

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

Aula Magna, Università degli Studi di Milano
Via Festa del Perdono 7 - 20122 Milano (Italy)
May 8-10, 2009

FINAL PROGRAM

Friday, May 08, 2009 (Morning)

09:30 Registration

10:00 Satellite symposium on:
**Molecular Mechanisms on Neurodevelopment
Axonal development and targeting**
(no registration required for this symposium)
Chairpersons: Roberto Maggi (Italy), Ivan De Curtis (Italy)

Plenary Lecture

10:15-11:00 **Patricia Salinas (UK)**
"Wnt signaling in the regulation of synapse formation and maintenance"

Session

11:00-11:30 **Ivan De Curtis (Italy)**
"Essential role of Rac GTPases in neuronal development"

11:30-12:00 **Flavia Valtorta (Italy)**
"Membrane trafficking in axonal growth cones"

12:00-12:30 **Giorgio Merlo (Italy)**
"Transcription factor regulations of axon targeting: the olfactory system"

Closing remarks

Friday, May 08, 2009 (Afternoon)

Opening Session

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

14:10-14:30 **Gianpiero Sironi** (Prorettore alla ricerca, Università di Milano)
Stefano Di Donato (Direttore Scientifico Emerito,
Fondazione Istituto Neurologico
"Carlo Besta" Milano)

Opening lecture

14:30-15:30 **Aaron Ciechanover (Israel)**
The Nobel Prize in Chemistry 2004
"The Ubiquitin Proteolytic System: From Basic Mechanisms via
Human Diseases and onto Drug Targeting"
Introduced by Ron R. Kopito (USA)

Session I: Cellular handling of aberrant neurotoxic proteins I

Chairpersons: Marcy McDonald (USA), Caterina Bendotti (Italy)

15:30-16:00 **Ron Kopito (USA)**

"Membrane penetration and prion-like propagation of amyloids"

16:00-16:30 **Ana Maria Cuervo (USA)**

"Degradation of misfolded proteins through the autophagic system in the central nervous system"

16:30-17:00 **Francesco Fornai (Italy)**

"The role of autophagy in selective neuronal degeneration: basic evidence and pharmacological manipulation"

17:00-17:30 *Coffee break*

Session II: Cellular handling of aberrant neurotoxic proteins II

Chairpersons: Piera Pasinelli, (USA), Mario Salmona (Italy)

17:30-18:00 **Christopher E. Shaw (UK)**

"TDP-43 mutations in familial and Sporadic Amyotrophic Lateral Sclerosis "

18:00-18. 30 **J. Paul Taylor (USA)**

"Insights into the pathogenesis of TDP-43-related disease from Drosophila models"

18:30-19:00 **Erich E. Wanker (Germany)**

"Modulation of amyloid formation pathways with chemical chaperones"

19:00-19:30 **Gianluigi Forloni (Italy)**

"Protein toxicity in AD and PD "

19:30 *Welcome Party*

Saturday, May 9, 2009 (Morning)

Session III: Mitochondria, oxidative stress, and genotoxicity I

Chairpersons: Silvia De Biasi (Italy), Massimo Stefani (Italy)

09:00-09:30 **Eva-Maria Mandelkow (Germany)**

"Modes of TAU toxicity and neuronal degeneration"

09:30-10:00 **Paolo Bernardi (Italy)**

"Toward a Mitochondrial Therapy of Collagen VI Muscular Dystrophies."

10:00-10:30 **Franco Taroni (Italy)**

"Mitochondrial maintenance and spinocerebellar degeneration"

10:30-11:00 *coffee break*

Session IV: Mitochondria, oxidative stress, and genotoxicity II

Chairpersons: Armando Gennazzani (Italy), Davide Trotti (USA)

11:00-11:30 **Robert H. Brown (USA)**

"Genetic determinants of Amyotrophic Lateral Sclerosis"

11:30-12:00 **Vittorio Calabrese (Italy)**

"Nitric Oxide in the brain: neuroprotection versus neurotoxicity"

12:00-12:30 **Keith W. Caldecott (UK)**
"Chromosomal Single-strand break repair and neurological disease"

12:30-13:30 *Lunch*

Saturday, May 9, 2009 (Afternoon)

13:30-15:00 *Poster Session I (P01-P66)*

Session V: Role of microRNAs in the neuronal functions and dysfunctions
Chairpersons: Robert M. Friedlander (USA), Robert G. Korneluk (Canada)

15:00-15:30 **Asa Abeliovich (USA)**
"A MicroRNA feedback circuit in midbrain dopamine neurons"

15:30-16:00 **Carlo Cogoni (Italy)**
"MicroRNA in the brain"

16:00-17:00 *Selected Free Communication*
Chairpersons: Franco Taroni (Italy), Giorgio Casari (Italy)

OC1 - Role of miRNAs regulation on CDK5R1 expression in Alzheimer's disease

Moncini S, Zuccotti P, Venturin M, Salvi A, Sabelli C, De Petro G, Barlati S, Riva P. (Italy)

OC2 - Identifying genes that modify Huntington's disease in the N171-82Q mouse model.

Corrochano-Sanchez S, Acevedo-Arozena A, Rubinsztein DC, Brown SDM (UK)

OC3 - Modifiers of aggregation in neurodegenerative diseases

Holmberg M, van Ham TJ, Thijssen KL, van Rheenen J, van Veluw J, Hofstra RMW, Nollen EAA (The Netherland)

OC4 - Analysis of inducible mouse model of Huntington disease combined with ubiquitin-proteasome system reporter mice.

