

Contributors

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Fred was the first person with haemophilia to have a knee replacement at Manchester Royal Infirmary. He has had a number of operations: left knee four times, right knee twice, and a full left ankle fusion. The quotations included in this booklet reflect his own experience and opinion, not those of Baxter Healthcare.



Preparing for surgery

Information for people with haemophilia

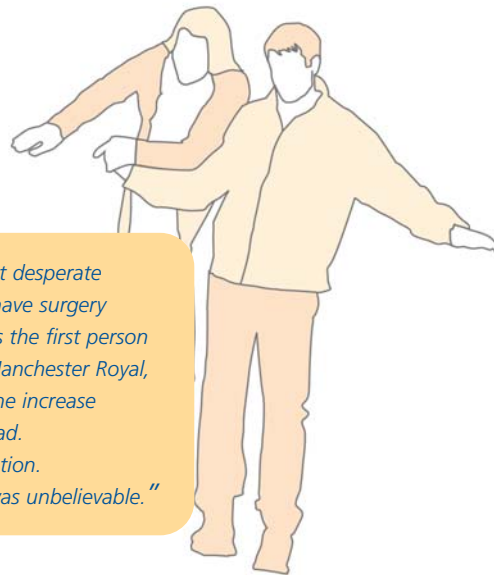
The need for surgery

AT SOME TIME IN THEIR LIVES, ALMOST EVERYBODY WILL NEED SOME SORT OF SURGERY, WHETHER IT IS MINOR, SUCH AS REMOVAL OF AN IN-GROWING TOENAIL, OR MAJOR, SUCH AS A KNEE REPLACEMENT.

While no one looks forward to surgery, we accept it because we know that the benefits will far outweigh the risks. This is also true for people with haemophilia or other bleeding disorders, even though surgery is never totally straightforward and may have a slightly higher risk of complications.

If you have haemophilia, surgery may also become necessary because of the damage done to target joints by repeated bleeds. Quite apart from the pain and restricted movement that a badly damaged target joint may cause, if left untreated it may also lead to problems in other joints.

For example, after repeated lower limb bleeds you may develop a limp, which can cause back pain, while using walking aids such as crutches can trigger bleeds in the arms and shoulders. Fused ankles due to severe joint damage could trigger knee bleeds in either the affected or unaffected sides. Consequences such as these may be avoided by timely surgery.



"I had so much pain in my knees, I was almost desperate for surgery. It was quite a simple decision – have surgery or end up in a wheelchair. Even though I was the first person with haemophilia to have the operation at Manchester Royal, the prospect of the pain disappearing, and the increase in mobility, overcame any doubts or fears I had. Would I recommend surgery? Without hesitation. The reduction in pain – almost overnight – was unbelievable."

When you need surgery

If you have haemophilia or another clotting disorder, you may be concerned by the prospect of surgery, because of the risk of bleeding during the procedure.

The special requirements of people with haemophilia during surgery are now well recognised, and prophylaxis regimes to prevent excessive bleeding are well established. Provided that all those involved are aware of the need for factor cover and it is given correctly, the risk is no greater than for any other patient.

Planned surgery

Surgery may be either planned or required in an emergency, but in either case proper communication between all those involved is vital. In the case of planned surgery this will not only be you and your surgeon, but also your haemophilia centre, your comprehensive care centre and the consultant haematologist and laboratory at the hospital where the operation is to be carried out.

Emergency surgery

If you need emergency surgery, for instance as the result of an accident or for appendicitis, then the operation will go ahead irrespective of your condition. It is important that you, or a relative or friend, makes sure that the surgeon knows about your haemophilia, and that he works in consultation with your haemophilia centre or the on-call haematologist to ensure you have the proper factor replacement cover. It is a good idea always to wear an alert bracelet and carry an emergency advice card.



The advantages of planned, early surgery

EVERY TIME YOU HAVE A BLEED INTO A JOINT, ITS LINING BECOMES SWOLLEN AND INFLAMED. NATURAL SUBSTANCES IN THE BLOOD ATTACK BOTH THE JOINT LINING AND THE CARTILAGE COVERING THE END OF THE BONES, SO THAT EVENTUALLY THE JOINT LINING BECOMES RAGGED AND THE CARTILAGE BREAKS DOWN, LEAVING ROUGH AREAS OF EXPOSED BONE. THESE CHANGES CAN LEAD TO MORE BLEEDING, PAIN, A DECREASED RANGE OF MOVEMENT AND ULTIMATELY TO SEVERE JOINT DAMAGE.

