
**V Meeting on the
MOLECULAR MECHANISMS OF NEURODEGENERATION**

Aula Magna, Università degli Studi di Milano
Via Festa del Perdono 7 - 20122 Milano (Italy)
May 13-15, 2011

FINAL PROGRAM

Friday, May 13, 2011 (Morning)

09:30 Registration

10:00 Satellite symposium on:
**Molecular Mechanisms on Neurodevelopment
New insights on synapse development and function**
(no registration required for this symposium)
Chairpersons: Roberto Maggi (Italy), Fabio Benfenati (Italy)

Plenary Lecture

10:15-11:00 **Fabio Benfenati**
"The key actors of synapse function and plasticity"

Session

11:00-11:30 **Michela Matteoli**
"From filopodia to synapsis: the cell biology of neuronal wiring"

11:30-12:00 **Alexander Dityatev**
"The dual role of the extracellular matrix in synaptic plasticity and homeostasis"

12:00-12:30 **Carlo Sala**
"Molecular organization and function of scaffold proteins at neuronal synapses"

**V Meeting on the
MOLECULAR MECHANISMS OF NEURODEGENERATION**

Friday, May 13, 2011 (Afternoon)

Opening Session

14:00-14:30 Introduction by the organizers
Angelo Poletti, Elena Cattaneo, Franco Taroni (Italy)

Prof. Fabio Celotti
(Director of the DEFIB, Università degli Studi di Milano)

Prof. Ferdinando Cornelio
*(Scientific Director of the Fondazione IRCCS Istituto Neurologico
"Carlo Besta", Milano)*

Kenneth H. Fischbeck (USA) and **Albert La Spada** (USA)
"Of thesis committees, birthday gels, and dubious reviewers: A personal recollection of the discovery of the first repeat expansion mutation"

Opening lecture

14:30-15:30 **Harry Orr** (USA)
"Polyglutamine disorders 20 years later"
Introduced by Kenneth H. Fischbeck

Session I: Cellular handling of aberrant neurotoxic proteins I
Chairpersons: Harry Orr (USA), Michela Matteoli (Italy)

15:30-16:00 **Ralph A. Nixon** (USA)
"Autophagy Failure in Alzheimer's Disease"

16:00-16:30 **Kenneth H Fischbeck** (USA)
"Therapeutic approaches to Spinal and Bulbar Muscular Atrophy"

16:30-17:00 *Coffee break*

Session II: Cellular handling of aberrant neurotoxic proteins II
Chairpersons: Harry Orr (USA), Michela Matteoli (Italy)

17:00-17:30 **Francesco Cecconi** (Italy)
"Caspase-3 triggers early synaptic dysfunction in a mouse model of Alzheimer's Disease"

17:30-18:00 **Sandrine Humbert** (France)
"Huntington's disease and beta-catenin homeostasis"

18:00-19.30 *Selected Oral Presentations*
Chairpersons: Roberto Maggi (Italy), Giorgio Battaglia (Italy)

OC-1 "USP14 functions in a non-catalytic manner to regulate synaptic activity"
Walters BJ, Dobrunz LE and Wilson SM (USA)

OC-2 "The role of Parkinson's disease associated Pink1 in the quality control mechanisms of mitochondrial dynamics and autophagy"
Mai S, Klinkenberg M, Auburger G, Bereiter-Hahn J, Jendrach M (Germany)

OC-3 "Identification of drosophila ortholog of HSPB8: implication of HspB8 loss of function in protein folding disease"
Boncoraglio A, Kanon B, Brunsting JF, Minoia M, Rana A, Vos MJ, Seidel K, Sibon OCM, Kampinga HH and Carra S (The Netherlands)

OC-4 "Pharmacological and molecular approaches to remove mutant misfolded androgen receptor in spinal and bulbar muscular atrophy"
Rusmini P, Crippa V, Giorgetti E, Sau D, Poletti A (Italy)

OC-5 "In vitro drug treatments aimed at rescuing the correct nuclear localization and transactivating function of polyala expanded PHOX2B proteins"
Di Zanni E, Bachetti T, Bocca P, Prigione I, Di Lascio S, Fornasari D, Ravazzolo R, Ceccherini I (Italy)

OC-6 "Involvement of peroxisomes in early hippocampal neurodegeneration in a transgenic mouse model of Alzheimer's disease"
Fanelli F, D'Angelo B, Benedetti E, Sepe S, Cristiano L, D'Amelio M, Bernardi C, Cecconi F, Cimini A, Cerù MP, Moreno S (Italy)

19:30 *Welcome Party*

Saturday, May 14, 2011 (Morning)

08:30-10:00 *Selected Oral Presentations*

Chairpersons: Fabrizio Tagliavini (Italy), Caterina Mariotti (Italy)

