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).