

Welcome to Waikiki and the Second International Symposium on IgG4-Related Disease and Associated Conditions

We are delighted to welcome you to Waikiki, and to the Second International Symposium on IgG4-Related Disease (IgG4-RD) and Associated Conditions.

Since the time of the First International Symposium on IgG4-RD in Boston less than two and a half years ago, the field of IgG4-RD has continued to expand at a remarkable rate. A quick look at PubMed confirms the dramatic growth of writing and interest pertaining to IgG4-RD and its associated conditions. Relevant topics have been added by the week. In short, it is no longer possible to have read all of the potentially important papers in this field: we truly need an international gathering such as the one we have planned in order to share the hottest ideas, focus and re-focus our efforts, and continue to move the field forward. No fewer than 100 abstracts will be presented at this Second Symposium. This fact, as much as any, testifies to the ferment of interest in IgG4-RD.

This Second International Symposium occurs at a time of significant change in IgG4-RD. It is safe to say that the clinical “descriptive” period of IgG4-RD is now coming to an end. Up until not long ago, it was a common occurrence to read, yet again, about another clinical manifestation of IgG4-RD – a new organ system manifestation detected; a novel radiology feature identified; another seemingly disparate, organ-specific syndrome pulled under the IgG4-RD umbrella. Although interesting clinical, radiology, and pathology reports continue to emerge and contribute to our nuanced understanding of this condition, we now witness the entry of new types of investigators into the field – immunologists, geneticists, cell biologists, and others. Their skills, melded with expertise from clinical disciplines, already promise to push the pace of progress ahead quickly.

The field of IgG4-RD investigation is fortunate originate in a time when a wealth of new scientific tools are available to help shine new light on this clinical area. We believe the collaborations between clinicians (including pathologists and radiologists) and an array of investigators with basic science expertise will unlock many of the mysteries surrounding IgG4-RD, lead to new questions and conundrums, and have implications for fields that are now regarded as tangential to IgG4-RD. The fruit of such collaborations, we hope, will lead to broader understanding of the issues fundamental to IgG4-RD, important hypotheses to test in the clinic, and better ways to diagnose, follow, and treat our patients.

It has been a pleasure to organize the Waikiki Symposium. This meeting is truly an international collaboration, not only in the sense of attracting investigators from around the world, but also a Symposium that has been international in its organization. Our Organizing Committee symbolizes the confluence of interest in IgG4-RD in both the United States and Japan. The larger Scientific Committee represents the worldwide interest and expertise that have contributed to pulling together the Second International Symposium.

We hope that you thoroughly enjoy the scientific proceedings of the Second International Symposium and have a marvelous time in Waikiki.

Yours truly,

The Symposium Committee

A Short History: How did we land in Hawaii?

At the time of the First International Symposium in Boston in October, 2011, a number of guests raised the possibility (partly in jest) of holding a Second International Symposium in Hawaii in a few years.

It seemed like a fun idea at the time, but the logistical issues of organizing began to appear daunting not long after guests had departed the first Symposium. We therefore began to plan to hold the Second Symposium in Boston again, in the summer of 2014.

It then became clear that the consortium of Japanese investigators had potential access to funds from the Japanese government's Ministry of Health, Labour and Welfare. These funds might be accessed if the dates of the Symposium were advanced by a few months – to February 2014. Of course, no one wants to have a Symposium in Boston in February so we decided to work together, merge fundraising efforts in both Japan and the United States, and hold the Symposium in a location that is mutually convenient, desirable, and (most importantly) warm. Hence, Hawaii.

We are thankful that circumstances helped to foster such collaboration, and believe that the Symposium itself has benefitted enormously from our groups working together.

We would like to give special thanks to our Symposium funders:

- Genentech
- Ministry of Health, Labour and Welfare, Japan
- Amgen

Finally, we wish to acknowledge the outstanding organization support of PINNACLE Marketing & Education Inc., based in Montreal, Canada. Our colleagues at PINNACLE have performed marvelous work in helping to put together the *Second International Symposium on IgG4-Related Disease & Associated Conditions*.

The Symposium Committee



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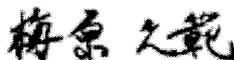
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The Boston Symposium

Group photo from the 2011 IgG4-RD and Associated Conditions



*Attendees are shown in front of the Bulfinch Building
(Massachusetts General Hospital)*

2011 IgG4-RD and Associated Conditions

Boston Symposium signed poster

INTERNATIONAL SYMPOSIUM IgG4-RELATED DISEASE

October 4-7, 2011
Massachusetts General Hospital
Boston, Massachusetts

http://www2.massgeneral.org/pathology/symposium/IgG4_related_systemic_dis.asp
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2014 Mission Statement and Objectives

Mission Statement:

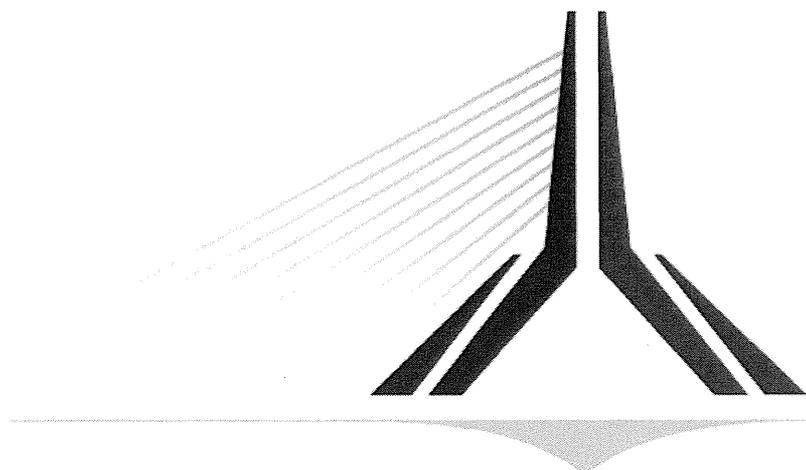
The purposes of the Second International Symposium on IgG4-RD & Associated Conditions are to gather together the world's investigators who have the expertise to define and address the most important questions confronting the field in 2014, and to conduct a scientific meeting in a collegial, cooperative, but rigorous environment that fosters the finest possible future international collaboration on IgG4-RD and related fields.

Symposium Objectives:

- To gather an international panel of experts with an interest IgG4-RD and its associated conditions.
- To present the latest information about the pathophysiology, clinical and pathology features, and treatment of IgG4-RD and its associated conditions
- To create a forum at which new research avenues can be discussed and developed.

Learning Objectives:

- To understand updates in clinical data and explore the most successful treatment methods in different IgG4-RD patient types.
- To learn about the most effective patient assessment tools and more effectively diagnose patients with IgG4-RD.
- To learn about and understand the pathology, pathophysiology, and epidemiology of IgG4-RD.



