

متغيرات صدى القلب في  
مرضى فقر الدم المنجلي  
**Echocardiographic changes  
in Patients with Sickle  
Cell Anaemia**

**By**

**Dr. Abdul Raheem H. Al-Humrani**  
**Assist Prof of Medicine**  
**Department of Medicine**  
**Medical college of Basrah university**

الدكتور عبد الرحيم الحمراني  
أستاذ مساعد | فرع الطب  
كلية الطب | جامعة البصرة

**Dr. Basim A. Motar**  
**M. B. Ch. B (postgraduate student)**

باسم عذيب  
دائرة صحة البصرة

## **SUMMARY:-**

A comparative study was carried out on 50 subjects, ( 25 patients with homozygous sickle cell anemia (SCA) and 25 healthy subjects with matched sex, age and surface area to patients group), their ages range from 15-30 years (mean age  $22.08 \pm 4.2$ ),

The aim of this study was to detect the echocardiographic changes in patients with sickle cell anemia.

The echocardiographic findings were abnormal in 9 (36%) patients. Mitral valve prolapse (MVP) was detected in 6 (24%) patients with SCA and 1 (4%) control.

An interesting observation that the ejection fraction was statistically significant lower in patients with SCA than the control group ( $53.63 \pm 8.45$  Versus (VS)  $62.22 \pm 10.75$ ) respectively. Despite that marked left ventricular (LV) function impairment was detected only in 1 (4%) patient (his ejection fraction 39.5%).

There was neither significant changes in left ventricular end diastolic volume (LVEDV) ( $95.92 \pm 23.08$  VS  $94.4 \pm 28.49$ ) milliliter (ml) , nor in left ventricular end systolic volume (LVESV) ( $57.64 \pm 24.45$  VS  $54.23 \pm 17.24$ ) ml .There was no significant changes in left atrial dimension (LAD) ( $24.19 \pm 7.01$  VS  $20.92 \pm 3.69$ ) millimeter (mm) , nor in aortic root dimension (AOD) ( $21.09 \pm 5.02$  VS  $19.98 \pm 3.23$  ) mm in the patients group and the controls group respectively.

**الخلاصة :**

الهدف من الدراسة :معرفة التغيرات القلبية بواسطة الموجات فوق الصوتية لدى المرضى المصابين بفقر الدم المنجلي

خلال الفترة من شهر آذار\_إلي كانون الأول عام 2001 ميلادية أجريت هذه الدراسة في مدينة البصرة على (50) شخصا منهم 25 شخصا مصابا بمرض فقر الدم المنجلي (12 ذكور, 13 إناث) و 25 شخصا من الأصحاء لغرض المقارنة . كان العمر والجنس لكلنا المجموعتين متطابق وتراوحت الأعمار من 15-30 سنة .

أظهرت الدراسة أن فحص صدى القلب بالموجات فوق الصوتية كان غير طبيعيا لدى

9مرضا (36%) في حين كان الفحص طبيعيا لدى جميع الأشخاص الأصحاء (مجموعة المقارنة) عدا شخص واحد فقط (4%) هذا الاختلاف كان ذو دلالة إحصائية اثبت هذا الفحص أن 6 مرضى أي نسبة (24%) كان لديهم تدلي الصمام التاجي بينما كانت هذه النسبة (4%) فقط لدي مجموعة المقارنة من الأصحاء. هذا الاختلاف ذو دلالة معيارية لم تظهر هذه الدراسة أي فروقات ذات دلالة معيارية في حجم البطين الأيسر أو الأيمن أو الأذين الأيسر أو الشريان الأبهري بين المجموعتين. ولكن ظهر انخفاض ملحوظ في نسبة التدفق الجزئي ذو دلالة معيارية كما أظهرت الدراسة اختلال ملحوظ في وظيفة البطين الأيسر في مريض واحد.

## Introduction:

Sickle cell anemia (SCA) is usually reserved for the homozygous state for HbS , which is characterized by substitution of a valine for

glutamic acid as the sixth amino acid in the B-globine chain<sup>1,2</sup>. SCD is a major public health problem in eastern countries with an estimated prevalence rate of 1.5 % to 1.75 %<sup>3</sup>.

The anemia of Hb SS disease is usually well tolerated by the cardiovascular system (CVS) for long periods. However, like other moderately severe anemias, cardiac function is ultimately affected<sup>4</sup>.

