those who responded partially evolved to liver cirrhosis, with a similar follow-up time in both groups (105 vs 113 months). 5 of the 6 patients who did not have a biochemical response developed liver cirrhosis. 7 of the 13 patients with cirrhosis (54%) already presented clinical or histological data of cirrhosis in the initial evaluation

Conclusions: PBC patients have frequently other autoimmune diseases such as Autoimmune Hepatitis, Sjögren syndrome or Scleroderma so we must actively seek the presence of these pathologies. The treatment with ursodeoxycholic seems to be useful in all patients but it is important to make an early diagnosis

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AB1144

HISTOLOGY OF ROSAI-DORFMAN DISEASE IN A SUBSET OF PATIENTS WITH ERDHEIM-CHESTER DISEASE: A DISTINCT ENTITY MAINLY DRIVEN BY MAP2K1

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Background: Diagnosis of Erdheim-Chester disease (ECD) is based on characteristic imaging of bone, retroperitoneal and/or cardiovascular involvement.¹ Biopsy is mandatory to exclude other diagnoses and confirm infiltration of histiocytes, but histology is not specific.² By contrast diagnosis of Rosai-Dorfman disease (RDD), a rare histiocytosis, is based on histology, which is characterised by infiltration by CD68 + CD1 a- S100+ histiocytes with large nuclei and abundant lesions of emperipolesis.² Up to 70% of ECD have *BRAF* or *MAP2K1* mutations,³ which are rare in RDD.

Objectives: We investigated patients harbouring an ECD phenotype but RDD histology.

Methods: We reviewed records of ECD patients followed in Pitié-Salpêtrière hospital (Paris, France) and Memorial Sloan Kettering Cancer Centre (New-York, NY, USA) between 2007 and 2018. Biopsy samples of the patients were systematically investigated for mutations of genes of MAP kinase pathway.

Results: Among 209 patients with ECD, we found 10 (4.7%) patients who had RDD histology. These 10 patients had typical ECD clinical and radiological presentation, in particular bones (n=7), vascular (n=5) and peritoneal (n=6) involvements. Patients also had typical neurological involvement of ECD (n=6). All patients except one had at least one biopsy with a compatible histology of ECD at diagnosis. ECD biopsies showed non-specific fibrosis (n=5), foamy CD 68 + CD1 a- histiocytes (n=3) and/or Touton cells (n=1). Biopsies disclosing RDD histology were performed during the course of the disease involving testes (n=5), stomach (n=1), tibia (n=2), cheek (n=1) and omentum (n=1). All tissues showed lympho-plasmocytic infiltrate with large histiocytes infiltration. Histiocytes were CD68 + CD1 a- S100 + with large nuclei and abundant lesions of emperipolesis. Five patient harboured *MAP2K1* mutation and one patient had *PIK3CA* mutation. None of the patients had *BRAF* mutation.

Conclusions: Some patients with ECD may also present the iconic histological lesions described by Rosai and Dorfman. Overlap forms of such distinct histiocytoses between ECD and RDD is mainly driven by *MAP2K1* but not by *BRAF*.

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AB1145

ACUTE POSTERIOR MULTIFOCAL PLACOID PIGMENT EPITHELIOPATHY: CLINICAL PATTERN AND MANAGEMENT OF 79 PATIENTS

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Background: Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE) is an uncommon inflammatory disease causing acute-onset chorioretinal bilateral disease. It typically affects the posterior pole of both eyes leading to visual blurring or scotomas. Although it is thought to be benign, APMPPE has been associated with central nervous system (CNS) manifestations: cerebral vasculitis, meningoencephalitis and cerebral vascular disease.

Objectives: The aim of this study was to define clinical features, systemic manifestations, treatment and outcomes of a review of 79 patients with APMPPE.

Methods: We retrospectively analysed the epidemiology, potential triggers, prodromes, clinical data, ophthalmological study, extraocular manifestations, treatment and outcomes of 79 patients collected through an extensive review of the literature from the first description of Gass JD up to the present time.

