

CHALLENGES CHOICES DECISIONS

A Guide on Orthopedic Surgery
for People with Hemophilia



Canadian Hemophilia Society
Help Stop the Bleeding



Canadian Hemophilia Society
Help Stop the Bleeding

The Canadian Hemophilia Society (CHS) is committed to improve the health and quality of life of all people with inherited bleeding disorders and ultimately to find a cure.

The CHS consults qualified medical professionals before distributing any medical information. However, the CHS does not practice medicine and in no circumstances recommends particular treatment for specific individuals. Brand names of treatment products are provided for information only. Their inclusion is not an endorsement of a particular product or company. In all cases, it is strongly recommended that individuals consult a hemophilia-treating physician before pursuing any course of treatment.

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Introduction

Hemophilia is a genetic bleeding disorder caused by low levels of clotting factor, a protein in blood that controls bleeding. Individuals with hemophilia bleed for a longer time than normal and experience frequent bleeding into joints, muscles and tissues.

Joint damage is one of the major complications of hemophilia. Damage is caused by prolonged bleeding into the joint cavity. It is normally the result of many bleeds into the same joint over time – the greater the number of bleeds and the more serious the bleeding, the greater the damage. However, just one serious bleed can cause major damage.

Almost all adults with severe hemophilia in Canada suffer from damage in the knees, ankles and/or elbows because they grew up in a time when treatment was less advanced. Fortunately, today many children with hemophilia in Canada are growing up with nearly normal joints due to preventive treatment.

Chronic joint damage causes pain and limits range of motion. When the pain is severe and interferes with the activities of daily living, orthopedic surgery is an option. Procedures may involve the removal of damaged joint tissue, bone grafting or excision, joint fusion or joint replacement. All surgical procedures for patients with hemophilia are done using replacement therapy to control bleeding.

There have been many successful outcomes for individuals with hemophilia who have had orthopedic surgery – specifically, reduced pain and discomfort and significantly improved quality of life. However, there can still be challenges and complications.

The goal of this resource guide is to help hemophilia patients and their families understand what is involved in orthopedic surgery. This guide does not have all the answers, as each patient is unique. It is therefore vital that patients and their families also consult with their physician and hemophilia treatment centre.

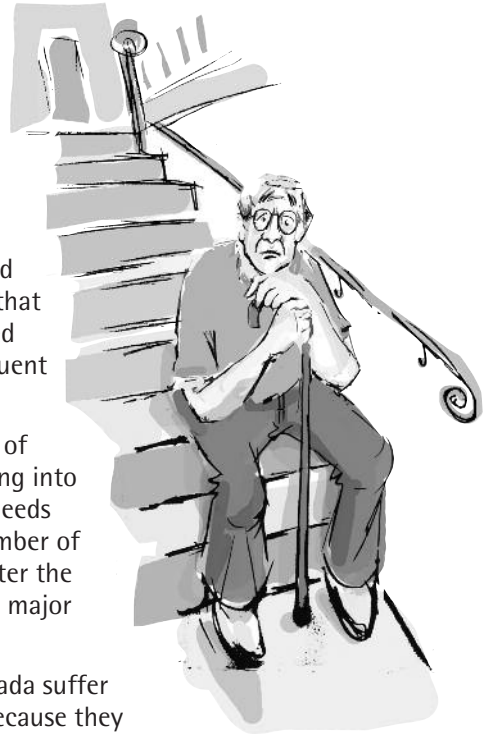


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Chapter 1

Joint Damage and Orthopedic Surgery

CHAPTER AT A GLANCE

- **Joint Damage in Hemophilia**
- **Preventing and Treating Joint Damage**
- **Orthopedic Surgery Explained**
- **The Orthopedic Surgery Team**
- **Chapter Summary**

Hemophilia is a genetic bleeding disorder caused by low levels of **clotting factor***, a protein in blood that controls bleeding (**hemorrhage**) and clotting (**coagulation**). Individuals with hemophilia bleed for a longer time than normal and experience frequent bleeding into joints, muscles and tissues. There are three levels of severity: **mild, moderate and severe**. The level of severity is defined by the amount of clotting factor missing from a person's blood.

*Words in bold are defined in the glossary. (see page 73)

Joint Damage in Hemophilia

Joint damage in individuals with hemophilia (**hemophilic arthropathy**) is caused by prolonged bleeding and accumulation of blood in the joint or joint cavity (**hemarthrosis**). It is normally the result of many bleeds into the same joint over time – the greater the number of bleeds and the more serious the bleeding, the greater the joint damage. However, just one serious **joint bleed** can cause lasting damage.

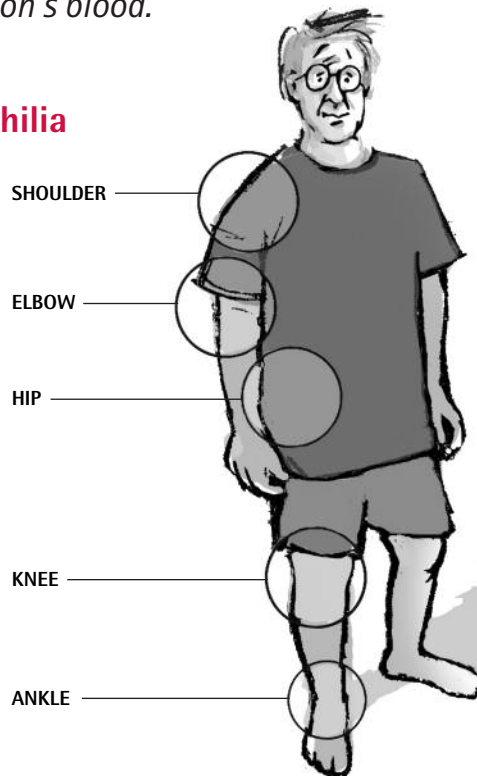


Figure 1:
Joints most commonly affected by bleeding

An insufficiently treated bleed can result in the development of a **target joint** – a joint that is continuously inflamed and vulnerable to recurrent, spontaneous bleeds. The joints most commonly affected by hemophilic bleeding, in decreasing order of frequency, are the ankles, elbows, knees, shoulders and hips. (See **Figure 1**, page 9.)

The damage occurs in two places in the joint: the lining (**synovium**) and the **cartilage**. Cartilage provides padding between the bones, allowing the joint to move smoothly and without pain. The synovium lubricates and nourishes the joint, reducing friction between the bones and preventing "wear and tear." There are a large number of blood vessels in the synovium – this is one of the reasons why joint bleeds and tissue or muscle bleeds (**hematomas**) are common in people with hemophilia.

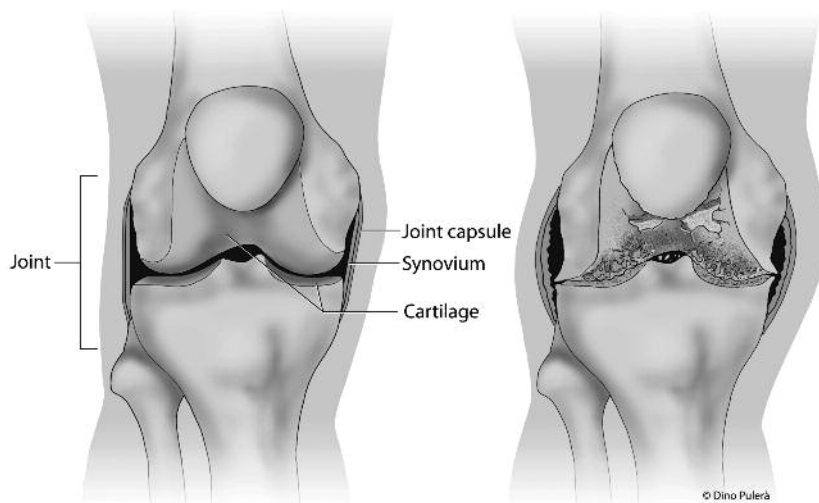


Figure 2:
Normal joint vs. damaged joint

A joint bleed causes the synovium to become inflamed, a condition referred to as **synovitis**. This makes the synovium vulnerable to being pinched between the bones, which causes further bleeding. Gradually, iron from destroyed red blood cells accumulates within the joint cavity and damages the bone's smooth cartilage surface. This results in pain that interferes with the joint's function. Bleeding into the joint cavity can also lead to the formation of **bone cysts**, fluid-filled lesions within the bone, or a **pseudotumour**, a slowly expanding cystic mass of blood in the joint tissues or along bone. In addition to damage to the joint itself, there is often shrinkage of the soft tissues, tendons and ligaments around the joint that diminishes the joint's range of motion.

