I Cardiorespiratory

1 Physiotherapy and the Adult with Non-Cystic Fibrosis Bronchiectasis

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INTRODUCTION

Bronchiectasis is defined as 'abnormal chronic dilatation of one or more bronchi' (Wilson 2003 **C**). The face of bronchiectasis is changing (Greenstone 2002 **C**). It used to be characterised by large volumes of purulent sputum, but today may also be characterised by a persistent and irritating non-productive cough. With the increasing use of antibiotics in the treatment of pulmonary infections in childhood, many patients with bronchiectasis have an underlying disease that predisposes them to chronic or recurrent infection, for example cystic fibrosis, immunodeficiency including HIV, primary ciliary dyskinesia, allergic bronchopulmonary aspergillosis and Mycobacterium avium complex (Rosen 2006 **C**). Diagnosis was by plain chest radiograph, with the extent of the disease assessed by bronchography (injection of contrast into the bronchial airway), but this was an invasive and unpleasant procedure. Today high-resolution computed tomography (thin slices taken through both lungs) allows identification of thickened bronchial walls, bronchial dilatation and ring opacities containing air-fluid levels (Copley et al. 2002 **C**) (see Figure 1.1).

This chapter will present two cases with diagnoses of bronchiectasis, referred for 'chest physiotherapy', one with severe bronchiectasis and one with mild bronchiectasis. Both patients had significant problems.

CASE REPORT I

Mrs AH, aged 58, presented with a chronic cough productive of copious amounts of purulent sputum and fatigue. Mrs AH's high-resolution computed tomography showed extensive bronchiectasis in both lower lobes associated with patchy consolidation and mucus plugging. The distribution was thought to be typical for a post-pertussis syndrome as the cause of her bronchiectasis. Her full lung function studies indicated severe airflow limitation with three-quarters of a litre of gas trapping and marked reduction in spirometric indices. Her gas transfer coefficient was 'reasonably' well preserved. End capillary carbon dioxide was at the upper limit of normal and there

Recent Advances in Physiotherapy. **Edited by C. Partridge**

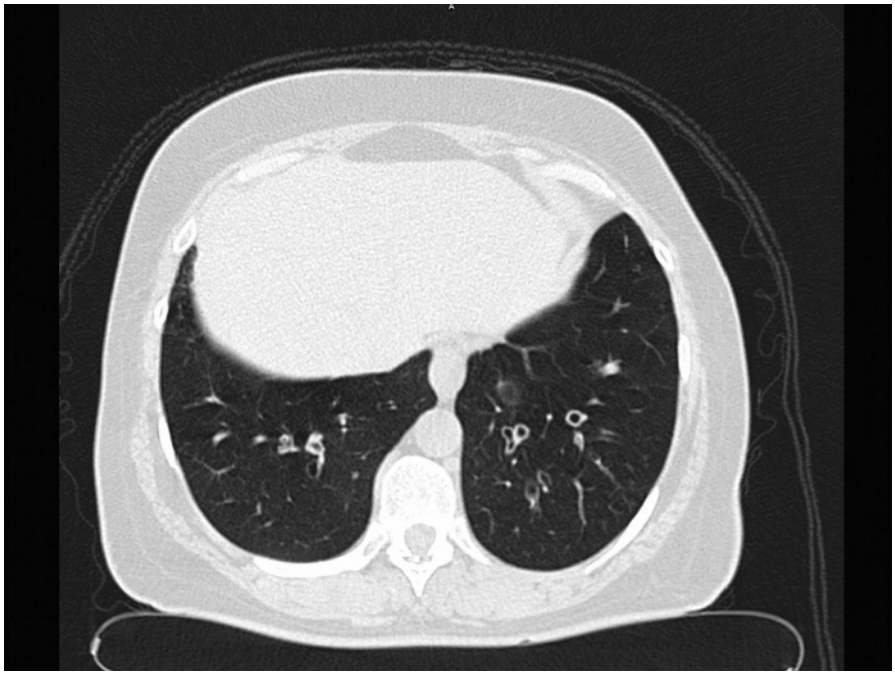

Figure 1.1. High-resolution computed tomography (CT) through the lower lobes, showing the classic signet ring sign (dilated bronchus with adjacent pulmonary artery of normal size) seen in established bronchiectasis.

was evidence of mild hypoxaemia. Haematological and biochemical indices showed mild microcytosis with no significant anaemia or abnormality in immunoglobulins. Her sputum cultured Pseudomonas aeruginosa. On auscultation there were coarse crackles throughout both lung fields.

