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## 31st PRINTo newsletter | September 2020

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### PRINTo news

#### **PRINTo General Assembly at PReS 2020**

We are glad to invite all of you to the upcoming **Annual PRINTo General Assembly at the PReS Congress 2020** !

Due to the COVID-19 restrictions, also the Annual PRINTo General Assembly will be held virtually, though the on line platform of the Congress.

The PRINTo General Assembly will be held on **Thursday 24 September 2020 from 13:00 to 14:00 CET** .

Here is the **agenda** :

- Introduction (**Alberto Martini** )
- The PRINTo/PRES research Grant (**Fabrizio De Benedetti** )
- Cross-cultural adaptation and validation of LoSQI in JLS (**Clare Pain** )
- The HyperPED-COVID Registry (**Roberta Caorsi**)
- The DAISY study (**Dragana Lazarevic** )
- The STARS trial (**Alessandro Consolaro** )
- The JIA classification study (**Nicola Ruperto** )
- Update on the PRINTo research activity (**Nicola Ruperto** )

The staff of the PRINTo Coordinating centre will be also available at the **PRINTo virtual booth** during the Congress.

We will be more than happy to speak with you at the following opening hours:

**Wednesday 23 September 2020: 14.30-16.00 CET**

**Thursday 24 September 2020: 11.00-12.30 and 14.30-16.00 CET**

**Friday 25 September 2020: 11.00-12.30 and 14.30-16.00 CET**

Thank you for your participation, and see you soon, hopefully in person and not virtually!

## PRINTO ongoing projects

### JIA classification study

**The JIA Classification study is a translational research** that aims to validate, in a prospective collection of at least 1,000 patients at disease onset, the proposed PRINTO consensus classification criteria for JIA. This will entail also the collection of biologic samples for ANA, anti CCP, RF, HLA B27 determinations and DNA analysis.

**As of today, more than 500 patients have been enrolled from 56 sites and 92 sites have been activated.** We are halfway there! We remind you that 1000 patients is the final goal to reach.

**PRINTO is providing the kits for the visits requiring samples collection** and laboratory instructions containing also storage and shipment information. If you need more kits please contact PRINTO and we will provide you with a refilling of the necessary material.

We remind you that the **biologic samples should be collected at Month 0 and after 3 months** .

In the next weeks you will be contacted to start with the procedures to export the biologic samples to the central Lab in Genoa.

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### The STARS Trial

**The comparison of Step-up and step-down therapeutic strategies in childhood ARthritiS trial (STARS)** is a new **interventional trial** financed by AIFA (Agenzia Italiana del Farmaco) and by the Italian Foundation Compagnia di San Paolo that will be conducted by PRINTO only in the Italian centres. This clinical trial has the aim **to investigate whether an early aggressive therapeutic intervention in children with JIA** , based on the initial start of synthetic and biologic DMARDs (Step-down strategy), **is superior to an approach based on treatment escalation conducted following the treat-to-target principle** (Step-up strategy). The effectiveness of the two strategies will be assessed by comparing their ability to induce sustained clinical disease remission on/off treatment.

As of today, out of the **22 participating sites** , **15 centres obtained their EC approvals** and **7 centres are currently in progress**. **The first 16 patients have been enrolled at 2 centres**.

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### PharmaChild

**Pharmachild** is a **pharmacovigilance project** which aims at observing children with JIA for 3-10 years undergoing treatment with MTX or biologic agents in order to collect moderate, severe or serious adverse events occurred. Pharmachild was started in 2011 thanks to a European Union (EU) grant within the FP7 framework and has obtained the ENCePP Study Seal ([ENCePP](#)).

As of today, **almost 9,000 patients have been enrolled from 88 centres in 31 countries** .

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### Eurofever

The Eurofever project was promoted in 2008 by the work group of autoinflammatory diseases of the Paediatric Rheumatology European Society (PRES) and was supported by the Executive Agency for Health and Consumers (EAHC).

