Vasculitis Working Party

PRES 2023
29th European Paediatric Rheumatology Congress
28 SEPTEMBER – 1 OCTOBER 2023
ROTTERDAM, THE NETHERLANDS

Vasculitis Working Party 2023

Marija Jelusic

Chair, Vasculitis Working Party of the PReS
Chair, Education and Training Committee of the PReS

DEPARTMENT OF PAEDIATRICS
DIVISION OF PAEDIATRIC RHEUMATOLOGY AND IMMUNOLOGY
Refferal Centre for Paediatric and Adolescent Rheumatology Republic of Croatia,
UNIVERSITY HOSPITAL CENTRE ZAGREB
UNIVERSITY OF ZAGREB SCHOOL OF MEDICINE, CROATIA

Vasculitis Working Party: 2023

MEETING AGENDA

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Marija Jelusic: Welcome and introduction, Progress of the Vasculitis Working Party, 2022-2023

ACTIVITIES IN SCIENCE & RESEARCH (ongoing/planned):

ONGOING PROJECTS- UPDATE:

1. Reima A Bakry. **Childhood Cogan Syndrome: Clinical manifestation, Treatment and Outcome: International multicentre study**
2. Özlem Akgün, Nuray Aktay Ayaz. **Safety and Efficacy of Biologic Therapies in Refractory/Severe Pediatric Behçet's Disease: An International Cohort**
3. Nuray Aktay Ayaz, Figen Çakmak. **The Nailfold Videocapillaroscopy in Pediatric Behçet's Disease**
4. Şengül Çağlayan, Betül Sözeri.**The Effect of the Initial Hyperinflammatory Condition on The Outcome of IgA Vasculitis**
5. David Cabral**: PedVas initiative projects: To comparatively evaluate CARRA-endorsed Consensus Treatment Plan options for pAAV in PedVas registry**

NEW PROJECTS (PROPOSALS):

1. Seza Ozen, Muserref Kasap Cuceoglu: **Pediatric Takayasu arteritis: a multicenter retrospective cohort study**
2. Marija Jelusic, Mario Sestan: **Comparison of different scoring systems for assessment of disease activity in childhood Takayasu arteritis (PRES – CARRA project)**
3. Isabelle Koné-Paut: **A retrospective observational study of the use of anakinra for the treatment of Kawasaki disease**
4. Tamás Constantin: **Addressing Diagnostic and Treatment Challenges in Pediatric Primary Angiitis of the Central Nervous System**
5. Teresa Giani: **Macrophage Activation Syndrome in Kawasaki disease: features, treatment, outcomes and prediciting factors** (Collaboration with MAS/sJIA WP)
6. Şengül Çağlayan, Betül Sözeri: **Capillaroscopy in ADA-2 deficiency**
7. Sara Stern/USA (Chair of the CARRA Childhood Sjögren's Disease Workgroup): **International Sjögren's Disease registry**

Vasculitis Working Party: 2023

MEETING AGENDA

EDUCATIONAL ACTIVITIES:

Teresa Giani: **1st International Kawasaki Disease Registry & EUROKiDs Joint Meeting, Bologna, November 2-4, 2023, Italy; Update - PReS Knowledge Base Exam**

Marija Jelusic: **PReS School webinars**

ELECTIONS :1. Lead of the science and research , 2. Lead of the training and education, 3. Lead of the clinical care, 4. EMERGE representative

Vasculitis Working Party: 2023

Chair: Marija Jelusic

Secretary: Teresa Giani

MEETING AGENDA

Marija Jelusic (Chair): Progress of the Vasculitis WP (2022-2023)

Vasculitis Working Party

- **1. PReS VASCULITIS WP Core team**
- 2. **SCIENCE AND RESEARCH ACTIVITIES**
- **3. EDUCATIONAL AND TRAINING ACTIVITIES**

Vasculitis Working Party Core Team

*Currently PReS Vasculitis WP mailing list contains **159** members*

Lead of clinical care: **Neil Martin** (Glasgow, UK), 2019-2023

EMERGE representative: **Mario Sestan** (Zagreb, Croatia), 2019-2023

Lead of science and research: **Ezgi Deniz Batu** (Ankara, Turkey) 2019-2023

Chair: **Marija Jelusic** (Zagreb, Croatia) 2021-2025

Secretary and Lead of education and training: **Teresa Giani** (Florence, Italy), 2021-2025

ELECTIONS (2023-2027)

Vasculitis Working Party: 2023

- **Lead of the training and education**: EZGI DENIZ BATU, Ankara, Turkey
  co-Lead : REIMA BAKRY, Jeddah, Saudi Arabia

- **Lead of the science and research**: MARIO SESTAN, Zagreb, Croatia

- **Lead of the clinical care**: JUDITH SANCHEZ MANUBENS, Barcelona, Spain

- **EMERGE representative**: CATERINA MATUCCI CERINIC, Genoa, Italy

VASCULITIS WP

SCIENCE AND RESEARCH ACTIVITIES

FINISHED and PUBLISHED RESEARCH PROJECTS (2021 – September 2023)

1. Histological predictors of outcome in patients with Henoch-Schonlein purpura / IgA vasculitis and nephritis"(2019-2021), *PI: Marija Jelusic and Nastasia Kifer (Journal of Nephrology)*

2. **COVID-19 associated pediatric vasculitis study,** (2022-2022) *PI: Ezgi Deniz Batu and Seza Ozen (Arthritis and Rheumatology)*

3. **Clinical features, treatment and outcome of patients with severe cutaneous manifestations in IgA vasculitis - multicenter study ,** (June 2021 – 2022), *PI: Mario Sestan and Marija Jelusic, (Seminars in Arthritis and Rheumatism)*

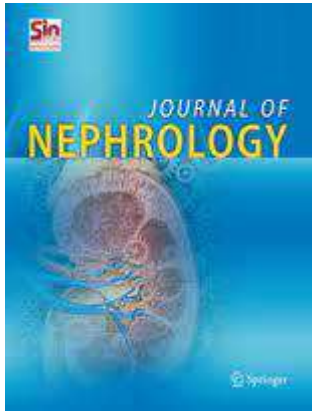
4. **Comparison of EULAR/PReS/PRINTO Ankara 2008 and adult classification criteria in patients with granulomatous polyangiitis (GPA)** *PIs – Ummusen Kaya and Seza Ozen (under revision)*

1. PROJECT: “Histological predictors of outcome in patients with Henoch-Schonlein purpura / IgA vasculitis and nephritis„ (*2018-2021)*

„Semiquantitative classification (SQC) and Oxford classifications predict poor renal outcome better than The International Study of Kidney Disease in Children (ISKDC) and Haas in patients with IgAV nephritis: a multicenter study”

Nastasia Kifer, Stela Bulimbasic, Mario Sestan, Martina Held, Domagoj Kifer, Sasa Srsen, Ana Gudelj Gracanin, Merav Heshin-Bekenstein, Teresa Giani, Rolando Cimaz†, Alenka Gagro, Marijan Frković, Marijana Coric, Marija Jelusic

J Nephrol. 2023;36(2):441-449. doi: 10.1007/s40620-022-01509-4



- Collaboration between Vasculitis WP members from Croatia, Italy, Israel

2. PROJECT:

The characteristics of patients with COVID-19-associated pediatric vasculitis: An international, multicenter study

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

*On behalf of the PReS Vasculitis Working Party

Arthritis Rheumatol, 2023, doi:10.1002/art.42411

Seminars in Arthritis and Rheumatism
Volume 61, August 2023, 152209

Clinical features, treatment and outcome of pediatric patients with severe cutaneous manifestations in IgA vasculitis: Multicenter international study

Mario Sestan [a], Nastasia Kifer [a], Betul Sozeri [b], Ferhat Demir [b], Kadir Ulu [b], Clovis A. Silva [c], Reinan T. Campos [c], Ezgi Deniz Batu [d], Oya Koker [e], Matej Sapina [f], Sasa Srsen [g], Martina Held [a], Alenka Gagro [h], Adriana Rodrigues Fonseca [i], Marta Rodrigues [i], Donato Rigante [j], Giovanni Filocamo [k], Francesco Baldo [k], Merav Heshin-Bekenstein [l], Teresa Giani [m], Janne Kataja [n], Marijan Frkovic [a], Nicolino Ruperto [o], Seza Ozen [d], Marija Jelusic [a] for the Vasculitis Working Party of the Pediatric Rheumatology European Society (PReS)

Results: Patients with IgAV/HSP and severe skin manifestations had higher frequencies of severe gastrointestinal complications like hematochezia, massive bleeding and/or intussusception (29.3% vs. 14.8%, p<0.001). D-dimer concentrations were significantly higher in these patients (4.60 mg/L vs. 2.72 mg/L, p = 0.003) and they had more frequent need for treatment with systemic glucocorticoids (84.4% vs. 37.2%, p<0.001) in comparison with the control group. Further multivariate analysis showed that severe cutaneous changes were associated with higher risk of developing nephritis [OR=3.1 (95%CI 1.04–9.21), p = 0.042] and severe gastrointestinal complications [OR=3.65 (95%CI 1.08–12.37), p = 0.038].

Conclusion: Patients with IgAV/HSP and severe skin manifestations had a more severe clinical course and more frequently required glucocorticoids compared to classic IgAV/HSP patients.

4. PROJECT (2021 – 2023)

Comparison of EULAR/PRINTO/PRES Ankara 2008 and 2022 ACR/EULAR classification criteria in childhood granulomatosis with polyangiitis

Under revision

PI: Ummusen Kaya and Seza Ozen, Hacettepe Univeristy, Ankara, Turkey


CONGRATS
YOU'RE FANTASTIC!

congratulations!

Vasculitis Working Party:
2023

Applied for the PRES-PRINTO RESEARCH GRANT (March, 2023):

1. Definition of disease status, outcomes and follow-up in IgA vasculitis: a PReS/PRINTO survey and consensus conference *(PI: Marija Jelusic and Mario Sestan)*

2. Clusters in pediatric Behçet's disease" (*PI: Ümmüşen Kaya Akca, Ezgi Deniz Batu, Seza Özen)*

Working with other societies/ Working Parties

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

Working with other societies/ Working Parties

CARRA Chronic Childhood Vasculitis (CCV) Workgroup (AAV ex)

INTERNATIONAL TAK STUDY GROUP (PReS; CARRA, India ...)

CARRA Refractory Kawasaki Disease Workgroup (first introductory zoom meeting , June 9 2023)

ERN-RITA (Vasculitis Working Group, Mark Little)

EUVAS (project : IgA vasculitis in adults, since 2023)

MAS/sJIA WP (project proposal: Macrophage Activation Syndrome in Kawasaki disease: features, treatment, outcomes and prediciting factors. PI: Teresa Giani)

SCIENCE AND RESEARCH ACTIVITIES

ONGOING RESEARCH PROJECTS

Vasculitis Working Party

ONGOING RESEARCH PROJECTS

1. Childhood Cogan Syndrome: Clinical manifestation, Treatment and Outcome: International multicentre study, *PI: Reima A Bakry*

2. Safety and Efficacy of Biologic Therapies in Refractory/Severe Pediatric Behçet's Disease: An International Cohort, *PI: Özlem Akgün and Nuray Aktay Ayaz*

3. The Effect of the Initial Hyperinflammatory Condition on The Outcome of IgA Vasculitis
PI: Şengül Çağlayan and Betül Sözeri

4. The Nailfold Videocapillaroscopy in Pediatric Behçet's Disease", *PIs: Nuray Aktay Ayaz, Figen Çakmak*

5. **KD-CAAP study "Multi-centre, randomised, open-label, blinded endpoint assessed, trial of corticosteroids plus IVIG and aspirin, versus IVIG and aspirin for prevention of coronary artery aneurysms in Kawasaki disease,** *PIs-Despina Eleftheriou and Paul Brogan*

6. Comparison and performance of Kobayashi and Kawanet IVIg resistance scores in Kawasaki disease (the KIWI study), *PI: Maria Vincenza Mastrolia and Vignesh Pandiarajan (PReS- PRINTO grant)*

7. To comparatively evaluate CARRA-endorsed Consensus Treatment Plan options for pAAV in PedVas registry, *PI: David Cabral*

Vasculitis Working Party

ONGOING RESEARCH PROJECTS - update

UPDATE:

1.Reima A Bakry. **Childhood Cogan Syndrome: Clinical manifestation, Treatment and Outcome: International multicentre study**

2.Özlem Akgün, Nuray Aktay Ayaz. **Safety and Efficacy of Biologic Therapies in Refractory/Severe Pediatric Behçet's Disease: An International Cohort**

3.Nuray Aktay Ayaz, Figen Çakmak. **The Nailfold Videocapillaroscopy in Pediatric Behçet's Disease**

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1. Childhood Cogan syndrome; clinical manifestation, treatment and outcome: A multicenter study for the PReS vasculitis working party

Reima A Bakry, MD

Consultant of Pediatrics & Pediatric Rheumatology

Maternity and Children Specialized Hospital

Sulaiman M Al-Mayouf, MD

Professor of Pediatric and Pediatric Rheumatology

King Faisal Hospital and Research Center-Riyadh

Introduction

- Cogan syndrome is a rare autoimmune vasculitis that falls into the variable-sized vasculitis category.
- There are around 250 cases of CS reported worldwide.
- The mean age at onset was 25 years (range: 5–63 years).

- In 1980, Haynes and colleagues proposed categorizing Cogan syndrome into two types:

  - Typical (non-syphilitic interstitial keratitis with vestibular involvement, similar to Ménière disease).

  - Atypical (involvement of the uvea, conjunctiva, sclera, optic disc, and retinal vessels) and audiovestibular symptoms do not resemble Ménière disease or appear before or after ocular symptoms for more than two years).

Objective

- 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.

Method

- This project is a cross-sectional, multicenter study.
- An invitation for participation was sent to all PReS vasculitis working party members.
- A comprehensive data sheet and ethical approval were sent to all members willing to contribute.
- The caring physician will be asked to evaluate the long-term assessment and outcome at the last follow-up visit, using the Pediatric Vasculitis Damage Index (PVDI).
- Upon completion of the clinical data sheet, it will be sent to the primary investigator for analysis.
- The estimated cohort is 50 patients.

Method

- **Inclusion criteria**
  - Patients who have been diagnosed with Cogan syndrome.
  - Patients with suspected Cogan syndrome.
  - Younger than 18 years old.

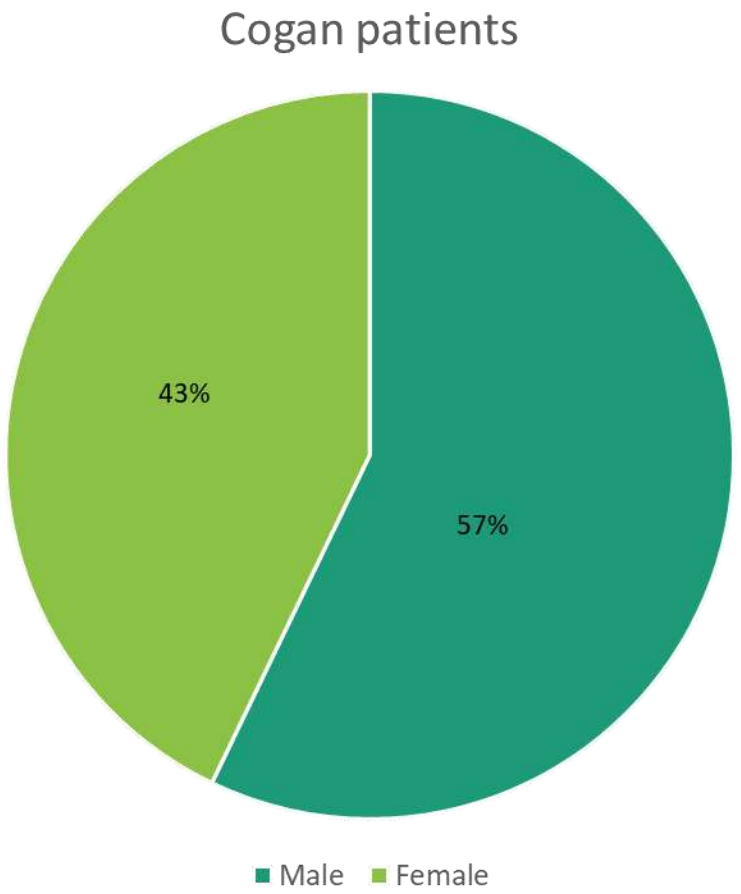
- **Exclusion criteria**
  - Patients with other systemic vasculitis.

Ethical Consideration

- All of the information gathered will be the result of routine medical procedures and will have been extracted from the patient's file.
- The confidentiality of the patients will be safeguarded, and all acquired data will be saved.
- No information identifying a specific person will be gathered for this study.
- Because the study protocol only involves the collection of routine patient data, formal submission of the research proposal to an ethics committee may not be necessary for most participating centers.
- Furthermore, we believe that consent is not required from the patient or parent.
- The study duration will be 12–18 months.