Ortega Z, Maynard CJ, Diaz-Hernandez M, Dantuma NP, Lucas JJ (Spain)

17:00-17:30 *Coffee break*

17:30-19:30 *Selected Free Communication*
Chairpersons: Franco Taroni (Italy), Giorgio Casari (Italy)

OC5 - Polyglutamine toxicity induces multiple photoreceptor degeneration pathways and complex retinal remodelling in a SCA7 mouse model

Trottier Y, Messaddeq N, Jacquard C, Weber C, Karem A, Wolfrum U, Jonet L, Jeanny JC, Yefimova MG (France)

OC6 - AFG3L2 mutations cause autosomal dominant ataxia SCA28 and reveal an essential role of the mitochondrial m-AAA AFG3L2 homocomplex in the cerebellum

DiBella D, Lazzaro F, Brusco A, Battaglia G, Pastore A, Finardi A, Fracasso V, Magri S, Plumari M, Cagnoli C, Tempia F, Brussino A, Gellera C, Mariotti C, Plevani P, DiDonato S, Langer T, Muzi-Falconi M, Taroni F (Italy)

- OC7 - Heat shock response and misfolded protein elimination associated with PHOX2B polyalanine expansion and GFAP mutations in congenital central hypoventilation syndrome and Alexander disease
Bachetti T, Di Zanni E, Bocca P, Borghini S, Prigione I, Caroli F, Filocamo M, Balbi P, Biancheri R, Ravazzolo R, Ceccherini I (Italy)
- OC8 - Molecular mechanism of the regulation of two ER stress-mediated degradation system, UPS and autophagy/lysosome, of the misfolded protein.
Kouroku Y, Fujita E, Momo YM, Momoi T (Japan)
- OC9 - Polyglutamine atrophin provokes autophagic neurodegeneration by repressing the tumour suppressor FAT
Charroux B, Occhi S, Nisoli I, Napoletano F, Chauvin JP, Calamita P, Volpi VG, Zanin V, Fanto M (Italy)
- OC10 - The nucleolus as source of oxidative damage and neurodegeneration.
Parlato R, Rieker C, Kreiner, Engblom D, Bierhoff H, Grummt I, Schutz G (Germany)
- OC11 - Aggresome-forming TTRAP mediates pro-apoptotic properties of Parkinson's disease-associated DJ-1 missense mutations
Zucchelli S, Vilotti S, Calligaris R, Lavina ZS, Biagioli M, Foti R, De Maso L, Pinto M, Gorza M, Speretta E, Casseler C, Tell G, Del Sal G, Gustincich S (Italy)
- OC12 - Role of dopamine-induced calcium signalling in neurodegeneration of control and PINK1 knockout neurons and astrocytes.
Vaarmann A, Abramov AY (UK)

20:30 Social Dinner

Sunday, May 10, 2009 (morning)

08:30-10.00 *Selected Free Communications*

Chairpersons: Angelo Poletti (Italy), Giorgio Battaglia (Italy)

- OC13 - Role of PPP4R2 in apoptosis and neuritogenesis
Bosio Y, Camera P, Berto G, Ambrogio C, Silengo L, Di Cunto F (Italy)
- OC14 - The Zing Finger protein ZPR1 deficiency causes activation of the JNK signaling cascade and neuron degeneration
Ahmad S, Gangwani L (USA)
- OC15 - Influence of post-translational modifications on the androgen receptor in SBMA
Jochum T, Ritz ME, Moss D, Cato ACB (Germany)
- OC16 - Effect of selective androgen receptor modulators in SBMA experimental models.
Bolzoni E, Orr CR, Liu Y, McDonnell D, Wilson E, Merry DE (USA).
- OC17 - Disruption of dynein/dynactin function in neurons in vivo does not lead to motor abnormalities in transgenic mice and improves lifespan in the SOD1-ALS mouse model
Teuling E, van Dis V, Wulf PS, Haasdijk ED, Akhmanova A, Hoogenraad CC, Jaarsma D (USA)
- OC18 - Spinal cord from an animal model of ALS: a potential source of adult neural stem cells in response to neurodegeneration.
Marcuzzo S, Preite L, Cappelletti C, Zingale A, Terragni B, Mantegazza M, Bernasconi P, Mantegazza R (Italy)

10:00-10:30 *coffee break*

*Session VI: Altered protein functions in neurodegenerative diseases
Chairpersons: Marian Di Figlia (USA), Neil Aronin (USA)*

10:30-11:00 **Angelo Poletti (Italy)**

"Clearance of misfolded proteins in SBMA and ALS"

11:00-11:30 **Daniela Rossi (Italy)**

"Role of astroglia on motor neuron cell survival and death in ALS"

11:30-12:00 **Elena Cattaneo (Italy)**

"BDNF, Huntingtin and neuronal cell death"

12:00-12:30 **Patrik Brundin (Sweden)**

"Can Parkinson's Disease pathology be propagated from one neuron to another?"

12:30-13:30 *Lunch*

Sunday, May 10, 2009 (afternoon)

13:30-15:00 *Poster Session II (67-133)*

*Session VI: Genetic determinants of Neurodegenerative diseases
Chairpersons: Elena Cattaneo (Italy), Stefano DiDonato (Italy)*

15:00-15:30 **Fabrizio Tagliavini (Italy)**

"Good Gene, Bad Gene: New APP Variant May Be Both"

15:30-16:00 **Piera Pasinelli (USA)**

"Alteration of mitochondrial functions in amyotrophic lateral sclerosis"

16:00-16:30 **James F. Gusella (USA)**

"Modifier genes in Huntington's disease"

16:30-17:00 **Michael R. Hayden (Canada)**

"Bad Boys and the Pathogenesis of HD"

17:00 *Closing Remarks*

*Program of the IV Meeting on the
Molecular Mechanisms of Neurodegeneration
Milan, Italy, May 8-10, 2009
web site: <http://users.unimi.it/triplet/2009/>
e-mail: triplets@unimi.it*

POSTERS

Poster Session I (P01-P66)

