International Journal of Orthopaedics Sciences

ISSN: 2395-1958
IJOS 2018; 4(3): 672-676

www.orthopaper.com
Received: 29-05-2018
Accepted: 30-06-2018

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Avascular necrosis of the femoral head due to sickle-cell disease in tribal population around Vishakhapatnam district, treated with core de-compression and non - vascularised fibular graft

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DOI: https://doi.org/10.22271/ortho.2018.v4.i3l.119

Abstract
Sickle-cell disease (SCD) is the most common cause of avascular necrosis (AVN) of the hip in childhood. It results in significant physical impairment and chronic pain, and ohen progresses to require hip replacement. Conservative therapy is ineffective. We evaluated whether core decompression can arrest progression of AVN. We performed 10 core decompression procedures in 6 patients with SCD and AVN. Patients ranged from age 12—18 years at diagnosis (mean, median age, 14 years); Six hips were stage II, Four hips were stage III, and. follow-up on these patients was 1.2 years. Efficacy of the procedure was evaluated by clinical improvement in pain, radiographic progression, and need for further surgery. All 6 stage II patients had substantial improvement in pain, and only one showed X-ray progression. Both stage III patients progressed on X-ray, but one was clinically improved. One patient was operated with Non-Vascularised Fibular Graft. Our results demonstrate that in early AVN, core decompression was beneficial for almost all patients, even with progression on X-ray. Core decompression should be considered in the management of SCD patients with early AVN. But in our study, only one pt was operated with Non-Vascularised Fibular Graft

Keywords: Sickle-cell disease, avascular necrosis, core decompression, non-vascularised fibullar graft

Introduction
Sickle-cell disease (SCD) is the most common cause of avascular necrosis (AVN) of the hip in childhood [1]. The overall prevalence of AVN in SCD is at least 109c, and homozygous SCD patients with concomitant alpha thalassemia are at particular risk, with a prevalence of >20% [2, 3]. The risk of developing AVN does not disap- pear with age, and by age 35 years nearly half of all homozygous SCD patients will have developed AVN. Avascular necrosis frequently progresses to total collapse of the femoral head, necessitating hip replacement [1, 4, 5]. Unfortunately, sickle-cell disease patients have had par- ticularly poor results with hip arthroplasty, with up to 59% failing within 6 years [6-8]. As the life expectancy of SCD patients increases, the morbidity of AVN will become increasingly important.

Avascular necrosis of the hip can often be detected at an early stage, and there is considerable interest in thera- pies to halt progression of AVN [9-11]. Unfortunately, conservative management, such as prolonged nonweight- bering or transfusion therapy, is not effective in preventing progression in most cases [12, 13]. Core decompression of the femoral head is an alternative therapy which has been reported to reduce pain and prevent progression of AVN [4, 14, 15]. There are, however, no reports of its use in children or solely in patients with SCD. We report the results of core decompression in SCD children afflicted with AVN of the hip.

Methods
Eight adolescent with SCD underwent core decompression in 10 hips. All patients are followed regularly at the Anil Neerukonda Hospital, NRI Institute of Medical Sciemces. Vishakapatnam. AP. (Affiliated To NTRUHS.A.P) and had their diagnosis of SCD confirmed by standard electrophoretic techniques. Eight patients had hemoglo- bin (Hb) SS, one patient had Hb SC, J30 thalassemia. Patients ranged from age 12—18 years (mean, median age, 14 years), and there were 7 males.

Patients were eligible for core decompression if they met the following criteria

1. severe pain localized to the hip or inguinal area requiring hospitalization with intravenous narcotics,
2. physical examination findings of pain and limitation on movement of the affected hip, and
3. Radiographic or Magnetic Resonance Imaging (MRI) findings diagnostic of AVN. After diagnosis with AVN, bed rest was recommended but not consistently followed. Patients had not received any other treatment for AVN (e.g., hydroxy rea or chronic transfusion) prior to the core decompression procedure. and most of the patients where Anaemic, where we Tran fused blood to improve the anaemia.

