Northern California End Duchenne Tour

Saturday November 3, 2018

Tina Duong
Physical Therapist
Stanford University
Acknowledgements

PPMD
Continually supports education locally, throughout communities, nationally, and internationally.

We appreciate all that they do to advance our understanding of this disease and bring families, professionals, and researchers together as a team!

Contributing Therapists

* Tina Duong * Claudia Senesac*
* Laura Case * Leslie Vogel*
Objectives

• Understand Therapists role throughout stages of DMD
• Overview of therapy focus throughout stages
• Review of Standards of Care in Rehab
• Stretching: Musculoskeletal Health
  – How/What
  – Stretching in DMD: CINRG natural history study
• Exercise
Stage 1: • Pre Symptomatic
Stage 2: • Early Ambulatory
Stage 3: • Transitional
Stage 4: • Early Nonambulatory
Stage 5: • Late Nonambulatory
Therapist’s Role
Encourage, Instruct, Educate Families and Child

• Maximize Range of Motion and Gross Motor Skills
  – Safety First
• Families should be instructed and involved in
  – STRETCHING
  – Safe activities for play, recreation
• Maximize Balance and Coordination
• DON’T OVER FATIGUE
Goals for Therapy and Home Program

- Maintain Range of Motion
- Maintain Postural Symmetry
- Improve Balance
- Improve Coordination
- Maintain fitness
- Recommendations for PE
  - Adaptations/Modifications
- Assisting with transitions
- Equipment
- Safety

PPMD: Helen Posselt PT
Participation in Activities

• “Therapeutic use of everyday life activities which enhance or enable participation in roles, habits and routines.” (AOTA, 2014, S1)

• Occupational Therapists (OT) or Physical Therapists enable participation in activities that children, adolescents and adults desire such as:
  – Activities of Daily Living
  – Leisure & Play
  – School & Work
  – Sleep
  – Socialization

• Why is this important?
  – Studies show that the more children with DMD participate in desired activities (such as roles, habits and routines), that it correlates to strength and function. (Bendixen et al., 2014)
Stage 1:
Pre-symptomatic Stage

• Your child may be having difficulty keeping up with peers
• He may appear clumsy, may fall more frequently than his peers
• You may see him sitting out on the sidelines
• He may have difficulty climbing stairs, or ask to be carried
• He may have motor and/or language delays
Presymptomatic Stage

Encourage Recreational Activities

stjohn.ca

Amazon.com
Stage 2: Early Ambulatory Stage

- Often just receiving diagnosis
- Elementary School
- Initiating Steroids
- Difficulty keeping up with peers
Stage 3
Transitional Stage

- Increasing gait deviations
- Increasing reports of back pain
- Limited community mobility
- Increased risk of falls
- Increased Fatigue
- Weight gain
- Power W/C purchase
Stage 4
Early Wheelchair Stage

- Increasing tightness
- Disuse Atrophy
- Increased Weight
- Loss of Independence
- Limited community participation
- Initiation of BiPAP
- Risk of Scoliosis
Stage 5
Late Wheelchair Stage/Non-ambulatory

• Increasing Pain
  – Positioning is critical
• Maximal Assistance
• Limited UE function
• Respiratory Support
• Reliance on Technology
• Decreased participation
• Decreased employment
• May need to employ caregivers
## Standards of Care

<table>
<thead>
<tr>
<th>Stages of</th>
<th>Stretching</th>
<th>Bracing</th>
<th>Exercises</th>
<th>Equipment</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pre Symptomatic</strong></td>
<td>Begin this routine now Make them enjoyable – encourage self-stretching</td>
<td>Introduce night bracing- as tolerated</td>
<td>Encourage recreational activities Bike, swimming, etc..</td>
<td>Nothing special in most instances</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Early Ambulatory</strong></td>
<td>Maximize ROM and Gross motor skills Safety first Stretching-regular routine daily if possible Include Arms and Legs Parent assisted and self-assisted stretching</td>
<td>Night bracing Establish a routine</td>
<td>SAFE activities for play and recreation Maximize balance and coordination DON’T OVER FATIGUE Maintain fitness, maintain symmetry Recommendations for PE for adaptations/modifications</td>
<td>Consider some mode of transportation for longer distances Stroller, manual WC, other</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Transitional</strong></td>
<td>Continue daily stretching routine-all extremities Be creative with stretching i.e. prone lying, standing frame, etc....</td>
<td>Continue routine-change style of night brace as indicated by comfort and ROM</td>
<td>Increased risk for falls Increased fatigue Possible increased weight gain Increased c/o Back pain Increased difficulty walking</td>
<td>Limited community mobility Consider power WC Consider use of standing frame Consider transfers and equipment if needed. Begin to explore technology</td>
</tr>
</tbody>
</table>