The main objective of the project has been the creation of a **registry of autoinflammatory diseases** .

A **recent amendment** has been approved in order to:

- add **new autoinflammatory diseases** according to the **new taxonomy** (Ben Chetrit et al Ann Rheum Dis 2018)
- add **new laboratory results and symptoms in the Case Report Forms**
- **update consent forms** according to the current **GDPR**

The Eurofever Registry is now collecting patients affected by 32 conditions ([click here to see the updated list of the diseases](#))

The PI Dr. Marco Gattorno thanks all the **120 centers in 43 countries** that collected information of **more than 4000 patients** with Autoinflammatory diseases.

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We are more than happy to receive any proposal for possible studies based on the Eurofever data! You may send your proposal to [printo@gaslini.org](mailto:printo@gaslini.org) and we will submit it to the Steering Committee (M. Gattorno, J. Frenkel, S. Ozen, N. Ruperto, H. Lachmann).

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### **The HyperPED-COVID Registry**

In the last few months the Novel Coronavirus has been reported as possibly linked to a paediatric **hyper-inflammatory syndrome** . A small number of children have developed a more serious inflammatory condition **in temporal association with COVID-19** in the community, often leading to hospitalization, and occasionally requiring intensive care.

In order to better characterize this new clinical phenotype, few month ago a **joined PRoS, ISSAID, ERN-RITA and PRINTO network's effort** has been started; an international survey has been launched asking the number of patients seen at with this clinical phenotype, the total number is now 470 from 87 pediatric centers in the world.

We are now happy to inform you that the project called **HyperPED-COVID, a retrospective observational study** , is now ready to start: the protocol and the Case Report Forms have been finalised by the Steering Committee and approved by the Coordinating center's Ethic Committee (Genoa). This project will be conducted in close collaboration with **ESID**.

**In a few days PRINTO Registry will be ready to collect clinical and laboratory data of these patients.**

For any further information please contact [printo@gaslini.org](mailto:printo@gaslini.org)

Marco Gattorno (Coordinator, RITA-ERN & ISSAID)

Nico Wullfraat (RITA-ERN), Sefi Uziely, Fabrizio De Benedetti (PRES research clinical affairs), Paul Brogan (ISSAID), Carine Wouters (ERN-RITA),

Claudia Bracaglia and Francesca Minoia (PRES MAS/sJIA working party)

Roberta Caorsi, Alessandro Consolaro (Survey Coordinators)

Nicola Ruperto and Alberto Martini for PRINTO

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### **The DAISY study**

Aim of the study is to investigate sensitivity and predictive value of the multi-biomarker panel (clinical examination of the joints, ultrasound and inflammatory biomarkers) and to evaluate if this panel could be applied in limited number of joints in patients with oligoarticular and polyarticular JIA course to be representative as **a new disease activity tool and outcome measure** .

This multicenter international study in JIA patients involves **selected pediatric rheumatology centers with expertise in the ultrasound examination of joints**. Main coordinating centers will be in Serbia (Nis, PI Dr Dragana Lazarevic) and Italy (Genoa, PI Dr Clara Malattia).

As of today, the study has been **approved in 5 centres** and the other submissions are ongoing.

**The first 7 patients have been enrolled at 2 centres.**

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### **Cross-cultural adaptation and validation of LoSQI in JLS**

**Winner of the 2019 PRoS/PRINTO call for grants** , the study (PI Dr Clare Pain) aims to formally evaluate the psychometric properties of the Localised Scleroderma Quality of Life Instrument (LoSQI) in a sample of patients with JLS.

**PRINTO centres, CARRA and PRES scleroderma working party members** , in partnership with patients and families, cooperate in the **first multi-national study of a disease-specific patient-reported outcome (PRO) in JLS** .

The PI team and PRINTO are currently working on the study set up.

