syndrome pts (15,3 per 100 patient-years). The probability of EN recurrence in *Löfgren syndrome pts* was significantly higher in subjects older than 40 y (OR 3,81; $p=0,03$), in subjects with late (≥ 3 months from EN onset) initiation of treatment (OR 8,94; $p=0,0008$) and in subjects with indurations >5 cm in size (OR 3,65; $p=0,03$). The probability of LDS recurrence was significantly higher in pts with confluent nodules, forming conglomerate masses (OR 4,33; $p=0,037$), and pts taking hydroxychloroquine at ≤ 200 mg/day during ≥ 6 months (OR 5,25; $p=0,019$).

Conclusions: Comprehensive examination is needed to identify the clinical type of Pn. Symmetrical involvement of all extremities is characteristic for EN in young subjects. Asymmetric nodular rash over lower extremities in subjects with abnormal BMI and CVI is specific for LDS. CWD usually presents with nodular rash on the trunk. Identified risk factor for EN and LDS recurrence should be taken into consideration while writing a treatment plan.

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AB1133 ROLE OF THE RHEUMATOLOGIST IN A REGIONAL REFERENCE HAEMOPHILIA UNIT

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Background: Haemophilia is an X-linked hereditary bleeding disorder caused by deficiency in coagulation factor VIII (FVIII), in haemophilia A (HA), and factor IX in haemophilia B (HB).

They are classified as severe, moderate or mild, depending on the level of coagulation factor deficiency. Men are affected clinically by the disease, and women, who are carriers, usually remain asymptomatic.

HA is more common than HB (from 80% to 85% of all cases).

Their bleeding complications primarily affect the musculoskeletal system. Hemarthrosis is the major hemophilia-related complication, responsible for a particularly debilitating chronic arthropathy, in the long term, affecting mainly the load joints (knees, ankles and elbows).

In addition to clotting factor concentrates, usually prescribed by the haematologist, The management of acute hemarthrosis and chronic arthropathy requires a close collaboration with rheumatologists. This collaboration is the key to effectively preventing hemarthrosis, managing acute joint bleeding episodes, assessing joint function, and actively treating chronic arthropathy.

Objectives: To analyse the clinical characteristics, extent of joint involvement and associated comorbidity of a cohort of patients with hemophilic arthropathy.

Methods: This is a retrospective study, carried out in the Haemophilia Unit of our hospital (regional reference), in patients with moderate to severe haemophilia A and B, with hemophilic arthropathy, seen in consultation with episodes of joint bleeding (2007–2017). Severity of haemophilia was defined based on determined by the percentage of FC activity (VIII and IX), moderate from 1% to 5%, severe $<1\%$. The number of episodes of hemarthrosis was collected in the 3 months before and after the radioisotopic synoviorthesis (intra-articular injection of a colloidal suspension of particles marked with a radioisotope, whose objective is the destruction of the synovial membrane).

Results: We included 88 patients (87 men and 1 symptomatic carrier woman with decreased levels of factor VIII), mean age 31 ± 17 years. HA (severe 56%, moderate 26%), HB (severe 14%, moderate 1%). The target joint: knee 51%, followed by ankle 26%, elbow 13% and other 7% (5 shoulders and 2 wrists). In 61 patients, magnetic resonance imaging (MRI) was performed: synovial hypertrophy 9%, hemosiderin deposits (in acute stage of joint bleeding) 2% and structural alteration (erosions and subchondral cysts, loss of focal cartilage) 68%. A radioisotope synoviorthesis was made to 18 patients: 12 with sulfide 186 Re colloidal (5 ankles, 4 knees, 2 elbows) and 6 with 90 Y coloidal citrate (4 knees, 3 ankles), having a decrease of 74% (range 59%–100%) in the number of hemarthrosis in the 3 subsequent months. Total knee replacement was needed in 13% of the patients (7 with HCV liver disease and in 6 HCV liver disease and coexistence with HIV). They have infection due to HCV 33%, HIV 25% and HBV 6%.

Conclusions: This study highlights the extent of joint damage in haemophilic patients as well as the high comorbidity of HCV and HIV infections.

