International Orthopaedics (SICOT) (2002) 26:157–161
DOI 10.1007/s00264-002-0337-5

ORIGINAL PAPER

F. Al-Mousawi · A. Malki · A. Al-Aradi · M. Al-Bagali
A. Al-Sadadi · M.M.Y. Booz

Total hip replacement in sickle cell disease

Accepted: 17 January 2002 / Published online: 5 April 2002

Abstract We report on 35 total hip replacement arthroplasties in 28 patients with avascular necrosis of the femoral head secondary to sickle cell disease (SCD). There were 15 men and 13 women with a mean age of 27.5 years. In all patients Harris hip scores improved from a mean of 36 pre-operative to 86 post-operative. However, at a mean follow-up of 9.5 (5–15) years six hips failed due to symptomatic aseptic loosening and one due to late deep infection. Our results support the decision to offer the procedure for patients with arthritic hips secondary to SCD. It is important that patients and surgeons should be aware of the wide varieties of complications.

Résumè Nous rapportons sur 35 arthroplasties totales de la hanche dans 28 malades avec nécrose avasculaire de la tête fémorale secondaire à drépanocytose. Il y avait 15 hommes et 13 femmes avec un âge moyen de 27.5 années. Dans tout le score de la hanche de Harris amélioré d'une moyenne de 36 préopératoire à 86 postopératoire. Cependant, à une suite moyenne de 9.5 (5–15) années six hanches ont manqué dû à descellement aseptique symptomatique et un dû à infection profonde tardive. Nos résultats supportent la décision d'offrir la procédure pour les malades avec les hanches arthritiques secondaire à drépanocytose. C'est important que les malades et chirurgiens doivent être informés des variétés larges de complications.

Introduction

Avascular necrosis of the femoral head is one of the significant complications affecting the musculoskeletal system in patients with sickle cell haemoglobinopathy [6]. The reported incidence of femoral head necrosis varies from less than 10% to more than 30% [15, 23]. In many patients both hips and other bones are affected [8].

Improved medical care of children with sickle cell disease (SCD) has led to prolonged life span. In these patients many young adults will be seeking treatment for avascular necrosis of the femoral head. Review of the literature showed few reports on hip replacement arthroplasties in SCD, with the number of patients ranging from one [9] to 30 [1] and a high complication rate [7, 12, 16, 22]. The incidence of sickle cell haemoglobinopathies presents major health care problems in Bahrain [18]. 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.

Patient and methods

Thirty-seven total hip replacements were performed between 1984 and 1995 for 30 SCD patients with avascular necrosis of the femoral head. One patient moved from the region and another died 3 years after hip replacement from acute pulmonary crisis. Thirty-five hips in 28 patients were available for follow-up. There were 15 men and 13 women with a mean age of 27.5 (19–42) years. Seven patients, five of whom were women, had bilateral hip replacements. The second side was replaced within 2 years of the first operation. The mean number of previous admission to hospital for medical problems related to SCD was 6.5 (range 3–22 admissions). Blood group was A+ve in three patients, O-ve in two, B+ve in one, and O+ve in 22. All patients had homozygous sickle cell anaemia, except one who had sickle cell trait. Mean haemoglobin was 9.1 (range 6.8–12.4) gm. Foetal haemoglobin of 5–15% was present in 25 patients.

Patients presented with pain, deformity, limb shortening, and stiffness of the hip joint. The mean duration of symptoms before operation was 2.3 (range 1–8) years. The decision to proceed to surgery was based on severity of pain and functional disability. Twelve patients were transfused for anaemia with 9 gm or less of blood before surgery. Prophylactic antibiotics and subcutaneous heparin were commenced on the day of surgery and continued for 3 days thereafter.