**Saturday, May 9, 2009
from 13:30 to 15:00**

- P01 - ALS GENES: MUTATIONAL ANALYSIS IN A LARGE COHORT OF ITALIAN PATIENTS**
Gellera C*, Ticozzi N, Ratti A, Corrado L, Castellotti B, Colombrita C, Plumari M, Carlomagno Y, Bragato C, Mazzini L, Taroni F, Silani V
- P02 - DOWN CRITICAL REGION-ENCODED PROTEIN TTC3 INHIBITS NEURONAL DIFFERENTIATION IN HIPPOCAMPAL NEURONS**
Berto G*, Camera P, Dotti CS, Di Cunto F
- P03 - AN OLD PROTEIN IN UNEXPECTED PLACES: ALPHA- AND BETA- GLOBIN EXPRESSION IN MESENCEPHALIC DOPAMINERGIC NEURONS AND GLIAL CELLS**
Biagioli M*, Pinto M*, Cesselli D, Zaninello M, Lazarevic D, Simone R, Vlachouli C, Plessy C, Bertin N, Beltrami A, Kobayashi K, Gallo V, Santoro C, Ferrer I, Rivella S, Beltrami CA, Carninci P, Raviola E, Gustincich S
- P04 - MUTATIONAL SCREENING AND FUNCTIONAL ANALYSES IN THE ZEBRAFISH MODEL OF GIGYF2 AS A CANDIDATE GENE FOR PARKINSON DISEASE**
Guella I*, Del Giacco L, Asselta R, Pistocchi A, Rimoldi V, Sironi F, Primignani P, Zini M, Pezzoli G, Duga S, Goldwurm S
- P05 - FMR1 GENE TESTING IN THE FAMILY WITH ATAXIA AND TREMOR**
Rajkiewicz M, Kaca-Orynska M, Sulek-Piatkowska A, Krysa W, Kozirowski D, Friedman A
- P06 - MODELING THE NEUROLOGICAL FEATURES OF TUBEROUS SCLEROSIS COMPLEX BY SELECTIVE TARGETING OF EMBRYONIC AND ADULT NEURAL STEM CELLS IN MICE**
Magri L, Galli R
- P07 - RELATIONSHIP BETWEEN AF4 EXPRESSION LEVELS AND THE TIME FRAME/ONSET OF NEURODEGENERATION IN THE ROBOTIC MOUSE**
Mattia J Finelli*, Emmanuelle Bitoun, Sheena Lee, Peter L Oliver, Kay E Davies
- P08 - A MODEL OF SPINOCEREBELLAR ATAXIA TYPE 2 (SCA2): THE (CAG)102-SCA2 KNOCK-IN MOUSE**
Damrath E*, Gispert S, Nonis FD, Lastres-Becker I, Nowock J, Auburger G
- P09 - MUTATION ANALYSIS OF THE SPG11 AND SPG15 GENE IN ITALIAN PATIENTS WITH SPASTIC PARAPLEGIA AND THIN CORPUS CALLOSUM**
Castellotti B*, Mariotti C, Pensato V, Fancellu R, Salsano E, Gellera C, Pareyson D, Taroni F
- P10 - ALPHA-SYNUCLEIN POINT MUTATION ANALYSIS IN ITALIAN PATIENTS WITH AUTOSOMAL DOMINANT PARKINSON DISEASE**
Francesca Sironi, Luca Trotta, Michela Zini, Silvana Tesei, Paola Primignani, Tiziana Brambilla, Domenico A Coviello, MD, Gianni Pezzoli, MD, Stefano Goldwurm
- P11 - ALTERNATIVE SPLICING ENHANCES LYSINE SPECIFIC DEMETHYLASE 1 (LSD1) EPIGENETIC TUNEABILITY IN THE MAMMALIAN NERVOUS SYSTEM.**
Zibetti C*, Mattevi A, Sala C, Battaglioli E.

- P12 - INCREASED SUSCEPTIBILITY OF CORTICAL SPREADING DEPRESSION IN ATP1A2 KNOCK-IN MOUSE MODEL OF FAMILIAL HEMIPLEGIC MIGRAINE TYPE2**
Loredana Leo, Lisa Gherardini, Virginia Barone, Tommaso Pizzorusso, Giorgio Casari
- P13 - HAPLOINSUFFICIENCY OF AFG3L2, THE GENE RESPONSIBLE FOR SPINOCEREBELLAR ATAXIA TYPE 28 (SCA28), CAUSES MITOCHONDRIAL-MEDIATED PURKINJE CELL DEGENERATION**
Maltecca F*, Magnoni R, Cox G, Quattrini A, Casari G
- P14 - ROLES OF ERYTHROPOIETIN AND SONIC HEDGEHOG IN NEUROPROTECTIVE EFFECTS OF BRAIN-DERIVED NEUROTROPHIC FACTOR AGAINST MITOCHONDRIAL DYSFUNCTION**
Ding-I Yang*, Chia-Lin Wu
- P15 - INTERPLAY BETWEEN GLUTAMINE AVAILABILITY, GLUTAMATE AND GLUTATHIONE IN A CELL MODEL FOR FAMILIAL FORMS OF AMYOTROPHIC LATERAL SCLEROSIS**
D'Alessandro G*, Calcagno E, Tartari S, Rizzardini M, Invernizzi RW, Cantoni L
- P16 - NEURAL TUBE DEFECTS**
Angeles Cavero
- P17 - NITROPROTEOMICS OF PERIPHERAL BLOOD MONONUCLEAR CELLS FROM PATIENTS AND RAT MODEL OF ALS**
Pozzi S*, Nardo G, Lauranzano E, Mantovani S, Garbelli S, Marinou K, Basso M, Mora G, Bendotti C, Bonetto V
- P18 - TOWARDS THE BIOCHEMICAL CHARACTERIZATION OF SELADIN-1/DHCR24: HETEROLOGOUS PRODUCTION OF THE HUMAN PROTEIN**
Zucchini D, Caprini G, Criaco L, Vanoni MA
- P19 - MORPHOLOGICAL CHANGES IN HUNTINGTON'S DISEASE MITOCHONDRIA SUSTAIN THEIR INCREASED SUSCEPTIBILITY TO APOPTOTIC STIMULI**
Veronica Costa*, Dmitry Lim, Ernesto Carafoli, Mauro Piacentini, Walter Malorni, Luca Scorrano
- P20 - STUDIES ON CONE CELL DEATH MECHANISMS IN THE CPFL1 MOUSE MODEL**
Trifunovic D*, Dengler K, Michalakis S, Biel M, Paquet-Durand F
- P21 - APOPTOSIS INDUCED BY PLATINUM COMPOUNDS IN B50 CELLS**
Santin G*, Bottone MG, Piccolini VM, Veneroni P, De Pascali SA, Migoni D, Marsigliante S, Fanizzi FP, Bernocchi G
- P22 - CISPLATIN ANALOGUES WITH NEW SUBCELLULAR TARGETS: A STRATEGY TO PREVENT NEUROTOXICITY INDUCED BY PLATINUM COMPOUNDS?**
Cerri S*, De Pascali SA, Migoni D, Marsigliante S, Fanizzi FP, Bernocchi G
- P23 - IN VIVO CHARACTERIZATION OF HUMAN NEURAL STEM ECLS IMMORTALIZED WITH v-MYC (IhNSC) BY TRANSPLANTATION INTO ISCHEMIC RAT BRAIN**
Lidia De Filippis, Laura Rota Nodari, Daniela Ferrari, Fabrizio Giani, Cristina Zalfa, Guido Santilli, Angelo Luigi Vescovi
- P24 - ABETA AGGREGATE TOXICITY DEPENDS ON MEMBRANE CHOLESTEROL CONTENT IN FAMILIAL ALZHEIMER'S FIBROBLASTS**
Evangelisti E*, Zampagni M, Wright D, Liguri G, Puccetti B, Bagnoli S, Nacmias B, Sorbi S, Cecchi C
- P25 - INVOLVMENT OF D-SERINE RACEMASE DURING NEURONAL CELL DEATH**

