standing, and lasts longer than six months. Pain is associated with dyspareunia and varicosities in lower limbs (LL) and genitals. It is challenging and important to consider atypical clinical presentations that simulate osteoarticular (OA) pathologies. It is usually diagnosed by Angio-CT, and a safe, definitive and successful treatment is the embolization of the affected vein.

Objectives: The objective of this study was to evaluate the characteristics of those patients diagnosed with PCS in our University hospital from January 2014 to May 2018, paying close attention to the atypical forms of presentation that simulate OA pathology.

Methods: We included all patients from our center who were operated by embolization due to a PCS from January 2014 to May 2018. Socio-demographic variables, forms of presentation, pain characteristics, associated symptoms, patient management and outcome data were collected.

Results: Sixty women were included with a mean age of 43 years at diagnosis, 87% (n=52) were multiparous, with a mean of 2 previous pregnancies. In 95% (57) of all cases the duration of symptoms until the diagnosis exceeded 6 months. Patients were classified according to presence and location of pain in 4 groups: 1. Women with AP pain, 23% (14); 2. Women with OA pain 5% (3); 3. Women with mixed AP and OA pain, 59% (35); and 4. Women with other symptoms 13% (8). Regarding patients from groups 2 and 3 (only OA pain or mixed pain) (38), 90% (34) of them presented low back pain, 53% (20) hip pain and 40% (15) sciatic pain. Only 5% (3) of all patients were evaluated by a rheumatologist.

As for the pain characteristics from groups 1, 2 and 3 (52), in 72% (37) of patients it was diurnal, in 48% (25) it worsened with menstruation, in 62% (32) it worsened with prolonged standing and in 35% (18) it worsened at rest. Among the associated manifestations, stand out the presence of LL varicose veins in 74% (38) of patients, genital varicosities in 58% (30), dyspareunia in 42% (22), dysmenorrhea in 40% (21), hemorrhoids in 37% (19) and dysuria in 18% (9). All patients underwent embolization of the affected vein, with an initial Visual Analogue Scale (VAS) mean of 7.38 over 10, and final VAS mean of 2.63. The mean recovery time was 36 days. The evolution was good or very good in 84% (32) of patients with mixed (AP and OA) pain and in 57% (8) of those who only had AP pain. Less than 2% (1) had a recurrence without the need for reoperation.

Conclusion: PCS is a rare entity, typically associated with long lasting AP pain, but, in more than half the cases, is accompanied with OA symptoms, mainly low back pain. It is important and challenging for the rheumatologist to identify these patients, since treatment is usually safe and effective, and diagnostic delay worsens their quality of life.

Disclosure of Interests: None declared

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27. BACK PAIN, MECHANICAL MUSCULOSKELETAL PROBLEMS, LOCAL SOFT TISSUE DISORDERS

THU0501

MUSCULOSKELETAL DISORDERS IN TYPE 1 AND 2 DIABETIC PATIENTS: PREVALENCE AND ASSOCIATION WITH MICROVASCULAR COMPLICATIONS OF DIABETE

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Background: Diabetes mellitus (DM), a worldwide high prevalence disease, is associated with a large variety of musculoskeletal (MS) disorders. They are poorly treated, as compared to microvascular complications. However, they are a common source of disability.

Objectives: We designed this study to assess the prevalence of MS disorders among diabetic patients and their relation to microvascular complications of diabetes.

Methods: A cross-sectional study enrolled consecutive subjects with diabetes seen in the Endocrinology department. We recorded age of patients, sex, body mass index, type and duration of diabetes, Long-term glycemic control assessed by hemoglobin A1c levels, and lipid profile. Musculoskeletal and microvascular disorders assessment was done by detailed history with clinical examinations and investigations if needed.

