To study physiotherapy service and techniques for people with cystic fibrosis and bronchiectasis

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Signed

Dated
ACKNOWLEDGEMENTS

I would firstly like to thank my colleagues that have supported my goals and visions, leading to being awarded with this fellowship. Thankyou to Nola Cecins, Dr Siobhain Mulrennan, and Prof. Phil Thompson, who all gave their time to help develop a project that would benefit myself, our patients, our colleagues, and the wider community.

Thankyou to the members of the CF team and Physiotherapy Department at Sir Charles Gairdner Hospital, who greatly supported my application, and to the Lung Institute of WA, who have assisted in publicising this fellowship and disseminating my findings. I must also thank the WA Department of Health for supporting my leave during the period of my fellowship.

I would like to thank the various centres, physiotherapists and other health professionals who welcomed me and openly shared their knowledge and skills, and who have remained in contact since my return. Thank you to the CF and non-CF bronchiectasis patients who allowed me to observe their physiotherapy treatment sessions.

I am grateful toward the Australian Winston Churchill Memorial Trust, as well as the WA regional and social committees for their effort and support, and their belief in this project. Without their work, people from Australia would not get this chance to experience such an amazing opportunity.

Finally I would like to thank my partner Lucinda for all of her patience and support, from the beginning of the application process to the completion of this fellowship. Your encouragement along the way has been a great help, and it has been wonderful to share this experience with you.
EXECUTIVE SUMMARY

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To study physiotherapy service and techniques for people with cystic fibrosis and non-CF bronchiectasis: UK, Sweden, and Belgium

Cystic Fibrosis (CF) is the most common inherited lethal disease affecting Caucasians, and affects approximately 1 in 3000 people in Australia. The most frequent cause of death in people with CF is respiratory failure as result of severe bronchiectasis. Non-CF bronchiectasis is a disease which affects a person’s lungs in a similar way to CF, but has many other causes. The aim of my fellowship was to study physiotherapy service provided by internationally recognised centres, and study physiotherapy techniques with world renowned physiotherapists to improve my skills and knowledge.

Conclusions
Cystic Fibrosis physiotherapy staffing levels at most of the UK and European centres visited reflects recommendations made by the Australian¹, UK⁴ and European⁵ CF Standards of Care. This allows the services to provide comprehensive, fully integrated inpatient, outpatient and community physiotherapy care. The CF physiotherapy service at SCGH meets most of the needs of the majority of its CF patients; however due to low staffing levels certain aspects of care are unable to be provided. Physiotherapy staffing for non-CF bronchiectasis was generally low at all centres visited, when compared with CF physiotherapy staffing. Preferred physiotherapy techniques varied slightly between the centres visited. Autogenic drainage was the most commonly preferred airway clearance technique, and can now be incorporated into clinical practice at SCGH.

Main Recommendations
1. Physiotherapy techniques provided to CF patients at SCGH should incorporate new techniques learned at centres in the UK and Europe, to enable a wider range of treatment options to be provided.

2. The CF physiotherapy service at SCGH should be reviewed by the Physiotherapy Department, with the potential for business case development, aiming to increase physiotherapy FTE towards benchmarks set in the Australian¹, UK⁴ and European⁵ CF Standards of Care.

3. Physiotherapy service for patients with non-CF bronchiectasis at SCGH should be reviewed by the Physiotherapy Department, with the potential for business case development to ensure the service provided meets guidelines set in the British Thoracic Society guidelines⁶, and Thoracic Society of Australia and New Zealand position statement² for non-CF bronchiectasis.

4. A post-graduate physiotherapy program, incorporating ACT commonly used in the UK and Europe, and now at SCGH, should be established through the Physiotherapy Department at SCGH to provide education and training to WA physiotherapists.
### LIST OF COMMON ABBREVIATIONS USED

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
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<tbody>
<tr>
<td>ACBT</td>
<td>Active Cycle of Breathing Technique</td>
</tr>
<tr>
<td>ACT</td>
<td>Airway Clearance Techniques</td>
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<tr>
<td>AD</td>
<td>Autogenic Drainage</td>
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<tr>
<td>B. Cepacia</td>
<td>Burkholderia Cepacia</td>
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<td>CFWA</td>
<td>Cystic Fibrosis Western Australia</td>
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<td>FET</td>
<td>Forced Expiration Technique</td>
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<tr>
<td>FTE</td>
<td>Full Time Equivalent</td>
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<td>HTS</td>
<td>Hypertonic Saline</td>
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<tr>
<td>IPPB</td>
<td>Intermittent Positive Pressure Breathing</td>
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<tr>
<td>IPV</td>
<td>Intrapulmonary Percussive Ventilation</td>
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<tr>
<td>IVAB's</td>
<td>Intravenous Antibiotics</td>
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<tr>
<td>KCH</td>
<td>Kings College Hospital</td>
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<td>LUH</td>
<td>Lund University Hospital</td>
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<tr>
<td>MRSA</td>
<td>Methicillin Resistant Staphylococcus Aureus</td>
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<td>MST</td>
<td>Modified Shuttle Test</td>
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<td>NIV</td>
<td>Non-Invasive Ventilation</td>
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<td>PAQ</td>
<td>Physical Activity Questionnaire</td>
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<td>PCD</td>
<td>Primary Ciliary Dyskinesia</td>
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<td>PEP</td>
<td>Positive Expiratory Pressure</td>
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<td>PMH</td>
<td>Princess Margaret Hospital</td>
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<td>PSA</td>
<td>Pseudomonas Aeruginosa</td>
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<td>PWC</td>
<td>Physical Working Capacity</td>
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<td>RBH</td>
<td>Royal Brompton Hospital</td>
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<td>RPH</td>
<td>Royal Perth Hospital</td>
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<td>SCGH</td>
<td>Sir Charles Gairdner Hospital</td>
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<td>UHB</td>
<td>University of Brussels Hospital</td>
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<td>UHSM</td>
<td>University Hospital of South Manchester</td>
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<tr>
<td>6MWT</td>
<td>6 Minute Walk Test</td>
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GLOSSARY OF TERMS

Acapella®
Airway clearance device which providing positive expiratory pressure, as well as oscillation of airflow aiming to loosen airway secretions

Autogenic Drainage
Airway clearance technique based on the principle of reaching high flow rates of air in different generations of bronchi, using controlled breathing

Aztreonam
Antibiotic used in the treatment of respiratory infections, which can be delivered intravenously or nebulised

Active Cycle of Breathing Technique
Airway clearance technique consisting of breathing control, thoracic expansion exercises, and the forced expiration technique

Burkholderia Cepacia
Colonising bacteria that can be found in the airways of CF patients

Colistin
Antibiotic used in the treatment of respiratory infections, which can be delivered intravenously or nebulised

Colomycin
Pharmaceutical brand of Colistin for nebulisation, used in the treatment of respiratory infections

E-Flow®
Mesh nebulisation device used to deliver medications directly into the patient’s airways. It delivers medication faster than conventional nebuliser systems

Flutter®
Airway clearance device which provides positive expiratory pressure, as well as oscillation of airflow aiming to loosen airway secretions

Gentamycin
Antibiotic used in the treatment of respiratory infections, which can be delivered intravenously or nebulised

Hi-PEP
Airway clearance technique which is a modification of PEP, combining forced expirations and positive expiratory pressure

Hypertonic Saline
Salt water solution, usually consisting of a salt concentration between 3% and 7%, nebulised to increase airway fluid levels and assist with airway clearance
GLOSSARY OF TERMS (Continued)

I-Neb®
Mesh nebulisation device used to deliver medications directly into the patient’s airways. It delivers the medication dose faster than conventional nebuliser systems and according to the patients breathing pattern. Not currently available for use with CF medications in Australia

Intrapulmonary Percussive Ventilation
Airway clearance technique combining nebulisation, positive expiratory pressure and percussive oscillatory vibrations aiming to loosen airway secretions

Intermittent Positive Pressure Breathing
A form of patient triggered, assisted inspiration produced by a ventilatory apparatus, which may also be combined with nebulised saline or hypertonic saline

Modified Shuttle Test
An externally paced, incremental test assessing maximal exercise capacity

Non-Invasive Ventilation
Assisted ventilation used to reduce respiratory muscle load and stabilise hypercapnic respiratory failure, also used as an adjunct to airway clearance techniques in cystic fibrosis

PEP (Positive Expiratory Pressure)
Airway clearance technique utilising positive expiratory pressure to increase the volume of air behind secretions which are obstructing airways, through collateral ventilation

Pari-Sinus®
Nebulisation device which delivers medication to the patient’s sinus in the treatment of sinusitis

Promixin
Pharmaceutical brand of Colistin for nebulisation, an antibiotic used in the treatment of respiratory infections

Pseudomonas Aeruginosa
Colonising bacteria that can be found in the airways of CF patients

Pulmozyme (Dornase Alfa)
Muco-active drug nebulised by CF patients aiming to thin airway secretions

Staphylococcus Aureus
Colonising bacteria that can be found in the airways of CF patients

Tobramycin
Antibiotic used in the treatment of respiratory infections. Can be delivered intravenously or nebulised

6 Minute Walk Test
A self-paced, sub-maximal exercise test
INTRODUCTION AND BACKGROUND

Cystic Fibrosis (CF) is the most common inherited lethal disease affecting Caucasians, and affects approximately 1 in 3000 people in Australia\(^1\). The most frequent cause of death in people with CF is respiratory failure as result of severe bronchiectasis. Non-CF bronchiectasis is a disease which affects a person’s lungs in a similar way to CF, but has many other causes. While the exact prevalence of non-CF bronchiectasis in Australians is not clear, it is known that it is greater than that of CF\(^2\), and even far greater in Indigenous Australians\(^3\).