- OC-7 "ER-mitochondria Ca²⁺ cross-talk in AD: (FAD-)PS2 tightens the link"
Zampese E, Filadi R, Kipanyula MJ, Capitanio P, Pozzan T, Fasolato C, and Pizzo P (Italy)
- OC-8 "Selective intracellular targeting and interference of Alzheimer's amyloid beta oligomers with conformational intrabodies"
Meli G, Lecci A, Krako N, Manca A, Albertini V, Ghidoni R, Cattaneo A (Italy)
- OC-9 "Misfolded protein oligomers formed by a model protein mimic the effect of A-beta oligomers on synaptic plasticity involved in Alzheimer's disease"
Tatini F, Pugliese AM, Traini C, Niccoli S, Maraula G, Eddami T, Mannini B, Pedata F, Casamenti F, Chiti F (Italy)
- OC-10 "Prodynorphin mutations cause the neurodegenerative disorder spinocerebellar ataxia type 23"
Bakalkin G, Jezierska J, Watanabe H, Depoorter C, Bazov I, Verschuuren-Bemelmans C, Artemenko KA, Van de Warrenburg BPC, Hauser KF, Sinke RJ and Verbeek DS (The Netherlands)
- OC-11 "Analysis of staminal and neuronal microRNA expression in spinal cord and ependymal stem cells from G93A-SOD1 amyotrophic lateral sclerosis mice"
Marcuzzo S, Tartari S, Cavalcante P, Bonanno S, Cappelletti C, Bernasconi P, Mantegazza R (Italy)
- OC-12 "Reversal of reduced neuronal and BDNF gene transcription in HD mice by dominant negative REST"
Conforti P, Mas Montes A, Zuccato C, Soldati C, Buckely N, Davidson B, Cattaneo E (Italy)

Session III: Non cell autonomous toxicity I

Chairpersons: Mario Salmona (Italy), Angelo Poletti (Italy)

10:00-10:30 **Jochen Herms** (Germany)

"Kinetics of A β -plaque formation and spine loss in Alzheimer's disease: Long-term in vivo two-photon imaging studies in transgenic mouse models"

10:30-11:00 **Marta Valenza** (Italy)

"Emerging roles for cholesterol in Huntington's disease"

11:00-11:30 *Coffee break*

Session IV: Non cell autonomous toxicity II

Chairpersons: Mario Salmona (Italy), Angelo Poletti (Italy)

11:30-12:00 **Antonio Musaro'** (Italy)

"The contribution of skeletal muscle to the pathogenesis of ALS"

12:00-12:30 **Maria Pennuto** (Italy)

"Phosphorylation of polyglutamine androgen receptor in skeletal muscle attenuates disease manifestations in SBMA mice"

12:30-13:30 *Lunch*

Saturday, May 14, 2011 (Afternoon)

13:30-15:00 *Poster Session A (P-A.01 / P-A.48)*

Plenary Lecture

15:00-16:00 **Andrea Ballabio** (Italy)

"Modulating cellular clearance in neurodegenerative diseases"
Introduced by Larry Wrabetz (Italy-USA)

Session V: Organelles alterations in neurodegeneration I

Chairpersons: Marian di Figlia (USA)

16:00-16:30 **J. Paul Taylor** (USA)

"VCP: the Rosetta Stone of Age-Related Degeneration"

16:30-17:00 **Maria Teresa Carri'** (Italy)

"Mitochondrial dysfunction in ALS"

17:00-17:30 *Coffee break*

Session VI: Organelles alterations in neurodegeneration II

Chairpersons: Franco Taroni (Italy), Valerio Magnaghi (Italy)

17:30-18:00 **Marian Di Figlia** (USA)

"Early dysfunction of recycling endosomes in Huntington's disease"

18:00-18:30 **Larry Wrabetz** (Italy)

"Translational homeostasis as a therapeutic target in ER stress diseases"

18:30-19:00 **Cecilia Bucci** (Italy)

"Molecular basis of Charcot-Marie-Tooth type 2B disease"

19:00-19:20 **Late Breaking News**

"Mutations in TTC19 cause neurodegeneration in humans and flies associated with impaired assembly and activity of mitochondrial respiratory chain complex III"

Ghezzi D, Arzuffi P, Diodato D, Lamperti C, Zordan M, Costa R, D'Adamo P, Zeviani M (Italy)

20:30 *Social Dinner*

Sunday, May 15, 2011 (morning)

08:30-09:30 *Selected Oral Presentations*

Chairpersons: Maria Teresa Carri' (Italy), Maria Pennuto (Italy)

OC-13 *"Function of the zinc finger protein ZPR1 in neurodegeneration associated with spinal muscular atrophy"*

Ahmad S, Wang Y, and Gangwani L (USA)

OC-14 *"PTEN signalling in motor neuron disease (ALS & SMA)"*

Ning K, Drepper C, Valori CV, Wyles M, Ismail A, Higginbottom A, Herrmann T, Pamela Shaw PJ, Michael Sendtner M and, Azzouz A (UK)

- OC-15 "Oxidatively modified wtSOD1 acquires mutSOD1 like toxic properties and induces mitochondrial degeneration in sporadic patients lymphoblasts"
Guareschi S, Cova E, Cereda C, Ceroni M, Bosco D, Trotti D, Pasinelli P (USA)
- OC-16 "TDP-43 post-transcriptionally regulates VEGF and PGRN genes by binding to their 3'UTR sequences"
Colombrita C, Onesto E, Buratti E, Pizzuti A, Silani V, Baralle FE, and Ratti A (Italy)

Session VII: Epigenetics and gene regulation diseases (Role of microRNAs in the neuronal functions and dysfunctions) I

Chairpersons: Paola Riva (Italy), Antonia Ratti (Italy)

- 09:30-10:00 **Albert La Spada** (USA)
"CTCF regulates ataxin-7 expression through promotion of a convergently transcribed, antisense non-coding RNA"
- 10:00-10:30 **Noel Buckley** (UK)
"Coordinate regulation of the transcriptome and epigenome by REST"
- 10:30-11:00 coffee break

