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Table 1. Baseline characteristics

	Mean (standard deviation) or %			P-value ^a
	App use (n=128)	App use + discussion (n=158)	No app use (n=1638)	
Age in years	42.44 (12.11)	43.62 (13.15)	47.77 (13.31)	<0.001
% Male	44.5%	45.9%	42.0%	0.572
Diagnosis				0.030
% RA	35.9%	38.2%	43.8%	
% PsA	21.1%	16.0%	21.0%	
% axSpA	43.0%	45.2%	43.8%	
Disease duration in years	16.10 (4.28)	16.49 (4.19)	16.53 (4.69)	0.596
Low disease activity at baseline	35.8%	40.3%	34.3%	0.356

#Test of difference between groups

*Low disease activity was defined as DAS28 < 3.2 in RA and BASDAI ≤ 4 in axSpA. In PsA, it was defined as fulfilling at least 5 of 7 criteria for minimal disease activity [2].

Table 2. Association of app use with patient satisfaction and disease management

Outcome	Odds Ratio (95% CI)	
	App use only	App use + discussion
Satisfaction with shared decision making ^a	0.79 [0.55, 1.17]	1.74 [1.14, 2.66]
Satisfaction with following evolution of disease ^a	0.73 [0.49, 1.09]	2.07 [1.36, 3.14]
Low disease activity at last visit ^a	1.08 [0.65, 1.72]	1.30 [0.82, 2.00]
Improved disease activity ^a	1.10 [0.68, 1.76]	1.54 [0.98, 2.40]
Treatment intensification ^a	1.82 [1.16, 3.01]	1.68 [0.80, 1.95]

^aTotal=1924 (app use only=128, app use + discussion=158, no app use=1638) & Total=1141 (app use only=106, app use + discussion=114, no app use=921)

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From the cradle to the grave – what does paediatric disease teach us about adult disease?___

OP0106

A NOVEL KNOCK-IN MOUSE MODEL OF CAPS THAT DEVELOPS AMYLOIDOSIS: THERAPEUTIC EFFICACY OF PROTON PUMP INHIBITORS

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Background: Cryopyrin associated periodic syndromes (CAPS) are a group of autoinflammatory diseases linked to gain-of-function mutations in the NLRP3

gene that cause uncontrolled IL-1β secretion. CINCA syndrome is the most severe CAPS disease characterized by central nervous system disabilities with a long-term risk of secondary amyloidosis.

Proton pump inhibitors (PPIs), commonly used as inhibitors of gastric acid production, also display anti-inflammatory properties, making them promising drugs in sepsis and in inflammatory disorders.

Objectives: To develop a novel NLRP3 knock-in (KI) mouse model of CAPS to evaluate amyloid deposition and to test alternative therapeutic approaches.

Methods: We generated KI mice by engineering N475K mutation associated with CAPS phenotype into mouse *Nlrp3* gene. KI and Wild Type (WT) mice received PPIs or PBS intraperitoneally and were analyzed for survival, inflammation, cytokine secretion, and amyloidosis development. Cytokines secretion from bone marrow derived dendritic cells (BMDCs) and peritoneal macrophages (PMs) was evaluated by ELISA. Histological analysis of all organs was evaluated by hematoxylin and eosin staining. Amyloid deposition was quantified through Congo Red staining.

Results: Mutant NLRP3 KI mice displayed features that recapitulates the immunological and clinical phenotype of CAPS. These mice had systemic inflammation, with high levels of serum pro-inflammatory cytokines compared to WT controls. Histological analysis revealed the presence of acute and chronic inflammatory cell infiltrates and amyloid deposits in spleen, liver and kidneys. As in CAPS monocytes, BMDCs and PM from KI mice showed a strong increase in IL-1β, IL-18, and IL-1α secretion and decreased levels in interleukin-1 receptor antagonist (IL-1Ra), the naturally occurring IL-1b inhibitor.

PPIs treatment of KI mice showed a clear clinical impact with improvement of inflammatory conditions and regression of amyloid deposits. Remarkably, BMDCs and PMs from PPI-treated mice presented reduced secretion of pro-inflammatory cytokines and re-established the levels of IL-1RA.

Conclusion: NLRP3 KI mice display a CAPS phenotype with many characteristics of autoinflammation, including amyloidosis. The therapeutic effectiveness associated with lack of toxicity indicates that PPIs could represent relevant adjuvants to the anti-IL-1 drugs in IL-1 driven diseases.

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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., Isabella Ceccherini: None declared, Marco Gattorno Grant/research support from: MG has received unrestricted grants from Sobi and Novartis, Anna Rubartelli: None declared, Sabrina Chiesa: None declared

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