Sir Charles Gairdner Hospital is the Western Australian state centre for CF, and provides a CF and non-CF bronchiectasis physiotherapy service. Physiotherapy is a major part of the daily treatment regime of people with CF and non-CF bronchiectasis, yet variations in techniques offered to patients exist between countries and centres throughout the world.

My aim was to observe the service offered by internationally recognised centres, and work with world renowned physiotherapists to improve my knowledge and skill base, and therefore be able to offer patients a wider range of physiotherapy treatment options. At UK centres I focussed on observing physiotherapy service delivery, while at European centres my focus shifted towards learning new airway clearance techniques, in particular autogenic drainage.

The contacts I have made, and professional relationships I have built while on this fellowship will allow me to continue sharing information with international colleagues, and will hopefully lead to new opportunities in areas of professional development and research for the CF physiotherapy service at SCGH.

I have incorporated much of the new skills and knowledge obtained into my clinical practice already, and will continue to strive to improve my understanding of physiotherapy for cystic fibrosis. While the benefit to me has been great, this will also greatly benefit my patients, colleagues and the wider CF community in Western Australia and Australia.
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<thead>
<tr>
<th>Date</th>
<th>Location</th>
<th>Hospital/Center</th>
<th>Team Leaders</th>
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<td>7-8 September 2010</td>
<td>London, United Kingdom</td>
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<td>London, United Kingdom</td>
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<td>Lund University Hospital</td>
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<tr>
<td>11-12 October 2010</td>
<td>Brussels, Belgium</td>
<td>University of Brussels Hospital</td>
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<tr>
<td>13-15 October 2010</td>
<td>De Haan, Belgium</td>
<td>Zeepreventorium</td>
<td>Fred Lessire (CF Physiotherapy Team Leader)</td>
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1. Overview of Cystic Fibrosis Staffing, Facilities and Service Structure

The adult CF service at RBH cares for approximately 650 adult CF patients, with a catchment area of greater London, north to Oxford, and east to Papworth.

1.1 Multidisciplinary Staffing

The CF team comprises:

- 3 Consultant Physicians
- 3 Specialist Registrars
- 2 Research Fellows
- 1 Clinical nurse consultant
- 2 Specialist Nurses
- 4.5 Physiotherapists
- 2 Dieticians

Other medical and allied health services are available via consultation.

1.2 Facilities

Cystic Fibrosis patients are admitted to the general respiratory ward, which contains 35 beds, with 28 single rooms. This includes 4 high dependency rooms and 2 negative pressure rooms. Up to 25 patients with CF can be admitted at any one time.

A separate CF day ward is used for patients commencing intravenous antibiotics (IVABs), but who do not require further inpatient admission. The ward is open Monday to Friday, and contains 6 single rooms. Cystic Fibrosis outpatient clinics are held in a dedicated respiratory outpatient department.

1.3 Outpatient Clinic Structure

Three CF outpatient clinics run weekly, with up to 25 patients attending. Each clinic is attended by 3 CF physicians, 2 registrars, a physiotherapist, dietician and 2 nursing assistants. Team members coordinate the clinic from a ‘control room’ to allow for good communication between staff.

At each outpatient clinic, the physiotherapist performs spirometry on each patient, and reviews their home treatment program. A sputum sample or cough swab is also collected from each patient by the
physiotherapist. Patients requiring test dosing of nebulised medication are referred to an outpatient respiratory medicine physiotherapist, separate to the CF physiotherapy team.

1.4 Annual Review Clinics
Annual review clinics are held on 3 days per week, with up to 4 patients attending on each day. Each patient is reviewed by all members of the CF multidisciplinary team. The physiotherapist reviews the patient’s home treatment program, including airway clearance, inhalation therapy, exercise, posture and incontinence. If appropriate, a 6 minute walk test (6MWT) or modified shuttle test (MST) is performed to assess exercise capacity and prescribe an exercise program.

1.5 Infection Control
1.5.1 Inpatient
Patients with Pseudomonas Aeruginosa (PsA) in sputum are usually admitted into single rooms on the ward. If a CF patient is admitted to a bay of 4 beds, it is ensured no other patients with CF are in the same bay. Patients with Burkholderia Cepacia (B.Cepacia) in sputum are always admitted to a single room. When exercising, CF patients must use the gym in isolation.

1.5.2 Outpatient
Cystic Fibrosis patients attend certain outpatient clinic days, depending on sputum microbiology. Patients attending the clinic are allocated a clinic room, with the CF team members rotating through the room to minimise patient crossover and cross-infection. A paging system is used to allow patients to wait in areas other than the clinic waiting area, before being allocated a clinic room.

1.6 Adolescent Transition to Adult Services at Royal Brompton Hospital
Royal Brompton Hospital accepts paediatric CF patients from its own paediatric service, as well as from Great Ormond St Hospital and Carshalton Hospital. Cystic Fibrosis adolescents first meet the adult team at the age of 14, and complete transition to the adult service between the ages of 15 and 16. Between 8 and 10 transition clinics are held per year with the paediatric services. A monthly ‘hospital school’ service is provided for newly transitioned adolescents, providing career advice and guidance.

1.7 Lung Transplantation
Patients requiring assessment for lung transplantation are referred to the transplant team at Harefield hospital, within the same hospital trust. Patients are still managed by the CF team at RBH
until transplantation, and are reviewed annually by the CF team post lung transplantation for ongoing CF issues.

2. Cystic Fibrosis Physiotherapy Staffing and Service
The CF physiotherapy team comprises 4.5 full time equivalent (FTE) physiotherapists (Team Leader, 2 senior, 2 junior) and 2 physiotherapy assistants. A respiratory research physiotherapist assists clinical staff with research projects. The team offers a comprehensive inpatient, outpatient and community service. Home visits, as well as phone and email contact, are utilised to provide further physiotherapy support. A fulltime respiratory outpatient physiotherapist, separate to the CF physiotherapy team, is available to see patients for outpatient needs including airway clearance review and nebuliser test dosing.

The CF physiotherapy team offer a weekend physiotherapy service, as well as an after hours service for appropriate patients. Physiotherapists may be called in after hours for patients with severe exacerbations requiring an extra session of airway clearance, or who are deteriorating and require urgent review. Physiotherapists familiar with the ward and CF patient group rotate through the on-call roster.

3. Physiotherapy Techniques
3.1 Airway Clearance Techniques (ACT)
The choice of ACT is individualised for each patient, with autogenic drainage (AD) and the active cycle of breathing (ACBT) being the most commonly used techniques. For inpatients, intermittent positive pressure breathing (IPPB) and non-invasive ventilation (NIV) are commonly used as adjuncts to treatment. Devices such as Acapella®, positive expiratory pressure (PEP) and Flutter® are used utilised where appropriate. Nebulised hypertonic saline (HTS) combined with PEP is used, but only as an adjunct to AD or ACBT. Daily sputum weight is measured and recorded throughout the inpatient admission.

Patients’ airway clearance regimens are usually performed at least twice daily. If required, patients are asked to complete a third session of airway clearance in the evening, independently. When at home, patients are expected to continue their airway clearance regimen at least daily, and this may be modified to accommodate lifestyle.
3.2 Inhalation Therapy
Pari nebuliser systems are supplied to patients by the hospital trust. If using Promixin, patients are provided with an I-Neb® by the pharmaceutical company.

