

**Total Hip Replacement On  
Sickle Cell Disease: Series  
Of 14 Prostheses.****Prothèse Totale De Hanche  
Sur Drépanocytose: Série  
De 14 Prothèses.**

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**ABSTRACT**

Avascular necrosis of femoral head is one of significant complications affecting the musculoskeletal system in patient with Sickle Cell disease (SCD). Total hip arthroplasty (THA) in SCD can be difficult and prolonged due skeletal changes in the form of osteitis and sclerosis of femoral canal. Twelve patients with SCD, who had osteonecrosis of the femoral head, were operated between 2006 and 2015. The mean age of patients was 31 years (range 19–48 years). There were seven females and five males. Bilateral cemented total hip replacement (THR) was performed in two patients (4 hips) and in the rest unilateral (10 hips). In all patients modified Harris hip

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scores improved from a mean 42 Preoperative to 92 postoperative. However, at a mean follow-up of 5.8 years (range 3-8 years), three hips failed, two due to symptomatic aseptic loosening and one due to septic loosening. Our paper results support the decision to offer the procedure for patients with arthritic hips secondary to SCD, and aims to highlight preoperative, intraoperative and postoperative difficulties encountered in performing THA in sicklers and the short term outcome using cemented implants.

**Keywords:** total hip arthroplasty, sickle cell, avascular necrosis, femoral head

**RESUME**

La nécrose avasculaire de la tête fémorale est l'une des complications importantes affectant le système ostéo-articulaire chez les patients atteints de drépanocytose. L'arthroplastie totale de la hanche (PTH) sur drépanocytose peut être difficile et prolongée en raison des changements de la structure osseuse à savoir l'ostéite et la sclérose du canal fémoral. Douze patients atteints d'ostéonécrose de la tête fémorale sur drépanocytose ont été opérés entre 2006 et 2015. L'âge moyen des patients était de 31 ans (extrêmes: 19-48 ans). Il y avait sept femmes et cinq hommes. Une prothèse totale de hanche cimentée a été réalisée chez 10 patients, deux patients ont bénéficié d'une arthroplastie totale bilatérale (4 hanches). Chez tous les patients, le score modifié de la hanche de Harris est passé d'un score moyen de 42 préopératoire à 92 en postopératoires. Cependant, à un suivi moyen de 5,8 ans (intervalle de 3 à 8 ans), trois arthroplasties ont échoué, deux en raison d'un descellement aseptique symptomatique et une en raison d'un descellement septique.

Les résultats de notre étude soutiennent la décision d'offrir cette procédure pour les patients souffrant de hanches arthrosiques secondaires à la drépanocytose. Egalement, ils mettent en évidence les difficultés préopératoires, peropératoires et postopératoires rencontrées pour réaliser une arthroplastie totale et les résultats à court terme pour les implants cimentés.

**Mots-clés:** arthroplastie totale de la hanche, drépanocytose, nécrose avasculaire, tête fémorale

## INTRODUCTION

Depending on the particular genotype and severity of the sickle cell hemoglobinopathy (1), the incidence of femoral head osteonecrosis in this patient population is estimated to be between 3% and 50% (2-4). Sickle Cell disease (SCD) patients have repeated sickling episodes leading to avascular necrosis of the femoral head. SCD leading to endarteritis induces skeletal changes in the form of osteitis, sclerosis of the femoral canal and osteonecrosis of femoral head (4). All these make surgery difficult and prolonged. Total hip replacement in cases of osteonecrosis of the hip secondary to SCD poses a considerable challenge to the treating orthopedic surgeon. The focal sclerotic marrow poses problem with reaming and placement of the implant. Narrow femoral canals may require the use of small/shorter femoral stems. The soft spongy bone in sicklers is at risk of fracture and bleeds a lot. There is increased risk of infection, SCD crisis and increased complication rate in these patients (5-8).

