HAEMOPHILIC ARTHROPATHY

Two types

Haemophilia A - deficiency in factor VIII

Haemophilia B (Christmas Disease) - deficiency in factor X

[Von Willebrands Disease - platelet disorder]

Epidemiology

Affects 1/10,000 people

Genetics

X -linked recessive- carried by females and manifest in males

Physiology

Factor VIII	Produced principally in the liver (also kidney, spleen) .
	Half life of 12 hours,
	In blood bound to von Willebrand's factor
	Normal level 0.50 - 1.50 iu/ml

Levels of factor VIII

>25%	Seldom have problem
Mild 5-25% available	Bleed at surgery or dental extraction
Moderate 1-5%	Minor injury
<1%	Spontaneous bleeding

Factor VIII should be:

20% = vigorous activity,

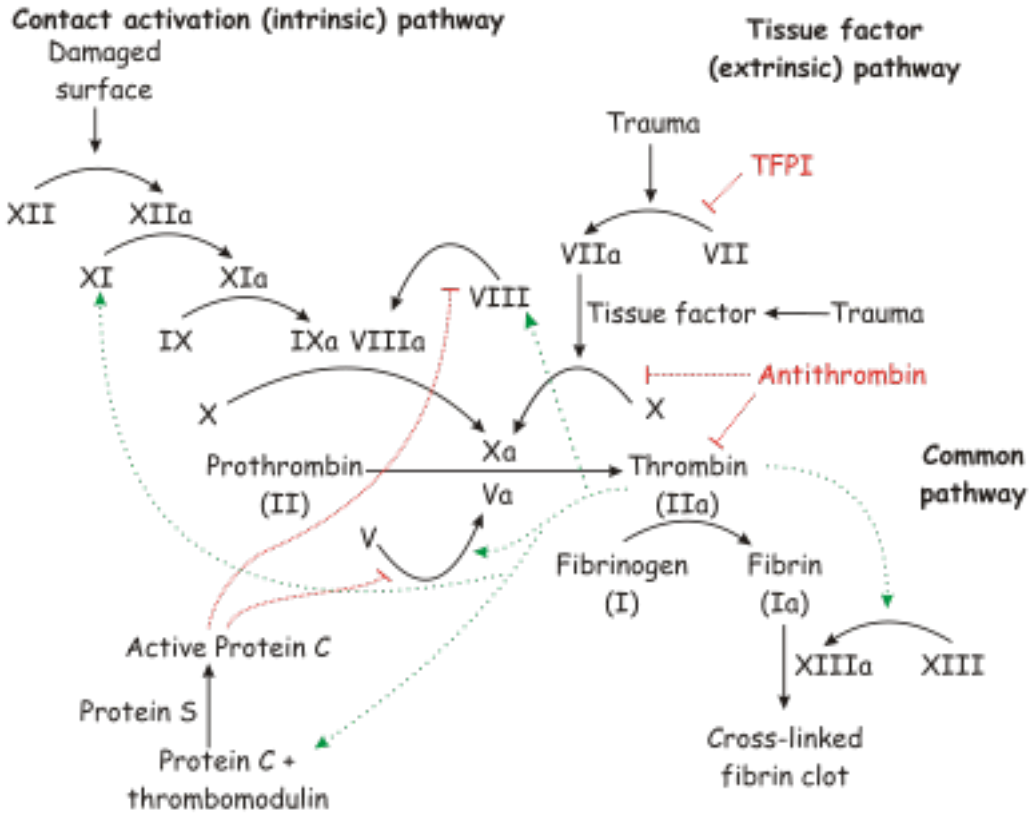
50% = Soft tissue surgery;

100% = Pre-op and 10^{th} post op [TJR]

Hemostasis: 2 pathways

Intrinsic: XII [Requires VIII] [APTT]

Extrinsic VII through tissue factor [INR]



Clinical Manifestations

1. Acute joint bleed

Most commonly affected joints: knees > elbows > ankles >hips

Pain; Warmth; Boggy swelling

Tenderness; Limited movement

Management

Purified clotting factor IV

Alternatively cryoprecipitate or FFP

Aspiration should be avoided unless infection is suspected

Rest in a splint and cold pack

Early physiotherapy with replacement factor cover

2.Acute bleed in the muscles

Danger of compartment syndrome and Volkmans ischaemia

Decompression ineffectual and unwise

Deep intramuscular bleeding can lead to pseudotumour (Blood cyst*)*.

