PT and Cystic Fibrosis: A Successful Team from Birth to Healthy Aging!

Part II: How to Bring Your Talents to the Needs of People with CF

Intro/Review of Part I

• Effects of CF on body systems
  – Lungs
  – Digestion & Endocrine
  – Musculoskeletal

• Challenges created for PT
  – Multiple systems changing across the lifespan

• Opportunities for involvement
  – Within and outside of CF Care Centers

Musculoskeletal Assessment
Musculoskeletal Considerations in the Cystic Fibrosis Population

Matthew Nippins PT, DPT, CCS
APTA Combined Sections Meeting 2013, San Diego, CA

CF and Musculoskeletal Impairments

- The onset of musculoskeletal changes can start as early as 8 years of age and are generally present at the end of puberty (Massery 2005).
- These abnormalities are caused by decreased bone mineral density and the abnormal respiratory workload superimposed on the developing musculoskeletal system of a developing child (Lanenefors 2004).

Theories of Musculoskeletal Impairments in CF

- Hyperinflation caused by obstruction causes flattening of the diaphragm and increased A-P diameter of the chest wall altering respiratory workload.
Theories of Musculoskeletal Impairments in CF

- All trunk musculature functions both as postural and respiratory muscles—even the diaphragm (Hodges, 2000–2003).
- Therefore if respiration is compromised or taxed, the effort of these trunk muscles in respect to posture/postural control will be reduced to support needs of respiration.
- This leads to postural adaptation during musculoskeletal development (Massery, 2005).

All the Theory Leads to Impaired.....

- Breathing Pattern
- Posture
- Strength
- Flexibility
- All of which further impair ventilation, endurance, aerobic capacity and more.....

The Musculoskeletal Assessment

- Breathing Pattern
  - Essential to linking the respiratory mechanics to the musculoskeletal system
  - This provides critical information to your postural assessment
Breathing Pattern

• Assessment (observation and palpation) of the following:
  – Accessory muscle use
  – Lateral costal expansion (the bucket handle)
  – Upper chest wall movement (the pump handle)
  – A-P to lateral diameter (barrel chest)
  – Chest wall expansion
  – Abdominal/Diaphragmatic expansion

Postural Assessment

• No different than a postural screen for any other patient
• Screening of posture in the, anterior, posterior and lateral planes
• Use of a plum line or points of contact against a wall can also be helpful in screening posture with this population

Postural Assessment

• Head & Neck
  – Forward head posture
  – Symmetry?
    • Accessory muscle use or tightness may cause asymmetry
• Shoulder Complex
  – Anterior view
    • Hand position (IR)
  – Posterior view
    • Scapular position (winged or anteriorly tilted)
Postural Assessment

• Shoulder Complex
  – Lateral view
  • Glenohumeral joint
    – Position of the humeral head in the GH joint
      ⇒ More anterior in this population
  • Shoulder girdle
    – Protracted/rounded

Postural Assessment

• Thoracic and Lumbar Spine
  – Lateral view
  • Thoracic kyphosis and lumbar lordosis
    – Increased kyphosis: due to anterior accessory muscle use and tightness
    – Increased lordosis: due to limited extension in the thoracic spine
  – Posterior view
    • Alignment of spine: scoliosis
      – May significantly affect ventilation and is usually compensated for when looking at breathing pattern

Postural Assessment

• Pelvis
  – Pelvic tilt
    • Anterior tilt with increased lumbar lordosis, excessive thoracic kyphosis, hip flexor tightness and/or weak abdominal muscles

• Lower extremities
  – Relationship to the postural impairments seen up the chain
Flexibility Assessment

- Chest Wall Expansion (also may be performed in the breathing pattern assessment)
  - Chest wall measurements (DeTurk & LaPier, 2004)
    - Overall expansion at the angle of Louis, xyphoid process and umbilicus
  - Rib Springing
    - Applied in the direction of chest wall movement to assess mobility
  - Side-bending
    - Determines quadratus tightness
  - Ruler test
    - Side-bending with observation of rib mobility

