

# Social Behavior Problems in Boys with Duchenne Muscular Dystrophy

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**ABSTRACT.** Duchenne muscular dystrophy (DMD) is a chronic, progressive pediatric disease that affects both muscle and brain. The objectives of the study were to examine parent reported behavior in children with DMD, investigate the influence of chronic illness, intellectual ability and etiology on behavior, and determine whether a specific behavioral profile is associated with DMD. Parental ratings of boys with DMD ( $n = 181$ ) on the Child Behavior Checklist behavior scales were examined and compared to reported findings of children with other chronic illnesses, unaffected siblings of boys with DMD ( $n = 86$ ), and children with cerebral palsy (CP) ( $n = 42$ ). Increased ratings of general behavior problems were reported, and neither physical progression nor intellectual level contributed to behavioral ratings. Among the children with DMD, the Social Problem behavior scale had the greatest number of "clinically significant" ratings (34%). Between-group comparisons showed significantly more boys with DMD were rated as having social behavior problems than either the sibling or CP comparison groups. In addition to the increase in reported behavioral problems likely related to the effects of chronic illness, boys with DMD may be at heightened risk for specific social behavior problems. The specificity of the findings of the behavior profile in DMD may be partially due to the lack of dystrophin isoforms in the central nervous system, and not solely a reactive response to the illness. *J Dev Behav Pediatr 27:470-476, 2006.* Index terms: *Duchenne muscular dystrophy, behavior, behavioral phenotype, social problems.*

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Duchenne muscular dystrophy (DMD) is a pediatric medical condition that presents with progressive physical disability and a shortened life span. DMD is primarily a disease of muscle, yet it also affects the brain. Individuals with DMD have a genetic mutation that prevents the production of the protein product dystrophin and of multiple dystrophin-like isoforms. In muscle, lack of dystrophin results in unstable muscle cell membranes that break down over time, causing progressive weakness. In the brain, a lack of dystrophin isoforms has been documented in the cerebral cortex and cerebellum, in specific cell types (especially pyramidal and Purkinje cells) and in specific cell areas

(especially the neuronal postsynaptic densities).<sup>1,2</sup> The exact functional role of dystrophin isoforms in the brain is unknown, but their absence is associated with a downward shift in I.Q.<sup>3</sup> Most affected boys present with specific mild cognitive deficits, characterized by poor verbal immediate memory and academic deficits.<sup>4,5</sup> The extent of the cognitive involvement is highly variable across individuals, but is not associated with physical severity, nor does it appear to be progressive. We have argued that the cognitive profile represents the brain's role in the illness and is not primarily due to consequences of the physical disability.<sup>6,7</sup> The goal of the current study was to examine behavioral traits reported by mothers of boys with DMD to determine whether there are specific characteristics associated with the illness.

Individuals with chronic medical conditions or physical disability have been shown to be at increased risk of behavioral problems. Chronically ill or physically disabled children are more likely to have significant internalizing

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and adjustment problems and more somatic complaints than their healthy peers, and there is little evidence of diagnosis-specific behavioral profiles among chronic illnesses.<sup>8-14</sup>

Increased presentation of behavior problems has also been noted in individuals with learning disabilities and mild cognitive impairment.<sup>15,16</sup> Generally, it is thought that the behavioral problems are a reactive result of the stress of the condition. However, it is also possible that the behaviors are a comorbid manifestation of the underlying etiology.

Indeed, the study of behavioral phenotypes of disorders of known etiology has increased dramatically over the past years. Understanding of behaviors that appear to be associated with developmental disabilities of known etiology such as fragile X or Prader-Willi syndrome has allowed for the conceptualization of how the underlying causal factors of the disorders may have contributed to the affected individuals' presenting behavior.<sup>17,18</sup> In DMD, where the cognitive involvement is generally milder than in the aforementioned syndromes, evidence indicative of the genetic mutation's effect on brain development provides a model system to study behavioral phenotypes.

