

# JOINT REPLACEMENT SURGERY IN HEMOPHILIA

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Dr. Sam Schulman

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## Introduction

In their severe forms, the inherited X-linked bleeding disorders hemophilia A and B are characterized by spontaneous bleeding into the joints. This is also a prominent clinical feature in patients with type 3 von Willebrand disease and, occasionally, in the severe forms of other coagulation factor deficiencies. The joints most frequently involved are the knees, ankles, and elbows [1, 2]. The consequences of recurrent bleeding into a joint are chronic synovitis and destruction of the articular cartilage and subchondral bone. This condition, called chronic hemophilic arthropathy, causes pain, stiffness, and deformity leading to severe impairment of function. With the advent of the availability of clotting factor concentrates, major surgical procedures can now be performed safely, allowing total joint replacements to be considered for correction of the severely involved joints. The purpose of this monograph is to review the indications for a joint replacement and discuss the pre-operative assessment and the peri-operative and post-operative management.

Of the major joints involved, the knee is associated with the most significant disability and quality of life issues. Since total knee replacement (TKR) is also the most common total joint replacement performed in people with hemophilia, this monograph will concentrate more on the knee replacement procedure; however, the general principles of management remain the same for all joint replacements.

## Indications and Contraindications

### Indications

The decision to proceed with joint replacement surgery can sometimes be difficult for the patient to make. It is usually not recommended until end-stage, bone-on-bone joint disease is present. Many patients, especially those with hemophilia, try to hold out as long as possible as their joint disease presents itself at a much younger age than what is common in the general population. Severe chronic pain that is limiting school, work, or activities of daily living and is unrelieved by pain management

is usually the symptom that causes the patient to seek orthopedic consultation. The indication for a prosthetic joint replacement is a joint with advanced degenerative disease that is painful and may have associated stiffness and deformity, which is causing functional impairment.

If there are no contraindications, a patient who is suffering from hemophilic arthropathy can look forward to the benefits provided by joint replacement surgery.

### Contraindications

Not everyone is a candidate for joint replacement surgery. The main contraindication to performing a joint replacement is the presence of an active infection. Other potential contraindications include local skin problems and co-morbid conditions that would affect outcomes, such as AIDS and liver disease [3-5].

A history of non-compliance with recommended hemophilia care may be a warning of an unsuccessful outcome, as total joint replacement requires a strong commitment to doing rehabilitation exercises. Medically speaking, this is not a contraindication to having the procedure done, however, this history should be considered by the patient, the orthopedic surgeon, and the hemophilia treatment centre (HTC) staff before proceeding with the surgery. There are also psychological barriers that may interfere with a successful outcome. A thorough psychosocial assessment of the patient should be done before scheduling the procedure.

## Pre-operative Assessment

To maximize the possibility of a good outcome, the patient should ideally be seen by the orthopedic/hemophilia treatment center team consisting of the nurse, social worker, and physical therapist (physiotherapist) a minimum of six weeks before the scheduled procedure [6].

### Nursing Considerations

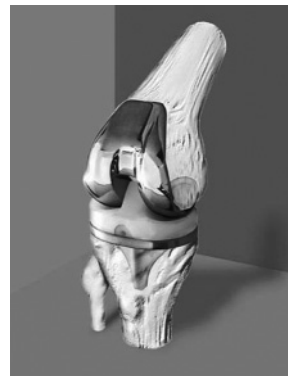
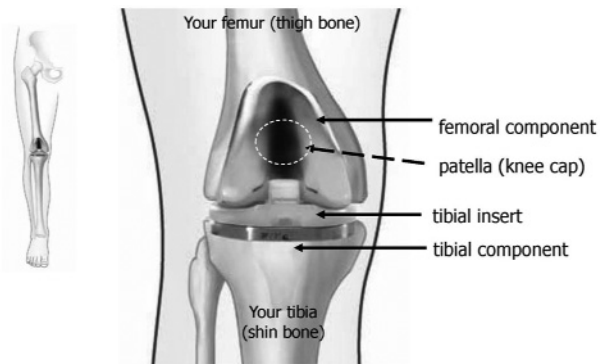
When a person with hemophilia is considering joint replacement, the nurse is often the first person with whom the patient speaks. The nursing role varies

from centre to centre and country to country, but in the comprehensive care setting, the nurse plays a pivotal role in determining whether or not surgery is pursued. Often it is the nurse's responsibility to recognize that a person may be a surgical candidate who would benefit from a joint replacement. It is then the nurse's role to guide the patient to the appropriate physician/healthcare provider to determine if surgery is the correct option.

