

# FREQUENCY OF HEART FAILURE IN PATIENTS WITH BETA THALASSEMIA MAJOR

Fazal-ur-Rahman Khan<sup>1</sup>, Muhammad Amin Jan Mahsud<sup>2</sup>, Tariq Ayub<sup>1</sup>,  
Muhammad Hussain Khan<sup>3</sup>, Syed Humayun Shah<sup>2</sup>

<sup>1</sup>Department of Paediatrics, <sup>2</sup>Pathology and <sup>3</sup>Community Medicine, DHQ Teaching Hospital, Gomal Medical College, D.I. Khan, Pakistan

## ABSTRACT

**Background:** Estimated prevalence of beta thalassemia is 3-8% in Pakistan. Over the past three decades, regular blood transfusions have significantly increased the survival of these patients. There has been an increase in the frequency of complications, mainly caused by iron overload. The aim of this study was to determine the frequency of heart failure in patients with beta thalassemia major in this region.

**Material and Methods:** It was a descriptive audit of ward record, carried out in the Department of Paediatrics, District Headquarter Teaching Hospital, D.I.Khan, Pakistan, from January 2004 to December 2005. Case records of all beta thalassemia major patients admitted during this period were analyzed for heart failure. Clinical data recorded was age, sex and cardiac status of patients. Investigations including ECG, chest x-ray and echocardiography were evaluated.

**Results:** Two hundred and twelve patients of beta thalassemia were admitted during the study period. Clinical congestive cardiac failure was observed in 33 (15.6%) patients with the age range of 8-21 years. Fifty four patients (25.5%) had cardiomegaly on chest x-ray but were not having clinical heart failure. Age range of these patients was 5-12 years. As a whole heart failure and cardiomegaly on chest x-ray was observed in 87 (41.1%) patients. All these patients were above the age of 5 years.

**Conclusion:** Heart failure is not uncommon in patients with beta thalassemia major in our setup. Regular transfusions and chelation therapy are recommended to reduce this complication.

**Key words:** Beta thalassemia, Heart failure, Cardiomegaly.

## INTRODUCTION

The estimated prevalence of beta thalassemia is 16% in Cyprus, 3-14% in Thailand and 3-8% in India, Pakistan, Bangladesh and China. Prevalence is low in African black people (0.9%) and Northern Europe (0.1%).<sup>1</sup> Over the past three decades, hyper-transfusion therapy has significantly increased the survival and prognosis of patients with beta thalassaemia. At the same time there has been an increase in the frequency of complications, mainly caused by iron overload; both because of repeated transfusions and because of increased gut iron absorption.<sup>2,3,4</sup> Cardiac complications of iron overload represent the main determinants of survival and primary cause of death in these patients.<sup>5,6,7</sup> Iron chelation therapy can eradicate or at least reduce cardiac and other complications but compliance with it is very deficient.<sup>8</sup> Heart disease can occur even in thalassemia intermedia, where no or minimal blood transfusions are needed.<sup>9</sup>

The aim of this study was to determine the frequency of heart failure in patients with beta thalassemia in this region.

## MATERIAL AND METHODS

It was a descriptive audit of the ward record, carried out in the Department of Paediatrics, District Headquarter Teaching Hospital, Gomal Medical Collage, Dera Ismail Khan, Pakistan, from January 2004 to December 2005. The case records of all the beta thalassemia major patients admitted during this period were analyzed for heart failure. Cases of beta thalassemia minor and intermedia were excluded. Clinical data recorded was age, sex and cardiac status of the patients. Investigations including ECG, x-ray chest and echocardiography were evaluated.

Results were analyzed by descriptive statistical methods.

**Table-1: Age and sex distribution of patients.**

|        | < 10 Years | 10-20 years | > 20 Years | Total       |
|--------|------------|-------------|------------|-------------|
| Male   | 87 (41%)   | 42 (19.8%)  | 2 (1%)     | 131 (61.8%) |
| Female | 51(24%)    | 29 (13.7%)  | 1 (0.5%)   | 81 (38.2%)  |
| Total  | 138 (65%)  | 71 (33.5%)  | 3 (1.5%)   | 212 (100%)  |

## RESULTS

During the study period of 2 years, 212 patients of beta thalassemia were admitted; 131 (61.8%) patients were male and 81 (38.2%) female. The youngest patient at diagnosis was 4 months old and the oldest patient of the study was 21 years old. Majority of the patients i.e. 138 (65%) were below 10 years of age. (Table-1)

Congestive cardiac failure was observed in 33 (15.6%) patients with the age range of 8-21 years. Echocardiography was performed in 21 of these patients and all of them had left ventricular systolic failure and diastolic dilatation. Fifty four (25.5%) patients had cardiomegaly on chest x-ray but they were not having other clinical features of heart failure. Age range of these patients was 5-12 years. Echocardiography was not performed in these patients. One of these patients had supraventricular tachycardia on ECG. As a whole heart failure and cardiomegaly on chest x-ray were observed in 87 (41.1%) patients. All these patients were above the age of 5 years.

## DISCUSSION

Cardiac failure is the commonest and leading cause of death in beta thalassemia.<sup>5,10</sup> In the present study the incidence of heart disease, particularly that of congestive cardiac failure is more than that of 13.5% reported from Greece.<sup>9</sup> The reasons are not clear but are probably multiple and include less frequent transfusions, lower pre-transfusion hemoglobin level and inadequate chelation therapy.<sup>11</sup> Another study from Greece has reported even lesser incidence of heart disease (3.8%), but it was on patients with beta thalassaemia intermedia in whom minimum transfusions are required.<sup>10</sup> It has also been documented that the presence of certain major histocompatibility antigens /alleles may protect (HLA-DRB1\*1401) or predispose (HLA-DRB1\*0501) to the development of congestive cardiac failure.<sup>10</sup> Other as yet unidentified factors (genetic, immune or infective) may also be important in the causation of congestive cardiac failure.<sup>10</sup> In some patients pericarditis has also been reported in association with congestive cardiac failure.<sup>5</sup>

Mean age of our patients with congestive cardiac failure was significantly lower than that of 20-25 years reported by Aessopes et al.<sup>5</sup> Chronic anaemia, malnutrition and iron overload because of inadequate chelation therapy are the probable reasons. Also in thalassaemia intermedia heart failure is mainly due to pulmonary hypertension,<sup>9</sup> while in the present study beta thalassaemia intermedia and minor were excluded. This may be one of the reasons for higher incidence of heart failure in the present study.

Although it was a retrospective study but it showed that heart failure is more common in our set up due to many reasons. Regular transfusions, chelation therapy and monitoring (comprehensive care) will decrease the rate of heart failure in beta thalassaemia but the possibility of genetically defined immune mechanisms playing a role in the pathogenesis of heart failure can not be ignored.<sup>10</sup>

## CONCLUSION

Heart failure is not uncommon in patients with beta thalassemia major in our setup. Regular blood transfusions and chelation therapy are recommended to reduce this complication.

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**Address for Correspondence:**

Dr. Fazal-ur-Rahman Khan  
Assoc. Prof. Paediatrics  
Gomal Medical College  
D.I.Khan, NWFP, Pakistan  
Cell: 03005792203  
Email: fazaldr@yahoo.com