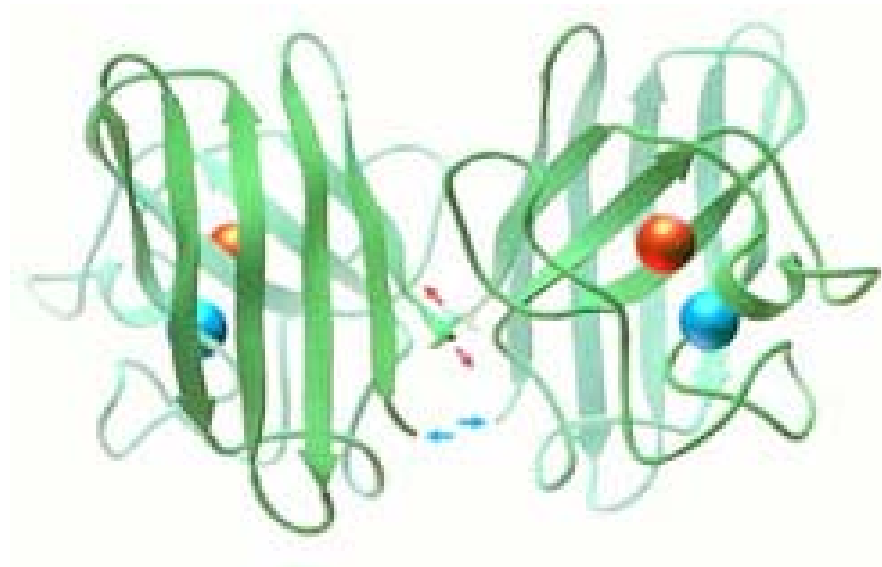


# Mutant SOD1 and familial ALS: from the molecule to man

Milan (Italy) September 13-16, 2007



## **Organizers**

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## **Scientific Committee**

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## **Secretariat**

Annalisa Volpe; [a.volpe@hsantalucia.it](mailto:a.volpe@hsantalucia.it)

*Dear Colleagues and Friends,*

*It is a great pleasure for us to welcome you to this first meeting on mutant SOD1 and familial ALS in Milano, Italy.*

*The conference is focussed on the selective neurotoxic properties of mutant SOD1, from biochemical and biophysical data in vitro to results in vivo from studies in animal models and patients.*

*Hopefully, this meeting will offer the opportunity to review and discuss existing evidence and new hypotheses on the mechanisms of mutant SOD1-linked ALS, and encourage scientific collaboration. We are certain that this will be of help both to all the neuroscientists already working in the field and to those that are now joining in the attempt to understand the cause of this rather obscure disease.*

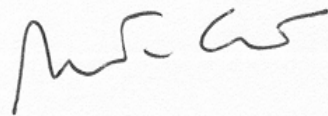
*Since the conference is running in Milano, we are also pleased to be able to offer you the exclusive opportunity to visit the famous "Last Supper" painted by Leonardo da Vinci from 1495 to 1498.*

*We wish to thank all participants, all the people who have assisted us in the organization of the meeting and our sponsors that made it possible.*

*Last, but not least, many thanks are due to Prof. Silvio Garattini, director of the Mario Negri Institute, for kindly hosting the meeting.*



*Caterina Bendotti*



*Maria Teresa Carri*

# Scientific Program

## Thursday 13 September

**Registration 3.30 pm**

**Welcome Ceremony 5.30 pm**

Caterina Bendotti, Maria Teresa Carri  
Silvio Garattini ( Director of Mario Negri Institute)  
Mario Melazzini ( President of AISLA)

**Opening lecture 6.00 pm**

**S1** - Giuseppe Rotilio (Italy)

*From erythrocyte to SOD1: memories of a Biochemist.*

**Welcome party 7.00 pm**

## Friday 14 September

**Session I 8.30 am – 10.45 am**

**Biochemistry of Mutant SOD1s**

**Chairpersons: J.S. Valentine – G. Rotilio**

**S2** - Joan S. Valentine (USA)

*How do ALS-associated mutations in superoxide dismutase 1 promote aggregation of the protein?*

**S3** - Mikael Oliveberg (Sweden)

*ALS-associated SOD1 mutations preferentially reduce the proteins repulsive charge*

**S4** - Lucia Banci (Italy)

*Interplay between structural and dynamical properties and protein aggregation in SOD1*

**S5** - Joseph S. Beckman (USA)

*Direct measurement of zinc-deficient SOD localized the ventral gray matter in G93A SOD rats*

**Selected oral communication**

**SO1** - Albrecht Clement (Germany)

*Dismutase-inactive mutant SOD1 has different functional properties when heterodimerized with wild type SOD1*

**Coffee break (10.45 am -11.15 am)**

**Session II 11.15 am – 1.00 pm**  
**Mutant SOD1s toxicity in cell models**  
**Chairpersons: S.L. Marklund – M.T. Carri**

**S6** - Pamela Shaw (UK)

*Mutant SOD1 in motoneuronal cells: mechanisms from the genomic and proteomic profile*

**S7** - Gen Sobue (Japan)

*Mutant SOD1s and protein aggregation/degradation in motoneuronal cells*

**S8** - Maria Teresa Carri (Italy)

*Mutant SOD1s and mitochondria in motoneuronal cells*

**Selected oral communication**

**SO2** - Mauro Cozzolino (Italy)

*Cysteine 111 affects aggregation of mutant SOD1 in NSC34 cells*

**Lunch (1.00 pm – 2.00 pm)**

**Session III 2.00 pm – 4.15 pm**  
**Mutant SOD1s toxicity in animal models**  
**Chairpersons: Z.S. Xu – W. Robberecht**