Preliminary Results (as of August 2023)

Total number : 14 patients



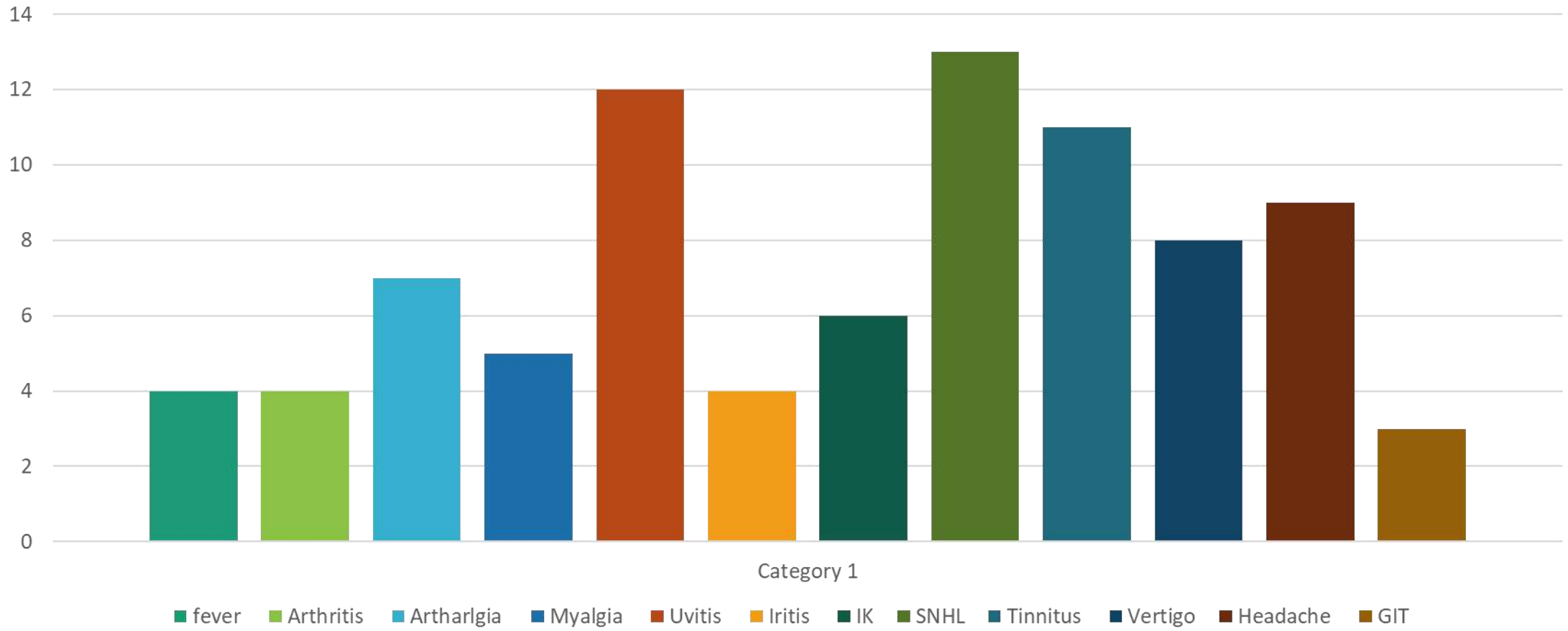
Contributing centers (till August 2023)

- Maternity and Children Specialized Hospital , Jeddah, Saudi Arabia (2 cases)
- Hacettepe University Hacettepe Üniversitesi, Ankara, Türkiye (2 cases)
- Royal Hospital for Children, Glasgow, United Kingdom (2 cases)
- University of Zagreb School of Medicine, Zagreb, Croatia (1 case)
- Leiter Zentrum für Pädiatrische Rheumatologie am Klinikum Stuttgart, Stuttgart, Germany (1 case)
- Tripoli Children Hospital , Tripoli, Libya (1 case)
- Al-Makassed Islamic Charitable Hospital, Jerusalem, Plastein (1 case)
- Fondazione Policlinico Universitario A. Gemelli IRCCS ,Università Cattolica Sacro Cuore, Rome, Italy (1 case) IT
- Hospital Universitari Parc Taulí Sabadell,Institut d'Investigació i Innovació I3PT, Universitat Autònoma de Barcelona, Barcelona, Spain (1 case)
- King Faisal Specialist Hospital and Research Center Riyadh, Saudi Arabia (2 cases)

Demographic Data

Mean Age of onset	11.2 y
Mean age of diagnosis	12 y
Mean disease duration	3.8 y
Mean follow-up duration	3.5 y

Clinical manifestation



Diagnostics :

- 50% has high inflammatory markers.
- Positive ANA was detected in 3 patient.
- C-P ANCA was positive in 1 patients.
- 4 patients had abnormal Echo findings.
- Abnormal Ear MRI findings in 3 patients.

Diagnostics

Test	Result n=14	Test	Result
Low Hgb	4 (28.5%)	+ve ANA	3 (21.4%)
High WBC	5 (35.7%)	+ve C-P ANCA	1 (.07%)
High plt	5 (35.7%)	High LFT	1 (.07%)
High ESR and CRP	7 (50%)	Abnormal ECHO	4 (28.5%)
Proteinuria	4 (28.5%)	Abnormal Brain MRI	4 (28.5%)

Treatment

Medication	Used or current use	Response
Steroids	13 (92.8%)	Only 2 pt had poor response
Mycophenolate Mofetile	1 (9%)	Improvement
Methotrexate	11 (78.5%)	7 improvement , 3 poor response, 1 S.E
Infliximab	6 (42.8%)	4 improvement , 2 poor response
Tocilizumab+ anti TNF	2 (14.2 %)	Improvement

Cochlear implantation was performed in 3 patients

PVDI

- 10 patients have hearing loss.
- 2 patients with visual impairment.
- 3 patients with valvular heart disease.

- 1 death was detected due to disease or treatment complications.

Invitation

I would like to invite all pediatric rheumatologists who have patients with Cogan syndrome to participate in this study.

Data collection forms and the study protocol will be emailed to all interested pediatric rheumatologists.

The manuscript will be written by me under the supervision of Prof.Soliman Almayouf

Please contact me at the following email address:

reimabakry@hotmail.com

Questions

Thank you

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2. Safety and Efficacy of Biologic Therapies in Refractory/Severe Pediatric Behçet's Disease: An International Cohort Study

Participating Centers

(Currently, 11 centers from Turkey, 3 from Italy, 2 from France, 1 from Croatia, and 1 from Sweden and Israel have stated that they want to participate in the study)

1) APHP, CHU de Bicêtre, Paediatric rheumatology and CEREMAIA, University of Paris Sud Saclay, Le Kremlin Bicêtre, France.
2) Rheumatology Unit, Meyer Children's University Hospital, Neurofarba Department, University of Florence, Firenze, **Italy**
3) Department of Internal Medicine and Clinical Immunology, Pitie Salpetriere Hospital, Paris France.
4) University of Zagreb School of Medicine, Division of Peadiatric Immunology, Rheumatology and Allergology, Zagreb, **Republic of Croatia**
5) Fondazione Policlinico Universitario Agostino Gemelli IRCCS Università Cattolica del Sacro Cuore, Rome, **Italy**
6) Astrid Lindgren Children's Hospital, Department of Paediatric Rheumatology, Karolinska, **Sweden**.
7) Department of Pediatrics, Università degli Studi di Milano, 20122 Milano, **Italy**.
8) Department of Pediatrics and Pediatric Rheumatology Service, Rambam Health Care Campus, Ruth Children's Hospital, Haifa, Israel

1) Ümraniye Education and Research Hospital, Pediatric Rheumatology, Istanbul, Turkey
2) Hacettepe University, Pediatric Rheumatology, Ankara, Turkey
3) Ankara Bilkent City Hospital, Pediatric Rheumatology, Ankara, Turkey
4) Kocaeli University, Pediatric Rheumatology, Kocaeli, Turkey
5) Erciyes University, Pediatric Rheumatology, Kayseri, Turkey
6) Ankara Etlik City Hospital, Pediatric Rheumatology, Ankara, Turkey
7) Pamukkale University, Pediatric Rheumatology, Denizli, Turkey
8) Karadeniz Technical University, Pediatric Rheumatology, Trabzon, Turkey
9) Göztepe Prof. Dr. Süleyman Yalçın City Hospital, Pediatric Rheumatology, İstanbul, Turkey
10) Diyarbakir Children's Hospital, Pediatric Rheumatology, Diyarbakır, Turkey
11) Istanbul Faculty of medicine, Pediatric Rheumatology, Istanbul, Turkey

Background & Objective

- There are a limited number of real-life data reporting issues about pediatric Behçet’s disease.
- Previous reports, particularly adult studies, mainly focus on effectiveness of biologics in single-organ involvements (eye, gastrointestinal system, etc.).
- However, more reliable evidence on the efficacy and safety of these drugs in pediatric Behçet's disease is still needed.

Background & Objective

- This project aims to evaluate the characteristics of biological treatment regimens, their efficacy, side effects and outcomes on any clinical involvement of pediatric BD.

- It is aimed to provide a comprehensive overview of the use of biologics in pediatric-onset BD, for which there is no worldwide consensus.

Material- Method

- The study is designed as a retrospective, observational, multicenter, and international research.
- Inclusion criteria: Patients under 18 years of age at diagnosis, patients classified as Behçet's Disease according to the "International Behçet's Disease Criteria" and/or "Pediatric Behçet's Disease" criteria, and patients who have used biologic drugs at some point in their treatment.
- Exclusion criteria: Patients over 18 years of age at diagnosis, patients who are not fulfilling International Behçet's Disease Criteria" and/or "Pediatric Behçet's Disease" criteria and patients who have not used biologic drugs at some point in their treatment.

Pediatric Behçet's Disease Criteria		
Items	**Description**	**Value**
Recurrent oral aphthosis	At least three attacks/year	1
Genital ulceration or aphthosis	Typically with scar	1
Skin involvement	Necrotic folliculitis, acneiform lesions, erythema nodosum	1
Ocular involvement	Anterior uveitis, posterior uveitis, retinal vasculitis	1
Neurological signs	With the exception of isolated headaches	1
Vascular signs	Venous thrombosis, arterial thrombosis, arterial aneurysm	1

Three of 6 items are required to classify a patient as having pediatric BD

ICBD criteria	
Symptoms	**Value**
Ocular lesions	2
Genital aphthosis	2
Oral aphthosis	2
Neurological manifestation	1
Vascular manifestation	1
Positive pathergy test	1*

*Pathergy test is optional and the primary scoring system does not include pathergy testing. However, where pathergy testing is conducted one extra point may be assigned for a positive result

Results

Age (years) (SD)	17±4.1
Gender (female/total) (%)	38/108 (35.2)
Age of diagnosis (years) (IQR)	13 (10.1-15.2)
Follow-up duration(month) (IQR)	33 (118.8-61.7)
Follow-up period under biological (months) (IQR)	18,5 (8,2-27,5)
HLA B51	n (%)
Positive	29 (65,9)
Negative	14 (31,8)
N/A	1 (2,3)
Result of pathergy test	n (%)
Positive	16 (14.8)
Negative	69 (63,9)
N/A	23 (21.3)
Comorbid disease	19 (17.5)
Diagnostic criteria	n (%)
Only ICBD	35 (32.4)
Only PedBD	0 (0)
Both ICBD and PedBD	67 (62)
İnkomplet BD	5 (4.6)

So far, data has been received from 17 centers, comprising a total of 108 patients.

	N (%)		N (%)
Ocular involement	44 (40.7)	Adalimumab Infliximab Etanercept Tofacitinib	28 (63.6) 14 (31.8) 1 (2.3) 1 (2.3)
Mucocutaneous involvement	23 (21.3)	Adalimumab Apremilast Infliximab Etanercept Tofacitinib Interferon	13 (56.5) 5 (21.7) 2 (8.7) 1 (4.3) 1 (4.3) 1 (4.3)
Neurological involvement	21 (19.4)	Infliximab Adalimumab	13 (61.9) 8 (38.1)
Vascular involvement	7 (6.5)	Infliximab Adalimumab Golinumab	4 (57.1) 2 (28.6) 1 (14.3)
GIS involvement	4 (3.7)	Adalimumab Infliximab	3 (75) 1 (25)
Musculoskeletal involvement	4 (3.7)	Adalimumab Tofacitinib	3 (75) 1 (25)
Cardiac involvement	4 (3.7)	Infliximab Adalimumab	3 (75) 1 (25)
Pulmonary involvement	1 (0.9)	İnfliximab	1 (100)

First choice biological agent

- ✓ 53.7% (58/108) adalimumab
- ✓ 35.2% (38/108) infliksimab
- ✓ 4.6% (5/108) apremilast
- ✓ 1.9% (2/108) etanercept
- ✓ 1.9% (2/108) tofacitinib
- ✓ 0.9% (1/108) golilumab
- ✓ 0.9% (1/108) interferon

Results

Treatment-related side effects developed in 11 patients.

- Infection

  Mild infection in 2 patients,

  Moderate infection in 2 patients,

  Severe infection in 1 patient,

- Allergic reaction

  Redness at the injection site in 2 patients

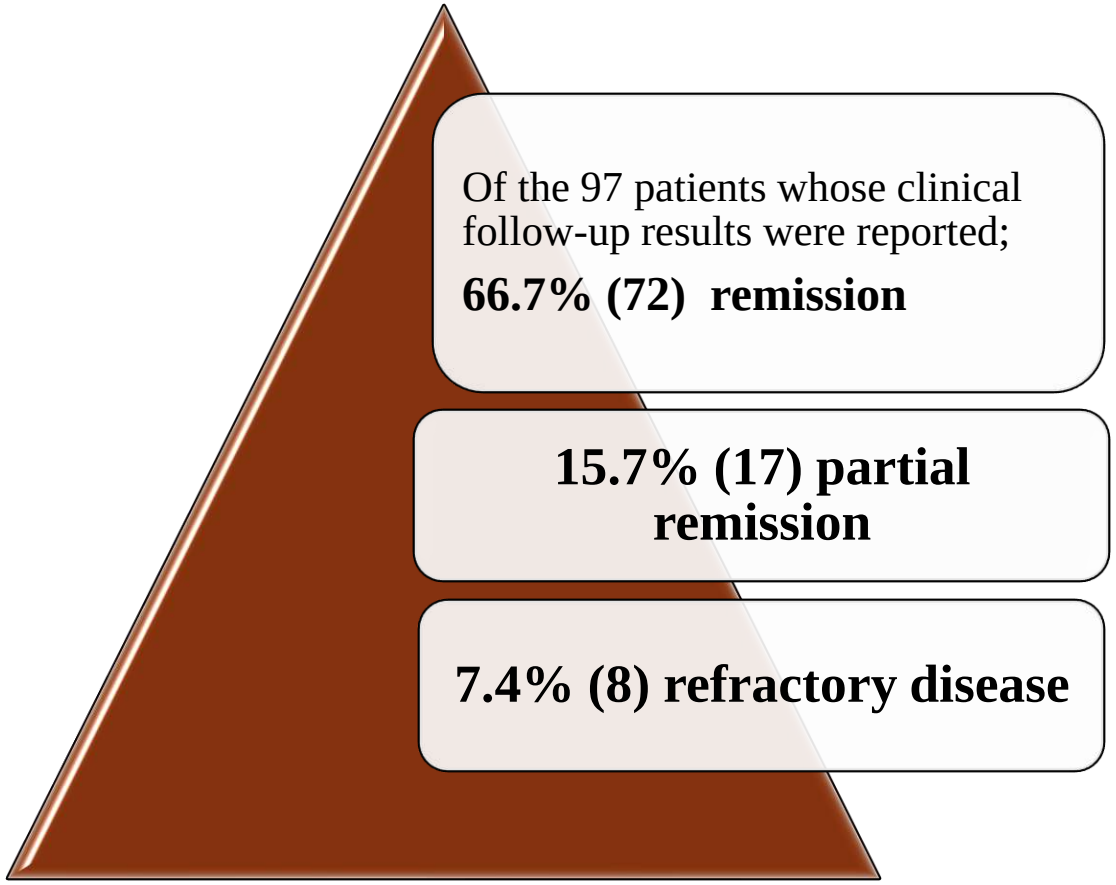
  Hypersensitivity reaction in 3 patients,

- Suspected demyelinating disease

  In 1 patient

No treatment changes were made in patients with mild infections and injection site reactions.

Results


Of the 97 patients whose clinical follow-up results were reported;
66.7% (72) remission
15.7% (17) partial remission
7.4% (8) refractory disease

Conclusion

- Biological drugs are increasingly used in the treatment of pediatric BD.
- In our study, it was observed that anti-TNFs were the first choice biological agent in 90.8% of the patients
- These treatments had an acceptable safety profile and high remission rates.
- Multicenter prospective controlled studies with long-term follow-up are required to confirm the observations that contribute to current treatment in pediatric BD and to confirm the efficacy and safety of these treatments.

Thank you for your collaboration!

Özlem Akgün (drozlemakgun@hotmail.com)
Fatma Gül Demirkan (fatmagy@gmail.com)
Nuray Aktay Ayaz (nurayaktay@gmail.com)

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3. The Nailfold Videocapillaroscopy in Pediatric Behçet's Disease: Multicenter International Study

Nuray Aktay Ayaz (PI), Rheumatology Unit, **Istanbul University, Faculty of Medicine**, Istanbul, Turkey

Figen Çakmak (Co- PI), Rheumatology Unit, **Istanbul University, Faculty of Medicine**, Istanbul, Turkey

Objective

- 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 BD activity scores and microvascular involvement
- To compare nailfold capillaroscopic alterations and NVC scores of patients diagnosed with BD with healthy volunteers.