The aim of this report is to present our experience, and the problems encountered, with total hip replacement in avascular necrosis of the femoral head in SCD patients.

## MATERIALS AND METHODS

Twelve patients with SCD, with osteonecrosis of the femoral head (fig 1),

operated between 2006 and 2015 were included in this study. The mean age of patients was 31 years (range 19–48 years). There were seven female and five male patients. All patients had homozygous sickle cell anemia, except one who had sickle cell trait. Bilateral cemented total hip replacement was performed in two patients (04 hips) and the rest had unilateral involvement (10 hips) (fig 2). The second side was replaced within two years of the first operation.

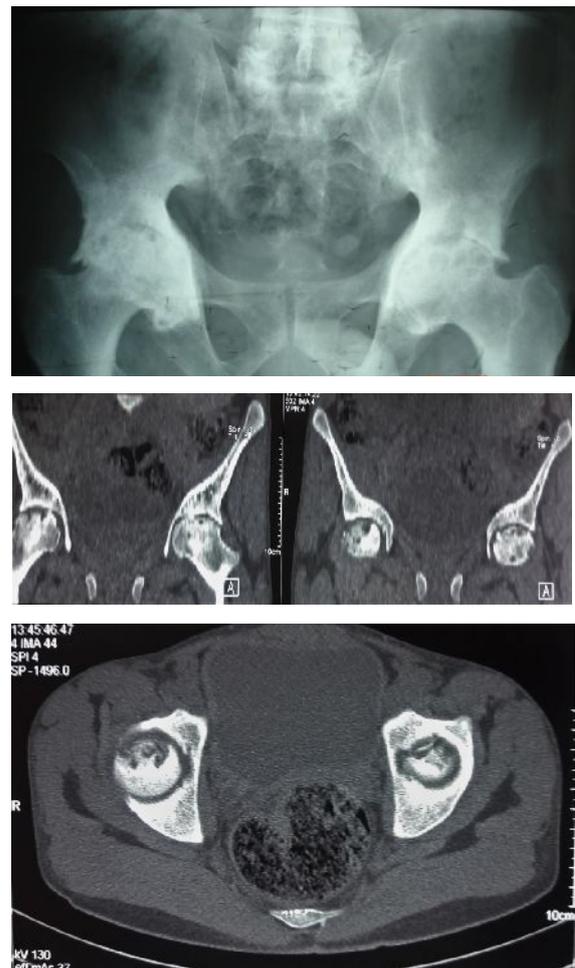


Figure 1a,b and c: bilateral osteoarthritis on bilateral necrosis of femoral heads  
a: appearance on standard radiography,  
b and c: CT appearance.

Main symptoms of our patients were pain, deformity, limb shortening, and stiffness of the hip joint. Severity of pain and functional disability were the main indications to proceed to surgery. All patients were

transfused for anemia before surgery. The preoperative HbS load was decreased by giving aggressive preoperative transfusions. Prophylactic antibiotics were commenced on the day of surgery and continued for 10 days thereafter. Anticoagulation was started 12 h postoperatively and continued for 35 days postoperatively.



Figure 2: Coxarthrosis with protrusio acetabuli

All patients were operated under general anesthesia. All operations were performed through the posterolateral approach with the patient in lateral position. In two hips dislocation of the femoral head was difficult due to severe adhesions or protrusio acetabuli (Fig. 3). In these cases the femoral neck was cut in situ to avoid the risk of femoral shaft fracture. Bone graft from the femoral head was used in one hip to reinforce the acetabulum before cementing the cup.



Figure 3: Treatment of the right hip with cemented THA.

In two cases femoral preparation of the medullary canal was complicated by sclerosis. In those circumstances drill bits were used under image intensifier until it was possible to introduce the guide for

medullar reaming. Power reaming of the canal was performed to a diameter of 9–11 mm. Following implantation of the cemented prosthesis, the wound was closed in layers and drained. During the operating, room temperature was maintained at 22°C. The patients were kept hydrated and warm to prevent hypothermia, and were given warm intravenous fluids.

