tertiary rheumatology centre. The frequency, temporal occurrence and the localization of arthritis was recorded. In addition, we searched for potential differences in other IgAV features between patients with and without arthritis.

Results: During the 132-month observation period we identified 328 new IgAV cases (59.5% males, median (IQR) age 64.3 (45.1; 76.1) years). Forty-eight (14.6%) patients developed arthritis. Arthritis was the first IgAV manifestation in 16 (4.9%) patients. Arthritis was mono-, oligo- and poly- articular (involving up to 15 joints) in 13 (4.0%), 25 (7.6%) and 10 (3.0%) patients, respectively. Arthritis was most common in wrists and ankles (each in 18 (37.5%) patients); metacarpophalangeal joints and knees (each in 11 (22.9%)); proximal interphalangeal joints (9 (18.8%)); elbows (8 (16.7%)) and metatarsophalangeal joints (4 (8.3%)). Clinical differences between IgAV patients with and without arthritis are presented in table 1. Patients with arthritis were significantly younger, more commonly developed gastrointestinal tract involvement compared to those without arthritis. Arthritis remitted in all with immunomodulatory treatment (given predominantly for necrotic skin purpura or visceral involvement). Follow up (FU) data accessible for 42/48 (87.5%) patients with arthritis showed that IgAV relapsed in 10 (23.8%) patients during a median (IQR) 24.5 (12.9; 40.7) month FU. Relapses were limited to skin and/or kidneys, there were no relapses of arthritis.

Table 1. Clinical characteristics of IgA vasculitis patients with and without arthritis

Clinical characteristics	Arthritis IgAV (48)	Non-arthritis IgAV (280)	P value
Male gender (%)	56.3	60.0	0.636
Age (years)*	49.9 (36.1-66.9)	65.4 (48.4-77.5)	<0.001
Ever smoker (%)	50.0	43.6	0.435
Prior infection (%)	43.8	30.4	0.094
Generalized purpura	50.0	50.0	1.0
Skin necroses (%)	35.4	49.6	0.085
GI involvement (%)	47.9	25.0	0.002
Renal involvement (%)	47.9	46.8	1.0
Elevated serum IgA (%)	33.3 (11/33)	52.0 (115/221)	0.061

Legend: * median and IQR; GI gastrointestinal;

Conclusion: Arthritis in adult IgAV was frequently oligoarticular, involved large and small joints of both upper and lower extremities, and was not prone to chronic course and recurrence. In addition, we found an association between arthritis and gastrointestinal tract involvement.

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POS0824

THE LONG-TERM CLINICAL COURSE OF MUSCULAR VASCULITIS DEPENDING ON THE ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY STATUS: A RETROSPECTIVE OBSERVATIONAL STUDY

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Background: Skeletal muscle is known as one of the organ involvements of primary systemic vasculitis.^{1,2} Muscle inflammation is detected by magnetic resonance imaging, and necrotizing vasculitis is proved by muscle biopsy.³ As with systemic vasculitis or single organ vasculitis, glucocorticoid (GC) and immunosuppressants are used in its treatment.⁴ There are not many reports about muscular vasculitis, and its long-term clinical course after initial treatment, including the rates of relapse and mortality, remains unclear.

Objectives: To identify the predictors of relapse and mortality in patients with muscular vasculitis, especially focusing on the status of anti-neutrophil cytoplasmic antibody (ANCA).

Methods: We retrospectively reviewed patients diagnosed with necrotizing vasculitis with muscle involvements in our hospital between 2004 and 2020. In all cases, muscular vasculitis was identified by muscle biopsy or magnetic resonance imaging. To focus on the clinical features of muscular vasculitis, we excluded patients with severe organ involvements as cardiovascular, abdominal, cerebral, severe renal, and severe pulmonary involvements. We compared the 5-year cumulative incidence of relapse, the overall survival rate, and the dose of GC over 5 years between the ANCA-positive and ANCA-negative groups. A relapse was defined as any new or worsened state of disease activity requiring an escalation of GC dose. Gray's method was used for assessing the cumulative incidence of relapse. The log-rank test was used for assessing overall survival. The Mann-Whitney U test was used for assessing the dose of GC. The possible factors for relapse in 5 years in a univariate analysis were selected for a multivariate analysis using the Cox proportional hazards model.