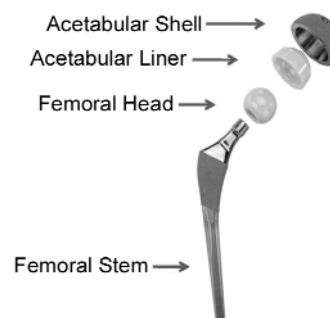
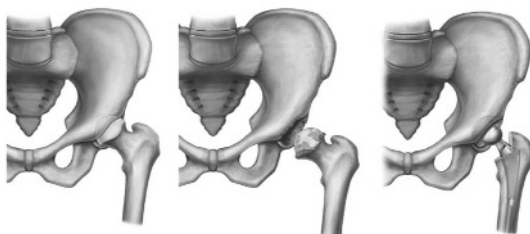
Once it has been decided that a person with hemophilia will undergo a total joint replacement, educating the patient is crucial to a successful outcome. The following topics must be discussed prior to total joint replacement.

1. The person undergoing surgery needs to have a good understanding of hemophilia; how it has impacted the health of the joint and ultimately led to the need for joint replacement. The person undergoing surgery must understand how treatment with factor replacement during and following the surgery will impact the outcome of the procedure.
2. The person undergoing surgery must be motivated to complete the required physiotherapy following the surgical procedure. While the surgery itself may improve range of motion and decrease pain, there is a period following the procedure that requires diligent attention to therapy and a great deal of hard work in order to achieve an optimal range of motion and strength.
3. The person undergoing the joint replacement should have a good understanding of the surgery. Pictures of the total joint replacement may help the individual visualize what the prosthesis looks like and how it will fit into the body (see Figure 1, Figure 2).
4. Outcomes should be discussed. Often, perceived outcomes and realistic outcomes are very different. The nurse should help the person undergoing total joint replacement verbalize the expected gain from surgery and determine if the expectations are realistic.

**Figure 1: Components of a Knee Replacement**



**Figure 2: Components of a Hip Replacement**



5. Time should be taken to review what will happen when the patient is admitted to the hospital, and during the hospital stay. The length of stay may vary greatly depending on the country in which the surgery is being performed. For example, the length of stay for a total knee replacement varies anywhere from 4 to 14 days in the United States. In many countries, the person undergoing a total joint replacement may be hospitalized for several weeks until factor replacement therapy is complete.
6. Laboratory studies must be completed prior to a surgery of this magnitude, including inhibitor testing, prothrombin time in those patients who are hepatitis C-antibody positive, and a full half-life and recovery study in those who have not been exposed to large amounts of clotting factor concentrate in the past. The HIV status of the person with hemophilia should be determined and, while this is not a contraindication to total joint replacement, the patient's HIV should be in good control with a cluster of differentiation 4 (CD4) count greater than 200.
7. While many people with hemophilia are well acquainted with emergency rooms or hospitalization, joint replacement may be their first surgical procedure. Topics such as IV access and continuous fluids, frequent laboratory assessment, vital signs, and anesthesia should be reviewed.
8. Pain management should be discussed at great length both before and following total joint replacement. If the person undergoing the surgery is already on large amounts of narcotic pain medication, pain management could be difficult. The anesthesiologist and orthopedic surgeon must be made aware of this.
9. How the person's hemophilia will be managed must be discussed prior to the surgery. Frequent factor replacement is essential to minimize bleeding and to promote healing following total joint replacement. Surgery of this nature should not be undertaken unless adequate factor supply is secured. This requires communication with the hospital pharmacy, blood bank and/or home care company that will be providing the factor.
10. There is frequently a need for blood transfusion following total joint replacement, and this need is even greater in people with hemophilia. The

person undergoing surgery needs to understand that this is likely, and be willing to receive whole blood and blood products, as well as clotting factor concentrates required for hemostasis.