(See **Figure 2**, page 10.)

Preventing and Treating Joint Damage

The key to preventing joint damage is to prevent bleeds whenever possible, and to treat bleeds quickly and completely when they do occur. Almost all adults with severe hemophilia have some degree of joint damage because they grew up in a time when treatment was not very advanced. Treatment has greatly improved over the past two decades and in Canada, the use of preventive treatment (**prophylaxis/prophylactic treatment**) has made it possible for most individuals with hemophilia to reach adulthood without developing joint damage.

Prophylaxis generally consists of factor replacement therapy several times a week to prevent bleeding. It is used to prevent bleeds and break a cycle of repeated bleeds to prevent joint damage. **Hemophilia A**, which is caused by low levels of factor VIII, is treated with factor VIII clotting concentrates. **Hemophilia B**, which is caused by low levels of factor IX, is treated with factor IX clotting concentrates. Early treatment of bleeds is essential – a joint bleed that is treated promptly and followed with **physiotherapy** to strengthen the muscles is less likely to cause damage.



Individuals with mild or moderate hemophilia have fewer joint bleeds than those with severe hemophilia – consequently, they have less joint damage. Nonetheless, it is important to prevent joint bleeds whenever possible, whether the hemophilia is mild, moderate or severe.

Before I had surgery on my left knee, I had very limited movement and was in almost constant pain. I was not able to walk more than a few hundred metres because my left leg was out of normal position and turned outward at the knee, resulting in about 4 cm of height loss. My quality of life was poor. A check-up with doctors at the hospital confirmed the need for joint replacement.

–Patient in his mid-sixties with hemophilia

Orthopedic Surgery Explained

Orthopedic surgery is a medical procedure performed to treat problems that develop in the bones, joints and ligaments. It is generally **elective surgery**, rather than required or emergency surgery.

Joint replacement with artificial parts (**prosthesis**) is common when there is irrevocable damage to a joint, usually the knee, hip or ankle. Replacement of the elbow or shoulder joint is performed less often due to the complexity of their structures and functions. Other surgical procedures are used to correct damage to these joints.

Orthopedic surgery may be suggested when:

- Bleeding into a joint becomes persistent and the pain interferes with everyday life, such as going to work, visiting friends, buying groceries, etc.
- Pain and loss of function is diminishing the individual's ability to perform **activities of daily living** necessary for normal self-care, including personal hygiene, dressing, eating, mobility and movement.
- Pain interferes with sleep at night.
- Different medications have been tried and do not alleviate the pain, or the medication no longer works.
- Other strategies such as rest, exercise, physiotherapy, **assistive devices** (sling, cane, crutches, walker, wheelchair, etc.), modified activity planning and/or non-invasive procedures are not effective at managing the pain.

Orthopedic surgery may not be required if:

- There is pain in a joint only after long periods of strenuous or weight-bearing activity.
- The pain is bothersome, but not severe enough to require pain medicine.
- There is pain from moving an affected joint or from weight-bearing activity that interferes with quality of life, but can be relieved by **analgesic** or **anti-inflammatory medication**.

It is important for the patient to monitor his condition carefully and discuss any changes with his physician.



The Orthopedic Surgery Team

Orthopedic surgery involves a number of different medical specialties. Depending on individual circumstances, the multidisciplinary team generally can include the following healthcare professionals:

- Hematologist/hemophilia physician
- Hemophilia nurse coordinator
- Physiotherapist
- Social worker
- Orthopedic surgeon
- Anesthesiologist
- Laboratory technician
- Psychologist
- Infectious disease specialist for patients with the **human immunodeficiency virus (HIV)**
- Hepatologist for patients with **hepatitis C (HCV)**



Hematologist/hemophilia physician

The hemophilia physician (hematologist) is responsible for the overall management of the patient's treatment and care. When a patient has severe joint damage that is interfering with activities of daily living and quality of life, assessment by the comprehensive care team may lead to a recommendation for orthopedic surgery. The hematologist ensures that there is good clinical backup for the surgery and helps make decisions about additional tests and other medical issues.

Hemophilia nurse coordinator

The hemophilia nurse coordinator ensures the availability of coagulation products for the procedure.

The nurse coordinator also educates the hospital staff on administering factor treatment, monitoring and responding to the patient's medical needs, and other aspects of hemophilia care. The nurse coordinator is the main liaison among the surgery team members and the patient's family. In most cases, the nurse coordinator also educates the patient, **caregiver** and nursing staff about preparing for surgery and **rehabilitation** at home.

My hemophilia nurse coordinator stayed with me in the operating room throughout the surgery to make sure that no one 'messed up' the administration of my clotting factor concentrate."

– Patient who underwent joint replacement surgery

Physiotherapist

The physiotherapist helps the patient prepare for surgery by prescribing specific exercises so that he is in as good physical shape as possible, with strong respiratory and cardiac function. The physiotherapist also works with the patient after surgery. Post-surgery exercises are essential for rehabilitation and achieving the best possible results from orthopedic surgery.



Social worker

The social worker coordinates a continuum of patient needs, providing guidance on work matters, medical coverage and other government programs (sick leave, disability, unemployment or welfare benefits, etc.), and resources for post-surgery care (rehabilitative or nursing services, domestic assistance, etc.).

Challenges, Choices, Decisions

A Guide on Orthopedic Surgery for People with Hemophilia

Orthopedic surgeon

The orthopedic surgeon uses non-invasive and/or surgical techniques to correct joint damage or other **musculoskeletal** problems. Many orthopedic surgeons specialize in specific areas (surgery for the shoulder, elbow, hip, knee, ankle, etc.), and/or procedures. The orthopedic surgeon must be experienced in surgery in hemophilia patients and work in a facility with a comprehensive hemophilia care program, usually a **hemophilia treatment centre (HTC)**.



Insight

As an orthopedic surgeon with a special interest in hemophilia, I have had the opportunity to meet some extraordinarily brave young men. The joint damage suffered as a result of repetitive bleeds into a joint (hemarthrosis) can be devastating. It may become so intolerable that orthopedic surgery has to be contemplated at an early age.

I am always deeply moved by the courage shown by my hemophilia patients and it is a real privilege to have been able to help them. It is my hope that the prophylactic use of 'factor' will dramatically reduce the need for surgical intervention and so far that appears to be coming true.

—Orthopedic surgeon

Anesthesiologist

An anesthesiologist provides pain treatment for surgery (**anesthesia**). Before the operation, the anesthesiologist will assess the patient's medical condition and discuss anesthesia and pain management with the patient and other members of the surgical team. During surgery, the anesthesiologist continually assesses the patient's medical status, monitoring vital life functions and managing pain. After surgery, post-anesthesia recovery is monitored with assistance from nurses.

Laboratory technician

The laboratory technician performs the complex laboratory tests required before, during and after surgery to measure patient bleeding time and response to the surgery and medications.

Psychologist

The psychologist helps the patient assess whether or not he is psychologically ready to undergo major surgery. The patient must be emotionally prepared and committed to having major surgery and adhering to post-surgery physiotherapy and exercises in order to achieve the best possible outcome. If the patient is not fully prepared to have surgery yet, he should not proceed.

Infectious disease specialist

For patients with HIV, the infectious disease specialist provides necessary expertise in the selection of antibiotics as well as preventive measures to take before surgery.

Hepatologist

For patients with HCV, the hepatologist provides necessary expertise in liver disease and related complications.



Insight

In 1998, my hematologist suggested we try total joint replacement of my two knees and one hip. The three surgeries were done about three months apart by the same orthopedic surgeon and were successful.

Part of the overall success came from the extensive post-operative physiotherapy required to regain as much joint range as possible. My physiotherapist helped keep me greedy for every degree of newfound joint range, and 10 years later I still do 30 to 40 minutes of daily physiotherapy to help maintain joint range.

The outcome of the elective joint replacement surgery was effectively a new lease on life. The change was from an electric scooter, a wheelchair and crutches, to being able to walk reasonable distances. Although there is no current shortage of pain from other destroyed joints, the three worst ones are history.

—Individual in his mid-sixties with hemophilia A

Chapter Summary

Patients and their caregivers should have an understanding of the basic aspects of orthopedic surgery in hemophilia patients.

