



THE FIFTH INTERNATIONAL SYMPOSIUM ON
IgG4-RELATED DISEASE

MILAN

4-6 APRIL
2024

Starhotels Rosa Grand
Piazza Fontana 3 Milan - Italy

The **5th International Symposium on IgG4-Related Disease** will be held from Thursday April **4th to Saturday April 6th**, 2024 at the Star Rosa Grand Hotel (Milan, Italy). The 1st International Symposium on IgG4-Related Disease was held in Boston (US) in 2011. At that Symposium, investigators agreed upon the name for the condition - “IgG4-Related Disease” - and codified other thinking around nomenclature and the pathology features of the disease. The 2nd and 3rd International Symposia on IgG4-Related Disease were held in Hawaii (US) in 2014 and 2017 respectively. At those gatherings, attendees elaborated upon the common clinical presentations of the diseases, established diagnostic standards for each organ involvement, formulated integrated diagnostic guidelines, and launched the first set of classification criteria for this condition. The 4th International Symposium on IgG4-Related Disease, held virtually in 2021 in Kitakyushu (Japan) during the pandemic, began to review treatment guidelines and ongoing investigational approaches to treatment.

The 5th International Symposium on IgG4-Related Disease will bring together international experts to inform, debate, and discuss novel aspects in the field. The Agenda topics include disease classification, organ-specific manifestations, diagnostic features, immunopathogenesis, and present and future treatment approaches.

Opportunities for multidisciplinary work on current and future trials will be highlighted.

In addition, an entire morning session will be dedicated to patients, patient associations, and patient dedicated initiatives.

Thus, over a remarkably short time, IgG4-Related Disease has been identified and characterized and effective targeted treatments capable of achieving regulatory approval now appear possible.

THURSDAY APRIL 4th

- 11.00 AM** WELCOME - OPENING CERIMONY
Moderators: *Emanuel Della-Torre/John Stone/Yoshiya Tanaka*
- 11.05 AM** Unmet needs and open research fields in IgG4-related disease *Emanuel Della Torre*
- 11.40 AM** IgG4-related disease: Twenty years from discovery to IgG4ward! *John H Stone*
- 12:00 PM** What's New (WIN) for PATIENTS
Presentation of the European Reference Network Working Group on IgG4-related disease *Tobias Alexander*
- 12:30 PM** Patient experience and PROMs definition: a round table with ERN-ReConnet and IgG4ward! ePAG representatives *Ilaria Galetti*
- 01:00 PM** *BUFFET LUNCH*

WHAT'S NEW IN THE PATHOGENESIS OF IgG4-RELATED DISEASE - PART I

Moderators: *Shiv Pillai/ Cory Perugino*

- 02.00 PM** Overview of IgG4RD pathogenesis *Cory Perugino*
- 02.20 PM** Updates on the role of Th / Tfh cells in the pathogenesis of IgG4-related disease *Shingo Nakayamada*
- 02.40 PM** Single cell interrogation of T-lymphocytes infiltrating IgG4-related disease affected tissues: a novel perspective into the pathogenesis of IgG4-related disease *Takashi Maehara*
- 03.00 PM** Unusual B-lymphocytes subsets in the pathogenesis of IgG4-related disease: is there a role for “double negative” B-cells?
Hugues Allard-Chamard
- 03.20 PM** Single cell interrogation of B-lymphocytes infiltrating IgG4-related disease affected tissues: a novel perspective into the pathogenesis of IgG4-related disease *Cai Shaozhe*

- 03.40 PM** From Fab-arm exchange to glycosylation: unique properties of IgG4 antibodies in the pathogenesis of IgG4-related disease and in vivo models to study them *Teo Riespens*
- 04.00 PM** Controversies around the complement system in the pathogenesis of IgG4-related disease *Guy Katz*
- 04.30 PM** *COFFEE BREAK*

WHAT'S NEW IN THE PATHOGENESIS OF IgG4-RELATED DISEASE - PART II

Moderators: *Hugues Allard-Chamard/Takashi Maehara*

- 05.00 PM** ABSTRACT/ORAL PRESENTATION (4)
- 06.00 PM** LECTURE: T-follicular regulatory cells at the cross-road between autoimmunity and allergy *Carola Vinuesa*
Introduced by: *Shiv Pillai*
- 06.30-08.30 PM** POSTER SESSION "WINE and CHEESE"

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FRIDAY APRIL 5th

WHAT'S NEW IN THE EPIDEMIOLOGY OF IgG4-RELATED DISEASE

Moderators: *Ulrich Beuers/Zachary Wallace*

- 08.30 AM** LECTURE: Sex-related immunological differences in autoimmunity: where does IgG4-related disease stand?
Coziana Ciurtin
- 09.00 AM** The changing epidemiology of IgG4-related disease. What we learned in the last 20 years *Zachary Wallace*
- 09.20 AM** Genetic susceptibility of IgG4-related disease *Emanuel Della Torre*
- 09.40 AM** ABSTRACT/ORAL PRESENTATION (4)
- 10.30 AM** *COFFEE BREAK*

