Dear Editor,

We read with great interest the recent paper entitled ‘A comparison of heart function and arrhythmia in clinically asymptomatic patients with beta thalassemia intermedia and beta thalassemia major’ by Amoozgar et al. [1] in Hematology journal. In their well-designed paper the authors compared the heart function and arrhythmias in clinically asymptomatic patients with beta thalassemia intermedia and beta thalassemia major (β-TM). According to their results, both atrial and ventricular arrhythmias were more common in beta thalassemia major than in beta thalassemia intermedia patients. Consequently, the authors suggested to perform serial ECG Holter monitoring in asymptomatic patients with beta thalassemia major and intermedia for the early diagnosis and the appropriate treatment of cardiac arrhythmias. Amoozgar et al. [1] by analyzing of the short-time 24-hour Holter recordings detected atrial fibrillation (AF) in one β-TM patient only (1.6%) among their study cohort, much lower than those reported in previous studies [2–4].

In the literature, little is still known about the true occurrence of atrial fibrillation in β-TM patients, because there are many heterogeneous studies including small number of patients, with different clinical characteristics, investigated using various methods for detecting AF. For the high supraventricular arrhythmias risk and its consequences, the early identification of β-TM patients with atrial fibrillation risk is of pivotal importance for the medical therapy optimization [5]. Non-invasive electrocardiographic and echocardiographic markers are useful in clinical practice to predict arrhythmias in some clinical conditions [6–15], including β-TM [9,10] and muscular dystrophy [11–15]. Our first observational study showed an atrial fibrillation incidence of 14% in β-TM cohort without cardiac dysfunction underwent ECG Holter monitoring every 3 months for 1 year follow-up. According to our results, a P wave dispersion (PD) cut-off value ≥35.5 ms and maximum P duration (P max) cut-off value ≥111 ms identified high-risk atrial fibrillation β-TM patients who needed a careful cardiac monitoring [2]. In a recent long-term prospective observational study [3], we revealed an overall AF prevalence of 20% in a large β-TM-population with normal cardiac function underwent long-period external loop recorder (ELR) monitoring every 6 months during the 5 years follow-up. We found that a cut-off value of 44.8 ms for inter-atrial electromechanical delay or intra-left AEMD cut-off value of 40.1 ms had a sensitivity of 81.2% and a specificity of 98.7% in identifying β-TM patients at increased AF risk.

Bell et al. [4] in a cross-sectional analysis of 80 β-TM patients followed over 12-month period showed an electrocardiographic evidence of atrial fibrillation in 27 patients, providing an overall prevalence of 33.8% within their cohort.

In consequence of the high atrial fibrillation occurrence in beta thalassemia major and the negative effects of cardio-embolic stroke in β-TM patients [16], the early identification of β-TM subgroup at high AF risk is of pivotal importance for the optimization of the clinical follow-up and medical therapy. We suggest to implement in our daily clinical practice the measurement of P wave parameters (P max and PD) on the electrocardiogram and the evaluation of the atrial electromechanical delay indices (inter-AEMD and intra-left AEMD) during the thoracacic echocardiogram in β-TM patients [17]. The β-TM subgroup who showed a PD cut-off value ≥35.5 ms or a P max cut-off value ≥111 ms at electrocardiogram or an inter-AEMD cut-off value of 44.8 ms or intra-left AEMD cut-off value of 40.1 ms should be considered at high AF risk and closely followed. In particular, we prefer to perform in this β-TM subgroup seriate long time ELR monitoring, even when the cardiac function is conserved, that proved to yield a higher diagnostic value than conventional electrocardiographic evaluation or Holter monitoring [18].

The early detection of AF onset offers us the opportunity of prophylactic antiarrhythmic therapy for patients with symptomatic AF and anticoagulant treatment for both symptomatic and asymptomatic AF β-TM subjects.

In conclusion for the several prognostic implications of AF in β-TM, we think the time is now ready to share worldwide databases on arrhythmic involvement in β-TM patients and to establish an evidence-based management and treatment of arrhythmic disorders in β-TM population.

Disclosure statement

No potential conflict of interest was reported by the authors.

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