Can occur in soft tissue or bone. Most common in lower extremity.

Diagnosis by USS.

Can lead to compression of adjacent nerves.

Compartment syndrome: Rapid transfusion; followed by Decompression

Aspiration: remains controversial for haemophiliacs. Pain relief rapid: However, removal of the tamponade affect and the introduction of infection have led many centres to discontinue this practice.

3. Pseudocyst

Pseudotumors typically are found in the vastus lateralis, soleus, or iliopsoas muscles

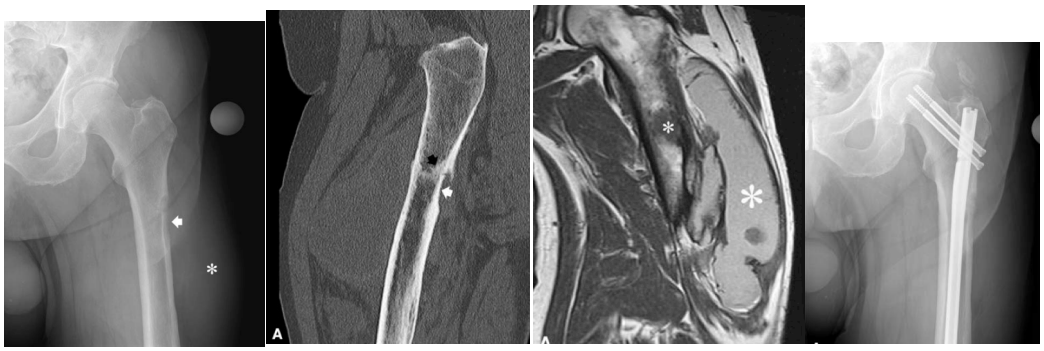
Pseduocyst: D/D ABC, OS

Encapsulated haematoma

Presents as a progressive painless, enlarging, hard mass over a period of 2–3 months.

Pathogenesis: Recurrent bleeds into soft tissue or bone.

Proximal bones (femur and pelvis) is the most common



A radiograph of the left hip showed a large nonmineralized mass in the lateral left thigh with remodeling of the adjacent lateral femoral cortex. MRI of the thigh

confirmed a large mass with relatively homogeneous high signal intensity on the T1-weighted spin-echo image and a rind of prominent decreased signal intensity at the periphery of the mass, and in multiple nodules, on a gradient MR image.
CT imaging performed to evaluate the osseous architecture more fully, identified the presence of an occult oblique fracture.

Natural course: Untreated pseudo-tumours will erode bone leading to pathological fractures

Surgical excision is the treatment of choice. It is associated with high morbidity and has a significant mortality rate (20%). Conservative management with replacement therapy and immobilisation may cause some regression, however, will not achieve a cure

Percutaneous drainage is not recommended owing to the potential for development of chronic draining fistulas and possibly fatal sepsis

4. **Articular deformities:**

- Realignment osteotomy

Progressive flexion of knee: Traction and orthosis – Hamstring release

or Supracondylar osteotomy

Large subchondral cysts: Curettage and bone graft.