For any further information please contact [printo@gaslini.org](mailto:printo@gaslini.org)

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### Patients enrolled in the PRINTO projects

| Country   | PHARMACHILD | EUROFEVER | JIA classification | The STARS trial |
|-----------|-------------|-----------|--------------------|-----------------|
| Argentina | 123         | 51        | 2                  | 0               |

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|                    |      |      |     |    |
|--------------------|------|------|-----|----|
| Armenia            | 0    | 101  | 0   | 0  |
| Australia          | 0    | 10   | 0   | 0  |
| Austria            | 31   | 58   | 0   | 0  |
| Belgium            | 0    | 2    | 0   | 0  |
| Brazil             | 408  | 16   | 1   | 0  |
| Bulgaria           | 56   | 5    | 7   | 0  |
| Canada             | 0    | 10   | 0   | 0  |
| Chile              | 0    | 5    | 0   | 0  |
| China              | 0    | 14   | 0   | 0  |
| Croatia            | 196  | 12   | 40  | 0  |
| Czech Republic     | 143  | 147  | 0   | 0  |
| Denmark            | 514  | 132  | 3   | 0  |
| Ecuador            | 25   | 1    | 0   | 0  |
| Estonia            | 0    | 0    | 9   | 0  |
| France             | 380  | 304  | 0   | 0  |
| Georgia            | 0    | 9    | 0   | 0  |
| Germany            | 0    | 288  | 31  | 0  |
| Greece             | 520  | 138  | 31  | 0  |
| Hungary            | 136  | 2    | 0   | 0  |
| India              | 119  | 1    | 11  | 0  |
| Israel             | 89   | 134  | 8   | 0  |
| Italy              | 1801 | 1494 | 201 | 16 |
| Japan              | 0    | 5    | 0   | 0  |
| Kazakhstan         | 0    | 0    | 1   | 0  |
| Latvia             | 256  | 4    | 2   | 0  |
| Libya              | 20   | 0    | 0   | 0  |
| Lithuania          | 320  | 2    | 0   | 0  |
| Mexico             | 161  | 0    | 0   | 0  |
| Netherlands        | 704  | 136  | 0   | 0  |
| Norway             | 361  | 2    | 27  | 0  |
| Oman               | 13   | 4    | 0   | 0  |
| Poland             | 31   | 7    | 0   | 0  |
| Portugal           | 0    | 0    | 1   | 0  |
| Romania            | 428  | 42   | 11  | 0  |
| Russian Federation | 492  | 37   | 0   | 0  |
| Saudi Arabia       | 71   | 38   | 0   | 0  |
| Serbia             | 276  | 5    | 4   | 0  |
| Singapore          | 50   | 0    | 14  | 0  |
| Slovakia           | 23   | 11   | 17  | 0  |
| Slovenia           | 0    | 18   | 0   | 0  |
| Spain              | 720  | 232  | 31  | 0  |

|                      |             |             |            |           |
|----------------------|-------------|-------------|------------|-----------|
| Sweden               | 0           | 1           | 0          | 0         |
| Switzerland          | 491         | 96          | 0          | 0         |
| Turkey               | 8           | 215         | 0          | 0         |
| Ukraine              | 0           | 5           | 70         | 0         |
| United Arab Emirates | 0           | 0           | 2          | 0         |
| United Kingdom       | 0           | 262         | 0          | 0         |
| United States        | 0           | 6           | 4          | 0         |
| <b>Total</b>         | <b>8966</b> | <b>4062</b> | <b>528</b> | <b>16</b> |

## [PRINTO overall enrollment status](#)

### [Latest PRINTO papers](#)

Giancane G, Swart JF, Castagnola E, Groll AH, Horneff G, Huppertz HI, Lovell DJ, Wolfs T, Herlin T, Dolezalova P, Sanner H, Susic G, Sztajn bok F, Maritsi D, Constantin T, Vargova V, Sawhney S, Rygg M, K Oliveira S, Cattalini M, Bovis F, Bagnasco F, Pistorio A, Martini A, Wulffraat N, Ruperto N for the Paediatric Rheumatology International Trials Organisation (PRINTO)

