End-Stage Hemophilic Arthropathy: The Journey to Joint Replacement

Husam Darwish, MD, FRCSC
Asst. Professor and Consultant
Department of Orthopaedic Surgery
King Abdulaziz University
Jeddah, Saudi Arabia
Outline

- Classification of hemophilic arthropathy
- Treatment options
- Pre and perioperative management
- Outcomes of joint replacement in hemophilia patients
Common Clinical Scenario

- A 24 y o man presents to clinic c/o left groin pain for 2 years

- known case of haemophilia A

- He has had multiple episodes of hemarthrosis since childhood treated with factor VIII replacement

- Currently he is unemployed due to the pain and uses Tramadol on a regular basis (150 mg TID)

- To date all conservative measures have failed to relieve his pain
On examinations:

- Limited left hip ROM with a $20^\circ$ flexion contracture
- The contralateral hip, both knees and both elbows are also affected
- Neurovascular examination is normal
Radiographic Classification

- Several Classifications
  - Kellgren And Lawrence
  - Pettersson, Hilgartner
Kellgren And Lawrence

0- Normal

1- Doubtful joint space narrowing and possible osteophytes

2- Definite osteophytes and possible narrowing of joint space

3- Moderate multiple osteophytes, definite narrowing of joint space, some sclerosis, and possible deformity

4- Larger osteophytes and definite narrow joint, sclerosis, and deformity
Modified Hilgartner

- Most commonly used clinically
- Structured to assist in surgical decisions
- Each grade represents significant alteration in disease progression influencing management

**Grade I:**
- Effusion
- Soft tissue thickening
- Juxta-articular osteopenia

Luck et al: JAAOS;12:235
Modified Hilgartner

- **Grade II:**
  - Widened epiphysis
  - Surface irregularities
  - Small erosions
  - *Normal joint space*

[Image: Radiograph showing bone changes associated with Modified Hilgartner grade II hemophilic arthropathy.]

Luck et al: JAAOS;12:235
Modified Hilgartner

- **Grade III:**
  - Narrowing of joint space
  - Bony cysts may be present
  - Extensive surface erosions

*Luck et al: JAAOS;12:235*
Modified Hilgartner

- **Grade IV:**
  - Complete loss of joint space
  - Marked surface irregularity
  - Significant angulation
  - Sclerosis & subluxation

Luck et al: JAAOS;12:235
Diagnosis

End-stage left hip osteoarthritis

Secondary to recurrent hemarthrosis
Treatment Options

Arthroplasty:
• Hemiarthroplasty (Bipolar)
• Hip Resurfacing
• Total Hip Arthroplasty (THA)
How should we manage this patient?
Multidisciplinary Approach

- Starts with the Hematologist:
  - Prevention of recurrent hemarthrosis
  - Prompt diagnosis and treatment
  - Development of a data base/registry

- Laboratory support:
  - Plasma derived or Recombinant clotting factors
  - Assessing for inhibitors
  - Man power on the day of surgery to assess plasma factor levels
  - Availability of PRBCs
Multidisciplinary Approach

• Nurses

• Physical therapy:
  • Pre-operatively to improve ROM and pain through modalities
    Atilla et al; Haemophilia 2012 “pre-op knee flexion contracture of 27.5° is an important threshold. Patients should be operated before that stage....”
  • Post-operatively
Multidisciplinary Approach

- Pain management:
  - Pre-operative consultation to assist in post-op Rx

- Anesthesia:
  - Cooperation and clear communication is key
  - Spinal/GA

  Englbrecht et al; Anaesthesist 2011. “Evidence-based recommendations for neuraxial anaesthesia in patients with hemophilia, vWD or ITP cannot be offered”

  “Supplementation of the missing factor to normal levels and monitoring during procedure is essential if neuraxial block is performed”
Multidisciplinary Approach

- Orthopedic Surgeon:
  - The “Maestro” coordinating between disciplines

- Technical consideration to address
  - Altered anatomy (pre-op CT)
  - Cysts and bony defects
  - LLD
  - Requiring individualized surgical approaches
  - Special instruments/implants
Peri-operative Considerations

- Timing of surgery
- Maintenance of coagulation factor level
- DVT prophylaxis
- Infection risk
- Outcomes of arthroplasty
Peri-operative Considerations

• Timing of Surgery:
  • Early in the week, early in the day

• Maintenance of coagulation factor level:
  • Factor VIII level 120% at surgery
  • Factor VIII level 60-80% for 72 hours
  • Then 50% from 72 hours to 2 weeks
    WFH
  • Factor VIII level at 100% from pre-op to 1 week
  • Factor VIII level 50-75% from 1-2 weeks
    Wong et al
Peri-operative Considerations

- DVT prophylaxis:
  - AAOS guidelines do not specifically address pt with haemophilia
  - ACCP recommends mech prophylaxis with chemoprophylaxis added when safe
Periprosthetic Infection

Perioperative clotting factor replacement and infection in total knee arthroplasty

- Systematic review of 19 retrospective case series
- 556 TKA in 455 patients
- Overall infection rate 7.9%
  - Maintenance of high levels of clotting factors for 2 weeks → 2.15% infection rate
  - Case series using the clotting factor replacement regime currently recommended in the World Federation of Hemophilia guidelines → 9.22% infection rate (P=0.00545)
- Conclusion:
  supports the maintenance of higher levels of clotting factor for a prolonged period of time

Wong et al; Haemophilia 2011
Outcomes

- **Norian et al; JBJS 2002**
  - Retrospective review of 53 TKA in 38 patients
  - 90% survival at 5 years with infection responsible for 7 of 11 failures
  - HIV status didn’t significantly affect the risk of infection

- **Zingg et al; Knee Surg Sports Traumatol Arthrosc 2012**
  - 43 consecutive TKA in 30 hemophilia patients
  - 34 knees were available at 9.6 yr f/u
  - 94% good or excellent results
  - 6% infection rate
  - 90% survival at 10 years
Outcomes

- Wang et al; Haemophilia 2012
  - Retrospective review of 65 consecutive joints in 45 patients
  - 40 TKA and 18 THA with 10.7 yr f/u
  - Knee Society clinical score, 83% good to excellent results
  - Harris Hip Score, 31% were good to excellent
  - Patient satisfaction with pain relief is higher than satisfaction with functional improvement
  - 88% of pt would have surgery again

- Cementing?
  - Kelley et al; JBJS 1995
    - 21% stems, 23% cups revised w/in 8 years
    - No loosening in press-fit
Summary

- Start with prevention
- Multidisciplinary **ownership** of the patient
- Clear communication between specialties
- “Haemophilia Center”
- Set patients’ expectations (pain not function)
- Adequate arthroplasty survivorship with high risk for infection
- extensive research regarding prophylactic protocols in hemophilic patients is needed before evidence-based recommendations can be made