Total hip replacement in patients with hemophilia

13 hips in 11 patients followed for 1–16 years

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During 1973–88, we performed 13 total hip replacements in 11 hemophilia patients, mean age 46 (25–65) years. During the operation, blood loss averaged 920 mL, and a mean of 120,000 units of factor VIII/IX were used. The mean duration of follow-up was 7 (1–16) years. 5 hips became loose within 6 years, and a further one after 13 years. 4 hips were revised, 2 of them due to infection in patients who were also seropositive for HIV. At the latest follow-up, 10 patients were alive. 6 had no hip pain and 7 could walk at least 1,000 meters at a time. Although these results are inferior to those obtained in arthrosis, total hip replacement should be considered in hemophiliac patients.

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In 1975, Ahlberg predicted that the near future would bring a broadening of indications for joint arthroplasty in hemophiliacs, but also expressed hope that such operations would become less and less common as a result of prophylactic measures. On the one hand, the development of factor VIII/IX substitution therapy has made it possible to perform even major surgical treatment—e.g., total hip replacement (THR)—in patients with hemophilia, but on the other hand, due to a prophylactic factor regime, the actual need for orthopedic surgical procedures has decreased over the past decades (Löfqvist et al. 1996). With reference to THR, only a few reports are available with varying results. Two recent reviews (Nelson et al. 1992, Kelley et al. 1995) showed that about half of the THRs were loose at an average follow-up of 8 years.

To compare our results with those of others and as to find out whether postoperative prophylactic factor VIII/IX treatment is of value, we present the outcome of hip joint replacements performed in hemophiliacs at the University Hospital, Malmö, Sweden, during a 16-year period.

Patients and methods

During 1973–88, 13 THRs were performed in 11 hemophilia patients; 9 with hemophilia A (6 severe, 1 moderate, 2 mild) and 2 with severe hemophilia B (Table 1). The mean age at surgery was 46 (25–65) years, 7 patients being under 50. 9 of the patients also had disabling arthropathy involving one or both knees and/or ankles. 1 patient was known to be seropositive for HIV at the time of primary surgery. 3 of the patients were found to be HIV-positive retrospectively. In patient 9, inhibitory antibodies to factor VIII were present and in 1985 this patient underwent tolerance induction according to the Malmö model, which resulted in disappearance of the antibodies and permitted surgery (Nilsson et al. 1986, 1988). The same patient also had hypertension and an aortic aneurysm. Patient 6 was referred to Malmö because of his inhibitor status. The patient had ongoing treatment with tolerance induction that made THR surgery possible.

Surgery

The indication for surgery was disabling pain due to advanced hemophilic arthropathy in all patients, except one who was referred to us for pseudarthrosis of a femoral neck fracture, not treated due to the presence of inhibitory antibodies.

The surgical technique was the same as for other patients; cemented Charnley prostheses were inserted, using the transtrochanteric approach. All patients were treated by senior orthopedic surgeons having special experience of problems related to hemophilia.

In the 1970s, in order to avoid postoperative bleeding complications, the patients were not mobilized as early as non-hemophiliacs. In the 1980s, mobilization routines were gradually brought into line with those for non-hemophiliacs. After 6–12 weeks with crutches, full weight bearing was allowed.

Table 1. THR 1973-88 in 11 patients with hemophilia

Patient no.	Age (years)	Year of operation	Hemophilia	Total factor requirement (units)	Peroperative bleeding (L)	Continuous postoperative prophylaxis	Radiographic loosening at follow-up (months)	Addenda
1	25	1976	A severe	144,000	2.9	No	Yes (53/66)	Bilateral THR simultaneously. Bilateral cup-loosening after 53 months. Bilateral stem-loosening after 66 months. Death 1986, AIDS.
2	26	1982	A severe	68,400	0.6	Yes	No (99)	Infection. Septic arthritis in ankle and hip. THR revised after 99 months.
3	34	1973	A severe	63,200	0.4	No	Yes (64)	Cup and stem loosening. Revised after 140 months. Death 1990, AIDS.
4	43	1975	B severe	82,000	0.5	Yes	Yes (162)	Cup loosening.
5	44	1980	A severe	89,000	1.1	Yes	No (126)	
6	46	1985	B severe inhibitor	375,000	1.8			Tolerance induction preoperatively. Lost to follow-up.
7	49	1978	A severe	52,000	0.5	No	No (142)	Infection. Cup and stem loosening. Revised after 153 months.
8	55	1985	A mild	102,000	0.8	No	No (12)	Non-A, non-B hepatitis
	55	1985		110,000	0.7	No	No (17)	postoperatively. Death 1988, esophageal cancer.
9	56	1986	A severe	228,000	0.4	Yes	No (25)	Death 1989, cerebrovascular hemorrhage.
10	56	1979	A mild	61,500	0.7	No	Yes (72)	Cup loosening. Revised after 116 months.
11	65	1988	A moderate	59,000	0.6	Yes	Yes (38)	Cup loosening. Death 1992, AIDS.

