



*Santiago de Compostela*

**PRION  
2018**

May, 22-25

[www.prion2018.org](http://www.prion2018.org)



# **Program**

## Tuesday 22: AI-APRI Workshop: Structural biology of prions

8:30	Accreditation desk opens.
9:30	Welcome and opening remarks: <b>Holger Wille</b> (University of Alberta) & <b>Jesús Requena</b> (University of Santiago de Compostela).
9:35	<b>Tomas Sneideris</b> (Vilnius University): Properties of prion self-replication.
9:50	<b>Vincent Béringue</b> (INRA, Université Paris-Saclay): Dynamics of prion replication and structural diversification.
10:05	<b>Patricia Aguilar-Calvo</b> (UCSD): Heparan sulfate modulates the brain distribution of fibrillar prions.
10:20	<b>Alejandro Sevillano</b> (UCSD): Recombinant PrP <sup>Sc</sup> as a surrogate of brain PrP <sup>Sc</sup> for structural studies.
10:35	<b>Byron Caughey</b> (RML, NIH): Profound seeding selectivities of tau aggregates associated with Alzheimer's disease and other tauopathies.
10:55	Additional Q&A time.
11:00	<b>Coffee break</b>
11:30	<b>Ilia Baskakov</b> (University of Maryland): Strain-specific PrP <sup>Sc</sup> structures and N-linked glycans.
11:50	<b>Witold Surewicz</b> (Case Western Reserve University): Structural aspects of prion protein conversion to the infectious form.
12:10	<b>Giovanni Spagnoli</b> (University of Trento): A new structural model of PrP <sup>Sc</sup> based on recent experimental data and computational techniques.
12:25	<b>Holger Wille</b> (University of Alberta): High-resolution structures from A $\beta$ (1-42), $\alpha$ -synuclein, PHF-tau, and HET-s(218-289) - why not from PrP <sup>Sc</sup> ?
12:45	<b>Lunch</b>
13:45	<b>Antoine Loquet</b> (CNRS, Bordeaux): A new solid-state NMR approach to determine 3D structures of prion amyloid fibrils at atomic resolution.
14:00	<b>Michael Overduin</b> (University of Alberta): Prion:lipid analysis using membrane nanodiscs bounded by amphipathic polymers.
14:15	<b>Frederic Halgand</b> (CNRS-Université Paris Sud): Prion protein conformational landscape studied by mass spectrometry and ion mobility.
14:30	<b>Werner Kremer</b> (University of Regensburg): Full-length human prion protein displays a whole variety of conformational transitions as observed by high hydrostatic pressure (HHP), xenon as a probe and spin labelling of the N-terminal domain.
14:45	<b>Michael Woodside</b> (University of Alberta): Differences in the folding dynamics of prion proteins from species with different disease susceptibility observed at the single-molecule level.
15:00	<b>Lyudmyla Dorosh</b> (University of Alberta): Structural biology of prions: Effects of mutations, polymorphisms, and other variants of the PRND gene on the PrP <sup>C</sup> structure from MD simulations analyzed with essential collective dynamics and machine learning methods.
15:15	<b>Dieter Willbold</b> (Forschungszentrum Jülich): High resolution fibril structure of amyloid- $\beta$ (1-42) by cryoelectron microscopy.
15:35	Additional Q&A time.
15:45	<b>Coffee break</b>
16:15	<b>Boran Uluca</b> (University of Düsseldorf): DNP-enhanced solid-state NMR at cryogenic temperatures: a tool to snapshot conformational ensembles of $\alpha$ -synuclein in different states.
16:30	<b>Martin Margittai</b> (University of Denver): Molecular basis for seeding barriers between three- and four-repeat tau.
16:45	<b>Cristina Fernández</b> (CIB-CSIC, Madrid): A bacterial prion-like protein as a synthetic model to study amyloid toxicity.
17:00	<b>Reed Wickner</b> (NIH, Bethesda): Five prion-curing systems in <i>Saccharomyces cerevisiae</i> : U <sup>pf</sup> proteins, inositol polyphosphates, B <sup>tn2p</sup> , C <sup>ur1p</sup> , and H <sup>sp104p</sup> .
17:20	Additional Q&A time. Final remarks.
17:45	<b>Welcome cocktail (Conference center, hall)</b>

