brief report

Revision hip arthroplasty in sickle cell disease

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Sickle cell disease (SCD) is a hemoglobinopathy causing osteonecrosis and collapse of the femoral head that results in secondary osteoarthritis.[1] Osteonecrosis is noted in 20% to 50% of patients and bilateral involvement is commonly encountered.[2,3] Vasoocclusive crisis results in osteoarthritis; total hip arthroplasty (THA) may be the only solution for these patients. However, it should be remembered that these patients are vulnerable to a variety of serious complications, such as acute chest syndrome, vasoocclusive crisis, perioperative and postoperative blood loss, wound abscess, and complications of transfusion associated with surgery.[3] There is also a tendency to early infection and aseptic loosening with arthroplasty procedures.[4-9] For the reasons mentioned, we rarely recommend THA to patients with SCD. However, we perform hip replacement quite often as the disease is prevalent in the territory. Most reports in the literature describe high complication rates with THA in patients with SCD,[3-5,7,9-11] but limited data are available about revision procedures in SCD. With these points in mind, we retrospectively reviewed the results of revision procedures of THA in patients with SCD.

PATIENTS AND METHODS

Ten patients with SCD underwent revision THA between 1988 and 1999. Eight patients exhibited hemoglobin SCD, 1 exhibited hemoglobin SS disease and 1 patient had hemoglobin S-thalassemia. Five patients were male, 5 were female and the average age was 35 years (range, 18-62 years). Involvement was right sided in 5, left sided in 4 and bilateral in one case. Revision was done on a single hip in 9 patients while one patient underwent revision of both hips. Eight cases were hybrid arthroplasties and 3 cases were cemented arthroplasties. The reason for revision was wearing of an insert in 4 cases (36.3%), symptomatic loosening of the femoral stem in 3 (27.3%), loosening of the acetabular cup in 2 (18.2%) and infection in 2 patients (18.2%). A diagnosis of infection was confirmed by clinical findings, positive aspiration culture and the presence of purulent fluid during exploration. Revisions due to infection were two-stage procedures. Implant removal and debridement with implantation of an antibiotic loaded acrylic spacer was done at the first stage. Reimplantation was done after an average interval of 13 weeks (range, 11-15 weeks) when the serologic markers of infection returned to normal limits. Appropriate antibiotics were administered parenterally for 6 weeks and antimicrobial chemotherapy continued with oral antibiotics for an additional 6 weeks under supervision of the department of infectious diseases.

Care was taken to keep the preoperative hemoglobin values between 9 and 11 g/dL and the hemoglobin S value under 30%. One gram of cefazolin was introduced preoperatively and continued twice a day for 48 hours for prophylaxis. The patients were kept well hydrated preoperatively. An oximetric central catheter was placed as it would be hard to find a venous portal during the procedure. Oversedation was avoided as it might result in hypoxemia due to respiratory inhibition. All procedures were done under general anesthesia in the lateral decubitus position. The revisions were done through an anterolateral approach in 3 cases and a posterior approach in 4 cases. Standard trochanteric osteotomy and an extended lateral trochanteric osteotomy was done for 2 cases each. The operative field was irrigated with diluted vancomycin solution throughout the procedure. Revision prosthesis were hybrid (cemented femoral component) in 4 cases, cemented in 2 cases and cementless in 5 cases. Sufficient analgesia and hydration was provided and the patients were kept warm postoperatively. Supportive oxygen was given for the first 12 to 48 hours. All patients were allowed to walk on the postoperative second day while weight bearing was avoided until 6 weeks in patients who underwent cementless revision. The patients were followed-up at 6, 12, 24 weeks and at 1-year intervals after the first year of the operation. Standard anterior-posterior and lateral radiographs were taken at each visit. Follow-up

time averaged 7 years (range, 5-16 years). The patients were evaluated clinically according to Merle D'Aubigné ve Postel hip scores.[12] De Lee and Charnley acetabular zones[13] and Gruen zones[14] in the femur were evaluated in the radiologic assessment.

