EOS in inhibitor pts

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PWH today…

- Adults (arthropathy)
- Kids on prophylaxis (Micro-bleeds) ??
- Inhibitor pts
Adult

- Management of daily life
- Physical activity
- Monitoring of target joint: RX, RMN, TAC
- Periodic cycles of FKT
- Pre-rehabilitation
- Post-surgical
Kids

- Sport education
- Monitoring target joints:
  - Baropodometry
  - Ultrasound
  - Gait analysis
European Study on the Orthopaedic Status of patients with haemophilia and inhibitors

“The burden of orthopaedic complications and the impact on quality of life are more severe in patients with haemophilia who have developed an inhibitor compared with those patients without inhibitors.”

Pts with INH with a history of orthopaedic procedures or surgery:

14-35 years: 34%
36-65 years: 66%

Morfini M et al., Haemophilia 2007
Immature joint
Our experience 2009-2016

- Total knee replacement
- Tkr revision
- Total hip replacement
- Thr revision
- Total ankle replacement
- Pseudotumor
- Flat foot
- Arthroscopy

Quantity
Our experience in PWH and INH

Pts **without INH** vs pts **with INH**
(last years)

<table>
<thead>
<tr>
<th></th>
<th>no INH</th>
<th>INH</th>
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<tbody>
<tr>
<td>2011</td>
<td>55</td>
<td>3</td>
</tr>
<tr>
<td>2012</td>
<td>67</td>
<td>5</td>
</tr>
<tr>
<td>2013</td>
<td>58</td>
<td>4</td>
</tr>
<tr>
<td>2014</td>
<td>63</td>
<td>5</td>
</tr>
<tr>
<td>2015</td>
<td>56</td>
<td>3</td>
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<tr>
<td>2016</td>
<td>66</td>
<td>3</td>
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Why?

- Successful ITI
- Other hematological treatment for inhibitor
- Cost
- Complication management
Factor influencing surgeries in PWH

- Factor replacement (or by-passing agents) availability
- Hemophilia center: number of pts
- Team learning curve
- Surgeon attitude
- Hematologist attitude
MDA: What the hematologist has to know about...

- Stiffness
- Axial deviation
- Deformity
- Soft tissue release
- Bone cut
- Synovectomy

Increase bleed expectation...
What the hematologist has to know about… Surgery

<table>
<thead>
<tr>
<th>Total knee replacement</th>
<th>Primary implant</th>
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<tbody>
<tr>
<td></td>
<td>Revision</td>
</tr>
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<table>
<thead>
<tr>
<th>Arthroscopy:</th>
<th>Synovectomy</th>
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<tbody>
<tr>
<td></td>
<td>Debridment</td>
</tr>
</tbody>
</table>

<table>
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<tr>
<th>Different bleeding expectation:</th>
<th>&lt; 500 cc</th>
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<tbody>
<tr>
<td></td>
<td>500 - 800 cc</td>
</tr>
<tr>
<td></td>
<td>800 - 1200 cc</td>
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Knee / Ankle
What the surgeon has to know

- Severity of coagulation factor deficiency
  - on demand or prophylaxis mild hemophilia?

- Comcominant liver disease
  - HBV+/HCV+
  - Liver dysfunction
  - Cirrhosis
  - Thrombocytopenia

- Concomitant HIV infection
  - CD4
  - CD8
  - HIV viremia
What the surgeon has to know

- Information on inhibitor history and current inhibitor status
  - Inhibitor titer
  - Anamnestic response
  - Choice of hemostatic therapy

- Choice of replacement therapy
  - Bolus vs CI
  - In inhibitor pts: high dose factor replacement by-passing agents
Hemophilia, 1998

Treatment of synovitis

Grade I
Transitory synovitis back to normal between bleeds
>3 bleeds in 6mo. Intervene

Grade II
Chronic synovitis does not return to normal between bleeds
Intervene

Grade III
Chronic arthropathy, axial deformities, rigidity
Caution! Unlikely to succeed

Grade IV
Bony of fibrous ankylosis
Not indicated

Fernández-Palazzi, Hemophilia, 1998
Treatment options

Prophylaxis upgrade treatment

Arthrocentesis

Synoviorthesis
  - Chemical synovectomy
  - Radio synovectomy

Synovectomy
  - Arthoscopic
  - Open

Angiographic embolization

TKR
Selective angiographic embolization of knee and elbow arteries is a feasible procedure that can prevent repetitive bleedings.
Synovectomy

bleeding tendency

progressive deterioration of the radiographic appearance
Chronic synovitis in advanced stage of arthropathy

**TKR**

- Higher expected bleeding
- Post-op swelling
- Drain management
- Fibrin seal
After synovectomy
Target joints

