

Core Decompression For Treatment of Avascular Necrosis of Femoral Head In Children With Sickle Cell Anemia

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Abstract

Sickle cell anemia (SCA) is a hereditary hemoglobin disorder that results in sickling of the red cell, leading to chronic hemolytic anemia, vascular occlusion and progressive organ damage.

Avascular necrosis (AVN) is a known and common complication in sickle cell anemia. There are very little studies addressing core decompression of femoral neck in children with SCA. Many studies are done for adult patients. We did core decompression (CD) of femoral neck in children with AVN due to sickle cell anemia. 39 hips in 29 patients are done between 2008 to 2018. The patients age ranges between 4 to 14 years of age. AVN is classified according to Ficat radiological classifications. Stage 1, 2A, and 2B are operated. Stage 3 and 4 hips were operated if the other hip is stage 1 or 2. We found a significant improvement of the operated patients compared to non-operated patients. Patients were assessed in follow-up in form of pain, gait, activity and radiological grading. Core decompression is done using a percutaneous cannulated drilling that do not cross the growth plate of the head.

Excellent results were found in 87%, good results in 9% and poor results in 4%. We recommend doing core decompression in all grades of AVN in sickle cell anemia children.

Conclusion

Core decompression in children with sickle cell anemia is a new surgical intervention. It is a simple surgical procedure that can be done by most of pediatric orthopedic surgeons. We found that core decompression is an effective method to stop and improve clinical and radiological stages of AVN of femoral head in children sickle cell anemia. We found out that core decompression in children has far much better outcome compared to adult's core decompression.

Introduction

Sickle cell anemia is a common hematological disorder that presents in the Eastern province of Saudi Arabia. The prevalence for sickle-cell trait was approximately 21% and for SCA was 2.6% [1].

Avascular necrosis is a well-known complication that occurs in sickle cell disease patients. Eastern province has a 27% risk of avascular necrosis of the femoral head compared with 8% to 12% in the African type [2,3].

Avascular necrosis of the head of femur can affect both children and adults. It is seen in both sickle cell trait and sickle cell anemia.

Incidence of AVN in SCA Minlner et al,1991 (USA) is 9.7%, Bishop,1994 (USA) is 19%, Bishop,1995 (USA) is 31%, Adekile AD, 2001(kuwait) is 26% , N'Dirk etal, 2000(France) is 30% based on adult patients CT scan of hips, Mukisi-Mukasa et al,2000(Guadeloupe) is 37% of adult patients having stage 1.

Sickle cell anemia patients with AVN may present with hip pain, abnormal gait, shortening of lower limb and in late cases they may present with pathological fracture [4-8].

It starts with ischemic necrosis of the bone of the femoral head due to obliteration of blood supply by sickling red cells and its process.

Material and Methods

We prospectively studied core decompression (CD) of the femoral neck between 2008 to 2018. Screening is done at the children hospital by the same hematologist. Suspected cases are referred to pediatric orthopedic clinic. Randomization is done at the outpatient department (OPD) appointment desk where random letter A or B is generated at appointment print. Letter A will go for core decompression and letter B will be treated as usual by supportive treatment. Patients refusing surgery or refusing to en-

roll in the study were eliminated from the study. Proper consents were obtained.

322 children with sickle cell disease were screened and referred to pediatric orthopedics. AVN was found in 59 patients Age of all children groups was ranging between 4-14 years. Average age is 8.54 years and standard deviation of +/- 2.2. Core decompression (CD) was done for 29 patients with 39 affected hips, Control group is the No core decompression (NO CD) group. They will be treated by supportive treatment like activity limitation, use of crutches and use of analgesics. In control group there were 30 patients with 38 hips (Table 1). Mean follow up for control group was 68.8 months with minimum requirement of 24 months. Longest follow up was 96 months STD 22.81. Harris score for Pain, activity, walking distance, ROM and gait). Plain x-ray AP and frog lateral pelvis views are done at each visit

Table 1: Demographic distribution of patients

	Male	Female	total
AVN +	37	22	59
AVN -	145	98	243
Total	182	120	322

In Pediatric hematology clinic, all patients do initial hip AP and frog lateral plane x-rays. Hip MRI is done for suspected or affected hips. The x-rays and MRI are read by the same pediatric radiologist.

