ventilation disorders.
- The exact physiological effects of IPV are not yet well demonstrated and despite significant progress, the level of evidence for the clinical effectiveness of IPV remains insufficient.
- Nevertheless, IPV may offer valuable assistance in the treatment of severe respiratory diseases that are refractory to conventional techniques of respiratory therapy. It may be used in:
  - advanced neuromuscular disorders with severe bronchial mucous accumulation
  - cystic fibrosis (stable or during exacerbations)
  - chronic obstructive pulmonary disease (stable, during exacerbations or after extubation)
  - atelectasis
  - other severe respiratory complications
- **The use of IPV is limited to individuals who have received proper training in their use** and it can be applied by both non-invasive and invasive interfaces:
  - It can be self-administered by home care patients through a mouthpiece or mask. Patient operated devices for chest physiotherapy allow greater independence for the adolescent and adult CF patients.
  - In the hospital, IPV can be administered by mouthpiece, mask or endotracheal tubes, as well as in combination with an intensive care ventilator.

Figure 3: Examples of intrapulmonary percussive ventilation (IPV) for hospital, intensive care, home care, and portable systems (Used with permission from Percussionnaire®)

- IPV are expensive devices and not reimbursed by health insurances in Switzerland. Most pulmonary ligues do not possess IPV. Other providers of respiratory materiel (e.g. SOS Oxygène) may rent such devices.

3.4. Non-invasive ventilation (NIV)

- Although CF is a multisystem disease, with increased disease severity **respiratory failure** may develop. Respiratory failure remains the primary cause of death in CF.
  - With non-invasive ventilation (NIV) **(see also Chapter "*Oxygen therapy and non-invasive ventilation*")**, positive pressure ventilator assistance can be delivered in the form of inspiratory pressure support (pressure pre-set) systems. The NIV system delivers a variable volume according to a pre-set inspiratory pressure.
  - NIV may reverse or stabilise hypercapnia and hypoxaemia by improving alveolar ventilation, reducing respiratory muscle fatigue, or both.

- The aim of NIV is to reduce hypoventilation and improve gas exchange by increasing minute ventilation and reducing the work of breathing without the associated complications of endotracheal intubation.

- Clinically, NIV has also been used as an **adjunct to ACT** in people with moderate to severe disease.
  - The exact mechanisms by which NIV may assist airway clearance are unclear.
  - It is postulated that: decreased respiratory muscle fatigue and prevention of airway closure during prolonged expirations → may lead to increased effective alveolar ventilation, better compliance with airway clearance and increased sputum clearance.
  - There is some limited evidence to support the use of NIV as a clinical treatment in CF. According to recent guidelines, NIV may be a useful adjunct to other ACT in cases of
    - respiratory muscle weakness or fatigue
    - desaturation during ACT
    - difficulty clearing secretions with other ACT

- NIV use may be justified during **exercise** to decrease dyspnea, increase oxygenation and improve exercise tolerance for example in patients in whom dyspnea and fatigue contribute to reconditioning and limit effective training. However, at present, there is no objective evidence to support this.

- A Cochrane review demonstrated that NIV, when used with overnight oxygen improves gas exchange **during sleep** to a greater extent than oxygen therapy alone in patients with moderate to severe CF.
  - One clinical trial showed that in CF patients experiencing daytime hypercapnia: use of NIV over a six-week period, compared to oxygen and room air, provided benefits in terms of exercise tolerance, dyspnea and nocturnal gas exchange.

4. INHALATION THERAPY AS AN ADJUNCT TO PHYSIOTHERAPY

- In general terms, inhalation therapy is an important component of CF treatment, as it allows direct deposition of the medication in the lungs (higher local concentration, less systemic side-effects). The main determinants of the deposition pattern for nebulized medications are:
  - the breathing pattern during inhalation
  - the droplet size
  - the age / condition of the lungs

- ACT and inhalation therapy may enhance the effectiveness of each other. Therefore, physiotherapists should be adequately skilled in both therapies.
  - There is insufficient research investigating the combination of inhalation therapy and airway clearance to make recommendations regarding this practice.
  - However, the combination of inhalation therapy with ACT could be considered in those patients who do not regularly perform any other form of airway clearance, or where adherence to ACT is low.