Flexibility Assessment

- Shoulder Complex
  - Functional screening
    - Scratch test
    - Butterfly position
    - Fastening of a bra (females) & reaching for a wallet (males)
  - All may lead to further specific tests and measures

Flexibility Assessment

- Cervical Spine
  - Screening of AROM in all planes
    - Forward head posture
      - Leads to increased tightness of the anterior accessory muscles (SCM) and shortened cervical extensors
    - Decreased lateral flexion and/or rotation from accessory muscle use (tight upper traps)
Flexibility Assessment

- Thoracic & Lumbar Spine
  - Screening AROM in all planes
  - Looking for compensatory motions from postural abnormalities
  - Decreased thoracic extension is common
    - Prone extension test
  - Manual assessment of individual spinal segment mobility

Muscle Fitness

- The CFTR defect may play a role in skeletal muscle wasting (Petrof 2009)
- This can lead to peripheral muscle weakness which may be exacerbated further by inactivity or steroid myopathy (Troosters 2009)
- Further this negatively effects functional activities and decreases the efficiency of these activities causing further fatigue, dyspnea or aerobic impairment (viscous cycle of inactivity)

Muscle Fitness: Strength

- Ventilatory Muscles
  - Maximal inspiratory pressure (MIP) & maximal expiratory pressure (MEP)
    - Measured in a PFT lab or PT clinic
    - Gives a cmH₂O pressure measurement of overall inspiratory and expiratory strength
Muscle Fitness: Strength

- Extremity screen
  - Standard MMT and/or hand dynamometry
    - Proximal weakness is common
- Trunk
  - Assessment of the accessory muscles of breathing, scapular adduction, thoracic/lumbar extensors & abdominal muscle complex

Muscle Fitness: Pelvic Floor

- Stress urinary incontinence is a frequent and underreported manifestation of CF
- Most commonly seen as stress urinary incontinence (SUI) which is unintentional urine leakage with cough, laugh, sneeze or strenuous activity
- More common in girls and women: between 30%-69% reported prevalence in various studies (Button 2004, Langman 2004)
  - Male prevalence ~15% (Button 2004)
- Intra-abdominal and intra-thoracic pressures increase with cough
- Chronic increased cough associated with CF pushes this pressure down against the pelvic floor muscles repeatedly which can stretch and/or weaken these muscles causing leakage
Exercise Capacity Assessment and Prescription

Practical Aspects of Exercise Testing and Prescription
Anne K. Swisher PT, PhD, CCS
West Virginia University

Exercise Testing: Ready, Set, Go!
Preparation for Maximal Exercise Test

- Screen health history
  - Esp. cardiovascular issues, metabolic (CFRD) issues, musculoskeletal issues (BMD, arthropathy)
- PFTs
  - Esp. maximal voluntary ventilation (MVV)
- Personnel and training
  - Minimum 2 people (1 for patient, 1 for equipment)
  - Experienced and trained in emergency management (BLS, ACLS as needed) and measuring exercise vitals
- Monitoring equipment
  - ECG, BP, pulse oximeter

During the Maximal Exercise Test

- BP every 1 or 2 min
- SpO₂ continuously
- 3-lead ECG continuously with someone watching it
- Rate of perceived exertion every 1 or 2 min
- Record vitals during recovery (1, 2, 5 min) and watch ECG closely following maximal effort
  - This is the highest risk time

Interpreting the Test

1. was it maximal?
   - RER > 1.1, meeting age-predicted HR max, RPE of very strong, etc
2. was it normal?
   - Predicted workload met
   - Normal vital sign response
   - Predicted VO₂ max met
Abnormal Test: Deconditioned

- HR reaches age-predicted max but low workload
- May also indicate underlying cardiac problem
- *If cardiac is OK, these patients can improve VO₂max with aerobic training*