Presented at the SICOT conference, Paris, France, 2001

F. Al-Mousawi · A. Malki (✉) · A. Al-Aradi · M. Al-Bagali
A. Al-Sadadi · M.M.Y. Booz
Salmanyia Medical Centre, PO Box 12, Bahrain
e-mail: ram81@batelco.com.bh
Tel.: +973-692660, Fax: +973-692660

A. Malki
PO Box 15805, Adlyia, Manama, Bahrain

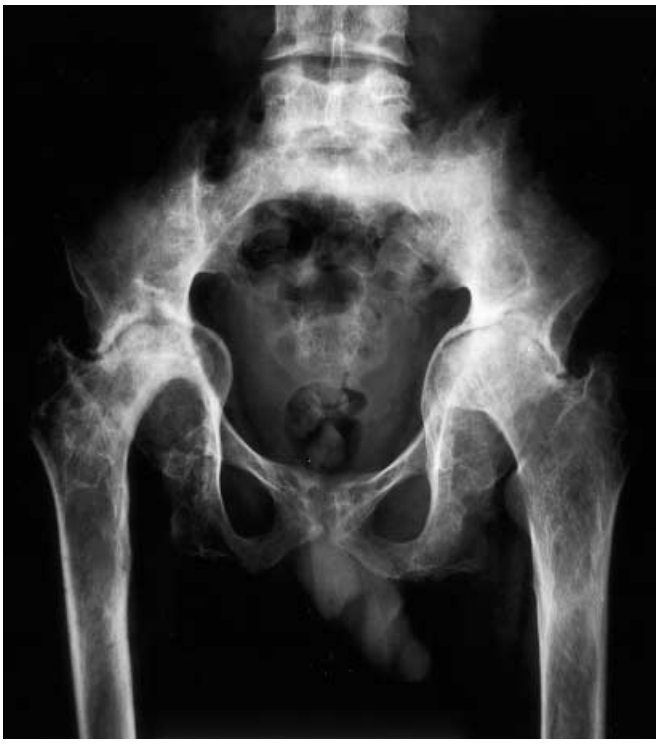

Fig. 1 Radiograph of a 27-year-old man with sickle cell disease showing bilateral protrusio-acetabuli, thin wall acetabulum, femoral head collapse, patchy sclerosis, and the step-sign appearance of fifth lumbar vertebra

Operative technique

Twenty-four patients were operated under general, and four under spinal, anaesthesia. All operations were performed through the posterolateral approach with the patient in lateral position. Subcutaneous adductor tenotomies were required in six hips before positioning on the table and in another three hips at the end of the operation.

Once exposed, the capsule was divided all around the hip joint. The psoas tendon was released by a subperiosteal dissection close to the lesser tuberosity. The tendon of the gluteus maximus was released close to its femoral attachment. In six hips dislocation of the femoral head was difficult due to severe adhesions or protrusio acetabuli (Fig. 1). In these cases the femoral neck was divided in situ to avoid forced dislocation. Bone graft from the femoral head was used in seven hips to reinforce the acetabulum before cementing the cup.

In ten femora 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 medullary reaming. Power reaming of the canal was performed to a diameter of 9–11 mm. A bone medullary plug was inserted in the femur to the desired level. A cement gun was used to fill the canal from the level of the plug upwards to the trochanteric level. Following implantation of the prosthesis, the wound was closed in layers and routinely drained.

Results

The mean duration of operation was 2.2 (1.5–4.5) h. Mean intraoperative blood loss was 1275 (700–2400) ml and from the drain 430 (190–780) ml. Mean blood replacement during and after the operation was 2.4 (1–5) units. Intravenous fluid at operation and during the first 48–72 h post-operation ranged from 4.5 to 12 (mean 9.4) l. Six patients were admitted to the special care unit for 48–72 h after the operation for critical care management of sickle cell crisis. The mean hospital stay was 18 (13–39) days.