Nadia Canu, Fulvio Florenzano, Anna di Luzio, Tiziana Borsello, Maria Teresa Ciotti

- P26 - MITOCHONDRIAL INVOLVEMENT IN CLN1 FIBROBLASTS**
Pezzini F*, Sembenini D, Vattemi G, Tonin P, Gismondi F, Marconi MG Carrozzo R, Santorelli FM, Simonati A
- P27 - PINK1 AND MITOCHONDRIAL DYSFUNCTION IN NEURONS AND SKELETAL MUSCLE CELLS**
Zhi Yao, Sonia Gandhi, Iain Hargreaves, Helene Plun-Favreau, Andrey Y Abramov, Nicholas W Wood
- P28 - CERULOPLASMIN DIFFERENTIAL EXPRESSION IN THE CEREBROSPINAL FLUID OF PARKINSON'S DISEASE REFLECTS OXIDATION AND FUNCTIONAL IMPAIRMENT FAVOURING INTRACELLULAR IRON OVERLOAD**
Olivieri S, Iannaccone S, Conti A, Cannistraci CV, Piccio L, Franciotta D, Cappa S, Alessio M
- P29 - ANALYSIS OF IRON HOMEOSTASIS IN SKIN FIBROBLASTS FROM PANTOTHENATE KINASE ASSOCIATED NEURODEGENERATION PATIENTS**
Campanella A, Rovelli E, Garavaglia B, Levi S
- P30 - BETA-AMYLOID ACTIVATES PARP THROUGH FREE RADICAL GENERATION BY THE NADPH OXIDASE**
Abeti R*, Abramov AY, Duchen MR
- P31 - GENOPOLIS CONSORTIUM FOR FUNCTIONAL GENOMICS- THE CORE FOR EXPLOITING MICROARRAY THECNOLOGIES**
Biancolini D, Papagna A, Lenzken C, Bonanno D, Barabino SML, Castagnoli-Ricciardi P, Calogero RA, Zolezzi F*
- P32 - MITOCHONDRIAL DYSFUNCTION IN EXPERIMENTAL CHARCOT-MARIE-TOOTH TYPE 1A (CMT1A) NEUROPATHY**
Lucilla Nobbio*, Fulvia Fiorese Valeria Prada, Silvia Ravera, Mariapaola Nitti, Angelo Schenone
- P33 - AXONUCLEAR COMMUNICATION SYSTEM IS ALTERED IN MOTOR NEURONS OF RODENT MODELS OF FAMILIAL AMYOTROPHIC LATERAL SCLEROSIS**
Peviani M*, Spano G, Pagani A, Bendotti C
- P34 - PURKINJE CELL AXONAL INJURY IN THE CEREBELLUM OF Ts65Dn MICE: ROLE OF BETA-AMYLOID OVERLOAD**
Lomoio S*, Necchi D, Scherini E
- P35 - PENTAPEPTIDES DERIVED FROM A-BETA 1-42 PROTECT NEURONS FROM THE MODULATORY EFFECT OF A-BETA OLIGOMER IN VIVO ELECTROPHYSIOLOGICAL STUDIES**
Chiovini B*, Barkùczi B, Juhçsz G, FflÜp L, Penke B, Szegedi V
- P36 - SYNAPTIC PLASTICITY STUDIES IN APOB-100 TRANSGENIC MICE IN CORRELATE TO ALZHEIMER'S DISEASE**
Chiovini B*, Barkùczi B*, Juhçsz G, Bereczki E, Scntha M, Penke B, Szegedi V
- P37 - MUTANT PRION PROTEIN EXPRESSION IS ASSOCIATED TO AN ALTERATION OF THE RAB GDP DISSOCIATION INHIBITOR ALPHA(GDI)/RAB11 PATHWAY**
Massignan T, Biasini E, Salmona M, Harris DA, Chiesa R, and Bonetto V
- P38 - CHARACTERIZATION OF DETERGENT-INSOLUBLE PROTEINS IN ALS MODELS AND PATIENTS INDICATES A POSSIBLE CAUSAL LINK BETWEEN NITRATIVE STRESS AND AGGREGATION IN PATHOGENESIS**

Manuela Basso, Giuseppina Samengo, Giovanni Nardo, Tania Massignan, Giuseppina D'Alessandro, Silvia Tartari, Lavinia Cantoni, Marianna Marino, Cristina Cheroni, Silvia De Biasi, Mario Salmona, Caterina Bendotti, Valentina Bonetto*

P39 - THE ACTIVITY OF NMDA RECEPTOR IS ENHANCED BY THE FIBRILLAR A-BETA 1-42 VIA THE INTEGRIN RECEPTOR PATHWAY

Barkùczi B*, Chiovini B, Juhász G, FflÜp L, Penke B, Szegedi V

P40 - ALPHA-SYNUCLEIN IS AN ACTIN BINDING PROTEIN AND ITS A30P MUTANT AFFECTS THE CYTOSKELETON STRUCTURE AND DYNAMICS

Bellani S*, Sousa VL, Giannandrea M, Meldolesi J, Valtorta F, Chieragatti E

P41 - ACUTE OR CHRONIC CO-TREATMENT WITH 1-METHYL-4-PHENYLPYRIDINIUM (MPP+) AND EPOXOMICIN IN ORGANOTYPIC CULTURES OF MOUSE MESENCEPHALON: SYNERGISTIC TOXICITY OR ALPHA-SYNUCLEIN AGGREGATES?