Abnormal Test: Ventilatory Limitation

- VEmax is > 85% of MVV
- Low peak HR (not always)
- Low peak workload

  - Full body training response will be limited by ventilation, however, interval training can significantly improve submaximal capacity
  - *? Role for non-invasive ventilation during training*

Abnormal Test: Gas Exchange Limitation

- SpO₂ drops below 85% (or high pCO₂)
- Low maximal workload achieved
- HR may be low or near maximal to compensate for low pO₂

  - *Important to train these patients with adequate levels of supplemental oxygen (keep SpO₂ > 90%)*
Translating Test Results to Behavior

Guidelines for Exercise/Physical Activity

- 60 min moderate-to-vigorous physical activity daily for children
- 30 min moderate physical activity 5 days per week for adults OR:
- 20 min vigorous physical activity 3 days per week for adults
- No specific CF guidelines
- More physical activity is better

Evidence-based intervention in physical activity: lessons from around the world

Graph showing data from various studies.
Exercise to Increase VO$_2$max
- Continuous activity
- Using large muscle groups
- 20-30 minutes per session
- 3-5 times per week
- 50-70% of VO$_2$max or 70-85% of HRmax
- Best for teens and adults who are ready for structured program

Exercise to Improve Anaerobic Capacity
- Intervals (30 sec to 2 min) of high-intensity (> 80% of max) followed by same or longer periods of recover (< 40% of max)
- If total exercise time is > 30 min, can also see improvements in VO$_2$max
- Best for young children, as this is developmentally appropriate
- Also good for ventilatory limited patients

Exercise to Improve Muscular Strength
- > 70% of 1-repetition maximum load
- 1-2 sets
- 8-15 repetitions
- Must have good form, including full range of motion and proper breathing
- Can start pre-teens on resistance training, but don’t expect large gains—focus on learning the movements
Change: The Problem of Inertia!

Exercise Prescription: Interview

- Interview
  - What are the patient’s likes & dislikes?
  - How does the patient perceive exercise/physical activity?
    - Facilitators/benefits
    - Barriers/drawbacks
  - Does the patient feel competent to be active?
- Use open-ended, non-judgmental questions to help the patient come to a plan
  - Consider: seasons/weather, changes in interests, time demands of other CF care (can aerobic exercise replace some/all of ACT?)

Example: Action Plan demo

<table>
<thead>
<tr>
<th>Physical Activity Action Plan</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Describe what you want to do:</td>
</tr>
<tr>
<td>2. Describe why:</td>
</tr>
<tr>
<td>Where:</td>
</tr>
<tr>
<td>When:</td>
</tr>
<tr>
<td>What:</td>
</tr>
<tr>
<td>Frequency:</td>
</tr>
<tr>
<td>3. Describe:</td>
</tr>
<tr>
<td>4. Describe who/what/why:</td>
</tr>
<tr>
<td>5. Consider confidence ratings (0-10):</td>
</tr>
<tr>
<td>6. Follow-up:</td>
</tr>
</tbody>
</table>
Exercise/Physical Activity Prescription
for the “Ages and Stages” of CF

Principles of Exercise Training

• Balanced program
  – Aerobic training
  – Resistance training
  – Flexibility

• Specificity of training for task
  – This will differ developmentally

• Avoid bad outcomes
  – Injury (e.g. muscle/joint)

Activity is Fun!
Goal Setting

- Set initial goal low to make it achievable
  - Rate confidence and conviction (action plan)
- Be specific (SMART goals)
  - Specific
  - Measurable
  - Achievable
  - Reasonable
  - Time-oriented
- Follow-up early (2-4 weeks) for accountability
  - Schedule a specific time to call/contact patient for this purpose