- In Canada, the use of factor replacement therapy several times a week to prevent bleeding (prophylactic treatment) has made it possible for most individuals with hemophilia, who do not have **inhibitors**, to reach adulthood without developing significant joint.
- Orthopedic surgery is performed to deal with problems that develop in the bones, joints and ligaments. It is generally considered after other strategies such as medication, rest, exercise, physiotherapy, assistive devices, modified activity planning and/or non-invasive procedures have not been successful.
- Orthopedic surgery may be suggested when bleeding into a joint becomes persistent and the pain and loss of function interfere with the individual's quality of life and ability to perform activities necessary for self-care (personal hygiene, dressing, eating, etc.).
- Orthopedic surgery may not be required if there is pain in a joint only after long periods of strenuous or weight-bearing activity, or if the pain can be relieved by pain or anti-inflammatory medications.
- It is important for patients to carefully monitor the condition of damaged joints and discuss any changes with their physician.

Chapter 2

Challenges: Considerations in Orthopedic Surgery

CHAPTER AT A GLANCE

- **Benefits and Risks of Orthopedic Surgery**
- **Additional Risks Due To HIV and/or Hepatitis C**
- **Surgery Pain and its Management**
- **Medical Coverage and
Employment Insurance Benefits**
- **Hospital Stay and Recovery at Home**
- **Leave of Absence From Work**
- **Obstacles to Successful Surgery**
- **Checklist – Key Considerations Before
Choosing to Have Orthopedic Surgery**

Orthopedic surgery is performed to correct joint damage. Most people who undergo major orthopedic surgery are significantly better after six weeks and continue to improve for several months. However, it is important to keep in mind that surgery may not necessarily improve function in all patients – and sometimes function is diminished.



The patient should not expect to be able to do more than was possible before the onset of joint damage. Surgery will not bring the capabilities of a normal and healthy joint, but it can free the patient from pain and restore substantial function. The physician and orthopedic surgeon will be able to tell the patient what improvement and progress can be expected based on his particular circumstances.

Before advising orthopedic surgery, the hemophilia team must consider a number of health factors that may affect surgical risks and outcomes. These include:

- General health (hypertension, heart or lung disease, immune disorders)
- Weight
- Age
- Bone density
- Bone and joint deformity and stiffness
- Nutrition

The patient's motivation and commitment to adhere to active, and sometimes very demanding, physiotherapy before and after surgery are also critical factors to whether surgery is advisable.

Benefits and Risks of Orthopedic Surgery

The most important benefits of orthopedic surgery are reduced pain and discomfort and improved quality of life. Individuals who have lived with a very painful joint may find that after surgery they are able to perform daily activities without medication and get back to "normal" life. Each individual must discuss the benefits and risks to expect with his physician and the orthopedic surgeon.

Overall, the benefits of orthopedic surgery for hemophilic joint damage may include:

- Pain relief
- Improved movement and use of a damaged joint
- Improved alignment of a damaged joint
- Reduced frequency of joint bleeds

Potential risks and complications that can affect a patient's recovery and outcomes include:

- Allergic reaction to anesthesia
- Infection at the **intravenous (IV)** or puncture site
- Bleeding complications during surgery
- Post-operative bleeding requiring further surgical intervention
- Acute post-surgery infection
- Surgical wound, bone or deep tissue infection
- Development of an inhibitor
- Nerve damage and impairment
- Chest infection (**pneumonia**) due to inadequate movement following surgery
- Dislocation of prosthesis

In addition, patients who have been successfully treated with **immune tolerance therapy** to overcome inhibitors and are about to undergo surgery with factor VIII or factor IX replacement, need to be aware that the presence of infection may increase the risk of inhibitors reoccurring.

Additional Risks Due To HIV and/or Hepatitis C

Patients who are infected with the human immunodeficiency virus (HIV) face a greater risk of post-surgery infection due to their suppressed immune systems. Patient management can be complicated by the complex interactions of different medication regimens, requiring close monitoring of drug levels with dosage adjustment as needed.

Surgery also brings additional risks for patients infected with hepatitis C (HCV), because any or all of the functions of the liver can be impaired. Surgery is reported to be safe in patients with mild chronic hepatitis, but patients with severe chronic hepatitis are at increased risk. A diseased liver is particularly susceptible to the changes in blood flow and circulation that accompany surgery – anesthesia and medications must be carefully administered to ensure proper control of bleeding and transformation and clearance of drugs.



Surgery Pain and its Management

Proper pain treatment is essential throughout the surgery and rehabilitation period. It is important that the patient not underestimate the severity and duration of pain that can accompany surgery. The patient and medical team should discuss how he can minimize post-operative pain and encourage healing.

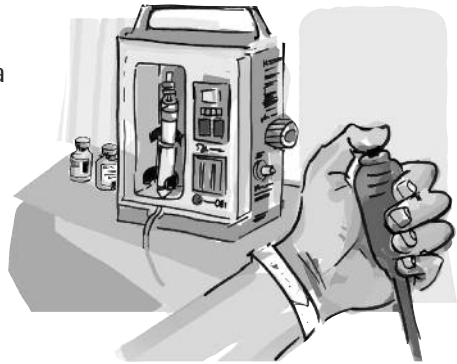
There is a broad variability in how much pain a patient will feel upon waking up from surgical anesthesia, depending on the procedure done. Sometimes there is quite substantial pain immediately after surgery, but it should subside within a few days. It is difficult to quantify how much pain is acceptable, since individuals have different levels of pain tolerance. The patient should not have to endure severe pain at any time after surgery. A good sign of proper pain management is if the patient is able to sleep restfully. However, even with pain medicine, he should expect some pain and discomfort in the weeks after surgery.

Managing pain effectively will differ from one patient to another. The hematologist and anesthesiologist will develop the management plan for all aspects of pain. This will include a surgery protocol that describes the required tests, **hemostasis** control, and detailed bleed records and **infusion** diaries necessary to ensure good pain management.

Often, pain medicine is administered pre-emptively, before the surgery begins, to prevent the nervous system from experiencing pain from the trauma of surgery. Pre-emptive pain treatment is usually given in addition to general anesthesia or other sedatives during surgery.

Post-operative pain can be treated with one pain medication or a combination of drugs. Pain medicine can be given orally (by mouth), intravenously (through a tube feeding into the vein) or by injection.

Following surgery, the patient may initially be attached intravenously to a patient-controlled analgesia machine. These devices allow the patient to administer pain relief as needed, and are calibrated to a maximum dosage to prevent overdose. Intravenous pain medicine is gradually withdrawn and replaced by oral tablets.



For more information on pain management, see the CHS publication, *Pain – The Fifth Vital Sign: A Resource on Managing Pain for People with Bleeding Disorders*.

Medical Coverage and Employment Insurance Benefits

Orthopedic surgery is expensive. The overall cost depends on the type of procedure being done, hospital services and resources used, and the costs of medications and other therapies. Other major factors include the particular province or territory's healthcare plan, and whether or not the patient has private medical insurance coverage.

There is considerable variation in medical coverage in the public healthcare plans of the individual provinces and territories, such as for outpatient treatment and rehabilitation. Depending on the circumstances and the extent of coverage provided by the provincial or territorial health plan, the patient may need to pay for a portion of the costs, either through private insurance or out-of-pocket. The patient must discuss each of these matters early on and in detail with his physician and HTC social worker, to gain a full sense of the medical coverage and costs to expect.

Patients who have private health insurance through their employers, professional affiliations or other private plans need to consult the insurance company well in advance about the procedures and treatments that are covered.



Specific questions to ask the physician and health insurance provider include:

- What is covered by medicare (e.g., hospitalization, diagnostic procedures, medications, physiotherapy, surgery, etc.)? Likewise, what is covered by private insurance and what are the deductible costs?
- Is in-hospital rehabilitation covered?
- Following hospital discharge, will the costs of rehabilitation, physiotherapy, medications and/or home health services be covered? For how long?
- Are costs for the purchase or rental of assistive devices or home modifications covered?

Some employment benefit plans provide paid sick leave. The patient should consult his human resources department as well as the health insurance provider to determine whether he is entitled to any short- or long-term sickness benefits. Those who do not have private health insurance can apply for employment insurance sickness benefits federally. Patients receiving social assistance should consult a social worker to determine their eligibility for the different benefits (welfare, sickness and disability) and other options.

We are fortunate to live in a country where we have some of the best and most dedicated healthcare professionals and advanced medical technology on the planet. I have been to several international congresses of the World Federation of Hemophilia and have seen and heard the alternative.