11. Discharge planning needs to begin prior to surgery. On discharge from the hospital, will the patient be going home or to a rehabilitation centre or nursing home for further rehabilitation? Is he going to need home health nursing or physical therapy? It is advantageous to explore the options with the patient prior to hospitalization.
12. The psychosocial impact of joint replacement should be reviewed with the person undergoing surgery. It is not uncommon for the person to actually grieve the loss of a body part. He may see his arthritic knee as part of his identity and will mourn its loss. It is important that these issues be covered either by the hemophilia nurse or physician or a psychosocial team member.

In addition to the points covered above, it is imperative that the person with hemophilia include his family and/or support network in the education process prior to surgery. A person undergoing major surgery needs the support of family and friends in order for it to be successful. Topics such as employment, income, and time lost from work need to be discussed. The person undergoing this type of surgery should anticipate at least six weeks of recovery time before returning to regular activity.

It is important to secure appropriate resources for the expense of total joint replacement. If insurance is needed to facilitate surgery, then appropriate authorizations may need to be obtained. It is very important that the person undergoing surgery be aware of the cost, both financial and in terms of the time commitment required, to ensure an optimal outcome.

### **Medical Considerations**

Having been exposed to contaminated blood products, many people with hemophilia have chronic HIV or hepatitis C infections. Pre-operative screening should be done to determine whether the current immune status is adequate for surgery and, if necessary, joint replacement should be delayed until adequate antiviral therapy is given. Table 1 shows routine testing that may be advisable prior to a total joint replacement. For patients with chronic hepatitis, the presence of abnormalities such as thrombocytopenia,

other coagulation defects, or cirrhosis-related pulmonary disease should be determined. Patients undergoing interferon and ribavirin therapy for hepatitis C should probably delay joint replacement until the side effects of the treatment have disappeared.

**Table 1. Pre-operative Screening Tests**

- |  |
|--|
| <ul style="list-style-type: none"> <li>• Inhibitor status</li> <li>• HIV antibody, viral load, and CD4 count</li> <li>• Hepatitis C and viral load</li> <li>• Fibrinogen, prothrombin time/INR, platelet count</li> <li>• Cardiopulmonary status</li> <li>• Inspection of venous access</li> </ul> |
|--|

Patients should also be screened for the presence of inhibitors and any past history of inhibitor development should be reviewed carefully. The person with severe factor VIII deficiency who has not had much factor exposure can pose a problem since the *de novo* induction of an inhibitor post-surgery may seriously compromise the end functional result.

A plan for venous access should be determined ahead of time. Many patients can perform peripheral venipuncture for the extensive period of time required for factor infusions. Others will need central access, for example, placement of a peripherally-inserted central catheter (PICC line). Arrangements should also be made pre-operatively for the out-patient infusion support from visiting nurses or other medical professionals.

The choice of factor product depends on the constraints between availability, cost, and patient preference. Successful joint replacements can be done with plasma-derived or recombinant factor and possibly with cryoprecipitate for patients with hemophilia A. Whenever possible, purified factor IX-containing product should be used in people with hemophilia B, since deep venous thrombosis (DVT) and pulmonary embolism were quite common in the past when prothrombin complex concentrate (PCC) was used for orthopedic surgery. Fresh frozen plasma (FFP) usage would probably require plasma exchange to achieve adequate factor levels without volume overload, and should not be used for orthopedic surgery in factor IX deficiency.

It is highly recommended that a laboratory capable of performing factor activity levels be available for

monitoring the patient post-operatively. Since the recovery of activity (factor IX activity in particular) post-surgery can be quite variable, especially in the early period after surgery, the factor infusions can then be specifically tailored to the patient to prevent bleeding and even reduce product usage. If factor activity levels are not available within a reasonable turnaround time (< 24 hours), then the activated partial thromboplastin time (aPTT) should be measured immediately after an infusion of factor. This should be calculated to increase the patient's factor activity to at least 80%, so that the aPTT can be used as a "rough guide" to adequate treatment. Unfortunately, the aPTT may appear prolonged despite factor activity values in the 50-70% range and thus monitoring with aPTT may lead to extra factor use. However, anecdotal experience at the University of Colorado has demonstrated that inadequate trough factor activity levels are frequently associated with post-operative swelling, suggesting that it is better to err on the higher end of the range to maximize the patient's functional outcome [7].