Special Situations for Exercise

Blood glucose levels & CFRD

- Need to monitor blood glucose (finger sticks) before, during, immediately after exercise AND at bedtime when beginning a new program
  - Work with endocrinologist & dietitian to modify calories and carbohydrates
- May need to assess for peripheral neuropathy and protective sensation
- May need to assess skin and fit of shoes for weight bearing activities
- Have an exercise partner
Low BMD

- Modify resistance training activities for more controlled situations (e.g. machines instead of free weights)
- Avoid vigorous spinal flexion, hip rotation
- Watch for pain with lifting, coughing, sneezing

- *Important to have weight bearing stress but safely*

High-velocity Loading

Jumping rope/plyometrics
- Recommend GRF 2-9 times body weight
- Needs to be progressive as bone adapts
- Unusual strains better than doing the same thing every time
- Multiple short sessions 2-3 times per week for total of 20-30 min (10-50 jumps per day)

Sports (volleyball, basketball)

Low-velocity Loading

Resistance training: Barbell squats, seated rows
- Safer for those with established osteoporosis or fractures
- Still engages mechanical stress of muscle on bone insertion sites
- Probably better for preventing further loss than gaining BMD

Walking, swimming

- Still engages mechanical stress of muscle on bone insertion sites
- Probably better for preventing further loss than gaining BMD
Movements to Avoid/Limit in Known Osteoporosis

- Twisting spine
- Coughing without support
  - both pelvic floor and thoracic spine
- Flexing forward with extended legs
- Lifting with flexion or rotation

Dehydration/electrolyte depletion

- Increase intake of salty drinks (sports drinks) and salty snacks for exercise involving significant sweating
- Limit exercise without a partner
- Work with dietitian here

Hemoptysis/pneumothorax

- Avoid high-intensity exercise until cleared
- New onset of blood in sputum should always be assessed by the CF team
- NO scuba diving, parachuting or bungee jumping for those at risk of pneumothorax
- Avoid skiing at high altitude and mountain climbing for those with low pO2
Very low BMI

– Must increase caloric intake to balance what is expended with physical activity/exercise
– Increased lean muscle mass means increased basal metabolic rate
  • Burn more calories all day long
– Work closely with CF dietitian to balance these issues
– Big concern of parents and patients, but exercise also stimulates appetite and may decrease overall inflammation, which consumes lots of calories

How to coordinate with ACT and inhaled medications?

– Exercise is a bronchodilator
  • Recommend 2 min of moderate walking before doing ACTs (Elkins et al)
– Exercise that increases VE causes mucus to be mobilized, thus these types are airway clearance
– Not ALL types of exercise mobilize mucus, so not all exercise is airway clearance

Exercise Assessment and Prescription for Adults with CF
Why Exercise Test?

- Peripheral muscle or non-pulmonary factors may predominate as limiting factors to exercise in patients with mild to moderate CF.


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Why Exercise Test?

- Patients with VO2 peak \( \geq 82\% \) predicted had a 83% higher survival rate than patients with VO2 peak 59–81%, or \( \leq 58\% \) of predicted.
- Survival rates were 51% and 28%, respectively.
- Age, sex, body mass index, FEV1, and PET CO2 at peak exercise were not independently correlated with mortality.
- Patients with higher levels of aerobic fitness had a 3 times greater survival than patients with lower aerobic fitness levels.


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Why Exercise Test?

- It has been suggested that in patients with mild pulmonary disease, parameters of exercise at peak of maximal performance may detect disease not identified by routine lung function testing or body mass index.

