

## Patients enrolled in the PRINTO projects

COUNTRIES	Pharmachild	EPOCA	Eurofever	MAS
Albania			1	
Argentina		309	33	39
Armenia			101	
Australia			10	1
Belgium			13	15
Bosnia				2
Brazil	68	289	10	34
Bulgaria		279		
Canada		7		38
Chile		47	5	
China			14	20
Colombia		10		
Croatia	83		11	13
Czech R.	67	203	193	
Denmark	30	402	128	6
Finland		200		
France	25		262	29
Estonia		76		
Georgia			9	6
Germany		425	263	34
Greece	176	204	62	39
Hungary		251	24	
India		238	3	55
Iran		311		
Israel		219	162	10
Italy	508	789	662	338

## Patients enrolled in the PRINTO projects

COUNTRIES	Pharmachild	EPOCA	Eurofever	MAS
Kuwait				
Japan			6	7
Latvia	9	304	5	47
Lithuania	14	213	6	
Lybia				42
Mexico		199		1
Netherlands	120	326	92	9
Norway	13	368		5
Oman	5	143	6	
Paraguay		101		
Poland	30	252	5	
Portugal		43		
Romania	193	411	23	
Russia	1		29	15
Saudi Ar.		49	39	3
Serbia	91	249	3	6
Slovakia	38	208	1	
Slovenia		117	13	14
S.Africa		32		
Spain	476	406	114	39
Sweden		50	1	4
Switzerland		100	92	1
Turkey		454	165	131
UK		200	263	2
USA		213	6	98
Venezuela				8
<b>Total</b>	<b>1947</b>	<b>8697</b>	<b>2835</b>	<b>1111</b>

September 2013

# Paediatric Rheumatology International Trials Organisation (PRINTO)



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20<sup>th</sup> NEWSLETTER

PRoS Congress 2013 Ljubljana, Slovenia  
Saturday September 28<sup>th</sup> 2013  
at 14.30-15.30, Linhart Hall  
Agenda for the annual PRINTO workshop

- Introduction (A. Martini)
- Update on PRINTO Projects (N. Ruperto)
- Pharmachild (N. Wulffraat, N. Ruperto)
- Share (P. Dolezalova, N. Wulffraat)
- The PRES EuroFever project (M. Gattorno)
- The EPOCA and MAS study (A. Ravelli)

#### The PharmaChild Project

This project is a retrospective and prospective registry with the aim to collect moderate-severe serious adverse events and related efficacy in JIA patients treated with biologics and/or MTX on a long-term period.

The project and related web database is now up and running (see attached table). The web data base has been created with the aim to be an international electronic resource for all PRINTO centres in order to collect safety and efficacy information on JIA patients. It will allow the immediate **quantification** of the disease activity (JADAS, ACR, flare, inactive disease etc) status as well as the possibility to autonomously **download electronic data** for local research or other related purposes. The web system is supposed to be friendly enough to be used in **daily clinical practice**.

As of September 2013, 65 PRINTO centres have been activated. A total of 6858 patients have been entered into the census, 1947 in the retrospective and 52 in the prospective. **As the grant will be over in March 2014, the retrospective data collection must be finished by the end of 2013 in order to obtain the funding.**

The project is financed by the European Union (PI Dr N. Wulffraat 2011-2014).

The EPOCA study (EPidemiology, treatment and Outcome of Childhood Arthritis). The study is aimed at photographing the current status of children with JIA across continents and countries. PRINTO national coordinators (NCC list at [www.printo.it](http://www.printo.it)) were asked firstly to translate and cross-culturally validate the parent and child versions of the Juvenile Arthritis Multidimensional Assessment Report (JAMAR) and secondly to assess 100 consecutive JIA patients and 100 healthy children. Additional centres may participate by collecting each 100 consecutive JIA pa-

tients. The national language translations of the JAMAR will be published in a dedicated supplement in Clinical and Experimental Rheumatology.

The aim of this study is therefore twofold:

- 1) to foster regular use of parent/patient-reported outcomes in paediatric rheumatology practice;
- 2) to obtain information about the frequency of ILAR categories, treatment modalities, and current outcome of JIA around the world.

