

**Saturday March 25**

16:40-17:00: Welcome and Welcome lecture (Room A)

**Diversity and Integration of Vasculitis**

Chairs: Toshihiko Nagasawa &amp; Shouichi Fujimoto

16:40-17:00 **W, WL:** Yoshihiro Arimura

17:00-18:00: Fokko Van Der Woude Lecture (Room A)

**ANCA-ASSOCIATED VASCULITIDES: FUTURE THERAPEUTIC STRATEGIES**

Chairs: Kazuo Suzuki &amp; Charles Pusey

17:00-18:00 **Wounded Lecture:** Loic Guillevin "ANCA-ASSOCIATED VASCULITIDES: FUTURE THERAPEUTIC STRATEGIES"

18:00-20:00: Welcome Reception (Exhibition)

**Sunday March 26**

09:00-11:20: Symposium 1 (Room A)

**Classification of AAV: clinical entity vs autoantibody profile**

Chairs: John L. Niles &amp; Caroline Savage

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| 09:00-09:35 | <b>SY1-1:</b> Ronald Falk "Classification of AAV Clinical Entity vs. Autoantibody Profile"  | 09:35-10:10 | <b>SY1-2:</b> Ken Smith "Genetic studies in ANCA-associated vasculitis point to a new, practical disease classification based on autoantibody specificity." |
| 10:10-10:35 | <b>SY1-3:</b> Naoyuki Tsuchiya "Insights from the Genetics of ANCA-associated Vasculitis in Japan."                                 | 10:35-11:00 | <b>SY1-4:</b> Jan Willem Cohen Tervaert "Classification of ANCA-associated vasculitis: Clinical entity versus autoantibody profile"                         |
| 11:00-11:20 | <b>SY1-5:</b> Yoshinori Komagata "Comparison of clinical features between MPO-ANCA- and PR3-ANCA-positive AAV in Japanese patients" |             |   |

10:10-11:20: Workshop 1 (Room B)

**ANCA and Other Biomarkers**

Chairs: Shunsuke Furuta &amp; Yasuaki Aratani

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| 10:10-10:20 | <b>WS1-1:</b> Parameswaran Nair "Sputum Anti-neutrophil Cytoplasmic Antibodies (ANCA) in Eosinophilic Granulomatosis and Polyangiitis (eGPA)"                                | 10:20-10:30 | <b>WS1-2:</b> Shunya Uchida "Clinical Significance of the Existence of Anti-Moesin Antibody in Systemic Vasculitides"  |
| 10:30-10:40 | <b>WS1-3:</b> Meghan Jobson "Immunosignature profiling of patients with ANCA vasculitis reveals novel patterns associated with disease activity and drug-associated disease" | 10:40-10:50 | <b>WS1-4:</b> Divi Corneec "Detection of Circulating PR3-Specific B Cells in Patients with ANCA-Associated Vasculitis and Their Association with Disease Activity" |
| 10:50-11:00 | <b>WS1-5:</b> Benjamin Wilde "GRANZYME B PRODUCING B-CELLS HAVE IMMUNOREGULATORY FUNCTION AND ARE DIMINISHED IN PATIENTS WITH ANCA-VASCULITIS"                               | 11:00-11:10 | <b>WS1-6:</b> Luc Mouthon "Skewed peripheral B and T lymphocytes compartments in patients with ANCA associated vasculitis"   |
| 11:10-11:20 | <b>WS1-7:</b> Anouk von Borstel "Increased CD38hiCD27+ Plasmablast Frequency in Remission Predicts Relapsing Disease in Granulomatosis with Polyangiitis Patients"           |             |  |

11:30-12:30: Case Discussion (Room A)

Chairs: Charles Jennette &amp; Eri Muso

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| 11:30-11:50 | <b>CD-1:</b> Shigeto Kobayashi "Large-vessel Giant cell arteritis: a report of three cases"<br><b>Commentator:</b> Maria Cid  | 11:50-12:10 | <b>CD-2:</b> Shuichiro Endo "Histological sign of smoldering ANCA-associated nephritis years before burst of RPGN in two cases of IgA nephropathy"<br><b>Commentator:</b> Franco Ferrario |
| 12:10-12:30 | <b>CD-3:</b> Akiko Hoshino "Relapse of pulmonary hemorrhage in a patient with pulmonary-renal syndrome with a sequential rise in MPO-ANCA and anti-GBM antibody levels"<br><b>Commentator:</b> Vladimir Tesar |             |   |

12:40-13:40: Luncheon Seminar 1 (Room A)

Chair: Yoshihiro Arimura

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12:40-13:40 **LS1:** David Jayne "Role of Rituximab in the current treatment of ANCA associated vasculitis"

