ventricular arrhythmias and 1 was found to have incidental abnormal myocardial uptake on PET imaging. 8 of the 9 patients had abnormal myocardial uptake on PET imaging. All but 1 patient had been initially treated with oral steroids (1 refused) and 7 of the 9 patients had also been given oral DMARDs; methotrexate (n=6), azathioprine (n=2), hydroxychloroquine (n=2) and mycophenolate mofetil (n=1). Biologics used were adalimumab (n=5), infliximab (n=3) and rituximab (n=1). The most common indication for biologics was progression of disease despite optimal doses of standard therapy, followed by intolerance or contraindication to standard therapy. 75% of the patients were noted to have marked clinical improvement with the addition of a biologic. 4 out of 9 patients had decreased myocardial uptake on PET following treatment with a biologic. One patient had no change on PET and 4 have not had repeat imaging done yet. None of the patients had worsening of left ventricular systolic function with the addition of a TNF alpha antagonist. There were no reported major infections or significant adverse events that were attributable to the use of biologics.

Conclusion: Based on our small cohort, biologics (mainly TNF alpha antagonists) appear to be safe and efficacious as salvage therapy for cardiac sarcoidosis. However, there is a need for prospective studies to further validate these findings as well as to identify the subset of patients that would benefit from early initiation of these therapies.

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FRI0610

CRYOPYRIN-ASSOCIATED PERIODIC SYNDROME: A SERIE OF CASES IN A ADULT'S PATIENTS COHORT WITH AUTOINFLAMMATORY SYNDROME IN A THIRD LEVEL HOSPITAL

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Background: The Cryopyrin-associated Periodic Syndrome (CAPS) or cryopyrinopathies are part of the spectrum of autoinflammatory diseases. The term cryopyrinopathy encompasses three clinical entities such as: CINCA/NOMID (chronic infantile neurologic, cutaneous and articular/neonatal onset multisystem inflammatory disease), Muckle-Wells disease and FCAS (familial cold-associated periodic syndrome). There are three different diseases that would be part of the same clinical spectrum, ranging from CINCA/NOMID (more serious) to FCAS (the less severe). It is more frequent in pediatric age although they can also be observed in adulthood. They are characterized by mutations in the NLRP3 gene and have an autosomal dominant inheritance pattern.

Objectives: To describe the clinical characteristics and genetic variants of a cohort of patients in adulthood diagnosed with cryopyrinopathy and with follow-up in a 3rd level hospital.

Methods: Retrospective descriptive study in adults patients with diagnosis of cryopyrinopathy since 2013 (year of introduction of genetic tests in the hospital laboratory) until now. The data was obtained from the review of medical records. All patients with mutations in NLRP3 gene and clinically compatible with this diagnosis were reviewed.

Results: Of a total of 44 patients in adulthood diagnosed with periodic fever syndromes (FMF, TRAPS, cryopyrinopathies, HIDS) and compatible genetic mutations, 7 patients (15.9%) were diagnosed with cryopyrinopathies, presenting 6 of them (13, 6%) mutations in NLRP3 gene. 1 patient was diagnosed with Muckle Wells based on clinical criteria without genetic test. 6 patients (13.6%) were women. 4 patients (9.1%) presented mutations in heterozygosis in exon 3 of NLRP3 gene (p.V198M). 1 patient presented heterozygous mutation in exon 3 of NLRP3 gene (p.R260W) and another patient presented a mutation in heterozygosis in exon 3 of the NLRP3 gene (p.S726G). The mean age at diagnosis was 38 years (IR 13-71 years). The totality of patients diagnosed with cryopyrinopathy showed elevation of acute phase reactants. 6 patients (13.6%) presented fever, joint symptoms (arthralgias and/or arthritis) and myalgias. 4 patients (9.09%) showed cutaneous involvement in the form of urticarial rash. 4 patients (9.09%) showed neurosensory deafness since childhood. 3 patients (6.8%) presented ocular involvement in form of conjunctivitis and/or uveitis. In 3 cases (6.8%), antiIL-1 (anakinra) or anti-TNF were used.

Conclusion: Cryopyrinopathies are autoinflammatory diseases that occur mainly in the pediatric age, but there are also cases in adulthood. We must consider its diagnosis in those cases with periodic fever, arthralgia or arthritis, sensorineural deafness, APR elevation and urticarial rash. The genetic diagnosis will help us to confirm the diagnosis and avoid a delay in it.

