Duchenne Muscular Dystrophy Part 3

Community and Home Environments

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Disclosures

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Chapter One

Keeping Your Patient Mobile and Active

Equipment Across the Life Span

- **Early Ambulatory stages:** few pieces of equipment are needed early on
- **Middle Ambulatory stages:** transitions to equipment for longer distances and to address fatigue
- **Late Ambulatory stage:** increased need for equipment for shorter distances, to facilitate independence, assistive devices to access environment
- **Non-Ambulatory stage**: assistive devices to meet the daily needs of the individual

Stretching: Active vs. Passive

Positioning for stretching is critical for results

- Hands-on therapy to preserve Range of Motion (ROM)
  - Active
    - Instruct individual how to stretch his own muscles, especially:
      - » Heel cords, hamstrings, hip flexors, hip abductors
      - » Elbows, forearms, wrists, hands, fingers
  - Active Assist: gentle traction will improve joint motion
    - Instruct parent/caregiver/sibling how to assist with stretching
  - Passive: Gentle traction will improve joint motion
    - Instruct parent/caregiver/sibling how to do stretching
      - » What does it feel like? How do you know when you have reached maximum stretch?

1. PPMD: YouTube videos : www.parentprojectmd.org StretchOUT
2. Stretch Instruction and Workout: www.cinrgresearch.org/stretchvideo2/index.cfm
3. www.youtube.com/watch?v=TVK75IzeLgML gM&t5sNf9Zfb0mTwJHfEB0wsGt

Stretching: Benefits

- Improves circulation
- Improves joint mobility
- Improves tissue extensibility
- Provides feeling of “well-being”

Precautions for Stretching

- Deep tissue stretching/myofascial release?
  - This can be damaging to the muscle and cell membrane
  - Can be traumatic to the tissue, increasing the inflammatory response and damage to muscle
- Taking a stretch to the end range with overpressure?
  - This can damage tissue and cell membrane
  - Avoid overpressure at a joint to decrease the response to trauma as noted above
- Should there be pain?
  - Pain indicates the stretch is too aggressive
  - Weigh the response, and determine if you have applied overpressure or used a technique that was too aggressive
  - Evaluate this response and determine origin

Devices to Help Stretching Program

Nada Chair

Assists in long sitting to stretch hamstrings when trunk control is present

StretchRite

Wedge

Prostretch

Assessment and Selection of Orthotic Intervention: Lower Extremity

- Lower Extremity
  - Braces for **stretching** heel cords
    - Daytime use infrequent
    - Nighttime use more common
  - Knee immobilizers to stretch hamstrings at the knee joint
- Bracing is recommended for periods of five to eight hours to be effective in improving ROM
  - Day use is often accomplished during school hours while sitting
    - Stretches Soleus not Gastrocnemius
  - Night use is a passive form of stretching
  - Weigh advantages for bracing day vs. night

Night Splints

- Maintain prolonged stretch for over six hours per night
- Shown to be effective[1]
- Best when forefoot is supported to keep the calcaneus aligned

Examples of Night Splints

- http://www.orthologix.com/
- www.cascadedafo.com/

1. Scott et al 1981, Hyde et al 2000

Serial Casting to Improve Ankle Range of Motion

Who	Ambulatory boys to regain ankle ROM
Criteria	Dorsiflexion less than neutral
Skills required	• Ability to stand up from the floor • Antigravity knee extension • Ability to safely stand and walk with cast
Schedule	Cast change every seven to ten days
No standard of care	Variable use and protocol throughout the world not standardized (less than half of centers surveyed use) **** Precaution: Child must be able to walk out of clinic**
Upper extremity	Typically addressed with splinting • Elbow and wrist and/or hand

Glanzman et al, 2011; Main et al 2007

Assessment and Selection of Orthotic Intervention: Upper Extremity

- Typical joints requiring special attention
  - **Elbow**: loss of elbow flexion/extension
  - **Wrist:** loss of wrist extension vs. flexion
  - **Fingers/Thumb:** loss of complete finger extension, thumb adduction
- Common contractures
  - Wrist flexion with deviation, finger extension, thumb abduction
- When should you refer to Occupational Therapy?
  - Splinting
    - Night vs. day
  - Adapting ADL
    - Modifying environments

Assessment and Selection of Orthotic Intervention: Upper Extremity (cont.)