Results: A total of 79 patients were reviewed (47 male). Mean age at diagnosis was 30 years (with a range of 8 to 58 years old). 27 patients (34.2%) presented with a previous triggering being flu-like illness the most frequent.¹⁵ However, the complete serological study was only requested in 28 patients (and the immunological study just in 22) Median time from trigger to overt APMPPE was 9 days. Main clinical symptoms were: decreased visual acuity/blurred vision (100%), headache (51.8%) and photophobia (12.2%). Average decreased visual acuity was 13/20. APMPPE is defined by the presence of multiple white-yellowish plaques in funduscopy, and early hypofluorescent areas with late hyperfluorescence in fluorescein angiography. The fundus was pathological and compatible with APMPPE in all cases (100%), as was fluorescein angiography in those that had been performed (59). CNS involvement appeared in up to 50.6% (40 patients). The CNS manifestations were divided into language disorders (11 patients), motor deficit,²⁰ sensory deficit²⁰ and other CNS manifestations.²³ The mean time from visual deficit to neurological manifestations was approximately 2 weeks. Cerebrospinal fluid was studied in 37 patients, with a predominance of lymphocytic pleocytosis (mean of 46 cells/mm3) and elevated proteins (mean of 111 mg/dl). Within the neuroimaging studies carried out (58) up to 69.7% were pathological. 67 patients (84.8%) received treatment with corticosteroids. 14 patients (17.7%) also received other immunosuppressants (mainly azathioprine and cyclophosphamide), especially if CNS involvement. Regarding the evolution, 55 patients (69.6%) presented improvement, 12 (15.2%) relapsed and 6 (7.5%) died due to APMPPE.

Conclusions: APMPPE is a rare inflammatory disease which primarily affects the retina. However, the CNS involvement could be more frequent than what is classically described. Also, it seems that there might be a trigger effect either inflammatory or infectious. Steroids and immunosuppressants should be considered in patients with CNS involvement from the beginning.

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AB1146

THE ASSOCIATION OF COMMON MEFV GENE MUTATIONS WITH AXIAL SPONDYLARTHROSIS IN FMF PATIENTS: A RETROSPECTIVE STUDY

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Background: Familial Mediterranean fever (FMF) is an autoinflammatory disease seen with autosomal recessive inheritance and is characterised by recurrent and self-limiting attacks with peritonitis, pleuritis, arthritis or fever alone. The association of spondylarthrosis and FMF is reported in some studies. There are few studies evaluating the association of MEFV gene mutations with axial spondylarthrosis in FMF patients.

Objectives: The aim of this study is to identify patients with FMF associated spondylarthrosis retrospectively and to compare the frequency of common MEFV gene mutations in FMF patients with and without axial spondylarthrosis.

Methods: We have reviewed 138 charts of FMF patients. The data of 116 patients (70 female, 46 male) who met the diagnosis of FMF with Tel Hashomer classification criteria and have the results of MEFV gene mutation were examined. Patients' age, sex, MEFV gene mutations were recorded. The presence of

inflammatory back pain (IBP) obtained from rheumatological system questionnaire in the patients' charts were recorded. The x-ray and MR images of sacroiliac joint registered in the PACS system were evaluated by a Rheumatologist. Patients with inflammatory low back pain and sacroiliitis detected by X-ray or MR were included in the FMF with axial spondylarthritis (FMF-SPA) group and others to FMF group. The frequency of MEFV gene mutations were compared between two groups.

Results: The frequency of M694V, M680I, V726A, E148Q and R202Q were 82.4%, 19.6%, 16.7%, 10.8% and 16.7% respectively in FMF group and 78.6%, 21.4%, 14.3%, 21.4% and 28.6% respectively in FMF-AS group. There was no significant difference between groups ($p=0.716$, $p=1.000$, $p=1.000$, $p=0.373$ and $p=0.279$ respectively). The frequency of M694V homozygotes and heterozygotes mutations were 27.5% and 21.6% respectively in FMF group and 35.7% and 7.1% respectively in FMF-SPA group and difference between groups was not significant ($p=0.537$, $p=0.296$ respectively).

Abstract AB1146 – Table 1. The characteristics and MEFV gene mutations of FMF patient with spondylarthritis

	Age, years	Sex	IBP	Sacroiliitis X-ray/MR	MEFV mutations
1	34	F	+	-/+	M694V/M680I
2	41	F	+	-/+	M694V/M694V
3	31	F	+	+/-ND	E148Q/R202Q
4	56	M	+	+/-ND	M680I/E148Q
5	49	M	+	+/-ND	M694V/-
6	47	M	+	+/-ND	M694V/E148Q
7	22	M	+	+/-ND	M694V/M680I
8	22	F	+	+/-ND	M694V/M694V
9	39	F	+	+/-ND	M694V/M694V
10	30	M	+	+/-ND	M694V/M694V
11	37	M	+	+/-ND	M694V/M694V/R202Q/R202Q
12	41	F	+	+/-ND	M694V/R202Q/A744S
13	24	F	+	-/+	V726A/K695R
14	31	F	+	+/-ND	M694V/V726A/R202Q

ND: Not done, IBD: inflammatory back pain

Conclusions: The frequency of common MEFV gene mutations in this study is not different between FMF patients with and without axial spondylarthritis. Increased frequency of axial spondylarthritis in FMF patients may not be associated with MEFV gene mutations.