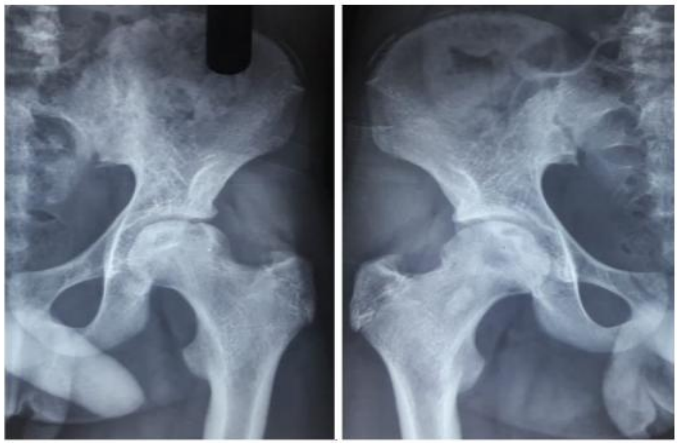


Fig 1: Radiological Staging

Diagnosis and Staging

All patients had a radiologic evaluation consisting of anteroposterior (AP) and frog-leg views on plain radiography and MRI after complaining of hip pain consistent with AVN. Conventional criteria were used to establish a diagnosis of AVN on plain radiographs, including sclerosis, increased radiolucency, and changes in femoral head shape [5]. When plain radiographs were normal, the diagnosis of AVN was made using MRI, which demon- strated increased signal intensity in the femoral epiphysis. Sixteen hips in 10 patients were found to be diagnostic of AVN. Six patients had bilateral disease, 3 had right- sided disease, and one had left-sided disease. Core decom- pression was performed in the 13 symptomatic hips, and these were staged according to the Ficat system [5]:

Stage I: Normal plain radiograph, abnormal MRI.

Stage II: Sclerosis and lytic areas on radiography.

Stage III: Flattening of femoral head.

Stage IV: Collapse of femoral head, joint space nar- rowing.
At time of surgery, six hips had stage II disease, four had stage III disease

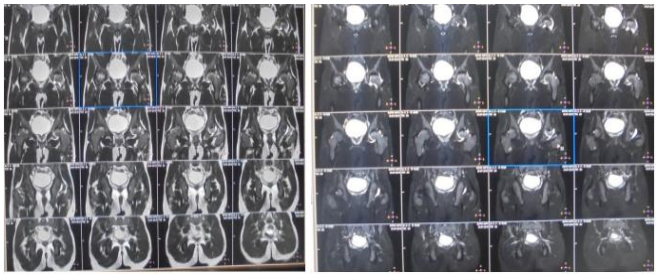
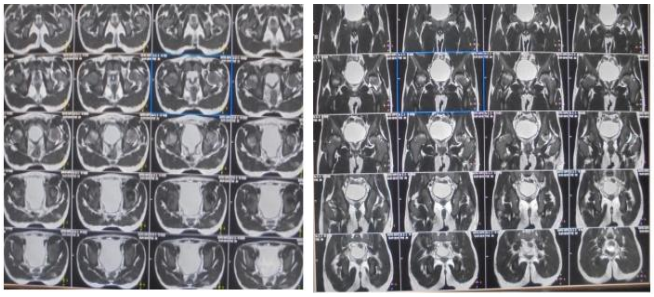


