utility of 18-FDG PET/CT to measure the disease activity by studying wall enhancement compared to the clinimetric assessment has been slightly studied.

Objectives: To explore the agreement between 18-FDG PET/CT and the clinimetric tools for the estimation of Takayasu activity in one national reference centre.

Methods: The clinical records of patients that had performed an 18-FDG PET/CT were consecutively included. The required information to fulfil the ITAS2010, ITAS.A, NIH and DR were gathered from clinical charts. The cut-off points we used are the following: SUVmax ≥ 2.1 for 18-FDG PET/CT, for ITAS2010 ≥ 2 points, for ITAS.A ≥ 4 points, for NIH ≥ 2 points and for DR ≥ 5 points. Kappa index was calculated, comparing SUVmax with all the clinimetric measures. As an exploratory exercise, ROC curves were performed. A p value less than 0.05 was considered statistically significant.

Results: Thirty six clinical records were reviewed. There was enough information to score ITAS2010 in 31 patients, ITAS.A in 28 patients, NIH and DR in 35 patients each. In our patients, moderate agreement was observed between 18-FDG PET/CT and DR score (Kappa=0.542, p=0.001). A tendency of weak agreement was observed with the NIH score (Kappa=0.215, p=0.086) and ITAS.A (kappa=0.351, p=0.063). There was no agreement with ITAS2010 (Kappa=0.107, p=0.519). Significant AUC were observed with DR (AUC=0.817, p=0.005) and NIH (AUC=0.756, p=0.025); however, this results were not obtained with ITAS2010 (AUC=0.675, p=0.124) and ITAS.A (AUC=0.697, p=0.083).

Conclusions: There was no strong agreement between 18-FDG PET/CT and any of these activity indices. On the other hand, these data suggest that the best disease activity tool in Mexican patients were DR and the NIH scores. Comparative studies in other populations are warranted.

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AB0656 CRYOGLOBULIN EVALUATION: ANALYSIS OF INTRA-LABORATORY AND INTER-LABORATORY VARIABILITY

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Background: Cryoglobulins (CRG) are immunoglobulins that precipitate in serum at temperatures below 37°C and resolubilize upon warming. The main reasons of interest of a clinical pathologist in the study of cryoglobulinemia are: 1) lack of standardisation in the preanalytical, analytical and postanalytical phases of the process (classification and reporting); 2) peculiarities of physiopathological mechanism 3) important clinical consequences. Vermeersch et al. studied these issues in 2008. To assess current practice in the detection, analysis, and reporting of cryoglobulins, a questionnaire was sent to 140 laboratories. They showed that only 36% of laboratories used standard procedures of analysis. Consequently, they concluded that standardisation was needed for cryoglobulin detection to avoid missed diagnoses and improve the comparability of results. Sargur et al. in 2010 reviewed the classification and clinical features of cryoglobulins and suggested “best practice” guidelines for laboratory detection and identification of cryoglobulins. They particularly highlighted the relevance of preanalytical and analytical phases: maintenance of the sample at a stable temperature of 37°C, especially throughout the initial steps (collection and transportation); centrifugation and separation methods; cryoprecipitate quantification; cryoprecipitate washing techniques; immunocharacterization of cryoprecipitates especially through immunofixation techniques (considered the “gold standard”).

Objectives: To verify and assess the variability of laboratory processes of CRG.

Methods: We checked laboratory databases of Hospital and University (Lab A and B) of Modena with long tradition in the cryoglobulin analysis (more than 6000 tests from 2002 to 2017). Concerning CRG testing, 734 patient samples were studied in both laboratories. We compared our results according to Brouet classification into subgroups: type I, II and III. Therefore, we evaluated intra-laboratory variability, compared to previous or more frequent results. Finally, we studied inter-laboratory variability based on non-concordant laboratory reports.

Results: In the following table, we have represented the comparison between labs about the same patient cohort in 734 patient samples:

Abstract AB0656 – Table 1.

