# Learning and Behavior

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# Learning and Behavior

- Some evidence that distinctive cognitive profiles and associated learning, behavioral, and emotional disorders exist in DMD.

# Prevalence of Developmental Psychopathologies

- Intellectual Disability (17-27%/40%)
- Learning Disability (26%)
  - Reading disability most common
- Autism Spectrum Disorder (32%)
- ADHD (12%-32%)
- Anxiety (27%)
- OCD (4.8%)

Care Considerations, Part 3, (Birnkrant, et. al., 2018); Pane, et. al. 2012; Snow, et. al., 2013; Battine, 2017)

# Phenotype-Genotype Correlations

- Cognitive impairments in DMD are associated with genetic findings likely to impact dystrophin expression in the brain.
- Mutations that impact Dp140 expression (exon 45-55) have been most reliably linked with cognitive deficits (not necessarily the psychological abnormalities).
- Correspondence of impairment and site of mutation is most significant for children with definitive diagnosis of cognitive delay (e.g. intellectual disability).

(Pane, et. al., 2012; Batinni et.al. 2017; Steele, et. al. 2006)

# Cognitive Concerns for Patients with DMD

(without a diagnosed psychological disorder)

- Problems in planning, directing and intending goal directed behavior found among boys with DMD
- Notable learning differences:
  - Executive Function deficits
    - Working memory deficits
    - Inhibiting responses
    - Difficulty in shifting tasks/sets, multitasking and switching tasks
  - Verbal working memory deficits
    - Verbal fluency deficits
    - Impaired phonological and semantic fluency

# Working Memory (WM)

- Part of our short-term memory capacities
  - WM goes beyond simple rehearsal & memorization
  - WM provides for manipulation and integration of new and previously learned information.
- Responsible for holding verbal and nonverbal information for processing and problem solving
- Critical component in reasoning and decision making
- Upwards of 80% of boys are showing low average WM skills, regardless of overall intellectual functioning (IQ).

# Inhibition

- Ability to “tune out” things that are irrelevant to our current task
- Developmentally sensitive cognitive process
  - Emerges in 3 and 4 year old's and grows in complexity over time
- Significantly impaired among boys with DMD and average intellectual functioning and no ADHD.
- Major symptom in ADHD.
- Often a core impairment in people diagnosed with OCD.

# Response Switch

- Often called task switching or set-shifting
- Ability to non-consciously (automatically) shift attention from one task to another task
  - Allows a child to rapidly and efficiently adapt to new situations
- Note: this is different than more intentional "cognitive shifting" (i.e. a deliberate & conscious change in attention)
- Significantly impaired among boys with DMD and average intellectual functioning.

# General Recommendations

- Short, clear verbal instructions.
- Segment multi-step tasks with shorter, circumscribed instructions and goals.
- Phonologically-based reading curriculums should be used.
- Consider early practice with dictation/speech-to-text software
- Lower threshold for referral to speech therapy.
- Preserve and expand cognitive abilities in anticipation of progression of muscle of pathology limiting physical activities.

# Psychosocial Concerns for Patients with DMD

- Other common emotional concerns could complicate educational management.
- IDD, ASD, and LDs are common.
- ADHD
  - Also known as a common comorbidity
- Obsessive Compulsive Disorder
  - Known to be common among youth diagnosed with DMD
- Anxiety and depression common among children and adolescents with chronic illness

# Intellectual Disability

**Core disability is an overall “slower than normal” developmental and learning rate.**
- Learning and developmental milestones are significantly below age expectations.

- Assessment
  - Often diagnosed earlier in development
  - Psychoeducational testing
  - Early language disorders are common
  - Term “Global Developmental Delay” often used in younger children.
- Impact
  - Learning and communication failures
  - Behavior problems (SIB, aggression)
  - Adaptive impairment & dependency

- Treatment
  - School: accommodations on IEP/504
  - Psychosocial: focus on adaptation
  - Medicines: only for co-occurring symptoms.
- Special Considerations for DMD
  - Interference with treatment regimen
  - Expectations are more limited for adaptation & independence
  - Increased parental & family burden

# Autism Spectrum Disorder

**Core social disability with repetitive and restricted behaviors**

- Joint attention, communication, & social reciprocity represent core set of development impairments.

- Assessment
  - Specialized multidisciplinary team evaluation
  - ASD symptom standardized assessment
  - Psychological and SLP assessment
- Impact
  - Developmental delays
  - Learning differences
  - Disruptive behaviors
  - Socialization impairments

- Treatment
  - Comprehensive developmental intervention using intensive behavioral intervention.
  - Social cognition/skills interventions
  - School: accommodations, aide, intensive learning, structure/transitions
  - Medicines: treat symptoms
- Special Considerations for DMD
  - Can development and overall learning rates be accelerated?
  - Can ASD interventions support expansion of PT interventions?

# Learning Disorders

**Core disability is learning "slower than expected" in reading, math, or writing.**

- Learning in academic areas is behind expectations for child's measured IQ.
- Impairments in phonological and semantic fluency have been reported in DMD boys.