Substitution

Patients with *severe* and *moderate* hemophilia A were given factor VIII (50–70 units/kg body weight) immediately before surgery. The dose was repeated peroperatively when surgery lasted for more than 1.5 hours or if the plasma concentration fell below 40% of normal. For 2–7 days postoperatively, the factor concentration was maintained at about 40–50% by repeated infusions at 4–8 hour intervals. Up to 2–4 weeks after surgery, the factor concentration was kept above 10–20% by infusions every 12 hours (Nilsson et al. 1987).

2 patients with *mild* hemophilia A were given factor concentrates in reduced doses adjusted according to the baseline level of factor VIII. Postoperative treatment continued for about 3 weeks.

In patients with hemophilia B, factor IX was given at higher initial doses, but at intervals longer than those used in the administration of factor VIII.

To suppress local fibrinolysis, the patients were also given tranexamic acid (Cyklokapron, Pharmacia, Stockholm, Sweden) at 6–8 hour intervals for about 7 days (Nilsson 1975).

4 patients with severe and 1 with moderate hemophilia have been given regular prophylactic factor treatment at average dosages of 40 units/kg body weight twice weekly after surgery.

Follow-up

Of the 13 THRs, 12 were reviewed radiographically and clinically with a mean follow-up of 7 (1–16) years. 1 patient (6) from Tunisia returned there after the operation and was lost to follow-up. During the follow-up, 4 patients were diagnosed as HIV seropositive. To date, 5 patients have died: 3 of AIDS, 1 of cerebral vascular hemorrhage, and 1 of esophageal cancer (Table 1).

Serial radiograms were studied for signs of loosening. A change in position of the socket or the femoral component or a progressive radiolucent zone after the 1-year follow-up was considered as a radiographical failure (Carlsson and Gentz 1984).

Results

Complications

1 patient (8) contracted non-A, non-B hepatitis postoperatively because of the treatment during surgery. No other serious complications occurred.

Hemorrhage and substitution (Table 1)

The mean total blood loss was 0.9 (0.4–2.9) L. Patient 1 underwent simultaneous bilateral hip replacement, which explains the excessive blood loss of 2.9 L. The mean factor requirement per THR was 120,000 (52,000–375,000) units of factor VIII/IX.

Function

All patients obtained early pain relief and an increase in hip motion. At the latest clinical follow-up, 6 patients (cases 2, 4, 7, 8, 9, 11) had no pain in the operated hip, 2 (cases 3, 10) had pain on walking, and 2 (cases 1, 5) pain at rest and walking. 3 (cases 5, 7, 11) of the patients needed crutches, partly because of multiple arthropathy. 7 (cases 2, 4, 7–11) patients could walk at least 1,000 meters at a time, whereas 3 (cases 1, 3, 5) patients had markedly reduced walking ability, due partly to pain in the hip operated on and partly to multiple arthropathy. At follow-up, full extension was recorded in all hips, except one where the reduction in extension was 30°. The flexion was, on average, 85° (70°–100°), abduction 30° (20°–40°), internal rotation 15° (10°–20°) and external rotation 25° (15°–40°).

Radiographic evaluation

1 patient was lost to follow-up. 2 patients (cases 8, 9) died within a few years postoperatively and radiograms of their 3 hips showed no sign of loosening after a follow-up of 1–5 years. Review of serial radiograms of the remaining 9 hips revealed loosening of the acetabular component in 6 cases after a mean of 6 (3–14) years. 3 were still well fixed after a mean interval of 10 (8–12) years. 3 of 9 femoral stems were radiographically loose after 6 years, but the remaining 6 were well fixed after 10 (3–16) years.

Of the 7 THRs performed in patients under 50 years of age, 3 had loosening within a mean of 5 years.

Revision surgery

4 hips were revised at a mean of 10 (8-12) years. 2 hips (cases 3, 10) were revised because of aseptic loosening and the other 2 revisions (cases 2, 7) were due to infection, both in HIV-positive patients. One of the infected hips was revised twice.

Prophylactic factor VIII/IX treatment/radiographic evaluation

Of 8 THRs in patients with prophylaxis with factor VIII/IX after the operation, 3 were radiographically loose after 3, 6 and 14 years. Of 4 THRs in patients without prophylaxis, 3 were loose after 4.5–5.5 years (Table 1).

Discussion

In hemophilia, arthropathy of the hip is less common than in the knee, elbow or ankles. Only a few reports have described THR in hemophilia. Augereau and coworkers (1987) reported good results in 10/13 hips over a 4-year period. Luck and Kasper (1989) reviewed 9 cemented THRs; 5 of them had to be revised, and 2 of these revisions were done because of infection. Nelson and coworkers (1992) reviewed 21 patients with 22 THRs; at a median follow-up of 8 years, 5 hips had been revised and another 3 manifested signs of impending failure. Kelley and coworkers recently (1995) presented a retrospective multicenter study of 27 patients with 34 THRs. At a mean follow-up of 8 years, 17/26 cemented acetabular components and 12/27 cemented femoral components had radiographic evidence of failure. The infection rate was 3/27. 10 THRs were revised. Our results concerning THR in hemophiliacs are comparable to those mentioned above. Some other groups have reported small series from which no conclusions can be drawn (Post and Telfer 1975, Arnold and Hilgartner 1977, Rana et al. 1986).

