reached disease remission by physicians' assessment along with rates of 40-61% absent disease activity in PGA. Patients reported stable low level disease activity, fatigue and Auto-Inflammatory Diseases Activity Index scores (AIDAI, figure 1). CAPS was impairing social life in 50% of patients and another 50% reported days off from school/work. Lab parameters were within normal limits.

Table 1. Patient and physician assessment of clinical CAPS disease activity and laboratory markers over time.

	Baseline		12 months		30 months	
	Total cohort	NOMID/CINCA	Total cohort	NOMID/CINCA	Total cohort	NOMID/CINCA
Number of patients, N	91	14	67	8	28	4
Number (%) of patients with days absent from work/school during last 6 months	30 (34)	4 (29)	28 (42)	2 (25)	17 (61)	4 (100)
Number (%) of patients in disease remission (physician assessment)	61 (68.5)	11 (78.6)	42 (66.7)	4 (66.7)	19 (67.9)	4 (100.0)
Physician Global Assessment, percentage of absent/mild-moderate/severe rating, %	40 / 53 / 2	57 / 36 / 0	33 / 60 / 2	33 / 50 / 0	61 / 39 / 0	75 / 25 / 0
Patient assessment of current disease activity; 0-10, median (min; max)	2.0 (0; 7)	1.0 (0; 6)	1.0 (0; 7)	1.0 (0; 5)	0.0 (0; 7)	0.0 (0; 4)
Patient assessment of current fatigue; 0-10, median (min; max)	3.0 (0; 9)	2.0 (0; 6)	3.0 (0; 8)	2.0 (0; 8)	1.0 (0; 8)	4.0 (0; 5)
Number (%) of patients without impairment of social life by the disease	32 (52.5)	4 (50.0)	31 (62.0)	3 (42.9)	11 (47.8)	1 (33.3)
CRP, median (mg/dl)	0.1	0.2	0.1	0.5	0.0	0.2
SAA, median (mg/dl)	0.3	0.4	0.5	0.9	0.3	0.1
ESR, median (mm/h)	5.0	6.0	5.0	3.5	5.0	5.5
SAE	Number of events		Incidence rate per 100 patient years			
	Total cohort	NOMID/CINCA	Total cohort	NOMID/CINCA		
All types of SAE	39	3	14.72	11.72		
SADR	20 ^a	0	10.69	0.00		

^a Alport's syndrome, appendicitis, blister, cardiovascular disorder, chest pain, circulatory collapse, erythema, febrile convulsion, glomerulonephritis, haemophilus test positive, pneumonia, premature delivery, skin discolouration, tonsillectomy, tonsillitis bacterial, tonsillitis streptococcal (all N=1 event), pyrexia (N=3 events), not yet coded (inpatient admission, N=1 event) CRP, c-reactive protein; ESR, erythrocyte sedimentation rate; n. a., not annotated; SAA, serum amyloid A; SADR, serious adverse drug reaction; SAE, serious adverse event

Figure 1: Median AIDAI score (0-390) of the CAPS cohort (N = 91 at baseline) over 30 months

Conclusion: The 30-month interim analysis of the RELIANCE study demonstrates that long-term canakinumab treatment is safe and effective in patients with any subtype of CAPS. However, impairment of social life and days off school/work still exists.

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OP0093

RETENTION RATE OF IL-1 INHIBITORS IN PATIENTS WITH SCHNITZLER'S SYNDROME

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Background: Schnitzler's syndrome is an autoinflammatory disease characterized by monoclonal gammopathy and recurrent episodes of urticaria accompanied by clinical and laboratory signs of acute inflammation. Interleukin (IL)-1 inhibitors proved to be useful in the treatment, but data on long-term safety and efficacy of these agents are sparse.

Objectives: To evaluate the retention rate of IL-1 inhibitors in patients with Schnitzler's Syndrome.

Methods: Retrospective analysis of an Italian multicenter cohort (9 Centers). All patients fulfilled Strasbourg diagnostic criteria. Data are expressed as median [IQR].

Results: We identified 15 patients (8 females, 7 males) who received a total of 24 treatment courses with IL-1 inhibitor treatment (16 anakinra and 8 canakinumab) between January 2001 and December 2019, with a median treatment duration of 19 months [8.5-51.3]. Median age at diagnosis was 64.0 years [56.0-72.5] and median follow up was 5.0 years [2.0-8.0]. Before the biological treatment, all patients were treated with corticosteroids and 11 with at least one conventional synthetic disease-modifying antirheumatic drug (csDMARD): methotrexate (5), colchicine (5), cyclosporine (3), azathioprine (1), mycophenolate mofetil (1), cyclophosphamide (1).

Fifteen patients received 16 courses of Anakinra, which was the 1st line biological treatment in 14 patients. Seven patients continued it with benefit, while 7 patients discontinued it: 3 for secondary inefficacy; 3 for adverse events (2 injection site reactions, 1 severe allergic reaction); 1 for secondary inefficacy and leukopenia. Anakinra was used as 2nd line treatment in 1 case (after tocilizumab failure); in 1 patient anakinra was resumed after temporary discontinuation and an attempt with infliximab. One patient died for multiple myeloma progression while on treatment with anakinra. The median duration of the courses with anakinra was 20.0 months [6.0-58.3].

Seven patients received 8 courses of canakinumab (150 mg/8weeks in 5 cases and 150 mg/4weeks in 3). In 5 cases the drug was administered as 2nd line biological treatment (after anakinra failure) and in 2 cases as 3rd line treatment (1 after tocilizumab and anakinra failures and 1 after anakinra and adalimumab failure). In 1 patient, it was resumed after temporary discontinuation and an attempt with etanercept. One patient died while on treatment with canakinumab due to a presumably unrelated adverse event. The median duration of canakinumab treatment courses was 19.0 months [13.5-31.0].

