Physiotherapy management of the paediatric CF patient

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Paediatric respiratory physiotherapist
Royal London Hospital

APCP Paediatric Physiotherapy Course
11th June 2017
**Manifestations of Cystic Fibrosis**

**Lungs**
- Bronchiectasis
- Bronchitis
- Bronchiolitis
- Pneumonia
- Atelectasis
- Hemoptysis
- Pneumothorax
- Reactive airway disease
- Cor pulmonale
- Respiratory failure
- Mucoid impaction of the bronchi
- Allergic bronchopulmonary aspergillosis

**Heart**
- Right ventricular hypertrophy
- Pulmonary artery dilation

**Liver**
- Hepatic steatosis
- Portal hypertension
- Biliary cirrhosis
- Neonatal obstructive jaundice
- Cholelithiasis

**Gallbladder**
- Cholelithiasis

**Bone**
- Hypertrophic osteoarthropathy
- Arthritis
- Osteoporosis

**Intestines**
- Meconium ileus
- Meconium peritonitis
- Rectal prolapse
- Intussusception
- Volvulus
- Fibrosing colonopathy (strictures)
- Appendicitis
- Intestinal atresia
- Distal intestinal obstruction syndrome

**Spleen**
- Hypersplenism

**Stomach**
- GERD

**Pancreas**
- Pancreatitis
- Insulin deficiency
- Symptomatic hyperglycemia
- Diabetes

**Clubbing**
- Infertility (aspermia, Absence of vas deferens)
- Amenorrhea
- Delayed puberty

**Inguinal hernia**
What do all these people have in common?
Cystic Fibrosis

Newborn screening

NBS nationwide July 2007
5 CF centres receive referrals (RLH, GOSH, KCH, RBH, Lewisham)

NBS Pathway

- Heel prick blood spot test 5-7 days old
- IRT
- Repeat IRT
- Blood test for Genotype
- Referred to CF Team
- HV by CF CNS
- Sweat test
- Diagnosis ≈ 3-6 weeks old
Cystic Fibrosis

Benefits of newborn screening

Mortality
Lungs
Nutrition
Brain
Early detection and specialist care
Pre-symptomatic treatment
Avoidance of early complications
Planning for future pregnancies
Financial to health-care

Balfour-Lynn 2008
Cystic Fibrosis

Potential downsides of newborn screening

No screening tool is perfect
Grief – lose your “perfect” baby
Early diagnosis of “mild” or “atypical” CF
False negatives
Carrier detection
Newborn screening

MDT management

Medical
Diagnosis, education: life expectancy, effects of CF, treatment / future
Meds: flucloxacillin, vitamins, reflux meds

Nursing
CF Trust leaflet “Finding out”, CF service leaflet, Team contact card

Dietician
Ax of feeding & reflux, Creon

Clinical Psychologist
Dealing with diagnosis
Newborn screening

Physiotherapy management

Education on CF
How CF affects lungs
Long-term implications of recurrent chest infections
Role of Physiotherapy – preventative / treatment
CF Trust Information leaflet

Symptomatic babies and ?asymptomatic babies
Chest Physiotherapy
PEP in infants
“Exercise in babies”
What is physiotherapy in CF
Aims of modern physiotherapy

Previously the primary aim of chest physiotherapy was to remove infected mucus. Now, the aim is to prevent clogging of the airways and slow disease progression. Modern physiotherapy is a combination of

- Inhalation therapy
- Airway clearance techniques
- Exercise and physical strength
- Education

Aim to achieve this while minimising the impact on quality of life and participation
Focus on Inhalation therapy
Inhalation therapy

Inhalers & nebulisers
Inhalation therapy

Hypertonic saline

Significant improvements in lung function, quality of life and ease of expectoration (Elkins et al, 2006)

A sterile saltwater solution in concentrations of between 3 and 7%

4mls of solution is normally nebulised just prior to airway clearance or with airway clearance

It restores the depleted airway surface liquid volume, which peaks almost immediately after a dose

There is an improvement in mucus rheology

Stimulation of cough is seen in many patients
People with cystic fibrosis could be encouraged to inhale hypertonic saline before or during airway clearance techniques to maximise perceived efficacy and satisfaction, even though these timing regimens may not have any better effect on lung function than inhalation after airway clearance techniques.

Given the long-term efficacy of hypertonic saline has only been established for twice-daily inhalations, clinicians should advise patients to inhale hypertonic saline twice daily.

However, if only one dose per day is tolerated, the time of day at which it is inhaled could be based on convenience or tolerability until evidence comparing these regimens is available.
Inhalation therapy

DNase

The enzyme recombinant human deoxyribonuclease (dornase alfa), has been shown to reduce the viscosity of sputum taken from people with CF by digesting the deoxyribonucleic acid (DNA) released from neutrophils (Lieberman 1968).

The proprietary name of dornase alfa is Pulmozyme® (produced by Genentech Inc).

Therapy with dornase alfa over a one-, six- or 12-month period is associated with an improvement in lung function in CF (Jones 2010).

Daily inhalation of dornase alfa has has been shown to reduce the number of pulmonary exacerbations, improve pulmonary function and is well-tolerated and safe in patients with mild, moderate and severe CF (Kearney & Wallis, 2000).
Inhalation therapy

DNase

55 studies, of which five studies met inclusion criteria.

Four trials compared dornase alfa inhalation before versus after airway clearance techniques.

Inhalation after instead of before airway clearance did not significantly change forced expiratory volume at one second.

