



## Can I Have Surgery? An Overview of Orthopedic Surgical Options

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## Preventing Joint Disease

- The primary goal for every joint bleed is to resolve the bleed as quickly as possible in order to prevent further bleeding and slow the progression of joint damage



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## Treatment Options: Stopping Hemarthrosis

- Administering clotting factor as soon as possible is important to stop the bleeding quickly

Rest  
Ice  
Compression  
Elevation

Complications of Hemophilia. In: Butler RB, Crudder SO, Riske B, Toal S, eds. *Basic Concepts of Hemophilia: A Self-Study and Planning Workbook for Families With a New Diagnosis of Hemophilia*. Atlanta, GA: Centers for Disease Control and Prevention; 2001; module 4.

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## Joint Bleeding: Hemarthrosis

- Hemarthrosis is any bleeding in a joint
- Tiny blood vessels called capillaries can break spontaneously or after an injury
- Bleeding in the joints is the most frequent type of bleeding episode in hemophilia



Acute bleeding of the joint is called hemarthrosis

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## Hemarthrosis: What Happens During a Joint Bleed

- A joint bleed occurs when blood accumulates into the joint space
  - The lining of the joint (synovium) is the source of the bleeding



Acute bleeding of the joint is called hemarthrosis

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## Joint Bleeding: Hemarthrosis

- The most common sites for hemarthrosis are
  - Knees
  - Elbows
  - Ankles
- Bleeding also occurs in other joints, such as
  - Hips
  - Shoulders

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## Hemarthrosis: Effect of Repeated Bleeding in Joints

- Over time, the body breaks down the blood
- Byproducts (iron, enzymes) damage cartilage and erode joint surfaces



Blood breakdown products damage joint cartilage

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## What Is Synovitis?

- Synovitis is recurring inflammation of the synovial membrane, resulting from repeated bleeding into a joint in hemophilia patients



Recurrent hemarthrosis leads to chronic synovitis

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## Impact of Chronic Synovitis

- Thickening of the synovial membrane inside the joint
  - Limits mobility
  - Can be easily pinched as the joint moves, causing recurrent bleeding
  - Very painful
- Hemarthrosis can lead to synovitis, which can eventually lead to joint disease

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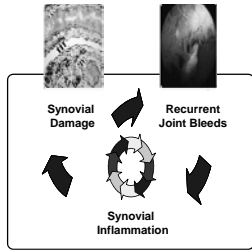
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## Hemarthrosis: Cycle of Joint Damage



- Just one bleeding episode can trigger a cycle of chronic synovitis, inflammatory arthritis, and progressive arthropathy

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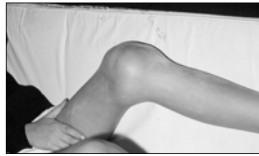
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## What Is Hemophilic Arthropathy?

- Hemophilic arthropathy is a state of limited mobility in a joint caused by the wearing away of cartilage from repeated prolonged bleeding episodes



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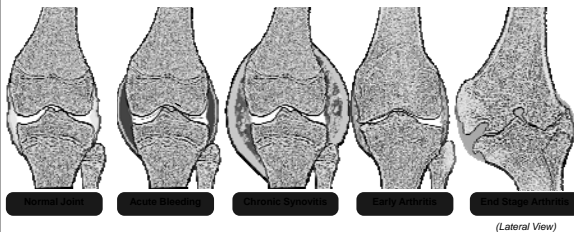
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## Hemophilic Arthropathy: Progressive Effect When Left Untreated



(Lateral View)

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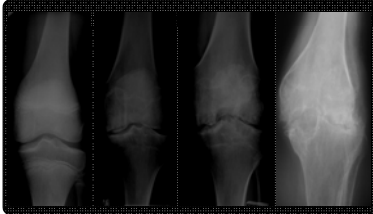
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## Hemophilic Arthropathy

Damage caused by the progression of hemophilic arthropathy can be detected and graded by radiographic images like these



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## Surgery: Potential Benefits

- Decreased pain
- Fewer bleeding episodes
  - Less time spent at the hospital
  - Less dependence on factor
- Potential for increased functional mobility
- Potential for increased range of motion
- Improved quality of life

Luck JV Jr, Silva M, Rodriguez-Merchan EC, et al. *J Am Acad Orthop Surg*. 2004;12:234-245.

