Total Hip Replacement in Patients with Severe Bleeding Disorders
A 30 Years Single Center Experience

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Introduction

Hemophilic arthropathy of the hip is less common in patients with bleeding disorders.

If the destruction of the hip joint is severe enough replacement of the joint is indicated.

Material and Methods

We performed a retrospective analysis of the cases done at our center to determine the outcome of hip replacements in this specific group of patients.

In July 1972 the first elective total hip replacement (THR) in a hemophilic patient was performed at the University Hospital in Frankfurt. Between then and 2002 in 13 patients 15 hips were found to be necessary to replace. In 10 hemophilic patients and 3 VWD patients a hip replacement was performed. The mean age of the patients was at the time of surgery 42 years. During that time we changed the implants used for the replacement. At the beginning we used only cemented cups and stems. Later hybrid THR was use with a cemented stem and a cementless cup. Recently we use only cementless implants even in those high risk patients.

The charts of the patients were evaluated for complications during surgery, early complications after surgery, survival of the implants, cases of revision surgery, and infections. To determine the functional outcome the widely accepted Harris Hip Score was calculated for the time before surgery and at the time of follow up. The earliest follow-up was 12 months after surgery.

Results

Good long term results after a follow up time of 132 months (12–363) were found. No perioperative complications like bleeding or infection were found. Only one aseptic loosening of a cemented cup occurred after 14 years as well as one septic loosening 14 months postoperatively in a HIV positive patient. Another HIV positive patient developed a hematogenic abscess on both operated hips without loosening.

The calculated survival time after a mean follow up of 11 years was 86.7%.

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The Harris Hip Score increased remarkably from 48 points (range 32 – 66) preoperatively to 89 points (range 76 – 100) at the time of follow up. We found excellent and good results in 72%, fair results in 19%, and bad results in 9% of the cases.

Discussion

End-stage hemophilic arthropathy is further complicated by arthrofibrosis and loss of motion as the hypertrophic synovium is replaced by dense fibrous scar tissue. Arthropathy of the hip is moderate in frequency but is less common than ankle, knee or elbow arthropathy. It has two modes of onset. In childhood, rapidly progressive severe arthropathy may result from a single hemarthrosis because of increased intracapsular pressure leading to osteonecrosis of the capital femoral epiphysis [1]. More often, hip arthropathy is the result of chronic synovitis similar to that which occurs in other joints. Between the second and fourth decades many hemophilic patients develop severe articular destruction. At this stage, possible treatments include arthrodesis and arthroplasty. For the hip, total hip arthroplasty remains the best solution [2].

Teitelbaum [3] reviewed pelvic radiographs of 34 patients complaining of pain in the hip (64 hips) from a population of 175 patients seen at one hemophilia centre. Sixteen (80%) of 20 hips that had an open proximal femoralepiohysis had a valgus deformity, but none had osteoarthritic changes. Fifteen (31%) of 48 skeletally mature hips had degenerative changes, including protusion acetabuli in eight hips. Thus, end-stage hemophilic arthropathy necessitating arthroplasty is infrequent in the hip.

There are few reports of the results of total hip arthroplasty in patients with hemophilia. The indication for a total hip arthroplasty in young patients who have hemophilic arthropathy of the hip should be severe disabling pain occurring both during activity and at rest that is unresponsive to non-operative treatment.

The first important series was published by Luck and Kasper of the Orthopedic Hospital, Los Angeles, USA in 1989 [4], announcing that the first prosthetic arthroplasty of the hip in a hemophiliac in the United States was a cup arthroplasty performed by J. Vernon Luck in 1968. Luck and Kasper reported that between 1968 and 1982, three more cup arthroplasties and ten primary, cemented hip arthroplasties were performed at the Orthopedic Hospital by six different surgeons. Prostheses varied with surgeon and include Müller (five cases), Ausfranc-Turner (two cases), Charnley (one case), Harris (one case) and Wagner resurfacing (one case). Since 1982, two cementless primary hip arthroplasties had been performed.

