

Behavior and Learning in DMD

James Poysky, PhD
Clinical Assistant Professor
Baylor College of Medicine

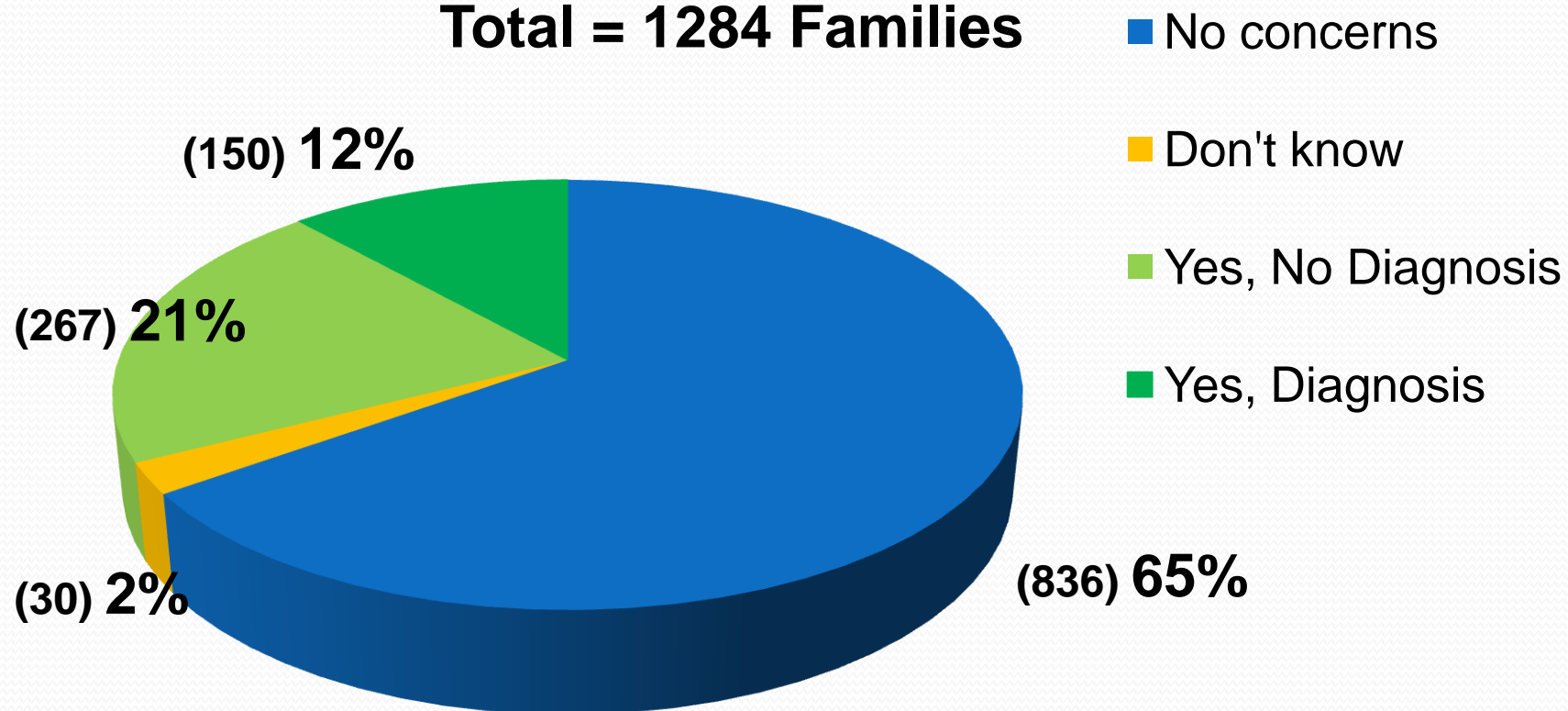
Overview

- How common are behavior and learning concerns in DMD?
- Causes of behavior and learning problems in DMD
- Behavior concerns in DMD
 - Coping with DMD
 - Neurobehavioral disorders
- Learning concerns in DMD
- Treatment recommendations

How Common are Behavior and Learning Concerns in DMD?