– Individual with hemophilia who has had several joint replacements

Hospital Stay and Recovery at Home

The hospital stay may be only a few days, but this depends greatly upon the type of procedure performed and the individual patient's post-surgery recovery. Hospital stay tends to be longer for patients with inhibitors because of hemostasis issues. The patient will remain in hospital until he is able to perform essential tasks independently, such as transfer himself from bed to chair and chair to toilet and move about safely (with assistive devices if necessary).

The patient may require round-the-clock assistance for the first 48 to 72 hours after leaving the hospital. Discharge from the hospital will depend on whether he will have a caregiver at home, or is going to a rehabilitation centre. Factor replacement at home may replace in-hospital treatment if the patient is mobilizing well. Some pain and discomfort should be expected following surgery and will be managed with pain medicine.

How long it takes to recover will vary depending on the individual patient and the joint that was treated. Generally, the best progress is made in the first two to three weeks following surgery, with continuing improvement possible for up to six months and more.



Leave of Absence From Work

If the patient's job is not too physically demanding, it may be possible for him to return to work six weeks after surgery. However, if the job requires prolonged walking, standing or lifting, it may be three months before this is possible.

Furthermore, there may be unanticipated complications due to the presence of inhibitors that could require a more extended rehabilitation period. The patient should discuss the recovery timeframe to be expected with his physician and orthopedic surgeon.



Some kinds of labour, may not be advisable following joint surgery. The patient should seek advice from an occupational therapist about whether or not he can safely return to his job following surgery. If the patient has a serious long-term disability that prevents him from working regularly at any job, it may be possible to obtain disability benefits through the Canada Pension Plan or the Quebec Pension Plan.

Obstacles to Successful Surgery

People with hemophilia have a higher risk of complications with orthopedic surgery than normal. The medical team carefully plans the surgical procedure in order to ensure the best outcomes. Some challenges are avoidable. These include:

- Insufficient education and preparation of the patient and caregiver
- Lack of patient motivation and commitment to follow the physiotherapy program

However, even with the best expertise and preparation, sometimes obstacles may arise over the course of surgery or afterwards. These challenges can be related to the higher surgery risks for patients with hemophilia, particularly those with inhibitors, or may be unrelated to hemophilia and common among patients who do not have the bleeding disorder.

These include:

- Susceptibility to viral or deep tissue infection
- Development of heart or lung complications during or after surgery
- Bleeding complications during surgery or post-operatively
- Failure of the procedure to achieve the desired results

It is essential for the patient to discuss the surgery options, benefits and risks with his physician and orthopedic surgeon. Together, the patient and the medical team will make the decision whether or not to proceed with orthopedic surgery.

Checklist

Key Considerations Before Choosing to Have Orthopedic Surgery

It is essential for the patient to have a good understanding of what to expect with orthopedic surgery in order to be able to make an informed decision. Issues and concerns should be discussed thoroughly with the medical team before proceeding.

1. What are the benefits of having orthopedic surgery?
2. What are the potential risks and complications of orthopedic surgery?
3. What are the outcomes or limitations that can be expected from orthopedic surgery?
4. Is there good clinical backup and ample access to factor replacement before, during and after surgery?
5. How much pain is involved in orthopedic surgery? How is pain monitored and treated?
6. What medical coverage and employment insurance benefits are available to help cover the costs of the surgery?

- 7. How long will the hospital stay and rehabilitation period be?
- 8. How much time is needed away from work? Is there a risk of long-term disability that would prevent return to work?
- 9. Will a caregiver be available to provide assistance at home following the surgery?
- 10. Is the patient physically and psychologically prepared for the demands of surgery and rehabilitation?

Surgery was the only alternative to eliminating the pain in my case. The operations (replacement / fusion) were godsend. I do not run or skate and I avoid stairs like the plague. My wife and I play golf (I still have a slice) and I am able to enjoy some travel and visit family and friends. Overall, I believe my life is good!

– 53 year-old man with severe hemophilia A who has had five joint replacements / fusions.

Chapter 3

Choices: Surgical Options for Joint Damage

CHAPTER AT A GLANCE

- **Types of Orthopedic Surgery**
- **Ankle Surgery**
- **Elbow Surgery**
- **Hip Surgery**
- **Knee Surgery**

A wide variety of surgical procedures are used for the treatment of hemophilic joint damage. The appropriate procedure will depend on a number of factors, including the degree of joint damage, condition of the bone or soft tissue to be treated and the patient's age, activity level and bleeding profile. Normally, major surgery on a joint may take two to three hours in the operating room. For patients with hemophilia, the procedure will take up to 50 per cent longer due to the careful vigilance and treatment needed to control bleeding.

Types of Orthopedic Surgery

The most commonly used surgical procedures to correct joint damage are:

- Arthroscopy
- Joint debridement
- Cheilectomy
- Synovectomy
- Arthrodesis
- Arthroplasty
- Osteotomy
- Resection
- Pseudotumour management
- Revision joint therapy



Arthroscopy: A relatively non-invasive technique in which a tiny camera and/or various surgical instruments are inserted through small incisions into the joint area to evaluate and repair damage.

Joint debridement: Minimally invasive surgery by arthroscopy to remove damaged cartilage, inflamed tissue and/or loose bone fragments from the joint. Arthroscopic debridement is used to treat bone cysts, by removing the bone's cystic contents and cyst lining, and the overlying unsupported cartilage.

Cheilectomy: Removal of small and abnormal bony growths around the bones of a joint that interfere with the joint's movement.

Synovectomy: Full or partial removal of damaged synovium or tissues lining the joint. Synovectomy is mainly performed in the knee, shoulder, elbow, wrist and hand. It can be done by arthroscopy or surgically. **Radiosynovectomy** is a non-surgical procedure using **intra-articular** injections of a radioactive compound to destroy the abnormal synovium. It is important to note that the synovium often grows back several years after surgery and synovitis can recur.

Arthrodesis: Fusion of two bones together with screws, steel rods or staples. The resulting fused joint loses flexibility but is stabilized and can bear weight better. Arthrodesis is performed in the ankle, spine, wrist, fingers and toes, where losing flexibility of one or more joints would not necessarily compromise normal motion. It is generally not recommended for larger joints such as the hip or knee, where joint replacement would be superior.

Arthroplasty: Partial or total joint replacement with an artificial prosthesis, usually made of plastic, metal and/or ceramic. Arthroplasty is the most common type of joint surgery, especially for the knee, hip and ankle. Replacement of other joints such as the shoulder, elbow and knuckles has not been perfected to the same extent but can sometimes still be effective. Patients should talk to the orthopedic surgeon about the advantages and disadvantages of the different materials used for prosthetic parts. It is important to note that prosthetic parts do not last forever – they usually wear out after 10 to 15 years. While replacement parts can be implanted, each subsequent procedure entails greater risk of complications.

Osteotomy: Correction of bone deformity and joint mal-alignment by cutting and repositioning the bone to improve the forces through the joint and shift stress from the damaged parts of the joint to healthy areas. Osteotomy is particularly useful for the weight-bearing knee and hip joints.

Resection: Removal of part or all of the damaged bone at the end of a joint that is causing discomfort. It is often performed when damaged joints in the foot cause painful swelling that makes walking very difficult. Resection and arthroplasty of the hip is referred to as a **Girdlestone Procedure**. Resection of parts of the elbow, wrist or thumb can help improve function and relieve pain.

Pseudotumour management: Surgical excision, aspiration, radiation or embolization of a pseudotumour, a potentially limb- and life-threatening condition unique to hemophilia. A pseudotumour occurs as a result of inadequate treatment of a soft tissue bleed, usually in muscle adjacent to bone. It is most commonly seen with a long bone or the pelvis. Untreated, a pseudotumour can reach an enormous size, causing pressure on the nerves and bone fracture. Management depends on the site of the pseudotumour, its size, rate of growth and effect on the adjoining bone and soft tissues. Some very small pseudotumours may be monitored when factor replacement therapy is used, but most require surgery.

Revision joint surgery: Replacement of artificial joints and damaged bone with new parts. This surgery is necessary when a previous joint replacement wears out. It is more difficult and takes longer than total joint replacement surgery.

