TOTAL KNEE REPLACEMENT IN HAEMOPHILIC ARTHROPATHY

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Eleven total knee replacements were performed in eight patients with severe haemophilia A and the patients were followed up for two to eight years. All the patients had disabling haemophilic arthropathy of one or both knees, which had not responded to conservative treatment. Postoperative complications occurred in 10 knees, including nose bleeding, haemarthrosis, anaphylactic reactions, urinary tract infection with haematuria, recurrent phlebitis at infusion sites, and fever for a few days. There were no wound infections.

The outcome, as determined by a standard scoring system, was rated as excellent or good in nine knees, fair in one and poor in one. Nevertheless, all patients were free of pain and all but one returned to full-time or part-time employment. Total knee arthroplasty appears to be a satisfactory procedure in the treatment of disabling haemophilic arthropathy of the knee.

In haemophilic arthropathy the knee is the most frequently affected joint. The early stage of the arthropathy is characterised by recurrent bleeding and synovial thickening; in the later stages pain, reduced range of movement and deformities, including posterior subluxation, are the characteristic features.

The current availability of Factor VIII replacement therapy enables the bleeding episodes to be managed and may prevent or delay the development of the arthropathy. It also makes surgical intervention possible when conservative measures fail.

This present study involved eight young men with haemophilia A (Factor VIII:C <1%), who underwent total knee replacement because of disabling haemophilic arthropathy of one or both knees. Careful selection and preparation preceded the elective procedure, and the patient was in the care of a multidisciplinary team of specialists including an orthopaedic surgeon, a haematologist and a physiotherapist.

MATERIALS AND METHODS

In the period 1978 to 1985 11 total knee arthroplasties were performed in eight men with severe haemophilia A. Their ages at operation ranged from 31 to 50 years. The knee-rating scale of the Hospital for Special Surgery was used for pre-operative evaluation (Insall et al. 1976). A score of 85 to 100 points was deemed to be an excellent result; 70 to 84 points, good; 60 to 69 points, fair; and less than 60 points, poor. In our series the pre-operative values varied from 25 to 56 (Table 1). Radiographic assessment was made using the Pettersson score (Pettersson, Ahlberg and Nilsson 1980). We found values between 6 and 11.

All patients in this series were disabled by persistent pain and dysfunction of one or both knees, caused by osteoarthritis secondary to progressive severe haemophilic arthropathy. They all had a concomitant haemophilic arthropathy of both ankles. Two patients were wheelchair-bound (Patients 2 and 3); another two patients used one or two crutches (Patients 5 and 7).

Four patients were dependent on analgesics, and two (Patients 1 and 7) had a history of prolonged bleeding induced by acetylsalicylates (Kasper and Rapaport 1972). Patient 6 had had a surgical synovectomy six years before replacement of the right knee and synovectomy by irradiation eight years before the left knee replacement. Patient 7 also had had the latter procedure three years before replacement.

Our principal indication for total knee arthroplasty was persistent, disabling pain for which arthrodesis had become the only rational treatment. Contra-indications were recent infections of the knee, bony ankylosis and the presence of Factor VIII inhibitor. All patients except Patient 1 were seen by us at follow-up, and all radiographs before and after operation were reviewed.

Haematological management. Before surgery, all patients had determinations of ABO-group, Rhesus factor, Factor VIII:C activity, Factor VIII:C inhibitor level, HBs antigen and antibodies. A survival study of Factor VIII was done in several patients.

Factor VIII was administered as cryoprecipitate in six patients; the other two patients were allergic to this so
Factor VIII concentrate was used. It was administered before operation to raise the circulating Factor VIII:C to normal level: the dose was 50 units of Factor VIII per kilogram body weight. Immediately after operation Factor VIII:C determinations were done to control further dosage; the level was maintained above 50% of normal during the first 10 to 14 days and above 20% in the following days and weeks. Twice daily administration of Factor VIII precipitate in the dose of 25 u/kg was necessary for the first three to five days, and thereafter 15 u/kg twice daily. From the fifteenth day Factor VIII once daily was usually sufficient. The average number of vials given was 192 (128 to 326), each containing cryoprecipitate or concentrate from four donors.

Operative procedure. All operations were performed under general anaesthesia in a conventional operating theatre with a vertical downflow system. A tourniquet was used. Prophylactic antibiotics, either fluoroquinolones plus ampicillin or cephalothin, were given at a dose of one gram every six hours, for 48 hours.