RESULTS

Postoperative hospital stays averaged 16 days (range, 6-42 days). Average blood loss was 1420 mL (range, 600-2850 mL) and patients received at least two units and up to eight units of transfusion. Operative time averaged 187±95 minutes. Merle D'Aubigné and Postel pain, range of motion and walking scores improved from 4.2/3.5/4.2 to 5.6/5.2/5.2, respectively. Radiologic assessment revealed progressive radiolucent lines of 2 mm in zones 2 and 3 of a cemented acetabular cup and revision was advised to the patient. A nonprogressive 1-mm radiolucent line in zones 2 and 3 was observed in the acetabular component of another cemented revision case. The femoral stem remained stable in both patients at 6 years follow-up. Femoral osteolysis occured in 4 patients at an average 5 years follow-up. Osteolysis was progressive in two cases.

Five of 10 revision cases required further revision procedures. The reason for re-revision were mechanical loosening in 3 cases and septic loosening in 2 cases. All components that exhibited loosening were cemented. One patient with acetabular loosening refused further revision. The average time from index surgery to primary revision procedures was 84 months (range, 9-132 months). The time for further revision surgery after first revision averaged 50 months (range, 20-82 months).

The most common complications encountered were hematoma formation (36.4%), wound drainage (27.3%) and infection (27.3%)(Table 1). Two patients with wound drainage responded well to local wound care. One superficial and two deep infections were observed. Superficial infection was detected in the early postoperative period in one patient. The isolated microorganism was *Staphlyococcus aureus*. Infection control was achieved by debridement of the wound and antibiotic therapy. One of the deep infections was caused by polymicrobial microorganisms, while the pathogen could not be isolated in the other patient. Both patients were treated by two-stage revision using antibiotic loaded cement spacers for the interval period. The microorganisms isolated were *Acinetobacter baumaniii* and *Klebsiella oxytoca* in the first stage of the revision. The patient developed progressive radiolucent lines through both components during follow-up. At 20 months postoperatively he underwent a second two-stage revision because of reinfection. This time the isolated microorganism was *Citrobacter freundii* which led us to consider a de novo infection (Figure 1, 2). Two patients developed heterotopic ossification. Vasoocclusive crisis was observed in two patients. The results were unsatisfactory because of complications requiring revision or obstacles that did not require operative intervention in 7 patients (63.6%).

Table 1. Complications in 10 cases of revision hip arthroplasty.

Complications	No.	%
Hematoma formation	4	36.4
Wound drainage	3	27.3
Vasooclusive crisis	2	18.2
Heterotopic ossification	2	18.2
Deep wound infection	2	18.2
Superficial wound infection	1	9.1

DISCUSSION

THA is effective for pain management in SCD, but has high rates of complications.[3,4,6,7,10,15-17] Complications like acute chest syndrome and vasoocclusive crisis are frequently encountered with arthroplasty procedures in SCD. The etiology of acute chest syndrome is multifactorial and includes infection, pulmonary infarction and pulmonary fat embolism.[3] Perioperative bleeding may be excessive and this brings out complications associated with transfusion. Vichinsky et al[3] have reported operative complication rates as high as 67%. To prevent such complications hemoglobin levels should be kept between 9-11 g/dL and hemoglobin S values below 30% by preoperative transfusion.[3,5,18] Transfusion preserves the oxygen carrying capacity and rheologic characteristics of thrombocytes.[3] If the patient has a history of febrile transfusion then leucocyte washed or filtered blood should be the choice.[3] As a venous approach might be problematic during the procedure, a central line should be placed priorly, the patient should be kept well hydrated, oximetric catheters should be used and excessive sedation leading to respiratory inhibition and thus hypoxia should be avoided.[19] Care should be taken for intraoperative complications such as hypo- and hypertension, hypo- and hyperthermia, hypoxia and acidosis.[3] The most common intraoperative complications are excessive bleeding followed by hypothermia.[3]