- The physiotherapist needs to be adequately skilled concerning:
  - the mixing of inhaled medications **(see Chapter "*Medications*")**
  - the timing and order of inhaled medications **(see Table 3 and Chapter "*Medications*")**

- the **maintenance and cleaning of the devices** (mainly nebulizers) (see **Table 4**).
  - Nebuliser devices are sources of bacterial contamination and can lead to an increased risk of infection.
  - All physiotherapists should be aware of the possible contamination of inhalation therapy equipment and the implications this may have for patients' health.
  - Cleaning methods should be taught and their use should be reinforced as part of routine assessment.
  - Nebulizer equipment and inhalation devices should be cleaned after every use according to the manufacturer's guidelines **(Table 4)** or according to the recommendations of the local CF center.
  - A review of the patients' cleaning technique should be incorporated into annual inhalation therapy reviews, outpatient clinic appointments or during a hospitalization.

- **Table 3** outlines some general recommendations concerning inhalation therapy.
- **Table 4** provides examples of the most commonly used nebulizers.

Table 3: General recommendations concerning inhalation therapy

Timing and order of inhaled medications (adapted from[1])
Should be individualized but some basic principles apply:
- Usually proposed order: Bronchodilators → hypertonic saline or dornase alpha → airway clearance → other inhaled medications (e.g. inh. corticosteroids) → inhaled antibiotics
- Hypertonic saline can be given before or during airway clearance
- Dornase alpha: can be given about 1h before airway clearance (it reduces the viscosity of the secretions and aids their elimination during airway clearance). Ideally, dornase alpha should be administered at least 30 min away from inhaled antibiotics.

Cleaning of the material: Nebulizer equipment and inhalation devices should be cleaned after every use according to the manufacturer's guidelines or according to the recommendations of the local CF center.

Maintenance of the material: Nebulizer bowls should be replaced frequently according to the manufacturer's guidelines and pumps should be checked (service) at regular intervals according to the manufacturer's instructions.

Material to use:
- Where possible, nebulized medication should be taken via a **mouthpiece**.
- An **expiratory filter** should be used when nebulizing antibiotics. Where this is not possible antibiotics should be administered in a well-ventilated room with the patient alone.

Safety precautions:
- As the physico-chemical properties of nebulized drugs vary, it is important to **use a specific nebuliser/compressor combination that has been proven to be effective for that preparation**.
- Inhaled medications **should not be routinely mixed** **(see Chapter "*Medications*")**
- **All new medications should be tested** in the presence of a suitably qualified health professional and inhalation therapy regimens should be reviewed at least annually.

Table 4: Examples of commonly used inhalation devices (not an exhaustive list)

Nebulizer	Devices	Technical data / Aerosol characteristics				Information on maintenance and cleaning
		TOR or AOR *[1]	MMD*[2]	MMAD*[3]	Mass percentage < 5µm	
Pari LC Sprint (blue insert)	Pari TurboBoy® SX	TOR: 600 mg/min	3.5 µm		67%	http://www.pari.com/us-en/education-ceu/cleaning-maintenance/
Pari LC Sprint (red insert)		TOR: 450 mg/min	2.2 µm		89%	
Pari LC Plus	Pari TurboBoy® SX	TOR: 440 mg/min	3.8 µm		65%	http://www.pari.com/us-en/products/nebulizers/
Pari LL	Pari TurboBoy® SX	TOR: 470 mg/min	3.7 µm		64%	
Pari LC Sprint	AKITA® JET	AOR: 0.23 ml/min		3.8 µm		https://henrotech.be/sites/default/files/field/manual/IFU%20Akita%20JET%20ENG.pdf
RF7 plus	MicroDrop®Pro 2	AOR: 0.23 – 0.53 ml/min		2.53-2.72 µm	77.8-79.6%	http://www.mpvmedical.com/fileadmin/user_upload/Produkte/Inhalation/Inhalationsgeraete/MicroDrop_Pro/GBA_MicroDrop_Pro2_Mail.pdf
SideStream	InnoSpire® Elegance	AOR: 0.38 ml/min		3.0 µm	78%	http://www.respironicsonline.co.uk/pdf/MLUK00040%20Reusable%20Nebuliser%20Care%20Card%20v1.pdf