Forced vital capacity and quality of life were not significantly affected; forced expiratory flow at 25% was significantly worse with dornase alfa inhalation after airway clearance, mean difference -0.17 litres (95% confidence interval -0.28 to -0.05), based on the pooled data from two small studies in children (seven to 19 years) with well-preserved lung function.

All other secondary outcomes were statistically non-significant.
### Analysis 1.1. Comparison of Pre-ACT versus Post-ACT, Outcome FEV₁ (L)

**Review:** Timing of domal α₁a inhalation for cystic fibrosis

**Comparison:** Pre-ACT versus Post-ACT

**Outcome:** FEV₁ (L)

<table>
<thead>
<tr>
<th>Study or subgroup</th>
<th>Mean Difference (SE)</th>
<th>Mean Difference (IV Fixed, 95% CI)</th>
<th>Weight</th>
<th>Mean Difference (IV Fixed, 95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Over 1 week and up to 2 months</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anderson 2009</td>
<td>-0.09 (0.1)</td>
<td>7.8%</td>
<td>-0.09 [-0.029, 0.011]</td>
<td></td>
</tr>
<tr>
<td>Bishop 2011</td>
<td>-0.04 (0.09)</td>
<td>9.7%</td>
<td>-0.04 [-0.022, 0.014]</td>
<td></td>
</tr>
<tr>
<td>Fitzgerald 2005</td>
<td>0.02 (0.038)</td>
<td>54.3%</td>
<td>0.02 [-0.005, 0.009]</td>
<td></td>
</tr>
<tr>
<td>van der Giessen 2007a</td>
<td>-0.0929 (0.0528)</td>
<td>28.1%</td>
<td>-0.09 [-0.020, 0.01]</td>
<td></td>
</tr>
</tbody>
</table>

**Subtotal (95% CI)**

- Heterogeneity: Chi² = 3.50, df = 3 (P = 0.32), P = 14%
- Test for overall effect: Z = 0.94 (P = 0.35)
- Test for subgroup differences: Not applicable
Nebuliser systems

Jet compressors
Pari Turboboy
Slower than e-flow
Loud!
Needs to be connected to the mains
Bulky
Sturdy

Assess technique to ensure therapeutic effect!
Nebuliser systems

Vibrating mesh nebulisers

E-flow

Liquid medication passes through a metal mesh to break up the liquid into a mist where each drop is a similar size.

They deliver the mist of medication constantly.
Nebuliser systems

Adaptive aerosol delivery

I-neb
Small, battery operated, silent
Vibrating mesh technology
Provided if a patient is on Promixin
Has two form of nebulisation
Tidal breathing
Targeted breathing
Can only be used with a mouthpiece
Focus on Airway Clearance
CF airways disease

CF airways display an exaggerated and prolonged inflammatory response to viral and bacterial pathogens (Ratjen and Doring, 2003).

The destructive cycle of inflammation, infection, and mucus plugging causing airway obstruction is well established (Rosenstein and Zeitlin, 1998).

This leads to a progressive reduction in lung function and aerobic capacity.
Airway clearance in CF

What’s the rationale?

The aim is to facilitate secretion clearance and therefore:

- Reduce bacterial load
- Decrease infection and inflammation
- Reduce airway damage
- Reduce chest wall changes
- Ultimately delay the disease process.

A systematic review has reported that ACTs have short-term beneficial effects on mucus transport in CF, however there is currently no robust evidence regarding their long-term effects compared with no airway clearance.

The routine should be optimised such that it provides the most benefit for the patient with the least burden.
PEP

Positive expiratory pressure

Creates a “back pressure” in the lungs that increases the functional residual capacity (FRC)

Tidal volume reaches above the opening volume for closed airways

Due to interdependence between the airways, the lung parenchyma and the elastic recoil of the lung tissue at this increased FRC level, closed airways should open and collateral ventilation increase

I.e. getting air behind the secretions
Positive expiratory pressure (PEP)

Creates a “back pressure” in the lungs that increases the functional residual capacity (FRC)

Opens up collateral ventilation getting air behind the secretions
Reduces hyperinflation

Recommended 12-15 breaths per cycle in a variety of positions
Can be used in babies and children of all ages

It is combined with the forced expiratory technique
PEP

Positive expiratory pressure

Sitting comfortably, elbows supported

A resistor that gives 10-20cm H20 during mid expiration

12-15 slightly active TV expirations

Followed by FET and cough
Long-term multicentre randomised controlled study of high frequency chest wall oscillation versus positive expiratory pressure mask in cystic fibrosis

Maggie Patricia McIlwaine, Nancy Alarie, George F Davidson, et al.

Thorax 2013 68: 746-751 originally published online February 13, 2013
doi: 10.1136/thoraxjnl-2012-202915
PEP v “The Vest”

The evidence

A randomised controlled trial across Canada and the US compared PEP and “The Vest” in children and adults with CF

Patients using the PEP had fewer pulmonary exacerbations (1.2) in a year compared to those using the vest (2.0) (p<0.05).

