were defined according to the type and the duration of symptoms (hyper-acute 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 of subacute female patients (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: <0.001).

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%, 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, acute symptomatically severe shoulder pain is found preferably 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:** DADA2 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 adenosine into inosine, a xenobiotic substance. B and T cell functions were impaired in DADA2 patients. Hence, we addressed if ADA2 mutation affects directly B-cell function as well.

**Objectives:** We addressed if ADA2 mutation affects directly B-cell function in DADA2 patients.

**Methods:** We analyzed immunophenotype by flow cytometry. B cells isolated from 14 patients carrying LOF mutations in ADA2 were examined. The mean age of the patients (54 years old) were females (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: <0.001).

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**Conclusion:** Clinical and demographic data cannot clearly predict the presence of RCC in patients presenting with shoulder pain. However, acute symptomatically severe shoulder pain is found preferably 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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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. Elisa Traggia Employee of: Novartis, Marco Gattorno Grant/research support from: MG has received unrestricted grants from Solbi and Novartis

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**Mast cell deficiency amplifies inflammatory response in a mouse model of Kawasaki’s disease**


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