



김 지 은

서울의료원 신경과

Update in Neurology 2 : Neuromuscular Disease

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Remarkable researches and recommendations from expert committees in the field of neuromuscular disease were made in 2016. In this review, we focused on the progression of main neuromuscular diseases - myasthenia gravis, inflammatory neuropathy and motor neuron disease - in this year. These include the results of multicenter, randomized controlled trial of thymectomy in generalized non-thymomatous myasthenia gravis, 2016 international consensus guidance for management of myasthenia gravis, recent conceptual changes in the pathogenesis of two types of Guillain-Barré syndrome (axonal and demyelinating types of Guillain-Barré syndrome) and chronic inflammatory demyelinating polyneuropathy with introduction of their newly suggested biomarkers (as antibodies against contactin-1, contactin-associated protein 1(Caspr), sialylated IgG-Fc). Furthermore, we present key novel genetic associations identified in amyotrophic lateral sclerosis(CCNF, NEK1 mutations, C21orf2 as gene associated with amyotrophic lateral sclerosis risk, variant within CAMTA1 gene as modifiers of survival, TTN as rapid functional decline marker), and several new outcomes of intra-spinal stem cell transplantation for amyotrophic lateral sclerosis. We also described newly suggested classification system of amyotrophic lateral sclerosis from experts and reappraised clinical characteristics as frequent amyotrophic lateral sclerosis plateaus/small reversal and changes in metabolism associated with hypothalamic melanocortin pathway. Among the neurological complications of Zika virus infection (microcephaly, Guillain-Barré syndrome, meningoencephalitis and myelitis), Zika virus-associated Guillain-Barré syndrome was also discussed in this section.

Key Words: Neuromuscular disease, Myasthenia gravis, Guillain-Barré syndrome, Chronic inflammatory demyelinating polyneuropathy, Inflammatory neuropathy, Amyotrophic lateral sclerosis. Zika virus

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