Pulmozyme is the preferred mucolytic agent for patients with CF, with fewer patients using nebulised HTS. Patients are encouraged to take Pulmozyme at any time of the day, except in the evening before bed. Colistin is the preferred nebulised antibiotic, with fewer patients using Tobramycin. To improve tolerability, nebulised antibiotics are often combined with either sterile water or Salbutamol.

3.3 Exercise Training
3.3.1 Inpatient
CF inpatients undertake a mixed program of aerobic and resistance exercise. Exercise programs are prescribed by physiotherapists at the start of an admission, but can be supervised by physiotherapy assistants as required. Patients are encouraged to exercise at least daily while admitted to the ward. An exercise bike can be provided for use in the patients room, otherwise exercise is performed in the physiotherapy gym.

3.3.2 Outpatient
Exercise is encouraged as part of each patient’s home program, and the choice of exercise is selected according to the patient’s preference with advice from physiotherapists. Outpatient assessment of exercise capacity, usually a MST or 6MWT, is performed during annual assessment clinics.

3.4 Musculoskeletal Dysfunction
Patients with musculoskeletal problems are referred to a respiratory outpatient physiotherapist, separate to the CF physiotherapy team, for assessment and treatment.

3.5 Incontinence
Female patients are screened for incontinence and taught pelvic floor exercises at annual review clinics. If further input is required, patients are referred to a women’s health physiotherapist, separate to the CF physiotherapy team, for further assessment.
4. Physiotherapy Service for non-CF Bronchiectasis

A full time non-CF bronchiectasis physiotherapist sees all patients admitted to the respiratory ward with an exacerbation of bronchiectasis. Patients with non-CF bronchiectasis can also be referred to the respiratory outpatient physiotherapist for airway clearance education. The physiotherapist is currently trying to develop a dedicated outpatient role for the service, aiming to be part of the weekly, physician-run, outpatient non-CF bronchiectasis clinic.

The preferred ACT for non-CF bronchiectasis patients at RBH are AD, ACBT, and Acapella®. Colistin and Gentamycin are the most commonly used nebulised antibiotics for patients with PsA in sputum. Nebulised HTS is not used regularly used as an adjunct to airway clearance.
Kings College Hospital (KCH), London, UK

1. Overview of Cystic Fibrosis Staffing, Facilities and Service Structure
Kings College Hospital provides a service for approximately 150 adult CF patients, with a catchment area including Brighton, Sussex, Kent and South London below the Thames River. This hospital also provides a paediatric CF service, however each service is staffed by separate multidisciplinary teams.

1.1 Multidisciplinary Staffing
The Adult CF team comprises:
2 Consultant Physicians
2 Registrars
3 Clinical Nurse Specialists
4 Physiotherapists (plus 1 recently approved Band 7)
1 Pharmacist, Dietician, Psychologist, Social Worker

1.2 Facilities
In September 2010, patients with CF were admitted to the general respiratory ward. Currently a dedicated CF inpatient ward is under development, with 8 single rooms, to meet infection control standards for CF. Physiotherapists have access to the physiotherapy gym, to provide exercise training for CF inpatients. Outpatient clinics are held in a dedicated outpatient department. A staff 'control room', plus 5 clinic rooms are available for CF outpatient clinics.

1.3 Outpatient Clinic Structure
CF outpatient clinics are attended by a consultant physician, physiotherapist, nurse, dietician and social worker. Separate clinics are held for patients with PsA, non-PsA and B.Cepacia in their sputum, to reduce the risk of cross-infection. Diabetic and liver clinics are also available for those with associated disease.

During outpatient clinics, the physiotherapist reviews the patient’s home program, including their airway clearance regimen, inhalation therapy and exercise. A sputum sample is collected by the physiotherapist at each clinic visit.
1.4 Annual Review Clinics
Annual review clinics are held on 3 days per week, with an individual patient attending each clinic for a full day. Strict times are allocated to each team member of the allied health team for their review of the patient. Patients are only reviewed by a member of the medical team if unwell. Six weeks after the annual review, the patient will meet with a CF consultant physician for feedback.

During the annual review, the physiotherapist will review the patient’s home program, as well as musculoskeletal problems, incontinence, oxygen therapy, non-invasive ventilation, exercise, smoking status and community physiotherapy requirements. Time can be taken to modify the patient’s airway clearance technique if necessary. If appropriate, patient will perform a MST to assess exercise capacity.

1.5 Infection Control
1.5.1 Inpatient
Patients are advised not to socialise with each other at KCH. Inpatients are admitted to a single room where possible, and never share a room with another CF patient if a single room is not available. Patients with B.Cepacia in their sputum are isolated and admitted to other wards. Patients always exercise in isolation, in the physiotherapy gym or outside.

1.5.2 Outpatient
Strict infection control is maintained in the outpatient setting. Five consultation rooms are available, with the patient remaining in the same room throughout the clinic. Each team member rotates through the patient’s room, to reduce patient crossover and potential spread of infection.

1.6 Adolescent Transition to Adult Services at Kings College Hospital
Kings College Hospital accepts paediatric CF patients from its own paediatric service, as well as the paediatric CF service in Brighton. CF adolescents first meet the adult team at the age of 12 or 13, and complete transition to the adult centre between the age of 15 and 18. Joint transition clinics between the adult and paediatric services at KCH are held 4 times per year, while the adult team also conducts transition outreach clinics in Brighton every 6 weeks.

1.7 Lung Transplantation
Patients requiring assessment for lung transplantation are referred mostly to Harefield Hospital, and less frequently to Papworth Hospital. Before lung transplantation, patients are still managed by the
team at KCH; however care is transferred to Harefield Hospital after transplantation.

2. Cystic Fibrosis Physiotherapy Staffing and Service
The CF physiotherapy team comprises 4 physiotherapists (Team Leader, 2 senior, 1 junior) and 1 physiotherapy assistant. Recent approval has been granted for an extra senior physiotherapist to commence soon. The physiotherapy team provide an extensive integrated inpatient, outpatient and community service, with the ability to phone or visit patients at home.

The CF physiotherapy team provide a weekend service. Patients who are considered self reliant with airway clearance, or who are not compliant with treatment, will not always be added to the list for weekend review. Patients who require treatment after normal working hours on any day of the week will be referred to the general on-call physiotherapy service for review and treatment.

A separate physiotherapy led NIV service provides input for CF patients requiring NIV as part of their treatment program. Patients who require assessment for musculoskeletal pain or injuries are referred to the outpatient musculoskeletal department.

3. Physiotherapy Techniques
3.1 Airway Clearance Techniques
The choice of ACT is individualised for each patient, with autogenic drainage the most commonly used technique. Devices such as PEP, Acapella® and Flutter®, as well as IPPB and NIV are also utilised as adjuncts to treatment. Inpatients are encouraged to perform an airway clearance regimen twice daily, with supervision from a physiotherapist. A formal exercise session may replace one of these sessions if appropriate. Patients’ airway clearance regimens are reviewed regularly in an outpatient setting, and they are encouraged to continue with their ACT at home. Home ACT regimens may be modified to accommodate their lifestyle.

3.2 Inhalation Therapy
Where considered appropriate, E-Flow® nebuliser systems are provided by the hospital trust. If using Promixin, patients are provided with an I-Neb® by the pharmaceutical company. Patients not using either of these devices will use Pari nebuliser equipment.

Pulmozyme is the preferred muco-active drug, taken once or twice daily, and is often introduced before a decline in lung function is observed. The use of nebulised HTS has reduced in recent years,
and may be prescribed 2-3 times per week rather than daily.

Colomycin is the most commonly used nebulised antibiotic, with Tobramycin and Aztreonam used less frequently. Nebulised antibiotics are recommended to be taken after airway clearance. Colomycin is used with a Pari-Sinus® nebuliser system, for patients with severe sinusitis. Nasal irrigation with normal saline is commonly used for treating sinusitis, or after sinus surgery.

3.3 Exercise Training
3.3.1 Inpatient
Patients are prescribed individualised exercise programs, consisting of aerobic and resistance exercise. Inpatients have an exercise program prescribed by a CF physiotherapist at admission, which can then be carried out daily under the supervision of a physiotherapy assistant. Inpatients have access to a physiotherapy gym under supervision.

3.3.2 Outpatient
In an outpatient setting, usually during the annual review clinic, exercise capacity is assessed using a MST. Exercise is encouraged as part of each patient’s home program, and the choice of exercise is often selected according to patient’s preference with advice from CF physiotherapists.

