



CASE REPORT

A 3D Printed Total Hip Arthroplasty in Adult with Sickle Cell Disease: A Case Report

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Abstract

Sickle cell disease (SCD) is an inherited blood disorder that distorts the red blood cells into a sickle-shaped. These deformed cells can interrupt the blood vessels supply to the bones and lead to Avascular Necrosis (AVN). Femoral head is the most vulnerable bone owing to vascular damage.

In patients with SCD, the condition progresses rapidly, the femoral head collapses and causes severe pain and limited range of motion. In the advanced stages of the femoral head necrosis where there is no existing standard medical management, the orthopaedic surgeons conventionally recommends Total Hip Arthroplasty (THA). This option is challenging and carries a high risk for serious complications both peri-operatively and post-operatively.

The present report illustrates a case of pre-operative 3D planning for ideal bone geometry and personalized care before, during, and after THA in a young adult which was a competent technique for the management of SCD.

Keywords: Articular range of motion; Femur Head Necrosis; Hip Replacement Arthroplasty; Sickle Cell Anaemia.

Introduction

Sickle cell disease (SCD) is a genetically transmitted disease, which features abnormally shaped red blood cells, that leads to a transient occlusion of the blood vessels. Femoral head Avascular Necrosis (AVN) is one of the commonest skeletal complications of SCD. Vaso-occlusive events in blood vessels that cross the femoral head causes bone infarction in the subchondrial regions where there is less collateral

circulation, gradually this leads to joint destruction and limited mobility, along with acute and chronic disabling pain.¹

Before the 1960s, the surgical options for advanced hip osteoarthritis were limited to osteotomy, arthrodesis, or excision arthroplasty. However, in the advanced stages of the disease, these options had limited outcomes and have led to multiple functional disabilities.²

Replacing the hip joint helps in alleviating the pain, promotes hip function, and the range of motion and thus improves the quality of life.³ Total Hip Arthroplasty (THA) can be followed for SCD cases which progresses over a period of five years, ranging from 42 months for (stage 1) femoral head AVN, and 30 months for (stage 2).³

SCD is relatively common in the Middle Eastern Arab countries, including Bahrain, and the eastern province of Saudi Arabia.⁴ In 1984–1985, the prevalence of SCD in Bahrain was estimated to be 2.1% of the total population, but declined to 0.9% in 2005.⁵ However, limited studies have been conducted in Bahrain concerning this group of patients, and there is a paucity of literature about the prevalence of THA among SDC patients.

Two studies were reported in 2002 and 2004 by Dr. Faisal Almosawi and his colleagues,^{2,4} describing the post-operative medical and orthopaedic complications and poor prognosis (infection and osteonecrosis).

Though conventional protocols of THA are associated with complications, cemented implants demonstrate a higher osteolysis rate followed by implant loosening requiring revision surgery, and uncemented implants pose a higher risk of intra-operative complications.⁶ The technical difficulties during the surgery are; perforation, fracture of the femoral shaft and excessive blood loss (greater than two litres). This kind of complications were expected due to the long-standing deformities, poor quality soft bone, and presence of sclerosis; which may obliterate the femoral canal making its preparation extremely difficult and hazardous.² Furthermore, the bones in these patients are usually smaller since sickle haemoglobinopathies affect the growth and development. Orthopaedic surgeons stated that soft tissue release and preparation of the acetabulum and the femur may significantly increase blood loss and prolong the operative time.⁷

As the morphology of femoral bones of SCD patients sometimes does not allow the insertion of a standard cemented or cementless stem, customized 3D printed hip implants promise an ideal insertion of the stem, limiting the risk of complications.

A continuous series of 232 THAs performed with a custom cementless femoral stem in patients aged less than 50, followed-up for twenty years, confirmed that the THA using this custom-designed stem can provide excellent clinical and radiographical outcomes. The individual 3D femoral stem and prosthetic neck restored extra and intra-medullary functional anatomy in this young and active cohort.⁸

The introduction of customized hip arthroplasty with 3D printing will enhance the surgical planning and benefit not only the surgeons but also the patients.