At last follow-up visit, all patients were on treatment with an IL-1 inhibitor: 8 with anakinra (7 at the dosage of 100 mg/day, 1 at the dosage of 200 mg/day) and 7 with canakinumab (2 at the dosage of 150 mg/8 weeks, 4 at the dosage of 150 mg/4 weeks and 1 at the dosage of 300 mg/4 weeks). Notably, in 3 patients the dosage of canakinumab was increased since the start of the treatment.

Among 9 patients who were on treatment with prednisone at the start of the last IL-1 inhibitor, the prednisone median dose was 12.5 mg/day [10.0-18.8] while at the last follow-up visit it was 5.0 mg/day [0-7.5] ($p=0.02$). The retention rate of IL-1 inhibitors was 73.4% [SE 9.4] at 1 year and 63.6% [SE 10.4] at 2 years (Figure 1a). There was no significant difference between the retention rate of anakinra (at 1 year: 67.0% [12.2]; at 2 years: 59.6% [12.9]) and canakinumab (at 1 year: 85.7% [13.2]; at 2 years 71.4% [17.1]) (log-rank test: $p=0.41$) (Figure 1b).

Figure 1. a) Retention rate of IL-1 inhibitors (24 courses); b) Retention rate of canakinumab (8 courses) and anakinra (16 courses).

Conclusion: In this multicentric cohort of patients affected by Schnitzler's syndrome, the treatment with IL-1 inhibitors as 1st, 2nd or 3rd line biological treatment permitted a good disease control and corticosteroid reduction in patients who did not respond to csDMARDs and/or to prior other biological DMARDs. The optimal dosage of these drugs needs to be tailored for every patient.

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OP0094

PULMONARY ARTERIAL HYPERTENSION IN ADULT-ONSET STILL'S DISEASE: A CASE SERIES OF 13 PATIENTS

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Background: Pulmonary Arterial Hypertension (PAH) is a rare but potentially fatal complication of Adult-Onset Still's Disease (AOSD) (1). To date, only isolated observations have been published.

Objectives: To establish the largest case series of AOSD patients with PAH, and to describe their clinical profile, evolution and response to treatments.

Methods: Cases were retrospectively identified from the French PAH network database and from an online call of the "Club Rhumatismes et Inflammation" (<http://www.cri-net.com>). To be included, all patients had to fulfil the Yamaguchi or Fautrel's criteria for AOSD and PAH had to be confirmed by right heart catheterization. The data were collected using a standardized questionnaire.

Results: Thirteen patients were identified. All were female, the mean age at PAH diagnosis was 32±12 years, 2 (15%) patients were Caucasian, 6 (46%) from

Sub-Saharan Africa, 1 (8%) from Asia and 4 (31%) from West Indies. Only 2 (15%) patients were smokers. All patients had a systemic onset of AOSD, 12 had a polycyclic and 1 a chronic articular evolution, and the mean delay between AOSD and PAH diagnosis was 2.9 (range 1.7-5.4) years. At PAH diagnosis, patients were receiving the following treatments: 13 (100%) corticosteroids (median dose 12 mg [interquartile range (IQR) 9-18]), 3 (23%) methotrexate, 8 (61%) interleukin (IL)-1 inhibitors (exposure median duration 6.7 months [IQR 3.6-8.5]), none IL-6 inhibitors, 2 (15%) TNF inhibitors. Six (46%) patients developed PAH during an AOSD flare. PAH was severe at diagnosis: 2 (15%), 7 (54%) and 4 (31%) patients were in NYHA functional class II, III and IV, respectively, with a median 6-minute walk distance of 289 m [IQR 0-448], a mean pulmonary arterial pressure of 41±12 mmHg, a mean pulmonary arterial occlusion pressure of 6±3 mmHg, a mean cardiac output of 3.9±1.2 L/min, a mean cardiac index of 2.5±0.9 L/min/m² and a median pulmonary vascular resistance of 7 Wood Units [IQR 6-11]. The treatment prescribed after PAH diagnosis is detailed in the table. The median follow-up was 34 months [IQR 7-42]. Five patients (38.5%) died. Figure 1 shows the overall survival. The haemodynamic response to PAH treatment seemed to be dissociated from the prognosis since several patients have died while their haemodynamic had improved or almost normalised.

Conclusion: PAH is a rare but potentially severe complication of AOSD, leading to death in 38.5% of our cases series. AOSD remission should be physicians' objective, since PAH seems to occur when the underlying disease is not controlled.

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Table 1. Therapeutic management

Treatment	n (%)
Inotropic therapy	5 (38%)
HTAP treatment	10 (77%)
• 0 Monotherapy	3
• 0 Initial oral dual combination therapy	3
• n Dual combination therapy including intravenous (IV) prostacyclin	1
• u Upfront triple combination therapy including IV prostacyclin	3
High-dose corticosteroids	9 (69%)
Interleukin 1 inhibitors initiation	2 (15%)
Interleukin 6 inhibitors initiation	5 (38%)

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Figure. Overall survival

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OP0095

NON-GONOCOCCAL PYOGENIC ARTHRITIS OF NATIVE JOINTS IN WESTERN AUSTRALIA. A LONGITUDINAL POPULATION-BASED STUDY OF FREQUENCY, RISK FACTORS AND OUTCOME.

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Background: The worldwide incidence of PyA is reportedly rising due to a combination of increased longevity, multi-comorbidity, iatrogenic complications and increasing use of immunomodulating therapies, while there is limited data on longterm outcomes of PyA.

Objectives: To describe the recent incidence, risk factors and long-term outcomes in adults hospitalised with non-gonococcal pyogenic arthritis (PyA) of native joints in Western Australia (WA).