3.4 Musculoskeletal Dysfunction
Patients are regularly screened for musculoskeletal problems. If further input is required, patients are referred to the musculoskeletal physiotherapy outpatient service for assessment and treatment.

3.5 Incontinence
Patients are screened for incontinence and taught pelvic floor exercises. If patients require further assessment and treatment of incontinence, they are referred to a women’s health physiotherapist.

4. Physiotherapy Service for non-CF Bronchiectasis
Respiratory physiotherapists aim to review non-CF bronchiectasis outpatients for airway clearance education, when they are reviewed by their physician. The preferred ACT for this group is AD or ACBT. As this is not a formalised physiotherapy clinic, it is difficult to capture all patients attending each day. Non-CF bronchiectasis inpatients are admitted to the general respiratory ward, where they will be reviewed by a respiratory physiotherapist.
1. Overview of Cystic Fibrosis Staffing, Facilities and Service Structure
The adult service at USHM cares for approximately 360 adult CF patients. The centre's catchment area includes the North West region of England, to Crewe, Cumbria, the Pennines and Northern Wales.

1.1 Multidisciplinary Staffing
The adult CF team comprises:
4 Consultant Physicians
4 Clinical Nurse Consultants
11 Physiotherapists
2 Pharmacists
4 Social Workers
2 Dieticians
2 Psychologists
Other medical and allied health services are available via consultation.

1.2 Facilities
New CF facilities have recently been opened which includes an inpatient ward, physiotherapy facilities and offices, an outpatient clinic and medical and nursing offices. The inpatient ward contains 22 single rooms. The CF ward has its own kitchen and chef roster, providing a specialised menu to inpatients. The physiotherapy facilities include 2 isolated exercise assessment rooms, and an isolated musculoskeletal treatment room. The outpatient clinic consists of medical offices, staff control room and 8 separate clinic assessment rooms.

1.3 Outpatient Clinic Structure
Outpatient Clinic Structure
Daily CF outpatient clinics are attended by up to 3 physicians, 3 nurses, 3 physiotherapists, 2 social workers and a dietician. Cystic Fibrosis patients are reviewed by a physician at each visit, and reviewed by members of the allied health team at least every 3 months. Physiotherapists also conduct weekly nebuliser challenge clinics and I-Neb® clinics. Diabetic clinics are held for those with cystic fibrosis related diabetes.
At each clinic visit, the physiotherapist performs spirometry, then reviews the patient’s airway clearance, inhalation therapy and exercise regimen. Airway clearance techniques can be checked if time permits. A sputum sample or cough swab is collected from each patient at every clinic visit.

During I-Neb® clinics, education is provided and adherence to the treatment regimen is checked. This is possible as the I-Neb® contains technology that records dose date, time, amount and if completed. During nebuliser challenge clinics, test dosing of nebulised medication such as hypertonic saline or antibiotics are administered to assess tolerability.

1.4 Annual Review Clinics
Annual reviews are conducted during the patient’s usual outpatient clinic appointment. More time is taken by the team members to review the patient’s treatment regimen. The patient also performs an exercise test if appropriate, usually a physical working capacity (PWC) test on a cycle ergometer. The following week, the patient's clinical status is discussed by the multidisciplinary team in the weekly annual review meeting.

1.5 Infection Control
1.5.1 Inpatient
Patients are always admitted to a single room on the ward, with 2 isolation rooms available for increased infection control. Patients with B.Cepacia in sputum are admitted to the general respiratory ward. Patients are advised not to socialise with each other, and always exercise in isolation.

1.5.2 Outpatient
Strict infection control is maintained in the outpatient setting. Several different clinics are held according to sputum microbiology, including Non-PsA, PsA, epidemic PsA, Methicillin-resistant Staphylococcus Aureus (MRSA) and B.Cepacia. Eight consultation rooms are available, with the patient remaining in the same room throughout the clinic. Each team member rotates through the patient’s room, to reduce potential cross-infection. A different spirometer, used to assess lung function, is used for each clinic according to sputum microbiology.

1.6 Adolescent Transition to Adult Services at the University Hospital of South Manchester
Currently approximately 50 adolescent CF patients transition to the adult centre at USMH each year, from various paediatric centres. Transition occurs at the age of 18, and patients are met by a
nurse and social worker from the adult CF team before attending the centre. Due to the large number of patients transitioning each year, it is difficult for physiotherapists at USMH to meet them before their first formal clinic appointment.

1.7 Lung Transplantation
Patients requiring assessment for lung transplantation are referred to either the transplant unit at UHSM or Papworth Hospital. Patients are still managed by the CF team at UHSM until they receive their lung transplantation.

2. Cystic Fibrosis Physiotherapy Staffing and Service
The CF physiotherapy team consists of 11 physiotherapists (3 Team Leaders, 5 senior, 3 junior). Recent approval has been granted for a new senior physiotherapist to commence. The physiotherapy team consists of separate inpatient, outpatient and musculoskeletal teams. The newly approved CF physiotherapy position will provide a community-based service.

The CF physiotherapy team provide an after hours service and a weekend service. Due to high numbers of inpatient admissions, weekend treatments are prioritised according to the patient's ability to perform ACT and their clinical status. A separate physiotherapy led NIV service provides input for CF patients requiring NIV as part of their treatment.

3. Physiotherapy Techniques
3.1 Airway Clearance Techniques
The choice of ACT is individualised for each patient, with the most common techniques being ACBT, AD, PEP, Acapella®, Flutter® and exercise. Patients in respiratory failure may use NIV as an adjunct to airway clearance. Often IPPB will be used with airway clearance before a patient requires NIV, to allow the patient to become familiar with positive pressure ventilation.

Inpatients are expected to complete 2 sessions of airway clearance daily, with increased supervision or assistance provided as required. Daily sputum weight is measured and recorded throughout the inpatient admission. Patients who are deemed suitable to complete independent airway clearance will perform exercise under supervision. Patients who are non-productive of sputum use exercise as the foundation of their physiotherapy treatment.
3.2 Inhalation Therapy
E-Flow® nebulisers are provided to all patients, at no cost, by the hospital trust. Patients using Promixin are provided with the I-Neb® by the pharmaceutical company. Promixin is the most commonly used nebulised antibiotic at the centre, followed by Bramitob (Tobramycin). Nebulised antibiotics are mixed with distilled water or Salbutamol to improve tolerability.

Pulmozyme is used more commonly than nebulised HTS; however the use of HTS is increasing. Pulmozyme is recommended to be taken at any time during the day that suits to the patient, and may be prescribed for use every 2nd day. The use of 3% hypertonic saline is more common than 6% or 7%, to improve tolerability.

3.3 Exercise Training
Two isolated exercise rooms are available for use by both inpatients and outpatients. Each room contains a treadmill, cycle ergometer and Nintendo Wii. Equipment to measure ventilation, heart rate and oxygen saturation is available.

3.3.1 Inpatient
Inpatients are recommended to exercise daily, and are able to use the exercise facilities under the supervision of a CF physiotherapist. More frequent exercise is recommended to patients using exercise as their main physiotherapy treatment for airway clearance. A portable NIV machine is available to assist patients in hypercapnic respiratory failure to exercise.

3.3.2 Outpatient
Outpatients are able to make appointments for an exercise review on the same day as their outpatient clinic appointments. Exercise capacity is assessed annually using the PWC test, performed on a cycle ergometer. Patients also complete a physical activity questionnaire (PAQ) annually to monitor changes in their ability to, and attitude towards exercise.

The CF service at UHSM provides financial assistance to patients joining gyms in the community, to encourage exercise as part of treatment regimens. Patients paying over £20 per month for gym membership, have the remaining value over this amount subsidised.

3.4 Musculoskeletal Dysfunction
Physiotherapists with CF and musculoskeletal experience, see both inpatients and outpatients for
ongoing assessment and treatment of patients' musculoskeletal problems. Posture, spinal and joint mobility, breathing pattern and core strength are routinely assessed and treated as required. Treatments such as joint mobilisation, acupuncture, stretching, strengthening exercises, taping and pilates are utilised.

3.5 Incontinence
Patients are referred to the musculoskeletal physiotherapists for management of incontinence issues. These physiotherapists also have experience using diagnostic ultrasound to assess and treat pelvic floor dysfunction.

4. Physiotherapy Service for non-CF Bronchiectasis
Patients with non-CF bronchiectasis are admitted to either the male or female respiratory medicine wards, and may be seen by any of the respiratory physiotherapy team. A respiratory physiotherapist at UHSM has 0.5 FTE dedicated to a weekly outpatient non-CF bronchiectasis service.