Scientific Committee

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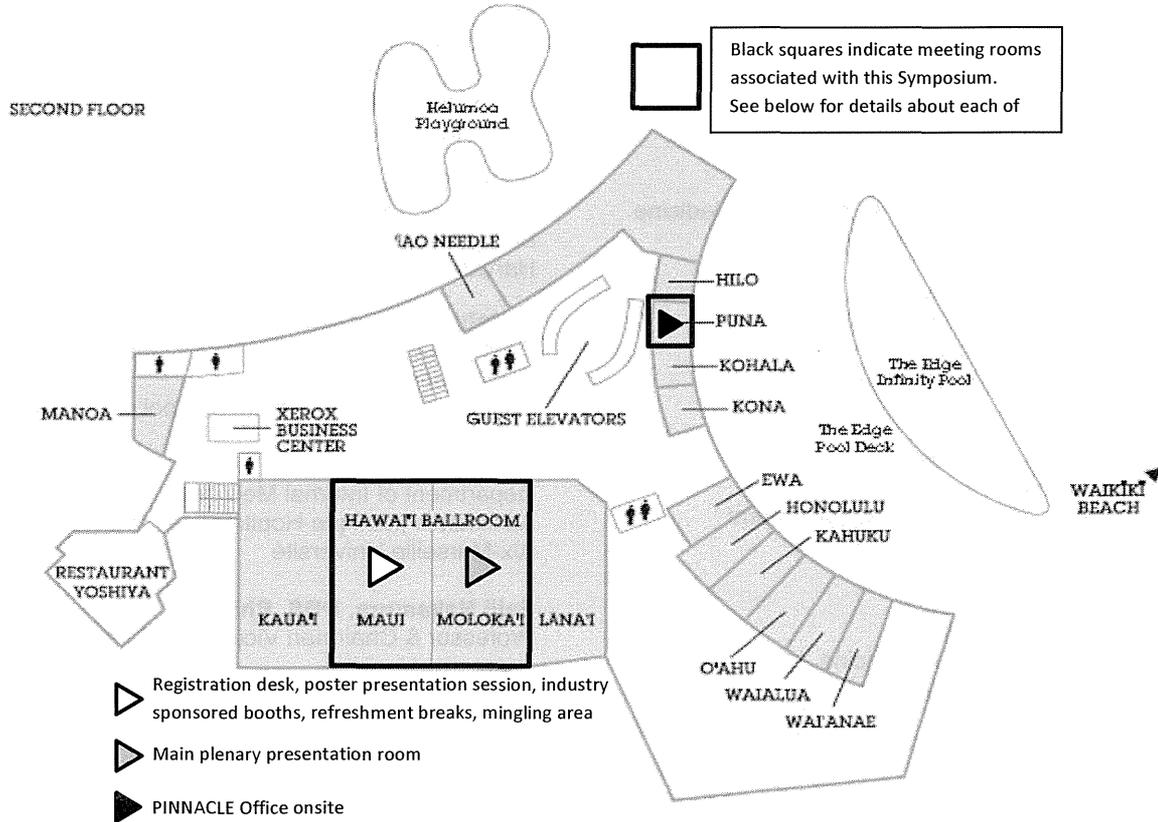
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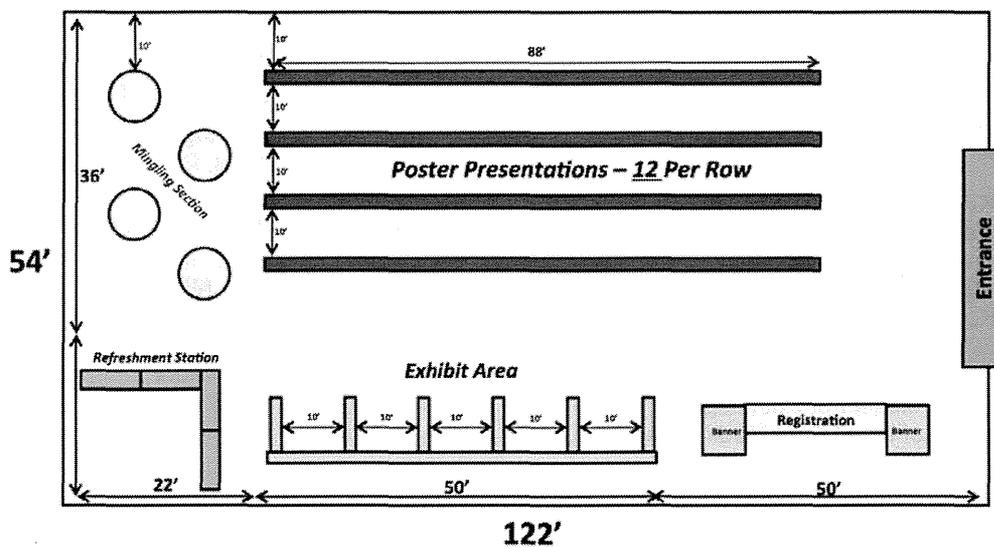
Zachary Wallace, MD

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Conference Hotel Map



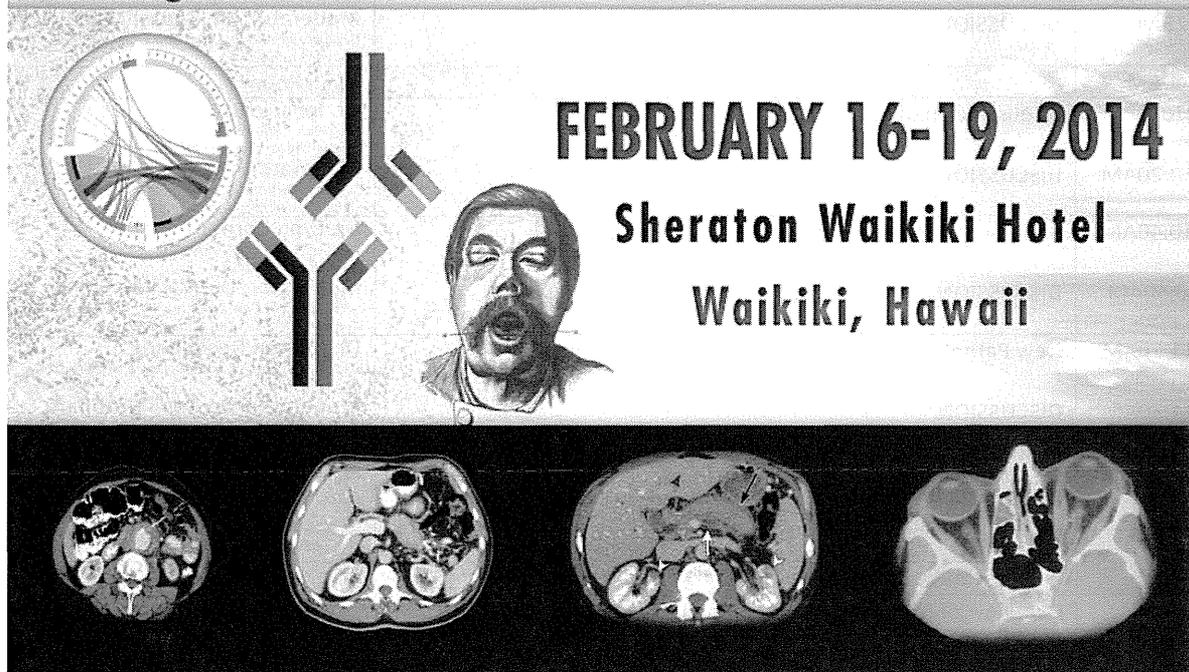
Maui Ball Room



Plenary Agenda

INTERNATIONAL SYMPOSIUM

on IgG4-Related Diseases & Associated Conditions



Scientific program

Sunday, February 16, 2014

Maui Room

1:00 – 5:00 Registration
5:00 Wine & Cheese / Poster Viewing

Moloka'i Room

6:30 Welcome – Dr. John Stone & Dr. Tsutomu Chiba
6:45 The Recognition of IgG4-related Autoimmune Pancreatitis – Dr. Shigeyuki Kawa
7:00 Overview of Pathophysiology – Dr. Shiv Pillai