*Consider other modes of exercise: ergometer, stationary bike, aquatic therapy, trunk stabilization.
<table>
<thead>
<tr>
<th>Stages of</th>
<th>Stretching</th>
<th>Bracing</th>
<th>Exercises</th>
<th>Equipment</th>
</tr>
</thead>
</table>
| Early Non-ambulatory | Continue daily stretching routine- all extremities  
Increased tightness, increased incidence of scoliosis | Continue routine – as long as ROM allows | Disuse atrophy- consider ergometer for some graded exercise without resistance  
Trunk stabilization exercises | May be using BiPAP  
May be using cough assist  
Possible use of standing frame  
Consider transfers and equipment if needed. i.e. sliding board, hoyer sling, people mover  
Rolling shower chair  
Consider options with technology. I.e. Blue tooth, computer access, accessing environment in home |
| Late Non-ambulatory | Stretching and positioning Increased pain | Continue bracing if possible. If not, positioning feet and ankles at nite to lessen deformity | Require maximum assist  
Increased difficulty with UE use  
Increased adaptations and technology | Reliance on technology  
May need caregivers  
Respiratory support |
Stretching: Maintaining Musculoskeletal Health
Muscle Function and Activity

Skeletal Alignment

Muscle Length
The Muscle-tendon unit

Svensson et al, 2017
Contractures

Abnormal shortening of muscles, skin or connective tissue with restrictions in active and passive range of motion

Muscle Imbalance around a joint: Muscle Weakness
Etiology of the Disease
Contracture Management: Purpose

- To Maintain or improve muscle length
- To prevent orthopedic deformity
- To decrease pain
- To improve function
Impairment and Function

**Contracture Development**
- Ankle Dorsiflexion
- Iliotibial Band
- Hip Flexors
- Knee flexor
- Wrist Flexors
- Elbow Flexors

**Function**
- **Biomechanics**
  - Changes in Torque
    - Length tension curve, Force production
  - Power
    - Velocity and association to prediction of loss of function
  - Difficulty with Walking, Getting up from floor/chair, Climbing Stairs
Contractures and Function

• Clinical Research Trials Inclusion/Exclusion Criteria

• Functional Criteria to enter into a trial
  • Ability to sit independently, Stand, Walk in certain time period

• “Limited By Contracture”

• Effectiveness of treatment
  • Would the contracture mean that even if strength improves, you are not able to see functional benefits?
Approaches to Manage Contractures

Conservative
- Short Duration Stretches <30min
  - Manual and Active Stretches
  - Standing frames: Depends on tolerance
- Long Duration Stretching
  - Splinting
    - Static/dynamic splints
  - Positioning
  - Serial Casting

Surgical
- Tendon lengthening and transfers
Stretching! Across all stages
Initiate before muscles are tight!

**Legs**
- Hip flexors
- Iliotibial band
- Hamstrings
- Quads/Rectus femoris
- Gastrocnemius

**Arms**
- Wrist/finger extensors
- Biceps
- Forearm rotators

**Use prolonged-sustained positions**
- Prone (stomach) lying
- Sitting with Velfoam strap around thighs
- Consider standing frames
- Elevated leg rests on WC
Daily stretching activities

- Parent assisted stretching
- Self assisted stretching
- Positioning
- Equipment to manage ROM
  - Night splints
  - Nada chair
  - Standing wedge

Photo by Helen Posselt PT
Where to start?