(Table 2). MRI staged according to Enneking's Stages of Osteonecrosis (Table 3). Ficat Stage 0 and 1 are confirmed by MRI. Stage 1, 2A, and 2B were operated. Stage 3 and 4 hips were operated only if the other hip is stage 1 or 2 and planned for surgery. Bilateral AVN is operated at the same time.

AVN is classified according to Ficat radiological classifications

Table 2: Ficat radiological classification

Stage	Clinical	Features	Radiographs
0	Preclinical	0	0
I	Preradiographic	+	0
II	Precollapse	+	Diffuse Porosis, Sclerosis Crescent Sign
III	Collapse	++	Broken Contour of Head Certain Sequestrum, Joint Space Normal
IV	Osteoarthritis	+++	Flattened Contour Decreased Joint Space Collapse of Head

Table 3: Enneking's MRI Stages of Osteonecrosis

Stage	Pain	Radiographs
I	None	Slight increased density
II	None	Reactive rim
III	Occasional	Crescent sign
IV	Limp	Step off flattening
V	Continuous	Collapse
VI	Severe	Deformed

For CD group, the patient is given a semi elective admission. Simple transfusion regimen is used for all case. No patient had needed a blood exchange transfusion before or after surgery (Figure 1 and Figure 2).



Figure 1a: Patient with stage 2b of left hip and 3 in right hip,

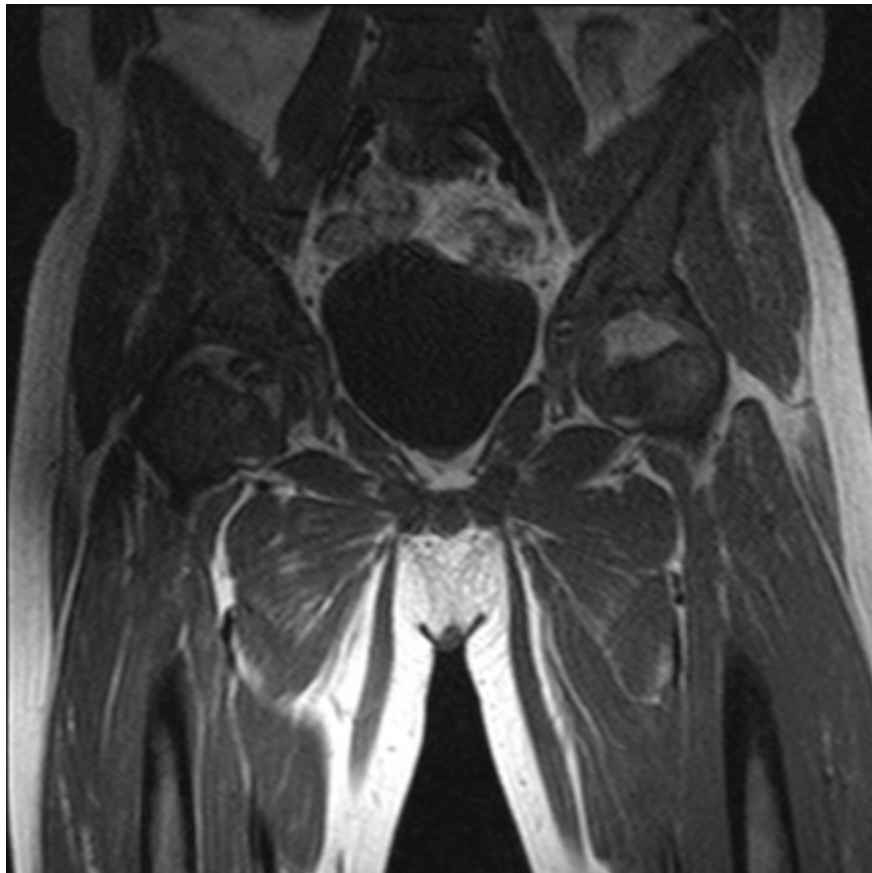




Figure 2b: MRI and c: post op

Core decompression surgery is done under general anesthesia on a fracture or translucent table. Percutaneous smooth wire is inserted under image intensifier into the center of femoral neck and advanced into the head of femur. Cannulated drilling of the femoral neck under image intensifier stopping just at the epiphyseal line of the femoral head. We don't drill through the epiphyseal plate. Cannulated drill (ACL reamer) size 5.0 mm to 6.5 mm depending on the size of the neck diameter. One central drill is