The experience of a monographic Haemophilia consultation, with the participation of different specialties (being fundamental the rheumatologist), benefits the multi-disciplinary approach of these patients, being the results obtained in our series concordant with the described in the literature.

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AB1134 DETERMINING FACTORS OF SEVERITY IN HEMOPHILIC ARTHROPATHY

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Background: In patients with haemophilia, the development of inhibitors to factor VIII/IX (Haemophilia A and B, HA/HB), prevents adequate replacement therapy and results in increased risk of serious bleeding episodes, poor control of joint bleeding, and progressive, debilitating joint disease.

Objectives: To describe, according to the Arnold-Hilgartner scale (AHRs), the radiological findings in a cohort of hemophilic arthropathy (HArth) patients and to analyse the relationship that may exist between the degree of joint involvement and HA/HB, severity, age and presence or absence of inhibitor.

Methods: This is a retrospective study, carried out in the Haemophilia Unit of our hospital, in patients with HArth (2007–2017). Severity of haemophilia: percentage of coagulation factor (CF) activity, moderate (1%–5%) and severe $<1\%$. The AHRs includes 5 levels.

For the study of the association of the type and degree of haemophilia; and the presence of inhibitor, with the level in the AHRs, a linear model was used. The significance in this regard of the linear contrasts of interest was studied using F tests, defining the first species error at 0.05.

Results: We included 88 patients. Characteristics of the patients in the attached chart. No significant association was detected between the type and severity of haemophilia with the development of inhibitor.

The results of the linear model only showed association ($p=0.01$) between the severity of haemophilia and AHRs, patients with moderate haemophilia presented a least square mean for the AHRs of 2.6 (0.3), whereas the serious ones had a value of 3.4 (0.2). The least squared means for patients with HA and HB were 2.9 (0.2) and 3.1 (0.4) respectively, the contrast did not reach statistical significance ($p>0.1$), and the same occurred between patients who had generated or not inhibitor 2.8 (0.2) Vs 3.1 (0.3). Apart from the severity of haemophilia, the other factor that showed a significant and significant effect ($p<0.001$) on the AHRs values was age. The estimated value for the regression coefficient of age on the radiological status was 0.05 (0.009).

Abstract AB1134 – Table 1

Type of Haemophilia	
Haemophilia A n (%)	73 (82%)
Haemophilia B n (%)	14 (15%)
Severity of Haemophilia	
Haemophilia A severe n (%)	50 (56%)
Haemophilia A moderate n (%)	23 (26%)
Haemophilia B severe n (%)	13 (14%)
Haemophilia B moderate n (%)	1 (1%)
Age m (SD)	31 (17)
Development of inhibitors to factor VIII/IX n (%)	16 (18%)
Haemophilia A severe with inhibitors n (%)	10 (11%)
Haemophilia A moderate with inhibitors n (%)	3 (3%)
Haemophilia B severe with inhibitors n (%)	3 (3%)
AHRs scale 1 n (%)	18 (20%)
AHRs scale 2 n (%)	17 (19%)
AHRs scale 3 n (%)	18 (20%)
AHRs scale 4 n (%)	15 (17%)
AHRs scale 5 n (%)	19 (21%)

Conclusions: Contrary to what has been observed in other studies, in our cohort it does not seem that the presence of inhibitor or the type of haemophilia has a negative effect on the severity of the radiological findings. The age and severity of haemophilia do seem to influence the radiological stage.

Disclosure of Interest: None declared

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AB1135 CARDIAC MAGNETIC RESONANCE FINDINGS IN PATIENTS WITH BIOPSY-PROVEN VIRUS-NEGATIVE LYMPHOCYTIC MYOCARDITIS

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Background: Cardiac magnetic resonance (CMR) is considered to be useful for non-invasive myocarditis diagnosis. Lake Louise Criteria (LLC) has been proved to have moderate accuracy in the diagnosis of acute or chronic myocarditis¹.

Objectives: to describe the CMR findings in patients with virus-negative lymphocytic myocarditis (VNLm) from a large monocentric Italian cohort.