- Understanding pathology of disease
- Understanding physiology of intervention
  - Stretching

Pathology  Physiology  Management
Effects of Stretching

• Things to Consider:
  – Improvement due to *actual increase in muscle or tendon length*
  – Improvement due to *increase tolerance to stretch*

• *Muscle extensibility*- “the ability of a muscle to extend to a predetermined endpoint” or capacity to lengthen, stretch or get longer

• Measured by end range joint angles

*Weppler, Magnusson. 2010*
Tendon/muscle contribution to total length change in muscle-tendon unit

Example: Ankle stretch

- Add picture of foot and tendons
  - Arthrokinematics of joint during stretch
    - Roll and glide
  - Tendon
Stretching

- **Purpose**
  - Increase tissue extensibility
    - 1. **Dynamic phase**
      - Viscous Deformation
      - Stretch Reflex
      - Lengthening Reaction
    - 2. **Static phase**
      - Structural Adaptations of muscle and surrounding soft tissue
        » Duration matters
        » Immobilization in lengthened position
        • Molecular changes in muscle length
        • Increase sarcomeres
Stretching

Stretch Reflex

Change in length, pressure Movement
- Golgi Tendon Organs
- Pacinian Corpuscles

Recording length and rate
- Muscle Spindles

Reflexive Muscle Contraction
- Spinal Signal

Lengthening Reaction
- Inhibits muscle contraction; allows elongation

Increase ROM
- Autogenic Inhibition
- Dampening neuro-reflexive mechanism
- Improving Stretch tolerance
The effect of 5 repeated 90-s static stretches on the human hamstring muscle group

<table>
<thead>
<tr>
<th>Variable</th>
<th>1</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dynamic phase (divided into thirds)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Energy (area)</td>
<td>0.4 ± 0.2</td>
<td>0.6 ± 0.1</td>
</tr>
<tr>
<td>First</td>
<td>2.6 ± 0.6</td>
<td>1.2 ± 0.3 b</td>
</tr>
<tr>
<td>Second</td>
<td>11.2 ± 2.0</td>
<td>7.8 ± 1.2 b</td>
</tr>
<tr>
<td>Third</td>
<td>14.2 ± 2.7</td>
<td>9.1 ± 1.5 b</td>
</tr>
<tr>
<td>Total</td>
<td>3.6 ± 1.2</td>
<td>1.4 ± 0.9 c</td>
</tr>
<tr>
<td>Stiffness (slope)</td>
<td>12.4 ± 2.1</td>
<td>8.3 ± 1.0 b</td>
</tr>
<tr>
<td>Static phase</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Torque (N-m)</td>
<td>41.1 ± 4.1</td>
<td>33.3 ± 3.2 b</td>
</tr>
<tr>
<td>Peak</td>
<td>29.8 ± 3.0</td>
<td>26.8 ± 2.7 b</td>
</tr>
<tr>
<td>Final</td>
<td>28 ± 2</td>
<td>20 ± 1 b</td>
</tr>
<tr>
<td>Delta (%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Magnusson et al, AJSM 1997
• 340 DMD participants aged from 2 to 28, followed up to 7 years longitudinally
• Recruitment in 20 Centers worldwide
• Longitudinal data (up to 7 years) on disease milestones and functional measures
• Accurate phenotype and standards of care data
• Specific aim to collect participant DNA and study genetic modifiers
  – (n = 275; 175 with sufficient DNA for SNP chip)
Study cohort, outcomes and methods

• **Clinical Outcomes**
  – 3 Timed Function tests: Supine to stand, Stairs, 10MWT
  – Covariates:
    • Glucocorticosteroid use, age

• **Frequency of Assessments**
  – Year 1
    • 5 timepoints (Baseline, 3mos, 6mos, 9mos, 12mos)
  – Year 2
    • 2 time points (18mos, 24mos)
  – Year 3 onward
    • 1 time point
Wrist Extension ROM

<table>
<thead>
<tr>
<th>Age interval</th>
<th>Ambulatory</th>
<th>Non-ambulatory</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N (Obs)</td>
<td>N (Ind.)</td>
</tr>
<tr>
<td></td>
<td>N (Obs)</td>
<td>N (Ind.)</td>
</tr>
<tr>
<td>&lt;4.9</td>
<td>62</td>
<td>29</td>
</tr>
<tr>
<td>5.0 to 8.9</td>
<td>681</td>
<td>216</td>
</tr>
<tr>
<td>9.0 to 12.9</td>
<td>370</td>
<td>157</td>
</tr>
<tr>
<td>13.0 to 16.9</td>
<td>145</td>
<td>70</td>
</tr>
<tr>
<td>17+</td>
<td>32</td>
<td>17</td>
</tr>
</tbody>
</table>
# Elbow Extension ROM

The image shows a box plot of elbow ROM (Range of Motion) by age (years) for both ambulatory and non-ambulatory patients. The normal range for elbow extension is indicated as 135-0 degrees.

