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A muscle-specific MuRF1-E2 Network requires stabilization of MuRF1-E2 complexes by Telethonin, a newly identified substrate

Cécile Polge, Stéphanie Cabantous, Christiane Deval, Agnes Claustre, Antoine Hauvette, Catherine Bouchenot, Julien Aniort, Daniel Béchet, Lydie Combaret, Didier Attaix, et al.

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PROTEOSTASIS



EUROPEAN COOPERATION
IN SCIENCE & TECHNOLOGY



Proteostatic Mechanisms in Health and Disease

The Final COST Action BM1307 Meeting

22-24 February 2018

Divani Palace Acropolis, Athens, Greece

Programme

| Thursday, 22 February 2018 | |
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| 09:00-16:00 | Registration |
| 09:00-11:45 | MC Meeting |
| 11:45-12:00 | <i>Welcome</i> |
| 12:00-12:45 | Keynote Talk 1: Aaron Ciechanover [Technion – Israel Institute of Technology, Haifa, Israel] <i>The Ubiquitin Proteolytic System - From Basic Mechanisms thru Human Diseases and on to Drug Development</i> |
| 12:45-13:15 | Thematic Talk 1: Manuel Rodriguez [CNRS, Toulouse, France] – Core Group Member; Networking co-Coordinator <i>Evidences for an Active Proteaphagy in Bortezomib Resistant Mantle Cell Lymphoma Cells</i> |
| 13:15-14:30 | Lunch |
| 14:30-14:45 WG-2 | <i>Complex Regulation of pro-atrophic Ubiquitin Ligase MuRF1 by the p53 Family in Amyotrophic Lateral Sclerosis and Cancer-related Sarcopenia</i> Barthe Anaïs , Von Grabowiecki Yannick, De Abreu Paula, Aina Venkatasami, Blanchard Orphée, Devignot Véronique, Mellitzer Georg and Gaidon Christian |
| 14:45-15:00 WG-2 | <i>PIP30/FAM192A, a Novel Regulator of the Nuclear 20S Proteasome / PA28γ Complex</i> Jonik-Nowak B, Menneteau T, Fesquet D, Baldin V, Bonne-Andrea C, Méchali F, Fabre B, Boisguerin P, de Rossi S, Henriquet C, Pugnière M, Ducoux-Petit M, Burlet-Schiltz O, Lamond AI, Fort P, Boulon S, Bousquet M-P, Coux O |
| 15:00-15:15 WG-2 | <i>Regulation of ERAD through Androgen Signaling in Prostate Cancer</i> Erzurumlu Y , Ballar Kirmizibayrak P |
| 15:15-15:30 WG-2 | <i>Proteasome Activation by Small Molecules</i> Ovaa H |
| 15:30-15:45 WG-2 | <i>ER-PM Contact sites Control EGFR Endocytosis and Signaling</i> Caldieri G, Barbieri E, Conte A, Raimondi A, Bonora M, Tacchetti C, Pinton P, di Fiore PP, Sigismund S |
| 15:45-16:15 | Coffee break |
| 16:15-16:45 | Thematic Talk 2: Nico Dissmeyer [Leibniz Institute of Plant Biochemistry, Halle, Germany] – Core Group Member; Dissemination co-Coordinator <i>Life and Death of Proteins after Protease Cleavage: Protein Degradation by the N-end Rule Pathway</i> |
| 16:45-17:00 WG-5 | <i>In silico Biophysics Accurately Predicts Protein Quality Control Targets</i> Stein A, Nielsen SV, Kampmeyer C, Abildgaard A, Clausen L, Lindorff-Larsen K, Hartmann-Petersen R |
| 17:00-17:15 WG-5 | <i>Structural Insights into the Regulation of an Unusual Viral DUB Enzyme: "Some Like to Move it, Move it!"</i> Jupin I , Ayach M, Jomat L, Fieulaine S, Bressanelli S |