Rossen S, Larsen TR, Gramsbergen JB

P42 - MIS-SPLICING OF MICROTUBULE-ASSOCIATED TAU IN MYOTONIC DYSTROPHY TYPE: IMPLICATION OF CUGBP1 AND MBNL1

Dhaenens CM, Tran H, Schraen-Maschke, Sistiaga, A, Goicoechea M, Obriot H, Ghanem D, Van Brussel E, Gavaert MH, Maurage CA, Buae L, Sablonniere B, Lopez de Munain A, Caillet-Boudin ML, Sergeant N

P43 - ATAXIN-3 BINDS TUBULIN WITH HIGH AFFINITY

Mazzucchelli S*, De Palma A, D'urzo A, Pozzi C, Pastori V, Comelli F, Fusi P, Vanoni M, Tortora P, Mauri PL, Regonesi ME

P44 - EXPERIMENTAL MODELS: TOWARD STUDYING MECHANISMS OF NEUROFIBRILLARY DEGENERATION

Hamdane M*, Desmerciares J, Burnouf S, Bouafia H, Belarbi K, Troquier L, Demeyer D, Grosjean ME, Caillierez R, Barbot B, Brion I, Fernandez Gomez FJ, Humez S, Sergeant N, Blum D, Buae L

P45 - INHIBITION OF CALPAINS PREVENTS TRANSLOCATION OF MUTANT ATAXIN-3 TO THE NUCLEUS AND REDUCES NEURODEGENERATION IN A LENTIVIRAL MOUSE MODEL OF MACHADO-JOSEPH DISEASE

Simoës AT*, Goncalves N, Nùbrega C, Deglon N, Bandeira Duarte C, Kugler S, Pereira de Almeida L

P46 - ADENOSINE A2A RECEPTORS ANTAGONISM DELAYS NEURODEGENERATION IN A LENTIVIRAL MOUSE MODEL OF MACHADO JOSEPH DISEASE

Goncalves N*, Simoës AT Cunha R, Pereira de Almeida L

P47 - COMPARATIVE ANALYSIS OF STRUCTURAL AND FUNCTIONAL PROPERTIES OF BETA-AMYLOID AGGREGATED SPECIES

Denise Ferrera*, Claudio Canale, Fabio Benfenati, Laura Gasparini

P48 - SIMILAR OLIGOMERS OF HYPF-N SHOW DIFFERENT CYTOTOXIC EFFECTS

Zampagni M*, Becatti M, Wright D, Campioni S, Mannini B, Cascella R, Chiti F, Stefani M, Cecchi C

P49 - CELL DAMAGE INDUCED BY EXPANDED ATAXIN-1 INVOLVES MECHANICAL INSTABILITY OF NUCLEAR MEMBRANE

Averaimo S, Canale C, Pesci D, Fortunati V, Paulis D, Gliozzi A, Relini A, Mazzanti M, Jodice C*

P50 - PORCINE NEURONAL MODELS THAT RECAPITULATE HD PATHOGENESIS IN VITRO

Hruska-Plochan M*, Vodicka P, Lackova M, Usvald D, Juhas S, Hrdlickova R, Miyanoohara A, Marsala M, Motlik J

- P51 - NEW INSIGHTS INTO THE FIBRILLOGENESIS PROCESS OF ATAXIN-3, A MODEL PROTEIN FOR STUDIES ON POLYQ DISEASES**
Invernizzi G, Frana AM, Aprile FA, Natalello A, Relini A, Doglia SM, Tortora P, Regonesi ME
- P52 - ATAXIN-3 PROTEIN CONTEXT AND CELL-SPECIFIC FACTORS MODULATE POLYQ-MEDIATED NEURONAL AGGREGATION IN A C ELEGANS MODEL OF MACHADO-JOSEPH DISEASE**
Andreia Teixeira-Castro, Richard Morimoto, Patricia Maciel
- P53 - EFFECT OF SOD1 EXPRESSION IN A MUSCLE CELL MODEL OF FAMILIAL AMYOTROPHIC LATERAL SCLEROSIS**
Onesto E, Galbiati M, Zito A, Sau D, Crippa V, Poletti A
- P54 - ROLE OF JNK IN EXCITOTOXIC CELL DEATH FOLLOWING SEIZURE ACTIVITY**
*G Spigolon, C Veronesi, T Herdegen, C Bonny, A F Vercelli
- P55 - VEGF PROTECTS MOTOR NEURONS AGAINST EXCITOTOXICITY BY UPREGULATION OF GLUR2**
Elke Bogert*, Philip Van Damme, Koen Poesen, Joke Dhondt, Nicole Hersmus, Dora Kiraly, Wendy Scheveneels, Wim Robberecht & Ludo Van Den Bosch
- P56 - ROLE OF JNK IN EXCITOTOXIC CELL DEATH FOLLOWING SEIZURE ACTIVITY**
*G Spigolon, C Veronesi, T Herdegen, Y Zhao, C Bonny, A E Vercelli
- P57 - LOSS OF PARKIN PROTEIN CAUSES ABNORMAL INTRACELLULAR CALCIUM LEVELS UPON GLUTAMATERGIC STIMULATION IN NEURONAL CELLS**
Sassone J, Ascardi C, Zippel R, Colciago C, Tiloca C, Silani V, Ciammola A
- P58 - RETINOIC ACID INHIBITS P38-PHOSPHORYLATION AND PROTECTS AGAINST PREFIBRILLAR BETA-AMYLOID AND GLUTAMATE TOXICITY**
Zoia CP*, Riva C, Proserpio P, Salerno D, Tremolizzo L, Ferrarese C
- P59 - CEFTRIAXONE TREATMENT IMPROVES PHENOTYPE IN A MURINE MODEL OF SPINAL MUSCULAR ATROPHY**
D Ronchi*, M Nizzardo, M Nardini, S Salani, C Simone, M Falcone, C Donadoni, NBresolin, GP Comi, S Corti,
- P60 - HIGH AFFINITY GLUTAMATE TRANSPORTERS GLAST AND EAAC1 ARE PRESENT IN SCHWANN CELLS**
Magnaghi V, Di Cairano ES, Ballabio M, Perego C
- P61 - MUSCLE-RESTRICTED OVEREXPRESSION OF IGF-1 ATTENUATES DISEASE IN A MOUSE MODEL OF SPINAL AND BULBAR MUSCULAR ATROPHY**
Pennuto M, Palazzolo I, Kong L, Sumner C, Hadachi H, Katsuno M, Sobue G, Taylor JP, Fischbeck K
- P62 - PROTEOMIC PROFILING OF PRP27-30-ENRICHED PREPARATIONS EXTRACTED FROM THE BRAIN OF HAMSTERS WITH EXPERIMENTAL SCRAPIE**
Giorgi A, Di Francesco L, Principe S, Mignogna G, Sennels L, Mancone C, Alonzi T, Sbriccoli M, De Pascalis A, Rappsilber J, Cardone F, Pocchiari M, Maras M, Schinine ME*
- P63 - CHARACTERIZATION OF THE MOLECULAR DIVERSITY OF PRION PROTEIN AGGREGATES**
E Biasini, L Tapella, R Chiesa
- P64 - A NEW TRANSGENIC MOUSE MODEL OF INHERITED PRION DISEASE**

