pediatric rheumatology european society

Paediatric Vasculitis Working Party

Chair: Marija Jelusic
Secretary: Teresa Giani
Lead of education and training: Ezgi Deniz Batu
Lead of clinical care: Judith Sanchez Manubens
Lead of science and research: Mario Sestan
EMERGE representative: Caterina Matucci Cerinic

October 16, 2023

To all Vasculitis Working Party members

Dear Colleagues,

Behind us is an active and project-rich year in which PReS Vasculitis Working Party members worked hard and thus we finished and published 4 projects, there are updates of 7 ongoing projects and we also have the announcement of 7 new ones.

So, please kindly find a short report of our Vasculitis Working Party meeting that we had on September 28th, from 15:00 to 16:15, during PReS 2023 Congress in Rotterdam.

1. ELECTIONS WITHIN THE WP

We have elected a new representatives for the following leadership positions for the period from 2023 to 2027: Lead of education and training: Ezgi Deniz Batu (Ankara, Turkey); Lead of clinical care: Judith Sanchez Manubens (Barcelona, Spain); Lead of science and research: Mario Sestan (Zagreb, Croatia) and EMERGE representative: Caterina Matucci Cerinic (Genoa, Italy)

Currently, the Core team of our Vasculitis WP has the following representatives:
1. Chair: Marija Jelusic (Zagreb, Croatia) (2021 -2025)
2. Secretary: Teresa Giani (Florence, Italy) (2021-2025)
3. Lead of education and training: Ezgi Deniz Batu (Ankara, Turkey) (2023-2027)
4. Lead of clinical care: Judith Sanchez Manubens (Barcelona, Spain) (2023-2027)
5. Lead of science and research: Mario Sestan (Zagreb, Croatia) (2023-2027)
6. EMERGE representative: Caterina Matucci Cerinic (Genoa, Italy) (2023-2027)

Our PReS Vasculitis WP mailing list currently contains 159 members.

2. SCIENCE AND RESEARCH ACIVITIES

FINISHED and PUBLISHED RESEARCH PROJECTS

1. Marija Jelusic, Nastasia Kifer ***"Histological predictors of outcome in patients with Henoch-Schönlein purpura / IgA vasculitis and nephritis"***
- Semiquantitative classification (SQC) proved to be the best, followed by Oxford classification; cellular crescents in SQC, mesangial hypercellularity and tubular atrophy in Oxford have shown significant contributions in outcome prediction

Kifer N, Bulimbasic S, Sestan M, Held M, Kifer D, Srsen S, Gudelj Gracanin A, Heshin-Bekenstein M, Giani T, Cimaz R†, Gagro A, Frkovic M, Coric M, Jelusic M. SQC and Oxford classifications predict poor renal outcome better than ISKDC and Haas in patients with IgAV nephritis: a multicenter study. J Nephrol. 2023;36(2):441-9.

2. Ezgi Deniz Batu, Seza Ozen *"COVID-19 associated pediatric vasculitis study"*

- COVID-19 associated IgAV had more severe disease course than non-COVID-19 associated IgAV: more prevalent renal involvement and less prevalent complete recovery.

Ezgi Deniz Batu, Seher Sener, Gulcan Ozomay Baykal, Elif Arslanoglu Aydin, Semanur Özdel, Alenka Gagro, Fatma Gül Demirkan, Esra Esen, Nilufer Akpınar Tekgöz, Kubra Ozturk, Olga Vougiouka, H. Emine Sonmez, Merav Heshin-Bekenstein, Maria Cristina Maggio, Ummusen Kaya Akca, Marija Jelusic, Aysenur Pac Kısaarslan, Banu Çelikel Acar, Nuray Aktay Ayaz, Betül Sözeri, Seza Özen. „The characteristics of patients with COVID-19-associated pediatric vasculitis: An international, multicenter study. Arthritis Rheumatol. 2023; 75(4):499-506.