In two patients the operation was performed in two stages, with a gap of 2 and 3 weeks, due to excessive bleeding. In the first stage the hip was dislocated and the

Table 1 Data on 28 replaced hips in 23 sickle cell disease patients. The seven failed hips are not included. *L* left, *R* right, *B* bilateral

Case and side	Age at operation	Duration of symptoms (years)	Pre-operative score	Post-operative score	Follow-up 1st side (years)	Follow-up 2nd side (years)
01-L	27	3.0	32	88	15	–
02-L	21	3.0	62	89	14	–
03-R	28	1.5	21	72	13	–
04-L	24	4.0	32	97	13	–
05-B	26	1.5	33	68	13	11
06-R	26	1.0	32	89	12	–
07-L	28	3.0	38	91	12	–
08-B	36	2.0	40	84	12	12
09-L	23	1.5	50	87	11	–
10-L	27	1.0	33	87	10	–
11-R	19	5.0	31	89	10	–
12-L	24	2.5	31	82	9	–
13-B	31	2.0	39	91	9	9
14-R	33	2.5	33	88	8	–
15-R	42	8.0	36	74	8	–
16-B	22	1.5	39	92	8	7
17-L	31	2.0	38	91	8	–
18-L	30	1.5	38	82	7	–
19-R	32	1.0	40	86	7	–
20-L	23	0.5	36	86	6	–
21-B	29	1.5	26	84	6	6
22-L	27	1.5	38	83	6	–
23-R	25	1.0	41	92	5	–

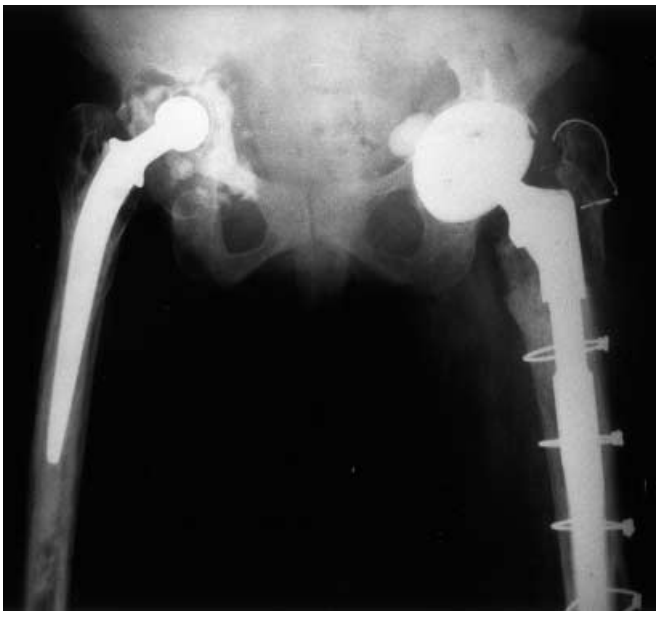

Fig. 2 Radiograph of a patient following revision of the left hip at 13 years due to symptomatic loosening. The loose right hip required revision a year later

femoral neck resected. In the second stage the prosthesis was implanted. During operation, three femoral shafts were accidentally perforated, one of which was fractured. Cerclage wires were used to stabilise the femoral shaft fracture. Delayed wound healing occurred in four patients.

On routine follow-up radiographs four hips showed development of heterotopic ossification. One patient experienced mild pain with extreme hip movements. The mean leg-length discrepancy pre-operatively was 1.8 (0–5) cm and on follow-up 1.1 (0–5) cm. All patients experienced post-operative improvement of pain, movement, and function. Modified Harris hip scoring system [13] was used for assessment on admission to hospital. The mean pre-operative hip score was 36 (21–62). Post-operatively the mean score was 86 (68–97) for 28 hips in 23 patients (Table 1).

Six hips failed due to aseptic loosening at 7, 9, 9.5, 11, 13, and 14 years. Two required revision for the cup and four for both components (Fig. 2). One patient developed deep infection after 10 years. This was treated by aspiration under ultrasound once and aggressive debridement twice but no cure from the infection resulted.