done in all patients (Figure 3). Closure is done by simple absorbable sutures. Follow up visits in 1, 3, 6, 12, 18, 24, 36, 72 and 96 months after surgery. The mean follow up for CD group is 85.7 months with minimum follow up requirement time was 24 months and longest follow up was 9 years STD +/- 16.58. Clinical evaluation of pain, limping, level of activity and gait (Harris score for Pain, activity, walking distance, ROM and gait). Plain x-ray AP and frog lateral pelvis views are done at each visit.



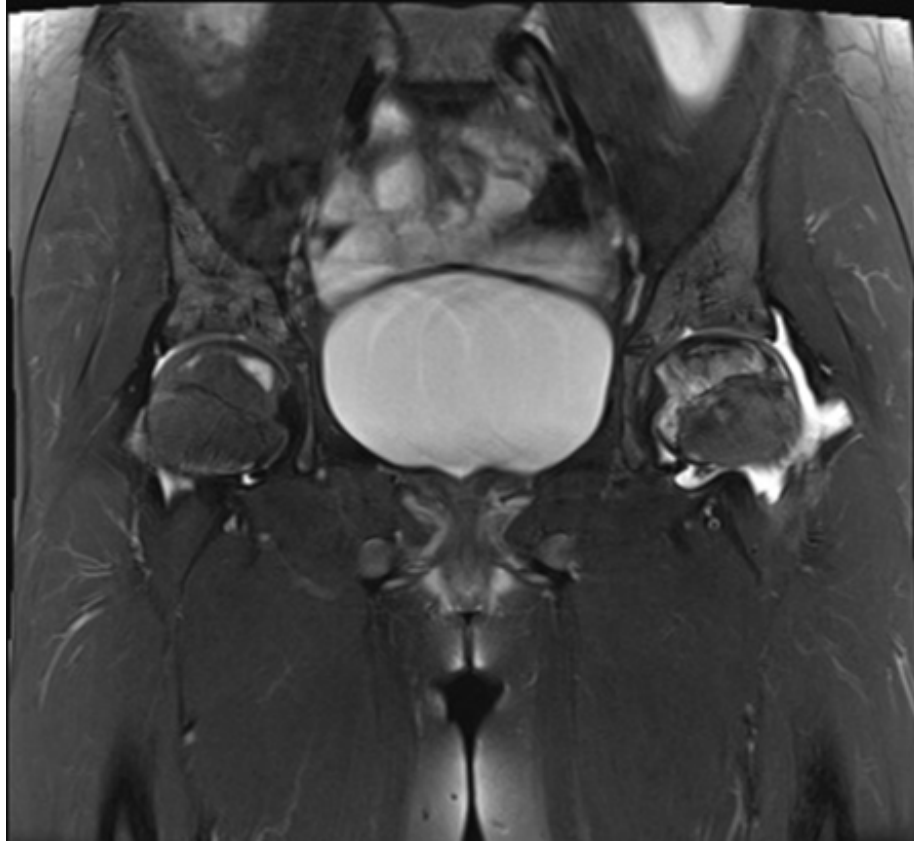


Figure 3a: Stage 2 both sides, b: MRI and c: post op

Results

AVN was found in 59 patients out of 322 sickle cell anemia patients with a prevalence of 18.4%. total of 77 hips are affected out of which 39 hips were enrolled in CD group. Bilateral hips in 10 patients. Stage 1, 2a, 2b are offered CD. Stage 3 are done

initially only if the other hip is in a lesser stage and planned for surgery.