3. Mario Sestan, Marija Jelusic *"Clinical features, treatment and outcome of patients with severe cutaneous manifestations in IgA vasculitis - multicenter study"*

- IgAV patients with severe skin manifestations more frequently developed IgAV nephritis, had worse outcome of renal disease and were more frequently treated with systemic glucocorticoids and were more likely to develop the most severe gastrointestinal manifestations

Sestan M, Kifer N, Sozeri B, Demir F, Ulu K, Silva Clovis, Campos Reinan T, Batu ED, Koker O, Sapina M, Srsen S, Held M, Gagro A, Fonseca A, Rodrigues M, Rigante D, Filocamo G, Baldo F, Heshin-Bekenstein M, Giani T, Kataja J, Frkovic M, Ruperto N, Özen S, Jelusic M. Clinical features, treatment and outcome of patients with severe cutaneous manifestations in IgA vasculitis: multicenter international study. Semin Arthritis Rheum. 2023;61:152209.

4. Ummusen Kaya, Seza Ozen *"Comparison of EULAR/PReS/PRINTO Ankara 2008 and adult classification criteria in patients with granulomatous polyangiitis (GPA)"*

Manuscript under revision

Ummusen Kaya Akca, Ezgi Deniz Batu, Marija Jelusic, Marta Calatroni, Reima Bakry, Marijan Frkovic, Nikol Vinšová, Reinan T. Campos, AnnaCarin Horne, Sengul Caglayan, Augusto Vaglio, Gabriella Moroni, Giacomo Emmi, Gian Marco Ghiggeri, Oya Koker, Renato Alberto Sinico, Susan Kim, Alenka Gagro, Caterina Matucci Cerinic, Elif Çomak, Zahide Ekici Tekin, Elif Arslanoglu Aydin, Merav Heshin-Bekenstein, Banu Celikel Acar, Marco Gattorno, Sema Akman, Betul Sozeri, Karin Palmblad, Sulaiman M Al-Mayouf, Clovis Artur Silva, Pavla Doležalová, Peter A Merkel, Seza Ozen on behalf of Vasculitis Working Party of the Pediatric Rheumatology European Society (PReS) ''Comparison of EULAR/PRINTO/PReS endorsed Ankara 2008 and 2022 ACR/EULAR classification criteria in childhood granulomatosis with polyangiitis''

- In March 2023 Vasculitis WP applied for the PRES-PRINTO RESEARCH GRANT with two projects: Definition of disease status, outcomes and follow-up in IgA vasculitis: a PReS/PRINTO survey and consensus conference *(Marija Jelusic and Mario Sestan)* and Clusters in pediatric Behçet's disease" (*Ümmüşen Kaya Akca, Ezgi Deniz Batu, Seza Özen).* The expert committee chose to support JIA WP project.

-Presented the PReS Vasculitis Working Party (Marija Jelusic) at the CARRA Annual Scientific Meeting, 26 - 29 March 2023, New Orleans, USA

ONGOING RESEARCH PROJECTS – UPDATE

1. Reima A Bakry. ''Childhood Cogan syndrome: Clinical manifestation, treatment and outcome: International multicentre study''

Objectives of the study: to report the spectrum and clinical manifestations of childhood Cogan syndrome, to highlight the current treatment strategies and propose guidelines for treatment, to highlight the long-term outcome of childhood Cogan syndrome
Patients and methods: cross-sectional, multicenter study
inclusion criteria: patients who have been diagnosed with Cogan syndrome, patients with suspected Cogan syndrome, younger than 18 years
Project progress: till now included 14 patients from 10 contributing centers,
the estimated cohort is 50 patients.