**S9** - Stefan L. Marklund (Sweden)

*Soluble misfolded SOD1 species in ALS*

**S10** - Jean-Pierre Julien (Canada)

*From secretion of SOD1 mutants to immunotherapeutic approaches*

**S11** - Jasna Kriz (Canada)

*Live imaging of disease progression in SOD1 mutant mice*

**S12** - Valentina Bonetto (Italy)

*Proteomics of spinal cord aggregates in G93A SOD1 transgenic mouse*

**Selected oral communication**

**SO3** - Avi Chakrabarty (Canada)

*Detection of misfolded SOD1 in sporadic and familial ALS*

**Coffee break (4.15 pm – 4.45 pm)**

**Session IV 4.45 pm – 6.45 pm**  
**Genetics of Familial ALS**  
**Chairpersons: C.E. Shaw – A. Chiò**

**S13** - Peter M. Andersen (Sweden)  
*Genetics of SOD1-linked fALS*

**S14** - Christopher E. Shaw (UK)  
*Genetics of non-SOD1-linked fALS*

**S15** - Stefania Battistini (Italy)  
*SOD1 in Italian ALS patients*

**S16** - Wim Robberecht  
*The pathogenesis of sporadic amyotrophic lateral sclerosis*

**Selected oral communication**

**SO4** - Richard Wroe (UK)  
*The Evolution of the Amyotrophic Lateral Sclerosis Database (ALSOD)*

**Social Event - Visit to Leonardo's "Last Supper" 7.30 pm**  
**Social Dinner – 8.30 pm**

**Saturday 15 September**

**Session V 8.30 am – 10.15 am**  
**Motor neuron death in SOD1-linked fALS: intra-cellular mechanisms**  
**Chairpersons: L. Cantoni – P. Pasinelli**

**S17** - Piera Pasinelli (USA)  
*The double life of two pro-survival proteins: when SOD1 and Bcl-2 get mutated, wild and pro-death*

**S18** - Claudia Crosio (Italy)  
*Role of Bcl2a1 in SOD1-linked fALS*

**S19** - Angelo Poletti (Italy)  
*The nucleus as target for mSOD1 toxicity*

**S20** - Massimo Tortarolo (Italy)  
*Role of MAP kinases cascade in SOD1-linked fALS*

**Selected oral communication**

**SO5** - Haining Zhu (USA)  
*Interaction between familial ALS-linked SOD1 mutants and the dynein complex*

**Coffee break (10.15 am – 10.45 am)**

**Session VI 10.45 am – 12.30 noon**

**Motor neuron death in SOD1-linked fALS: inter-cellular mechanisms**

**Chairpersons: M. Bentivoglio – L. Barbeito**

**S21** - Luis Barbeito (Uruguay)

*SOD1 mutations and astrocyte dysfunction in ALS*

**S22** - Marina Bentivoglio (Italy)

*Neuroinflammatory signalling in the motor circuit in SOD1-linked fALS*

**S23** - Davide Trotti (USA)

*Post-translational processing of the glial glutamate transporter EAAT2 in ALS*

**Selected oral communication**

**SO7** - Giambattista Bonanno (Italy)

*Excessive exocytotic release of glutamate in the spinal cord of SOD1 G93A mutant mice*

**Lunch (12.30 noon – 1:30 pm)**

**Session VII 1:30 pm – 3.15 pm**

**Motor neuron death in SOD1-linked fALS: remote contributions**

**Chairpersons: J.P.Loeffler – A.Musarò**

**S24** - Antonio Musarò (Italy)

*Muscle control of motor neuron survival and activity*

**S25** - Linda Greensmith (UK)

*Axonal Transport deficits in models of motor neuron disease*

**S26** - Jean-Philippe Loeffler (France)

*Energy homeostasis and denervation processes at the neuro-muscular junction in ALS*

**Selected oral communication**

**SO8** - Valeria Padovano (Italy)

*Effects of physical exercise and the steroid nandrolone on neuromuscular junctions in G93A -SOD1 transgenic mice*

**Coffee break (3.15 pm – 3.45 pm)**

**Session VIII 3.45 pm – 5.45 pm**

**Poster Session**

## **Sunday 16 September**

**Session IX 8.30 am – 12.30 noon**  
**Therapeutics in SOD1 mutant mice**  
**Chairpersons: V. Silani – C. Raoul**

**S27** - Cedric Raoul (Switzerland)

*Lentiviral and adeno-associated viral vectors for motoneuron gene therapy*

**S28** - Zuoshang Xu (USA)

*An RNAi approach to understanding the mechanism and therapy of ALS caused by mutant SOD1*

**S29** - Vincenzo Silani (Italy)

*Translational research: possible explanations for the failure in ALS*

**Coffee break (10.00 am -10.30 am)**

**S30** - Jonathan Glass (USA)

*SOD1-People: It's time for a trial*

**S31** - Lucie Bruijn (USA)

*Drug discovery and development for ALS*

**Selected oral communication**

**SO9** - Rita Perlingeiro (USA)

*The potential of endothelial progenitors to recover the stem cell niche in ALS*

**Closing remarks**

Caterina Bendotti (Italy)

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**Individual departure** (Sandwiches available)