Patients are referred to this outpatient service from respiratory physicians at USHM. The ACT most commonly chosen is ACBT or AD. Nebulised HTS is commonly used as an adjunct to ACT. Nebulised Colistin and Tobramycin are the preferred antibiotics for management of patients with PsA in sputum. Airway clearance devices and nebulisation equipment is not provided to this patient group by the hospital trust; therefore patients must purchase their own airway clearance devices if recommended by the physiotherapist.

Due to a lack of available staffing, patients are reviewed once but are not provided with routine follow up appointments. Patients requiring further physiotherapy review must be re-referred by a respiratory physician at a later date.
Lund University Hospital (LUH), Lund, Sweden

1. **Overview of Cystic Fibrosis Staffing, Facilities and Service Structure**

Lund University Hospital provides care for approximately 120 adults CF patients, and 80 paediatric CF patients. The adult and paediatric services are staffed by separate multidisciplinary teams, and located in different areas of the hospital.

1.1 **Multidisciplinary Staffing**

The adult CF team comprises:

- 2 Consultant physicians
- 3 Physiotherapists
- 1 Psychologist

Other medical and allied health services are available via consultation.

1.2 **Facilities**

Patients who require IVAB’s stay in the patient hotel located on the hospital site. The hotel can also accommodate the patient’s family. Patients staying in the hotel attend the outpatient clinic for medical and physiotherapy review until they are able to return home to complete their treatment independently. The outpatient clinic contains medical clinic rooms, 2 airway clearance rooms, an exercise room, and a waiting room. Patients requiring an inpatient admission for a severe exacerbation are admitted to the general respiratory ward.

1.3 **Outpatient Clinic Structure**

Patients staying in the hotel attend the outpatient clinic twice daily for physiotherapy review, and once daily for medical review. Patients may also visit the centre for outpatient medical and physiotherapy review when they are well. Routine outpatient clinics are not held for review of large numbers of patients on particular days.

1.4 **Annual Review Clinics**

‘Annual health controls’ are held yearly to review the patient’s home program in more detail. Patients attend for a full day to have medical and physiotherapy review, as well as multiple medical tests.
1.5 Infection Control

1.5.1 Inpatient
Cystic Fibrosis inpatients are admitted to a single room on the general respiratory ward. Patients are advised not to socialise with each other to reduce potential cross-infection.

1.5.2 Outpatient
Only 1 or 2 CF patients attend the outpatient clinic at any one time, reducing the risk of cross-infection. Airway clearance is reviewed in isolated clinic rooms, and furniture and equipment are cleaned before the next patient is reviewed.

1.6 Adolescent Transition to Adult Services at Lund University Hospital
The adult centre at LUH accepts patients from the paediatric unit at the same centre. The adult CF physiotherapists all have paediatric experience, and often have previously treated the transitioning adolescent as a child. The adult and paediatric teams work closely together throughout the year to ensure an effective process.

1.7 Lung Transplantation
A lung transplantation team is also located at LUH. Referrals are accepted from any location in Sweden, and between 20 and 30 bilateral lung transplants are performed each year. Assessment for lung transplantation is performed at the centre, or in the patient’s home location via a GP in contact with the team. Before transplantation, patients continue to exercise independently at home, however may need occasional advice from a CF or transplantation physiotherapist. The transplantation team takes over care of the patient post transplantation.

2. Cystic Fibrosis Physiotherapy Staffing and Service
The adult CF physiotherapy team consists of 3 physiotherapists. The CF physiotherapists review patients staying in the hotel up to twice daily for airway clearance and inhalation therapy, exercise and joint mobility. If patients are admitted to the ward, a physiotherapist will review the patient up to twice daily. Physiotherapists have the capacity to visit patients in the community if required, but this is rare. Any CF patient receiving general anaesthetic for a procedure will be accompanied by a CF physiotherapist, who will perform assisted autogenic drainage while the patient is under sedation.
3. Physiotherapy Techniques

3.1 Airway Clearance Techniques

The foundation of airway clearance for CF patients in Sweden is exercise, combined with or followed by, autogenic drainage or the forced expiration technique. Patients are taught these techniques from a young age, and if competent, can manage airway clearance independently throughout adulthood. Patients that require increased assistance with airway clearance routines are introduced to other techniques, such as PEP, Hi-PEP, Flutter® and NIV. Side lying positioning and upper limb stretching are also incorporated into routines.

It is common for patients attending the outpatient clinic to incorporate several techniques into their routine. This may include using NIV, PEP, autogenic drainage, intermittent inhalation therapy (Salbutamol and HTS) and positioning as part of a routine to assist in airway clearance. High NIV inspiratory pressures of up to 40cmH₂O are common, without any reported complications.

3.2 Inhalation Therapy

Patients are provided with E-Flow® or Aero-Neb nebuliser systems by the hospital. If using Promixin, the I-Neb® is provided by the pharmaceutical company.

Nebulised HTS is the preferred muco-active medication used, with less patients using Pulmozyme. The use of 3% hypertonic saline, as opposed to stronger concentrations, is common to improve tolerability. Hypertonic saline is also used intermittently throughout the airway clearance regimen rather than at the beginning of airway clearance only.

Nebulised antibiotics are less commonly used at the centre, when compared to UK centres. The preferred nebulised antibiotic is Colistin, with Tobramycin less commonly used. If necessary, antibiotics are diluted with normal saline to improve tolerability.

3.3 Exercise Training

3.3.1 Inpatient

Inpatients are expected to exercise daily as part of their physiotherapy management. As inpatients generally have more severe exacerbations, choice of exercise may be more limited.

3.3.2 Outpatient

Patients staying in the patient hotel attend the outpatient clinic for supervised exercise. Patients
perform aerobic training on an exercise bike or treadmill, light resistance exercises, and various thoracic and trunk mobility exercises. Exercise capacity is assessed using a PWC test. At home patients are expected to perform an intensive daily aerobic and resistance exercise program, which can also be combined with AD or FET, to be used as the patient’s ACT.

3.4 Musculoskeletal Dysfunction
Due to a preventative approach to CF musculoskeletal dysfunction in Sweden, few complications regarding posture, osteoporosis and pain are seen. Patients are encouraged to be active, exercise regularly and maintain good posture from an early age. If a patient presents with an acute musculoskeletal problem which cannot be managed by the CF physiotherapists, they are referred to a specialist musculoskeletal physiotherapist.

3.5 Incontinence
Patients are taught pelvic floor exercises and screened for incontinence. If a patient presents with an incontinence problem that cannot be managed by the CF physiotherapists, they are referred to a specialist incontinence physiotherapist.

4. Physiotherapy Service for non-CF Bronchiectasis
There is no dedicated physiotherapy service for patients with non-CF bronchiectasis at LUH. CF physiotherapists are only funded to see patients with CF and primary ciliary dyskinesia (PCD).
1. Overview of Cystic Fibrosis Staffing, Facilities and Service Structure
The University of Brussels hospitals provides care for approximately 160 paediatric and adult CF patients.

1.1 Multidisciplinary Staffing
The CF team comprises:
4 Physicians
2 Physiotherapists
1 Social Worker
1 Psychologist
Endocrinology and Ear Nose & Throat input is available via consultation

1.2 Facilities
CF patients are admitted to the general respiratory ward at the hospital. Outpatient clinics are held in the respiratory outpatient department.

1.3 Outpatient Clinic Structure
Weekly CF outpatient clinics are held, with approximately 15 patients attending each clinic. Clinics are segregated according to sputum microbiology, and are attended by a physician, physiotherapist and social worker.

1.4 Infection Control
1.4.1 Inpatient
Inpatients are admitted to single rooms where possible, however often will be admitted into a room with 2 or 4 beds. Patients with B.Cepacia in sputum will not share a room with another patient.

1.4.2 Outpatient
Separate outpatient clinics are held for patients with different sputum microbiology.

1.5 Adolescent Transition to Adult Services at the University of Brussels Hospital
As the hospital is a paediatric and adult centre with the same CF team, no formal transition program is in place.
1.7 Lung Transplantation

CF patients are managed by the CF team at UBH until they receive transplantation. Care is then transferred to the transplantation team.

2. Cystic Fibrosis Physiotherapy Staffing and Service

The CF physiotherapy team consists of 2 physiotherapists, who see both adult and paediatric CF inpatients and outpatients, as well as other non-CF paediatric patients admitted to the respiratory ward. CF physiotherapists do not provide community assistance to CF patients, however the Belgian government funds up to 60 physiotherapy treatment sessions per year for patients.