Session VIII: Epigenetics and gene regulation diseases (Role of microRNAs in the neuronal functions and dysfunctions) II

Chairpersons: Paola Riva (Italy), Antonia Ratti (Italy)

- 11:00-11:30 **Peng Jin** (USA)
"Noncoding RNAs in Neurodegenerative Disorders"
- 11:30-12:00 **Ernest Fraenkel** (USA)
"Integrating multiple 'omic strategies to identify early changes in Huntington's disease"
- 12:00-12:30 **Udai Bhan Pandey** (USA)
"Genetic interaction between FUS and TDP-43 influences the neurodegenerative phenotype in a drosophila model of ALS"

12:30-13:30 Lunch

Sunday, May 15, 2011 (afternoon)

13:30-15:00 Poster Session B (P-B.01 / P-B.47)

Session VI: Pharmacological perspectives in neurodegenerative diseases
Chairpersons: Elena Cattaneo (Italy), Neil Aronin (USA)

- 15:00-15:30 **Brent Stockwell** (USA)
"Probing Cell Death with Small Molecules"
- 15:30-16:00 **Andrea Caricasole** (Italy)
"Knocking-down the druggable genome for HD target discovery"
- 16:00-16:30 **Paolo Paganetti** (Switzerland)
"Small molecule inhibitors of beta-amyloid oligomer toxicity"
- 16:00-17:00 **Bernhard Landwehrmeyer** (Germany)
"Clinical trials in HD: an update and a perspective"
- 17:20 Closing Remarks

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POSTERS

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POSTER SESSION - A

Saturday, May 14, 2011 (Afternoon)
13:30-15:00 Poster Session A

P-A.1 / GENE EXPRESSION CHANGES IN THE BLOOD OF THE R6/2 MOUSE MODEL OF HUNTINGTON'S DISEASE

Diamanti Daniela, Tarditi Alessia, Magnoni Letizia, Mori Elisa, Malusa Federico, Fondelli Costanza, Incarnato Danny, Giuseppe Pollio and Caricasole Andrea (Italy)

P-A.2 / A C. ELEGANS MODEL OF SPINAL MUSCULAR ATROPHY NEURODEGENERATION

Gallotta I, Bazzicalupo P, and Di Schiavi E (Italy)

P-A.3 / ATXN8 -62 G/A PROMOTER POLYMORPHISM AND RISK OF TAIWANESE PARKINSON'S DISEASE

Guey-Jen Lee-Chen*, Siou-Jia Yang, I-Cheng Chen, Yih-Ru Wu, and Chiung-Mei Chen (Taiwan)

P-A.4 / THE NEED FOR HUNTINGTIN

Alan Tartakoff* and Purnima Jaiswal (USA)

P-A.5 / ATP13A2 VARIABILITY IN TAIWANESE PARKINSON'S DISEASE

Chih-Hsin Lin*, Yih-Ru Wu, Chiung-Mei Chen, Hsueh-Fen Juan, and Guey-Jen Lee-Chen (Taiwan)

P-A.6 / THE ROLE OF RNA BINDING IN TDP-43 MEDIATED DISEASE

Freibaum BD*, Taylor JP (USA)

P-A.7 / CLINICAL HETEROGENEITY IN ITALIAN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

Piacentini S*, Piaceri I, Nacmias B, Bagnoli S, Tedde A, Del Mastio M, Paganini M, Latorraca S, Sorbi S. (Italy)

P-A.8 / AMYLOID BETA ACCUMULATION IN THE MCGILL-R-THY1-APP RAT MODEL OF ALZHEIMER'S DISEASE

Canneva F, Melis T, Leon WC, Dobner J, Ducatenzeiler A, Schilling S, vonHürsten S and Cuellar AC (Canada, Germany)

P-A.9 / NEUREGULIN 1 TYPE III CAN RESTORE MYELIN THICKNESS IN POQ215X CONGENITAL HYPOMYELINATION MICE

Ornaghi F, Saccucci S, Schwab M, Nave KA, Feltri ML, Wrabetz L (Italy)

P-A.10 / F1 AND F2 GENERATIONS OF TRANSGENIC MINIATURE PIG EXPRESSING THE N-TERMINAL TRUNCATED MUTANT HUMAN HUNTINGTIN

Baxa M (1*), Hruska-Plochan M, Juhas S, Vodicka P, Pavlok A, Juhasova J, Solc P, Klima J, Marsala S, Miyano-hara A, Weiss A, Vrtel R, Kubickova S, Musilova P, Cattaneo E, DiFiglia M, Marsala M and Motlik J (Czech Republic)

P-A.11 / CEREBELLAR ABNORMALITY AND PROGRESSIVE NEUROLOGICAL PATHOLOGY IN MICE CONDITIONAL MUTANT FOR THE SOX2 TRANSCRIPTION FACTOR GENE

Baseggio E, Scherini S, Verzeroli C, Ferri A Favaro R, De Biasi S and Nicolis SK (Italy)

P-A.12 / GABA RECEPTORS IN SCHWANN-LIKE ADULT STEM CELLS: A POSSIBLE TOOL TO IMPROVE NERVE REGENERATION?

