Patients enrolled in the PRINTO projects

Country	Pharmachild	EPOCA	Eurofever	MAS
Albania				
Algeria		140		
Argentina	123	473	55	39
Armenia			101	
Australia			10	1
Austria	25			
Belgium			13	15
Bosnia				2
Brazil	375	299	13	34
Bulgaria	57	300		
Canada		7	1	38
Chile		119	5	
China			14	20
Colombia		10		
Croatia	100	100	11	13
Czech R.	124	203	194	
Denmark	524	402	131	6
Finland		276		
France	150		284	29
Egypt		34		
Estonia		210		
Georgia		190	9	6
Germany		424	275	34
Greece	432	375	96	39
Hungary	126	297	24	
India	103	375	3	55
Iran		320		
Israel	81	216	162	10

Patients enrolled in the PRINTO projects

Country	Pharmachild	EPOCA	Eurofever	MAS
Italy	1161	1304	772	338
Japan			6	7
Latvia	259	304	6	47
Lithuania	301	217	7	
Lybia		148		42
Mexico	12	199		1
Netherlands	466	317	97	9
Norway	331	375		5
Oman	16	143	6	
Paraguay		151		
Poland	29	248	5	
Portugal		82		
Romania	409	411	38	
Russia	470		43	15
Saudi Ar.	62	59	39	3
Serbia	272	349	4	6
Slovakia	124	208	1	
Slovenia	53	117	13	14
S.Africa		65		
Spain	662	566	196	39
Sweden		50	1	4
Switzerland	457	100	94	1
Turkey		460	170	131
UK		200	296	2
Ukraine		200		
USA		412	6	98
Venezuela				8
Total	**7304**	**11455**	**3201**	**1111**

June 2015

Paediatric Rheumatology INternational Trials Organisation (PRINTO)

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22nd NEWSLETTER

PReS Congress 2015 Rome, Italy

Annual PRINTO General Meeting:

Thursday, June 11th, 2015

at 18:15 in Room Vivaldi,

after the Annual PReS General Meeting

Agenda

- Introduction (A. Martini)
- Update on PRINTO Projects (N. Ruperto)
- MYPAN (P. Brogan)
- Pharmachild (N. Wulffraat, J. Swart, N. Ruperto)
- SHARE (P. Dolezalova, N. Wulffraat)
- The PRES EuroFever project (M. Gattorno, S. Federici)
- EPOCA/MAS studies (A. Ravelli, A. Consolaro, F. Minoia)

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 **graphic 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 has been designed to be friendly enough to be used in **daily clinical practice**.

As of May 2015, 93 PRINTO centres have been activated. 10968 patients have been entered into the census, 7304 in the retrospective and 2052 in the prospective. The European Union grant (PI Dr N. Wulffraat 2011-2014) has expired in September 2014 and we are looking for other funding**.**

The Abatacept JIA Registry

This registry **is a sub-study of the PharmaChild project (scientific data property of PRINTO)** meaning that the registry will use exactly the same prospective case report forms and web resources with a complete freedom to publish the related results. This project is an observational registry financed by Bristol-Myers Squibb (Prot. IM101240) that aims to describe the long-term safety of the abatacept treatment for JIA in routine clinical practice. The study is conducted by PRINTO in collaboration with PRCSG (for North American sites). Administrative aspects (ethics committees, contracts and local monitoring) will be handled by the CRO inVentiv Health. As of May 2015, 14 PRINTO centres from 8 countries have been activated and 110 patients have been entered in the study web system.

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) 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 are participating by collecting each 100 consecutive JIA patients. The national language translations of the JAMAR will be published in a dedicated supplement on a rheumatologic journal

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 52 translations have been completed and other 3 are in progress (see **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 almost 8000 JIA patients and 3500 healthy controls have been collected.

The PReS EuroFever project.