It is appropriate to provide sufficient hydration and analgesia, keep the body warm and supplement oxygen inhalation for the first 24 to 48 hours postoperatively.[20] PO_2 should not drop below 95 mm Hg and should be checked by evaluating arterial blood gases. It should be

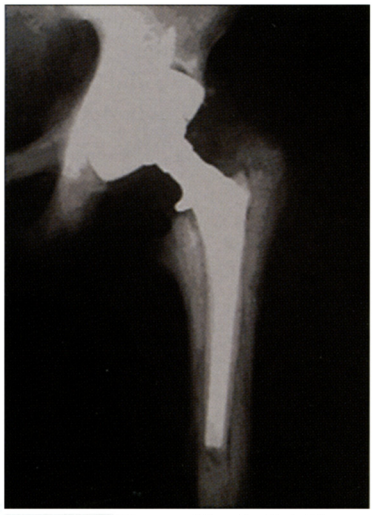

Figure 1.A 38-year old male. Hybrid arthroplasty (femur cemented). Septic loosening at postoperative 9 months.

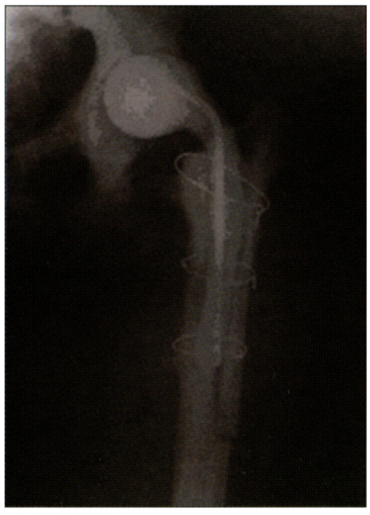

Figure 1.B Application of acrylic spacer with extended trochanteric osteotomy.

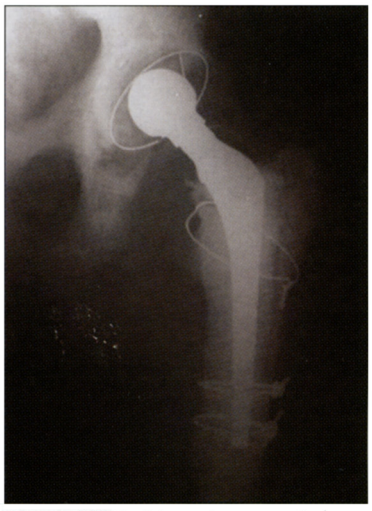

Figure 1.C Revision using cemented components. Early postoperative radiograph.

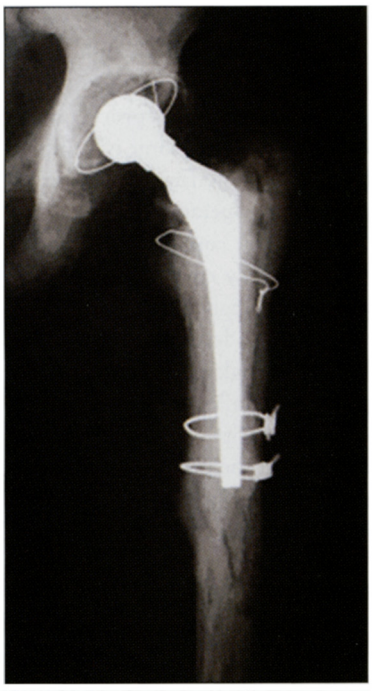

Figure 2. A Reinfection in the same patient at postoperatively 20 months.

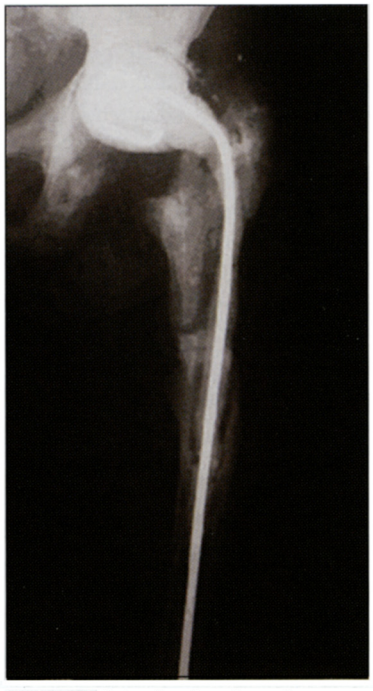

Figure 2. B Application of acrylic spacer for the second time. Development of segmental bone defect.