	I type (n)	II type (n)	III type (n)
Lab A	21	242	108
Lab B	42	270	108
Chi-quadro	p=0.0016	p=0.0004	Ns

Intra-laboratory variability: 14% Lab A, 16% Lab B (ns). Inter-laboratory variability: non-concordance in 25% of cases, considering 133 patients studied in both laboratories (Chi-square test).

Conclusions: No data about variability in CRG analysis are reported in literature. National and international guidelines are not explicative enough. Furthermore, many doubts about classifications are established. Our experience is unique but limited in two laboratories. Given the variability of testing conditions used in different laboratories and the lack of test standards and reference values, we confirm the need of further investigations into standardisation of CRG testing. New guidelines are fundamental, in order to optimise all phases of CRG research (pre and post analysis) and to ensure correct diagnosis and adequate treatments of the associated diseases.

Disclosure of Interest: None declared

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AB0657 CYCLOPHOSPHAMIDE-SPARING ROLE OF AN INTENSIFIED B-CELL DEPLETION PROTOCOL IN ANCA-ASSOCIATED VASCULITIS: A CASE-CONTROL STUDY

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Background: The management of ANCA-associated-vasculitis (AAV) requires the use of immunosuppressive drugs with potential toxicity. Recently, two trials demonstrated the efficacy of Rituximab (RTX) for the therapy of AAV.

Objectives: this case-control study aims to evaluate the immunosuppressive-sparing effect of a RTX-based protocol compared to the standard CYC treatment.

Methods: 26 patients with AAV and extracapillary glomerulonephritis were prospectively enrolled. Thirteen patients received an intensified protocol of B-cell depletion therapy (IBCDT) consisting of 4-weekly infusions of 375 mg/sm RTX followed by 2 infusions after 1 and 2 months, 3 pulses of methylprednisolone followed by prednisone tapered to 5 mg/day in three months and 2 pulses of 10 mg/kg CYC, without further maintenance therapy. Thirteen patients treated with 2 mg/kg/day CYC followed by azathioprine as a maintenance therapy served as controls.

Results: A significant improvement (p<0.05) of B-VAS, ESR, CRP and ANCA was observed in the IBCDT-group at 3, 6 and 12 months, with decrease of mean creatinine values from 4.81±6.4 mg/dl to 2.21±3.8 mg/dl.

When compared to controls, no difference was observed in terms of complete and partial response. However, the IBCDT regimen achieved a 1 g/month reduction of CYC cumulative dose (p<0.001).

Conclusions: in the treatment of this sample of severe AAV patients, the IBCDT protocol appeared to be noninferior to CYC-based regimen. Notably, the IBCDT regimen allowed a significant reduction of CYC cumulative dose.

Disclosure of Interest: None declared

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AB0658 DIFFERENT ORBITAL MANIFESTATIONS OF GRANULOMATOSIS WITH POLYANGIITIS. COMPARATIVE STUDY

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Background: Ophthalmic manifestations are typical for granulomatosis with polyangiitis (GPA), and occur in 28.6%–60% of patients. In 8% of cases they lead to permanent visual loss. According to different studies orbital lesion develops in 5%–30.6% of GPA patients and is considered to be the second most prevalent ophthalmic manifestation after conjunctivitis/episcleritis.

Objectives: to study clinical features of different orbital manifestations of GPA.

Methods: 74 GPA patients with orbital involvement were studied and compared. 3 types of orbital involvement were proposed: orbital mass (45 patients),

dacryoadenitis (26 patients) and myositis (3 patients). Patients with orbital mass (group 1) and dacryoadenitis (group 2) were included into further investigation.

