

PHYSICAL THERAPIST MANAGEMENT OF NEUROMUSCULAR CONDITIONS

CMT, LGMD, POMPE, IBM

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CU PHYSICAL THERAPY PROGRAM



Physical Therapy

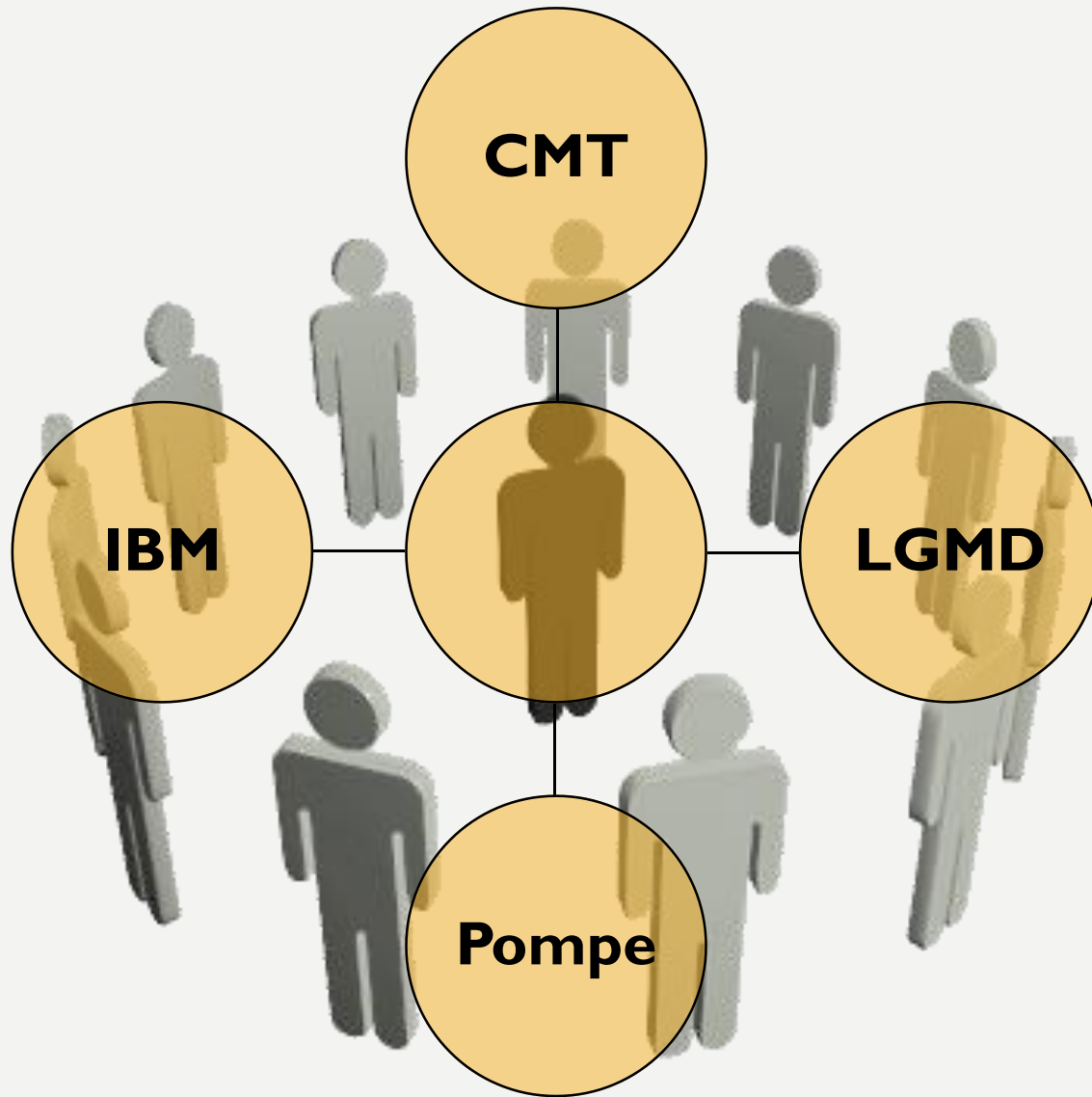
SCHOOL OF MEDICINE

**UNIVERSITY OF COLORADO
ANSCHUTZ MEDICAL CAMPUS**

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, Szczesny-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 than 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 strengthen may be particularly helpful