The medial parapatellar exposure was used in all cases. In all but one knee (Patient 7) extensive intra-articular fibrosis and sub-total obliteration of the suprapatellar pouch was found with localised chronic synovitis and widespread deposits of haemosiderin. The peri-articular tissues in all knees were taut. A total synovectomy was done in one knee (Patient 7). In another four knees (Patients 1, 2, 3 left and 5) a partial synovectomy was performed.

Depending on the state of preservation of the collateral and/or cruciate ligaments, a resurfacing or a semi-constrained total prosthesis was implanted. A narrow femoral or tibial shaft gave some problems in aligning the stemmed prosthesis, causing slight varus deformity (Patients 2 and 8). Release of the dorsal capsule was necessary in the knees with pre-operative flexion contracture of more than 25°, in order to achieve full extension. All knees had at least 90° of flexion. A retropatellar button was used in nine knees.

During the operative procedure, blood loss was negligible due to the use of the tourniquet. In one knee (Patient 7) release of the tourniquet resulted in considerable blood loss (1500 ml), despite meticulous haemostasis by electrocautery; in this patient a total synovectomy was performed.

The usual postoperative treatment for total knee arthroplasty was followed. If necessary a cane or crutch was used for three months. Seven patients convalesced in a rehabilitation centre for six weeks.

RESULTS

Results were evaluated using the knee-rating score (Table I). The follow-up ranged from two to eight years (average 3 years 9 months).

All patients were pleased with their arthroplasty. Walking ability, without a cane or crutch, was unrestricted in all but two patients, one of whom was able to walk for more than half an hour while the other (Patient 2) could walk indoors. Climbing stairs and getting up out of a chair did not give rise to problems or pain. Gait was improved dramatically; only a slight limp was seen in two patients with bilateral arthroplasty and in one patient who before operation had a stiff knee. None of the patients complained of pain in the knee and none depended on analgesics.

On examination all the knees had healed well; a small effusion was seen in two. Quadriceps strength was good, and the range of movement had improved in all but two knees (Table I). Slight lateralisation of the patella on flexion was seen in two knees. There was no instability. Three patients could participate in some sporting activities, including swimming, cycling, and occasionally table tennis.

Table I. Pre-operative clinical details and the results of total knee replacement in eight patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>L/R</th>
<th>Prosthesis</th>
<th>Pett. score</th>
<th>Range of flexion (degree)</th>
<th>Manipulation</th>
<th>Knee score</th>
<th>F VIII vials</th>
<th>Follow-up</th>
<th>Complications</th>
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<td>Post-op</td>
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<td>Post-op</td>
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<td>+3</td>
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Dad, Dadurian; GSB, Gschwend-Scheier-Bahler; Kin, Kinematic
Patients 3 and 7 had bilateral procedures; patient 8 had a one-stage bilateral procedure
*Petterson score: radiographic assessment of the haemophilic arthropathy (see text)
Full length radiographs showed no signs of loosening of the prostheses. Radiolucent lines were seen in five knees with hinged prostheses, but all the prostheses remained in position. The Dadurian prosthesis showed a number of lucent lines around the bone cement in the distal part of the femoral stem and in the proximal part of the tibial component. Other radiographic features were: some para-articular ossification in two knees, a slight periosteal reaction at the distal femur in two knees and severe haemophilic arthropathy of the contralateral knee (if there was no arthroplasty) and in all ankle joints.

Three patients were able to start full-time or part-time work. Two patients were able to continue their previous work normally, and two could do their work at 50% of normal capacity. One patient was very crippled by other arthropathies and other complications of the haemophilia and remained unemployed.

At follow-up there was a considerable improvement in comparison with the pre-operative score. Five knees were classified as excellent, four as good, one as fair and one as poor (Table I).

Complications
Most of the patients had temperatures above 38°C during the first four to six days after operation. Only one positive blood culture was obtained, which yielded *Staphylococcus aureus*: appropriate antibiotics were given, though there were no signs of infection in this patient.

In Patient 8, after operation, the arterial pulse distal to the knee disappeared at full extension: slight flexion of the knee was sufficient to restore the peripheral circulation. At follow-up full extension was achieved without compromising the circulation.

All patients had difficulties regaining movement from 90° of flexion to full extension. Eight of the 11 knees were manipulated under anaesthesia in the second or third week. Sometimes the procedure was combined with open drainage of a haemarthrosis (Table I). These procedures were done under full cover of Factor VIII.

Prostatic problems in Patient 6 caused urine retention, which was treated initially by an indwelling urethral catheter and later by a suprapubic catheter inserted under local anaesthesia, requiring supplements to bring the Factor VIII up to 60%. A transient haematuria did not present any problems. Urine cultures grew *Actinobacillus anitratus*. Nevertheless, on the advice of the urologist no special therapy was given.