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| 17:15-17:30 WG-5 | <u>Biochemical Properties and Cellular Function of the E3 Ubiquitin Ligase HECTD1</u> Vaughan N, Byrne R, Kessler B, Lindon C, Licchesi JDF |
| 17:30-17:45 WG-5 | <u>Understanding the Dynamics of Stress-induced Protein Aggregates in Plants</u> Li L, Popa C, Malgrem Hill S, Valls M, Nyström T, Sanchez Coll N |
| 17:45-18:00 WG-5 | <u>The Deubiquitinating Enzyme ATXN3 is Relevant for Photoreceptor Ciliogenesis</u> Toulis V , do Carmo Costa M, Marfany G |
| Friday, 23 February 2018 | |
| 09:00-09:45 | Keynote Talk 2: Daniel Finley [Harvard Medical School, Boston, USA] <u>Recognition and Processing of Ubiquitin Conjugates by the Proteasome</u> |
| 09:45-10:15 | Thematic Talk 3: Michael H. Glickman [Technion – Israel Institute of Technology, Haifa, Israel] – MC Member; Networking co-Coordinator <u>“and Proteasome, would you like any Ubiquitin with your Substrate?”</u> |
| 10:15-10:45 | ☕ Coffee break |
| 10:45-11:00 WG-1 | <u>Metabolic Regulation of the Proteasome</u> Meul T , Berschneider K, Schmitt S, Mayr CH, Schiller HB, Lukas C, Eickelberg O, Prehn C, Adamski J, Perocchi F, Kukat A, Trifunovic A, Kremer L, Prokisch H, Popper B, von Toerne C, Hauck SM, Zischka H, Meiners S |
| 11:00-11:15 WG-1 | <u>Intrinsically Disordered Regions Fine-tune AUX/IAA Ubiquitylation</u> Niemeyer M , Kölling M, Hellmuth A, Winkler M, Wilde V, Reimers M, Calderón Villalobos Luz IA |
| 11:15-11:30 WG-1 | <u>Progress towards Using in vivo Biotinylation and CRISPR/Cas9 Genome Editing for Novel Exploration of the SUMOylation Pathway</u> Sutherland JD , Giordano I, Elortza F, Barrio R |
| 11:30-11:45 WG-1 | <u>Interplay between SUMOylation and NEDDylation Regulates RPL11 Localization and Function</u> EI Motiam A , Vidal S, de la Cruz-Herrera CF, Da Silva Álvarez S, Baz-Martínez M, Vidal A, Rodríguez MS, Xirodimas DP, Carvalho AS, Beck HC, Matthiesen R, Collado M, Rivas C |
| 11:45-12:00 WG-1 | <u>SUMO and Ubiquitin Cooperate to Drive Mitosis</u> Eifler K, Cuijpers S, Liebelt F, González-Prieto R, Willemstein E, Vertegaal A |
| 12:00-13:30 | 🍽️ Lunch |
| 13:00-15:00 | Poster Session 1 |
| 15:00-15:45 | Keynote Talk 3: Nektarios Tavernarakis [IMBB-FORTH & University of Crete, Heraklion, Greece] <u>Regulation and Roles of Autophagy in Mammalian Brain Proteostasis</u> |
| 15:45-16:15 | Thematic Talk 4: Nico Dantuma [Karolinska Institutet, Stockholm, Sweden] – MC Member; Dissemination co-Coordinator <u>Interplay Between Polyglutamine Proteins and the Ubiquitin/Proteasome System</u> |
| 16:15-16:45 | ☐ Coffee break |
| 16:45-17:00 WG-4 | <u>Sphingolipid- and Conformation-dependent Partitioning of Yeast Transporters into Starvation-protective Membrane Domains</u> Gournas C , Gkionis S, Carquin M, Twyffels L, Tyteca D, André B |
| 17:00-17:15 WG-4 | <u>The Ubiquitin Enzymes IAPs in the Regulation of Autophagy</u> Ebner P, Ikeda F |
| 17:15-17:30 WG-4 | <u>Evolution of Selective Autophagy as a Mechanism of Oxidative Stress Response</u> Carroll B, Otten EG, Manni D, Stefanatos R, Menzies FM, Smith GR, Jurk D, Kenneth N, Wilkinson S, Passos JF, Attems J, Veal EA, Teyssou E, Seilhean D, Millecamps S, Eskelinen E-L, Bronowska AK, Rubinsztein DC, Sanz A, Korolchuk VI |
| 17:30-17:45 WG-4 | <u>Regulation of Basal Mitophagy by Deubiquitylases</u> Urbé S , Marcassa E, Jardine J, Kallinos A, Martínez A, Bertsoulaki E, Clague MJ |