Results: A total of 376 subjects were studied, (84.6% had type 2 diabetes). The mean age was 52.5±13.9 years, 41% had one or more

microvascular complications, among which retinopathy was present in 28.2%, nephropathy in 16.1% and neuropathy in 12.8%. Moreover, 23.4% of the patients had one or more musculoskeletal disorders. Shoulder capsulitis was present in 12.5%; carpal tunnel syndrome in 8.8%; trigger finger in 5.9%; and 2.9% had diabetic cheiroarthropathy. Dupuytren's contracture and Charcot foot, were found in 0.5% and 0.3% of the cases respectively. Symptomatic osteoarthritis was found in 19.4%. Musculoskeletal disorders prevalence increased with age, diabetes duration, presence of dyslipidemia and various microvascular complications.

Abstract THU0460 –Table 1. Distribution of cases according to MS disorders in relation to type of diabetes

MS disorders	Type of diabetes					Total N =376	
	Type 1 N = 58		Type 2 N = 318		P		
	No.	%	No.	%	%	No.	%
Osteoarthritis	2	3.4	71	22.3	0.001*	73	14.9
Shoulder capsulitis	2	3.4	45	14.2	0.023*	47	12.5
Carpel Tunnel Syndrome	5	8.6	28	8.8	0.969	33	8.8
Limited joint mobility	0	0	11	3.5	0.151	11	2.9
Trigger Finger	2	3.4	20	6.3	0.397	22	5.9
Dupuytren's contracture	0	0	2	0.6	0.545	2	0.5
Charcot's Foot	1	1.7	0	0	0.154	1	0.3
Total MS disorders	10	17.2	78	24.5	0.149	88	23.4

Conclusion: this study shows a high prevalence of musculoskeletal disorders in diabetic's patients which were significantly associated with advanced age, longer duration of diabetes, presence of dyslipidemia and microvascular complications.

Disclosure of Interests: : None declared

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THU0502

DIFFERENTIAL DIAGNOSIS OF MONOARTHRITIS: THREE CASES WITH PIGMENTED VILLONODULAR SYNOVITIS

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Background: Pigmented Villonodular Synovitis (PVNS) is a proliferative disorder of synovium that affects synovial joints, tendon sheaths and bursas. The estimated incidence is around 1.8 cases per million people in a population. PVNS is usually found in adults aged 20-50 years, but it may occur also in children. As it is a rare pathology in children, diagnosis is often delayed, and it is difficult to distinguish from Juvenile Idiopathic Arthritis (JIA), hemophilic arthropathy, tuberculosis, and other neoplastic processes.

Objectives: The aim of this case series is to emphasize PVNS in the differential diagnosis of monoarthritis, and the importance of interpretation of imaging, i.e. MRI.

Methods: Three pediatric PVNS cases who were misdiagnosed as monoarticular JIA and familial Mediterranean fever (FMF) are presented as case series.

Results: Case-1. A 14 year-old male had swelling and pain on his left knee for one year. An MRI of knee was reported as joint effusion in suprapatellar bursa, and he was referred to our clinic as monoarticular JIA. He was on methotrexate (MTX) 15 mg/m²/wk. SC, and despite effective treatment there was no improvement. MRI was performed by a pediatric radiologist who was an expert on musculoskeletal diseases. Joint effusion and a lesion compatible with PVNS were observed in left suprapatellar recess (Figure-A). Arthroscopic synovectomy was performed and histopathological result was PVNS.

Case-2: A one year-old female had swelling on her right knee for three months after a minor trauma. On physical examination, she had effusion of the right knee which was confirmed on USG. She was diagnosed as monoarticular JIA and intraarticular triamcinolone hexacetonide injection was performed. She was unresponsive to adequate non-steroid drug therapy, and MTX was started. Despite intensive therapy, there was no change. MRI showed focal lesions with high signal in T2 weighted images in suprapatellar and intraarticular areas, and findings were evaluated as compatible with PVNS (Figure-B). Total synovectomy was performed and PVNS was confirmed in pathological evaluation (Figure-C). she is in complete remission without treatment.