Mrs AH's medical management included the introduction of an aggressive cyclical antibiotic regimen to reduce the bacterial load and an inhaled corticosteroid was introduced to suppress airway inflammation. She had received physiotherapy for her chest, in the form of airway clearance, in the Middle East. This had comprised the head-down tilt position with chest clapping from an assistant, and coughing when secretions reached the upper airways. The physiotherapist visited twice a week, no airway clearance was undertaken in between times and there was no encouragement to undertake a programme of physical exercise.

QUESTION 1

Which airway clearance regimen should be recommended for an adult with bronchiectasis?

A search for the evidence for airway clearance in bronchiectasis was undertaken in February 2006 using the key words 'physiotherapy' or 'physical therapy'

and 'bronchiectasis'. This revealed nothing on the Cochrane database but using 'bronchiectasis' alone, two systematic reviews of interest were identified: 'Bronchopulmonary hygiene physical therapy for chronic obstructive pulmonary disease and bronchiectasis' (Jones & Rowe 2006 **R**) and 'Physical training for bronchiectasis' (Bradley et al. 2006 **R**). Jones and Rowe identified seven trials, which were said to be small and not generally of high quality. The authors said that in most comparisons, bronchial hygiene physical therapy produced no significant effects on pulmonary function, apart from clearing sputum. They concluded that there was not enough evidence to show whether there are benefits from chest physiotherapy to remove secretions from the lungs of people with chronic obstructive pulmonary disease or bronchiectasis.

The key word 'bronchiectasis' was used in the PEDro physiotherapy evidence database and identified 16 studies, 14 in English. Ten of these studies related to airway clearance and two to exercise. This database is one of the most efficient ways for the busy clinician to access some of the evidence, but not all clinical trials of relevance are included and it is therefore important to be aware of related publications in the field which can be accessed via Medline, Embase and the Cumulative Index to Nursing and Allied Health Literature (CINAHL). A systematic review requires evidence from randomised controlled trials and few have been undertaken in cardiorespiratory physiotherapy. This does not mean the evidence from other types of trial is invalid, but rather it means that valid 'low-grade' evidence, which may be of clinical significance, will probably not have been included in any systematic review.

The reviews on airway clearance do not address the physiological benefits of the removal of excess purulent secretions from the airways. Hypothetically, airway clearance techniques can decrease mucus plugging and aid in removing secretions containing inflammatory cells and by-products, thus decreasing damage to epithelia. In addition, movement and removal of bronchial secretions containing bacteria, especially Pseudomonas, may decrease local inflammatory responses and delay the change of Pseudomonas to mucoid morphology (Lapin C (2006) Personal communication **C**). Clinical expertise would support the practice of using an airway clearance technique in people with chronic sputum production and it is important to remember the definition of evidence by Sackett et al. (1996 **C**), that is, the integration of clinical expertise and the best available evidence from systematic research.