For boys affected with DMD, each of the above conditions, physical disability/chronic illness, cognitive impairment and underlying etiology, may potentially contribute to behavioral phenotype. Prior work examining behavior in children with DMD has indicated two general areas which the boys appear to be susceptible to: internalizing/depressive disorders and social problems. Studies using projective measures and no comparison groups indicated few depressive disorders, some social immaturity and insecurity, and anxiety.<sup>19,20</sup> Data collected using parent questionnaires documented depressive signs that were associated with age and less well-characterized antisocial tendencies.<sup>21-23</sup> Data collected from child self reports indicated social inhibition to be the dominant behavioral feature.<sup>24</sup>

Previous research examining behavior in DMD has not directly examined the relationship of cognitive level and behavior, nor has it examined behavior in children with DMD compared to their unaffected siblings to control for background variation and family functioning. Further, most work in this area was done prior to understanding of direct brain involvement. None of the available literature on behavior of boys with DMD hypothesized that there may be a behavior profile that is characteristic of the underlying brain pathology.

The goal of the current work is to investigate behaviors associated with DMD, and to examine the influences of general physical and cognitive ability and underlying etiology to the behaviors reported. First, we examine behavior ratings in a large sample of boys with DMD, and evaluate of the possible contributing influences of disease progression and intellectual level on reported behavior. Next, to control for environmental and rater influences, we compare behavior of boys with DMD to that of their unaffected siblings. Third, to control, in part, for the effects of developmental motor disability, we compare the behavioral ratings of boys with DMD to those of children with cerebral palsy (CP).

We hypothesize that in children with DMD there will be an increased level of general behavior problems relative to

controls that is accounted for by the combined chronic illness/physical and intellectual disability aspects of illness. We hypothesize, based on prior research, that boys with DMD will be at increased risk of somatic, depressive, and social behavioral problems. Additionally, independent of prior published reports, and fueled by clinical experience with the children, we hypothesize that there will be some observed behaviors that are not due solely to the generalized effects of physical and intellectual impairment. We suggest that given the involvement of the CNS in development that there may be behavioral characteristics that are associated with DMD, and are not due to reactive responses to the illness.

## METHOD

### Participants

*Duchenne muscular dystrophy probands.* One hundred eighty-one boys with Duchenne muscular dystrophy (DMD) were studied. As part of a large ongoing neuropsychological study of DMD, participants were recruited from private physicians associated with the Muscular Dystrophy Association clinics and through announcements and mailings through the Muscular Dystrophy Association and the Parent Project Muscular Dystrophy. All participants were male, between 6 and 16 years of age, and in otherwise good general health; spoke English; and were willing to participate. Diagnosis of DMD was based on clinical onset of progressive weakness before 5 years of age, elevated serum creatine kinase levels, and either molecular assessment of mutation in the DMD gene or muscle biopsy that was deficient in dystrophin and compatible with DMD. Only one affected child from each family was included. Sixty-four boys were in wheelchairs at the time of testing, 117 were still walking. Sixty-two children reported taking steroid medications regularly. Mothers of all participants reported having a high school or above level of education. The majority self-reported they were white (88%), and there were fewer Hispanic (7%) and African American or Asian (3% each). Cognitive data on many of these children have been published previously.<sup>5-7</sup>

*Probands versus siblings.* Where possible, one healthy sibling without DMD was also recruited for each proband. Selection criteria included 6 to 16 years old, age within 5 years of the proband's age, good general health, English as primary language, and willingness to participate. Where more than one control participant was available, preference was given first to male gender and then to closeness of age. A total of 86 siblings met these criteria and participated. All sibling pairs were from separate families. Forty control participants were male and 46 participants were female.

*Duchenne muscular dystrophy versus CP.* Because of the presumed contributing effects on behavior of having a developmental motor disability, data from 42 children diagnosed with CP were included as a comparison sample. All these children were enrolled in an ongoing study examining the school-age cognitive outcome of children born at very low birth weight (i.e., <1500 g) as described in Arad et al.<sup>25</sup> These children were originally enrolled at

**Table 1. Participant Characteristics**

Analysis	Participants	Age, yr		PPVT-R		Ravens SS		CBCL Total T	
		Mean	SD	Mean	SD	Mean	SD	Mean	SD
DMD	181 DMD	9.03	2.95	101.14	20.57	99.79	13.03	55.29	11.40
DMD: siblings	86 DMD	8.93	2.51	101.89	7.04*	99.11	12.57*	53.63	11.12*
	86 siblings	9.75	3.17	109.39	5.63*	106.30	10.83*	45.11	10.76*
DMD: brothers	40 DMD	9.25	2.61	102.95	7.42*	100.52	13.03*	52.12	11.86*
	40 brothers	10.05	3.17	113.83	3.36*	108.15	10.29*	48.53	11.07*
DMD: CP (6–7 yr)	51 DMD	6.60	0.49	99.68	17.23			56.16	10.42*
	42 CP	6.40	0.63	95.24	16.15			49.29	11.15*

\*Between-group *t* test,  $p < .05$ .