As of today 46 translations have been completed and other 6 are in progress (see [www.printo.it/jamar.asp](http://www.printo.it/jamar.asp)). The data collection phase started in 2011 through a dedicated web database on the PRINTO member area and as of today more than 8000 patients have been collected. We have planned to close officially the EPOCA data collection at the latest by the end of 2013.

The PRES Eurofever project. The Eurofever Registry (PI Dr M. Gattorno) aims to increase the knowledge on the clinical presentation, response to treatment and complication of Autoinflammatory Diseases. The registry is fully enrolling; a total of 2835 patients (see table) have been enrolled so far: among them 225 with CAPS, 402 with CRMO, 815 with FMF, 171 with MKD, 581 with PFAPA and 244 with TRAPS. New important papers have been recently published (see list on next page). Other publications are ongoing: description of CAPS, MKD and CRMO populations, evidence-based classification criteria for periodic fevers.

#### The MAS criteria study

The MAS Criteria Study is an international and multinational project, aimed to develop a new set of classification criteria for MAS complicating sJIA. The data collection is now over with 1111 patients collected: 362 patients with sJIA-associated MAS, 404 with acute flare of sJIA, 345 with febrile systemic infection. Statistical analysis on these data are underway.

We are now moving forward the third step of the project: a consensus conference on MAS classification criteria is planned for March 2014.

#### The SHARE project

The "Single Hub and Access point for paediatric Rheumatology in Europe" (SHARE) is a European project whose main objective is to provide the European countries with recommendations for the care of children

with rheumatic disease. These recommendations will be based on systematic literature reviews and on the online surveys sent by PRINTO to individual centres belonging to its network all over the world.

To identify the specific needs for the optimal care in PRD an online survey has been implemented ([www.printo.it/SHARE](http://www.printo.it/SHARE)) and already sent to all the PRINTO and PRES members.

Another important aim of the project is to update and enrich the PRINTO website for families. The site (<http://www.printo.it/pediatric-rheumatology>), actually visited by more than 11.000 per day, offers scientific information regarding the pediatric rheumatic diseases (PRD), the list of centres dealing with PRD, and the list of the family associations in more than 50 languages. We are now starting the procedures for the renewal of the website, and the revision and update of the PRD description (PI Dr N. Wulffraat, EU project number 2011 1202).

#### New onset JDM trial:

The enrollment of the trial is now closed with 139 patients randomized from 55 centres in 23 countries. No major safety concern identified since last year. The analysis of the data has been completed. The results of the trial will be presented during the PRES meeting on Saturday September 28 at 15.30 pm in the Linhart Hall (PRoS13-ABS-1281)

Title: A RANDOMIZED TRIAL IN NEW ONSET JUVENILE DERMATOMYOSITIS: PREDNISONE VERSUS PREDNISONE PLUS CYCLOSPORINE VERSUS PREDNISONE PLUS METHOTREXATE.

#### Membership

In 2013 PRINTO has reached 982 effective members in 451 centres from 81 countries. (with an increase of 10%).

**WELCOME ABOARD!**

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/membership.asp>

Your cooperation will be more than welcome and your effort will be essential for the research in the field of paediatric rheumatic diseases.

For more info: [printo@ospedale-gaslini.ge.it](mailto:printo@ospedale-gaslini.ge.it)

## Recent PRINTO publications

For reprints go to [www.printo.it](http://www.printo.it) and click on "papers".