Materials- Methods

- Behçet's Disease according to the following criteria;
  - Pediatric Behçets' Disease criteria
  - International study group (ISG) criteria
  - The international criteria for Behçet's Disease (ICBD)

  (The inclusion criteria were broad to permit generalizability to a wider BD population)
- Age of the participants
  - 5-21
  - Demographic and clinical features will be recorded
- Activity of BD measured by BD current activity form (BDCAF)
- Laboratory findings
  - CBC, ESR, CRP
  - Patergy testing, HLA B51 typing (if present)
- Treatment modalities will be recorded

NVC examination

200 * magnification CapillaryScope

4 images from 8 fingers (excluding thumbs)

A drop of immersion oil placed on the nailfold bed to improve resolution

A booklet illustrating definitions of the capillaroscopic alterations

A detailed history of

- Dominant hand
- Jobs and hobbits
- Sports and physical activities
- Recent finger trauma, nail biting (onychophagy) and/or habit of self-injuring the cuticles
- Spesific medications (i.e. beta-adrenergic blockers, vasodilators, anticoagulants and antihypertensive drugs) and other agents with vasoactive effects (i.e. marijuana, cocaine,amphetamines)
- Comorbidities will be recorded

Materials- Methods

For standardization of NVC examinations

Interactive online workshop

To ensure optimal reliability in the assessment, staff at all centers planning to be trained on the assessment of capillaroscopic images in an interactive online workshop with practical use of the capillaroscopy devices)

- The images was analyzed **at each participating center**
- The steering committe evaluated **the quality of NVC images** of the first 3 patients at each center.
- The results of the study was evaluated collectively.

Capilary morphology → Capillary measurements → Capillary alterations

Participating centers

1. Turkey
    1. Ümraniye Education and Research Hospital, Pediatric Rheumatology, Istanbul, Turkey
    2. Hacettepe University, Pediatric Rheumatology, Ankara, Turkey
    3. Erciyes University, Pediatric Rheumatology, Kayseri, Turkey
    4. Dokuz Eylül University, Pediatric Rheumatology, İzmir, Turkey
    5. Kocaeli University, Pediatric Rheumatology, Kocaeli, Turkey
2. Italy
    1. Genoa University Genoa, Italy
    2. AOU Meyer Children University Hospital, University of Florence, Italy
    3. Bambino Gesu 'Pediatric Hospital, Roma, Italy
3. Egypt
    1. Al-Azhar University, Rheumatology and Rehabilitation Centre, Cairo, Egypt
4. Spain
    1. Sant Joan de Déu Children's Hospital, Pediatric Rheumatology Unit, Barcelona, Spain
5. Croatia
    1. Department of Pediatrics, University of Zagreb School of Medicine

Enrollment

- The enrollment of the study was closed.

- The data of patients and healthy controls will completed before January 2024.

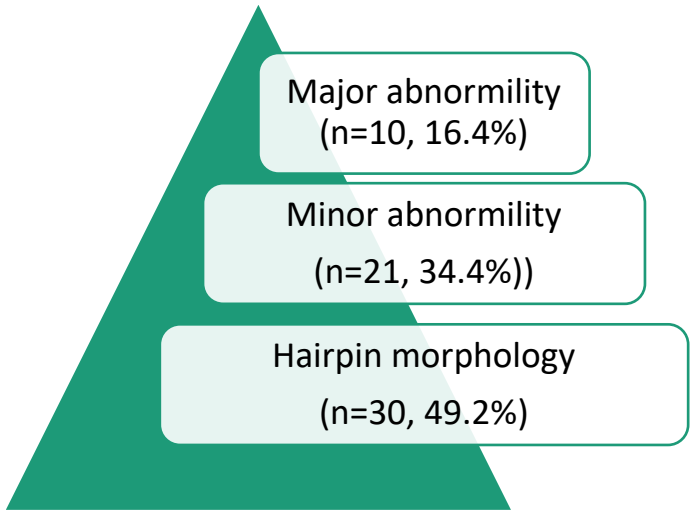
Preliminary results

7 centers

61 patients

Results

- 61 patients
- The mean age was 17 years (IOR 13-19)
- 32 (52.5%) of them were girls



CLINICAL PRESENTATION

- Mucocutaneous involvement (n=61, 100%)
- Uveitis (n=16, 21.3%)
- Vascular and neurological involvement (n=19, 31.1%)
- GIS involvement (n=4, 6.6%)

- Capillary density=8 (IQR:7-8.3)
- Capillary length= 370 µm (IQR:320-431)
- Arterial width = 11 µm (IQR:9.1-13.5)
- Venous width = 17 µm (IQR:12-17.2)
- Apical loop width= 17 µm (IQR:12-23)
- Capillary width = 37 µm (IQR:31.5-43)
- Intercapillary distance= 101 µm (IQR:84-122)

Results

- Neoangiogenesis (n=16, 26.2%)
- Enlarged capillary (n=14, 23%)
- Capillary meandering (n=11, 18%)
- Bushy capillary (n=8, 13.1%)
- Bizarre capillary (n=4, 6.6%)
- Microhemorrhage (n=4, 6.6%)

- 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).

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- We are waiting for the data of healthy peers of the participating centers…
- Thanks to all researchers and vasculitis working group who participated and contributed....

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THANK YOU

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pediatric rheumatology european society

Paediatric Vasculitis Working Party

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4. The Effect of the Initial Hyperinflammatory Condition on The Outcome of IgA Vasculitis

University of Health Sciences, Umraniye Training and Research Hospital, Pediatric Rheumatology

Dr. Şengül Çağlayan

Prof. Dr. Betül Sözeri

Background

- IgA vasculitis (IgAV) is the most common primary systemic vasculitis in children.
- The most common and typical characteristic feature of the disease is non-thrombocytopenic, palpable purpura.
- Other clinical findings are arthritis, arthralgia, gastrointestinal system findings, and renal involvement
- Renal involvement is the most important cause of chronic complications and long-term morbidity
- However, in the studies conducted so far, a factor that predicts renal involvement in which patients has not been defined.

Aim of the study

Initial inflammatory parameters

- Pentraxin 3
- Serum Galectin
- NLR
- CRP/albumin
- SII (Trombosit count x nötrofil count/lenfosit count)
- SIRI (Nötrofil count x monosit count/lenfosit count)

Can it predict multisystemic involvement affecting disease prognosis?

Pentraxin-3

Pentraxins are an evolutionarily rooted family of proteins involved in the innate immune response

Expressed at inflammatory sites

- Regulation of tissue hemostasis
- Fertility
- Cancer biology
- Autoimmunity
- Regulation of angiogenesis

PTX3 is thought to be a more effective biomarker than CRP and SAP in predicting the prognosis of inflammatory diseases.

Studies on SLE, AAV, PMR, RA, TA, GCA

Galectin-3

Galectins are a family of lectins that bind to β-galactoside

They play critical roles in maintaining homeostasis

- Modulating cell apoptosis
- Proliferation
- Cell cycle
- Immune response.

There are some studies in the literature that galectins can be used as biomarkers for inflammatory bowel disease, AAV, and SLE

NLR

Neutrophil-lymphocyte ratio has been used as an inflammation marker in several diseases

Increased NLR has been associated with poor prognosis in many cancer patients

Studies about IgAV support that NLR can be used as a marker of gastrointestinal system involvement

CRP/albümin ratio

CAR is a newly identified and more valuable and reliable marker than CRP or albumin alone in predicting inflammatory status and prognosis in various diseases.

It was found to be associated with predicting prognosis in patients with

- Colorectal cancer
- Coronary artery disease
- Inflammatory bowel disease
- Takayasu arteritis
- Rheumatoid arthritis
- Sepsis

SII and SIRI

SII and SIRI are newly defined scores used as local and systemic inflammation markers

SII is currently used as a prognostic marker in malignancy studies

SIRI has been shown to have a strong predictive value in cardiovascular diseases, infectious diseases, cancer, and trauma patients

Materials and Methods

- The study will be conducted at **:** University of Health Sciences**,** Umraniye Training and Research Hospital, İstanbul, Turkey after obtaining approval from institute ethics committee.
- Informed written consent will be received from patients/parents as appropriate.

Inclusion criteria

- Patients <18 years
- All patients must meet the 2008 Ankara PReS/EULAR/PRINTO IgAV classification criteria
- The study will include patients with a new IgAV diagnosis (Patients who present with relapse at the start of the study will be excluded)
- Before beginning treatment, biological samples should be collected and stored
- Prior to enrolment, the patient's parents and/or children must be willing to participate and assigned informed consent

Exclusion criteria

- Patients with systemic diseases causing leukocytoclastic vasculitis such as **connective tissue diseases** (systemic lupus erythematosus, Sjogren's syndrome), **inflammatory bowel disease**, **Behçet's disease**, **cryoglobulinemic vasculitis**, **hypocomplementemic urticarial vasculitis**
- Any major illness or condition, as well as evidence of an unstable clinical condition (eg, **cardiovascular**, **cerebrovascular**, **neurologic**, **metabolic**, **immunologic**, **infectious**, **hepatic**, **renal condition**, **uncontrolled diabetes mellitus** or **hypertension**)
- Patients who have a history of **kidney disease**, **proteinuria**, or **kidney failure**

Materials and Methods

The study will be conducted in the 1,5 year period between February 2023 – August 2024.

- Patients diagnosed with IgA vasculitis between February 2023 and February 2024
- The study will be completed in August 2024
- The last patient will be included in February 2024

Visits

- 1. day (blood samples will be stored, and data entry will be made)
- 6. Months (data entry will be made)

Materials and Methods

- Before the treatment, complete blood count, CRP, and biochemical parameters will be studied at their centers.
- In addition, for pentraxin-3 and galectin-3, patient 6 mL serum samples will be taken, centrifuged, and stored until studied.
- At the end of the study, the collected samples will be analyzed in the Umraniye Training and Research Hospital.
- The target number of patients: 200
- Number of control patients: 50

Serum sample

1. Allow serum to clot for 10-20 minutes at room temperature.
2. Centrifuge at 2000-3000 RPM for 20 minutes.
3. Collect the supernatants carefully. When sediments occurred during storage, centrifugation should be performed again.
4. Hemolysis can greatly impact the validity of test results. Take care to minimize hemolysis.
5. Samples can be stored at -20°C for a maximum of 1 month, then at -80°C. Avoid repeated freze thaw cycles.
6. Serum samples will be shipped on dry ice.

The current situation

- So far, 114 patient samples have been collected from Turkey
- Patient recruitment will continue until February 2024
- Afterwards, samples collected from all centres will be sent to Umraniye Training and Research Hospital and serum samples will be studied collectively
- DHL codes will be disseminated to international centers for the purpose of shipping their respective samples.

- Thank you for your collaboration….

Şengül Çağlayan (sengulturkercaglayan@gmail.com)
Betül Sözeri (drbetulsozeri@gmail.com)
umraniyepedrhestudy@gmail.com

5. Pediatric Vasculitis Initiative: PedVas

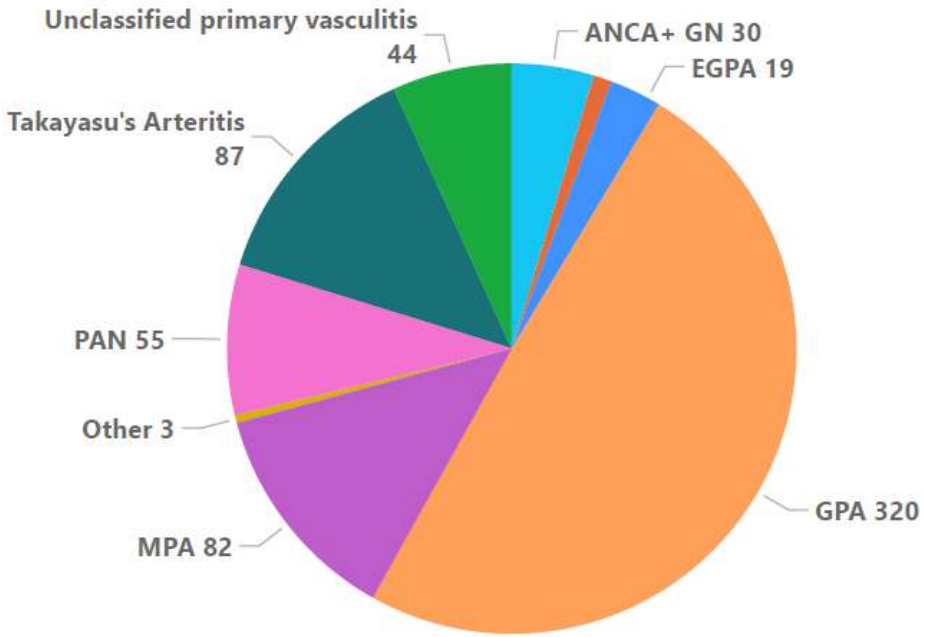
Patients in the registry:

647

Collected samples from

269

Patients



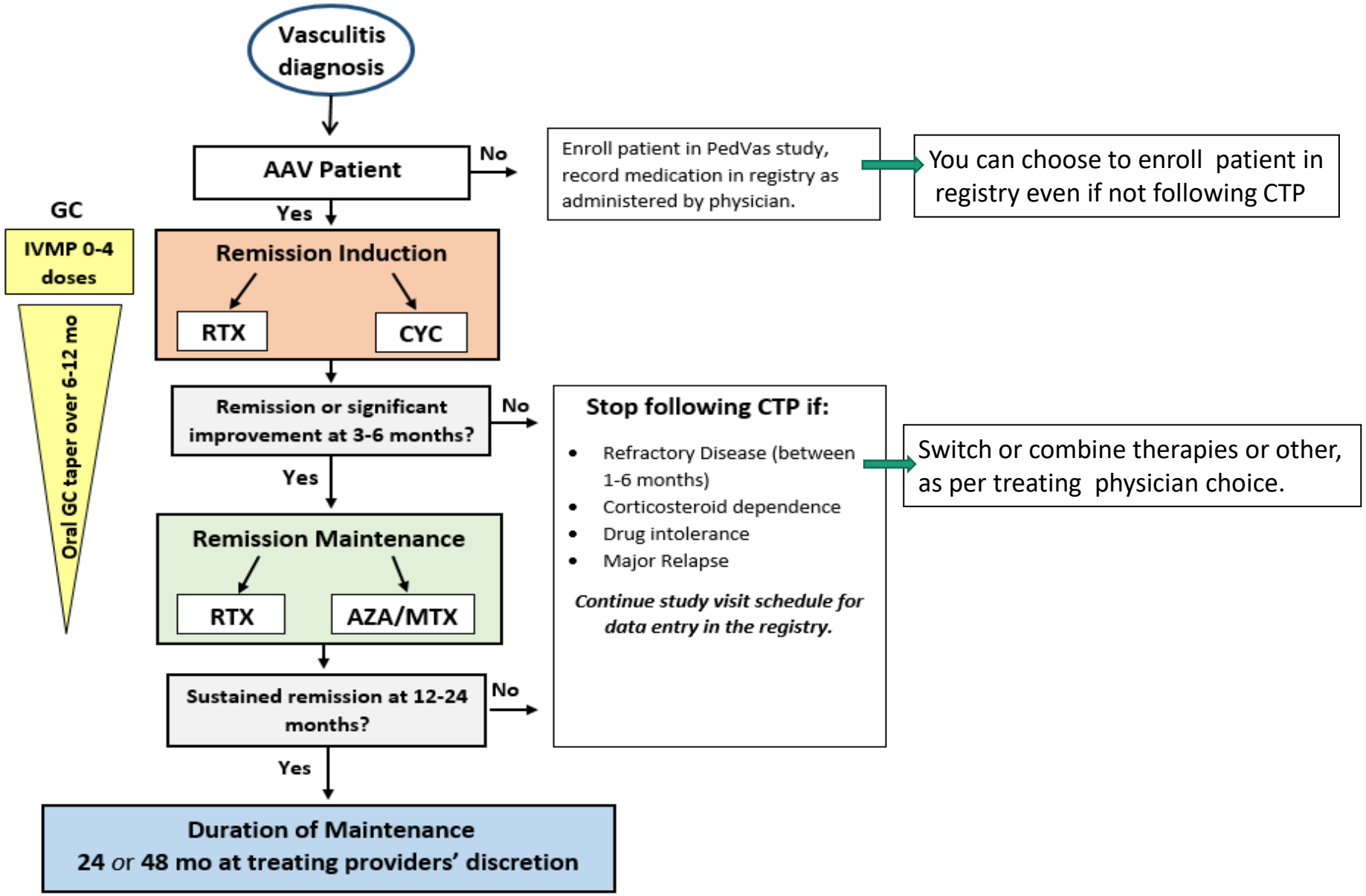
Abstract: **PReS 23-ABS-1616** Adult ACR/EULAR AAV criteria tested in pediatrics

Poster P158: displayed on Friday

Current collaborative PedVas studies

- *Two PRES –CARRA proposals*
  1. Comparison of different scoring systems for assessment of disease activity in childhood TAK (PRES – CARRA proposal)
     - *Within PedVas, or using a modified REDCap data-set from PedVas*
     - *Marija to discuss this New proposal*
  2. *Review baseline features & one year outcome of PedVas TAK patients*
     - *In collaboration with above retrospectively filling in data gaps*
- To comparatively evaluate CARRA-endorsed Consensus Treatment Plan options for pAAV in PedVas registry
  - *presented last year - Summary slide follows:*

Comparative evaluation of AAV CTP alternatives



How to participate in PedVas?