## Tuesday 22: AI-APRI Workshop: Animal prion disease

- 8:30 Accreditation desk opens.
- 9:30 Welcome and Opening Remarks: **Judd Aiken** (University of Alberta) and **Debbie McKenzie** (University of Alberta).
- 9:35 **Candace Mathiason** (Colorado State University): An Overview-Chronic Wasting Disease mother to offspring transmission studies conducted at Colorado State University.
- 10:05 **Hermann Schätzl/Sandor Dudas** (University of Calgary): Oral transmission of CWD into Cynomolgus macaques: signs of atypical disease, prion conversion and infectivity in macaques and bio-assayed transgenic mice.
- 10:35 **Sylvie Benestad** (Norwegian Veterinary Institute): How is the CWD situation in Norway?
- 10:50 Additional Q&A time.

### 11:00 Coffee Break

- 11:30 **Camilo Duque Velasquez** (University of Alberta): Evolution of Chronic Wasting Disease prion conformers.
- 11:45 **Nate Denkers** (Colorado State University): Assessing CWD prion neuro-invasion by longitudinal CSF analysis in deer.
- 12:00 **Jifeng Bian** (Colorado State University): Advances in CWD prion research.
- 12:15 **Judd Aiken** (University of Alberta): Interaction of CWD prions with environmental samples.
- 12:30 **Rodrigo Morales** (University of Texas Health Science Center, Houston): PMCA as a platform for pre-symptomatic CWD diagnosis. Validation in field samples.

### 12:45 Lunch

- 13:45 **Juan Carlos Espinosa** (CISA-INIA, Madrid): Pig-PrP as a paradigm of resistance to prion propagation.
- 14:15 **Gabriele Vaccari** (Istituto Superiore di Sanità, Rome): Prion disease in dromedary camel.
- 14:45 **Anne Balkema-Buschmann** (Friedrich-Loeffler-Institut): Absence of classical and atypical (H- and L-Type) BSE prions in the blood of clinical bovine cases as confirmed by cattle to cattle blood transfusions.
- 15:00 **Giuseppe Ru** (Istituto Zooprofilattico Sperimentale Piemonte, Liguria e Valle d'Aosta, Turin): A scrapie cohort study shows the success of breeding for resistance.
- 15:15 **Khalid Salamat** (The Roslin Institute): Detection of seeding activity in preclinical blood samples from BSE-infected sheep using protein misfolding cyclic amplification (PMCA).
- 15:30 Additional Q&A Time.

### 15:45 Coffee Break

- 16:15 **Pierre Sibille** (INRA-Université Paris-Saclay): Prion strain selection and evolution.
- 16:30 **Jo Moore** (USDA, Ames): The agent of chronic wasting disease from pigs is infectious in transgenic mice expressing human PRNP.
- 16:45 **Ermias Belay** (CDC, Atlanta): Monitoring potential CWD transmission to Humans.
- 17:00 **Hermann Schätzl** (University of Calgary): Vaccination of transgenic elk PrP mice and reindeer with recombinant prion protein overcomes self-tolerance and protects mice against CWD infection.
- 17:15 Final remarks.

### 17:45 Welcome cocktail (Conference center hall)

## Wednesday 23

- 8:30 Accreditation desk opens.  
9:30 **Opening Plenary:** Chairs: **Jesús Requena** (University of Santiago de Compostela, Conference Chair) & **Emmanuel Comoy** (CEA.Université Paris-Saclay, NeuroPrion Association).  
9:30 Opening addresses.  
10:00 Plenary lecture: **Stanley Prusiner** (UCSF): A unified view of prion biology and diseases.

### 11:00 Coffee break

- 11:30 **Session: PrP<sup>Sc</sup> prions: animal disease (I)**, Chair: **Juan María Torres** (CISA-INIA, Madrid); Junior Co-chair: **Alba Marín-Moreno** (CISA-INIA, Madrid).  
11:30 Invited talk: **Sylvie Benestad** (Norwegian Veterinary Institute): Unusual types of CWD in Norwegian cervids.  
12:30 **Gabriele Vaccari** (Istituto Superiore di Sanità, Rome): Prion disease in dromedary camels.

