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Background: Tenosynovitis can occur in patients with Juvenile Idiopathic Arthritis (JIA) and may be clinically difficult to distinguish from joint synovitis. The role of musculoskeletal ultrasound (msk-US) is still discussed in the management of JIA but recent studies supported the utility of msk-US especially in detection of tenosynovitis. There is no consensus treatment for tenosynovitis in children with JIA and almost all studies focused on tendon sheath injection with glucocorticoids.

Objectives: The aims of the study were: i) to define the prevalence of tenosynovitis of the ankle in JIA patients and ankle swelling; ii) to describe the clinical characteristics of patient with tenosynovitis and to analyze different response to treatment.

Methods: We conducted an observational cross-sectional study of a group of patients with JIA followed at the Rheumatology Service of the Maternal and Child Institute "Burlo Garofolo" of Trieste. We enrolled all the patients who reported a swelling of the ankle at least once during the follow-up period and, among these patients, we included only those who underwent msk-US. Based on both clinical and sonographic examination, we identified patients with tenosynovitis and we described their demographical and clinical characteristics as well as the therapeutic approach undertaken in this group of patients.

Results: On December 31st 2019, 56 swollen ankles of 48 patients were assessed with msk-US: 22 ankles showed sonographic signs of synovitis (39%), 16 ankles of both synovitis and tenosynovitis (28%), 14 ankles of tenosynovitis only (25%). Overall, tenosynovitis was detected on twenty-seven (56%) out of 48 children with at least a swollen ankle. The majority of patient were females (70%) and the most affected tendon was the tibialis posterior (66%). Twenty-five patients with tenosynovitis (92%) achieved clinical and radiological remission: seven out of 26 patients (26%) treated with methotrexate achieved clinical and radiological remission without the addition of other therapies; fifteen out of seventeen patients (88%) treated with a biological drug responded to the therapy, of which eleven (73%) were in combination therapy with methotrexate.

Conclusion: We observed that more than 50% of the patients with a swelling of the ankle presented a tenosynovitis and among these patients about 50% did not show sonographic sign of synovitis. Msk-US was decisive in order to identify tenosynovitis and to characterize ankle swelling in JIA patients. Among patients with tenosynovitis biological therapy alone or in association with immunomodulating therapy showed effectiveness in inducing disease remission.

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POS1334 RHEUMATOLOGY PATIENT ATTENDANCE TO THE PAEDIATRIC EMERGENCY DEPARTMENT

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Background: A large proportion of patients present to the paediatric emergency department (PED) with non-urgent problems [1]. With growing pressures on services, there is a need to reduce unnecessary attendances. Children with rheumatological conditions may attend PED with flares of their condition or unrelated presentations. These patients may be immunosuppressed due to steroid, disease-modifying or biologic therapy; PED attendance may therefore pose a serious infection risk.

Objectives: To evaluate why children with rheumatological conditions attend the PED and whether these attendances are avoidable.

Methods: This study was run in Royal Manchester Children's Hospital, a UK hospital with a PED and tertiary paediatric rheumatology services. Patients under rheumatology who attend PED are usually seen directly. A dataset of all patients referred directly to paediatric rheumatology between 01/01/19 and 31/12/19 was

obtained from electronic records (n=59). The age, sex and reason for attendance were collected, as well whether they were admitted/discharged. Further data on management was collected from discharge letters for those with joint pain/swelling.

Results: This cohort had 36 females and 23 males, with an average age of 11.9 years (range 1-18). 28 were discharged (47.5%) and 30 were admitted (50.8%). One patient left before being seen. The average waiting time was 3.97 hours.

Table 1 summarises reasons for attendance to PED and final outcome. The most common reason for attendance was joint pain/swelling (n=14, 23.7%). Of these, nine were discharged with advice on analgesia or steroid dosing and for early review in clinic. There was no documentation on whether there had been any attempt to access the available rheumatology helpline prior to attending PED. Non-rheumatology-related reasons for attendance are also shown in Table 1, and notably include 20 patients with infections, 75% of whom were admitted to hospital. Chicken pox was the second most common reason for attendance (n=5, 8.5%); all patients attending with chicken pox or shingles (n=6) were admitted.

Table 1. Reasons for attendance to PED and final outcome

Reason for attendance	Number admitted	Number discharged
Joint pain/swelling	5	9
Chicken pox	5	0
Viral URTI	1	4
Rash	0	4
Pneumonia	3	0
SLE flare	3	0
Influenza A	2	0
GPA flare	2	0
Asked to attend due to blood results from clinic	2	0
Drug reaction	0	2
Minor injury	0	2
Shingles	1	0
Oral herpes simplex	1	1
Orbital inflammation	1	0
Urinary tract infection	1	0
Dysphagia	1	0
Gastroenteritis	1	0
Cerebral infarcts	1	0
Constipation	0	1
Chest pain	0	1
Abdominal pain	0	1
Enlarged lymph node	0	1
Picking up prescription	0	1
Unresponsive episode	0	1

