

**Mental Health Considerations**  
**for**  
*Adult Patients*  
**with Neuromuscular Disease**

# Created in Collaboration with



## **Mary Holt, RN, MS, CPLC**

Mental Health Nurse — MDA/ALS Center of Hope at Temple University  
Clinical Nurse Educator — Ashfield Healthcare, LLC

# Program Agenda

## 1. Overview of mental health in NMD

- Varying degrees of cognitive and psychological symptoms in NMD

## 2. Unique Aspects of Psychological Care in Adult NMD Patient Population

- Cognition, psychosocial considerations, and psychological issues

## 3. Management of mental health issues for adults with NMD

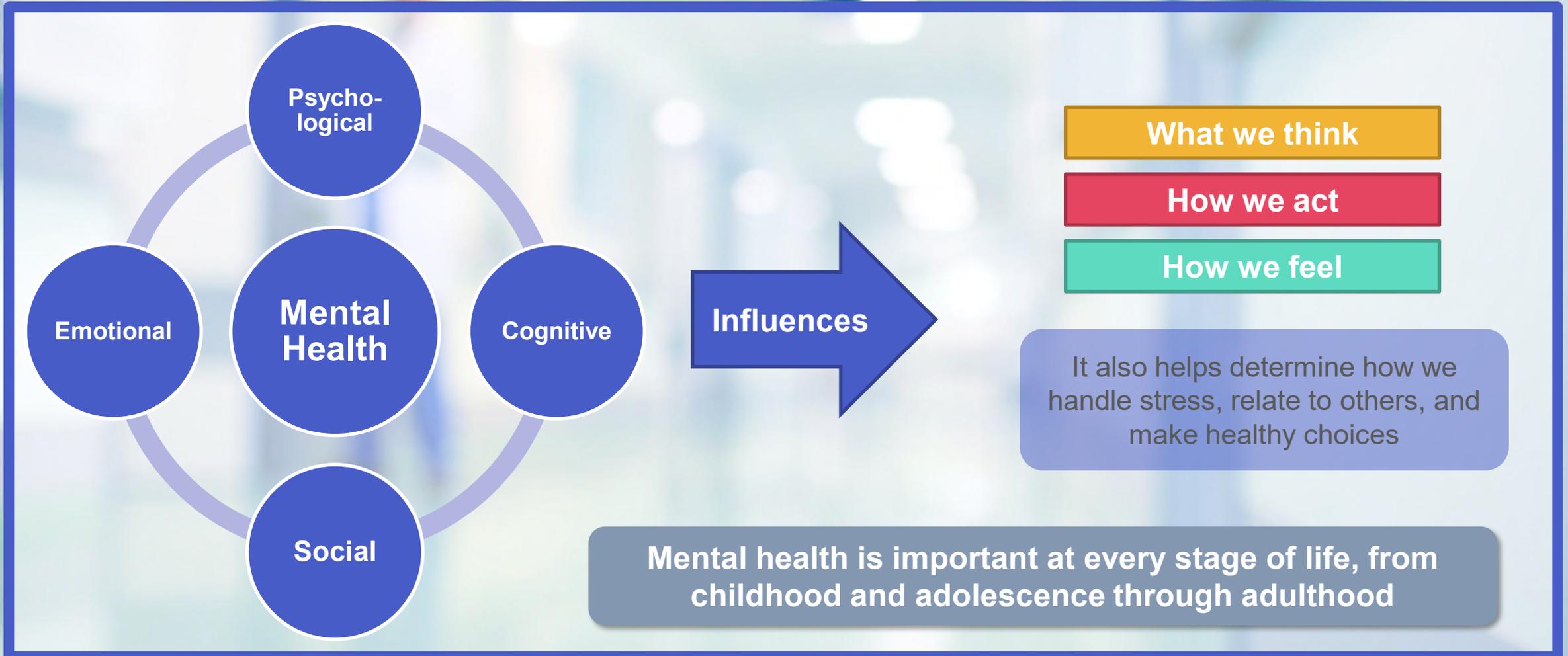
- Multidisciplinary team collaboration, community resources, education