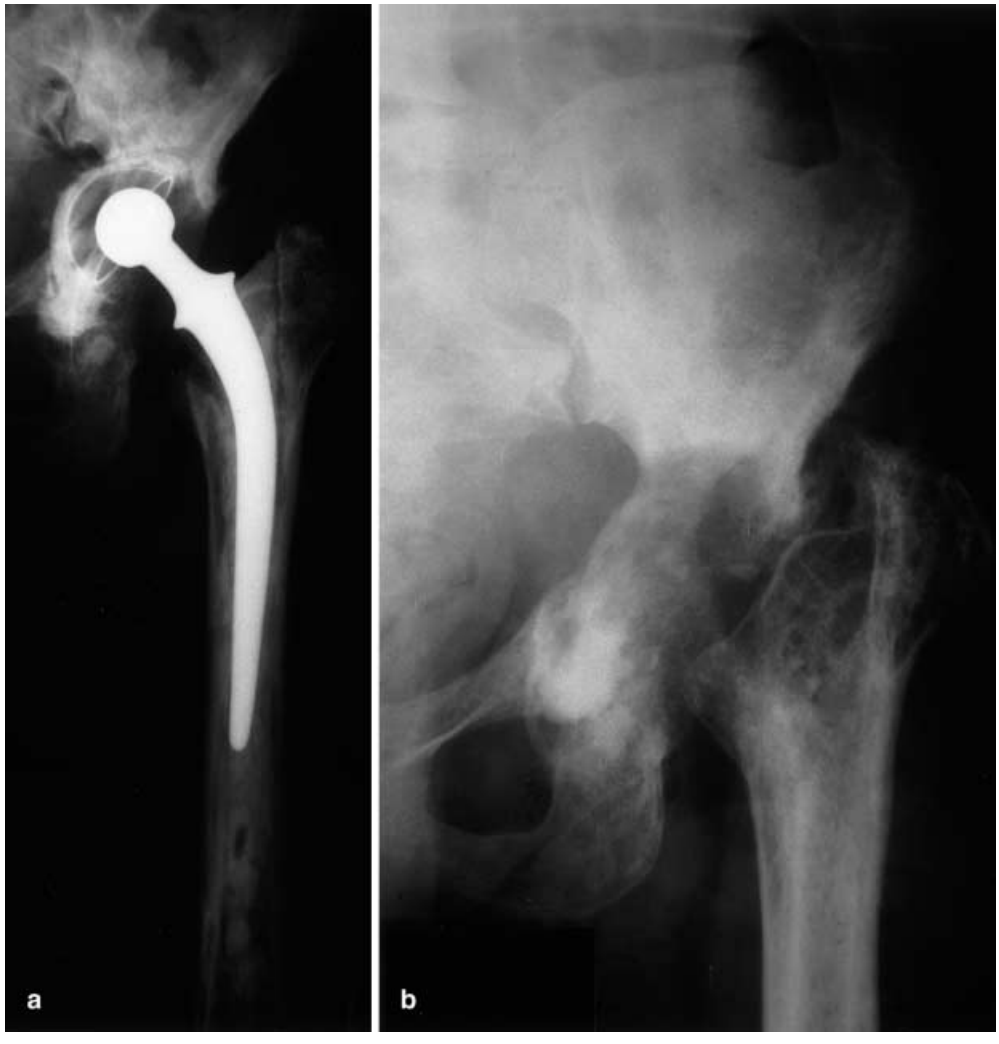

Fig. 3 Radiograph of a patient with **a** replaced left hip that developed deep infection after 10 years; **b** following excision arthroplasty

One year later excision arthroplasty was performed (Fig. 3). Three years later the patient was free from infection and pain. Limb shortening was compensated for by raising the shoe.

Discussion

Specific symptoms related to SCD, including those due to bone infarction, are usually obvious in the first decade of life [3]. Many young adults with SCD are developing disabling problems due to the effect of the disease on the skeletal system [21]. Avascular necrosis of the femoral head is a common sequela of vaso-occlusive attacks. Hip symptoms are commonly seen in the second and third decades [10]. Hip replacement arthroplasty is becoming a more frequent operation in the management of those patients who have passed the stage of more conservative surgery. Decision for surgery is based on severity of pain and functional disability.