Methods: CMR findings in 40 patients (mean age 45.43 ± 14.7 years; male:female ratio 1:1) diagnosed with endomyocardial biopsy-proven VNLM at our Centre from January 2015 to December 2017, were retrospectively evaluated. CMR was performed at time of clinical presentation and before EMB. At contrast-enhanced CMR we analysed: ventricular function, T2-weighted sequences, short inversion time inversion recovery sequences (STIR), early (EGE) and delayed gadolinium enhancement (LGE). Demographic, clinical and histological data were correlated with CRM findings.

Results: Mean left ventricular (LV) ejection fraction (EF) was $52.5\% \pm 10.8\%$. 50% of patients had reduced LVEF ($<55\%$). Mean right ventricle EF was $55.25\% \pm 7.2\%$. Akinetic or hypokinetic areas were detected in 3 (7.5%) and 17 (42.5%) patients, respectively. LLC were identified in 39 patients (92.5%). LGE was the most common CMR finding. 20 patients (50%) met one LLC, 16 patients (40%) 2 LLCs. 1 patient met all 3 LLCs. STIR abnormalities were identified in the LV lateral wall in 8 (19%), septal wall in 9 (21.4%), LV inferior wall in 9 (21.4%), in the apex in 3 (7.1%), LV posterior wall in 3 (7.1%) and LV anterior wall in 2 (4.8%) patients. Myocardial oedema on T2 images was detectable in 21 (52.5%), EGE in 1 (2.5%) and LGE in 36 (90%) patients. LGE was intramural in 22 (55%), subepicardial in 23 (57.5%) and subendocardial in 4 (10%) patients. Multiple LGE patterns were disclosed in 10 (25%) patients. Pericardial effusion was detected in 9 patients (22.5%). 2 patients did not meet any LLC at CMR. When comparing CMR findings with Holter-ECG tape, clinical presentation, histology, biochemistry and autoimmunity results we found CRM oedema positively correlated with the detection of active myocarditis on EMB (11 vs 4, $p=0.027$). Serum anti-heart antibodies positively correlated with the detection of pericarditis at CMR (3 vs 1, $p=0.050$). Patchy/intramural LGE positively correlated with rhythm abnormalities at Holter-ECG tape (12 vs 6, $p=0.037$).

Conclusions: VNLM is an overlooked disease defined by EMB established histological, immunological and immune-histochemical criteria. Although the diagnostic power of CMR needs to be further investigated, it seems to correlate with EMB results and arrhythmic burden.

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AB1136 CLINICAL FEATURES OF ADULT CHINESE PATIENTS WITH PERIODIC FEVER, APHTHOUS STOMATITIS, PHARYNGITIS, AND ADENITIS: REPORT FROM AN SINGLE CENTRE

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Background: Periodic fever, aphthous stomatitis, pharyngitis, and cervical adenopathy (PFAPA) syndrome is a polygenic or multifactorial autoinflammatory disease (AUIDs) with unknown etiology. It mainly affects children, though recently it has been also recognised in adulthood. PFAPA syndrome has not been reported in the Chinese population up till now.

Objectives: We aimed to investigate the clinical features, treatment and prognosis of the first cohort of adult Chinese patients with PFAPA syndrome.

Methods: We prospectively evaluated clinical data of adult patients (≥ 14 years) with recurrent fever of unknown origin who were suspected monogenic AUIDs in the period April 2015 to December 2017, at the adult AUIDs centre, Department of Rheumatology, Peking Union Medical College Hospital. Gene tests of AUIDs associated pathogenic genes were examined to exclude monogenic AUIDs. The clinical features were evaluated for patients with PFAPA syndrome, and were compared with a newly reported large Italian cohort of adult patients with PFAPA syndrome.¹

Results: During the study period, a total of 7 adult patients with PFAPA syndrome were diagnosed (table 1), and the male to female ratio was 6:1. All these patients had a disease onset in adulthood, and the mean age of onset was 27 ± 10 years, and the mean time of delay of diagnosis was 4.9 ± 4.0 years. Clockwork periodicity fever was the main feature of PFAPA syndrome, and the median duration of attacks was 3.5 days, and the median interval between attacks was 4 weeks. Pharyngitis, adenitis and aphthous stomatitis were present in 85.7%, 57.1% and 42.9%, respectively. Other common symptoms included fatigue (100%), headache (71.4%), and myalgia (71.4%). The presence of arthralgia/arthritis was less frequent compared to the Italian cohort ($p=0.012$) (table 2). Corticosteroids given during attacks were effective in 3 patients. NSAIDs were partially effective but tonsillectomy was of no effect. Colchicine was rarely used compared to the Italian cohort ($p=0.046$).

