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 Kit^{W-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