If you are interested in participating, please contact PIs: Reima A. Bakry (reimabakry@hotmail.com)

2. Özlem Akgün, Fatma Gül Demirkan, Nuray Aktay Ayaz. ''Safety and efficacy of biologic therapies in refractory/severe pediatric Behçet's disease: an International cohort''
Objectives of the study: to evaluate the characteristics of biological treatment regimens, their efficacy, side effects and outcomes on any clinical involvement of pediatric BD and to provide a comprehensive overview of the use of biologics in pediatric-onset BD, for which there is no worldwide consensus
Patients and methods: retrospective, observational, multicenter and international study
Inclusion criteria: patients under 18 years of age at diagnosis, who have used biologic drugs at some point in their treatment
Project progress: data has been received from 17 centers, comprising a total of 108 patients; it was observed that anti-TNFs were the first choice biological agent in 90.8% of the patients
*For more information please contact PIs: Özlem Akgün (*drozlemakgun@hotmail.com), *Fatma Gül Demirkan*(fatmagy@gmail.com) *and Nuray Aktay Ayaz* (nurayaktay@gmail.com)

3. Şengül Çağlayan, Betül Sözeri. ''The effect of the initial hyperinflammatory condition on the outcome of IgA vasculitis''
Aims of the study: to evaluate initial inflammatory parameters: pentraxin 3, serum galectin, NLR, CRP/albumin, SII and SIRI
Patients and methods: patients <18 years, all patients must meet the 2008 Ankara PReS/EULAR/PRINTO IgAV classification criteria, only new IgAV diagnosed between February 2023 and February 2024
- 6 ml serum should be collected and stored at -80°C before beginning treatment, and shipment will be organized by the principal investigator at the end of the project (February 2024)
- the target number of patients: 200
- number of control patients: 50
Project progress: 114 patient samples have been collected from Turkey

*If you are interested in participating, please contact PIs: Şengül Çağlayan (*sengulturkercaglayan@gmail.com*) and Betül Sözeri (*drbetulsozeri@gmail.com)

4. Nuray Aktay Ayaz, Figen Çakmak: ''The Nailfold Videocapillaroscopy in Pediatric Behçet's Disease'' -
Aims of the study: to evaluate the microvascular involvement in juvenile Behçet's disease, to find out the correlation between clinical findings and microvascular involvement, to find out the correlation between Behçet's disease activity scores and microvascular involvement, to compare nailfold capillaroscopic alterations and nailfold videocapillaroscopy scores of patients diagnosed with Behçet's disease with healthy volunteers
Project progress: the enrollment of the study was closed, it was included 61 patients from 7 centers,
Preliminary results: neoangiogenesis was found to be significantly more common in the NVC evaluation of patients with lower hemoglobin values at the time of diagnosis (p=0.014)
- Thanks to all researchers and Vasculitis working group who participated and contributed
For more information please contact PIs: Nuray Aktay Ayaz (nurayaktay@gmail.com) and Figen Çakmak (figenatamancakmak@gmail.com)

5. David Cabral: ''PedVas initiative projects: to comparatively evaluate CARRA-endorsed consensus tratment plan options for pAAV in PedVas registry''
- how to participate:
- join PedVas (IRB +/- contract) - contact Else at ebosman@cw.bc.ca; dcabral@cw.bc.ca;
- commit to using specified regimens for remission-induction and remission-maintenance treatment of moderate-severe GPA/MPA: limited dosing and duration of IV and oral prednisone; train in use of PVAS (with testing) if not proficient using on-line training module; go to: https://redcap.link/PVAStraining to leave your information to create an account; capture damage with pVDI PLUS additional specific drug toxicities; collect clinical data +/- biological samples at specified times according to schedule

If you are interested in participating, please contact else.bosman@cw.bc.ca or dcabral@cw.bc.ca

NEW RESEARCH PROJECTS (PROPOSALS)

1. Seza Ozen, Muserref Kasap Cuceoglu: ''Pediatric Takayasu arteritis: a multicenter retrospective cohort study''
- retrospective multicenter study
- data will be compiled in the form of an Excel File
Aims of the study: to evaluate childhood-onset TAK patients in all centers around the world (age of disease onset, clinical course, laboratory data, imaging findings, concomitant diseases, medical and surgical treatments) will contribute to the literature; Determination of the general characteristics of pediatric TAK patients will increase the awareness of physicians on this issue; Assessing the treatment response
Inclusion criteria: all pediatric TAK patients under 18 years of age
- till now 64 pediatric TAK patients data collected from Turkey