Monday, February 17, 2014
Moloka'i Room

7:30 – 8:00AM	BREAKFAST	(Provided by Conference)
8:00 – 8:05AM	Welcome	Dr. Arezou Khosroshahi <i>Division of Rheumatology Emory University, Atlanta, GA USA</i>
	Pathology	
8:05 – 8:35AM	The Pathology of IgG4-RD: What have we learned since 2011?	Dr. Yoh Zen <i>King's College Hospital, Institute of Liver Studies, London UK</i>
8:35 – 8:45AM	DISCUSSION	Moderator: Dr. Giuseppe Zamboni <i>Dept of Pathology, S. Cuore-Don Calabria Hospital, University of Verona, Negrar, Italy</i>
8:45 – 9:15AM	Can the diagnosis of IgG4-RD be made by lymph node biopsy?	Dr. John K.C. Chan <i>Dept of Pathology, Queen Elizabeth Hospital Kowloon, Hong Kong, China</i>
9:15 – 9:25AM	DISCUSSION	Moderator: Dr. Tadashi Yoshino <i>Dept of Pathology, Okayama University Medical School Okayama, Japan</i>
9:25 – 9:40AM	BREAK	Maui Room
9:40–10:10AM	B cells, plasmablasts, and plasma cells in IgG4-RD	Dr. Ignacio Sanz <i>Division of Rheumatology Emory University, Atlanta, GA USA</i>
10:10 – 10:20AM	DISCUSSION	Moderator: Dr. Zhan-Guo Li <i>Dept of Rheumatology and Immunology, Beijing University Medical School People's Hospital</i>
10:20 – 10:40AM	Immune complexes in IgG4-RD	Dr. Lynn Cornell <i>Dept of Pathology, Mayo Clinic, Rochester, MN USA</i>
10:40 –10:50AM	DISCUSSION	Moderator: Dr. MH Kim <i>Asan Medical Center/University of Ulsan College of Medicine, Asan Medical Center, Seoul, Korea</i>
10:50 – 11:20AM	New Pathology Techniques	Dr. Vikram Deshpande <i>Dept of Pathology, Massachusetts General Hospital Boston, MA USA</i>
11:20 – 11:30AM	DISCUSSION	Moderator: Dr. Kenji Notohara <i>Dept of pathology, Kurashiki Central Hospital, Kurashiki, Japan</i>
11:30 – Noon	Abstract presentation (#142 and #230) 1- Epstein-Barr virus reactivation in IgG4-RD – <i>Dr. Takeuchi</i> 2- Active IgG4-RD Is Characterized By Substantial Elevations In Circulating Plasmablasts – <i>Dr. Wallace</i>	Moderator: Dr. Tooru Shimosegawa <i>Dept of Gastroenterology, Tohoku University, Japan</i>
Noon – 1:30PM	LUNCH	At Leisure
	Challenging Individual Organ Manifestations	
1:30 –2:00PM	Overview of New Entities within the IgG4-RD spectrum	Dr. Mitsuhiro Kawano <i>Dept of Rheumatology & Internal Medicine, Kanazawa University Hospital, Kanazawa, Japan</i>
2:00 – 2:10PM	DISCUSSION	Moderator: Dr. Nicolas Schleinitz <i>Dept of Internal Medicine, AP-HM, Aix Marseille Université Marseille, France</i>
2:10 – 2:30PM	Pulmonary manifestations of IgG4-RD and their management	Dr. Jay H. Ryu <i>Pulmonary and Critical Care Medicine, Mayo Clinic Rochester, MN USA</i>
2:30- 2:40	Abstract 152 Proposal for diagnostic criteria for IgG4-related lung disease – <i>Shoko Matsui</i>	Moderator: Dr. Shoko Matsui <i>Toyama Medical University, Toyama, Japan</i>
2:40 – 2:50PM	DISCUSSION	
2:50 – 3:15PM	BREAK	Maui Room
3:15 – 3:45PM	Cancer & IgG4-RD: Is there a real relationship?	Dr. Tsutomu Chiba <i>Dept of Gastroenterology and Hepatology Graduate School of Medicine, Kyoto University, Kyoto, Japan</i>
3:45 – 4:00PM	DISCUSSION	Moderator: Dr. John K.C. Chan <i>Dept of Pathology, Queen Elizabeth Hospital Kowloon, Hong Kong, China</i>
4:00 – 4:30PM	Thoracic and abdominal aortitis	Dr. James Stone <i>Anatomic Pathology, Massachusetts General Hospital Boston, MA USA</i>
4:30 – 4:40PM	DISCUSSION	Moderator: Dr. Satomi Kasashima <i>Dept of Pathology and Clinical Laboratory, National Hospital Organization, Kanazawa Medical Center, Kanazawa, Japan</i>
4:40 – 5:10PM	IgG4 and the Kidney	Dr. David Salant <i>Dept of Medicine, Division of Nephrology, Boston University, Boston, MA USA</i>
5:10 – 5:20PM	DISCUSSION	Moderator: Dr. Takako Saeki <i>Nagaoka Red Cross Hospital, Rheumatology & Internal Medicine & Nephrology, Niigata, Japan</i>
5:20 – 6:05PM	Abstract presentation (#202, #160, #179) 1- Phenotypic differences between IgG4+ & IgG1+ B cells – <i>Dr. Rispens</i> 2- Inhibition of APRIL suppresses disease progression: Analysis from a novel mouse model for IgG4-related disease – <i>Dr. Yamada</i> 3- Polarized M2 macrophages promote fibrosis of glandular tissue in IgG4-related dacryoadenitis and sialoadenitis – <i>Dr. Furukawa</i>	Moderator: Dr. Hisanori Umehara <i>Dept of Hematology and Immunology Kanazawa Medical University</i>

