

# Hip replacement in sickle cell anemia patient and the effect of Musculoskeletal changes other than hip joint on outcome

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Received: May 2021; Accepted: June 2021; Published: July 1, 2021.

Citation: Mohammed Lafi Al-Otaibi. Hip replacement in sickle cell anemia patient and the effect of Musculoskeletal changes other than hip joint on outcome. World Family Medicine. 2021; 19(7): 20-26 DOI: 10.5742/MEWFM.2021.94074

## Abstract

**Introduction:** Sickle cell anemia is endemic in south west of Saudi Arabia, and patients with hip pain and sickle cell anemia are frequently seen in orthopedic clinics and are a challenge to treat. Total hip replacement is proven to be of benefit for patients with secondary hip joint degeneration due to avascular necrosis. Patient's other musculoskeletal manifestations of sickle cell anemia have their impact on patient's outcome following hip replacement.

**Objective:** To compare the effects of disease manifestation other than that on the replaced hip on outcome of hip replacement in sickle cell anemia patients.

**Material and Methods:** Retrospective study of 20 patients who underwent total hip procedure with 6 month follow up divided into two groups, to see the effect of other musculoskeletal changes on outcome.

**Results:** The total hip procedure was done for all 20 patients; all patients were aged 25 to 52-years old; females were 13 patients and Males 7 patients; Left hip was affected in 12 patients and right hip in 8 patients. All patients had uneventful post-operative recovery; all patients' data is shown in Table one. 6 months post op showed less satisfaction in patients with manifestation of sickle cell anemia involving, in addition of the replaced hip, spinal disorders such as scoliosis or back pain due to infarct and collapse or soft tissue contractures of hips or knees.

**Conclusion:** Categorizing all sickle cell hip avascular necrosis in one group is unfair for patients and surgeons. Patients with multiple sites of disease manifestation must be considered a difficult primary hip and handled with care, while some patients with isolated hip involvement have comparable results to hip replacement in other conditions.

**Key words:** Sickle cell anemia, Total hip, hip scores, hip replacement, avascular necrosis.

## Introduction

Sickle cell anemia is endemic in the south west of Saudi Arabia; patients with hip pain and sickle cell anemia are frequently seen in orthopedic clinics and are a challenge to treat. Although medical treatment has been developed over recent years, surgical treatment namely total hip replacement, is the last resort to address the changes happening to the hip joint due to avascular necrosis and secondary osteoarthritis in sickle cell anemia and there are less invasive procedures such as core decompression and trap door procedure that can and should be offered earlier to alter the natural history and delay the need for total hip replacement. Once total hip replacement is offered it should be remembered that its outcome correlates to the presence of soft tissue changes such as contracture or infection and skeletal findings like the loss obliteration of the medullary canal variation of bone quality, bone load resistance and spine vertebral segmental collapse with avascular necrosis. Patients who only have their changes limited to the hip joint are expected to have a better outcome, while those with other musculoskeletal changes have varying outcomes.

Nearly over 50 years of progressing success, total hip replacement is one of the most rewarding surgeries and named the century surgery (1). In the 1960s, total hip replacement started in an attempt to restore hip function and therefore restore activities of daily living. Since then, improvement in many aspects of this procedure have brought it to today's current situation of high-quality prosthetic implants and well proven surgical indication and contraindication together with evidence proof pre- and post-operative management, with reproducible results in so many hands thanks to improvement in the surgical technique and surgeons training. In general, survival of hip replacement ranges from 85 to 95% (2). Proven to be a complex procedure, total hip replacement for sickle anemia is of concern due to its medical, intraoperative and postoperative complications. The success required to optimize patients medically as well as improving surgical techniques and prosthetic implant manufacture have led recently to comparable outcomes for the sickle cell anemia patient. Adherence to adequate perioperative measures such as hydration, maintenance of body temperature, oxygenation and above 10 mg/dl hemoglobin level factors positively in the outcome and are somewhat easy to apply. Dealing with intraoperative challenges such as soft tissue contractures, bony fracture or perforations are difficult and the major factor for poor outcomes (3). The effects of other bony involvement like scoliosis or soft tissue conditions such as Periarticular Contracture on hip replacement is discussed for other conditions in literature for example degenerative hip and spine in elderly but for sickle cell anaemia patients there is need to study this effect further to improve outcomes for hip replacement for these patients.