DMD, Duchenne muscular dystrophy group; CP, cerebral palsy group; PPVT-R SS, Peabody Picture Vocabulary Test standard score; SS, standard score; CBCL Total, Child Behavior Checklist Total T score.

birth in the Developmental Epidemiology Network cohort.<sup>26</sup> Each child received a standardized and reliable neurological evaluation from an experienced pediatric neurologist who made a diagnosis of whether the child had cerebral palsy (CP).<sup>27</sup> The children with CP were between 6 and 7 years old at the time of participation, and all were ambulatory. Twenty-two participants were male and 20 were female. All participants were from separate families. The CP children were compared to all the boys with DMD who were of comparable age (between 6 and 7 years old). Fifty-one children with DMD were included. The majority of children with DMD were ambulatory (only six of the 51 were in wheelchairs). Maternal levels of education were comparable between the groups, with all mothers having completed high school. The majority of participants in each group were white (DMD: 88%, CP: 75%), with fewer Hispanics (DMD: 8%, CP: 7%) and African Americans (DMD: 4%, CP: 18%).

## Procedure

The present study was approved by the home institution's Institutional Review Board. Prior to data collection, parents of all participants provided written informed consent and all participants gave verbal assent. Parents completed questionnaires while their children were being tested.

## Measures

As estimates of intellectual function, participants were individually administered a version of the Peabody Picture Vocabulary Test revised (PPVT-R),<sup>28,29</sup> an untimed test of receptive vocabulary, and the Ravens Coloured Progressive Matrices,<sup>30</sup> a nonverbal untimed spatial reasoning task. Both tests are appropriate for use across a wide age range as well as range of intellectual function and do not involve any significant motor response that might confound performance among the physically disabled children. Raw scores are converted to age-referenced standard scores with a mean of 100 and SDs of 15. Ravens data were not collected on the CP sample.

Parents completed the Child Behavior Checklist (CBCL),<sup>31</sup> a widely used measure that consists of 118 problem behaviors. Parents rate, on a 0 (never) to 2 (very much) scale, how often their child engages in each behavior. The CBCL

yields a Total T score and two factor scores (Internalization and Externalization). Additionally, the CBCL yields eight narrow band subscales of problem behaviors (Withdrawn, Somatic Complaints, Anxiety/Depression, Social Problems, Thought Problems, Attention Problems, Delinquent Behavior, Aggressive Behavior). For the purpose of these analyses, the eight Problem Scales were used to examine specificity of reported behavior profiles. Content validity of the scales was established using a T score cut-point of 67 (or at the 96th percentile) that effectively discriminated between matched clinically referred and nonreferred samples.<sup>31</sup>

## Data Analysis

For the different groups, mean ages, PPVT-R standard scores, Ravens scores, and CBCL Total T scores were calculated. Participant characteristics are presented in Table 1.

*Duchenne Muscular Dystrophy Probands.* To determine whether some problem behaviors are selectively reported in DMD, the percentage of subjects scoring in the "clinically significant" range on each of the eight problem behavior scales was determined. Because the CBCL is a continuous scale designed to examine behaviors that normally occur across populations, a cutoff at the 96th percentile (or a T value of 67) was set for the eight behavior problem scales. This was based on the finding that this value was sufficient to discriminate between

**Table 2. CBCL Subscale T Scores in 181 Boys with DMD**

CBCL Subscale	% T score ≥ 67	T Score Range	T Score	
			Mean	SD
Social Problems	34	50–83	61.13	9.04
Attention	24	50–86	59.09	8.46
Withdrawn	22	50–81	58.21	8.00
Thought	22	50–82	57.24	7.82
Anxiety/Depression	17	50–94	56.75	8.45
Somatic	17	50–82	56.39	7.32
Aggressive	12	50–89	54.71	7.33
Delinquent	6	50–70	53.12	4.74

DMD, Duchenne muscular dystrophy group; CBCL, Child Behavior Checklist.