If this happens, pain and bleeding into the joint may become constant and interfere with everyday life. Sometimes, because of their understandable concerns about surgery, people with haemophilia put up with this increasing pain and disability until there is no other option. But, if you are suffering pain and impaired movement, it is better to discuss the possibility of early orthopaedic referral with your haemophilia centre as soon as possible, because there is a lot that can be done.



When treatments such as physiotherapy and painkillers are no longer adequate, there are a number of operations which can be done before the joint is badly damaged. These include removing the damaged joint lining, cleaning up the joint surfaces and removing any loose fragments or clearing away any bony growths from around the joints. Many of these operations can be done through tiny incisions using 'keyhole' techniques.

Taking action at this early stage has great advantages. These are much simpler operations, with less likelihood of complications and, because they are much shorter, there is less risk from bleeding. They may also prevent or slow the progress of joint damage and avoid the need for joint replacement later on

In the long run, relatively minor, planned surgery early on not only relieves pain and lets the joint work better to keep you mobile, but may also reduce the likelihood of needing major surgery later and the risk of damage to other joints.

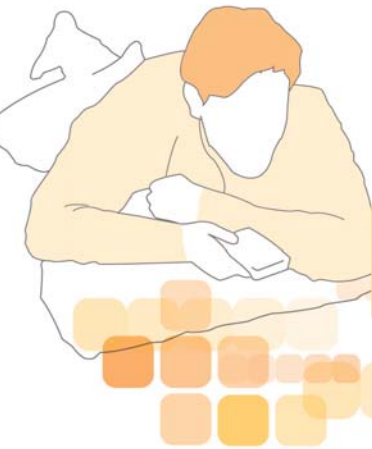


Planning for surgery

WHEN YOU MAKE THE DECISION TO OPT FOR SURGERY, IT GIVES YOU THE FREEDOM TO PLAN AND TO MAKE YOUR OWN CHOICES, IN CONJUNCTION WITH YOUR HAEMOPHILIA CENTRE OR COMPREHENSIVE CARE CENTRE.

Different hospitals have different policies about surgery for people with haemophilia and different expectations of what they can achieve, so it's important to find one which has the facilities you need and will undertake the surgery you want. Your comprehensive care centre will be able to advise you.

They will also help you find a surgeon who has the skills, experience and success rate you are looking for. Even though your surgeon may not have operated on someone with haemophilia before, good haemophilia care from a haematology department accustomed to dealing with it, will make the operation no different from normal.



"I used a muscle stimulator (similar to a TENS machine) a couple of times a day for about 6 weeks before my operation. That built up a bit of muscle that then helped with the post-op physiotherapy."



Once you have made the decision, your surgeon will give you a full explanation of what the surgery involves and what you need to do to prepare yourself. This will involve putting in place a plan for your stay in hospital: when you will go in, when the operation will be done, what blood tests by the laboratory will be needed while you are in and how long you will have to stay. He or she will also sort out any pain relief you require in the meantime.

In some cases it may also be necessary for the Hospital Trust to negotiate the funding for the operation from your Primary Care Trust.



"Never be afraid to ask. My orthopaedic surgeon was very open and honest with me about all aspects of the operation. Even though we discussed worst case scenarios I could tell he was only thinking of a positive outcome and that really put my mind at ease."

Minimising the risks

Surgery for people with haemophilia is becoming more and more common. As surgeons, haematologists and hospitals have gained more experience, they have been able to progressively reduce the risks of the operations.

To make sure the risk level is kept as low as possible, all surgery is performed only after extensive consultation with your comprehensive care centre, and following exhaustive checks to make sure you are fit for the operation. During the operation itself, you will be under the care of an experienced haematologist, who will make sure that bleeding is kept under control.