- Ruperto N, Quartier P, Wulffraat N, Woo P, Ravelli A, Mouy R, Bader-Meunier B, Vastert SJ, Nosedà E, D'Amrosio D, Lecot J, Chakraborty A, Martini A, Chioato A for PRINTO. A phase II study to evaluate dosing and preliminary safety and efficacy of canakinumab in systemic juvenile idiopathic arthritis with active systemic features. *Arthritis Rheum* 2012;64(2):557-567.
- Demirkaya E, Ozen S, Pistorio A, Galasso R, Ravelli A, Hajsija R, Baskin E, Dressler F, Fischbach M, Garcia Consuegra J, Igaru N, Pasic S, Scarpato S, van Rossum MAJ, Apaz MT, Barash J, Calcagno G, Gonzalez B, Hoppenreijts E, Ioseliani M, Mazur-Zielinska H, Vougiouka O, Wulffraat N, Luqmani R, Martini A, Ruperto N, Dolezalova P, for PRINTO. Performance of the Birmingham Vasculitis Activity Score and Disease Extent Index in childhood vasculitides. *Clin Exp Rheumatol* 2012;30(Suppl. 70):S162-168.
- Toplak N, Frenkel J, Ozen S, Lachmann HJ, Woo P, Koné-Paut I, De Benedetti F, Neven B, Hofer M, Dolezalova P, Kümmerle-Deschner J, Touitou I, Hentgen V, Simon A, Girschick H, Rose C, Wouters C, Vesely R, Arostegui J, Stojanov S, Ozgodan H, Martini A, Ruperto N, Gattorno M, for PRINTO. Eurotraps and Eurofever Projects. An International registry on Autoinflammatory diseases: the Eurofever experience. *Ann Rheum Dis* 2012;71:1177-1182.
- Ruperto N, for PRINTO. Classification criteria and diagnostic tests for vasculitides. *J Rheumatol* 2012;39:1503-1505.
- De Benedetti F, Brunner HI, Ruperto N, Kenwright A, Wright S, Calvo I, Cuttica R, Ravelli A, Schneider R, Woo P, Wouters C, Xavier R, Zemel L, Baildam E, Burgos-Vargas R, Dolezalova P, Garay SM, Merino R, Joos R, Grom A, Wulffraat N, Zuber Z, Zulian F, Lovell DJ, Martini A, for PRINTO and PRCSSG. Randomized Trial of Tocilizumab in Systemic Juvenile Idiopathic Arthritis. *N Engl J Med* 2012;367:2385-2395.
- Ruperto N, Brunner HI, Quartier P, Constantin T, Wulffraat N, Horneff G, Brik R, McCann L, Kasapcopur O, Rut-

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- Consolaro A, Bracciolini G, Ruperto N, Pistorio A, Magni-Manzoni S, Malattia C, Pederzoli S, Davi S, Martini A, Ravelli A, for PRINTO. Remission, minimal disease activity and acceptable symptom state in juvenile idiopathic arthritis. *Arthritis Rheum* 2012;64(7):2366-2374.
- Gerss J, Roth J, Holzinger D, Ruperto N, Wittkowski H, Frosch M, Wulffraat N, Wedderburn L, Stanevicha V, Mihaylova D, Harjacek M, Len C, Toppino C, Masi M, Minden K, Saurenmann T, Uziel Y, Vesely R, Apaz MT, Kuester RM, Rua Elorduy MJ, Burgos-Vargas R, Ioseliani M, Magni-Manzoni S, Unsal E, Anton J, Balogh Z, Hagelberg S, Mazur-Zielinska H, Tauber T, Martini A, Foell D for PRINTO. Phagocyte-specific S100 proteins and high-sensitivity C-reactive protein as biomarkers for a risk-adapted therapy to maintain remission in juvenile idiopathic arthritis: a comparative study. *Ann Rheum Dis* 2012;71:1991-1997.
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- Ruperto N, Vesely R, Saint-Raymond A, Martini A for PRINTO. The impact of the European paediatric legislation in pediatric rheumatology: past present and future. *Ann Rheum Dis* 2013; 0:1-4. doi:10.1136/annrheumdis-2013-204168. [Epub ahead of print]
- Lachmann HJ, Papa R, Minden K, Obici L, Touitou I, Cantarini L, Frenkel J, Anton J, Kone-Paut I, Cattalini M, Bader-Meunier B, Insalaco A, Hentgen V, Merino R, Modesto C, Toplak N, Berendes R, Ozen S, Cimaz R, Jansson A, Brogan P, Hawkins PN, Ruperto N, Martini A, Woo P, Gattorno M for PRINTO, the Eurotraps and the Eurofever Project. The phenotype of TNF-receptor associated autoinflammatory syndrome (TRAPS) at presentation: a series of 158 cases from the EUROFEVER/EUROTRAPS international registry. *Ann Rheum Dis* 2013; 0:1-8. doi:10.1136/annrheumdis-2013-204184. [Epub ahead of print]
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