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. A new longitudinal section dedicated to treatment efficacy, safety and quality of life has just been implemented and is available online in the new version of the web system. The registry is fully enrolling: a total of 3201 patients have been enrolled so far: among them 288 with CAPS, 425 with CRMO, 905 with FMF, 192 with MKD, 650 with PFAPA and 271 with TRAPS. New papers have been recently published (see list on next page). Other publications are ongoing: first paper on PAPA and CRMO, new evidence based PFAPA criteria, validation of the recently published classification criteria for monogenic periodic fevers.

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, 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 ensure the patient perspective is put forward, a dedicated survey has been implemented online in 14 different languages.

Thanks to the data gathered through SHARE, PRINTO is updating the website for families. The site will offer scientific information regarding the paediatric 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 translating the disease information in all languages. The new website will have customized illustrations, dynamic features and it will be accessible by various devices, such as smartphones and tablets. You can have a look at the draft of the new website for families at the following link: www.printo.it/pediatric-TEST/home

(PI Prof Nico Wulffraat, EU project number 2011 1202).

The Abirisk project

This project (Anti-Biopharmaceutical Immunization: Prediction and Analysis of Clinical Relevance to Minimize the Risk, www.abirisk.eu, funded by Innovative Medicines Initiative, Grant Agreement nr. 115303) aims to provide an integrated approach to investigate anti-drug antibody formation in JIA treated with biopharmaceuticals. The Abirisk project is conducted by PRINTO as a Pharmachild registry substudy (same case report forms) and the goal is to collect, besides clinical information, biosamples (Serum and RNA) of 200 children with JIA, newly treated with adalimumab, etanercept or tocilizumab (both naïve or after failure with other biologics) at start of therapy and follow-up, for a total of 6 study visits. As of today, 32 patients have been enrolled in 4 different countries.

The MAS criteria study

The MAS Classification Criteria Study is a multinational project, which is aimed at developing and validating a new set of classification criteria for systemic JIA-associated MAS. The data obtained in the data collection part of the project have been published in 3 papers (Davì S at al. Arthritis Rheumatol. 66:2871-2880; Minoia F et al. Arthritis Rheumatol. 66:3160-3169; Minoia F et al. J. Rheumatol 2015 Apr 15 [Epub ahead of print]). The new classification criteria have been established in the International Consensus Conference on MAS Classification Criteria, which was held in Genoa, Italy on March 21-22, 2014. The paper that presents the new criteria has been submitted for publication. The last step of the project, which is planned to be started shortly, will lead to the validation of the new classification criteria in a prospective cohort of MAS patients. In the last year, the collaboration with the Histiocyte Society has led to start a new project aimed at comparing the features of the 362 patients with sJIA-associated MAS collected in the data collection part of the project with a large sample of patients with familial hemophagocyitic lymphohistiocytosis. The results of this study will be presented as poster at EULAR 2015.

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 has been identified since last year. The analysis of the data has been completed and the first paper accepted by Lancet. We would like to continue to follow-up all patients enrolled in order to have for each 5-10 years of follow-up data.

The MYPAN trial in Childhood Polyarteritis Nodosa (PAN)

An Open Label Randomised Controlled Trial of Mycophenolate Mofetil (MMF) Versus Cyclophosphamide (CYC) for the Induction of Remission of childhood PAN sponsored by University College London and coordinated by the Children Hospital in Liverpool and PRINTO is underway. MYPAN will investigate the comparative efficacy and safety of MMF (experimental treatment) vs CYC (standard treatment) for induction of remission of childhood PAN. This will be the first ever randomized trial for childhood PAN. As of today 35 centres have shown interest in participating in MYPAN and we are currently working to start the submission procedures to the regulatory authorities and ethics committees in these centres (PI Dr P. Brogan).

Membership

In 2015, PRINTO has reached 1318 effective members in 516 centres from 83 countries.

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**

Recent PRINTO publications

For reprints go to www.printo.it and click on "papers".

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