Results: Forty-nine patients were enrolled. The median age of onset was 77 (69-82) years and 71.4% were women. There were 30 ANCA-positive patients (90.0%

with anti-myeloperoxidase) and 19 ANCA-negative patients. The median age and the number of patients with renal involvements were higher in the ANCA-positive group than in the ANCA-negative group (73.0 ± 9.29 years vs. 79.5 ± 20.28 years, $p=0.0062$ and 7/30 [23.3%] vs. 0/19 [0.0%], $p=0.034$, respectively). The Birmingham Vasculitis Activity Score (ver. 3), the induction dose of GC, and the rate of immunosuppressants use were not significantly different between the two groups. During the observational period, 24 patients relapsed. The 5-year cumulative incidence of relapse was significantly higher in the ANCA-positive group than in the ANCA-negative group ($p=0.026$) (Figure 1). The Cox proportional hazards model revealed that the presence of ANCA was an independent risk factor for relapse (hazard ratio: 3.15; 95% confidence interval 1.06–9.38; $p=0.040$). During the observational period, 9 patients died (3 died from cancer, 1 from interstitial pneumonia, 1 from cerebral hemorrhage, 1 from infection, and 3 from unknown reasons). The ANCA-positive group exhibited a higher mortality rate than the ANCA-negative group without a statistical significance ($p=0.12$). The 5-year cumulative dose of GC was larger in the ANCA-positive group than in the ANCA-negative group without a statistical significance (14786 [11246–19138] mg vs. 10088 [7129–12634] mg, $p=0.12$).

Conclusion: In muscular vasculitis, the presence of ANCA is an independent risk factor for long-term relapse. Stratified treatment depending on the ANCA status may reduce the relapse rate and the occurrence of side effects of GC in patients with muscular vasculitis.

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POS0825

DEVELOPMENT OF A NEW AND SIMPLIFIED FORMULA TO PREDICT MORTALITY IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS ADMITTED TO THE INTENSIVE CARE UNIT

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Background: ANCA-associated vasculitis (AAV) can present with a wide range of symptoms, including acute kidney injury (AKI) requiring renal replacement therapy or diffuse alveolar hemorrhage (1). These two manifestations often require admission to an intensive care unit (ICU) and are associated with increased mortality. To predict ICU mortality, the Simplified Acute Physiology Score version 2 (SAPS2) is often used but has not been formally tested in AAV patients (2). In addition, it is cumbersome to assess.

Objectives: To develop a novel, simplified formula to predict ICU mortality in an AAV ICU population from an academic tertiary care center.

Methods: We retrospectively recorded clinical and laboratory parameters in patients admitted to our ICU from 2000-2018. We performed risk factor analysis using univariate and multivariate logistic regression. In the multivariate case we applied the least absolute shrinkage and selection operator (lasso) method for variable selection. We considered average marginal effects and partial dependence plots in order to describe the influence of various independent variables on the probability of death more specifically. We evaluated our new score by comparing the corresponding area under the curve (AUC) to the AUC corresponding to the established SAPS2 score.

Results: We analyzed 58 patients with AAV (39 granulomatosis with polyangiitis, 19 with microscopic polyangiitis) with a mean age of 74 ± 14 (GPA) and 73 ± 12 (MPA). 19/39 (48.7%) of GPA and 9/19 (47.4%) were female. Reasons for admission included disease manifestations or infectious complications from treatment (e. g. pneumonia, urinary tract infection). In total, 13/58 (22.4%) patients died throughout the study (10 GPA, 3 MPA patients). Using a cut-off threshold of 40 for SAPS2, sensitivity and specificity for mortality were 0.92 and 0.60, respectively. Confidence interval for the AUC was [0.68,0.95]. In the fitted multivariate logistic regression model, lasso was applied for variable selection. The identified variables included: disease duration, pH, procalcitonin, hemoglobin, leukocytes on admission, coronary heart disease, and pneumonia on admission. The estimated mortality is given by the formula $f(\beta_0 + \beta_1\chi_1 + \dots + \beta_7\chi_7)$, where $f(u) = 1/(1 + \exp(-u))$. Table 1 shows the estimated mortality for various values of the new score.

Table 1. Example scores predicting mortality using the novel formula.

Score	Predicted mortality
-2.2	0.1
-1.1	0.25
0	0.5
1.1	0.75
2.2	0.9

Testing if the AUC corresponding to the new model is significantly larger than the one corresponding to the SAPS2 score as independent variable resulted in p-value of 0.037. To identify possible overfitting, a 5-fold cross validation was performed. This resulted in a CI for the AUC of [0.64,0.96], suggesting that the new score allows for simpler prediction of mortality.

Conclusion: We developed a novel formula corresponding to a score which is able to simpler predict mortality in patients with AAV admitted to the ICU. We will test our formula in the available ICU database MIMIC III, which comprises a large dataset of ICU patients.

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POS0826 SKIN LIMITED IGA VASCULITIS IN ADULTS

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Background: IgA vasculitis (IgAV) could be limited to skin or evolve into a systemic disease, affecting characteristically joints, gastrointestinal tract and/or kidneys.