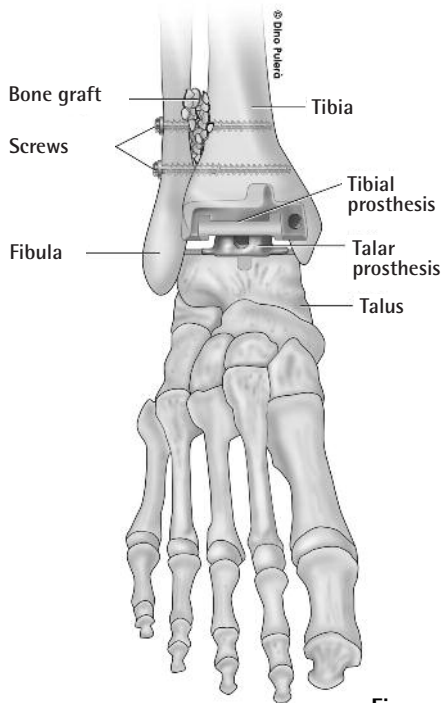


Figure 3: Ankle replacement

Ankle Surgery

Damage to the ankle joint is treated by synovectomy, arthroplasty, cheilectomy or arthrodesis.

Synovectomy: Removal of damaged synovium in the ankle by open surgery or arthroscopy.

Arthroplasty: Partial or total replacement of the ankle joint using prosthetic parts. (See **Figure 3.**)

Cheilectomy: Removal of small bony growths around the bones of the ankle joint.

Arthrodesis: Fusion of the ankle joint into immobility (for better weight-bearing strength, greater stability and reduced pain).

Challenges, Choices, Decisions

A Guide on Orthopedic Surgery for People with Hemophilia

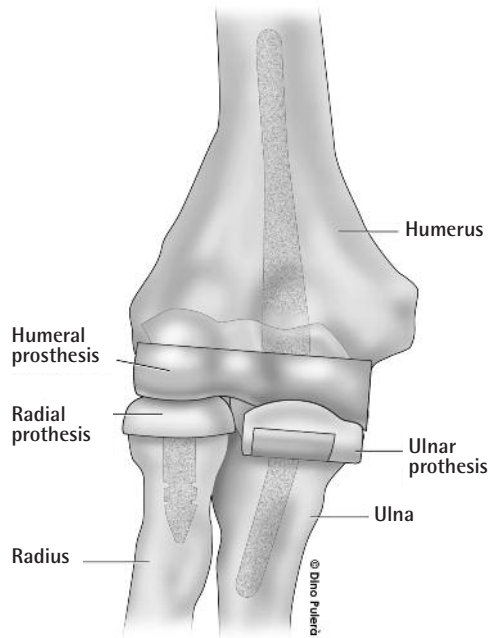


Figure 4: Elbow replacement

Elbow Surgery

Damage to the elbow joint is treated by synovectomy, removal of the radial head, joint debridement or arthroplasty. Joint replacement is rare due to the complexity of the elbow joint.

Synovectomy: Removal of damaged synovium in the elbow by open surgery or arthroscopy.

Removal of radial head: Removal of the damaged end of the shorter and thicker of the two bones in the forearm (**radius**) to improve elbow rotation.

Joint debridement: Removal of damaged cartilage, inflamed tissue and/or loose bone fragments from the elbow by arthroscopy.

Arthroplasty: Partial or total replacement of the elbow joint using prosthetic parts. (See **Figure 4**.)

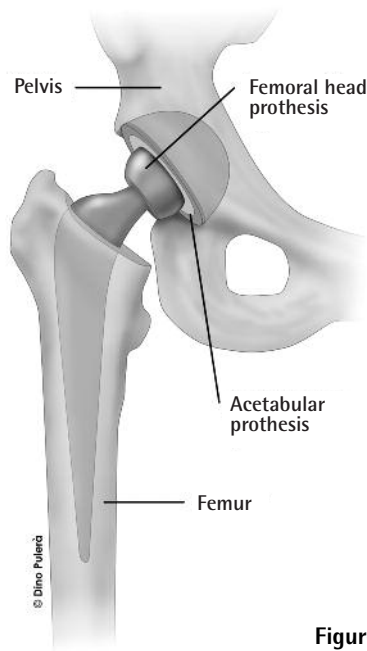


Figure 5: Hip replacement

Hip Surgery

The most common surgeries for hip damage are total joint replacement and osteotomy. On rare occasions, the Girdlestone Procedure is performed.

Total hip replacement: Complete replacement of the hip joint with a prosthesis consisting of a plastic socket and a metal "ball" at the head of the thigh bone (**femur**).

(See **Figure 5**.)

Osteotomy: Correction of deformity and joint mal-alignment by cutting and repositioning the femur.

Girdlestone Procedure: Removal of part of the head of the femur and allowing it to fuse with the socket of the hip (a rare procedure done only following a failed total hip replacement due to repeated dislocation or deep infection).

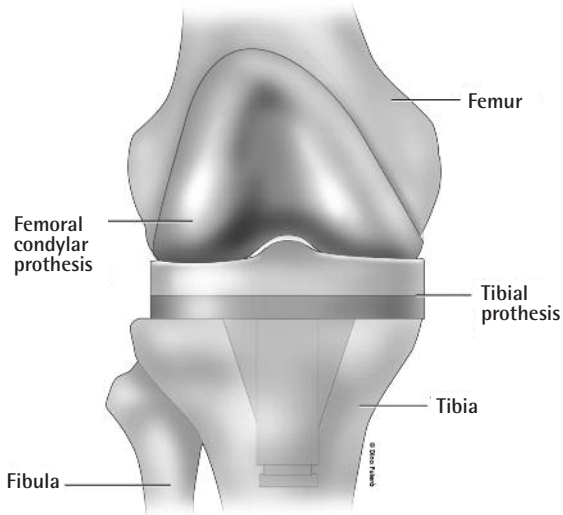


Figure 6: Knee replacement

Knee Surgery

Damage to the knee joint is treated by synovectomy, osteotomy, partial or total knee replacement or, in the worst-case scenario, arthrodesis.

Synovectomy: Removal of damaged synovium in the knee by arthroscopy or open surgery (often done before considering total knee replacement).

Osteotomy: Removal of a wedge of bone from the femur or lower leg bone (**tibia**) to realign the limb.

Unicondylar knee replacement: Partial knee replacement done when only one of the knee's three joint compartments is severely damaged.

Total knee replacement: Replacement of the damaged parts of the femur and the (tibia) with metal components. (See **Figure 6.**)

Challenges, Choices, Decisions

A Guide on Orthopedic Surgery for People with Hemophilia

Insight

Going into the surgery, my most notable concerns were about bleeding problems and infection risks. The thought of pain during recovery also crossed my mind.

I had a total knee replacement and my recovery was amazing. There was little if any bleeding, very little pain and no infection. I was in the hospital for about five days. I used a walker for about one week, crutches for about two weeks and then a cane for about one week. Physiotherapy improved movement from 0 to 110 degrees over five weeks. I was told that would be the maximum movement to expect after nearly 40 years of very limited range.

–67-year-old patient who underwent total knee replacement

Chapter 4

Decisions: Having Orthopedic Surgery

CHAPTER AT A GLANCE

- **Getting Informed About the Surgery**
- **Pre-Surgery Physiotherapy**
- **Other Health Preparations**
- **Modifications and Preparations at Home**
- **Post-Surgery Care Arrangements**
- **Compassionate Care Benefits and Support**
- **Rehabilitation Facilities**
- **Medical Preparations**
- **Pertinent Medical Information**
- **Checklist
Preparations in the Months Before Surgery**
- **Checklist
Preparations in the 24 Hours Before Surgery**

Deciding to undergo major surgery is a decision to be made together by the patient, physician and orthopedic surgeon. Overall, orthopedic surgery should be regarded as a comprehensive, demanding and relatively lengthy process. It can take about six months from the decision to have orthopedic surgery to the actual surgery itself. During this time, the patient should prepare physically and mentally, and take care of financial and domestic arrangements for the surgery.

Getting Informed About the Surgery

The medical team will give the patient and caregiver a full explanation of what to expect from the surgery, including what is expected of the patient and caregiver before, during and after the operation. Many hospitals and clinics also provide group education sessions. It is important for caregivers to be included in education sessions whenever possible. These sessions inform patients and caregivers about the different aspects of orthopedic surgery, recovery and rehabilitation, and their responsibilities towards achieving the best possible outcomes.



Most hospitals and hemophilia treatment centres provide brochures and fact sheets on joint damage and orthopedic surgery, or can recommend literature and educational videos available in public libraries or online.

