Physiotherapy for Patients with CF:
During clinic all patients who attend out-patient clinic have access to a qualified physiotherapy assessment and review of their current physiotherapy regimen. Once a year they should receive a comprehensive Annual Review, including exercise testing, for example step testing in younger children and modified shuttle walk testing or 6 minute walk test in adolescents and adults. They should have access to community therapy or open access clinic for advice and support when required. Access to specific transition clinics when age appropriate.

As a in patient access to qualified physiotherapy staff to provide treatment as necessary throughout a 24 hour period at least twice per day, everyday. If a patient is admitted after the normal working day or at the weekend the on-call physiotherapist should be notified. During an exacerbation the patients treatment sessions should be adjusted accordingly by a qualified physiotherapist and timed with the delivery of patients drugs e.g. Pulmozyme.

Assessment:
This is a continuous process, which highlights the individual problems and needs of the patient. It enables effective treatment planning and the close monitoring of these treatment plans.

The following are objective measurement tools used to monitor disease progression and treatment intervention:
- Lung function tests.
- Auscultation.
- Oximetry.
- CXR or CT scans.
- ABG’s.
- Body Mass Index (BMI).
- Sputum or cough swab culture.
- Number of exacerbations.
- Exercise tolerance.
- Quality of wellbeing measures.
- Breathlessness.

Most patients will need a combination of an airways clearance techniques and an adjunct.

Airways clearance techniques:
- The Active Cycle of Breathing Technique - ACBT:
  - This is a combination of Breathing Control (BC); Thoracic Expansion Exercises, emphasising inspiration with relaxed expiration; Forced Expiration Technique or Huff, used to mobilise and clear secretions. The ACBT is not a rigid treatment method and is modified to suit all ages and individual needs. Pryor et al 1979; Webber et al 1986; Webber 1990.

- Postural Drainage - PD:
  - This may be used in conjunction with other techniques e.g. ACBT, Positive Expiratory Pressure (PEP) and Percussion. It involves positioning the patient in different ways to enable gravity to assist in the drainage of secretions, from the peripheries of the lung to the larger more central
airways. This in turn aids with easier expectoration.

- **Percussion:**
  - This can be performed with cupped hand(s) over the area being drained, in conjunction with ACBT and PD, if the patient can tolerate this. Patients can be taught self percussion in conjunction with the above techniques, but during times of exacerbation assisted treatment by qualified physiotherapist (or family member who has been taught by a qualified physiotherapist) is preferable.

  - **Precautions to Percussion:**
    - Osteopaenia
    - Osteoporosis.
    - Uncontrolled thoracic pain.
    - Rib fractures/Flail segments.
    - Episodes of haemoptysis - see separate Peninsula guideline on haemoptysis.

  - **Percussion should not be used:**
    - Directly over incisions.
    - Directly over implantable venous access devices.
    - Those who have low bone mineral density.

- **Positive Expiratory Pressure - PEP:**
  - These devices are used to open up and recruit obstructed airways, allowing air to move behind secretions and assist in mobilising them. Breathing out against a slight resistance (10-20 cmH2O) prevents the smaller bronchial tubes from collapsing and therefore permits the continuing upward movement of secretions Tyrell et al,1986; Falk & Anderson 1991 PEP can be in the form of an Astra Mask or as a PARI PEP System which consists of a mouthpiece and manometer as shown in the picture above. In children PEP can be introduced in the form of Bubble PEP, bringing play to chest treatment. Young children who are unable participate in any adjuncts to physiotherapy e.g. PEP will have postural drainage and percussion from parents/carers as their main physiotherapy care.

- **Oscillating Positive Expiratory Pressure - Flutter:**
  - The flutter consists of a plastic pipe like device containing a stainless steel ball bearing. The patient exhales through the device causing the ball bearing to oscillate up and down causing a cyclic oscillatory PEP and vibratory effect. The flutter can have benefits bringing independence to the patient. The flutter can be tipped upwards from horizontal to increase PEP. CF Trust Guidelines 2002.

- **Oscillating Positive Expiratory Pressure - Acapella:**
  - Acapella combines the benefits of both PEP therapy and airway vibrations to mobilise secretions. Unlike the flutter the acapella is not gravity dependant, therefore can be used in a variety of positions, for example in a side lying postural drainage position. Some patients may find this easier to use. Volsko et al 2003. The resistance can be altered to find the setting that is optimal for the patient by turning the dial at the end of the
- **R C Cornet:**
  - The cornet is a curved tube that contains a plastic flexible inner tube. On expiration through the tube there is a positive expiratory pressure and an oscillation effect is felt within the airways. The Cornet can be introduced from around the age of 2 years and can also be used in a variety of positions as it is not dependant upon gravity, to work effectively. CF Trust Guidelines 2002.

- **High frequency chest wall oscillation - HFCWO/The Vest - Hill-Rom:**
  - A mechanical air pulse generator which connects via two tubes to a vest that inflates around the patients’ thorax. Increasing oscillatory frequencies are used during the course of one treatment session, ranging from 5-20Hz. CF Trust Guidelines 2002. The treatment can be combined with other adjuncts to physiotherapy for example, postural drainage, ACBT, PEP and Exercise. The device is widely used in the USA and is new to the UK.

- **Incentive spirometry:**
  - The Mediflow duo can be used as an incentive spirometer in patients who have recently had surgery e.g. Totally Implantable Venous Access Device (T.I.V.A.D) and are experiencing difficulties in taking deep breaths post-op. The patient inhales through the device aiming to keep the ball hovering for as long as possible, therefore taking a large breath in. This device can also be used as an introduction to PEP, by changing the placement of the valve. The patient breaths out through the device, aiming to keep the ball hovering for as long as possible.

- **Autogenic Drainage - AD:**
  - A series of breathing exercises devised by the Belgium physiotherapist Jean Chevallier. The aim is to dislodge and collect mucus from the lungs and then clear these secretions by breathing at various lung volumes. Chevallier, 1984; Schoni 1989. There are three phases – the unstick, collect and evacuate when breathing at low, mid and high lung volumes to mobilise, collect and expectorate secretions respectively. The technique should be taught by a person skilled in its use. This technique is not suitable for young children but may be of good use in adolescents who can co-operate fully.

- **Exercise.**
  - Regular strenuous activity is beneficial and should be encouraged in patients with CF. A wide variety of activities should be encouraged however caution should be taken with patients who have implantable venous access devices who wish to play contact sports. It has been suggested that aerobic exercise should replace chest physiotherapy in some patients Andreasson et al 1987 however more recent studies Webb et, al,1995; Baldwin et al 1994; Bilton 1992 demonstrated that exercise should compliment conventional chest physiotherapy, and that a combination of the two treatment techniques increased sputum clearance.

**References:**

1. Andreasson B, Jonson B, Kornfalt R, Normark E, Sangstrom S. long term effects
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