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## Types of Surgery

- Synovectomy
  - Radioactive/isotopic, arthroscopic, open
- Elbow surgery
  - Radial head excision
- Joint replacement
- Joint fusion

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## Synovectomy

- For the treatment of chronic synovitis
- Purpose: to remove swollen synovium
- Helps decrease pain and frequency of bleeding episodes
- Three types
  - Radiosynovectomy/radionuclides
  - Arthroscopic synovectomy
  - Open synovectomy

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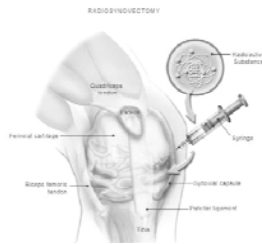
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## Radiosynovectomy/Radionuclides

- Outpatient medical procedure
- Small amount of radioactive substance injected into the joint
- Minimally invasive
- Less clotting-factor treatment needed
- Less need for rehabilitation
- Must consider potential risks



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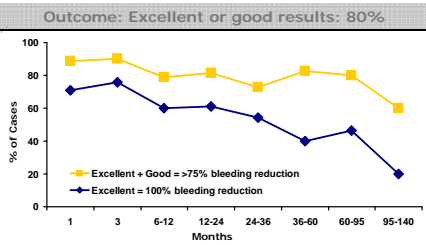
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## Radiosynovectomy/Radionuclides

- Effectively reduces frequency of bleeding



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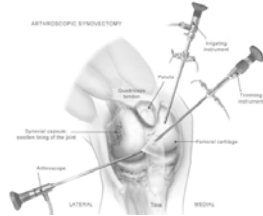
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## Arthroscopic Synovectomy

- Surgical procedure
- Arthroscope is inserted into joint through small incision
- Performed most often on knees, shoulders, and ankles, and can be done on elbows



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## Arthroscopic Synovectomy

- Arthroscopic synovectomy has largely replaced open synovectomy as the option of choice
- It is associated with reduced risk of loss of range of motion

Luck JV Jr, Silva M, Rodriguez-Merchan EC, et al. *J Am Acad Orthop Surg*. 2004;12:234-245.

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## Open Synovectomy

- Open synovectomy has an 80% success rate in preventing further bleeding
- Inpatient surgical procedure
- Knee is opened to remove the synovial membrane
- More tissue can be removed compared with arthroscopic option
- Regular, rigorous, physical therapy is required to regain range of motion

Rodriguez-Merchan E. *Haemophilia*. 2002;8:360-368.  
Luck JV Jr, Silva M, Rodriguez-Merchan EC, et al. *J Am Acad Orthop Surg*. 2004;12:234-245.

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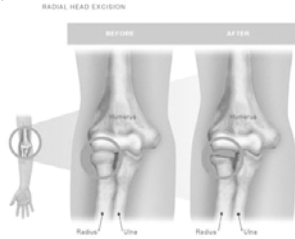
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## Radial Head Excision

- Radial head excision may improve symptoms associated with elbow hemophilic arthropathy, including
  - Pain
  - Recurrent bleeding
  - Limitation in range of motion
  - Not usually recommended in pediatrics



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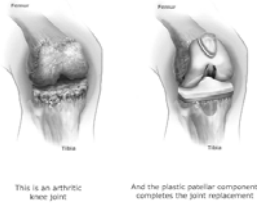
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## Joint Replacement

- A long-lasting solution for end-stage arthropathy
- Involves the removal and replacement of damaged areas of the joint



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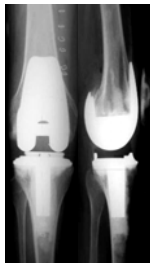
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## Joint Replacement

- Involves removing the damaged bone and joint tissue and replacing it with metal, ceramic, and plastic parts
  - Followed by an intensive rehabilitation program
  - Procedure takes about 2 hours per joint
  - Hospital stay will be extensive
- Can result in dramatically decreased pain and bleeding, increased mobility, and improved quality of life once the joint is fully healed



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## Joint Replacement

- Shoulder
- Elbow
- Hip
- Knee
- Ankle?



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## Joint Replacement

- Requires extensive PT:
  - Isometric, muscle tightening, exercises that do not involve moving the joint
  - Joint flexion and extension exercises to promote increased range of motion (ROM)
  - Active ROM and progressive resistive exercises to gain strength
  - Clotting factor therapy before each PT session
- Prognosis
  - Resume most normal daily activities within 8-12 weeks
  - Activity program should be gradual



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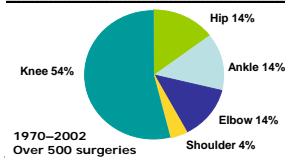
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## Treatment Options: Early Arthritis and Advanced Arthropathy

- The most effective means available to treat advanced joint damage is surgery

To date, many surgical procedures have been performed to correct damage caused by hemophilic arthropathy.