### 12:45 Lunch buffet & poster viewing

- 14:15 **Session: PrP<sup>Sc</sup> prions: animal disease (II)**, Chair: **Juan Badiola** (University of Zaragoza); Junior co-Chair: **Alba Marín-Moreno** (CISA-INIA, Madrid)  
14:15 **Romolo Nonno** (Istituto Superiore di Sanità, Rome): Different chronic wasting disease strains circulate in North America and Europe.  
14:30 **Glenn Telling** (Colorado State University): The strain properties of prions causing chronic wasting disease in Norwegian cervids are distinct from those causing disease in North America.  
14:45 **Glenn Telling** (Colorado State University): Gene targeted mice reveal an essential role for PrP residue 226 and an accurate means to model CWD peripheral pathogenesis.  
15:00 **Alvina Huor** (INRA, Toulouse): Permeability of the bovine transmission barrier to Atypical / Nor98 scrapie.  
15:15 **Alba Marín-Moreno** (CISA-INIA, Madrid): Zoonotic potential of atypical BSE prions: a systematic evaluation.

### 15:30 Coffee break

### 16:00 Poster session: poster presenting authors will be at their posters

- 17:00 **Session: PrP<sup>Sc</sup> prions: animal disease (III)**, Chairs: **Juan Badiola** (University of Zaragoza) and **Juan María Torres** (CISA-INIA, Madrid).  
17:00 **Wen-Quan Zou** (Case Western Reserve University): Preclinical detection of prions in skin samples of scrapie-infected animals by serial PMCA and RT-QuIC assays.  
17:15 Invited talk: **Rosa Bolea** (University of Zaragoza): Experimental studies on prion transmission barrier and TSE pathogenesis in large animals.

### 17:45 End of scientific sessions for the day

## Thursday 24

- 9:00 **Session: Structure and basic biology of PrP<sup>Sc</sup> and other mammalian, and fungal prions.** Chair: **Jesús Requena** (University of Santiago de Compostela); Junor Co-chair: **Alejandro Sevillano** (UCSD).
- 9:00 Discussion table: The structure of PrP<sup>Sc</sup>; Chair: **Jesús Requena**; Junior Co-chair: **Alejandro Sevillano** (UCSD). Experts: **Holger Wille** (University of Alberta), **Witold Surewicz** (Case Western Reserve University), **Byron Caughey** (RML-NIH), **Iliia Baskakov** (University of Maryland).
- 10:30 **Iaria Vanni** (Istituto Superiore di Sanità, Rome): Purified 7 kDa PrP<sup>res</sup> aggregates from GSS-A117V are infectious.
- 10:45 **Christina Sigurdson** (UCSD): Generation of novel neuroinvasive prions following intravenous challenge.

### 11:00 Coffee break

- 11:30 Invited talk: Sven Saupe (CNRS, Bordeaux): Prion forming domains in cell death signaling.
- 12:30 **Sarah Kane** (Colorado State University): a novel means of epitope unmasking due to incomplete N-glycan attachment establishes that PrP<sup>Sc</sup> is underglycosylated relative to PrP<sup>C</sup> and affords prion detection.

### 12:45 Lunch buffet & poster viewing

- 14:15 **Session: A $\beta$ , tau,  $\alpha$ -synuclein, SOD1, TDP43, FUS...and other prion and/or prion-like proteins causing human disease.** Chair: **Mathias Jucker** (DZNE-Tübingen); Junior Co-chair: **Hasier Eraña** (CIC-bioGUNE, Bilbao).
- 14:15 Discussion table: are they prions, prion-like proteins, or what? Chair: **Mathias Jucker** (DZNE, Tübingen); Junior Co-chair: **Hasier Eraña** (CIC-bioGUNE, Bilbao). Experts: **Stanley Prusiner** (UCSF), **Claudio Soto** (University of Texas Houston Medical School), **Erdem Tamguney** (DZNE, Bonn), **Corinne Lasmezas** (Scripps, Florida).

### 15:45 Coffee break

- 16:15 **Natalie Beschorner** (DZNE, Tübingen): Amyloid polymorphisms constitute distinct clouds of conformational variants in different etiological subtypes of Alzheimer's disease.
- 17:00 **Amanda Woerman** (UCSF): Multiple system atrophy prions replicate in Tg(SNCAA53T) mice and induce glial pathology throughout the limbic system.
- 17:15 **Jacob Ayers** (University of Florida, Gainesville): Prion strain-like properties of misfolded superoxide dismutase-1 (SOD1).

