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POS1367 THE PREVALENCE AND RISK FACTORS FOR CARDIAC DISEASE IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean fever (FMF) is a genetic disorder manifested 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 (16.1 % vs 12.8% p- 0.022, 9.3% vs 5.1 p- 0.01 and 4.5% vs 0.2% 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.
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POS1368 DIVERSITY OF HEMODYNAMIC TYPES IN CONNECTIVE TISSUE DISEASE ASSOCIATED PULMONARY HYPERTENSION: MORE THAN A SUBGROUP OF PULMONARY ARTERIAL HYPERTENSION

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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<15mmHg) according to hemodynamic features. Patients with obvious 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& post-capillary PH had higher mPAP and larger right ventricle diameter. Association of mild to 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.
Conclusion: Pre-capillary PH is not the only hemodynamic type of CTD-PH. Different types of CTD-PH present different clinical phenotypes and outcome. Carefully phenotyping PH in CTD-PH patients is important.
Disclosure of Interests: None declared
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POS1369 PULMONARY ARTERY WALL THICKNESS IS INCREASED IN BEHÇET'S DISEASE

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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 wideness, thin-walled, increased compliance, and low resistance [3]. We have recently shown increased venous wall thickness in lower extremity 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-portion of the main PA (approximately 1 to 2 cm distal to the pulmonary valve) as demonstrated in Figure 1.

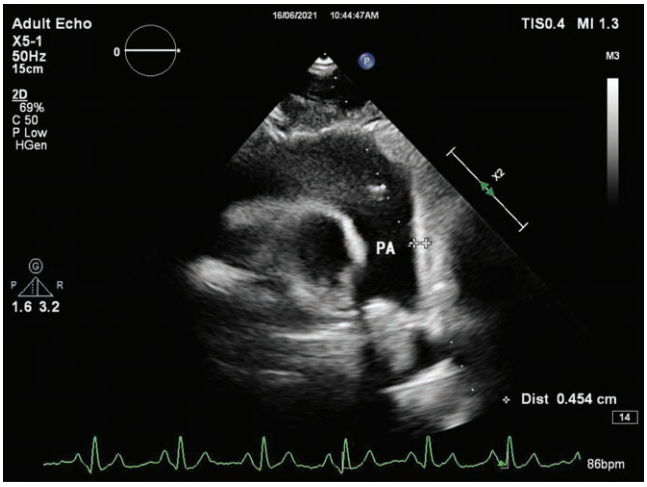

Figure 1. Measurement of pulmonary artery wall thickness by Transthoracic Echocardiography

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:0.06) (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 0.36 mm, 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

involvement regardless of major organ involvement type. These results suggest that increased PA wall thickness in BD may be the predictor of the major organ involvement during follow-up.

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POS1370 INCREASED INFERIOR VENA CAVA WALL THICKNESS AS A SIGN OF VENOUS INFLAMMATION IN BEHÇET'S DISEASE

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Background: Vascular involvement of Behçet's disease (BD) involves both arterial and venous vessels of all sizes [1]. Femoral (superficial, deep, and common) and popliteal veins are the most frequently affected veins. We have previously shown that femoral wall thickness is increased in BD patients and can be used as a diagnostic test [2].
However, many other sites including vena cava inferior/superior and pulmonary arteries may also be involved [3]. Despite the dominance of venous vessel involvement, there is limited data assessing the large veins in BD.
Objectives: In this study, we aimed to assess inferior vena cava wall thickness (IVC) by transthoracic echocardiography (TTE) in BD compared with healthy controls.
Methods: Patients with BD (n=70) and age and sex-matched healthy controls (n=51) were included in this study. Assessment of inferior vena cava (IVC) wall thickness was performed by an experienced cardiologist blinded to cases. Measurement of IVC wall thickness was made at end-expiration and approximately 0.5 to 2.0 cm proximal to the ostium of the right atrium as demonstrated in Figure 1.

