Knee arthroplasty in hemophilia

5–12 year follow-up of 15 patients

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Between 1979 and 1987, 15 knee arthroplasties were performed in 15 Norwegians with congenital disorders of blood coagulation. 10 patients with a median follow-up of 7 (5–12) years had an almost painless joint, without hemorrhage. Flexion contractures were corrected, but total range of motion was not improved. There was a radiolucent zone at the bone-cement interphase of the tibial stem in 2 knees. The placement of the implants was correct and the alignment not changed. There were no fractures. 1 prosthesis had been removed because of a chronic infection. 4 patients had died.

We conclude that arthroplasty can be safely performed with excellent relief of pain and improvement of function in patients with congenital disorders of blood coagulation.

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Arthropathy is a well known complication in severe hemophilia and von Willebrand's disease and is related to recurrent hemarthroses in childhood and adolescence. The knee is the most common site of hemophilic arthropathy (Arnold and Hiltgartner 1977). The availability of the coagulation factor concentrates permits reconstructive surgery in patients with chronic hemophilic arthropathy, and knee arthroplasty has been recommended for relief of pain caused by severe arthropathy (Marmor 1977, Goldberg et al. 1981, Lachiewicz et al. 1985, Augereau et al. 1987). We report our experience with total knee arthroplasty in patients with congenital disorders of blood coagulation.

Patients and methods

Patients

Between March 1979 and May 1987, 15 knee arthroplasties were performed in 15 patients (14 men and 1 woman) with hemophilia A and von Willebrand's disease (Table 1). The median age at the time of operation was 30 (25–48) years. Cases 1 and 2 had a synovectomy performed 9 and 3 years prior to the arthroplasty, respectively. All patients had recurrent hemarthroses in the order of 2–4 per month during the preceding year.

Orthopedic surgery in hemophiliacs is performed at one center (Oslo Sanitetsforenings Rheumatism Hospital/Rikshospitalet) in Norway. Approximately 180 Norwegians have severe congenital disorders of blood coagulation, i.e., hemophilia A and B and von Willebrand's disease.

Indications

The indication for total joint replacement was disabling pain in grossly damaged knees. The decision to perform arthroplasty was made from the radiographic findings, adjusted by the findings made by the surgeon during the operation. The radiographic changes were interpreted and graded according to the recommendations of the Orthopedic Advisory Committee of the World Federation of Hemophilia (Pettersson et al. 1980, Greene et al. 1989). Osteoporosis, enlarged epiphysis, irregular subchondral surface, narrowing of joint space, subchondral cyst formation, erosions of joint margins, gross incongruence of articular bone ends, and joint deformity were allotted 0–2 points according to their existence and severity. Zero points correspond to a normal joint, and the maximum score was 13 points, corresponding to the most advanced degree of degeneration. In 3 patients the preoperative radiographic grading was 6, but during the operation the knees were found to be more damaged than expected and arthroplasty was found to be preferable.

Table 1. 15 knee arthroplasties in 15 patients with Hemophilia A and von Willebrand's disease

Case	Age	Disease	Preoperative radiographic joint score	Consumption of Factor VIII (thousand IU)	Time in hospital (days)	Follow-up (months)
1	26	Hem A < 1%	10	55	28	148
2	25	Hem A < 1%	11	39	17	144
3	30	vWD[1]	6	22	93	133
4	30	Hem A < 1%	NA	50	34	90 [3,4]
5	28	Hem A < 1%	NA	39	21	17 [3,5]
6	34	Hem A < 1%	10	43	30	75 [3,4]
7	29	Hem A 2%	6	43	23	31 [3,4]
8	48	Hem A < 1%	NA	51	35	87
9	49	Hem A <1%	13	49	30	85
10	29	Hem A < 1%	10	52	17	84
11	34	Hem A < 1%	10	40	17	72 [4]
12	28	vWD[2]	6	40	30	74
13	30	Hem A < 1%	13	37	31	108
14	34	Hem A <1%	10	37	17	60
15	34	Hem A <1%	10	34	16	59

NA Not available
[1] Bleeding time > 30 minutes, F VIII C: 2%, F VIII Ag: <5%, Ristocetin coF: <5%
[2] Bleeding time > 30 minutes, F VIII C: 1%, F VIII Ag: <5%, Ristocetin coF: <5%
[3] Time from operation to death
[4] AIDS
[5] Suicide

Substitution

Reconstituted lyophilized Factor VIII concentrates were infused before the operation and on the subsequent 10–14 days. Only patients with documented adequate elevation of Factor VIII levels following the preoperative coagulation factor concentrate infusion were accepted for operation. The goal of the substitution was to maintain the level of the deficient coagulation factor above 70 percent of normal during the operation, keep the level above 40 percent during the first postoperative days, and then gradually reduce replacement therapy. The patients also received tranexamic acid. The mean Factor VIII consumption was 42,000 (22,000–55,000) IU in the 15 patients (Table 1).

Surgery

1 Guepar prosthesis, 1 Attenborough prosthesis and 13 Richards Maximum Contact (RMC) prostheses were used. The arthroplasties were performed through a parapatellar medial incision in a bloodless field. The tourniquet time was 90–120 minutes. All the prostheses were fixed by cement impregnated with gentamycin. Operations were performed in an operating theater with vertical laminar airflow. The patients did not receive a prophylactic antibiotic regimen.