Exercise Testing: Now What

- 10-12.5% of program use formal exercise prescriptions.
- Prescribe safe levels of exercise especially in those with advanced lung disease.
- Describe the need for supplemental oxygen.
- Describe level of disability


Exercise Testing Considerations

- Space limitations
- Equipment cost/availability
- Patient preference/safety
- Knowledge of staff on how to perform/interpret/utilize equipment

How To Exercise Test

- Submaximal
  - 6-minute walk test
  - Step tests
  - Incremental tests
- Maximal
  - Bicycle
    - Godfrey
    - Wingate
  - Treadmill
    - Modified Balke
    - Bruce
6-Minute Walk Test (6MWT)

- Allows clients to self pace
- Distance is primary outcome measure
- Good for persons with moderate to severe lung disease

3-Minute Step Test (3MST)

- Uses a 15-cm step at 30 steps per minute
  - Metronome set at 120
  - Leading leg is changed at 1.5 minutes
- More sensitive than 6MWT in detecting desaturation
- Easy to perform for young children

Incremental Shuttle Walk Test

- Incremental test
- Full test takes 12 minutes to complete
- Utilizes a 10-meter oval course marked with two cones
- More challenging than 6MWT and 3MST
Sub/Maximal Tests

- **Submaximal test benefits/limitations**
  - Easy
  - Low cost
  - No MD presence needed
- **Maximal**
  - Expensive
  - Needs equipment
  - Need MD presence

Norms

- Currently no norms exist for persons with CF
- Regression equations do exist for persons without specific health conditions

What’s “Normal”

- These regression equations can be used to compare exercise tests for children 8-18 years of age
  - **Boys**: Peak Work = (20*age) - 94 (SE = 26, r=0.91)
  - **Girls**: Peak Work = (13*age) - 23 (SE=24, r=0.84)
- Peak work was measured in watts.

Cystic Fibrosis – Related Diabetes (CFRD)

- Effects up to 20% of adults with CF
- As many as 75% of adults with CF have glucose intolerance
- Insulin deficiency occurs due to scarring of the pancreas
- Chronic infections can cause insulin resistance
- Use of steroids to treat lung disease causes increase in cortisol level contributing to diabetes

Exercise and CFRD

- Regular aerobic exercise can improve blood glucose control and delay onset of complications of type II diabetes
- Due to concern of peripheral neuropathy, sensation (especially on the soles of the feet) should be assessed
Exercise and CFRD

- Blood glucose levels should be checked before, during and after exercise
- Insulin shots should be avoided in exercising muscles
- Source of carbohydrate should be available
- Exercise should not be done when alone until blood glucose response to exercise is stable

Exercise and CFRD

[Diagram showing blood sugar levels and exercises]

Dyspnea

- “Abnormal sensation of awareness of breathing out of proportion to the given ventilatory or metabolic demand”

- Clinical measurement:
  - To assess impact of dyspnea on functional status
  - To identify efficacy of treatment

**MMRC Dyspnea Scale**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Not troubled with breathlessness except with strenuous exercise</td>
</tr>
<tr>
<td>1</td>
<td>Troubled with shortness of breath when hurrying on the level or walking up a slight hill</td>
</tr>
<tr>
<td>2</td>
<td>Walks slower than people of the same age on the level because of breathlessness or has to stop for breath when walking at own pace on the level</td>
</tr>
<tr>
<td>3</td>
<td>Stops for breath after walking about 100 yards or after a few minutes on the level</td>
</tr>
<tr>
<td>4</td>
<td>Too breathless to leave the house or breathless when dressing or undressing</td>
</tr>
</tbody>
</table>


**Dyspnea Index**

- Patient is asked to count to 15, while the number of breaths (including the first) are counted
- Rating is assigned based on total number of breaths:
  - 0 single breath
  - 1 two breaths
  - 2 three breaths
  - 3 four breaths
  - 4 unable to count

**Rate of Perceived Exertion**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>no breathlessness at all</td>
</tr>
<tr>
<td>.5</td>
<td>very very slight breathlessness (just noticeable)</td>
</tr>
<tr>
<td>1</td>
<td>very slight breathlessness</td>
</tr>
<tr>
<td>2</td>
<td>slight breathlessness</td>
</tr>
<tr>
<td>3</td>
<td>moderate breathlessness</td>
</tr>
<tr>
<td>4</td>
<td>somewhat severe breathlessness</td>
</tr>
<tr>
<td>5</td>
<td>severe breathlessness</td>
</tr>
<tr>
<td>6</td>
<td>very severe breathlessness</td>
</tr>
<tr>
<td>7</td>
<td>very, very severe breathlessness (almost max)</td>
</tr>
<tr>
<td>8</td>
<td>maximum breathlessness</td>
</tr>
</tbody>
</table>
Dyspnea and exercise