There is widely varying opinion as to whether people with hemophilia undergoing joint replacement should have prophylaxis against DVT with heparin or warfarin. Because the total factor activity levels achieved, especially in factor VIII deficiency, are not as high as those naturally occurring in people without hemophilia in a similar post-operative situation, many centres do not use DVT prophylaxis and have had excellent results without it. If PCC is chosen for factor IX replacement, then guidelines for DVT prophylaxis in non-hemophiliacs should be followed.

In anticipation of the need for blood transfusions during surgery, patients who meet blood bank criteria may donate one or two units of autologous blood in the weeks before surgery.

### Physiotherapy Considerations

The physiotherapist should obtain baseline measurements of range of motion and strength, and note the presence of muscle atrophy or a discrepancy in leg length. Any deficits in other joints should be noted, as these may impact the patient's ability to bear weight properly or may make weight bearing through the arms difficult if crutches or a walker are needed for moving about. The patient's gait should also be assessed and he should be fitted with crutches or a walker.

Any mobility issues and transfers that will be needed should be identified. Adaptive equipment that may be needed in the post-operative period can be ordered in advance of the procedure to allow time for delivery to the patient. These may include items such as a wheelchair, raised toilet seat, shower chair or commode, an abduction wedge pillow, or a gripper for reaching for objects on the floor. Platform attachments for the walker or crutches for forearm weight-bearing should be arranged for in advance of the procedure for patients with elbow problems who might have difficulty bearing weight through extended elbows.

A history should be taken of the person's present need for pain medication and what he is currently taking, as pain management post-operatively can be affected by a patient's medication requirements. The person with hemophilia should be instructed that clotting factor and pain medication are required prior to doing therapy sessions and he should be informed about how pain will be managed during therapy sessions.

The intensity of the therapy sessions and the commitment required to physiotherapy should also be discussed with the patient. It will be easier to handle the challenging times in physiotherapy if the person with hemophilia knows that he may have difficult periods and that he will be able to work through them. It is also recommended that the patient and his family be aware before the surgery that family members may be asked to leave the room during therapy sessions. Since therapy can be very hard work, it may sometimes be difficult for family members to watch a loved one work through the pain that may accompany a therapy session.

During the pre-operative evaluation, the patient should be taught the exercises that will be done immediately after surgery. For those having surgery on the hips or knees, the proper way of moving using crutches or a walker should be taught. Weight-bearing status following surgery will be ordered by the surgeon. A review of any adjunct therapies that might be used should also be explained. These may include the use of ice, continuous passive motion (CPM) for knee replacements, wrapping with a pressure bandage or splinting, electrical stimulation, and transcutaneous electrical nerve stimulation (TENS). Although generally not used in the peri-operative period, whirlpool and superficial heat may be used

in follow-up out-patient therapy once the incision is well healed. Heat should only be used under strict guidelines by the therapist and surgeon to minimize the risk of increased bleeding.

If the person with hemophilia has a significant loss of range of motion and strength, he may benefit from a pre-operative out-patient physiotherapy program to get him in the best possible shape prior to the procedure [8]. Although the exercises are easily done at home, many patients may be reticent about doing them independently for fear of causing increased pain or bleeding. The physiotherapist at the HTC is the best resource for a regular out-patient program, but since many patients do not live within close proximity to their HTC, the hemophilia physiotherapist can contact a therapist in the patient's local community to arrange for the therapy. It is recommended that the HTC therapist talk directly with the local physiotherapist to provide education on the special needs of treating someone with hemophilia, specifically regarding the use of factor replacement therapy prior to the physiotherapy session. The hemophilia therapist can continue to act as a resource as questions or problems may arise, either pre- or post-operatively. Establishing a relationship between the physiotherapist at the HTC and the local physiotherapist will optimize the physiotherapy experience and lead to a better outcome for regaining range of motion and strength.

### **Surgical Considerations**

The evaluation of a person with hemophilia who is a candidate for a joint replacement requires a thorough physical examination. This should include observing the person's gait and performing activities of daily living such as climbing stairs and changing positions (i.e. from sitting to standing). Although the majority of joint procedures will involve the lower extremity, upper extremity function should also be evaluated in order to assess all the patient's needs and determine the best way to meet them, particularly in the immediate post-operative period.