### Table: Elbow ROM by Age

<table>
<thead>
<tr>
<th>Age Interval</th>
<th>Ambulatory N (Obs)</th>
<th>Ambulatory N (Ind.)</th>
<th>Non-Ambulatory N (Obs)</th>
<th>Non-Ambulatory N (Ind.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;4.9</td>
<td>62</td>
<td>29</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5.0 to 8.9</td>
<td>683</td>
<td>216</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>9.0 to 12.9</td>
<td>375</td>
<td>158</td>
<td>189</td>
<td>96</td>
</tr>
<tr>
<td>13.0 to 16.9</td>
<td>144</td>
<td>70</td>
<td>322</td>
<td>138</td>
</tr>
<tr>
<td>17+</td>
<td>32</td>
<td>17</td>
<td>525</td>
<td>147</td>
</tr>
</tbody>
</table>

The box plot visually represents the distribution of elbow ROM values for each age group, with separate boxes for ambulatory and non-ambulatory patients.
### Knee ROM by Age (years)

<table>
<thead>
<tr>
<th>Age interval</th>
<th>Ambulatory N (Obs)</th>
<th>Ambulatory N (Ind.)</th>
<th>Ambulatory Median</th>
<th>Ambulatory Min</th>
<th>Ambulatory Max</th>
<th>Non-ambulatory N (Obs)</th>
<th>Non-ambulatory N (Ind.)</th>
<th>Non-ambulatory Median</th>
<th>Non-ambulatory Min</th>
<th>Non-ambulatory Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;4.9</td>
<td>103</td>
<td>51</td>
<td>0</td>
<td>-5</td>
<td>15</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>-30</td>
<td>-30</td>
</tr>
<tr>
<td>5.0 to 8.9</td>
<td>684</td>
<td>216</td>
<td>0</td>
<td>-45</td>
<td>20</td>
<td>4</td>
<td>3</td>
<td>-30</td>
<td>-30</td>
<td>0</td>
</tr>
<tr>
<td>9.0 to 12.9</td>
<td>365</td>
<td>154</td>
<td>0</td>
<td>-50</td>
<td>25</td>
<td>48</td>
<td>35</td>
<td>-20</td>
<td>-70</td>
<td>5</td>
</tr>
<tr>
<td>13.0 to 16.9</td>
<td>140</td>
<td>69</td>
<td>0</td>
<td>-65</td>
<td>15</td>
<td>87</td>
<td>52</td>
<td>-30</td>
<td>-90</td>
<td>20</td>
</tr>
<tr>
<td>17+</td>
<td>29</td>
<td>15</td>
<td>0</td>
<td>-15</td>
<td>5</td>
<td>131</td>
<td>69</td>
<td>-40</td>
<td>-120</td>
<td>0</td>
</tr>
</tbody>
</table>

**Normal Range:** 140-0
Ankle Dorsiflexion ROM

Ankle ROM by Age (years)

Not on GC
On GC
# Ankle Dorsiflexion ROM

<table>
<thead>
<tr>
<th>Age interval</th>
<th>Ambulatory</th>
<th></th>
<th></th>
<th></th>
<th>Non-ambulatory</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>N (Obs)</td>
<td>N (Ind.)</td>
<td>Median</td>
<td>Min</td>
<td>Max</td>
<td>N (Obs)</td>
<td>N (Ind.)</td>
<td>Median</td>
<td>Min</td>
</tr>
<tr>
<td>&lt;4.9</td>
<td>100</td>
<td>51</td>
<td>10</td>
<td>-10</td>
<td>30</td>
<td>0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5.0 to 8.9</td>
<td>685</td>
<td>216</td>
<td>5</td>
<td>-50</td>
<td>25</td>
<td>3</td>
<td>2</td>
<td>-7</td>
</tr>
<tr>
<td>9.0 to 12.9</td>
<td>365</td>
<td>155</td>
<td>0</td>
<td>-52</td>
<td>20</td>
<td>42</td>
<td>35</td>
<td>-20</td>
</tr>
<tr>
<td>13.0 to 16.9</td>
<td>137</td>
<td>67</td>
<td>0</td>
<td>-70</td>
<td>15</td>
<td>61</td>
<td>39</td>
<td>-25</td>
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<tr>
<td>17+</td>
<td>29</td>
<td>15</td>
<td>-10</td>
<td>-35</td>
<td>5</td>
<td>87</td>
<td>46</td>
<td>-30</td>
</tr>
</tbody>
</table>

