Chapter 4

Discussion

Echocardiography has been providing a simple noninvasive means of assessing cardiac structure and function. Left ventricular ejection fraction was normal (65-85%) in all but five patients in our study. As demonstrated earlier (8), the resting ejection fraction and percent fractional shortening of our patients were similar and did not differ statistically from that found in the normal group, although the thalassemic patients tended to have a lower EF. Although B-thal/HbE patients have larger hearts, stroke index is high and the cardiac index increased. LV systolic function was normal in almost all patients, even in the patient who died in severe congestive heart failure within 3 months of the non-invasive study. Cardiologist did not have the opportunity to perform echocardiography at the time of the terminal episode; it is possible that serial echocardiographic studies may reveal terminal deterioration of LV performance in thalassemic patients dying of cardiac failure. Nonetheless, systolic function of the LV is preserved until very late in the course of the disease. It could be argued that the presence of a larger heart without a further increase in systolic function (as compared between cases and controls) indicates an incipient abnormality of LV systolic performance in the patients, but these changes are small and are certainly not those of a severe congestive cardiomyopathy. Diastolic function of the LV is difficult to quantitate by non-invasive technique and was not studied in these patients. So, common symptom of dyspnea in these patients may be due to anemia, diastolic LV dysfunction, chronic pulmonary arterial thromboembolism and pulmonary hypertension etc. Most patients show hyperkinetic heart and exhibit variable cardiac chamber enlargement (Table 5). The relative contributions of low hemoglobin level and

tissue hypoxia versus myocaridal iron deposition in producing this change can not be readily determined in our study. Stroke volume and cardiac output are increased (Table 6.2). Chronic anemia in man usually increases the cardiac output when the hemoglobin level is 7 gm per 100 ml of blood or less (35). In this aspect, like other chronic anemia, B-thal/HbE patients increase their cardiac output principally by increasing cardiac stroke volume, since tachycardia is frequently not found.

Nienhuis, A.W.et al (17), suggested that cardiac function of thalassemia-major patients as reflected in the resting ventricular ejection fraction were well preserved during most of the course of cardiac iron deposition and that a fall in resting EF was a rather ominous prognostic sign. Their all four patients with low EF died within a short period of time. In contrast to their study, our five patients with low EF (< 65%) tolerated quite well. Meanwhile, another patient with EF of 80% and intermittent circulatory congestive state died one month later with congestive heart failure. Thus, some caution must be exercised in interpreting the significance of low EF in B-thal/HbE patients.

Our data concerning cardiac arrhythmia is quite limited because we performed only 12 leads cardiogram. Supraventricular premature beats was reported to be the most common arrhythmia in patients over the age of 14 (9). By 12 leads cardiogram we detected 1 case with atrial premature beat in a patient aged 17 and 1 case of premature ventricular beats in a patient aged 29. Neither case has other ECG abnormality.

The most frequent abnormal ECG findings in our thalassemic patients are ventricular hypertrophy (6 cases) and ST- T wave changes (2 cases). These abnormal ECG findings were reported to be associated with the patients who were younger, more anemic and had bigger heart size (36). If definite LVH , probable right ventricular hypertrophy, probable biventricular hypertrophy and nonspecific ST-T wave changes were considered abnormal ECG; the age, average hemoglobin

level, cardiothoracic ratio, serum ferritin, and serum iron of the patients with normal and abnormal ECG were not different : 26.3±7.8 vs 24.8±10.5 years, 7.0±1.3 vs 6.6±1.9 gm/dl, 52.8±6.6 vs 53.1±7.7 percent, 2932.4±2261.3 vs 2612±3301.3 ug/litre and 225.6±66.2 vs 226.2±127.1 ug/decilitre respectively ($p > 0.05$).

Splenectomy was found to be associated with pulmonary thrombus in an autopsy study(29). We found that pulmonary hypertension detected from chest x-ray was related to splenectomy but causal relationship can not be concluded by such a crossectional study. It may be that splenectomy is usually performed in a severe case, and that severe case (by some unknown mechanism) is more likely to develop pulmonary hypertension than a mildly affected one.

Though roentgenogram identified a large number of ventricular hypertrophy (20 cases), this was not to be true by electrocardiogram. This may be due to the strict criterion of ECG diagnosis applied and/or counter balance of force from right ventricular hypertrophy (usually secondary from chronic pulmonary thromboembolism common in B-thal/HbE patients) and left ventricular hypertrophy or enlargement secondary to anemia. Further investigation on pulmonary hypertention, diastolic LV function, systolic LV function at the terminal stage of disease, early non-invasive detection of LV dysfunction are needed for better care of these patients.