HIP ARTHROPLASTY IN PATIENTS WITH SICKLE-CELL HAEMOGLOBINOPATHY

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We reviewed retrospectively 25 hip arthroplasties in 25 patients with sickle-cell haemoglobinopathy and osteonecrosis. The mean age of the ten women and 15 men at the onset of hip symptoms was 25 years, and at surgery 30 years (16 to 45); 66% had either SS or S-thal disease, 20% sickle-cell trait, and the remainder SC disease. The mean follow-up was 8.6 years (two to 18).

Fourteen (40%) of the arthroplasties had been revised at a mean of 7.5 years after the primary procedure and nine other hips were either radiographically and/or symptomatically loose. The overall complication rate was 49% and the infection rate 20%.

The risk-to-benefit ratio of hip arthroplasty in sickle-cell haemoglobinopathy is high.

Sickle-cell haemoglobinopathies include a variety of diseases which have been linked to the development of osteonecrosis of the femoral head. Sickle-cell disease (SS) is prevalent among black people in North America, the West Indies, Africa and other regions around the world, and the most common type is sickle-cell trait (AS) (Chung and Ralston 1969). Others include sickle thalassaemia (S-thal) (0.06%) and sickle C (SC) (0.2%); some types are occasionally seen in people of Mediterranean descent.

Osteonecrosis has been reported to occur in 20% to 50% of patients with SC disease (Chung and Ralston 1969), but there is a lower incidence in SS disease. This is, however, the more prevalent form and patients with SS disease and osteonecrosis are therefore more likely to be encountered.

In sickle-cell haemoglobinopathy the immune system is altered possibly due to a defect in the complement cascade (Chung, Alavi and Russell 1978). Patients also have functional asplenism secondary to autoinfection, poor bone perfusion, and experience multiple crises which are of four types: vaso-occlusive, aplastic, haemolytic, and sequestration.

During a vaso-occlusive crisis, alterations in the amino-acid chains of haemoglobin S result in conformational changes when in the deoxygenated state. Molecular stacking occurs, and hollow rigid rods form due to the irregular aggregation of molecules in the individual cells. This causes sickling of the individual red blood cells, initially reversible on reoxygenation. With repeated cycles, however, this process becomes irreversible and the sickled cells block the sinusoids and small capillaries of metaphyseal bone (Middlemiss and Raper 1966; Chung et al 1978; Sennara and Gorry 1978).

The small blood vessels of the femoral head, with its specific blood supply and lack of collateral circulation, are particularly liable to occlusion by sickled cells. Local thrombosis gives a further reduction of the oxygen tension, resulting in increased sickling. This vicious cycle of continued hypoxia and sickling eventually produces infarction, necrosis, femoral head collapse, and joint destruction (Chung et al 1978).

Because of the shortened survival time of red cells, chronic anaemia develops. In response to this, red marrow proliferates in many areas of the skeleton, the medullary canal widens, and there is thinning and weakening of the cortices (Middlemiss and Raper 1966; Haddad 1967; Chung and Ralston 1969). This is especially noticeable in the metaphyseal region of long bones such as the proximal femur (Chung et al 1978), producing weakness, increased chance of fracture, and a less than optimal environment for a femoral prosthesis. All these changes may lead to the early failure of a hip arthroplasty in these patients.

Short-term results of such hip arthroplasties have been reported (Bishop et al 1988; Hanker and Amstutz 1988); we have determined the long-term clinical and radiographic results.

PATIENTS AND METHODS

We reviewed 25 patients with osteonecrosis secondary to sickle-cell haemoglobinopathy who had undergone 35 hip arthroplasties at the Medical University of South Carolina, Department of Orthopaedic Surgery, Charleston, South Carolina 29425, USA.

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0301-620X/92/$2.00
RESULTS

Two patients had died, one after a traffic accident and one from bladder cancer, but their records were complete and were included.

The mean age of the ten women and 15 men at the onset of symptoms was 25 years (eight to 43), and at the initial hip arthroplasty 30 years (16 to 45). None of the patients was significantly overweight, but most had an activity level consistent with their age and not that routinely recommended for patients with a hip arthroplasty. The mean follow-up was 8.6 years (two to 18).

No risk factors for osteonecrosis other than sickle-cell disease were present in any of the patients except for a femoral neck fracture in one. Eleven (44%) had SS disease, six (24%) S-thal disease, five (20%) sickle-cell trait, and three (12%) SC disease.

All 23 patients completed a detailed questionnaire, and had physical and radiographic examinations. Pain, function, and range of motion of the involved hip were evaluated by the Harris hip rating system (Harris 1969). Failure of the arthroplasty was defined as clinical or radiographic loosening, with or without revision.

RESULTS

Of the 35 primary hip arthroplasties, 20 were total (16 cemented), 11 were bipolar (one cemented), two were Austin-Moore hemi-arthroplasties, and two were cup arthroplasties. In all, 17 were implanted with methylmethacrylate cement; 18 were cementless.