There are several airway clearance techniques which have been shown to aid the mobilisation and clearance of excess mucus from the airways. These include postural drainage and percussion (the regimen Mrs AH had been using, with assistance, in the Middle East) (Pryor et al. 1979 **A**), the active cycle of breathing techniques (Pryor et al. 1979 **A**; Thompson & Thompson 1968 **A**), autogenic drainage (Schöni 1989, **C**), positive expiratory pressure (Falk et al. 1984 **A**), oscillating positive expiratory pressure (Cegla et al. 1997 **A**; Konstan et al. 1994 **A**), high frequency chest wall oscillation (Warwick & Hansen 1991 **A**), intrapercussive pulmonary ventilation (Newhouse et al. 1998 **A**; Varekojis et al. 2003 **A**) and resistive inspiratory manoeuvres (Chatham et al. 2004 **A**; Patterson et al. 2004 **A**). Over 27 years ago, postural drainage and percussion was shown to be less effective than the active cycle of breathing techniques (Pryor et al. 1979 **A**) and yet it is still practised in many countries.

Many of the airway clearance studies have been undertaken in people with cystic fibrosis. Extrapolation to people with non-cystic fibrosis bronchiectasis must be with caution, but it is likely that the regimens of the active cycle of breathing techniques, autogenic drainage, positive expiratory pressure, oscillating positive expiratory pressure and high frequency chest wall oscillation are equally effective (Accurso et al. 2004 **A**; Patterson et al. 2005 **A**; Pryor 2005 **A**; Thompson et al. 2002 **A**). The choice of regimen may be one of personal preference, but this is likely to be influenced by the knowledge and experience of the physiotherapist. It is also likely that adherence to treatment will be increased if the airway clearance regimen is one which appeals to the patient and if they have been involved in the selection process. What is as yet unknown is whether a change of regimen, at intervals, will increase adherence to treatment.

Many countries use the sitting position for airway clearance. A study by Cecins et al. (1999 **A)**, in people with bronchiectasis associated and not associated with cystic fibrosis, concluded that the side-lying position was as effective as the head-down tipped position and was preferred by the patients. Cystic fibrosis, in the early stages, is a disease which primarily affects the upper lobes bilaterally (Tomashefski et al. 1986 **B**). Bronchiectasis not associated with cystic fibrosis often presents with a middle and/or lower lobe distribution, indicative of a childhood viral infection. Generalised changes suggest an underlying host defence defect and an upper lobe unilateral problem, either post-tuberculosis or allergic bronchopulmonary aspergillosis (Greenstone 2002 **C**). The sitting position may be effective for people with cystic fibrosis, but this is not necessarily the best position for people with bronchiectasis not associated with cystic fibrosis and affecting the middle and/or lower lobes. In the individual patient, it is not difficult to solve this clinical problem. The patient should begin by using the selected airway clearance regimen in the sitting position. When the patient and the therapist have decided that continuing the treatment will not result in further expectoration of sputum, side lying with positioning for the affected segments should be tried. If more sputum is mobilised and cleared this will indicate there is benefit in using a side lying (lower lobes) or side lying 1/4 turn from supine (middle zones) position.

Traditionally the emphasis for the use of gravity assisted positioning has been on the drainage of secretions (Ewart 1901 **C**). Wong et al. (1977 **A**), using radionuclide imaging techniques in patients with cystic fibrosis, demonstrated that an abnormal tracheal mucus clearance approached normal when the patients were placed in a 25 degree head-down tipped position. More recent work, using inhaled radiolabelled particles, found during postural drainage in people with cystic fibrosis that mucus clearance was greater from the dependent lung than from the uppermost lung (Lannefors & Wollmer 1992 **A**). This suggests that in mucus clearance the effect of the increase in regional lung ventilation may be greater than the direct effect of gravity.

An abscess cavity is likely to drain more effectively when the opening of the cavity points downwards, but today many people with bronchiectasis have only minor dilatation of the airway walls and the movement of mucus along these bronchiectatic airways may be better facilitated by the increase in airflow in the dependent lung than

by the drainage effects of gravity in the uppermost lung, which were useful in the past. Theory would therefore indicate a patient with minimal right lower lobe bronchiectatic changes should be positioned in right side lying first, to increase ventilation, and then changed to left side lying.