The PEP system is considerably cheaper to use £20 versus thousands

A typical PEP session takes 15-20 mins
A typical Vest session takes 20 mins
Nebulized Hypertonic Saline Via Positive Expiratory Pressure Versus Via Jet Nebulizer in Patients With Severe Cystic Fibrosis

Oisin J O’Connell MD, Carmel O’Farrell, Mike J Harrison MB, Joseph A Eustace MD, Michael T Henry MD, and Barry J Plant MD

BACKGROUND: Nebulized hypertonic saline is a highly effective therapy for patients with cystic fibrosis (CF), yet 10% of patients are intolerant of hypertonic saline administered via jet nebulizer. Positive expiratory pressure (PEP) nebulizers split open the airways and offers a more controlled rate of nebulization. METHODS: In 4 consecutive adult CF patients who were intolerant of hypertonic saline via jet nebulizer, we nebulized 6% hypertonic saline via a PEP nebulizer. We measured the number of days the patients required intravenous antibiotics from enrollment to study end, compared to an equal period before PEP, and the mean time between the patients’ 3 most recent infective pulmonary exacerbation episodes before PEP to their next exacerbation after PEP. Patients also completed a Likert-scale adverse-effects questionnaire on hypertonic saline via PEP versus jet nebulizer. RESULTS: The 4 patients had severe CF pulmonary disease and all fully tolerated hypertonic saline via PEP, for 77, 92, 128, and 137 days, respectively until the study end date. There were fewer days of antibiotics in 3 of the 4 patients, from 45 to 20 days, 66 to 14 days, and 28 to 0 days (mean relative risk reduction 53%, $P = .11$). The other patient had 63 days of antibiotics during both the PEP and the jet nebulizer periods. There was a mean 3.6-fold longer time to next infective pulmonary exacerbation during the PEP period ($P = .07$). Adverse effects were less with PEP; chest tightness 68% ($P = .04$), bad taste 62% ($P = .06$), cough 47% ($P = .10$), and sore throat 50% ($P = .20$). CONCLUSIONS: Hypertonic saline via PEP nebulizer benefits CF patients who do not tolerate hypertonic saline via jet nebulizer. Key words: cystic fibrosis; CF; hypertonic saline; nebulizer; positive expiratory pressure; PEP; antibiotics. [Respir Care 2011; 56(6):771–775. © 2011 Daedalus Enterprises]

Introduction

Cystic fibrosis (CF) is the most common life-threatening autosomal recessive disease in Ireland, with an incidence of 1 in 1,461 births.\textsuperscript{1} CF is characterized by mucus retention, bacterial infection, and inflammation, leading to lung damage and ultimately respiratory failure. CF therapies aim to improve mucociliary clearance, reduce bacterial load, and lower airway inflammation.\textsuperscript{2}

A mutation in the CF transmembrane receptor gene results in abnormal ion transport across the respiratory epithelium.\textsuperscript{3} The primary pathophysiological defect is thought to be depletion of the airway surface liquid. The isotonic volume

\begin{figure}
\centering
\includegraphics[width=\textwidth]{figure.png}
\caption{A figure related to the content of the text.}
\end{figure}

\textbf{See the Related Editorial on Page 886}
Oscillating PEP

Acapella

The Acapella has a counterweighted plug and magnet to create airflow oscillation.

As you inhale the pressure builds up in the airways until the mechanism moves and gas escapes.

The oscillations that occur from the occlusion and opening of the gas pathway are transmitted through the tracheobronchial tree.

10-15 breaths with an FET and cough.
Oscillating PEP

Acapella

West et al 2010 found no difference between acapella and PEP during a 10 day chest exacerbation

Comes largely down to patient preference and adherence
Practical!
Autogenic drainage

Jean Chevallier
Belgian physiotherapist 1970s

Asthmatic children laughing

“Flow and breathing modulation concept”
Autogenic drainage

**Principles**

Slow inspiration
Inspiratory pause
Breathe out at correct lung volume for the position of the mucus in airways
Balance expiratory force with airway stability
Productive cough only
Good huff / cough technique
Changing lung volume level
MPD and Percussion

“traditional physiotherapy”

PD with head-down tip & percussion x2/day

Reflux (acid & non-acid)
18 – 40% of normal babs
35 – 81% infants with CF

? appropriate for babies with CF now
NO head down tip

Mcllwaine 1997 – PEP superior in adults
Airway Clearance

Other types of ACT

Active Cycle of Breathing Technique
High frequency chest wall compression (HFCWC)
Non-invasive ventilation
Percussion (used in infants)
Modified postural drainage
Used in combination with another technique
Intermittent positive pressure breathing
Trampolining
Main ACT in a Swedish centre
Role of the physiotherapist

Infants

Education

• Physiotherapists should meet the parents soon after diagnosis
• Usually the following day

Help the family get into a routine

Act as a resource

Teach airway clearance techniques if appropriate

• The importance of exercise should be instilled early
• Explain the benefits of exercise as an adjunct to airway clearance
• Promote an active family lifestyle
• Promote active play
• Interspersed with airway clearance
Role of the physiotherapist

**Toddlers and small children**

Adapt physiotherapy as the child grows and becomes more active in treatment

Liaise with MDT about starting new treatments as needed

Make physiotherapy fun – blowing games, trampoline, sticker charts

Start educating the child about their lungs and why they do physio

School visits with the dieticians and CNSs when the child starts school

Modify treatments during times of exacerbations
Role of the physiotherapist

Older child and adolescent

Continue to optimise airway clearance techniques

Recognise the growing independence of the child

Assist the family during times of change, such as starting secondary school

Often difficulties with compliance due to “physio is boring”

- Education important

Prepare the family for transition into adult services

Promote exercise
Focus on Exercise
Exercise in CF

A modern staple

The role of exercise is well established as a physiotherapy management strategy in CF

The ‘safety’ of exercise in CF was first documented in the early 1980’s

Exercise is now seen as ‘medicine’ for CF

Children are encouraged to exercise on a daily basis
## Benefits of Exercise in Cystic Fibrosis

