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Sommario/riassunto	Amyotrophic lateral sclerosis (ALS), which was described since 1869 by Jean Martin Charcot, is a devastating neurodegenerative disease characterized by the selective and progressive loss of upper and lower motor neurons of the cerebral cortex, brainstem and the spinal cord. The cognitive process is not affected and is not merely the result of aging because may occur at young ages. The only known cause of the disease is associated with genetic mutations, mainly in the gene encoding superoxide dismutase 1 (familial ALS), whereas there is no known cause of the sporadic form of ALS (SALS), which comprises >90% of cases. Both ALS types develop similar histopathological and clinical characteristics, and there is no treatment or prevention of the disease. Because effective treatments for ALS, as for other neurodegenerative diseases, can only result from the knowledge of their cellular and molecular pathophysiological mechanisms, research on such mechanisms is essential. Although progress in neurochemical, physiological and clinical investigations in the last decades has identified several mechanisms that seem to be involved in the cell death process, such as glutamate-mediated excitotoxicity, alterations of inhibitory circuits, inflammatory events, axonal transport deficits,

oxidative stress, mitochondrial dysfunction and energy failure, the understanding of the origin and temporal progress of the disease is still incomplete and insufficient. Clearly, there is a need of further experimental models and approaches to discern the importance of such mechanisms and to discover the factors that determine the selective death of motor neurons characteristic of ALS, in contrast to other neurodegenerative diseases such as Parkinson’s and Alzheimer’s disease. Whereas studies in vitro in cell cultures, tissue slices or organotypic preparations can give useful information regarding cellular and molecular mechanisms, the experiments in living animal models obviously reflect more closely the situation in the human disease, provided that the symptoms and their development during time mimics as close as possible those of the human disease. It is necessary to correlate the experimental findings in vitro with those in vivo, as well as those obtained in genetic models with those in non-genetic models, aiming at designing and testing therapeutic strategies based on the results obtained.