- Contribute clinical data +/- biological samples (TOD + FU) for chronic systemic vasculitis or selected diagnoses
- To enrol your site, contact coordinator **else.bosman@cw.bc.ca** or **dcabral@cw.bc.ca**
- requirements
    - REB/IRB (templates available)
    - Signed inter-institutional agreement (multi-site consensus document established)
    - Letter of transfer of funds
    - Basic laboratory infrastructure if providing biosamples

Overview: Registry-based evaluation of comparative effectiveness of CTP options for treating moderate to severe AAV (GPA/MPA)

Study Phase		Dichotomous comparisons	Patient target group	Primary outcome
1	Remission-induction	**CYC** Or **RTX**	Mod-Severe AAV	Significantly improved or inactive Disease at 6m (not refractory /intolerant)
2	Remission-maintenance	**AZA/MTX** Or **RTX**	Phase 1 patients achieving primary outcome	Time to first major relapse/relapse frequency
3	Duration of maintenance	**2 years** Or **4 years**	Phase 2 patients with inactive disease at 24m	Time to first major relapse after 24m /relapse frequency

REMISSION-INDUCTION ALTERNATIVES

Cyclophosphamide

Regimen 1: *(adapted EULAR)*

- 15mg/kg 2 weekly X 3 doses then 3 weekly

OR

Regimen 2: (adapted NIH SLE protocol)

- 750 mg/m^2 4 weekly for 4-7 doses

until sustained disease inactivity (pVAS=0)

Rituximab

Regimen 1: *(adapted from adult trials)*

- 375 mg/m^2 - <u>4 doses</u> one week apart

OR

Regimen 2: (most common in pediatrics)

- 750 mg/m^2 – <u>2 doses</u> at 2 week interval

Maximum individual dose 1000mg

REMISSION-MAINTENANCE ALTERNATIVES

Azathioprine regimen:

- 2-3 mg/kg PO daily, max 200 mg/day

OR

Methotrexate regimen:

- 0.5-0.7 mg/kg (max 25 mg) once weekly

Rituximab regimen 1:

- 375 mg/m^2 IV (max 500mg) day 0 & 14, ev 6 mo

OR

Rituximab regimen 2:

- 750 mg/m^2 IV (max 1000mg) day 0, every 6 mo

GLUCOCORTICOID USE FOR ALL PATIENTS

IV Methyl prednisolone (at induction onset)

- 30mg/kg/dose (max 1gm); not >4 in weeks 0-3

Oral Prednisone

- Weeks 2-4: 1-2mg/kg/d before weaning
- Wean to 15-30mg/d (or <0.5mg/kg/d if <40kg) by 12 weeks
- Wean to 0-10 mg/d (or <0.2mg/kg/d if <40kg) by 6 months

Vasculitis Working Party: 2023

MEETING AGENDA

ACTIVITIES IN SCIENCE & RESEARCH (ongoing/planned):

NEW PROJECTS (PROPOSALS):

1.Seza Ozen, Muserref Kasap Cuceoglu: **Pediatric Takayasu arteritis: a multicenter retrospective cohort study**

2.Marija Jelusic, Mario Sestan: **Comparison of different scoring systems for assessment of disease activity in childhood Takayasu arteritis (PRES – CARRA project)**

3.Isabelle Koné-Paut: **A retrospective observational study of the use of anakinra for the treatment of Kawasaki disease**

4.Tamás Constantin: **Addressing Diagnostic and Treatment Challenges in Pediatric Primary Angiitis of the Central Nervous System (cPACNS)**

5.Teresa Giani: **Macrophage Activation Syndrome in Kawasaki disease: features, treatment, outcomes and prediciting factors** (Collaboration with MAS/sJIA WP) 5.

6. Şengül Çağlayan, Betül Sözeri: **Capillaroscopy in ADA-2 deficiency**

7. Sara Stern/USA (Chair of the CARRA Childhood Sjögren's Disease Workgroup): **International Sjögren's Disease registry**

PRES Vasculitis Working Party

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ROTTERDAM, THE NETHERLANDS

1. Pediatric Takayasu Arteritis: A Multicenter Retrospective Cohort Study

PReS Vasculitis Working Group

National Vasculitis Group of Turkey

Prof. Dr. Seza OZEN

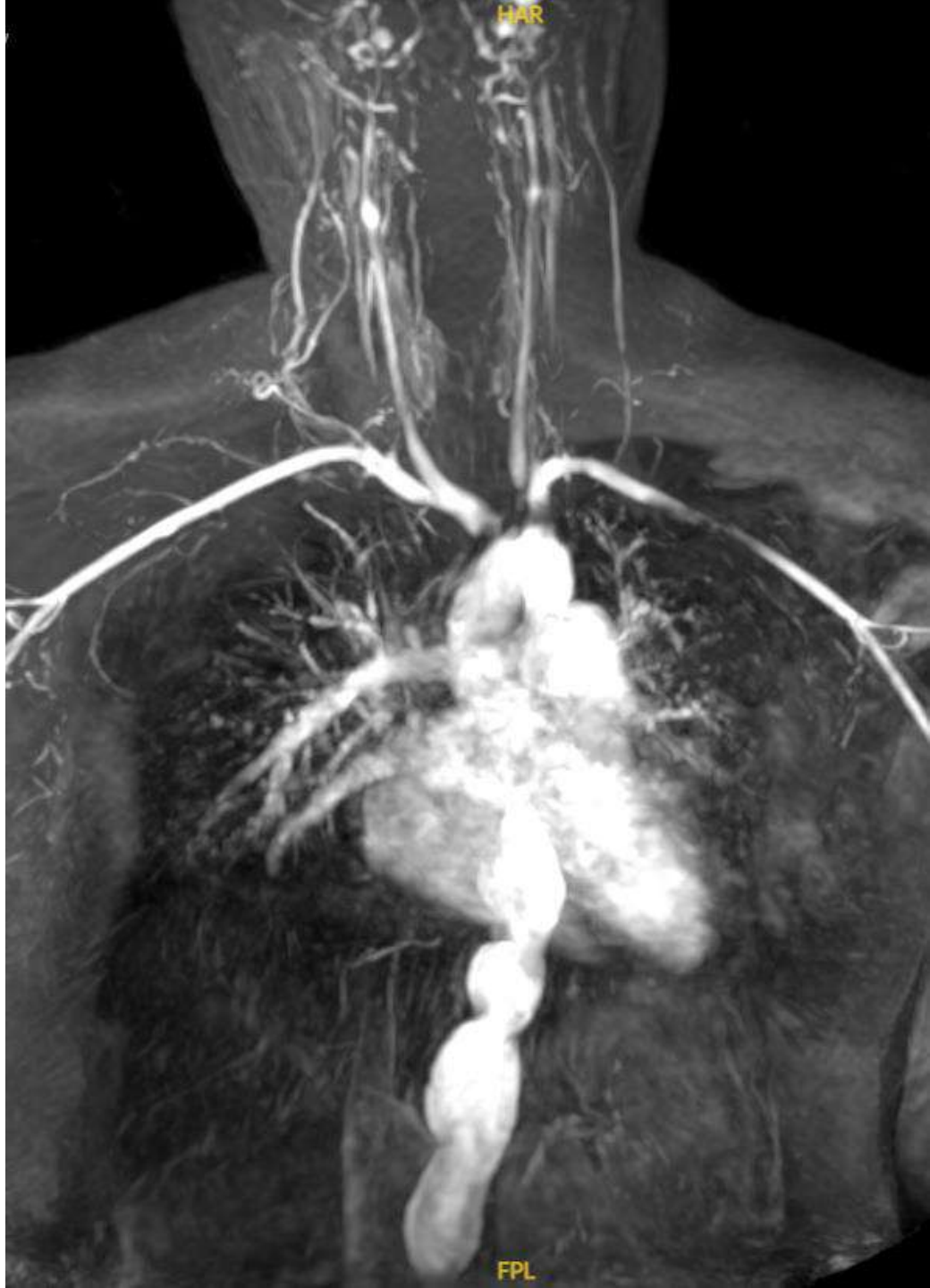
Assoc. Prof. Dr. Ezgi Deniz BATU

Dr. Muserref Kasap CUCEOGLU

Hacettepe University Faculty of Medicine, Pediatric Rheumatology Unit

BACKGROUND

- Takayasu Arteritis is a large vessel vasculitis involving the aorta and its associated branches with life-threatening clinical manifestations.
- It can cause serious conditions such as syncope, intracranial thrombus and sudden cardiac death.
- Early recognition of the disease and initiation of effective treatment is essential.



STUDY DESIGN

Aim:

- Evaluation of 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.

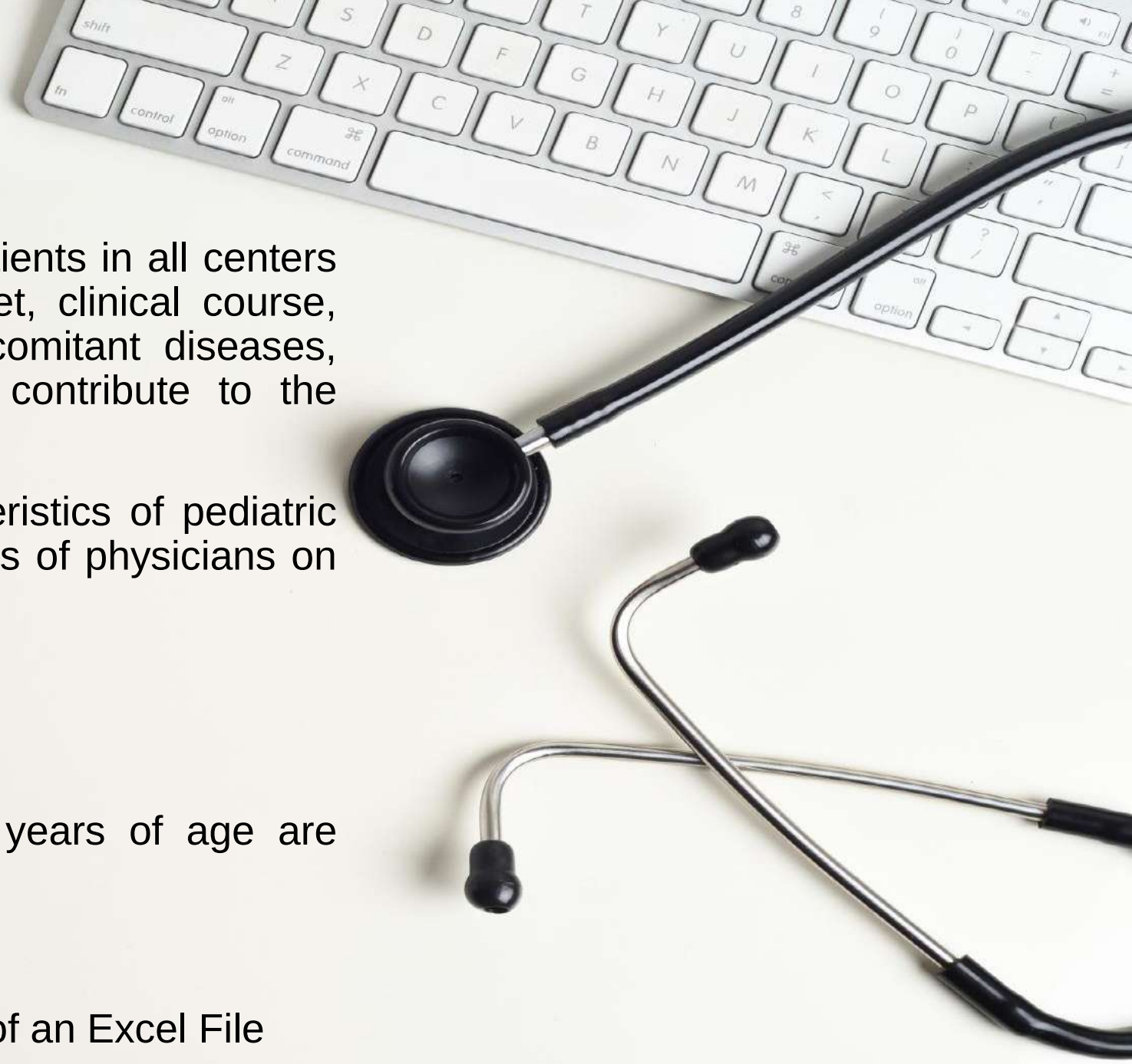
- Assessing the treatment response

Inclusion Criteria:

- All pediatric TAK patients under 18 years of age are included to the research.

ITAS will be used as activity score

Data will be compiled/stored in the form of an Excel File



Currently

- 64 pediatric TAK patients data is collected from Turkey
- New centers participate in our project.
- We encourage you to collaborate in this project.

Contact us via

Muserref Kasap Cuceoglu — drmuserref@gmail.com

Seza Ozen — sezaozen@gmail.com

PReS Vasculitis Working Party

CARRA Childhood Arthritis and Rheumatology Research Alliance

New PReS-CARRA research proposal

2. Comparison of different scoring systems for assessment of disease activity in childhood Takayasu arteritis

Professor Marija Jelusic, MD, MSc, PhD,
Mario Sestan, MD, PhD
University of Zagreb School of Medicine
Division of Paediatric Immunology, Rheumatology and Allergology
Centre of Reference for Paediatric and Adolescent Rheumatology Republic of Croatia
University Hospital Centre Zagreb, Croatia

Takayasu arteritis - disease activity

- Assessment of disease activity is intrinsic to the management of autoimmune inflammatory diseases. Active disease is generally treated with the initiation or intensification of immunosuppressive therapy

- The distinction of active disease is challenging in Takayasu arteritis (the sites of pathology are not easily accessible for clinical examination or histopathological evaluation)

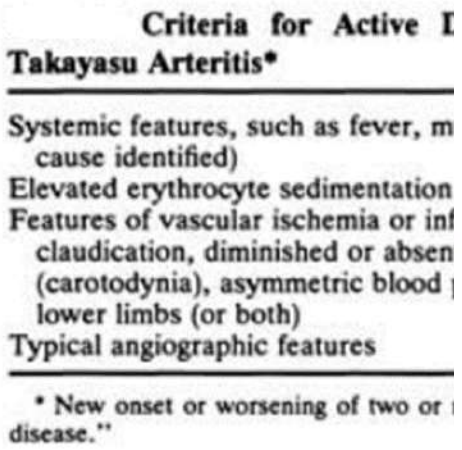
- Clinical assessment of disease activity in TAK relies on a composite assessment of clinical features, inflammatory markers, and serial imaging

Misra DP, et al. *Diagnostics (Basel)*. 2022;12:2565.
Aeschlimann FA, et al. *Front Pediatr*. 2022;10:872313.

Takayasu arteritis - disease activity

- several scoring systems are used: NIH Score, the Disease Extent Index in TAK (DEI.TAK), the Indian TAK Clinical Activity Score (ITAS2010), EULAR criteria for active LVV, PVAS...

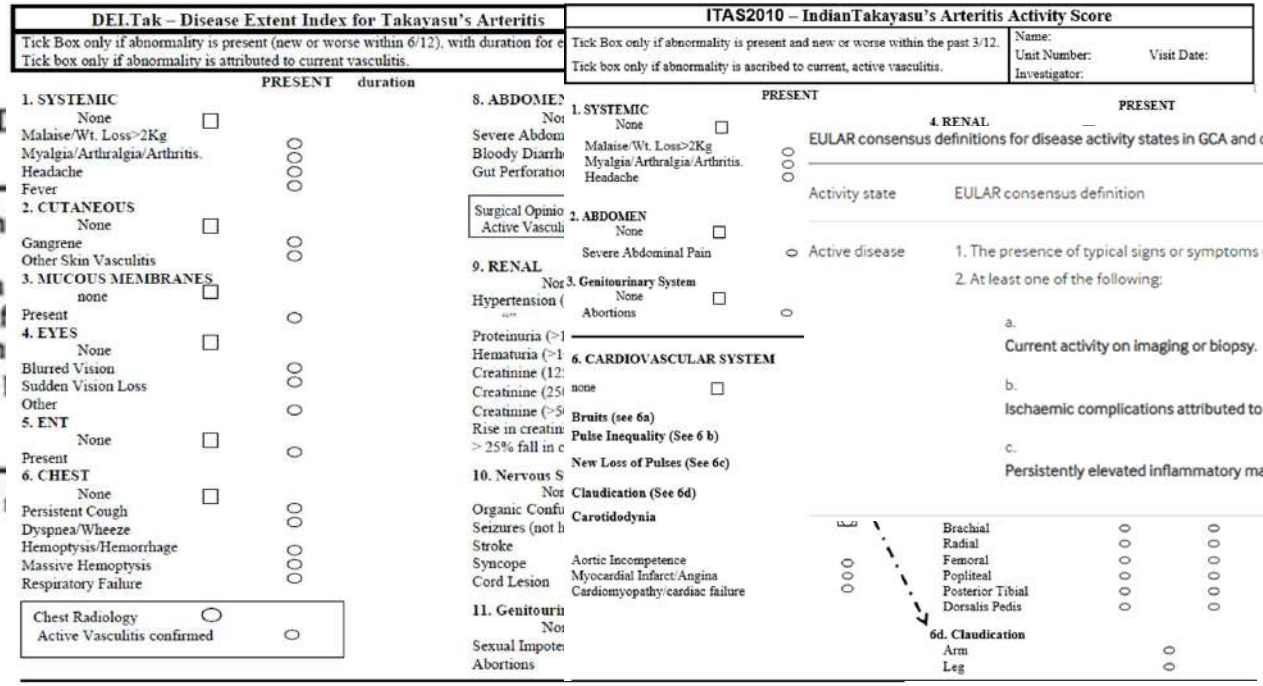
- only PVAS has been validated in children



Criteria for Active I
Takayasu Arteritis*

Systemic features, such as fever, m
cause identified)
Elevated erythrocyte sedimentation
Features of vascular ischemia or inf
claudication, diminished or absen
(carotodynia), asymmetric blood
lower limbs (or both)
Typical angiographic features

* New onset or worsening of two or
disease.**



DEI.Tak – Disease Extent Index for Takayasu's Arteritis

Tick Box only if abnormality is present (new or worse within 6/12), with duration for e
Tick box only if abnormality is attributed to current vasculitis.