- Assessment
  - Often diagnosed at school
  - Psychoeducational testing
  - Could be evaluated outside of school
  - Early language disorders are common
- Impact
  - Learning failure
  - Executive function deficits
  - Self-esteem, self-efficacy deficits
  - Behavior problems, school avoidance
  - Limited friendships

- Treatment
  - School: accommodations on IEP/504 plans
  - Psychosocial: "get a Master's degree"
  - Medicines: only for co-occurring symptoms.
- Special Considerations for DMD
  - Absences for healthcare limiting access to curriculum.
  - Impacts on treatment engagement and adherence
    - understanding, motivation

# ADHD

**Core disability is one of "intentional behavior"**
- Sustained attention, distractibility, impulsivity, and hyperactivity

- Assessment
  - Often diagnosed in primary care.
- Impact
  - Learning differences
  - Executive function deficits
  - Behavior problems
  - Negative Parent-Child interactions
  - Social immaturity
  - Limited friendships

- Treatment
  - Medicines: psycho-stimulants very commonly prescribed.
  - Psychosocial: parent training
  - School: accommodations on IEP/504 plans
- Special Considerations for DMD
  - Are medicines as effective for ADHD in DMD as in a general ADHD population?
  - Common protective factors limited.

# OCD

**Core disability is characterized by obsessions and/or compulsions that are time consuming and cause significant distress or impairment.**

- **Obsessions are recurrent and persistent thoughts, urges, or images that are experienced as intrusive and unwanted,**
- **Compulsions are repetitive behaviors or mental acts that a person feels driven to perform**

- Assessment
  - Often diagnosed by mental health professionals.
  - Clinical interview and standardized questionnaires.
  - tends to emerge in late childhood or early adolescence
- Impact
  - inflated sense of personal responsibility
  - thought-action fusion, which exacerbate their symptoms.
  - Tics, Trichotillomania, & Excoriation Disorder associated

- Treatment
  - Psychosocial: Exposure and response prevention (EX/RP) is a first-line behavioral treatment
  - Tics, trichotillomania, and excoriation can be treated using behavioral techniques such as self-monitoring and habit reversal training (also with medications like risperidone or guanfacine).
  - Medicines: SSRIs can be effective for OCD (with EX/RP).
- Special Considerations for DMD
  - Important to model flexibility and acceptance early on in treatment and coping with a chronic medical condition.

# Anxiety Disorders

**Core disability is marked by worry, physical symptoms, & avoidance.**

- Generalized anxiety, social anxiety, panic, separation anxiety, selective mutism, and specific phobias

- Assessment
  - Often diagnosed by mental health professionals.
  - Clinical interview and standardized questionnaires.
- Impact
  - Avoidance patterns narrow opportunities and shrink a child's world.
  - Anxiety reinforces itself
  - Anxiety is very treatable
    - Relaxation training with exposure, cognitive restructuring, & interoceptive exposure

- Treatment
  - Psychosocial: preferred, first level intervention is psychotherapy (CBT) & counseling with parent training.
    - Exposure Treatment
  - Group treatments may be effective.
  - Medicines: SSRI's and other medications that help reduce anxiety.
  - CBT with SSRI led to better anxiety reduction than either treatment alone
- Special Considerations for DMD
  - Medical anxiety, medical trauma stress
  - Parents can model and trigger anxiety
  - Reinforces parental over-protection

# Depression and Mood Regulation

**Core disability is a stable (episodic) sad, hopeless, globally negative, irritable, & hostile (or, even elevated/elated) mood.**

- Distinct periods of “abnormal’ mood that are excessive in intensity and duration for the situation.

- Assessment
  - Often diagnosed by mental health professionals.
  - Clinical interview and standardized questionnaires.
- Impact
  - May not talk about sadness – uses terms like empty, bored, feels nothing, cranky, angry, somatic complaints
  - Decline in school performance
  - Anhedonia/social withdrawal
  - Somatic complaints
  - Concerns about suicide risk should be carefully managed.

- Treatment
  - Medicines: SSRIs (flouextine/Prozac)
    - atypical antipsychotics, neuropleptics
  - Psychosocial: CBT, IPT (Interpersonal therapy)
  - Combination treatments most effective.
- Special Considerations for DMD
  - Need to compensate for escalating limitations on activity and social inclusion
  - Deliberately develop family communication strategies about emotions;
    - “Start the Conversation”
    - Talk about your feelings because we don’t wear our heart “on our sleeves”.

# Words for Feelings

- EMOTION: a feeling state involving thoughts, physiological changes, and an outward expression or behavior
  - "Mom/Dad, this is how I feel right now . . . "
- MOOD: *pervasive and sustained* emotion that colors the individual's perception of the world
  - "Mom/Dad, this is how I've been feeling lately . . ."
  - Mood Words: depressed, irritable, anxious, angry, expansive, euphoric, empty, guilty, fearful
- AFFECT: an individual's current emotional responsiveness
  - "How I see you right now, son?"