Patients were mobilized next day, and were followed clinically and radiologically at 2 weeks, 1 month, 3 months and then every year. Preoperative and postoperative modified Harris hip score was evaluated (Tableau 1).

Tableau 1: Preoperative and follow-up modified Harris hip score

Case and side	Age of operation	Preoperative score	Follow-up score
1-L	28	51	103
2-L	36	41	91
3-R	32	55	102
4-B	19	35	84
5-R	45	35	86
6-R	38	41	91
7-B	22	32	82
8-R	33	39	90
9-L	22	40	90
10-R	27	51	102
11-L	49	32	82
12-R	23	53	102
/Mean	<b>31</b>	<b>42</b>	<b>92</b>

## RESULTS

The average followup was 5.8 years (range 3-8 years). The average blood loss in

patients was 920 ml (range 650–1300 ml). Mean blood replacement during and after the operation was 2.4 (1–5) units. One patient was admitted to the special care unit for 72 h after the operation for critical care management of sickle cell crisis. All the patients showed an improvement in Harris Hip score which improved from average 42 points preoperatively to average 92 points at latest followup.

Two patients had persistent wound discharge for 5–7 days. Cultures from their wounds were sterile. None of the patients had infection. Three patients developed limb length discrepancy less than 1 cm. Three hips failed, the first due to a septic loosening at 18 months (fig 4) which required a revision on two stages at an interval of 6 months using a spacer of antibiotic impregnated cement, two due to aseptic loosening at 6 and 7 years, both required revision for the cup. There were no early or late dislocations. No heterotopic ossification was seen in any of the cases. There were no cases of sciatic nerve palsy.

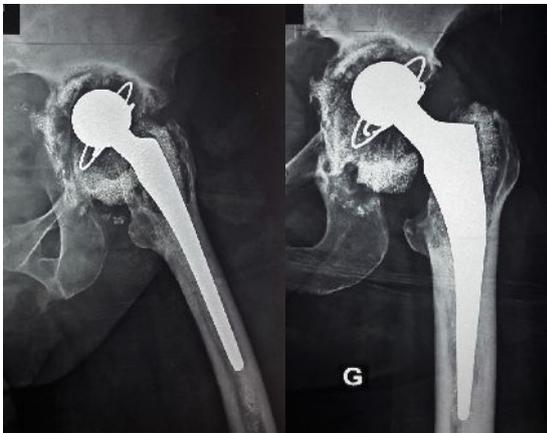


Figure 4: Bipolar septic loosening of the total hip prosthesis.

## DISCUSSION

Sickle cell disease is the most common cause of avascular necrosis in childhood. Depending on the particular genotype and severity of the sickle cell hemoglobinopathy (1), the incidence of femoral head osteonecrosis in this patient population is estimated to be between 3% and 50% (2–4). Hip replacement

arthroplasty is becoming a more frequent operation in the management of those patients when more conservative surgery is no longer an option and the decision for surgery is based on severity of pain and functional disability (9).

Preoperative correction of the anemia to attain a hemoglobin level of 9–11 gm% reduces the risk of a postoperative sickle cell crisis (6). In fact, a conservative preoperative transfusion, to bring Hb 9–11 gm%, is as effective as an aggressive transfusion regimen in which the HbS level was reduced to <30% (8). In our series, despite all patients required blood transfusion before surgery, one required intensive care for sickle cell crisis. Awareness of the risk of sickle cell crisis in the postoperative period is essential, as early recognition and immediate correct measures are mandatory to avoid unnecessary morbidity and mortality (9).

Surgery in SC can be prolonged due to increased blood loss, difficulty in dislocation secondary to protrusion or adhesions. Bleeding occurs from the vascular channels that enter the bone from the capsule. Some authors found bone wax very useful to block these vascular channels (10).