Airflow is essential for airway clearance (Lapin 2002 **B**). There are similarities across most of the airway clearance regimens. All except autogenic drainage include the forced expiratory manoeuvre of huffing (Thompson & Thompson 1968 **A**), which increases expiratory flow, and this is now recognised as the most effective component of airway clearance (van der Schans 1997 **B**). Autogenic drainage utilises an unforced manoeuvre to augment expiratory flow (Schöni 1989 **B**), and the increase in expiratory flow of both the huff and an autogenic drainage breath should reduce the viscosity of mucus. This can be explained by its thixotropic property (Selsby & Jones 1990 **B**). The movement of secretions along the airways is said to be by either slug or annular flow (Lapin 2002 **B**; Selsby & Jones 1990 **B**). In addition, with the forced expiratory manoeuvre of the huff there is an oscillation of the airway walls (Freitag et al. 1989 **B**) which should further help to loosen secretions from them. Most of the regimens include a technique to increase lung volume and this is said to increase airflow via the collateral ventilatory channels (Macklem 1971 **B**), allowing air to flow behind secretions and to assist in mobilising them.

To return to Mrs AH, it was ethical to introduce an airway clearance regimen independent of an assistant to give her the opportunity to take responsibility for her management, and one which had been shown to be more effective than that of postural drainage and percussion. The two regimens not only independent of an assistant but also independent of a device are the active cycle of breathing techniques and autogenic drainage. The therapist's selection of one or other is probably influenced by their familiarity with the regimens.

For Mrs AH the active cycle of breathing techniques was chosen. The physiology behind the techniques of the active cycle of breathing was explained to Mrs AH. This included the loosening effect of the thoracic expansion exercises, utilising collateral ventilation to get the air in behind the mucus; the rest periods of breathing control; and the squeezing up of the excess bronchial secretions, from the choke points proximal to the equal pressure points, with huffing (the forced expiration technique (Pryor et al. 1979 **A**)). The techniques were practised with effect, initially in the sitting position and then in alternate side lying as the change in posture led to an increase in audible crackles from the airways. It was not long before Mrs AH developed an appreciation of how short or long a huff was required, dependent on the position of secretions within the airways, and a moderately copious amount of purulent secretions was expectorated. Mrs AH expressed her disappointment that she had not received any chest clapping and initially was not enthusiastic about continuing the regimen twice daily herself.

Self-chest clapping, in the stable clinical state, has not been shown to increase the expectoration of sputum (Webber et al. 1985 **A**). It could be argued that Mrs AH was not in a stable clinical state, but it was important to introduce a regimen which she could continue on her return to the Middle East and the introduction of

self-chest clapping was likely to increase the work involved and detract from effective huffing.

Mrs AH returned for reassessment the following week. She had conscientiously undertaken the airway clearance regimen twice a day. Her sputum had decreased in purulence and quantity and she said that she was feeling much better and had more energy. The improvement is likely to have been owing to the combination of the medical management and adherence to an effective self-airway clearance regimen.

Additional techniques which may increase airway clearance in people with bronchiectasis include the nebulisation of normal saline and hypertonic saline (Kellett et al. 2005 **A**), humidification (Conway et al. 1992 **A**) and adrenoceptor agonists (Sutton et al. 1988 **A**). These, used together with airway clearance techniques, may enhance mucus clearance. Dornase alfa has not been shown to be of benefit in non-cystic fibrosis bronchiectasis and may lead to a reduction in lung function (Wills et al. 1996 **A**). Oral mucolytics, combined with antibiotics, may help sputum production and clearance (Crockett et al. 2006 **A**).

QUESTION 2

What is the evidence for physical training in an adult with bronchiectasis?

The fatigue experienced by Mrs AH is a characteristic of chronic chest infection and is usually associated with a decrease in exercise capacity together with increasing breathlessness on exertion, leading to a vicious cycle of increasing inactivity. Bradley, Moran and Greenstone (2006 **R**), in their systematic review on physical training for bronchiectasis, identified only two reports suggesting some benefits from inspiratory muscle training on exercise capacity, quality of life and respiratory muscle function. They concluded that further research is needed to assess the benefits of other types of physical training and pulmonary rehabilitation in bronchiectasis.