Serial Casting

- No standard of care (based on survey of 16 sites throughout US, Canada, Australia)
- 50 percent of therapists use occasionally
- No standard for number of weeks the casting should be repeated
- No standard for pre or post assessments

**Survey Terri Carry 2015 Physical Therapist Children's Hospital of Colorado

Chapter Two

Exercise Guidelines

What We Know About Exercise in This Population

- No exercise leads to muscle atrophy
  - Encourage self-modulation
  - Schedule rest breaks
- Too much exercise hastens muscle breakdown
  - Rhabdomyolysis
- Eccentric contractions are more damaging to the muscle cell
  - **** Isometric→Concentric→Eccentric****
- Younger boys benefit more than older boys
- Boys with DMD are 40 percent less active than age-matched peers[1] supported by other studies

McDonald, 2000

Exercise Recommendations

- Age-appropriate recreational activities as opposed to strengthening regimes
- Concentric low load versus eccentric high load: **stay submaximal**
- Balance activity with rest: don't overdo
  - Children should not experience muscle soreness or excessive fatigue
  - Distributed practice
    - Rest period is greater or equal to the exercise period
- Incorporate balance and coordination skills
- Activities should be fun and promote self-esteem and social interaction

UE Ergometer vs. Active ROM

- 24 boys aged 8-12 years
- **Control Group:** Home program with ROM exercises, 40 minutes, five times per week
- **Treatment Group:** UE ergometer training at 50 percent max, 40 minutes, three times per week
- Ergometer had improved NSAA, supine to stand, elbow flexion endurance
- Control had improved grip and endurance

Alemdaroglu I, et al, Muscle Nerve 51, 2015

Cycling

Assisted cycling and cycling without excessive resistance can be beneficial

- Avoid excessive resistance and fatigue
- Avoid hills, or give assistance on hills or with signs of fatigue or effort
- Add power when needed for energy conservation to keep submaximal

Jansen M, van Alfen N, Geurts AC, de Groot IJ. Assisted bicycle training delays functional deterioration in boys with Duchenne muscular dystrophy: the randomized controlled trial "no use is disuse". Neurorehabil Neural Repair. Nov-Dec 2013;27(9):816-827

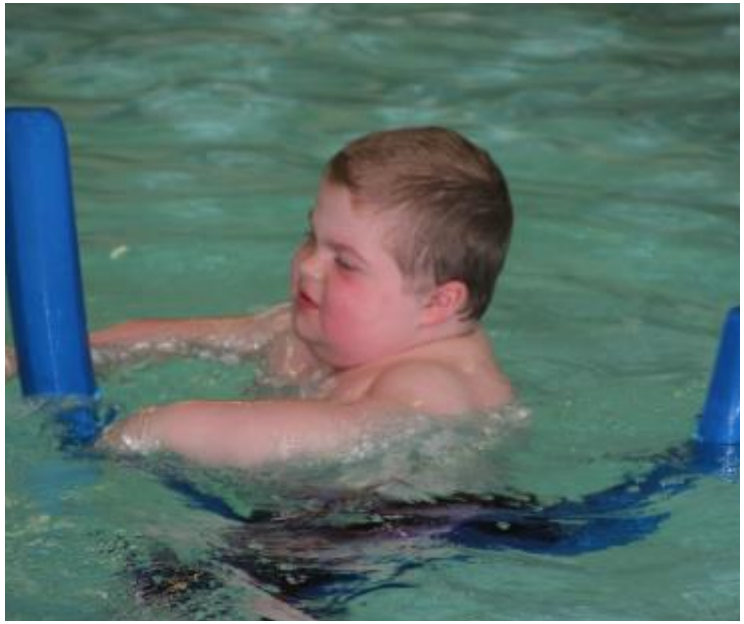
Cycling (cont.)