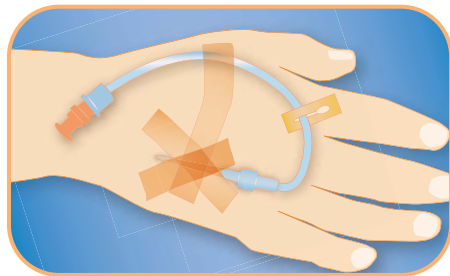
As a result, surgery today carries less risk for people with haemophilia than it did previously.

Factor cover for surgery

AS YOU WOULD EXPECT, BLEEDING DURING SURGERY WILL BE KEPT UNDER CONTROL BY GIVING YOU FACTOR. THE DOSE YOU NEED WILL BE CALCULATED ON AN INDIVIDUAL BASIS TO KEEP YOUR BLOOD FACTOR LEVELS WITHIN THE NORMAL RANGE THROUGHOUT.

When you have been given your pre-operative factor cover, your blood levels will be checked very carefully, to make sure the concentration is up to the target. If your haematologist is satisfied, the operation can start. If not, more factor will be given.

Your factor levels will also be monitored closely if you are under-going lengthy surgery, so that they can be topped up if necessary. Under certain circumstances, some haematologists are now giving factor by continuous infusion, to maintain a constant factor level.



Some hospitals allow patients to perform their own infusions as soon as they are well enough, and will prescribe factor for self-administration.

After your operation your factor levels will continue to be checked at regular intervals to ensure satisfactory levels. You will also be given prophylaxis to cover you until your wound has healed and during your post-operative physiotherapy.

Because this regime means there will be a need for frequent infusions, you may be given a temporary venous access device, such as a peripheral cannula, which is inserted into a vein.

"I had blood tests before surgery, and my factor cover was adjusted. On the day of the operation, I had 4,000 units in the morning and 2,000 in the evening. The next day it was 2,000 in the morning and 1,000 in the evening and for the following eight days, a total of 2,000 units a day. After that, to cover the period of physiotherapy, I had 2,000 units every other day for six weeks."

If you have an inhibitor

Inhibitors make the control of haemophilia much harder, but even these are not necessarily a bar to surgery. If you have an inhibitor, you will need to work closely with the specialists at your comprehensive care centre to devise the best individual approach for you.

If you only have a low titre inhibitor, it may be possible to proceed in the same way as for a person with no inhibitors. You will, however, need a somewhat higher factor dosage to achieve the target blood factor level, and this will probably need more frequent topping up.

Even if your inhibitor titre is high, it may be possible to temporarily bring it down. More usually, treatment with either recombinant factor VIIa (rFVIIa) or activated prothrombin complex (APCC) is used to bypass the inhibitor activity.

Both these treatments are expensive, and your comprehensive care centre may have to agree funding with your Primary Care Trust prior to surgery being undertaken.



After the operation

WHEN YOU HAVE HAD YOUR OPERATION, THERE IS STILL A LOT MORE TO BE DONE TO COMPLETE YOUR RECOVERY. IF YOU HAVE HAD GENERAL SURGERY, YOU MAY NOT REQUIRE PHYSIOTHERAPY BUT YOU WILL STILL NEED TO GET YOURSELF UP AND ABOUT AS SOON AS POSSIBLE.

Pain management

After the operation, you will be given whatever pain relief you need. Good pain control is important throughout your recovery. To start with you may be given pain relief by intravenous infusion or by a pump that allows you to control your own dosage (patient-controlled analgesia or PCA).

Intravenous painkillers are then gradually withdrawn and replaced with tablets. Alternative methods such as ice packs or acupuncture may also be used. Discuss the options with your haemophilia centre.



Physiotherapy

If you have had orthopaedic surgery, you will have to put in a lot of hard work. Immediately after your operation and (depending on the type of operation) for some days or weeks after you will have individually planned physiotherapy in addition to continuing factor cover. The physiotherapy will differ for different operations but the main objective is to get you mobile as soon as possible.

While you are in hospital your physiotherapist will see you once or twice per day. They will measure how well your joint is moving and keep you informed of how you are improving. You will be able to rely on his or her support and encouragement throughout.

