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Original Research Article

Evaluation of Abdominal Organs in Children with Sickle Cell Anemia using Ultrasonography

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Abstract: Sickle-cell disease (SCD), also known as sickle-cell anemia (SCA), is a hereditary blood disorder, caused by an abnormality in the oxygen-carrying protein hemoglobin found in red blood cells. This leads to a propensity for the cells to assume an abnormal, rigid, sickle-like shape under certain circumstances. Sickle-cell disease is associated with a number of acute and chronic health problems, such as severe infections, attacks of severe pain ("sickle-cell crisis"), stroke, and an increased risk of death. The study was aimed to evaluate the abdominal organs in children with sickle cell anemia using ultrasonography. This was cross sectional descriptive study; conducted in a total of 103 patients with sickle cell anemia, their age was range 10 months -16 years (mean age 6.1 years ± 4.3 SD), in different hospital in Khartoum from January - August 2016. All patients scanned by ultrasound according to international guidance and protocol, their ultrasound findings and history was recorded in data collection sheet and these data was analyzed by Statistical package of social sciences program version 15. The commonest transabdominal ultrasound finding in this study was splenic abnormality in 74 (71.9%) flowed by hepatomegaly in 53 (51.5%) patient, gallbladder abnormality in 22 patients (21.3%), and enlarged kidneys in 23 patients (22.3%). There was positive relation between spleen size and the duration of disease, in early stage of disease the spleen is enlarge and after that become small with time until disappear in adulthood. This study provides that ultrasound have excellent role in detected abdominal abnormality in sickle cell anemia patients and to follow the complications of disease. The study recommended that, ultrasound should be used as routine exam to follow the stages of disease because it is noninvasive and repeatable.
Keywords: Sickle-cell disease (SCD), blood disorder, ultrasonography, abdominal organs

INTRODUCTION:

Sickle-cell disease (SCD), also known as sickle-cell anemia (SCA), is a hereditary blood disorder, caused by an abnormality in the oxygen-carrying protein hemoglobin found in red blood cells. This leads to a propensity for the cells to assume an abnormal, rigid, sickle-like shape under certain circumstances. Sickle-cell disease is associated with a number of acute and chronic health problems, such as severe infections, attacks of severe pain ("sickle-cell crisis"), stroke, and an increased risk of death. [1]Red blood cells that contain normal hemoglobin are disc shaped (like a doughnut without a hole). This shape allows the cells to be flexible so that they can move through large and small blood vessels to deliver oxygen [2].

Normally, humans have haemoglobin A, which consists of two alpha and two beta chains, haemoglobin A2, which consists of two alpha and two delta chains, and haemoglobin F, consisting of two alpha and two gamma chains in their bodies. Of these, haemoglobin F dominates until about 6 weeks of age then A dominates throughout life [2]. People who have SCD inherit two abnormal hemoglobin genes, one from each parent. In all forms of SCD, at least one of the two abnormal genes causes a person's body to make hemoglobin S. When a person has two hemoglobin S genes, Hemoglobin SS (homozygous) the disease is called sickle cell anemia. This is the most common and often most severe kind of SCD. If the person is heterozygous for genes (as the parent of infants with sickle cell anemia), he is said to have sickle cell trait here the

RBCs contain mixture of HbA and HbS and sickling does not occur except if individual expose to hypoxia resulting from shock [1].

Hemoglobin SC disease and hemoglobin Sβ thalassemia are two other common forms of SCD. Some Forms of Sickle Cell Disease includes, Hemoglobin SS, Hemoglobin SC, Hemoglobin Sβ0 thalassemia, Hemoglobin Sβ+ thalassemia, Hemoglobin SD and Hemoglobin SE. Sickle cells can't change shape easily, so they tend to burst apart or hemolysis. Normal red blood cells live about 90 to 120 days, but sickle cells last only 10 to 20 days [2].

