The goal of the NINDS Neuromuscular Cognitive CDE subgroup was to review the literature and make recommendations for the use of cognitive and behavioral instruments that would be informative across the range of diseases that fall into this category and have been used in clinical studies of these disorders. After extensive review of the instruments available, the advisory group came to the conclusion that there are no data to support one instrument for general use across all neuromuscular diseases. The group agreed that the cognitive and behavioral aspects of neuromuscular diseases are important, and that there are multiple domains that may be considered in the measure of cognition and behavior. The group came up with the following recommendations:

1. Cognitive and behavioral outcomes should be considered in studies of neuromuscular disorders, yet few have been validated for use among individuals with neuromuscular disease. Selection of individual measures should be chosen with the particular etiology, functional ability and developmental stage of the individuals in mind.
2. When choosing scales/instruments, the neuromuscular limitations of the subjects must be considered, such as motor limitations or associated sensory deficits that may affect the performance of the testing or the rating of answers by the subject.
3. General domains that should be considered in the selection of measures include: general intellectual function, learning and memory, language, visuospatial, executive functions, and academic achievement. Additional domains that may need to be evaluated because they may have an impact on cognition in these diseases include adaptive skills, behavior, pain and sleep. In some neuromuscular populations, specific cognitive issues are prominent. When possible, cognitive and behavioral testing should be chosen to address the specific and expected deficits of the studied neuromuscular population. When they become available, disease-specific outcome measures validated in select neuromuscular population should be favored over generic measures.
4. No specific instrument is mandated for use among the neuromuscular diseases, and selection of measures should be guided by relevant expertise. However, multiple instruments have been described and can be chosen from the other CDEs, having been tested in those other disorders.
5. A table of instruments that may be considered for study of pediatric neuromuscular disease is provided for guidance. Please note, however, the table is provided to offer suggestions of possible instruments. No instrument listed is required to be used, and not all instruments suggested are expected to be of use in all studies. The table is not comprehensive and does not reflect all possible instruments that would be of value in studying pediatric neuromuscular disease.
6. Further study and/or development of scales/outcomes for specific neuromuscular disorders is important for future research.

This subgroup strongly endorses activities to develop and validate instruments to assess critical cognitive domains in these patient populations.