The mean hemoglobin (HB) was 7.6 in AVN group \pm 0.96 STD compared to 7.9 in no AVN \pm 1.04 STD (Table 4)





Figure 4b, 4c: arm drill position, c: wound size, d: post op

Table 4: The average HB was 7.6 in AVN group compared to 7.9 in no AVN

	AVN + (59) X + SD		AVN - (243) X + SD		t test	Pvalue
Age	8.5	2.2	7.8	2.0	1.58	>.05
Hb	7.6	1.6	7.9	1.7	0.7	>.05
Hb F	27.2	8	28	8.8	0.63	>.05
VOC	3.8	2.39	3.3	2.1	1.1	>.05

Left hip was found to be slightly more affected by AVN than right hip 59% to 41%. Males were affected more than females in about 62% (Table 5).



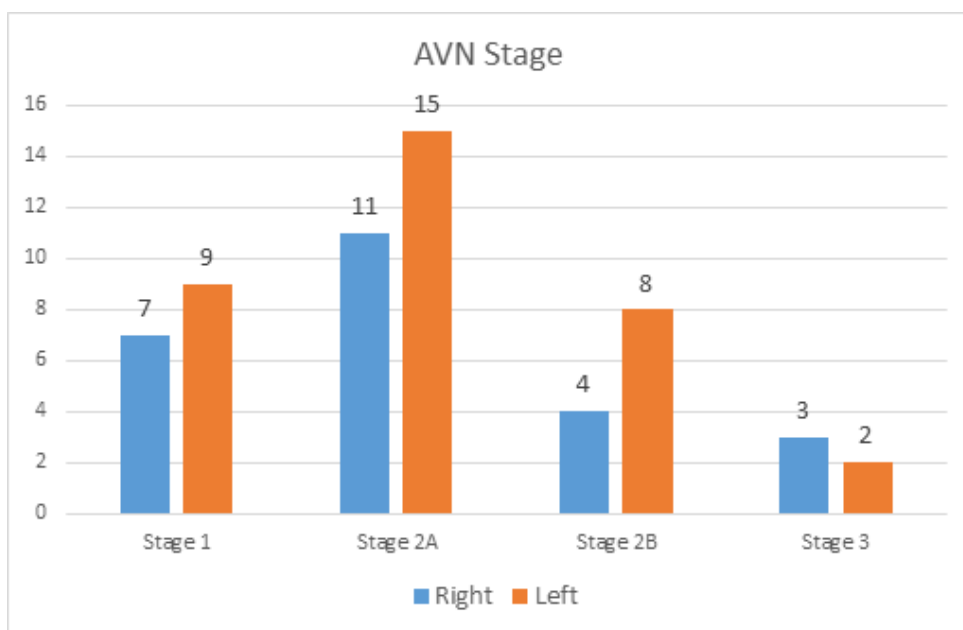
Figure 5a: Surgical setup and positioning

Table 5: Age and side distribution of AVN in SCA

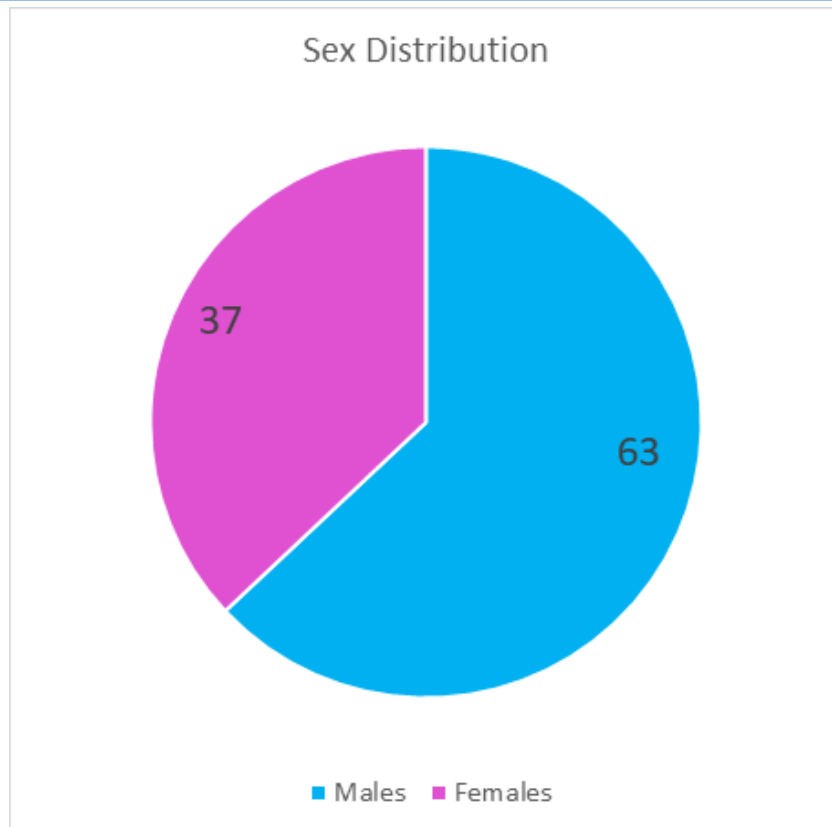
	Right	Left	
Male	15 (25.6%)	22 (38.5%)	37 62.8%
Female	9 (15.4%)	13 (20.5%)	22 37.2%
Total	24 (41.0%)	35 (59.0%)	59