3. Physiotherapy Techniques

3.1 Airway Clearance Techniques

The preferred ACT is AD, which is usually taught in a sitting position. In infants, assisted autogenic drainage is performed with the assistance of a physiotherapist, usually on a physiotherapy ball. The Flutter® and PEP devices are used only as an adjunct to autogenic drainage, to prevent early airway closure. Children may use bubble-PEP in the same manner. Patients perform airway clearance twice daily under the supervision of a physiotherapist. All inpatients use nasal irrigation with normal saline before airway clearance.

Outpatients are encouraged to continue AD at home at least daily, preferably twice daily. Autogenic drainage technique is corrected during inpatient admissions and at outpatient clinics, to ensure it is being performed correctly.

3.2 Inhalation Therapy

Patients are provided with E-Flow® nebuliser systems by the hospital, with a 5 year supply of aerosol heads at a time. Patients must purchase their own E-Flow® aerosol heads if they exhaust this supply early. Occasionally patients will use Pari nebuliser equipment to nebulise HTS, to improve tolerability.

All CF patients over the age of five take Pulmozyme daily, and twice daily if sputum is difficult to expectorate. Nebulised HTS is also commonly used, at a concentration of 3% or 6% according to tolerability. Nebulised antibiotics are prescribed for use where appropriate.
3.3 Exercise Training

3.3.1 Inpatient
A gymnasium containing aerobic and resistance exercise equipment is available for use by inpatients, under the supervision of a physiotherapist. If time is available for supervision, exercise is performed daily by inpatients.

3.3.2 Outpatient
Patients are encouraged to incorporate exercise into their home physiotherapy program. Exercise capacity is assessed annually at the centre using a PWC test on a cycle ergometer.

3.4 Musculoskeletal Dysfunction
Patients with musculoskeletal problems are referred to a specialist musculoskeletal physiotherapist for assessment and treatment.

3.5 Incontinence
Patients with incontinence problems are referred to an incontinence physiotherapist for assessment and treatment.
Zeepreventorium, DeHaan, Belgium

1. Overview of Cystic Fibrosis Staffing, Facilities and Service Structure

Zeepreventorium (The Preventorium) is a government funded CF, obesity, burns and chronic fatigue rehabilitation centre located on the Western coast of Belgium. Up to 9 adult and 21 paediatric CF patients can attend the centre at any one time. Patients must stay for a minimum of 4 weeks, and are accepted from Belgium, France, Germany, and occasionally other countries. CF patients listed for lung transplantation are allowed to stay at the centre until their surgery. Zeepreventorium is usually not the primary care centre for a CF patient, but rather a facility for longer admissions to improve patients' airway clearance, inhalation therapy and exercise regimens.

1.1 Multidisciplinary Staffing

The CF team comprises:
2 Consultant Physicians
2.75 Physiotherapists
1 Social Worker

Pulmonary Physiologists are available to conduct spirometry and exercise tests

1.2 Facilities

Adult and paediatric CF patients stay in separate wings of the centre. In the adult wing, patients have a shared kitchen, living and games area. The physiotherapy department contains 4 airway clearance rooms, a large shared gymnasium, and a 25 metre salt water pool. Two ovals are also available for outdoor sport.

1.3 Infection Control

Patients with B.Cepacia in sputum are not permitted to attend the centre. Patients with PsA or Staphylococcus Aureus in sputum can be admitted but are only allowed to socialise with patients with similar sputum microbiology. Physiotherapy sessions are conducted in groups, but again according to sputum microbiology.

2. Cystic Fibrosis Physiotherapy Staffing and Service

The physiotherapy team provides a service to all patients admitted to the centre, including those without CF. Physiotherapists with CF experience conduct 3 airway clearance sessions, 1 stretching and mobility session, and 1 sport session daily for CF patients. A 7 day service is offered, however
most patients take weekend leave to return home.

3. Physiotherapy Techniques
3.1 Airway Clearance Techniques
The preferred choice of ACT is autogenic drainage, developed by the previous senior physiotherapist at the centre, Jean Chevalier. Autogenic drainage is performed in sitting, supine or side lying, and combined with a Flutter® or PEP if early airway closure is observed. Patients complete three 90 minute sessions of inhalation therapy and autogenic drainage daily.

3.2 Inhalation Therapy
Patients at the centre are provided with E-Flow® nebuliser systems where appropriate, or Pari nebuliser systems, and do not bring their own nebulisation equipment. Nebulised HTS is the most commonly used muco-active medication, used intermittently during airway clearance sessions. Hypertonic saline is mixed with Salbutamol to improve tolerability as required. Patients who are prescribed Pulmozyme continue to use it at the centre, usually after the morning airway clearance session. Prescribed nebulised antibiotics are used at the end of the morning and afternoon airway clearance sessions.

3.3 Exercise Training
Patients complete 60 minute group sport sessions, as well as a 30 minute stretching and mobility session daily. Sport sessions consist of indoor swimming, soccer, basketball, and beach sports in summer. Aerobic capacity is assessed using a maximal PWC test on a cycle ergometer, or a 6MWT.

3.4 Musculoskeletal Dysfunction
Daily stretching and mobility sessions aim to improve posture and reduce musculoskeletal pain. Patients with ongoing musculoskeletal dysfunction will be managed by their primary care centre.

3.5 Incontinence
Patients with incontinence issues are managed by their primary care centre.
CONCLUSIONS

1. Cystic Fibrosis

1.1 Multidisciplinary Staffing
Multidisciplinary teams at the centres visited were generally well staffed, and closer to meeting recommended staffing levels published in Australian (2008)\(^4\), European (2005)\(^5\) and UK (2001)\(^6\) CF Standards of Care.

Allied health staffing (physiotherapy, pharmacy, diet therapy, social work, psychology) in CF at SCGH is well below the recommended levels in these guidelines, which limits aspects of service that is able to be delivered to CF patients. Comparisons of physiotherapy staffing and service will be reviewed on page 37.

1.2 Facilities
CF inpatients were admitted to general respiratory wards, with the exception being the University Hospital of South Manchester, which has a new dedicated CF ward. Most inpatient wards contain single rooms, however cannot always provide a single room for CF inpatients.

Outpatient clinics were held in general respiratory outpatient departments, with the exception being the University Hospital of South Manchester and Lund University Hospital. Most centres visited have enough clinic rooms available for patients to stay in the same room during clinic, with staff rotating through to reduce patient cross over.

At all centres visited, physiotherapists and CF patients had access to a gym or exercise rooms containing aerobic and resistance exercise equipment, and some of the centres are able to provide exercise equipment for use in the patients room.

All CF patients at SCGH are admitted to the general respiratory ward, in single rooms. Inpatients have limited access to a gymnasium with aerobic and resistance exercise equipment, however aerobic exercise equipment is provided for use in patients’ rooms. Outpatient clinics are held in the respiratory medicine outpatient department.

1.3 Outpatient Clinic Structure
A multidisciplinary team is present during outpatient clinics at all hospital centres visited. The
number of staff varies according to the number of patients attending, as well as which members of the team attend. Professions present during outpatient clinics include physicians, physiotherapists, nurses, pharmacists, dieticians and social workers. Separate outpatient clinics are held for patients with different sputum microbiology. Allied health professionals aim to see patients at least every 3 months in most centres, and often review the patient at each visit.

*Two weekly PsA outpatient clinics at SCGH are attended by a consultant physician, nurse practitioner, and a physiotherapist. During one of these clinics a dietician is present. All patients are seen by each member of the CF team at each clinic. During non-PsA clinics and Cepacia clinics, no allied health team members are present due to staffing constraints, while pharmacy and social work are currently unable to provide any CF outpatient service at SCGH.*

### 1.4 Annual Review Clinics

All hospital centres visited have implemented multidisciplinary annual review clinics, aiming at reviewing the patient’s current health status and home program in more detail. Physiotherapists are allocated enough time to review the patient’s airway clearance and inhalation technique, musculoskeletal dysfunction, incontinence and exercise tolerance. Most centres provided the patient with a follow up appointment a number of weeks after the annual review to discuss the results with a consultant physician. At UHSM, the multidisciplinary team discuss the patient in the week following the annual review.

