(n=5, 13.5%), major airway cartilage loss (n=5, 13.5%), auricular collapse (n=2, 5.4%), nasal collapse (n=3, 8.1%), subglottic stenosis (n=2, 5.4%), hearing loss (n=5, 13.5%), vision loss (n=1, 2.7%) and deforming arthritis (n=1, 2.7%). In the univariate analysis, only major airway involvement had a relationship with complication development. (OR 12, 95% CI 1.97–72.8, p=0.007)

Conclusions: RP is a rare disease which diagnosis is commonly delayed and lead to cartilage loss. RP developed after trauma in 4 patients, possibly trauma can be blamed as an etiologic factor. Awareness and early diagnosis results with better prognosis

Disclosure of Interest: None declared

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FRI0374 THE DIAGNOSTIC VALUE OF ALPHA-1-ANTITRYPSIN PHENOTYPE IN PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS (WEGENER'S GRANULOMATOSIS)

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Background: Deficiency of alpha-1 protease inhibitor, or alpha-1-antitrypsin (A1AT), predisposes to chronic lung diseases and some extra-pulmonary pathology. Besides classical implications, such as pulmonary emphysema and other associated lung diseases, A1AD is also known to be associated with granulomatosis with polyangiitis (GPA, or Wegener's).

Objectives: The aim of our study was to evaluate the frequency of mutated A1AT forms and its clinical significance among GPA patients.

Methods: We analysed 38 samples of sera, collected from GPA patients. Detailed clinical data, including Birmingham Vasculitis Activity Score (BVAS), incidence of lung involvement, anti-(Pr3) antibodies concentrations and other laboratory data were collected. We also studied control group of 46 healthy donors. All collected samples underwent A1AT phenotyping by isoelectrofocusing (IEF) and turbidimetric A1AT measurement.

Results: Mutated A1AT variants were revealed in 18.4% (7/38) cases and included following phenotypes: 1ZZ, 4MZ and 2MF. The mean A1AT concentration in samples with pathological genetic A1AT forms was significantly lower (P=0,0038), than in normal A1AT phenotype. According to reference values, only 2 of 7 pathological samples demonstrated decreased A1AT concentration. Comparing BVAS activity, we found that patients with mutated A1AT phenotype had significantly higher vasculitis activity, as well as anti-PR3 antibodies concentration. When analysis of nonspecific systemic inflammation markers was performed, the significant difference ($P<0.05$) was found only between means of erythrocyte sedimentation rate (ESR)

Conclusions: Laboratory testing to find pathological A1AT forms are not usually ordered for patients with systemic vasculitides. During medical examination of GPA patients the possibility of A1AT deficiency should be considered and may be verificated by IEF.

Disclosure of Interest: None declared

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FRI0375 CLINICAL IMPROVEMENT ACCORDING TO PATIENT REPORTED OUTCOMES IN PATIENTS WITH POLYMYALGIA RHEUMATICA: A LONGITUDINAL ANALYSIS FROM ROUTINE CARE

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Background: Several outcomes and clinical response measures have been used in patients with polymyalgia rheumatica (PMR), but there is no consensus about the optimal endpoint for evaluating response to treatment. A PMR activity score has been proposed which includes three patient reported outcomes (PROs)[1]. RAPID3 is an index found on a multidimensional health assessment questionnaire (MDHAQ) only including PROs, which is effective to document improvement not only in RA but also in other rheumatic diseases[2]

Objectives: To prospectively evaluate the performance of RAPID3 and other PROs included on the MDHAQ to document improvement in clinical status over time in patients with PMR.

Methods: This study was conducted at an academic rheumatology center at which all patients complete a MDHAQ, which includes 0–10 scores for physical function (FN), pain (PN), and patient global estimate (PATGL), compiled into a 0–30 RAPID3. The MDHAQ also scores fatigue, morning stiffness, self-reported joint counts and demographic data. Data collection included MDHAQ, acute-phase reactants, and prednisone dose. PMR patients with complete data seen between 2010 and 2014 were included. A baseline visit and the most recent visit were compared using paired t-test and McNemar's test. Spearman correlation analysis for non-normally distributed variables was performed between RAPID3 and ESR and CRP.

Results: 34 PMR patients seen in routine care were included: 59% were females, 71% Caucasian, and mean age was 71.6 years. The mean duration from a baseline visit to most recent visit was 15.5 months. At initial presentation, RAPID3 was 12.2, FN 2.2, pain 5.3, and PATGL 4.7, fatigue 3.9, and morning stiffness 63.1 minutes; 64.7% of the patients had painful hips, 79.4% had painful shoulders, 73.5% had abnormal ESR, and 70.6% had abnormal CRP. Significant improvement was seen between baseline and last visit in mean level of RAPID3 and all other MDHAQ measures, except fatigue ($p<0.05$), as well as ESR and CRP (Table). The mean dose of prednisone was decreased from 12.2 mg at first visit to 4.3 mg at most recent visit. The RAPID3 was significantly correlated with ESR (rho=0.52), and with CRP (rho=0.50).