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AB1147 EVOLUTIONARY STUDY OF 45 CASES OF UNDIFFERENTIATED NEGATIVE HLA B27 SERONEGATIVE OLIGOARTHRITIS

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Background: The prognosis of patients with undifferentiated arthritis may vary from self-limited to severe destructive rheumatoid arthritis. Early diagnosis is important, specially in seronegative oligoarthritis in order to start a treatment as early as possible.

Objectives: To describe the evolution of patients older than 16 years diagnosed with negative HLA B27 seronegative oligoarthritis without axial involvement.

Methods: We retrospectively studied 45 patients (23 women, 22 men) with negative HLA B27 seronegative oligoarthritis without axial involvement who debuted between 1985 and 1990 and who did not meet the criteria for any of the rheumatic diseases at the time of debut: rheumatoid arthritis (AR), psoriatic arthropathy (PsA), spondyloarthropathy, enteropathic arthritis, reactive arthritis, microcrystalline arthritis or connective tissue disease.

Results: The mean age at onset of oligoarthritis was 42.2 years (range 17–66). The mean follow-up time was 13.7 years (range 1–32). In its evolution, a definitive diagnosis was reached in 21 (46.6%) patients, with the mean time between debut and diagnosis being 5.47 years (range 1–25): 8 AR, 4 APs (3 with involvement) peripheral and 1 mixed), 2 undifferentiated spondyloarthritis, enteropathic arthritis, 5 gouty arthropathies and one SLE. In the case of RA, the diagnosis was made on an average of 4.8 years after the debut (range 1–16); the RF was positive in 4 patients a mean of 7.6 years (range 3–11) after the debut, and the anti-CCP were positive in 3 of the patients with positive RF. Within PsA, one developed skin psoriasis, another psoriatic onychopathy at 4 years after debut and 2 continue without skin involvement but with a family history of psoriasis, all met CASPAR criteria. From the other 24 patients (53.3%), only 3 patients (12.5%) continued to be followed up, with an average of 21.3 years (range 18–26) without meeting the criteria that allow us to define diagnosis. With the rest of the patients (40.8%), followed for an average of 4.5 years, a diagnosis was not achieved by resolution of the clinical picture or loss of follow-up.

Conclusions: In our series, 46.6% of the patients with a diagnosis of negative HLA B27 seronegative oligoarthritis began to meet diagnostic criteria for rheumatic disease after a mean time of 5.47 years, with RA being the most frequent diagnosis (38%) after an average of 4.8 years after the arthritis onset.

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AB1148 BIOLOGICAL TREATMENT OF NON ISCHAEMIC OPTIC NEURITIS ASSOCIATED TO IMMUNE-MEDIATED INFLAMMATORY DISEASES. MULTICENTER STUDY

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Background: Non ischaemic optic neuritis (NION) is a severe inflammation of the optic nerve that may lead to blindness. It can be primary or associated to immune-mediated inflammatory diseases (IMIDs). The treatment of the NION is based on systemic corticosteroids and conventional immunosuppressive drugs.

Objectives: To assess the efficacy of the biological treatment in refractory NION to conventional treatment.

Methods: Multicenter study of 8 patients diagnosed with NION refractory to systemic corticosteroids and at least one conventional immunosuppressive drug.

The main outcomes were visual acuity (VA) and OCT of the optic nerve and the ganglionar cells. Comparisons were made between baseline and the 1st week, 1st and 6th month and 1st year. (STATISTICA, StatSoft Inc. Tulsa, Oklahoma, USA).

Results: We studied 8 patients (12 affected eyes) (4♀/4♂); mean age of 34.37 ±13.30 years. The underlying diseases were SLE (n=1), neuromyelitis optica (n=1), neuroretinitis (n=1), relapsing polychondritis (n=1), idiopathic (n=2) and Behçet's disease (n=2).

Before biological treatment and besides oral corticosteroids patients had received intravenous (IV) methylprednisolone boluses (n=6), cyclosporine A (CyA) (n=1), cyclophosphamide (n=2), micophenolate (n=2), hydroxychloroquine (n=1), methotrexate (MTX) (n=4) and azathioprine (AZA) (n=2).

Biological treatment was based on rituximab (n=2) (2 IV. doses of 1 g/very 2 weeks and every 6 months), adalimumab (n=2) 40 mg/week, tocilizumab (n=2) 8 mg/kg/2–4 weeks and infliximab (n=2) 5 mg/kg at 0, 2 and 6 week and then every 8 weeks.

The characteristics of the 8 patients are shown in the **TABLE**

After biological treatment we observed an improvement in the ocular parameters: VA [0.60±0.33 to 0.76±0.41, $p: 0.04$], OCT of the optic nerve [130.63±60.54 to 102.60±18.17, $p: 0.1$] and OCT of the ganglionar cells [404.60±184.73 to 243 ±18.38, $p: 0.17$] at one year. After a mean follow-up of 27±14.47 months there were no severe adverse effects.