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ORIGINAL ARTICLE

Cardiac Status of Patients with Thalassemia Major on Echocardiography

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ABSTRACT

Background: Beta-thalassemia is a common genetic disorder in which red blood cell fail to perform normal function due to alterations in structure of red blood cells.

Objective: To determine the cardiac status (mean left ventricular end diastolic diameter, left ventricular end systolic diameter and interventricular septal thickness) of patients with thalassemia major on echocardiography.

Study Design: Cross-sectional study.

Place and Duration of Study: Department of Pediatric Medicine, GMMMC Hospital, Sukkur from 2nd October 2020 to 1stApril 2021.

Methodology: Ninety eight patients of both genders with age 2-18 years having thalassemia major. Thalassemia major were assessed and demographic variables including age, gender, family history of cardiac diseases, socio-economic status, other comorbid condition, duration since diagnosis of thalassemia was determined. All patients were also assessed for cardiac status using echocardiography.

Results: Mean age of patients was 7.73±5.01 years. Mean duration since diagnosis of thalassemia was 6.12±4.80 years. There were 65 (66.33%) males and 33 (33.67%) female patients. There were 18 (18.37%) patients having low, 52 (53.06%) middle and 28 (28.57%) high status. Family history of cardiac disease was found in 15 (15.31%) patients. Mean left ventricular end diastolic diameter was 38.42±3.75 mm, left ventricular end systolic diameter was 24.45±2.40 mm and interventricular septal thickness was 6.23±1.45 mm.

Conclusion: Left ventricular diastolic dysfunction (LVDD) occurs in all studied patients of thalassemia major. There was no systolic dysfunction and intraventricular septal thickening in thalassemia major patients.

Keywords: Left ventricular end diastolic diameter, Left ventricular end systolic Diameter, Interventricular septal thickness, Thalassemia major, Echocardiography.

INTRODUCTION

Thalassemia is appeared to be the common genetic disorder in which red blood cells stops functioning normally due to alteration in structure of red blood cells (RBCs). Patients' require regular blood transfusion which cause increase in serum iron level. This higher concentration of iron starts depositing in heart, liver and cells of other organ that can cause serious diseases and complications. Iron chelators used for the removal of additional iron.[1,2]

Cardiac diseases are widely associated with thalassemia due to excess accumulation of iron in heart cells. Over the last few decades, new treatment plan and therapeutic interventions has been employed which greatly influence the quality of life and well-being of thalassemic patients.[2-4] Cardiac diseases caused with thalassemia can be categorized into two clinical categories such as iron overload complications and non-iron overload complications.[5-7]

Pal et al[8] conducted a study on children with thallasemia major and divided then into two groups on basis of serum ferritin level as <2500 ng/dl and >2500 ng/dl. They found that cardiomegaly was found in 8 patients in those with ferritin level <2500 ng/dl and in 22 patients with level >2500 ng/dl. Also left ventricular hypertrophy was found in 3 patients with levels <2500 ng/dl and in 23 patients with level >2500 ng/dl. In another study, left ventricular end diastolic diameter was found to be 38.14±4.59 mm, left ventricular end systolic diameter was 25.13±3.45mm and literventricular septal thickness was 7.20±1.12 mm among those having thalassemia major.[5]

As thalassemic patients are commonly encountered in pediatric practice, however, we yet don't know about the cardiac status of these patients in our population. As these patients are unfortunately exposed to many factors including continued transfusions and higher serum iron and ferritin levels, these all make these patients more prone to develop cardiac complications. So it is important to determine the cardiac status of these patients so that we may make a holistic plan of management for them. Therefore, I want to conduct this study to unreveal its different aspects in our own population. This study will help us to find the cardiac status of these patients in our population and we will be better able to anticipate and educate our patients.