Much of the research in pulmonary rehabilitation has been in people with chronic obstructive pulmonary disease but people with bronchiectasis whose quality of life has been reduced by chronic breathlessness may also benefit (British Thoracic Society Standards of Care Subcommittee 2001 **A**). Newall et al. (2005 **A**), in people with bronchiectasis, compared pulmonary rehabilitation plus sham inspiratory muscle training, pulmonary rehabilitation with targeted inspiratory muscle training, and a control group with no intervention. They concluded that exercise training (pulmonary rehabilitation) improved exercise capacity in this group of patients and that inspiratory muscle training conferred no additional benefit.

Access to a full pulmonary rehabilitation programme is not always available and the vicious cycle of increasing inactivity can be broken by the simple progressive stair climbing programme designed by McGavin et al. (1977 **A**) and modified by Webber for use on the flat (Pryor 2004 **C**; Webber 1980 **C**). As Mrs AH was to return to her own country, which was different from that in which she was receiving treatment, the McGavin programme on the stairs was selected. The programme encourages the patient to exercise to breathless, in a defined and short period of time (eight weeks), with the understanding that breathlessness in this context is uncomfortable but not harmful. In between this daily exercise, breathlessness on exertion can be lessened

by the introduction of breathing control (Rose 1999 **A**) to minimise the work of breathing. Positions which encourage the use of breathing control are said to be effective by altering the length tension status of the diaphragm, but the evidence is controversial (Gosselink et al. 1995 **A**) and it is important to assess and reassess the outcomes in the individual patient.

OUTCOME MEASUREMENTS

Outcome measurements for Case I could include: sputum volume or weight, sputum purulence (Miller 1963 **C**) (but sputum purulence is also likely to be affected by the antibiotic regimen), a field exercise test to measure exercise capacity (six-minute walking test (Butland et al. 1982 **A**) or shuttle walking test (Singh et al. 1992 **A**)) in association with a Borg scale (Borg 1982 **A**) of breathlessness and limb fatigue, and lung function.

CASE REPORT II

Mr SB, aged 30, presented with an irritating non-productive cough of 12 months, with each episode of coughing lasting for several minutes at a time, and being particularly troublesome at night on lying down. His partner had moved to a separate bedroom as she was unable to sleep with the persistent coughing. Stress, a change in air temperature and a change in posture could all precipitate bouts of coughing. Mr SB was a life-long non-smoker. There was no abnormality on his plain chest radiograph, and he had been given several courses of antibiotics and asthma management (British Thoracic Society & Scottish Intercollegiate Guidelines Network 2005), including inhaled sympathomimetic bronchodilators and inhaled corticosteroids, without effect. There was no evidence of a post-nasal drip or gastro-oesophageal reflux. He was finally referred to a specialist respiratory physician. High-resolution computed tomography revealed some changes in the right middle zone which just met the diagnostic criteria for bronchiectasis. His full lung function studies and gas transfer coefficient were all within the normal ranges. End capillary carbon dioxide was normal, and haematological and biochemical indices were normal with no immunoglobulin abnormality. His sputum culture was reported as 'No significant bacterial growth' and his chest was clear on auscultation, with normal breath sounds and no added sounds. The cause of his bronchiectasis was unknown, but may have been related to an episode of pneumonia in childhood. He was referred for physiotherapy.

QUESTION 1

Which is the evidence-based airway clearance regimen for an adult with bronchiectasis?

The literature search was as for Case I, but most of the subjects in the studies were expectorating sputum. Mr SB was not expectorating any sputum.

From previous clinical experience, the active cycle of breathing techniques was introduced with positioning for the right middle lobe. The first position was that of right side lying 1/4 turn from supine to increase ventilation to the right middle zone. Mr SB's huff was initially dry sounding and non-productive, but with the breathing exercises it became moist sounding and Mr SB said that he could feel mucus coming up into the back of his throat, which he was aware of swallowing. The exercises were continued in left side lying 1/4 turn from supine with similar results. The treatment time was about 15 minutes shared between the two positions, twice daily, and each session concluded with one or two huffs combined with breathing control in the sitting position.