**Table 3. No Influence of Age or Intellectual Ability on Total Problem Scores from the Child Behavior Checklist for Boys with Duchenne Muscular Dystrophy ( $F_{4,155} = 0.47$ )**

Model	Beta	t	p
Constant		6.07	.00
Age	0.01	0.13	.90
PPVT-R SS	-0.08	0.87	.39
Ravens CPM SS	0.11	1.28	.20
Wheelchair use	-0.02	0.17	.86

PPVT-R SS, Peabody Picture Vocabulary Test Revised Standard Score; Ravens CPM SS, Ravens Coloured Progressive Matrices Standard Score.

referred and nonreferred samples while minimizing the number of false positives.<sup>31</sup>

To determine whether physical ability or intellectual level influenced reported behavior among the boys with DMD, a linear regression was run. The effects of age (which in DMD also serves a proxy measure for disease progression), wheelchair use, PPVT-R, and Ravens performance was examined on the CBCL Total T score.

*Duchenne muscular dystrophy versus siblings.* To control for potential rater and environmental confounds, proband behavior ratings were compared to those of their unaffected siblings. Two sets of analyses were done. First, matched proband-sibling pairs (n = 86) were examined, followed by proband-brother pair comparisons to control for potential effects of gender (n = 40). Paired t tests were calculated for age, PPVT-R, Ravens standard scores, and the CBCL Total T score to examine group characteristics. To determine whether the likelihood of scoring above cutoff on each problem behavior scale was similar for the proband and sibling groups, eight chi-square analyses were run. Alpha was adjusted using the Bonferroni method (.05/8) and set at .006.

*Duchenne muscular dystrophy versus cerebral palsy.* To control for potential motor confounds, proband behavior ratings were compared to those of a group of children diagnosed with CP. Since children in the CP (n = 42) group were between 6 and 7 years old, only boys with DMD who were of comparable age were included (n = 51). Independent t tests were calculated for age, PPVT-R, and Ravens standard scores, and CBCL Total T score to examine similarity of the groups. Eight chi-square analyses were run to examine between-group differences on CBCL

problem behavior scales. Alpha was adjusted using the Bonferroni method (.05/8) and set at .006.

**RESULTS**

**Duchenne Muscular Dystrophy Probands**

Examination of problem behavior scales indicated that among the individuals with Duchenne muscular dystrophy (DMD), some scales were more likely to fall in the “clinically significant” range than others. For the group of 181 probands, the percentages of participants who scored above the T score  $\geq 67$  cutoff for each problem behavior scale are presented in Table 2. Most notable is the finding that 34% (or 64 individuals) of the DMD sample had significantly elevated Social Problem scores. In addition, 24% had elevated Attention scores, and 22% had elevated Withdrawn and Thought Problem scores.

Neither disease progression (as indicated by age and wheelchair use) nor intellectual level (as measured by PPVT-R and Ravens tests), contributed significantly to reported general elevated behavior problems (Table 3). Linear regression analysis indicated that the contributions of age, wheelchair use, PPVT-R and Ravens test performance on the CBCL Total T scores were not statistically significant, ( $df = 155, 4; F = 0.47$ , not significant).

*Probands Versus Siblings.* Comparison of probands to unaffected siblings and to unaffected brothers using paired t tests showed that the matched groups differed on measures of intellectual estimates and total behavior problems, but were comparable in age (Table 1).

Comparisons of 86 boys with DMD to their unaffected siblings demonstrated the boys with DMD were more often rated as falling in the “clinically significant” range on the Social Problem scale ( $\chi^2 = 16.82, p = .000$ ) and the Attention scale ( $\chi^2 = 10.40, p = .001$ ) (Table 4). No other between-group differences were observed. When only probands with brothers were examined (n = 40), no between-group comparison was found to be significant at an alpha of .006, although the Social Problem scale had the greatest between-group difference (20% vs 2%,  $\chi^2 = 6.13, p = .01$ ).

*Duchenne Muscular Dystrophy group Versus Cerebral Palsy.* No differences were observed between boys with DMD and children with CP on age or PPVT-R scores (Table 1).