Distribution By Stage

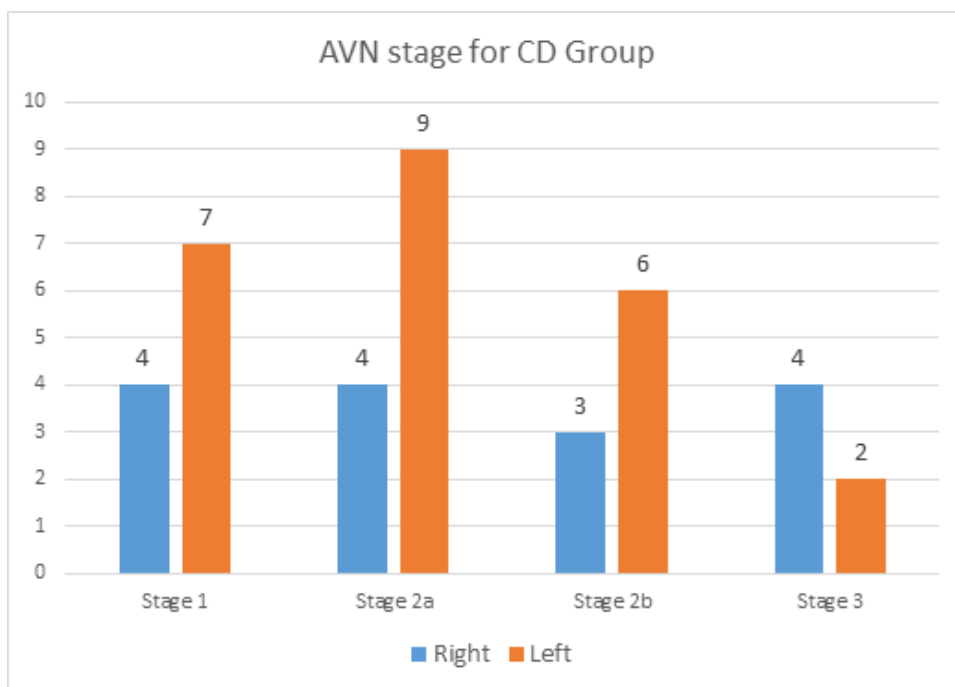
In all stages we found that left side is affected more than right side except in stage 3, it is more in the right side. (Graph 1 and 2)



Graph 1a: Hip side distribution



Graph 1b: Sex distribution.