Neglected joints
X-ray: HA evolution monitoring
Surgery timing
X-ray: HA evolution monitoring
Surgery timing
Correlation between x-ray and clinical exam

Advanced deformity and joint stiffness

Advanced stage of arthropathy
No correlation between x-ray and clinical exam

good function-no pain

advanced stage of arthropathy
<p>| | |</p>
<table>
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<tbody>
<tr>
<td>NSAIDs</td>
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</tr>
<tr>
<td>Crural analgesia</td>
<td>No</td>
</tr>
<tr>
<td>Antithromboembolic prophylaxis</td>
<td>No/yes</td>
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<td>Early Rehab</td>
<td>No</td>
</tr>
<tr>
<td>Guideline</td>
<td>No</td>
</tr>
<tr>
<td>Pts and bleeding related</td>
<td>Yes</td>
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Factors influencing the long-term outcome of primary total knee replacement in haemophiliacs: a review of 116 procedures at a single institution

- 22 years: 1993 – 2007
- 116 primary TKR / 92 pts (INH and no INH)
- different types of implants (considering bone stock, axial deviation and instability)
  - cemented or cementless,
  - cruciate-retaining
  - posterior-stabilized
  - constrained
- Lost follow—up: none

TKR, in the past...
Risk of complications was related to:

- Presence of inhibitors
- Continuous infusion
- Cementless implant
- Different primary surgeons

Conclusions: TKR

TKR, in the past...
Our experience in PWH and INH

16 years: 1997-2016

53 major surgeries / 32 pts

- 21 TJR: 18 TKR
  1 THR
  2 TAR
- 4 Revision
- 11 Arthroscopic Procedures
- 17 Miscellaneous
Surgery in inhibitor pts

1997-2001
• TKR: 3/6 infections
• TKR: 1/6 aseptic loosening

2006-2016
• TJR: 3/21 infections
• TJR: 1/21 aseptic loosening
Complication rate

Same post-op bleeding complication

Different management of post-op bleeding complication

8% 25%
Complications

Arthroscopic procedures

Total joint replacement

Perioperative Bleeding

Orthopedic
Complications: arthroscopic procedures

- Prolonged post-op bleeding
- Haematoma
- Blood transfusion needed
- Delayed rehab
Complications: bleeding after arthroscopic procedures

**Hematological management**
- Tranexamic acid
- Increase rFVIIa dosage
- Decrease administration interval
- Shift to APCC
- Add APCC

**Orthopaedic treatment**
- Ice
- Elevation
- Bendage
- Splint
- Delay rehab
- Drain management
- Arthrocentesis
- Post-op embolization

**Prevention:** embolization
Complications in Total Joint Replacement

**Early**
- Knee
  - Post-op bleeding
  - Haematoma
  - Early infection: superficial, deep
- Ankle
  - Fracture
  - Wound healing

**Late**
- Knee/Ankle
  - Aseptic loosening
  - Septic loosening

__Sir John Charnley__

“Hematoma means death of surgeon”

Life style related ??
Complications: bleeding after TKR

Hematological management:
- Tranexamic acid
- Increase rFVIIa dosage
- Decrease administration interval
- Shift to APCC
- Add APCC

Orthopaedic treatment:
- Ice
- Elevation
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- Post-op embolization

Prevention: embolization
• **Major bleeding**
  after 17 procedures on 18 TKR
  after 4 procedures (36%) in 4 patients who underwent to arthroscopy

• **TKR**
  100% advanced arthropathy (Pettersson score 10-13)
  100% flexion deformity
Comments

median **drop in hemoglobin** levels after surgery was:

- 7.3 g/dl (IQR: 3.7-10.8) for TKR
- 4.8 g/dl (IQR: 1.2-6.2) for arthroscopy

**red blood cell transfusions** were required following:

- 17 on 18 TKRs (92%)
- 4 arthroscopic procedures (36%, 50% of knee procedures).
Comments

• This surgical series shows how the **results changed during years** according to changes in haematological treatment and peri-operative management.

• During the first years of experience 3 infections after knee replacement were registered (1 early – 2 late).

• Lower median age of inhibitor patients with non-inhibitor patients confirmed the **higher severity of arthropathy**.

• The **type of used implants** is not different in the two population.

• ** Longer period of hospital stay** registered shows the need of a careful post-operative management.
According to our experience

- **knee arthroscopy**: risk for bleeding uneffective
- **ankle arthroscopy**: effective in order to reduce joint bleedings pain

- In order to reduce bleeding complication after replacement and arthroscopic surgery of the knee, it could be advisable to use angiographic embolization.
Thank u for your attention!