*At SCGH, annual review clinics have been implemented over the past 2 years. Patients attend for a half day, to have routine tests as well as review by the multidisciplinary team. The physiotherapist reviews the patient’s home program and performs an exercise test if appropriate. Due to time constraints, the physiotherapist is unable to always review airway clearance and inhalation technique. The patient is provided with a follow up appointment with a consultant physician 2 weeks after the annual review.*

### 1.5 Infection Control

Hospital centres aim to admit CF inpatients into single rooms, yet this is not always possible. At UHSM, and the new CF ward at KCH, all patients are or will be admitted to single rooms. It is generally accepted that CF inpatients should not share rooms where possible. Airway clearance, inhalation therapy and exercise are performed in isolation, with the exception of Zeepreventorium. Apart from Zeepreventorium, CF patients are encouraged not to socialise with each other, and
exercise alone with or without physiotherapy supervision.

In the UK, outpatient clinics have sufficient clinic rooms available to allow the patient to stay in the same room, with members of the CF team rotating through the room. This is done to reduce patient crossover and potential cross infection.

*All CF inpatients at SCGH are admitted into single rooms to reduce potential cross infection. During outpatient clinics, multiple rooms are used when possible to reduce patient cross over and potential cross infection. A pager system is also available for use, to allow patients to wait outside of the clinic waiting room.*

### 1.6 Adolescent Transition to Adult Services

The UK centres visited all had established adolescent transition services. At RBH and KCH, CF patients meet the adult CF team at the age of 12 or 13, then transition to the adult centre at the age of 15 or 16. The early transition age occurs as the centres feel it is important for care to be transferred before the age of other life milestones such as university entrance exams and beginning work are reached. At UHSM, adolescent CF patients transition to the adult centre at the age of 18, with less multidisciplinary team input prior to this occurring. At LUH and UBH paediatric and adult CF services are linked, resulting in less need for a formal transition program.

*A successful adolescent transition program runs between SCGH and WA's paediatric CF centre, Princess Margaret Hospital (PMH). CF teams from both centres maintain open communication, and regular transition clinics are held at PMH, allowing the adult team to meet adolescent CF patients. Adolescent CF patients currently transition to the SCGH at approximately age 18, which is slightly later than the international centres visited.*

### 1.7 Lung Transplantation

CF teams in the UK, Sweden and Belgium continue to provide care for CF patients up until lung transplantation is performed. However, assessment and workup for transplantation by the transplanting team begins many months and even years before this. Post transplantation, patients are managed by the transplanting team, with the exception being RBH, who still provide care for non-respiratory CF issues.

*Royal Perth Hospital (RPH) is the WA state heart and lung transplantation centre. CF patients from*
SCGH are referred to RPH for lung transplantation assessment and workup. Close communication is maintained between the two centres regarding patients awaiting lung transplantation. After surgery, RPH manages the patient's ongoing care. SCGH also has a lung transplant physician on site.

2. Physiotherapy Staffing and Service

CF physiotherapy staffing levels at centres visited are higher than at SCGH, and generally reflect recommendations made in Australian (2008)\(^1\), European (2005)\(^4\) and UK (2001)\(^5\) CF Standards of Care. Figure 2.1 illustrates centre staffing and Standards of Care comparisons, listing actual physiotherapy FTE for each centre, and the recommended physiotherapy FTE for the centres number of patients.

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Figure 2.1: *CF physiotherapy staffing comparison between centres visited and SCGH (Zeepreventorium excluded as not a primary CF care centre)

* Combined adult and paediatric CF physiotherapy service

S.O.C = Standards of care

Rec. = Recommended

All primary care centres visited provide an inpatient and outpatient service, with the level of service varying due to physiotherapy staffing. Most centres visited also provided a community service to patients, with the exception of UBH. In this case a government funded community physiotherapy service is available.

All inpatient centres provide a weekend physiotherapy service. Most patients receive weekend physiotherapy review, with the exception of those who may be independent with airway clearance.
and exercise, or those who may be non-compliant with their physiotherapy treatment regimen. All inpatient centres also provide an after hours service, for patient’s who are admitted to the ward outside of normal working hours, who have severe exacerbations requiring increased input, or who are clinically deteriorating and require urgent review.

Centres with high levels of physiotherapy staffing are able to provide a more complete physiotherapy service. Increased areas of CF physiotherapy service observed included twice daily physiotherapy review of airway clearance and exercise for all inpatients, increased outpatient clinic representation including physiotherapy led clinics, routine exercise assessment and prescription, and musculoskeletal and incontinence assessment and treatment. This increased staffing also enables for structured education and training of junior physiotherapists, to ensure appropriate care is being provided by all physiotherapists working with this patient group.

Physiotherapy staffing in CF at SCGH is well below recommended levels (Figure 2.1). It was observed that most centres visited in the UK and Europe meet these recommendations. Despite low staffing levels, a CF physiotherapy inpatient and outpatient service is provided at SCGH, but an integrated community service is not possible. Patients requiring review after discharge are reviewed to Cystic Fibrosis Western Australia, who can provide physiotherapy and homecare worker assistance. A weekend and after-hours CF physiotherapy service is provided as part of a weekend respiratory service at SCGH. While the CF physiotherapy service at SCGH meets most of the needs of the majority of its CF patients, some aspects of care are unable to be delivered. These aspects of care are discussed in Section 3 Physiotherapy Techniques.

3. Physiotherapy techniques

3.1 Airway Clearance Techniques

The choice of ACT prescribed to patients is individualised, with AD being the most common technique. In Sweden exercise is the foundation of physiotherapy treatment, combined with AD or the FET. Other techniques commonly used are ACBT, PEP, Acapella® and Flutter®, NIV and IPV. Airway clearance is most commonly performed in a sitting position, while patients at LUH and Zeepreventorium also incorporated supine and side-lying positions. No patients were observed using head down positioning or ‘tipping’, and no manual techniques such as percussion or vibrations were utilised. ‘The Vest’, a device which delivers high frequency chest wall oscillation, is not preferred as an ACT, and was not seen at any of the centres visited.

Most inpatients are recommended to perform airway clearance at least twice daily, however in some
centres one or both sessions may be substituted with exercise. Physiotherapy supervision of airway clearance is dependent on the patient’s ability to perform and clinical status, with some requiring more assistance than others. At home patients are expected to continue their prescribed ACT regimen, which is often modified to suit lifestyle.

At SCGH, patient’s airway clearance regimens are individualised, with the preferred ACT being PEP, Acapella®, Flutter® and ACBT. Since the completion of this fellowship, I have incorporated AD into patient’s regimens, with airway clearance devices used as adjuncts to this technique. Inpatients are expected to complete airway clearance twice daily, however due to staffing constraints supervision is not always able to be provided to patients twice daily. For patients who are non-productive of sputum, exercise and/or nebulised HTS is recommended.

Cystic fibrosis patients at SCGH are expected to continue their airway clearance regimen at home up to twice daily. Airway clearance techniques that empower the patient to be able to continue independent airway clearance after discharge are encouraged. The ACT regimen may be modified to suit their lifestyle, and education is provided regarding how to modify their regimen during an exacerbation. A small number of CF patients at SCGH use intensive daily exercise as their ACT.

3.2 Inhalation Therapy

All centres visited provided E-Flow® or Pari nebulisation devices at no cost to the patient. Patients using Promixin are provided with an I-Neb® by the pharmaceutical company. Fully subsidising the cost of equipment may improve adherence in some cases, but may also result in patients caring for equipment poorly. At UHSM, I-Neb® adherence measured in the outpatient clinic ranged from 3% to 97%, and physiotherapists reported many patients did not care for nebuliser equipment appropriately.

In the UK, Colomycin is the most commonly used nebulised antibiotic, followed by Tobramycin. Nebulised antibiotics are mixed with distilled water, normal saline or Salbutamol to improve tolerability as indicated.

Pulmozyme is the preferred muco-active medication, with nebulised HTS used less frequently. Physiotherapists at UK centres reported the use of nebulised HTS is increasing. In Lund, nebulised HTS is used frequently, with fewer patients using Pulmozyme. Nebulised antibiotics are less common in Lund than other centres visited. In Brussels and DeHaan, patients were using both HTS
and Pulmozyme regularly. HTS with a 3% concentration is commonly used to improve tolerability.

Test dosing of nebulised medications is performed by physiotherapists at all centres to ensure no bronchospasm occurs during treatment. Pulmozyme is given either as a 1 month or 3 month trial to assess effectiveness, before commencing the drug long term. At UHSM, test dosing is performed during a dedicated weekly physiotherapy outpatient clinic.