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FRI0611

ACUTE AND LONG-TERM EFFICACY OF IL-1 INHIBITOR ANAKINRA IN IDIOPATHIC AND SECONDARY PERICARDITIS IN PATIENTS REFRACTORY OR INTOLERANT TO CONVENTIONAL THERAPY

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Background: In contrast to its use in idiopathic recurrent pericarditis (IRP), efficacy and optimal treatment duration of Anakinra are unclear in pericarditis secondary to known autoimmune or inflammatory conditions.

Objectives: a) Assess the efficacy of Anakinra in secondary and idiopathic pericarditis, refractory or intolerant to conventional therapy (CT; colchicine and corticosteroid [CS]).

b) Study the effect of extended Anakinra continuation on post-treatment relapse risk.

Methods: Retrospective chart review of 12 adults hospitalised between January 2016 and October 2018 treated with Anakinra 100mg SQ daily for pericarditis refractory or intolerant to CT (CT+A group), and 22 consecutive hospitalised patients treated with CT only (CT group).

Results: Diagnosed autoimmune or inflammatory condition was present in 83.3% (10/12) of CT+A and 40.9% (9/22) of CT group (p=0.07, Table 1). The most common etiologies in CT+A group were RA and Undifferentiated Connective Tissue/Overlap Disease (Figure 1).

Despite CT-intolerance or resistance (Figure 2), all CT+A (12/12) patients had symptom relief at discharge vs. 72.7% (16/22) in CT group (p=0.04). There was a strong trend in CT+A compared to CT group towards shorter time to symptom improvement (1.75±1.29 vs. 3.55±3.06 days; p=0.06) and symptom relief (3.75±1.87 vs. 5.63±3.28 days; p=0.08). CS were tapered successfully in 71.4% (5/7) vs. 66.7% (6/9) of patients in the CT+A and CT groups (p=0.56). Anakinra was associated with a longer mean length of stay (13.0±16.2 vs. 8.68±5.07 days; p=0.25).

No recurrence during treatment was noted in CT+A group (0/12) vs. 40.9% (9/22) in CT group (p=0.009). There was a trend towards reduced post-treatment relapse risk in CT+A group [16.7% (2/12) vs. 41.7% (5/12), p=0.18]. In relapsed patients, there was no significant difference in time to relapse (1.75±0.34 vs. 1.72±0.71 months, p=0.93). Within CT+A group, extended Anakinra use in 6 patients (mean: 5.58±6.5 months) did not reduce post-treatment relapse risk [16.7% (1/6) vs. 16.7% (1/6), p>0.9]. Adverse effects were infrequent; elevated transaminases (n=1) and local injection-site reaction (n=1). Neither required permanent Anakinra discontinuation.

Table 1. Baseline Characteristics

	Anakinra plus conventional therapy % (n=12)	Conventional therapy only % (n=22)	p- value
Age (years +/- SD)	60.3±17.3	56.8±17.8	0.590
Female	66.7 (8)	50 (11)	0.349
Diagnosed autoimmune or inflammatory condition	83.3 (10)	40.9 (9)	0.074
Recurrent pericarditis (>1 episode)	33.3 (4)	27.3 (6)	0.710
Time since initial episode (months +/-SD)	49.0±79.9	9.21±11.7	0.441
Prior NSAID use	25.0 (3)	18.2 (4)	0.638
Prior Colchicine use	25.0 (3)	9.09 (2)	0.210
Prior CS use	50.0 (6)	18.2 (4)	0.051
Pericardial effusion on ECHO	75.0 (9)	81.8 (18)	0.638
Treatment during hospitalisation			
NSAID	50.0 (6)	54.5 (12)	0.799
Colchicine	58.3 (7)	63.6 (14)	0.761
CS	58.3 (7)	40.9 (9)	0.331
Risk factors for pericarditis			
PCI, cardiac surgery or chest trauma	16.7 (2)	45.5 (10)	0.093
Chest radiation	8.33 (1)	4.55 (1)	0.653

Malignancy	0 (0)	0 (0)	
Pericardial drainage	25.0 (3)	13.6 (3)	0.406
GFR<30 ml/min/1.73m ²	8.33 (1)	13.6 (3)	0.133
Family history of IRP	8.33 (1)	4.55 (1)	0.653
CRP, mg/L (mean+/-SD)	136+/-115	95.9+/-87.7	0.309
DM	8.33 (1)	27.3 (6)	0.191
HTN	25.0 (3)	63.6 (14)	0.031

Conclusion: In hospitalised patients with new-onset or recurrent pericarditis (secondary or idiopathic) refractory or intolerant to CT, Anakinra is associated with improved symptom relief and decreased recurrence risk. Extended continuation of Anakinra after symptom relief does not reduce post-treatment relapse risk.