**Table 4. Comparison of Percentage of Clinically Significant CBCL Subscale T Scores in Boys with DMD Versus Siblings**

CBCL Subscale	86 DMD % T > 67	86 Siblings % T > 67	$\chi^2$	40 DMD % T $\geq$ 67	40 Brothers % T $\geq$ 67	$\chi^2$
Social Problems	26	6	16.82*	20	2	6.13
Attention	22	5	10.40*	13	2	2.88
Withdrawn	16	7	3.30	17	10	0.95
Thought	17	6	5.16	15	5	2.22
Anxiety/Depression	15	6	3.59	13	7	0.56
Somatic	13	3	5.87	5	0	2.05
Aggressive	7	9	0.07	5	10	0.72
Delinquent	6	9	0.33	2	7	1.05

\* $p \leq .006$ .

DMD, Duchenne muscular dystrophy group; CBCL, Child Behavior Checklist.

**Table 5. Comparison of Percentage of Clinically Significant CBCL Subscale T Scores in Children with DMD Versus CP**

Problem Scale	DMD ( <i>n</i> = 51)	CP ( <i>n</i> = 42)	$\chi^2$
	% T $\geq$ 67	% T $\geq$ 67	
Social Problems	46	14	14.24*
Attention	33	17	5.13
Withdrawn	23	5	0.01
Thought	25	10	4.96
Anxiety/Depression	9	5	0.90
Somatic	14	5	2.99
Aggressive	11	3	3.02
Delinquent	5	7	0.05

\* $p \leq .006$ .

DMD, Duchenne muscular dystrophy group; CP, cerebral palsy group; CBCL, Child Behavior Checklist.

Table 5 shows the DMD and CP groups differed only on the social problems scale ( $\chi^2 = 14.24$ ,  $p = .000$ ), with more participants in the DMD group (46%) scoring above the T  $\geq$  67 cutoff than in the CP group (14%).

## DISCUSSION

These data of parental reports of behavior of boys with DMD demonstrate that (1) boys with Duchenne muscular dystrophy (DMD) have an increased number of behavioral problems; (2) neither age (a proxy for illness progression), wheelchair use, or overall general estimates of intellectual level add significantly to the report of behavioral problems; and (3) the behavioral profile associated with DMD is characterized, in part, by report of increased social behavior problems.

As expected, the boys in the sample were reported to have higher rates of general behavior problems than normative data and than their unaffected sibling controls. Further, the elevation in scores on the Child Behavior Checklist (CBCL) was concentrated in some domains and was not spread equally across the scales. When the CBCL data from boys with DMD are examined by problem scale with stringent cutoffs, parents report a substantial number of social behavior problems among their sons with DMD, and this is the most significant behavior finding in the group. Thirty-four percent of the boys with DMD were rated by their parents as above the cutoff (i.e., compared to an expected 4% by statistical definition) on the Social Problems scale. Further, when compared to either their unaffected siblings or a disabled comparison group, the Social Problems behavior scale was the one area that consistently showed a significant difference. In the matched sibling comparisons, 26% of the boys with DMD and 6% of their siblings (20% vs 2% when the smaller proband brother group was studied) were rated as having clinically significant Social Problems, even when their family backgrounds did not differ. Further, although the sample of children diagnosed with cerebral palsy (CP) also had an elevated Social Problems scale relative to normative data, the percentage of children with CP whose scores were above the clinical cut-point (14%) was still significantly lower than the

percentage of participants (of comparable age) with DMD who scored above cut-point (46%).

The finding of poor social and interpersonal behavior skills is substantially more robust than the finding of increased depressive and anxiety behaviors among boys with DMD. Among the current sample, parents did not rate higher levels of depressed and anxious affect in the boys with DMD than their siblings. Although early studies of DMD have suggested increased depressive and anxiety symptoms in some boys with the illness when evaluated either through parent interview or direct projective techniques,<sup>19-23</sup> our data indicate that this is not the predominant behavioral characteristic reported by parents. Indeed, earlier work from our group analyzing wish content of boys with DMD found their wishes in general to be similar to those of their siblings and a healthy comparison group of boys, and they did not reflect increased depressive or health-related content relative to the other two groups, contrary to what we had originally hypothesized.<sup>32</sup> Instead, among the boys with DMD, elevated total T scores from the CBCL were associated with increased wishes for interpersonal relationships, perhaps also reflecting the social limitations reported in the current analyses.