Abstract THU0502 – Figure 1. A. Hypo intense mass lesion in the left knee of the first case and on the in the suprapatellar and intraarticular areas (T-1-weighted MRI image). B. Intraoperative image of open approach total synovectomy (second case). C. Suprapatellar and intraarticular lesions compatible with PVNS right knee of second case (T-2 weighted image). D. Suprapatellar and intraarticular focal hyper intense lesions (T-2-weighted MRI image).

Case-3: An eleven-year-old female was referred to our clinic with recurrent swelling and pain on her right knee. Attacks lasted for 2-3 days and resolved spontaneously, accompanied by abdominal pain and vomiting. FMF attack was suspected, for she had signs of peritonitis with knee arthritis, and elevated acute phase reactants. A744S/R202Q compound heterozygote mutations were detected in the MEFV analysis. Colchicine was started 1 mg/kg/ per day. Abdominal pain attacks were resolved with colchicine, but articular symptoms persisted. MRI of the right knee detected focal hyper intense lesions in T2 weighted images compatible with PVNS. Total synovectomy was performed and confirmed PVNS.

Conclusion: PVNS is a rare but serious reason for joint effusion, presented as chronic arthritis. It should be considered in the differential diagnosis of mono-arthritis, especially when the patient is unresponsive to specific treatment for JIA. MRI is the best diagnostic tool in imaging, and histopathological evaluation should be performed to confirm the diagnosis.

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THU0503 ICONOGRAPHIC STUDY OF ACUTE MECHANICAL HIP PAIN IN YOUNG PATIENTS: CLINICAL-ULTRASONOGRAPHIC CORRELATE AND PROPOSAL OF DIAGNOSTIC-THERAPEUTIC ALGORITHM

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Background: Coxarthrosis is the first cause of mechanical pain and functional limitation of the hip joint, however its prevalence in subjects over 65 years is much higher than the incidence of these symptoms so its causal link is not complete. In a young population, mechanical groin pain of acute origin is usually linked to periarticular processes mainly related to adductor muscles, psoas and tendons with pertrocanteric entheses. In the routine approaches of these patients, when they consult, the simple radiological study is included and it is usually treated with NSAIDs, assuming tendinopathy as a diagnostic process.

Objectives: The purpose of this study is to determine the ultrasound characteristics of the coxofemoral joint of patients who consult for acute inguinal pain, correlate it with its clinical presentation and propose a reasoned strategy for its management.

Methods: We reviewed the clinical records of patients under the age of 50 who consulted for direct non-traumatic hip joint pain. Only cases with less than one week of evolution were included. Patients who consulted on weekends or in the afternoon were not included in this register because of the lack of immediate ultrasound study during these periods of time. We excluded those patients who, during the consultation, acknowledged having used anti-inflammatory drugs in the last 24 hours and those whose records are incomplete. The ultrasounds were performed with a portable Logiq e ultrasound machine, equipped with a linear probe of up to 12MHz.

Results: Between 2014 and 2017, 211 patients under 50 years of age consulted for non-traumatic mechanical groin pain. Excluding patients who consulted outside of the immediate ultrasound study access hours, who had recently taken NSAIDs and whose clinical records were incomplete, we reviewed 116 clinical and image records. The mean age of the patients whose records were included was 37.8 SD 8.5. The mean time, in days, between the consultation and the onset of symptoms according to history constancy was 4 SD 2. 59 subjects were male (50.9%). In the physical examinations described in the reports, 29 patients (25%) presented painful passive manoeuvres, 38 (32.8%) presented painful active manoeuvres, and 22 (19%) could not maintain standing due to the impossibility of loading due to coxofemoral pain. Regarding the ultrasound findings, 5 patients presented unequivocal capsular distension at the level of the upper anterior labrum (4.3%) and 32 (27.6%) presented distension of the anterior recess. One patient with capsular distension was finally diagnosed with ONA and another with APso.

Abstract THU0503 – Table 1. summarizes the prevalence of clinical findings depending on the existence or not of distension of the anterosuperior labrum or of the previous recess in the ultrasound scan performed.

