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Special Issue on Motor Neuron Disorders: Molecular Mechanisms of Pathogenesis and Therapeutic Developments

Motor neuron diseases (MNDs) are a group of most devastating neurological disorders (sporadic and inherited) characterized by degeneration of motor neurons that regulate the function of the skeletal muscles to control essential voluntary movements of body parts. This issue will focus on the recent advances in the fields of spinal muscular atrophy (SMA), amyotrophic lateral sclerosis (ALS), and motor neuron diseases. Both SMA and ALS share common pathogenesis caused by the loss of spinal motor neurons. However, the two diseases are quite different in terms of age and onset. SMA is a childhood disease with early onset, whereas ALS is an adulthood disease with middle to late age onset. SMA is an autosomal recessive genetic disorder caused by mutation of the survival motor neurons 1 (SMN1) gene. Insufficient levels of SMN protein result in degeneration of lower motor neurons and lead to wasting of the skeletal muscle. ALS has both inherited (familial) and sporadic forms and the familial forms have been linked to several distinct genes, including superoxide dismutase (SOD1) and C9orf72. ALS has degeneration of upper as well as lower motor neurons. Studies have indicated that the possibility of common defects such as altered RNA metabolism might play a role in the pathogenesis of ALS and SMA. At present, no effective treatment is available to cure or reduce the burden of illness because of limited understanding of the molecular mechanisms of motor neuron degeneration associated with MNDs. However, recent advances in basic and clinical research have indicated exciting possibilities of commonalities between MNDs. Despite differences in genetics and upstream molecular events that trigger the onset of these diseases, the fact is that the motor neurons are specifically affected which raises the possibility of identification of common downstream targets for therapeutic intervention.

We invite investigators to contribute original research work as well as review articles that will advance our understanding of the molecular mechanisms of motor neuron degeneration and muscle atrophy and knowledge to design novel strategies for the treatment of MNDs.

Potential topics include, but are not limited to:

- ▶ Molecular mechanisms of motor neuron degeneration
- Molecular mechanisms of muscle wastage and neuromuscular junctions (NMJs) pathology
- > Development of new vertebrate and invertebrate models of ALS and SMA
- Testing of therapeutic potential of new compounds using cell or animal-based models
- Identification and characterization of new molecular targets
- Genetic and pharmacological methods for rescue of phenotype
- ► Identification and characterization of disease modifier genes
- ▶ Function of proteins associated with pathogenesis of diseases
- ▶ Identification of additional genetic loci associated with familial ALS

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