Table 1

	PMR patients (n=34)			Mean change	% Improvement
	Baseline	Most recent visit	P value		
MDHAQ: patient self-reported measures					
RAPID3, mean (SD)	12.2 (7.0)	8.5 (7.2)	0.02	3.7	30.7%
MDHAQ – Function, mean (SD)	2.2 (2.1)	1.5 (1.7)	0.03	0.6	27.2%
MDHAQ – Pain, mean (SD)	5.3 (2.9)	3.4 (3.4)	0.002	1.9	35.8%
MDHAQ – PATGL, mean (SD)	4.7 (2.9)	3.1 (3.1)	0.01	1.6	34.0%
RADAI – painful hip, n (%)	22 (64.7%)	12 (35.3%)	0.02	29.4	45.4%
RADAI – painful shoulder, n (%)	27 (79.4%)	17 (50%)	0.005	10	37.0%
MDHAQ – Fatigue, mean (SD)	3.9 (3.6)	3.5 (3.3)	0.54	0.4	10.5%
Morning stiffness duration, minutes, mean (SD)	63.1 (97.7)	19.1 (34.1)	0.05	43.9	69.5%
Laboratory measures					
Abnormal ESR, n (%)	25 (73.5%)	14 (41.1%)	0.002	32	43.5%
Abnormal CRP, n (%)	24 (70.6%)	13 (38.2%)	0.01	32	45.3%
Medication					
Prednisone dosage, mg, mean (SD)	12.2 (6.8)	4.3 (3.5)	<0.001	7.9	64.7%

Conclusions: In patients with PMR, improvement was seen according to PROs included on a self-reported MDHAQ questionnaire in a similar range to ESR and CRP, documenting effective response to prednisone.

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FRI0376 COMPARISON OF CLINICAL CHARACTERISTICS OF PATIENTS WITH TAKAYASU'S ARTERITIS BY AGE AND GENDER

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Background: Takayasu's arteritis (TA) is a rare and heterogeneous disease that can be difficult to diagnose.

Objectives: In this study we distinguish clinical characteristics in the different groups with TA and aimed to improve the diagnosis ability of this disease in different people.

Methods: We retrospectively analyzed the clinical manifestations and laboratory and angiographic findings of 53 TA patients divided into groups by age and by gender.

Results: Ratio of incidence in males and females was 1:4; mean age at onset was (35 ± 11) years in males and (28 ± 11) years in females. In 17% of patients, at onset was over the age of 40. The most common symptom at onset was chest pain and reduced glomerular filtration rate (GFR) in males [7 (63.6%) *vs.* 12 (28.6%), *P*=0.031; 8 (72.7%) *vs.* 14 (33.3%), *P*=0.034], especially in young males [age≤40 years, 5 (62.5%) *vs.* 8 (22.2%), *P*=0.024; 7 (87.5%) *vs.* 12 (33.3%), *P*=0.006]. Significantly more male patients, especially males over the age of 40, had multi-vessel involvement [10 (90.9%) *vs.* 24 (57.1%), *P*=0.038; 2 (66.7%) *vs.* 0, *P*=0.033]. Significantly more young males had aortic insufficiency [5 (62.5%) *vs.* 8 (22.2%), *P*=0.024]. Pulselessness [2 (66.7%) *vs.* 0, *P*=0.03] and radial artery weakening [2 (66.7%) *vs.* 0, *P*=0.03] were found in more males over 40 years of age at onset. There were no differences in laboratory data between males and females. Chest pain [13 (29.5%) *vs.* 6 (66.7%), *P*=0.031] and fever [0 *vs.* 1 (11.1%), *P*=0.027]occurred significantly more often in patients over the age of 40., and patients over 40 years of age at onset had significantly less renal artery [1 (11.1%) *vs.* 21 (47.7%), *P*=0.042], abdominal aortic [0 *vs.* 16 (38.1%), *P*=0.030], and multi-vessel involvement [2 (22.2%) *vs.* 32 (72.7%), *P*=0.004]. Patients over 40 years of age were rarer in type V [1 (11.1%) *vs.* 22 (50%), *P*=0.032)] while more common in type IIa [3 (33/3%) *vs.* 4 (9.1%), *P*=0.050] than young ones. Multivariate analysis showed that thoracic aortic involvement was an independent risk factor for developing hypertension (OR =3.918, 95% confidence interval [CI] 1.616–1566.185, *P* =0.026), and ascending aortic involvement was an independent risk factor for both aortic insufficiency (OR =3.674, 95% CI 2.734–567.621, *P* =0.007) and aneurysm formation (OR =7.255, 95% CI 7.255 ~ 1,628,614; *P* =0.044).

Conclusions: Patients with disease at onset after age 40 are more likely to present with chest pain and fever and only rarely have renal artery, abdominal aortic or multi-vessel involvement. Males are likely to present with multi-vessel involvement and reduced GFR, and these differences are more prominent in young males, who are more likely to develop aortic insufficiency. Aortic stenosis was an independent risk factor for hypertension, while ascending aortic involvement was an independent risk factor for aortic valvular insufficiency and aneurysm formation. Aortic valve lesion and ascending aorta involvement may play reciprocal role to aggravates destructions of each other and induce chest pain.

Disclosure of Interest: None declared