PSYCHOSOCIAL ISSUES IN DMD

End Duchenne Tour
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OUTLINE

1) Causes
2) Learning Problems
   ▶ General Learning Problems
   ▶ Specific Learning Disorders
3) Neurobehavioral Disorders
   ▶ Externalizing
   ▶ Internalizing
4) Coping
5) What to do about it
POTENTIAL CAUSES

- Psychological
  - Coping with DMD

- Psychosocial Factors
  - Family stress/conflict
  - Parenting
  - Peer interactions
  - Teachers/adults

- Medical factors
  - Steroids
  - Fatigue/sleep
  - Medical procedures
  - Blood sugar
  - Clinical Trials

- DMD impact on brain functioning
FULL-LENGTH DYSTROPHIN (DP 427)

Usually found in different brain areas:
- Cerebral Cortex
- Subcortical structures
- Hippocampus
- Cerebellum – Purkinje Cells
- GABA neurons – primary inhibitory mechanism of brain
- Also glutamate, acetylcholine, NMDA
DYSTROPHIN IN THE BRAIN

- Everyone with DMD is missing Dp427
MISSING DYSTROPHIN IN THE BRAIN

- **Neurons**
  - Ion channels not clustered correctly
    - less efficient in sending signals to each other
    - less ready for new signals
  - Changes in shape and size
  - Fewer neurons
  - Less new “connections”

- **Structural**
  - Blood brain barrier less “tight”
  - Smaller grey matter
  - Less white matter density

- **Functional**
  - Differences in metabolism
  - Increase in choline-containing compounds
  - Higher incidence of abnormal EEG

Knuesel et al., 1999; Vaillend & Billard, 2002; Kueh, Head, Morley, 2008; Doorenweerd et al., 2014
MISSING DYSTROPHIN IN THE BRAIN

- Some may also be missing smaller versions
- 55% of mutation breakpoints occur between introns 45 - 51

Pane et al 2012

Region for Dp140
- Missing Dp140 related to smallest brain grey matter
- Greater risk for cognitive/behaviour problems
Dystrophin in the Brain

Why does my son/daughter with DMD have problems......(fill in the blank)?

Answer: It’s neurological.
GENERAL LEARNING PROBLEMS: INTELLIGENCE

- Most have IQ in the normal range
- Average IQ score lower (80)
- Increased amount in the intellectual disability range (35% below 70)
- Verbal scores lower than visual-spatial scores

Cotton, S., N. Voudouris, and K. M. Greenwood. 2001
GENERAL LEARNING PROBLEMS: LANGUAGE

- Greater chance of delays in language development
- Greater problems in younger children
- Normal vocabulary
- Problems with:
  - complex comprehension and verbal fluency
  - About two years behind

Cyrulnik, S., Fee, R., De Vivo, D.C., & V.J. Hinton. 2007
GENERAL LEARNING PROBLEMS:
SHORT-TERM MEMORY

- Weaknesses in short-term memory
  - Worst for verbal information
  - Pattern seen at all IQ levels and ages

SPECIFIC LEARNING DISORDERS IN DMD

- About 40% have one or more learning disorders:
  - Dyslexia: Difficulty learning to read
  - Dyscalculia: Difficulty learning math
  - Dysgraphia: Difficulty with writing

Hendriksen & Vles 2006
Other Signs of a Learning Disorder

- Anxiety related to specific subject
- Shutting down, overwhelmed
- Frustration, exploding
- Poor performance relative to other subjects
- Says “I hate _______!” (reading, math, writing)
NEUROBEHAVIORAL DISORDERS IN DMD

Attention-deficit disorder: 12% – 44% in DMD (with or without hyperactivity-impulsivity)

Signs to look for:

Impulsive
Blurts things out
Interrupts
Impatient
Fidgets
Easily frustrated
Too loud

Avoids work
Overly focused on fun
Easily distracted
Messy and disorganized
Forgetful
Daydreams
Difficulty following directions

Hendriksen & Vles 2008; Poysky & Lotze, 2008; Hinton et al. 2006; Pane et al. 2012; Ricotti et al. 2015
NEUROBEHAVIORAL DISORDERS IN DMD

Autism: 3-21%

Signs to look for:

- Delayed language development
- Excessive and unusual interests/obsessions and routines
- Impaired understanding of social interactions

NEUROBEHAVIORAL DISORDERS IN DMD

Oppositional, argumentative, & explosive behavior: 15% - 52% of boys with DMD?