Two days later, Mr SB was no longer complaining of a cough. The ongoing programme was a short daily check, in the sitting position, using the active cycle of breathing techniques. In the presence of any audible crackles on huffing, Mr SB was to progress to the side lying positions and to increase the time for treatment. He was also to follow this regimen if he thought he was getting, or if he developed, a chest infection. An alternative airway clearance regimen to that of the active cycle of breathing techniques could have been used dependent on the therapist's knowledge and expertise, and patient preference.

Using the forced expiration technique of the active cycle of breathing techniques, patients can be taught to recognise early crackles on huffing as a sign of excess mucus in the airways. The forced expiratory manoeuvre of huffing can be explained using the concept of the equal pressure point (West 1997 **B**). The equal pressure point (EPP) is the point where the pressure within the airway is equal to the pressure surrounding the airway. The airway downstream of the equal pressure point, towards the mouth, is compressed. This dynamic compression is an important mechanism which determines the efficacy of cough (Macklem 1974 **B**) and also applies to the forced expiratory manoeuvre of the huff. Proximal to the equal pressure point is the choke point (Dawson & Elliott 1977 **B**; Selsby & Jones 1990 **C**) and it is from this point, up towards the mouth, that there is a squeezing effect on the airway owing to the higher pressure outside the airway.

The positions of the equal pressure points are dependent on lung volume (West 1997 **B**). During normal tidal breathing and at a high lung volume, for example a spontaneous cough, the equal pressure points are said to be at the level of the carina or larger bronchi (Mead et al. 1967 **B**). As lung volume decreases, the equal pressure points move peripherally, allowing progressively deeper parts of the airways to be cleared. Without the need for a stethoscope, excess bronchial secretions produce audible coarse crackles during huffing. Crackles which occur with high lung volume huffing represent secretions in the larger proximal upper airways. If they occur with huffing at low lung volumes, secretions are likely to be in the smaller more peripheral airways and can be mobilised from bronchiectatic lung segments to non-bronchiectatic lung segments, where the normal mucociliary escalator should be effective in the cephalad movement of bronchial secretions.

Mr SB was not complaining of any increase in shortness of breath on exertion and was attending the gymnasium at his work place five days a week.

OUTCOME MEASUREMENTS

With computed tomography, bronchiectasis can be identified before the patient has developed a productive cough and the amount of sputum expectorated may not be an appropriate outcome measure for the effectiveness of treatment in these patients. Outcome measurements for Case II could include a visual analogue scale of cough or a valid and reliable cough-specific health-related quality of life instrument (Irwin et al. 2006 **A**).

COMMENT

The evidence and, in particular, systematic reviews alone are not yet able to answer many clinical questions in cardiorespiratory physiotherapy. The randomised controlled trial is not necessarily the best research methodology for clinical research questions in physiotherapy, but usually only research using the randomised controlled trial is considered for inclusion in systematic reviews. Recently the Cochrane Reviews have included the generic inverse variance method for meta-analysis of data from cross-over trials and data from parallel-designed trials, but even with these included the systematic review data for physiotherapy in bronchiectasis is limited.

Physiotherapy, rather than being 'evidence-based practice', should be 'practice-based evidence' (Lewis E (2004) Personal communication **C**), where the clinician generates the research questions for the researcher. This approach will lead more quickly to effective patient management and patient benefit. If the current approach to evidence-based practice, which has not itself been validated, is to continue, many physiotherapy techniques will be lost, not because they are ineffective but either because the randomised controlled trial has not been undertaken or because the right measurement tool has not been used or is not yet available. Future generations of physiotherapists must be very cautious in their interpretation of the evidence and take into consideration not only A grade evidence but also C grade evidence, of clinical experience and expertise.

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