Haemorrhagic Arthropathy

Andy Langdown
July 2008
Haemophilia

• Commonest congenital coagulation disorder
  – 1:10,000 worldwide

• Haemophilia A
  – Factor VIII deficiency

• Haemophilia B
  – Factor IX deficiency

• X-linked but 1/3 new mutations
Orthopaedics & haemophilia

• Recurrent spontaneous haemarthroses
  – Knee 44%
  – Elbow 25%
  – Ankle 15%

• Chronic synovitis
  – ↑ risk of bleeds

• Muscle bleeds

• Joint contractures

• Secondary arthropathy
# Factor levels & bleeds

<table>
<thead>
<tr>
<th>Blood factor level (IU/l)</th>
<th>Haemorrhagic manifestations</th>
</tr>
</thead>
<tbody>
<tr>
<td>50-100</td>
<td>Normal level – no bleeding problems</td>
</tr>
<tr>
<td>25-50</td>
<td>Tendency to bleed after major surgery</td>
</tr>
<tr>
<td>5-25</td>
<td>Severe bleeding after surgery or injury</td>
</tr>
<tr>
<td>2-5</td>
<td>Severe bleeding after minor injury; occ spontaneous haemarthrosis</td>
</tr>
<tr>
<td>&gt;2</td>
<td>Severe haemophilia; recurrent spontaneous bleeding into muscles &amp; joints</td>
</tr>
</tbody>
</table>
Pathogenesis of joint damage

• Bleeding from subsynovial venous plexus
  – ↓thromboplastic activity
• Recurrent bleeds cause synovial hypertrophy
  – Villous formation
  – ↑vasculaarity
  – Chronic inflammation
• Some blood remains in joint
  – Fibrous adhesions formed
• Haemosiderin accumulation in cells
  – Lysosomal enzyme release
  – Articular cartilage damage
  – Synovial proliferation

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Treatment of haemophilia

• A job for the haematologists!
• Factor VIII & IX concentrates
  – Problems with HIV transmission
• Recombinant factors
• Dose & frequency dependent on severity & site of bleeding
• Problems with inhibitory antibodies
• FFP in emergency

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Prophylaxis of haemophilia

• To ↓ incidence & severity of bleeds

• Cradle to college theory
  – Primary prophylaxis
  – From age of independent mobility till skeletal maturity
  – Factor levels >1% normal
  – 2-3 infusions/week
  – Cost $2.5-3 million per child in US

• Secondary prophylaxis
  – Treat after bleeds have occurred
Treatment of acute haemarthrosis

- X-rays of no use
- Diagnosis clinical
  - Obvious in knee & elbow
  - Hip: pain, spasm & ↓ROM
- Prompt IV Factor VIII or IX
- Severe bleeds may need repeat doses
- Aspiration
  - Controversial
  - Removes effect of tamponade
  - Speeds rehab
- Rest
- Physio

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Muscle bleeds

• 30% all musculoskeletal bleeds
• Minor injury
• Slow to resolve
• Features depend on size, location & fascia
  – Compartment syndrome
  – Iliopsoas
    • Hip contractures
    • Pelvic tilt & compensatory scoliosis
    • Femoral nerve compression
Treatment of muscle bleeds

- Factor replacement
  - 50-70% normal
- Rest 5-7 days
- Gentle rehab
- Surgery if nerve/compartment compromise
- Failure to resolve
  - ?antibodies to factor
  - ?cyst formation

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Contractures

- From joint damage or muscle bleeds
  - Equinus ankle
  - FFD knee
  - Elbow flexion
  - Hip flexion
- Splints, orthoses & physio to prevent
- Serial casting/dynamic splints
- Surgery
Surgery for contractures

**Soft-tissue**
- Hamstring & posterior capsule release
- TA lengthening
  - Beware ankle osteophytes

**Osseous**
- Extension osteotomy of distal femur
  - Problems with future growth
- Ilizarov techniques

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Chronic synovitis

- Fernandez-Palazzi et al 1996

<table>
<thead>
<tr>
<th>Grade</th>
<th>Classification</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Transitory synovitis, no bleeding sequelae, &gt;3 episodes in 6/12</td>
</tr>
<tr>
<td>2</td>
<td>Permanent synovitis, ↑ joint size, synovial thickening &amp; ↓ ROM</td>
</tr>
<tr>
<td>3</td>
<td>Chronic arthropathy with deformity &amp; muscle atrophy</td>
</tr>
<tr>
<td>4</td>
<td>Bony/fibrous ankylosis</td>
</tr>
</tbody>
</table>
Treatment of synovitis

• Medical
  – Factor replacement
  – Steroids

• Physio
  – Rest
  – Splints/orthoses

• Invasive
  – Aspiration
  – Radioisotope injection
  – Chemical injection
  – Surgical synovectomy
    • Open/arthroscopic
Pseudotumours

- Encapsulated haematoma
- ↓ incidence with factor Rx
- Painless hard mass ↑ size over months
- Can cause:
  - Pathological #
  - Local compression syndromes
  - Infection which can rupture
- Treatment
  - Excision
  - Amputation

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Perioperative issues

• In experienced centres
• Admit 1-2/7 pre-op
• Infusions for 7-10/7 post-op
• No such thing as minor surgery
• Infuse 1hr pre-op
  – Pre- & post-op level checks
  – bd infusions 5/7 post-op then od until wound healed
  – No IM injections!
  – Change dressings after infusions
• DVT not an issue with Type A but Type B need high purity concentrates

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Infection & haemophilia

- ↑ risk of pyarthrosis
  - Repeated aspirations
  - After surgery
  - Immunosuppression secondary to HIV

- Post-operative infection
  - 15% in 1 series
  - Linked with HIV

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Surgical options

• Elbow
  – Radial head excision
    • After skeletal maturity
    • For deformed radial head
  – Arthroscopy
    • For synovectomy
    • Difficult!
  – Open synovectomy
  – Arthroplasty
    • concerns re long term survivorship
  – Arthrodesis
Surgical options

• Knee
  – Arthroscopy
    • For synovectomy
  – Open synovectomy
  – Arthrodesis
    • Post-sepsis
  – Arthroplasty
    • Commonest joint replacement
    • Associated synovectomy
    • Expensive (14x)
    • ↑infection rate (esp HIV +ve)
Surgical options

• Shoulder
  – MUA
    • Early stages
  – Arthroscopy
    • With synovectomy
  – Arthroplasty
Surgical options

• Ankle
  – Arthroscopy
    • Synovectomy
    • Drilling of articular defects
    • May need skeletal distraction
  – Cheilectomy
    • Anterior osteophytes
  – TA lengthening
  – Arthrodesis
    • For end stage

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Summary

• Haemophilia commonest cause of haemorrhagic arthropathy
• Factor concentrates essential
  – Prophylaxis & treatment
• Multi-disciplinary problem
• Surgery may be indicated
  – Done in special centres
  – HIV is an issue
  – ↑ risk of complications

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