Case presentation

A twenty six-year-old female patient, presented to a private hospital out-patient department in October 2018, reported with the chief complaint of limping and worsening of right side groin pain. The pain began when the patient was about thirteen years. The pain was intermittent and sharp, increased in intensity, and affected her mobilization during daily activities. At the age of twenty-four years, she underwent conservative treatment including physiotherapy which with focused on exercises to maintain the joint mobility and strengthen the muscles around the affected joints, but it resulted in temporary relief only.

The patient was a known case of SCD. The patient had received blood transfusion at the age of thirteen, which presented with vaso-occlusive crisis (VOC) at a range of one episode a month. The most recent crisis that required hospital admission was in June 2018. Regular medications were as follows: Folic Acid 5mg, one tablet per day, Ibuprofen 200mg and Paracetamol 1gm, when required. On physical examination, the patient was alert, oriented to time place and person, antalgic gait with limping on the right side. Vital signs were within the normal range as follows: blood pressure 110/73mmHg, heart rate 86 beats/minute, temperature 37.2°C, respiratory rate 16 breaths/min, oxygen saturation 99%. The body mass index range was normal (19.8 kg/m²). The cardiovascular examinations, Electrocardiography, and Doppler were well within the normal limits. Trendelenburg test was negative for the lower limbs, Thomas test revealed limited hip

flexion 90 degrees right side, along with restricted external rotation and internal rotation 0 degrees of the right side. On comparison of both limbs, right side leg length discrepancy of 3cm was observed, and the neurovascular examination was well within the normal limits.

Radiographic evaluation of both the hips was done, the right hip joint revealed severe deformation of the femoral head which was impinging on acetabulum (AVN), compared to the left hip, which showed an early stage of the disease (Figure 1).

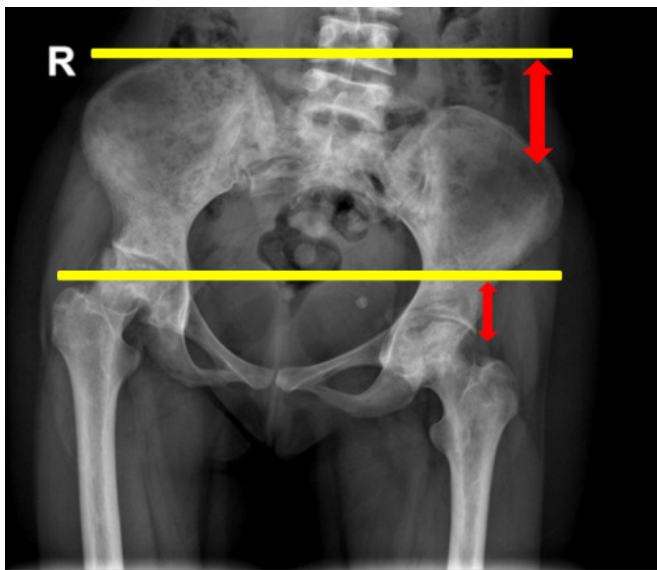


Figure 1: Pre-operative bilateral pelvis digital X-ray image, impinging on the right acetabulum (AVN), compared to the left hip, with an early stage femoral head flattening

After consideration of the physical examinations and x-ray findings, the final diagnosis and plans had been made to perform THA. Computerized Tomography (CT) was done to proceed with the 3D design plan of the stem. The CT report showed that the cancellous bone density was similar to the cortical bone density 700-8000 HU. Planned correction of Patellofemoral contact point (CP): +11mm, Mediolateral (ML): -3mm. The data from the 3D scan, was analysed using 3D planning software (HipPlan) used to plan the operation on a computer, which was then dispatched to the Symbios headquarters in Switzerland, a company specialized in manufacturing 3D implants. The detailed reports provided by Symbios showed the density of the bones, and fit of the implant inside the bone, to avoid the weak bone areas. Custom

hip cementless implant was commissioned with the following details: 2/3 partial Titanium and full Hydroxyapatite coating, Acetabular shell size was 46, Acetabular Liner was size 46 / 28mm/10 Degree, and the Delta Head was 28mm/-3.5mm.