PRESENT duration

1. SYSTEMIC
None
Malaise/Wt. Loss>2Kg
Myalgia/Arthralgia/Arthritis.
Headache
Fever
2. CUTANEOUS
None
Gangrene
Other Skin Vasculitis
3. MUCOUS MEMBRANES
none
Present
4. EYES
None
Blurred Vision
Sudden Vision Loss
Other
5. ENT
None
Present
6. CHEST
None
Persistent Cough
Dyspnea/Wheeze
Hemoptysis/Hemorrhage
Massive Hemoptysis
Respiratory Failure

Chest Radiology
Active Vasculitis confirmed

8. ABDOME
Nor
Severe Abdom
Bloody Diarrh
Gut Perforatio

Surgical Opinio
Active Vascul

9. RENAL
Nor
Hypertension (
Proteinuria (>1
Hematuria (>1
Creatinine (12
Creatinine (25
Creatinine (>5
Rise in creatin
> 25% fall in c

10. Nervous S
Nor
Organic Confu
Seizures (not h
Stroke
Syncope
Cord Lesion

11. Genitouri
Nor
Sexual Impote
Abortions

ITAS2010 – IndianTakayasu's Arteritis Activity Score

Tick Box only if abnormality is present and new or worse within the past 3/12.
Tick box only if abnormality is ascribed to current, active vasculitis.

Name:
Unit Number: Visit Date:
Investigator:

PRESENT

1. SYSTEMIC
None
Malaise/Wt. Loss>2Kg
Myalgia/Arthralgia/Arthritis.
Headache

2. ABDOMEN
None
Severe Abdominal Pain

3. Genitourinary System
None
Abortions

6. CARDIOVASCULAR SYSTEM
none
Bruits (see 6a)
Pulse Inequality (See 6 b)
New Loss of Pulses (See 6c)
Claudication (See 6d)
Carotidodynia

Aortic Incompetence
Myocardial Infarct/Angina
Cardiomyopathy/cardiac failure

4. RENAL
PRESENT

EULAR consensus definitions for disease activity states in GCA and other types of LVV

Activity state	EULAR consensus definition
Active disease	1. The presence of typical signs or symptoms of active LVV (tabl 2. At least one of the following: a. Current activity on imaging or biopsy. b. Ischaemic complications attributed to LVV. c. Persistently elevated inflammatory markers (after other

Brachial
Radial
Femoral
Popliteal
Posterior Tibial
Dorsalis Pedis

6d. Claudication
Arm
Leg

	None	Active
1. General	○	
Myalgia		○
Arthralgia or arthritis		○
Fever ≥ 38.0 °C		○
Weight Loss ≥ 5% body weight		○
2. Cutaneous	○	
Polymorphous exanthema		○
Livedo		○
Panniculitis		○
Purpura		○
Skin nodules		○
Infarct (nail edge lesion, splinter haemorrhage)		○
Ulcer (full-thickness necrosis)		○
Gangrene (extensive necrosis)		○
Other skin vasculitis (specify below)		○
3. Mucous membranes/eyes	○	
Mouth ulcers/granulomata		○
Genital ulcers		○
Adnexal inflammation		○
Significant proptosis		○
Red eye (Epi)scleritis		○
Red eye conjunctivitis/ biepharitis/keratitis		○
Uveitis		○
Blurred vision		○
Sudden visual loss		○
Retinal vasculitis/retinal vessel thrombosis/ retinal exudates/haemorrhages		○
4. ENT	○	
Nasal discharge/crusts/ulcers/granuloma		○
Paranasal sinus involvement		○
Subglottic stenosis/ hoarseness /stridor		○
Conductive hearing loss		○
Sensorineural hearing loss		○
5. Chest	○	
Wheeze or expiratory dyspnea		○
Endobronchial/endotracheal involvement		○
Nodules or cavities		○
Pleural effusion/pleurisy		○
Infiltrate		○
Massive haemoptysis/Alveolar haemorrhage		○
Respiratory failure		○

	None	Active
6. Cardiovascular	○	
Loss of pulses		○
Bruits over accessible arteries		○
Blood pressure discrepancy		○
Claudication of extremities		○
Ischaemic cardiac pain		○
Cardiomyopathy		○
Congestive cardiac failure		○
Valvular heart disease		○
Pericarditis		○
7. Abdominal	○	
Abdominal pain		○
Peritonitis		○
Blood in stools or bloody diarrhoea		○
Bowel ischaemia		○
8. Renal	○	
Hypertension >95th centile (for height)		○
Proteinuria >0.3 g/24h,>20mmol/mg creatinin		○
Haematuria >2+ or 5 rbc/hpf or red cell casts		○
GFR 50-80ml/min/1.73 m²		○
GFR 15-49 ml/min/1.73 m²		○
GFR <15 ml/min/1.73m²		○
Rise in creatinine > 10% or Creatinine clearance (GFR) fall > 25%		○
9. Nervous system	○	
Headache		○
Meningitis/encephalitis		○
Organic confusion/cognitive dysfunction		○
Seizures (not hypertensive)		○
Stroke		○
Cord lesion		○
Cranial nerve palsy		○
Sensory peripheral neuropathy		○
Motor mononeuritis multiplex		○
10. OTHER	○	
		○
		○
NO NEW/WORSE DISEASE :		
Tick here if there is no new/worse abnormality present in ANY of the systems above and active items represent low grade grumbling disease		

Misra DP, et al. *Diagnostics (Basel)*. 2022;12:2565.
Aeschlimann FA, et al. *Front Pediatr*. 2022;10:872313.

Takayasu arteritis - disease activity

- As of now, the PVAS stands as the sole validated disease activity measurement tool in childhood vasculitis, commonly employed for clinical research.

- However, it might not be the most suitable option for assessing disease activity in pediatric large vessel vasculitis.

- While the ITAS2010 has been specifically designed for evaluating disease activity in TAK patients, it has not yet undergone validation in children. As a result, it is not routinely utilized for assessing childhood TAK cases.

Aeschlimann FA, et al. *Arthritis Res Ther*. 2017;19:255.

International Takayasu Study Group Meeting (May - August 2023)

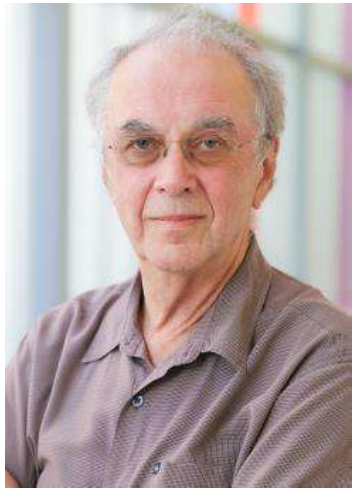
PRES Vasculitis Working Party

CARRA Childhood Arthritis and Rheumatology Research Alliance

Professor Marija Jelusic, PhD, MSc, MD

Associate Professor Vidya Sivaraman, MD

Associate Professor Linda Wagner-Weiner, MD, MS



Professor David Cabral, MBBS, FRCPC

Else Bosman, PhD

Mario Sestan, PhD, MD

James Bistolarides, MD

Idea was born at the CARRA meeting, New Orleans, March 2023

Research proposal (1)

OBJECTIVE: To assess the performance of the PVAS in pediatric patients with TAK and compare it with ITAS2010 and EULAR criteria for active LVV

- the correlation with physician global assessment
- the agreement between scores to assess active disease
- predictive value for the angiographic progression?
- respond to therapy?

PATIENTS: Pediatric patients with TAK diagnosed by EULAR/PRINTO/PReS classification criteria for childhood-onset TAK.

The control group: patients with other primary systemic vasculitis or diseases that mimic vasculitis.

Estimated number: ~ 80 patients with TAK and ~ 100 controls

Research proposal (2)

DATA COLLECTION: 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: RedCap or Excel Spreadsheet

REDCap
Research Electronic Data Capture

TIMELINE: 1.5 years (2024 - Spring 2025) (RedCap registry: 87 TAK patients, some missing data)

- the data will be presented in national and international conferences and will be reported in the **final manuscript**

Invitation to participate in the study

- We would like to invite all clinicians and scientists with patients with Takayasu arteritis to join our project!

- Please contact: Marija Jelusic: marija.jelusic@mef.hr, marija.jelusic.drazic@gmail.com

3. Kawasaki and anti-IL1:

A retrospective observational study of the use of anakinra for the treatment of Kawasaki disease

Prof. Isabelle Koné-Paut

Evidences from the literature

- **Main treatment of KD = IVIG + ASA** → administration of these treatments within the first 10 days following fever onset has been associated with a fivefold reduction in the risk of coronary artery aneurysms (CAA).
- However, 10–20% of patients do not respond to standard treatment and have an increased risk of cardiac complications and death.
- Therefore, the **2017 AHA guidelines**

➢ Suggest for resistant KD a **second dose of IVIG or a short course of high-dose steroids** or **infliximab**.

➢ mention **anakinra** among the different drugs that can be used in refractory forms

Interleukin-1 (IL-1) plays a key role in the pathogenesis of Kawasaki disease (KD), especially in the development of coronary artery aneurysms

❖ Up to now **many case reports** have reported the efficacy and safety of anakinra in the treatment of refractory KD, both on the inflammatory complications (MAS, KSS) and on coronary aneurisms

❖ **KAWAKINRA**: open label phase 2a*: 16 patients refractory to IvIg treated w/sc anakinra with good results

Retrospective observational multicentric study

OBJECTIVES:

- To identify KD patients treated with anakinra (demographics, KD characteristics; i.e; cardiac involvement, MAS)
- To analyse 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 realeted to treatment with anakinra

- **Inclusion criteria:**
- Patients with KD diagnosis according to AHA criteria for either complete or incomplete KD)
- Treatment with anakinra

Exclusion criteria:

-Patients suspected with another disease than KD (ex: MIS-C)

Methods

- We will use the JIR cohort as data repository
- 24 patients already identified with KD and anakinra treatment in France
- 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).
- Then we have to submit the project to the JIR scientifc committee
- It will be an opportunity for more countries to participate in the KD registry in general

INTERESTED???

Please contact:

isabelle.kone-paut@aphp.fr

perrine.dusser@aphp.fr

4. Addressing Challenges in Pediatric Primary Angiitis of the CNS (cPACNS)

A Multidisciplinary Approach

Dr. Tamás Constantin

Semmelweis University, Budapest, Hungary

Introduction

- Is it really vasculitis?

- Is it really necessary?

Diagnostic Challenges

- Overlapping phenotypes leading to misdiagnosis
- Limitations of MRI and lack of definitive laboratory tests
- The importance of accurate diagnosis for effective treatment

Treatment Challenges

- Lack of evidence-based guidelines
- Absence of newer immunomodulatory agents
- Concerns over
  - high cumulative doses of steroids and
  - optimal choice for anticoagulation

Objective

- Establish an **international multidisciplinary** working group

- Improve patient outcomes by refining diagnostic precision and evolving treatment paradigms

Plan of Action

- Systematic Review of the Literature
- Consensus Conference (online?) to develop unified diagnostic and treatment protocols
  - Expected to last 1.5 days with professionals from neurology, radiology, and rheumatology

Join the Working Group!

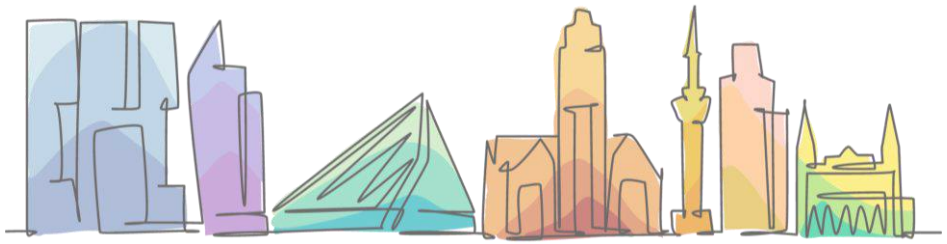
Collaborate to Refine Diagnosis and Treatment for cPACNS

- **Who Can Join?**
  - Neurologists, Radiologists, Rheumatologists, and other interested healthcare professionals with experience or interest in childhood vasculitis.
- **Why Join?**
  - Be a part of a global collaboration aimed at improving patient outcomes through refined diagnostic and treatment paradigms.
  - Contribute to the development of unified, evidence-based diagnostic and treatment protocols for cPACNS.

- **How to Join?**
  - Express your interest by contacting Tamás Constantin at **tamas.constantin@gmail.com**

PRES 2023

Rotterdam



Pediatric Vasculitis WP
&
MAS WP

Project Proposal:

5. Macrophage Activation Syndrome in Kawasaki disease: features, treatment, outcomes and predicting factors

Teresa Giani, AOU Meyer IRCCS, Florence, Italy
Francesca Minoia, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico Milan, Italy

Background

- ❑ <3%: MAS in KD
- ❑ Mortality and CAAs
- ❑ Risk factors:
  - Persistent fever
  - Splenomegaly
  - Hyperferritinemia
  - Thrombocytopenia
  - Elevated aspartate aminotransferase

- ❑ clinical and laboratory features overlapping
  underestimation of MAS

Aim

1. Epidemiological, clinical, and laboratory characteristics, management and out-comes in KD/MAS

2. Identify potential risk factors and diagnostic criteria for MAS in KD

Inclusion criteria:

- KD & MAS
- age 4 weeks - 17 years
- > January 2000

Exclusion criteria:

- non-confirmed KD (mimickers)
- primary HLH
- age < 4 weeks or >17 years
- no patient's consent.
- MAS diagnosis > 30 d

*

- ✓ Definitions ("complete", "incomplete", and "atypical" KD and "IVIG resistance") from American Heart Association doi/10.1161/CIR.0000000000000484
- ✓ MAS diagnosis: treating pediatric rheumatologist's expert opinion
- ✓ MAS within 30 days of KD first line treatment administration

Control group

KD age- and gender-related KD without MAS

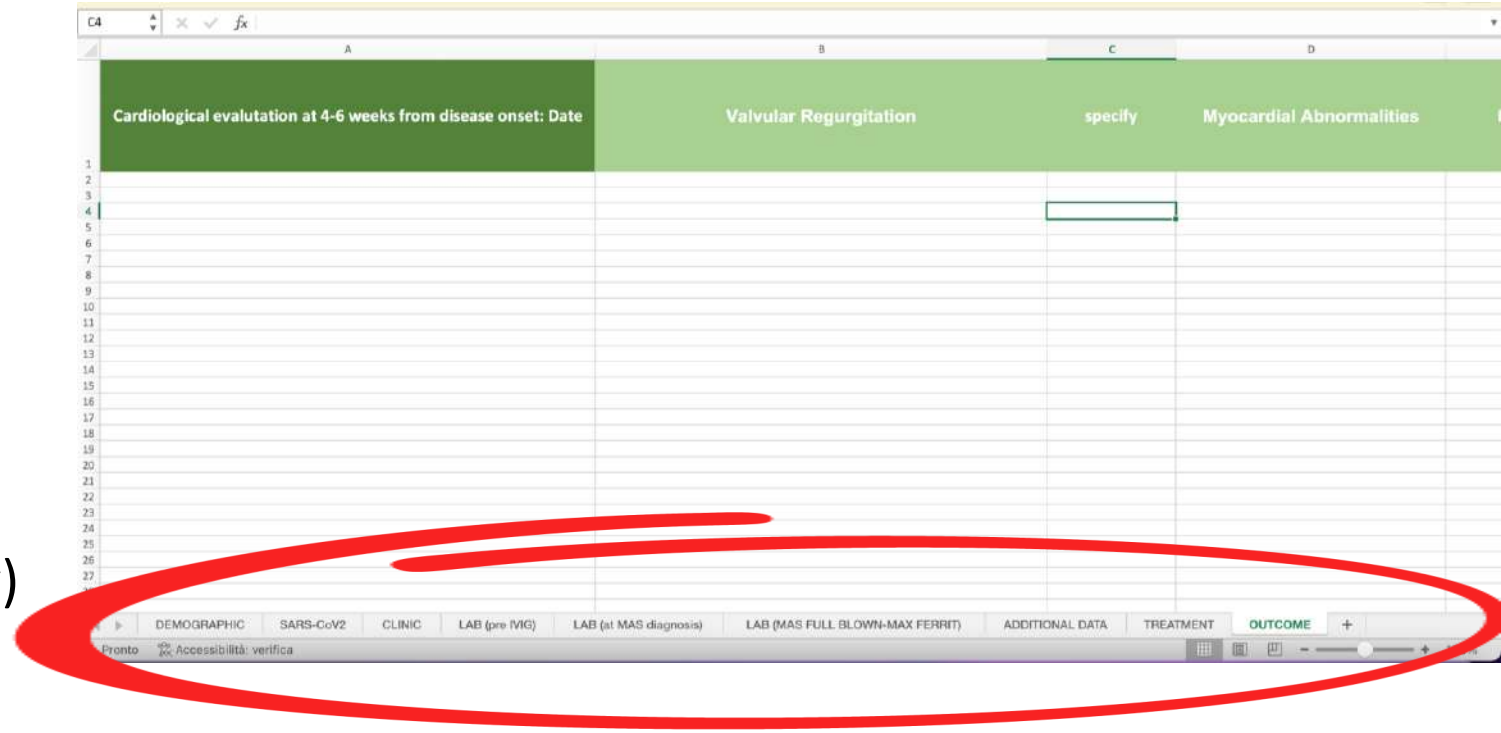
Any KD presentation (classic, incomplete, atypical, super KD)