This part of the evaluation is very important to assist the medical team in making a decision as to whether combining surgical procedures would be a consideration. Since most patients have multiple joint involvement and are potentially facing a decision about the necessity for more than one operation, considering combining at least two procedures is very appropriate. Examples of combining procedures

include bilateral total knee (most common); a total hip and a total knee replacement; a total hip or total knee and an ankle procedure. The decision to consider combining procedures should be based on the obvious need and also on the expectations that the recovery will not be compromised for any of the procedures performed.

Specific aspects of the affected joints examination should include active and passive range of motion, stability, deformities (fixed or correctable), and strength of muscle function.

Radiographic evaluation should include standard views of the joints involved. In the lower extremity, weight-bearing films are recommended as well as a long antero-posterior hip-to-ankle film. The important radiographic features to recognize are the deformities and bone loss. The information gathered from this observation will help with the surgical planning, including the need for soft tissue releases and managing bone loss (bone graft material or prosthetic augmentation).

## Peri-operative Treatment

### Medical Considerations

#### *Factor replacement for factor VIII deficiency*

The factor level should be corrected to 120% corresponding to 60 international units (IU)/kg of FVIII, as close to induction of anesthesia as possible (see Table 2). If the patient is still in the operating room

at four hours post-dose, another 20 IU/kg should be given. Factor VIII activity should be assayed when the patient is in the recovery room. The goal is to have a factor VIII activity level between 60-80% for about 72 hours post-surgery. This can be achieved either by bolus treatment every 12 hours or by continuous infusion of factor VIII. A daily trough factor VIII activity will help the clinician adjust the doses. In the recovery room, some patients will require an immediate bolus to bring them to the required range, but most will achieve the required levels with 25-40 IU/kg every 24 hours. Continuous infusion of factor VIII is started at 2 IU/kg/hour for adults and 4 IU/kg/hour for children after determining that the baseline level is at least 70%. The patient may require bolus therapy to achieve adequate basal level on which to start the continuous infusion. The monitoring of the continuous infusion can be done at any time and the drip is titrated up or down in order to maintain the desired level. At 72 hours, the doses are changed to maintain a trough factor VIII activity level of 50%. Many patients on continuous infusion will be changed to bolus dosing at this time.

#### *Factor replacement for factor IX deficiency*

The range of half-life and recovery of the purified and recombinant factor IX products is extremely variable from patient to patient and in different clinical situations. Therefore, it is especially important to have a laboratory monitoring plan. The factor level should be corrected with 120 IU/kg on induction of anesthesia. If the patient is still in surgery four hours after the dose, then another 40 IU/kg should be given and factor IX activity should be obtained in

**Table 2. Factor Replacement for Orthopedic Surgery**

	Factor VIII Deficiency	Factor IX Deficiency
Pre-operative (induction of anesthesia)	120% (60 IU/kg)	120% (120-140 IU/kg)
After 4 hours in operating room	Extra 40% (20 IU/kg)	Extra 40% (40 IU/kg)
72 hours post-operative	Maintain trough of 60-80%	Maintain trough of 60-80%
Up to 14 days post-operative	Trough of 50%	Trough of 50%
Week 3-4	Trough of 30-40%	Trough of 20-40%
Week 5-6	Before each PT-session 40%	Before each PT-session 40%

PT: Physiotherapy

the recovery room. Similar factor activities as described in the above section on factor VIII deficiency are desirable, with trough levels of 60-80% expected during the first 72 hours, followed by 50% for two weeks, then 40% for the next week or two depending on the status of the replaced joint. Usually, the factor IX-deficient patient can be managed with a daily infusion but the dose can be quite variable, ranging from 50-80 IU/kg daily then decreasing gradually to 40-50 IU/kg. Some patients with excellent resolution of wound swelling and healing may be able to receive factor every other day at 40 IU/kg after the third week.

Prior to every physiotherapy session, patients with factor VIII deficiency or factor IX deficiency should receive a minimum of 40% correction. A similar dose should be given prior to suture removal.

