Amyloid: Insoluble, but Solvable

XIVth International Symposium on Amyloidosis
April 27 – May 1, 2014
Indianapolis, Indiana, USA

Sunday, April 27, 2014	
12:00 noon	Registration opens, 3rd floor JW near escalators
7:30 pm – 9:30 pm	Welcome reception , Eiteljorg Museum

Monday, April 28, 2014		
8:00 am– 9:00 am		**Keynote lecture: Daniel Kastner (NIH, Bethesda, MD, USA) (Grand Ballroom 5)**
9:00 am – 10:30 am		**Plenary session 1. Inflammatory disease: From SAA to AA amyloid Chairmen: Kisilevsky and Livneh**
9:00	OP-1	Mechanism of IL-6 induced SAA production and amyloid A deposition in AA amyloidosis patients with RA. K. Yoshizaki (Osaka, Japan)
9:15	OP-2	Soluble, recombinant, receptor for advanced glycation end-products (RAGE) binds AA amyloid in vivo. J. S. Wall (Knoxville, TN, USA)
9:30	OP-3	Endemic and highly prevalent systemic amyloid A (AA) amyloidosis in endangered island foxes (Urocyon littoralis). P. M. Gaffney (Davis, CA, USA)
9:45	OP-4	Obesity induced chronic inflammation in C57Bl6J mice, a novel risk factor in the progression of amyloid formation? B. P. C. Hazenberg (Groningen, the Netherlands)
10:00	OP-5	Obesity as a determinant in the development and progression of AA amyloidosis. B. Kluve-Beckerman (Indianapolis, IN, USA)
10:15	OP-6	International randomized, double-blind, placebo-controlled, phase 3 study of the efficacy and safety of Kiacta™ in preventing renal function decline in patients with AA amyloidosis: An update on study progress. D. Garceau (New York, NY, USA)
10:30 am – 11:00 am		Coffee and tea break

11:00 am – 12:30 pm		**Plenary session 2. Fibril and amyloid formation** **Chairmen: Saraiva and Vidal**
11:00	OP-7	Polymorphism and cryo-EM structures of peptide fibrils from AL proteins. M. Fandrich (Ulm, Germany)
11:15	OP-8	The effect of post-translational modifications on the aggregation of the cardiovascular amyloid protein medin. J. Madine* (Liverpool, UK)
11:30	OP-9	Identification of glycosaminoglycan linkage regions and attachment sites – implications for amyloid accumulation. F. Noborn (Gothenburg,Sweden)
11:45	OP 10	Oligomeric light chains in urinary exosomes as detection method for organ response in light chain amyloidosis: 3 cases. M. Ramirez-Alvarad (Rochester, MN, USA)
12:00	OP-11	The cellular protein homeostasis network strongly influences the stability of secreted tetrameric TTR. X. Zhang* (La Jolla, CA, USA)
12:15	OP-12	Elucidating the mechanism of D76N B2-microglobulin amyloidogenesis and its inhibition. V. Bellotti (London, UK & Pavia, Italy)
12:30 pm – 2:00 pm		**Lunch and poster viewing** **Nomenclature Committee meeting**
2:00 pm – 4:00 pm		**Plenary session 3. Diagnosis and typing: Histochemistry and proteomics** **Chairmen: Hazenberg and Phillips**
2:00	OP-13	The challenging diagnosis of transthyretin amyloidosis. P. Westermark (Uppsala, Sweden)
2:15	OP-14	Classification of amyloidoses using antibodies. Essentials of reliable and vices of unreliable immunohistochemistry. R. P. Linke (Martinsried, Germany)
2:30	OP-15	Immuno-electron microscopy in the classification of systemic amyloidoses: Experience in 423 patients from a single institution. C. F. de Larrea (Pavia, Italy and Barcelona, Spain)
2:45	OP-16	Fluorescence detection of amyloid in subcutaneous abdominal fat tissue. D. Sjolander* (Linkoping, Sweden)
3:00	OP17	Is it possible to use the proteome of amyloid n fat to predict cardiac and renal tropism? A. Dispenzieri (Rochester, MN, USA)
3:15	OP-18	Proteomic analysis of suspected amyloid in different tissues. J. D. Gillmore (London, UK)
3:30	OP 19	Proteome of amyloidosis: Mayo Clinic experience in 4139 cases. P.J. Kurtin (Rochester, MN, USA)
3:45	OP-20	Identification of hereditary fibrinogen A a-chain amyloidosis by proteomic analysis. R. H. Sayed* (London, UK)

