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Neurotoxic mutant misfolded proteins in motor neuron diseases

**Angelo Poletti**

Dipartimento di Endocrinologia, Fisiopatologia e Biologia Applicata (DEFIB)  
Centro di Eccellenza per lo studio delle Malattie Neurodegenerative (CEND)  
Universita' degli Studi di Milano

## **ABSTRACT OF THE TALK**

The neurotoxic events at the basis of several types of inherited neurodegenerative diseases are often a consequence of aberrant conformations (misfolding) generated by the insertion of mutations in the affected protein(s). The protein misfolding might trigger and perturb a wide variety of downstream processes affecting different neuronal functions.

Motor neuron diseases (MNDs) are a class of neurodegenerative diseases in which cortical and/or bulbar and spinal motor neurons are affected. Two different types of MNDs, Spinobulbar Muscular Atrophy (SBMA or Kennedy's disease) and some familial forms of Amyotrophic Lateral Sclerosis (fALS), have been linked to mutant proteins that seem to acquire gain of neurotoxic functions as a consequence of protein misfolding. The major aspects related to mutant proteins aggregation, degradation via the proteasome and the autophagic system, as well as the role of chaperones in mutant protein turnover will be presented.