The manifestation of disease does not appear in first 3-4 months as HbF products against sickling [1]. Epidemiology of disease almost 300,000 children are born with a form of sickle-cell disease every year, mostly in sub-Saharan Africa but also in other parts of the World such as the West Indies and in people of African origin elsewhere in the world. In 2013 it resulted in 176,000 deaths up from 113,000 deaths in 1990. The condition was first described in the medical literature by the American physician James B. Herrick in 1910 [2].

Distribution of sickle cell anemia among the Sudanese, studies showed that sickle cell gene frequencies vary from region to another in Sudan as well as within the same region. Central Sudan: Sickle cell gene is known to be prevalent in the Khartoum area, which is the capital of the country and situated in central Sudan. In the 1980s when drought and famine struck western Sudan, a huge number of migrations took place and many tribes settled around Khartoum. This unique situation made Khartoum a multiethnic area, with a blend of almost all the Sudanese tribes. Among 632 patients attending various clinics at the Khartoum Teaching Hospital, there were 5.1% with Hb AS and 0.8% with Hb SS. The rate is highest in Western Sudanese ethnic groups particularly in Mesearia tribes in Darfur and Kordofan regions. In the Blue Nile area, where groups of indigenous population live, the prevalence ranges from 0-5% in addition to a rate of 16% among some immigrant tribes from western Sudan and West Africa [3].

Northern Sudan: Although the data about sickle cells gene in the north of Sudan is incomplete, it seems that this area shows a low frequency of SCA. A study conducted in the north of Sudan in Shagia and Manaseer tribes confirmed that the sickle cell gene is lower in the north of Sudan than in other areas. Shagia are partly nomadic, isolated, and an agricultural population. Therefore, it is difficult to determine significantly whether they are Arab or African [3].

Eastern Sudan: Most studies on sickle cell gene done in this region were conducted in Gedarif state, since the majority of the population migrated to and settled in this state in different decades in the past century. Blood samples tested for SCA among 100 individuals from different tribes in Gedarif state showed that 20 samples had HbSS, 55 samples had HbAS and 25 samples had HbAA. The results of this study cannot be generalized for the population of the area due to the low sample number. Showing that sickle cells trait Hb AS was found in approximately 35% of study subjects in Hausa and 24% in Massaleet, whereas HbSS was reported as 6% and 5% in Hausa and Massaleet respectively [3].

Western Sudan: The presence of HbS is already well documented among Kordofan and Darfur region inhabitants, especially Albaggara, an Afro-Arab constellation of tribes with a predominantly African descent. Some findings of a study conducted in Elobied hospital in north Kordofan state, showed that sickle cell trait in relatives of patients suffering from sickle cell disease (SCD) who were referred to this Hospital, was 54% of target samples, which concentrated mainly in two tribes, Bederia and Fulani. Sickle cell disease in Messeryia of Darfur and Messeryia Hummer of Kordofan showed a prevalence of 30.4% and 18% respectively. It is estimated that one in every 123 children born in Messeryia tribe is at risk of having SCD [3].

Ultrasound is noninvasive technique that can use easily to evaluate abdominal organs for abnormality that determine the management of disease and reduce the complication. Ultrasound is doing for spleen, hephatiobillary system and kidney. Splenic enlargement may occur transiently with the sequestration syndrome, where rapid pooling of blood occurs in the spleen, resulting in intravascular volume depletion, with potential for cardiovascular collapse [2].

The slow, tortuous micro-circulation of the spleen renders it susceptible to infarction and subsequent functional asplenia (Autosplenectomy). Hepatobiliary: hepatic iron deposition secondary to multiple transfusions. Hepatomegaly +/- coarsened echotexture with portal hypertension Cholelithiasis +/- choledocholithias & multiple liver abscesses. Renal: kidneys are often large early in the disease, with variable Echogenicity on ultrasound, but shrink with development of renal failure. Bilateral Echogenic pyramids are frequently seen in sickle cell disease.

Gastrointestinal tract: approximately 40% patient may develop peptic ulcers due to reduced mucosal resistance and bowel ischemia [2].