Faroni A, Terenghi G, Magnaghi V (UK)

P-A.13 / GENERATION OF A SERIE OF KNOCK-IN MOUSE NS (NEURAL STEM) LINES AS A TOOL FOR THE SCREENING OF MOLECULES USEFUL IN HD

Camnasio S *, Conforti P, Mutti C, MacDonald M and Cattaneo E (Italy)

P-A.14 / THE NEUROACTIVE STEROID ALLOPREGNANOLONE UP-REGULATES EAAC1-MEDIATED GLUTAMATE UPTAKE IN SCHWANN CELLS: A NEUROPROTECTIVE MECHANISM?

Perego C*, Di Cairano ES, Fino E, Ballabio M, Magnaghi V (Italy)

P-A.15 / ZEBRAFISH AS A POWERFUL TOOL TO STUDY HUNTINGTIN'S FUNCTION AND DOMAINS

Gaudenzi G*, Zuccato C*, Strippoli L, Pistocchi A, Bassi I, Molinari E, Giunta M, Cotelli F, Cattaneo E (Italy).

P-A.16 / HD MOUSE-IPS RECAPITULATES SOME FEATURES OF HUNTINGTON'S DISEASE

Castiglioni V., Onorati M., Rochon C. and Cattaneo E. (Italy).

P-A.17 / LT-HESNS CELLS FOR HUMAN DEVELOPMENTAL STUDIES AND DISEASE MODELING: GENERATION, CHARACTERIZATION, AND DIFFERENTIATION TO DARPP32+ NEURONS

Andrea Faedo, Stefano Camnasio, Alessia Delli Carri, Valentina Castiglioni, Marco Onorati, Stefano Di Donato, Caterina Mariotti, Outi Hovatta, Marisa Jaconi, Anis Feki, Elena Cattaneo (Italy).

P-A.18 / EXTRINSIC SIGNALS DRIVE HUMAN EMBRYONIC STEM CELLS DIFFERENTIATION TOWARDS FULLY FUNCTIONAL STRIATAL DARPP32+ NEURONS

Delli Carri A, Onorati M, Castiglioni V, Faedo A, Spaiardi P, Biella G, and Cattaneo E (Italy).

P-A.19 / EZ LAYER: A SELF-RENEWING AND LONG-TERM EXPANDABLE HUMAN STEM CELL SYSTEM WITH WIDE MULTILINEAGE NEURO POTENCY

Onorati M, Gravati M, Biella G, Svendsen C, Cattaneo E (Italy).

P-A.20 / NF-KB/RELA SPECIFIC ACETYLATION AND NEURONAL INJURY DURING ISCHEMIA: A POSSIBLE LINK BETWEEN RELA ACTIVATION AND IRON TRANSPORT IN ISCHEMIA-ASSOCIATED NEURODEGENERATION.

Rosaria Ingrassia, Ilenia Sarnico¹, Anna Lanzillotta, Laura Borgese, Laura Depero, Pier Franco Spano and Marina Pizzi (Italy).

P-A.21 / THE POLYGLUTAMINE-EXPANDED MUTANT HUNTINGTIN SELECTIVELY ACTIVATES THE AMP-ACTIVATED PROTEIN KINASE IN STRIATAL NEURONS OF HUNTINGTON'S DISEASE

Ju T-C and Chern Y (Taiwan)

P-A.22 / DJ-1 KNOCK-DOWN IMPAIRS ASTROCYTES MITOCHONDRIAL FUNCTION

Giulia Ambrosi*, Niccole J. Larsen, Steven J. Mullett, Sarah B. Berman, David A. Hinkle (Italy)

P-A.23 / 5-S-CYSTEINYLDOPAMINE AFFECTS THE EXPRESSION OF ALPHA-SYNUCLEIN AND OF THE THIOL OXIDOREDUCTASE ERp57

Mosca L*, Aureli C, Masci A, Francioso A, Faraldi M, Gaetani S, Ferraro A, D'Erme M, Cassano T (Italy).

P-A.24 / PANK2 SILENCING CAUSES GENERAL CELL GROWTH REDUCTION AND A CELL SPECIFIC FERROPORTIN UPREGULATION.

Dario Finazzi*, Maura.Poli, Sara Luscieti, Maura Derosas, Roberto Dominici, Alessandro Campanella, Paolo Arosio (Italy).

P-A.25 / A NEW INTEGRATED BIOCHEMICAL APPROACH SHOWS A DECREASE OF OXIDATIVE STRESS IN IDEBENONE-TREATED FRDA PATIENTS AFTER ORAL TOCOTRIENOL SUPPLEMENTATION

Marini M, Pini A, Abruzzo PM, Malisardi G, Marchionni C, Modesti A, Gamberi T, Morganti E, Ferreri C, Vertuani S, Fortuna F, Ghezzi A, Manfredini S (Italy).

P-A.26 / INSULIN AND IGF-1 PRECLUDE OXIDATIVE STRESS IN HUNTINGTON'S DISEASE KNOCK-IN STRIATAL CELLS

Rosenstock TR *, Ribeiro M, Oliveira CR, Rego AC (Portugal)

P-A.27 / DEVELOPING CENTRAL NERVOUS SYSTEM AND VULNERABILITY TO PLATINUM COMPOUNDS: MOLECULAR MARKERS OF CELL PROLIFERATION AND DEATH, AND CELL DIFFERENTIATION

*Piccolini VM, *Dal Bo V, De Pascali S, Migoni D, Fanizzi PF, Barni S, Bottone MG, Fenoglio C (3, 4), Bernocchi G (Italy).