## Material and Methods

Ethical approval Number [ECM#2019-20] was approved by King Khalid University, Research Ethical Committee. Retrospective study took place at our hospital during the period between November 2007 to April 2011. Total number of patients was 20 patients who underwent total hip procedure divided into two groups:

**Group A patient inclusion criteria:** no previous back pain or spine clinic follow up, no history of skeletal infection or joint contractures. All patients had pathology localized to the joint articulation only.

**Group B patient inclusion criteria:**

- 1: radiological evidence of vertebral column changes requiring previous spine unit care (Chronic back pain, vertebral column changes e.g., scoliosis, collapse or infection)
- 2: radiological evidence of femoral canal obliteration
- 3): evidence of flexure contractures of the knees or the hips  
(Unable to stand or stand with difficulty, uses wheel chair or walking aid)

## Results

The total hip procedure was done for all 20 patients; all patients were age 25 to 52 years old. Female 13 patients and Males 7 patients, Left hip affected in 12 patients and right hip in 8 patients. All patients had uneventful post-operative recovery. All patient data is shown in Table one. 6 months post op Harris Hip Score obtained for all patients (Table 1).

**Table 1: 6 weeks Post-operative all patients involved**

|   | Group A             | Group B                   |
|---|---------------------|---------------------------|
| Total patients                                  | 10                  | 10                        |
| Males   | 4                   | 3                         |
| Females   | 6                   | 7                         |
| Site of skeletal changes                        | Hips only           | Hips/spine/femoral canal  |
| Presence of soft tissue contractures            | None                | Hip and knee contractures |
| Number of admissions per year                   | Less than 4         | More than 4               |
| Wound infections                                | 1                   | 3                         |
| Transient femoral nerve palsy                   | 1                   | 4                         |
| Aseptic loosening                               | 0                   | 2                         |
| Septic loosening                                | 0                   | 1                         |
| Femoral canal violation                         | 0                   | 2                         |
| Medial acetabular violation                     | 0                   | 1                         |
| Leg length discrepancy                          | 0                   | 2                         |
| Average intraoperative blood loss               | 1100(+/-150ml)      | 1500(+/-200ml)            |
| Average intraoperative blood loss               | 120 (+/-30) minutes | 180(+/-30) minutes        |
| Average Harris hip score: Preoperative          | 45.22+/-3.021       | 25.94±4.437               |
| Average Harris hip score: 6 weeks postoperative | 92.53±1.419         | 88.82±1.845               |

In Table 2 the females dominated the younger group of patients although the oldest patient in the study was female.

**Table 2: Age Distribution of Patients (n=20)**

| Age Distribution (n=20)          |                |       |         |       |       |     |
|----------------------------------|----------------|-------|---------|-------|-------|-----|
| Age Groups (years)               | Males          | %     | Females | %     | Total | %   |
| 20-25                            | 0              | 0.00  | 3       | 15.00 | 3     | 15  |
| 26-29                            | 5              | 25.00 | 7       | 35.00 | 12    | 60  |
| 30-35                            | 2              | 10.00 | 2       | 10.00 | 4     | 20  |
| 36-39                            | 0              | 0.00  | 0       | 0.00  | 0     | 0   |
| 40-45                            | 0              | 0.00  | 0       | 0.00  | 0     | 0   |
| 46-49                            | 0              | 0.00  | 0       | 0.00  | 0     | 0   |
| 50-55                            | 0              | 0.00  | 1       | 5.00  | 1     | 5   |
| Total                            | 7              | 35%   | 13      | 65%   | 20    | 100 |
| Median Age (years)               | 29             |       | 27      |       |       |     |
| t test-unpaired (p value)=0.9315 |                |       |         |       |       |     |
| 95% CI                           | (6.07 to 5.58) |       |         |       |       |     |

[Unpaired t test was used to compare between the Groups A and B]

In Table 3 most complications happened to the patients in group B which is the group with disease manifestation which included other musculoskeletal sites with the hip involvement.