# Overview of Mental Health in NMD

Varying degrees of cognitive and psychological  
symptoms in NMD

# Mental Health Includes Our Emotional, Psychological, Cognitive, and Social Well-Being

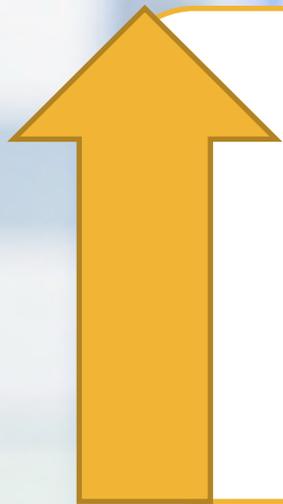


# Mental Health and NMD Patient Care

» Mental and physical health are equally important components of overall health<sup>1</sup>

*“Health is a state of complete physical, mental, and social well-being and not merely the absence of disease or illness.”<sup>2</sup>*

– World Health Organization (WHO), 2006



Mental illness, especially **depression**, increases risk for<sup>1</sup>...

Stroke  
Type 2 diabetes  
Heart disease  
Other long-term illnesses

The presence of chronic conditions can also increase the risk for developing mental illness<sup>2,3</sup>

Mental health challenges can pose barriers to optimum patient care if neglected and/or poorly managed<sup>1</sup>

1. Centers for Disease Control and Prevention. Learn about mental health. <https://www.cdc.gov/mentalhealth/learn/index.htm>. Access

2. World Health Organization. Mental health: strengthening our response. <https://www.who.int/en/news-room/fact-sheets/detail/mental-health-strengthening-our-response>. Accessed June 23, 2020.

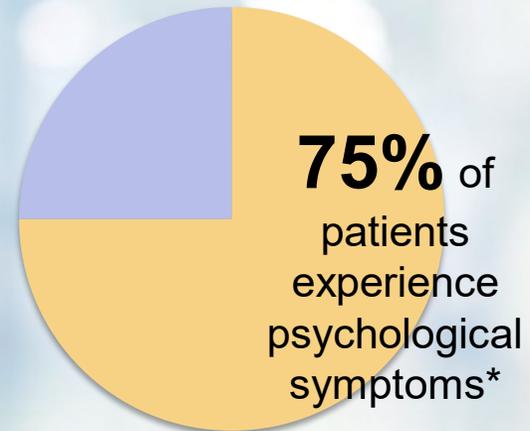
3. National Institutes of Health. Chronic illness and mental health. <https://www.nimh.nih.gov/health/publications/chronic-illness-mental-health/index.shtml>. Accessed June 23, 2020.

# Mental Health Issues Comorbid With NMD Are Increasingly Being Recognized

## Cognitive deficits in NMD<sup>1,2</sup>

- » Cognitive deficits are common in patients with NMD
  - Irritability, personality change, executive dysfunction, visuospatial dysfunction, and daytime sleepiness have been reported
  - Symptoms can progress with worsening disease

## Psychological symptoms in NMD<sup>2</sup>



### Drivers:

- Decreasing ability to physically keep up with daily life
- Isolation
- Uncertainty about the future
- Pervasive fatigue

Unmet need in managing psychological symptoms in patients with NMD has been rated **high or medium-high**<sup>2,\*</sup>

\*According to Neuromuscular Disease Healthcare Provider Survey respondents. Responses from 51 neurologists and 39 non-neurologists, including physical medicine and rehabilitation clinicians, neuromuscular nurses, nurse practitioners, physical therapists, and social workers. June 2018.

# Unique Aspects of Psychological Care in Adult NMD Patient Population

*Cognition, psychosocial considerations, and  
psychological issues*

# Cognition

» The mechanisms by which essential motor dysfunctions can affect cognitive performance remain poorly understood

Three large, heterogeneous groups of neuromuscular diseases have been implicated in cognitive alterations

**Motor neuron diseases**

**Muscular dystrophies**

**Mitochondrial disorders**

# Motor Neuron Disease and Cognition

## Amyotrophic lateral sclerosis (ALS)

- **>50%** of patients, at some point in the disease course, will develop phenotypic picture of cognitive or behavioral impairment
- **~13%** will exhibit concomitant behavioral-variant frontotemporal dementia (b-FTD)