P-A.28 / PARP ACTIVATION MODULATES APOPTOSIS EVENTS IN CELLULAR AND ANIMAL MODELS OF ALZHEIMER DISEASE

Mosca L, Martire S, Fuso A, Beyaie D, Masci A, Rotili D, De Leo A, Consalvi V, Giordano C, Scarpa S, d'Erme M (Italy).

P-A.29 / LYSOZYME INTERACTION WITH NEGATIVELY CHARGED LIPOSOMES RESULTS IN PROTEIN AGGREGATION AND VESICLE FUSION

Russo E*, Al Kayal T, Nappini S, Berti D, Bucciantini M, Berti D, Stefani M, Baglioni P (Italy).

P-A.30 / AXON PATHOLOGY IN MOUSE MODELS OF HUNTINGTON'S DISEASE

Martina Marangoni, Robert Adalbert, Lucie Janeckova, ^Michael Coleman and ^Laura Conforti (UK)
^equal contribution

P-A.31 / A NH2 TAU FRAGMENT TARGETS AND IMPAIRS NEURONAL MITOCHONDRIA AT AD SYNAPSES

G. Amadoro*, V. Corsetti*, F.Florenzano, R.Bussani, D. Mercanti, P. Calissano (Italy).

* These authors equally contribute to this work

P-A.32 / PROTEIN AGGREGATION IS PREDICTIVE OF A FASTER DISEASE PROGRESSION IN MOUSE MODELS OF AMYOTROPHIC LATERAL SCLEROSIS.

Marino M*, Papa S, Peviani M, Cheroni C, Ferraiuolo L, Shaw P, Fontana E, DeBiasi S, Bendotti C (Italy).

P-A.33 / CURCUMIN AND CEFTRIAXONE: TWO DIFFERENT MOLECULES INDUCING COMMON NEUROPROTECTIVE EFFECTS IN AN IN VITRO MODEL OF ALEXANDER DISEASE

Bachetti T*, Di Zanni E, Balbi P, Sechi GP, Ceccherini I (Italy).

P-A.34 / TRANSCRIPTIONAL CONTROL OF THE SMALL HEAT SHOCK PROTEIN B8 (HSPB8) IN MOTORNEURONS

Crippa V*, Boncoraglio A, Rusmini P, Sau D, Giorgetti E, and Poletti A (Italy)

P-A.35 / INTRASTRIATAL VIRAL VECTOR DELIVERY OF MUTANT HUNTINGTIN AS RAT MODEL OF HUNTINGTON'S DISEASE.

Ceccarelli I, Scali C, Rossini L, Pollio G, Biotti I, Cappelli A, Miragliotta V, Fiengo P, Caricasole A and Gaviraghi G. (Italy).

P-A.36 / PERIPHERAL GANGLION NEURONS FROM HUMAN MUTANT P301S TAU TRANSGENIC MICE TO INVESTIGATE TAU PATHOLOGY

Mellone M*, Andrews MR, Dassie E, Richardson JC, Tolkovsky AM, Goedert M, and Spillantini MG (UK)

P-A.37 / HUMAN STEM CELL-DERIVED NEURONS AS A MODEL TO STUDY TAU FUNCTION AND DYSFUNCTION

Mariangela Iovino, Rickie Patani, Siddharthan Chandran, and Maria Grazia Spillantini (UK)

P-A.38 / FORMATION AND DEGRADATION OF ER MEMBRANE-DERIVED CYTOPLASMIC INCLUSIONS GENERATED BY THE AMYOTROPHIC LATERAL SCLEROSIS-ASSOCIATED MUTANT FORM OF VAPB

Papiani G *, Ruggiano A, Fossati M, Fasana E, Francolini M, Borgese N, and Navone F (Italy).

P-A.39 / A NOVEL MAPT MUTATION ASSOCIATED TO LATE-ONSET SPINAL MUSCULAR ATROPHY WITHOUT DEMENTIA IN A LARGE ITALIAN FAMILY

Ronchi D (1,3,*), Di Fonzo A, Gallia F, Fassone E, Donadoni C, Salani S, Bordoni A, Rizzuti M, Del Bo R, Corti S, Sacilotto G, Nobile-Orazio E, and Comi GP (Italy).

P-A.40 / PRP90-231 INTRACELLULAR AGGREGATION INDUCES CELL DEATH THROUGH LYSOSOMAL LOSS OF IMPERMEABILITY

Thellung S *, Corsaro A, Villa V, Vella S, Pagano A and Florio T (Italy).

P-A.41 / STRUCTURAL BASIS FOR CHAPERONIN-MEDIATED INHIBITION OF PROTEIN AGGREGATION IN HUNTINGTON'S DISEASE REVEALED BY CRYO-ELECTRON TOMOGRAPHY

Shahmoradian SH*, Galaz JG*, Cong Y, Chen BY, Schmid MF, Frydman J, Ludtke SJ, Chiu W (USA).

*Both authors contributed equally to this work.

P-A.42 / MITOCHONDRIAL FUNCTION OF JNK ISOFORMS DURING STATUS EPILEPTICUS-INDUCED NEURONAL DAMAGE

Spigolon G, Zhao Y, Bonny C, Herdegen T, Vercelli A (Italy).