Conclusion: Our study shows that attendance to PED with joint pain/swelling is usually avoidable in patients known to paediatric rheumatology services. Appropriate alternative services are needed to reduce hospital attendances; nurse-led helplines are beneficial, but still face some challenges in both availability and accessibility [2]. It is also important that patients and parents are given action plans for acute flares, which would ideally involve early clinic review rather than PED attendance. Our data also showed that patients are spending a long time waiting in PED, which needs to be avoided particularly in the context of immunosuppression. This is especially poignant now in light of the Covid-19 pandemic. Direct referral to rheumatology with non-rheumatological problems likely reflects a desire for specialist advice in the context of complex conditions and medications, for example with regards to infections whilst on immunosuppressive medications, or uncertainty about whether new symptoms are related to the underlying condition. Expansion of helpline services and new approaches such as remote consultations should be explored.

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Other orphan diseases

POS1335 CLINICAL CHARACTERISTICS OF THE PULMONARY ARTERY INVOLVEMENT IN PATIENTS WITH BEHÇET'S SYNDROME: A SINGLE-CENTRE EXPERIENCE OF 61 PATIENTS

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Background: Behçet's Syndrome (BS) is a vasculitis of unknown origin. Vascular involvement, so-called vascular Behçet's syndrome (VBS), may involve blood vessels of all sizes belonging to both venous and arterial system, with pulmonary artery involvement (PAI) being the most frequent form of arterial involvement. PAI in BS occurs in the form of aneurysms or thrombosis, and results in significant mortality.

Objectives: To report the clinical characteristics of PAI in patients with BS and to define the predictors of relapses.

Methods: We performed a retrospective analysis of BS patients with PAI who fulfilled international study group criteria. Among 460 patients with VBS, 66 were diagnosed with PAI. For final analyses, 61 patients with PAI, who had at least 2 follow-up visits (72.1% male, mean age at BS diagnosis 29.34 (SD 10.1) years), were included. The data of the patients were recorded. Relapse was defined as the reoccurrence of vascular event at any vascular structure. Factors associated with relapse were assessed by logistic regression analysis.

Results: There were not any differences considering demographic and clinical features of the patients with and without PAI in VBS group except for that intracardiac thrombosis was more common in the patients with PAI than the patients without (19.7% vs 0.3%, respectively) (Figure 1). Among 61 patients with PAI, 50 (82.0%) had isolated pulmonary artery thrombosis (PAT), whereas 11 (18.0%) had pulmonary artery aneurysm (PAA) with or without PAT. The characteristics of 61 patients with PAI were shown in Table 1. A total of 37 relapses occurred in 24 (39.3%) patients during follow-up a median of 65.9 (IQR 20.1-109.0) months. To define the factors associated with relapses, the patients with isolated PAT were analysed. In multivariable logistic regression analysis, older age at BS diagnosis and anticoagulation usage seemed to be protective, even though they could not reach statistical significance (OR: 0.92 95% CI 0.86-1.02, OR: 0.34 95% CI 0.09-1.33, respectively).

Figure 1. A) Axial CT image shows thrombus formation in the left upper lobe pulmonary artery branch. B) Giant thrombus is seen in the right ventricle (black arrow). Note the thrombus formation in the left lower lobe pulmonary artery branches (white arrows).

Table 1. Characteristics of the patients with pulmonary artery involvement

	All n= 61	Isolated PAT n= 50	PAA n= 11
Sex (male), n (%)	44 (72.1)	35 (70.0)	9 (81.8)
Age at diagnosis of BS, years (SD)	29.3 (10.1)	28.8 (10.4)	31.9 (8.0)
Age at diagnosis of PAI, years (SD)	36.3 (12.83)	36.7 (13.8)	34.7 (6.7)
Symptoms, n (%)			
Dyspnea	28 (45.9)	23 (46.0)	5 (45.5)
Hemoptysis	26 (42.6)	19 (38.0)	7 (63.6)
Vascular event before PAI, n (%)	21 (34.4)	20 (40.0)	1 (9.1)
Other vascular events at PAI, n (%)	33 (54.1)	28 (56.0)	5 (45.5)
Thrombophlebitis, n (%)	7 (21.2)	6 (21.4)	1 (20.0)
Upper and/or lower extremity DVT, n (%)	20 (60.6)	16 (57.1)	4 (80.0)
SVCS, n (%)	1 (3.0)	1 (3.6)	0 (0)
IVCS, n (%)	3 (9.1)	1 (3.6)	2 (40.0)
Intracranial thrombosis, n (%)	4 (12.1)	4 (14.3)	0 (0)
Intracardiac thrombosis, n (%)	8 (24.2)	7 (25.0)	1 (20.0)
Arterial event other than PAI, n (%)	4 (12.1)	3 (10.7)	1 (20.0)
Relapse, n (%)	24 (39.3)	20 (40.0)	4 (36.4)
Time to the first relapse, median (IQR), month	49.8 (13.2-102.6)	54.1 (24.4-117.9)	3.9 (1.7-67.9)
Death, n (%)	2 (3.3)	0 (0)	2 (18.2)

BS= Behçet Syndrome, PAI= pulmonary artery involvement, PAT= pulmonary artery thrombosis, PAA= pulmonary artery aneurysm, DVT= deep vein thrombosis, SVCS= superior vena cava syndrome, IVCS= inferior vena cava syndrome

Conclusion: Our results indicate that there is a higher frequency of intracardiac thrombosis in BS patients with PAI and an increasing current trend of PAT in patients with PAI. In addition, there might be a possible efficacy of anticoagulation usage in preventing relapses, which needs confirmation with further studies.