- Stationary exercise bike
- Tandem bike
- Adaptive bike
- Ex-n-Flex: Active Assist

Examples of wheelchair cycles can be found at the following websites

- www.exnflex.com/
- www.freedomconcepts.com
- http://www.edmontonbikes.ca/

Aquatic Exercise



Aquatic Activities

- Safest form of exercise
  - Non-weight-bearing, low-load activity
  - Able to move through full range of motion
  - Improves aerobic function
- Develops independence and confidence
- Fosters a lifelong recreational activity
- Freedom of movement in later years

Chapter Three

Equipment Changes and Challenges Across the Stages of DMD

Overview of Equipment Important Points to Consider

- Consider for recreation and outdoor play
  - Requires a helmet for safety outdoors
  - Child needs appropriate safety judgement
- When child unable to keep up with peers
- Independence for long distances for community events
- Can be transported in most vehicles
- Can the child use the device independently?
- Will it grow with child?
- Will it meet the individual's needs for five years
- Mobility devices
  - Scooters
  - Wheelchairs
    - Manual vs. Electric
- Standing Devices
- Toileting
- Showering
- Transfers
- Home adaptations/modifications

Scooters and Alternative Motorized Systems

Scooters

- More portable than wheel chair

Negatives

- Easily tips, especially with decreased trunk strength
- Large turning radius
- Difficult transfers on/off
- Poor/no seating support
- UEs may get tired from reaching for handles

Use Caution!

Portable Power Assist Wheelchairs

Examples of Electric Wheel Chairs

- Efix by Frank Mobility
  - http://www.frankmobility.com
- Smart Drive
  - http://www.max-mobility.com
- E Motion Power Assist
  - http://www.alber-usa.com/produkte-rollstuhl-zusatzantrieb/zusatzantriebe-fuer-rollstuehle/aktivrollstuhl-emotion.html

Power Wheelchair Purchase

- Power options
  - Power standing feature
  - Power tilt
  - Power seat elevation
  - Power recline
  - Separately elevating power elevating leg rests
- Cushion to allow for scooting forward
- Seating to maximize postural alignment
- Safety features to decrease injury
- Joystick modifications

Supported Standing

Standers, Stand and Drive chairs

- EasyStand hydraulic stander
  - www.EasyStand.com
- Permobil F5 Stand and Drive motorized wheelchair
  - www.permobilus.com/f5vs.php
- Redman standing powerchairs
  - www.redmanpowerchair.com/

***Bluetooth is now becoming a more standard feature on motorized chairs**

Toileting and Bathing

- Bathing
  - Tub vs. shower
  - Shower chair
- Toileting
  - Transfer to toilet
  - Use of urinal

Video

Shower and Toilet Transfer Patient

Adaptations

- **Computer**
  - Mouse
  - Touch screen
  - Table/desk access
    - With wheelchair
    - With upper extremity access
- **Games**
- **TV**
- **Light switches**
- **Call button**
  - Doorbell
  - Medical alert
- **Environmental control systems**
- **Bluetooth devices**
- **Siri/Dragon Speak-voice-activated systems**
- **Portable Amazon Echo-"Alexa"**

Video

Participation Patient

Chapter Four

Case Examples

Elements of an Assessment

- Use of objective measures
- Observational gait and posture
- Functional skills
- Balance skills
- ROM

Use of Objective Measures

- Objective measures were discussed in Course Two. I encourage to go back and refresh your memory about what these measures are and when to use them.

Observational Gait and Posture

- Walking
- Running
- Standing posture
- Posture during activities

Functional Skills

Strength assessment during functional skills

- Getting up from the floor
- Getting into sitting
- Hop, jump, step
- Climbing stairs
- Squatting in play
- Transitions

Balance Skills

- Ankle strategy
- Hip strategy
- Stepping strategy
- Equilibrium and righting reactions

ROM

- Lower extremity ROM
  - Hip, knee, and ankle ranges
- Upper extremity ROM
  - Shoulder, elbow, forearm, wrist, fingers/thumb
- Trunk
  - Screen for scoliosis

Stretching

- Instruction of a stretching program is critical, with focus initially on areas of tightness in new families: hamstrings and heel cords
- Over time other muscle groups are added that become tight as the disease progresses

Stretching: Public Access

Stretching Videos

- PPMD: YouTube videos
- www.parentprojectmd.org

StretchOUT Stretch Instruction and Workout

- www.cinrgresearch.org/stretchvideo2/index.cf
- https://www.youtube.com/watch?v=TVK75IzeLgML

Chapter Five

Participation and Quality of Life (QOL)

Assessment Tools for Quality of Life

- DASH: Disability of the Arm, Shoulder, and Hand: patient self-inventory
- PedsQL: Multiple modules for family and child available for specific age ranges
- PEDI: Pediatric Evaluation and Disability Inventory: assesses level of functional capabilities
- Depression
  - Center for Epidemiological Studies Depression Scale (CES-D) is a brief self-report questionnaire that is designed to measure depressive symptoms in the general population
  - Anxiety and Depression Scales[1]
- Other: New assessment tools are developed all the time. Stay abreast of the latest assessments; look for population-specific tools
  - Behavioral screening and comprehensive neuropsychological/ developmental evaluation is recommended for boys with DMD