Long-standing deformities, poor quality soft bone (2), and presence of sclerosis (5, 11) should be expected and account for technical difficulties during operation. Hernigou *et al.* performed hip arthroplasty in 244 patients (312 hips) with SCD and found medullary sclerosis in 46 cases (12). This increases the risk of perforation and fractures. Hanker *et al.* and others have described increased perforation rates (5, 6). In our case, femoral preparation of the medullary canal was complicated by sclerosis in two patients. In those circumstances drill bits were used under image intensifier until it was possible to introduce the guide for medullar reaming. Then power reaming of the canal was performed to a diameter of 9–11 mm.

Furthermore, since sickle haemoglobinopathies affect growth and development, the bones in sicklers are usually smaller (13). Mosawi *et al.* recommend keeping a small size implant available if needed for implantation to avoid over-preparation at the expense of bone stock (9). We did not need to use specialized short stems in our cases.

The presence of protrusion and thin floor in the acetabulum are relatively common in SCD patients with avascular necrosis (14). Therefore, Bone graft should always be considered. Sufficient time and care should be given to the preparation of the acetabulum to have good cement hold.

A great care should also be given to the intraoperative and postoperative management to avoid sickle cell crisis, including replacement of blood, hydration, and oxygenation until the patient is stable (8, 9).

Patients with SCD are more prone to infection due to compromised immune status and poor circulation of blood in the bone. Prolonged operative time added to the risk (15, 16). There is a high risk of osteomyelitis in sicklers. It is recommended to use antibiotic impregnated PALACOS® cement in cemented THA (5). Hernigou *et al.* in their series of THA performed in SC hemoglobinopathy reported postoperative infection rate of 3%. They advocated a two-stage revision at an interval of 45 days (12) which is contrasting to earlier series that reported a high infection rate (15-17). The authors attribute this to shorter operative time, meticulous hemostasis, laminar air flow, preoperative correction of anemia, and SC load. In our study one hip was developing a septic loosening which revised on two stages at an interval of 6 months using a spacer of antibiotic impregnated cement.

The selection of prosthesis fixation in patients with SCD is controversial. In one hand, good results have been demonstrated

using cementless THA (7, 18, 19). Cementless fixation has potential advantages in patients with SCD, including mainly a lower rate of aseptic loosening (7,18,19), in addition to the fact that polymethylmethacrylate cement has been implicated as a source of high infection rates and septic loosening (11), the use of cement may cause thermal necrosis, further predisposing the bone to infection and loosening (19). In the other hand, a more recent retrospective study reported better results with cemented components (12). There are some advantages that cemented fixation may provide, including additional hemostasis, decreased risk of femoral perforation and avoidance of biologic fixation in avascular/necrotic bone (20). Furthermore, the use of cementless components relies on bony ingrowth for fixation in bone that may be largely necrotic. We used cemented implants in all our patients.

The rate of aseptic loosening is reported to be higher in THA in SCD patients compared to other populations (6, 11, 18). Hernigou *et al.* in their review of 312 hip arthroplasties performed in 244 patients with SC disease at mean followup of 13 years reported an aseptic loosening rate of 8% (20 cups) and 5%(16 stems). They also mentioned that the risk of aseptic loosening was less than that reported in earlier studies (12), we observed two cases of symptomatic aseptic loosening which represents around 14% of our patients but it is still premature to speak of exact loosening outcome at a short term follow-up.

## CONCLUSION

THA in patients with SCD remains a successful treatment choice for improvements in functions and pain relief in this difficult to treat patient population. Contrary to earlier reports, THA in sicklers now has a predictable outcome, less complications and failure rates especially with a better multidisciplinary management involving hematologists, anesthetists and

orthopedic surgeons. Most complications are preventable by appropriate preoperative preparation, attention to surgical detail, anticipating the potential difficulties, and careful post-operative care.

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