<table>
<thead>
<tr>
<th>Benefit</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Improve physical function, improve cardiovascular performance, improve</td>
<td>Muscle strength</td>
</tr>
<tr>
<td>Slow the rate of lung function decline or increase lung function</td>
<td></td>
</tr>
<tr>
<td>Increase the effectiveness of airway clearance therapy</td>
<td></td>
</tr>
<tr>
<td>Improve aerobic capacity</td>
<td></td>
</tr>
<tr>
<td>Improve quality of life</td>
<td></td>
</tr>
<tr>
<td>Potential effects on CF related diabetes</td>
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</tr>
</tbody>
</table>

(Rand, 2012)
Review Article

Exercise Training in Children and Adolescents with Cystic Fibrosis: Theory into Practice

Craig A. Williams,¹ Christian Benden,² Daniel Stevens,¹ and Thomas Radtke¹,²

¹ Children’s Health and Exercise Research Centre, School of Sport and Health Sciences, University of Exeter, Exeter EX1 2LU, UK
² Division of Pulmonary Medicine, University Hospital Zurich, 8091 Zurich, Switzerland
<table>
<thead>
<tr>
<th>Recommended activities</th>
<th>Patients with mild to moderate CF lung disease</th>
<th>Patients with severe CF lung disease</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Cycling, walking, hiking, aerobics, running, rowing, tennis, swimming, strength training, climbing, roller-skating, (trampolining)</td>
<td>Ergometric cycling, walking, strengthening exercises, gymnastics, and day-to-day activities</td>
</tr>
<tr>
<td>Method</td>
<td>Intermittent and steady-state</td>
<td>Intermittent</td>
</tr>
<tr>
<td>Frequency</td>
<td>3–5 times per week</td>
<td>5 times per week</td>
</tr>
<tr>
<td>Duration</td>
<td>30–45 minutes</td>
<td>20–30 minutes</td>
</tr>
<tr>
<td>Intensity</td>
<td>70%–85% HRmax; 60%–80% peak ( \dot{V}O_2 ); LT; GET</td>
<td>60%–80% HRmax; 50%–70% peak ( \dot{V}O_2 ); LT; GET</td>
</tr>
</tbody>
</table>
Exercise in CF

Age appropriate exercise

Infants
- Swimming
- Swiss ball

Toddlers
- Swimming
- Swiss ball
- Games
- Obstacle course
- Circuits
- Trampoline
- Dance classes
Exercise in CF

Age appropriate exercise

School age

- Swimming
- Sports specific training
- Cardiovascular machines
- Circuits
- Weights
- Trampoline
- Core-conditioning
- Pilates
- Yoga
- Stretching
- Dance classes
Exercise in CF

Potential barriers to exercise

Breathlessness
Cough
Wheeze
CFRD
CF liver disease – contact sports
Haemoptysis
Pulmonary hypertension
Exercise in CF

How do we promote it?

Start early!

Baby gym groups, dance classes, swimming, trampoline clubs, sports clubs

Promote family exercise. It’s not of benefit just for the child with CF

The whole team needs to have exercise as an agenda – not just the physiotherapist
Other important factors

Things to consider

- Burden of treatment
- Fluid intake
- Bone density
- Nutritional status
- Adherence
“Assessment is a continuous process, which identifies individual problems and needs of the patient, providing information essential for effective clinical reasoning and formulation of treatment plans. It enables the monitoring of clinical management and evaluation of treatment intervention. It is the process that forms the basis of professional autonomy and competence and should include assessment of respiratory status and functional ability.”

CLINICAL GUIDELINES FOR THE PHYSIOTHERAPY MANAGEMENT OF CYSTIC FIBROSIS, 2008
Focus on Exercise Testing
Cardiopulmonary Exercise Testing

CPET is considered the “gold standard” for assessing aerobic capacity in the CF population

(Saynor et al., 2013)

Cardiopulmonary exercise testing (CPET) is a comprehensive assessment of respiratory, cardiovascular and metabolic changes on exercise usually performed on a treadmill or cycle ergometer using a ramp protocol

(Urquhart et al., 2012)

In healthy children and adults exercise capacity is usually limited by the cardiovascular system. However, CF children with severe lung disease (FEV1%pred <40%) may be limited by ventilatory factors during exercise.
## Exercise testing

<table>
<thead>
<tr>
<th>Cardiopulmonary exercise test</th>
<th>Field tests</th>
</tr>
</thead>
<tbody>
<tr>
<td>General fitness level</td>
<td>General fitness level</td>
</tr>
<tr>
<td>• Anaerobic threshold</td>
<td></td>
</tr>
<tr>
<td>• VO2 peak</td>
<td></td>
</tr>
<tr>
<td>What is limiting the patient</td>
<td>Response to exercise and breathlessness</td>
</tr>
<tr>
<td>• Ventilation</td>
<td></td>
</tr>
<tr>
<td>• Cardiovascular system</td>
<td></td>
</tr>
<tr>
<td>Changes from baseline</td>
<td>HR and SpO2 during exercise</td>
</tr>
<tr>
<td>Response to training</td>
<td>Changes from baseline</td>
</tr>
<tr>
<td></td>
<td>Response to training</td>
</tr>
</tbody>
</table>
THE PROGNOSTIC VALUE OF EXERCISE TESTING IN PATIENTS WITH CYSTIC FIBROSIS

PATRICIA A. NIXON, PH.D., DAVID M. ORENSTEIN, M.D., SHEeryl F. KELSEY, PH.D.,
and CARL F. DOERSHUK, M.D.