Behavior Concerns in DMD

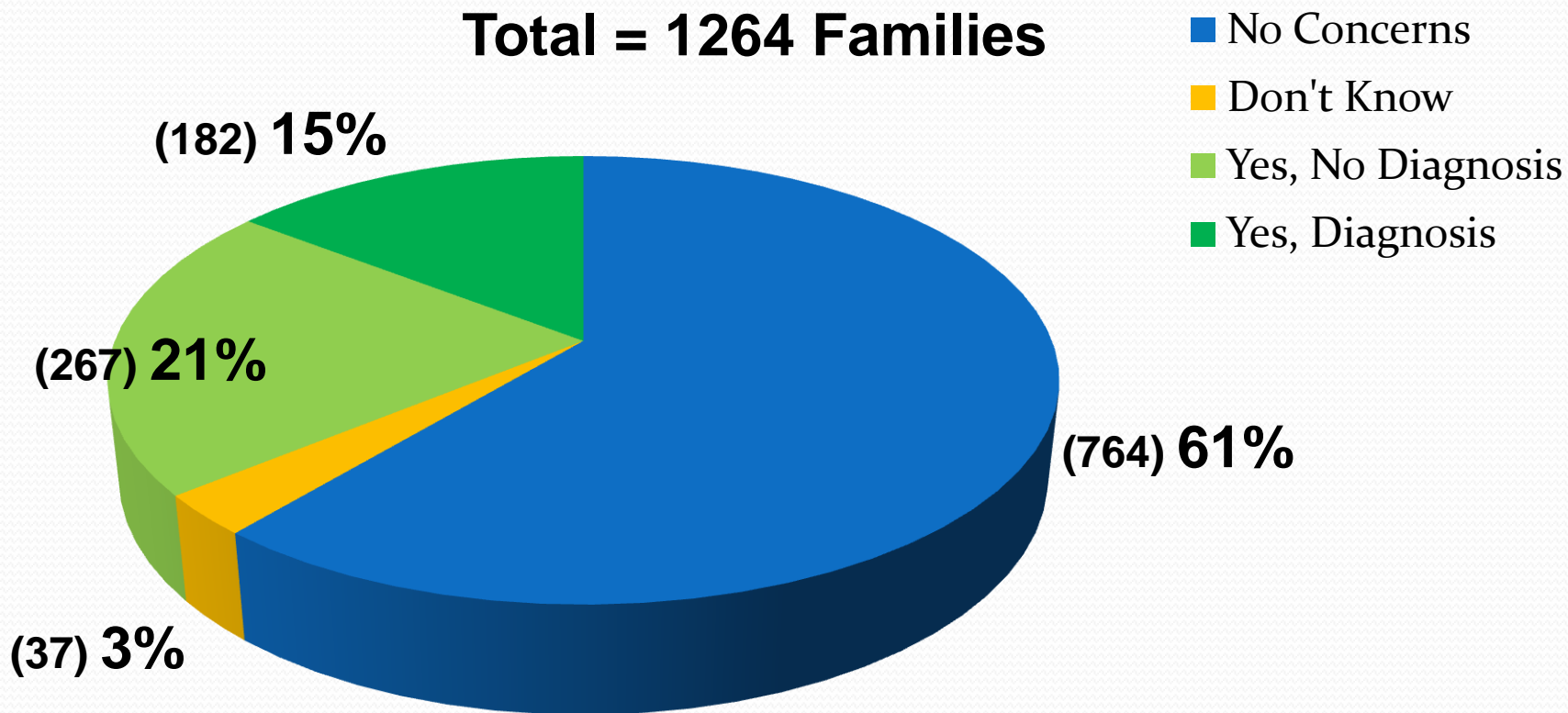
Total = 1284 Families



DuchenneConnect (preliminary data that may change before publication)

Learning Concerns in DMD

Total = 1264 Families



DuchenneConnect (preliminary data that may change before publication)

Causes of Behavior and Learning Problems in DMD

Potential Causes

- DMD impact on brain functioning
- Medical factors
 - Steroids
 - Fatigue
 - Medical procedures
- Psychological
 - Coping skills
 - Disposition
- Social
 - Family stress/conflict
 - Peer interactions
 - Teachers/adults
- Economic resources
- Behavior and Cognitive interactions

Behavior Concerns in DMD

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 might be extra difficult

COPING WITH DMD

- Social isolation can be problematic for all ages
- Peer relationships decline with age
 - Due to increased physical limitations and health problems?
 - Boys on steroids had better peer relationships than those off steroids.