Advanced arthropathy: Hip - THR

Knee - Debridement - TK**R if necessary**

Issues with joint replacement

1. Younger patients
2. Deformity and soft tissue balancing
3. Small joints
5. HIV and infection: total joint arthroplasty [Note: HIV: high infection following TJR: when CD4 counts of 200 mm3 or less]
6. Factor replacement at the time surgery
7. Antibodies : relative contraindication

8. Osteoporosis and careful dislocation

4. Hemophilic arthritis

Bilateral symmetrical joint arthritis in young male at 20 yrs: In Hemophilia

Multiple bleeds, deformity: valgus, varus, flexion, decreased ROM

Synovial thickening are common

Treatment

a. Synovectomies: Arthroscopic or

Open or

Medical: Yittrium, Phosporous

Open better than arthroscopic better than medical treatment. Aim: to prevent recurrent joint bleed and? Prevention of OA

Investigations

Blood

VIII levels and IX

APTT: low;[Intrinsic]

INR= Normal [extrinsic]

Antibody level: <10 units can be treated with high VIII dose

Important Considerations

Antibody inhibitors present in 5-20% haemophiliacs. Relative contraidication to surgery.

Large levels of factor : required to counteract these inhibitors.

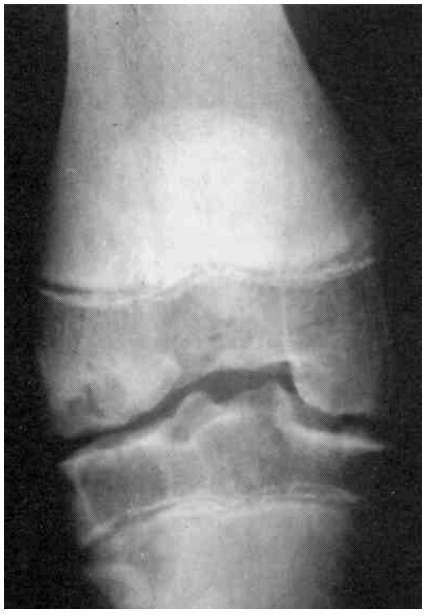
<10 Bethseda units - treatable with factor eight

>10 bethesda units - requires more sophisticated treatment

HIV A [85%]

Hep B

Hep C



X rays

Periarticular osteoporosis

Progressive joint erosion

Squaring of patella and condyles,

Squaring of intercondylar notch.

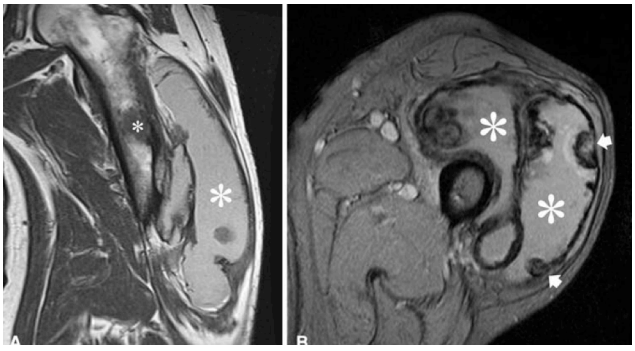
Epiphyseal overgrowth

Limb length discrepancy

Ultrasound

CT scan

MRI



Biopsy

Hyperplastic synovium

Copious iron deposition (Hemosiderin stained by Prussian blue)

D/D: RA, PVNS, Hemochromatosis, Ochronosis

No characteristic Lyphoplasmacytic infiltrate like Rheumatoid arthritis

No nodular mononuclear nodule or giant cells like PVNS

Hemosiderin pigments are seen in the chondrocytic lacunae

Facts

1. Plasma activity will increase 2% for every unit/kg of factor VIII
   1% for factor IX given

2. Patients who have factor levels > 1% rarely develop significant hemophilic arthropathy

3. **Primary prophylaxis** is to maintain factor levels between 1-5%

Most bleeding problem: Soft tissue surgery 50% level should be fine.
THR/TKR: 100%

4. Antibodies: 33% in Type A
   3% in Type B

5. HIV transmission not documented in those receive blood after 1986