The mean hospital stay for the primary arthroplasty was 20 days (nine to 58), and for revision cases 33 days (ten to 102). The average blood loss was 997 ml (350 to 4900) for the primary arthroplasties and 1585 ml (200 to 2500) for revision procedures. The average transfusion requirement for the primary procedures was three units (0 to seven) and for revision 5.5 units (0 to 17).

Before operation the Harris hip rating averaged 66 points (19 to 98), as both pain and function were limited. The mean pain score was 30 (ten to 44) out of a possible 44, and function averaged 29 (five to 45) out of a possible 47.

At latest review, only 11 patients (34%) had a good or excellent rating, and therefore 66% had an unacceptable result after their primary hip arthroplasty.

Of the 35 primary operations, 14 (40%) required revision surgery. The relative failure rates were 50% for THA, 50% for cup arthroplasty, 50% for hemi-arthroplasty, and 18% for bipolar arthroplasty. Four patients were revised twice, three revised three times, and one four times. The average time from the primary procedure to the first revision was 90 months (nine to 180), from the first to second revision 58 months (24 to 93), and from the second to third revision 30 months (five to 48).

The 14 revised arthroplasties had a total of 22 procedures, including nine resection arthroplasties and 13 revision THA (Fig. 1). Of the resection arthroplasties, five were performed for infection, three for severe bone loss, and one for suspected infection in which the cultures were subsequently negative. Five infections occurred in the cemented arthroplasties and only two in the uncemented. The organisms cultured at the time of surgery are shown in Table I.

Of the 17 hips that have been cemented, ten (59%)

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*Fig. 1a*  
Radiographs of a 23-year-old man with sickle-cell disease and bilateral osteonecrosis, first affecting the left hip at 16 years of age. Figure 1a – Three months after a cemented left total hip arthroplasty. Figure 1b – Four years postoperatively, there is loosening, osteolysis and migration of both components.
had revision surgery, while only four of the 18 (22\%) uncemented hips underwent revision (p < 0.05). Of the nine additional primary replacements which were clinically or radiographically loose, six were cemented and three were not. In total, 16 of the 17 cemented hips were either loose or had undergone revision (94\%), while only seven of the 18 uncemented hips (39\%) were in this category (p < 0.05). The revision rate at 8.6 years was 40\%, with definite loosening in another 26\%, giving failure in 23 of 35 (66\%).

The revision rate in 15 followed for less than seven years was 13\% (one bipolar and one THA out of nine bipolar, four THA, one cup and one hemi-arthroplasty), while it was 60\% in those followed for more than seven years (9/16 THA, 1/2 bipolar, one cup, and one hemi-arthroplasty).

The 17 complications included seven infections (six deep and one superficial), three genito-urinary tract infections, three sickle-cell crises, two intra-operative femoral fractures, and two episodes of excessive blood loss, giving a total rate of 49\%.

Radiographs of revision hip arthroplasties undertaken for aseptic loosening all showed severe bone loss with osteolysis around the femoral component, and similar changes with protrusio on the acetabular side (Fig. 2).

DISCUSSION

Osteonecrosis occurs in patients with haemoglobin SS, SC, and S-thal but is said to be rare in those with sickle-cell trait (Chung and Ralston 1969; Sennara and Gorry 1978). It is seen most commonly in the femoral head, but also in the humeral head, the knee, the spine, and the diaphyses of long bones. Of our patients, 20\% had sickle-cell trait; the incidence of osteonecrosis in this group may be higher than previously reported.

Epps and Castro (1978) reported 45 hip arthroplasties performed for osteonecrosis secondary to sickle-cell disease. Their complication rate was 63\% with a mean follow-up of only 3.3 years, but they concluded that short-term pain relief had been successful. Gunderson, D'Ambrosia and Shoji (1977) reported 11 hip arthroplasties in seven patients with a complication rate of 27\%.

Bishop et al (1988) had four infections in 11 hip arthroplasties at a mean follow-up of 7.5 years, and considered that there was a high risk for both early and late infection. Despite this, they considered that THA was not too great a risk in this group of patients.

Hanker and Amstutz (1988) reviewed 14 hips at 6.5 years with a complication rate of 100\%, increased blood loss and transfusion requirements, and prolonged hospitalisation. They warned of the considerable risk of postoperative morbidity and recommended that the risk-to-benefit should be carefully assessed for each individual patient.

The infection rate in our series was 20\%, comparable with the mean 16\% infection rate reported by Epps and Castro (1978), Bishop et al (1988) and Hanker and Amstutz (1988). This rate is unacceptably high.