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| 17:45-18:00 WG-4 | <u><i>Epigallocatechin-3-Gallate Promotes Autophagy-Dependent Survival via Influencing the Balance of mTOR-AMPK Pathways upon Endoplasmic Reticulum Stress</i></u> Holczer M, Besze B, Zámbo V, Csala M, Bánhegyi G, Margittai É , Kapuy O |
| 18:00-20:00 | Poster Session 2 |
| 20:30 | ☪ Depart for Gala Dinner |
| Saturday, 24 February 2018 | |
| 09:00-09:45 | Keynote Talk 4: Kelvin J.A. Davies [University of Southern California, Los Angeles, USA] <u><i>The Decline of Adaptive Proteostasis in Ageing</i></u> |
| 09:45-10:15 | Thematic Talk 5: Elah Pick [University of Haifa, Tivon, Israel] – MC Member; Dissemination co-Coordinator <u><i>COP9 Signalosome Subunit 5 Maintains Both Proteostasis and Lipidostasis</i></u> |
| 10:15-10:30 | Rosa Barrio [CIC bioGUNE, Functional Genomics Unit, Spain] - Chair <u><i>An Assessment of COST PROTEOSTASIS</i></u> |
| 10:30-11:00 | ☪ Coffee break |
| 11:00-11:15 WG-3 | <u><i>KHNYN, a Multitasking Protein that Potentially Orchestrates Protein Trafficking and RNA Metabolism</i></u> Castagnoli L, Mandaliti W, Nepravishta R, Giordano F, Valentini E, Mattioni A, Ferrentino F, Procopio R, Polo S, Paci M, Cesaren G, Santonico E |
| 11:15-11:30 WG-3 | <u><i>NEDD8 Promotes Reversible Nuclear Protein Aggregation and Protects the Ubiquitin Proteasome System upon Proteotoxic Stress</i></u> Maghames CM, Lobato-Gil S, Perrin A, Trauchessec H, Rodriguez MS, Urbach S, Marin P, Xirodimas DP |
| 11:30-11:45 WG-3 | <u><i>DENEDDYLASE1 (DEN1) Removes NEDD8 from NEDD8 Conjugates that have become Neddylated by Non-enzymatic Neddylation</i></u> Mergner J, Schwechheimer C |
| 11:45-12:00 WG-3 | <u><i>Nrf2 as a Main Hub in the Wiring of Proteostatic, Mitostatic and Metabolic Pathways</i></u> Tsakiri EN, Gumeni S, Iliaki KK, Benaki D, Vougas K, Sykiotis GP, Gorgoulis VG, Mikros E, Scorrano L, Trougakos IP |
| 12:00-13:00 | Rapid Oral Poster Presentations (5 minutes) 1. A. Busturia (WG-1): <u><i>Epigenetic and Non-epigenetic Functions of dRYBP, a Ubiquitin Binding Protein</i></u> 2. L.I.A. Calderón Villalobos (WG-1): <u><i>Auxin-programmed Ubiquitylation as a Master Signal Integration System</i></u> 3. P. Domingos (WG-1): <u><i>The Fbox Protein CG6758 Regulates Xbp1-induced Cell Death in the Drosophila eye</i></u> 4. I.E. Kammerl (WG-2; STSM participant): <u><i>Proteasome Activity Profiling as a Biomarker Approach for Patients with Chronic Obstructive Pulmonary Disease (COPD)</i></u> 5. G. Bánhegyi (WG-4): <u><i>The Role of PERK Pathway in Life-and-Death Decision upon Endoplasmic Reticulum Stress</i></u> 6. M.H. Ogmundsdottir (WG-2): <u><i>A Rare Germline Coding Variant Affects ATG7 Function</i></u> 7. A. Sibirny (WG-3): <u><i>Gss1 Glucose Sensor is Involved in Pexophagy Induction in the Yeast Komagataella phaffii (Pichia pastoris)</i></u> 8. V. Welk (WG-2; STSM participant): <u><i>Proteasome Activator 200 (PA200) is Dysregulated in Fibrotic Tissue Remodeling</i></u> 9. T. Zoladek (WG-2): <u><i>Using Yeast vps13 Models of Chorea-acanthocytosis to Isolate Genetic and Chemical Suppressors</i></u> |
| 13:00-14:15 | ☪ Lunch |
| 14:15-14:45 | Thematic Talk 6: Wiep Scheper [Vrije University, Amsterdam, Netherlands] – Core Group Member; Working Group Leader <u><i>The Neuronal Unfolded Protein Response and Neurodegenerative Disease</i></u> |