Tuesday, February 18

Moloka'i Room

7:30 – 8:00AM	<i>BREAKFAST</i>	<i>Maui Room</i>
8:00 – 8:05AM	Welcome	Dr. Kazuichi Okazaki <i>The Third Dept of Internal Medicine, Division of Gastroenterology and Hepatology, Kansai Medical University, Osaka, Japan</i>
	Pathophysiology	
8:05 – 8:45AM	What can we learn about IgG4-RD from parasitic diseases?	Dr. Thomas A. Wynn <i>Immunopathogenesis Section, NIH, Bethesda, MD USA</i>
8:45 – 9:00AM	DISCUSSION	Moderator: Dr. Tomohiro Watanabe <i>Dept of Gastroenterology, Kyoto University, Kyoto, Japan</i>
9:00 – 9:30AM	The Genetic Determinants of Pulmonary Fibrosis	Dr. David Schwartz <i>University of Colorado, Denver, Colorado USA</i>
9:30 – 9:45AM	DISCUSSION	Dr. Vinay Mahajan <i>MGH Center for Cancer Research, Massachusetts General Hospital, Boston MA USA</i>
9:45 – 10:00AM	<i>BREAK</i>	<i>Maui Room</i>
10:00 – 10:30AM	T cells and IgG4-RD	Dr. Hamid Mattoo <i>MGH Center for Cancer Research, Massachusetts General Hospital, Boston, MA USA</i>
10:30 – 10:40AM	DISCUSSION	Moderator: Dr. Seiji Nakamura <i>Kyushu University, oral surgery, Fukuoka, Japan</i>
10:40 – 11:05AM	B-T cell interaction in IgG4-RD	Dr. Vinay Mahajan <i>MGH Center for Cancer Research, Massachusetts General Hospital, Boston MA USA</i>
11:05 – 11:15AM	DISCUSSION	Moderator: Dr. Kenji Notohara <i>Dept of pathology, Kurashiki Central Hospital, Kurashiki, Japan</i>
11:15 – 11:45AM	IgE: How does it fit into the IgG4-RD milieu?	Dr. Frances Lee <i>Division of Pulmonary and critical care., Emory University, Atlanta, GA USA</i>
11:45 – Noon	DISCUSSION	Moderator: Dr. Emanuel Della Torre <i>Massachusetts General Hospital, Boston MA USA</i>
Noon – 1:30PM	<i>LUNCH</i>	<i>At Leisure</i>
	Epidemiology & Clinical Assessment	
1:30 – 1:50PM	Genome-wide association studies in IgG4-RD	Dr. Fumihiko Matsuda <i>Kyoto University Graduate School of Medicine, Kyoto, Japan</i>
1:50 – 2:00PM	DISCUSSION	Moderator: Dr. Shiv Pillai <i>Massachusetts General Hospital, Boston MA USA</i>
2:00 – 2:30PM	Imaging in IgG4-RD	Dr. Dai Inoue <i>Dept of Radiology, Kanazawa University Graduate School of Medical Science, Kanazawa, Ishikawa, Japan</i>
2:30 – 2:45PM	DISCUSSION	Moderator: Dr. Yoh Zen <i>King's College Hospital, Institute of Liver Studies, London UK</i>
2:45 – 3:00PM	<i>BREAK</i>	<i>Maui Room</i>
3:00 – 3:20PM	Multi-centric Castleman's disease	Dr. Yasuharu Sato <i>Dept of Pathology, Okayama University, Okayama, Japan</i>
3:20 – 3:40PM	DISCUSSION	Moderator: Dr. Hiroki Takahashi <i>Dept of Gastroenterology, Rheumatology and Clinical Immunology, Sapporo Medical University School of Medicine, Sapporo, Japan</i>
3:40 – 4:00PM	Inflammatory Myofibroblastic Tumors	Dr. Adrian Bateman <i>University Hospital Southampton NHS Foundation Trust, Dept of Cellular Pathology, Southampton, Hampshire, UK</i>
4:00 – 4:10PM	DISCUSSION	Moderator: Dr. Vikram Deshpande <i>Dept of Pathology, Massachusetts General Hospital Boston MA USA</i>
4:10 – 4:30PM	Monitoring IgG4-RD activity through the blood	Dr. Arezou Khosroshahi <i>Division of Rheumatology, Emory University, Atlanta, GA USA</i>
4:30 – 4:45PM	DISCUSSION	Moderator: Dr. Kazuichi Okazaki <i>The Third Dept of Internal Medicine, Division of Gastroenterology and Hepatology, Kansai Medical University, Osaka, Japan</i>
4:45 – 5:30PM	Abstract presentation (#233 and #167) 1- Aberrant activation of Th2 cells by IL-33 in IgG4-related dacryoadenitis and sialoadenitis – <i>Dr. Nakamura</i> 2- Activation of Toll-like receptors and NOD-like receptors in monocytes and basophils is involved in the immuno-pathogenesis of IgG4-related disease – <i>Dr. Watanabe</i>	Moderator: Dr. Terumi Kamisawa <i>Dept of Gastroenterology, Tokyo Metropolitan Komagome Hospital, Tokyo, Japan</i>
7:00PM	Symposium Dinner: Private Patio – Kai Market, Ground Floor	Ticket Purchase <i>Dressy Casual (no jackets please)</i>