Fig 2: MRI Scanning: Staging of Avascular Necrosis



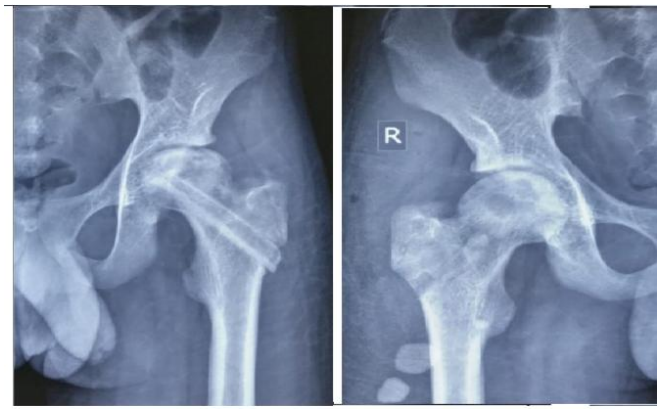
Operative Procedure

Patients were admitted the day before the scheduled coring procedure and received intravenous hydration and red blood cell transfusion, following a standard protocol for SCD patients undergoing elective surgery [16]. Core decompression was performed according to a previously described procedure [151 Briefly, in this procedure a guide pin is inserted into the femoral head from below the trochanteric ridge and directed toward the necrotic area.

A. Incission B. Guidewire, C. Core De-Compression

Fig 3

Correct placement of the guide pin is established with two-plane imaging, and then an 8-mm hollow coring device is used over the guide pin to remove a core of medullary bone. Intramedullary pressure measure- ments and biopsies were not regularly performed. Patients were discharged with instructions to complete 2 weeks of strict no weight-bearing followed by increasing activ- ity as tolerated. No other specific treatments, such as antiinflammatory agents or hydroxyurea, were used post- operatively.



A. Fibular Graft in Situ B. Core De-compression

Fig 4

Follow-Up Evaluation

After surgery, patients were seen for a follow-up visit at 2 weeks with the hematology service, and again at 6 weeks postoperatively by the orthopedic service. Thereafter, patients were followed with routine comprehensive health care visits every 6-12 months. Patients were evalu- ated by three criteria: clinical improvement in pain and activity, radiographic progression, and need for further surgery. Clinical efficacy was scored using a four-point scale assigned on the basis of patient report and physician examination according to the following criteria: 1) patient pain free and no limitation of activities or range of motion on exam, 2) much improvement in pain and minimal limitation in activities or range of motion, 3) little or no change in pain, significant limitation of activities or range of motion, and 4) worse pain, more limitation in activities or range of motion. Radiographic evaluation was made approximately every 6-12 months, or more frequently if the patient developed symptoms suggestive of progres- sion. Radiographic follow-up protocol was for plain radiographs on all patients and, in most cases, MRI as well. The decision for further surgery was based on severity of pain and impairment of patient activity: specifically, continued dependence on narcotics and lack of ambula- tion (unable to bear weight on affected limb).

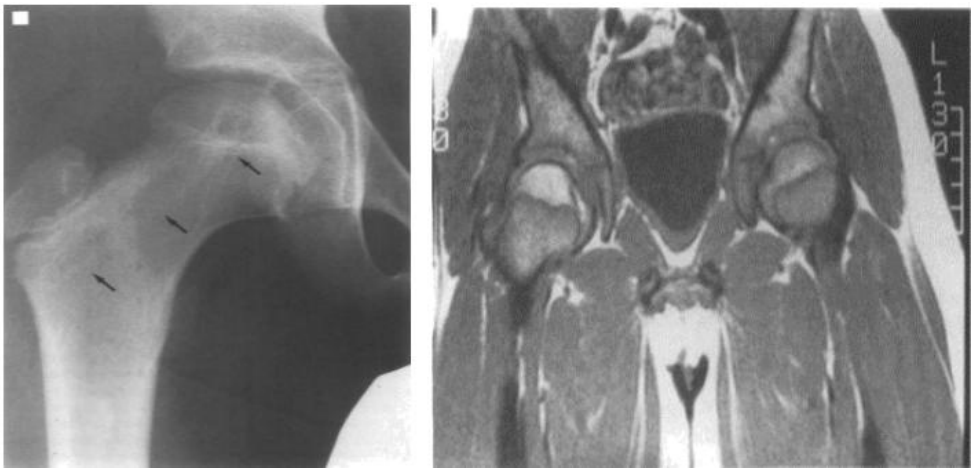


Fig 6: Radiographic appearance of avascular necrosis of the hip before and aher core decompression. Plain radio- graph (lower leh) demonstrating stage II avascular necrosis, with mild sclerosis of right femoral head (arrows), and MRI (upper left) with increased, asymmetric uptake in the right femoral head. Right: Same patient 1 year later, after core decompression, with stable disease and prominent core de-compression track (arrows).