Dossena S, Imeri L, Mangieri M, Garofoli A, Ferrari L, Senatore A, Restelli E, Balducci C, Fiordaliso F, Salio M, Bianchi S, Fioriti L, Morbin M, Pincherle A, Marcon G, Villani F, Carli M, Tagliavini F, Forloni G, Chiesa R

P65 - RRS1 IS INVOLVED IN ENDOPLASMIC RETICULUM STRESS RESPONSE IN HUNTINGTON'S DISEASE

Carnemolla A*, Fossale E*, Agostoni E, Michelazzi S, Calligaris R, De Maso L, Del Sal G, MacDonald ME, Persichetti F

P66 - MUTANT PRION PROTEIN EXPRESSION CAUSES IMPAIRMENT OF VOLTAGE-GATED CALCIUM CHANNELS IN A TRANSGENIC MOUSE MODEL

Senatore A, Colleoni S, Verderio C, Restelli E, Mantovani S, Condliffe S, Congi L, Prestori F, Forloni G, D'Angelo E, Matteoli M, Gobbi M, Chiesa R

Poster Session I (P67-P133)

**Sunday, May 10, 2009
from 13:30 to 15:00**

P67 - MOLECULAR AND FUNCTIONAL ANALYSIS OF PARAPLEGIN GENE (SPG7) MUTATIONS IN PATIENTS WITH FAMILIAL AND SPORADIC SPASTIC PARAPLEGIA

D DiBella*, M Plumari, S Magri, V Fracasso, F Lazzaro, M Muzi-Falconi, R Fancellu, S DiDonato, D Pareyson, C Gellera, C Mariotti, S Baratta, F Taroni

P68 - EXPRESSION MODULATION OF A SMALL HEAT SHOCK PROTEIN INVOLVED IN NEURODEGENERATIVE DISEASES

Crippa V*, Boncoraglio A, Simonini F, Rusmini P, Poletti A

P69 - ROLE OF ALTERED PROTEOLYSIS IN THE DEGENERATION OF MOTOR NEURONS IN IN VIVO MODELS OF FAMILIAL AMYOTROPHIC LATERAL SCLEROSIS INVOLVEMENT OF CONSTITUTIVE AND IMMUNO PROTEASOMES

Fontana E*, Vitellaro-Zuccarello L, Cheroni C, Marino M, De Biasi S, Bendotti C

P70 - TDP-43 REDISTRIBUTION IS AN EARLY EVENT IN SPORADIC AMYOTROPHIC LATERAL SCLEROSIS

Giordana MT, Piccinini M, Grifoni S*, De Marco G, Vercellino M, Magistrello M, Pellerino A, Buccinnè B, Lupino E, Rinaudo MT

P71 - AUTOPHAGIC CLEARANCE OF THE PARKINSON'S DISEASE-ASSOCIATED GPR37 RECEPTOR

Di Pietro C, Marazziti D, Golini E, Mandillo S, Matteoni R, Tocchini-Valentini GP

P72 - GEC1 : A NEW AUTOPHAGY EFFECTOR HIGHLY EXPRESSED IN THE SNC

Chakrama FZ*, Seguin S, Fraichard A, Delage-Mourroux R, Jouvenot M, Boyer-Guittaut M

P73 - GREY-LETHAL MOUSE: A MURINE MODEL FOR MALIGNANT OSTEOPEetrosis AND A NEW NEURONAL LYSOSOMAL STORAGE DISEASE

Frattini A, De Biasi S, Strina D, Vitellaro Zuccarello L, Fontana E, Gioria M, Valnegri P, Sala C, Costantino-Ceccarini E

P74 - BRAIN NEURODEGENERATION AND INTERFERON GENE EXPRESSION ANALYSIS IN THE CEREBRO-SPINAL FLUID OF AICARDI-GOUTIRES PATIENTS

Izzotti A, Cartiglia C, Longobardi M, Pulliero A

P75 - TRANSCRIPTIONAL SIGNATURES IN NEURODEGENERATIVE DISEASES

Daniela Diamanti*, Pollio Giuseppe, Tarditi Alessia, Remelli Rosaria, Caricasole Andrea

P76 - THE EXPRESSION OF THE STEROIDOGENIC ACUTE REGULATORY PROTEIN (STAR) IS INCREASED IN THE DEGENERATING TRAIT OF SPINAL CORD OF WOBBLER MICE

Tartari S*, Bigini P, Orly J, Mennini T, Cantoni L

P77 - REST, THE TRANSCRIPTION REPRESSOR THAT GOVERNS NEUROSECRETION EXPRESSION, APPEARS TO HAVE A ROLE ALSO IN THE EXPRESSION OF GLIOSECRETION