Results: Patients in group 1 had systemic disease in 51.1% compared to 7.7% in group 2. Patients with orbital mass presented with more severe clinical manifestations, including significant exophthalmos (82.2%), periorbital oedema (95.5%) and hyperemia (46.7%), strabismus (31.1%), globe movement restriction (71.1%), chemosis (8.9%). Orbital mass was accompanied by necrotizing scleritis in 7 and PUK in 6 patients. Symptoms included orbital pain (88.9%) and binocular diplopia (73.3%). BCVA in this group was 0.67. Clinical outcome was relatively unfavourable: in 24.4% BCVA was less than 0.3, in 5 patients enucleation was performed, in 24.4% strabismus developed and in 73.3% globe movement restriction occurred. Clinical course and outcome were significantly more favourable in patients with dacryoadenitis. Periorbital oedema, exophthalmos and chemosis did not show significant difference in groups, though orbital pain, binocular diplopia, globe movement restriction, strabismus, optic neuropathy, necrotizing scleritis, PUK were observed in group1 significantly more often. Clinical outcome in group 2 was much more favourable with no cases of enucleation and persistent visual loss. They only had moderate exophthalmos 1–3 mm (65.4%), strabismus (7.7%) and mild globe movement restriction (19.2%).

Conclusions: Three types of orbital involvement in GPA were proposed: orbital mass, dacryoadenitis and myositis. Patients with orbital mass are characterised more often with systemic disease, more severe clinical course not uncommon associated with other ocular manifestations (necrotizing scleritis, PUK, orbital walls destruction), relatively unfavourable outcome with high level of morbidity (optic nerve atrophy, anophthalmos, strabismus) and higher rate of recurrences.

Disclosure of Interest: None declared

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AB0659 PREVALENCE AND CLINICAL FEATURES OF CRANIAL AND EXTRACRANIAL GIANT CELL ARTERITIS

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Background: Giant cell arteritis (GCA) is the most common systemic vasculitis in adults. Although it typically affects the cranial branches derived from the aortic arch, there is increasing evidence of the damage occurring to large vessel extracranial arteries, which is usually misdiagnosed. This subset of large vessel extracranial GCA may have specific clinical features that could hinder the diagnosis, which highlights the need for a different treatment and follow up.

Objectives: To analyse the frequency and clinical and analytical features in cranial and extra-cranial GCA.

Methods: An observational retrospective descriptive study analysing data from patients with GCA was performed in our hospital. Colour duplex sonography (CDS) studies carried out in the last 29 months were revised. Standardised CDS images from the frontal and parietal branches of the temporal superficial artery (TA) and axillary artery (AXE) with GCA compatible image with intra or extra-cranial involvement (hypochoic, homogeneous and circumferential thickening of the artery wall ≥ 0.34 mm in TA, and ≥ 1 mm in AXE, respectively). SPSS version 23 were used to analyse a total of 29 variables.

Results: Out of the 119 patients that were tested in our fast track GCA clinic, with standardised CDS of TA and AXE, 22 had GCA. All GCA patients was halo sign in CDS. From them, 7 had large vessel involvement (31.8%), from whom 6 were mixed and 1 had exclusively extra-cranial involvement. Mean age at diagnosis was 76 ± 9.7 SD in the extracranial GCA arteritis and 79 ± 5.7 SD in the cranial GCA. The extracranial subset had a greater number of men than the cranial (28.6% and 26.6%, respectively) and suffered usually more fever (28.5% > 6.6%, respectively), constitutional syndrome (85.7% > 40%, respectively) or polymyalgia rheumatica (42.8% > 40%, respectively). However, they suffered with more frequency from headache (85.7% vs 93.3%) and jaw claudication (28.5% vs 33.3%). None of them suffered from visual impairment or central nervous system involvement; both do appeared in the cranial GCA group in 13.3% and 6.6%. In the blood tests, mean $\pm$ standard deviation was: ESR 87.6 ± 36 mm/h in cranial GCA and 89.5 ± 19.5 SD mm/h in extracranial GCA, CRP 65 ± 57.6 and 82.6 ± 48.4 mg/L and Hb 11.4 ± 1.3 and 12.1 ± 1.3 g/dl, respectively. Patients with large vessel involvement met ACR criteria in 80% opposite 92.8% from these with solely cranial GCA. AT biopsy was performed in 7 patients in the cranial subset and 2 in the extra-cranial, testing positive for GCA in 5 (71.4%) in the first group, and none in the second one. CT-PET was performed in 2 patients with cranial GCA with negative results, and in 4 with large vessel involvement, testing positive in 50%. Despite the large differences, the statistical analysis didn't reach statistical significance due to the small sample size.