12:40-13:40: Luncheon Seminar 2 (Room B)

Chair: Hirofumi Makino

Sponsored by NIPRO CORPORATION

12:40-13:40 **LS2:** Maarten Segelmark "1st, 2nd and 3rd generation of ANCA testing"

13:50-15:55: Symposium 2 (Room A)

**Kawasaki Disease: history and step to next stage**

Chairs: Seza Ozen & Tsutomu Saji

13:50-14:25 **SY2-1:** Tomisaku Kawasaki "Recognition of Kawasaki Disease"

14:25-14:50 **SY2-2:** Yoshikazu Nakamura "Epidemiologic features of Kawasaki disease and lessons from them"

14:50-15:15 **SY2-3:** Yoshihiro Onouchi "Genetic factors of Kawasaki Disease"

15:15-15:40 **SY2-4:** Tohru Kobayashi "Acute management of Kawasaki disease"

15:40-15:55 **SY2-5:** Hideyuki Nakaoka "MicroRNA-145-5p and microRNA-320a upregulate inflammatory cytokine expression levels and contribute to the progression of vasculitis in acute Kawasaki Disease."

14:30-15:20: Workshop 2 (Room B)

**Epidemiology**

Chairs: Maarten Segelmark & Hirofumi Makino

14:30-14:40 **WS2-1:** Raaashid Luqmani "The UK & Ireland Vasculitis Registry (UKIVAS): Cross-sectional data on the first 2290 Patients with anti-neutrophil cytoplasm (ANCA) associated vasculitis (AAV)"

14:40-14:50 **WS2-2:** Nishkantha Arulkumaran "Outcomes in ANCA-Associated Vasculitis - a 40 Year Retrospective Cohort Study"

14:50-15:00 **WS2-3:** Zdenka Hruskova "Chronic Damage in Patients with ANCA-Associated Vasculitis: Data from a National Registry"

15:00-15:10 **WS2-4:** Mara Felicetti "Change of Perspective: Anca Pattern Based Analysis in Granulomatosis with Polyangiitis and Microscopic Polyangiitis."

15:10-15:20 **WS2-5:** Nobuyuki Ono "Characteristics of MPO-ANCA positive GPA comparing to PR3-ANCA positive GPA and MPA."

15:25-15:55: Workshop 3 (Room B)

**Genetics**

Chairs: Ronald Falk & Naoyuki Tsuchiya

15:25-15:35 **WS3-1:** Toshiki Nakajima "The Synergistic Effects of HLA-B\*52 and Risk Allele at a SNP in IL12B Region on Complication of Aortic Regurgitation in Patients with Takayasu Arteritis"

15:35-15:45 **WS3-2:** Joshua Ooi "In Anti-GBM Disease, HLA-Peptide Conformation Determines Susceptibility or Protection from Disease"

15:45-15:55 **WS3-3:** Peter Merkel "Identification of Functional and Expression Polymorphisms Associated with Risk for Anti-Neutrophil Cytoplasmic Autoantibody-Associated Vasculitis"

16:30-17:30: Poster 1 (Posters)

P1-1—P1-185

18:00-19:00: Evening Seminar (Room B)

Chair: Masayoshi Harigai

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18:00-19:00 **ES:** Yoshikazu Nakaoka "Update on Large Vessel Vasculitis ~Pathogenesis, Diagnosis and Treatment~"

## Monday March 27

09:00-11:20: Symposium 3 (Room A)

**Pulmonary-limited MPA: a new entity of MPO-AAV or not?**

Chairs: Ulrich Specks & Sakae Homma

09:00-09:35 **SY3-1:** Keishi Sugino "Clinical characteristics of pulmonary-limited microscopic polyangiitis"

09:35-10:00 **SY3-2:** Charles Pusey "PULMONARY INVOLVEMENT IN MICROSCOPIC POLYANGIITIS"

10:00-10:25 **SY3-3:** Yasuyuki Kurihara "Pulmonary Imaging of Microscopic Polyangiitis"

10:25-10:50 **SY3-4:** Tamiko Takemura "Pathologic features of myeloperoxidase anti-neutrophil cytoplasmic antibody positive interstitial pneumonia in comparison with idiopathic pulmonary fibrosis: New insight into the cystic lesions"

10:50-11:15 **SY3-5:** Luis Felipe Flores-Suarez "Pulmonary-limited MPA: a new entity of MPO-AAV or not?"

11:15-11:30 **SY3-6:** Takeshi Saraya "Pulmonary involvement in myeloperoxidase antineutrophil cytoplasmic antibody (MPO-ANCA) related disease. CT findings in 18 patients."

