Discussion.— Patient education should be targeted to provide, as quickly as possible, reliable information on the diagnosis, clinical treatment, and outcomes. There do not appear to be any differences in informational needs between UK and North American patients, suggesting that the needs reflect disease requirements more than cultural differences or the two methods of surveying the patients.

Conclusion.— This study highlights the high informational needs of patients with AAV and that patients prefer education on a one-to-one basis with a doctor.

http://dx.doi.org/10.1016/j.lpm.2013.02.201

P131
Proposal of an algorithm for the diagnosis and aetiological identification of diffuse alveolar haemorrhage (DAH)
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Introduction.— One of the major causes of mortality in some small-vessel vasculitides (SVV) is DAH. Indeed, SVV are often the main cause. However, DAH may be secondary to other aetologies like infections, blood dyscrasias, haemodynamic disorders, other autoimmune diseases, neoplastic, idiopathic, etc. Interestingly, no defined and validated algorithm to establish its presence and causes exists. Our purpose is to propose an algorithm for the diagnosis and aetiological identification of HAD.

Methods.— Based on literature review, a systematic search was done of all articles (PubMed) in English, Spanish, German and French, plus abstracts in English with the terms “alveolar hemorrhage”, “diffuse alveolar hemorrhage”, “lung hemorrhage”, “diffuse lung hemorrhage”, “diagnosis” and “algorithm” in the last 10 years. The search was limited to adult population. Selection of articles to be evaluated was agreed after discussion by at least three authors.

Results.— Forty-four articles were identified. Two included information relevant to the purpose of study. With it, plus data from the other published studies, a proposed algorithm is presented in the figure (Supplementary data). On the left side, procedures to confirm DAH are presented, and on the right, those tending to disclose the cause. Approximate times for the algorithm implementation and results retrieval are offered.

Discussion.— No consensus or validated diagnostic approach for DAH exist. In our setting, previous experience has been published for DAH in the context of SLE [1], which allowed for reduction of mortality by 50%. A systematic approach, at least in our environment, would be important to optimise the work-up of DAH and avoid delay regarding its diagnosis and cause.

Conclusion.— We offer an algorithm for studying DAH which needs prospective validation. Its implementation may reduce mortality of this life-threatening condition by identifying the cause of DAH and installing timely treatment.

Supplementary data associated with this article can be found on the website of La Presse Médicale (http://www.em-consulte.com/revue/lpm).

Reference
http://dx.doi.org/10.1016/j.lpm.2013.02.202

P132
Validation of the Birmingham Vasculitis activity score (BVAS) and the Vasculitis Damage Index (VDI) in a population of patients with ANCA-associated vasculitis (AASV) in Mexico
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Introduction.— BVAS and VDI are validated tools for evaluation of activity and damage in the vasculitides. Transcultural adaptation is recognised as important for optimal application. After translation into Spanish for Mexico, reliability, reproducibility, validity and responsiveness were tested.

Methods.— Longitudinal study in which diagnosis of the 3 AASV were made according to the 1990 ACR criteria and/or 2012 Chapel Hill nomenclature, supported by the EMEA algorithm. Patients with doubtful diagnosis, other autoimmune diseases, or lost follow-up were excluded. Transcultural adaptation included translation, back translation and review by a committee for the purpose. Internal consistency (with Cronbach’s alpha for BVAS), reproducibility (Spearman’s correlation on 40 stable patients) and responsiveness (Wilcoxon’s in two groups, one with improvement after treatment and the other on relapsing patients) were done.

Results.— Sixty-seven patients were included (June-November 2012); 41 females, 26 males; GPA-49, MPA-12, EGPA-6; 15 improved, six relapsed, 46 stable. BVAS results: internal consistency: 0.813; reproducibility: same numbers were obtained at both evaluations, with mean ± SD of 0.06 ± 0.26 (P = 1); responsiveness: for those who improved mean ± SD at time 1 4.2 ± 2.6 and at time 2, 0.9 ± 1.3, Wilcoxon’s Z value -3.4 (P = 0.001); for relapsing patients at time 1 mean ± SD of 0 (both) and time 2, 2.5 ± 1.5, Z value -2.2 (P = 0.02). VDI results: reproducibility: mean ± SD at both 2.2 ± 1.5 (P = 1); responsiveness: for those without further damage, mean ± SD of 1.9 ± 1.4 at both times, and for six patients with further damage after 6 months, mean ± SD at time 1 were 1.8 ± 1.6 and 2.1 ± 1.7 for Z -1.4 (P = 0.15) at time 2.

Discussion.— A larger number of patients with larger follow-up is needed to confirm our initial results, especially regarding VDI.

Conclusion.— BVAS and VDI translations showed good performance and reliability when applied to our patients. They can be used in Mexico and possibly other Spanish-speaking countries, as they are comparable to the original instruments.

http://dx.doi.org/10.1016/j.lpm.2013.02.203

P133
Illness perceptions and fatigue in systemic vasculitis
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Reference
http://dx.doi.org/10.1016/j.lpm.2013.02.202
Introduction. Illness perceptions, the organized cognitive beliefs patients hold about their illness, have not been assessed in vasculitis. Fatigue is highly prevalent in vasculitis, yet underlying mechanisms are unclear. This study compared illness perceptions among different vasculitides, identified risk factors for negative illness perceptions, and determined associations between illness perceptions and fatigue.

Methods. Participants were recruited from an online registry in vasculitis to complete the revised Illness Perception Questionnaire (IPQ-R) [1]. Scores on IPQ-R dimensions were compared across types of vasculitis. Cluster analysis and stepwise regression was used to identify predictors of negative illness perceptions. Fatigue was measured using the general subscale of the Multidimensional Fatigue Inventory (MFI). Patient-reported measures of disease activity and IPQ-R dimensions were assessed in relation to MFI scores using linear regression in sequential, additive models.