Abstract AB1136 – Table 1. Summary of the clinical characteristics of our patients.

No. of patients	7
Male	6 (85.7%)
Female	1 (14.3%)
Age at onset (years)	27 ± 10 (16-46)
Age at diagnosis (years)	31 ± 11 (17-53)
Delay in diagnosis (years)	4.9 ± 4.0 (1.0-11.0)
Adult onset	7
Positive family history	0
Duration of febrile attacks (days)	3.5 (0.5-7.0)
Duration of attack intervals (days)	4 (1-12)
Frequency of attacks per year	10 (4-20)
Peak temperature (°C)	39.2 ± 0.6 (38.5-40)
Periodic Fever with 3 major clinical features	2 (28.6%)
Periodic Fever with only 2 major clinical features	2 (28.6%)
Periodic Fever with only 1 major clinical features	3 (42.9%)

Clinical features	Ours (n, %) (n=7)	Italian (n, %) (n=30)	p value
Pharyngitis	6 (85.7)	23 (76.7)	0.601
Cervical adenopathy	4 (57.1)	22 (73.3)	0.399
Aphthous stomatitis	3 (42.9)	15 (50)	0.734
Fatigue	7 (100)	19 (63.3)	0.056
Headache	5 (71.4)	13 (43.3)	0.181
Myalgia	5 (71.4)	16 (53.3)	0.384
Conjunctivitis	2 (28.6)	4 (13.3)	0.325
Arthralgia/arthritis	1 (14.3)	20 (66.7)	0.012
Nausea and vomiting	1 (14.3)	5 (16.7)	0.878
Chest pain	0 (0)	6 (20)	0.196
abdominal pain/diarrhea	0 (0)	8 (26.7)	0.123
Periorbital edema	0 (0)	5 (16.7)	0.245
Rash	0 (0)	5 (16.7)	0.245
Treatment			
Glucocorticoid	3/3 (100)	15/17 (88.2)	0.531
NSAIDs	1/3 (33.3)	10/12 (83.3)	0.080
Colchicine	0/1 (0)	3/3 (100)	0.046
Tonsillectomy	0/2 (0)	1/3 (33.3)	0.361

Abstract AB1136 – Table 2. Comparison of with an Italian cohort of adult patients with PFAPA syndrome

Conclusions: PFAPA syndrome is one of the main causes of adults Chinese patients with recurrent fever of unknown origin and might be misdiagnosed. Clinicians, not only paediatricians, should take into account this clinical entity, especially for patients with pharyngitis, cervical adenopathy and aphthous stomatitis.

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AB1137 INFLAMMATORY AND EROSIVE ARTHRITIS, A COMMON UNKNOWN MANIFESTATION OF VOGT-KOYANAGI-HARADA DISEASE (VKHD)

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Background: VKHD is a rare cause of bilateral granulomatous panuveitis leading to significant visual loss in patients who present it. VKHD is a multisystemic, autoimmune, inflammatory disease that usually displays extraocular manifestations that include meningismus, vitiligo, poliosis, and hearing loss. Arthritis is an unknown manifestation of the disease. In Mexico, VKHD affect mainly young women and is responsible for 13% to 27% of all uveitis.

Objectives: To describe the presence of inflammatory and erosive arthritis in patients with VKHD.

Methods: Observational study of patients with established VKHD that fulfilled the 2001 revised diagnostic criteria for VKHD was rolled out. Articular ultrasonography was performed to all patients by a trained rheumatologist in carpal, metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints evaluating