Occupational therapy

Your occupational therapist may visit you at home before the operation so that they can see what help you might need afterwards to cope with everyday tasks. After the operation you may be shown the techniques you will need to look after yourself on your own and help you get ready to be sent home.



"I knew I wouldn't be pain-free straight after the operation, but it wasn't as sore as I thought. And, after 3 or 4 weeks of physiotherapy I felt comfortable and didn't experience any pain. The first time in over 10 years. But the physiotherapy didn't stop there. It had to continue to keep the mobility."

Common orthopaedic procedures

Knee replacement

For knee replacements intensive pre-operative physiotherapy can be very helpful in addition to post-operative treatment.

On the first day after your operation you will be treated as normal, with routine chest physiotherapy if necessary. After that, providing your factor levels are adequate, knee bending exercises will be introduced. You will usually be encouraged to get out of bed, with a splint or brace on your knee to start with. Once you are able to lift your leg off the bed while keeping it straight, this brace will be removed.

Once you have good muscular control, can bend your knee to a reasonable degree and can move well enough to manage stairs, you can be sent home. You will continue to receive regular outpatient physiotherapy until you have achieved as great a range of movement as you can. Even if the operation does not give you any great increase in range of movement, it should reduce the amount of pain you suffer.



Ankle surgery

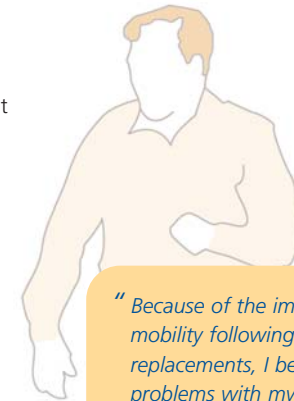
Ankles are commonly involved as target joints and can be the source of severe pain and disability. Due to their complexity, however, ankle joint replacement is currently not performed. There are a number of other procedures which can provide significant relief.

The most commonly used form of ankle surgery is called arthrodesis. Arthrodesis involves fusing the surfaces of the joint together. This leaves the joint completely frozen, but you will still be able to walk and the joint will be painless.

Synovectomy is the removal of damaged joint lining and can be carried out either by open or 'keyhole' surgery.

Cheilectomy is the removal of small bony growths around the joint margins, which gives improved range of movement with less pain. This too may be performed using keyhole techniques.

After all these procedures your physiotherapist will help you get moving as quickly after the operation as possible.



" Because of the improvement in mobility following my knee replacements, I began to have problems with my ankles. But an ankle fusion has solved that now."

Less common orthopaedic procedures

Shoulder Replacement

After this operation recovery is more complex, because a shoulder joint replacement can dislocate quite easily.

Following the operation your shoulder may be kept in a shoulder immobiliser for up to six weeks. This will only be removed for exercises. On the other hand you will be encouraged to be active straight away and on the first day you will undertake active finger, wrist and elbow movements, with chest physiotherapy if needed.

While you remain in hospital, you will have frequent passive exercises performed by the physiotherapist. After six to seven days you will probably be sent home. You will then continue your physiotherapy as an outpatient. The regimen will be strict to allow the muscles, tendons and ligaments around the joint to heal. The whole recovery period can take up to three months of intensive physiotherapy.

Elbow Surgery

Postoperative treatment will vary, depending on the operation performed. Usually your surgeon will recommend getting the joint moving as soon as possible, supervised by the physiotherapist. In the initial stages the joint may be immobilised in a back-slab, which is only removed for exercises.

Activities can be gradually increased but carrying any weight should be avoided. Continuing outpatient physiotherapy will be necessary to increase your muscle strength and range of movement. During this time ice therapy or heat treatment may be used in addition to other physiotherapy techniques to reduce swelling and help with pain relief.



Personal details

Name: DoB:

Address:

Hospital number:

Diagnosis:

Severity/Factor level:

Inhibitor Status:

Treatment: (Name of product)

Dosage by bleed type

Life or Limb Threatening:

Major:

Minor:

Other information

Emergency contact/next of kin:

Relationship: Telephone:

Haemophilia Centre:

Consultant:

Telephone: Bleep/Pager:

Nurse:

Telephone: Bleep/Pager:

Comprehensive Care Centre:

Consultant:

Telephone: Bleep/Pager: