ORIGINAL ARTICLE

Echocardiographic Findings In Thalassemia Major Patients: An Experience From Jammu

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Abstract

Current Study was done to assess the echocardigraphic changes in Thalassemia Major patients. All Thalassemic Major patients above 10 years of age registered with Thalassemia day care centre at SMGS hospital, Government Medical College Jammu with normal electrocardiogram and no clinical features of heart failure were included in this retrospective study. Their medical records were analyzed in terms of age, gender, average hemoglobin levels, serum ferritin levels and echocardiographic findings. 95 patients who were more than 10 years of age and had normal electrocardiogram and no clinical features of heart failure were included in this study. Out of these 95 patients 55 showed abnormal findings on echocardiography. All patients except one showed normal systolic function. Echocardiography in 54 patients showed only diastolic dysfunction and one had tricuspid regurgitation (TR) in addition . 55 Thalassemia major patients with abnormal echocardiographic findings comprised of 30 males and 25 females. All the patients in the age group of more than 20 years and 12 patients in the age group of 10 years to 20 years showed abnormal echocardiographic findings. Average hemoglobin and serum ferritin in these children were around 5-7gms /dl and more than 3000 ng/ml respectively. Echocardiographic findings suggestive of left ventricular diastolic dysfunction were the commonest findings indicating early cardiac dysfunction in patients with Thalassemia major in 2nd and 3rd decade of their life.

Key Words

Echocardiography, Thalassemia, Serum Ferritin, Diastolic Dysfunction

Introduction

Thalassemia is one of the most common genetic disorders.(1) Approximately 1 lakh children with thalassemia major are born world over every year, of which 10,000 are born in India. The carrier rate of beta thalassemia gene is around <1 - 17% in India. (2,3) Beta thalassemic patients need regular blood transfusion for their survival which leads to iron overload. This excess of iron gets deposited in heart, liver, endocrinal glands and other organs and iron chelators are required to remove this excess of iron. Cardiac disease constitutes the major cause of death in thalassemics.In last few decades the quality and duration of life of patients with thalassemia has improved.(4,5) It is now expected that with adequate and regular care a patient with thalassemia will live a good quality life even beyond middle age and can raise a family of their own too. Earlier the major complication affecting the heart was heart failure due to accumulation of iron within heart muscle cells but now other complications of thalassemia have also become apparent with increased survival due to better care. The

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cardiovascular complications of thalassemia can be considered in two major clinical categories: 1. Iron overload complications (Reversible myocyte failure, Arrhythmia, including heart block, Arterial changes - loss of vascular compliance.). 2. Non-iron overload complications (Pulmonary hypertension, Arrhythmia - particularly Atrial fibrillation later in life, Thrombotic stroke, linked to AF, Cardiac function changes due to restriction/diastolic dysfunction/fibrosis, Arterial changes - loss of vascular compliance). (6)

The Current study was done to assess the echocardigraphic findings in Thalassemia Major patients.

Material And Method

All Thalassemia Major patients above 10 years of age registered with Thalassemia day care centre at SMGS hospital, Government Medical College Jammu with normal electrocardiogram and no clinical features of heart failure were included in this retrospective study. Their medical records were analyzed in terms of age, gender, average hemoglobin levels, serum ferritin levels and echocardiographic findings. All thalassemic patients more than 10 years of age were subjected to echocardiography every year on routine basis. Their clinical data and echo findings (both M mode and Doppler indices) were analyzed.

Results

Out of a total of 279 Thalassemia patients registered with thalassemia day care centre, SMGS hospital Government Medical College Jammu 95 patients who were more than 10 years of age and had normal electrocardiogram and no clinical features of heart failure were included in this study.

These 95 patients comprised of 52 males and 43 females with a male to female ratio of 1.2:1. Majority 52 (54.7%) of them were in the age group of 11 to 20 years followed by 43 (45.2%) in the age group of more than 20 years.