#### *Other blood product support*

Although intra-operative cell saver devices can salvage red blood cells during joint replacement surgery, many patients will have major drops in hemoglobin during the first two to three post-operative days. It is not unusual to see a 6-8 g/dl decrease in hemoglobin, which may require a transfusion of packed red blood cells. Oral or IV iron replacement should be considered to facilitate erythropoiesis. The major hemostatic challenge of orthopedic surgery can unmask defects of protein synthesis in chronic liver disease. Patients with chronic hepatitis should have fibrinogen activity, prothrombin time/INR, and platelet count monitored in the recovery room and for at least the first post-operative day. Fibrinogen should be maintained above 150 mg/dl, INR < 1.5, and platelets >50,000 for the first couple of days. Vitamin K can be given to improve hepatic synthesis. Fibrinogen can be replaced with fibrinogen concentrate or cryoprecipitate and fresh frozen plasma can be used to correct the prolonged prothrombin time. If a severely anemic patient refuses red cell transfusion, aggressive IV iron infusion (up to 1,000 mg) with erythropoietin injections can be used depending on the severity of the anemia.

### **Physiotherapy Considerations**

Physiotherapy usually begins the day after the procedure. Therapy is ordered by the orthopedic surgeon and will vary depending on the surgeon's preference and the nature of the procedure being done. In the recovery room, adjunct therapies of ice, compression, and elevation will be initiated in addition to the previously mentioned factor replacement and

pain management protocols. With total knee replacements CPM is usually started the evening of the surgery and is used intermittently during the inpatient admission. If a patient is having difficulty regaining range of motion, CPM may also be used in the home after discharge. If maintaining good extension is an issue, an extension splint may be worn periodically throughout the day or for sleeping at night. With total hip replacements an abduction wedge pillow will be placed between the legs. Generally, physiotherapy will be started the day following the surgery, with therapy progressing slowly as the patient is able to tolerate it. The first day may only consist of sitting or standing at bedside or transferring with help into a chair, if that is all that can be tolerated. Each day the patient will do more, starting with isometric exercises and progressing to the active exercises that were learned pre-operatively. The distance that the patient is able to walk will also increase each day. If continuous infusion is not being done, therapy sessions should be scheduled around factor infusion and pain medication schedules. Ideally the patient should receive physiotherapy twice daily during the inpatient admission.

Therapy sessions focus on regaining joint range of motion and strengthening muscles adjacent to the joint that was replaced, in addition to increasing endurance when moving about. For knee or hip replacements, the amount of weight that can be put on the leg will be determined by the surgeon and the type of joint replacement performed. The length of time that crutches or a walker will be required is determined by the surgeon but usually ranges from six to eight weeks. There will also be restrictions on driving a vehicle during the immediate post-operative period. If factor levels are monitored daily and adequate levels are achieved, post-operative exercises for people with bleeding disorders are similar to exercises for the general population following joint replacement [9, 10]. To prevent the formation of motion-limiting adhesions, it is important to begin joint mobility exercises in the early post-operative days and expect increasingly aggressive therapy, including resistive exercise, through the first several months. Early mobilization and aggressive work on regaining motion are critical to achieving a successful outcome. Once the patient is discharged from the hospital, clotting factor infusion will still be necessary prior to physiotherapy sessions. This will continue for a minimum of six weeks, with the length of time determined by the surgeon, hematologist, treatment

nurse, and physiotherapist, based on how he is doing in his therapy program.

### **Surgical Considerations**

A detailed pre-operative evaluation should provide the operating surgeon with enough information to avoid any unexpected encounters during the surgery. An experienced surgeon knows clearly that the deformities and the bony changes encountered in hemophilic arthropathy are unique. This once again emphasizes the importance of these procedures being performed in a centre experienced in treating patients with hemophilia.

The knee is the joint that will challenge the surgeon the most. A complex set of deformities are common in the advanced stage of arthropathy, particularly in neglected cases. In more severe cases, a flexion contracture exists with restricted motion. Angular deformity with translation of the tibia is a common finding. Patellofemoral abnormalities always exist. The most common are lateral subluxation or dislocation, central location of the patella with deepening of the femur groove, and patella baja. These complex deformities contribute to a difficult surgical procedure, which in turn leads to a great variation in the eventual outcome [11-14].

