Lifelong Physical Therapy Needs

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Disclosures

- Program Manager ImagingDMD Studies
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- Instructor MedBridge Educational Courses
Signs and Symptoms

- Delayed motor skills
- Head lag with pull to sit
- Delayed walking skills
- Rising from the floor
- ↓ed Squat to stand
- ↓ed Walking up stairs
- ↓ed Jumping/Hopping
- Calf hypertrophy
- ↓ed running speed
- ↑ed falling
- Language delays
- Cognitive delays
- Social Difficulties
- Hamstring tightness
- Heel cords tightness
- Diagnosis of Autism

www.childmuscleweakness.org

Differential Diagnosis

Rule out the following in your assessment

- **Idiopathic Toe Walking**
  - Toe walking, often fall, difficulty stopping, poor trunk strength

- **Cerebral Palsy**
  - Falling, difficulty walking, difficulty getting up from the floor

- **Developmental Coordination Disorder**
  - Falling, clumsy, gait difficulties, delays
Classic Signs-Observed in Clinic
To set this diagnosis apart from others during differential diagnosis

- Gowers sign
- Posture
- Calf Hypertrophy (Pseudohypertrophy)

DMD Clinical Milestones: *Ambulatory*

- Gowers Sign
- Loss of Standing from the Floor
- Loss of Stair Climbing
- Loss of Standing from the Chair
- Loss of Walking Independently
DMD Clinical Milestones: Non-Ambulatory

- Loss of ability to reach head
- Loss of ability to roll in bed
- FVC ~ 50%: Need for Cough assist
- Loss ability to feed oneself
- Loss of ability to get hands to table
- Loss of ability to use computer/phone


Stage 1 • Pre-Symptomatic
Stage 2 • Early Ambulatory
Stage 3 • Late Ambulatory-Transitional
Stage 4 • Early Non-ambulatory
Stage 5 • Late Non-ambulatory
Pre Symptomatic Observations Commonly Reported

Difficulty noted with:

<table>
<thead>
<tr>
<th>Activity</th>
<th>Observation/Sign</th>
</tr>
</thead>
<tbody>
<tr>
<td>Climbing Stairs</td>
<td>Getting up from the Floor-Partial Gowers Sign</td>
</tr>
<tr>
<td>Jumping</td>
<td>Running: slow, no flight phase</td>
</tr>
<tr>
<td>Toe Walking</td>
<td>Frequent Falling</td>
</tr>
<tr>
<td>Weakness</td>
<td>Leg Cramps</td>
</tr>
<tr>
<td>Clumsiness</td>
<td>Fatigue</td>
</tr>
<tr>
<td>Large Calf Muscles</td>
<td>Muscle Tightness-Hamstrings/HC</td>
</tr>
<tr>
<td>Head lag on pull to sit- difficulty lifting head from supine</td>
<td>Decreased score on BSID III Bailey Scale of Infant Development (Connolly et al, 2013)</td>
</tr>
<tr>
<td>Delayed Motor Skills</td>
<td>Delayed Speech</td>
</tr>
</tbody>
</table>

Begin A Regular Routine

Concentration Areas

- Heel Cords
- Hamstrings
- Hip Flexors
- IT Band
- Consider Initiation of Night Braces

Stretching

- Incline Standing Board
- Long Sitting
- Prone Lying
- Side Lying Stretch
- Consider Behavior

Make it Enjoyable.....Make it Fun!!!!
Stretching
May Be Preventative-Start Before Tightness

- May maintain length of the muscle over time
  - Single and two joint muscles
- May maintain symmetrical postural alignment
- May prolong the ability to walk
- May improve positioning in wheelchair
- ROM assist with use of a standing frame/chair
- Improve comfort and positioning in bed
- Improve hygiene
- May improve fit of shoes
- May improve hand manipulation
- Improves Well Being
- Improves Circulation

Stretching Resources for Parents

- PPMD Website
  Stretches for Duchenne Muscular Dystrophy (CD)-View online
  http://www.parentprojectmd.org/site/PageServer?pagename=Care_resources_materials
  Instructional Photographs -View online

- YouTube
  Stretches for Duchenne Muscular Dystrophy-YouTube Video
  https://www.youtube.com/watch?v=6eHLTkOAOG

- CINRG Website
  StretchOUT Stretch Instruction and Workout http://www.cinrgresearch.org/stretchvideo2/index.cfm
Pre-Symptomatic

Encourage Recreational Activities

Understanding Disease Progression Validates our Observations
Sensitivity: MR measures can detect muscle disease at a young age are sensitive to disease progression, detecting change over 1 year even in boys that are functionally stable (no change in 6 min. walk). The figure above shows images and spectra (MRS) from the upper leg muscles.