	Aeroneb® Go	AOR: 0.4 ml/min		2.1 μm	83%	**https://www.activeforever.com/content/manuals/aeroneb_7090_manual.pdf**
Mesh nebulizer	Pari eFlow® rapid	TOR: 610 mg/min	4.1 μm		69%	**http://www.pari.com/de-en/products/lower-airways/eflow-rapid-nebuliser-system/** **https://www.youtube.com/watch?v=7UpEelITSak**
Mesh nebulizer	Velox® *(not suitable for antibiotic inhalation)*	TOR: 497 mg/min	3.8 μm		74%	**http://www.pari.com/de-en/products/lower-airways/velox/**
Mesh nebulizer (*Aeroneb® solo*)	Aerogen® Ultra	AOR: 0.30 ml/min		3.9 μm	depends on medication	**http://www.aerogen.com/wp-content/uploads/2015/11/Aerogen-Solo-System-Instruction-Manual-EN-UK-2.pdf**

Abbreviations: TOR=total output rate, AOR=aerosol output rate, MMAD = Mass Median Aerodynamic Diameter, MMD = mass median diameter

*[1]The TOR is the amount of inhalation liquid in mg that is supplied at the mouthpiece per minute.
The AOR is the mass per minute of particles of aerosol produced by the nebulizer.

*[2]The MMD is the diameter of a particle for which half the mass of the aerosol has smaller diameter particles and half larger.

*[3]The MMAD refers to the behavior of aerosol particles in an air flow and takes into account their geometric diameter, shape and density. It is the diameter of a sphere of unit density that has the same aerodynamic properties as a particle of median mass from the aerosol (the higher MMAD of an aerosol, the more likely to deposit in the upper airway).

5. PHYSICAL ACTIVITY AND EXERCISE

- **Exercise should be considered part of the overall physiotherapy management in CF, it should be encouraged and incorporated in daily routine.**
  - Participation in regular exercise throughout life is a critical part of CF management.
  - CF patients must be educated about the importance of physical activities to increase and/or maintain adherence.
  - Annual assessment of exercise capacity is indicated in all CF patients.
- Potential benefits of physical activity and exercise in CF are presented in **Table 5.**
- Specific situations in which exercise prescription should be considered are presented in **Table 6.**

Table 5: Potential benefits of physical activity and exercise in CF

Quality of life: Most individuals find exercise enjoyable as it may improve their cardiorespiratory fitness, leading to a higher degree of perceived competence, improved self-esteem and quality of life.
Survival: Higher levels of physical fitness have been associated with better survival.
Lung function: Medical literature supports the importance of regular aerobic exercise to maintain good lung function. Recent studies demonstrated that exercise should complement conventional chest physiotherapy and that a combination of the two treatment techniques increases sputum clearance.
Posture: respiration and posture have a coupled relationship. Common postural impairments in CF include increased chest anterior–posterior diameter, shoulder elevation and protraction, trunk flexion and thoracic kyphosis. Physical activity and exercise may improve posture.
Bone mineral density: Higher level exercise capacity and physical activity are associated with higher bone mineral density in CF patients, suggesting that exercise may have an important role in maintaining bone health.

Table 6: Specific situations as indications for exercise prescription

Reported reduction in exercise tolerance/involvement in normal activities
Reduction in muscle mass/strength
Osteopenia/osteoporosis
Onset of CF-related diabetes
Patients awaiting lung transplantation

- Aerobic exercise prescription should follow the same principles as those used in healthy individuals and patients with other chronic respiratory diseases
  - Exercise at least 3 days per week
  - Duration of 30 minutes per session, consisting of shorter intervals if required

- Ideal intensity is similar to healthy persons (70-85% of maximum heart rate or 60-80% of maximal oxygen consumption).
- Exercise should be performed in a safe way and it may be necessary to be supervised or controlled by a physiotherapist.
- Exercise should be individually tailored.