Nevertheless, appropriate planning for surgery should prevent adverse outcomes. Some of the common issues to consider are recognizing the oversized dimension of the distal femur and proximal tibia due to overgrowth associated with frequent bleeding episodes in childhood, as well as identifying bone defects and loss that require bone grafts or prosthetic augments. Another consideration is creating a surgical exposure to manage these complexities. Antibiotic loaded cement should be used in all cases where cement fixation is used, wound closure should be meticulous and tight, and drains should be in place for 24 hours. Once the procedure is completed, the surgeon will have a good understanding of the potential range of motion the patient can expect.

## **Post-operative Treatment**

### **Nursing Considerations**

If factor or blood products are given continuously, it is important that they be administered through a dedicated IV. Blood samples should not be drawn through this same IV line in order to assure accurate factor levels.

If the patient will be infusing his own factor in the home setting, it may be necessary to have a PICC line or some other temporary form of venous access in place. However, many people with hemophilia do not require this, as they prefer to give themselves daily injections.

### **Medical Considerations**

#### *Factor Replacement*

Two weeks from the date of surgery, the dose of factor replacement therapy is modified to maintain a trough of 40% and patients may be switched to a daily infusion. For the third and fourth weeks post-operatively, patients receive 20-40 IU/kg/daily depending on the clinical status of the joint replaced. Any increase in swelling or pain is promptly treated with full dose factor concentrate for several days until it subsides. Factor concentrate should be given prior to suture removal if a regularly scheduled dose was not recently infused. All physiotherapy sessions should be pretreated with 20 IU/kg for up to six weeks post-operatively. Some patients with mild factor VIII deficiency may be able to use nasal desmopressin alternating with factor infusions after the first week or two, but an adequate response should be demonstrated by assaying factor activity.

#### *Pain Management*

Post-operative pain management is important to facilitate the mobilization and physical therapy of the replaced joint. Regional anesthesia with epidural catheters can be quite useful in the first 24-48 hours after surgery. There is, however, a risk of spinal/epidural hematoma, which can result in paraplegia, if high levels of the deficient factor are not maintained. Peripheral nerve block (i.e. femoral nerve block) for total knee arthroplasty can also provide very good pain control in the immediate post-operative period [15].

Patient-controlled intravenous analgesia is a very effective method and can usually be converted to oral narcotics in two to three days. Patients with severe joint damage may be taking high doses of narcotics as out-patients and consultation with a pain service may help transition to the doses necessary for post-operative relief of pain. Some patients experience a remarkable relief of pain when the offending joint is removed and do not require long-term pain management.

### **Surgical Considerations**

The drains are removed at 24 hours following surgery, and the first dressing is changed at 48 hours. Continuous full-leg compression stocking and frequent ice packing are critical in the initial post-operative period. Daily wound/leg inspections determine how long local treatments are necessary. Range of motion exercises can be started immediately post-surgery, or it may be appropriate to keep the knee in an extended, resting, splinted position for the first 24 to 48 hours.

### **Physiotherapy Considerations**

For the more than 30 years that total joint replacements have been done in people with bleeding disorders, the length of hospitalization in the United States has significantly decreased and now averages from four to 14 days. This may vary in other countries where patients may remain as in-patients until factor replacement therapy is complete. Because of the very short in-patient opportunity for physiotherapy and the need for an aggressive exercise program, some patients may be admitted to a rehabilitation facility or be seen in the home by a homecare physiotherapist. The degree of post-operative follow-up will depend upon how quickly the patient regains range of motion and strength in the joint. If the patient is doing well and is able to get transportation, it is optimal for him to be seen in an out-patient clinic, which is better equipped to provide therapy equipment such as a stationary bicycle, balance equipment, and therapy methods. A therapist comfortable treating someone with hemophilia is preferable, but if this is not an option then it is recommended that the therapist from the HTC speak directly with the therapist who will be following the patient, as is recommended pre-operatively. If the therapist sees the patient regularly, two to three times per week, any problems that might arise will be noted early and the appropriate intervention can be initiated more quickly.

Physiotherapy after joint replacement surgery may require a commitment of several months of dedication to the exercise program. With regular attendance at therapy sessions and by working independently at home, patients can expect a good outcome.