Out of 95 patients with thalassemia major 55 showed abnormal findings on echocardiography. All patients except one showed normal systolic function. Echocardiography in 54 patients showed only diastolic dysfunction and one showed tricuspid regurgitation (TR) in addition. Out of these 55 Thalassemia major patients with abnormal echocardiographic findings 30 were males and 25 females. All the patients in the age group of more than 20 years and 12 patients in the age group of 10 years to 20 years showed abnormal echocardiographic findings. Average hemoglobin and serum ferritin in these children were around 5-7gms /dl and more than 3000 ng/ml respectively.

Discussion

In Thalassemics heart failure due to anemia can occur at any age but heart failure due to cardiomyopathy occurs usually in or after second decade of life. Due to regular blood transfusions iron starts getting deposited in the myocardium causing cardiac disorders in the later part of their life. On the other hand children on irregular and inadequate treatment will have chronic anemia which leads to development of sign and symptoms of hyperdynamic circulation finally resulting into congestive cardiac failure. The anemia and ineffective erythropoesis along with iron overload, nutritional deficiencies, chronic oxidative stress and lung diseases have their effect on cardiac changes in thalassemia major.(7)

Therefore in thalassemic children who are poorly chelated or unchelated heart failure can occur even in first decade of life. The diagnosis of heart failure in thalassemic patients is often difficult due to presence of already existing symptoms of anemia such as exercise intolerance, fatigue and hepatomegaly. So their baseline clinical status should be recorded for future reference and any change in symptoms should be seen as a red

Table 1. M Mode and Doppler indices on echocardiography measurement in Thalassemic patients with abnormal findings (N=55).

Parameter	Mean value	Standard deviation
LVEDD(mm)	45.31	5.34
LVESD(mm)	29	4.12
EF%	68.09	4.19
FS%	32.17	2.12
E/A	2.13	0.71
Dt (msec)	120	20.24

Table 2. M Mode and Doppler indices on echocardiography measurement in Thalassemic patients with normal findings (N=40).

Parameter	Mean Value	Standard Deviation
LVEDD (mm)	44.32	4.12
LVESD (mm)	28.03	3.23
EF%	70.06	4.56
FS%	36.42	3.02
E/A ratio	1.62	0.23
Dt (msec)	141.62	30.41

signal such as increase in dyspnea on exertion. Similarly a patient not tolerating the regular blood transfusion should also undergo cardiac investigations to rule out heart failure.(7)

So early detection of cardiac dysfunction is very important for preventing myocardial damage and echocardiography is an important noninvasive tool to detect early cardiac dysfunction changes.(7,8)

In this study we evaluated the patients with thalassemia major with normal electrocardiogram and no sign/ symptoms of heart failure for cardiac dysfunction (systolic function and diastolic indices of the left ventricle) by echocardiography.

A total of 95 patients with thalassemia were enrolled in this study comprising of almost equal number of males and females similar to the observation made by Yuksel *et al*.(9) All of the enrolled patients except one had normal systolic LV function on echocardiography.

Out of 95 enrolled thalassemia major patients 40 had normal echocardiographic findings. Their hemoglobin and serum ferritin were 9-11 gms% and less tha 2000 ng/ml. 55 patients showed abnormal echocardiographic findings suggestive of diastolic LV dysfunction. The average hemoglobin in these children was 5-7gms/dl and average serum ferritin was more than 3000 ng/ml. Yuksel also reported a negative association between S. ferritin levels and LV dysfunction. (9)

The diastolic indices in these patients showed higher early diastolic filling of LV and E/A ratio suggestive of restrictive diastolic pattern and stiff myocardial wall. Garadah *et al* , Yaprak *et al* also reported similar findings. (10,11) Silviliarat *et al* also reported impaired diastolic function parameters (E/A ratio and deceleration time) in patients with serum ferritin higher than 2500 ng/ml.(12)

Spirito et al observed in their study that patients with beta thalassemia major had restrictive pattern with no heart failure.(13) Oki *et al.* also reported that high E/A ratio was the most common echocardiographic finding in beta thalassemia major patients.(14)

Conclusion

Echocardiographic findings suggestive of left ventricular diastolic dysfunction i.e high early diastolic filling velocity, increased E/A ratio and decreased deceleration time were the commonest findings indicating cardiac dysfunction in patients with Thalassemia major in 2nd and 3rd decade of their life in absence of clinical features of heart failure.

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