Wednesday, February 19
Moloka'i Room

7:30 – 8:00AM	BREAKFAST	Maui Room
8:00 – 8:05AM	Welcome	Dr. John Stone <i>Division of Rheumatology, Massachusetts General Hospital Harvard Medical School, Boston, MA USA</i>
	Treatment Session	
8:05 – 8:25AM	Glucocorticoid Treatment in IgG4-RD	Dr. Yasufumi Masaki <i>Kanazawa Medical University, Hematology and Immunology, Kahoku, Ishikawa, Japan</i>
8:25 – 8:30AM	DISCUSSION	Moderator: Dr. Suresh Chari <i>Dept of Gastroenterology, Mayo Clinic, Rochester, MN USA</i>
8:30 – 8:50AM	Conventional Steroid sparing agents for IgG4-RD	Dr. Mark Topazian <i>Division of Gastroenterology, Mayo Clinic Rochester, MN, USA</i>
8:50 – 9:00AM	DISCUSSION	Moderator: Dr. Arezou Khosroshahi <i>Division of Rheumatology Emory University, Atlanta, GA USA</i>
9:00 – 9:20AM	B cell depletion strategies	Dr. John Stone <i>Division of Rheumatology, Massachusetts General Hospital, Harvard Medical School, Boston, MA USA</i>
9:20 – 9:25AM	DISCUSSION	Moderator: Dr. Hisanori Umehara <i>Dept of Hematology and Immunology Kanazawa Medical University</i>
9:25 – 9:45AM	Current & Future Treatment for Pulmonary Fibrosis	Dr. David Schwartz <i>University of Colorado, Denver, Colorado USA</i>
9:45 – 9:50AM	DISCUSSION	Moderator: Dr. Hamid Mattoo <i>Cancer Center, Massachusetts General Hospital, Boston, MA USA</i>
9:50 – 10:00AM	BREAK	Maui Room
10:00 – 11:45AM	Panel Discussion on Treatment Drs. Chari, Khosroshahi, Kamisawa, Topazian, Hart, Umehara, Kawano, Saeki, Ryu, Yamamoto, Shimosegawa, Masaki, Schwartz, and others	Moderator: Dr. John Stone <i>Division of Rheumatology, Massachusetts General Hospital Harvard Medical School, Boston, MA USA</i>
11:45 – Noon	Wrap up for treatment	
Noon – 1:30PM	LUNCH	At Leisure
1:30 – 5:00PM	Biliary Session	Chair: Dr. Suresh Chari <i>Dept of Gastroenterology, Mayo Clinic, Rochester, MN USA</i>
1:30 – 1:40 PM	Clinical Features: Are there clinical features that distinguish IgG4-SC from other biliary pathologies?	Dr. Roger Chapman <i>Dept of Gastroenterology, Oxford University, Oxford, UK</i>
1:40 – 1:45 PM	DISCUSSION	Dr. Atsushi Tanaka <i>Teikyo University, Japan</i>
1:45 – 1:50 PM	Consensus summary: Role of clinical features in distinguishing IgG4-SC from other biliary pathologies.	Moderator: Dr. Suresh Chari <i>Dept of Gastroenterology, Mayo Clinic, Rochester, MN USA</i>
1:50 – 2:20PM	Do imaging findings distinguish IgG4-SC from other biliary pathologies? <i>CT/MRI, ERCP, EUS</i>	Dr. Dai Inoue <i>Dept of Radiology, Kanazawa University Graduate School of Medical Science, Kanazawa, Ishikawa, Japan</i>
2:20 – 2:25PM	DISCUSSION	Dr. Naoki Takahashi <i>Dept of Radiology, Mayo Clinic Rochester, MN, USA</i>
		Dr. Takahiro Nakazawa <i>Dept of Gastroenterology, Nagoya City university, Japan</i>
		Dr. George Webster <i>Dept of Gastroenterology and Hepatology University College London Hospitals, London, UK</i>
		Dr. Kazuo Inui <i>Dept of Gastroenterology, Fujita Health University, Second Teaching Hospital, Nagoya, Japan</i>
		Dr. Michael Levy <i>Dept of Gastroenterology, Mayo Clinic Rochester, MN, USA</i>
2:25 – 2:35PM	Consensus summary: Role of imaging in distinguishing IgG4-SC from other biliary pathologies	Moderator: Dr. Suresh Chari <i>Dept of Gastroenterology, Mayo Clinic, Rochester, MN USA</i>
2:35 – 2:45PM	Do IgG4 levels distinguish IgG4-SC from PSC?	Dr. Hirotaka Ohara <i>Dept of Gastroenterology, Nagoya City university, Japan</i>
2:45 – 2:50PM	DISCUSSION	Dr. Roger Chapman <i>Dept of Gastroenterology, Oxford University, Oxford, UK</i>
2:50 – 2:55PM	Consensus summary: What is the role of serum IgG4 levels in distinguishing IgG4-SC from PSC?	Moderator: Dr. Kazuichi Okazaki <i>The Third Dept of Internal Medicine, Division of Gastroenterology and Hepatology, Kansai Medical University, Osaka, Japan</i>

2:55 – 3:10PM	BREAK	
3:10 – 3:50PM	Does histology distinguish IgG4 SC from PSC? <i>Resection</i> <i>Bile duct Biopsy</i> <i>Ampulla Biopsy</i> <i>Bile duct brush</i> <i>Liver Biopsy</i>	Dr. Yasuni Nakanuma <i>Kanazawa University, Faculty of Medicine, Institute of Medical, Pharmaceutical and Health Sciences, Japan</i> Dr. Lizhi Zhang <i>Dept of Pathology, Mayo Clinic, Rochester, MN USA</i> Dr. Kenji Notohara <i>Dept of Pathology, Kurashiki Central Hospital, Kurashiki, Japan</i> Dr. Giuseppe Zamboni <i>Dept of Pathology, S. Cuore-Don Calabria Hospital, University of Verona, Negrar, Italy</i> Dr. Kensuke Kubota <i>Yokohama City University, Graduate School of Medicine, Yokohama, Japan</i> Dr. Thomas Smyrk <i>Dept of Pathology, Mayo Clinic, Rochester, MN USA</i> Dr. Vikram Deshpande <i>Dept of Pathology, Massachusetts General Hospital, Boston MA USA</i> Dr. Yoh Zen <i>Dept of Pathology, King's College Hospital, London UK</i>
3:50 – 4:00PM	DISCUSSION	
4:00 – 4:10PM	Consensus Summary: Role of histopathology in distinguishing IgG4-SC from PSC	Moderator: Dr. Suresh T. Chari <i>Dept of Gastroenterology, Mayo Clinic, Rochester, MN USA</i>
4:10 – 4:20PM	Does response to steroids distinguish IgG4-SC from PSC? <i>IgG4-Sclerosing cholangitis</i> <i>Primary sclerosing cholangitis</i>	Dr. Myung-Hwan Kim <i>Asan Medical Center/University of Ulsan College of Medicine, Asan Medical Center, Seoul, Korea</i> Dr. Terumi Kamisawa <i>Dept of Gastroenterology, Tokyo Metropolitan Komagome Hospital, Tokyo, Japan</i> Dr. Roger Chapman <i>Dept of Gastroenterology, Oxford University, Oxford, UK</i> Moderator: Dr. Tooru Shimosegawa <i>Dept of Gastroenterology, Tohoku University, Japan</i>
4:20 – 4:25PM	DISCUSSION	
4:25 – 4:30PM	Consensus summary: Role of steroid response in distinguishing IgG4-SC from PSC	Moderator: Dr. Tooru Shimosegawa <i>Dept of Gastroenterology, Tohoku University, Japan</i>
4:30 – 4:35PM	What OOI is seen in IgG4-SC and PSC?	Dr. Suresh Chari <i>Professor of Medicine, Mayo Clinic, Rochester, MN USA</i>
4:35 – 4:45PM	DISCUSSION	
4:45 – 4:50PM	Consensus: Role of OOI in distinguishing IgG4-SC from PSC?	Moderator: Dr. Suresh Chari <i>Dept of Gastroenterology, Mayo Clinic, Rochester, MN USA</i>
4:50 – 5:10PM	Wrap up for Diagnostic criteria for IgG4-SC	Moderator: Dr. Suresh Chari <i>Dept of Gastroenterology, Mayo Clinic, Rochester, MN USA</i>

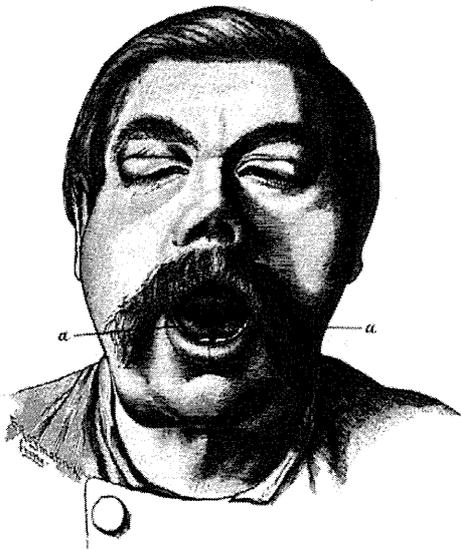
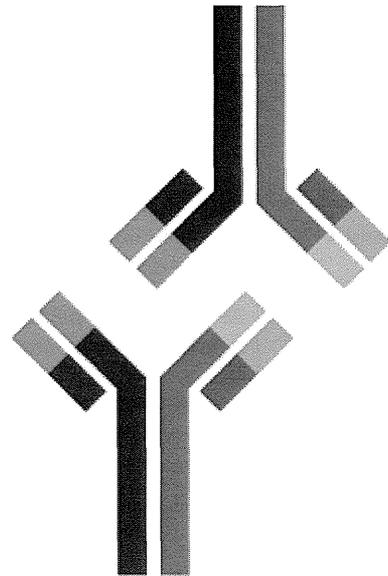


Fig. A.