Abstract Background. Previous studies have shown female sex, impaired pulmonary function, older age, malnutrition, and colonization of the respiratory tract by *Pseudomonas cepacia* to be associated with a poor prognosis in patients with cystic fibrosis. We sought to determine the prognostic value of exercise testing in addition to the other prognostic factors.

Methods. A total of 109 patients with cystic fibrosis, 7 to 35 years old, underwent pulmonary-function and exercise testing with subsequent follow-up: 107 patients were followed for eight years. Regression analysis identified standard life-styles, the age, body-mass index, FEV1, end-tidal PCO2 at peak exercise (VO2peak), and the levels of aerobic fitness (VO2peak) as the most significant predictors of survival. The survival curve shows the percentage of patients surviving at each year of follow-up, categorized by low, medium, and high levels of aerobic fitness. Patients with low levels of aerobic fitness had a survival rate of 83 percent at eight years, as compared with rates of 51 percent and 28 percent for patients with middle (VO2peak, 59 to 81 percent of predicted) and lowest (VO2peak, ≤58 percent of predicted) levels of fitness, respectively. After adjustment for other risk factors, patients with higher levels of aerobic fitness were more than three times as likely to survive than patients with lower levels of fitness. Colonization by *P. cepacia* was associated with a risk of dying that was increased fivefold. Age, sex, body-mass index, FEV1, and end-tidal PCO2 at peak exercise were not independently correlated with mortality.

Conclusions. Higher levels of aerobic fitness in patients with cystic fibrosis are associated with a significantly lower risk of dying. Although better aerobic fitness may simply be a marker for less severe illness, measurement of VO2peak appears to be valuable for predicting prognosis. Further research is warranted to determine whether improving aerobic fitness through exercise programs will result in a better prognosis. (N Engl J Med 1992;327:1785-8.)

![Figure 1. Survival among 109 Patients with Cystic Fibrosis, According to Fitness Level.](image-url)

See the Results section for definitions of the fitness levels.
Aerobic fitness in CF

Why assess?

Greater aerobic fitness has been shown to correlate with a lower risk of hospitalisation (Perez et al 2013)

VO2peak is known to be an important predictor of mortality in the CF population (Nixon et al., 1992; Pianosi et al., 2005)

Known benefits of exercise in children with CF:
- improvements in VO2peak
- Increased strength
- Slow decline of FEV1 and FVC
Statement on Exercise Testing in Cystic Fibrosis

Helge Hebestreit\textsuperscript{a}  Hubertus G.M. Arets\textsuperscript{b,c}  Paul Aurora\textsuperscript{d}  Steve Boas\textsuperscript{f}  Frank Cerny\textsuperscript{g}  Erik H.J. Hulzebos\textsuperscript{c}  Chantal Karila\textsuperscript{i}  Larry C. Lands\textsuperscript{k}  John D. Lowman\textsuperscript{h}  Anne Swisher\textsuperscript{i}  Don S. Urquhart\textsuperscript{e}

for the European Cystic Fibrosis Exercise Working Group

\textsuperscript{a}University Children’s Hospital Würzburg, Würzburg, Germany; \textsuperscript{b}Department of Pediatric Pulmonology, and \textsuperscript{c}Child Development & Exercise Center, Wilhelmina Children’s Hospital, University Medical Center Utrecht, Utrecht, The Netherlands; \textsuperscript{d}Paediatric Respiratory Medicine and Lung Transplantation, Great Ormond Street Hospital for Children, and Portex Respiratory Unit, UCL Institute of Child Health, London, and \textsuperscript{e}Department of Paediatric Respiratory and Sleep Medicine, Royal Hospital for Sick Children, Edinburgh, UK; \textsuperscript{f}Northwestern University Feinberg School of Medicine, Chicago, Ill.; \textsuperscript{g}Women’s and Children’s Hospital of Buffalo and University at Buffalo, Buffalo, N.Y.; \textsuperscript{h}Department of Physical Therapy, University of Alabama at Birmingham, Birmingham, Ala., and \textsuperscript{i}Division of Physical Therapy, West Virginia University, Morgantown, W.Va., USA; \textsuperscript{j}Service de Pneumologie et Allergologie pédiatriques, Centre de Ressources et Compétences dans la Mucoviscidose, Hôpital Necker Enfants Malades, Université Paris V – Descartes, Paris, France; \textsuperscript{k}Montreal Children’s Hospital – McGill University Health Centre, Montreal, Que., Canada
### Table 2. Reasons for exercise testing in CF, recommended testing modes and test protocols

<table>
<thead>
<tr>
<th>Indication for test</th>
<th>Level of recommendation</th>
<th>Test and measurements</th>
</tr>
</thead>
<tbody>
<tr>
<td>Routine monitoring and assessment of exercise-related symptoms&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Preferred test</td>
<td>Cycle ergometry with pulse oximetry and with gas exchange measures using the Godfrey protocol&lt;sup&gt;a&lt;/sup&gt;</td>
</tr>
</tbody>
</table>
|                                                               | Second best options     | Cycle ergometry with pulse oximetry but without gas exchange measures using the Godfrey protocol<sup>a</sup>  
|                                                               |                         | Treadmill exercise with pulse oximetry and gas exchange measures using the Bruce protocol<sup>a</sup> |
|                                                               |                         | Treadmill exercise with pulse oximetry but without gas exchange measures using the Bruce protocol<sup>a</sup> |
| Pretransplant assessment                                      | Preferred tests         | Cycle ergometry with pulse oximetry and with/without gas exchange measures using the Godfrey protocol<sup>a</sup>  
|                                                               |                         | Treadmill exercise with pulse oximetry and with/without gas exchange measures using the Bruce protocol<sup>a</sup> |
|                                                               |                         | 6-min walk test with pulse oximetry                                                  |
| Physical activity counselling/recommendations/exercise prescription | Preferred tests         | Cycle ergometry with pulse oximetry and with/without gas exchange measures using the Godfrey protocol<sup>a</sup>  
|                                                               |                         | Treadmill exercise with pulse oximetry and with/without gas exchange measures using the Bruce protocol<sup>a</sup> |
|                                                               |                         | Maximal (incremental) field tests                                                    |
| Interim functional assessment                                 | Preferred tests         | Maximal (incremental) tests                                                          
|                                                               |                         | Submaximal tests                                                                     
|                                                               |                         | Task-specific tests                                                                  