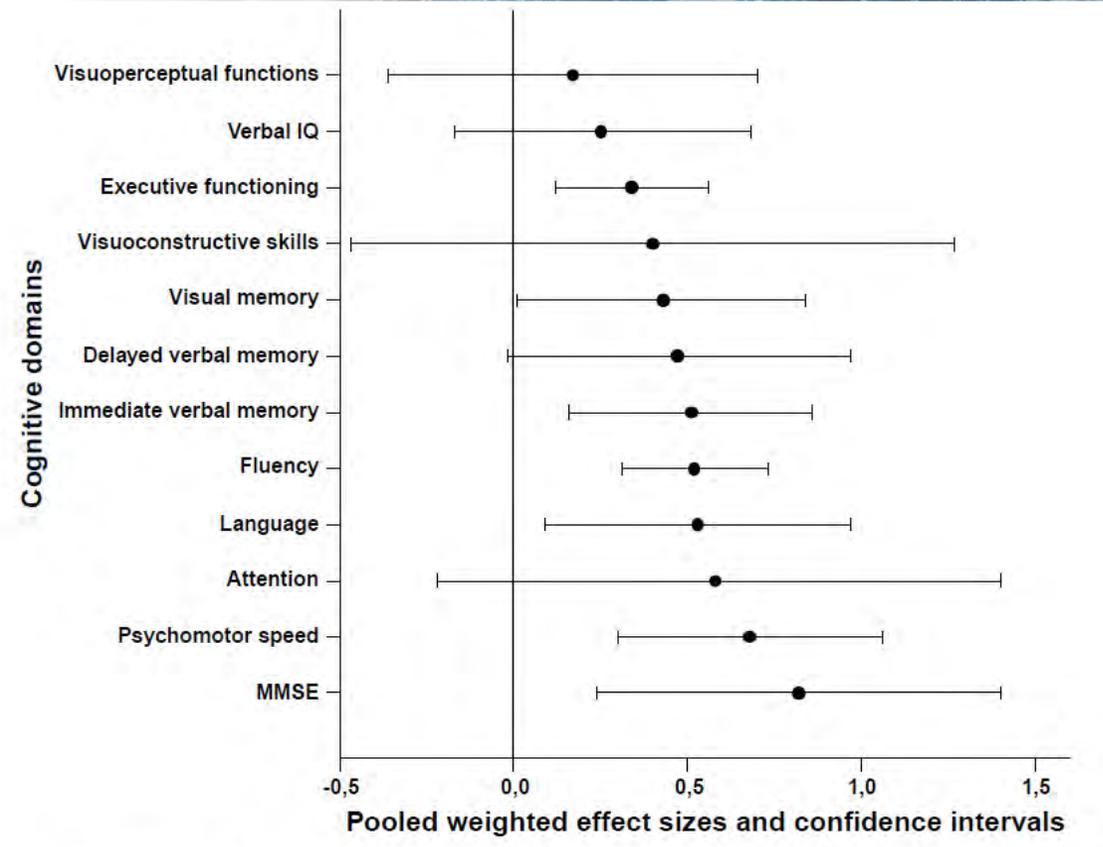
## Bulbospinal muscular atrophy or Kennedy disease (KD)

- Cognitive impairment with a predominant impairment in long-term memory and selective attention
- Clinical characteristics:
  - Frontal lobe dysfunction with disinhibition
  - Impaired executive functions
  - Reduced affective modulation

# Motor Neuron Disease and Cognition (cont.)

- » Meta-analysis of 16 published studies confirmed that patients with non-demented ALS (N=554) experience cognitive impairment in multiple domains

Pooled weighted effect sizes (•) and 95% confidence intervals of the cognitive domains calculated from available studies. A positive effect size indicates a worse cognitive performance of patients with ALS compared to controls for that domain. If the horizontal line does not bracket the vertical bar (0), the effect is significant at the 0.05 level.



