PSYCHOSOCIAL CONCERNS IN DMD

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PSYCHOSOCIAL CONCERNS IN DMD

• OVERVIEW
  • Introduction and causative factors
  • Neurobehavioral Disorders in DMD
  • Coping and Emotional Adjustment
  • Interventions
BEHAVIOR CONCERNS IN DMD

Total = 1284 Families

- No concerns: 836 (65%)
- Don't know: 150 (12%)
- Yes, No Diagnosis: 267 (21%)
- Yes, Diagnosis: 30 (2%)

DuchenneConnect (unpublished data)
POTENTIAL CAUSES OF BEHAVIOR PROBLEMS IN DMD

- Psychological
  - Coping with DMD

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

- DMD impact on brain functioning

- Medical factors
  - Steroids
  - Fatigue/sleep
  - Medical procedures
  - Blood sugar
Dystrophin is not just in muscles
- 6 kinds are also in the brain
  - 3 full versions
    - Dp427
  - 3 smaller versions
    - Dp140
    - Dp116
    - Dp71
DYSTROPHIN IN THE BRAIN

- Everyone with DMD is missing Dp427
FULL-LENGTH DYSTROPHIN (DP 427)

Usually found in different brain areas, including:

- Cerebral Cortex
- Subcortical structures
- Hippocampus
- Cerebellum – Purkinje Cells
- GABA neurons – primary inhibitory mechanism of brain
MISSING DYSTROPHIN IN THE BRAIN

• Neurons
  • Ion channels not clustered correctly
    • less efficient in sending signals
    • 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
NEUROBEHAVIORAL DISORDERS IN DMD

• Brain dysfunction results in recurrent pattern of behavior problems.

• “Nature” > “Nurture”
  • Brain dysfunction results in weaknesses in processing/problem solving skills and/or ineffective/inappropriate behavior.
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

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

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

Poysky, Hodges, Lotze – unpublished data; Ricotti et al 2015
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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

• 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
  • Changes from day to day
  • Associated with being rigid/stubborn in thought process
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 “Wie geht’s?”
  • 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
COPING AND EMOTIONAL ADJUSTMENT
RESPONDING TO THE IMPACT OF DMD ON ONE’S LIFE
COPING WITH DMD

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

Hendriksen, Poysky, Schrans, Shouten, Aldenkamp, Vles, 2008; Fitzpatrick et al 1986; Liebowitz et al 1981
COPING WITH DMD

- 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*
PEER INTERACTIONS IN DMD

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
• **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)
FAMILIES AND DMD

• 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


Mental health specific recommendations under review in Pediatrics (Molly Colvin, PhD)
TREATMENT RECOMMENDATIONS

• 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

• Mental health and quality of life screening at each visit, using standardized questionnaires (PARS-III).

• Meet with mental health clinician at least once a year for evaluation.

• Neuropsychological evaluation at baseline and when concerns are identified.

• Speech/language and Autism evaluations as needed.

• Caregiver coping and resources.
TREATMENT RECOMMENDATIONS

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
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
TREATMENT RECOMMENDATIONS

• Start focusing on independence at diagnosis
  • Talking about DMD: child, peers, teachers, etc.
  • Developing interests and staying involved!
  • Set goals for the future (working, living, social/romantic)
  • Pushing them out of their comfort zone.

• Parents need to take care of themselves too!
  • Get support, get involved, get a break.
  • Don’t forget about brothers and sisters.
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
THANK YOU!!!

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