OBJECTIVES:

To evaluate the abdominal organs in children with sickle cell anemia using Ultrasonography.

MATERIAL & METHODS

Descriptive cross sectional study deal with the evaluation of children have sickle cell anemia. The study was conducted in different hospitals in Khartoum. The sample size were103 children diagnosed with sickle cell anemia, the selection of participation was through simple random sampling technique.

Study variables includes, children age from 10 months -16 years with gender, different race, duration of disease and family history. Determine the size, echogenicity and lesion in liver, Spleen and kidney by using ultrasound. Data was collected by data collection sheet which designed to include all variables that satisfy the study. Ultrasound scanning of abdominal organs, liver, GB, spleen and kidney following the international guidance and protocol of ultrasound scanning.

DISCUSSION:

The results of this study showed that the prevalence of disease in male (52.4%) & female (47. 6%), there is no significant difference between them. The study found that most of the patients have positive family history 79 (76.7%). This result was considered with literature "sickle-cell anemia (SCA) is a hereditary blood disorder, caused by an abnormality in the oxygen-carrying protein hemoglobin found in red blood cells" .[1]

The results showed that the most affected age group was 1-5.5 years which represented 40 cases (38.8%) and the least one more than 15 years 3 patients (2. 9%). The mean age = 6.8 years, minimum=10 months, maximum =16 years and SD =4). This result agreed with (Nahid Mahmoud 2014), she found that the common age group affected by SCA was in the range of 1-5 years [19], and in line with (Usmanu Danfodiyo 2010), he found that the mean age 6.1 and SD =4.3[15]. Concerning the residence of patients, the study found that sickle cell anemia was high in the center about 89 (86.4%). This result agreed with literature "Sickle cell gene is known to be prevalent in the Khartoum area, which is the capital of the country and situated in central Sudan This unique situation made Khartoum a multiethnic area, with a blend of almost all the Sudanese tribes"[3].

Regarding patient's tribe, the most tribe affected with disease is western Sudan Mesearia tribe 31 (30.1%), This result agrees with (Majdi Mohammed Sabahelzain, & Hanan Hamamy 2014), they found that the highest rates reported from Western and Eastern Sudan where one in every 123 children born in Messeryia tribe in Western Sudan is at risk of having SCD [3]. Regarding to duration of disease, the study found most patients 27 with percentage (26.2%), the duration was 1-3 years and when the patients age increases the duration spontaneously increase.

About the spleen size the study found that 29 patients with percentage (28.2%) have normal spleen size ,46 patients have abnormal spleen size with percentage (44.7%), (enlarged in 22 patients (21.4%) and shrunk in 24 patients (23.3%) and autosplenectomy in 28 patients (27. 2%).this results were consistent with (Usmanu Danfodiyo 2010), he found that splenomegaly was found in 15 (21.1%), and disagree with them about Autosplenectomy was demonstrated in 3 (4.2%) patients[15], and (Bakhieta Ibrahim Attalla 2010), She found that Autosplenectomy (asplenia) was detected in 43 (47.8%) patients[18], and (Ali Balcı *et al.;* in 2008), who found that autosplenectomy was found by percentage (33.3%), and splenomegaly (17.4%) [3].

Regarding to spleen echogenicity, most of the patients have normal spleen echogenicity 72 patients (69. 9%).This finding consistent with literature "in case of splenomegaly the sonographic texture and echogenicity often remain normal despite the presence of disease"[9] and most of my patient have splenomegaly and autosplenectomy. About splenic lesion most of the patients about 71 patients (68.9%) have no splenic lesion, and hypoechoic splenic lesion (infarction) was found in 4 patients (3.9%) this results matching with (Bakhieta Ibrahim Attalla 2010), who found that Seven (7.8%) patients had hypoechoic lesions and Hyperechoic lesions with splenomegaly were found in 6 (6.6%) patients [18]. Regarding to liver size the most number of patients 53 have hepatomegaly (51.5), this results in line with (Usmanu Danfodiyo 2010), who reported that Hepatomegaly was found in 70 (98.6%) (15), and also matching with (Ali Balcı et al 2008), she found that hepatomegaly was reported in (71.6%) [11].

