uniform. Recently, based on previous clustering analysis and clinical, histopathological, serological and prognostic aspects three subcategories of AAV have been proposed and named as: non-severe AAV, severe PR3-AAV and severe MPO-AAV [1].

Objectives: In line with these attempts to subcategorize AAV we decided to use latent class analysis (LCA) on a large multicenter cohort of Polish AAV patients from POLVAS [2] registry to identify potential new subphenotypes or confirm already proposed ones.

Methods: Latent Class Analysis (LCA) approach was used as a model based clustering method of objects described by dichotomous (e.g., gender; ANCA status – cANCA, pANCA; organ involvement – skin, eye, ENT, respiratory, heart, GI, renal, urinary, CNS, peripheral nerves) and polytomous (number of relapses) variables supported by quantitative covariates (e.g., age at diagnosis, CRP at diagnosis, maximal serum creatinine concentration ever).

Results: Results of LCA on our AAV group returned four class model of AAV subphenotypes, confirming existence of the previously proposed by Mahr et al. [1] and revealed fourth – previously not described clinically relevant subphenotype. To this fourth class – belong patients only with GPA, diagnosed at young age, with multiorgan involvement, high relapse rate and relatively high risk of death.

Table 1. AAV subcategorization – summary of clinical characteristics and ANCA specificity

	LCA Class 1	LCA Class 2	LCA Class 3	LCA Class 4
No of patients	130	194	102	97
AAV type	Mainly GPA	Mainly GPA	mainly MPA	Only GPA
Age at diagnosis	Middle age	Middle age	Old	Young
Male/female ratio	1:2	2:1	1:1	1:1
Main organ involvement	ENT, respiratory, eye	Renal, respiratory, ENT	Renal, respiratory, skin	Multiorgan involvement
Relapse rate	intermediate	intermediate	low	high
Modified class description (based on ref. [1])	Non severe AAV	Severe PR3 AAV	Severe MPO AAV	Severe non-renal PR3 AAV

Conclusion: Based on multiple clinical and serological variables LCA methodology identified 4-class subphenotypes model of AAV. Fourth-class is a new clinically important subphenotype including exclusively PR3-positive young AAV patients with multiorgan involvement, high risk of relapse and distinct mortality.

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IDENTIFICATION OF RISK AND PROGNOSTIC FACTORS FOR POLYARTERITIS NODOSA PATIENTS WITH DIGITAL GANGRENE

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Background: Polyarteritis nodosa (PAN) is a segmental, necrotizing vascular disease that primarily impacts medium-sized muscle arteries. The estimated annual incidence of PAN is still lacking in China. Digital gangrene is an ischemic manifestation of the limb. However, the causes and the treatment methods vary from case to case, and the outcome is unpredictable. These features emphasize

the need to identify measurable variables that accelerate digital gangrene development in PAN patients. However, little effort has been made to identify the clinical and laboratory factors that affect PAN patients with digital gangrene to anticipate their natural history and response to therapy.

Objectives: Many patients with polyarteritis nodosa (PAN) complicated with digital gangrene have poor outcomes and related research information is limited. This study was carried out to identify the associated risk and prognostic factors.

Methods: We conducted a retrospective study of 148 PAN patients admitted to Peking Union Medical College Hospital (PUMCH) from September 1986 to December 2018. The characteristics, therapeutic regimens, and outcome data for patients with and without gangrene were compared. The Kaplan–Meier method and Cox hazard regression model were used to evaluate the prognostic factors.

Results: Forty-seven (31.8%) PAN patients had digital gangrene complications. The average age was 40.4±17.9 years and the average disease duration was 11 (4–27) months. The presence of digital gangrene was correlated with smoking history [odds ratio (OR), 4.27; 95% confidence interval (95% CI), 1.56–11.66] and eosinophil elevation (28.12; 10.30–76.8). Thirty-two (68.1%) gangrene patients received methylprednisolone pulse therapy and all of these patients were treated with cyclophosphamide. Nine patients suffered irreversible organ injury and two died. Disease duration ≥ 24 months and elevated serum C-reactive protein (CRP) were identified as hazardous factors for poor prognosis in patients with gangrene ($P=0.003$, HR=8.668, 95% CI 2.11, 35.55 and $P=0.042$, HR=27.062, 95% CI 1.13, 648.57, respectively).

Conclusion: Smoking history and eosinophil elevation in PAN patients were more prone to digital gangrene and high serum CRP level predicted poor outcomes. PAN patients with smoking history and elevated eosinophils need to be seriously evaluated by clinicians. Furthermore, the CRP level should be efficiently controlled for good prognosis.

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THE CLINICAL FEATURES AND OUTCOME OF VENA CAVA INVOLVEMENT IN BEHCET'S DISEASE

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Background: Behcet's disease (BD) is a systemic disease that can affect vessels of any size and type. However, only limited cases of BD patients with vena cava involvement have been reported.

Objectives: To investigate the clinical features and outcome of vena cava involvement in BD patients.

Methods: We retrospectively reviewed the clinical data of BD patients with vena cava involvement in our institute from August 2001 to October 2019. The treatment and outcome of these patients were also analyzed.

Results: Fifty BD patients with vena cava involvement were included. The median interval between BD onset and diagnosis of vena cava involvement was 2.8 (range 0–19.4) years. Superior vena cava (SVC) involvement was detected in 22 (44.0%) patients, and 21 patients had typical manifestations of SVC syndrome. Inferior vena cava (IVC) was detected in 35 (70.0%) patients, including 7 patients diagnosed with Budd-Chiari syndrome. Seven patients had both superior and inferior vena cava involvement. Forty-five (90.0%) patients had venous involvement other than vena cava, including 19 patients with common iliac thrombosis, 12 patients with common femoral vein thrombosis, 11 patients with external iliac vein thrombosis, etc. For the other BD manifestations, oral ulceration was presented in all patients, followed by genital ulceration (35, 70.0%), Erythema nodosum (27, 54.0%) and pathergy reaction (25, 50.0%). Thirteen (26%) patients had eye involvement. Ten (20%) patients had pleural and/or pericardial effusions. Eleven (22.0%) patients had pulmonary thromboembolism, and 4 (8.0%) patients had arterial involvement.