The recommendations are based on expert consensus. <sup>a</sup> Other measurements such as ECG, blood pressure and/or blood gas analysis might also be indicated.
Exercise testing

Guiding exercise programmes

An exercise can indicate the limiting factor of the patient’s aerobic capacity.

Diagram showing the interaction between the respiratory system, cardiovascular system, and muscles during exercise, with labels for VCO₂, VO₂, CO₂ production, and O₂ consumption.
What is their PA level?

Global Recommendations on Physical Activity for Health

5–17 years old

The scientific evidence available for the age group 5–17 years supports the overall conclusion that physical activity provides fundamental health benefits for children and youth. Appropriate levels of physical activity contribute to the development of:

- healthy musculoskeletal tissues (e.g., bones, muscles and joints);
- healthy cardiovascular system (i.e., heart and lungs);
- neuromuscular awareness (i.e., coordination and movement control); and
- is also facilitates maintenance of a healthy body weight,

Moreover, physical activity has been associated with psychological benefits in young people by improving their control over symptoms of anxiety and depression; and being practiced by promoting opportunities for self-expression, building self-confidence, social innovation and integration.

Recommendations:

For children and young people, physical activity includes play, games, sports, transportation, daily chores, physical education, and planned exercise, on an ongoing basis, that are suitable for their age, abilities, and interests, provide enjoyment, and are safe. The key objectives are to improve cardiovascular and muscular fitness, bone health, and coordination and reduce health hazards are:

1. Children and youth aged 5–17 should accumulate at least 60 minutes of moderate-to-vigorous intensity physical activity daily.
2. Amounts of physical activity greater than 60 minutes provide additional health benefits.
3. Most of the daily physical activity should be aerobic. Vigorous-intensity activities should be incorporated, including those that strengthen muscle and bone, at least 3 times per week.

These recommendations are relevant to all healthy children aged 5–17 years, unless specific medical conditions indicate the contrary. In the absence of gender, race, ethnicity, or income level, whenever possible, children and youth with disabilities should meet these recommendations; however they should work with their health care provider to understand the types and amounts of physical activity appropriate for them considering their disability.

If children are currently doing no physical activity, doing amounts below the recommended level will bring more benefits than doing none at all. They should start with small amounts of physical activity and gradually increase duration, frequency and intensity over time.

The concept of accumulation refers to meeting the goal of 60 minutes per day by performing activities in multiple shorter bouts spread throughout the day (e.g. 3 bouts of 20 minutes), then adding together the time spent doing each of these bouts.

Assessing perceived exertion

The Children's OMNI-walk/run Scale of Perceived Exertion

The Children's OMNI-walk/run Scale of Perceived Exertion (category range, 0-10) was evaluated using male and female children (6-13 yr of age) during a treadmill graded exercise test.

Significant correlations were found between OMNI-walk/run Scale RPE responses and VO(2), %VO(2max), HR, V(E)/VO(2) ratio, and RR throughout the maximal treadmill exercise test.
Assessing perceived exertion

The Children's OMNI-walk/run Scale of Perceived Exertion

The strongest correlations were found between RPE and %VO(2max) ($r = 0.41-0.60$, $P < 0.001$) and HR ($r = 0.26-0.52$, $P < 0.01$).

The psychophysiological responses provide validity evidence for use of the Children's OMNI-walk/run Scale over a wide range of exercise intensities during both walking and running.
Key assessment

**Holistic approach**

As per acute Ax
Cough
Cough strength
Sputum production
Microbiology
Infection control status
Sinuses
Auscultation
Clubbing

CXR & HRCT
Spirometry
Chest shape & movement
Musculoskeletal Ax
Posture
Exercise capacity
Incontinence
Quality of life
Objective assessment

Baseline as per acute Ax

Heart rate
Temperature
Blood pressure
Respiratory rate
Auscultation
SpO2
Pain – acute or ongoing
Oxygen requirement
NIV requirement
Questions to address in cough evaluation

How did the cough start?
When did the cough start?
What is the quality of the cough?
  - Productive
  - Paroxysmal
  - Haemoptysis
  - Dry
  - Barking

Is the cough an isolated symptom?

What triggers the cough?
  - Exercise
  - Cold air
  - Lying down
  - Feeding
  - Early morning cough
Cough effectiveness

Can the patient huff and is it effective?

Can the patient cough and is it effective?

Patients with bronchiectasis often have an ineffective cough due to the diseased airway’s inability to maintain patency during the high pressures generated by a cough.
Focus on sampling
Sputum

What can it tell us?

One of the defining features of suppurative lung disease is the production of sputum.

Volume, colour, tenacity and daily pattern should all be assessed.

Ideally, an induced sputum should be performed to isolate any microorganisms growing.
Sputum

What can it tell us?