P-A.43 / MUSCLE CELLS AND MOTONEURONS DIFFERENTIALLY REMOVE MUTANT SOD1 IN TWO CELL MODELS OF AMYOTROPHIC LATERAL SCLEROSIS.

Onesto E (*), Rusmini P, Crippa V, Ferri N, Zito A, Galbiati M, Poletti A (Italy).

P-A.44 / PARKIN ABSENCE IMPACTS MICROTUBULE STABILITY AND AXONAL TRANSPORT IN KNOCKOUT MICE.

Cartelli D, Sassone J, Amadeo A, Ciammola A, And Cappelletti G (*) (Italy).

P-A.45 / A MAJOR ROLE FOR SIDE-CHAIN POLYGLUTAMINE HYDROGEN BONDING IN IRREVERSIBLE ATAXIN-3 AGGREGATION.

Natalello A, Frana AM, Relini A, Apicella A, Invernizzi G, Casari C, Gliozzi A, Doglia SM, Tortora P* , Regonesi ME (Italy).

P-A.46 / THE ROLE OF THE MITOCHONDRIAL m-AAA PROTEASE COMPLEX IN THE PATHOGENESIS OF HEREDITARY SPINOCEREBELLAR DEGENERATIONS

Di Bella D*, Fracasso V, Magri S, Lazzaro F, Plumari M, Mariotti C, Gellera C, Muzi-Falconi M, Taroni F (Italy)

P-A.47 / DROSOPHILA MODEL OF TDP-43 NEUROPATHY

Rebecca B. Smith*, J. Paul Taylor (USA)

P-A.48 / CHOLESTEROL LOADED NANOPARTICLES INJECTED SYSTEMICALLY REACH THE BRAIN AND LOCALIZE INTO SPECIFIC CELL TYPES

Valenza M*, Tosi G, Cepeda C, Bondioli L, Brilli E, Ruozi B, Joshi PR, Chen JY, Singh S, Vandelli MA, Levine MS, Cattaneo E (Italy)

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POSTER SESSION - B

Sunday, May 15, 2011 (Afternoon)
13:30-15:00 Poster Session B

- P-B.1 / A PROLONGED PHARMACOLOGICAL BLOCKADE OF TYPE-5 METABOTROPIC GLUTAMATE RECEPTORS PROTECTS CULTURED SPINAL CORD MOTOR NEURONS AGAINST EXCITOTOXIC DEATH**
Simona D'Antoni, Antonio Berretta, Giovanna Seminara, Patrizia Longone, Anna Maria Giuffrida-Stella, Giuseppe Battaglia, Maria A. Sortino, Ferdinando Nicoletti, Maria V. Catania* (Italy).
- P-B.2 / A NOVEL APPROACH TO THE STUDY OF DEGENERATION IN THE NEUROMUSCULAR JUNCTION (NMJ)**
Simone D., Colosimo A (Italy)
- P-B.3 / SCA14 MUTATIONS AFFECT THE INTRA-MOLECULAR CLAMP BETWEEN THE C1 DOMAIN AND CARBOXYL-TERMINUS OF PKC-GAMMA**
Jeziarska J *, Goedhart J, Westerink JT, Reits EA and Verbeek DS (The Netherlands).
- P-B.4 / INTRINSIC BRAIN INFLAMMATION AND ENHANCED SUSCEPTIBILITY TO KAINATE-INDUCED SEIZURES IN A TRANSGENIC MOUSE MODEL OF INHERITED CREUTZFELDT-JAKOB DISEASE**
Bertani I, Maroso M, Mantovani S, Iori V, Vezzani A, Chiesa R (Italy).
- P-B.5 / ALTERATION OF SECRETORY PROTEIN TRAFFICKING AS POSSIBLE PATHOGENETIC MECHANISM IN INHERITED PRION DISEASES**
Restelli E *, Mantovani S, and Chiesa R (Italy).
- P-B.6 / GENERATION OF A TRANSGENIC MOUSE MODEL OF FATAL FAMILIAL INSOMNIA**
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- P-B.7 / CELLULAR MODELS TO INVESTIGATE THE PATHOGENIC MECHANISMS OF CEREBRAL AMYLOIDOSIS ASSOCIATED WITH NONSENSE MUTATIONS OF THE PRION PROTEIN**
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- P-B.8 / ZINC MODULATES COPPER COORDINATION MODE IN PRION PROTEIN OCTA-REPEAT SUBDOMAINS**
Francesco Stellato, Ann Spevacek, Olivier Proux, Velia Minicozzi, Glenn Millhauser, Silvia Morante (Italy).
- P-B.9 / PERIPHERAL MARKERS OF NEUROINFLAMMATION IN PARKINSON'S DISEASE**
Bazzini E *, Armentero MT, Cerri S, Levandis G, Ghezzi C, Pacchetti C, Cristina STassorelli C, Zangaglia R, Blandini F (Italy).
- P-B.10 / NEW INSIGHTS INTO THE ROLE OF HSP90 IN HUNTINGTON'S DISEASE: INFLUENCE ON PROTEIN CLEARANCE AND STABILITY**
Barbara Baldo*, Andreas Weiss, Christian Parker, Jens Richter, Gabi Schutzius, Paolo Paganetti and Klemens Kaupmann (Switzerland).