COPING WITH 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

Abi Daoud, Dooley, Gordon 2004; Bothwell , Dooley , Gordon , MacAuley, Camfield 2002; Poysky & Kinnett , 2009; Nereo, Fee, Hinton, 2003

Neurobehavioral Disorders in DMD

Attention-deficit disorder: 12% – 30% 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



Neurobehavioral Disorders in DMD

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

Hard-headed, gets stuck on things

Rigid expectations

Difficulty adjusting to unexpected /unfavorable outcomes

Difficulty controlling anger

Blames others

Difficulty anticipating consequences

Doesn't learn from mistakes

Punishment escalates behavior

(Poysky, Hodges, Lotze – unpublished data)



Neurobehavioral Disorders in DMD

Increased risk of:

- Anxiety
 - Worries
 - Excessive fears
 - Avoids new situations or people
 - Fearful of being alone
- OCD
 - Rituals and excessive routines
 - Very particular about things being even, lined up, etc.
 - Repetitive behaviors
 - Intrusive thoughts/images

Neurobehavioral Disorders in DMD

Autism: 3-19%



Signs to look for:

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

Wu et al. 2005; Hendriksen & Vles, 2008; Darke, Bushby, Le Couteur, McConachie, 2006; Hinton et al 2006

Neurobehavioral Disorders in DMD

Social Problems: 34%

Signs to look for:

Immaturity

Overly dependent

Social skills deficits



Hinton, Nereo, Fee, Cyrulnik, 2006

Learning in DMD

Cognitive Skills

- Increased risk for weaknesses in:
 - IQ
 - Language development
 - Short-term memory
 - Social reasoning (perspective-taking, social judgment, “reciprocity”)
 - Executive functioning (flexibility, planning, organization, insight)
 - Attention/Impulse Control
 - Fine Motor Skills?

Cotton, Voudouris, Greenwood 2001; Hinton, De Vivo, Nereo, Goldstein, Stern 2000; Hinton, De Vivo, Nereo, Goldstein, Stern 2001; Cyrulnik, Fee, De Vivo, Goldstein, Hinton 2007; Hendriksen, Vles 2006; Hinton, Nereo, Fee, Cyrulnik 2006; Cotton, Crowe, Voudouris 1998; Wicksell, Kihlgren, Melin, Eeg-Olofsson 2004; Donders, Taneja 2009

Learning Disorders

40% may have a learning disorder despite normal intelligence.

Dyslexia: Difficulty learning to read

Dyscalculia: Difficulty learning mathematics

Dysgraphia: Difficulty with writing



Treatment Recommendations

Bushby K, Finkel R, Birnkrant DJ, Case L, Clemens P, Cripe L, Kaul A, Kinnett K, McDonald C, Pandya S, Poysky J, Shapiro F, Tomezsko J, Constantin C, DMD Care Considerations Working Group. The diagnosis and management of Duchenne muscular dystrophy – part 1. Diagnosis, pharmacological and psychosocial management. *Lancet Neurology* 2010;9(1):77-93.

Treatment Recommendations

- Effective treatment options!
- Same interventions as non-DMD kids.
- Early interventions work best.

Treatment Recommendations

Recommended testing/assessments

- Neuropsychological/developmental
 - All kids with DMD at diagnosis or prior to starting school
- Language and autism
 - If concerns arise
- Emotional/coping screening
 - Annually, in clinic

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

Coping/Social interventions

- Promoting patient independence and self-advocacy
- Talking about DMD: child, peers, teachers, etc.
- Developing interests and staying involved!
 - Modified/adapted sports, summer camps, and youth groups/programmes
 - Art groups, equestrian, and aqua therapies, use of service dogs, nature programmes, and internet/chat rooms, etc.

Treatment Recommendations

Educational interventions

- Learning needs
- Behavior needs
- Modifying potentially harmful activities (physical education)
- Saving energy
- Safety (e.g., climbing on playground)
- Private tutoring

Treatment Recommendations

Psychiatric Medication

- For moderate to severe problems



jamespoysky@gmail.com

PPUK Toolkit

Education Matters: Learning and Behavior
Guide

(parentprojectmd.org)