

Mental Health Issues in  
*Pediatric*  
Neuromuscular Disease

# Created in Collaboration with



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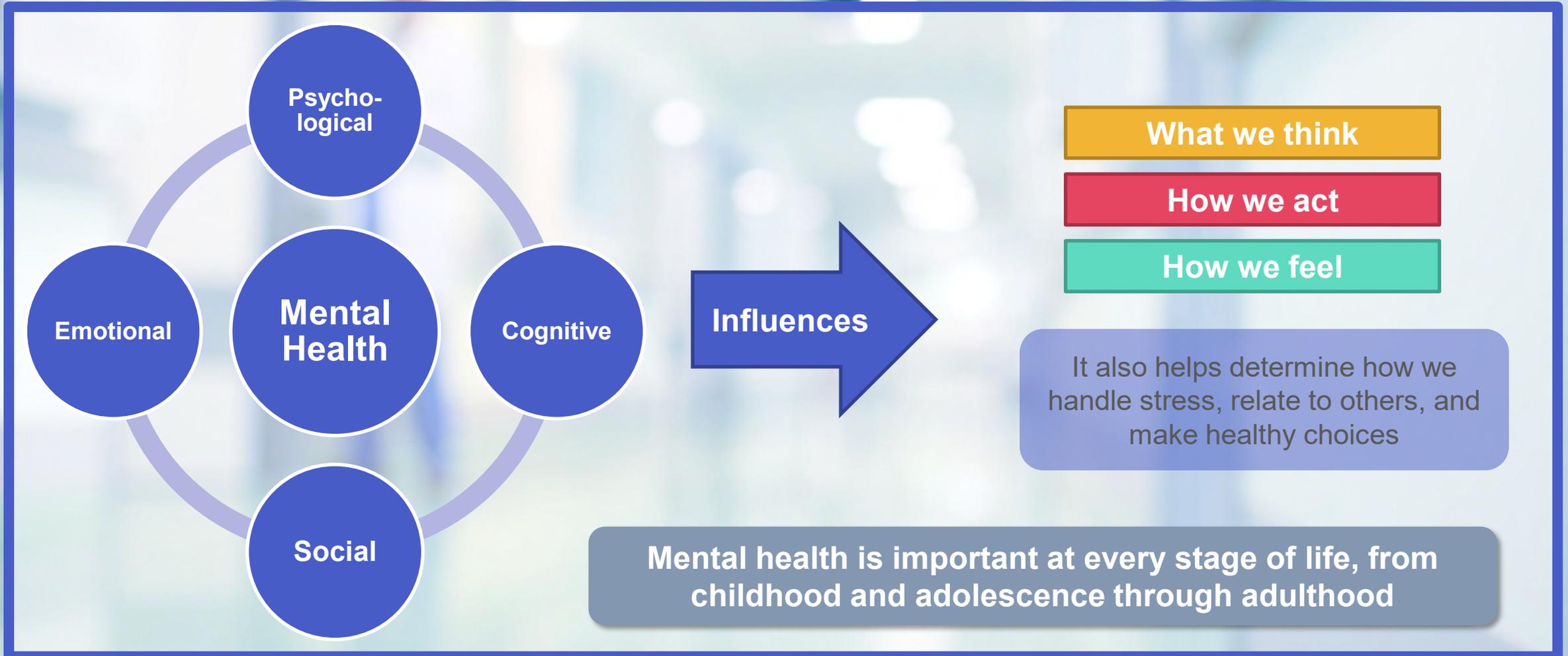
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# Overview of Mental Health in NMD

Varying degrees of cognitive and psychological  
symptoms in NMD

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

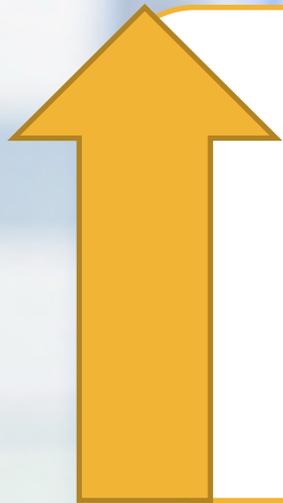


# Mental Health and NMD Patient Care

» 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.