Abstract Submissions

We received a total of 96 outstanding abstracts from all around the globe for the Second International Symposium on IgG4-RD and Associated Conditions. All the abstracts were evaluated by the Abstract Review Committee and accepted for either oral or poster presentations. This not only speaks to the growing interest in the area of IgG4-RD, but also the quality of data that is being produced.

We would like to thank you all for your interesting and thought-provoking abstracts, and also thank the Abstract Review Committee members for their excellent effort in evaluating the many submitted abstracts.

We are looking forward to both the poster presentations on the opening night of our Symposium, and the oral presentations during the meeting. All attendees will receive a USB flash drive containing the submitted abstracts in PDF format.

Oral Abstract Presentations

Abstracts that were accepted for oral presentation will be presented throughout our meeting agenda. See the meeting agenda to follow for more details. See below for a summary of oral abstract presentations.

Presenter Name	Abstract title	Topic	Category
Sachiko Furukawa	Polarized M2 macrophages promote fibrosis of glandular tissue in IgG4-related dacryoadenitis and sialoadenitis (IgG4-DS).	Pathophysiology	Investigator initiated
Shoko Matsui	Proposal for diagnostic criteria for IgG4-related respiratory disease	Clinical Aspects & Treatment	Investigator initiated
Seiji Nakamura	Aberrant activation of Th2 cells by IL-33 in IgG4-related dacryoadenitis and sialoadenitis	Pathophysiology	Investigator initiated
Theo Rispens	Phenotypic differences between IgG4+ and IgG1+ B cells	Pathophysiology	Investigator initiated
Mai Takeuchi	Epstein-Barr virus reactivation in IgG4-related lymphadenopathy with comparison to extranodal IgG4-related disease	Pathology	Investigator initiated
Zachary Wallace	Active IgG4-Related Disease Is Characterized By Substantial Elevations In Circulating Plasmablasts	Clinical Aspects & Treatment	Investigator initiated
Tomohiro Watanabe	Activation of Toll-like receptors and NOD-like receptors in monocytes and basophils is involved in the immunopathogenesis of IgG4-related disease	Pathophysiology	Investigator initiated
Kazunori Yamada	Inhibition of APRIL suppresses disease progression: Analysis from a novel mouse model for IgG4-related disease	Pathophysiology	Investigator initiated

Poster Presentations

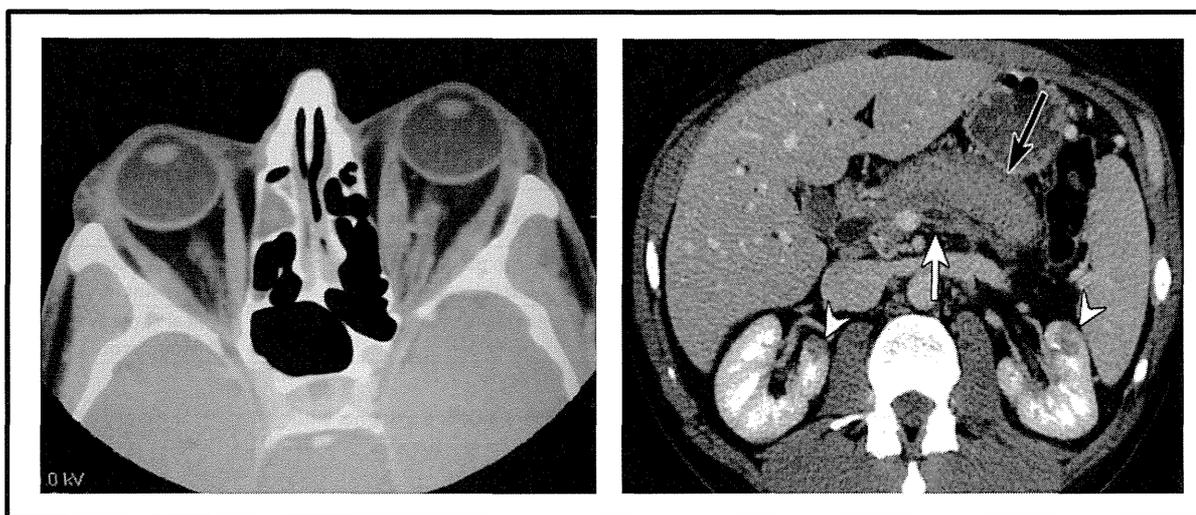
The poster presentation session will be taking place on Sunday February 16th from 5:00pm-6:30pm in the Maui Ballroom. Below you will find a list of all poster numbers, presenters, and topics. If you are presenting a poster, please make sure that the poster has been installed prior to 4:30pm on Sunday February 16th.

Poster #	Presenter Name	Abstract Title	Topic	Category
101	Takashi Akamizu	Clinical characteristics of Graves' patients with elevated serum IgG4 concentrations	Clinical Aspects & Treatment	Investigator initiated
102	Yasuyuki Arai	Plasmacytoid dendritic cells activated by neutrophil extracellular traps (NETs) contribute to the pathophysiology of IgG4-RD	Pathophysiology	Investigator initiated
103	Luke Chen	Polyclonal hyperviscosity syndrome in IgG4-Related Disease	Clinical Aspects & Treatment	Investigator initiated
104	Emma Culver	Gene expression analysis identifies immune signaling and complement pathways in IgG4-Related Disease	Pathophysiology	Investigator initiated
105	Emma Culver	Elevated serum IgG4 and abundant IgG4-positive plasma cells in the colon of patients with ulcerative colitis in the presence and absence of IgG4-Related Disease	Pathology	Investigator initiated
106	Emma Culver	Elevated serum IgG4 and IgE in IgG4-RD and other disease mimics	Pathophysiology	Investigator initiated
107	Emma Culver	Autoimmune pancreatitis and IgG4-related sclerosing cholangitis is associated with extra-pancreatic organ failure, malignancy and mortality in a prospective UK cohort.	Clinical Aspects & Treatment	Investigator initiated
108	Emanuel Della Torre	Diagnostic value of cerebrospinal fluid IgG4 indices in IgG4-Related hypertrophic pachymeningitis	Clinical Aspects & Treatment	Investigator initiated
109	Emanuel Della Torre	Prevalence of atopy, eosinophilia and IgE elevation in IgG4-Related Disease	Clinical Aspects & Treatment	Investigator initiated
110	Emanuel Della Torre	IgG4-Related Midline Destructive Lesion	Clinical Aspects & Treatment	Investigator initiated
111	Mikael Ebbo	FDG-PET/CT in staging and evaluation of treatment response in IgG4-Related Disease: analysis of 46 FDG-PET/CT examinations	Clinical Aspects & Treatment	Investigator initiated
112	Mikael Ebbo	Serum immunoglobulin free light chain in IgG4-related disease: a potential novel biomarker?	Pathophysiology	Investigator initiated
113	Sachiko Furukawa	Clinical relevance of Küttner tumor and IgG4-Related dacryoadenitis and sialoadenitis	Pathophysiology	Investigator initiated
114	James Garrity	Diagnostic confusion regarding orbital IgG4-RD	Clinical Aspects & Treatment	Investigator initiated
115	Jovan Popovich	IgG4 Related retroperitoneal fibrosis with involvement of the spleen- clinical, radiological and pathologic correlation	Clinical Aspects & Treatment	Investigator initiated
116	Shin Hamada	Comprehensive analysis of serum miRNA in autoimmune pancreatitis	Pathophysiology	Investigator initiated
117	Shoko Hamaoka	Cases of IgG4-related ophthalmic disease without serum IgG4 elevation: the probable cases according to the comprehensive diagnostic criteria	Pathology	Investigator initiated
118	Tomohiro Handa	Evaluation of 18F-fluorodeoxyglucose uptake by positron emission tomography in IgG4 Related Disease	Clinical Aspects & Treatment	Investigator initiated
119	Satoshi Hara	Components and distribution of interstitial fibrosis in IgG4-related kidney disease; an autopsy series of 5 cases	Pathology	Investigator initiated
120	Ikuko Haruta	Analysis of a commensal bacteria triggered autoimmune pancreatitis mouse model	Pathophysiology	Investigator initiated
121	Noriko Hayami	IgG subclass and PLA2R in membranous nephropathy associated with IgG4-Related Disease	Pathology	Investigator initiated
122	Kenji Hirano	Intrapancreatic biliary stricture in autoimmune pancreatitis should not be classified into IgG4 related sclerosing cholangitis	Clinical Aspects & Treatment	Investigator initiated