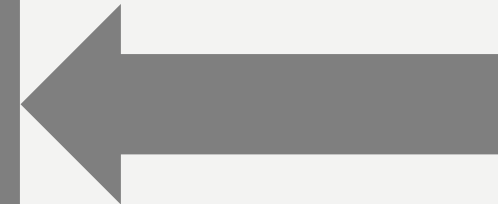
PT CONSIDERATIONS

- Bracing and assistive devices are important considerations
 - Maintain independence
 - Prevent contractures or deformity
- Scoliosis prevention and management
 - Strengthening, stretching, bracing
 - Pain management
- Osteopenia and osteoporosis is common
 - Weightbearing exercise can be helpful in preventing/ managing



MODIFIED BORG SCALE

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



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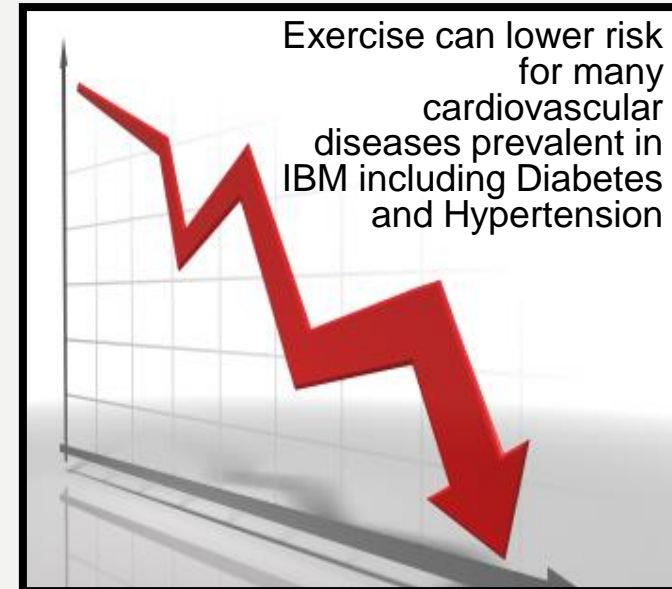
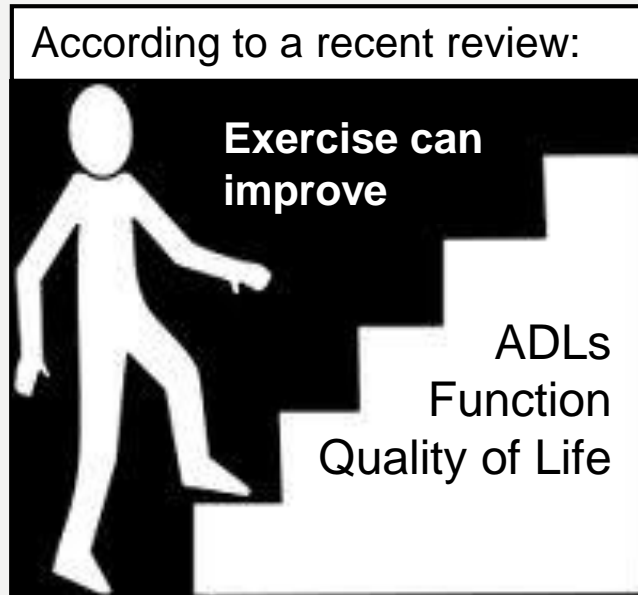
INCLUSION BODY MYOSITIS

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



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





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”



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

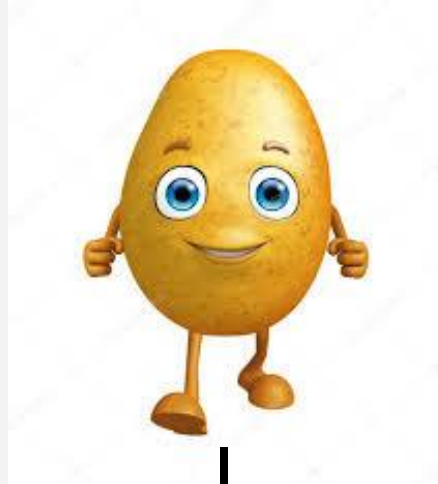
- As ability to stand and safely balance changes, exercises will need to be adapted
- There are many ways to exercise that do not include walking
 - Stationary arm and leg bikes
 - Recumbent steppers
 - Seated weight machines
 - Seated exercise classes
 - Pool therapy and aquatic therapy
 - Pilates and Yoga



BEING SEDENTARY IS A BAD OPTION



Couch potato



Exercise or
activity that
makes you
feel good



Over-exercised potato



Our neuromuscular
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