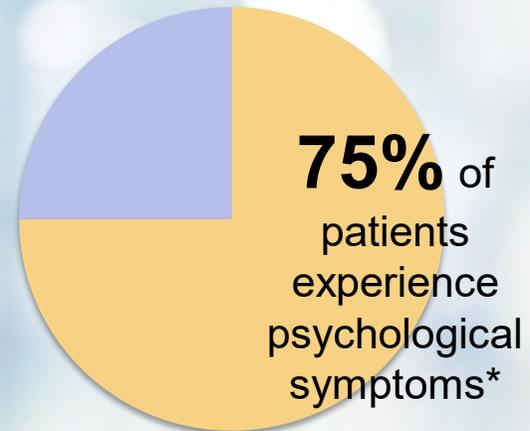
3. National Institutes of Health. Chronic illness and mental health. <https://www.nimh.nih.gov/health/publications/chronic-illness-mental-health/index.shtml>. Accessed June 23, 2020.

# 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

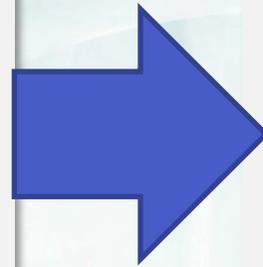
Unmet need in managing psychological symptoms in patients with NMD has been rated **high or medium-high**<sup>2,\*</sup>

\*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.

# Overview of Mental Health in Pediatric NMD

# Mental Health Issues Comorbid With NMD Are Increasingly Being Recognized

The emergence of disease-modifying treatments are **improving and prolonging** the lives of individuals with NMD.

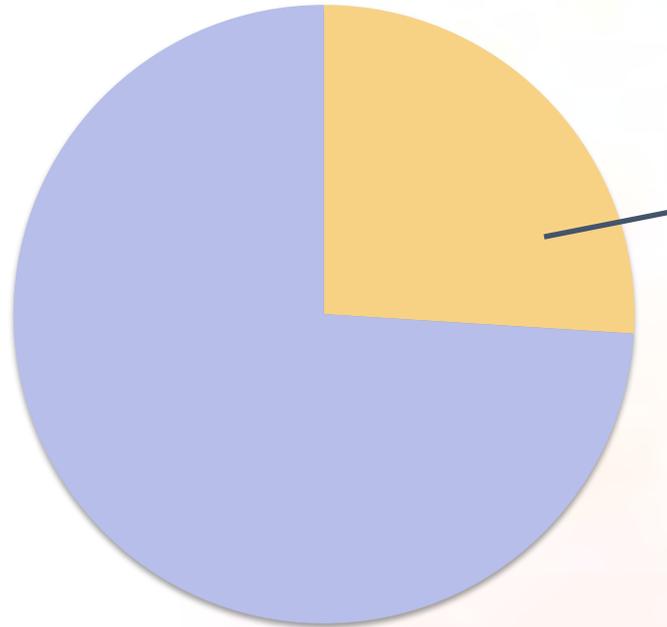


Prolonged lives means that the **transition to adulthood has become more important** than any other time in the care of patients with neuromuscular disease.



# Mental Health in the Pediatric Population With NMD

## Strengths and Difficulties Questionnaires



In one study of British boys with NMD, approximately **26%** scored above the threshold in composite questionnaires, which enquire about children's behavior in emotional symptoms, conduct, hyperactivity, peer problems, and prosocial behavior<sup>1</sup>

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# Impact of Behavioral Health Goes Beyond Just the Patient

## Family and caregivers



- » Family and caregivers of pediatric patients are negatively impacted by behavioral health issues added to NMD
  - Families of children with behavioral, social, and communication problems reported the greatest levels of unmet needs for these services, despite reporting high satisfaction with professionals they were consulting about the child's behavior

