Introduction.– Ear, nose and throat (ENT) symptoms of EGPA often precede the diagnosis by several years, and occur in 70–85% of patients. We aimed to design a composite activity/damage-scoring tool for the objective assessment of ENT features in EGPA, to facilitate disease assessment in clinical studies.

Methods.– A prospective review of patients with known or suspected EGPA over 18 months was carried out. For each patient, a full ENT history and examination, including otoscopy, anterior rhinoscopy and flexible nasendoscopy was obtained, and pure tone audiometry and tympanometry performed where indicated.

An objective assessment tool (the AD Tool) for recording the presence of disease activity, damage or infection was developed, using BVAS and VDI, the literature, and the experience of ENT surgeons, and physicians with a specialist interest in vasculitis. ENT manifestations were recorded using the AD Tool and an “AD score” was calculated. AD scores were then correlated with disease state and systemic disease activity after a multi-disciplinary review of each patient. Performance of the AD score was compared to BVAS.

Results.– Thirty patients were reviewed at a total of 57 visits. The AD score was associated with disease state but did not correlate well with systemic disease activity. As disease control deteriorated, activity scores increased. The detailed item list in the AD tool (30 activity items and 20 damage items) showed increased sensitivity as compared to BVAS (5 ENT items) in monitoring ENT activity and predicting systemic flare (Supplementary data).

Conclusion.– A detailed scoring system which is sensitive for monitoring ENT disease activity in EGPA has been developed. This allows for early recognition of flaring disease, rapid treatment, and prevention of systemic disease progression. Of all ENT items assessed, seven activity items have shown close correlation with deteriorating disease control. Work is needed to validate the tool and define its role in the management of EGPA.

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P42
A scoring tool for ear, nose and throat disease activity and damage in eosinophilic granulomatosis with polyangiitis (Churg-Strauss)
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Introduction.– Ear, nose and throat disease (ENT) symptoms of eosinophilic granulomatosis with polyangiitis (Churg-Strauss) often precede the diagnosis by several years, and occur in 70–85% of patients. We aimed to design a composite activity/damage-scoring tool for the objective assessment of ENT features in EGPA, to facilitate disease assessment in clinical studies.

Methods.– A prospective review of patients with known or suspected EGPA over 18 months was carried out. For each patient, a full ENT history and examination, including otoscopy, anterior rhinoscopy and flexible nasendoscopy was obtained, and pure tone audiometry and tympanometry performed where indicated.

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P43
Serum biomarkers during relapsing disease in eosinophilic granulomatosis with polyangiitis
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Introduction.– Previous studies suggest a role for serum levels of eosinophil-3, TARC/CCL17 and IgG4 in newly diagnosed active patients with eosinophilic granulomatosis with polyangiitis (Churg-Strauss, EGPA). This study investigated the role of these biomarkers in patients with EGPA as measures of disease recurrence.

Methods.– Serum levels of TARC/CCL17, eosinophil-3 and IgG4 were determined in EGPA patients during follow-up. All study subjects (n = 25) were randomly selected from a longitudinal cohort of patients with EGPA. Epidemiological, clinical and laboratory data including disease activity (BVAS and BVAS/WG) were available for all visits.

Results.– TARC/CCL17, eosinophil-3, and IgG4, as well as IgG4/IgG ratio were determined in serum samples from 105 visits of 25 patients with EGPA (median 4 visits, range 2–8). At the first visit, 20 patients (80%) were using glucocorticoids and 17 (68%) additional immunosuppressive drugs. Disease flares were seen at 18 visits and remission was observed in the remaining 87 visits. The median BVAS score in patients with relapsing...
P44 Eosinophilic granulomatosis with polyangitis (EGPA): Clinical and immunologic expression in a single center cohort
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Introduction. – The purpose of this study was to determine whether epidemiologic, clinical, and analytical features might modulate disease expression in a single center cohort of patients with eosinophilic granulomatosis with polyangitis (EGPA).