The intraoperative loss of blood, together with the loss through Hemovac drainage, was minor and did not require replacement in any patient.

The patients started active exercises on the first postoperative day. Partial weight bearing was permitted after 6–8 days.

Follow-up

The last follow-up examination was made during the spring of 1992. 4 patients were dead—1 had commited suicide, and 3 had died of AIDS. 1 patient had had the prosthesis removed because of infection and an arthrodesis performed. 10 patients with a median follow-up of 7 (5–12) years were subjected to an interview and clinical and radiographic examinations. Pain was evaluated by a visual analog scale and given a score from 0 to 10, from no pain to very severe pain. Measurements of range of motion were carried out.

Results

Complications. Case 8 developed high-titered antibodies to Factor VIII within 7 days of the operation, contracted a major wound hemorrhage and was transfused with 6 units of packed red cells. Previously, joint hemorrhages in this patient had been treated successfully with Factor VIII concentrates, and the preoperative elevation of the Factor VIII level was as expected. Infusion of highly purified porcine Factor VIII C (Hyate C—Speywood Laboratories Ltd, UK) con-

trolled the bleeding. Case 9 developed a chronic infection with fistulae, antibiotic treatment was unsuccessful. The implant, an Attenborough prosthesis, was removed and an arthrodesis was performed 2 years after the index operation.

Pain and hemorrhage. Chronic joint pain was considerably reduced in all patients and completely relieved in 8. The arthroplasty had eliminated hemorrhage in the operated joints. 2 additional patients (Cases 4 and 6), who later died of AIDS, were evaluated in 1987 after a follow-up of 4 and 3 years, respectively. They were both completely pain-free.

Range of motion. Before surgery, the range of motion was considerably impaired. All patients had a flexion contracture, mean 23° (5–35°), impaired maximal flexion, mean 77° (40°–110°), and impaired total range of motion, mean 54°. With arthroplasty, extension deficiency was corrected, maximal flexion was reduced, mean 50° (30°–90°), and the total range of motion was unchanged, mean 50°. Case 13 had been reoperated after 24 months with release of the quadriceps muscle and improvement of maximal flexion.

Function. All of the patients had a normal level of activity on follow-up. 9 patients were either full-time employed or full-time students. 1 patient was unemployed, due to advanced HIV infection. All patients achieved full, unprotected weight bearing, and they were able to walk without crutches or orthoses. 2 patients (Cases 4 and 6), who died 9 and 6 years after the arthroplasty, respectively, never complained about their knees.

Radiographic evaluation. At follow-up, all 10 patients had their implanted joints radiographed. In one knee with a RMC prosthesis, a radiolucent zone of 1–2 mm at the bone-cement interphase of the tibial stem was demonstrated, and in another with a Guepar prosthesis, a radiolucent zone of 1 mm was found. These changes were not present in 1987. None of these patients complained of pain. In all 10 knees there was correct placement of both components of the prosthesis, with no change of alignment, compared to radiographs taken within the first week postoperatively. There were no fractures. Furthermore, Cases 4 and 6, who later died, were available for evaluation in 1987. Neither radiolucency nor change of alignment was detected in these 2 patients.

Discussion

The availability of coagulation factor concentrates has made arthroplasty feasible in patients with congenital disorders of blood coagulation. In this series, pain was considerably reduced in all patients and completely relieved in 8 patients. Joint-related hemorrhage was abolished. This is in agreement with previous reports (McCollough et al. 1979, Goldberg et al. 1981, Lachiewicz et al. 1985, Augereau et al. 1987). Thus, knee arthroplasty offers excellent relief of pain. On the other hand, no improvement in range of motion was demonstrated in our patients. Our experience is not unique (McCollough et al. 1979, Goldberg et al. 1981, Beeton et al. 1992). Encouraging, however, was the finding of correction of the preoperative flexion contracture in all patients. Together with relief of pain, this explains why the patients were very pleased with the result of the arthroplasty. Goldberg et al. (1981) speculated that the extensive involvement of periarticular soft tissue, recurrent intramuscular bleeding and residual weakness about the knee, as well as marked distortion of bone may explain the lack of improvement of range of motion in hemophiliacs, in contrast to the improvement usually seen in patients with rheumatoid arthritis and arthrosis after arthroplasty (Insall et al. 1979, Goldberg et al. 1981, Laskin 1981).

A radiolucent zone at the bone-cement interphase was detected in 2 knees. This is less than that reported by others, even in studies with a shorter follow-up time (Lachiewicz et al. 1985, Figgie et al. 1989) and less than that reported in knee arthroplasty for arthrosis (Insall et al. 1983).

Some degree of bone stock impairment is found in most of the patients accepted for arthroplasty in our center. This is the reason why we still use, though not exclusively, cemented prostheses. At present the Tricon II C is utilized.

It seems fair to conclude that total knee arthroplasty can be safely performed, with relief of most symptoms of disabling arthropathy in patients with congenital disorders of blood coagulation. A prerequisite for an optimal result is the close cooperation between orthopedic surgeons and hematologists experienced with this group of patients and adequate facilities for monitoring the substitution therapy. However, it should be emphasized that, despite severe destructive arthropathy, many hemophiliacs continue to function quite well, and total knee replacement is primarily indicated when the pain is disabling. We do not recommend that arthroplasty be performed with the sole objective of improving the range of motion in the knee.

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