Prada I*, D'Alessandro R, Stucchi L, Meldolesi J

- P78 - PARK2 PATIENT FIBROBLASTS TRANSCRIPTOME ANALYSES INDICATE A ROLE OF PARKIN IN ENDOCYTOSIS**
Klinkenberg M*, Gispert S, Morales-Gordo B, Walter M, Auburger G
- P79 - AMYOTROPHIC LATERAL SCLEROSIS: A HIGH THROUGHPUT ANALYSIS USING PHARMACOLOGICAL AND GENETICALLY MODIFIED CELLULAR MODELS**
Lenzken SC*, Della Beffa C, Bonanno D, Zolezzi F, Vivarelli S, Carri' MT, Calogero R, Barabino SML
- P80 - TNF-ALPHA mRNA AND PROTEIN EXPRESSION IN SPORADIC ALS PATIENTS**
Guareschi S, Cova E, Baiocchi C, Milani O, Federici Canova D, Cuccia M, Ceroni M, Cereda C*
- P81 - REGULATION OF MRNA STABILITY BY THE RNA BINDING PROTEIN TNRC4 IN NEURONAL DIFFERENTIATION**
Viero G*, Vessey J, Savino M, D'Apolito M, Grifa A, Macchi P, Quattrone A
- P82 - TRANSCRIPTIONAL REGULATION OF GLYCOSPHINGOLIPIDS DEGRADING ENZYMES**
Urbanelli L, Trivelli F, Magini A, Tancini B, Ciccarone V, Polidoro M, Emiliani C*
- P83 - PATHOGENETIC MECHANISMS IN THE CONGENITAL CENTRAL HYPOVENTILATION SYNDROME (CCHS)**
Benfante R*, Di Lascio S, Bachetti T, Ceccherini I, Fornasari D
- P84 - TRANSCRIPTIONAL CHANGES IN A CELLULAR MODEL OF HUNTINGTON'S DISEASE**
Rosenstock TR*, Oliveira CR, Rego AC
- P85 - MICROARRAY ANALYSIS UNVEILS THE INVOLVEMENT OF ADDITIONAL PATHWAYS IN DJ-1 MEDIATED NEURODEGENERATION**
Foti R, Zucchelli S, Biagioli M, Roncaglia P, Pinto M, Carrieri C, Vilotti S, Krmac H, Calligaris R, Gustincich S
- P86 - GENE DYSREGULATION IN FRAGILE X-ASSOCIATED TREMOR/ATAXIA SYNDROME**
F Tassone F, Garcia-Arocena D, Iwahashi C, Greco C, Hagerman RJ, PJ Hagerman PJ
- P87 - STUDIES ON THE PATHOPHYSIOLOGY OF NEUROLOGICAL INVOLVEMENT IN A MUCOPOLYSACCHARIDOSIS TYPE I MOUSE MODEL**
Marika Salvalaio, Rosella Tomanin, Elisa Legnini, Francesca D'Avanzo, Antonello D'Arrigo, Alberta Leon, Maurizio Scarpa
- P88 - OXIDATIVE AND ANTIOXIDANT MARKERS IN PATIENTS WITH NEURODEGENERATIVE DISEASES: A PILOT STUDY TO ESTIMATE RISK AND PROTECTIVE FACTORS**
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- P89 - EFFECTS OF BEE VENOM ON SYMPTOMATIC AMYOTROPHIC LATERAL SCLEROSIS MODEL**
Eun Jin Yang, Jing Hua Jiang, Hye Suk Hwang, Myeong Soo Lee, Sun Mi Choi
- P90 - POSTTRAUMATIC GABA(A)-MEDIATED [Ca²⁺]_i INCREASE IS ESSENTIAL FOR THE INDUCTION OF BDNF-DEPENDENT SURVIVAL OF MATURE CENTRAL NEURONS**
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- P91 - ACETYLCHOLINE INDUCES CHANGES IN PROTEIN FOLDING REGIMEN AND CYTOSKELETON ORGANIZATION**
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- P92 - CHRONIC TREATMENT WITH AMYLOID beta1-42 CHANGES AGONIST BINDING AT M1 AND M3 MUSCARINIC RECEPTORS EXPRESSED IN CHO CELLS**
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- P93 - THE GTPase AND THE KINASE DOMAINS OF THE PARKINSON'S DISEASE ASSOCIATED LRRK2: A CLOSE RELATIONSHIP**
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- P94 - ROLE OF ATAXIN-3 PHOSPHORYLATION IN SPINOCEREBELLAR ATAXIA TYPE 3 (SCA3) PATHOGENESIS**
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- P95 - ANTIBODIES AGAINST NEURONAL NICOTINIC ACETYLCHOLINE RECEPTORS: POTENTIAL ROLE IN NEURODEGENERATION**
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- P96 - GENETIC REPROGRAMMING OF DISEASE-SPECIFIC FIBROBLASTS**
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- P97 - FUNCTIONAL CONSEQUENCES OF WASH-RESISTANT XANOMELINE BINDING AT M2 AND M4 MUSCARINIC RECEPTORS**
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- P98 - INFLUENCE OF ALCURONIUM ON ACTIVATION OF M1-M4 MUSCARINIC RECEPTORS SIGNALLING BY CARBACHOL**
Randakova A, Jakubik J, El-Fakahany EE, Dolezal V
- P99 - MATHEMATICAL MODELING AND SIMULATION OF NEURO-CHIPS**
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- P100 - INFLUENCE OF MEMBRANE CHOLESTEROL CONTENT ON MUSCARINIC RECEPTOR STIMULATION OF GQ/11 G-PROTEIN SIGNALING**
Michal P, Rudajev V, El-Fakahany EE, Dolezal V
- P101 - CEREBELLAR IMMUNOLocalIZATION OF THE GUANINE EXCHANGE FACTOR RASGRF1 AND CHANGES IN THE PROTEOME PROFILE INDUCED BY ITS GENETIC DELETION IN MICE**
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- P102 - 17-(ALLYLAMINO)-17-DEMETHOXYGELDANAMYCIN (17-AAG) A PHARMACOLOGICAL APPROACH TO INDUCE DEGRADATION OF MISFOLDED PROTEINS**
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- P103 - MOLECULAR BIOMARKER/S OF HUNTINGTON'S DISEASE PATHOGENESIS IN HUMAN PERIPHERAL BLOOD AND CLONALLY STRIATA-DERIVED RAT CELLS**
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- P104 - THE ROLE OF REST AND ITS ASSOCIATED COFACTOR BHC80 IN MODULATING NEURONAL FUNCTIONS**