- P-B.11 / ALTERED AXONUCLEAR COMMUNICATION IN MOTONEURONS OF RODENT MODELS OF FAMILIAL AMYOTROPHIC LATERALE SCLEROSIS:ROLE OF IMPORTIN BETA AND VIMENTIN.**
Peviani M, Spano G and Bendotti C (Italy).
- P-B.12 / ASSESSMENT OF CHAPERONE-MEDIATED AUTOPHAGY IN CIRCULATING LYMPHOMONOCYTES OF PATIENTS WITH PARKINSON'S DISEASE**
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- P-B.13 / VASCULAR INFLAMMATION AND LEUKOCYTE TRAFFICKING IN AN EXPERIMENTAL MODEL OF ALZHEIMER'S DISEASE**
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- P-B.14 / HUMAN INDUCED PLURIPOTENT STEM CELLS FROM PARKINSON'S DISEASE AFFECTED PATIENTS**
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- P-B.15 / ACETYLATION OF NF-KAPPA B P50/RELA IN THE EPIGENETIC REGULATION OF TRANSCRIPTION: A NEUROPROTECTIVE TARGET IN BRAIN ISCHEMIA**
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- P-B.16 / TARGETING NF-KAPPAB AND EPIGENETIC CHROMATIN REMODELLING IN THE THERAPY OF POST-ISCHEMIC BRAIN INJURY**
C. Branca *, A. Lanzillotta, I. Sarnico, M. Benarese, G. Pignataro, O. Cuomo, L. Annunziato, G. Faraco, F. Blasi, A. Chiarugi, PF. Spano, M. Pizzi (Italy).
- P-B.17 / LYSOSOMAL GLYCOHYDROLASES ARE ASSOCIATED TO LIPID RAFTS OF MOUSE BRAIN: POTENTIAL IMPLICATION FOR NEURODEGENERATION**
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- P-B.18 / NOVEL DYSFUNCTIONS AT THE ORIGIN OF HYPOMYELINATION, A FEATURE OF NIEMANN PICK DISEASE TYPE A**
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- P-B.19 / ATAXIN-2 DEFICIENCY CAUSES CHANGES IN THE EXPRESSION OF GRB2 AND SRC**
Drost J*, Nonis D, Damrath E, Auburger G (Germany).
- P-B.20 / NEUROPROTECTIVE EFFECT OF NOVEL HDAC INHIBITORS ON SCA3 CELLS**
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- P-B.21 / EXPRESSION OF PHOX2B TARGET GENES IN THE CONGENITAL CENTRAL HYPOVENTILATION SYNDROME (CCHS)**

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P-B.22 / THE C-TERMINUS OF P/Q TYPE VOLTAGE GATED CALCIUM CHANNEL (CAV2.1) ALPHA1A SUBUNIT MODULATES THE TRANSCRIPTION OF OTHER NEURONAL GENES.

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P-B.23 / NEURONAL LSD1, AN EPIGENETIC ENZYME DEVOTED TO NEURONS

Paganini L, Toffolo E, Zibetti C, Rusconi F, Romanelli L, Tedeschi G, Sala C, and Battaglioli E (Italy).

P-B.24 / AN IMMUNOHISTOCHEMICAL ANALYSIS OF HUNTINGTIN AND REST EXPRESSION IN HUMAN CONTROL AND HD BRAIN

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P-B.25 / miR-103 AND miR-107 ARE INVOLVED IN CDK5R1 EXPRESSION MODULATION: POSSIBLE IMPLICATIONS IN ALZHEIMER'S DISEASE

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P-B.26 / ERK MODULATION IN FIBROBLASTS FROM ALZHEIMER'S DISEASE PATIENTS: INVOLVEMENT IN APP METABOLISM AND EAAT1 PROCESSING

Zoia CP, Compagnoni P, Cereda D, Isella V, Tremolizzo L and Ferrarese C (Italy).

P-B.27 / DOWNREGULATION OF THE CHROMATIN-REMODELLING FACTOR BRAHMA IN SOD1 G93A CELLS MODIFIES TRANSCRIPTION AND ALTERNATIVE SPLICING OF NRG1 AND P15RS, TWO GENES INVOLVED IN AXON GROWTH AND GUIDANCE

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P-B.28 / PROTEIN KINASE A ATTENUATES THE TOXICITY OF POLYGLUTAMINE ANDROGEN RECEPTOR

Scaramuzzino C, Parodi S, Sambataro F, Contestabile A, Gozes I, Pennuto M (Italy).

P-B.29 / NEUROPROTECTIVE POTENTIAL OF ADENOSINE A2A AND CANNABINOID CB1 RECEPTOR ANTAGONISTS IN AN ANIMAL MODEL OF PARKINSON'S DISEASE

Cerri S*, Levandis G, Ambrosi G, Mfller CE, Blandini F, and Armentero MT (Italy).

P-B.30 / STUDY OF PI3/AKT PATHWAY IN A HUMAN NEUROBLASTOMA CELL LINE AFTER PHARMACOLOGICAL TREATMENTS

Ghezzi C*, Oda A, Blandini F, Armentero MT (Italy).

P-B.31 / PLASMA CYSTATIN C AND RISK OF DEVELOPING ALZHEIMER'S DISEASE IN SUBJECTS WITH MILD COGNITIVE IMPAIRMENT

Stoppani E *, Benussi L, Paterlini A, Glionna M, Albertini V, Levy E, Emanuele E, Binetti G, Ghidoni R (Italy).