Our patients had a mean age of 30 years at the primary arthroplasty, and it is therefore important to compare the results with those for THA in young patients with diagnoses other than sickle-cell haemoglobinopathy. Dorr, Takei and Conaty (1983) reported 108 THAs in patients under 45 years of age (mean 30.5, range 14 to

Fig. 2a
Radiographs of a 31-year-old man with sickle-cell disease and bilateral osteonecrosis. Symptoms began at 19 years. Figure 2a - Ten years after bilateral THA there are radiolucencies around the acetabular components, and osteolysis around the proximal parts of the femoral stems. Figure 2b – The right THA showed severe osteolysis, migration, and component failure on the acetabular side, and resorption of the calcar femorale. Resection arthroplasty was performed because of suspected sepsis, although cultures were subsequently negative. Figure 2c – At 16 years postoperatively, the left THA shows acetabular component failure, osteolysis, and migration. An aspiration arthrogram cultured Staphylococcus epidermidis: resection arthroplasty was performed.
was 50% at a mean follow-up of ten years, and the infection rate was 10%.

The other surgical options for this condition include arthrodesis, resection arthroplasty, osteotomy, and uncremented hip arthroplasty. Arthrodesis may be difficult to achieve, but could be recommended for young patients with unilateral disease who wish to remain active; sickle-cell patients, however, frequently have bilateral involvement. Resection arthroplasty is not indicated as a primary treatment, but has a place as a salvage procedure.

Osteotomies cannot be considered in patients with extensive involvement of the femoral head, and are not recommended as a long-term solution for the sickle-cell patient, since the underlying cause remains. An uncremented arthroplasty, possibly allowing porous ingrowth, or with some form of biological fixation, is an available option, but the safety is unknown and long-term follow-up is essential.

A number of recommendations can be made regarding hip arthroplasty for osteonecrosis secondary to sickle-cell haemoglobinopathy (Chung and Ralston 1969; Chung et al 1978; Epps and Castro 1978; Hanker and Amstutz 1988; Clarke et al 1989):
1) a thorough pre-operative evaluation, including a haematological consultation;
2) pre-operative transfusion, or plasmapheresis, to achieve a circulating level of haemoglobin A greater than 50%;
3) peri-operative antibiotics;
4) adequate hydration and oxygenation intra-operatively to prevent a sickling crisis; and
5) caution in the use of methylmethacrylate cement as this may be a major contributing factor to the high infection and revision rates.

We conclude that patients with sickle-cell haemoglobinopathy have an excessively high infection rate, failure rate, and complication rate after hip arthroplasty for osteonecrosis. With failure, there is extensive osteolysis and bone loss, which makes revision much more difficult. There is an increased risk of early mechanical as well as septic loosening, and the risk-to-benefit ratio may be unacceptably high. Sickle-cell patients should be informed of the risks involved, and surgeons must be prepared to deal with the many possible complications.

The authors wish to thank Vicki Bradshaw for help in the preparation of this manuscript, Mark Foster, MD, for gathering some of the patient data, and M. C. Miller, PhD, for the statistical analyses. No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

REFERENCES


Table I. Organisms cultured from seven THAs, infected after replacement for osteonecrosis due to sickle-cell haemoglobinopathy

<table>
<thead>
<tr>
<th>Organism</th>
<th>Number</th>
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<tr>
<td>Staphylococcus epidermidis</td>
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<tr>
<td>Staphylococcus aureus</td>
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<tr>
<td>Acinetobacter calcoaceticus</td>
<td>1</td>
</tr>
<tr>
<td>Proteus vulgaris</td>
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<tr>
<td>Pseudomonas herrelea</td>
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![Kaplan-Meier survivorship curve](image)

Fig. 3

The Kaplan-Meier survivorship curve for 35 hip arthroplasties for osteonecrosis due to sickle-cell haemoglobinopathy, in which failure is defined as radiological loosening, with or without revision.

45), with a mean follow-up of 4.5 years. Nine of their patients had either sickle-cell disease or trait, and two of these developed postoperative infections. The overall infection rate for their series was 7% and the failure rate 17.5%. Chandler et al (1981) reported 33 hip arthroplasties in 29 young patients (mean 23 years), at a mean follow-up of 5.6 years. The failure rate was 21%, but there were no infections. At follow-up, 57% had evidence of actual or potential loosening of their prosthesis.

White (1988) reported 44 hip arthroplasties in 33 patients with a mean follow-up of 7.5 years, and a mean age of 38 years. The infection rate was 2% and revision rate 14%. Relief of pain and improvement in function were obtained in 81% and 68% respectively.

The revision rate of 40% in our series is nearly three times that reported by White. Our overall failure rate was 66%, unacceptably high when compared with other series with similar age ranges. We performed a statistical analysis using a Kaplan-Meier survivorship curve on our revision rate, which showed a 50% revision rate at 9.6 years (Fig. 3). For the 20 THA in our series, the revision rate was 50% at a mean follow-up of ten years, and the infection rate was 10%.


