Summary & Conclusions

Beta-thalassemia is a major health problem in Egypt and other countries. It constitutes 39% of the total hematological patients and 84% of chronic hemolytic pediatric patients. Over 1400 of the annual 1.5 million newborns are expected to be affected with this disorder.

Repeated blood transfusion is the main option in keeping those patients alive, but it has complications of increased iron burden and consequent multi-organ affection due to deposition of iron in their tissues. Patients with B-thalassemia who develop pulmonary hypertension (PHT) frequently have cardiac and pulmonary deterioration, which can result in significant morbidity and mortality.

While PHT is increasingly recognized as a part of the clinical spectrum for B-thalassemia, little is understood about the mechanisms and risk factors for its development. Consequently, prevention and treatment efforts are inadequate. Most patients do not undergo routine monitoring of right heart pressure to detect PHT, and reports of treatment efficacy have been anecdotal.

There are currently limited data on the effect of any treatment modality as data supporting a selective treatment approach have been insufficient. Several studies have suggested that chronic hypoxia and lung injuries due to infections and iron depositions are common causes.whereas others suggest a hypercoagulable state causing thrombotic lesions in the lungs as a risk factor.

This study was designed to compare between splenectomized and non splenectomized patients with transfusion-dependant β-thalassemia major through echocardiographic study and its correlation with hematological data.

This study was conducted on 30 cases with beta thalassemia major (TM) of those attending to pediatric hematology clinic, Damnhour national inistitute. Fifteen patients were surgically splenectomized and the other half wase none splenectomized.

The following parameters were used:

1- Echocardiography for all patients.

From these echo parameters we calculate E\A ratio , FS , level of PHT,IVSTS,IVSTD,LVSD,LVSS,LVPWT,Aortic root diameter and Left atrium diameter.

2-Laboratory investigation:

Including CBC,Retics,Serum ferritin,Blood suger level,Liver function tests(AST,ALT),Kidney function tests(blood urea,serum creatinine).

The results of this study revealed that:

- Splenectomized patients with TM are at high risk of having impaired diastolic left ventricular function and this can occur in patients with reserved left ventricular systolic function.
- PHT exists in significant numbers in splenectomized patients with TM than non splenectomized patients.

- Serum ferritin levels were more increased inTM patients with raised PHT, indicating that iron overload is a major contributing factor for development of PHT.
- Transfusion interval was shorter in non splenectomized patients with TM than splenectomized group which show increase in total numbers of blood transfusion and the possibility of iron overload and infection with HCV increases.
- Tissue Doppler echocardiography is important for early diagnosis of cardiomyopathy in TM patients especially above nine years.
- Diastolic dysfunction usually proceedes the systolic ventricular dysfunction.It may be an alarming sign.

Conclusion

The removal of the spleen stabilizes the hemoglobin concentration at higher levels and reduces transfusion needs. This may contribute to the prevention of additional cardiac deterioration by limiting myocardial hypoxia and iron overload.

But splenectomy increases the risk of development of PHT. These results may be useful for the design of therapeutic interventions, since early application of intensive therapy inTM have been shown to prevent the complications related to chronic anemia.