Eight patients had required revision, and one required an excision arthroplasty. The revisions included three cup arthroplasties because of pain and bleeding, three cemented total hips for aseptic loosening, two cemented total hips for infection, and one Wagner resurfacing for a fracture of the femoral head sustained in a fall 5 years postoperatively. One patient had a Girdlestone resection arthroplasty for a pseudomonas aeruginosa infection. Thus, five of nine conventional, cemented total hip arthroplasties had failed over the last 16 years. Including the three cup arthroplasties, eight revisions had been performed. Seven of these patients were symptom-free. One patient developed Serratia marcescens infection 1 year after revision. In

another of the revision case, there was radiographic evidence of component loosening. This cup arthroplasty was revised to a cemented Charnley low-friction prosthesis in 1972. The femoral component loosened at 3 years and subsided, but it had stabilized and remained asymptomatic over the last 12 years.

The long-term experience of Luck and Kasper with various types of hip prostheses can be rated as only fair, with a revision rate of about 60% over 20 years. However, the clinical status of all patients was quite satisfactory in that all were free from hip pain and were capable of unlimited, unassisted, community ambulation. They were substantially better than they had been prior to their initial hip surgery. End-stage hip disease in hemophilic patients poses an unsolved problem. Primary cemented prostheses have a 33% aseptic failure rate 5-14 years after operation, which is higher than would be expected in a comparable group of patients with another form of polyarthritis. One reason for the loosening of cemented hip prostheses in hemophilic patients may be the increased stresses of a stiff-legged gait.

In 1992, Nelson et al. [5] reported the experience of the Nuffield Orthopedic Centre, Oxford, UK. From 1969 until 1985, 39 total hip arthroplasties were performed in 38 patients for hemophilic arthropathy. The median age of the patients at operation was 48 years. In 21 patients, 22 hip replacements were reviewed clinically and radiographically, with a median follow-up of 7.6 years. Five of the 22 hips had been revised and three were likely to require revision in the near future. The incidence of revision was compared to other studies of total hip arthroplasty in young patients and the influence of HIV infection was examined. Total hip arthroplasty was believed to be an appropriate operation for disabling hemophilic arthropathy.

As hemophilic arthropathy infrequently affects the hip joint, Kelley et al. [6] in 1995 reported a multicenter retrospective study to determine the results of hip arthroplasty in hemophilic patients. Thirty-four hip arthroplasties were performed in 27 male patients at four major hemophilia centres between October 1972 and September 1990 in the United States.

The mean age of patients at the time of operation was 38 years (range 15-73 years). Four patients were seropositive for HIV at the time of the operation, and 16 patients were seropositive at the latest follow-up examination. Nine patients (33%) died before the latest review, seven of whom had been seropositive for HIV. The mean duration of follow-up was 8 years, with a minimum of 2 years for all patients who were still alive at the latest review.

Surgery was carried out as follows: 26 total hip arthroplasties performed with cement, six total hip arthroplasties performed without cement, one total hip arthroplasty in which the femoral component was inserted with cement and the acetabular component was inserted without it (so-called hybrid arthroplasty), and one bipolar arthroplasty performed with cement. There were no early infections after these 34 primary arthroplasties. Three late infections occurred around prostheses inserted with cement, all of which led to a resection arthroplasty. Six (21%) of the 28 cemented femoral components and six (23%) of the 26 cemented acetabular components were revised because of aseptic loosening.

Of the 24 cemented femoral components for which radiographs were available and which were still in place at the time of latest review or at the time of death, ten were definitely loose, two were probably loose, five were possibly loose and seven had

no evidence of loosening. Of the 23 cemented acetabular components for which radiographs were available and which were still in place at the time of review, ten were definitely loose, seven were probably loose, three were possibly loose, and three were not loose. None of the cementless prostheses was loose. There was a high rate of loosening of the cemented hip prosthesis in this series. There was also a high rate of mortality overall, and a high rate of late deep infection in the patients who were seropositive for HIV. Kelley et al. advised caution when a total hip arthroplasty is considered for a patient with hemophilia. Despite the aforementioned complications, Kelly et al. stated that total hip arthroplasty has a continuing role in the treatment of hemophilic arthropathy in patients who have severe pain and disability.