4:00 pm – 4:30 pm	Coffee and tea break
4:30 pm – 6:00 pm	**Poster viewing and presentations by junior investigators***
6:00 pm	**Picnic buffet and baseball game at Victory Field**

Tuesday, April 29, 2014		
8:00 am – 10:30 am		**Plenary session 4. Imaging in diagnosis and organ disease** **Chairmen: Linke and Schonland**
8:00	OP-21	SPECT-based semi-quantitative assessment of ^{123}I-SAP scintigraphy in patients with amyloidosis. R. W. J. van Rheenen* (Groningen, the Netherlands)
8:15	OP-22	Detection of cardiac amyloidosis by SPECT/CT imaging using both ^{125}I-serum amyloid P-component and the novel ^{125}I- p5R+14 peptide. E. B. Martin* (Knoxville, TN, USA)
8:30	OP-23	High affinity amyloid-reactive peptide, p5R, binds non-uniformly to large amyloid deposits due to a binding site barrier effect. J.S. Wall (Knoxville, TN, USA)
8:45	OP-24	Non-coding genetic variation of the transthyretin gene in senile systemic amyloidosis. J. L. Sikora* (Boston, MA, USA)
9:00	OP-25	MR-neurography: In-vivo detection of nerve injury in systemic light chain (AL) amyloidosis. J. Kollmer* (Heidelberg, Germany)
9:15	OP-26	Light chain monoclonal immunoglobulin rapid accurate mass measurement (miRAMM) in patients with a monoclonal gammopathy. D. Murray (Rochester, MN, USA)
9:30	OP-27	Clarifying immunoglobulin gene usage in immunoglobulin light chain amyloidosis by mass spectrometry of amyloid in clinical tissue specimens. T.V. Kourelis (Rochester, MN, USA)
9:45	OP-28	Characterization of a novel peptide, p43 optimized for renal and pancreatic amyloid detection. J. S. Wall (Knoxville, TN, USA)
10:00	OP-29	The SMART-Amy Project: A smart guide towards the diagnosis of systemic amyloidosis. P. Russo* (Pavia, Italy and Republic of Korea)
10:15	OP-30	Human monoclonal antibodies specific for amyloid species. R. D. Puligedda* (Wynnewood, PA, USA)
10:30 am – 11:00 am		**Coffee and tea break**
11:00 am – 12:30 pm		**Plenary session 5. AL amyloidosis: Biology, clinics, and prognosis** **Chairmen: Kyle and Abonour**
11:00	OP-31	Characteristics and outcomes of 714 patients with systemic AL amyloidosis – analysis of a prospective study (ALChemy study). A. Wechalekar (London, UK)
11:15	OP-32	Risk and response adapted conventional treatment strategy in 147 patients with AL amyloidosis. A. Jaccard (Limoges, France)