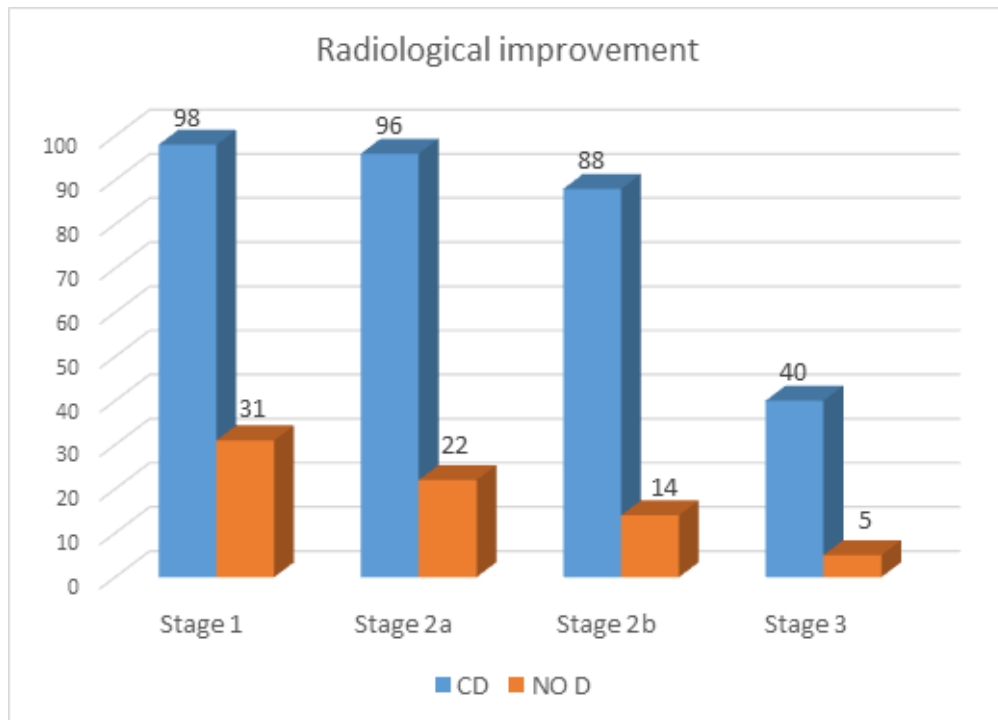


Graph 2: Graph distribution by stage for CD group

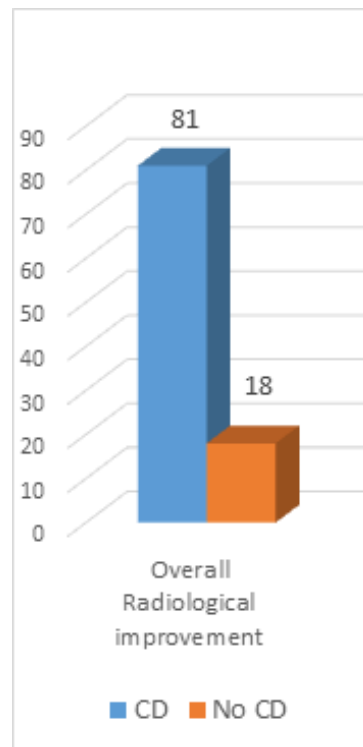
Improvement

Radiological evaluation is done by the same radiologist and the patients had follow up in 1, 3, 6, 12, 18, 24, 36, 72 and 96 months after surgery. Plain x-ray AP and frog lateral pelvis views. Radiological restaging is done at each visit. The radiological improvement was noticed after 5 months after core decompression,

some patients had recurrent AVN within 2 to 3 years after core decompression, redo core decompression was offered to them. Radiological improvement was found in 30 hips (77%) of CD group compared to 6 hips (16%) of no CD group (self-recovery) (Graph 3).