Physical examination results		Joint capsule				Joint recess			
		Normal		Distended		Normal		Distended	
		N	% of row	N	% of row	N	% of row	N	% of row
Inability to stand	NO	94	100,0%	0	0,0%	79	84,0%	15	16,0%
	YES	17	77,3%	5	22,7%	5	22,7%	17	77,3%
Painful passive maneuvers	NO	87	100,0%	0	0,0%	72	82,8%	15	17,2%
	YES	24	82,8%	5	17,2%	12	41,4%	17	58,6%
Pain to mobility against resistance	NO	78	100,0%	0	0,0%	65	83,3%	13	16,7%
	YES	33	86,8%	5	13,2%	19	50,0%	19	50,0%

Conclusion: Our results show that in the studied population (<50 years old, non-traumatic coxofemoral pain of sufficient intensity to be consulted in the emergency department in less than 7 days of evolution) synovitis is a finding present in a quarter of patients and that it is severe in approximately 4%. Its strong association with incapacity for load and passive joint dysfunction would condition the incorporation of these findings in therapeutic decision making, including -in these cases- rest in unloading. On the other hand, the absence of synovitis in $\frac{3}{4}$ parts of the patients strongly suggests a muscular etiology that with high probability will respond positively only to NSAIDs. The incorporation of ultrasound, in our opinion, plays a role of diagnostic clearance when the semiological exploration is doubtful.

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THU0504 ROTATOR CUFF CALCIFICATION AND SHOULDER PAIN: A CLINICAL AND ECHOGRAPHIC STUDY OF 465 PATIENTS

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Background: Shoulder pain is a common cause for medical consultation and is commonly linked to lesions of the rotator cuff. Rotator cuff calcification (due to BCP crystal deposition) is also a common finding and can give rise to chronic pain as well as acute inflammatory episodes

Objectives: The goal of the study was to compare the clinical and the echographic characteristics of patients with BCP calcification in the rotator cuff and shoulder pain to those with no signs of calcification

Methods: A retrospective case-control study of 465 patients whose primary complaint was shoulder pain presenting between 1997 and 2011 and seen by one rheumatologist. 125 patients who had rotator cuff calcification (RCC) were identified and constituted the study group. We compared the patients with RCC with 125 patients without calcification who were randomly extracted from the same registry which constituted the control group. All had detailed demographic and clinical documentation as well as a precise description of the echographic findings. Subgroups

were defined according to the type and the duration of symptoms (Hyperacute painful shoulder, subacute inflammatory symptoms and chronic mechanical pain). Short-term response (up to 6 weeks) to treatment was also analyzed for the two first subgroups

Results: 25% of the patients consulting for shoulder pain had evidence of RCC as demonstrated by ultrasound. Hyperacute and subacute inflammatory symptoms were not linked to calcification (Hyperacute: 16 pts with 13pts without (p value : ns), subacute: 32 pts in both groups). The mean age (54 years) and female predominance (60%) were also similar in patients with or without calcification. However, patients with hyperacute painful shoulder with RCC were younger (mean years: 49 against 58 years; p: 0.007). None were <35 or > 60 years old and 75% of them were females. In 60% of cases of hyperacute painful shoulder linked to calcification, the acute flare was the first shoulder complaint compared to <30% in the overall sample (p: <0.0001).

More than a 1/3 of the patients with RCC (35% in chronic and 43% in acute symptom groups) had no other echographic lesion compared to < 5% of patients without RCC. Calcifications were rarely associated with partial or total rotator cuff rupture (<10% against 25% without, p: 0.007). Steroid infiltrations were mostly performed with rapid short-term response in the acute and subacute groups linked to calcification: 15/16, 24/34 against 4/13 p:0.001, 10/34 p:0.001 without calcification. Surgery was necessary in 7/101 pts with chronic symptoms without calcification against only 1/95 when calcifications were present (p:0.03).