At SCGH, patients are provided with Pari nebuliser equipment at no cost. Patients using the E-Flow® system purchase equipment at their own cost, with only approximately 10% of patients using this device. All nebulised medications are prescribed after test dosing and trials. Pulmozyme and nebulised HTS are both commonly used muco-active agents. Patients use nebulised HTS once or twice daily, and may combine with a PEP device as an adjunct to airway clearance. Tobramycin and Colistin are the preferred nebulised antibiotics used at the centre.

3.3 Exercise Training

Individualised inpatient exercise programs were prescribed by physiotherapists at all centres visited, and supervised by either physiotherapists or physiotherapy assistants. Patients are encouraged to exercise at least daily during admissions. Exercise equipment was available in gymnasiums or in patient's rooms at all centres visited. Exercise programs consisted of either aerobic or resistance exercise, or a combination of both, and varied according to equipment available. Aerobic exercise was most commonly performed on an exercise bike or treadmill, with free or fixed weights used to perform resistance exercise. Exercise tests observed being used in inpatient settings were the 6MWT and PWC test.

All centres visited aimed to assess each patient's exercise capacity annually. An exercise test is usually performed at annual review clinics, with the exception being UHSM. At UHSM patients were able to have exercise capacity assessed at any time of the year by outpatient appointment. Exercise tests used at the centres are the 6MWT, MST and PWC test. UHSM, UBH and Zeepreventorium have facilities allowing physiotherapists to assess ventilation during exercise. All CF patients are encouraged to exercise as part of their home treatment regime, with individualised exercise prescribed to fit the patient's lifestyle and interests.

At SCGH, inpatients are encouraged to complete inpatient exercise at least daily, with physiotherapy supervision provided where possible. Patient's can be provided with an exercise bike
in their room, or can use a treadmill and resistance equipment in a gymnasium. Outpatient exercise assessments are completed at annual review clinics, with either a 6MWT or MST. Patients are encouraged to perform an individualised exercise program as part of their home treatment program.

3.4 Musculoskeletal Dysfunction

The level of musculoskeletal assessment and treatment provided to patients by CF physiotherapists varies according to staffing and level of expertise. In most cases, patients are taught postural exercises and screened for musculoskeletal issues. If more severe musculoskeletal issues are present, patients are referred to a musculoskeletal outpatient physiotherapist for further assessment and treatment. At UHSM, a dedicated service was provided by CF musculoskeletal physiotherapists.

The CF physiotherapy service at SCGH screens patients for postural problems and provides postural exercises as required. Patients requiring further assessment and treatment by a musculoskeletal physiotherapist are referred to the public hospital physiotherapy department closest to their home location. Currently the CF physiotherapy service at SCGH does not have the resources to expand this part of the service.

3.5 Incontinence

Physiotherapists at most centres routinely teach pelvic floor exercises to patients, and screen patients for incontinence issues. Patients are generally referred to incontinence or women’s health physiotherapists for further assessment and treatment if required. At UHSM, CF musculoskeletal physiotherapists have experience with diagnostic and feedback ultrasound.

Currently at SCGH patients are screened for incontinence issues at annual review clinics, or may raise this with a physiotherapist or physician at outpatient review. Patients are taught pelvic floor exercises as required, and can be referred on to a women’s health physiotherapist for further assessment and treatment. Currently the CF physiotherapy service does not have the resources to expand this part of the service.

4. Non-CF Bronchiectasis

Physiotherapy staffing for non-CF bronchiectasis is generally poor at all centres visited, when compared to that provided for CF. At UK centres, dedicated outpatient physiotherapy services are
developing, with varying staffing of up to 1 FTE physiotherapist aiming to provide inpatient and outpatient physiotherapy review for airway clearance, inhalation therapy, exercise and incontinence. It is acknowledged by physiotherapists at all UK centres, that they are unable to provide a complete physiotherapy service for non-CF bronchiectasis due to a lack of resource.

In most centres, the preferred ACT for this patient group is AD or ACBT. The Acapella® is used where appropriate at RBHT, but not at UHSM as they are not provided free by the hospital trust. Nebulised HTS is increasing in use, as are anti-pseudomonal nebulised antibiotics.

Non-CF bronchiectasis is the same lung disease that people with CF develop, although often less severe, with similar physiotherapy treatments recommended for this patient group. This includes ACTs, inhalation therapy, exercise training and incontinence review, which require considerable patient education and in many cases ongoing review by a physiotherapist.

At SCGH, a dedicated non-CF bronchiectasis physiotherapy outpatient service has been running for the past 2 years. Patients, referred by respiratory physicians and physiotherapists, are taught airway clearance techniques, trial nebulised HTS and antibiotics, and are given advice on exercise and incontinence. Patients are reviewed within 3 months of their initial assessment, and follow-up review is provided as required. The service is also used to screen for patients suitable for pulmonary rehabilitation programs. At present this service remains an outpatient service only, with more resources required to expand into a more comprehensive inpatient and outpatient service.

The British Thoracic Society guidelines, and Thoracic Society of Australia and New Zealand position statement, for non-CF bronchiectasis recommend patients are reviewed by a physiotherapist for assessment of ACT, inhalation therapy, continence, musculoskeletal dysfunction and suitability for pulmonary rehabilitation. It also recommended that patients are provided with a follow up appointment within 3 months, with ongoing review provided according to the patient’s clinical status.
DISSEMINATION OF FINDINGS

The skills and knowledge learned, and the findings of this fellowship will be disseminated to CF patients and families via clinical practice and CF community media. Information has been shared with the CF team and fellow colleagues at SCGH, and with further scheduled presentations this information will be shared with colleagues at PMH, RPH and LIWA. This report will be published and sent to key stakeholders including the Health Department of WA, CFWA and the Cystic Fibrosis Special Interest Group of the Thoracic Society of Australia and New Zealand.

This fellowship presents an opportunity for all Australian CF centres to benchmark their physiotherapy services against those provided by the centres visited in the UK, Sweden and Belgium. Furthermore, the Australasian CF Conference in Melbourne, 2011, provides a forum capable of ensuring this information is received and discussed by the wider Australian CF physiotherapy community. This will also allow discussion regarding how the observations from this fellowship compare to current evidence based guidelines.

Links with Curtin University of Technology will allow this information to be shared with physiotherapy students, and this report will be discussed with other WA universities offering physiotherapy courses. Links with rural and remote health centres in WA, currently being established, will see this information shared with physiotherapists and patients in areas unable to access sufficient ongoing education and training.

Furthermore, the knowledge and skills obtained from this fellowship, more specifically relating to ACT, provides an excellent opportunity for an ongoing post-graduate physiotherapy course to be established, to provide education and training to WA physiotherapists.
RECOMMENDATIONS

1. Physiotherapy techniques provided to CF patients at SCGH should incorporate new techniques learned at centres in the UK and Europe, to enable a wider range of treatment options to be provided.

2. The CF physiotherapy service at SCGH should be reviewed by the Physiotherapy Department, with the potential for business case development, aiming to increase physiotherapy FTE towards benchmarks set in the Australian, UK and European CF Standards of Care.

3. CF allied health service at SCGH should be considered for review by SCGH allied health departments, with potential for business case development, aiming to increase allied health FTE towards benchmarks set in the Australian, UK and European CF Standards of Care.

4. Physiotherapy services for patients with non-CF bronchiectasis at SCGH should be reviewed by the Physiotherapy Department, with the potential for business case development to ensure the physiotherapy service provided meets guidelines set in the British Thoracic Society guidelines, and Thoracic Society of Australia and New Zealand position statement for non-CF bronchiectasis.

5. Western Australian universities offering physiotherapy courses should take the opportunity to provide students with increased exposure to autogenic drainage, an ACT commonly used in the UK and Europe, and which will be incorporated into clinical practice at Sir Charles Gairdner Hospital.

6. A post-graduate physiotherapy course, incorporating ACT commonly used in the UK and Europe, and to be incorporated in clinical practice at SCGH, should be established through the physiotherapy department at SCGH to provide education and training to WA physiotherapists.

7. Physiotherapists working with patients who have CF, or non-CF bronchiectasis, should be considered for financial support from their employer, toward travel to improve their knowledge and skill base, to ensure their patient group has access to the widest possible range of physiotherapy treatment options.
8. Equipment required for the nebulisation of medications, such as Pari and E-Flow® devices, should be considered for subsidisation by CF centres, which may improve adherence to treatment regimes by reducing nebulisation treatment time.

9. This fellowship provides an opportunity for further measurement and comparison of CF service inputs, outputs and outcomes between SCGH and the centres visited, to assess the effectiveness of service offered to CF patients in each centre.
REFERENCES


