Myositis, Myopathy, Muscular Dystrophy and Charcot-Marie Tooth Disease: A Therapeutic Overview

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Objectives

- Recognize common signs and symptoms of Myositis, Myopathy, MD, and CMT that may lead to a patient presenting to OP Rehab
- Identify potential interdisciplinary and care collaboration needs for a patient with Myositis, Myopathy, MD, and CMT receiving OP Rehab
- PT: Understand considerations for evaluation and interventions including safe exercise and activity recommendations for this patient population
- OT: Discuss specific Occupational Therapy assessment, evaluation, and intervention techniques in an outpatient setting for this patient population

Myopathy – disorder affecting the channels, structure or metabolism of skeletal muscle.

Acquired

- Toxic
- Inflammatory
- Associated with other diseases

Hereditary

- Dystrophies
- Metabolic
- Myotonias

Common Muscle Symptoms

- Myalgia
- Weakness
  - Often proximal but not always
  - Lack sensory symptoms
- Cramps
- Exercise intolerance
- Atrophy
- Myoglobinuria – excess myoglobin in the urine
- Elevated CPK

Rhabdomyolysis - Acute muscle breakdown

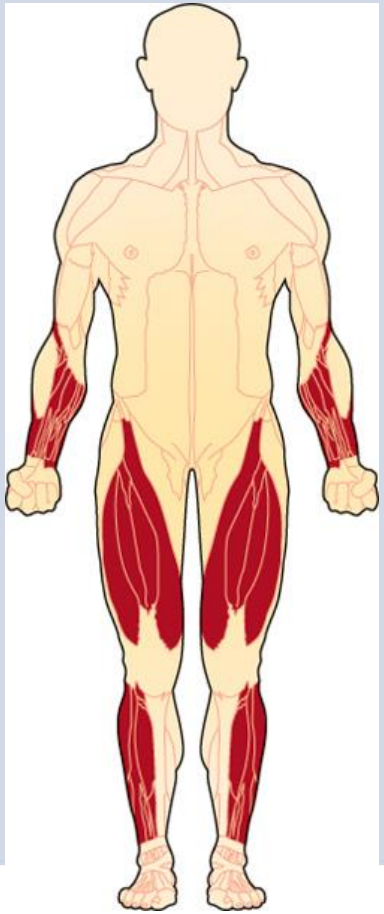
- Symptoms
  - Muscle Pain/Edema
  - Myoglobinuria
  - Elevated CPK (10K+)
  - Renal Failure
  - Electrolyte abnormalities
- Intense Exercise
  - Recruits/Athletes
  - Recurrent = Metabolic myopathies
- Injury/Compression
- Muscle Disease
  - Inflammatory

Acquired Myopathies

- Toxic
  - Drugs of Abuse
  - Prednisone
  - Statins
    - Myalgia and CK elevation
      - Resolves after discontinuation
    - Statin Induced Necrotizing Myopathy
      - Treat as autoimmune
- Systemic Diseases
  - Hypothyroid
  - Critical Care Myopathy
- Inflammatory
  - Dermatomyositis/Polymyositis
    - Elevated CPK
    - Proximal Weakness
    - Myalgia
    - Rash
  - Treatment
    - Prednisone
    - IVIG
  - Slow improvement/Remission

Inclusion Body Myositis

- Onset 50s
- Finger Flexors/Knee extensors
- Dysphagia
  - Cricopharngeal Hypertrophy
- Slowly progressive
  - Some rapid symptoms
- Treatment Resistant

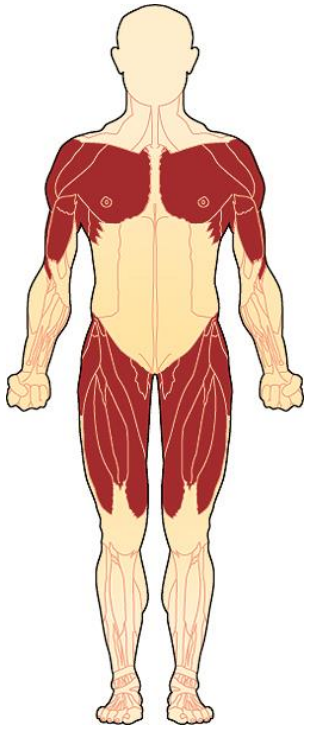


Inherited Myopathy – General Principles

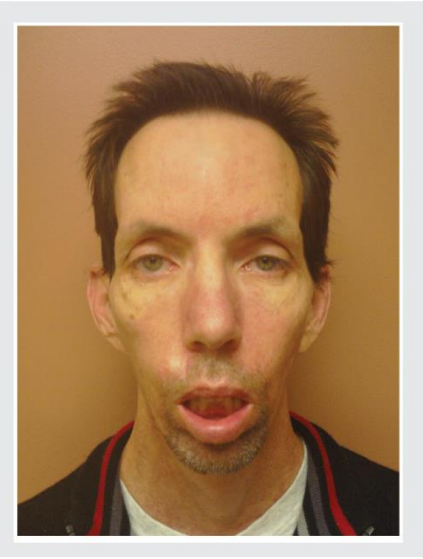
- Variable Patterns and Onset
- Contracture Possible
- Slowly Progressive
- Cardiac or Respiratory Involvement
- Diagnosis
  - Muscle Biopsy
  - Genetic Testing
- Rarely Treatable

Inherited

- Limb Girdle Dystrophies (LGMD)
  - 25 Subtypes
    - AD/AR
  - Proximal Weakness
  - Slowly Progressive
  - Dx – genetics/biopsy
  - Supportive Treatment



- Myotonic Dystrophy 1
  - 2 types
- Myotonia
- Distal Weakness
- Facial Weakness/Atrophy
- Dysarthria/Dysphagia
- Systemic
  - Mood
  - Cognition
  - Cataracts
  - Frontal Balding
  - Diabetes
  - Cardiac Conduction (90%)



Summary Myopathies

Treatable Inflammatory Diseases	Monophasic Insults/Injuries	Inherited
• Dermatomyositis • Polymyositis • Statin-Induced Necrosis • Improvement with Relapses	• Compression • Toxic • Critical Care • Improvement	• LGMD, MD • Variable Presentations • Slowly Progressive

Know their myopathy and course
- Respiratory or cardiac involvement
- Exercise intolerance

Address needs for specific disease and its stage

Myositis, Myopathy, Muscular Dystrophy: General Therapeutic Approach

- Compensation strategies for energy conservation
- Low to moderate intensity strengthening and aerobic exercises recommended
- Bracing/splinting
- Assistive devices and home modifications for safety
- Counseling resources for support and coping strategies

Myositis, Myopathy, Muscular Dystrophy: General Precautions

- Fall risk
- Contractures
- Fatigue
- Pain
- Respiratory Dysfunction
- Bone health/fracture risk
- Dysautonomia/Orthostatic hypotension
- Depression/anxiety/stress
- AD recommendations

OT screening

- Fine motor coordination
- UE weakness
- Difficulties with ADLs
- Issues with home accessibility, use of technology or work/occupation

SLP screening

- Facial weakness
- Impaired Swallowing
- Impaired cognition

PT Evaluation Considerations

- Balance: FGA vs BERG
- Gait: 10MWT
- Endurance: 2MWT vs 6MWT
- Strength: 5XSTS, MMT, dynamometer
- ROM

PT Treatment Considerations in Myositis, Myopathy, Muscular Dystrophy

Exercise has been found to improve QOL, ROM, and flexibility.