11:30	OP-33	Soluble ST2 (sST2) is a novel valuable prognostic marker among patients with immunoglobulin light hcain (AL) amyloidosis. A. Dispenzieri (Rochester, MN, USA)
11:45	OP-34	Outcomes of primary systemic light chain (AL) amyloidosis in patients treated upfront with bortezomib or lenalidomide and the importance of risk adapted strategies. E. Kastritis (Athens, Greece)
12:00	OP-35	Comparison of the N-latex and Freelite assays for serum free light chain: Clinical performance in AL amyloidosis. G. Palladini (Pavia, Italy)
12:15	OP-36	A clinicopathological and long-term follow-up study of AH amyloidosis patients in Japan. M. Yazaki (Matsumoto, Japan)
12:30 pm – 2:00 pm		**Lunch and poster viewing**
2:00 pm – 4:30 pm		**Plenary session 6. AL therapy: Chemotherapies** **Chairmen: Hawkins and Roodman**
2:00	OP-37	Lenalidomide / melphalan / dexamethasone chemotherapy in 50 patients with newly diagnosed and advanced amyloid light chain amyloidosis: Results of aprospective single center phase 2 study (LEOMEX). S. O. Schonland (Heidelberg,Germany)
2:15	OP-38	Treatment of AL amyloidosis with two cycles of induction therapy with bortezomib and dexamethasone followed by bortezomib-high dose melphalan conditioning and autologous stem cell transplantation. V. Sanchorawala (Boston, MA, USA)
2:30	OP-39	Event-free and overall survival following risk-adapted melphalan stem cell transplant and consolidation for systemic light chain amyloidosis. H. Landau (New York, NY, USA)
2:45	OP-40	Long-term follow-up of patients with systemic light chain amyloidosis after bortezomib based treatment. R. F. Cornell (Nashville, TN, USA)
3:00	OP-41	A real world experience with cyclophosphamide, bortezomib, and corticosteroid combinations in patients with high-risk AL amyloidosis. A. Dispenzieri (Rochester, MN, USA)
3:15	OP-42	The addition of bortezomib to standard melphalan/dexamethasone improves the quality of response but does not reduce the rate of early deaths in AL amyloidosis: A matched case control comparison. G. Palladini (Pavia, Italy)
3:30	OP-43	Outcome of patients with immunoglobulin light chain amyloidosis with lung, liver, gastrointestinal, neurologic and soft tissue involvement after autologous hematopoietic stem cell transplantation. A. Afrough* (MD Anderson Cancer Center, USA)
3:45	OP-44	Once weekly subcutaneous bortezomib based induction therapy in systemic AL amyloidosis. J. Valent (Cleveland, OH, USA)

4:00	OP-45	Outcomes of AL amyloidosis patients treated with first line bortezomib: A collaborative retrospective Israeli multicenter assessment. M. E. Gatt (Jerusalem, Israel)
4:15	OP-46	Autologous stem cell transplant for AL amyloidosis: Impact of light chain isotype on transplant-related mortality. M. T. Cibeira (Barcelona, Spain)
After 6:00 pm		**No scheduled dinner. Open for consensus panels, corporate presentations**

4:30 pm – 6:30 pm	International Kidney Monoclonal Gammopathy – Satellite symposium

1. Introduction. Nelson Leung, IKMG President
2. MGRS: When the monoclonal gammopathy is no longer insignificant. N. Leung (Mayo Clinic
3. Rochester, MN, USA
4. Monoclonal gammopathy beyond amyloid – what you need to know. M. M. Picken (Loyola University, Chicago, IL, USA)
5. Renal response criteria in amyloidosis. G. Merlini (University of Pavia, Pavia, Italy)
6. IKMG membership group meeting.

6:00 pm – 7:30 pm	Symposium (Sponsored by Pfizer, Inc) Advancing knowledge and sharing experience in transthyretin cardiomyopathy Chair: Rapezzi
6:10 pm	Welcome and Introduction -- Rapezzi
6:15 pm	Cardiomyopathy (TTR-CM) -- Rapezzi
6:35 pm	TTR-CM case studies: Practical aspects of identification and diagnosis -- Hanna
7:00 pm	TTR-CM clinical trials -- Maurer
7:15 pm	Summary and Q & A -- Rapezzi