MATERIALS AND METHODS

This cross-sectional study was conducted in Pediatric Medicine Department, GMMMC Hospital, Sukkur from 2ndOctober 2020 to 1stApril 2021 and 98 patients were enrolled. All the patients of both genders with age 2-18 years having thalassemia major were included. Patients who have undergone any type of cardiac surgery before (medical records) (as the cardiac indices may be different in these patients) were excluded. Approval from Ethical Review Board of the hospital was obtained. The demographic variables including age, gender, family history of cardiac diseases, socio-economic status, other comorbid condition and duration since diagnosis of thalassemia were noted. All patients were assessed for cardiac status and data was recorded. The collected data was entered and analyzed accordingly using SPSS-21.

RESULTS

Mean age of patients was 7.73±5.01 years, mean duration since diagnosis of thalassemia was 6.12±4.80 years, mean left ventricular end diastolic diameter was 38.42±3.75 mm, mean left ventricular end systolic diameter was 24.45±2.40 mm and mean interventricular septal thickness was 6.23±1.45 mm (Table 1).

There were 65 (66.33%) males and 33 (33.67%) female patients. According to socioeconomic status, 18 (18.37%) patients have low, 52 (53.06%) patients have middle and 28 (28.57%) patients have high. Family history of cardiac disease was found in 15 (15.31%) patient and it was not found in 48 (48.98%) patients (Table 2).

Table 1: Descriptive statistics of patients (n=98)

Variable	Mean±SD
Age (years)	7.73±5.01
Duration since diagnosis of thalassemia	6.12±4.80
Left ventricular end diastolic diameter (mm)	38.42±3.75
Left ventricular end systolic diameter (mm)	24.45±2.40
Interventricular septal thickness (mm)	6.23±1.45

Table 2: Demographic information of the patients (n=98)

Variable	No.	%
Gender		
Male	65	66.33
Female	33	33.67
Socioeconomic status		
Low	18	18.37
Middle	52	53.06
High	28	28.57
Family history of cardiac disease		
Yes	15	15.31
No	83	54.69

DISCUSSION

Cardiac involvement represents an important complication of b-thalassemia major, and results in increased mortality and morbidity rates. Disease mechanisms implicate iron infiltration of the heart structures due to frequent blood transfusions. Transfusion-dependent patients receive 20 times the normal intake of iron, which leads to iron accumulation and damage in the liver, heart, and endocrine organs.[1] Although iron chelating therapy has markedly improved outcomes, cardiac failure remains an important cause of death in thalassemic patients.[2]

Tendency of cardiovascular complications are higher in patients who were poorly chelated or not chelated at all. Late and inappropriate diagnosis could also prove fetal because both share common symptoms. Therefore, each and every symptoms needs to be rule out carefully.[3] Timely diagnoses would minimize the risk of heart failure and saves patients' life. Echocardiography is considered as a potential tool for exact diagnosis as it is a non-invasive procedure.[4]

In present study, 98 diagnosed cases of thalassemia were enrolled. In present study, the mean LVDD was 38.41±3.74 mm, LVESD was 24.4±2.40 mm and intraventricular septal thickness (IVST) was 6.23±1.44 mm. Maheshwari et al[5] in a similar study, evaluated the echocardiographic parameters in thalassemia patients and reported LVDD in 39.25±6.47 mm, LVESD in 25.23±4.62 mm and IVST in 7.95±1.43 mm.[5]

Another study by Manzoor et al[6] reported LVDD 45.31±5.34 mm, LVESD in 29±4.12 mm in patients of thalassemia major and LV dysfunction. Diastolic functions impairment is observed in large number of patients. This fact has been investigated in various studies and proved that, serum ferritin level was significantly higher in patients who showed impairment of diastolic parameters.[7-9] Higher E/A ratio was the common echocardiographic evaluations in majority of the patients.[10,11]

CONCLUSION

Left ventricular diastolic dysfunction (LVDD) occurred in all studied patients of thalassemia major. There was no systolic dysfunction and intraventricular septal thickening in thalassemia major patients.

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