Conclusions: One third of the patients in our study had large vessel involvement, making axillary vessel CDS an important tool for the clinical exam of this disease. These patients tend to be younger and start more often with fever or general

syndrome and less with GCA typical symptoms like headache, jaw claudication or visual loss.

Disclosure of Interest: None declared

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AB0660 THE ROLE OF ANTI-NEUTROPHIL CYTOPLASMIC AUTOANTIBODY SPECIFICITY FOR MPO OR PR3 IN PHENOTYPE OF ANCA ASSOCIATED VASCULITIS: KNOWING THE AUTOIMMUNITY IN LATIN AMERICA

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Background: Antineutrophil cytoplasmic antibodies (ANCA) are present in up to 90% granulomatosis with polyangiitis, 80% microscopic polyangiitis and 70% eosinophilic granulomatosis with polyangiitis. MPO-ANCA has been associated with vasculitis limited to the kidney, chronic renal damage and less frequent gastrointestinal or respiratory tract involvement. PR3-ANCA are characterised by destructive lesions of the ears, nose and throat, alveolar haemorrhage, combination of upper and/or lower respiratory tract involvement with renal compromise and increased number of relapses. The frequency of pulmonary involvement is similar in both serotypes, and most ANCA associated vasculitis (AAV) patients are diagnosed between ages 50 and 70 years.

Objectives: To describe differences in clinical profiles of patients with AAV regarding ANCA specificity against MPO or PR3 in a Colombian based adult population

Methods: All medical records of patients with a diagnosis of AAV in two high complexity hospitals in Medellín, Colombia from January 1, 2014 to December 31, 2016 were reviewed. The clinical and demographic characteristics were abstracted and analysed with descriptive and inferential statistics in SPSS.22

Results: Of 59 cases of AAV, 44 were positive for MPO or PR3-ANCA with male predominance (65.5% men vs 34.5% women) and similar age at diagnosis (47 years in MPO-ANCA vs 50 in PR3-ANCA). MPO-ANCA group had more fever and weight loss (34.8% vs 20%), arterial hypertension (34.5% vs 26.7%), hematuria (34.5% vs 26.7%), proteinuria (31% vs 26.7%), creatinin higher than 5.6 mg/dL (20.7 vs 13.3%), myalgias (13.8 vs 0%) pachymeningitis (7% vs 0%) and skin compromise. PR3-ANCA patients had more arthralgias/arthritis (40% vs 31%), escleritis (33% vs 13.8%), episcleritis (13.3% vs 0%) and uveitis (10% vs 7%).

Conclusions: In this Latin American population ANCA specificity affected the phenotype of clinical disease. MPO-ANCA patients had more constitutional symptoms, renal and central nervous system compromise while PR3-ANCA patients showed more articular and ocular involvement.

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AB0661 ASSESSMENT OF DAMAGE AND PROGNOSIS IN PATIENTS WITH ADULT IGA VASCULITIS: RETROSPECTIVE MULTICENTERED COHORT STUDY

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Objectives: IgA Vasculitis is a leukocytoclastic vasculitis involving small vessels with depositions of immune complexes containing IgA. IgA Vasculitis is a predominantly paediatric vasculitis. There is limited data for the prognosis of adult IgA Vasculitis, with also no damage assessment. In this study, we aimed to evaluate