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