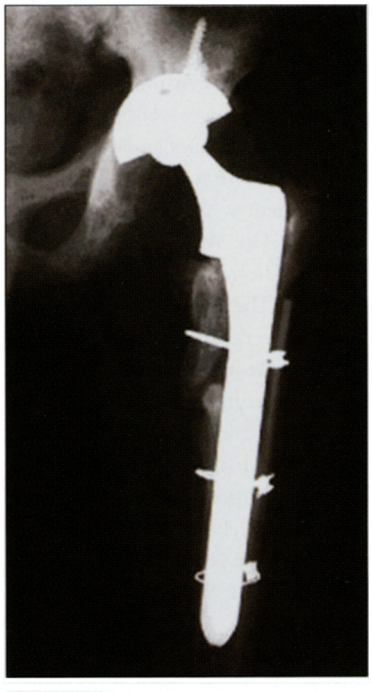

Figure 2. C Revision using cementless components and strut allografts.

remembered that atelectasia, pneumonia, an increased risk of vasoocclusive crisis, hemolytic transfusion reactions due to alloimmunization, acute renal failure, wound infections and sepsis may also be encountered.[3,4,8,16,17,19,21] Hospitalization times are prolonged in this group of patients which further increases the risk of complications.[3,5,7] Mortality is generally secondary to pulmonary complications. Epidural anesthesia should be preferred whenever possible. This provides vasodilation and thus decreases the risk for sickling and also decreases perioperative bleeding.[11] Although the cases in the current series are revision procedures, we did not experience major complications except two vasoocclusive crises during the perioperative period. Precautions taken in preoperative work-up and postoperative care avoided occurance of possible complications.

The literature on hip replacement in SCD reveals increased rates of complications and revisions. Epps and Castro[21] reported a 63% complication rate in 45 patients in a 3.3-year period. Gunderson et al[10] reported a 27% complication rate in 11 patients in 7.5 years. Al-Mausawi et al[6] reported 6 aseptic and 1 septic revision in 35 hips in a 9.5-year period. Hanker and Amstutz[5] reported a 100% complication rate and 5 revisions in a series of 9 patients in 6.5 years. Clarke et al[11] reported the necessity for revision as 59% in 5.5 years, 90% of which had been cemented (one of these had a revision procedure for aseptic loosening while three patients had resection arthroplasty for infection). Bishop et al[4] peformed 4 (33%) revisions in 11 patients during a 7.5-year follow-up. Moran et al[8] revised 5 (38%) of 13 patients in a 4.8-year follow-up. White[17] reported satisfactory pain and functional outcome in 44 hips of 33 patients as 81% and 68%, respectively with 7.5 years follow-up. In a series of 25 patients, Acurio[7] reported the rate of problematic hips as 66% with a revision rate of 40%. Their revision rate for cemented hips was 59% compared with 22% for uncemented replacement. Moran et al[22] reported three failures (43%) in a series of 7 revision THA in 5.3 years. The causes of failure were femoral loosening, acetabular loosening and deep sepsis in one case each. Hickman et al[9] reported that 5 of 8 revisions had one or more early complications, but one patient required re-revision for aseptic loosening of the acetabular component at 11 years. Acurio and Friedman[7] reported 14 revisions in a total of 22 procedures, including nine resection arthroplasties and 13 revision THA. Clarke et al[11] reported 10 revision hip arthroplasties. Average time to revision was only 42 months after the primary procedure, seven for loosening and three for secondary bacterial infection.

Our results for revision hip arthroplasty in SCD were unsatisfactory because of complications requiring revision or obstacles that did not require operative intervention in 7 (63.6%) patients. Kaplan-Meier analysis revealed 5-year survival of arthroplasties as 64%. This rate is extremely high, especially compared with revision THA procedures undertaken for other groups of patients.