09:40-10:30: Workshop 4 (Room B)  
**Molecular and Cellular Mechanisms 1**  
 Chairs: Peter Heeringa & Akihiro Ishizu

09:40-09:50	<b>WS4-1:</b> Veronique Witko-Sarsat "Role of the cyclin-dependent inhibitor p21/waf1 in proteinase 3-induced macrophage activation"	09:50-10:00	<b>WS4-2:</b> Judith Everts-Graber "Proteomic analysis of neutrophils from GPA demonstrates a dysregulation in apoptotic pathways and a defect in annexin-A1-mediated anti-inflammatory mechanisms"
10:00-10:10	<b>WS4-3:</b> Scott Henderson "Proteinase-3 (PR3) Auto-antigen Drives Giant Cell and Granuloma Formation in ANCA-Associated Vasculitis (AAV)"	10:10-10:20	<b>WS4-4:</b> Lucas Lintermans "Kv1.3 Channel Blockade Modulates the Effector Function of B Cells in Granulomatosis with Polyangiitis "
10:20-10:30	<b>WS4-5:</b> Su-Fang Chen "Myeloperoxidase Influences the Complement Regulatory Activity of Complement Factor H"		

10:35-11:25: Workshop 5 (Room B)  
**TAK, GCA, KD, PAN**  
 Chairs: Elisabeth Brouwer & Kei Takahashi

10:35-10:45	<b>WS5-1:</b> Fatma Alibaz-Oner "Assessment of the Frequency of Cardiovascular Risk Factors in Patients with Takayasu Arteritis "	10:45-10:55	<b>WS5-2:</b> Yoshikazu Nakaoka "Efficacy and Safety of Subcutaneous Tocilizumab in Japanese Patients with Refractory Takayasu Arteritis: A Randomized, Double-Blind, Placebo-Controlled, Phase 3 Trial"
10:55-11:05	<b>WS5-3:</b> Matthew Koster "Differences in Presentation and Outcome in Patients with Giant Cell Arteritis Based on Temporal Artery Biopsy Positivity"	11:05-11:15	<b>WS5-4:</b> Tamihiko Kawakami "Relationship of anti-phosphatidylserine-prothrombin complex antibodies and anti-moesin antibodies in patients with polyarteritis nodosa"
11:15-11:25	<b>WS5-5:</b> Yoshiro Kitagawa "Increased levels of plasma pentraxin 3 predict IVIG resistance and coronary artery lesion formation in patients with Kawasaki disease "		

11:50-12:50: Luncheon Seminar 3 (Room A)  
 Chair: Shouichi Fujimoto  
 Sponsored by Zenyaku Kogyo Co., Ltd.

11:50-12:50	<b>LS3:</b> John Stone "Achieving and Maintaining Remission in AAV:What are we missing?"
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11:50-12:50: Luncheon Seminar 4 (Room B)  
**Behcet's Disease**  
 Chair: Masayoshi Harigai  
 Sponsored by Mitsubishi Tanabe Pharma Corporation

11:50-12:20	<b>LS4-1:</b> Annabelle Okada "Behcet's disease ; Ocular Manifestations and Advances in Treatment"	12:20-12:50	<b>LS4-2:</b> Shunsei Hirohata "Recalcitrant complications of Behcet's Disease"
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13:00-15:30: Symposium 4 (Room A)  
**TAK and GCA: a spectrum within the same disease or not?**  
 Chairs: Peter Merkel & Hajime Yoshifuji

13:00-13:35	<b>SY4-1:</b> Raashid Luqmani "Giant cell arteritis (GCA) and Takayasu arteritis (TAK): a spectrum within the same disease or not? A report from the DCVAS project."	13:35-14:00	<b>SY4-2:</b> Tanaz Kermani "Takayasu arteritis and Giant Cell Arteritis: A Spectrum Within the Same Disease or Not? Review of Epidemiological and Clinical aspects."
14:00-14:25	<b>SY4-3:</b> Yasuhiro Maejima "TXNIP accumulation caused by single nucleotide polymorphism of MLX gene enhances inflammasome formation, thereby promoting the development of Takayasu arteritis"	14:25-14:50	<b>SY4-4:</b> Carol Langford "Takayasu's Arteritis and Giant Cell Arteritis: A Spectrum Within the Same Disease or Not?"
14:50-15:15	<b>SY4-5:</b> David Saadoun "The roles of cytokines in the pathogenesis of TAK and GCA"	15:15-15:30	<b>SY4-6:</b> John Stone "Efficacy and Safety of Tocilizumab in Patient Subgroups With New-Onset and Relapsing Giant Cell Arteritis From a Randomized, Double-Blind, Placebo-Controlled, Phase 3 Trial"

