Physical Therapy Management of the Patient with Hemophilia Across the Lifespan

Janet B. Tankersley, PT, DPT, PCS
Ryan
Objectives

• Identify key impairments, activity limitations and participation restrictions attributed to aging with hemophilia.
• Discuss health-related concerns of aging with hemophilia.
• Describe patient-reported outcome measures specific to hemophilia.
• Discuss preventative and wellness interventions for patients with hemophilia.
• Guidelines for referral to outpatient physical therapy.
### Demographics

#### HTC Population Profile Patient Characteristics, Factor VIII and Factor IX

Data reported from 1/1/2012 through 9/30/2014

<table>
<thead>
<tr>
<th>Age</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
<th>Severity Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;2</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2–10</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11–19</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>20–44</td>
<td>1171 (29)</td>
<td>631 (34)</td>
<td>2338 (39)</td>
<td>69 (41)</td>
</tr>
<tr>
<td>45–64</td>
<td>653 (14)</td>
<td>232 (13)</td>
<td>620 (10)</td>
<td>23 (14)</td>
</tr>
<tr>
<td>65+</td>
<td>296 (7)</td>
<td>68 (4)</td>
<td>92 (2)</td>
<td>8 (5)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Gender†</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
<th>Severity Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>3402 (86)</td>
<td>1822 (89)</td>
<td>5958 (99)</td>
<td>119 (7)</td>
</tr>
<tr>
<td>Female</td>
<td>575 (14)</td>
<td>28 (2)</td>
<td>31 (1)</td>
<td>49 (29)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Ethnicity</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
<th>Severity Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hispanic, Latino, or Spanish origin</td>
<td>770 (19)</td>
<td>334 (18)</td>
<td>973 (16)</td>
<td>37 (22)</td>
</tr>
<tr>
<td>Not Hispanic, Latino, or Spanish origin</td>
<td>3190 (80)</td>
<td>1512 (82)</td>
<td>5005 (84)</td>
<td>131 (78)</td>
</tr>
<tr>
<td>Unknown</td>
<td>17 (0)</td>
<td>4 (0)</td>
<td>11 (0)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Race</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
<th>Severity Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>American Indian/Alaska Native</td>
<td>64 (2)</td>
<td>13 (1)</td>
<td>53 (1)</td>
<td>3 (2)</td>
</tr>
<tr>
<td>Asian</td>
<td>84 (2)</td>
<td>74 (4)</td>
<td>280 (5)</td>
<td>9 (5)</td>
</tr>
<tr>
<td>Black or African American</td>
<td>242 (6)</td>
<td>276 (15)</td>
<td>939 (16)</td>
<td>20 (12)</td>
</tr>
<tr>
<td>Native Hawaiian or other Pacific Islander</td>
<td>7 (0)</td>
<td>8 (0)</td>
<td>31 (1)</td>
<td>1 (1)</td>
</tr>
<tr>
<td>White</td>
<td>3528 (89)</td>
<td>1449 (78)</td>
<td>4602 (77)</td>
<td>135 (80)</td>
</tr>
<tr>
<td>More than one of these</td>
<td>20 (1)</td>
<td>9 (0)</td>
<td>40 (1)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Unknown</td>
<td>32 (1)</td>
<td>21 (1)</td>
<td>44 (1)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

(CDC, 2014)
Hemophilia A and B population

- Individuals 45 years and older comprise 15% of the total hemophilia population

- Individuals older than 65 years comprise 2%
  - very little clinical data to guide recommendations
Complications of aging

- Intracranial Hemorrhage
  - death rate of 30% in individuals with hemophilia age 50 and older (Darby, 2004)
- Joint disease (Mauser, 2009)
- Cardiovascular disease (age, hypertension, diabetes, and hyperlipidemia risk factors) (Kulkarni, 2005)
- Viral Disease (Philipp, 2010)
  - HIV, Hepatitis C
- Cancer
  - HCV-associated hepatocellular carcinoma
  - HIV-associated non-Hodgkin’s lymphoma
ICF Components

- **Body functions**: Physiological functions of body systems
- **Body Structures**: Structural or anatomical parts of the body
- **Activities**: Execution of a task or action by an individual
- **Participation**: Persons involvement in a life situation
- **Environmental Factors**: All aspects of the external world that effect the person’s functioning
- **Personal Factors**: Aspects internal to the person that effect the person’s functioning
Body Functions

• Intra-articular hemorrhage (>90%)
• Intracranial hemorrhage
• Hematomas
• Intramuscular hemorrhage
• Intraneural hemorrhage (6%, (Katz, 1991)
Body Structures

- Fibrosis and contractures of joints
- Compartment syndrome
- Nerve palsies
- Heterotopic ossification
- Pseudotumors

