PHYSICAL THERAPIST
MANAGEMENT OF
NEUROMUSCULAR
CONDITIONS

CMT, LGMD, POMPE, IBM

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4 NEUROMUSCULAR CONDITIONS

• Charcot Marie Tooth/ Hereditary Sensory Motor Neuropathy
• Limb Girdle Muscular Dystrophy
• Pompe Disease
• Inclusion Body Myositis
MULTI-DISCIPLINARY REHABILITATION IS ESSENTIAL
PHYSICAL THERAPIST IS AN IMPORTANT PART OF TEAM

• Usually goal is to preserve and optimize function
  – Maximize quality of life (QOL)
  – Not necessarily to improve strength dramatically

• Common interventions include:
  – Strengthening and aerobic training
  – Stretching
  – Mobility aides and orthotics
  – Balance training
  – Safety evaluations
CHARCOT MARIE TOOTH
HEREDITARY MOTOR SENSORY NEUROPATHY
PT CONSIDERATIONS

• Multidisciplinary care (McCorquodale 2016)
  – Maintain function and QOL
  – Assess and be proactive regarding work

• Mobility aides and orthotics (Bird 2018)
  – May require canes/ crutches; < 5% require WC
  – AFOs

• General exercise – aerobic and resistance (Sman 2015)
  – Guidelines unknown, but likely safe at mod intensities and/or unaffected muscles
  – Some recent evidence that high intensity
PT CONSIDERATIONS

• UE function, fine motor
  – Refer out to OT if needed for adaptive equipment

• Pain
  – Neuropathic vs MSK
  – Joint preservation/ MSK derangement prevention

• Balance
  – Fall risk, safety

• Home/ work modifications
PES CAVUS AND FOOT CARE

- Manual self-stretching of toes and ankle
- Hammer toe pads
- Arch supports
- Footwear considerations/ toe boxes
- Skin protection
  - Daily foot checks
  - Address blisters, red spots immediately!
LIMB GIRDLE MUSCULAR DYSTROPHY
PT CONSIDERATIONS

• No clear guidelines on best type of exercise or intervention

• Long-term management
  – To maximize QOL, function, and independence
  – Spine deformities/ scoliosis
  – Anticipate need for assistive devices
PT CONSIDERATIONS

• Resistance training
  – Both higher and lower intensity resistance training may improve strength in arms over 6 months
    • But not clear if that also improves function

• Aerobic and endurance training
  – May also improve aerobic capacity
    • And also may improve functional performance such as walking

(Siciliano 2015)
PT CONSIDERATIONS

• Anti-gravity or supported treadmill training (Berthelsen, Sczesny-Kaiser)
  – 3 patients over 8 weeks
    • Modest improvements in gait speed and endurance (up to 80% support)
  – 10 patients over 10 weeks did walking and resistance
    • Walking and weight bearing resistance exercises
    • Modest improvements in strength and function
POMPE DISEASE
PT CONSIDERATIONS

• Some evidence that sub-maximal aerobic training may improve muscle strength and function (Cupler 2012)
  – 60-70% of HR Max
  – Closely monitored for HR, RR, and effort level

• Less evidence for strength training but proximal weakness (trunk and hips) tends to be more severe then in legs (Cupler 2012, Case 2006)
  – Hip abduction and flexion weakness are strong predictors of slower walking speeds (Ada 2018)
  – Interventions that target trunk and hip muscle endurance or strength may be especially helpful
  – Interventions that stabilize versus aggressively strength may be particularly helpful
PT CONSIDERATIONS

• Bracing and assistive devices are important considerations
  – Maintain independence
  – Prevent contractures or deformity

• Scoliosis prevention and management
  – Strengthening, stretching, bracing
  – Pain management

• Osteopenia and osteopersosis is common
  – Weightbearing exercise can be helpful in preventing/ managing
## MODIFIED BORG SCALE