Colour – In adults with bronchiectasis, green purulent sputum is associated with bacterial infection (Woodhead et al., 2005)

Volume – an increase in volume can indicate infection

Tenacity – dry sticky secretions can indicate mucolytics are required and fluid intake may not be optimal
Sputum

What can it tell us?

Patients should be encouraged to expectorate to facilitate assessment

If sputum cannot be produced, regular cough swabs should be sent

Patients presenting with a viral illness on the background of CLD should have a viral and MC&S swab sent
Microbiological sampling

Lower & Upper Airways

MC&S Cough Swab
MC&S Sputum sample
NP Virology Swab or aspirate sample
Non-bronchoscopic BAL
Bronchoscopic BAL
Sinus washout
Microbiological sampling

Lower & Upper Airways

A positive cough swab is a strong predictor of sputum culture.

However, a negative cough swab does not rule out infection. Persistent symptoms should be further investigated.

RBH PCD guidelines recommend upper and lower airway samples at each clinic – this will be discussed in a lecture later on in module.
Microbiological sampling

Why sample?
To aid diagnosis
To guide therapy/management
To aid hospital infection control
To reduce patient length of stay and hospital costs
Infection control

What is the IC status of the patient?

Respiratory infections v age

Overall percentage in 2004:
- P.aeruginosa 57.3%
- H.influenza 16.2%
- B.cepacia 2.9%
- S.aureus 51.7%
- S.maltophilia 11.6%
- MRSA 14.6%
Infection control

Does the patient have any highly transmissible infections?

**Burkholderia Cepacia Complex**
Originally considered to be part of the Pseudomonas species
It is an obligate aerobic gram-negative rod bacteria
Associated with “Cepacia syndrome” in CF

**Mycobacterium abscessus**
A member of the non-tuberculous mycobacterium family
Upper airways

Sinus disease in chronic resp conditions

Patients with CF and PCD often grow pathogens in their upper airways.

CF patients often develop nasal polyps that can make nasal breathing difficult.

Sinusitis & rhinitis can also be troublesome for patients with chronic respiratory conditions.
Clubbing

Indication of a variety of diseases
Cardiac, respiratory, digestive
Focus on spirometry
Spirometry

Spirometry is used to:
- establish baseline lung function
- evaluate dyspnoea
- detect pulmonary disease
- monitor effects of therapies used to treat respiratory disease
- evaluate respiratory impairment
- evaluate operative risk
- perform surveillance for occupational-related lung disease.
Flow-volume curves

Examples of expiratory flow-volume curves from a healthy subject, and in obstructive, restrictive and mixed pulmonary defects. Confirmation of restrictive defects (quite rare in general practice) requires measurement of total lung capacity in a lung function laboratory (right panel). (Reproduced from Quanjer et al.9 with permission from the editor of the European Respiratory Journal).
Spirometry

Questions to ask

What are the FEV1 and FVC?
Are they reduced?

Is there scooping on the curve?
Is it reversible?

Look at the FEV1/FVC ratio – remember this ratio is different for different age groups

Think about the small airways
CXR & HRCT
Chest observation

Chest shape

Look for deformities

Is the chest symmetrical?

Is the AP diameter increased – barrel chest

Abnormalities of the chest wall can increase the WOB and indicate an underlying disease process

• Bell shaped chest in SMA
Chest observation

Chest movement

- Symmetrical
- Reduced
- Paradoxical
  - Can indicate a respiratory muscle weakness
  - Seen in SMA type 1
Chest observation

Breathing pattern

Is the patient using accessory muscles to breath?
  - apical breathing

Is the tummy moving our on inspiration?
  - diaphragmatic breathing
Focus on posture
Posture

What is posture?

“The mechanical relationship of the parts of the body to each other. It can be divided into static and dynamic posture. It changes with positions and movements of the body, and is influenced by many factors including general health, sex, body build, strength, personal habits, environment and mood”

(Kendall et al, 1952)
Posture

What is ideal posture?

The head in neutral
The ribcage is able to expand easily
The scapulae lie flat
The natural curves of the spine are present
Neutral pelvis
The knees are not hyperextended
Posture

Why is posture important?

The muscles of the trunk have a postural and a respiratory element to their function.

We should assess posture in patients with chronic respiratory disease

- In CF, this is recommended at annual review and for inpatient stays
Posture

Spinal curves
Head posture
Shoulder restrictions
Chest wall shape and mobility
Muscle imbalances
Neuromuscular control – motor strategies
Lung function base line measurements
Which Posture do you have?

Correct  Hollow Back  Flat Pelvis  Slumping  Military  Round Shoulders  Correct  High Shoulder  High Hip  Head Tilt  Severe Scoliosis
Musculoskeletal assessment

Not just the lungs

Musculoskeletal pain is associated with a reduction in QoL and ability to perform ACT (Botton et al, 2002)

Assess:
Muscle strength
Joint ROM
Muscle flexibility
Thoracic mobility
Muscle Length and Joint Range of Motion in Children with Cystic Fibrosis Compared to Children Developing Typically

Allison Mandrusiak, Donnæe Giraud, Julie MacDonald, Christine Wilson, Pauline Watter

ABSTRACT

Purpose: To explore range of motion in children with cystic fibrosis (CF) compared to children developing typically. With the increasing longevity of people with CF, musculoskeletal concerns are becoming more prevalent and should be identified and considered in management plans. Reduced range of motion (ROM) in older people with CF has been demonstrated, and the age at which these changes begin to occur must be explored.