Pre-Surgery Physiotherapy

Pre-operative physiotherapy and exercises are essential to prepare the patient physically for surgery and achieve successful surgery results. Most members of the orthopedic surgery team – the hematologist, surgeon, nurse coordinator, physiotherapist and social worker – will be involved in some aspect of the patient's pre- and post-surgery physiotherapy and rehabilitation.

Before the surgery, the physiotherapist will teach the patient specific exercises to strengthen his body for surgery. For example, a patient having hip or knee replacement surgery will benefit from exercises to strengthen the upper body in order to help him cope with crutches or a walker after surgery. Other exercises help strengthen the muscles surrounding a damaged joint.

The physiotherapist will also show the patient the exercises to be done post-surgery, so that he can practice them beforehand. The physiotherapist or a nurse will also teach the patient how to get out of bed, use the bathroom, shower, dress and move about post-surgery.



Challenges, Choices, Decisions

A Guide on Orthopedic Surgery for People with Hemophilia

Physiotherapy intervention is critical both before and after surgery to maximize the impact of the operation. By starting before the surgery takes place, there is less apprehension about what is going to be required in the post-operative phase in order to build as much strength, movement and function as possible. The surgery must be viewed as a process that begins months before the date of the operation itself if true success is to be achieved.

—Physiotherapist

Other Health Preparations

Physical and nutritional preparation can significantly improve surgical outcomes and recovery time. Patients about to undergo surgery should:

- **Have tooth or gum problems fixed.** Dental problems should be treated before surgery to help reduce the risk of infection in the new joint.
- **Eat a well-balanced diet.** A patient who is overweight may be advised by his physician to lose weight before surgery. However, the patient should not diet during the month before surgery.
- **Reduce or quit smoking.** Smoking changes blood flow patterns, delays healing and slows recovery.
- **Don't drink alcohol.** The patient should not have any alcohol for at least 48 hours before surgery.
- **Report any infections.** The medical team must be notified if the patient comes down with a fever, cold, infection or any other illness in the week before the surgery. Surgery cannot be performed until all infections have cleared up.

Modifications and Preparations at Home

It is important to prepare the home for the patient's rehabilitation period. A number of modifications or preparations at home could be done, depending on the joint involved.

- Remove any rugs or loose carpets and tape down electrical cords to avoid slips and falls.
- Rearrange furniture as necessary to widen passages and improve accessibility. If the patient will need to use an assistive device such as a walker or wheelchair after surgery, borrow the equipment beforehand to see how well he can move through the house.
- Install a raised toilet and safety handrails in the bathroom, and a chair and gripping bars in the shower.
- Set up a "recovery centre" where the patient will spend most of his time, with a comfortable bed or armchair and footstool. Items such as medications, a water pitcher and glass, facial tissues, wastebasket, telephone, radio, television remote control and reading materials should be easy to reach.
- Assemble items that will make personal tasks easier during the recovery period, such as a long-handled shoehorn, long-handled sponge for bathing, grabbing tool or reacher to avoid bending too far, and a hip pack for carrying things around.
- Stock up on snacks, prepared meals and disposable dishes and cutlery to minimize food preparation and cleanup.

- Apply for a temporary disabled parking permit (also called accessible parking permit). Most people are able to resume driving about six weeks after surgery but the patient must first regain adequate muscle control for braking and accelerating.

As the caregiver, you need to be ready for unexpected surprises and creative solutions once your husband returns home. We improvised by using landscaping bricks under the sofa and bed legs to adjust the height and make it possible for him to sit or lie down.

– Wife and caregiver of a patient who had several joint replacements

Post-Surgery Care Arrangements

Care and assistance at home is important to help the patient recover more quickly. The patient will need to arrange for a caregiver to take him home and stay with him for several days after the surgery, or possibly longer. The availability of a caregiver will influence when the patient is discharged from hospital. The caregiver (usually a family member or friend) helps make sure that post-surgery instructions are followed, monitors the patient's symptoms, and reports adverse developments. The caregiver also helps with tasks such as cooking, childcare, laundry and other domestic chores to prevent the patient from over-straining.



Compassionate Care Benefits and Support

Canada's employment insurance program provides compassionate care benefits to individuals who have to be away from work temporarily to provide care or support to a family member who is gravely ill. Caregivers who work while providing care and support can receive compassionate care benefits. A social worker can provide information about employment insurance and compassionate care benefits. Private employment benefit programs may also offer compassionate care leave or benefits. The patient and caregiver should consult their insurance providers about coverage and conditions.

The demands and responsibilities of providing patient care often take a toll on the physical and emotional well-being of caregivers. It is important to provide the caregiver with support and resources. A psychologist can provide counseling and advice on how to cope as a caregiver, while a social worker can coordinate access to caregivers' support group, temporary substitute care (**respite care services**), or financial assistance.

There were no particular challenges before or after surgery. He was co-operative with physiotherapy while being aware and respectful of his limitations. Gaining confidence as a non-professional was important to me. While I have been used to the infusion process, the extra steps involved with the peripherally inserted central catheter (PICC), although simple, were new. It is beneficial to be patient, encouraging but firm in assisting the patient with physiotherapy routines and exercises post-surgery and beyond.

– Caregiver of a patient who underwent joint replacement surgery

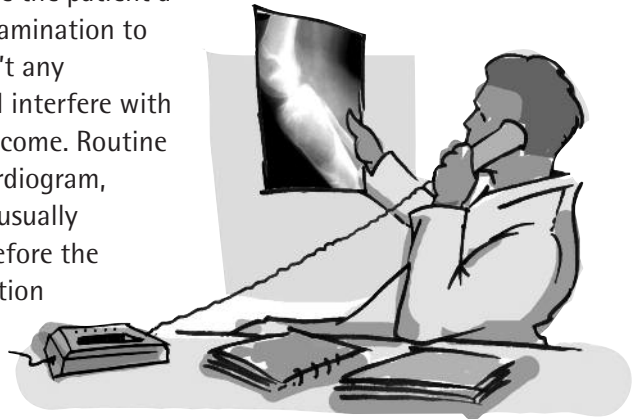
Rehabilitation Facilities

A patient who lives alone and does not have caregiver support, or who has special needs, may need a transition at a specialized rehabilitation centre after discharge from the hospital and before going home. The physician and social worker will refer the patient to an appropriate rehabilitation centre. The patient may want to visit the rehabilitation centre before the surgery to meet some of the staff and tour the facilities. The social worker can also identify resources and financial assistance to support the patient with rehabilitation at home.

Medical Preparations

The patient should consult the physician and orthopedic surgeon about the medications he is taking. The physician may advise the patient to stop taking certain medications before the surgery. It is also important to ask the physician about the appropriate pain medications to keep on hand for the recovery period.

Several weeks before the surgery, the physician will give the patient a complete physical examination to make sure there aren't any conditions that could interfere with the surgery or its outcome. Routine tests (blood work, cardiogram, chest x-ray, etc.) are usually carried out a week before the surgery. The examination results and clearance for surgery are then forwarded to the orthopedic surgeon.



Shortly before the scheduled surgery, the patient will have an orthopedic examination. The physician and orthopedic surgeon will review the procedure and address any outstanding questions or concerns. The patient will also meet with the anesthesiologist to discuss the type of anesthesia that will be used. The nurse coordinator will provide support and information throughout the process and help the patient plan for discharge from the hospital.

Pertinent Medical Information

The hospital will require the patient's medical information before the surgery including:

- The name of the family member or friend designated as the patient's primary contact with the medical team.
- A list of all current physicians (with names, addresses and phone numbers) and the reasons for seeing them.
- A list of medical conditions and all previous operations.
- A list of any allergies or adverse reactions to drugs or anesthesia taken in the past.
- Any dietary restrictions or other health problems such as diabetes, asthma, HIV or hepatitis infection.
- A list of insurance coverage and plans, including the name of the insurance company, the plan or group number and contact information.
- Information about any legal arrangements, such as a living will or power of attorney, in the event that the patient is not mentally or physically able to make decisions regarding his treatment.

Your mindset going into orthopedic surgery and rehabilitation must be an unwavering belief that it's going to go your way.

—Individual who has had several joint replacements

Checklist

Preparations in the Months Before Surgery

Orthopedic surgery should be regarded as a comprehensive, demanding and relatively lengthy process. In the months between the decision to have surgery and the scheduled operation, the patient should prepare physically and mentally for surgery and the rehabilitation period.