- Points to consider when prescribing exercise are presented in **Tables 7 and 8.**

Table 7: Points to consider when prescribing exercise

Identify the indications and main targets of exercise
– Define realistic goals
– Take into consideration: the age, the interests and the resources of the patient
Identify contraindications for exercise
– Fever
– Unstable cardiovascular status or a cardiac event in the last month
Identify situations that necessitate precautions
– Exercise-induced desaturation: supplemental oxygen should be considered during training*
– Exercise-induced bronchoconstriction: prescribed bronchodilators should be administered prior to exercise
– Hemoptysis
– Pulmonary hypertension
– Cor Pulmonale
– Low bone mineral density: attention notably in case of resistance training
– Arthropathy or other musculoskeletal problems
Decide the appropriate 'dosage': frequency, duration, intensity of exercise
Follow guidelines for monitoring safe exercise levels

see also Chapter *"Oxygen therapy and non-invasive ventilation"

Table 8: Activities not recommended in CF

Activity	Comment
Contact sports	Discouraged if the patient has an indwelling venous access device
Scuba-diving	Discouraged due to the increased risk of pneumothorax (snorkeling is an enjoyable and acceptable alternative)
Heavy weight training	Discouraged if the patient has an implantable venous access device (the catheter may become dislodged). Risk of inguinal hernia for all patients.

6. PHYSIOTHERAPY DURING PREGNANCY, LABOUR AND THE POSTNATAL PERIOD

- **Pregnancy** often poses problems with airway clearance, particularly in later stages when the diaphragm is displaced and its movement is restricted due to the gravid uterus → decline in functional residual capacity → decrease airway caliber at the lung bases → retention of secretions.
  - It is important that chest physiotherapy is continued throughout pregnancy.
  - As pregnancy progresses, modifications or even changes in ACT may be necessary. In a severely compromised patient NIV may be required.
  - Assistance and advice should be offered on coping with breathlessness.
  - Positioning during airway clearance therapy should be done with caution:
    - An upright position seems to be the most comfortable for pregnant women.
    - A head down position should be avoided due to the risk of gastro-oesophageal reflux.
    - Left and right side lying, horizontal or with the head up might be possible (of note, lying on the right side may potentially cause compression of the inferior vena cava and this should be taken into consideration). Focus should be on a neutral lumbar spine position.
- **Labor:** As in healthy women, common symptoms such as pain, shortness of breath and low oxygen saturation may occur.
  - Pain and desaturation should be anticipated and treated appropriately.
  - A normal vaginal delivery is desirable in order to minimize post-delivery complications.
- **Postnatal period:** In the postnatal period the support for the mother is very important in order to perform appropriate inhalation therapy, airway clearance therapy, as well as post-natal exercises.

7. PHYSIOTHERAPY MANAGEMENT OF INCONTINENCE

- The relationship between CF and incontinence has been recognized since the late 1990s, and studies are still being done to understand its cause.
  - Chronic cough, paroxysms of prolonged coughing or higher pressure on the pelvic floor during ACT are possible reasons.
- The prevalence of urinary incontinence (UI), defined as involuntary leakage of urine, is higher in the CF population compared to non CF **(see also Chapter "*Urinary disease*")**.
  - Studies have shown that the problem is not very common before the age of 20, but after this age, up to 68% of women with CF have urine leakage to some degree.
  - Men do not usually experience UI as a symptom of CF.
- As patients are generally embarrassed to talk about UI and with the knowledge of the high prevalence of UI, the CF team should routinely assess this item.
- Physiotherapists working with CF should teach the patients strategies to prevent and /or resolve the problem and pelvic floor exercises:
  - As a basic therapy or lifelong habit **"the Knack" maneuver** should be taught to women with CF. It consists of a voluntary contraction of the pelvic floor prior to activities which increase the load on the pelvic floor e.g. coughing, huffing, sneezing, lifting heavy things.

- **Strength and endurance training** for the pelvic floor and lower abdominal muscles.
- **Optimal position of the lumbar spine** when performing ACT in an upright sitting position. The lumbar spine should be in a neutral position to help lower abdominal and pelvic floor muscle activity to prevent urinary leakage.

- If necessary, the patient should be referred to a physiotherapist specialized in UI, where further therapy methods including biofeedback, electrical stimulation and behavioral pattern are used.