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- P105 - WALLERIAN-LIKE DISTAL AXONAL DEGENERATION FOLLOWING LOCAL AXOTOMY IN A NOVEL MICROFLUIDIC CULTURE SYSTEM**
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- P106 - SOD1 EFFECT ON CELL CYCLE PROGRESSION IN AN ALS MODEL**
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- P107 - TEMPORAL EVOLUTION OF SPINAL NETWORK ACTIVITY IN WILD TYPE AND G93A MUTANT MOUSE ORGANOTYPIC CULTURES**
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- P108 - PROTEINS RELATED TO NEURODEGENERATION ARE DIFFERENTIALLY EXPRESSED IN FIBROBLASTS FROM FAMILIAL ALZHEIMER'S DISEASE MUTATION CARRIERS, BOTH SYMPTOMATIC AND PRE-SYMPTOMATIC**
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- P109 - 9,10-ANTHRAQUINONE INTERFERES WITH Abeta-PEPTIDE AGGREGATION: MECHANISM OF ACTION**
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- P110 - ALTERED AXONAL GROWTH OF CULTURED SYMPATHETIC NEURONS FROM DYSTROPHIC MDX MICE**
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- P111 - ECTOPIC ATP SYNTHESIS: A NEW CORRELATION BETWEEN MITOCHONDRIAL DISORDERS AND NEURODEGENERATIVE DISEASES**
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- P112 - EFFECT OF GABA-A RECEPTOR SIGNALLING MODULATION ON NEURAL PRECURSORS**
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- P113 - PHENOTYPICAL CHARACTERIZATION OF THIRD TRIMESTER HUMAN AMNIOTIC FLUID CELLS WITH POTENTIAL FOR THERAPY IN NEURODEGENERATIVE DISORDERS**
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- P114 - MOTONEURON TRANSPLANTATION RESCUE THE PHENOTYPE OF SPINAL MUSCULAR ATROPHY WITH RESPIRATORY DISTRESS TYPE 1 (SMARD1)**
Monica Nizzardo*, Stefania Corti, Martina Nardini, Chiara Donadoni, Sabrina Salani, Dario Ronchi, Roberto Del Bo, Dimitra Papadimitriou, Federica Locatelli, Nereo Bresolin, Giacomo P Comi,
- P115 - NEURAL STEM CELLS DERIVED FROM GENETICALLY ENGINEERED, LINEAGE-SELECTABLE ES CELLS IMPROVES THE PHENOTYPE OF A MOUSE MODEL OF SPINAL MUSCULAR ATROPHY**
C Simone*, S Corti, M Nizzardo, S Salani, M Nardini, C Donadoni, D Ronchi, M Falcone, N Bresolin, GP Comi,
- P116 - MATRIX METALLOPROTEINASE-9 INCREASES IN THE STRIATUM AND SUBSTANTIA NIGRA OF A MOUSE MODEL OF PARKINSONISM: AN mRNA EXPRESSION, PROTEIN LEVEL AND IMMUNOLocalIZATION STUDY**
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- P117 - AN IN VITRO MODEL OF GLIA ACTIVATION TO DISSECT THE SPECIFIC ROLES OF ASTROCYTES AND MICROGLIA IN NEURODEGENERATIVE DISEASES**
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- P118 - NERVE GROWTH FACTOR (NGF) RESTORES NEURONAL AND GLIAL FUNCTION IN THE SPARED NERVE INJURY (SNI) MODEL OF NEUROPATHIC PAIN**
Cirillo G, Bianco MR, Colangelo AM*, Cavaliere C, De Luca D, Viggiani S, Fragni M, Alberghina L, Papa M
- P119 - IS MICROTUBULE DYNAMICS IMBALANCE AN EARLY EVENT IN MPTP-INDUCED NEUROGENERATION?**
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- P120 - THE MODULATION OF D-SERINE CONCENTRATION IN THE BRAIN AS A MOLECULAR MECHANISM IN SCHIZOPHRENIA SUSCEPTIBILITY**
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- P121 - NEUROMYELITIS OPTICA AUTOANTIBODIES SELECTIVELY RECOGNIZE AQUAPORIN-4 ORTHOGONAL ARRAYS OF PARTICLES**
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- P122 - SOLIDITY OF THE REDUCTION OF THE CHOLESTEROL BIOSYNTHESIS PATHWAY IN HD AND POTENTIAL REGULATORY ROLE OF NORMAL HUNTINGTIN**
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- P123 - ROLE OF IGF-1 AUTOCRINE SECRETION AND SIRTUINS REDISTRIBUTION IN THE RESISTANCE OF NEURONAL DIFFERENTIATED CELLS TO STS-INDUCED CELL DEATH**
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- P124 - FETAL-DERIVED NEURAL STEM (NS) CELLS RETAIN THE IN VIVO POSITIONAL INFORMATION CODE AND DIFFERENTIATE IN APPROPRIATE NEURONAL LINEAGES**
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- P125 - A NEW SYSTEMATIC APPROACH TO DETECT BDNF MRNA IN HUMAN BLOOD SAMPLES**
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- P126 - AN EVOLUTIONARY APPROACH TO THE STUDY OF HUNTINGTIN, THE PROTEIN CAUSING HUNTINGTON'S DISEASE**
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- P127 - ASTROCYTES DERIVED FROM NEURAL PRECURSOR CELLS OF WOBBLER MICE INDUCE APOPTOTIC DEATH OF MOTOR NEURONS THROUGH REDUCED GLUTAMATE TRANSPORT**
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- P128 - GENERATION OF iPS CELLS FOR MECHANISTIC STUDIES IN HUNTINGTON'S DISEASE**

Christelle Rochon*, Valentina Castiglioni, Alessia Delli Carri, Luciano Conti, Caterina Mariotti, Stefano DiDonato, Marisa Jaconi, Outi Hovatta, Anis Feki, Elena Cattaneo

P129 - THE DIVERSITY OF LIFE: AN APPROACH

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P130 - GENETIC INTERACTION BETWEEN THE m-AAA PROTEASE ISOENZYME REVEALS NOVEL ROLES IN NEURODEGENERATION

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P131 - SPASTIN: A BRIDGE BETWEEN CYTOSKELETAL REMODELING AND MEMBRANE TRAFFICKING VIA PROTRUDIN-RAB11 PATHWAY

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P132 - ROLE OF THE AXONAL-SMN (α -SMN) PROTEIN IN AXON GROWTH AND SPINAL MUSCULAR ATROPHY (SMA) PATHOGENESIS

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P133 - SELADIN-1 AND BETA-AMYLOID TOXICITY ARE MODULATED BY GnRH.

Zito A, Cariboni A, Piva F, Maggi R, Galbiati M