### **Long-term Joint Replacement Management**

A person undergoing a total joint replacement using current techniques and prosthetics can expect the artificial replacement to last for many years – more than 20 years. The long-term survival of a prosthetic

joint replacement depends on getting it right to start with, meaning good surgical techniques, plus using current materials to prolong the wear characteristics of the implant, and finally, avoiding complications that could cause failure [16-22].

A person with hemophilia who undergoes a total knee or hip replacement can feel confident that he will not wear out the prosthesis. What will really determine long-term success is avoiding complications.

### **Complications**

Complications from joint replacement surgery can be classified as operative and post-operative. Operative complications include those associated with the anesthetic and those related to the surgical procedure. These risks/complications are not unique to the people with hemophilia, but are what any person undergoing these operations would face. Bleeding is the one complication for which people with hemophilia may have an increased risk.

Post-operative complications include those occurring in the immediate post-operative period (first two weeks) and those occurring later and indefinitely. The immediate post-operative complication most commonly seen is excessive wound hematoma with swelling. In rare cases, a return to the operating room for evacuation of a wound hematoma may be necessary. Failure to gain motion may also be recognized in the early post-operative period. A few cases have required manipulations under anesthesia.

The most common complication that can lead to a catastrophic failure is infection. This can occur any time from immediately post-operatively through the rest of a person's life. The risk of an immediate post-operative infection in a person with hemophilia should be no higher than the general population. Surgeons using peri-operative antibiotics as well as antibiotic-loaded cement during surgery, and performing surgery in an operating room for joint replacements will result in a less than 1% risk of infection. However, it is well documented that people with hemophilia are at a greater risk of delayed secondary infection [23-30]. The reason for this higher risk in a person with hemophilia is not clear. What usually causes a delayed infection is a bacteremia from a distant or local infection. A real potential source of bacteremia in a person with hemophilia is through contamination during intravenous factor replacement. Another source of bacteremia is a dental procedure.

For long-term prevention of infection, people with hemophilia should use sterile precautions with IV access and take prophylactic antibiotics at the time of dental procedures [31].

An important scenario that a person with hemophilia who has had a successful total joint replacement may experience is a sudden onset of pain and swelling of the replaced joint. One might think that this may be an acute bleeding episode but it is uncommon to have a spontaneous bleeding occurrence in a totally replaced joint. Failure to respond to factor replacement should be a clear signal that this may be something else other than a bleed. Infection is the diagnosis until proven otherwise. Immediate aspiration of the joint and studies of the fluid including gram stain and cultures should be performed. It is strongly recommended that an aspiration be performed by an orthopedic surgeon who is available to participate in the appropriate management.

Long-term complications of prosthetic failure are low risk. They include prosthetic wear, loosening, and breakage.

Needless to say, it is very important for the person with hemophilia to strive for a healthy lifestyle. Maintaining a strong immune system and good dental hygiene as well as receiving immediate treatment of any infection will significantly improve the chances of long-term success of a total joint replacement.

### Joint Replacement in Factor VIII-Deficient Inhibitor Patients

Joint replacement can be successful in inhibitor patients on immune tolerance therapy who are able to achieve a factor VIII activity of 50-100% with reasonable doses of factor concentrate. Such patients should have intensive monitoring of factor VIII activities post-operatively. The inhibitor patient with a low titer and history of a low-responding inhibitor might also be able to achieve the two to four weeks of hemostasis required for good surgical results. However, many inhibitor patients will have titers too high for adequate factor VIII therapy. There are reports of patients undergoing joint replacement with bypassing agents, particularly with recombinant factor VIIa. This requires a major use of resources and, since it is potentially life threatening, should only be undertaken with full

understanding of the risks. Such surgery should be managed only by hemophilia centres with extensive experience in inhibitor treatment [32-44].

### Conclusions

Total joint replacement has been successfully performed in people with hemophilia for over 30 years. By far the greatest numbers have been total knee replacements. The benefits of pain relief and improved function provided by total joint replacement make this procedure the most successful orthopedic operation for managing chronic hemophilic arthropathy. To increase the chances of a successful outcome and decrease the risks, these procedures should be performed in hospitals where there is an established HTC with all the specialists available. 🌐

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