- Low to moderate intensity aerobic training
- Low to moderate intensity strength training shown to improve strength without negative effects
  - Moderate resistance strength training appears to be safe with supervision and can reverse disuse weakness
  - High intensity exercise shown to increase fatigue
- Concentric exercises preferred over eccentric exercises
- Pairing aerobic and resistance training
  - Level II evidence benefitting over aerobic conditioning alone

PT Treatment Considerations in Myositis, Myopathy, Muscular Dystrophy

Consider proximal versus distal weakness patterns

- Inclusion body myositis
  - Slow progressing muscle weakness, especially in finger flexors, quads, and distal LE
  - Drop foot
    - AFO recommendations
  - Potential need for powered mobility with proximal hip weakness
    - Referral for Wheelchair Clinic
  - Home modifications
    - Elevated seat heights due to difficulty with sit to stand
    - Ramped entrances
    - Referral for AT clinic

PT Treatment Considerations in Myositis, Myopathy, Muscular Dystrophy

Goal Oriented/Task Specific Approach	• Discuss patient centered goals for functional task focus
Balance training	• Improve function and reduce risk of falls
PROM/Stretching	• Lower extremity weight bearing can reduce risk of contractures
Assessment for assistive devices	• Cane, Walker, Potential wheelchair clinic evaluation
Bracing	• AFO for foot drop • Headmaster for Head drop

PT POC Considerations in Myositis, Myopathy, Muscular Dystrophy

- Limited research on POC recommendations
  - 50–85% of the predicted maximal heart rate
  - 15–45 min per session
  - 2–4 sessions per week
  - 8– 18 weeks
- 11-22% improvements in ADL scores
- 5-20% reduction in mean exercise heart rate
- 5-30% increase in mean maximal VO2
- No evidence of change in CK levels with reduction in lactate levels

(Aboussouan, 2009)

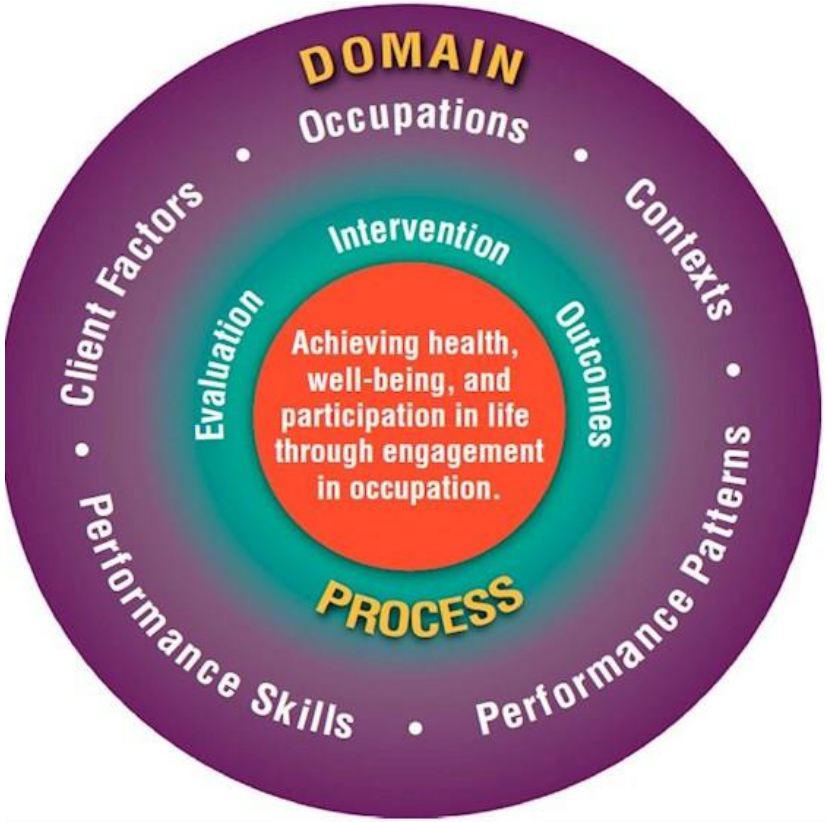
Occupational Therapy for Myositis/Myopathy/MD - POC Considerations

- ADL/IADL performance, modifications and adaptive equipment use
- Caregiver participation and burden
- Splinting/orthotics
- AT/Wheelchairs
- Exercise/Activity Tolerance
- Sleep
- Respiratory function
- Home set-up
- Cognitive skills, sensory regulation
- FMC
- Swallowing/Oralmotor function
- Academics and transition

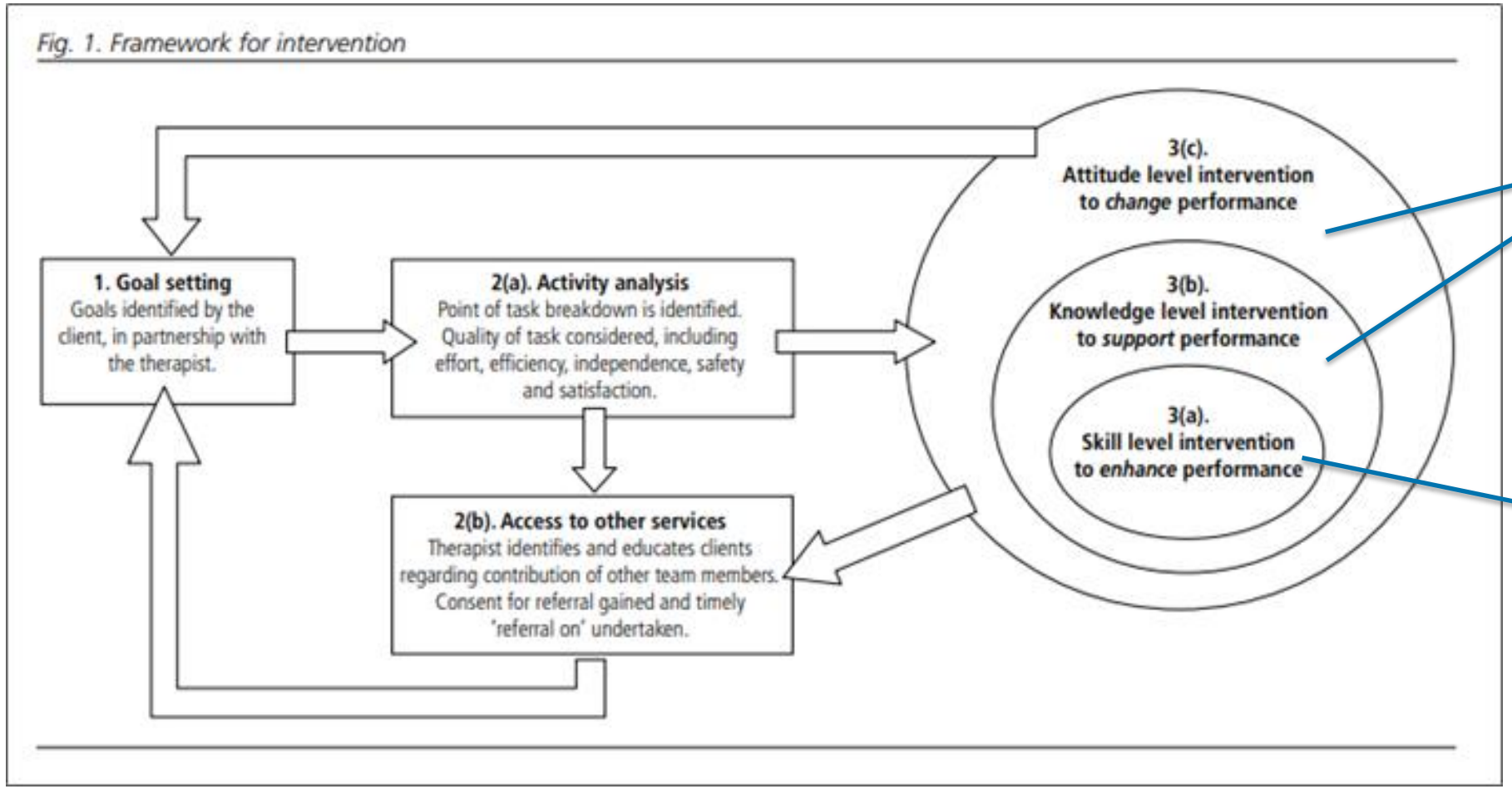
Note: Research in this area is limited and older.