There is still no consensus on application of THA for SCD. Pierce,[23] Sennara and Gorry[24] recommended postponing surgical intervention. Sebes and Kraus[25] recommended THA in SCD. They reported the results of six hemiarthroplasty and six THA. There was one infection requiring resection arthroplasty. The maximum duration of follow up was less than two years.

The reasons for loosening starts in the perioperative period. Patients have poor bone quality and potential for remodelling. The procedure is technically demanding due to sclerosis and bone marrow hyperplasia in the femoral canal. Fractures and perforations are quite frequent.[5,6,11,18,20] This creates problems while using press-fit cementless stems. Preoperative planning and choosing a narrow stem can reduce the incidence of this complication. The quality of acetabular subchondral bone is also poor due to pelvic marrow hyperplasia. For the reasons mentioned it is wise to do reamerization with flexible reamers under floroscopic control.[11] Long-term fixation is problematic.[8] The bony bed is prone to infection.[3] Rehabilitation is troublesome because of painful crisis. As a conscequence of these factors septic or aseptic loosening and poliethilen wear are problems encountered.

The rate of infection for arthroplasty in SCD is reported to be between 16% to 20%.[4,5,7,21] These rates are quite high compared to arthroplasty procedures undertaken for other groups of patients.[16,17,26] It was even higher (27.3%) in our current series of revision. These patients are prone to infection because of functional asplenia, abnormal immunity, decreased blood flow through bony substances secondary to sickling and common intravenous procedures during the treatment of their primary disease. Wound drainage and hematoma formation creates an increased tendency for infections. Prolonged operative periods because of technical difficulties further increases the risk.[4,5,8] For this reason introduction of preoperative antibiotics for prophylaxis is of great importance.[2,4,5,7,11,15,21] Care should be taken in administration of antibiotics prior to invasive procedures such as catheterization or dental procedures. The surgeon should also be suspicious of an infection as unexplained pain may be the only presenting symptom, which is quite common due to the primary disease in this group of patients.

Perioperative complications may be avoided by precautions. However, implant choice seems to be an important factor influencing outcome and rate of complications in the long term. Hickman and Lachiewicz[9] reviewed 15 hip arthroplasties with SCD after an average follow-up of 6 years. No infection or loosening was observed with cementless components, although polyethylene wear and osteolysis was a concern. Sanjay and Moreau[20] reported 26 sickle cell hip disease patients with uncemented bipolar hip replacement with a mean follow up of 4.6 years and reported no femoral stem loosening. Acurio and Friedman[7] reported 17 hips that had been cemented; 10 (59%) ended with revision surgery, while only 4 of the 18 (22%) uncemented hips required revision. Nine of 10 arthroplasties that underwent revision were cemented in the series reported by Clarke et al.[8] In the current series all loosenings were observed in the cemented components. The use of cement is likely to cause thermal necrosis of the already infarcted bone, predisposing to a higher incidence of infection and loosening. Now there is almost universal agreement for the use uncemented THA.

Functional recovery is satisfactory in patients with SCD undergoing THA because of functional limitation secondary to osteonecrosis and degenerative arthritis of the hip joint. However, because of high rates of infection and other complications the procedure probably ends up with harm to the patient in the long term. Patients should be carefully selected and managed by a team including a hematologist, an anesthesiologist and an orthopedic surgeon to decrease the risk of complications. Care should be taken while making decisions on the implant type to be used. Cementless systems should be preferred. Revision is far more difficult because of excessive osteolysis and loss of bone stock. The surgeon dealing with arthroplasty in SCD should be cautious against specific complications. The high rates of complications with arthroplasty procedures lead surgeons to consider different treatment options such as arthrodesis, resection arthroplasty or osteotomy. Arthrodesis is usually not prefered as the disease commonly involves both hips. Resection arthroplasty may be considered as a primary measure, but it should be remembered that it is primarily a salvage procedure.

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