**Variability Across the Population**

Longitudinal Evaluation of Muscle Composition Using Magnetic Resonance

![Images of muscle composition using magnetic resonance in DMD patients and controls](image)

**UE Images**

What do you see?
What does it mean?

Early Ambulatory

- Often just receiving diagnosis
- Elementary School
- Initiating Steroids
- Difficulty keeping up with peers
- Increased falling
- Occasional leg pain or cramping
- ROM (tightness may begin to develop or worsen)

Common Measures

Clinic vs. Clinical Trial

- Manual Muscle test (proximal weaker than distal; extensors < flexors; LE<UE)
  - Myometry
  - Goniometry
  - Postural and Gait assessment
  - Timed Performance Tests
  - North Star Ambulatory Assessment
  - 6 minute walk test
  - PUL
  - Brooks Scale- UE
  - Vignos Scale- LE
Why Are Assessments So Important

- Provide evidence for status and change in status
- Provide feedback for change in medication or treatment
- Demonstrate pattern of weakness and decline
- Predict need for intervention
  - Loss of ambulation: Equipment, home access, transportation, transfer teaching, support
  - Help explain status to parents and others

Postural and Gait Deviations

- Increased lordosis
- Steppage gait with reduced stride length
- Widening of base of support
- Increased lateral trunk lean
- Rising up on toes *not always seen
- Toeing in
- Pronation/Supination
Therapist’s Role
Encourage, Instruct, Educate Families and Child, Be available to Consult

- Maximize Range of Motion and Gross Motor Skills
  - Safety First
- Families should be instructed in
  - STRETCHING-Keep building a routine
  - Safe activities for play, recreation
- Maximize Balance and Coordination
- Maintain Overall Fitness
- Consult with School: Adaptations/Modifications
- DON’T OVER FATIGUE-Rest periods are “OK”

How To Fit Everything In

- Make stretching part of the daily routine
  - Brushing teeth
  - During homework position for success
  - Active assistive stretching- get child involved

- Equipment to improve/maintain ROM
  - Night splints
  - Nada chair
  - Standing wedge
**Night Splints - Across All Stages**

*Consider Goals*

- Maintains prolonged stretch (6+ hours/night)
  - Shown to be effective (Scott et al 1981, Hyde et al 2000)

- Medial trim lines - support forefoot
  - Shoes vs no shoes - consider as they get older

- Night vs Day use
- Increased padding

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**Serial Casting**

- Ambulatory Boys to regain ankle ROM
- Dorsiflexion less than neutral *no clear cut off
- Ability to stand up from the floor - pre cast
- Antigravity knee extension
- Ability to safely stand and walk with casts before leaving facility
- Cast change every 7-10 days - variable across facilities
- No standard of care: variable use and protocol, no common assessment used pre/post, varied criteria for selection

(Glanzman et al, 2011; Main et al 2007)
Late Ambulatory-Transitional
A Time of Change

- Limited community mobility
- Increased risk of falls
- Increased fatigue
- Consider power W/C purchase
- Needs change: across life span
- Provider(s), location of services change with needs, and frequency
- Continual assessment of status and needs
- Goals for this population are unique and challenging

Why Predicting Loss of Ambulation is Important

- The impact on families is huge and life changing
- Discussion: start early and repeat often
- Equipment often takes 6 months to receive
- Home modifications are costly and stressful
- Transportation becomes challenging
- Transfers are challenging
- School needs time to prepare (hoyer, classroom location, evacuation plan, etc).
Fracture? Now what?

Fractures

- Most often in the lower extremities
- Generally in boys older than 9 years
  - Falls while ambulating
  - Falls from wheelchair (seat belts!!)
- May result in loss of ambulation
- Boys need to avoid immobilization
- Families need to contact their specialists

James et al, 2015; Subasi et al, 2016 Abstract MDA Clinical Conference
Consult “NEW” Care Guidelines www.thelancet.com or www.parentprojectmd.org
Remobilization After Fracture

- Mat Exercise
- Aquatic Therapy
- Alter-G Treadmill
- Body Weight Supported TM
- Up and Free walker - May be an option
- Introduce weight bearing ASAP

http://www.alterg.com/

Early Non-Ambulatory

- Family education!!!
- Encourage activity
  - Aquatic activities
  - Ergometers – active assist
- Assistance for Transfers
- Equipment needs changing
- Increased tightness
- Disuse atrophy
- Increased weight gain
- Loss of functional independence
- Changes in community participation
- Initiation of BiPAP/ Cough Assist
- May increase risk of scoliosis
Late Non-Ambulatory

- Increase c/o Pain
- Maximal Assistance
- Limited UE Function
- Respiratory Support
- Reliance on Technology
- Decreased Participation
- Decreased Employment
- May need to employ caregivers