- Responsive
- Resistant

Collection data

9 sections:

- Demographic variables
- Clinical signs
- Laboratory values
- SARS-CoV2 information
- Additional data (genetics, histology)
- Treatment
- Outcome



LAB TIMING

KD & MAS:

(1) before I IVIG
(2) MAS onset
(3) MAS full-blown

KD control group:

a. KD responsive I line treatment:
(1) before I IVIG
(3) at 96 h from the end IVIG

b. KD resistant :
(1) before I IVIG
(2) before II line treatment
(3) at 96 h from last (II line) successful treatment

Target numbers:

- 50 KD/MAS patients
- 50 KD first line-resistant patients
- 150 first line-responsive KD patients

Questionnaire

Since specific criteria for MAS in KD are not available, respondents are asked to

→ Identify 10 most important features suggestive for “MAS in KD”

→ Assign a score to all features and rank-order the top 10 ones

Happy to collaborate with you!

teresa.giani@gmail.com

francesca.minoia@policlinico.mi.it

pediatric rheumatology european society

Paediatric Vasculitis Working Party

PRES 2023

29th European Paediatric Rheumatology Congress

28 SEPTEMBER – 1 OCTOBER 2023

ROTTERDAM, THE NETHERLANDS

Project Proposal

6. Determining the Relationship Between DADA-2 Clinical Findings and Capillaroscopy

University of Health Sciences, Umraniye Training and Research Hospital, Pediatric Rheumatology

Dr. Şengül Çağlayan

Prof. Dr. Betül Sözeri

Background

- Deficiency Adenosine deaminase 2 (DADA2) is an inherited monogenic autoinflammatory disease caused by loss-of-function mutations in the ADA2 gene
- DADA2 patients exhibit two distinct phenotypes.
  - PAN
  - Bone marrow failure
- The diagnosis of DADA2
  - ADA2 enzymatic activity
  - ADA2 mutation

Aim of the study

- Since the clinical features of DADA2 are related to vascular inflammation, direct visualization of capillaries may provide a sensitive diagnostic test as well as a valid indicator of disease activity.

- This study aims to delineate the characteristics of nailfold capillary changes in individuals with DADA2 and explore potential correlations with the clinical features of the disease.

Methods

- The study will start, after the approval of the ethics committee at the Umraniye Training and Research Hospital in November 2023.

- 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

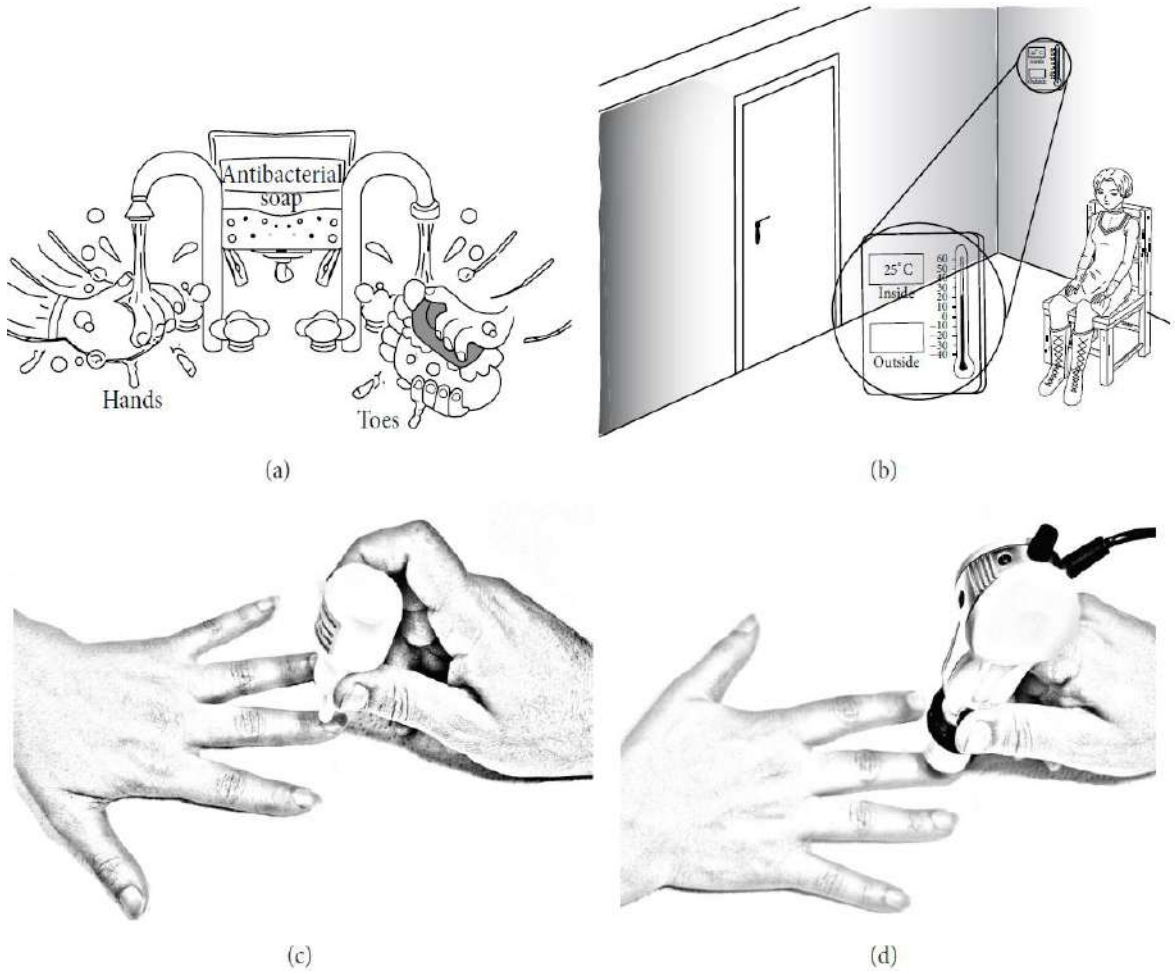
- The study will include patients with DADA2 diagnosis
- Prior to enrolment, the patient's parents and/or children must be willing to participate and assigned informed consent.

Exclusion criteria

- Patients with systemic diseases (systemic lupus erythematosus, Sjogren's syndrome), inflammatory bowel disease, Behçet's disease
- Any major illness or condition, as well as evidence of an unstable clinical condition (eg, cardiovascular, cerebrovascular, neurologic, metabolic, immunologic, infectious, hepatic, renal condition, uncontrolled diabetes mellitus or hypertension)
- Exposure to chronic trauma to the nailbed (farming, nail biting, musical instruments that may cause vibration in the finger)
- Having had a manicure in the last 1 month.

Preparations before the NVC procedure

- The patient is advised to avoid smoking and caffeine at least 4-6 hours before the examination
- Hands are cleaned with soap and water before the examination
- Wait 15-20 minutes at room temperature (20-25 degrees) before the procedure
- Optimum dose of oil (olive oil, immersion oil) is dripped.



NVC evaluations

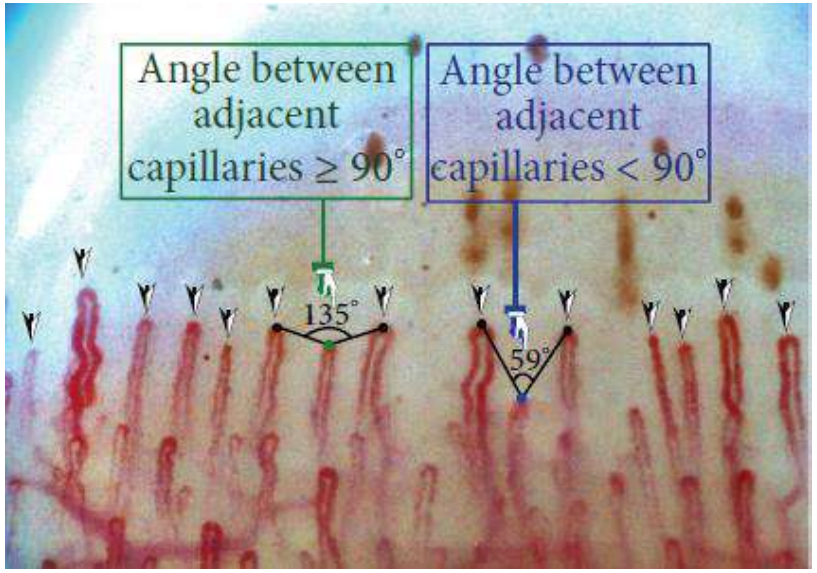
- 200x magnification lens will be employed
- The examination focuses on the eight fingers, excluding the thumbs
  - To calculate the total score for the nailbed evaluation, the following steps are performed
  - For each finger (fingers 2 to 5 on each hand), 4 images are captured
  - The scores obtained from these 2 or more images for each finger are averaged
  - The average scores from the 8 separate fingers are then summed
  - Finally, the summed score is divided by 8 to calculate the total score for the nailbed evaluation.

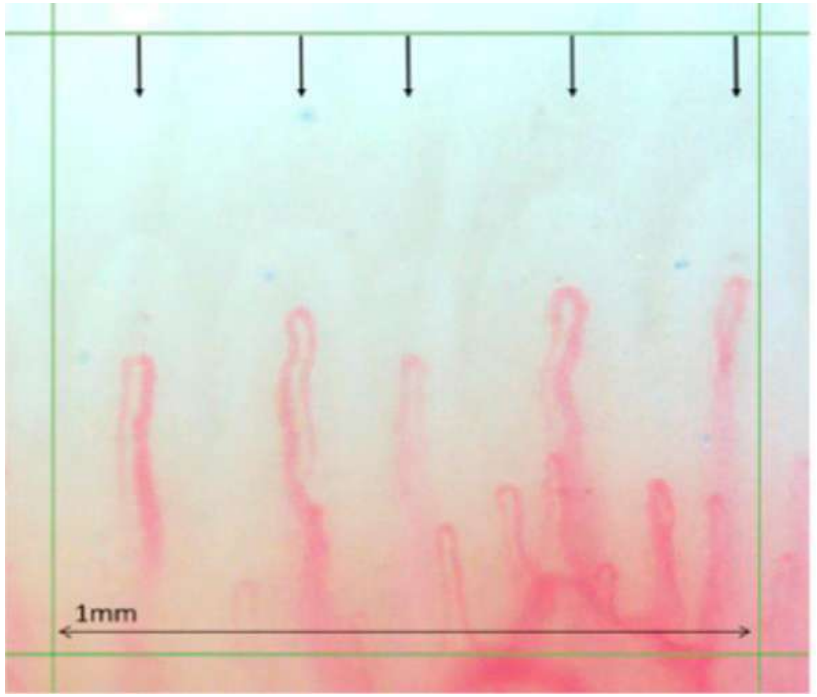
NVC evaluations

- Capillary morphology
- Capillary density
- Capillary width
- Inter-capillary distance
- Microhemorrhage
- Avascular area
- Neovascularisation

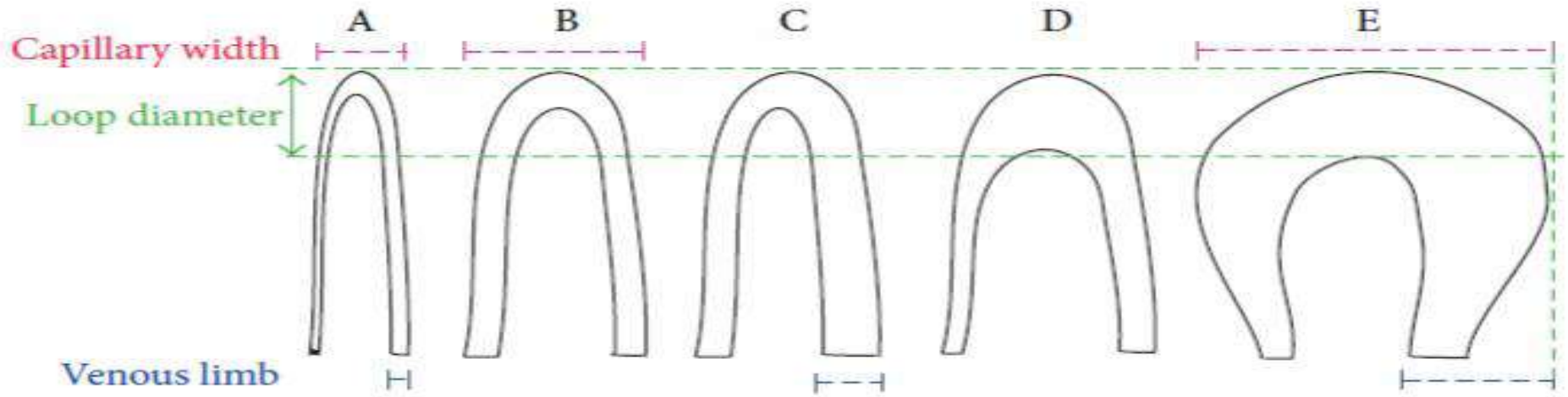
Capillary density

- The number of capillaries 1 mm area in the distal row of each finger
- Despite age-related variations, more than 9 capillaries are considered normal
- A capillary ring is considered to be in the distal row if the angle between the apex of one capillary ring and two adjacent capillary rings is greater than 90°.





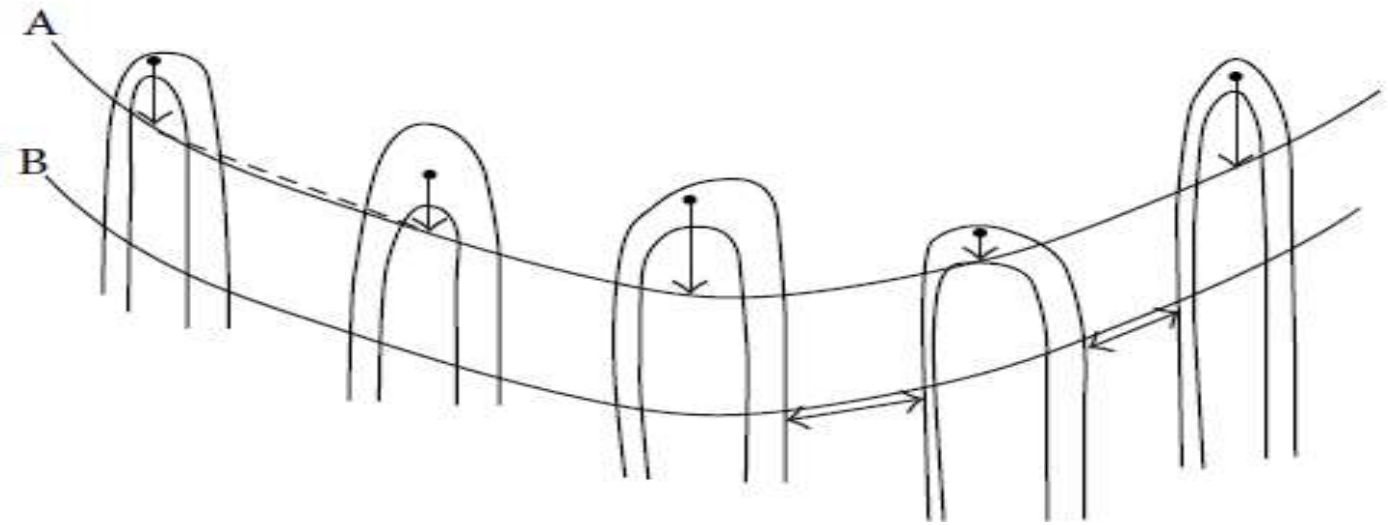
Capillary Width



Normal Dilate capillaries ≥20 microns, ≤ 50 microns Giant capillary≥50 mikron

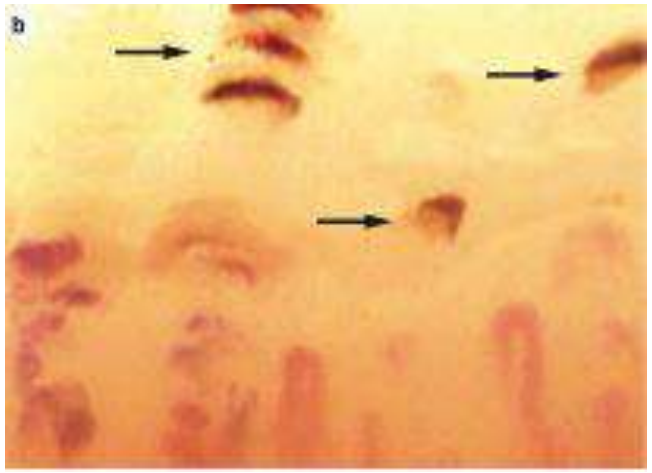
Intercapillary distance

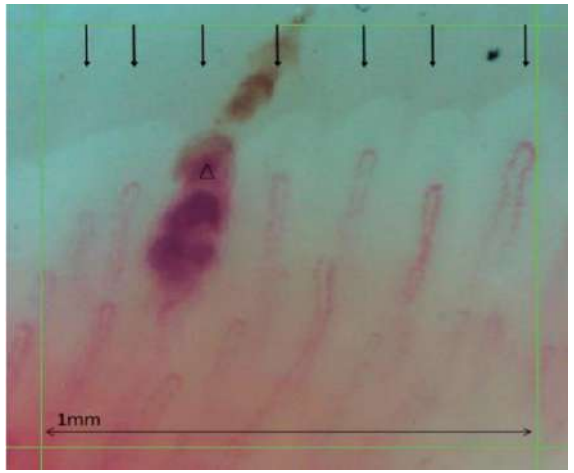
- It is the distance between the apical ends of two capillaries
- Varies between 96 and 166 microns