1. Be sure to be well informed about the different aspects of the surgery and rehabilitation, and what is needed achieve the best results.
2. Follow the exercise program prescribed by the physiotherapist to strengthen muscles and respiratory and cardiovascular function in preparation for surgery. Practice some of the exercises to be done during rehabilitation.
3. Get in good physical health before surgery. Fix dental problems to reduce the risk of infection. Eat a well-balanced diet. Avoid cigarettes and alcohol.
4. Report any infections or other illnesses that arise in the week before surgery.
5. Some rooms at home may need safety modifications and rearrangement of furniture to improve access and mobility.

- 6. Arrange for a caregiver to provide assistance at home following surgery. If there is no caregiver support, arrange to stay at a rehabilitation centre.
- 7. Consult the physician and orthopedic surgeon about the medications being taken and whether any should be discontinued.
- 8. In the weeks before surgery, see the physician for a complete physical examination and routine tests and the orthopedic surgeon for an orthopedic examination.
- 9. Discuss pain management with the anesthesiologist, including the appropriate pain medications for the rehabilitation period.
- 10. Assemble the pertinent medical information that will be needed by the hospital before the surgery. Be sure to raise any outstanding questions or concerns to the medical team.

Checklist

Preparations in the 24 Hours Before Surgery

There are a number of basic preparations and precautions for the patient in the 24 hours before surgery.

- ✓ 1. Take a shower or bath the night before the surgery. This will help reduce the risk of infection.
- ✓ 2. Do not shave the area of the surgery. If this is necessary, the physician will take care of it.
- ✓ 3. Do not eat or drink anything after midnight the night before surgery. This will help prevent nausea from the anesthesia, and more importantly, prevent gastric contents from entering the lungs while under anesthesia (**aspiration pneumonia**).
- ✓ 4. Prepare an overnight bag with items such as:
 - Slippers
 - Pyjamas
 - Bed robe
 - Personal care items (comb, toothbrush, toiletries, shaving kit, etc.)
 - Medications
 - Medical and insurance information
 - Loose-fitting clothes
 - Comfortable shoes



Chapter 5

Orthopedic Surgery, Recovery and Rehabilitation

CHAPTER AT A GLANCE

- **Surgery Day**
- **Patient Self-Care Following Surgery**
- **Symptoms to Report Immediately**
- **Post-Surgery Physiotherapy**
- **Hospital Discharge**
- **Activities to Avoid Until Recovery**
- **Surgery and Medical Follow-Up**
- **Checklist – Important Steps for Rehabilitation**

***P**roper preparation is essential to help ensure a smooth surgery, optimal recovery and maximum rehabilitation.*

Surgery Day

On the day of the surgery, the patient will go to the hospital several hours before the scheduled operation for the admission process. He will meet the anesthesiologist and nursing staff to prepare for surgery and discuss any concerns about the process.

Depending on the individual case, the surgery may take several hours and the patient will then spend an additional two to three hours in the recovery room before being transferred to a hospital room. A member of the medical team will meet with family members immediately after the surgery to discuss how the operation went.

If general anesthesia has been administered, the patient may awaken wearing an oxygen mask, a venous drip to replace fluids, an arm cuff to monitor blood pressure, and a sensor on the fingertip that records pulse and oxygen levels. Post-surgery pain medicine may initially be administered intravenously via a patient-controlled analgesia machine, and will eventually be replaced by oral pain medication. The surgeon and hematologist will make rounds daily to monitor the patient's progress and make any required changes to treatment.



Challenges, Choices, Decisions

A Guide on Orthopedic Surgery for People with Hemophilia

The surgical wound will be checked and the dressing changed daily. The hematologist will also be on call 24 hours a day, should any medical attention be required.

Patient Self-Care Following Surgery

- Administer factor concentrate as prescribed. Treatment should be given in the morning and at the same time each day, and prior to physiotherapy.
- Take pain medications as prescribed. Most patients will require a short-term course of pain medicine for several weeks after discharge.
- Breathe deeply and cough frequently to avoid lung congestion.
- Do not stay in bed once at home. It is important to rest when needed but also to move about frequently, with the help of assistive devices if necessary, gradually increasing the activity level to help the joint heal.
- Keep the incision clean and dry. To avoid accidental reopening of the surgical wound, stitches and surgical staples should be kept in place for a longer time than for normal patients. Staples are usually removed about two weeks after surgery.
- Eat a balanced diet, take any vitamins or iron supplements recommended by the physician, and be sure to drink plenty of fluids. Good nutrition will help the tissues heal and muscles regain strength.
- Follow the exercise program developed by the physiotherapist to help the muscles regain maximum mobility and strength.

Symptoms to Report Immediately

The patient and caregiver must monitor his condition for the development of symptoms or complications. These include:

- An unusually red or hot wound
- An accidental opening at the wound or drainage from the wound
- Pain that is not relieved by medication
- Unusual pain, swelling or redness in the joint area
- A temperature over 101°F / 38.5°C
- Chest pains or breathing problems



Post-Surgery Physiotherapy

While the patient is in hospital, the physiotherapist will visit daily to assess the joint and measure range of motion. The patient will also begin physiotherapy sessions, which are vital to optimal recovery. Factor replacement must be given prior to post-surgery physiotherapy sessions.

Physiotherapy can consist of exercise sessions three times a week for the first two or three weeks after surgery, then twice a week for a month. During a typical session, the physiotherapist might apply ice to reduce pain and swelling and prescribe specific exercises for the patient to do on his own to restore muscle strength and improve joint function.

Exercise is an extremely important aspect of recovery. The patient must be prepared to follow the exercise program prescribed by the physiotherapist for at least three months after surgery. It could require as much as six months to a year to achieve optimal strength and range of motion, depending on the joint involved, its condition at the time of surgery, the type of procedure performed, and how well it went. The physiotherapist will design an exercise program suitable to the joint that was operated upon and the patient's lifestyle.

Hospital Discharge

The patient's discharge from hospital will depend on whether he is going home or to another facility to recuperate. Close contact and regular communication with the physiotherapist and medical staff is important during the rehabilitation period. Most HTC's have a dedicated physiotherapist experienced in hemophilia management, and hospitals generally provide outpatient physiotherapy to patients who have had orthopedic surgery. The nurse coordinator will follow up by telephone on the patient's progress, adherence to medications and physiotherapy, and address any concerns about rehabilitation at home.

Activities to Avoid Until Recovery

Inevitably, there will be some restrictions after surgery. The physiotherapist may initially restrict certain positions and movements, but the patient should be able to resume most light activities within three to six weeks. Following a hip or knee replacement, the patient should not take part in high-impact sports or other rigorous activities (jumping, heavy lifting, etc.) that will threaten the integrity of the new joint. Once recovered from surgery, patients may be able to participate in low-impact sports (cross-country skiing, swimming, cycling, golf, etc.), but it is important not to over-exert or aggravate the joint with vigorous movements. The patient should be sure to clear any intended physical activities with his physician before beginning.



I now have practically unlimited capacity to walk. I still cannot ride a bicycle, due to lack of range of motion in my right knee. Nonetheless, I am now a strong proponent of knee replacement.

– Patient following total knee replacement

Surgery and Medical Follow-Up

About two weeks after leaving the hospital, the patient will return to have the surgical staples removed. At the end of the rehabilitation period, there will be follow-up appointments with the orthopedic surgeon and the physiotherapist. The patient may then be allowed to resume vigorous sports and activities – however, it is possible that certain high-impact activities will need to be avoided because they could put extra wear on the joint. The patient will be assessed again by the medical team at six and twelve months post-surgery.

Checklist

Important Steps for Rehabilitation

For optimal recovery and the best possible surgery results, the patient must adhere to the replacement therapy, pain medication and physiotherapy prescribed by the medical team.

1. Administer factor concentrates, and/or pain medication prescribed.
2. Keep the incision clean and dry.
3. Breathe deeply and cough frequently to avoid lung congestion.
4. Eat a balanced diet, take any vitamins or iron supplements recommended by the physician and drink plenty of fluids.
5. Move about frequently, using assistive devices if necessary, gradually increasing the activity level.
6. Report adverse symptoms immediately. These include an unusually red or hot wound, an accidental opening or drainage from the wound, pain that is not relieved by medication, unusual swelling or redness in the wound area, fever, chest pains or breathing problems.

- ✓ 7. Follow the exercise program developed by the physiotherapist to strengthen muscles and improve mobility. Avoid high-impact activities and sports.
- ✓ 8. Be patient and allow time for the joint to recover and heal.
- ✓ 9. Schedule follow-up appointments with the orthopedic surgeon, physician and physiotherapist to discuss surgery results and maintaining joint health after surgery.