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FRI0612 IMMUNE-RELATED ADVERSE EVENTS ASSOCIATED WITH IMMUNOTHERAPY. LONG-TERM FOLLOW-UP OF 102 PATIENTS FROM A REFERRAL SINGLE CENTER

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Background: Immune checkpoint blockade therapy (ICBT) has shown remarkable benefit in different cancer types. Blockade of intrinsic down-regulators of immunity increases the activity of the immune system, which can lead to different immune-related adverse events (irAEs). Our aim was to assess the immune-related adverse events in patients who received anti-cytotoxic T-lymphocyte antigen 4 (CTLA-4), anti-programmed cell death 1 (PD-1) or anti-programmed cell death ligand 1 (PD-L1).

Objectives: Our aim was to assess the incidence, treatment and evolution of irAEs due to ICBT.

Methods: We set up an observational study of patients treated with Nivolumab and Pembrolizumab (anti-PD1), Atezolizumab (anti-PD-L1) and Ipilimumab (antiCTLA-4) for solid organ tumors. All these patients were

followed in a single reference University Hospital from March-2015 up to December-2018. The main outcome was to determinate the incidence of irAEs.

Results: We studied 102 patients (63♂/39♀) with a mean age of 60.6 ±9.7 with lung (n=63), melanoma (21), kidney (11), gastric (3), bladder (1), and colon cancer (n=3). Only 7 patients had a previous diagnosis of an immune-mediated disease: psoriasis (n=2), psoriatic arthritis (1), systemic lupus erythematosus (1), spondyloarthritis (1), rheumatoid arthritis (1) and skin lupus (1). ICBT was performed as follows: nivolumab (n=52), pembrolizumab (35), atezolizumab (10) and ipilimumab (5). After a median of 5 [2.5-10.5] months since the ICBT onset, we observed 87 (85.3%) patients with different irAEs, summarized in **TABLE**. ICBT discontinuation was required in 39 patients. 36 patients received specific treatment (prednisone, antihistamine, levothyroxine and thiamazol), obtaining a good response in 31 cases (79.5%). ICBT was reintroduced in 28 patients (71.8%) after resolution of the adverse event, with an appropriate tolerance in all cases.

Immune-related adverse event	Anti CTLA-4 n= 5	Anti PD1-/PD-L1 n= 97	TOTAL n=102
Gastrointestinal, n (%) (diarrhoea, colitis, mucositis)	1 (1.1)	38 (40)	39 (41.1)
Skin, n (%) (rash, erythema nodosum, psoriasis, vitiligo and alopecia)	1 (1.1)	12 (12.6)	13 (13.7)
Thyroid, n (%) (thyroiditis)	0 (0)	18 (18.9)	18 (18.9)
Articular, n (%) (arthralgia, arthritis)	0 (0)	14 (13.7)	14 (13.7)
Vasculitis, n (%) (aortitis)	0 (0)	1 (1.1)	1 (1.1)
LFTs alteration, n (%)	0 (0)	8 (8.4)	8 (8.4)
Nephritis, n (%)	0 (0)	6 (6.3)	6 (6.3)
TOTAL	2 (2.1)	97 (97.9)	99 (100)

+LFT = Liver function tests.

Gastrointestinal irAEs were the most frequent (n=39, 41.1%), with severe manifestations in 4.9% of the patients. Among these 39 patients, only 11 of them required specific treatment, with oral prednisone in all cases. The diagnosis of gastrointestinal irAEs was based on the clinical manifestations and the laboratory tests. The cutaneous most frequent irAE was rash (n=7, 6.8%), followed by vitiligo (n=2, 1.9%). 15 patients developed a rheumatological irAE (14 arthralgias/arthritis and 1 aortitis). Patients who developed any type of irAE had a better answer to ICBT (70.1%) than those who did not (20%).

Conclusion: In our study, the majority of autoimmune side effects due to ICBT were gastrointestinal, thyroiditis and rheumatological. The develop of any irAE could be an indicator of good response to ICBT.

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