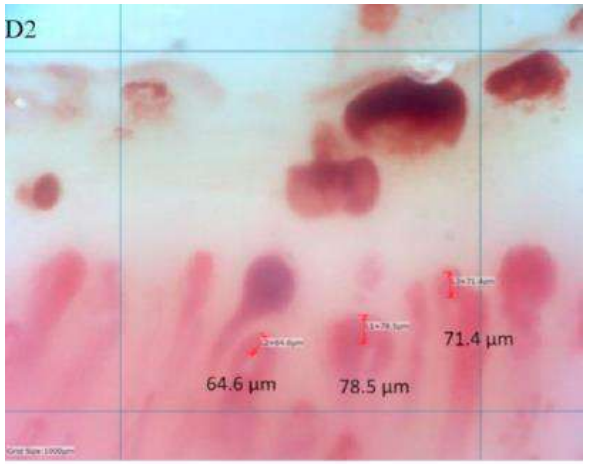


Microhemorrhages

- Reddish-brown punctate lesions in the nail bed
- They represent early vascular damage.

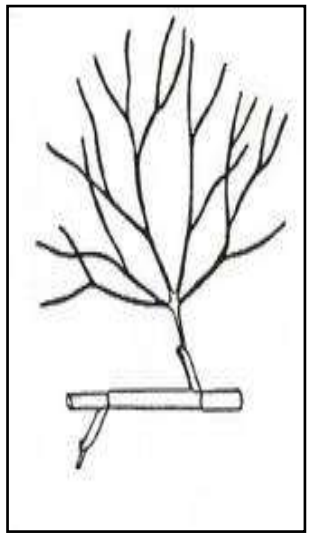


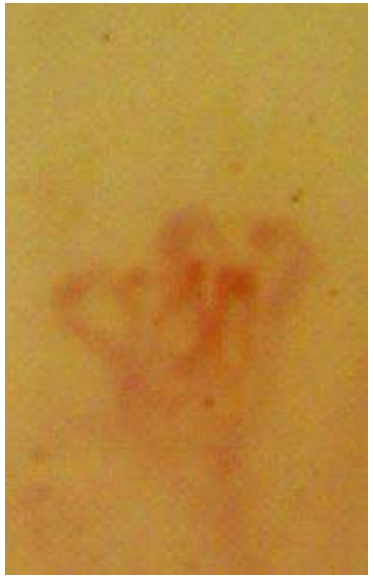


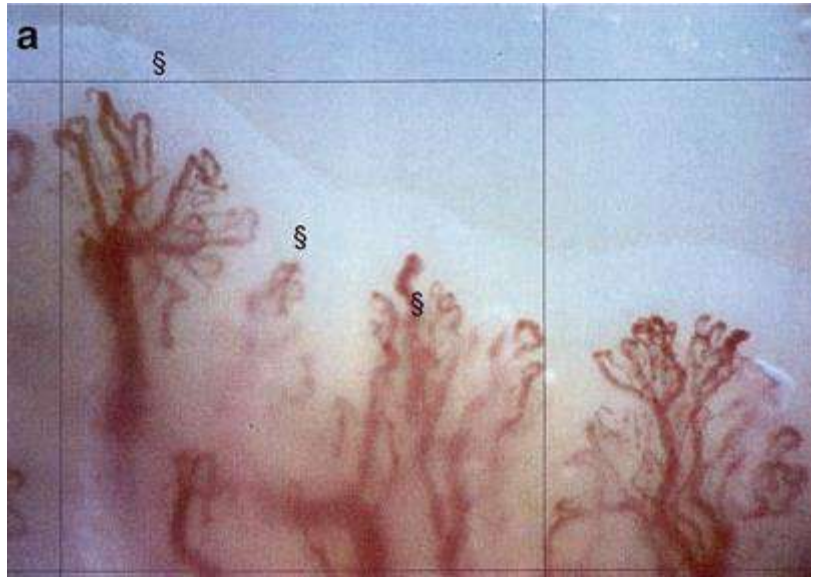


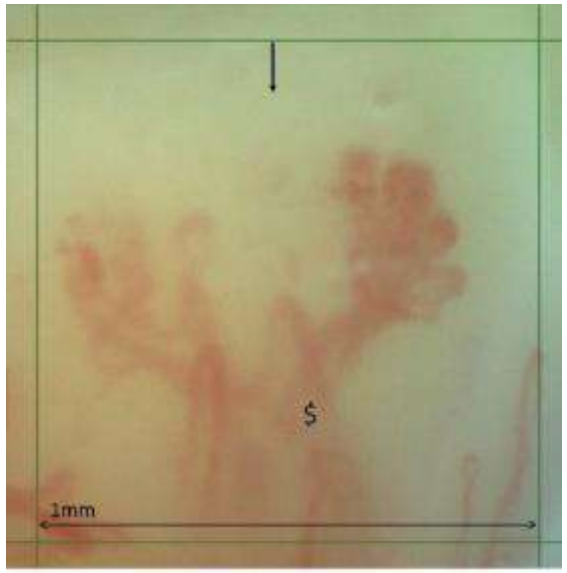
Neoangiogenesis

- ≥4 capillaries in a single dermal papilla
- Thin and branched interconnected capillaries arising from a single capillary loop
- Increase in bushy, branching, ramified capillaries

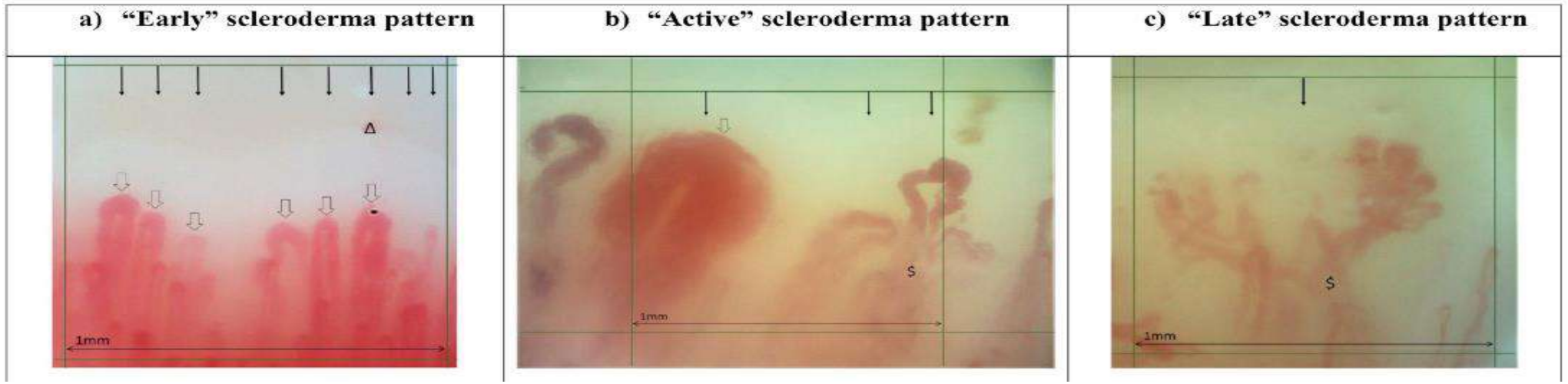








Scleroderma Pattern



	Early	Active	Late
Capillary density	≥7	4-6	≤3
Capillary width	>50 microns	>50 microns	(-)
Abnormal morphology	(-)	+	++
Haemorrhage	+/-	+-/-	(-)

- Thank you for your collaboration….

Şengül Çağlayan (sengulturkercaglayan@gmail.com)
Betül Sözeri (drbetulsozeri@gmail.com)
umraniyepedrhestudy@gmail.com

Sara Stern, Chair of the CARRA Childhood Sjögren's Disease Workgroup:

7. **International Sjögren's Disease registry**

Do these children have Sjögren Disease?

A 7-year-old girl

- Seronegative
- Recurrent bilateral parotitis
- Ultrasound consistent with Sjogren Disease
- Recurrent dental caries

A 15-year-old-girl

- High titer positive ANA and SSA
- Joint pain
- Fatigue

Which children will progress to Sjögren Disease and When?

Working Together and Making Magic in Childhood Sjögren Disease

Our history together:

- Basiaga et al. Childhood Sjögren syndrome: features of an international cohort and application of the 2016 ACR/EULAR classification criteria.
- Randell et al. Pediatric rheumatologists' perspectives on diagnosis, treatment, and outcomes of Sjögren disease in children and adolescents.
- Stern et al. Evaluating a Diagnostic Algorithm for Childhood Sjögren's Disease.
- Treemarcki et al. Medication Use in Childhood Sjögren Disease.

Opportunities for the Future

- **Prospective Registry**
  - Sjögren Disease based off criteria
  - Recurrent or Persistent Parotitis without an underlying diagnosis
  - Elevated SSA/Ro or SSB
  - Subject clinically suspected of having Sjögren Disease in evolution for another reason
- **Become more involved with the International Workgroup**

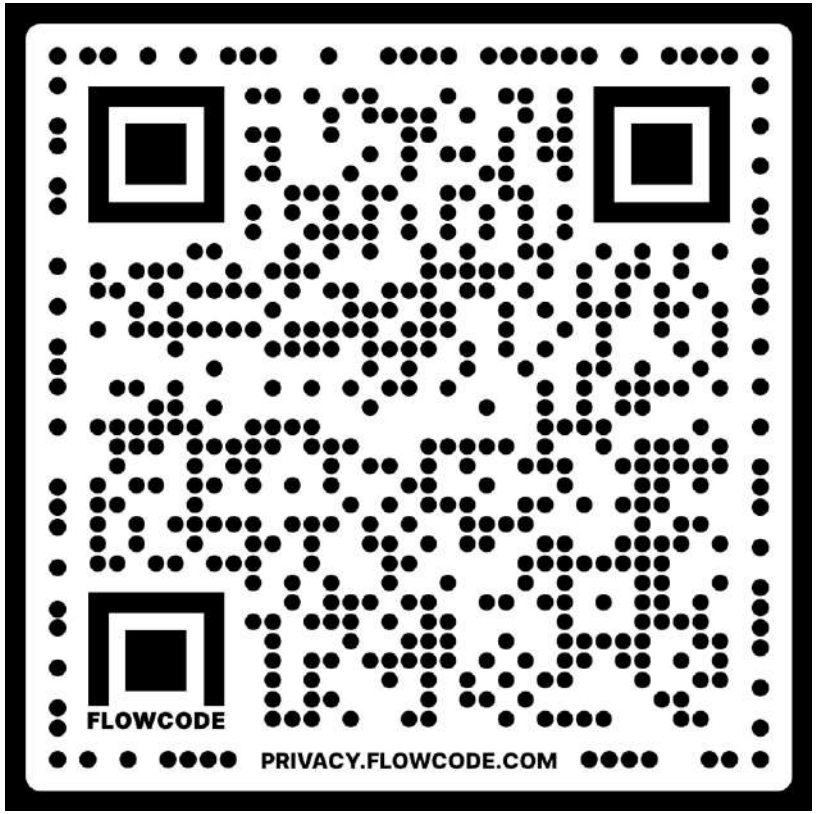
CHildhood Onset Sjögren disease Outcomes Network (CHOSEN)

- This prospective registry was developed to fill this gap and we need your help.
- **We need your help.**
- If you are interested in participating, please contact Matt, Scott, or Sara

Matthew L. Basiaga, DO MSCE (basiaga.matthew@mayo.edu)

Scott M. Lieberman, MD PhD (scott-lieberman@uiowa.edu)

Sara M. Stern, MD (sara.stern@hsc.utah.edu)

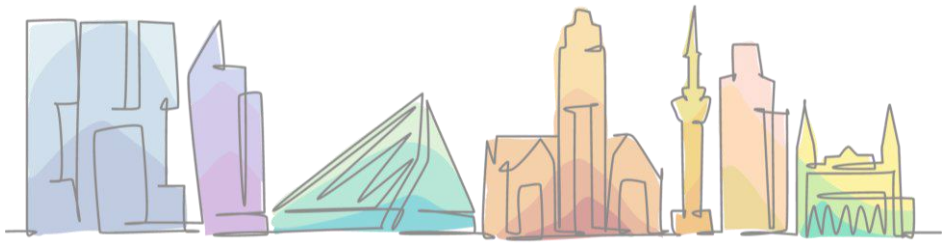


Vasculitis Working Party

- 1. PReS VASCULITIS WP Core team
- 2. SCIENCE AND RESEARCH ACTIVITIES
- 3. **EDUCATIONAL AND TRAINING ACTIVITIES**

PRES 2023

Rotterdam



Vasculitis working party

Educational updates

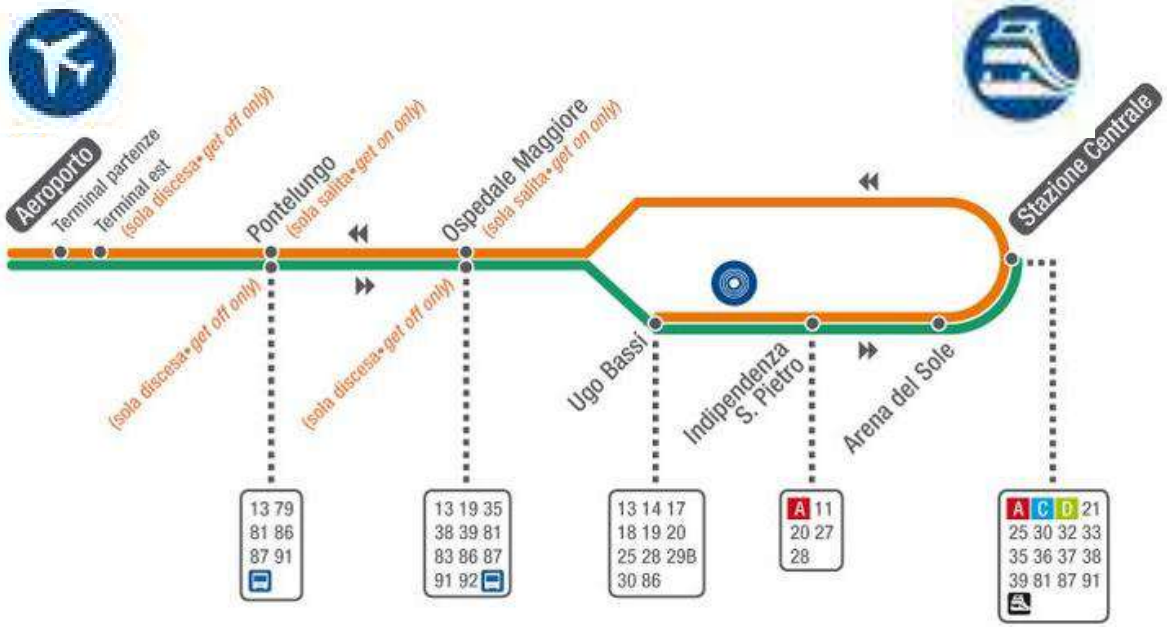
1st International Kawasaki Disease Registry&EUROKiDs Joint Meeting

Inflammation and the Young Heart

Through the Lens of MIS-C and Kawasaki Disease
Cardiology and Rheumatology Perspectives