The overall pain level is much reduced. Life is about 1000% better. I am looking forward to being able to do some physical activity with my grandkids. Our country home, which was previously on the 'endangered' list due to my lack of mobility, is now where I hope to spend my rocking chair years.

—Patient following several joint replacements



Glossary

Glossary

Activities of daily living: A defined set of activities necessary for normal self-care: the activities of personal hygiene, dressing, eating, using the toilet, and transferring from a bed to chair.

Analgesic: A type of medication to alleviate pain. Treatment to control pain is called analgesia.

Anesthesia: Induced loss of sensitivity to pain in part or all of the body and sometimes awareness using medication, done for many surgical procedures.

Anti-inflammatory medication: Treatment to reduce the inflammatory response to infectious agents, trauma, surgical procedures, or in musculoskeletal disease.

Aspiration pneumonia: A type of pneumonia while under anesthesia that develops due to the entry of foreign materials into the lungs, usually oral or gastric contents (food, saliva or nasal secretions).

Assistive device: Any device that is designed, made or adapted to help a person perform a particular task. Assistive devices include slings, canes, crutches, walkers and wheelchairs.

Bone cyst: A fluid-filled lesion within the bone that weakens the bone.

Cartilage: Padding between the bones that allows the joint to move smoothly and without pain.

Caregiver: A family member or friend who provides care to the patient at home and makes sure that post-surgery instructions are followed.

Clotting factor: A protein in blood that controls bleeding. Individuals with hemophilia do not have enough clotting factor. Therefore, they bleed for a longer time than normal and experience frequent bleeding into joints, muscles and tissues.

Clotting factor concentrate: A **lyophilized** preparation of clotting proteins that is dissolved in sterile water for infusion to correct a coagulation disorder. Factor concentrates can be manufactured from human plasma or by recombinant technology.

Coagulation: A complex process that makes it possible to stop torn blood vessels from bleeding. Coagulation tests are needed to correctly diagnose the different bleeding disorders, including hemophilia and its severity.

Elective surgery: Surgery that is not urgent or an emergency. The patient, physician and surgeon discuss the options, benefits and risks, and decide whether or when to proceed with the surgery.

Femur: The main bone in the human thigh, also the strongest bone in the body.

Girdlestone Procedure: Removal of the head of the femur and allowing the bone to fuse with the hip socket, a rare procedure done only following a failed total hip replacement due to repeated dislocation or deep infection.

Hemarthrosis: Accumulation of blood in a joint or joint cavity.

Hematoma: A bleed into tissues or a muscle, causing swelling and bruising.

Hemophilia: A genetic bleeding disorder caused by low levels of clotting factor, a protein in blood that controls bleeding. Individuals with hemophilia bleed for a longer time than normal, and experience frequent bleeding into joints, muscles and tissues. Hemophilia is treated with clotting factor concentrates.

Hemophilia A: A genetic disorder characterized by frequent bleeding into joints, muscles and tissues. The prolonged bleeding is caused by low levels of factor VIII and is treated with factor VIII clotting concentrates. The disorder is also called classical hemophilia and factor VIII deficiency.

Hemophilia B: A genetic disorder characterized by frequent bleeding into joints, muscles and tissues. The prolonged bleeding is caused by low levels of factor IX and is treated with factor IX clotting concentrates. The disorder is also called Christmas Disease and factor IX deficiency.

Hemophilia treatment centre (HTC): A medical clinic that provides comprehensive care for people with hemophilia.

Hemophilic arthropathy: Joint damage caused by acute or recurrent bleeding into the joint.

Hemorrhage: The escape of blood from blood vessels, either on the surface of the body or internally.

Hemostasis: Stoppage of bleeding or hemorrhaging in an organ or body part.

Hepatitis C: A viral disease that leads to swelling (inflammation) and scarring (cirrhosis) of the liver. It is transmitted by the exchange of contaminated needles and bodily fluids. In very rare cases, it can still be transmitted by fresh blood components.

Human immunodeficiency virus (HIV): the virus responsible for AIDS.

Immune tolerance therapy: The infusion of high doses of the missing clotting factor concentrate numerous times per week for very long periods of time, usually months or years. The objective of the therapy is to allow the body's defenses to become accustomed to the foreign factor and to stop producing antibodies (inhibitors) against it, so that normal doses will be effective in stopping bleeding.

Infusion: The administration of clotting factor concentrates intravenously using a syringe and butterfly needle, or a central venous access device.

Inhibitor: An antibody produced to attack infused factor VIII or IX or other clotting factor proteins, perceived as foreign by the body's immune system.

Intra-articular: Administered or occurring within a joint.

Intravenous (IV): The infusion of a medication through a tube inserted directly into a vein.

Joint bleed: Caused by a tear in the synovium, blood escapes from the blood vessels and accumulates in the joint cavity.

Lyophilized: Freeze-dried.

Mild hemophilia: A genetic coagulation disorder characterized by bleeding after trauma or surgery. The level of factor VIII or IX in the bloodstream is from 5 to 30 per cent of normal.

Moderate hemophilia: A genetic coagulation disorder characterized by bleeding after minor injury, more serious trauma or surgery. The level of factor VIII or IX in the bloodstream is from 1 to 5 per cent of normal.

Musculoskeletal: Relating to or involving the muscles and the skeleton.

Physiotherapy: The treatment of injuries and musculoskeletal conditions using exercise to strengthen the body or rehabilitate weakened muscles or damaged joints.

Pneumonia: Inflammation of one or both lungs, frequently but not always due to infection. Symptoms may include fever, chills, cough with phlegm, chest pain and shortness of breath.

Prophylaxis/prophylactic treatment: The use of factor replacement therapy several times a week to prevent bleeding, effective for decreasing joint bleeds and delaying the onset of chronic joint damage in individuals with hemophilia and no inhibitors. Prophylaxis is used to prevent joint damage and to break a cycle of repeated bleeds.

Prosthesis: An artificial body part usually made of plastic, metal and/or ceramic that replaces a limb or joint, common when there is irrevocable damage to the knee, hip or ankle joint.

Pseudotumour: An abnormal, gradually expanding cystic mass of blood that occurs as a result of inadequate treatment of a soft tissue bleed, usually in muscle adjacent to bone. A pseudotumour is a potentially limb- and life-threatening condition unique to hemophilia and causes damage to adjacent muscles, nerves and bones.

Radiosynovectomy: Non-surgical synovectomy using intra-articular injections of a radioactive compound to destroy the abnormal synovium.

Radius: The shorter and thicker of the two bones in the forearm.

Rehabilitation: The process of restoration of skills to a person who has had an illness or injury so as to regain maximum self-sufficiency and function in a normal or as near normal a manner as possible.

Respite care services: Temporary substitute care that can be arranged by a social worker to provide the patient's caregiver with a break from the demands of providing care.

Severe hemophilia: A genetic coagulation disorder characterized by spontaneous bleeding and bleeding after minor injury, more serious trauma or surgery. The level of factor VIII or IX in the bloodstream is less than 1 per cent of normal.

Synovitis: Inflammation of the joint lining (synovium) caused by bleeding into the joint.

Synovium: The joint lining, which helps reduce friction and wear on the joint.

Target joint: A joint that is continuously inflamed and vulnerable to recurrent, spontaneous bleeds.

Tibia: The inner and larger of the two bones of the lower leg, extending from the knee to the ankle.



Additional Resources

Additional Resources

Canadian Hemophilia Society

www.hemophilia.ca

- All About Hemophilia: A Guide for Families
- All About Inhibitors
- Assessment and Treatment of Joint and Muscle Bleeds
- Hepatitis C: An Information Booklet for People Infected with the Hepatitis C Virus, and Their Families and Friends
- Hepatitis C: Common Disabling Symptoms and Treatment Side Effects
- Information Booklet on Mild Hemophilia
- Pain Management: The Fifth Vital Sign
- Passport to Well-Being

World Federation of Hemophilia

www.wfh.org

- Emergency Care Issues in Hemophilia
- Exercises for People with Hemophilia
- Guidelines for the Management of Hemophilia
- HIV and HCV Co-infection in Hemophilia
- Musculoskeletal Complications of Hemophilia: The Joint
- The Pain Management Book for People with Hemophilia and Related Bleeding Disorders
- What is Hemophilia?

Challenges, Choices, Decisions

A Guide on Orthopedic Surgery for People with Hemophilia

Notes



Canadian Hemophilia Society
Help Stop the Bleeding