- May limit ability to exercise at higher workloads
- Increased work of breathing may cause high heart rate
- Pursed lip breathing should be encouraged
- Supplemental oxygen is indicated during exercise to maintain SpO₂ above 90%

Lung transplantation

- Patients considering or listed for lung transplantation should exercise regularly to maximize:
  - Flexibility
  - Strength
  - Mobility and endurance
- 6 minute walk test distance of less than 400 meters is associated with poor prognosis

Factors that influence adherence with exercise program

- Knowledge
- Motivation
- Support
- Access/availability
- Severity of disease
Questions

• What do you typically do for exercise?
• How many days a week do you exercise?
• How long do you exercise?
• Describe the level of your exercise:
  Light – my breathing is very comfortable
  Moderate – my breathing is deeper and more rapid
  Vigorous – my breathing is deep, rapid and it is difficult to speak in full sentences
• What are your barriers to exercise?
• Do you think exercise is important?
• Do you want to exercise more than you currently are
• Do you have a plan to increase your exercise?

Common barriers

• Time
• Coughing
• Need for oxygen
• Not feeling well
• Lack of knowledge
• Limited access to equipment
• Lack of interest

Home Exercise Program

• Identify barriers to exercise
• Develop and discuss possible solutions
• Facilitate problem solving
• Set realistic goals
• Identify and recruit supports
• Provide feedback
• Prepare for setbacks
Case Studies

Infant case

- Tommy is an 8 week old infant with CF
  - diagnosed through new born screening
  - lives with parents and 2 year old sibling
  - patient’s mother has two cousins with CF
- Normal physical exam:
  - No signs of respiratory distress, RR 46 breaths/min, SpO2 100%
  - Afebrile, HR 154 bpm
  - BS clear throughout, no adventitious sounds
  - CXR: normal
  - Throat culture: normal upper respiratory flora, few staphylococcus aureus
- Medications:
  - Creon 3000 (one capsule per feeding)
  - Aquadek 1 ml per day

Infant case

- Assessment
  - respiratory status
  - developmental screening
- PT intervention
  - parental instruction in chest PT
  - discussion of future options for airway clearance treatments
  - discussion of importance of exercise and active lifestyle
Infants

- Encouraged “tummy time” and reaching in prone for developing neck and back muscle strength antigravity
  - during times of relatively empty stomach to limit reflux problems
  - daily for 30 minutes (brief intervals) as Luke shows he is comfortable

Gaining Upright Posture

Encourage both back extensors and weight bearing

Case #2: Preschooler: “Ashley”

- 4-year-old girl with CF diagnosed at birth, BMI is at the 50th percentile
- She enjoys swinging, running and climbing on her backyard play set and dancing to the radio. She attends pre-school 3 mornings a week.
Exercise assessment

Ashley is asked to run a short distance in the clinic hallway, climb a flight of stairs and do 5 jumping jacks.
She is assessed via pulse oximetry at rest and following each activity as well as being asked about her breathlessness and noting any coughing with activity.
Her responses are normal and age-appropriate.

Ashley’s Prescription

• Ashley and her parents are encouraged to make physical activity a habit for Ashley and the family as a whole.
  – This is a critical time for setting activity patterns.
  – She has normal capacity for exercise at this point.

• She and her family are especially encouraged to incorporate jumping for bone density, climbing and monkey bar hanging for chest and shoulder flexibility, trampolining or bouncy-ball use for encouraging deep breathing.

• Physical activity is recommended daily for 20-30 minutes, but to be done in short bursts of 1-2 minutes, as this is a developmentally appropriate pattern.

