Ageing and haemophilia

Dr. Michelle Lavin
Senior Clinical Research Fellow
NCC/RSCI
Ageing and haemophilia

- Joint damage
- Bleeding risk
- Operations
- Inhibitors
- Dentition
- Bone health
Ageing and general health

- General health
- Cardiovascular disease
- High blood pressure
- Poor kidney function
- Prostate issues
Arthropathy

• Physiotherapy and Exercise
• Maintain muscle strength, balance, bone health
• Orthotics
• Adaptation/Aids
• Radiosynovectomy
• Orthopaedic surgery
Bleeding

Variable bleeding patterns

• Less joint bleeding due to decreased activity, synovium burnt out
  BUT
• Any bleeding into damaged joints may be problematic

• Prophylaxis
  • Primary ?continue
  • Secondary
Pain management

• Paracetamol ± Codeine
• Etoricoxib/Arcoxia – anti-inflammatory
• Lignocaine patches
• Opiates

• Non-medications techniques – exercise, sleep, meditation
Operations

• Surgical procedures increase with age

• Particular importance people with mild haemophilia
Inhibitor risk with age

Hay et al. Blood 2011
Your own Factor VIII

None
Your own Factor VIII
Administered
Factor VIII
Body recognises as a foreign substance and makes an inhibitor
Binding of inhibitor to administered Factor VIII stops it working as effectively
Binding of inhibitor to your own Factor VIII may reduce Factor VIII levels.
• Risk of developing inhibitor with intense exposure to factor concentrate

• Mild haemophilia - may cross react with your own Factor VIII
  • Lower Factor VIII levels
Osteoporosis
Why does this matter in haemophilia

• UK, case control, n=37
  • PWH = 5% osteoporosis, 24% osteopenia; (controls = 0%, 3%)
  • 91% vs 92% controls had low Vitamin D
  • ? Peak BMD reached

• Low Vitamin D, reduced BMD
• Greece (n=104); Italian study (n=78); Arizona (n=30)

1. Wells et al. Haemophilia 2014
3. Linari et al. Haemophilia 2013
• Meta-analysis, 13 studies
• Men with haemophilia -significant reduction in both lumbar spine and hip BMD
• Appears to begin in childhood

Paschou et al. Osteoporos Int 2014
NCC Screening programme

• Bloods and DXA scan **all** patients with moderate and severe Haemophilia A & B

• Updated on results
Osteoporosis Prevention

Maximise and maintain bone mass – weight bearing or resistance exercise (adjust the factor treatment if needed)
Osteoporosis – More Prevention!

Vit D – Get out in the Sun (but not too much!)

67.3% of 153 patients admitted with hip fracture had low Vita D levels

No excess alcohol, No Smoking
Calcium in your diet

SMART
Safe Sun Exposure NHS Advice

• **S**pend time in the shade between 11.00 am and 3.00 pm.
• **M**ake sure you never burn.
• **A**im to cover up with a T-shirt, hat and sunglasses.
• **R**emember to take extra care with children.
• **T**hen use factor 15+ sunscreen.
You should have >400 IU of Vitamin D per day......

<table>
<thead>
<tr>
<th>Food Item</th>
<th>Vitamin D Content</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cow’s milk</td>
<td>3–40 IU/L</td>
</tr>
<tr>
<td>Fortified infant formula</td>
<td>400 IU/L</td>
</tr>
<tr>
<td>Butter</td>
<td>35 IU/100g</td>
</tr>
<tr>
<td>Egg yolk</td>
<td>20–25 IU/yolk</td>
</tr>
<tr>
<td>Cod liver oil</td>
<td>1360 IU/tsp</td>
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<tr>
<td>Fish</td>
<td>44–624 IU/100 g</td>
</tr>
<tr>
<td>Yoghurt</td>
<td>89 IU/100g</td>
</tr>
<tr>
<td>Cheese</td>
<td>12–44 IU/100g</td>
</tr>
</tbody>
</table>
Vitamin D – the top 10.....
Oral health

- Importance of dental health
- Bleeding is not normal
- Affect diet, nutrition
- Microbiome may be linked to cardiovascular disease risk
Oral health

Solution?

• Make friends with your Dentist

• Dental letters for Mild and Severe bleeding disorders available
General health

• Cardiovascular disease
• High blood pressure
• BMI
• Bowel screen
• Prostate cancer
Cardiovascular disease - (CVD)

- Systematic review - toward protection from CVD – not significant
- Case control study
  - 100 hemophilia A and B patients; 200 unaffected controls
  - No significant difference in deaths from CVD over 10 years
  - Despite more diabetes and high blood pressure
  - ? protective effect of the hemophilia

CVD

• Carotid intimal thickness - 69 PWH\(^1\)
  • not protected against the development of atherosclerosis
  • related to the traditional cardiovascular risk factors

• HIV - accelerated CVD\(^2\)

% Population obese
BMI and obesity

• 2011 figures

• Additional 34% Irish adults overweight ............

IUNA National Adult Nutrition Survey 2011
BMI

• Increased weight
  • Joints
  • Bleeds
  • How to dose prophylaxis/treatment

• Heart disease and stroke.
• High blood pressure.
• Diabetes.
• Some Cancers
• Breathing problems
Does haemophilia reduce CVD risk

• What to believe......

• Screening risks

• Arcoxia

• No smoking
• If you develop cardiovascular disease – what next?
The dreaded....
• Cardiovascular disease
  • Anticoagulation
  • Antiplatelet
Kidney disease

• ? more common in haemophilia population

• 6 US states - 1993-98
  • x50 fold increased risk death from renal disease

• Cardiovascular risk factors

• Medications (Arcoxia)

1. CKD Prognosis consotium, Lancet 2010 Jun 12;375(9731):2073-81
Blood pressure and bleeding

• Risk for CVD

• High blood pressure may increase bleeding risk

• Uncontrolled high BP – risk of brain haemorrhage
Take home message on cardiovascular disease

• GP checks

• Discussion

• Prevention is better!
Bowel screen

• Postal samples for FOB as part of the national screening programme
  • 60-69 years old every 2 years

• If positive for blood – colonoscopy scheduled

• Advise to contact screening programme and SJH
Risk of prostate cancer

- American Cancer Society
Prostate check

- Annual check 45 - 70 years of age

- PSA alone is **not** a prostate screen
All doom and gloom?

Factor VIII PK may increase with age, not Factor IX

A lot changes in 20 years.....
Future of haemophilia

• Personalised programmes
  • Improved understanding

• Novel treatment
  • Long acting agents
  • Emicizumab
  • Gene therapy
John Glenn, 77
Oldest man in space
• Michael Caine, 84
• 9 films since 2010
• Harriette Thompson, 92
• Completing her 15th marathon (started running in her 70s)
Dr. Frank Schearer, 101
Oldest known water skier
• Age is an issue of mind over matter. If you don’t mind, it doesn’t matter
  • Mark Twain