Methods. – Patients with a diagnosis of EGPA made according to the 1990 ACR classification criteria for the disease were enrolled in this retrospective study. Data collected included:
- gender;
- age at diagnosis;
- age at inclusion;
- cumulative clinical features retrospectively assessed according to the Birmingham Vasculitis Activity Score (BVAS) glossary.
The following parameters were also recorded: eosinophil count, ANCA status, rheumatoid factor (RF) positivity. Statistical analysis was performed using SPSS 13 (SPSS Inc., Chicago IL, USA). A 2-tailed value of P < 0.05 was taken to indicate statistical significance.

Results. – Forty-seven EGPA patients (23F; mean age 47 ± 15 yrs; mean follow-up 7 ± 5 yrs) were enrolled. The prevalence of the clinical and laboratory features observed in our EGPA cohort was consistent with the larger cohorts of the literature. More specifically, ANCA-MPO were detected in 21/47 (44%) patients. Statistical analysis showed that EGPA female patients had a lower frequency of nasal polyps (P = 0.02) and a higher frequency of arthralgia (P = 0.05), urticaria (P = 0.008) and RF positivity (P = 0.03). Elderly-onset patients had a higher prevalence of mononeuritis multiplex or polyneuropathy (P = 0.001). Finally, the subset of patients with ANCA-MPO positivity had less frequent cardiac manifestations (P = 0.04) and a higher prevalence of relapses (P = 0.006).

Conclusion. – Epidemiologic and immunologic features have a significant impact on the clinical presentation of EGPA, influencing the prevalence and diversity of systemic involvement and should be taken into account in the assessment of EGPA patients in the clinical setting.

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P45 Sinonasal involvement in patients with eosinophilic granulomatosis with polyangitis (EGPA, ex Churg Strauss Syndrome): A modern look to an ancient problem
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Introduction. – “Paranasal sinus abnormalities” are commonly reported in granulomatosis with polyangitis (EGPA) representing one of the six 1990 ARA classification criteria for the disease. Although frequently encountered, the definition of “paranasal sinus abnormalities” in EGPA has been based on a number of often-vague patients’ symptoms making it uncertain to assess the real prevalence, the presentation pattern and the clinical course of ENT involvement in EGPA. The purpose of this study was to describe the frequency and the clinical presentation of ENT involvement in a series EGPA patients. We focused on sinonasal involvement and on cytological analysis, as a tool to better-diagnosed sinonasal inflammatory diseases.

Methods. – Thirty-seven EGPA pts (20F; mean age 57.7 ± 14 yrs) were enrolled in this cross-sectional study. Fiber-optic nasal endoscopy was performed in all cases leading to the following diagnosis:
- normal;
- allergic (AR) or non allergic rhinitis (NAR);
- CRSwNP or CRSSNP. In all cases, nasal cytologic analysis was performed.

NAR, CRSwNP and CRSSNP pts were further subclassified accordingly to predominant nasal cellular population. The impact of sinonasal involvement on Qol was evaluated by the SF-36 and the Sino-Nasal Outcome Test-22. Correlations between the different variables were analyzed using linear regression and the Spearman coefficient (P < 0.05).

Results. – AR was diagnosed in five, CRSSNP in nine, CRSwNP in 13, NAR in eight patients [of which five with eosinophilia (NARES), two with neutrophils (NARNE), and one without any cytological alteration], and normal in only two patients. Health-related Qol was deeply impacted by sinonasal involvement (mean SNOT22: 26.9; mean ISF-36: 42.2; mean ISM-36: 49.8).

Conclusion. – CRS represents the “clinical prototype” of ENT involvement in EGPA. Sinonasal involvement greatly affects patients’ QOL, therefore, multidisciplinary efforts are required in order to optimize nasal symptoms treatment and to improve the management of EGPA patients in clinical practice.

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P46 A case of eosinophilic granulomatosis with polyangitis (EGPA) who relapsed with saddle nose
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Introduction. – Saddle nose is a typical symptom of Granulomatosis with polyangitis (GPA). We report here an interesting case of EGPA, who was

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