CLINICAL MANIFESTATIONS OF IgG4-RELATED DISEASE: "ANOTHER GREAT MIMICKER" - PART II - UEG/EPC/EASL JOINT SESSION

Moderators: *Matthias Lohr/ Luca Frulloni/ Gabriele Capurso/ Pietro Invernizzi*

- 11.00 AM** Differentiating type 1 and type 2 autoimmune pancreatitis: diagnostic and clinical implications *Matthias Lohr*
- 11.20 AM** IgG4-related cholangitis and hepatitis: controversies in the diagnosis and management *Ulrich Beuers*
- 11.40 AM** Inflammatory Bowel "IgG4-related disease": myth or reality? *Ferdinando D'Amico*
- 12.00 PM** Role of endoscopy for biliary tree and pancreatic involvement of IgG4-related disease: a practical appraisal
Mariana Arvanitaki
- 12.20 PM** Post-surgery outcomes of pancreato-biliary IgG4-related disease *Miroslav Vujasinovic*

12.40 PM Organ damage in IgG4-related disease and management of secondary comorbidities *Emma Culver*

01.00 PM *BUFFET LUNCH*

CLINICAL MANIFESTATIONS OF IgG4-RELATED DISEASE: “ANOTHER GREAT MIMICKER” - PART II

Moderators: *Mikael Ebbo/Mitsuhiro Kawano*

2.00 PM Differentiating “fibrotic” and “lymphoproliferative” IgG4-related disease: why it matters *Wen Zhang*

2.20 PM Primary and secondary IgG4-related disease *Nicholas Schleinitz*

2.40 PM IgG4-related disease as a novel form of systemic vasculitis *James Stone*

03.00 PM Retroperitoneal fibrosis: when is IgG4-related and when is not *Augusto Vaglio*

03.20 PM Renal involvement of IgG4-related disease *Dario Roccatello*

03.40 PM Salivary gland involvement of IgG4-related disease: the role of salivary gland biopsy and ultrasound in the diagnostic process *Masafumi Moriyama*

04.00 PM Lymph-node involvement of IgG4-related disease: how to extricate in the differential diagnosis *Luke Chen*

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SATURDAY APRIL 6th

WHAT'S NEW IN THE DIAGNOSIS OF IGG4-RELATED DISEASE - PART I - HOW TO ACHIEVE A CORRECT DIAGNOSIS

Moderators: *Fernando Martinez Valle/Tobias Alexander*

- 08.30 AM** Pathology of IgG4-related disease (when IgG4-related disease is not IgG4-related disease) *Yoh Zen*
- 08.50 AM** Update on available biomarkers of IgG4-related disease *Marco Lanzillotta*
- 09.10 AM** Distinguishing active/fibrotic IgG4-related disease. The role of 18FDG-PET and new tracers *Andreas Ramming*
- 09.30 AM** How to image active and fibrotic IgG4-related disease *Diego Palumbo*
- 09.50 AM** IgG4-related disease clinical phenotypes and their impact on patient management *Andreu Fernandez-Codina*
- 10.10 AM** Clinical utility of the 2019 ACR/EULAR classification criteria for IgG4-related disease 5 years down the road *Viniciane Rebours*
- 10.30 AM** *COFFEE BREAK*

WHAT'S NEW IN THE DIAGNOSIS OF IGG4-RELATED DISEASE - PART II - OVERLOOKED DIFFERENTIAL DIAGNOSIS

Moderators: *Nicholas Schleinitz/Marco Lanzillotta*

- 11.00 AM** Castleman's Disease *Éric Oksenhendler*
- 11.30 AM** Autoimmune lymphoproliferative disease *Frederic Rieux-Laucat*
- 12.00 PM** Systemic Histiocytoses *Lorenzo Dagna*
- 12.30 PM** Mesenchymal tumors and sclerosing mesenteritis *Alessandro Gronchi*
- 01.00 PM** *BUFFET LUNCH*

WHAT'S NEW IN THERAPY OF IGG4-RELATED DISEASE – PART I

Moderators: *Dario Roccatello/Ling Li Dong*

- 02.00 PM** Current available treatment guidelines for IgG4-related disease: is it time to change? *Fernando Martinez Valle*
- 02.20 PM** Glucocorticoids - what the best scheme for induction and maintenance of remission of IgG4-related disease *Yoshiya Tanaka*
- 02.40 PM** Is there still a role for DMARDs in the treatment of IgG4-related disease? *Fei Yunyun*
- 03.00 PM** How to define IgG4-related disease flare: do we have reliable “flare criteria”? *Matthew Baker*
- 03.20 PM** Is there a time to stop immunosuppressive therapy in patients with IgG4-related disease? *Arezou Koshroshahi*
- 03.40 PM** Is spontaneous remission of IgG4-related disease possible and what does it teach us about disease pathogenesis?
Mitsuhiro Kawano
- 04.00 PM** Long-term outcomes of IgG4-related disease: is there an increased risk of solid or hematologic malignancies? *Jan Van Laar*
- 04.30 PM** *COFFEE BREAK*

WHAT'S NEW IN THERAPY OF IgG4-RELATED DISEASE – PART II

Moderators: *Wen Zhang / Emma Culver*

- 05.00 PM** ABSTRACT/ORAL PRESENTATIONS (4)
- 06.00 PM** B-cell depletion in IgG4-related disease: pros and cons from a 10 years experience *Mikael Ebbo*
- 06.20 PM** Repurposing biologic agents for treating IgG4-related disease: what can we learn from available case series *Ling Li Dong*
- 06.40 PM** Updates from clinical trials in IgG4-related disease *John H Stone*
- 07.00 PM** CONCLUSIVE REMARKS *Emanuel Della Torre*
- SOCIAL DINNER - CLOSING CERIMONY*
Lecture *Salvatore Mangione*

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