### 17:30 Poster competition winner announced

### 17:35 End of scientific sessions of the day

### 19:30 Cultural event: visit to the cathedral and *botafumeiro*

### 20:30 Gala Dinner & Neurovision dance extravaganza (Pazo de San Lourenzo)

## Friday 25

- 9:00 **Session: PrP<sup>Sc</sup> prions: human disease (I).** Chair: **George Carlson** (UCSF); Junior Co-chair: **Aušrinė Areškevičiūtė** (University Hospital, Copenhagen).
- 9:00 **Emmanuelle Viré** (University College, London): Epigenetics in prion diseases: identification of novel markers for misfolded protein-associated neurodegenerative disorders.
- 9:15 **Emmanuel Comoy** (CEA-Université Paris-Saclay): Experimental transfusion of variant CJD-infected blood reveals novel class of prion disorders.
- 9:30 **Jean-Yves Douet** (INRA, Toulouse): Evaluation of iatrogenic risk of CJD transmission associated with corneal transplantation.
- 9:45 **Session: Patients & families: a dialogue with scientists;** Chair: **Raffaella Robello** (Italian Association on Prion Diseases).
- 9:45 **Suzanne Solvyns & Deana Simpson** (CJD-ISA): Reflections and testimonials.
- 10:20 **Debbie Yobs** (CJD Foundation-USA): Announcement of the CJDF 2018 call for research projects.
- 10:25 **Alice Anane** (CJD Foundation-Israel): Creating a large database of gCJD samples in Israel.

### 10:30 Coffee break

- 11:00 **Session: PrP<sup>Sc</sup> prions: human disease (II).** Chair: **George Carlson** (UCSF); Junior Co-chair: **Aušrinė Areškevičiūtė** (University Hospital, Copenhagen).
- 11:00 **Cécile Voisset** (INSERM-Brest University): AP326, a new FDA-approved drug active against PrP<sup>Sc</sup>.
- 11:15 Discussion table: Prion therapy: where do we stand? Chair: **Joaquín Castilla** (CIC-bioGUNE, Bilbao); Junior Co-chair: **Akin Nihat** (University College, London); Experts: **John Collinge** (University College, London), **Fabrizio Tagliavini** (Carlo Besta Institute, Milan), **Kurt Giles** (UCSF), **Kenta Teruya** (Tohoku University).

### 12:45 Lunch buffet & poster viewing

- 14:15 **Session: special, hot and/or late-breaking.** Chair: **Jiyan Ma** (Van Andel Institute); Junior Co-chair: **Alzbeta Cardova** (Cambridge University).
- 14:15 Brain junk clearing mechanisms: **Hermann Schätzl** (University of Calgary): Autophagy has various roles in the life cycle of prions.
- 14:30 Prion precursor biology & functions: **Cathryn Haigh** (University of Melbourne): Redox regulation of adult neurogenesis by PrP and PrP N-terminal cleavage fragments.
- 14:45 **Joaquín Castilla** (CIC-BioGUNE, Bilbao): Shaken, not sonicated: A strategy to generate large amounts of recombinant bona fide prions for definitive structural studies.
- 15:00 **Andrew Fang** (University of Alberta): Rationally designed, structure-based vaccine candidates targeting Chronic Wasting Disease.
- 15:15 **Giuseppe Legname** (SISSA, Trieste): Serpina3n plays a critical role in prion infection.

### 15:30 Coffee break

- 16:00 **Closing plenary**
- 16:00 Winning poster presentation.
- 16:15 Presentation: Iberian Prion Meeting 2018, Laguardia, Spain, **Joaquín Castilla** (CIC-BioGUNE, Bilbao).
- 16:25 Presentation: PRION 2019 Edmonton: **Hermann Schätzl** (University of Calgary) & **David Westaway** (University of Alberta).
- 16:40 Concluding remarks from NeuroPrion: **Emmanuel Comoy**.
- 16:50 Concluding remarks: **Jesús Requena**.

### 17:00 Farewell

*“Prudens quaestio dimidium scientiae”*

*Francis Bacon*



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*Cover: Triple baroque staircase,  
San Domingos de Bonaval, Santiago de Compostela  
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*Laguardia, Spain  
25-26 Oct. 2018.*