Procedure and anaesthesia consent was obtained from the patient to proceed for the surgery, besides an informed consent to utilize her data anonymously in this case report.

Laboratory studies were done pre-operatively, haemoglobin level was 11.8gm/dL, Serum Creatinine was 20.9 U/L, haemoglobin electrophoresis showed sickle cell anaemia (HbSS), Hgb S 76.1%, Hgb F 21.3%, Hgb A 22.6%. One Pack of Red Blood Cell (PRBC) (O+) was transfused pre-operatively as a preventive exchange transfusion, to reduce the concentration of sickle cells without increasing the haematocrit or whole-blood viscosity.⁹

One day before the surgery, the patient was optimised by adequate hydration and blood exchange transfusion. Additionally, for the prevention of deep vein thrombosis (DVT), she received an injection of Enoxaparin Sodium 40mg subcutaneously, 12 hours before the operation. Management of hypothermia started in the pre-operative phase using intravenous fluid warmer and continued into the post-operative phase with a forced-air warming blanket.

THA right hip (customized stem) was done using cementless implantation in a posterolateral approach with no muscle incision, under intravenous general anaesthesia. The estimated blood loss during the operation was approximately 700ml, no hemovac drain was used. One PRBC was transfused intra-operatively and two packs post-operatively were indicated. Intravenous fluids (Lactated Ringer) were given at the rate of 100 ml/hour to maintain adequate hydration. Supportive humidified oxygen was given 2 liters via nasal cannula for 24 hours post-operatively, oxygen saturation was $\geq 99\%$, and incentive spirometry started as soon as the patient was awake.

The patient was able to mobilize on the first post-operative day, with immediate full weight-bearing, and was permitted to lie on the operated side.

However, crossing of the operated leg and internal rotation were prohibited. By the day of discharge, she was able to walk with one elbow crutch, use high and low stairs, transfer in and out of bed independently, and sit on a chair for prayer. Comparatively, patients with normal THA, mobilization usually starts on the second postoperative day due to pain and discomfort, despite the analgesics.

Pain visual analog scale (VAS) score was mild to moderate in nature; therefore the stronger analgesics were avoided. For post-operative pain management, transdermal Fentanyl patch 25mcg was applied on the day of the surgery and continued for 72 hours, besides intravenous Paracetamol 1gm every 8 hours and tablet Etoricoxib 90mg orally once daily.

As for DVT prevention, she continued with anti-embolic stockings and blood-thinning medications, Dabigatran 220 mg once daily for 14 days post-operation. The patient had a smooth recovery with no post-operative complications. The wound was healthy, edges were intact, no exudates. Normal healing was observed, and 90 days surveillance post-operatively showed no evidence of surgical site infection.

Pelvic x-ray images were taken on the second post-operative day and after six weeks, and was evaluated by the chief surgeon, no signs of implant loosening or early dislocation was observed (Figure 2). Harris hip score was used to assess the surgery outcome and a score of “excellent” (90 points) was observed.

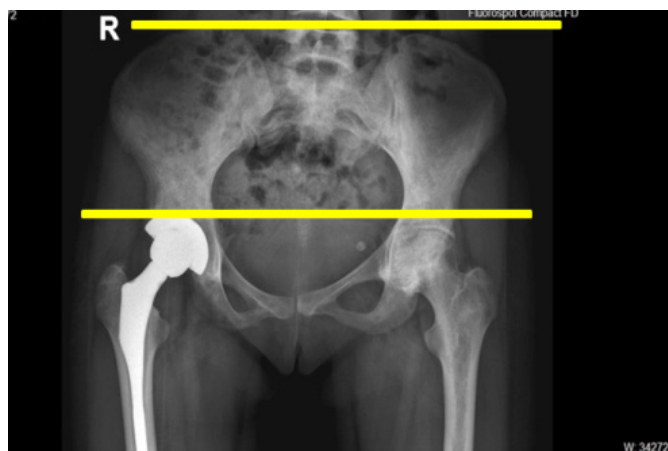


Figure 2: Second post-operative day: Antero-posterior digital x-ray image of patient’s hip, the femoral stem with the fixed head articulates with the native acetabulum, neutral alignment with the longitudinal axis of the shaft, and corrected length discrepancy of < 1 cm