Graph 3a: Radiological improvement

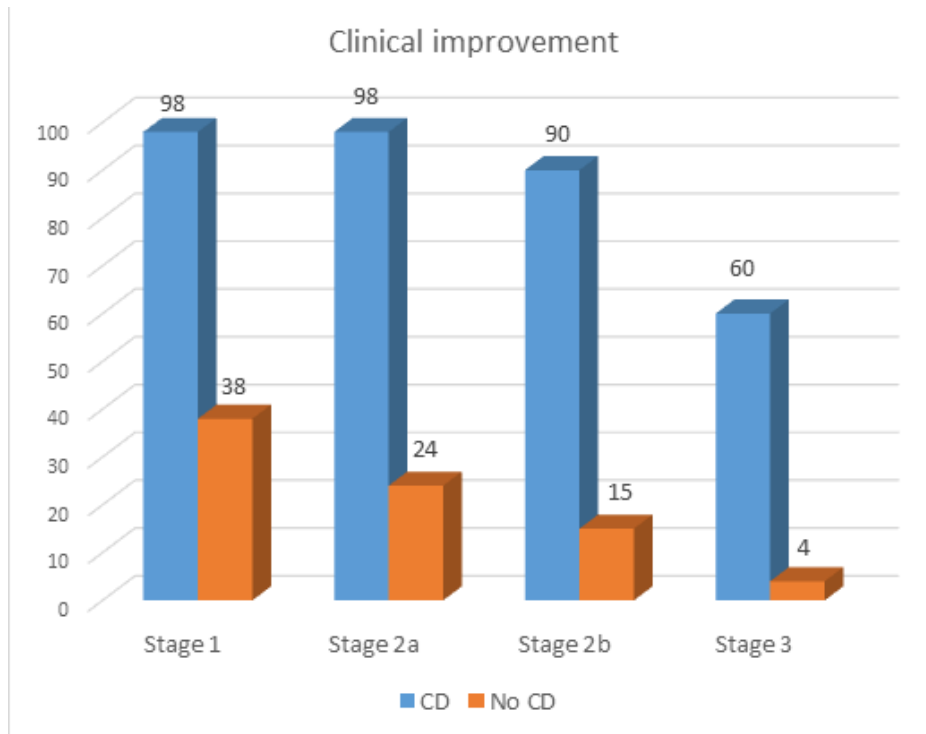


Graph 3b: Radiological overall stages improvement

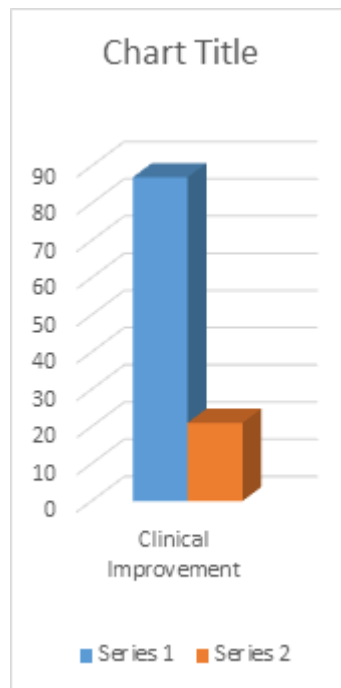
Clinical Improvement

Clinical evaluation is done in pediatric orthopedic clinic by the same surgeon unit using Harris score for Pain, activity, walking

distance, range of motion and gait. We found that there is 87% in CD group clinical improvement compared to 20% for no CD group (graph 4).



Graph 4a: Clinical improvement



Graph 4b: Overall stages Average Clinical improvement

Discussions

International prevalence of AVN of the femoral head is ranging from 5% to 40%. Our prevalence is 18.4%. very little studies were published regarding core decompression in sickle cell anemia in children. AVN in LCPD recommendation is to perform early core decompression with bone grafting combined with a shelf acetabuloplasty. there were only 2 cases report of delay in collapse of femoral head in AVN in sickle cell anemia [7,8].

AVN in males was found higher 62.8%. Left side is more affect-

ed 59% than right side. Asymptomatic AVN in 3 patients. Routine pelvic x-ray is recommended every 4-6 months for patients with SCA to screen for AVN of femoral head.

Stage 1 disease is only detected by MRI in symptomatic cases with normal hip x-rays. Self-recovery was noticed in 20% of patients in no core decompression group.

Stage 1, 2a and 2b have 98% excellent clinical and radiological outcome after CD. Stage 3 and 4 improved by 40% radiological-

ly and 60% clinically compared to 5% and 4% without surgery. Overall Excellent prognosis after CD 81.5% compared to 17.2 for no CD.

Recommendations

Stage 1, 2a and 2b have an excellent outcome after CD. Stage 3 and 4 showed good improvement radiologically and clinically compared to no CD group [3]. We recommend to do core decompression as early as possible to have the best outcome. We also recommend to do core decompression for all stages of AVN including stage 3 and 4 with or without the other hip involvement. Bilateral hip core decompression is recommended if one hip is affected.

Early detection and treatment of AVN provide the best results. We recommend AVN screening for all SCA patients by regular pelvis 2-views x-rays and MRI for suspected cases. We recommend Core decompression for all stages of AVN of femoral head in SCA in children. Core decompression may be an effective method to stop and improve AVN of femoral head in SCA in children.

Establishment of unified management protocol for comprehensive care of sickle cell anemia patients that include pediatric hematology, radiology and pediatric orthopedic.

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