It is possible that the children who agree to participate in our research are those whose families are best adjusted to the illness; the low rates of depressive symptoms may be attributed (in part) to selection bias. It is also possible that many children with this illness do adjust well and are not depressed, contrary to what might be expected from an unaffected person's understanding of the illness. This idea was given support in a study that compared ratings of quality of life and affect of patients with DMD who are on ventilators and found that a group of health care professionals rated the DMD subjects as having a significantly poorer quality of life, greater hardship, and more depressed affect than the DMD individuals who rated themselves.<sup>33</sup>

Interestingly, the finding of increased Social Problems was also considerably more robust than increased Somatic Complaints among the boys with DMD, contrary to reports in other chronic illnesses.<sup>34</sup> It has been suggested that the CBCL is of limited use with chronically ill children, in part because of the inflated scores due to increased somatic complaints,<sup>34</sup> yet our analyses suggested that although somatic complaints may be elevated among boys with DMD, they are not the distinguishing behavioral characteristics commonly observed among the sample.

Research has shown that some children taking oral corticosteroids may experience adverse psychological side effects.<sup>35</sup> Since 34% of the boys in the DMD sample reported taking steroid medication regularly, the possibility that the steroids contributed to the increased ratings of behavior problems was explored in a post hoc analysis. Children with DMD were segregated into those on steroid medication ( $n = 62$ ) and those not taking steroid medication ( $n = 119$ ) and multiple chi-square analyses were run across the eight different behavior scales on the CBCL split at the T  $\geq$  67 cut-point. None of the analyses was significant at the .05 level, indicating that steroid use is not the main factor contributing to the finding of elevated scores on the Social Problems scale.

Items that are included on the CBCL Social Problem Scale

- Clings to adults
- Complains of loneliness
- Doesn't get along with other kids
- Easily jealous
- Feels others are out to get him
- Gets hurt a lot
- Gets teased a lot
- Not liked by other kids
- Poorly coordinated or clumsy
- Prefers being with younger kids
- Speech Problems

**FIGURE 1.** Items that are included on the Child Behavior Checklist Social Problems scale.

The majority of the items that make up the social problems scale are labels of immaturity and poor peer relationships (Fig. 1). However, some items, such as “clumsy”, may reflect physical disability associated with DMD. Item analysis of randomly chosen DMD/sibling pairs indicated that the items in the social problem scale that accounted for differences included “clings to adults” and “prefers playing with younger children”, suggesting an immature behavior style. The scale's items appear to reflect social delay more than social deviancy, but the items are too general to offer much insight into the underlying nature of the problems.

Research using the CBCL has found the social problem scale to be elevated in different groups of children selected by behavioral characteristics. For example, children identified by their mothers as having peer relationship problems had elevated social problem and withdrawal scale scores,<sup>36</sup> children with pervasive developmental disorder had elevated social problems and withdrawal scores,<sup>37</sup> and autistic children had elevated Attention, Social Problems, and Thought scale scores.<sup>38</sup>

At face value, these groups appear to be substantially different from those with DMD, as they are characterized more by behavioral attributes than physical disability. Yet cases of autistic children with DMD have been reported,<sup>39-41</sup> and we have suggested that many boys with DMD meet

criteria for pervasive developmental disability and autism spectrum disorder.<sup>42,43</sup> The findings of increased social skill problems may be associated with the underlying developmental brain aberrations found in DMD. The increased endorsement of behaviors on this scale along with the Attention, Withdrawn, and Thought Problems scales may reflect behavioral characteristics associated with DMD.

The current examination of behavior in children with DMD is limited by the fact that only one scale was used (the CBCL), and it relied solely on maternal report. Moreover, no teacher report was collected and no clinical interview was given, so the data are sparse and need validation. The CBCL has been shown to have significant limitations in assessing the behavioral adjustment of children with chronic illness, including heightened scores due to physical symptoms and lower sensitivity for problems associated with adjustment to the illness.<sup>34,44,45</sup> We attempted to control for these limitations by setting a stringent cutoff to increase specificity of behavioral problems (at the expense of sensitivity) and by comparing data across different subject groups. Thus, although these analyses may not be an ideal screen for children at risk of adjustment problems (as they would likely miss some), they add support for the findings of the specificity of the observed profile (as scoring above the cut-point is unusual). The observation that one third of the participants with DMD scored above the 96th percentile on the Social Problems scale highlights the differences between the behaviors reported by these parents and those used in the original standardization sample. Our ongoing work using additional indices of children's behavior and more robust scales of social communication will more thoroughly characterize the behavioral profile associated with DMD.

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