Discussion

AVN is a major complication seen in SCD patients; it starts with phases of healing and infarction ultimately leading to osteoarthritis, which could lead to functional disability. Classically these patients are relatively younger age, than the typical osteoarthritis patients.⁴

We reported this case to highlight the importance of pre-operative planning for ideal geometry and fit of the implant among patients with a femoral head deformity like SCD patients, to help preserve the hip mechanics and restore the leg length. A review of the literature revealed that this was a pioneer case report of a 3D customized hip arthroplasty, performed for a sickle cell patient in Bahrain.

The patient presented with a 3cm length difference along with limping and pain. Hip conservation procedures in this case would have been ineffective and arthroplasty was the only option. Challenges suspected to occur during the operation were related to the quality of the bone, besides that if the pre-operative shortening was more than 3 cm, complete equalization of leg length could have caused a high risk of the sciatic nerve irritation.⁶

The implant could have settled in a very different position than planned or may have posed difficulties in insertion or stability. Considering these outcomes, a meticulous pre-operative treatment plan for stem insertion was outlined. Thus, a 3D customized stem was recommended (Figure 3).

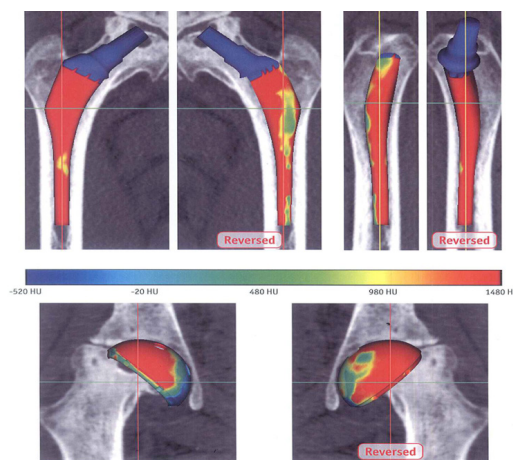


Figure 3: CT images of the right hip, and shows the Hounsfield scale of implants density, areas with (<400 HU) are associated with a higher risk of implant failure whereas normal bone (400–1,000 HU) represents a safe zone (400–1,000 HU) represents a safe zone

Table 1: Comparison between conventional Total Hip Arthroplasty (THA) and 3D customized THA

	Conventional THA	3D Customized THA
Average operation duration (minutes)	67	134
Average blood loss(ml)	300	500
Average length of hospital stay (days)	≥6	≤6
Mobilization with full weight bearing	2nd post-operative day	1st post-operative day
Average pain visual assessment scale (VAS)	Moderate-severe	Mild-moderate
Analgesics demand	Patient controlled analgesia narcotic	Non steroidal anti-inflammatory drug
Leg length difference	Required custom made foot orthotics	Corrected

As observed during the general physical examination and radiography, the leg length discrepancy decreased post-operatively and a marked improvement in the gait was observed.

The hospital performed 54 THA surgeries from 2014-2019, of which 26(48%) were females. Among these patients, 12 (22.2%) were females with sickle cell disease and AVN femoral head, with an average age of 38 years. The same surgical procedure was followed for four female patients with SCD who underwent THA, using the same model of the implant by the same surgeon, which were studied and compared (Table 1). Surgeries conducted by different surgeons and with different implant models were excluded from this compilation (eight surgeries).

The traditional THAs are performed in a shorter operation time and with less blood loss. On the other hand, length of hospital stay is lengthened along with a pain score of moderate to severe. Current strategies for severe and acute pain management typically includes an opioid given by Patient-controlled analgesia (PCA).¹⁰

Conclusion

SCD is a common disease among the Middle Eastern Arab population, which could lead to femoral head AVN as a complication among this group of patients, which eventually requires a THA. Pre-planning for ideal geometry and fit of the implant among the patients with a femoral head deformity helps to preserve the hip mechanics and restores leg length.

Even though custom-made prostheses are more expensive than standard stems, their use offers a reasonable chance of improved results compared to what would be obtained using standard stems.

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