# Dystrophinopathies and Cognitive Impairment

## Myotonic dystrophinopathies (DM1/2)

- DMs are autosomal dominant multisystemic disorders that also affect the brain
- Cognitive deficits have been observed in 24%-75% of patients with DM1 and depend on age of onset of disease
  - **Congenital onset:**
    - Mental retardation
  - **Childhood onset:**
    - Learning disabilities due to:
      - Low IQ
      - ADHD
      - Autistic behavior
      - Visuospatial impairment
  - **Adult onset:**
    - Dysexecutive syndrome
    - Visuospatial deficit
  - **Elderly onset:**
    - Age-dependent memory deficit

# Dystrophinopathies and Cognitive Impairment (cont.)

## Duchenne muscular dystrophy (DMD)

- Dystrophin plays an important role in the architectural organization of the CNS
- Can lead to neurobehavioral disorders:
  - ADHD
  - Autism spectrum disorders
  - Obsessive-compulsive disorder
- Can lead to specific learning disabilities:
  - Dyslexia
  - Impaired executive functions (e.g., problem solving, inhibition, and working memory)

## Becker muscular dystrophy (BMD)

- In BMD, there is a production of partially functional dystrophin
- Compared to DMD, cognitive impairment and neurobehavioral comorbidities are not as common or severe

# Mitochondrial Disorders and Cognitive Impairment

## Mitochondrial disorders

- A genetically heterogeneous group and variable phenotype — listed are the most frequent mitochondrial diseases with cognitive impairments:
  - MELAS
  - MERRF
  - MIDD
  - CPEO
  - KSS
  - NARP
- Cognitive function in these patients has been evaluated with neuropsychological testing, and the domains impaired were:
  - Abstract reasoning
  - Verbal memory
  - Visual memory
  - Language
  - Executive/constructive function
  - Calculation, attention, or visuospatial functions

# Psychosocial Considerations

## » Socialization

- Access to family/friends
- Assimilating
- Isolation
- COVID-19

## » Relationships

- Intimacy (physical/emotional)