Occupational Therapy for Myositis/Myopathy/MD – General Treatment Approach

- Activity Analysis
- Balance between remediation and compensation
- Client-centered goals, occupation-based, addressing quality of life
- Prevention of secondary deficits (ROM loss, learned helplessness, isolation)
- Outcome Measures: MMT, dynamometry, COPM, ADL performance, Muscular Dystrophy Functional Rating Scale, Functional Index in Myositis, Borg, timed motor performance, Brooke UE Scale, Abilhand Scale



Occupational Therapy for Myositis/Myopathy/MD – ADL/IADL Performance



Compensation

Remediation

Occupational Therapy for Myositis/Myopathy/MD – ADL/IADL Performance

- Focus on task modification and adaptive equipment balanced with remediation depending on chronicity
- Utilization of activity analysis
- Energy conservation / Health Management
  - Modified Parkwood Pacing or other intentional tracking system
- Adaptive equipment
  - Focus on AE recommendations that can be consistently used even with changing function
  - Changing needs for DME, prepare for lifelong home set-up changes in some
- Connections with organizations for equipment, support groups, resources, applications

Parent Project Muscular Dystrophy

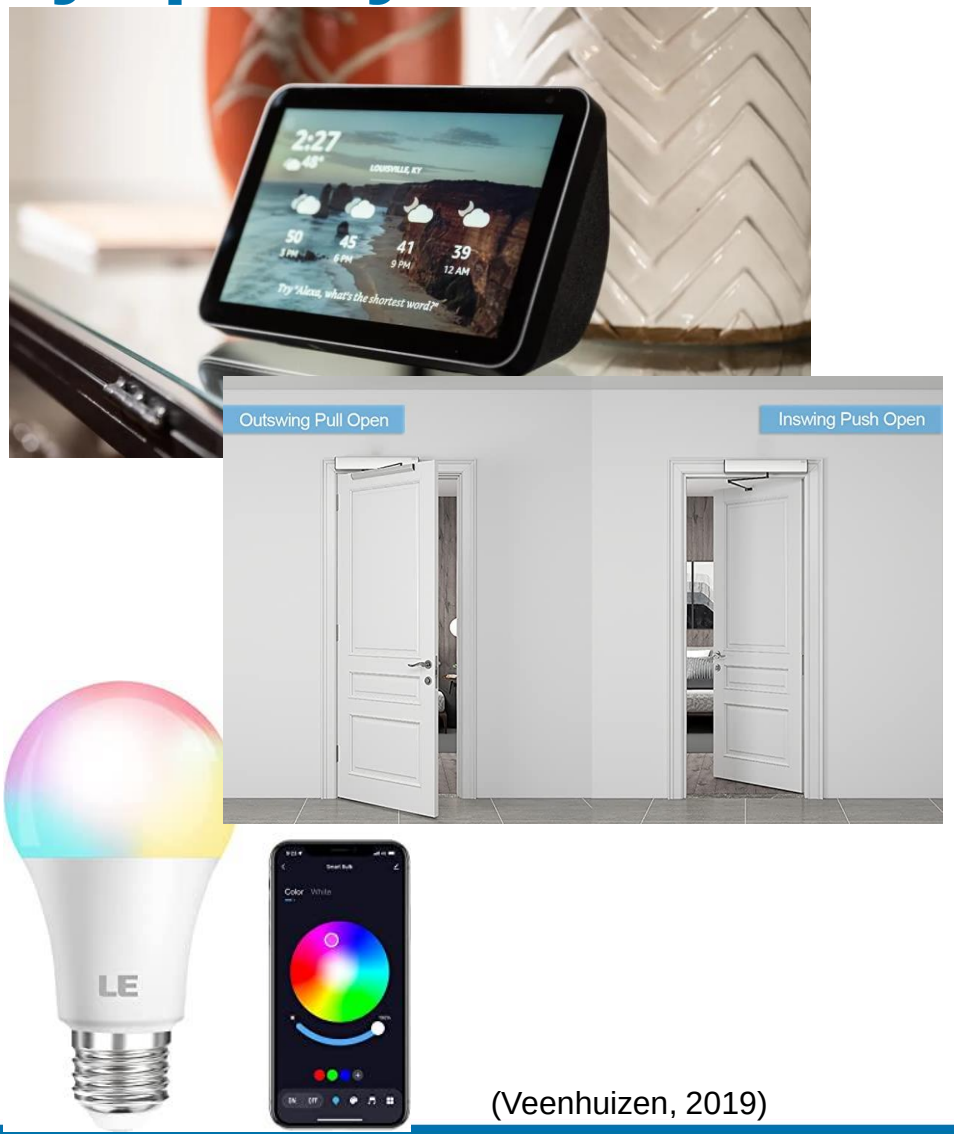
MDA Muscular Dystrophy Association

The Myositis Association

(Veenhuizen, 2019)

Occupational Therapy for IBM/Myopathy/MD – Assistive Technology

- Referral to AT clinic
- Use of phone and computer modifications
- Use of EADL devices for safety in home (accessing lights, emergency services, door openers)
- Switch control, joystick control of computer, voice activation
- Low-tech adaptive equipment (cups, writing utensils, lap trays)
- Connection with BVR, DODD for education and workplace accommodations and technology



(Veenhuizen, 2019)

Occupational Therapy for Myositis/Myopathy/MD – ADL/IADL Performance

- Home modification recommendations
  - Safe At Home Checklist
  - Home Safety Self Assessment Tool (HSSAT)
  - In-Home Occupational Performance Evaluation (I-HOPE) Kit
  - The Safety Assessment of Function and Environment for Rehabilitation (SAFER) Tool
  - Use of video visits or photos

Safe AT HOME Checklist

Created in partnership with the Administration on Aging and the American Occupational Therapy Association

Rebuilding Together
1899 L Street NW, Suite 1000
Washington, DC 20036
800-473-4229
www. rebuildingtogether.org

Rebuilding Together has long recognized that greater attention must be given our elderly population, so they may age-in-place and safely in their homes. We have also built lasting national partnerships with Area Agencies on Aging, AARP, American Occupational Therapy Association, National Association of Home Builders, National Council on Aging, and others.

Use this list to identify home safety, fall hazards and accessibility issues for the homeowner and family members. Home safety, fall prevention and accessibility modification interventions on the reverse side of this page can help prioritize your work. Underline or use a highlighter to note. problems and add comments.

1. EXTERIOR ENTRANCES AND EXITS
- ☐ Note condition of walk and drive surface; existence of curb cuts
- ☐ Note handrail condition, right and left sides
- ☐ Note light level for driveway, walk, porch
- ☐ Check door threshold height
- ☐ Note ability to use knob, lock, key, mailbox, peephole, and package shelf
- ☐ Do door and window locks work easily?
- ☐ Are the house numbers visible from the street?
- ☐ Are bushes and shrubs trimmed to allow safe access?
- ☐ Is there a working door bell?

2. INTERIOR DOORS, STAIRS, HALLS
- ☐ Note height of door threshold, knob and hinge types; clear width door opening; determine direction that door swings
- ☐ Note presence of floor level changes
- ☐ Note hall width, adequate for walker/wheelchair
- ☐ Determine stair flight run: straight or curved
- ☐ Note stair rails: condition, right and left side
- ☐ Examine stairway light level
- ☐ Note floor surface texture and contrast
- ☐ Note if clutter on stairway

3. BATHROOM
- ☐ Are sink basin and tub faucets, shower control and drain plugs manageable?
- ☐ Are hot water pipes covered?
- ☐ Is mirror height appropriate, sit and stand?

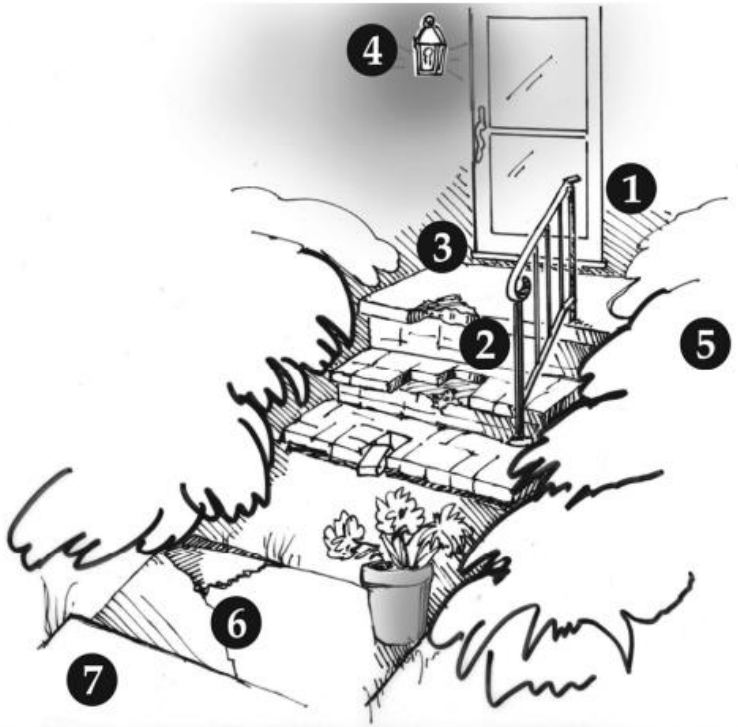
8. TELEPHONE AND DOOR
- ☐ Phone jack location near bed, sofa, chair?
- ☐ Able to get phone, dial, hear caller?
- ☐ Able to identify visitors, hear doorbell?
- ☐ Able to reach and empty mailbox?
- ☐ Wears neck/wrist device to obtain emergency help?
- ☐ Is there an answering machine?
- ☐ Is there a wireless phone system?

9. STORAGE SPACE
- ☐ Able to reach closet rods and hooks, open bureau drawers?
- ☐ Is there a light inside the closet?

10. WINDOWS
- ☐ Opening mechanism at 42 inches from floor?
- ☐ Lock accessible, easy to operate?
- ☐ Sill height above floor level?
- ☐ Are storm windows functional?

11. ELECTRIC OUTLETS AND CONTROLS
- ☐ Sufficient outlets?
- ☐ Are there ground fault outlets in kitchen and bathroom?
- ☐ Light switch at the entrance to each room
- ☐ Outlet height, wall locations
- ☐ Low vision/sound warnings available?
- ☐ Extension cord hazard?
- ☐ Are there any uncovered outlets or switches?

Entrance to Front Door and Front Yard



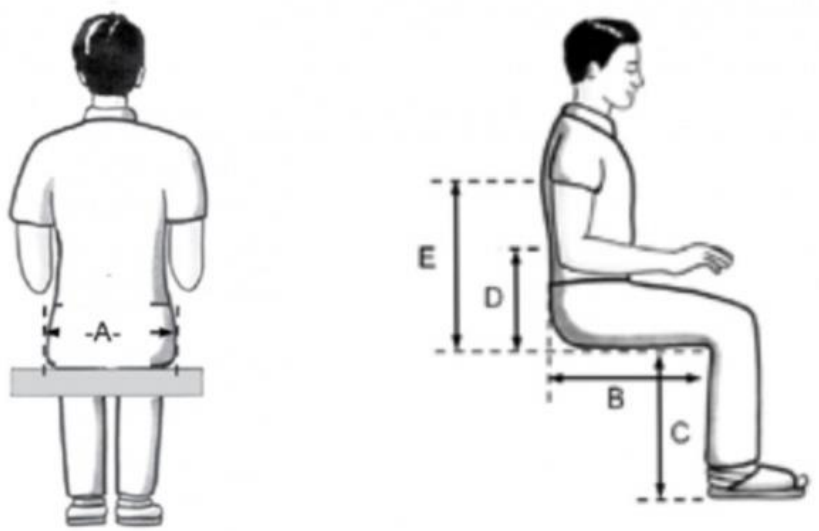
Are these problems present in your home?
If yes, please check in the box next to the problem,
then add the total number of checks and enter it in the box below.

- ☐ 1. Lack of railings
- ☐ 2. Unsafe steps (too steep/cracked/chipped)
- ☐ 3. Unmarked or raised threshold
- ☐ 4. Lack of lighting at night
- ☐ 5. Lack of a ramp for a wheelchair
- ☐ 6.Uneven/cracked Pavement
- ☐ 7.Ice or snow on driveway/walkway
- ☐ Other_____

Number of total problems ☐

Occupational Therapy for Myositis/Myopathy/MD – Wheelchairs

- Referral to wheelchair clinic
- Refer especially if:
  - High fall risk (Tinetti, Tug, Berg, 30s Sit to Stand)
  - Progressive change in function
  - Unable to utilize assistive device consistently due to UE/LE function
  - Significant fatigue
  - Limited in independence in ADL/IADL due to mobility limitations
  - Utilizing a wheelchair (power or manual) that does not fit the client

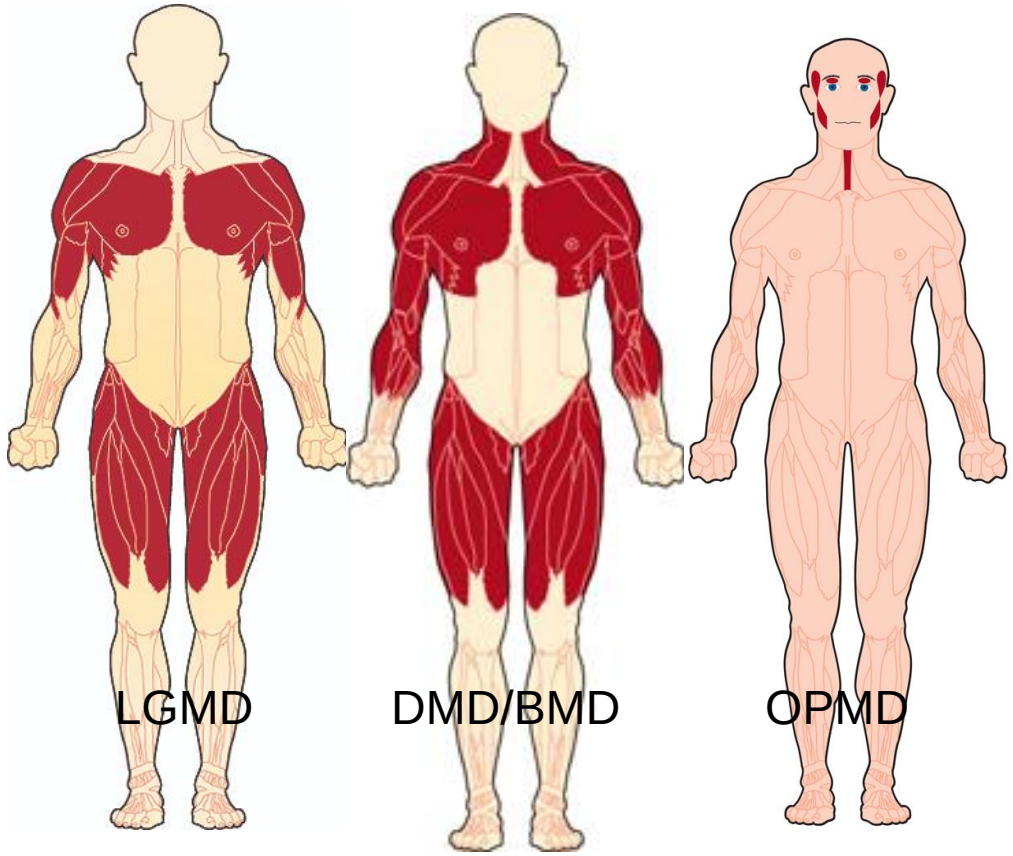


Occupational Therapy for Myositis/Myopathy/MD – BVR / Employment / Academics

- Significantly lower employment rates for people with MD than the able-bodied population (Carter 2010)
- Referral to BVR or engaging with workplace accommodations or employment training
- BVR and DODD can support funding of technology or adaptations needed for participation
- MDA Young Adults Program – Supports transition age young adults into workplace and academics

Occupational Therapy for Myositis/Myopathy/MD – Weakness Presentations

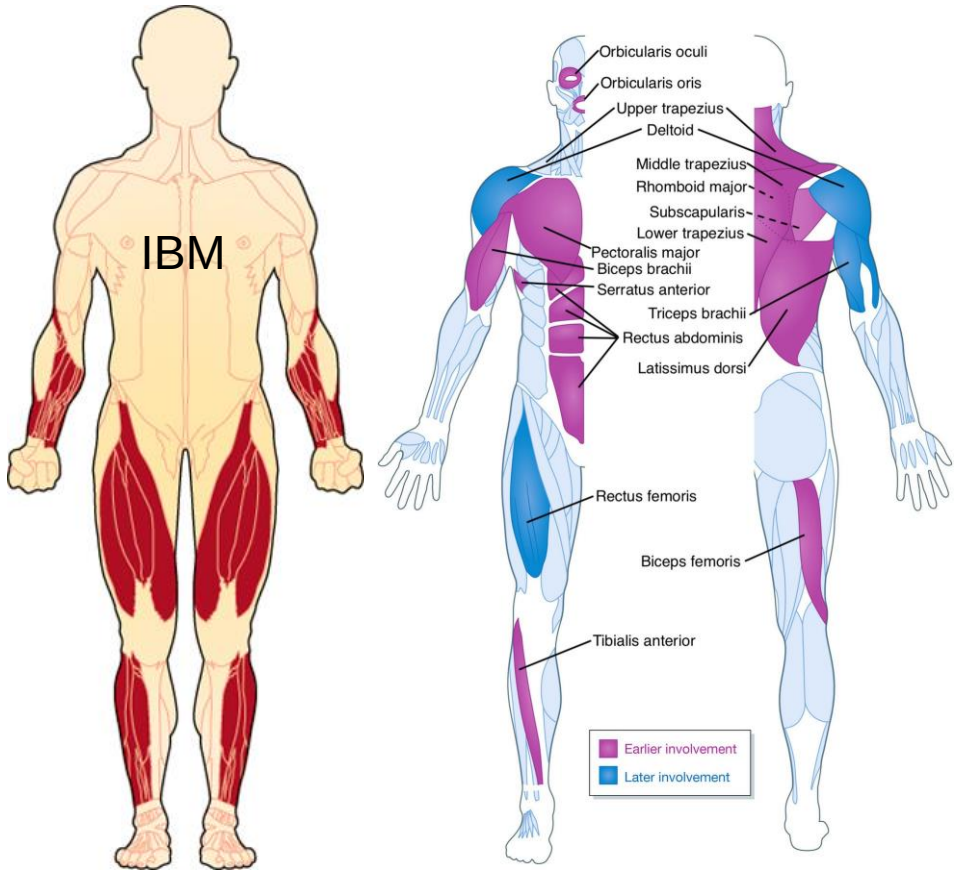
- LGMD – Symmetrical weakness, less severe than LE weakness in UE, proximal>distal
- DMD/BMD- Trunk weakness with progression to LE then UE, often with scoliosis and cardiac considerations; Sensation and FMC can be affected
- Oculopharyngeal - typically effects eyes and throat first, with shoulders, upper legs, and hips possibly showing weakness later



(Cup et al., 2007)

Occupational Therapy for Myositis/Myopathy/MD – Weakness Presentations

- Myopathy/Myositis - Varies depending on type of disorder; Typically muscle weakness, cramps, proximal and symmetrical, muscle wasting. At times can have respiratory and bulbar onsets. Varied prognosis. Many are progressive.
- IBM - Progressive weakness in finger flexors, quads, distal LE
- FSHMD – Proximal>distal UE weakness, facial weakness, affects deltoid, lats, ER, extensor muscles>flexors



(Cup et al., 2007)

Occupational Therapy for Myositis/Myopathy/MD – Strengthening / Stretching

- See PT considerations for strengthening
  - Myopathies/Myositis: Moderate to low intensity resistance exercises with gentle and safe aerobic exercise (walking and cycling), focus on concentric > eccentric (Abresch, 2009; Adaikina, 2020; Hart, 2014)
  - IBM: Slow progressive muscle weakness in fingers, quads, distal LE
  - MD: Submaximal exercise and self-paced endurance
  - General consideration: Perform submaximal activity to avoid overuse and fatigue
- PROM/stretching for reduction of deficits
  - Caregiver and patient education on full ROM stretches and use of splints as needed; goal of prevention of contractures
  - Weight-bearing if safe (Carter 1997)
  - Hold for 15s, release, complete x4 times for 60s total

(Cup et al., 2007)

Occupational Therapy for Myositis/Myopathy/MD – Splinting and Orthotics

- Nighttime orthotics for prolonged UE stretch
  - Resting hand, wrist cock-up, cone splint
  - Focus on splinting to accommodate or prevent deformities and provide a slight stretch but not lead to risk of further injury (See photo)
- Shoulder supports
  - Ottobock, Zenkeyz
- Cervical supports
  - Headmaster, soft collars, wheelchair positioning considerations

(Datta Gupta, 2009; Hasegawa, 2015)

Occupational Therapy for Myositis/Myopathy/MD – Splinting and Orthotics

- Robotics
  - Robotic options available but often not covered by insurance
  - Can be used as an intervention or a daily occupational tool
    - Myomo, Jaco, Armeo, ReoGo
- Mobile arm supports (specifically for LGMD)
  - Demonstrated overall improved upper limb function and independence (Cruz 2011)
  - Training required for optimal positioning and reduction of further deficits in shoulder
  - Powered by resistance bands, tension, electronically powered
    - Jaco, Jaeco, Saebo

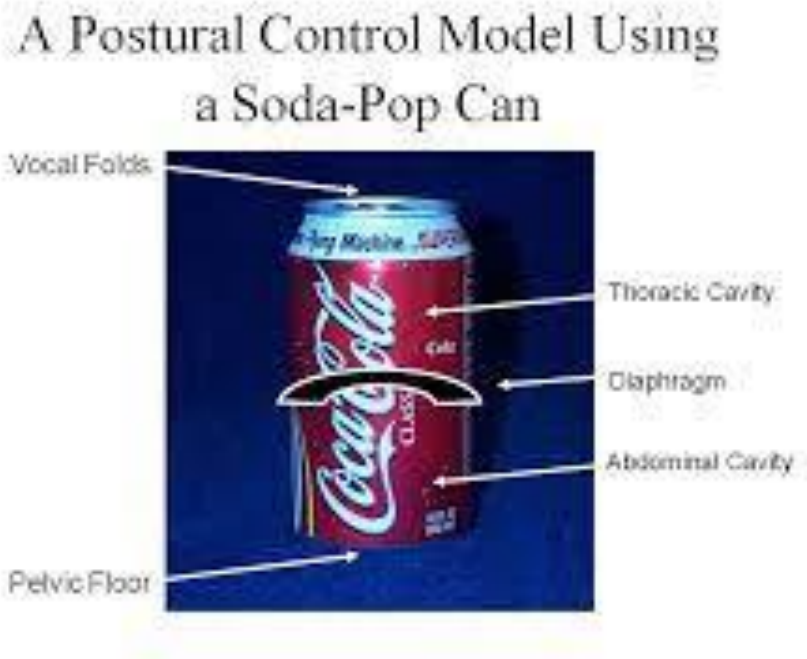
(Datta Gupta, 2009; Hasegawa, 2015)

Occupational Therapy for Myositis/Myopathy/MD – Modalities

- Ice, ultrasound, manual mobilization for less pain and improved functional recovery with myositis traumatica (Buselli 2010)
- Heat, ultrasound, ice for myositis ossificans for improved pain and mobility, but noted concern for increased bone growth (De C 1992)
- Heat and cold for improved tendon plasticity and acute inflammation for inflammatory myositis (Hicks 1998)

Occupational Therapy for Myositis/Myopathy/MD – Respiratorv Function

- Importance of pop can visual
  - Closed system
  - Intra-thoracic pressure > atmospheric pressure
  - Pelvic floor and vocal folds serve as pressure regulators; Diaphragm decreases intrathoracic pressure, increases abdominal pressure and allows for breathing
- Typically, patients following one of the following patterns:
  - Diaphragm (Low abs)
  - Lateral (Intercostals, low rib expansion)
  - Upper Chest (anterior-superior; SCM, scalenes, upper intercostals)
  - Upper chest (superior; upper trap)



(Massery, 2021)

Occupational Therapy for Myositis/Myopathy/MD – Respiratory Function

- Identify breathing patterns
  - What expands most when breathing? Upper vs lower chest, abdomen
  - Do the ribs move or stay stationary?
    - .mmchestwallexcursion
  - Does respiratory rate change in different positions?
    - .mmdynamicbreathingobs
  - How is the patient's posture? What do you notice about their landmarks?
    - .mmstaticobschart

	Anterior View	Lateral View	Posterior View
Shape	thoracic shape		
Rib rotation/orientation	rib orientation	rib orientation	rib orientation
Symmetry	symmetry	symmetry	symmetry
Head position	ant view head position	lat view head position	
Neck	neck space		
Shoulders	ant shoulder view	lat shoulder view	
Sternum	sternal angle		
Ribs 8-10	rib flare		
Pelvis		pelvic tilt	pelvic obliquity
Spinal curve		lat view spinal curve	post view spinal curve
Scapulae			scapula positioning

(Massery, 2021)

Occupational Therapy for Myositis/Myopathy/MD – Respiratory Function

- Simple Interventions
  - Promote appropriate posture and evaluate alignment
  - Pair breathing with movement (inhale on effort, exhale on release)
  - Promote another type of breathing if current method is not effective for a good breath (if patient is dominant chest breather, promote diaphragmatic, for example)
    - Promote chest (fast twitch)
      - shoulder flexion, abduction, ER; Open, expanded chest, APT; overhead reach
      - Higher pitched, faster cues
    - Promote diaphragmatic (slow twitch)
      - shoulder extension, adduction, IR; Rounded shoulders; flexed thoracic spine, PPT; reaching low
      - Lower pitched, calmer cues
    - Inhibit through manual overpressure

(Massery, 2021)

Occupational Therapy for Myositis/Myopathy/MD – Respiratory Function

- Simple Interventions
  - Strengthen the preferred breathing strategy
    - Diaphragmatic resistance training
    - Core strengthening
    - Pelvic floor / ENT / respiratory referrals as needed
  - Trial supports for pressure (binders)
  - Vocalizations
    - If noticing breath-holding, this typically signifies an attempt at core engagement. Try having the patient hum, sing, or count when doing an activity that involves substantial effort
  - Loop in someone who took the Massery Course!

(Massery, 2021)

Screening for SLP – Myositis/Myopathy/MD

- Dysphagia
  - Noted with DMD, MD1, OPMD, IBM leading to aspiration
- AAC Needs
  - DMD, MDM
- Facial muscle weakness
  - FSHD
- Cognitive impairment
  - More prevalent in DMD/BMD
  - Can experience attention deficits, sensory processing disorders, dyslexia, dysgraphia, dyscalculia

Patient Resources- Myositis/Myopathy/MD

Handouts/websites:

- See sharepoint for patient education and quick guides to diseases

Associations:

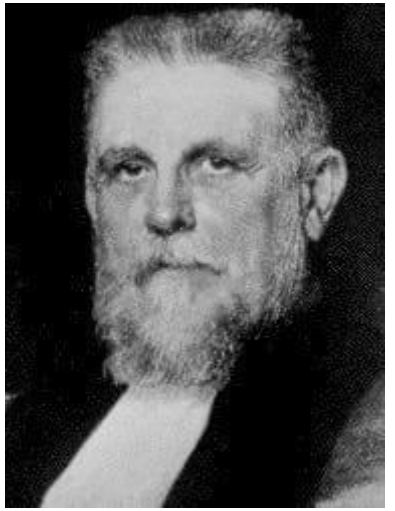
- Muscular Dystrophy Association
  - https://www.mda.org/
- Myostitis Association
  - https://www.myositis.org/
- Parent Project Muscular Dystrophy
  - https://www.parentprojectmd.org/

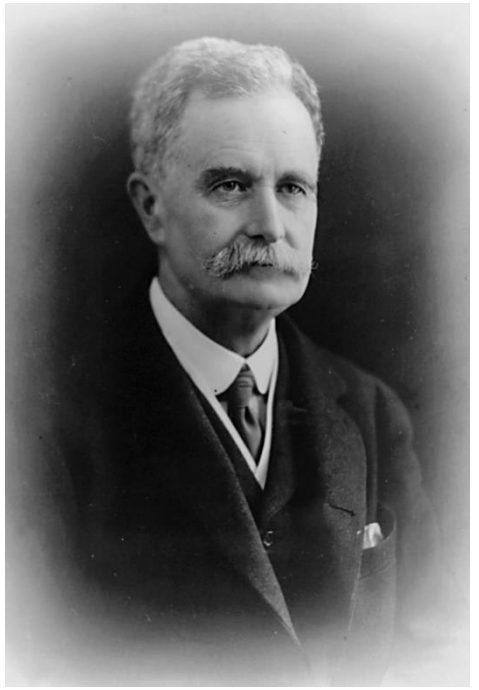
Support Considerations:

- Patient Support - The Myositis Association
  - https://www.myositis.org/patient-support/
- Providing Help and Hope | Muscular Dystrophy Association (mda.org)
  - https://www.mda.org/services

CMT

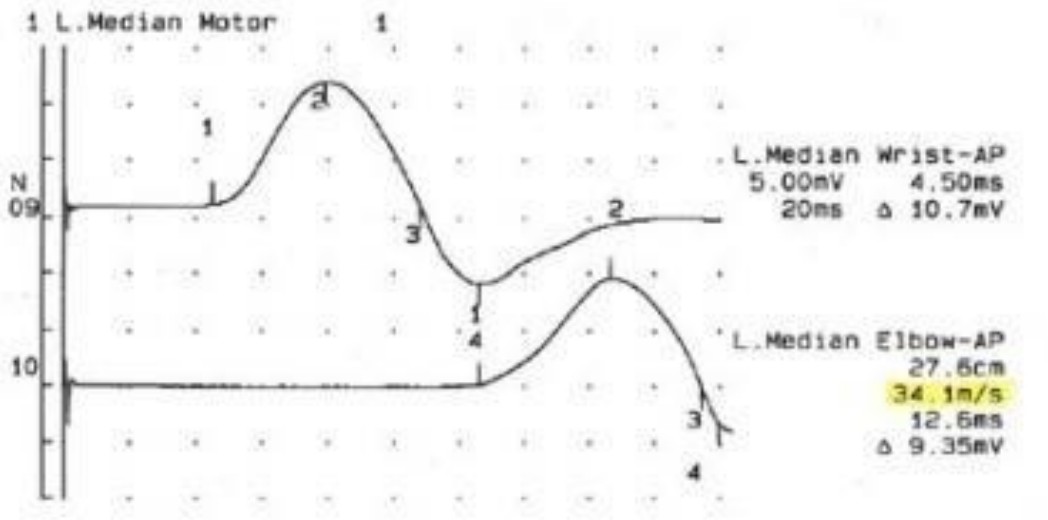
- Charcot Marie Tooth
- 1886
- Distal atrophy attributed to nerve damage

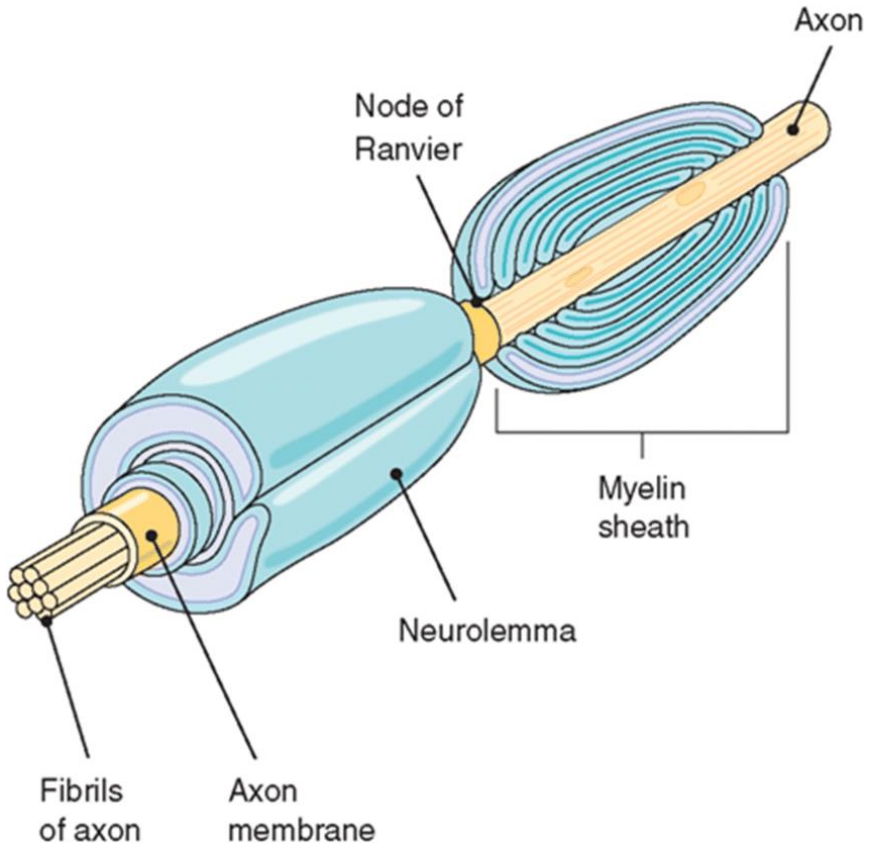




CMT – inherited sensory motor neuropathy

- 1968 – 2 distinct subtypes
  - Type 1 - Demyelinating
  - Type 2 - Axonal





Clinical

Complaints

- Variable Age
- Weakness – foot drop
- Falls
- Atrophy
- Neuropathic Pain
- +/- Numbness

Exam

- Distal Weakness and Atrophy
- High Arches/Flat Feet
- Hyporeflexia
- Sensory Deficits
- Symmetric

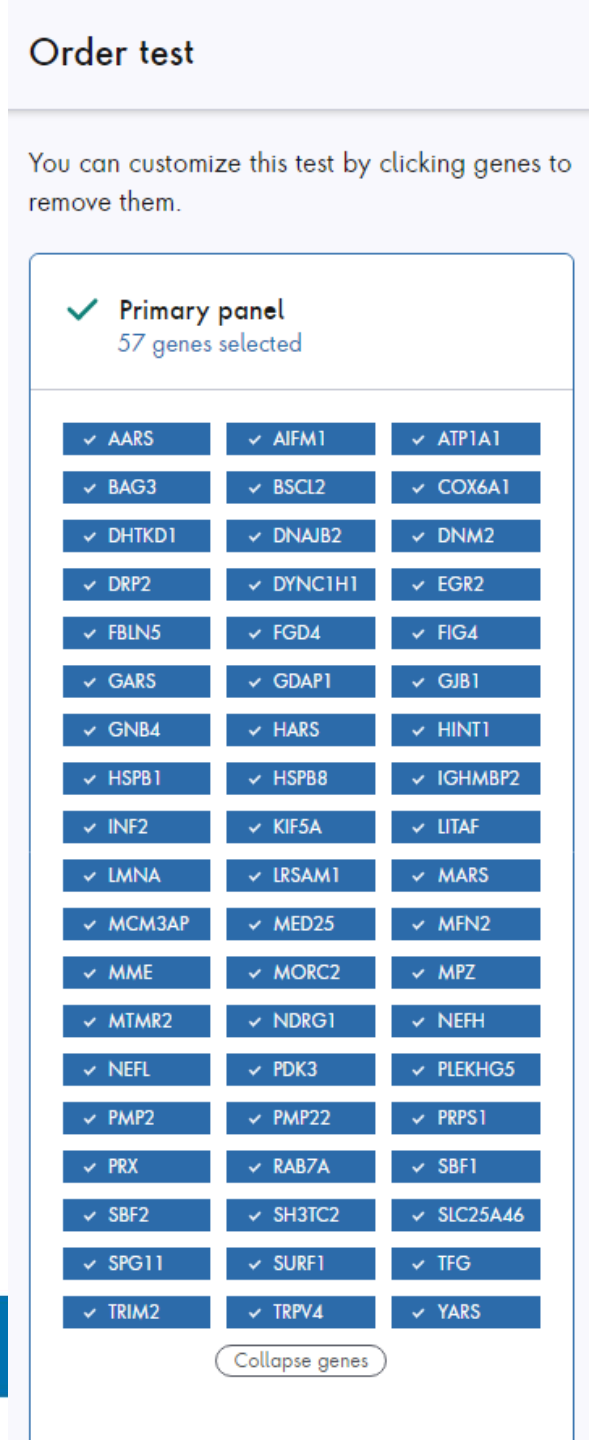
Diagnosis

Nerve Conduction Study/EMG

Rule out other causes

Appropriate History

- Insidious Onset
- Slow Progression
- Historical Hints
- Family History



Prognosis/Course

- Variable but progressive
  - Mild to Severe
- Prone to injury or infections
- Hereditary Neuropathy with Pressure Palsies
  - Recurrent injuries of variable severity

- No direct treatments
  - Trials ongoing
- Symptom Management
- Rehab Needs

CMT: General Therapeutic Approach

- Resistance exercise led to positive changes in strength, functional activities and muscle fiber size
- Aerobic training led to positive changes in strength, functional activities, and aerobic capacity
- Combined exercise led to changes with flexibility, balance, agility, and mobility
- Intensity in studies less than recommended for healthy older adults
  - Lack of consistency across studies
- Stretching, proprioceptive exercises, and treadmill training had objective benefit with CMT without causing overwork weakness
- Bracing, orthotics, assistive device recommendation
- Balance retraining

Sman AD et al. 2015

CMT: General Precautions

- Autonomic dysfunction
  - Specifically cardiac
- Skin/wound assessment
- Pain
- Skeletal deformities
- Depression
- Dysphagia, dyspnea, and OSA
  - Patient's with CMT rarely have swallowing difficulties
- Cognitive dysfunction
  - Neuropsychology or SLP needs
- Weakness of hands/feet
  - Foot drop
- Fall risk assessments

PT Evaluation Considerations in CMT

- Balance: FGA, BERG, Community balance and mobility scale
- Gait: 10MWT
- Endurance: 2MWT vs 6MWT
- Strength: 5XSTS, MMT, dynamometer
- ROM, sensory testing
- QOL: Fatigue severity scale, SF-36

CMT: PT Treatment Considerations

Aerobic Training

- Moderate- to high-intensity
  - 60%–80% HRM
  - RPE 14–17
- Mode:
  - Ergometer
  - Stationary cycling
  - Treadmill
  - Swimming
- Duration:
  - 30-minute session
- Frequency
  - 4–5 times per week
    - improvements seen with 2x/week

Strength Training

- General intensity prescribed less than healthy older adults
  - 8-12 repetitions
- Frequency
  - 2 times per week
- Combination of aerobic and strength led to most significant benefits

(Corrado B et al 2016)

Skin Assessment in CMT

- Screening for protective sensation in feet
  - Monofilament testing – 5.07 monofilament wire
    - Remove patient's socks/shoes in sitting
    - Touch the patient with the 5.07 monofilament in an unaffected area to demonstrate sensation
    - With the patient's eyes closed, hold the monofilament perpendicular to the foot and apply pressure until the monofilament bends into a "C" shape
    - Instruct the patient to report when feeling sensation
    - Typically performed at the heel, 1st/3rd/5th met heads, and 1st/3rd/5th plantar aspect of the toes
  - If patient is lacking protective sensation, education on importance of performing skin assessments to prevent skin breakdown

Skeletal deformities in CMT

Pes Cavus

- Imbalance in foot musculature causing high arched foot
- Weakness at Tibialis Anterior
- Strength of Peroneus Longus

Toe Deformities

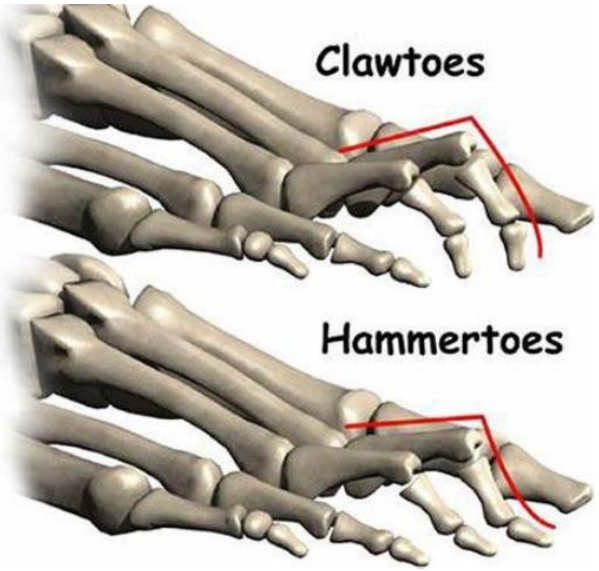
- Hammer toes
- Claw toes

Surgery for foot deformities

- Indications:
  - Realigning foot
  - Muscle imbalance
  - Pain management
- Individualized per patient, surgery earlier is better