P-B.32 / DETERMINATION OF BETA-AMYLOID PEPTIDE SIGNATURES IN CEREBROSPINAL FLUID OF FAMILIAL ALZHEIMER DISEASE MUTATED PATIENTS

Albertini V *, Stoppani E, Paterlini A, Benussi L, Squitti R, Bruno A, Bernardini S, Cassetta E, Rossini PM, Squitieri F, Binetti G, Ghidoni R (Italy).

P-B.33 / FISHING FOR A MECHANISM: ZEBRAFISH AS A MODEL FOR THE STUDY OF MITOFUSIN 2 MUTATIONS CAUSING CHARCOT-MARIE-TOOTH TYPE 2A

Bergamin G*, Vettori A, Moro E, Vazza G, Polo G, Tiso N, Argenton F, Mostacciolo ML (Italy)

P-B.34 / TFPI IS A USEFUL BIOMARKER LINKING ENDOTHELIAL PERTURBATION TO NEURODEGENERATION AND ANTI-Abeta42 ANTIBODIES IN AD PATIENTS

Fabrizio Piazza*, Daniela Galimberti, Gloria Galimberti, Elisa Conti, Simona Andreoni, Elio Scarpini, Guido Cavaletti and Carlo Ferrarese (Italy).

P-B.35 / NEUROPROTECTION IN SPINAL MUSCULAR ATROPHY (SMA) USING NEURAL STEM CELLS AS A THERAPEUTIC APPROACH

Falcone M, Rizzo F, Nizzardo M, Simone C, Ronchi D, Donadoni C, Salani S, Ulzi G, Riboldi G, Bresolin N and Corti S (Italy).

P-B.36 / IDENTIFICATION OF SMALL MOLECULES INHIBITORS OF THE REST/NRSF COMPLEX AS A POSSIBLE THERAPY FOR HUNTINGTON'S DISEASE

Ieraci A, Conforti P, Mutti C, Stefano Camnasio S, Zuccato C, Contini A and Cattaneo E (Italy).

P-B.37 / INCREASED CYTOPLASMIC MISLOCALIZATION OF TAR DNA BINDING PROTEIN 43 (TDP-43) IN CIRCULATING LYMPHOMONOCYTES OF ALS PATIENTS RECAPITULATES THE MAJOR DYSFUNCTION FEATURING MOTOR NEURONS IN THE DISEASE

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P-B.38 / PC12 CELL GROWN ON NANOSTRUCTURED TITANIUM OXIDE

E. Maffioli*, S.Nonnis, S.Forti, N.Cuevas Polo, A. Negri, L.Cannizzaro, C.Lenardi, P.Milani, G.Tedeschi (Italy).

P-B.39 / ENDOGENOUS IMMUNE RESPONSE AGAINST ABETA PEPTIDE: INFLUENCE OF ACHETYL-CHOLINESTERASE INHIBITORS

Conti E, Tironi M, Galimberti G, Zoia CP, Tremolizzo L and Ferrarese C (Italy).

P-B.40 / EX VIVO VISUALIZATION OF TRANSFECTED HUMAN MESENCHYMAL STEM CELLS AFTER TRANSPLANTATION: A RELIABLE CELL-LABELING PROTOCOL

Diana V. *, Cova L., Libani I.V., Ottobriani L., Armentero M-T., Blandini F., Lucignani G., Silani V (Italy).

P-B.41 / ASTROCYTES INFLUENCE CHOLESTEROL-DEPENDENT NEURONAL PROPERTIES IN HUNTINGTON'S DISEASE

Marullo M*, Valenza M, Cattaneo E (Italy).

P-B.42 / REGULATION OF SOD1 GENE EXPRESSION IN A CELLULAR MODEL OF NEURODEGENERATION.

Milani P, Amadio M, Gagliardi S, Sardone V, Ghiroldi A*, Cova E, Pascale A, Cereda C (Italy).

P-B.43 / KETOGENIC DIET ACCELERATES THE ONSET OF DISEASE IN A PLA2G6 KO MOUSE MODEL.

D. Brunetti, S. Dusi, F. Moda, A. Uggetti, C. Lamperti, Y. Tsujimoto, M. Morbin, V. Tiranti (Italy)

P-B.44 / MICROTUBULE STABILITY IS PRECOCIOUSLY AFFECTED IN A MOUSE MODEL OF PARKINSON'S DISEASE.

Cartelli D(*), Casagrande F, Battaglia G, Busceti C, Molinaro G, Cappelletti G (Italy)

P-B.45 / A NOVEL MUTATION IN SACS GENE ASSOCIATED TO A COMPLICATED FORM OF HEREDITARY SPASTIC PARAPLEGIA

Gregianin E*, Pegoraro E, Mostacciuolo ML, Vazza G (Italy).

P-B.46 / TDP-43 SUB-CELLULAR LOCALIZATION AND KINETICS OF TRANSPORT IN CULTURED CORTICAL NEURONS

Alami NH*, Taylor JP(USA)

P-B.47 / LIVING CELLS AS DRUG DELIVERY VEHICLES FOR TREATMENT OF NEURODEGENERATIVE DISORDERS

Batrakova E*, Haney MJ, Zhao Y, Suresh P, Vetro JA, Klyachko NL, Mosley RL, Kabanov AV, Gendelman HE (USA)