Wednesday, April 30, 2014		
8:00 am – 9:30 am		**Plenary session 7. More AL amyloidosis** **Chairmen: Merlini and Skinner**
8:00	OP-47	High response rates and minimal toxicity with subcutaneous bortezomib in combination regimens in newly diagnosed patients with systemic AL amyloidosis. G. Shah* (Boston, MA, USA)
8:15	**OP 48**	Long term results of pomalidomide and dexamethasone for patients with relapsed or refractory AL amyloidosis. A. Dispenzieri (Rochester, MN, USA)
8:30	OP-49	Identification of reversible renal damage and early markers of response to chemotherapy in two independent cohorts of patients with light chain amyloidosis: A longitudinal study on 732 newly diagnosed patients. G. Palladini (Pavia, Italy)
8:45	OP-50	Phase II trial of lenalidomide, dexamethasone and cyclophosphamide (LENDEXAL) for previously untreated patients with light-chain amyloidosis. M. T. Cibeira (Barcelona, Spain)
9:00	OP-51	Impact of induction therapy on the outcome of immunoglobulin light chain amyloidosis after autologous hematopoietic stem cell transplantation. A. Afrough* (M. D. Anderson Cancer Center, USA)
9:15	OP-52	Hereditary systemic immunoglobulin light-chain (AL) amyloidosis. M. D. Benson (Indianapolis, IN, USA)
9:30 am – 11:00 am		**Plenary session 8. ATTR: Genetics and basic biology.** **Chairmen: Ando and Buxbaum**
9:30	**OP 53**	Hereditary amyloidosis: A single institution experience with 284 patients. P.L. Swiecicki* (Rochester, MN, USA)
9:45	OP-54	Genealogic studies of the Swedish hereditary transthyretin V30M amyloidosis population – differences in age at onset within the population. O. B. Suhr (Umea, Sweden)
10:00	OP-55	The origin of the transthyretin V122I allele in Africa: A study of 2,620 DNA samples. D. R. Jacobson (Boston, MA, USA)
10:15	OP-56	Quantification of transthyretin kinetic stability in human plasma using subunit exchange. I. Rappley (La Jolla, CA, USA)
10:30	OP-57	Fragmentations of TTR in cultured cells. M. Ueda (Kumamoto, Japan)
10:45	OP-58	Understanding the role of proteolysis in transthyretin amyloidosis. V. Bellotti (London, UK and Pavia, Italy)

11:00 am – 11:30 am		**Coffee and tea break**
11:30 am – 1:00 pm		**Plenary session 9. ATTR: Diagnosis and pathogenesis** **Chairmen: Obici and Cummings**
11:30	OP-59	Neuropathy progression rate in patients with familial amyloidotic polyneuropathy. D. Adams (Paris, France)
11:45	OP-60	Early in-vivo detection of lower limb nerve injury in hereditary transthyretin familial amyloid polyneuropathy using high-resolution MR-neurography. J. Kollmer* (Heidelberg, Germany)
12:00	OP-61	Place of skin biopsy in asymptomatic and paucisymptomatic amyloidogenic TTR mutation gene carriers (TTR-GC). D. Adams (Paris, France)
12:15	OP-62	Global gene expression profiling of sex-specific inflammatory triggers of the transthyretin amyloidoses. S. M. Kurian (La Jolla, CA, USA)
12:30	OP-63	Haplotype analysis: modulation of AO through a trans-acting mechanism in familial amyloid polyneuropathy. M. Alves-Ferreira* (Porto, Portugal)
12:45	OP-64	Linking extracellular matrix remodeling genes and age-at-onset variability in familial amyloid polyneuropathy. D. Santos* (Porto, Portugal)
1:00 pm – 2:30 pm		**Lunch and poster viewing**
2:30 pm – 4:30 pm		**Plenary session 10. ATTR: Prognosis and therapy** **Chairmen: Ikeda and Seldin**
2:30	OP-65	Survival in patients with transthyretin familial amyloid polyneuropathy receiving tafamidis treatment. G. Merlini (Pavia, Italy)
2:45	OP-66	Interim analysis of long-term, open-label tafamidis treatment in transthyretin amyloid cardiomyopathy after up to 5 years of treatment. M. S. Maurer (New York, NY, USA)
3:00	OP-67	Familial amyloid polyneuropathy treatment with tafamidis – evaluation of one year treatment at Porto, Portugal. T. Coelho (Porto, Portugal)
3:15	OP-68	A phase II study of doxycycline plus tauroursodeoxycholic acid in transthyretin amyloidosis. L. Obici (Pavia, Italy)
3:30	OP-69	The prevalence of cardiac amyloidosis in familial amyloidotic polyneuropathy with predominant neuropathy: The diflunisal trial. C. C. Quarta* (Bologna, Italy and Boston, MA, USA)
3:45	OP-70	Safety and efficacy of long-term diflunisal administration in familial amyloid polyneuropathy – Summary of ten years therapeutic experience. Y. Sekijima (Matsumoto, Japan)