#### **Opportunistic Infections in Immunosuppressed Patients with Juvenile Idiopathic Arthritis. Analysis by the Pharmachild Safety Adjudication Committee**

Arthritis Res Ther 2020 Apr 7;22(1):71 [PubMed](#)

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Brunner HI, Tzaribachev N, Cornejo GV, Joos R, Gervais E, Cimaz R, Calvo Penadés I, Cuttica R, Lutz T, Quartier P, Gandhi Y, Nys M, Wong R, Martini A, Lovell DJ, Ruperto N for the Pediatric Rheumatology Collaborative Study Group (PRCSG) and the Paediatric Rheumatology International Trials Organisation (PRINTO)

#### **Maintenance of antibody response to diphtheria/tetanus vaccine in patients aged 2-5 years with polyarticular-course juvenile idiopathic arthritis receiving subcutaneous abatacept.**

Pediatr Rheumatol Online J 2020 Feb 22;18(1):19 [PubMed](#)

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Nordal E, Pistorio A, Rygg M, Giancane G, Maghnie M, Di Iorgi N, Flemming K, Hofer M, Melo-Gomes JA, Bica BERG, Brunner J, Dannecker G, Gerloni V, Harjacek M, Huppertz HI, Pratsidou-Gertsi P, Nielsen S, Stanevicha V, Ten Cate R, Vougiouka O, Pastore S, Simonini G, Ravelli A, Martini A, Ruperto N. for the Paediatric Rheumatology International Trials Organisation (PRINTO)

#### **Growth and puberty in juvenile dermatomyositis: a longitudinal cohort study.**

Arthritis Care Res 2020 Feb;72(2):265-273 [PubMed](#)

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Lovell DJ, Brunner HI, Reiff AO, Jung L, Jarosova K, N?mcová D, Mouy R, Sandborg C, Bohnsack JF, Elewaut D, Gabriel C, Higgins G, Kone-Paut I, Jones OY, Vargová V, Chalom E, Wouters C, Lagunes I, Song Y, Martini A, Ruperto N. for the Pediatric Rheumatology Collaborative Study Group (PRCSG) and the Paediatric Rheumatology International Trials Organisation (PRINTO)

#### **Long- term outcomes in patients with polyarticular juvenile idiopathic arthritis receiving adalimumab with or without methotrexate.**

RDM open 2020 Jul;6(2):e001208 [PubMed](#)

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Brunner HI, Abud-Mendoza C, Viola DO, Calvo Penades I, Levy D, Anton J, Calderon JE, Chasnyk VG, Ferrandiz MA, Keltsev V, Paz Gastanaga ME, Shishov M, Boteanu AL, Henrickson M, Bass D, Clark K, Hammer A, Ji BN, Nino A, Roth DA, Struemper H, Wang ML, Martini A, Lovell D, Ruperto N for the Paediatric Rheumatology International Trials Organisation (PRINTO) and the Pediatric Rheumatology Collaborative Study Group (PRCSG).

#### **Safety and efficacy of intravenous belimumab in children with systemic lupus erythematosus: results from a randomised, placebo-controlled trial.**

Ann Rheum Dis 2020 Oct;79(10):1340-1348. [PubMed](#)

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Brunner HI, Quartier P, Alexeeva E, Constantin T, Koné?Paut I, Marzan K, Schneider R, Wulffraat NM, Chasnyk V, Tirosh I, Kallinich T, Kuemmerle?Deschner J, Wouters K, Lauwerys B, Nikishina I, Trachana M, Vougiouka O, Martini A, Lovell DJ, Levy J, Vritzali E, Ruperto N, for the Paediatric Rheumatology International Trials Organisation (PRINTO) and the Pediatric Rheumatology Collaborative Study Group (PRCSG)

## **Efficacy and Safety of Canakinumab in sJIA Patients with and without Fever at Baseline: Results from an Open-label, Active Treatment Extension Study**

Arthritis Rheum [Epub ahead of print] [PubMed](#)