About liver echogenicity, the study was showed most of the patients have normal liver echogenicity (79.6%) and small number patients 21 (20.4%) have increased liver echogenicity. This result attributed to literature "patients with SCD have often received

multiple transfusions, placing them at risk for viral hepatitis & iron overload sonographically, a patient with hepatitis may initially have a completely normal-appearing liver"[11] and also agrees with (Bakhieta Ibrahim Attalla 2010) who found that Bright liver was identified in 6 patients (5.9%) "[18].

According to GB abnormality, 81patients (78.6%) have no GB abnormality and 22 patients (21.3%) have abnormal GB ranged from (distended GB (10.7%), GB stones (4.9%), (1.9%) have stone and thick wall and (1%) have stone &distended GB, sludge (1.9%) & thick wall (1. 0%). this results matching with, (Usmanu Danfodiyo 2010), who reported that gallbladder wall thickening found in 7 (9.7%), and gallbladder stone seen in 4 (5.6%) patients and gallbladder sludge was seen in 2patients, (15) and also with (Bakhieta Ibrahim Attalla 2010) The overall prevalence of gallstones was 11.1%, and it increased with age. Two patients (2.2%) had thickened gallbladder wall, and only 1 patient (1.1%) had a sludge gallbladder] 18]. About kidney size, the researcher reported that high number of patient 79 (76.7%) have normal kidney size and 23 patients (22.3%) have enlarged kidneys, this results consistent with (Ali Balcı et al 2008), who reported that renal enlargement was reported in (30.4%) [11].

The study found that high number of patient 79(76.7%) have normal kidney echogenicity, following by 16 patients (15. 5%) increase kidneys & 8 patients have decrease kidneys (7.8%). This result was matching with (Brachiate Ibrahim Attalla 2010), she found that normal renal echogenicity was reported in 86 (95.6%) of patients (18) and (Ali Balcı et al 2008), who reported that medullary or diffusely increased renal echogenicity was observed in 16 patients (15. 7%). Sonographic findings typical of renal papillary necrosis were observed in one patient [11]. The study found that, there was no correlation between liver size and the duration of disease shows no significant difference (p value 0.026).

The study found that there was strong correlation between spleen size and the duration of disease (p value 0.002). These results attributed to literature "promising new therapies are being developed; many homozygous sickle cell patients become a Splenic in late childhood or early adulthood. This results in a small spleen, often difficult to visualize, with a diffuse echogenic appearance"[12]. The study reported that there was no correlation between spleen size and patients gender (p value 0.656). 103 children were diagnosed to had homozygous sickle cell disease were scanning with ultrasound for abdominal organs. The mean age of the study subjects was (6.8) years old; the minimum age was 10 months old and the maximum age was 16 years old. Forty-five (52.4%) of the study subjects were males and forty nights (47.6%) were females.

Table (1.A) Spleen Size * Duration of disease Cross tabulation

Spleen size	Duration of disease					Total
	Less than one years	one to 3 years	3.1 to 6 years	6.1 to 9 years	more than 9 years	
Normal	5	9	6	7	2	29
Enlarge	2	10	3	4	3	22
Shrunk	0	7	6	5	6	24
Autosplenectomy	0	1	6	7	14	28
Total	7	27	21	23	25	103

Table (1, b) Chi-Square Tests

	Value	df	Asymp. Sig. (2-sided)
Pearson Chi-Square	30.638(a)	12	.002
Likelihood Ratio	35.266	12	.000
Linear-by-Linear Association	21.376	1	.000
N of Valid Cases	103		

a 7 cells (35.0%) have expected count less than 5. The minimum expected count is 1.50.