Normal Range: 20 degrees
Functional =10 degrees
12 month change in Ankle ROM
Progression of Contractures
• Contracture progression did not significantly differ between groups
  – Compared GC

<table>
<thead>
<tr>
<th>Age interval</th>
<th>Not on GC at visit</th>
<th>On GC at visit</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N (Obs)</td>
<td>N (Ind.)</td>
</tr>
<tr>
<td>&lt;4.9</td>
<td>45</td>
<td>33</td>
</tr>
<tr>
<td>5.0 to 8.9</td>
<td>74</td>
<td>51</td>
</tr>
<tr>
<td>9.0 to 12.9</td>
<td>13</td>
<td>9</td>
</tr>
<tr>
<td>13.0 to 16.9</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>17+</td>
<td>4</td>
<td>3</td>
</tr>
</tbody>
</table>
ROM Stratified by GC Use

Wrist ROM by Age (years)

Elbow ROM by Age (years)

Knee ROM by Age (years)

Ankle ROM by Age (years)
## Knee Strength and ROM: Predictors of Timed Function Tests

<table>
<thead>
<tr>
<th>Outcome</th>
<th>N (obs)</th>
<th>N (indiv.)</th>
<th>Knee extensor</th>
<th>Ankle ROM</th>
<th>Age</th>
<th>Steroid use</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Coefficient (p-value)</td>
<td>Coefficient (p-value)</td>
<td>Coefficient (p-value)</td>
<td>Coefficient (p-value)</td>
</tr>
<tr>
<td>Run/walk velocity</td>
<td>1073</td>
<td>265</td>
<td>0.042 (p&lt;0.001)</td>
<td>0.009 (p&lt;0.001)</td>
<td>-0.086 (p&lt;0.001)</td>
<td>0.193 (p&lt;0.001)</td>
</tr>
<tr>
<td>Climb velocity</td>
<td>1033</td>
<td>265</td>
<td>0.012 (p&lt;0.001)</td>
<td>0.002 (p&lt;0.001)</td>
<td>-0.017 (p&lt;0.001)</td>
<td>0.039 (p&lt;0.001)</td>
</tr>
<tr>
<td>Stand velocity</td>
<td>954</td>
<td>264</td>
<td>0.007 (p&lt;0.001)</td>
<td>0.002 (p&lt;0.001)</td>
<td>-0.020 (p&lt;0.001)</td>
<td>0.036 (p&lt;0.001)</td>
</tr>
</tbody>
</table>
Prediction of Loss of Functional Milestones

Log rank P value < 0.0001
Stretching and Bracing

CINRG DNHS

NIFD Survey
CINRG DMD Natural History Study (NIFD Study)

• Use of Night time splints and Stretching

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All subjects N=340</th>
<th>Ambulatory only n=194</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Night Splints</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Never Prescribed</td>
<td>125 (36.8%)</td>
<td>73 (37.6%)</td>
</tr>
<tr>
<td>Have used in the past</td>
<td>74 (21.8%)</td>
<td>19 (9.8%)</td>
</tr>
<tr>
<td>Uses occasionally</td>
<td>18 (5.3%)</td>
<td>14 (7.2%)</td>
</tr>
<tr>
<td>Uses regularly or daily</td>
<td>111 (32.8%)</td>
<td>83 (37.6%)</td>
</tr>
<tr>
<td>No data</td>
<td>12 (3.5%)</td>
<td>5 (2.6%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>All subjects N=340</th>
<th>Ambulatory only n=194</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Stretching</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Never Prescribed</td>
<td>95 (27.9%)</td>
<td>58 (29.9%)</td>
</tr>
<tr>
<td>Have used in the past</td>
<td>46 (13.5%)</td>
<td>10 (5.2%)</td>
</tr>
<tr>
<td>Uses occasionally</td>
<td>56 (16.5%)</td>
<td>33 (17.0%)</td>
</tr>
<tr>
<td>Uses regularly or daily</td>
<td>135 (39.7%)</td>
<td>89 (29.9%)</td>
</tr>
<tr>
<td>No data</td>
<td>8 (2.4%)</td>
<td>4 (2.1%)</td>
</tr>
</tbody>
</table>
Are there differences in ankle ROM: Night splints vs. Stretching?