<table>
<thead>
<tr>
<th>RPE Scale</th>
<th>Rate of Perceived Exertion</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td><strong>Max Effort Activity</strong></td>
</tr>
<tr>
<td></td>
<td>Feels almost impossible to keep going. Completely out of breath,</td>
</tr>
<tr>
<td></td>
<td>unable to talk. Cannot maintain for more than a very short time.</td>
</tr>
<tr>
<td>9</td>
<td><strong>Very Hard Activity</strong></td>
</tr>
<tr>
<td></td>
<td>Very difficult to maintain exercise intensity. Can barely breath</td>
</tr>
<tr>
<td></td>
<td>and speak only a few words</td>
</tr>
<tr>
<td>7-8</td>
<td><strong>Vigorous Activity</strong></td>
</tr>
<tr>
<td></td>
<td>Borderline uncomfortable. Short of breath, can speak a sentence.</td>
</tr>
<tr>
<td>4-6</td>
<td><strong>Moderate Activity</strong></td>
</tr>
<tr>
<td></td>
<td>Breathing heavily, can hold short conversation. Still somewhat</td>
</tr>
<tr>
<td></td>
<td>comfortable, but becoming noticeably more challenging.</td>
</tr>
<tr>
<td>2-3</td>
<td><strong>Light Activity</strong></td>
</tr>
<tr>
<td></td>
<td>Feels like you can maintain for hours. Easy to breathe and carry</td>
</tr>
<tr>
<td></td>
<td>a conversation</td>
</tr>
<tr>
<td>1</td>
<td><strong>Very Light Activity</strong></td>
</tr>
<tr>
<td></td>
<td>Hardly any exertion, but more than sleeping, watching TV, etc.</td>
</tr>
</tbody>
</table>
PT CONSIDERATIONS

- Weakness typical in knees, also hands
  - Stairs, curbs, sit-to-stand
  - Grip and ADLs
- Attempts to improve strength are indicated
  - But as weakness progresses, adaptations are usually needed
  - Knee braces, assistive devices, sit-to-stand assist, power wheelchairs
PT CONSIDERATIONS

- Resistance exercise *may or may not* improve strength,
  - *But* it can prevent loss of strength at least in the short term
- Regular aerobic exercise can improve aerobic capacity

According to a recent review:

- Exercise can improve ADLs, Function, Quality of Life
- Exercise can lower risk for many cardiovascular diseases prevalent in IBM including Diabetes and Hypertension
Higher intensity exercise in IBM

- Higher intensity strength training has been studied in people with IBM
  - Likely does not increase inflammation or damage muscles
  - May lead to improvements in strength and function
- Some studies are starting to examine Blood Flow Restriction
  - Cuff placed over limb to restrict blood flow, and training occurs at lower intensities
  - Results show no harm, but not clear it is effective

Jørgensen
GENERAL CONSIDERATIONS FOR EXERCISE
CMT, LGMD, POMPE, IBM
EXERCISE IS GENERALLY SAFE

• Under consultation of a physical therapist
• However no guidelines exist for any of these conditions
• Generally speaking aerobic exercise should be undertaken at submaximal levels
  – More intense exercise should only be done under supervision of PT and with clearance from MD
• Strength training is also likely safe at submaximal levels (2-3 sets of 10-15 reps)
  – In muscles that can already work against gravity
  – Avoiding muscle soreness or excessive fatigue
  – Overall goal is usually to maintain/preserve function, not to “bulk up”

DON’T OVERDO IT!

Physical Therapy
TYPICAL PLAN OF CARE

• Long-term management is key
• Regular check-ins, preferably in an interprofessional setting
• Periodic short episodes of PT as needed
  – To address acute changes, pain
  – To learn/ modify exercise programs
• Address mobility needs and help with planning ahead
SAFETY WITH EXERCISE

• As ability to stand and safely balance changes, exercises will need to be adapted

• There are many ways to exercise that do not include walking
  – Stationary arm and leg bikes
  – Recumbent steppers
  – Seated weight machines
  – Seated exercise classes
  – Pool therapy and aquatic therapy
  – Pilates and Yoga
BEING SEDENTARY IS A BAD OPTION

Couch potato

Exercise or activity that makes you feel good

Over-exercised potato
Our neuromuscular providers are involved in transformative research to improve lives.

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www.medschool.ucdenver.edu/neuromuscular/clinicaltrials