Cardiac determinants of survival in beta-thalassemia

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We read with great interest the article by Telfer et al concerning survival of medically treated thalassemia patients in Cyprus.[1] Although the authors have analyzed data on a large cohort of patients with beta-thalassemia and presented very encouraging data on survival for the patients who were followed-up after the year 2000, there are some issues to be addressed. It is well known that congestive heart failure is the main cause of death in transfusion-dependent patients with beta-thalassemia. Cardiac function in these patients remains normal for many years, but once symptoms of heart failure become evident, death usually occurs within 1 year.[2,3] This is in accordance with the results reported by Telfer *et al*,[1] where the 53.4% of deceased patients died due to cardiac causes. Although cardiac death is the most frequent cause of death in patients with beta-thalassemia, early markers of cardiac function with prognostic impact on survival have not yet been adequately defined. In a recently published study we investigated the impact of the Doppler-demonstrated left ventricular restrictive filling pattern on survival in a cohort of 45 asymptomatic adult patients with beta-thalassemia and normal left ventricular systolic function throughout a 15-year long observation period.[4] Our data indicates that the presence of left ventricular restrictive filling pattern predicts worse prognosis in patients with beta-thalassemia. The Kaplan – Meier survival analysis showed that the mean survival in patients with restrictive filling pattern was 11.1±1.2 years (95% CI: 8.7 to 13.4 years), compared to 14.3±0.5 years (95% CI: 13.3 to 15.2 years) in patients with non-restrictive filling pattern. The 15-year cumulative survival rate was 58% in patients with restrictive filling pattern and 88% in patients with non-restrictive filling pattern (log-rank statistic=6.02, p=0.014). We also found that patients' poor compliance with chelation therapy was significantly associated with restrictive filling pattern (p=0.007). In conclusion, Doppler-echocardiography is the main tool in the era of long-term follow-up of patients with beta-thalassemia and should be performed routinely in these patients. It would be very interesting if the authors of this study, which includes more than 500 patients with beta-thalassemia, provided the Doppler-echocardiographic data of the patients who died due to cardiac causes.

Efthimiadis GK,[1] Hassapopoulou HP,[2] Tsikaderis DD,[1] Karvounis HI,[1] Giannakoulas G,[1] Parharidis GE[1]

[1]Department of Cardiology, Aristotle University, AHEPA Hospital, Thessaloniki, Greece

[2]Thalassemia Unit, AHEPA Hospital, Thessaloniki, Greece

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