Conclusion: Clinical and demographic data cannot clearly predict the presence of RCC in patients presenting with shoulder pain. However, acutely symptomatic RCC is found preferentially in middle aged women, often with no degenerative or traumatic echographic lesions. Steroid infiltration appears to be the treatment of choice when acute inflammatory symptoms linked to RCC are present

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Basic science in paediatric rheumatology

THU0505 INTRINSIC AND EXTRINSIC B CELL DEFECT IN DADA2 PATIENTS

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Background: ADA2 Deficiency is an autoinflammatory disease characterized by systemic vasculopathy, strokes and mild immunodeficiency. The defect is due to a mutation in ADA2 gene. It regulates the catabolism of extracellular adenosine, which is an important regulator of Class Switch Recombination in B lymphocytes.

Objectives: We addressed if ADA2 mutation affects directly B-cell function and the capacity of helper T cells to support B cells.

Methods: 14 patients carrying LOF mutations in ADA2 were examined. We analyzed immunophenotype by flow cytometry. B cells isolated from DADA patients or HD have been cultured alone or in co-culture with CD4+ T cells and *in vitro* B cell proliferation has been evaluated by CFSE dilution, whereas B cell differentiation to Immunoglobulins secreting cells in response to TLR9 agonist and T cell help has been evaluated by ELISA assay.

Results: Flow-cytometric analysis showed a significant reduction of switched memory B cells and of CD4+/CD8+ T cells. We identified an significant expansion of circulating Tfh cells in DADA2. Then we investigated a role of ADA2 in B cells: we found that it is expressed, secreted but the activity is strongly impaired in DADA2. We also addressed the interaction between B and T cells; we found that proliferation and differentiation of patients' B cells were not sustained from patient's T cells.

Moreover DADA2 T cells showed an impairment in the IL21 production and a downregulation of CD40L.

Conclusion: Our findings suggest that ADA2 mutation could affects directly B cell function and T cell help functions.

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I perform consultancy activities on behalf of the Gaslini Institute for the companies listed below:

AbbVie, Biogen, Boehringer Ingelheim, Bristol-Myers Squibb, EMD Serono, Janssen, Novartis, Pfizer, R-Pharm.

The money received for these activities are directly transferred to the Gaslini Institute's bank account. Before March 2016, I was the head of the Pediatric Rheumatology Department at the G. Gaslini Hospital, where the PRINTO Coordinating Centre is located. For the coordination activity of the PRINTO network, the Gaslini Hospital received contributions from the industries listed in this section. This money has been reinvested for the research activities of the hospital in fully independent manners besides any commitment with third parties., Elisabetta Traggiai Employee of: Novartis, Marco Gattorno Grant/research support from: MG has received unrestricted grants from Sobi and Novartis

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THU0506 MAST CELL DEFICIENCY AMPLIFIES INFLAMMATORY RESPONSE IN A MOUSE MODEL OF KAWASAKI'S DISEASE

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Background: Kawasaki's disease is a pediatric medium vessel vasculitis causing coronary arteritis and potentially coronary artery aneurysms. Higher systemic levels of Interleukin-6 (IL-6) in the first week of disease has been shown to be associated with a higher risk of coronary artery aneurysms (1, 2). Mice injected with *Candida albicans* water-soluble fraction (CAWS) develop coronary and aortic arteritis like that seen in Kawasaki's disease. Our prior work has demonstrated that mast cell degranulation can inhibit lipopolysaccharide (LPS)-induced IL-6 gene expression in the aorta and systemic IL-6 production (3).

Objectives: The objective of this study was to determine if mast cells play a role in the IL-6 homeostasis in an established mouse model of Kawasaki's disease (CAWS model).

Methods: 8-10-week-old male wild type controls (WT) and KitW-sh/W-sh mice (mast cell deficient, MCD) of C57Bl/6 background were randomly distributed into four groups (WT-PBS, WT-CAWS, MCD-PBS, MCD-CAWS). They were either injected intraperitoneally with PBS or CAWS (2 mg/mouse) daily for a period of 5 days. Eight animals from each group