Pre-operative correction of the anaemia reduces the risk of a post-operative sickle cell crisis [12]. In our series 12 patients required blood transfusion before surgery. Despite all these precautions six of our patients required intensive care for sickle cell crisis. Awareness of the development of sickle cell crisis in the post-operative period is essential, as early recognition and immediate correct measures are mandatory to avoid unnecessary morbidity and mortality.

Technical difficulties during operation should be expected due to long-standing deformities, poor quality soft bone [6], and presence of sclerosis [7, 17], which may obliterate the femoral canal making its preparation extremely difficult and hazardous. Furthermore, the bones in these patients are usually smaller since sickle haemoglobinopathies affect growth and development [19]. To avoid over-preparation at the expense of bone stock, a range of small-sized prosthesis should be available when needed for implantation.

Adductor tenotomy, complete capsulotomy, partial capsulectomy, tenotomy of psoas tendon, and subperiosteal release of gluteus maximus tendon, were frequently required in our patients to facilitate the operation. Dislocation of the hip is difficult in the presence of large osteophytes, protrusio acetabuli, and intra-articular adhesions between the acetabulum and the head of the femur. After removal of osteophytes, if dislocation is still difficult it is safer to divide the femoral neck in situ and remove the femoral head in fragments rather than taking the risk of femoral shaft fracture. The acetabulum is usually full of adhesions, and sufficient time and care should be given to its preparation for good cement hold. Bone graft for the acetabulum is needed in the presence of protrusio and thin floor, which are relatively common in SCD patients with avascular necrosis [15].

Variation in the quality of bone stock was remarkable in our patients. Difficulty with drilling of hard sclerotic femur frequently was encountered. Care should be taken in the preparation of an obliterated femoral canal by using a drill bit under image intensifier until it is possible to insert the guidewire for the flexible reamers. Despite this precaution perforation occurred thrice in our series and one of them fractured. Incidence of fractures and perforations were higher in other reports [7, 20].

Soft tissue release and preparation of the acetabulum and the femur may significantly increase blood loss [22] and prolong operative time. If sickle cell crisis is to be avoided replacement of blood, hydration, and oxygenation should be monitored with great care, both during operation and post-operatively until the patient is stable. Sickle cell patients are at high risk for infection due to compromised immune status and poor circulation of blood in bone [1, 4, 5, 11]. Prolonged operative time adds to the risk of infection. Late deep infection occurred in one of our patients. The protective role of foetal haemoglobin against infection and crisis in SCD patients is well recognised [2, 14]. This may have contributed to our relatively favourable results, as it was detected in 25 of our patients. Quantitative estimation of foetal haemoglobin in Bahrain is reported to be 4–20% in 76% of SCD patients [18].

The number of hip replacement arthroplasties for SCD patients in the major published reports is relatively small, ranging from 11 to 35 hips. Furthermore, the mean follow-up periods ranges from 4.6 to 8.6 years [1, 4, 7, 12, 16, 17, 20]. There is a need to encourage reports on more patients with longer follow-up periods.

In conclusion, patients with SCD are at a high risk during total hip replacement surgery and in the early post-operative period. Approximately a 20% or higher failure rate is expected at a mean follow-up of 10 years. A multidisciplinary approach involving haematologist, anaesthetist, and the orthopaedic surgeon should be used to reduce the incidence and severity of complications. Most complications are preventable by appropriate pre-operative preparation, attention to surgical detail, anticipating the potential difficulties, and careful post-operative care.

We have confirmed that hip replacement is a challenging problem in SCD patients. However, it is a useful procedure and could be relatively safe in patients suffering from avascular necrosis of the femoral head secondary to SCD.