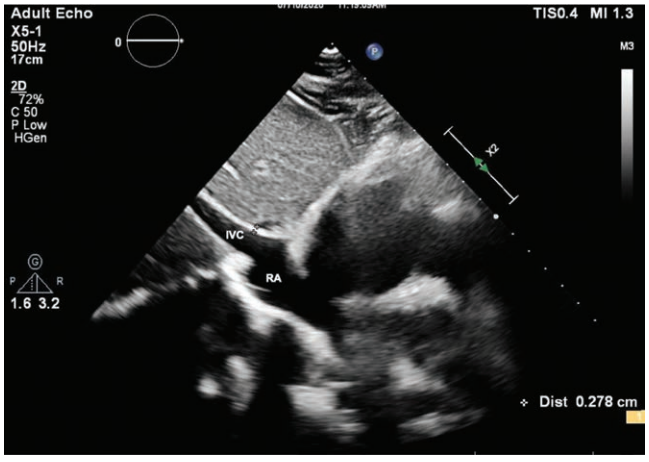

Figure 1. Measurement of inferior vena cava wall thickness by transthoracic echocardiography

Results: IVC wall thickness of patients with BD (0.29 mm (SD: 0.03) was significantly higher than healthy controls (0.26 mm (SD: 0.03) (p<001). Although IVC wall thickness was higher in patients with BD with vascular involvement (0.30 mm (SD:0.04) and history of pulmonary embolism (0.30 mm (SD:0.04)), the difference did not reach statistical significance. There was no difference between IVC wall thicknesses in patients who used immunosuppressive and anti-TNF treatments due to major organ involvement, compared to those who did not. Similarly, no difference is observed between IVC thicknesses among Behçet's patients according to age, gender, and activity status at the last visit. Although no correlation was found between IVC wall thicknesses, disease duration, and BDCAF scores at the last visit in the BD group, there was a low-grade correlation between age and IVC wall thickness (r=0.31, p=0.09)
Conclusion: Increased IVC wall thickness shows vasculitic involvement of large venous structures in BD and can be easily measured by TTE which is an easily accessible, noninvasive modality without radiation. The role of IVC wall thickness assessment for the diagnosis or management of BD requires further studies.

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POS1371 HYPOMETHYLATION OF CIRCULATING IMMUNE CELLS IN PATIENTS WITH GRAVES' ORBITOPATHY – A PRELIMINARY STUDY

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Background: Graves' orbitopathy (GO) is an eye disease occurring in patients with autoimmune thyroid disorders (AITD), most commonly Graves´ disease. It is characterized by inflammation affecting soft tissues of the orbit. A recent study demonstrated an association between fibroblast hypomethylation and disease activity in GO (Virakul *et al*., Front Endocrinol, 2021). Because procurement of fibroblast from GO patients require an invasive sampling, we wondered whether analysis of global DNA methylation in circulating immune cells obtained from peripheral blood could contribute to early detection of GO from patients with AITD.
Objectives: To compare global DNA methylation pattern in circulating immune cells obtained from AITD patients with GO and without GO history and healthy controls.
Methods: Global DNA methylation was quantified in circulating immune cell populations by flow cytometry using 5-methylcytosine antibody in patients with GO (n=10), AITD without GO history (n=9) and healthy controls (n=8). Immune populations ($CD4^+$ and $CD8^+$ T cells, B cells, monocytes and $CD56^{dim/bright}$ NK cells) and their activation status were identified using CD3/4/8/14/16/19/25/45/56/69 antibodies.
Results: In patients with GO, global DNA methylation was reduced by ~50% in the activated ($CD25^+$) $CD8^+$ T cells and by ~35% in the whole $CD8^+$ T cell population compared to patients with AITD (p=0.006). Moreover, percentage of $CD8^+$ T cells, but not activated subpopulation, was higher in GO when compared to AITD (p=0.04). Hypomethylation by ~20% was detected in monocytes as well as in $CD56^{dim}$ NK cells and their activated ($CD69^+$) subpopulation when GO was compared with AITD (p≤0.02). Of these cell populations, percentage of monocytes was also higher in GO when compared to AITD (p=0.04). Global methylation in B cells, $CD4^+$ T cells and $CD56^{bright}$ NK cells did not differ between patients with GO and patients with AITD (p>0.05). Of these populations, higher percentage of B cells was detected in GO when compared to AITD group (p=0.02). Analysis of larger patient cohorts is in progress with particular emphasis on the relationship of methylation patterns to GO disease activity.
Conclusion: This is the first study identifying the different global methylation profile of circulating immune cells in patients with GO characterized by DNA hypomethylation in $CD8^+$ T cells, $CD56^{dim}$ NK cells and monocytes compared to patients with AITD. Our study nominates hypomethylation as a non-invasive biomarker of GO and should be validated in a larger cohort of patients.
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