Daily Routines Changing

- Transfers-more dependent
- Continue LE stretching
- Include UE stretching
- Sustained positioning
  - ¼ - ¾ Prone lying if tolerated
  - Side Lying
  - Standing frames
  - Elevating leg rests
  - Reclining in WC
Assistive Technology
Exciting and Changing all the Time

- Blue tooth devices
- Siri/Dragon Speak
- Environmental control systems
  - Google home & Amazon Echo
- Computers / tablets / smart phones
  - Alternative keyboards/ touch pad screens
  - Electronic pointing devices
- Voice assist (amplifiers)
- Glassouse
- TouchTapSwipe guide
  (dmdpathfinders.org.uk)

MedTrade Expo
Largest Medical Equipment Tradeshow and Conference

Spring: Las Vegas February 2018
Fall: Atlanta October 2018

https://www.medtrade.com/
Exercise

Historically

- Exercise recommendations have been based on rodent studies that induced injury
- Resultant recommendations were: exercise may cause damage - exercise with caution
- Current studies suggest
  - Exercise may be beneficial - BUT questions remain
    - Age?
    - How much?
    - What type?
Exercise

What do we know.....

- No exercise leads to muscle atrophy
  - Important for bone health
  - Important for self-esteem
- Too much exercise increases muscle breakdown
  - Rhabdomyolysis
- Younger boys benefit from exercise more than older boys * however most of the research is with this group
- Boys with DMD are 40% less active than age matched peers (McDonald, 2000) ** supported by other studies

Exercise

- Staying active is KEY
- Stretch may maintain ROM allowing activity and participation
- Age appropriate recreational activities as opposed to “resistive” strengthening regimes
- Concentric low load or isometric versus eccentric high load - stay submaximal-add power if needed/energy conservation
- Don’t overdo!! Build in Rest Periods
  - Self modulation
  - Structured breaks
- Fatigue is REAL, Differences in endurance
- Incorporate balance and coordination skills
Assisted bicycle training delays functional deterioration in boys with Duchenne muscular dystrophy: the randomized controlled trial "no use is disuse".  
Jansen M, van Alfen N, Geurts AC, de Groot IJ

Arm and Leg Ergometer Ex-n-Flex (active assist)
- 24 boys age 8-12 years (amb and non-amb)
- Training at 50% max, 40 minutes, 3 day/week
- 24 weeks

Assisted bicycle training
- Delays functional deterioration in boys with DMD
  - MFM and Assisted 6min cycling test - remained stable
- No serious adverse events
- Safe and feasible
- May decline the deterioration due to disuse


Different types of upper extremity exercise training in Duchenne muscular dystrophy: effects on functional performance, strength, endurance, and ambulation.
Alemdaroglu I, Karaduman A, Yilmaz ÖT, Topaloglu H

- Subjects = 24 boys ages 8-12 y/o, ambulatory
- Study group n=12 * assistive UE ex/PT-Arm Ergometer
  - 40 minutes per session, 3x/week x 8 weeks
  - positive effects on subjects' muscular endurance, performance of ADL's arm function, ambulation status- NO significant change in muscular strength
- Control group n=12 *strengthening ROM ex/Family
  - 40 minutes per session, 5x/week x 8 weeks
  - Improved grip strength and endurance only
  - Improved NSAA score
- Summary-Both groups improved to varying levels

Pilot Study for Mild to Moderate-Intensity Resistance Exercise in Boys with Duchenne Muscular Dystrophy

DJ Lott, KD Cooke, HJ Park, BM Black, SC Forbes, BJ Byrne, GA Walter, K Vandenborne

Subjects: 8 ambulatory boys w/DMD (8.3 ± 0.7yr)

Safety Measures: 48 hr before/after
- T2 weighted MRI
- Pain assessment
- Clinic Exam & CK levels

Protocol: 6 reps/4 sets (isokinetic dynamometer) 4 exercises
- Isometric maximal voluntary contraction (MVC)
  - Quads and Hams at 30 and 60 degrees of knee flex
- Isometric exercise-Mild level (30%MVC) n=4
- Isometric exercise-Moderate level (50%MVC) n=4

Example of protocol
Exercise Hamstrings- 30 and 60 degrees
Exercise Quadriceps- 30 and 60 degrees

Data Analyses/ Results
T2 changes assess potential muscle inflammation/damage

Cross-section of thigh with 8 muscles of interest outlined in color: rectus femoris, vastus medialis, vastus intermedius, vastus lateralis, short head biceps femoris, long head biceps femoris, semitendinosus, semimembranosus

Preliminary Results- awaiting publication
THANK YOU!

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PPMD: www.parentprojectmd.org