4:00	OP-71	Clinical development of ISIS-TTR_{Rx}: A second generation antisense therapy for the treatment of transthyretin-associated diseases. E. J. Ackermann (Carlsbad, CA, USA)
4:15	OP-72	Further analysis of phase II trial of patisiran, a novel RNAi therapeutic for the treatment of familial amyloidotic polyneuropathy. O. B. Suhr (Umea, Sweden)
4:30 pm – 5:00 pm		**ISA members meeting: Report on the journal Amyloid (Per Westermark, editor). Election results. Future symposia.**
5:00 pm – 6:30 pm		**Poster viewing and presentations by junior investigators*.**
6:30 pm – 6:45 pm		**Board buses from JW to IMA**
7:00 pm – 11:00 pm		**Congress dinner and awards presentations. Indianapolis Museum of Art**

Thursday, May 1, 2014		
8:00 am – 9:30 am		**Plenary session 11. Transplantation for amyloidosis** **Chairmen: Ericzon and Breall**
8:00	OP-73	Liver transplantation for hereditary ATTR-amyloidosis, any indication for non-V30M patients? O. B. Suhr (Umea, Sweden)
8:15	OP-74	Domino liver transplantation using familial amyloidotic polyneuropathy liver grafts; proposal for an international multicentre study to assess risk of *de novo* amyloidosis in the domino recipients. A. J. Stangou (Birmingham, UK)
8:30	OP-75	Who should receive the TTR domino liver? H. H.-J. Schmidt (Munster, Germany)
8:45	OP-76	Experiences of domino liver transplantations in Germany. A. P. Barreiros (Mainz, Germany)
9:00	OP-77	Regression of gastroduodenal mucosal amyloid deposits in FAP patients after combined therapy with oral intake of diflunisal followed by liver transplantation. T. Yoshinaga* (Matsumoto, Japan)
9:15	OP-78	Outcomes from an international registry of cardiac transplantation for light chain (AL) and transthyreitin (TTR) amyloidosis. M. Semigran (Boston, MA, USA)
9:30 am – 11:00 am		**Plenary session 12. Models and emerging therapies** **Chairmen: Palladini and Pickens**
9:30	OP-79	The small molecule Systebryl™ (PTI-110) causes potent disaggregation/reduction of AL, TTR and AA amyloid fibrils. A. D. Snow (Kirkland, WA, USA)
9:45	OP-80	Development of NPT088 for the treatment of amyloidosis disorders. V. C. Cullen (Cambridge, MA, USA)
10:00	OP-81	Plerixafor and G-CSF moblization for autologous stem cell transplantation in AL amyloidosis. E. Kaul* (Boston, MA, USA)
10:15	OP-82	Establishment of a *C. elegans* model to study amyloidogenesis of human B2-microglobulin *in vivo*. V. Bellotti (London, UK and Pavia, Italy)
10:30	OP-83	Differences in NT-proBNP release in patients with cardiac m-ATTR depend on the specific transthyretin mutation. S. Perlini (Pavia, Italy)

10:45 OP84	Human mesenchymal stromal cells protect human cardiomyocytes from primary amyloid light chain induced cytotoxicity. Y. Lin (Rochester, MN, USA)
11:00 am – 11:30 am	**Coffee and tea break**
11:30 am – 12:30 pm	**Closing session.** **Symposium overview and prospects: Skinner**
12:30 pm – 1:30 pm	**Lunch**