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Right angle dorsiflexion</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>Mean ± SD</td>
</tr>
<tr>
<td>Stretching</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Have used</td>
<td>43</td>
<td>-6.0 ± 13.10</td>
</tr>
<tr>
<td>Daily use</td>
<td>89</td>
<td>1.0 ± 10.7</td>
</tr>
<tr>
<td>Night splints</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Have used</td>
<td>33</td>
<td>-7.7 ± 15.0</td>
</tr>
<tr>
<td>Daily use</td>
<td>83</td>
<td>0.4 ± 9.7</td>
</tr>
</tbody>
</table>

* p-value from non-parametric comparison using Wilcoxon rank sum test
DMD Natural History Study Summary

• **DMD ROM**
  – High variability in ROM with overall greater variability in the non-ambulatory
  – Better ROM for those on GC
  – No clinical difference in ROM progression with respect to GC use
    • 12-months may be too short to observe clinically meaningful changes in ROM
  – Knee extension and ankle ROM significantly associated with all 3 timed tests
    • Knee strength > ankle ROM

• **Stretching and Braces**
  – 30% never prescribed stretching
  – Daily/Frequent reported use of stretching/bracing significantly associated with ankle ROM
Stretching in Neuromuscular Disease

- **Systematic Review**: Effects of Casting, orthoses, stretching, and supported standing devices
- 16 studies – effectiveness of stretching
- Populations: CP, CMT, DMD
- Age: 20 months – 30 years old
- Outcome for stretching: prevention of contractures
  - Insufficient evidence to support/refute PROM or positioning
  - Generally studies demonstrated limited/weak evidence: improved ROM immediately after stretching and loss of ROM when stretching stopped

*Effectiveness of Stretch Interventions for Children with Neuromuscular Disabilities: evidence-Based Recommendations Craig J. et al. 2016.*
Stretch for the treatment and prevention of contractures

\[49\] studies with \[2135\] participants met the inclusion criteria

- ‘We do not recommend further studies looking at the short-term effects of stretch on joint mobility in either people with neurological or non-neurological conditions because the quality of evidence indicating that stretch is ineffective is high’

- ‘... there is no theoretical basis upon which to believe that stretch may have long-term effects on joint mobility in the absence of a short-term effect.’

- ‘There may be worth in examining the effectiveness of stretch administered with other interventions’

Harvey et al, Cochrane 2017
Benefits of Stretching…Besides preventing contractures

- Stretching may not be proven to improve or prevent contractures but has a multitude of benefits that indirectly affect range of movement and function
  - Increased blood flow to the muscles reducing arterial stiffness
  - Increased lubrication to articular surfaces of the joints
  - Muscle stiffness may lead to increased injury
  - Stretching related to amounts of collagen with known effects of collagen on exercise and aging
    - Movement decreases stiffness---Improves mobility; ease of movement
- Stretching Recommendation
  - Most studies agree on a total of 90 second stretch (30 sec x 3 reps) with rest period of 20-30 seconds to decrease stiffness
Take Home…

• Contractures have an effect on
  • Muscle Force Production
  • Mobility and Function
  • Pain
  • “Stiffness”
• Prevention is KEY
  • If you have muscle weakness, then you need to be sure to stretch opposing muscle group
  • Positioning is a good way to maintain alignment
  • Ensuring good muscle length is key to best possible benefits of drug treatments
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=6eHLt3KAOtg

• CINRG Website
StretchOUT Stretch Instruction and Workout
http://www.cinrgresearch.org/stretchvideo2/index.cfm
Night Splints- Across all stages

• Begin the routine early
• Maintains prolonged stretch 6+hours/night
• Shown to be effective (Scott et al 1981, Hyde et al 2000)
• Best when forefoot is supported
  – Medial trim lines
  – Shoes vs no shoes-consider as they get older
• Night vs Day use
• Increased padding
Exercise
What we know now!
Exercise

What do we know.....

