THU0517

THE LONGITUDINAL EUROFEVER PROJECT: AN UPDATE ON ENROLLMENT

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Background: In 2008 the Paediatric Rheumatology European Society (PReS) promoted an International Project for the study of Autoinflammatory Diseases (AIDs) named Eurofever, whose main purpose is to create a web-based registry for the collection of information in AIDs patients.

Objectives: To implement the Registry with the new recently described AIDs and to increase the collection of longitudinal data.

Methods: The data were extracted from the Eurofever registry, which is hosted in the PRINTO website (<http://www.printo.it>). From February 2015 we started the longitudinal collection of follow-up data with particular focus on treatment, modification of the clinical picture, onset of complication/adverse events. We have enrolled patients included in the registry up to 28 September 2018.

Results: Up to date 4175 patients have been enrolled (3843 of them with complete demographic information, 1903 M e 1940 F) from 62 countries. For 3356 (87%) patients also clinical data from onset to diagnosis, collected during the first visit performed at referred pediatric or adult center, are available. For each disease the number of enrolled patients is: FMF 1086 pts (951 with complete clinical data); TRAPS 273 pts (256 complete); CAPS 298 pts (279 complete); MKD 205 pts (190 complete); Blau's disease 49 pts (26 complete); PAPA 42 pts (41 complete); NLRP12 mediated periodic fever 13 pts (11 complete); DADA2 14 pts (9 complete); DIRA 3 pts (all complete); SAVI 3 pts (all complete); CANDLE 1 pt (complete) and Majeed 4 pts (all complete). Among multifactorial autoinflammatory diseases: PFAPA 676 pts (551 complete); CNO 581 pts (540 complete); Behcet 214 pts (186 complete), undefined periodic fever 368 pts (292 complete) and Schnitzler 13 pts (all complete). The median onset age is 4 years (range 1 month – 75 years), the median diagnosis age is 8 years (range 1 month – 78 years). Most of patients (3509, 91%) presented disease onset during pediatric age (<16 years), 334 (9%) during adult age (81 FMF, 31 CAPS, 53 TRAPS, 40 CRMO, 12 Schnitzler syndrome e 90 unknown fever). 405 of 3509 (12%) patients with pediatric onset received diagnosis during adult age. The median diagnostic delay is 5 years; diseases with longer diagnostic delay are: NLRP12 (24 years, range 4-76), CAPS (15 years, range 0-77), PAPA (14 years, range 2-57), TRAPS (12 years, range 0-77). 396 patients have been treated with at least one biologic drug, 1031 with DMARDs, 427 with systemic steroid and 686 with others drugs. The most frequent diseases treated with biologic drugs are: CAPS (38%), multifactorial diseases (22%), TRAPS (14%), MKD (11%), rare monogenic (8%: 1 CANDLE, 2 DIRA, 2 NALP12, 3 Majeed, 8 DADA2 and 14 PAPA), and FMF (7%). Since February 2015, longitudinal visits have been inserted for 477 (12%) patients, with detailed data on treatment and safety.

Conclusion: The enrollment in Eurofever Registry is still ongoing. The analysis of data will improve our knowledge both on the natural history of the single disease and on the efficacy/safety of treatment commonly used in the clinical practice.

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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., Nicolino Ruperto Grant/research support from: The Gaslini Hospital, where NR works as full-time public employee, has received contributions (> 10.000 USD each) from the following industries in the last 3 years: BMS, Eli-Lilly, GlaxoSmithKline, F Hoffmann-La Roche, Janssen, Novartis, Pfizer, Sobi. This funding has been reinvested for the research activities of the hospital in a fully independent manner, without any commitment with third parties., Consultant for: Received honoraria for consultancies or speaker bureaus (< 10.000 USD each) from the following pharmaceutical companies in the past 3 years: Ablynx, AbbVie, Astrazeneca-Medimmune, Biogen, Boehringer, Bristol-Myers Squibb, Eli-Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, SanofiServier, Sinergie, Sobi and Takeda., Speakers bureau: Received honoraria for consultancies or speaker bureaus (< 10.000 USD each) from the following pharmaceutical companies in the past 3 years: Ablynx, AbbVie, Astrazeneca-Medimmune, Biogen, Boehringer, Bristol-Myers Squibb, Eli-Lilly, EMD Serono, GlaxoSmithKline, Hoffmann-La Roche, Janssen, Merck, Novartis, Pfizer, R-Pharma, SanofiServier, Sinergie, Sobi and Takeda., Marco Gattorno Grant/research support from: MG has received unrestricted grants from Sobi and Novartis

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EFFICACY AND SAFETY OF ETANERCEPT IN JUVENILE IDIOPATHIC ARTHRITIS – 18 YEAR EXPERIENCE USING DATA OF THE BIKER REGISTRY

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Background: Since 2000, Etanercept is approved for polyarticular juvenile idiopathic arthritis (JIA). Subsequently, approval has been extended to additional JIA categories.

Objectives: To describe efficacy and safety of Etanercept in clinical practice in JIA patients in comparison to a biologic-naïve JIA cohort using the German Biologics registry (BiKEr).

Methods: Baseline demographics and disease activity parameters were documented. Efficacy was determined using the JADAS10. Safety assessments were based on adverse events reports (AE) processed according to MedDRA.

Results: Until October 1, 2018, 2645 JIA patients treated with Etanercept were registered, representing 5820 patient-years (PY) of exposure. The total observation time (from date of first dose until last follow-up, censored, if another biologic was started) was calculated as 8080 PY.

In general, the cohort treated with Etanercept had experienced disease duration of 4.2±/3.7 years (mean±/SD). 2303 patients (87%) were pre-treated with methotrexate, 100 with alternative biologics. Concomitantly, 1822 patients (69%) received methotrexate, 2098 (79%) NSAIDs, 949 (36%) systemic corticosteroids as well as further drugs in lower numbers. In the control cohort, 1517 biologic naïve JIA patients started methotrexate.

At last follow-up, 68%/62%/50%/34% of patients reached JIAACR30/50/70/90 criteria. The median (IQR1-3) JADAS10 score decreased from 15.0 (10-20.4) at baseline to 3.5 (0.7-10.1). 60% of patients achieved a JADAS defined minimal disease activity, 40% reached a JADAS remission.

1924 AE were observed (33.1/100PY [95% CI 31.6-34.6]). 221 qualified as SAE (3.8/100PY [3.3-4.3]). These figures were compared to 1292 AE reported during 3714PY in the control cohort (34.8/year [32.9-36.8]) and 52 SAE (1.4/100PY [1.1-1.8]).

The most frequent SAE in the Etanercept cohort were grouped in the MedDRA SOC infections and infestations (n=54) followed by musculoskeletal and connective tissue disorders (n=27), eye disorders (n=21), gastrointestinal disorders (n=17) and nervous system disorders (n=15). There were 5 deaths. Serious infections were of bacterial and viral origin and mostly occurred once only.