Complications related to haemophilia were:
1. Haemarthrosis (three cases), requiring open drainage in two knees.
2. Prolonged nose bleeding in one patient after aspirin was unfortunately administered after manipulation of the knee. Bleeding started more than 24 hours later at a Factor VIII:C level of 39%, and was treated by a nasal haemostatic catheter device, and e-tranexamine medication.
3. Allergic reaction to cryoprecipitate in two patients, treated by clemastine and triamcinolone medication.

The cryoprecipitate was replaced by Factor VIII concentrate. Fortunately no antibodies to Factor VIII developed, and no hepatitis appeared in the postoperative period.

In one patient recurrent thrombophlebitis occurred at the infusion sites. The Factor VIII supplement had been stopped on the fifth day, resulting in a bleed in the knee two days later. Open drainage of the haemarthrosis was not necessary. Successful prevention of thrombophlebitis was achieved by combining the Factor VIII supplements with appropriate antibiotics (cephalothin). In the long term, occasional bleeding, less than three times a year, was seen in the knees of Patients 7 and 8.

DISCUSSION
Patients suffering a severe haemophilic arthropathy of the knee are usually relatively young men. Nevertheless, the age of the patients was not considered to be a contra-indication. Concomitant joint involvement in the same or the contralateral extremity protects the prosthesis from misuse.

Surgical procedures in haemophilia are considered to be hazardous. Only a few small series of total knee replacements in haemophiliacs have been reported (Post and Telfer 1975; Arnold and Hilgartner 1977; London et al. 1977; Marmor 1977; McCollough et al. 1979; Goldberg et al. 1981). The results are variable and there is a fairly high complication rate, mostly related to the haemophilia. In our series more than the half of the patients had one or more complications; most of these might have been prevented by a more accurate regime of Factor VIII administration. Stopping the Factor VIII therapy a few days after operation, as occurred with Patient 8, resulted in bleeding in the knee; in this case no open drainage was necessary, but the haemarthrosis delayed healing and rehabilitation.

The technical problems of the arthroplasty are formidable (Lachiewicz et al. 1985). The intra- and peri-articular tissues are taut and fibrotic; this may be the reason for nerve palsies (Goldberg et al. 1981) and also for the circulatory problems in Patient 8. In addition, this fibrosis is the basis of the problems in regaining full extension, and flexion. A minimal range of movement (Patients 4 and 5) is not considered a contra-indication, nor is a severe flexion contracture: Patient 2 had a contracture of 50° and Patient 3 had 30° of contracture in both knees. By traction and mobilising exercises these flexion contractures were reduced pre-operatively to 40° and 25° respectively (Table I). The operation may be more difficult with severe contractures, and the results can be expected to be inferior to those with more pre-operative function.

The choice of knee prosthesis, whether semi-constrained or resurfacing, depends on the pre-operative and per-operative findings. With posterior subluxation the normal anatomy is seriously disturbed, and we think
that the semi-constrained is useful in preventing re-subluxation. In securing good alignment and an acceptable range of movement, we prefer to resect more bone rather than performing reconstructive procedures such as tendon lengthening or capsular release. We found no disadvantages in bone resection.

Manipulation of the knee should be avoided. It has undesirable effects on the healing process, causes a painful postoperative period, requires more Factor VIII, and disturbs the psychological process of conquering the pain on exercise. The haemophiliac associates pain with the start of bleeding. He must be convinced that in the postoperative period pain is a quite normal finding and under these circumstances it is not related to bleeding. Otherwise he tends to use large amounts of Factor VIII although there are no objective signs of bleeding (Small et al. 1983).

Despite the problems, we feel that total knee replacement provides an excellent quality of life, without bleeding episodes and without compromising the adjacent and contralateral joints. The haemophiliac is able to regain his former lifestyle without pain, bleeding and restrictions other than the prevention of bleeding into other joints.

The risks of post-transfusion hepatitis, haemolytic anaemia, development of antibodies to Factor VIII and, recently, infection with the AIDS virus increase with the amount of Factor VIII administered. Meticulous screening of blood donors and advanced preparation techniques are the only means of achieving “clean” Factor VIII, as long as no synthetic product is available.

**Conclusion.** Total joint replacement can be a very satisfactory solution to patients with disabling haemophilic arthropathy of the knee. The procedure is not as hazardous as it may appear to be, if performed under the strict supervision of a haematologist in a special centre, and if meticulous attention is paid to pre-operative selection, the technical problems of the operation, all aspects of Factor VIII supplementation, and rehabilitation.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

**REFERENCES**