1. http://www.childfirst.ucla.edu/Resources.html Neuromuscular Disorders 17 (2007) 986–994

Things You Need to Know in a School Environment

- School environments are educationally driven, **not** medically driven
  - Be sure to work with the school therapists and personal care assistants
  - Integrate goals into IEP or 504 plan through professional and college training
- Work cooperatively as a team with school personnel
- Multiple resources for teachers and parents are available *see next slide
- Sport/PE periods
  - Self Modulation, adaptive PE, stretching
  - Advocate for child

IEP vs. 504 Plans

IEP

- The Individualized Educational Plan (IEP) is developed to ensure that a child with a diagnosed disability under the law, attending an elementary or secondary educational institution, receives specialized instruction and related services.

504 Plan

- The 504 Plan is developed to ensure that a child with a diagnosis identified under the law, attending an elementary or secondary educational institution, receives accommodations.
  *** No specialized instruction required.**

Educational Environment

Think about the following:

- Services direct or consultative
- Modifications: what type?
- Accessibility issues
- PE/ field trip participation
- Safety plans
- Equipment for transfers
- Toileting equipment

Resources for Schools and Families

- PPMD Website
  - www.parentprojectmd.org
- Education Matters: A teacher's guide to Duchenne Muscular Dystrophy
- Education Matters: Adaptive Physical Education: A PE teacher's guide to Duchenne Muscular Dystrophy
- Education Matters: An introduction for parents
- Learning and Behavior in DMD for parents and educators

www.parentprojectmd.org

Higher-Level Education

- MDA Transitions Center
  - http://transitions.mda.org
- Do-IT program Disabilities, Opportunities, Internetworking, and Technology
  - http://www.washington.edu/doit
- Going to college handbook
  - http://www.going-to-college.org

Participation in Outside Activities

Coordinate with families and the person providing activities in the community.

Participation in Outside Activities (cont.)

Warning Signs to Be Aware of

Depression

- **Patient with DMD:** increased stress levels during periods of transition and changes in function, surgeries
  - Despondent, change in behavior, irritable, perseveration on negatives
- **Family members:** increased stress levels day to day and during periods of transition
  - Despondent, change in behavior, disruption of sleep pattern, hypervigilance, paranoia
- **Caregivers: all of the above**
  - Stress, irritability, unable to relax, not taking time for self

Role of Physical Therapist

- Assessment, plan of care, intervention
- Interpretation of medical information and research
- Referral to medical team specializing in DMD/coordinator for clinic
- Referral to research studies/coordinator
- Referral to counselor/social worker
- Home/environmental assessment
- Connecting family with support
  - PPMD, MDA, FACES, Duchenne Connect, clinical care, local support
- Supporting family and child through progression of disease

Summary of Course Three

- Equipment needs change across the lifespan
  - Individually assessed
- Stretching is important: Active > Passive
  - Improves circulation, joint mobility, tissue extensibility
- Night bracing beneficial
  - Walking braces not usually used
  - Upper extremities may need splinting
  - Serial casting may improve ROM but strength necessary to be successful

Summary of Course Three (cont.)

- Exercise: Stay Active
  - Inactivity leads to muscle wasting and atrophy
  - Don't overdo
    - Self-modulate
  - Eccentric contractions are the most harmful
  - Consider all the pros and cons to exercise when developing an exercise program for a child with DMD
- Mobility devices improve access
  - Assess individually considering environmental factors

Summary of Course Three (cont.)

- Assess environment for modifications and/or adaptations
  - School
  - Home
  - Community
- Quality of life
  - Multiple assessments available
- Role of therapist
  - Team member with child, family, and other health care professionals

Chapter Six

Parent and Therapist Discussion

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References Template

Duchenne Muscular Dystrophy Part 3: Community & Home Environments, Claudia Senesac, PT, PhD, PCS

For this course to be approved by the accredition bodies, please make sure there are at least **five** references published in a peer-reviewed journal within the past **five** years. Please create full text citations in AMA or APA style for each reference, including websites.

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