**Table 3: Surgical Complications**

| Complications               | Group A       | Group B                |
|-----------------------------|---------------|------------------------|
| Wound infections            | 1             | 3                      |
| Transient Femoral N Palsy   | 1             | 4                      |
| Septic loosening            | 0             | 2                      |
| Femoral Canal Violation     | 0             | 2                      |
| Medial Acetabular Violation | 0             | 1                      |
| Leg length Discrepancy      | 0             | 2                      |
| Mean                        | 0.33          | 2.33                   |
| SD                          | 0.52          | 1.03                   |
| SEM                         | 0.21          | 0.42                   |
| t test (unpaired)           | 4.2426        | (95% CI=-3.05 to-0.95) |
| p value                     | <b>0.0017</b> |                        |

[Unpaired t test was used to compare between the Groups A and B]

Table 4 showed the statically significant changes between the two groups marking the group with other musculoskeletal manifestations low satisfaction due to more frequently occurring complications.

**Table 4: Comparison between Groups A and B**

| Categories                               | GROUP A | GROUP B | t test<br>(unpaired) | 95% CI               | p value |
|--|---------|---------|----------------------|----------------------|---------|
| Average Intra-operative Blood loss (ml)  | 1100    | 1500    | 5.6882               | (-547.74 to -252.26) | <0.0001 |
| Average operative Time (min)             | 120     | 180     | 9.2445               | (-73.64 to-46.36)    | <0.0001 |
| Average Harris hip Score: Preoperative   | 45.22   | 25.94   | 27.5206              | (17.80 to 20.74)     | <0.0001 |
| Average Harris Hip score: Post operative | 92.53   | 88.82   | 5.66                 | (2.33 to5.08)        | <0.0001 |

[Unpaired t test was used to compare between the Groups A and B]

In Table 5 functional scores favoured those with solitary hip manifestations over patients with multiple musculoskeletal manifestations.

**Table 5: Comparison between Pre and Post-operative Harris Hip Scores**

|                    | Preoperative Hip Score | Postoperative Hip Score |
|--------------------|------------------------|-------------------------|
| Statistical Values | Group A (n=10)         | Group A (n=10)          |
| Mean               | 45.22                  | 92.53                   |
| Paired t test      | 89.4035                |                         |
| 95% CI             | (-48.60 to -46.01)     |                         |
| p value            | <0.0001                |                         |
|                    | Group B (n=10)         | Group B (n=10)          |
| Mean               | 25.94                  | 88.82                   |
| Paired t test      | 119.5067               |                         |
| 95% CI             | (-64.0264 to -61.7336) |                         |
| p value            | <0.0001                |                         |

[Paired t test was used to compare pre and post operative Hip scores]

## Discussion

The prevalence of sickle cell anaemia in southern region Saudi Arabia is one of the largest among a patch incidence ranging from 2 to 27 % with the eastern region being the second with higher incidence [4].

