

The clinical manifestation of hereditary thrombophilia in patients with unprovoked venous thrombosis

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Background and Aims: Hereditary thrombophilia (HT) is a condition increasing the risk to develop venous thrombosis. The prevalence of HT varies among different ethnic groups, but there are few studies of that in Korean patients. We aimed to estimate the frequency and clinical characteristics of HT in Korean patients with unprovoked VTE. **Methods:** A total of 369 consecutive patients with thromboembolic event who underwent thrombophilia tests were reviewed retrospectively. Arterial thromboembolism and VTE with predisposing factors were excluded. Patients with acquired thrombophilia were also excluded. Patients who had low levels of natural anticoagulants were assessed by DNA sequencing of the corresponding gene to confirm HT. **Results:** Among 222 Korean patients with unprovoked VTE, 66 patients were suspected HT. 62 of 66 underwent genetic molecular test and 33 were finally confirmed to have HT (53.2%; 33/62). The most common type was antithrombin III deficiency (14 of 222, 6.3%) followed by protein C deficiency (12, 5.4%), protein S deficiency (4, 1.8%) and dysplasminogenemia (3, 1.4%). Compared with patients without HT, those with HT were associated with higher proportion of male (69.7% vs 47%; $p=0.013$), younger age (37 [32-50] vs 52 [43-65] years; $p=0.000$), more history of VTE (57.6% vs 31.7%; $p=0.004$) and family history (43.8% vs 1.9%; $p=0.000$). Multivariate analysis showed age <45 years and presence of family history were strong predictor of unprovoked VTE with HT (odds ratio [OR]=9.435; 95% confidence interval [CI]=2.45-36.35; $p=0.001$ and OR=92.667; 95% CI=14.95-574.29; $p=0.000$, respectively). The cumulative incidence of recurrent VTE were not significantly different in both two group ($p=0.987$), however all two events of recurrence in HT group have occurred under oral anticoagulant treatment. **Conclusion:** About 15% of patients with unprovoked VTE had HT, and antithrombin III deficiency was the most common cause. Above all unprovoked VTE patients who are less than 45 years old and have positive family history of VTE should be tested for HT. Furthermore, we need to closely monitor the recurrence of VTE despite maintaining anticoagulation, especially in patients with HT.

Prognostic Value of Electrocardiographic Parameters and Longitudinal Strain in Cardiac Amyloidosis

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Background: Abnormal electrocardiographic findings in cardiac amyloidosis were suggested that they could predict mortality. Also longitudinal strain was a strong prognostic factor in comparison with any other echocardiographic parameter. The aim of the present study was to confirm a prognostic value of abnormal electrocardiographic findings and longitudinal strain. **Methods:** A total 32 patients with cardiac amyloidosis and mean left ventricular wall thickness ≤ 14 mm were enrolled retrospectively. A longitudinal strain was measured by 2-D speckle tracking technique in basal and global segment. A standard 12-lead electrocardiography was reviewed to find abnormal findings. Patients were divided into two groups according to the presence of all-cause mortality ($n=20$), or the absence of all-cause mortality ($n=12$). All electrocardiographic abnormalities and longitudinal strain were analyzed and the relationship between electrocardiographic abnormalities and longitudinal strain also confirmed. We invented the ECG Scoring System additionally to predict mortality. **Results:** The prevalence of abnormal electrocardiographic findings was similar to results of previous studies, except for interventricular conduction delay. All abnormal electrographic findings were not associated with adverse outcomes directly. Patients with all-cause mortality reported higher N-terminal pro-brain natriuretic peptide levels (16286 pg/ml vs 6174 pg/ml, $p<0.002$), lower average basal longitudinal strain (5.8 vs 9.1, $p<0.008$) and lower global longitudinal strain (8.6 vs 11.2, $p<0.009$). Multivariate analysis showed that the average basal longitudinal strain and NT pro-BNP levels were the only independent predictor of all-cause mortality. Among abnormal electrocardiographic findings, low voltage in limb leads, pseudo-infarction, extreme QRS axis deviation, and Total ECG Score were related with poor longitudinal strain values. **Conclusions:** The average basal longitudinal strain was an independent prognostic factor in cardiac amyloidosis. Low voltage in limb lead, pseudo-infarction, and Total ECG Score has the possibility of predicting poor outcomes in cardiac amyloidosis but, need larger study to prove it.