## » Ongoing chronicity of condition

- Rapidity vs. slowness of progression

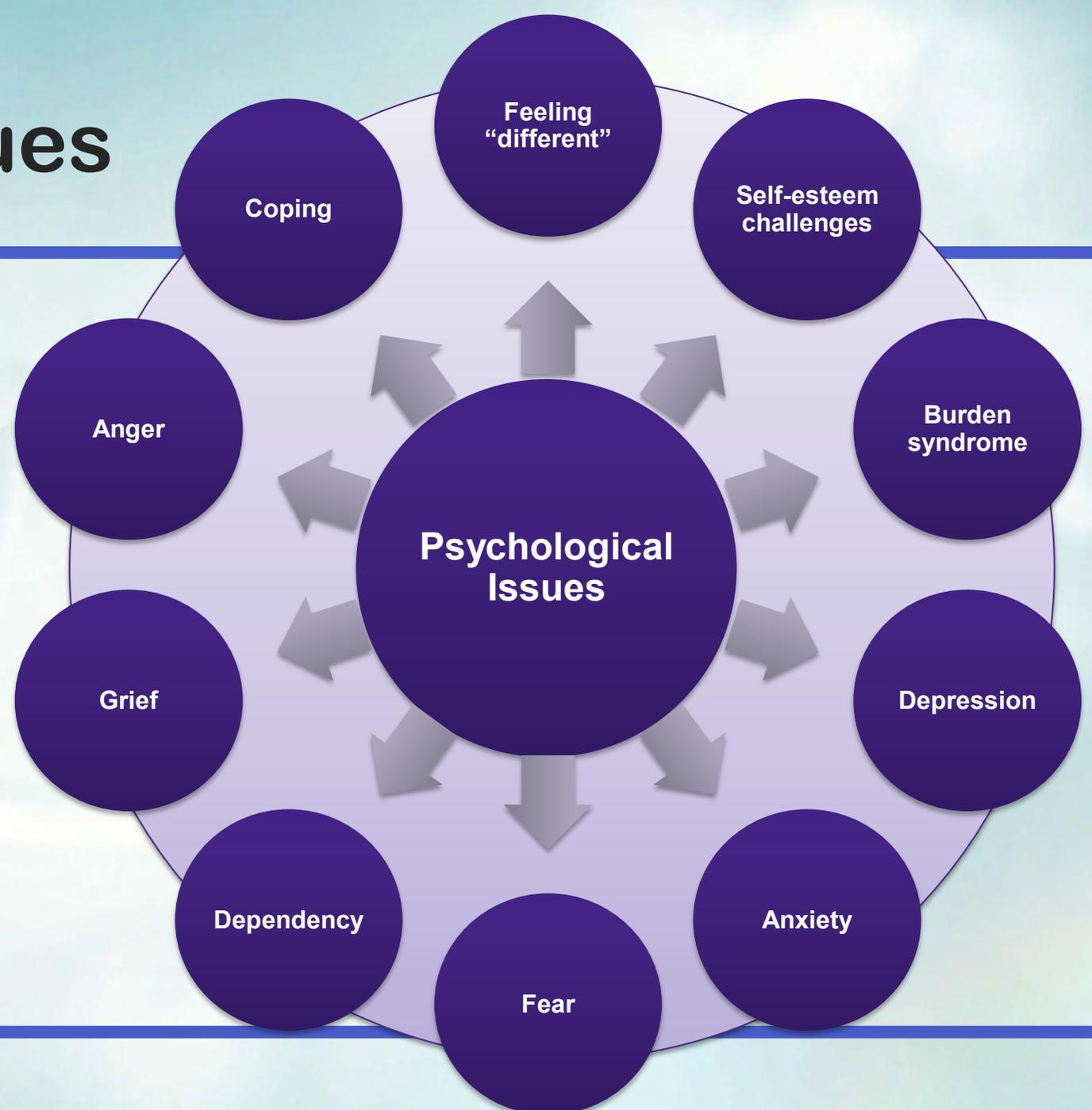


# Psychosocial Considerations (cont.)

- » Financial issues
  - Income challenges/options
  - Health insurance
- » Life needs along the continuum
  - Access to medical/home care
  - Housing, food, etc.
  - Work
- » Life-stage specific needs related to EOL issues
  - Care along the continuum
  - Options/choices



# Psychological Issues



# Management of Mental Health Issues for Adults With NMD

Multidisciplinary team collaboration,  
community resources, education

# Mental Health Screening

## Incorporating mental health assessment into multidisciplinary/interdisciplinary treatment plan

Consider who on the team will manage mental health

- Mental health specialist
- Nurse
- Physician
- Social worker

## Determining what to treat

- Are there underlying or comorbid cognitive issues?
- Mental health history
  - Hereditary issues/diagnoses
- Specifics due to disease (helping to cope with):
  - Physical disabilities
  - Pain from disease or from medical complications/intervention
  - Emotional responses to physical condition



# Mental Health Screening (cont.)

## Initial assessments for mental health history

- Initial mental health intake should include information about:
  - Patient and family mental health history
  - Stressors
  - Support
  - Coping strategies
  - Lifestyle — sleep/diet/self-care
  - Assessment for S/I — assess for LW/AD completion

## Other testing options

- Mini-Mental State Examination (MMSE)/ALS-Cognitive Behavioral Screen (ALS-CBS)/Mini-Cog/Montreal Cognitive Assessment (MoCA)
- Beck Depression Inventory (BDI)
- Patient Health Questionnaire (PHQ-9)
- State-Trait Anxiety Inventory
- Health-related Quality of Life (HRQoL)
- Center for Neurologic Study-Lability Scale (CNS-LS)
- Nottingham Health Profile
- Self-efficacy/motivation scales



AD, advanced directive; LW, living will.

# Mental Health Screening (cont.)

## Ongoing assessments

During each clinic visit and prn — assess for:

- New stressors
- Changes to support network
- Mental health changes (mood/sleep/coping)

## Ongoing education and collaboration

- With multi/interdisciplinary team
  - Discussion among interdisciplinary team members about mental health concerns/ possible barriers to treatment (PT/OT/SP/MD/RN/SW)
- With patient/caregivers/family
  - Education about mental health issues related to:
    - Disease:
      - Signs and symptoms
      - Cognitive/behavioral changes/challenges
    - Stress management/coping
    - Behavior modification strategies
    - Resource options and access to those options



# Life-Stage Specific Needs and End of Life

» Consider specific needs for an adult with NMD:

- Independence (physical/financial) and barriers
- Socialization/intimate relationships
- Reproduction/legacy
- Schooling/employment/career advancement
- Sense of purpose
- Physical care concerns/options along the continuum

» EOL issues:

- As they relate to specific disease process
- Continual education and discussions about individual choices/fears/concerns
- Living will/advanced directive/power of attorney discussions
- Resources — palliative care and hospice options/estate planning