There is conflicting reports about cardiac changes in SCA<sup>5-13</sup>,  
The aim of this study was to study the echocardiographic changes of SC patients during rest.

## **PATIENTS AND METHODS:-**

A comparative study was carried out on 50 subjects, 25 with homozygous SCA (12 male and 13 female) who were attended or admitted at Basrah general hospital, and 25 healthy control subjects with matched sex and age to the patients during the period from March 2001-October 2001 were included in this study. Their ages ranged from 15-30 years (mean 22.08±4.2). One patient was excluded from the study because he was discovered to had silent mitral valve stenosis.

A complete history and full clinical examination with particular attention to CVS were performed for every patient and healthy subject. None of the patients or controls that were included in this study had evidence of known valve disease, hypertension or cardiac ischemia. All subjects had Hb %, Hb electrophoresis, chest x-ray and ECG. The diagnosis of SCA was confirmed by Hb electrophoresis.

M-mode and two-dimensional echocardiograms with 2-4MHZ Probe using Kretz technique volusion<sup>(R)</sup> 530-D were obtained at rest. Subjects were not febrile, dehydrated or in SC crisis during the study, and

echocardiograms were obtained at least 1 month after the SC crisis. All measurements were made according to the recommendations of the American society of Echocardiography<sup>14</sup>.

The following dimensions were measured : Left atrium (LAD), aortic root (AOD), left ventricular end diastole (LVEDD), left ventricular end systole (LVESD), right ventricular diastole (RVDD), right ventricular systole (RVSD), left ventricular posterior wall thickness during diastole (LVPWTD) and during systole (LVPWTS), interventricular septal thickness during diastole (IVSD) and systole (IVSS). Left ventricular end diastolic volume (LVEDV) and left ventricular end systolic volume (LVESV) LV end diastolic volume index (LVEDVI) and LV systolic volume index (LVESVI), Shortening fraction (SF%), ejection fraction (EF%) and stroke volume (SV) .

Differences were considered statistically significant if the P value was less than 0.05.on the basis of Chi-square test analysis.

## **RESULTS:-**

Fifty subjects were included in this study, 25 patients with SCA their ages range 15-30 years (mean  $22.08 \pm 4.2$  SD), 12 patients were males, 13 patients were females, and 25 healthy subjects as a control with matched sex, age to the patients.

The echocardiographic findings were abnormal in 9 (36%) patients with SCA, 2 male and 7 female VS 1 (4%) female in control group (Table II), these differences are statistically significant (P value = 0.0476).

An interesting observation that 6 (24%) patients had mitral valve prolapse VS 1 (4%) pt in the control, this was statistically significant.

One (4%) patient had marked LV function impairment, with an ejection fraction (39.5%).

Table (I) demonstrated no significant changes in LVEDV (It was  $(95.92 \pm 23.08)$  ml in the patients group VS  $(94.4 \pm 28.49)$  ml in the controls group), or in the LVESV ( $57.64 \pm 24.45$  ml in the patients group VS  $54.23 \pm 17.24$  ml in the controls group), and there were no statistical significant differences in LVEDVI, or LVESVI. The magnitude of stroke volume was lower in the patients group ( $39.08 \pm 15.67$ ) ml than in the controls group ( $42.92 \pm 22.41$ ) ml, but this was statistically not significant. There were highly significant differences in the (EF) between patients group ( $53.63 \pm 8.45$ ) VS ( $62.22 \pm 10.75$ ) in the controls group (p value 0.029).

Table I also demonstrated neither statistical significant differences in LAD nor in AOD ( $24.19 \pm 7.01$  VS  $20.92 \pm 3.69$  and  $21.09 \pm 5.02$  VS  $19.98 \pm 3.23$ ) mm respectively.

Table II demonstrates an increase in both IVS thickness and LVPW thickness during systole. 11 (44%) patients VS 2 (8%) controls had IVS thickness and LVPWT  $\geq 12$  mm respectively, this was statistically significant, but there were no statistical significant differences in IVS thickness, LVPW thickness during diastole between patients and controls group (Table III).

The RV was more dilated in patients group than controls group both in diastole and systole. 12 (48%) patients VS 9 (36%) controls had RV diastolic dimension above 25 mm and 6 (24%) patients VS 2 (8%) controls had RV systolic dimension above 25 mm. These differences were statistically not significant. (Figure 1&2).

