**AB0623**

**RATE AND PREDICTIVE FACTORS ASSOCIATED WITH SUSTAINED REMISSION IN IDIOPATHIC INFLAMMATORY MYOSITIS**

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**Background:** Idiopathic inflammatory myositis (IIM) is a group of heterogeneous connective tissue diseases, characterised by chronic muscle inflammation, myositis-specific or myositis-associated autoantibodies and different extra-muscular features. Achieving low disease activity or remission in patients with IIM has proven to be difficult due to the wide clinical spectrum of the different IIM types.

**Objectives:** To retrospectively assess any predictive factors for sustained remission in IIM patients.

**Methods:** We retrospectively analyzed data taken from medical charts, which included age at disease onset, gender, laboratory data as well as clinical features present at onset, organ involvement and treatment history. A total of 151 adult patients with IIM followed-up for >1 year were retrospectively enrolled. Remission was defined as no clinical and laboratory evidence of disease activity persisting for more than 6 months during follow-up, while undergoing myositis therapy or under no medication. The remission of cutaneous involvement was defined as no current activity of skin rash, absence of Gottron’s papules as well as heliotrope rash and erythema, whereas the remission of pulmonary involvement was considered as no requirement for intensification of immunosuppressive therapy during follow-up. Likewise, absence of muscle weakness or hypostenia was taken into account for evaluating muscle involvement. Moreover, the clinical features were accompanied by normalization of myogenic enzymes such as creatine kinase (CK) and lactate dehydrogenase levels.

**Results:** Among all 151 patients, 89 (58.9%) patients achieved sustained remission. By univariate analysis, overlap myositis (79% vs 27.4%; p=0.003; OR:0.22), cancer-associated myositis (CAM) (78% vs 19.35%; p=0.046; OR:0.3), as well as the presence of anti-Ku (3.3% vs 12.9%; p=0.05; OR:0.23) and anti-TIF-1 gamma (1.1% vs 8%; p=0.043; OR:0.13) antibodies and polyclonality (11.2% vs 24.19%; p=0.045; OR:0.397) at onset were significantly associated with active IIM, not achieving remission. Out of 89 patients in remission, 79 (88.8%) achieved long-term sustained remission, lasting at least 2 years, whereas 10 patients (11.2%) showed relapse. Most of relapsed patients showed a relapse/remitting disease, with no evident trigger for the relapse. We divided our cohort of 89 patients in remission in 2 subsets: 83 patients in remission undergoing therapy (92.3%) and 6 patients in drug-free remission (6.7%). No significant differences were found between two groups, except for younger age at onset (p=0.0002) found in patients achieving drug-free remission.

**Conclusion:** Sustained remission occurs in about one half of patients with IIM. The presence of anti-Ku and anti-TIF-1 gamma antibodies as well as polyclonality at onset lowers the chance of achieving sustained remission. Younger age at diagnosis has proved to predict drug-free long-lasting remission.

**Disclosure of Interests:** None declared

**AB0625**

**IS PULMONARY ARTERIAL HYPERTENSION, ASSOCIATED WITH SYSTEMIC SCLEROSIS REVERSIBLE?**

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**Background:** Systemic sclerosis (SSc) is one of the frequent causes of the pulmonary arterial hypertension (PAH) which found in 6-15% patient with SSc. Patients with PAH, associated with SSc have a poorer prognosis than other forms of PAH. 6 World Symposium of PH lowered diagnostic cut-off to 21 mm Hg in hope of improved survival. PAH reversibility is described in congenital heart defects, HIV and some tumors

**Objectives:** The aim of the study to detect reversibility of PAH associated with SSc.

**Methods:** Hemodynamics (mean pulmonary artery pressure (MPAP), pulmonary artery wedge pressure (PAWP), cardiac output (CO) and pulmonary vascularity resistance (PVR)), functional class (NYHA), 6-minute walk distance (6MWMD), biomarkers and DCO were assessed. Patients with pulmonary fibrosis and left heart diseases were excluded.

**Results:** The study includes 56 patients receiving start-up monotherapy with 1st generation PAH-specific drugs: bosentan (25 patients) and sildenafil (31 patients). The median age of the patients was 51.5 (37, 58) years. At the time of diagnosis, the MPAP in the sildenafil group was 49 (30; 50) mm Hg, bosentan - 50 (42; 56) mm Hg, differences not significant (p = 0.11). During observation against the background of sildenafil intake,