13:40-14:50: Workshop 6 (Room B)  
**Molecular and Cellular Mechanisms 2**  
 Chairs: Alan Salama & Junichi Hirahashi

13:40-13:50	<b>WS6-1:</b> Peiqi Hu "Anti-MPO antibodies cause granulomatosis in mice: an animal model of GPA"	13:50-14:00	<b>WS6-2:</b> Adrian Schreiber "Neutrophil-gelatinase associated lipocalin (NGAL) attenuates ANCA-induced glomerulonephritis by inhibiting Th17 immunity"
14:00-14:10	<b>WS6-3:</b> Kim Maree O'Sullivan "Deoxyribonuclease I Reduces Glomerular Injury and Modulates anti-Myeloperoxidase Autoimmunity in Experimental anti Myeloperoxidase Glomerulonephritis"	14:10-14:20	<b>WS6-4:</b> Holly Hutton "Targeting the NLRP3 Inflammasome in ANCA Vasculitis"
14:20-14:30	<b>WS6-5:</b> Mira Choi "Endothelial NF-kB Blockade by E-selectin Targeted Liposomes Containing P65 SiRNA Abrogates Anti-MPO Antibody-induced Glomerulonephritis"	14:30-14:40	<b>WS6-6:</b> Megan Huynh "HLA-DR15 Inhibition Attenuates Experimental Autoimmune Anti-Glomerular Basement Membrane Disease"
14:40-14:50	<b>WS6-7:</b> Yusuke Nishioka "Rat type II NKT cell clone pathogenic for small vessel vasculitis recognizes sterol carrier protein 2."		

14:55-15:30: Workshop 7 (Room B)  
**QOL and Health Economics**  
 Chairs: Min Chen & Toshiko Ito-Ihara

14:55-15:05	<b>WS7-1:</b> Maria Weiner "Organ Damage and Hospitalization in Elderly Patients with Anti-Neutrophil Cytoplasmic Antibody (ANCA)-Associated Vasculitis"	15:05-15:15	<b>WS7-2:</b> Christian Pagnoux "Impact of Vasculitis on Employment and Income"
15:15-15:25	<b>WS7-3:</b> Loic Guillevin "Economic evaluation of rituximab versus azathioprine for maintenance treatment of ANCA-associated vasculitis: the MAINRITSAN trial"		

16:00-17:00: Poster 2 (Posters)  
**P2-1—P2-177**

19:00-21:00: GALA Dinnar (Hotel Chinzanso Tokyo)

**Tuesday March 28**

09:00-10:50: Symposium 5 (Room A)  
**Behcet Disease**  
 Chairs: Eric Matteson & Yoshinari Takasaki

09:00-09:35	<b>SY5-1:</b> Nobuhisa Mizuki "Genome-wide association study for Behcet's disease"	09:35-10:00	<b>SY5-2:</b> Alfred Mahr "MHC class I antigen and Behcet's disease"
10:00-10:25	<b>SY5-3:</b> Mitsuhiro Takeno "Vascular involvement of Behcet's disease"	10:25-10:50	<b>SY5-4:</b> Shunsei Hirohata "Neurological involvement in Behcet's disease"

09:40-10:50: Workshop 8 (Room B)  
**Head and Neck Involvement and Thrombosis in AAV**  
 Chairs: Tamihiko Kawakami & Yasuaki Harabuchi

09:40-09:50	<b>WS8-1:</b> Kan Kishibe "Otitis media with ANCA-associated vasculitis (OMAAV): A retrospective multicenter study in Japan, Clinical findings."	09:50-10:00	<b>WS8-2:</b> Yuka Morita "Otitis Media with ANCA-associated Aasculitis (OMAAV): A Retrospective Multi-center Study in Japan, Clinical feature of the OMAAV Patients with Facial Nerve Palsy and/or Hypertrophic Pachymeningitis"
10:00-10:10	<b>WS8-3:</b> Kaori Tateyama "Clinical differences of otitis media with ANCA-associated vasculitis based on the type of ANCAs: a retrospective multi-center study in Japan"	10:10-10:20	<b>WS8-4:</b> Naohiro Yoshida "Audiological Findings and Outcome after Immunosuppressive Therapy in Otitis Media with ANCA-associated Vasculitis (OMAAV): A Retrospective Multi-center Study in Japan"
10:20-10:30	<b>WS8-5:</b> Ashima Makol "Clinical Characteristics of Inflammatory Eye Disease in Patients with Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis: A Retrospective Cohort Study"	10:30-10:40	<b>WS8-6:</b> Pavel Novikov "Venous Thrombotic Events in Patients with ANCA-Positive Vasculitides"
10:40-10:50	<b>WS8-7:</b> Charles Pusey "ANCA-Associated Vasculitis is Associated with a High Rate Of Arterial and Venous Thrombosis"		