Our study describes peculiar findings of the medullary obliteration figure, acetabular periarticular infarct Figures (1A&B) and spine vertebral column collapse with kyphoscoliosis Figure (2) and presence of soft tissue hip and knee contractures Figure (3) in sickle cell disease patients correlates proportionally with the severity of pre-operative symptoms, intraoperative difficulties and the patient post operative Harris Hip Scores. Severe soft tissue contractures, leg length discrepancy, poor bone quality and medullary cavities obliteration are found to be other factors of lower hip score. Sickle cell disease is quite common in our region and is associated with orthopaedic pathologies of which osteonecrosis of the femoral head is common. Lumbar spine involvement with osteonecrosis and collapse with secondary sclerosis contributes to leg length discrepancy. Intraoperative blood loss which can be anticipated in patients with high difficulty index should be minimized as possible and blood transfusion is given as necessary. Sickle cell anaemia vascular necrosis involves hip joint early in patients life and with improvements in sickle cell anaemia patients survivor it is highly possible hip replacement will be required early than anticipated as well as revision surgeries than in other conditions [5]. Presentation of bony infarct without loss of congruency which accounts for the majority of cases pain is due to infarcts with intraosseous compartmental pressure of the

femoral head and may benefit from core decompression [6]. Hips become complex because by the time they eventually present, the pathology has worsened due to weakness, stiffness and lack of motivation.

Duration of surgery comparable to time required for difficult primary hip and our study matches what other authors established [7]. Technical difficulties encountered in the course of the surgery related to extent of local changes of the disease with stiffness, femoral canal obliteration and acetabular bone stock variation from cysts, sclerosis and protosio in rare cases. Intraoperative blood loss in our study varies according to these technical difficulties and thus the need for more blood transfusions. Compared to works on primary THR where mean operative time of 89 minutes [14] and 123±28 minutes [15], duration in our study is higher than expected. Mean intra operative blood loss of 1600 ml is higher than 1090 ml, 984 ml and about 371 ml respectively reported in primary THR [ 8,9 & 10].

Perforation or fracture is of either the acetabulum or the femur. The patients that had femoral perforation did not need any further intervention. The canal was eventually located and stem bypassed the area of perforation. Acetabular perforation occurred in a sickler and a young man with steroid induced AVN. The perforation occurred during reaming due to extensive irregularity and weak acetabular floor respectively. Al-Mousawi reported acetabular perforation, femoral perforation and fracture similar to our report in sicklers [11].

**Figure 1: Pelvic plane x-ray**



**A** **B**  
**A (advanced bilateral avascular necrosis beyond the hips)**  
**B (unilateral avascular necrosis isolated to the left hip)**

**Figure 2: Scoliosis of the spine secondary to avascular necrosis contributes to leg length discrepancy**



**Figure 3: Presence of soft tissue hip and knee contractures**



## Conclusion

The primary total hip in sicklers should be prepared well; we think categorizing all sickle cell hip avascular necrosis in one group is unfair for patients and surgeons. While some of these patients can be dealt with low difficulty others have higher difficulty, index indicated by the presence of the musculoskeletal changes other than those restricted to the hip joint. Careful history, physical and radiological examination exploring these factors of poor outcome must be exercised to differentiate between the two groups. Strict pre-operative optimization and in terms of implants one should have a well-equipped armamentarium for hip replacement. Sickler hips with high difficulty index is challenging technically; difficulties should be anticipated and dealt with to avoid high incidence of intraoperative complications, increased operation time and blood loss. The complexity of primary hip replacement in sickle cell disease patients is common and poses a surgical challenge. The main limitation of this study is that it is a single center study; multicenter future studies are needed to increase the number of cases in this specific condition. We recommend routine categorization of these patients into low and high difficulty index groups as well as multidisciplinary approach in preoperative optimization for patients for surgery and if possible to have this procedure done in an experienced center familiar with its technical difficulties and ready to deal with complications in case they occur. The use of image intensifiers to deal with the femoral canal preparation in case of its obliteration to avoid canal violation and to seat the acetabular cup during acetabular reaming can avoid violation of medial wall. Patient understanding of the effect of remote manifestation away from the hip such as vertebrae infarct and lower limb joints and soft tissue contracture on outcome helps alleviate the unpleasant high expectation feelings of the patients and gathers efforts toward maximum cooperation among all members of the multidisciplinary treating team.

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