*If you are interested in participating, please contact PIs: Muserref Kasap Cuceoglu (*drmuserref@gmail.com*) and Seza Ozen (*sezaozen@gmail.com*)*

2. Marija Jelusic, Mario Sestan: ''Comparison of different scoring systems for assessment of disease activity in childhood Takayasu arteritis (PRES-CARRAinitiative)'
Objectives of the study: to assess the performance of the PVAS in pediatric patients with Takayasu arteritis (TAK) and compare it with ITAS2010 and EULAR criteria for active large vessel vasculitis
Patients and methods: pediatric patients with TAK diagnosed by EULAR/PRINTO/PReS classification criteria for childhood-onset TAK and control group consisting of patients with other primary systemic vasculitis or diseases that mimic vasculitis; estimated number: ~ 80 patients with TAK and ~ 100 controls
- a retrospective evaluation of the data of patients - demographic characteristics, detailed clinical symptoms and organ involvements, laboratory features, angiography features and treatment
- 2 points: diagnosis and 12-month follow-up
- data entry in form of RedCap or Excel Spreadsheet

If you are interested in participating, please contact: Marija Jelusic (marija.jelusic.drazic@gmail.com)

3. Isabelle Koné-Paut: ''A retrospective observational study of the use of anakinra for the treatment of Kawasaki disease''
Objectives of the study: to identify Kawasaki disease (KD) patients treated with anakinra (demographics, KD characteristics; i.e. cardiac involvement, MAS); to analyze when and how anakinra was used (reasons for use, concomittant treatments, delay to treatment, doses, duration); to evaluate the efficacy of anakinra on KD clinical signs, CRP, and essentially coronary Z scores and cardiac function, to assess any side effects related to treatment with anakinra

Inclusion criteria: patients with KD diagnosis according to AHA criteria for either complete or incomplete KD treated with anakinra
Project progress: 24 patients already identified with KD and anakinra treatment in France and ethical procedures have been agreed already at least for the already participating centers 87 centers from 11 countries (France, Germany, Switzerland, Belgium, Netherlands, Morocco,Tunisia, Poland, Austria, Armenia, Greece)

If you are interested in participating, please contact PI: Isabelle Koné-Paut: (isabelle.kone-paut@aphp.fr) or Perrine Dusser (perrine.dusser@aphp.fr)

4. Tamás Constantin: ''Addressing diagnostic and treatment challenges in pediatric primary angiitis of the central nervous system (cPACNS)''
Objectives of the study: to establish an international multidisciplinary working group that brings together neurologists, radiologists, and rheumatologists from various institutions. Through this global collaboration, the goal is to improve patient outcomes by refining diagnostic precision and evolving treatment paradigms for cPACNS, achieved through systematic literature research and consensus building

If you are interested in participating, please contact PI Tamás Constantin: (tamas.constantin@gmail.com)

5. Teresa Giani, Francesca Minoia: Macrophage activation syndrome in Kawasaki disease: features, treatment, outcome and predicting factors (Collaboration with MAS/sJIA WP)
Aims of the study: to evaluate epidemiological, clinical and laboratory characteristics, managment and outcomes in KD/MAS and to identify potential risk factors and diagnostic criteria for MAS in KD
Patients and methods: inclusion criteria: patients with KD and MAS at the age of 4 weeks to 17 years
control group: KD age- and gender-related KD without MAS
collection data: demographic variables, clinical signs, laboratory values, SARS-CoV-2 information, additional data (genetics, histology), treatment, outcome
Target numbers: 50 KD/MAS patients, 50 KD first line-resistant patients, 150 first line-responsive KD patients
- data will be compiled in the form of an Excel File