*Complications increase with aging

(Vanderhave, 2012)
Articulations most susceptible to Hemophilic Arthropathy
Impairments

- Decreased ROM
- Decreased strength
- Edema
- Pain
- Decreased proprioception
- Decreased coordination
- Balance deficits
- Decreased endurance
Activity Limitations

• Lying/sitting/kneeling/standing
• Walking
• Self-care
• Household tasks
• Leisure activities and sports
• Driving
Participation Restrictions

- Work or School Life
- Family Life
- Social Life
- Team sports
Chronic Hemophilic Arthropathy

Conservative management

• Serial casting to assist in correcting deformities
• Bracing and orthotics to support painful and unstable joints
• Walking aids or mobility aids to decrease stress on weight-bearing joints
• Adaptations to the home, school, or work environment to allow participation in community activities and employment and to facilitate activities of daily living.

(Alhaosawi, 2014)
Preventative Strategies

• Patient and Family Education
• Acquiring crutches before a bleed
• Maintain objective data to follow the clinical process
• Musculoskeletal ultra-sound for assessment and to guide intervention (Wyasure, 2016)
Outcome Measures
**Table 1 Clinical evaluation score of hemophilic arthropathy (0–12)**

<table>
<thead>
<tr>
<th>Clinical sign</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Swelling</td>
<td>0 = None</td>
</tr>
<tr>
<td></td>
<td>2 = Present</td>
</tr>
<tr>
<td>Muscle atrophy</td>
<td>0 = Absent</td>
</tr>
<tr>
<td></td>
<td>1 = Present</td>
</tr>
<tr>
<td>Axial deformity</td>
<td>0 = No deformity</td>
</tr>
<tr>
<td></td>
<td>1 = Less than 10°</td>
</tr>
<tr>
<td></td>
<td>2 = More than 10°</td>
</tr>
<tr>
<td>Crepitus on motion</td>
<td>0 = None</td>
</tr>
<tr>
<td>Flexion contracture</td>
<td>1 = Present</td>
</tr>
<tr>
<td></td>
<td>1 = Less than 15° fixed flexion contracture</td>
</tr>
<tr>
<td></td>
<td>2 = More than 15°</td>
</tr>
<tr>
<td>Range of motion</td>
<td>0 = Loss of up to 10% of full range of motion</td>
</tr>
<tr>
<td></td>
<td>1 = Loss of 10–33.33% of motion</td>
</tr>
<tr>
<td></td>
<td>2 = Loss of more than 33.33% of motion</td>
</tr>
<tr>
<td>Instability</td>
<td>0 = None</td>
</tr>
<tr>
<td></td>
<td>1 = Present but normal function possible</td>
</tr>
<tr>
<td></td>
<td>2 = Creates functional deficit, requires bracing</td>
</tr>
</tbody>
</table>

Clinical Evaluation Score of the World Federation of Hemophilia, (Gilbert, 1993; Gurcay, 2006)
Hemophilia Joint Health Score

http://www1.wfh.org/docs/en/Publications/Assessment_Tools/HJHS_Summary_Score.pdf

- 11-Item scoring tool for assessing joint impairment
- For ages 4-18 years of age
- Focuses on the six joints most affected in hemophilia (Ankles, Knees, Elbows)
- Global gait score
- Scores range from 0 to 124; 0= no identifiable joint impairment

(Hilliard, et al, 2006)
The HAL measures the impact of hemophilia on self-perceived functional abilities in adults. It contains 42 multiple choice questions in seven domains:

- Lying/sitting/kneeling/standing (8 items)
- Functions of the legs (9 items)
- Functions of the arms (4 items)
- Use of transportation (3 items)
- Self-care (5 items)
- Household tasks (6 items)
- Leisure activities and sports (7 items)

(van Genderen, 2004)
Paediatric Haemophilia Activities List (PedHAL)  

The PedHAL should be selected to measure the impact of hemophilia on self-perceived functional abilities in children. The current version (0.11) consists of 53 items in seven domains:

- Sitting/kneeling/standing (10 items)
- Functions of the legs (11 items)
- Functions of the arms (6 items)
- Use of transportation (3 items)
- Self care (9 items)
- Household tasks (3 items)
- Leisure activities and sports (11 items)
Haemophilia-specific QoL

• For children with hemophilia and their parents
• I: 21 items for children aged 4-7
• II: 64 items for children aged 8-12
• III: 77 items for adolescents aged 13-16
• physical health, feeling, view of yourself, family, friends, perceived support, others, sports and school, dealing with hemophilia, treatment, future, relationships, global health
Referral to Outpatient PT

- Patient could benefit from a supervised exercise program
- Patient has activity or participation limitations that need further evaluation and development of treatment strategies
- Patient could benefit from a wellness program
Questions ?
References