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POS1336

RETROPERITONEAL FIBROSIS IN ERDHEIM-CHESTER DISEASE HAS UNIQUE PRESENTING AND PROGNOSTIC FEATURES: A SINGLE CENTRE RETROSPECTIVE COMPARATIVE COHORT STUDY

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Background: Erdheim-Chester disease (ECD) is a multisystemic non-Langerhans cell histiocytosis. Retroperitoneal fibrosis (RPF) is a common ECD feature, found in up to 60% of patients. Besides ECD, RPF differential diagnosis includes other clinical entities of rheumatological interest, such as idiopathic RPF and IgG4-related disease (IgG4-RD). Since ECD management does not include glucocorticoids and mainly relies on the use of anti-neoplastic small molecules, a prompt diagnosis is critical.

Objectives: To characterize the main differential clinical, radiologic and prognostic features of a monocentric cohort of patients with RPF. To provide practical learning points useful to recognize ECD among patients with RPF.

Methods: Medical records of ECD patients with radiologic signs of RPF and of patients with non ECD-related RPF followed up at our Rheumatology Clinic were retrospectively reviewed. Final diagnosis relied either on analysis of histologic samples or on fulfilment of appropriate classification criteria. Main clinical, radiologic and laboratory features at diagnosis and long-term outcomes were evaluated and compared between the two groups. Non parametric tests were used for comparison.

Results: 31 (41%) ECD and 45 (59%) non-ECD patients (IgG4-RD, n=28; idiopathic RPF, n=11; peri-aortitis with large-vessel involvement, n=6) were included. All but one (97%) ECD patients and 56% non-ECD patients had a biopsy-proven diagnosis. ECD patients were younger at disease onset (51±14 vs 59±10 years, P=0.024) and diagnosed with a greater delay (48 [IQR, 13-92] vs 4 [IQR, 2-7] months, P<0.001). In the majority of ECD patients (84%), RPF was clinically silent and was disclosed in the context of the diagnostic work-up; conversely, 82% non-ECD patients had at least one RPF-related symptom (mainly, abdominal pain, n=19, and low-back pain, n=16). At diagnosis, C-reactive protein (50±34 vs 33±31 mg/L, P=0.052) and serum creatinine (1.31±0.53 vs 1.34±0.79, P=0.76) levels were not statistically different. In all ECD patients, fibrotic tissue surrounded both kidneys symmetrically, involving the ureters in half of the cases (Table 1); signs of hydronephrosis were present in 55% of patients (mostly bilaterally, 77%). In the non-ECD group, kidneys were rarely affected, and ureteral involvement was mostly asymmetric; hence, hydronephrosis incidence was similar (76%), but more frequently monolateral (65%). In non-ECD patients, fibrotic tissue more frequently involved inferior vena cava and iliac arteries. FDG uptake by RPF was disclosed in 39/40 (98%) non-ECD and 5/12 (42%) ECD patients who underwent a PET before therapy start (P<0.001). Ureteral stenting was required by a similar fraction of patients (ECD, 29%; non-ECD, 38%, P=0.47), but it was bilateral in 100% of ECD vs 29% of non-ECD patients (P<0.001). No differences in the rate of chronic kidney failure was found (26% vs 18%, P=0.41), but only ECD patients progressed to a terminal stage requiring hemodialysis (13% vs 0%, P=0.025).

Table 1.

	ECD patients (n=31)	Non-ECD patients (n=45)	P-value
Left kidney	30 (97%)	4 (9%)	<0.001
Right kidney	31 (100%)	6 (13%)	<0.001
Left ureter	16 (52%)	16 (36%)	0.24
Right ureter	16 (52%)	24 (53%)	1
Abdominal aorta	21 (68%)	40 (89%)	0.04
Inferior vena cava	5 (16%)	29 (64%)	<0.001
Left iliac artery	10 (32%)	27 (60%)	0.02
Right iliac artery	10 (32%)	29 (64%)	0.001
Left renal artery	9 (29%)	6 (13%)	0.14
Right renal artery	6 (19%)	4 (9%)	0.3
Mesenteric artery	8 (26%)	4 (9%)	0.06
Celiac trunk	7 (23%)	3 (7%)	0.08

Conclusion: Our retrospective study suggests that ECD-related RPF has a significantly worse renal prognosis than non ECD-related RPF. A complete baseline work-up including a critical evaluation of the extension of the fibrotic tissue and the sites involved is fundamental to avoid patients' misdiagnosis and mistreatment.

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