Table 1: Results of Core Decompression in Patients with Sickle-Cell Disease

Patient	Age/sex	Stage	Follow-up	Clinical score	X-Ray progress
1	14 yrs/M	II Stage	10 months	2	Yes -IV
3	16 yrs/M	II Stage	12 months	2	No
4	10 yrs/M	II Stage	11monyhs	2	No
5	10 yrs/M	III Stage(L)	14 months	1	No
6	15 yrs/F	II Stage(R)	11months	3	Yes-IV
7	18 yrs/M	III Stage	10months	2	No
8	13 yrs/F	II	10 months	3	No
9	16 yrs/M	III Stage	14months	1	Yes IV
10	13 yrs/M	III Stage	14 months	2	Yes IV

Stage at time of core decompression; R, right; L, left. 'Postoperative results as of last follow-up visit. Clinical score: 1, no pain or limitation of mobility; 2, substantial decrease in pain and increased mobility; 3, little charl8• ' n pain or mobility; 4, worse pain or more limitation in activities.

Results

Stage II

All five hips had a significant reduction in pain (clinical score = 2), although none was pain-free (Table I). The most common remaining complaint was of vague aching in the joint which did not require regular narcotic use or hospitalization. Limitation in activity and range of motion was not seen in the absence of pain. Four of the 5 patients' disease was stable on radiologic evaluation. One of the 5 patients progressed on plain radiography to stage IV disease over the 18 months following core decompress- sion. Despite progression, this patient reported substantial improvement in pain and mobility. No patient has required further surgery. Clean follow-up for this group is 2 years and 8 months (median, 3 years, 3 months; range, 9 months—3 years, 5 months).

Stage III

Five of the six hips (83%) had significant improvement in pain (clinical score = 2) as with stage I patients, and the remaining patient reported little change. Four of the 6 patients showed no radiographic progression of disease. The remaining 2 patients progressed on radiography, one to stage III and one to stage IV. The patient who pro- grassed to stage HI had improvement in pain, but the patient who went on to develop stage IV disease reported no change after core decompression. None of these 6 patients has needed further surgery, and mean follow-up

Stage IV

Both hips in this group progressed on plain radiographs. One had significant clinical improvement despite X-ray progression, and the other was clinically unchanged. Nei- ther patient has needed further surgery.

In analyzing all of the groups together, radiographic progression was not consistently associated with change in symptoms after the coring procedure. Three of the 6 patients (60%) who progressed on plain film had substantial clinical improvement, with reduction of pain and increased mobility. Clinical failure, however, did correlate with radiographic failure with both patients who failed clinically, also showing radiographic progression.

Complications

Surgery, in general, was well-tolerated, and patients recuperated quickly. Postoperatively, one patient devel- oped a wound infection and another developed an acute chest syndrome which did not require transfusion. A third patient with a history of severe auto- and alloimmuniza- tion was discharged after 4 days but was readmitted 3 days later for a severe haemolytic transfusion reaction due to receiving incompatible blood. Estimated blood loss was minimal in almost every case and less than 10 ccl kg in all patients. Mean length of hospitalization was 7 days (median, 6.5 days). There were no subtrochanteric fractures and no growth disturbance in the 3 patients with open epiphyses at time of surgery.

