Outcome measures for older people with haemophilia

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European Haemophilia Consortium
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## Life expectancy in haemophilia

<table>
<thead>
<tr>
<th>Period</th>
<th>Country</th>
<th>Type</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>1830-1920</td>
<td>Sweden</td>
<td>severe</td>
<td>11</td>
</tr>
<tr>
<td>1920-1940</td>
<td>Sweden</td>
<td>severe</td>
<td>23</td>
</tr>
<tr>
<td>1940-1960</td>
<td>Sweden</td>
<td>severe</td>
<td>28</td>
</tr>
<tr>
<td>1930-1957</td>
<td>Sweden</td>
<td>severe</td>
<td>25</td>
</tr>
<tr>
<td>1960-1980</td>
<td>Sweden</td>
<td>severe</td>
<td>57</td>
</tr>
<tr>
<td>1975-1980</td>
<td>United Kingdom</td>
<td>severe</td>
<td>69</td>
</tr>
<tr>
<td>1973-1986</td>
<td>The Netherlands</td>
<td>severe</td>
<td>63</td>
</tr>
<tr>
<td></td>
<td></td>
<td>moderate</td>
<td>65</td>
</tr>
<tr>
<td></td>
<td></td>
<td>mild</td>
<td>69</td>
</tr>
</tbody>
</table>

Life expectancy (1992-2001)

<table>
<thead>
<tr>
<th>Severity</th>
<th>Year 1</th>
<th>Year 2</th>
<th>Year 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>All</td>
<td>67</td>
<td></td>
<td></td>
</tr>
<tr>
<td>severe</td>
<td>59</td>
<td></td>
<td></td>
</tr>
<tr>
<td>moderate</td>
<td>67</td>
<td></td>
<td></td>
</tr>
<tr>
<td>mild</td>
<td>73</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

(Plug, J Thromb Haemost 2006)

(Dutch men: 76 yr)
Similar data from Italy

(Tagliaferri, Haemophilia 2010)
Consequences

- consequences of increased life-expectancy: more older patients

- age-distribution result of
  - current and past birth rates
  - current and past death rates
Birth excess
General population 2015
Haemophilia population

(Plug, Blood 2004)
Conclusion

• Life-expectancy has increased much
  • similar to population

• Consequence: age-distribution changing
  • proportionally more older patients

• Age distribution will vary from place to place
  • dependent on birth rates last decades (baby-boom)
  • dependent on previous excess deaths due to HIV and HCV
  • dependent on reproductive choices
Current haemophilia population

- born before 1960
  - 55+
  - major sequelae from haemophilia
  - viral infections

- born 1960-1990
  - 25-55 yr
  - moderate sequelae haemophilia
  - viral infections

- born 1990-2016
  - <25 yr
  - mild sequelae haemophilia

- born after 2016
  - generation that may be cured
Changing perspectives

The 50-year old patient with severe haemophilia

• in 1965
  • was not alive

• In 1990
  • severe arthropathy
  • HIV

• in 2016
  • moderate arthropathy
  • HIV, HCV

• In 2040
  • No or mild arthropathy
  • No viral infection
There is not one Europe

(WFH, 2014)
The ageing patient

- the bleeding disorder
  - type and frequency of bleeding

- the ageing process
  - diseases of advancing age

- sequelae of previous events
  - viral infections
  - arthropathy

- interactions between all the above
Uncharted territory
The bleeding disorder

European CHESS project (n=541)

<table>
<thead>
<tr>
<th>Age Range</th>
<th>Annual Bleeding Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>18-30</td>
<td>9.99</td>
</tr>
<tr>
<td>31-40</td>
<td>10.85</td>
</tr>
<tr>
<td>41-50</td>
<td>12.38</td>
</tr>
<tr>
<td>51-60</td>
<td>13.85</td>
</tr>
<tr>
<td>61+</td>
<td>13.42</td>
</tr>
</tbody>
</table>

(O’Hara, personal communication)
Diseases of advancing age

- malignancies
- atherosclerotic diseases
  - atrial fibrillation
  - ischaemic stroke
  - myocardial infarction
  - heart failure
- Alzheimer diseases
- chronic obstructive pulmonary disorder (COPD)
- osteoarthritis
- frailty
- ....
Sequelae of previous events

European CHESS project (n=541)

Mean number of target joints

<table>
<thead>
<tr>
<th>Range</th>
<th>Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>18-30</td>
<td>1.04</td>
</tr>
<tr>
<td>31-40</td>
<td>1.16</td>
</tr>
<tr>
<td>41-50</td>
<td>1.25</td>
</tr>
<tr>
<td>51-60</td>
<td>1.43</td>
</tr>
<tr>
<td>61+</td>
<td>0.89</td>
</tr>
</tbody>
</table>
Interactions

• stenting for coronary stenosis in haemophilia
  • dual antiplatelet treatment?

• type 2 diabetes in haemophilia
  • what type of exercise?

• malignancies in virus-infected patients
  • surgical approaches?
  • effects of radio- en chemotherapy?
Conclusion

- ageing haemophilia patients offer complex medical questions
- ageing patients differ by cohort and location
- the number of older patients grows rapidly
- targeted research needed
  - gerontologists
  - epidemiologists
  - patients