Table (1, C) Symmetric Measures

		Value	Asymp. Std. Error(a)	Approx. T(b)	Approx. Sig.
Interval by Interval	Pearson's R	.458	.075	5.175	.000(c)
Ordinal by Ordinal	Spearman Correlation	.456	.077	5.152	.000(c)
N of Valid Cases		103			

a Not assuming the null hypothesis.
b Using the asymptotic standard error assuming the null hypothesis.
c Based on normal approximation.

Table (2.A) Crosstab between liver size and duration of the diseases

Liver size	Duration of disease					Total
	Less than one years	one to 3 years	3.1 to 6 years	6.1 to 9 years	more than 9 years	
Normal	6	11	7	9	17	50
Enlarge	1	16	14	14	8	53
Total	7	27	21	23	25	103

Table (2.b) Chi-Square Tests between liver size and duration of the diseases

	Value	df	Asymp. Sig. (2-sided)
Pearson Chi-Square	11.080(a)	4	.026
Likelihood Ratio	11.594	4	.021
Linear-by-Linear Association	.471	1	.493
N of Valid Cases	103		

Table (2.C) Symmetric Measures between liver size and duration of the diseases

		Value	Asymp. Std. Error(a)	Approx. T(b)	Approx. Sig.
Interval by Interval	Pearson's R	-.068	.099	-.684	.495(c)
Ordinal by Ordinal	Spearman Correlation	-.081	.101	-.812	.419(c)
N of Valid Cases		103			

a Not assuming the null hypothesis.
b Using the asymptotic standard error assuming the null hypothesis.
c Based on normal approximation.

Table (3.A|) Patient gender * Spleen Size Cross tabulation

Gender	Spleen Size				Tot al
	Normal	Enlarged	Shrunk	Autosplenectomy	
Male	17	10	14	13	54
Female	12	12	10	15	49
Total	29	22	24	28	103

Table (3.b) Chi-Square Tests Patient gender * Spleen Size

	Value	df	Asymp. Sig. (2-sided)
Pearson Chi-Square	1.614[a]	3	.656
Likelihood Ratio	1.618	3	.655
Linear-by-Linear Association	.397	1	.529
N of Valid Cases	103		

a. cells (.0%) have expected count less than 5. The minimum expected count is 10.47

Table (3.C) Symmetric Measures Patient gender * Spleen Size

		Value	Asymp. Std. Error[a]	Approx. T[b]	Approx. Sig.
Interval by Interval	Pearson's R	.062	.098	.628	.531[c]
Ordinal by Ordinal	Spearman Correlation	.065	.098	.650	.517[c]
N of Valid Cases		103			

a. Not assuming the null hypothesis.
b. Using the asymptotic standard error assuming the null hypothesis
c. Based on normal approximation.

CONCLUSION:

The study concluded that Ultrasound is reliable, repeatable and noninvasive examination used to detect and follow up the sickle cell disease and their complication on the abdominal organs. Most of patients with sickle cell anemia come to hospital with severe anemia and episodes pain and other on stable condition, come for follow up. The commonest transabdominal ultrasound finding in this study was splenic abnormality in flowed by hepatomegaly, gallbladder abnormality and the least common finding enlarged kidneys. The residence of patient with sickle cell anemia was high in the center of Sudan with the Most tribe affected with disease was Mesearia tribe in western Sudan. The study found positive relation between spleen size and the duration of disease, in early stage of disease the spleen is enlarged and become small with time until disappear in adulthood (autosplenectomy) and no correlation between liver size and the duration of disease. The study found there was two patients underwent to cholecystectomy due to acute cholecystitis and one patient to splenectomy due to hypersplenism.

RECOMMENDATIONS:

An investigation before marriage for affected tribe's member should be performed; patients with sickle cell anemia should be investigated by ultrasound regularly as part of routine follow up exam. Encourage each woman, man, and couple affected by sickle cell anemia to have a reproductive life plan. Remind people with sickle cell anemia, their families, and caregivers to seek immediate medical attention whenever acute abdominal pain and recurrent infections were occurring for immediate treatment and reduce the complications.

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