Discussion

Avascular necrosis of the hip results from severe isch- emia of the medullary bone. Blockage of the osseous circulation results in necrosis and in an increase in intra- medullary pressure, which then causes further vascular compromise [5, 17]. Without treatment, AVN often prog- resses to total collapse of the femoral head. Core decom- pression was originally reported by Ficat and Utheza [18] to prevent progression of AVN when performed early in the disease. In this procedure a core of bone is removed from the femoral head, reducing intramedullary pressure and thereby preventing further vascular impingement and allowing for new bone formation [5, 15]. Stulberg *et al*. [14] recently reported a randomized trial comparing core decompression with prolonged nonweight-bearing. They documented that 70% of patients with early AVN treated with core decompression showed improvement, com- pared with only 20% of those treated with prolonged bed rest. Twenty-eight percent of patients treated with core decompression needed hip replacement, as compared to 1509c in those treated no operatively. Other series have not had as much success with core decompression, and have reported complications such as sub trochanteric frac- tures from the procedure [19-21]. It is difficult to compare these different reports, as there were no separate analyses by aetiology of AVN and no standard criteria to define efficacy of the procedure.

We have attempted to perform core decompression in all SCD patients with early AVN, and our results docu- ment that core decompression was effective for almost all patients with stage II or III AVN. Many of these patients were nearly pain-free after the coring procedure. In this series, 2 patients underwent core decompression when their disease was relatively advanced (stage III). With only 2 patients in this group it is not possible to determine if core decompression is beneficial in patients with ad- vanced AVN. Our results, however, do concur with other series and indicate that stage III disease may be too late for intervention [14].

In our study, the patient's report of reduced pain and increased activity was chosen as the primary determinant of efficacy. It is interesting that although 10 of 11 patients with early disease had significant clinical improvement in pain and activity, 3 of 11 (27%) progressed on plain radiography. This discrepancy between clinical and radio- graphic findings underscores the need for uniform criteria to define efficacy.

Because of the morbidity associated with hip replace- ment and the historically poor results of hip replacement in SCD, the need for further surgery is an important secondary endpoint. Though the length of follow-up is relatively short, no patient treated with core decompression has required further surgical intervention. Core de- compression is a much simpler procedure than hip arthroplasty and patients, generally, do not experience the more serious complications seen with total hip replace- ment. The only reported complication of core decompress- sion has been intra- or perioperative fractures of the femur. Total hip replacement has a high rate of associated throm-botic events, and patients often require a large number of transfusions. Sickle-cell disease patients are at particu- lar risk of all immunization, and the Preoperative Trans- fusion study in SCD [16, 22] documented increasing rates of all immunization as the number of units transfused in- creased.

Progression on X-ray was the third endpoint used to evaluate patients. The Cooperative Study of Sickle Cell Disease reported that radiographic findings in AVN were not a good indicator of clinical symptoms (2). Similarly, radiographic progression was often not associated with clinical failure in our patients, as 60% of those who had X-ray changes had significant improvement in pain and mobility. Progression on plain radiographs may be im- portant, however, in long-term outcome, and extended follow-up of these patients is needed to answer this question.

All our SCD patients with early AVN underwent core decompression and, therefore, the expected outcome with conservative management alone cannot be determined. Natural history studies in SCD children with AVN docu- mented that 80% remain symptomatic from diagnosis, and continue to have pain with follow-up as long as 18 years [1, 2]. Despite the risks and high failure rate, almost one third of SCD patients with AVN will undergo hip replacement [1, 2]. From these historical data it appears that the short period of bed rest after the coring procedure was probably not responsible for our patients' improve- ment, and supports our conclusion that core decompression improves the outcome of AVN in SCD. Due to the increased morbidity of surgery in SCD patients, however, a prospective, randomized trial is needed to better define the natural history of AVN in SCD, and to definitively recommend core decompression over more conserva- tive therapies.

In summary, core decompression was effective in re- ducing pain and increasing mobilty in SCD patients with stage II or

III AVN. Symptomatic relief of pain often did not correlate with radiographic findings, as several pa- tients had clinical success but progressed on radiography. Core decompression was associated with few complica- tions, and no patient has required furhter surgery. How- ever, due to the lack of natural history data and the risks of surgery in patients with SCD, a prospective, random- ized trial is needed to definitively establish the benefit of core decompression in SCD patients with AVN.

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