THE PREVALENCE AND RISK FACTORS FOR CARDIAC DISEASE IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER

Y. Brutbul1, L. Gabay2, Y. Braun-Moscovic1,2,3. Pediatric Rheumatology Service, Ruth Rappaport Children's Hospital, Rambam Medical Center, Haifa, Israel, Pediatric B, Haifa, Israel; 1The Ruth and Bruce Rappaport Faculty of Medicine, Technion - Israel Institute of Technology, Haifa, Israel, 2The Ruth and Bruce Rappaport Faculty of Medicine, Haifa, Israel; 3Maccabi Institute for Research and Innovation, Maccabi Healthcare Services, Tel Aviv, Israel.

Background: Familial Mediterranean fever (FMF) is a genetic disorder manifest by recurrent attacks of peritonitis, pleuritis and arthritis, and characterized by clinical and laboratory evidence for localized and systemic inflammation. Colchicine treatment usually prevents the attacks and the associated inflammation. Inflammation may play an important role in the initiation and progression of atherosclerosis. Recently colchicine was suggested as a therapy that help to prevent coronary heart disease.

Objectives: To study the effect of FMF and colchicine treatment on the cardiovascular morbidity and the overall mortality.

Methods: We studied using the data base from health insurance in Israel (Maccabi Healthcare Services-MHS) the presence of IHD and its risk factors in 492 FMF patients aged 40 years or more, and in a control groups matched by age gender and socioeconomic status.

Results: The incidence of cardiac disease in FMF patients was similar to the control group (6.5% vs 5.7% p = 0.594), smoking kidney disease and gout were higher in FMF compared to the control group (16% vs 12.6% p = 0.022, 9.3% vs 5.1 p = 0.01 and 4.5% vs 2.2% p<0.001 respectively), but hypertension and diabetes were similar. The overall mortality in average follow up of 3174.37 ±1738.84 days was similar in both groups.

Conclusion: The incidence of cardiac disease among FMF patients was not increased compared to the control group, despite the exposure to recurrent inflammation. We suggest that colchicine may have a protective role in these patients. Further studies are required.

Disclosure of Interests: None declared

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PULMONARY ARTERY WALL THICKNESS IS INCREASED IN BEHÇET’S DISEASE

S. Kutlu Ali Asag˘ ir1, M. Sunbul2, D. Kocakaya3, S. Kayacic, H. Direskeneli1, F. Alibaz-Oner1, 1Marmara University Faculty of Medicine, Department of Internal Medicine, Division of Rheumatology, Istanbul, Turkey; 2Marmara University Faculty of Medicine, Department of Cardiology, Istanbul, Turkey; 3Marmara University Faculty of Medicine, Department of Pulmonology, Istanbul, Turkey.

Background: Behçet’s Disease (BD) is a unique systemic vasculitis that mainly involves veins, in contrast to other vasculitides [1, 2]. Prior studies showed that pulmonary arteries have a similar structure with systemic veins in terms of widening, thin-walled, increased compliance, and low resistance [3]. We have recently shown increased venous wall thickness in lower extremities veins of BD patients.

Objectives: In this study, we aimed to assess pulmonary artery (PA) wall thickness by transthoracic echocardiography (TTE) in BD compared to healthy controls and patients with non-inflammatory pulmonary embolism (NIPE).

Methods: Patients with BD (n=77), NIPE (n=33) and healthy controls (n=57) were included in the study. PA wall thickness was measured with TTE by a cardiologist blinded to cases. PA wall thickness was measured from the mid-portio of the main PA (approximately 1 to 2 cm distal to the pulmonary valve) as demonstrated in Figure 1.

Results: PA wall thickness was significantly lower in controls (0.36 mm (SD:0.03) compared to NIPE (0.44 mm (SD:0.05) and BD (0.44 mm (SD:0.00) (p<0.001 for both). PA wall thickness was also found to be significantly higher in BD patients with major organ involvement (0.47 mm (SD:0.04) compared to healthy controls and NIPE (p<0.001 and p=0.006, respectively). PA wall thickness was similar between BD and NIPE (p= 0.6). Among patients with BD, PA wall thickness was significantly lower in patients with only mucocutaneous involvement compared to patients with major organ involvement (0.37 mm vs 0.47 mm, p= 001), it was also similar between patients with only mucocutaneous involvement and healthy controls (0.37 mm vs 0.36mm, p= 0.3). PA wall thickness was comparable between patients with vascular and non-vascular major organ involvement (0.46 mm vs 0.47 mm, p= 0.3). Patients with vascular and non-vascular major organ involvement had significantly higher PA wall measurements compared with NIPE patients (p= 0.04, p= 0.02, respectively).

Conclusion: We found that PA wall thickness was significantly higher in BD with major organ involvement compared to BD patients with only mucocutaneous

Disclosure of Interests: None declared


DIVERSITY OF HEMODYNAMIC TYPES IN CONNECTIVE TISSUE DISEASE ASSOCIATED PULMONARY HYPERTENSION: MORE THAN A SUBGROUP OF PULMONARY ARTERIAL HYPERTENSION


1Pediatric Rheumatology Department, Medical College Hosp., Department of Rheumatology, Beijing, China; 2Medical College Hosp., Medical Intensive Care Unit, Beijing, China; 3Peking Union Medical College Hosp., Department of Pulmonology, Beijing, China.

Background: Connective tissue disease (CTD) associated pulmonary hypertension (PH) is classified as a subgroup of WHO group 1 PH, also called pulmonary arterial hypertension (PAH). However, not all CTD-PH fit the hemodynamic definition of PAH. This study investigates the diversity of hemodynamical types of CTD-PH, their different clinical characteristics and outcomes.

Objectives: This study investigates the diversity of hemodynamical types of CTD-PH, their different clinical characteristics and outcomes.

Methods: We performed a retrospective cohort study. CTD-PH patients underwent right heart catheterization (RHC) were enrolled and divided into WHO group1 PH, WHO group 2 PH and high output PH (PVR<3WU and PAWP<15m mHg) according to hemodynamic features. Patients with pus lung diseases, left heart disease and pulmonary embolism were excluded. Baseline characteristics, inflammatory markers, autoantibodies, cardiac function status, echocardiogram parameters, hemodynamics and survival rates were compared.

Results: 207 CTD-PH patients were included, including 139 in WHO group 1 PH, 36 in WHO group 2 PH and 32 in high output PH. Incidence of anti-ribonucleoprotein antibody was lower in WHO Group 2 PH. High output PH is less severe, presenting lower NT-proBNP level, better WHO functional class, lower mPAP and PVR, higher cardiac output, and less cardiac remodeling. Among patients with elevated PAWP, combine pre-capillary PH had higher mPAP and larger right ventricle diameter. Association of more moderate interstitial lung disease didn’t show significant difference in disease characteristics. Short-term survival was significantly worse in WHO group 2 PH, yet 5-year survival rates didn’t differ between groups.

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Figure 1. Measurement of pulmonary artery wall thickness by Transthoracic Echocardiography.