Objectives: We aimed to look for differences between adult IgAV patients with disease limited to skin compared to systemic IgAV.

Methods: Medical records of histologically proven adult IgAV cases, diagnosed between January 2010 and December 2020 at our secondary/tertiary rheumatology centre were analyzed.

Results: During the 132-month observation period we identified 328 new IgAV cases (59.5% males, median (IQR) age 64.3 (45.1; 76.1) years). Ninety-four (40.2%) patients had skin limited disease, and the rest systemic IgAV.

Clinical differences between skin limited and systemic adult IgAV are presented in table 1. Adults with IgAV limited to skin were significantly older, had less commonly skin lesions above the waistline and a lower level of C reactive protein compared to patients with a systemic disease. There were no differences in the frequency of skin necroses between the compared IgAV subgroups. The frequency of potential vasculitis triggers (prior infections, new medications, malignancy) was similar between the compared subgroups.

Table 1. Clinical characteristics of IgA vasculitis patients with skin limited and systemic disease

Clinical characteristics	Skin limited IgAV (94)	Systemic IgAV (234)	P value
Male gender (%)	54.3	61.5	0.263
Age (years)*	68.0 (55.0-80.5)	61.5 (41.7-75.8)	0.007
Current smoker (%)	13.8	21.8	0.123
Antecedent infection (%)	28.7	33.8	0.434
New medication	23.4	23.5	1.0
History of cancer	12.8	10.7	0.569
Symptom duration (days)*	7 (5-21)	8 (5-14)	0.756
Purpura above waistline	36.2	55.6	0.002
Skin necroses (%)	52.1	45.7	0.329
ESR (mm/h) *	32 (18-52)	34 (17-53)	0.873
CRP (g/l) *	13.5 (1-32)	30 (11-68)	<0.001
Elevated serum IgA (%)	50.6	49.1	0.892

Legend: * median and IQR;

Follow up data were available for 250 (76.2%) patients. During the follow up of median (IQR) 12.5 (6.8 – 22.4) months 35 patients relapsed (13/70 (18.6%) with skin limited IgAV and 22/180 (12.2%) with systemic IgAV, p= 0.224).

Conclusion: Skin limited IgAV was associated with older age and less extensive skin purpura in adults. However, relapses of purpura were as common as in systemic IgAV.

Disclosure of Interests: None declared

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POS0827 HEPATOTROPIC VIRUSES WITH HIGHER RHEUMATOID FACTOR, BUT NOT RHEUMATIC DISEASES LINK TO PREVALENT CRYOGLOBULINEMIA. CORRELATION OF CLINICAL AND SEROLOGICAL MARKERS WITH ETIOLOGICAL CAUSES

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Background: Cryoglobulinemia (CG) is a rare phenomenon, which is defined as the persistent presence in serum of abnormal immunoglobulins (Igs) that precipitate in vitro at less than 37°C and dissolve when the temperature rises again. Is related to hematological disorders, infections and autoimmune diseases.

Objectives: To describe the differential clinical features, serological and demographics in a cohort of patients diagnosed with CG.

Methods: We describe a retrospective cohort of 252 cryoglobulin (Cg) positive samples, obtained from a database from the immunology laboratory of a tertiary hospital (November 2018-November 2019). We obtained 182 patients with CG, classified according to their etiology into 4 groups: 1) Rheumatic diseases (RD) that includes rheumatoid arthritis, Systemic lupus erythematosus, Sjögren's syndrome and Systemic sclerosis, 2) Hepatotropic viruses (HV) with patients diagnosed with Hepatitis C virus, B virus and both, 3) Hematological diseases (HD) and 4) Essential cryoglobulinemia (CGE). Demographic variables, clinical and serological data were collected. A comparative analysis was performed with the Mann-Whitney U test and the multivariate Kruskal-Wallis test, nonparametric variables were compared using a Wilcoxon test. Ten patients, with more than one disease from 4 groups, were excluded from the study.

Results: Out of 182 reviewed patients, 172 were included in the study. Mean age at diagnosis was 59.7(±14.0). Demographic, clinical and laboratory characteristics are described in table 1. Mixed CG was the predominant subtype, in 116 (67.4%) patients. The most prevalent CG-associated diseases were HV infection with 91(53%) patients. CGE mostly presented with cutaneous manifestations (p=0.0001), particularly purpura. In RD group the presence of Raynaud and non-erosive arthritis (p=0.0001) was relevant. Laboratory findings showed that CG titration varies according to the etiology, being HD the one with the highest values with 292.2 (±546.2). There is significant difference in