Bologna


marconi express


Aeroporto
Terminal partenze
Terminal est
(sola discesa•get off only)
Pontelungo
(sola salita•get on only)
Ospedale Maggiore
(sola salita•get on only)
Stazione Centrale
(sola discesa•get off only)
(sola discesa•get off only)
Ugo Bassi
Indipendenza
S. Pietro
Arena del Sole
13 79
81 86
87 91
13 19 35
38 39 81
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Verona
Venezia
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Mare Adriatico
Pescara
Corse
Roma
Napoli
Salerno
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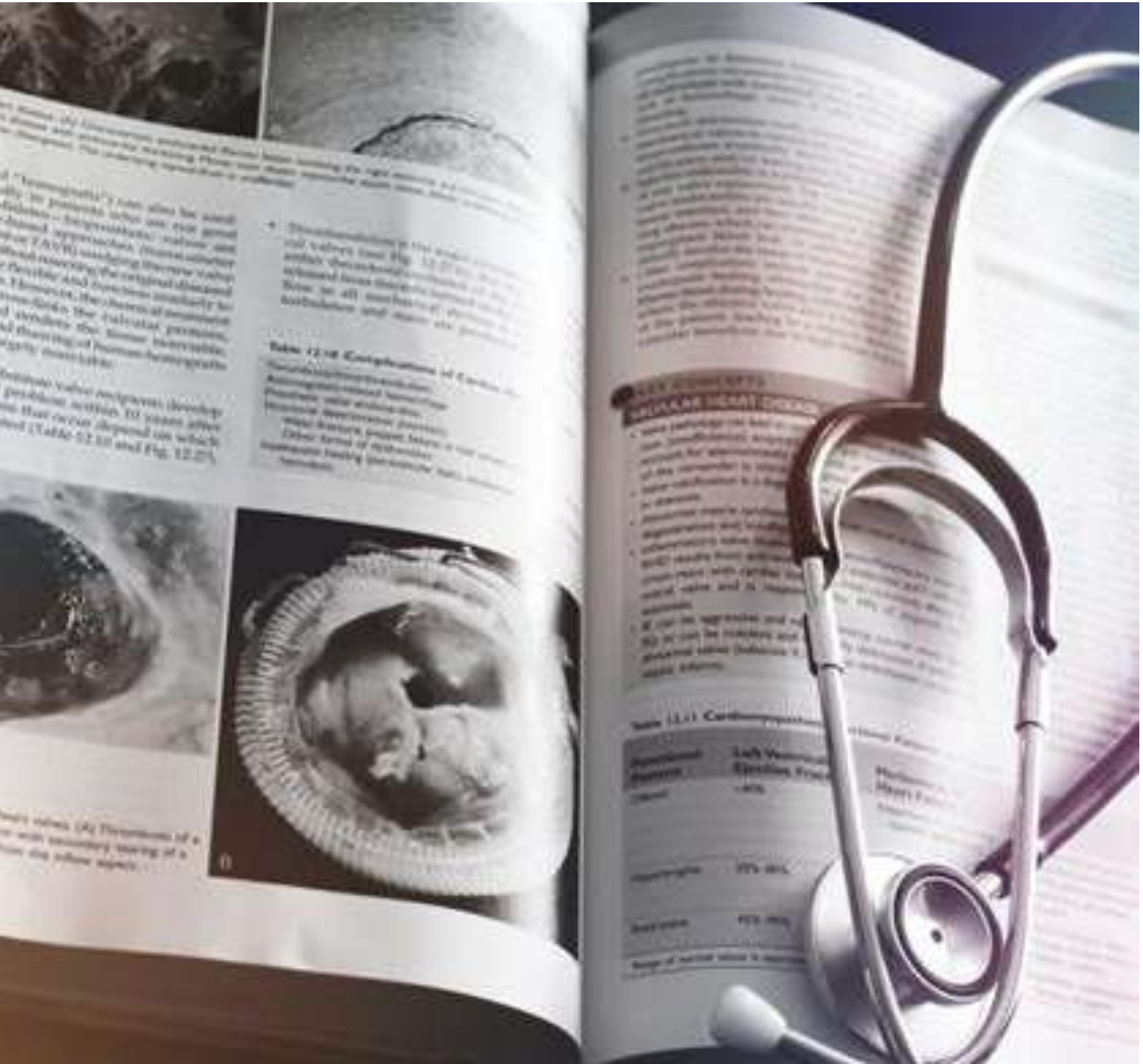
Mar Tirreno
Google

Website:

www.ikdm.info

Email address:

kawasaki.disease23@gmail.com



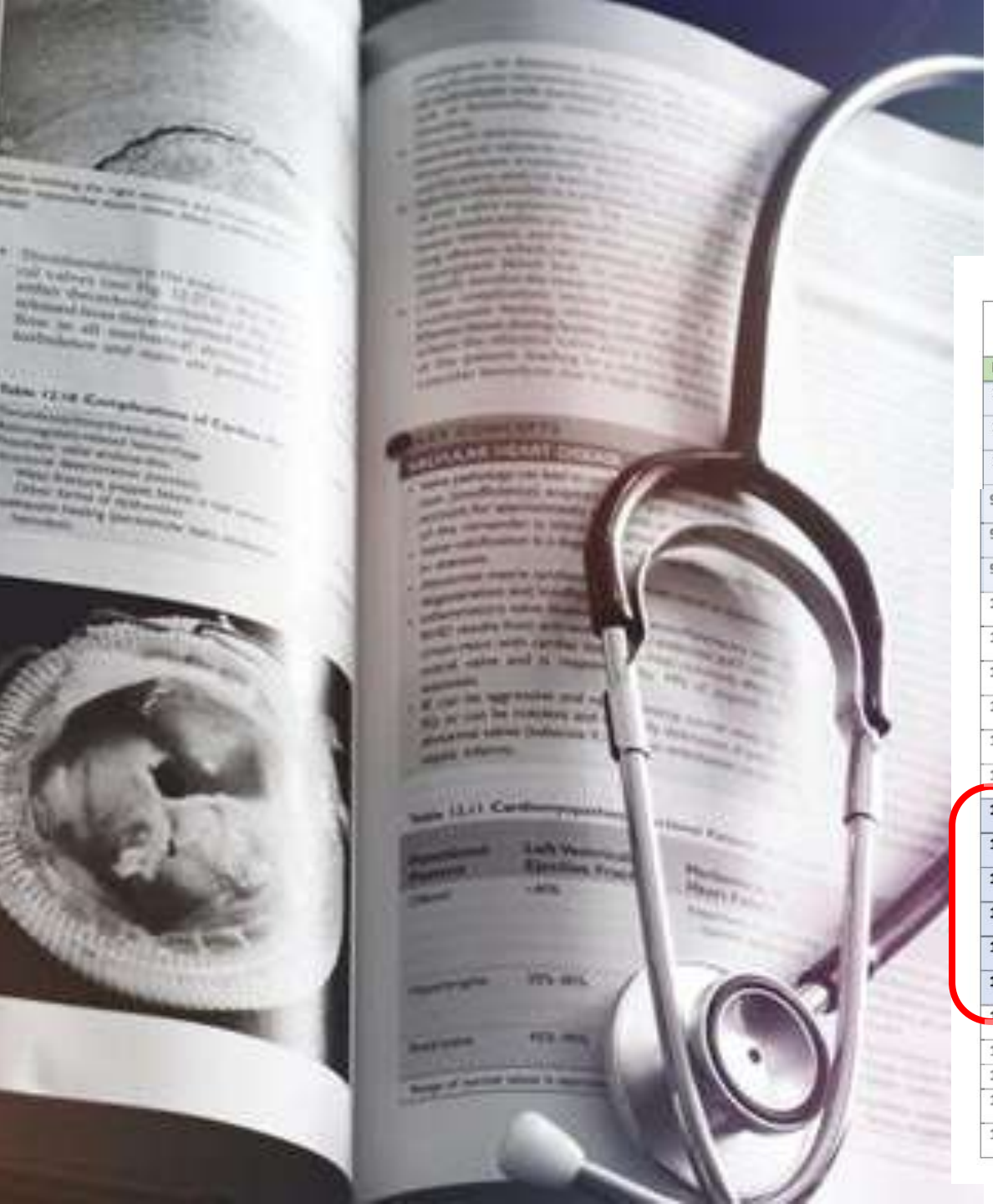
PReS Knowledge Base Exam

To test current knowledge.

Results strictly confidential!

Open to trainees, and to each practicing pediatric rheumatologist from around the world

→ project approved by the Council
→ endorsed by the General Assembly



PReS KBE

Summary of Questions Draft, Need Revision, Rejected, Deleted and Accepted *January 8th, 2023*

	Draft	Submitted	Need Revision	Accepted	Rejected	Deleted	*Total*
DISEASES							
1. JIA (except SJIA) >> Clinical presentation, differential	0	7	2	2	0	0	*11*
1. JIA (except SJIA) >> Diagnosis and classification criteria	1	7	3	1	0	0	*12*
1. JIA (except SJIA) >> Epidemiology and prognosis	0	6	3	0	0	0	*9*
9. JDM >> Etiology, physiology, pathology	0	0	1	4	0	0	*5*
9. JDM >> Exams (Labs, imaging...)	0	1	0	6	1	1	*9*
9. JDM >> Treatment	0	1	0	10	0	0	*11*
10. Scleroderma (systemic and localized) >> Clinical presentation, differential	0	0	13	7	0	0	*20*
10. Scleroderma (systemic and localized) >> Diagnosis and classification criteria	0	0	0	12	0	2	*14*
10. Scleroderma (systemic and localized) >> Epidemiology and prognosis	0	0	1	7	0	0	*8*
10. Scleroderma (systemic and localized) >> Etiology, physiology, pathology	0	0	0	3	0	0	*3*
10. Scleroderma (systemic and localized) >> Exams (Labs, imaging...)	0	0	3	6	0	0	*9*
10. Scleroderma (systemic and localized) >> Treatment	0	0	0	8	1	0	*9*
11. Vasculitis >> Clinical presentation, differential	1	1	4	34	0	22	*62*
11. Vasculitis >> Diagnosis and classification criteria	1	1	1	7	0	8	*18*
11. Vasculitis >> Epidemiology and prognosis	0	0	1	3	0	7	*11*
11. Vasculitis >> Etiology, physiology, pathology	0	0	1	3	0	4	*8*
11. Vasculitis >> Exams (Labs, imaging...)	0	0	3	10	0	8	*21*
11. Vasculitis >> Treatment	1	0	2	9	1	6	*19*
13. Arthritis related to infection >> Clinical presentation, differential	0	1	0	6	0	0	*7*
13. Arthritis related to infection >> Diagnosis and classification criteria	0	0	0	0	0	0	*0*
13. Arthritis related to infection >> Epidemiology and prognosis	0	0	0	0	0	0	*0*
13. Arthritis related to infection >> Etiology, physiology, pathology	0	0	0	1	0	0	*1*
13. Arthritis related to infection >> Exams (Labs, imaging...)	0	0	0	2	0	0	*2*

PRES SCHOOL WEBINARS

- **PRES school webinars:**
  - 3rd Tuesday each month , 12:00 – 13:00 CET

- **Goals:**
  - To review knowledge in pediatric rheumatology from medium to advance level (Aimed to trainees and already practicing pediatric rheumatologists)
  - Important to include transition, patients' perspective, combined sessions with HP,…
  - To build community between PRES members across the world
    - PRES contact point in between congresses
    - To include in one or two slides key messages from society (upcoming courses, recommendations, congress dates,…)

PRES webinars

- **Program:**
  - **1st edition:**
  - One session - **each WP (currently 10 WP** + Global MSK Health WP, Sjogren WP?)
  - 2 speakers, 2 moderators (1 senior, 1 EMERGE)

Template:

- Welcome and Introduction: 3 min
- Brief summary of the WP (strictly brief!): 5 min, focusing on activities and projects that members could join.*The goal of these first edition webinars is not to present the work of the WP.*
- 2 talks of 20 minutes each, 10-15 minutes for discussion.

Further editions:

- To organize a comprehensive program to develop in 2-3 years according to our PRES syllabus
  - Important: To *include transition, patients' perspective*, combined sessions with HP,…
  - To consider *joint sessions* (with other scientific societies ISSAID, ESID, EUVAS, ERN,…)
  - Supported by PRES WP education leaders, but the program must cover all pediatric rheumatology knowledge (beyond diseases included in the WP)

JIR- CliPS project

Kawasaki disease

Vasculitis Working Group

Improving outcome of Juvenile Inflammatory Rheumatism via universally applicable clinical practice strategies, JIR-CliPS

cost EUROPEAN COOPERATION IN SCIENCE & TECHNOLOGY

Funded by the European Union

JIR-CliPS:
Real Life Clinical Practice Strategie

Objectives

The JIR-CliPS project aims to gather real-life clinical practice strategies from physicians globally.

Purpose

Develop a library from which physicians can select the most suitable strategy for their local clinical practice.

WG1: LUPUS NEPHRITIS management in children

WG2: Immunoglobulin A VASCULITIS and KAWASAKI disease management in children

WG3: Use of biological drugs for treatment of AUTOINFLAMMATORY DISEASES

WG4: PFAPA and SURF Syndrome

WG5 : Systemic Juvenile Idiopathic Arthritis (sJIA) and adult onset still disease (AOSD)

JIR CliPS

KAWASAKI DISEASE

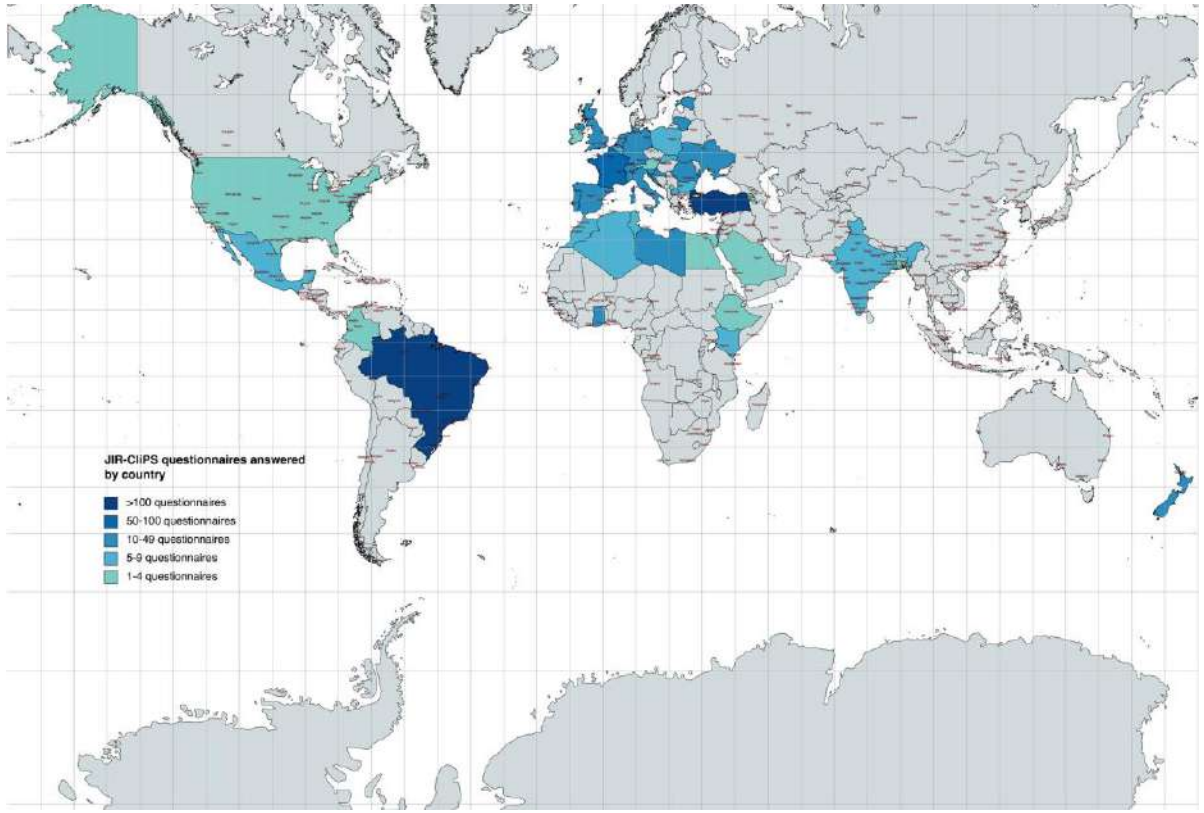
1. Diagnosis
2. First and second line treatment
3. Diagnosis >10° day
4. Follow-up

JIR CLIPS - SEPTEMBER 2023

MORE THAN 1000 QUESTIONNAIRES COMPLETED BY PHYSICIANS FROM 48 COUNTRIES

Didn't share your CliPS yet ?

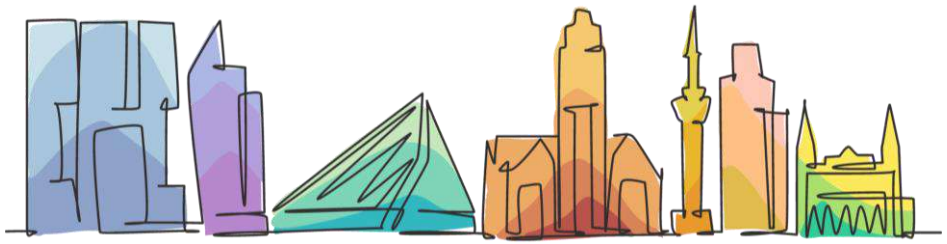
Scan to request the questionnaire




21ST INTERNATIONAL
VASCULITIS
WORKSHOP
BARCELONA
7 - 10 APRIL 2024

PRES 2023

Rotterdam



thank you

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