• *No exercise* leads to muscle atrophy
  – Encourage self-modulation
  – Schedule rest breaks
• Too much exercise increases *muscle breakdown*
  – *Rhabdomyolysis*
• *Eccentric contractions* are more damaging to the muscle cell
  ** Isometric → Concentric → Eccentric**
• Younger boys benefit from exercise more than older boys
• Boys with DMD are 40% less active than age matched peers
  *(McDonald, 2000)* **supported by other studies**
Exercise Recommendations

• Age appropriate recreational activities as opposed to strengthening regimes
• Concentric low load or isometric versus eccentric high load - stay submaximal
• Balance activity with rest: Don’t overdo
• Incorporate balance and coordination skills
• Activities should be fun and promote self esteem and social interaction
Subjects: 8 ambulatory boys with DMD (8.3 + 0.7 yrs)

Time Points: Each subject tested at 3 time points over 5 days

Safety Measures: performed 48 hours before and 48 hours after
- T₂ weighted magnetic resonance imaging (MRI)
- Pain assessment
- Clinical examination & serum creatine kinase (CK) levels

Mild to Moderate - Intensity Resistance Exercise:
6 reps/4 sets (isokinetic dynamometer) 4 exercises

- Isometric maximal voluntary contraction (MVC) was determined for the knee extensors and knee flexors at both 60 and 30 degrees of knee flexion
- Isometric exercise mild level 30% MVC (n=4)
- Isometric exercise moderate level 50% MVC (n=4)
Data Analyses

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

• Subjects tolerated all testing very well
• Participants @ 30% MVC worked at 31.5% ± 4.7% of their MVC
  * Able reach that level
• Participants @ 50% MVC worked at 41.8% ± 3.3% of their MVC
  * Worked under level
• Average $T_2$ change only increased 1.8% ± 2.6% (not significant)
• 48 hrs post-exercise no pain reported
• CK values were not indicative of muscle injury
Phase 2

• In home exercise program followed weekly for 12 weeks
  – Closely Monitored
  – Remotely followed by Skype 2 hr/day, 3 days week

• Follow up in person
  – Return for 1 day visit after 6 weeks
  – Return for 1 day visit after 12 weeks
  – All previous testing repeated on these visit days
    • CK levels, MRI, Pain scale

Future work may provide information of proper dose/intensity of exercise that is safe and feasible for boys with DMD
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, Topaloğlu H

- **Subjects** = 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**
Many Options for Cycling!!  
Family Fun 😊

Summary

Assisted cycling and cycling without excessive resistance can be beneficial.
Avoid excessive resistance
Avoid hills or give assistance
Add power when needed
Aquatic Exercise

Aquatic Therapy or Just Family Fun
Water is Fun!!!

Permission from Wavetherapies.com
Seattle, WA
Aquatic Activities

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

• Stay active
• Stretch to maintain ROM so you can stay active and participate

• **Don’t overdo!!** Build in **Rest Periods**
  – Self modulation

• **Fatigue** is *REAL*, Differences in endurance
• Add power when needed for energy conservation to keep submaximal
• Research is promising in this arena with results indicating exercise may help
“It is what we think we know that keeps us from learning”
-Claude Bernard

Thank you....
Transfers when needed: Considerations

- Stand pivot with maximum assist
- Sliding board (Supervised → Max Assist)
- Maximum lift of caretaker-
  - “Watch your back”
- Use of mechanical transfer lift
  - Hoyer lift
  - Ceiling lift
- Minimize transfers with rolling bath chair
- Mover
Assistive technology

- Blue tooth devices
- Siri/Dragon Speak
- Alexa [www.amazon.com](http://www.amazon.com)
- Environmental control systems
  - Google home & Amazon Echo
- Computers / tablets / smart phones
  - Alternative keyboards/ touch pad screens
  - Electronic pointing devices
- Voice assist (amplifiers)
- Glassouse [http://glassouse.com](http://glassouse.com)
- TouchTapSwipe guide
  [www.dmdpathfinders.org.uk](http://www.dmdpathfinders.org.uk)
Occupational Therapy
How is the goal of independence achieved?

