CHRONIC
HAEMOPHILIC
ARTHROPATHY

A/PROFESSOR JOHN YORK
INTERNATIONAL HAEMOPHILIA TRAINING CENTRE
ROYAL PRINCE ALFRED HOSPITAL
SYDNEY
<table>
<thead>
<tr>
<th>SEVERITY</th>
<th>FREQUENCY OF HAEMARTHROSES</th>
<th>JOINT INVOLVED</th>
</tr>
</thead>
<tbody>
<tr>
<td>FACTOR LEVEL (%NORMAL)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>SEVERE (50% of cases)</td>
<td>KNEE</td>
</tr>
<tr>
<td>1-5</td>
<td>MODERATELY SEVERE</td>
<td>ELBOW</td>
</tr>
<tr>
<td>5-20</td>
<td>MODERATE</td>
<td>ANKLE</td>
</tr>
<tr>
<td>20-60</td>
<td>MILD</td>
<td>SHOULDER</td>
</tr>
<tr>
<td></td>
<td></td>
<td>HIP</td>
</tr>
<tr>
<td></td>
<td></td>
<td>OTHER</td>
</tr>
</tbody>
</table>
HAEMOPHILIC ARTHROPATHY

ACUTE HAEMARTHROSIS

- PRODROMAL STIFFNESS AND PAIN.
  TENSE, SWOLLEN, HARD, HOT, TENDER.
  SHINY RED SKIN.

- KNEES
  ELBOWS
  ANKLES
  HIPS
  SPINE - RARELY
  HAND - MAINLY POST TRAUMA

- RAPID PAIN RELIEF
  WITH APPROPRIATE FACTOR REPLACEMENT
CLIN. PATHOLOGY

- ACUTE BLEEDS
- SUB-ACUTE ARTHROPATHY
- CHRONIC ARTHROPATHY
SUB ACUTE HAEMARTHROSIS

- Usually after 2 or more acute bleeds.
- Persists despite treatment.
- Thick boggy synovium.
- Moderate restriction of joint motion.
- Pain not prominent feature.
CHRONIC HAEMARTHROSIS

- SUB ACUTE CHANGES FOR 6/12
- PROGRESSIVE CHANGES UNTIL END STAGE FIBROTIC
  - CONTRACTED
  - TOTALLY DESTROYED
- BLEEDING OFTEN ABSENT OR INFREQUENT IN THE LATE STAGES.
MANAGEMENT

- PROPHYLAXIS
- ACUTE HAEMARTHROSES
- SUB-ACUTE ARTHROPATHY
- CHRONIC ARTHROPATHY
PROPHYLACTIC CLOTTING FACTOR REPLACEMENT

- THREE OR MORE TIMES WEEKLY-
  FACTOR LEVEL $\approx 10\%$
- PHYSIOTHERAPY
  STRONG MUSCLES
    CYCLING
    SWIMMING
    WALKING
- NON-BODY CONTACT SPORTS
ACUTE BLEEDS

1. ADEQUATE HAEMATOLOGICAL CONTROL
2. REST 48 HOURS
3. ASPIRATION
4. MOBILISATION - ACTIVE MOVEMENTS ONLY
   - ICE PACKS
   - ULTRASOUND
   - HYDROTHERAPY
DRUG TREATMENT

- ANALGESICS
- ANTI-INFLAMMATORY AGENTS
  - NSAID’S, LOCAL I/A AGENTS,
  - CORTICOSTEROIDS
- DMARD’S, SPECIFIC AGENTS
- IMMUNOSUPPRESSANTS AND CYTOTOXIC DRUGS
- GENERAL SUPPORTIVE AGENTS
  - ANTI-DEPRESSANTS
  - MUSCLE RELAXANTS
  - BONE PROTECTIVE MEDICATIONS
M_x SUBACUTE ARTHROPATHY

Medical synovectomy - synoviorthesis

(DXRT)

radio-colloids (^90Yt)

(thiotepa)

(osmic acid)

rifampicin

tetracyclines

ethamoline
SURGERY

- TERTIARY HAEMATOLOGY CENTRE
- ADEQUATE SUPPLIES AND HAEMATOLOGICAL CONTROL
- ORTHOPAEDIC SURGEON EXPERIENCED IN HAEMOPHILIA SURGERY
PSEUDOTUMOURS

MASSIVE HAEMORRHAGE
SKIN ULCERATION
GANGRENE
PAIN
SEPSIS
FRACTURE
AMPUTATION
(BOWEL OBSTRUCTION)
PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY
PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY
INHIBITORS

PREVENTION

rF FACTOR VIIa

MASSIVE REPLACEMENT

IMMUNOSUPPRESSIVE AND CYTOTOXIC DRUGS

- RETUXIMAB “MabThera”

PLASMAPHERESIS
PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY
PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY
PROBLEMS

• INHIBITORS
• SEPSIS
• HIV / HEP C / CJD / ? OTHERS
• THE ANKLE
• THE ELBOW
• OSTEOPOROSIS
• THE ELUSIVE GENE THERAPY
PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY
PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY
PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY
PAIN RELIEF AND DRUG ADDICTION

- NON-ADDICTIVE ANALGESICS
- TRANSCUTANEOUS N STIMULATION
- RELAXATION THERAPY, HYPNOSIS
- PAIN PROGRAMS
- LIAISON PSYCHIATRY    CBT
- CONSULTATION WITH LAW ENFORCEMENT AGENCIES
AN OLD BUT NEW PROBLEM
AGEING

- GENERAL MEDICAL PROBLEMS
  CARDIOVASCULAR, NEUROLOGICAL

- DEGENERATIVE JOINT DISEASE
  HIP, KNEE, SPINE

- TRAUMA, FRACTURES
AN OLD BUT NEW PROBLEM
AGEING

- GENERAL MEDICAL PROBLEMS
  CARDIOVASCULAR, NEUROLOGICAL

- DEGENERATIVE JOINT DISEASE
  HIP, KNEE, SPINE

- TRAUMA, OSTEOPOOROSIS, FRACTURES,
AN OLD BUT NEW PROBLEM
AGEING

- GENERAL MEDICAL PROBLEMS
  CARDIOVASCULAR, NEUROLOGICAL

- DEGENERATIVE JOINT DISEASE
  HIP, KNEE, SPINE

- TRAUMA, OSTEOPOROSIS, FRACTURES,
AN OLD BUT NEW PROBLEM
AGEING

- GENERAL MEDICAL PROBLEMS
  CARDIOVASCULAR, NEUROLOGICAL

- DEGENERATIVE JOINT DISEASE
  HIP, KNEE, SPINE

- TRAUMA, OSTEOPOROSIS, FRACTURES,
ACKNOWLEDGEMENTS

- JANE BLEASEL
- SCOTT DUNKLEY
- STEVE MATTHEWS
- LEONE MUDGE
- KEVIN RICKARD
- (FIONA RENNISON)
- AND...