Poster #	Presenter Name	Abstract Title	Topic	Category
123	Kenji Hirano	Incidence of malignancies in patients with IgG4-Related Disease	Clinical Aspects & Treatment	Investigator initiated
124	Kenji Hirano	Prognosis of patients with AIP in whom steroid was ceased after at least three-year maintenance therapy	Clinical Aspects & Treatment	Investigator initiated
125	Kenji Hirano	Indication for steroid therapy in AIP patients without obstructive jaundice	Clinical Aspects & Treatment	Investigator initiated
126	Shintaro Harata	Characterization of peripheral blood immune cell subsets in patients with IgG4 related disease: A comparative analysis with patients with Sjögren syndrome and healthy donors	Pathophysiology	Investigator initiated
127	Soon Auck Hong	Xanthogranulomatous cholecystitis manifesting IgG4-related cholecystitis	Pathology	Investigator initiated
128	Abbie Husman	Increased IgG4+ plasma cells and IgG4/IgG ratio may be seen in lymph nodes from patients without IgG4-Related Disease	Pathology	Investigator initiated
129	Satoshi Ikeda	Interstitial pneumonia with marked IgG4 positive plasma cells infiltration: are they classified as a spectrum of IgG4-related lung disease?	Clinical Aspects & Treatment	Investigator initiated
130	Joanne E. Yi	Cavitating lung disease: A novel presentation of IgG4 pulmonary involvement	Clinical Aspects & Treatment	Investigator initiated
131	George Webster	Cerebral manifestations of IgG4-Related Disease	Clinical Aspects & Treatment	Investigator initiated
132	Keita Kanai	Two siblings with type 1 autoimmune pancreatitis	Clinical Aspects & Treatment	Investigator initiated
133	Atsushi Kanno	The diagnosis of autoimmune pancreatitis by EUS-FNA based on the International Consensus Diagnostic Criteria (ICDC)	Pathology	Investigator initiated
134	Fuminori Kasashima	IgG4-Related periaortitis associated with rupture during low-dose steroid administration and aortoduodenal fistula following endovascular repair	Clinical Aspects & Treatment	Investigator initiated
135	Yuri Kato	Is corticosteroid administration not always required in patients with type 1 autoimmune pancreatitis?	Clinical Aspects & Treatment	Non-investigator initiated
136	Arezou Khosroshahi	Distinct phases of IgG4-RD, exhibit unique phenotype of B cell activation and clonal expansion	Pathophysiology	Investigator initiated
137	Sigeru Ko	Autoimmune pancreatitis: Reorienting the goals of steroid therapy to repair, regeneration, and functional recovery of pancreatic tissues	Clinical Aspects & Treatment	Investigator initiated
138	Toshinobu Kubota	Monitoring IgG4-RD activity through the blood in Ophthalmology	Pathophysiology	Investigator initiated
139	Katsutoshi Kuriyama	Clinical characteristics of IgG4-RD patients; multicenter retrospective cohort study in Japan	Clinical Aspects & Treatment	Investigator initiated
140	Jong Kyun Lee	Comparison of ERCP with papillary biopsy and EUS-guided pancreatic biopsy in the diagnosis of autoimmune pancreatitis	Clinical Aspects & Treatment	Investigator initiated
141	Wen Zhang	IgG4-RD in China: A prospective cohort study of 118 patients	Clinical Aspects & Treatment	Investigator initiated
142	Wen Zhang	B cell subsets and dysfunction of regulatory B cells in IgG4-related diseases and primary Sjögren's syndrome: the similarities and differences	Pathophysiology	Investigator initiated
143	Takashi Maehara	IL-21 contributes to ectopic germinal center formation and IgG4 production in IgG4-Related dacryoadenitis and sialoadenitis	Pathology	investigator initiated
144	Vinay S Mahajan	Individuals with IgG4-Related Disease do not have an increased frequency of the K409 variant of IgG4 that compromises Fab-arm exchange	Pathophysiology	Investigator initiated
145	Vinay S Mahajan	De novo oligoclonal expansions of circulating plasmablasts in active and relapsing IgG4-Related Disease	Pathophysiology	Investigator initiated
146	Masahiro Maruyama	Long term outcomes of type 1 autoimmune pancreatitis: development to pancreatic stone formation and chronic pancreatitis	Clinical Aspects & Treatment	Investigator initiated
147	Hamid Mattoo	Response to rituximab in IgG4-Related Disease is mediated by depletion of clonally expanded CD4+ effector T cells	Pathophysiology	Non-investigator initiated