The fact that the plasma concentration of factor VIII/IX in mild hemophilia without prophylactic treatment in general is sufficient to prevent bleedings, makes these patients comparable to severe/moderate hemophiliacs with prophylactic treatment. For this reason mild hemophiliacs were included among the 8 THRs with postoperative prophylaxis in our series. If the 2 with shortest follow-up, less than 2 years, are excluded, the remainder had a loosening rate of 3/6. Of 4 THRs in patients without prophylaxis, mild hemophiliacs excluded, 3 were loose at follow-up. Theoretically, factor prophylaxis should reduce early and late microbleedings at the cement/bone interface. Although there seems to be some difference between the 2 groups of patients as regards radiographic loosening, the groups are too small to draw any statistical conclusions from. Until the opposite is proven, however, we still believe that postoperative prophylaxis should be recommended to patients with THRs. Further research will hopefully bring clarity to this question.

The average requirement of factor VIII/IX was somewhat lower in the 1970s than in the 1980s. Today the approximate requirement per THR is roughly 100,000 units of factor concentrate.

Postoperative mobilization is possible with intensive factor replacement therapy during the first postoperative weeks. In contrast, Nelson and coworkers (1992), who used a postoperative spica cast during the wound-healing period, gave only about half of the

amount of factor treatment compared to our praxis.

2 THRs became infected, and both patients were HIV-positive. They had no radiographic signs of loosening and the hips functioned well. The risk of infection in THRs in hemophiliacs with HIV appears higher than usual (Luck and Kasper 1989, Greene et al. 1990, Kelley et al. 1995).

We conclude that hemophilia patients can expect a fairly high frequency of aseptic loosening after THR. HIV positive patients also seem to have an increased infection rate. However, according to our findings THR is of value in some hemophiliacs.

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References

Ahlberg Å. New horizons in reconstructive joint surgery. Recent advances in hemophilia (Ed. Aledort L M). Ann N Y Acad Sci 1975; 240: 291-4.

Arnold W, Hilgartner M. Hemophilic arthropathy. J Bone Joint Surg (Am) 1977; 59 (3): 287-305.

Augereau B, Travers V, Le Balch T, Witvoët J. Les arthroplasties totales de hanche et de genou chez l'hémophile. À propos de 27 cas. Rev Chir Orth 1987; 73: 381-94.

Carlsson Å S, Gentz C F. Radiographic versus clinical loosening of the acetabular component in non-infected total hip arthroplasty. Clin Orthop 1984; 185: 145-50.

Greene W, De Gnore L, White G. Orthopaedic procedures and prognosis in hemophilic patients who are seropositive for human immunodeficiency virus. J Bone Joint Surg (Am) 1990; 72 (1): 2-11.

Kelley S S, Lachiewicz P F, Gilbert M S, Bolander M E, Jankiewicz J J. Hip arthroplasty in hemophilic arthropathy. J Bone Joint Surg (Am) 1995; 77 (6): 828-34.

Löfqvist T, Nilsson I M, Petersson C. Orthopedic surgery in hemophilia—20 years' experience in Sweden. Clin Orthop 1996 (in press).

Luck J V, Kasper R. Surgical management of advanced hemophilic arthropathy. Clin Orthop 1989; 242: 60-82.

Nelson I, Sivamurugan S, Latham P, Matthews J, Bulstrode C. Total hip arthroplasty for hemophilic arthropathy. Clin Orthop 1992; 276: 210-3.

Nilsson I M. Local fibrinolysis as a mechanism for haemorrhage. Thromb Diathes Haemorrh 1975; 34: 623-33.

Nilsson I M, Berntorp E, Zettervall O. Induction of split tolerance and clinical cure in high-responding hemophiliacs with factor IX antibodies. Proc Natl Acad Sci USA 1986; 83: 9169-73.

Nilsson I M, Larsson S A, Bergentz S E. The use of blood components in the treatment of congenital coagulation disorders. Wld J Surg 1987; 11: 14-9.

Nilsson I M, Berntorp E, Zettervall O. Induction of immune tolerance in patients with hemophilia and antibodies to factor VIII by combined treatment with intravenous IgG, cyclophosphamide, and factor VIII. N Engl J Med 1988; 318: 947-50.

Post M, Telfer M C. Surgery in hemophilic patients. J Bone Joint Surg (Am) 1975; 57 (8): 1136-45.

Rana N A, Shapiro G R, Green D. Long-term follow-up of prosthetic joint replacement in hemophilia. Am J Hematol 1986; 23: 329-37.