Methods: Participants were 38 children with CF (aged 7–14 years) and 38 children developing typically, matched for age and gender. Muscle length and joint ROM measurements were compared, using analysis of variance, in the thoracolumbar region (extension, flexion, lateral flexion), upper limb (pectoralis minor, pectoralis major, shoulder horizontal abduction), and lower limb (hip internal and external rotation, hamstrings, gastrocnemius-soleus).

Results: Children with CF had significantly shorter pectoralis minor, pectoralis major, and gastrocnemius-soleus muscles and significantly greater range of shoulder horizontal abduction than matched controls.

Conclusion: Altered ROM was identified in children with CF, strengthening the rationale to support proactive physiotherapy management of the musculoskeletal system, aiming to optimize function across the increasing lifespan. Evaluation and management of ROM and its impact on activity and participation are important areas for future research and clinical practice.

Key Words: children, cystic fibrosis, ICF, joint range of motion, muscle length

Focus on incontinence
Incontinence

What is urinary incontinence (UI)?

There are two types of incontinence:

- Urge incontinence happens when the muscle of the bladder is over sensitive and there is not enough warning of the need to empty the bladder. Certain drugs (e.g. caffeine) may make the bladder muscle ‘twitchy’ and cause urge incontinence.

- Stress incontinence happens during actions such as coughing and sneezing and this is the type most commonly reported by patients with chronic cough.

- A combination of the two causes can also occur and is known as mixed incontinence.
Incontinence

What is urinary incontinence (UI)?

Urinary incontinence has been found to be common in girls and women with cystic fibrosis (CF).

This often can have a marked impact on daily activities and quality of life.

It may also lead to potential deterioration of respiratory status due to reduced adherence to forced expiratory manoeuvres (cough, lung function tests and exercise) by the girls trying to avoid occurrence of urinary incontinence (UI).
Incontinence

What is urinary incontinence (UI)?

National guideline recommendations in the management of CF in Australia and the United Kingdom have in recent years changed to incorporate screening and management of UI.

ACPCF national standards of care for CF advise screening for incontinence

Assessed at annual review or if any concerns

At RLH, we screen every CF patient over the age of 10
Incontinence

Assessing incontinence

Can be an awkward subject for teenage girls, but it is important to establish if incontinence present

They may be embarrassed if siblings are present – try to ensure there a few people in the room

Explain that it is something they can speak to the physiotherapist about
New ways to engage

Using technology

Squeezy App
Quality of Life

There are many questionnaires that are used to assess Quality of life in children with respiratory conditions:

Paediatric Quality of Life Inventory

The Cystic Fibrosis Questionnaire revised (CFQ –R)
Quality of Life

Paediatric Quality of Life Inventory

The PedsQL Measurement Model is a modular approach to measuring health-related quality of life in healthy children and adolescents and those with acute and chronic health conditions.

The survey integrates generic core scales and disease-specific modules.

There are four scales: Physical Functioning (8 items), Emotional Functioning (5 items), Social Functioning (5 items), and School Functioning (5 items).
In the past ONE month, how much of a problem has this been for you ...

### ABOUT MY HEALTH AND ACTIVITIES (problems with...)

<table>
<thead>
<tr>
<th>Problem</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Often</th>
<th>Almost Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>It is hard for me to walk more than one block</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>It is hard for me to run</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>It is hard for me to do sports activity or exercise</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>It is hard for me to lift something heavy</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>It is hard for me to take a bath or shower by myself</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>It is hard for me to do chores around the house</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I hurt or ache</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I have low energy</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>

### ABOUT MY FEELINGS (problems with...)

<table>
<thead>
<tr>
<th>Feeling</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Often</th>
<th>Almost Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>I feel afraid or scared</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I feel sad or blue</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I feel angry</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I have trouble sleeping</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I worry about what will happen to me</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>

### HOW I GET ALONG WITH OTHERS (problems with...)

<table>
<thead>
<tr>
<th>Problem</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Often</th>
<th>Almost Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>I have trouble getting along with other kids</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Other kids do not want to be my friend</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Other kids tease me</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I cannot do things that other kids my age can do</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>It is hard to keep up when I play with other kids</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>

### ABOUT SCHOOL (problems with...)

<table>
<thead>
<tr>
<th>Problem</th>
<th>Never</th>
<th>Almost Never</th>
<th>Sometimes</th>
<th>Often</th>
<th>Almost Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>It is hard to pay attention in class</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I forget things</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I have trouble keeping up with my schoolwork</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I miss school because of not feeling well</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>I miss school to go to the doctor or hospital</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>
Quality of Life

The Cystic Fibrosis Questionnaire revised (CFQ –R)

Disease-specific instrument designed to measure impact on overall health, daily life, perceived well-being and symptoms.

Developed specifically for use in patients with a diagnosis of cystic fibrosis.
Quality of Life

The Cystic Fibrosis Questionnaire revised (CFQ –R)

Four versions of the instrument have been developed

- one for adults and adolescents 14 years of age and older (CFQ Teen/Adult)
- two for assessing children ages 6-13 years, one to be completed by the child and one to be completed by parent (CFQ Child and CFQ-Parent respectively)
- One for preschool children
Useful guidelines

Standards of Care and Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis
Third edition – April 2017

The CF Trust national standards of care for children with CF

The “Blue booklet” by the IPG/CF
References

Key references

Physiotherapy for respiratory and cardiac problems – adults & paediatrics. Pryor & Prasad 2008

Clinical guidelines for the physiotherapy management of cystic fibrosis, 2008

Recommendations for the assessment and management of cough in children. Shields et al., 2008


References

Key references