HTC at Orthopedic Hospital Los Angeles



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## Fusion

- Joint fusion is often recommended for the ankle joints, due to limited experience in ankle-joint replacement
- Joint fusion is often the “gold standard” for treatment of significant orthopedic changes in the ankle
- The goal is to relieve pain
- The fused joint is no longer flexible, but pain is decreased



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## Patients With Inhibitors: Challenges

- Patients with inhibitors may have more difficulty preventing and stopping bleeding and may be more prone to bleeding and developing target joints
- Inhibitors typically develop early in life and may result in early onset arthropathy due to challenges in treating joint bleeds

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## Considerations When Making a Decision About Surgery

- Other medical conditions
- Frequency of bleeds in target joints
- Degree of joint damage and pain
- Problems with mobility
- Time, determination, and endurance
- Risks of surgery vs benefits
- Risk of bleeding and response to factor therapy
- Patient’s goals and motivation to complete the recovery process
- Insurance coverage

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## Surgery Risks and Considerations

- People with hemophilia have specific concerns when it comes to surgery
  - Bleeding
  - Infection
  - Blood clots
  - Pain
  - Physical therapy



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## Surgery Risks and Concerns: Bleeding

- In patients with hemophilia, bleeding must be controlled during and after the procedure
- Excessive bleeding after surgery may lead to complications such as
  - Hematoma(s)
  - Infection
- To control bleeding, factor therapy must be used before, during, and after the procedure

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## Surgery Risks and Concerns: Bleeding

- Hemophilia patients and their family members need to know how bleeding will be controlled during surgery
  - Discuss clotting-factor treatment
  - Is the treatment approved for surgical use?
  - What is the plan before, during, and after surgery?
  - What is the plan for IV access before, during, and after surgery?

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### \*\*\* **Surgery Risks and Concerns: Infection**

- Infection is a universal risk after surgery
- Infections can
  - Prolong recovery
  - Require repeat surgery (when severe)
  - Result in removal of prosthesis or amputation
  - Occur after a long while, even after full recovery
- The risk of late-onset infection is higher for people with hemophilia than the general population

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### \*\*\* **Surgery Risks and Concerns: Blood Clots**

- Blood clots are a concern for all patients undergoing joint replacement, including those with hemophilia
  - Although rare in people with hemophilia, they can occur
  - A very serious concern, because venous clots can move and block an artery that provides blood to the lungs
- A prevention program may be needed, including
  - Periodic elevation of the legs
  - Lower leg exercises (to increase circulation)
  - Compression stockings

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### \*\*\* **Surgery Risks and Concerns: Pain**

- Pain control is essential
- Postoperative pain differs from arthritic pain
- Discuss pain management with your HTC and surgeon prior to the procedure
- Adjunct measures
  - Ice/cold compression
  - TENS

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## **Surgery Risks and Concerns: Physical Therapy**

- Physical therapy is essential for a successful outcome
- Intensive physical therapy requires a strong commitment
- Rehab schedule and duration of PT depend on
  - Type of surgery
  - Progress/complications
  - Risk of bleeding
  - Damage in multiple joints may require a slower rehabilitation or special equipment

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## **Patients With Inhibitors: Opportunities**

- Advances in medical treatments and therapies are providing more options for people with hemophilia and inhibitors
  - Surgery is becoming a reality for many patients with inhibitors
  - Surgical success in trials and studies is promising

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## **Speaking With Your HTC Team About Surgery**

- Discuss whether surgery is an option with your HTC team
  - Before committing to surgery, ask questions about expectations
  - Meet with your HTC team about all aspects of the surgery
  - Keep in close contact with the surgeon and other team members

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## Additional Resources

### Surgery and Hemophilia

Fact-based information, professional commentary, and personal experiences about surgical opportunities in hemophilia patients with inhibitors: [www.surgeryandhemophilia.com](http://www.surgeryandhemophilia.com)

### Inhibitor Patient Education Summits

Regional, patient-education meetings where patients, families, caregivers, and professionals can share information and experience: [www.inhibitorsummits.org](http://www.inhibitorsummits.org)

### National Hemophilia Foundation

A non-profit, support, advocacy, and educational organization for people whose lives are affected by hemophilia and other bleeding disorders: [www.hemophilia.org](http://www.hemophilia.org)

### Hemophilia Federation of America

A national, non-profit organization that assists and advocates for the blood-clotting disorders community: [www.hemophiliated.org](http://www.hemophiliated.org)

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## Acknowledgement

Much of the content for this presentation has been provided by

**Mauricio Silva, MD**  
Assistant Professor, Department of Orthopedics  
David Geffen School of Medicine, University of California  
Hemophilia Treatment Center, Orthopaedic Hospital  
Los Angeles, CA

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