Social workers are regularly assessing the psychosocial issues that may get in the way of patients' care, **even if it is not obvious**

# Contributors to Mental Health Issues in NMD

*Pediatric Patients*

# Contributors to Mental Health Issues in NMD

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

**Heterogeneous groups of neuromuscular diseases have been implicated in cognitive alterations**

**Muscular  
dystrophies**

**Mitochondrial  
disorders**

# Dystrophinopathies and Cognitive Impairment

## Duchenne muscular dystrophy (DMD)

- Dystrophin plays an important role in the architectural organization of the CNS
- Can lead to neurobehavioral disorders, including:
  - Attention-deficit/hyperactivity disorder (ADHD)
  - Autism spectrum disorders
  - Obsessive-compulsive disorder (OCD)
- 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

# Myotonic Dystrophy and Cognitive Impairment

## Myotonic Dystrophy (DM1/2)

- DMs are autosomal dominant multisystemic disorders that affect also the brain
- Cognitive deficits have been observed in 24%-75% of patients with DM1 and depends on age of onset of disease
  - **Congenital onset:**
    - Intellectual developmental disabilities
  - **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

# Congenital Muscular Dystrophy

- » Congenital muscular dystrophies (CMDs) are clinically and genetically heterogeneous inherited disorders that compass muscle weakness typically manifested at birth or in infancy
- » Besides muscle compromise, white matter and structural abnormalities of the brain, seizures, cognitive impairment, and eye abnormalities may occur depending on the genetic cause

## Congenital muscular dystrophy and cognitive/visual impairments\*

Fukuyama congenital muscular dystrophy	Cobblestone lissencephaly, visual impairment, and retinal dysplasia
Severe Walker-Warburg syndrome	Cobblestone lissencephaly, microphthalmia, buphthalmos, cataracts
Merosin-deficient congenital muscular dystrophy	Disruption of the blood brain barrier (BBB), resulting abnormal white matter signal intensity

\* Not a complete list.

# Mitochondrial Disorders

## Mitochondrial disorders

- A genetically heterogeneous group and variable phenotype; listed are the most frequent mitochondrial diseases with cognitive impairments:

MELAS

CPEO

MERRF

KSS

MIDD

NARP

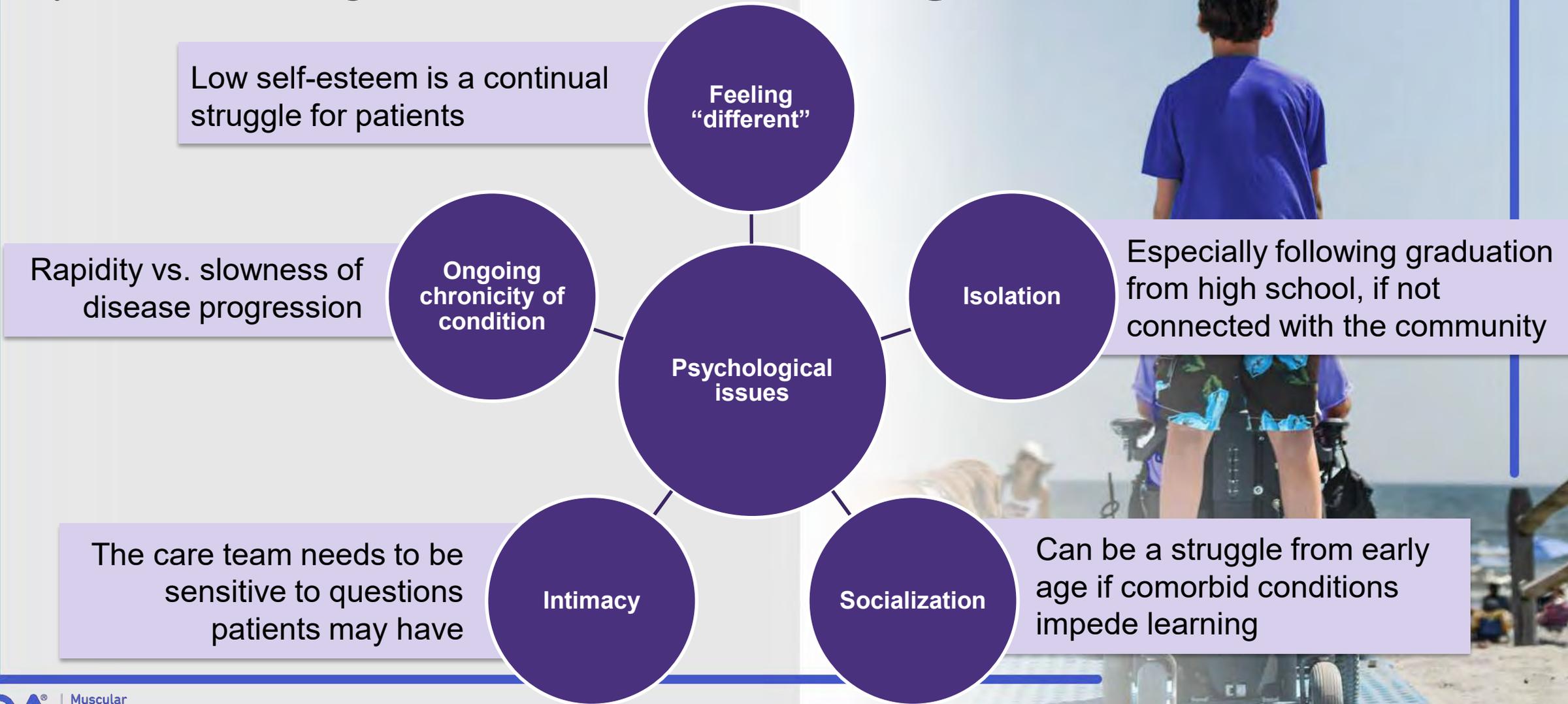
# Mitochondrial Disorders and Cognitive Impairment

**Cognitive function in these patients is evaluated with a neuropsychological test; domains impaired are typically:**

- Abstract reasoning
- Verbal memory
- Visual memory
- Language
- Executive/constructive function
- Calculation, attention, or visuospatial functions

# Unique Aspects of Psychological Care in the Pediatric NMD Patient Population

# Specific Psychosocial Challenges



# Parent and Caregiver Patterns of Help-Seeking

In a survey of patients and caregivers (N=82) of children aged 5 to 13 years with diagnosed NMD:

Parents are **more likely to seek advice** about their children's behavioral, social, and communication difficulties from the educational sector or at the regional muscle clinic than from mental health professionals (such as local community child psychiatrists, clinical psychologists, or mental health nurses)

**Most common issues for which parents sought help:**

- Behavior problems
- Developing child's communication skills



# Management of Mental Health Issues for Children With NMD

# Mental Health Screening and Management

## Determining what to treat

- Are these underlying or comorbid cognitive issues?
- Mental health history
  - Hereditary issues/diagnosis
- Complications due to disease, helping to cope with:
  - Physical disabilities
  - Pain from disease or medical complications/interventions

## Incorporating assessments into multidisciplinary/interdisciplinary treatment plan

- Consider who on the team will manage mental health?
  - Mental health specialist
  - Nurse
  - Physician (neurologist, pulmonologist, cardiologist, etc)
  - **Social worker**
  - **Care manager**



# Mental Health Screening and Management (cont.)

## Initial assessments for mental health history

- Initial mental health intake form should include information on:
  - Patient and family mental health history
  - Stressors
  - Coping strategies
  - Support