Recurrent ankle sprains

Fractures of the 5th metatarsal



(Piscoiotta, C. et al 2021)

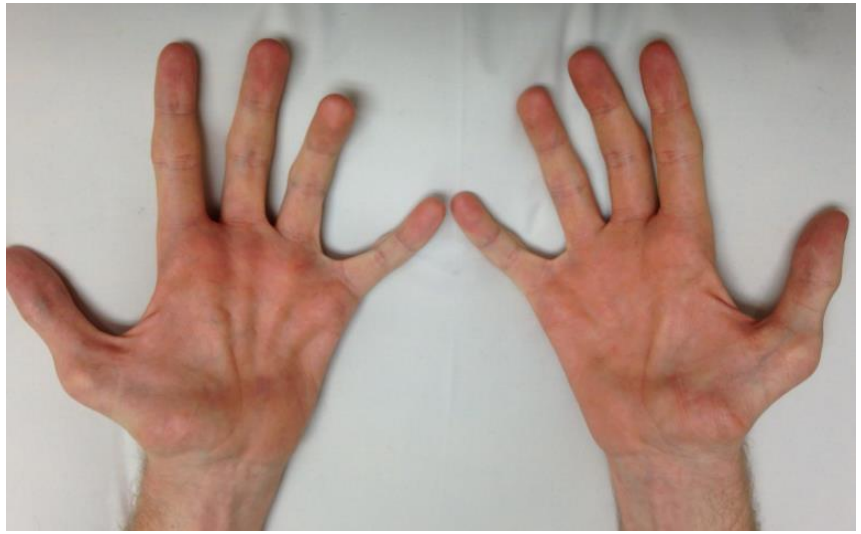
Skeletal deformities in CMT

Hand tendon transfers

- Improve grip and grasp function
  - Rare

Scoliosis

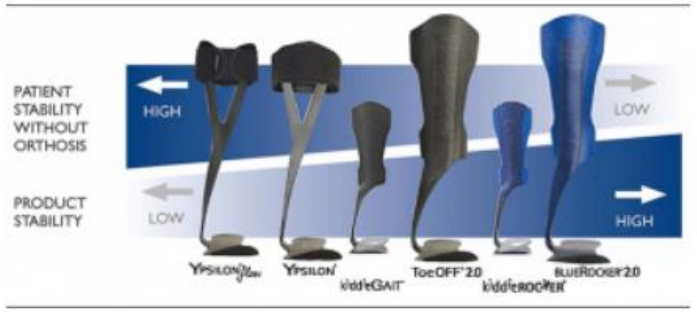
- Bracing and PT suffice
  - Surgery rarely needed



(Piscoiotta, C. et al 2021)

Orthotics in CMT

- Frequently used for lower extremities
  - AFO
    - Improved balance, gait mechanics, gait speed
    - Decrease in hip flexion
      - steppage gait
    - See orthotic guide for CMT on CMTA website
      - Bracing for CMT | Charcot–Marie–Tooth Association (cmtausa.org)
  - Shoe inserts
    - Insole, toe spreaders
      - Foot misalignment
      - Pain
      - Callouses



(Piscoiotta, C. et al 2021)

Assistive device recommendations in CMT

- Cane
- Lofstrand crutches
- Walker
  - 2WW versus rollator
- Wheelchair or Scooter
  - Referral to Wheelchair Clinic

Occupational Therapy for CMT - POC Considerations (Reference Previous Slides)

- ADL/IADL performance, modifications and adaptive equipment use
- Caregiver participation and burden
- Splinting/orthotics
- AT/Wheelchairs
- Home set-up
- FMC
- Pain management/ Stretching
- Exercise/Activity Tolerance
- Sleep

Occupational Therapy for CMT - UE

Table 2
Rehabilitation protocol followed by professionals.

Strengthening (1-4 week)		
		Muscles involved
Abduction of the fingers with a submaximal effort	5 times per hand	Interosseous
Adduction of the fingers with a submaximal effort	5 times per hand	Interosseous
Thumb opposition with a submaximal effort	5 times per hand	Thenar eminence
Extension of the fingers with a submaximal effort	5 times per hand	Extensors
Opposition of all fingers with a submaximal effort	5 times per hand	Thenar and Hypothenar eminence
Stretching (1-4 week)		
Fingers flexors	5 times per finger	
Wrist flexors	5 times per wrist	
Pollicis adductor	5 times per hand	
Interosseous and lombrical (dorsal)	5 times per hand	
Interosseous and lombrical (palmar)	5 times per hand	
Proprioception (1-2 week)		
Turn 2 marbles in the palm per 60 sec	2 times per hand	
Theraputty manipulation: making stripes	4 times per hand	
Theraputty manipulation: little balls modeling (6 balls)	2 times per hand	
Proprioception (3-4 week)		
Turn 4 marbles in the palm per 60 sec	2 times per hand	
Theraputty manipulation: making stripes	4 times per hand	
Theraputty manipulation: little balls modeling (6 balls)	2 times per hand	
Extraction of 4 marbles from theraputty with pinch	2 times per hand	

Occupational Therapy for CMT – Tremor Management

- Compensatory techniques
  - Proximal support
  - Orthotics
    - Upcoming robotics and tremor-dampening orthoses
  - Weighted objects / wrist weights
  - Adaptive equipment such as pens, silverware
  - Stress management strategies such as deep breathing, mental imagery
  - Similar interventions to essential tremor
    - https://essentialtremor.org/resource/assistive-devices/

Occupational Therapy for CMT – Orthotics

- Oval 8s
  - IP support and resisting flexion
- Intrinsic Plus orthosis
  - Stretch to intrinsics and for functional grasp
- Resting hand if appropriate
  - Specifically in supportive intrinsic plus positioning
- Thumb positioning orthosis
  - For functional thumb positioning

Occupational Therapy for CMT – Modalities and Pain Management

- E-Stim and Spinal Stimulators
  - A case study by Hassel (1998) shows that e-stim potentially safe and effective for improving muscle strength
  - A case study by Skaribas et al (2009) shows successful use a spinal stimulator for pain
- No distinct literature for specific modalities for pain management in CMT, clinical judgement and physician clearance should be used for use of ultrasound, e-stim, etc.

Speech Therapy Screening for CMT

- Vocal cord paresis
- Executive function deficits
- Rare swallowing dysfunction
- Rare oral/facial involvement

(Benson, 2010)

Patient Resources- CMT

Handouts/websites:

- See sharepoint for patient education and quick guides to diseases

Associations:

- Home | Charcot–Marie–Tooth Association (cmtausa.org)
  - https://www.cmtausa.org/

Support Considerations:

- Emotional Support Group | Charcot–Marie–Tooth Association (cmtausa.org)
  - https://www.cmtausa.org/emotional-support-group/

OHIOHEALTH PHYSICIAN GROUP

Neuromuscular Clinic

Neuromuscular medicine is an area of neurology that manages disease affecting the peripheral nerves and muscles. Common diseases include Myasthenia Gravis, Guillain-Barre Syndrome, Chronic Inflammatory Demyelinating Polyradiculoneuropathy, and Inclusion Body myositis. Other rare acquired or hereditary disorders affecting the nerves or muscles are managed within this neurology specialty. A neuromuscular diagnosis often brings complex life challenges and this interdisciplinary neuromuscular clinic offers support for patients with an existing diagnosis.

Preston J. Eibling, DO
Neuromuscular Medicine

John C. Novak, MD
Neurophysiology

Timothy J.Rust, MD
Neurology

Xiaosong Zhao, MD
Neurology

Neurologists with neuromuscular expertise

+ Review your study results, perform a detailed medical history and physical examination to develop the best plan with you.

Physical Therapists

+ Assess risk for falls and helps to improve overall balance and strength focusing on general mobility including transfers, walking and stairs.

Occupational Therapists

+ Focus on safety and mobility with activities of daily living, including improving independence with bathing, dressing, eating, driving and household chores.

Speech Therapists

+ Focus on strategies to assist with swallowing, communication, memory and cognition.

OhioHealth Neuromuscular Clinic
XXXX Address, Suite 210, Westerville, OH 43082
Questions? Please call (614) 533-5500.

LEARN MORE

For class and support group dates and times, visit OhioHealth.com/DempseyCenter.

OhioHealth

Questions?

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