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| 14:45-15:00 WG-6 | <u>Quantitative Proteomics Reveals Neuronal Ubiquitination of Rngo/Ddi1 and Several Proteasomal Subunits by Ube3a, Accounting for the Complexity of Angelman Syndrome</u> Ramirez J, Lectez B, Osinalde N, Sivá M, Elu N , Aloria K, Procházková M, Perez C, Martínez-Hernández J, Barrio R, Grantz Šašková K, Arizmendi JM, Mayor U |
| 15:00-15:15 WG-6 | <u>Conditional Loss of FBXO7/PARK15 Results in a Parkinson's-like Dopaminergic Degeneration via a p53 Dathway</u> Stott SRW, Randle SJ, Rowicka P, Xia J, Dalley JW, Barker RA, Laman H |
| 15:15-15:30 WG-6 | <u>Novel Protein Interactions and Functional Roles of USP14 in Neurodegenerative Diseases</u> Srinivasan V, Bruelle C, Lindholm D |
| 15:30-16:00 | ☕ Coffee break |
| 16:00-16:15 WG-6 | <u>A Muscle-specific MuRF1-E2 Network Requires Stabilization of MuRF1-E2 Complexes by Telethonin, a Newly Identified Substrate</u> Polge C, Cabantous S, Deval C, Claustre A, Hauvette A, Bouchenot C, Aniort J, Béchet D, Combaret L, Attaix D, Taillandier D |
| 16:15-16:30 WG-6 | <u>Nrf2 Couples Antioxidant Defense with Thyroglobulin Production and Iodination in the Thyroid Gland</u> Ziros PG, Habeos IG, Chartoumpakis DV, Ntalampyra E, Somm E, Renaud CO , Bongiovanni M, Trougakos IP, Yamamoto M, Kensler TW, Santisteban P, Carrasco N, Ris-Stalpers C, Amendola E, Liao X-H, Rossich L, Thomasz L, Juvenal GJ, Refetoff S, Sykiotis GP |
| 16:30-16:45 WG-6 | <u>Regulation of the Ebolavirus VP24 Protein by SUMO</u> Vidal S , El-Motiam A, Ruibal P, Seoane R, Rodríguez MS, Rejas MT, Rodríguez D, Muñoz-Fontela C, Rivas C |
| 16:45-17:00 | Awards & Closing Remarks |

*WG-Working Group

*The Chair of each WG Session will be its corresponding Thematic Speaker