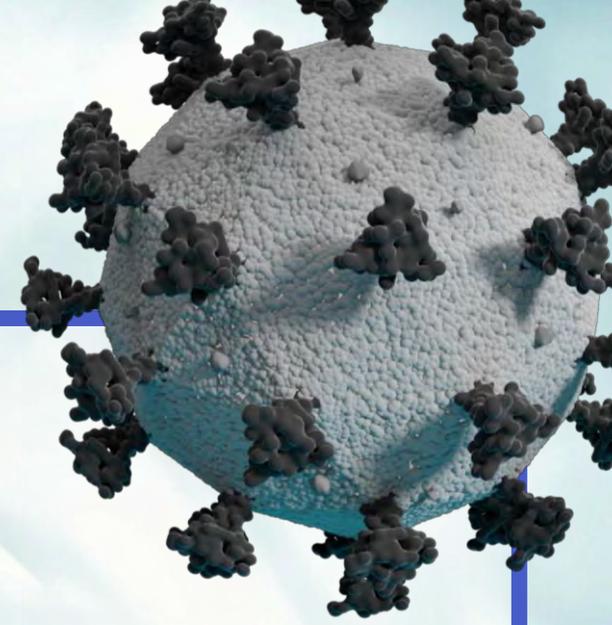
## Many testing options

- A number of cognitive assessments have been developed, which can yield helpful information in diagnosis and management
- Cognitive/behavioral deficits can manifest in a number of ways; because of this, no one test will cover all potential cognitive/behavioral issues



# Management of Mental Health During COVID-19 for Children With NMD

# Helping Children Cope With Changes Due to COVID-19



- Most schools, places of public gathering, and nonessential businesses are closed; parents and other caregivers are faced with helping their families adjust to the new normal which includes:
  - Trying to keep children occupied
  - Keeping children feeling safe
  - Attempting to keep up with schoolwork

**Resources are available to help parents and caregivers:**

**National Association of School Psychologists**

<https://www.nasponline.org/>

**National Federation of Families for Children's Mental Health**

<https://www.ffcmh.org/covid-19-resources-for-parents>

# Signs a Child Is Having Difficulty Coping With the Pandemic

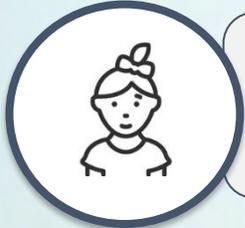
- Most children will manage well with the support of parents and other family members, even if showing signs of some anxiety or concerns, such as difficulty sleeping or concentrating

Some children, however, may have risk factors for more intense reactions, including:



# Signs a Child Is Having Difficulty Coping With the Pandemic (cont.)

» Parents and caregivers should contact a professional if children exhibit significant changes in behavior or any of the following symptoms for more than 2 weeks



## Preschoolers

- Thumb sucking, bedwetting, clinging to parents, sleep disturbances, loss of appetite, fear of the dark, regression in behavior, and withdrawal



## Elementary school children

- Irritability, aggressiveness, clinginess, nightmares, school avoidance, poor concentration, and withdrawal from activities and friends



## Adolescents

- Sleeping and eating disturbances, agitation, increase in conflicts, physical complaints, delinquent behavior, and poor concentration