Poster #	Presenter Name	Abstract Title	Topic	Category
148	Hamid Mattoo	Circulating Th2 memory cells in IgG4 Related Disease are restricted to a defined subset of subjects with concomitant atopy	Pathophysiology	Investigator initiated
149	Katsuyuki Miyabe	Comparison study of the immunohistochemical stainings for the diagnosis of type 1 autoimmune pancreatitis	Pathology	Investigator initiated
150	Masaki Miyazawa	A clinical study on steroid therapy for AIP	Clinical Aspects & Treatment	Investigator initiated
151	Ichiro Mizushima	Clinical course after corticosteroid therapy	Clinical Aspects & Treatment	Investigator initiated
152	Sung-Hoon Moon	Clinical differences between primary and IgG4-related sclerosing cholangitis	Clinical Aspects & Treatment	Investigator initiated
153	Masafumi Moriyama	The diagnostic usability of labial salivary and submandibular gland biopsy in IgG4-Related dacryoadenitis and sialoadenitis	Clinical Aspects & Treatment	Non-investigator initiated
154	Alfredo Musumeci	Isolated abdominal aortitis in the ER	Clinical Aspects & Treatment	Investigator initiated
155	Yoshimasa Nakazato	Lung cancer complicated with IgG4-Related Disease of the lung; A case report	Clinical Aspects & Treatment	Investigator initiated
156	Takayoshi Nishino	Incidence of cancer and risk factors for complication by cancer in IgG4-Related Diseases	Clinical Aspects & Treatment	Investigator initiated
157	Susumu Nishiyama	Features of quantitative salivary gland scintigraphy in patients with IgG4-Related sialadenitis, so-called Mikulicz disease	Clinical Aspects & Treatment	Investigator initiated
158	Takaya Oguchi	Genome-wide association study for lachrymal/salivary gland lesion susceptibility genes in type I autoimmune pancreatitis	Pathophysiology	Investigator initiated
159	Miho Ohta	A case of malignant lymphoma strongly suspected IgG4-Related dacryoadenitis and sialoadenitis	Clinical Aspects & Treatment	Investigator initiated
160	Masao Ota	Risk HLA allele for predisposition to type I autoimmune pancreatitis	Pathophysiology	Investigator initiated
161	Eva Amin	IgG4 related disease with great vessels and cardiac involvement	Clinical Aspects & Treatment	Investigator initiated
162	Erika Resetkova	IgG4-Related Sclerosing Mastitis (IgG4-RSM)	Pathology	Investigator initiated
163	Jikon Ryu	Clinical and Pathological Differences between Serum IgG4-positive and IgG4-negative type 1 autoimmune pancreatitis	Clinical Aspects & Treatment	Investigator initiated
164	Takako Saeki	Initial corticosteroid therapy in IgG4-Related kidney disease	Clinical Aspects & Treatment	Investigator initiated
165	Aurelie Grados	Increase of T regulatory and T follicular helper cells in peripheral blood of IgG4 RD patients	Pathophysiology	Investigator initiated
166	Aurelie Grados	Characterization of IgG4 RD patients- subgroups by multiple correspondence and cluster analysis	Clinical Aspects & Treatment	Investigator initiated
167	Yusuke Sekino	Therapeutic strategy for patients with type 1AIP developed with cyst formation	Clinical Aspects & Treatment	Non-investigator initiated
168	Masahiro Shiokawa	Undifferentiated IgG4-positive cholangitis; primary sclerosing cholangitis or IgG4-Related sclerosing cholangitis?	Clinical Aspects & Treatment	Investigator initiated
169	Masahiro Shiokawa	Risk of cancer in patients with autoimmune pancreatitis	Clinical Aspects & Treatment	Investigator initiated
170	Evgeniya Sokol	Unusual site of IgG4-RD manifestation: vertebral lesions	Clinical Aspects & Treatment	Investigator initiated
171	Jin Woo Song	Clinical course and outcome of patients with pulmonary involvement of immunoglobulin G4 related sclerosing disease	Clinical Aspects & Treatment	Investigator initiated
172	John Stone	The Diagnostic utility of serum IgG4 concentrations in IgG4-Related Disease	Clinical Aspects & Treatment	Investigator initiated
173	Kimi Sumimoto	Circulating CD19+CD24hiCD38hi regulatory B Cells in patients with type 1 autoimmune pancreatitis	Pathophysiology	Non-investigator initiated

Poster #	Presenter Name	Abstract Title	Topic	Category
174	Junko Tahara	Assessment of the rate of change in the serum IgG4 level of autoimmune pancreatitis as a predictor of a relapse.	Clinical Aspects & Treatment	Investigator initiated
175	Masayuki Takahira	Outcome of steroid therapy for cases with IgG4-related ophthalmic disease	Clinical Aspects & Treatment	Investigator initiated
176	Kenichi Takano	Evaluation of submandibular versus labial salivary gland fibrosis in IgG4-related disease	Pathology	Investigator initiated
177	Christin Tiegs-Heiden	Imaging of IgG4-Related Disease of the orbit	Clinical Aspects & Treatment	Investigator initiated
178	Kazushige Uchida	Comparison of IgG4-positive plasma cell infiltration between type 1 autoimmune pancreatitis and pancreatic ductal adenocarcinoma based on comprehensive diagnostic criteria for IgG4-Related Disease, 2011	Clinical Aspects & Treatment	Investigator initiated
179	Kaori Uchino	Comparison of macrophage morphology and phenotypes in type 1 autoimmune pancreatitis and other inflammatory diseases of the digestive organs	Pathology	Investigator initiated
180	Vladimir Vasilyev	IgG4-Related Disease: patients group characterization and rituximab therapy	Clinical Aspects & Treatment	Investigator initiated
181	Zachary Wallace	Ophthalmic manifestations of IgG4-Related Disease: single-center experience and literature review	Clinical Aspects & Treatment	Investigator initiated
182	Takayuki Watanabe	Clinical features of a new disease concept, IgG4-Related thyroiditis	Clinical Aspects & Treatment	Investigator initiated
183	Kazunori Yamada	Role of APRIL in IgG4-Related kidney disease	Pathophysiology	Investigator initiated
184	Motohisa Yamamoto	Can only the monitoring serum levels of IgG4 prevent the relapse in IgG4-RD?	Clinical Aspects & Treatment	Investigator initiated
185	Satoshi Yamamoto	Problems related to steroid therapy in patients with autoimmune pancreatitis	Clinical Aspects & Treatment	Non-investigator initiated
186	Hiroaki Yamashita	Evaluation of IgG4/IgG immunostaining and counting method for the supporting diagnosis of type 1 autoimmune pancreatitis: A comparative biopsy study from 8 organs	Clinical Aspects & Treatment	Investigator initiated
187	Hajime Yoshifuji	Subclass-based analysis of anti-nuclear antibody in IgG4-Related Disease and systemic autoimmune diseases	Clinical Aspects & Treatment	Investigator initiated
188	Wajeeha Yousaf	IgG4 Related Disease: A 23-year follow up.	Clinical Aspects & Treatment	Investigator initiated



VIII. 研究成果の刊行物・別刷

Clinical diagnostic criteria of IgG4-related sclerosing cholangitis 2012

**Hiroataka Ohara, Kazuichi Okazaki,
Hirohito Tsubouchi, Kazuo Inui,
Shigeyuki Kawa, Terumi Kamisawa,
Susumu Tazuma, Kazushige Uchiyama et al.**

**Journal of Hepato-Biliary-Pancreatic
Sciences**

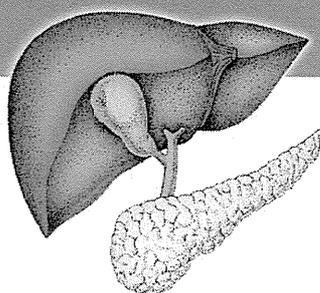
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controlled trial in pancreatic
surgery

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