11:00-12:00: Special Lecture (Room A)  
**Control of immune responses by regulatory T cells**  
 Chairs: Wolfgang Gross & Yoshihiro Arimura

11:00-12:00	<b>SL:</b> Shimon Sakaguchi "Control of immune responses by regulatory T cells"
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12:20-13:20: Luncheon Seminar 5 (Room A)  
Chair: Akihiro Ishizu  
Sponsored by TEIJIN PHARMA LIMITED

12:20-12:50 **LS5-1:** Takashi Kanda 12:50-13:20 **LS5-2:** Tsutomu Saji

12:20-13:20: Luncheon Seminar 6 (Room B)  
Chair: Toshiko Ito-Ihara  
Sponsored by Thermo Fisher Diagnostics K.K.

12:20-13:20 **LS6:** Xavier Bossuyt "New paradigm for ANCA detection in vasculitis."

13:30-15:25: Symposium 6 (Room A)  
**New drugs and Ongoing clinical trial for Vasculitis**  
Chairs: Yoshihiro Arimura & Shigeto Kobayashi

13:30-14:05 **SY6-1:** Michael Walsh 14:05-14:30 **SY6-2:** Rona Smith "Mepolizumab for the treatment of eosinophilic granulomatosis with polyangiitis."  
14:30-14:55 **SY6-3:** Loic Guillevin "New therapeutic approaches in eosinophilic granulomatosis with polyangiitis (Churg-Strauss)" 14:55-15:10 **SY6-4:** Benjamin Terrier "Rituximab Versus Azathioprine to Maintain Remission of ANCA-Associated Vasculitides (MAINRITSAN): Follow-Up At 60 Months"  
15:10-15:25 **SY6-5:** Janneke Tuin "Mycophenolate mofetil versus Cyclophosphamide for the Induction of Remission in Non-life-threatening Relapses of PR3 and MPO-ANCA Associated Vasculitis"

14:10-15:20: Workshop 9 (Room B)  
**Renal Pathology, Classification and Scoring**  
Chairs: Ingeborg Bajema & Kensuke Joh

14:10-14:20 **WS9-1:** Emma van Daalen "International Validation Study for the Histopathological Classification of ANCA-Associated Glomerulonephritis (AAGN)" 14:20-14:30 **WS9-2:** Yayoi Ogawa "Pathological Analysis on the Role of Small Vessel Vasculitis and Tubulo-interstitial Lesions in ANCA-associated Nephritis"  
14:30-14:40 **WS9-3:** Tomomi Endo "Reevaluation of Combining Chronic Tubulo-interstitial Parameters on the Glomerular Histological Classification to Predict the Long Term Outcome in Japanese Cohort of MPO-ANCA Associated GN" 14:40-14:50 **WS9-4:** Noriko Uesugi "What glomerular morphology causes interobserver disagreement?"  
14:50-15:00 **WS9-5:** Layla Alharbi "Prevalence of Immunofluorescent Deposits in ANCA Associated Vasculitis" 15:00-15:10 **WS9-6:** Gerjan Dekkema "Combined Measurement Of Urinary sCD25 And sCD163 As Biomarker For Active ANCA Associated Renal Vasculitis"  
15:10-15:20 **WS9-7:** Sarah Moran "Utility of Measurements of Urinary Soluble CD163 & MCP1 in the Identification of Subtle Renal Flares in ANCA Vasculitis"

15:30-16:25: Closing Session (Room A)  
**Ongoing and Future Vasculitis Studies**  
Chairs: David Jayne & Shouichi Fujimoto

15:30-15:45 **CS-1:** Elena Csernok "Revised international consensus recommendation on testing of anti-neutrophil cytoplasm antibodies (ANCA) in small vessel vasculitis: evaluation of ANCA testing by antigen specific assays" 15:45-16:00 **CS-2:** Masayoshi Harigai "A clinical trial of tocilizumab for microscopic polyangiitis and granulomatosis with polyangiitis"  
16:00-16:25 **CS-3:** Peter Merkel "Current and Future Clinical and Translational Research Studies in Vasculitis"

16:25-16:30: Closing Remarks (Room A)  
**Acknowledgements and Information of Next Workshop**  
Chairs: David Jayne & Shouichi Fujimoto

16:25-16:30 **CR:** Yoshihiro Arimura