# What Can Parents and Caregivers Do?

## Be a role model

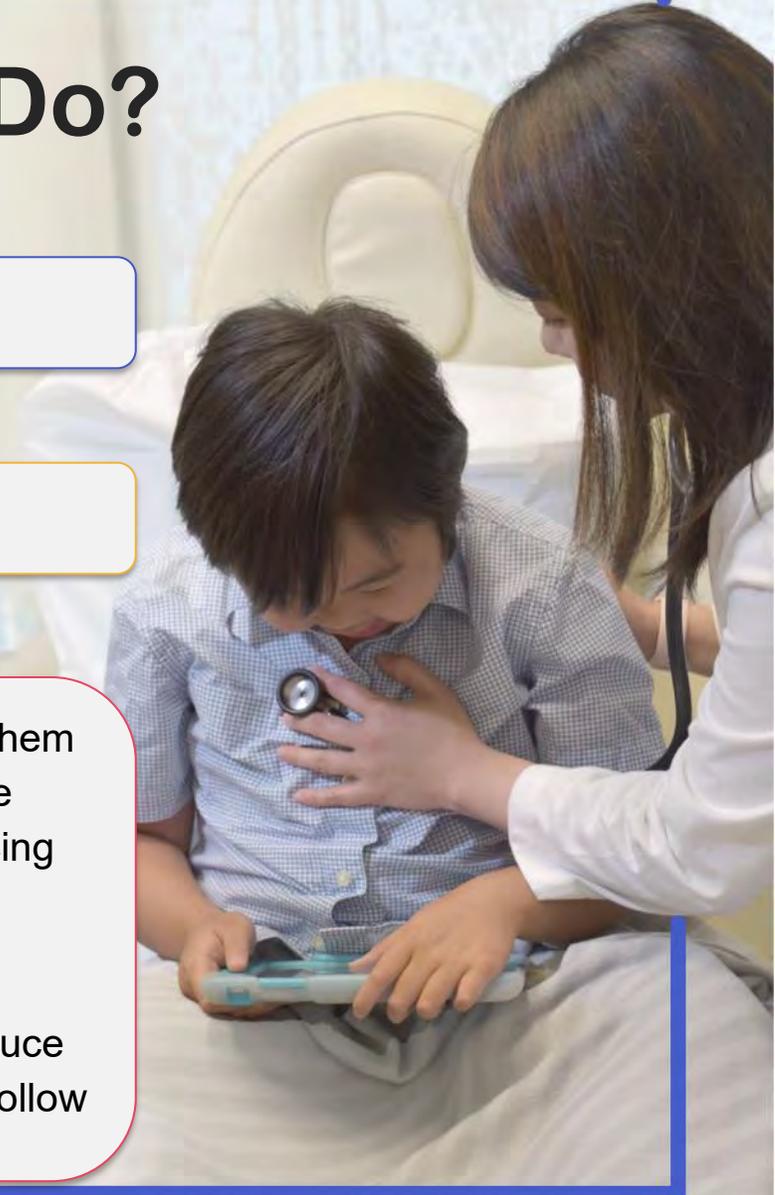
Children will react to and follow your reactions. They learn from your example

## Be aware of how you talk about COVID-19

Your discussion about COVID-19 can increase or decrease your child's fear

## Explain social distancing

- Children probably don't fully understand why parents/guardians aren't allowing them to be with friends. Tell your child that your family is following the guidelines of the Centers for Disease Control and Prevention (CDC), which include social distancing
- Showing older children, the "flatten the curve" charts will help them grasp the significance of social distancing
- Explain that while we don't know how long it will take to "flatten the curve" to reduce the number of those infected, we do know that this is a critical time — we must follow the guidelines of health experts to do our part



# What Can Parents and Caregivers Do? (cont.)

## Demonstrate deep breathing

Deep breathing is a valuable tool for calming the nervous system. Do breathing exercises with your children

## Focus on the positive

Celebrate having more time to spend as a family and try to make it as fun as possible. Do family projects

## Establish and maintain a daily routine

Keeping a regular schedule provides a sense of control, predictability, calm, and well-being



# What Can Parents and Caregivers Do? (cont.)

## Identify projects that may help others

This could include writing letters to healthcare workers or neighbors/others who may be stuck at home alone; sending positive messages over social media; or reading a favorite children's book on a social media platform for younger children to hear

## Offer plenty of love and affection!



# Summary

- » Mental health is a high unmet need in pediatric patients with NMDs
- » Specific NMDs can drive disease-mediated cognitive impairments that cause mental health issues
- » Mental health screening is an important part of management for pediatric patients with NMDs
- » Referrals to appropriate resources is important for optimal management
- » Social workers and mental workers are important for identifying mental health issues and educating patients, families, and all stakeholders (schools, community, etc.)
- » Early detection of mental health issues is important for optimal management and increasing quality of life for patients