• Maintaining Body Functions (as much as possible)
  – Physical, Neuromuscular & Cardiovascular
  – Mental
  – Sensory

• Addressing the performance patterns and context of the occupation
  – Environmental modification
  – Social Modifications
  – Habit, Routine, Ritual and Role Analysis
Splinting
Addressing Body Function

Purpose:
• Prevent contractures
• Prevent pain
• Maintain function and mobility of a joint
• Allow increased function by placing joint in ideal position

Thumb support
Elbow Flexion Contracture Block Splint
Night resting hand splint
Adaptive Equipment
Addressing Occupations

Purpose
• Enable independence
• Improve quality of life
• Increase accessibility

Note:
• Insurance coverage varies
• Options are endless
Socialization

- Engaging in enjoyable socialization (especially as the physical limitations progress) is critical.
  - mental health well-being
  - improving quality of life
- Places to explore socialization include:
  - social skills group
  - support groups
  - special interest clubs
  - adaptive sports programs


Equipment

Medical Vendor

Therapist

Insurance Funding

Family
Early Mobility Devices

- 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 distance community events
- Can be transported in most vehicles
Scooters and alternative motorized systems

GO-GO scooter
- more portable than w/c
- negatives:
  - large turning radius
  - poor/no seating support
  **poor posture in scooter can contribute to contractures if used too much
- UE’s may get tired from reaching for handles

Go-Go® Elite Traveler Plus

http://www.pridemobility.com/gogo/
Scooters and alternative motorized systems

EZ Lite Cruiser

http://www.ezlitecruiser.com/
Scooters and alternative motorized systems

Zappy
- more portable than w/c
- can stand or sit
- ~13 mph

EV Stand/Ride

http://www.zapworld.com/vehicles/zappy-pro-flex-500
Portable power assist wheelchairs

http://www.alber-usa.com

Efix

E motion power assist
or
twion power assist

Smart Drive
Power Wheelchair purchase

• Drive Mechanism
  – Front wheel
  – Mid-wheel
  – Rear wheel

• Power Options
  – Power standing feature
  – Power tilt and/or recline
  – Power seat elevation
  – Separately elevating power elevating leg rest
Seating for young men

- Emphasis on comfort
- Pressure relieving cushion
- Larger head rest
- Wider arm rests
- Elbow blocks
- Padded footrests
- Joystick modifications
- Attendant controls
Supported Standing

Permobil F5 stand & drive
www.permobilus.com/f5vs.php

Stand & Drive chairs
MedTrade Expo and ATIA Conferences

Showcases products like home medical equipment, latest innovations in mobility, respiratory, sleep, rehab, and aids etc…

ATIA Conferences:
Where the Assistive Technology Community Meets to...
Network, Learn, and Share

Assistive Technology Industry Association
Save the Date for ATIA 2018
Orlando, Florida
Pre-Conference Seminars: January 30 -31, 2018
Conference: January 31 – February 3, 2018
School and Beyond
Common Campus Modifications

• Extra time to get to class
  – Classes close together if possible
• Individualized PE modifications
  – With extended breaks as needed
  – Adaptive sports (involve the whole class for socialization)
• Use of accessible bathroom
• Key for elevator (for those still ambulating)
• Staff trained in transfers (using a lift)
• Assistance for lunch set up
• Safety evacuation plan
Common Classroom Modifications

- Copies of PowerPoints or notes
- Extra set of books for home
- Use of laptop/tablet or scribe
- Modified homework to limit fatigue
- Extra time for standardized or timed tests
- Oral tests if writing difficult
- One-on-one aide
- Individualized and preferential seating or desk
Transition to Higher Education

Resources

- State Vocational Rehabilitation Department STAR program
- Scholarships & Financial Aids
  - https://www.mda.org/young-adults/resources
- College or University Office of Student Disability Services
- Center for Independent Living (dependent on location)
What is Vocational Rehabilitation?

http://www.rehabworks.org

**What is it?**

Federal program designed to:

- Enhance independence of people with disabilities by helping them find and maintain employment

Located throughout the state by county and city

**Services**

- Can vary at each location
- Ranges from:
  - Career Counseling
  - Job Training & Placement
  - Supported Employment
  - Assistive Tech training & support
  - Medical & Psychological Assessment
- STAR Program
  - Ages 15-21 for students
School Resources: PPMD

• **Education Matters**: A Parent’s Guide

For Duchenne Muscular Dystrophy

• **Education Matters**: A Teacher’s Guide

• **Education Matters**: Adaptive Physical Education:

  A PE teacher’s guide to Duchenne Muscular Dystrophy

• **Education Matters**: Learning and Behavior in DMD for parents and educators

• Support through others- Network with families that have been through it!

• Get your rehabilitation team involved
THANK YOU!

Tina Duong: trduong@Stanford.edu
PPMD: www.parentprojectmd.org