Child-sized Equipment
Case: Pre-adolescent: Hunter

- Hunter is an 11-year-old boy diagnosed at age 1 year. He is in good nutritional condition. FEV₁ is 80% predicted.
- Mom reports he is very active, including “running around”, playing soccer and bike riding.
- He does activity every day that the weather allows. He also plays the “Wii” on inclimate days (boxing, tennis games)

Assessment

- Incremental Shuttle Walk Test
  - He performs the Incremental Shuttle Walk Test, completing level 10. Ending HR was 192 bpm (resting 102). His dyspnea and leg fatigue ratings were both 3/10. Occasional nonproductive cough noted during test, no desaturation.

Hunter’s Prescription

- Exercise prescription: Encouraged continued involvement with sports, especially those that can be done in the winter season.
- Watch sitting posture!
Posture Correction with Seating Change

Adolescent Case

Transplant Case: Mr. M
- The patient is a 42 yo male with CF s/p cadaveric double lung transplantation 1 year ago.
- Declining function for about 2 years prior to the transplant.
  - Still was quite active with a resistive exercise program and (with some PT coaxing) an aerobic exercise program on a stationary bike with supplemental O2 (4L) up to 2 weeks prior to transplant
  - Was quite active prior to that with regular jogging, tennis and golf
- PMH: CF (pancreatic insufficient), CFRD, pulmonary HTN (prior to tx- now resolved), osteopenia and renal dysfunction
Patient #4: Mr. M

• Since the transplant:
  – He has returned to his prior resistive strengthening program
  – Walking on the treadmill for 30 minutes 4-5x/wk at 3.5-4.8 mph
  – Aerobic activity is currently limited by right medial foot pain

Patient #4: Mr. M

• Musculoskeletal
  – Posture
    • Protracted shoulder girdle, increased thoracic kyphosis, increased lumbar lordosis, anteriorly seated humeral heads and UE's IR at rest standing
  – ROM
    • Grossly WNL except:
      – DF: R 0 degrees; L 4 degrees
      – HS: R 30 degrees; L 45 degrees (both in SLR position)
  – Strength
    • Grossly WNL except:
      – B Hip Flexion 3+/5, Hip ABD 4/5, Hip ADD 3+/5

Patient #4: Mr. M

• Integument
  – WNL except for the nicely healed clamshell scar and chest tube site incisions
• Neurologic
  – Grossly WNL as observed with functional activity
Patient #4: Mr. M

- Cardiopulmonary
  - VS: HR 81  BP 112/64  RR 16  O₂Sat 98% RA
  - Auscultation: CTA
  - Breathing pattern: Coordinated, symmetric, without accessory muscle use and good diaphragmatic expansion

Patient #4: Mr. M

- Cardiopulmonary
  - Aerobic capacity: 6MWT

<table>
<thead>
<tr>
<th>Total Distance</th>
<th>615 Meters (3.8 mph)</th>
<th>119% of predicted distance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Predicted Distance</td>
<td>518 Meters predicted (3.2 mph)</td>
<td></td>
</tr>
<tr>
<td>Limiting Symptom</td>
<td>R foot pain and B anterior shin pain</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Hemodynamic Response</th>
<th>(see image)</th>
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</thead>
<tbody>
<tr>
<td>Pre</td>
<td>HR 91</td>
</tr>
<tr>
<td>Peak/Post</td>
<td>HR 118</td>
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</tbody>
</table>

Healthy Aging Case
Adult case

- Donald is a 52 year old man with CF
  - by report is relatively symptom free
  - baseline PFT: FVC 78%, FEV1 54% of predicted normal values
  - active lifestyle, cycling regularly, working full time
  - history of CFRD
  - medications include: Pulmozyme, Cayston/TOBI (alternating months), azithromycin, albuterol, pancreatic enzymes, insulin

- Presented with decline in pulmonary status:
  - PFT: FVC 52%, FEV1 27%
  - CT scan revealed severe bronchiectasis of right upper lobe
  - sputum culture positive for stenotrophomonas maltophilia;
  - cough productive of sputum each morning
  - reports exercise is difficult due to dyspnea

- Hospitalized to initiate course of IV antibiotics; airway clearance treatments.