## **DISCUSSION:-**

Abnormal echocardiographic findings in this study were detected in 9 (36%) patients with SCA, this consistent with a study was done by Kapoor RK, et al. <sup>15</sup>.

MVP was detected in 6 (24%) patients with SCA and 1 (4%) control. (P-value 0.072). This consistent with Lippman et al. <sup>16</sup> and the findings of Markiewicz et al. <sup>17</sup> but in contrast with Husain A, et al. <sup>18</sup> who reported no significant differences in his study, the drawback of his study that the high incidence of MVP in the controls group (13.3%) and this is in contrast with most studies <sup>16-18</sup>.

This study showed 4% incidence of MVP in control group , there was no study in Iraq determined the prevalence of MVP in general population ,but this was consistent with other study carried outside <sup>16-18</sup>This study demonstrates no significant changes in LVEDV, LVESV, LAD and AOD in SCD patients and controls group. This consistent with most studies <sup>16-18</sup> in contrast with James FW study <sup>19</sup> who showed significant differences but his study was done by using exercise test echocardiography, while we did echocardiography at rest.

The ejection fraction was significantly lower in the patients with SCA ( $53.63 \pm 8.45$ ) than the controls group ( $62.22 \pm 10.75$ ) despite these measured within lower limit of normal range <sup>16</sup>.

Significant impairment of LV function (EF less than 40 %) was detected only in 1 (4%) patient with SCA who hadn't any other cause of LV function impairment other than SCA. This suggest that sickle cell cardiomyopathy despite it is rare but still occur in severe disease <sup>1</sup>.

IVS and LVPW thickness were above 12 mm in 11 (44%) patients and 2 (8%) controls during systole. This consistent with other study<sup>18</sup>, these may be explained by fact that a number of factors (like decreased

myocardial compliance, impaired systolic function, hyperdynamic circulation, LV hypertrophy) may be contributing to these abnormalities<sup>13</sup>.

RV diastolic dimension was more than 25 mm in 12 (48%) patients VS 9 (36%) controls, these consistent with other study<sup>18</sup>. This may be explained by fact that SCD patients may had intrapulmonary crisis or recurrent emboli that lead to pulmonary hypertension and RV overload<sup>1,2</sup>.

## **CONCLUSIONS :**

- 1- The heart may be the target of sickling process directly or indirectly.
- 2- Echocardiography can detect significant changes in LV function or dimensions. The significant echocardiographic changes which were detected in this study include : impaired LV function, increase LV wall thickness, increase RV dimension and MVP which was more prominent in SCA than the controls group.

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Table I the LV size and function LA&AO dimension

PARAMETER	PATIENTS GROUP	CONTROLS
LVEDV(ml)	95.92 ±23.08	94.4±28.49
LVEDVI	61.09 ± 13.5	59.69±19.11
LVESV(ml)	57.64 ± 24.45	54.23 ±17.24
LVESVI	38.46 ± 15.54	33.81± 11.16
SV (ml)	39.08 ± 15.67	42.92 ± 22.11
EF % *	53.63 ± 8.45	62.22 ± 10.75
LAD (mm)	24.19 ± 7.01	20.92 ± 3.69
AOD (mm)	21.09 ± 5.02	19.98 ± 3.23

\*Highly significant P value 0.0029

Table 2 The IVS &LPW thickness in systole

THICKNESS	IVSS			LVPWS		
	Patients	Controls	P value	Patients	Controls	P value
Less than 10	2	13	>0.05	2	13	> 0.05
10- 10.9	4	4	>0.05	8	4	>0.05
11- 11.9	8	6	>0.05	4	6	>0.05
>12	11	2	<0.05*	11	2	<0.05*
Total No of the cases	25	25		25	25	

mm: millimeter, \* statistically significant

Table III the IVS &LPW thickness in diastole

Thickness (mm)	IVSD			LVPWD		
	Patient	Control	P value	Patient	Control	P value
Less than 10	11	19	>0.05	19	18	>0.05
10-10.9	11	2	<0.05*	4	6	>0.05
11-11.9	1	4	<0.05*	2	1	>0.05
>12	2	0	>0.05	0	0	>0.05
Total	25	25		25	25	

\* statistically significant