# Mental Health Interventions



Provide a supportive and nonjudgmental environment for open discussion of mental health issues

Discussion and implementation of medical intervention, if deemed appropriate, using resources/referrals to other mental health professionals prn



Connecting patient and care partners with support resources as needed and deemed appropriate:

- Groups/other patients and families/local therapists/stress management tools/socialization/spiritual
- Peer support (NAMI)

# Mental Health Interventions (cont.)



Education about mental health concerns, stress management, and other coping mechanisms, such as behavior modification strategies; access to these tools



## **Life-stage-specific needs: adulthood through EOL**

- Continual discussion about EOL concerns/resources/planning

# Management of Mental Health

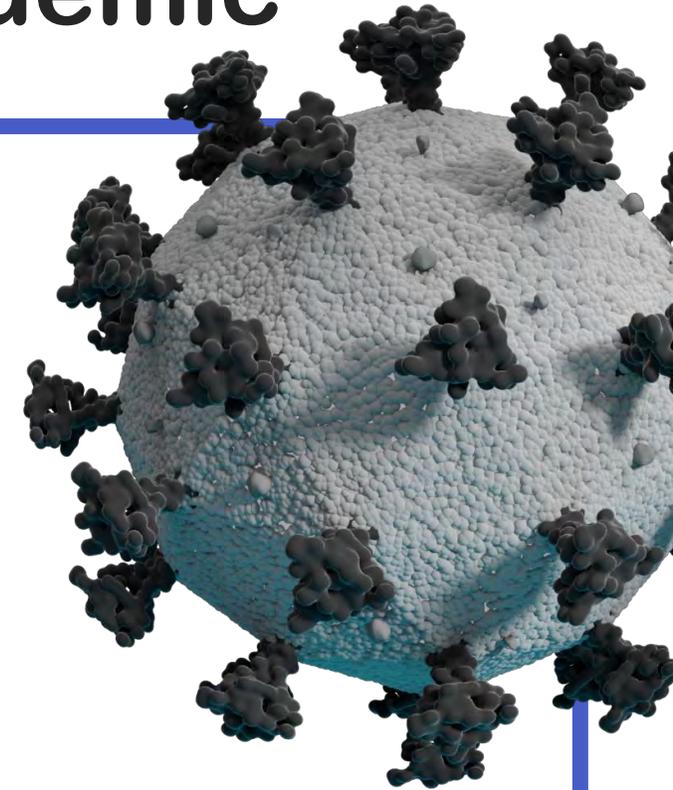
*During COVID-19*

## for Adults With NMD

# Anxiety During the COVID-19 Pandemic

## Anxiety/stress during a pandemic can include:

- Fear and worry about one's own health and the health of loved ones
  - Isolation limiting ability to see family
- Changes in sleep or eating patterns
- Difficulty sleeping or concentrating
- Worsening of chronic health problems
  - Worried about continuing their care for chronic conditions such as MDA - getting to the pharmacy, keeping appointments, using telehealth services, etc.
- Worsening of mental health conditions
- Increased use of alcohol, tobacco, or other potentially harmful substances/coping strategies



People with underlying health conditions might respond differently to the stress related to COVID-19

# Coping During COVID-19

General recommendations for managing anxiety and stress include:



Healthy eating



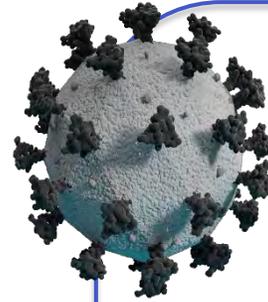
Exercise



Mindfulness and relaxation



Get enough sleep



Recommendations specifically related to COVID-19 include:

- Follow CDC guidelines to keep self safe
- Educate patients on what to do if they start to feel sick
- Take breaks from exposure to news stories
- Talk to trusted people about concerns

[www.cdc.gov](https://www.cdc.gov) has more information and resources

# Summary

- » Mental health is a high unmet need in patients with NMD
- » Mental health can impact physical health of patients
- » Mental health challenges can impact care, long-term functioning and quality of life
- » Specific diseases can increase susceptibility to mental health issues
- » Management of mental health in patients with NMD involves:
  - Continual evaluation
  - Close communication with providers
  - Connecting with the appropriate support resources