*If you are interested in participating, please contact PI Teresa Gi**ani: (teresa.giani@gmail.com)** or Francesca Minoia **(francesca.minoia@policlinico.mi.it)***

6. Şengül Çağlayan, Betül Sözeri: ''Determining the relationship between DADA-2 clinical findings and capillaroscopy''
Aim of the study: to delineate the characteristics of nailfold capillary changes in individuals with DADA2 and explore potential correlations with the clinical features of the disease
Patients and methods: the study will involve patients who have been confirmed to have ADA2 deficiency based on ADA2 enzyme activity measurements and/or genetic testing results
Inclusion criteria: patients with DADA-2 diagnosis

If you are interested in participating, please contact PI Şengül Çağlayan (sengulturkercaglayan@gmail.com) or Betül Sözeri (drbetulsozeri@gmail.com)

7. Sara Stern (Chair of the CARRA Childhood Sjögren's disease workgroup): ''International Sjögren's disease registry''
CHildhood Onset Sjögren disease Outcomes Network (CHOSEN) wants to establish prospective international registry of childhood Sjögren disease. They would like to include children with Sjögren disease based off criteria, but also children with recurrent or persistent parotitis without an underlying diagnosis, elevated SS-A/Ro or SS-B, subjects clinically suspected of having Sjögren's disease in evolution for another reason.

However, they need help and ask for the participation of other centers in terms of including patients in the registry, so that it will have an international character.

If you are interested in participating, please contact Matthew L. Basiaga (basiaga.matthew@mayo.edu), Scott M. Lieberman (scott-lieberman@uiowa.edu) or Sara M. Stern (sara.stern@hsc.utah.edu)

8. Muthana Al Obaidi: Proposal for the establishment of the PReS Sjogren Syndrome Working Party (PReS Sjogren sy WP).

The objectives of the newly proposed Sjögren syndrome WP are to promote excellence in clinical care, research, training, and education related to Sjogren's syndrome with childhood onset and to address the unique challenges associated with Sjogren's syndrome in children, which include: under-recognition and under-diagnosis of autoimmune rheumatic diseases, limited understanding of the natural history of the disease, delayed diagnosis due to children less frequently reporting dryness, lack of validated classification criteria, differences in clinical presentation between childhood and adult-onset Sjogren's syndrome.
Everyone with the interest for Sjögren syndrome WP, please contact Muthana Al Obaidi (*Muthana.AlObaidi@gosh.nhs.uk*) or Sezgin Sahin (sezgin@istanbul.edu.tr).

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3. EDUCATIONAL AND TRAINING ACTIVITIES

1st International Kawasaki disease Registry and EUROKiDs Joint Meeting, Bologna, November 2-4, 2023, Italy - hybrid conference

contact: kawasaki.disease23@gmail.com, www.ikdm.info

PReS Knowledge Base Exam - open to trainees, and to each practicing pediatric rheumatologist from around the world, project approved by the Council and endorsed by the General Assembly

PReS School Webinars - 3rd Tuesday each month, 12:00 - 13:00 CET

PReS School webinars will be held 3rd Tuesday each month, from 12:00 to 13:00 CET, 1 hour duration - 2 moderators (1 senior, 1 EMERGE), and 2 speakers (20 minutes presentation each), organized by the Working Party. The goal is to review knowledge in pediatric rheumatology from medium to advanced level, at the same time that we build community between PReS members across the world.
PReS School webinars will start on October 24, 2023, from 12:00 to 13:00 CET and the organizer of the first webinar will be Vaccination Working Party.

CliPS Project: collection of real life clinical practice strategies from worldwide physicians through the questionnaire of the following medical conditions: lupus nephritis, Kawasaki disease, IgA vasculitis, monogenic autoinflammatory diseases, PFAPA, SURF, sJIA/Still's disease
contact: info@jircohorte.ch, www.jircohorte.org/clips

Dear PReS Vasculitis Working party members, please feel free to join in some ongoing and new research projects.

Sincerely yours,

Marija Jelusic, Chair of the Vasculitis WP