Initial GXT – while inpatient

<table>
<thead>
<tr>
<th>stage</th>
<th>speed (mph)</th>
<th>grade (%)</th>
<th>duration (min)</th>
<th>METS</th>
<th>HR (bpm)</th>
<th>BP (mmHg)</th>
<th>SpO2</th>
<th>RPE (1-10)</th>
<th>RPD (1-10)</th>
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<tr>
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<tr>
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<td>87</td>
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<tr>
<td>recovery</td>
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<td>20</td>
<td>97</td>
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</tbody>
</table>

Home exercise Program

Warm up by progressively increasing the speed on the equipment or walking faster for 5-10 minutes. RPE should be at 1-2 METS. Increase your walking speed until you feel like you are working at a RPE of 3-4/10 or stay within the range of 2.4-3.1 METS when using treadmill or other equipment. Continue walking at this speed for 20-30 minutes.

Cool down by progressively slowing your walking or equipment speed for 5-10 minutes. RPE should slowly decrease to 1-2/10.

Monitor oxygen saturation while exercising and use supplemental oxygen, 1-2 liters/minute to keep above 90%.

* Over following 6 months PFT continued to improve:

<table>
<thead>
<tr>
<th>stage</th>
<th>FVC</th>
<th>FEV1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previous baseline</td>
<td>78%</td>
<td>54%</td>
</tr>
<tr>
<td>Initial presentation</td>
<td>52%</td>
<td>27%</td>
</tr>
<tr>
<td>6 weeks after hospitalization</td>
<td>61%</td>
<td>36%</td>
</tr>
<tr>
<td>3 months after hospitalization</td>
<td>65%</td>
<td>38%</td>
</tr>
<tr>
<td>6 months after hospitalization</td>
<td>65%</td>
<td>39%</td>
</tr>
</tbody>
</table>

* Donald continued home exercise program:
  - Cycling for 30 minutes, 3-5 times/week
  - Weight lifting, 3 times/week
  - Yoga, 2 times/week
  - Acapella daily; CPT 5 times each week; inhaled hypertonic saline initiated
  - Reported continued improvement in symptoms
### GXT at 6 month follow-up

<table>
<thead>
<tr>
<th>stage</th>
<th>speed (mph)</th>
<th>grade (%)</th>
<th>duration (minutes)</th>
<th>METS</th>
<th>BP (120/86)</th>
<th>RR (bpm)</th>
<th>SpO2</th>
<th>RPE (1-10)</th>
<th>RPE (1-10)</th>
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</thead>
<tbody>
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<td>97</td>
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<td>95</td>
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<tr>
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</table>

**Home Exercise Program**

Intensity increased to working at a RPE of 3-4/10 or within the range of 3.5-4.5 METS when using treadmill or other equipment, 30 minutes.

### PT Reimbursement Issues

**Challenges in Reimbursement**

- Need for prior authorization for PT services
  - Have patient get lunch or other time-consuming activity between clinic visit and PT evaluation to allow time to get authorization
  - Primarily an issue with HMOs
- Co-pays
  - Need to convince patient/care team that PT services are important to their function
Important Aspects of Documentation

- Demonstrate skilled intervention (trained hands)
  - Use evidence-based tests & measures
  - Set goals based on objective findings
- Diagnosis based on movement/functional limitations
  - Not enough to list "CF" as PT diagnosis (list complicating factors)
  - Must be tied to patient's specific functional issues
- Document education/home program
- Indicate why PT is medically necessary

Thank You

www.cftrust.org.uk
www.cfww.org