8. PHYSIOTHERAPY FOR END-STAGE DISEASE

- The management of the severe, end-stage lung disease is an important component and challenging situation for all involved persons **(see also Chapters "Transplantation" and "*Palliative and end-of-life care*")**.
- Long before this point is reached it has to be clarified whether transplantation is an option for the patient.
- **If an organ transplant is planned, it is most likely that NIV will be necessary.** NIV reduces the work of breathing, gives the respiratory muscle time to recover, increases alveolar ventilation and improves gas exchange. It can therefore be used as a bridge to transplant.
  - The pulmonologist or the CF specialist does the installation and specifies the NIV settings.
    - Often NIV is installed first during sleep in order to correct hypoxia and hypercapnia. These alterations in gas exchange are frequently seen in moderate to severely affected CF patients. NIV results in reduced $PaCO_2$, respiratory rate and dyspnea.
    - NIV can be a useful adjunct to ACT. For this, the NIV-settings have to be adjusted. The aim is to provide as much pressure support as necessary by increasing the inspiratory pressure and to increase expiratory flow rate by reducing expiratory pressure.
  - A correct and well-fitted mask is necessary, as well as sufficiently heated and humidified air.
  - Inhalation therapies can be carried out in the usual way, if breaks from NIV are possible. Otherwise connectors for metered dose inhalers are available, as well as connectors for nebulization, when a facial mask is used.
  - A close coaching by the physiotherapist is necessary, so that the patient is able to perform forced expiratory maneuvers, cough and expectorate the sputum. A nasal mask will facilitate expectoration, a facial mask usually has to be taken off for a short period of time.
- **NIV can be used to assist severely ill patients during exercise training**:
  - Improved exercise endurance, reduced oxygen consumption, reduced dyspnea, improved oxygenation and reduced work of breathing has been observed in these patients.
- **In the situation where the patient no longer wishes life extending treatment (end-of-life care)**:
  - The physiotherapist has some possibilities to reduce dyspnea by
    - positioning the patient in a work of breathing reducing position,

- helping to relax mainly tense breathing muscles, using soft tissue techniques, massages and heat applications to support relaxation and for pain relief.
- ACT which can be performed to relieve symptoms and should be based on patient's preference.

◦ The physiotherapist can teach family and caregivers to assist with many of these techniques.

◦ Physiotherapy treatment at the end of life requires knowledge in palliative care and can be performed in the hospital or in the home setting. All treatments should be tailored to each patient's wishes and needs.

9. REHABILITATION IN CF

- Regardless of the type of chronic respiratory disease, patients experience substantial morbidity from secondary impairments, such as peripheral muscle, cardiac, nutritional, and psychosocial dysfunction, as well as suboptimal self-management strategies.
- ''Pulmonary rehabilitation is designed to improve the physical and psychological condition of people with chronic respiratory disease and to promote the long-term adherence to health-enhancing behaviors''.
  - A Cochrane review shows improvements in exercise capacity, strength, and quality of life after exercise training, with some evidence of a slower decline in lung function. However, these effects are not consistent across trials.
  - Rehabilitation programs in CF have a long-standing tradition in some European countries but until today there have been no high-quality randomized controlled trials of pulmonary rehabilitation in CF → the efficacy of systematic rehabilitation in CF is unknown and validated and structured rehabilitation programs have not yet been developed.
- Pulmonary rehabilitation may be initiated at any stage of the disease
  - during periods of clinical stability or
  - during an exacerbation or
  - directly after an exacerbation
- The goals of pulmonary rehabilitation in CF are summarized in **Table 9.**
- Local **infection control policies** may exclude the participation of people with CF in standard group-based pulmonary rehabilitation programs **(see also Chapter "*Infection control*")**.

Table 9: Goals of pulmonary rehabilitation

Minimizing burden of symptoms
Maximizing exercise performance
Promoting autonomy
Increasing participation in everyday activities
Enhancing health-related quality of life

- Home-based programs
  - Exercise at home-based programs may be necessary for some patients
  - These programs consist mainly of
    - exercises to maintain or increase the flexibility of the intervertebral and costovertebral joints,
    - stretching of the overloaded accessory respiratory muscles and
    - strengthening of the thoracic extensor muscles and scapular stabilizers in order to help and keep the normal posture.

10. CF AND AGE-RELATED COMPLICATIONS AND PHYSIOTHERAPY

- With increasing life expectancy, the possibility of developing CF and age-related complications also increases.
- Complications present a new challenge for adult CF patients and the physiotherapist should be informed of these problems on a regular basis. This should result in :
  - Adaptation of the patient-specific program.
  - Referral, if necessary, to a specialized physiotherapist (e.g. in case of musculoskeletal issues or urinary incontinence).

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