References

1. Acurio MT, Friedman RJ (1992) Hip arthroplasty in patients with sickle cell haemoglobinpathy. J Bone Joint Surg [Br] 74:367–371
2. Ali SA (1970) Milder variant of sickle cell disease in Arabs in Kuwait associated with unusually high level of foetal haemoglobin. Br J Haematol 19: 613–619
3. Bainbridge R, Higgs DR, Maude GH, Serjeant GR (1985) Clinical presentation of homozygous sickle cell disease. J Paediatr 106: 861–885
4. Bishop AR, Roberson JR, Eckman JR, Fleming LL (1988) Total hip arthroplasty in patients who have sickle cell hemoglobinopathy. J Bone Joint Surg [Am] 70:853–855
5. Booz MMY, Hariharan V, Aradi AJ, Malki AA (1999) The value of ultrasound and aspiration in differentiating vaso-occlusive crisis and osteomyelitis in sickle cell disease patients. Clin Radiol 54:636–639

6. Chung SMK, Ralston EL (1969) Necrosis of the femoral head associated with sickle cell anaemia and its genetic variants. J Bone Joint Surg [Am] 51:33–58
7. Clarke HJ, Jinnah RH, Brooker AF, Michaelson JD (1989) Total replacement of the hip for avascular necrosis in sickle cell disease. J Bone Joint Surg [Br] 71:465–470
8. David HG, Bridgman SA, Davies SC, Hine AL, Emery RJH (1993) The shoulder in sickle cell disease. J Bone Joint Surg [Br] 75:538–545
9. Dumarey N, Martin P, Jayankura M, Putz P, Verhas M, Peretz A (2000) A 'made in one piece' skeleton in a 22-year-old man suffering from sickle cell anaemia. Clin Rheumatol 19:287–290
10. Ebong WW, Kolawole TM (1986) Aseptic necrosis of the femoral head in sickle cell disease. Br J Rheumatol 25:34–39
11. Epps CH, Bryant DD, Coles MJM, Castro O (1991) Osteomyelitis in patients who have sickle cell disease; diagnosis and management J Bone Joint Surg [Am] 73:1281–1293
12. Hanker GJ, Van Nuys, Amstutz HC (1988) Osteonecrosis of the hip in sickle cell disease; treatment and complications. J Bone Joint Surg [Am] 70:499–506
13. Harris WH (1969) Traumatic arthritis of the hip after dislocation and acetabular fracture; treatment by mould arthroplasty; an end result study using a new method of result evaluation. J Bone Joint Surg [Am] 51:737–755
14. Hawker H, Neilson H, Hayes RJ, Serjeant GR (1982) Haematological factors associated with avascular necrosis of the femoral head in homozygous sickle cell disease. Br J Haematol 50:29–34
15. Hernigou P, Galacteros F, Bachir D, Goutallier D (1991) Deformities of the hip in adults who have sickle cell disease and had avascular necrosis in childhood. J Bone Joint Surg [Am] 73:81–92
16. Hichman JM, Lachiewicz PF (1997) Results and complications of total hip arthroplasties in patients with sickle cell hemoglobinopathies. Role of cementless components. J Arthroplasty 12:420–425
17. Moran MC, Huo MH, Garvin KL, Pellicci PM, Salvati EA (1993) Total hip arthroplasty in sickle cell hemoglobinopathy. Clin Orthop 294:140–148
18. Nadkarni KV, Al-Arrayed SS, Bapat JP (1991) Incidence of genetic disorders of haemoglobins in the hospital population of Bahrain. Bahrain Medical Bulletin 13:19–24
19. Platt OS, Rosenstock W, Espeland MA (1984) Influence of sickle hemoglobinopathies on growth and development. N Eng J Med 311:7–12
20. Sanjay BKS, Moreau PG (1996) Bipolar hip replacement in sickle cell disease. Int Orthop 20:222–226
21. Sennara H, Gorry F (1978) Orthopaedic aspects of sickle cell anemia and allied hemoglobinopathies. Clin Orthop 130:154–157
22. Vichinsky EP, Neumayr LD, Haberkern C, Earles AN, Eckman J, Koshy M, Black DM (1999) The perioperative complication rate of orthopaedic surgery in sickle cell disease: Report of the National Sickle Cell Surgery Study Group. Am J Hematol 62:129–138
23. Ware HE, Brooks AP, Toye R, Berney SI (1991) Sickle cell disease and silent avascular necrosis of the hip. J Bone Joint Surg [Br] 73:947–949