- Difficulty controlling emotional reactions
- Easily irritated, angry outbursts
- Blames others
- Hard time predicting consequences
- Punishment increases anger/bad behavior
- Rigid expectations
- Difficulty adjusting to unexpected outcomes

Poysky, Hodges, Lotze – unpublished data; Ricotti et al 2015; Conway et al 2015
NEUROBEHAVIORAL DISORDERS IN DMD

- INCREASED RISK OF ANXIETY
- Generalized Anxiety
  - Worrying, stressing, feeling tense, about what might happen or has already happened
    - May seek reassurance from others repetitively
    - Too hard on themselves for mistakes
    - Keep replaying things in their mind
    - Has to know what is going to happen
  - Easily “thrown off”, agitated, overwhelmed by change in routine or unexpected events
    - Often presents as anger, criticism of others, arguing
    - Associated with being rigid/stubborn in thought process
  - Changes from day to day
NEUROBEHAVIORAL DISORDERS IN DMD

- INCREASED RISK OF ANXIETY
- Social Anxiety
  - Very shy, uncomfortable around people they don’t know well
  - Avoids eye contact, won’t say much, may ignore other’s when they say “What’s up?”
  - Worried about others watching/judging them
  - Afraid they will do/say something embarrassing
  - Avoid/leave situations where they might have to interact
  - Use electronics so they don’t have to have conversations
NEUROBEHAVIORAL DISORDERS IN DMD

- INCREASED RISK FOR ANXIETY

- Obsessive-Compulsive Disorder
  - Rituals and excessive routines
  - Very particular about things being even, lined up, etc.
  - Repetitive behaviors
  - Intrusive thoughts/images
  - Too sensitive to how things feel
“HANGRY” = Hungry + Angry

- Blood sugar starts to drop
  - Angry
  - Irrational
  - Mean/aggressive
  - Emotionally sensitive/labile

- Don’t feel hungry
- Blood sugar may still technically be in the normal range
How are the boys coping?

- Same as boys with other chronic medical conditions
  - Being sad and frustrated *at times* due to DMD is a normal reaction
  - Coping gets better with age
  - Ages 8-10 and adolescence might be extra difficult
  - Some boys may become depressed/distressed

**COPING WITH DMD**

Hendriksen, Poysky, Schrans, Shouten, Aldenkamp, Vles, 2008; Fitzpatrick et al 1986; Liebowitz et al 1981
Many boys and young men not as “independent” as they could be

- Increased focus on transition to adulthood
  - Living independently
  - Making decisions in medical care
  - Employment
  - Romantic relationships*
Social Problems: 34%

- Social skills weaknesses
- Social anxiety
- Teasing/bullying
- Peer inclusion

Peer relationships decline with age

Hinton, Nereo, Fee, Cyrulnik, 2006; Hendriksen et al 2008
DEPRESSION

- 24% have “Internalizing” problems (anxiety and depression) – Ricotti et al 2015
- 19% of adult men with DMD report symptoms of depression – Pangalila 2015
Signs of Depression

- Increased irritability, moodiness
- Loss of interest in fun activities
- More withdrawn (socially), wanting to be alone
- Feeling “blah”, not happy but not sad, “no emotions”
- More negative about things (everything is terrible, not right, not good enough)
- Guilty feelings
- Low self-esteem
- Crying spells
- Changes in appetite (more/less)
- Drop in motivation
- Changes in sleeping (more/less)
Family Adjustment

- Increased rates of parental depression and isolation
- Behavior problems can be as stressful for parents as physical aspects of DMD
- Sibling adjustment

Stress related to clinical trials

LEARNING AND BEHAVIOR TREATMENT RECOMMENDATIONS


David J Birnkrant, Katharine Bushby, Carla M Bann, Susan D Apkon, Angela Blackwell, Mary K Colvin, Linda Cripe, Adrienne R Herron, Annie Kennedy, Kathi Kinnett, James Naprawa, Garey Noritz, James Poysky, Natalie Street, Christina J Trout, David R Weber, Leanne M Ward. Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan. The Lancet Neurology Published online: January 23, 2018

Effective treatments! Same as for people without DMD
Don’t wait if you have concerns
Mental health professional
  - Does not need to be “expert” in DMD
  - Willing to learn from, and listen to, patient, parents and other professionals
  - It is helpful if they have worked with other medical conditions
**Evaluations**

- Neuropsychologist
- School Psychologist
- Special Education Case Manager
- Psychiatrist

There may be more than one problem going on.

Don’t wait to see if he/she will “grow out of it”
LEARNING PROBLEMS: WHAT CAN I DO ABOUT IT?

PREVENTION (so it doesn’t become a problem)
INTERVENTION (trying to fix the problem)
ACCOMMODATION (minimizing the impact)
MODIFICATION (changing teaching and/or evaluation methods)
Psychotherapy

- Parental behavior management training
  - Noncompliance, disruptive behavior, temper meltdowns

- Individual therapy
  - Low self-esteem and depression, anxiety, obsessive-compulsive disorder, coping

- Group therapy
  - Social skills deficits

- Applied Behavior Analysis
  - Autism

NEUROBEHAVIORAL TREATMENT RECOMMENDATIONS
TREATMENT RECOMMENDATIONS

Psychiatric Medication

- Remember, it is neurological!
  - Stimulants or alpha-agonists for ADHD
  - SSRI’s for anxiety, depression, emotional reactivity

Ritalin + Prozac combination becoming more common
ADDITIONAL RESOURCES:

treat-nmd.eu
  TREAT NMD: Family Care Guidelines

parentprojectmd.org
  “DMD Learning and Behavior Guide”  (Poysky)
  “Psychology of Duchenne”  (Hendriksen)

A Guide to Duchenne Muscular Dystrophy: Information and Advice for Teachers and Parents -Janet Hoskin – Editor. (Amazon)

The Explosive Child – Ross Greene