During 1973-88, at the University Hospital, Malmö, Sweden, 13 total hip replacements were performed in 11 hemophilic patients with a mean age of 46 years. According to Löfqvist et al. [7], the indication for surgery was disabling pain due to advanced hemophilic arthropathy. The surgical technique was the same as for other patients: cemented Charnley prostheses were inserted, using the transtrochanteric approach. The mean duration of follow-up was 7 years. Five hips became loose within 6 years, and a further one after 13 years. Four hips were revised, two of them due to infection in patients who were also seropositive for HIV. At the latest follow-up, ten patients were alive, six had no hip pain, and seven could walk a distance of at least 1000 meters. Although these results were inferior to those obtained in arthrosis, total hip replacement should be considered in patients with hemophilia. Löfqvist et al. concluded that this group of patients can expect a fairly high frequency of aseptic loosening after total hip replacement. HIV-positive patients also seem to have an increased infection rate. However, according to their findings, they concluded that total hip replacement is of value in some hemophilic individuals.

In 1998 Heeg et al. [8] evaluated the long-term results of three total hip arthroplasties. One hip was revised after 9 years for aseptic loosening.

During 1976-99, six total hip arthroplasties were performed in six hemophilic patients at La Paz University Hospital, Madrid, Spain. The indication for total hip arthroplasty in people with hemophilia was severe disabling pain, both during activity and at rest, that was unresponsive to non-operative treatment. The mean age of the patients was 42 years (range 35-47). Four Harris-Galante hybrid prostheses (acetabular component without cement and a precoated femoral component with cement), one uncemented isoelastic prosthesis and one cemented Charnley arthroplasty (for an ankylosed hip; see next section in this chapter) were used in these procedures. At the time of the index hip arthroplasty, no patient was known to be seropositive for HIV. The mean duration of follow-up for all the patients was 7 years (range 1-15 years). One patient (the one with the isoelastic prosthesis) died before the time of this review, and five were alive. So far, both clinical and radiographic results are satisfactory.

Studies from several hemophilia centres suggest that between 33% and 92% of patients with hemophilia B, carry the HIV antibody [9]. In two studies of hip arthroplasty for hemophilic arthropathy with more than 20 patients and more than 5 years follow-up, approximately 50% of the patients were known to be seropositive for HIV, contributing to an overall mortality rate at median 7-year follow-up of 20% to 33% [5,6]. Patients with CD4 levels of greater than 500 cells/mm^3, a positive re-

action with energy testing to intradermal skin antigens, platelet count greater than 60 000, total leukocyte count greater than 1000, serum albumin greater than 25g/l, and no history of opportunistic infections of neoplasm have a postoperative complication risk similar to the general population.

In addition to thorough preoperative medical preparation of the patient, considerable surgical preparation is also required. Depending on the age at which significant bleeding began, the proximal anatomy of the femur can be distorted and, in the most severe cases, there can be an extremely small femoral medullary canal, valgus and excessive anteversion of the head and neck, and protrusion acetabuli [10].

Conclusion

Despite FVIII prophylaxis and better joints in hemophilic patients this group of patients will get older and arthritis of the hip is an increasing problem in the older generation. Regardless hemophilic arthropathy hemophilic patients will develop hip and knee arthritis and the need for surgery will increase.

We conclude after more than 30 years of experience with THR in hemophilic patients and based on our positive results that for advanced severe hemophilic hip arthropathy, THR results in pain-relief and an improvement in quality of life. Thus, total hip replacement should be considered regardless the age of the patient and the surgical intervention should be performed in specialized centers with experience in the treatment of bleeding disorders.

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