Results. Six hundred and ninety-two participants with nine forms of vasculitis completed the IPQ-R. For six out of eight IPQ-R dimensions, there were no signiﬁcant differences in mean scores between the different vasculitides. Scores in identity and timeline-cyclical dimensions were higher in Behcet’s disease compared to other vasculitides (13.5 vs 10.7; 4.0 vs 3.2, \( P < 0.05 \)). Younger age (OR = 1.04; 95%CI 1.02–1.06), depression (OR = 4.94; 95%CI 2.90–8.41), active disease status (OR = 2.05; 95%CI 1.27–3.29), and poor overall health (OR = 3.92; 95%CI 0.88–17.56) were associated with negative illness perceptions. IPQ-R dimensions and disease activity explained equally to the variability in fatigue scores.

Conclusion. Illness perceptions are similar across different vasculitides. Younger age is a risk factor for negative illness perceptions. Illness perceptions explain differences in fatigue scores beyond what can be explained by measures of disease activity suggesting that illness perceptions may have a causal and modifiable role in fatigue among patients with vasculitis.

Reference

http://dx.doi.org/10.1016/j.lpm.2013.02.204

P134
Successful treatment with rituximab in patients with Schnitzler syndrome

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Introduction. Schnitzler syndrome is a rare inﬂammatory disease characterised by chronic urticaria and monoclonal IgM gammapathy. The treatment is empirical.

Methods. We present the case of effective treatment with RTX.

Results. In 2004, 47-years-old male developed recurrent urticaria with high fever, shivering, bone pain, myalgia and arthralgia. Investigation showed no signs of neoplasm, infection or systemic disease. Antibodies and NSAID were ineffective. Blood test: high WBC, CRP and IgM, negative tests for viral hepatitis and HIV. Skin biopsy: non-speciﬁc vasculitis, no amyloid. Serum and urine immunoelectrophoresis: monoclonal IgMx. Bone marrow: no signs of malignancy. We diagnosed Schnitzler syndrome according to Lipsker criteria. The patient was treated with PRED 30 mg and HCQ 400 mg with slight effect. AZA, MXM and colchicine were also ineffective. Fever and skin rash diminished after pefloxacin (PEF) treatment (800–1200 mg). The dose of PRED was reduced to 10 mg. In spite of clinical improvement patient still presented with recurrent urticaria, bone pain, active inﬂammation and monoclonal IgM level. Later he developed peripheral neuropathy. In 2010 after second treprostinib the patient was treated with RTX (2000 mg, than 1000 mg every 6 months for 2 years) that signiﬁcantly improved clinical and laboratory data and reduced monoclonal secretion. After 2 years RTX was well tolerated (table 1).

Conclusion. Schnitzler syndrome has favourable prognosis though benign IgM gammapathy can transform into Waldenström’s macroglobulinemia, lymphoma or other blood dyscrasias. It is presumed that treatment can reduce the risk of late complications. The efﬁcacy of RTX is not well established. We showed that RTX can lead to improvement of clinical and laboratory parameters in Schnitzler syndrome.

http://dx.doi.org/10.1016/j.lpm.2013.02.205

P135
Rituximab and vascular function in granulomatosis with polyangiitis (GPA)

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Introduction. GPA is associated with 6.7 odds-ratio of cardiovascular events compared to the general population [1], mostly accounted by the degree of immune-mediated inﬂammation rather than traditional vascular risk factors. Rituximab, an alternative induction therapy for GPA, was associated with beneﬁcial vascular effects in mouse models of atherosclerosis [2], human lupus [3] or rheumatoid arthritis [4,5]. We hypothesised that rituximab would improve endothelial function in GPA to a greater extent compared to conventional immunosuppression.

Methods. Pilot data from 11 active GPA patients (nine treated with rituximab (R), two treated with cyclophosphamide (C) as per routine clinical care) are presented. Vascular studies were scheduled pre- and 6 months post-immunosuppressive therapy, with follow-up data available in 4/11 patients (n = 3 (R), n = 1 (C)). Data was expressed as mean ± SEM.

Forearm blood flow responses to intra-arterial infusions of acetylcholine (ACh, endothelium-dependent vasodilatation independently correlated to cardiovascular outcomes), sodium nitroprusside (SNP) and Nmonomethyl-L-arginine were measured using venous plethysmography [6]. Arterial stiffness was measured non-invasively, by recording arterial pulse wave velocity (PWV) using a SphygmoCor device.

Results. Mean age was 59 ± 3 years (8:3 male:female), systolic BP 145 (±6) mmHg/diastolic BP 84 (±2) mmHg and total cholesterol 5 ± 0.6 mmol/L. Baseline PWV was elevated at 9.0 (±0.7) m/s versus normal value for age (8.0 m/s) [7]. The mean FBF response to 15 mcg ACh improved after 6 months in 3 R patients but worsened in the C patient (ﬁgure 1).

| Table I |
| Labotatory tests after treatment |
| 2007 (no treatments) | 2008 (PRED + HCQ) | 2010 (PRED + HCQ+PEF) | 2011 (PRED + after 1st RTX) | 2012 (low dose PRED + RTX) |
| WBC | 20.5 | 24.5 | 23.3 | 14.7 | 11.6 | 4–10 |
| ESR, mm/h | 92 | 84 | 44 | 46 | 28 | 2–30 |
| CRP, mg/dl | 12.8 | 8.5 | 7.6 | 2.3 | 1.6 | 0–0.8 |
| IgM, mg/ml | 560 | 1219 | 884 | 559 | 311 | 60–405 |
| IgMv, g/l | 4.7 | 4.9 | 5.2 | 3.6 | 2.3 | 0 |

Figure 1