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Brunner HI, Chen C, Bovis F, De Benedetti F, Espada G, Joos R, Akikusa J, Chaitow J, Boteanu AL, Kimura Y, Rietschel C, Siri D, Smolewska E, Schmeling H, Brown DE, Martini A, Lovell DJ, Huang B, Ruperto N for the Paediatric Rheumatology International Trials Organisation (PRINTO) and the Pediatric Rheumatology Collaborative Study Group (PRCSG)

## **Functional Ability and Health-Related Quality of Life in Randomized Controlled Trials of Tocilizumab in Patients With Juvenile Idiopathic Arthritis**

Arthritis Care Res [Epub ahead of print] [PubMed](#)

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Quartier P, Alexeeva E, Tamàs C, Chasnyk V, Wulffraat N, Palmblad K, Wouters C, Brunner H, Marzan K, Schneider R, Horneff G, Martini A, Anton J, Wei X, Slade A, Ruperto N, Abrams K for the Pediatric Rheumatology International Trials Organization (PRINTO), the Pediatric Rheumatology Collaborative Study Group (PRCSG).

## **Tapering Canakinumab Monotherapy in Patients with Systemic Juvenile Idiopathic Arthritis in Clinical Remission: Results from an Open-label, Randomized Phase IIIb/IV Study.**

Arthritis Rheum [Epub ahead of print] [PubMed](#)

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Brunner HI, Nanda K, Toth M, Foeldvari I, Bohnsack J, Milojevic D, Rabinovich CE, Kingsbury DJ, Marzan K, Chalom E, Horneff G, Kuester RM, Dare JA, Trachana M, Jung LK, Olson J, Minden K, Quartier P, Bereswill M, Kalabic J, Kupper H, Lovell DJ, Martini A, Ruperto N; Paediatric Rheumatology International Trials Organisation (PRINTO) and the Pediatric Rheumatology Collaborative Study Group (PRCSG)

## **Safety and Effectiveness of Adalimumab in Patients With Polyarticular Course of Juvenile Idiopathic Arthritis: STRIVE Registry 7-Year Interim Results**

Arthritis Care Res [Epub ahead of print] [PubMed](#)

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Gattorno M, Hofer M, Vanoni F, Federici S, Cantarini L, Ruperto N for the Paediatric Rheumatology International Trials Organisation (PRINTO) and the Eurofever Registry

## **Response to: 'Do we need the PFAPA syndrome in adults with non-monogenic periodic fevers?' by Fayand et al**

Ann Rheum Dis [Epub ahead of print] [PubMed](#)

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Gattorno M, Hofer M, Federici S, Vanoni F, Ruperto N for the Paediatric Rheumatology International Trials Organisation (PRINTO) and the Eurofever Registry.

## **Some clarifications on the new classification criteria for recurrent fevers**

Semin Arthritis Rheum [Epub ahead of print] [PubMed](#)

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## **PRINTO membership**

As of today, PRINTO has reached 1511 effective members in 679 centres from 93 countries.

If you wish to become a PRINTO member

and receive regular updates about our research activity and invitations to our projects

please go to:

<https://www.printo.it/contact/apply-membership>

Your cooperation will be more than welcome

and your effort will be essential for the research in the field of paediatric rheumatic diseases.

**WELCOME ABOARD!**

## **PRINTO advisory council & contacts**

**Chairman**

**Alberto Martini, MD, Prof - Genoa, Italy**

**Counsellors**

**Tadej Avcin, MD, PhD - Ljubljana, Slovenia**  
**Michael Hofer, MD, Prof - Lausanne, Switzerland**  
**Seza Ozen, MD, Prof - Ankara, Turkey**  
**Pierre Quartier, MD - Paris, France**  
**Claudia Saad Magalhães, MD, Prof - Botucatu, Brazil**  
**Joos Swart, MD - Utrecht, The Netherlands**

**Senior Scientist**

**Nicolino Ruperto, MD, MPH - Genoa, Italy**

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