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Resilience in Children Diagnosed with a Chronic Neuromuscular Disorder

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Abstract

Objective—To examine what contributes to resiliency in children living with Duchenne muscular dystrophy (DMD), a chronic, progressive neuromuscular disorder that also influences cognitive ability. We hypothesized that family and social support will moderate the effects of individual symptoms of illness severity and influence positive adjustment in boys with DMD.

Method—146 boys with DMD were included. Child adjustment, as determined by parent ratings of their son's behavior using the Total Behavior score from the Child Behavior Checklist (CBCL), was examined as an outcome measure. The contributions of individual variables (including age (which serves also as a proxy for degree of physical disability), wheelchair use, and estimated verbal IQ), family variables (the Parental Distress score from the Parent Stress Index), and social environment variables (the Social Competence score from the CBCL) on child adjustment were examined in a linear regression analysis.

Results—Both family and social environment variables significantly contributed to the variance in the CBCL total behavior score. In contrast, individual factors that are related to illness severity (age, degree of physical involvement and estimated verbal IQ) were not associated with child adjustment.

Conclusion—Increased children's social networks and decreased parents' stress levels positively contributed to good child adjustment, whereas degree of individual clinical severity did not. Thus, emphasis on providing opportunities for friendships and social support and on parents' adjustment will aid in children's resilience, ensuring they can live well, even while living with the significant burdens associated with DMD.

Keywords

Duchenne Muscular Dystrophy; resilience; social support; psychosocial impact

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INTRODUCTION

Despite the significant stressors of medical illness, many chronically ill children and their families remain productive and happy in their lives¹⁻³. Somehow they are resilient, living well even when living with illness and its associated burdens. Individual response to significant adversity raises questions as to why some achieve positive adaptation and others develop behavioral and emotional dysfunction. Research has examined the concept of resilience across multiple at-risk populations and the answers have yet to be solidified. Yet determining what is associated with good outcomes in the face of chronic illness may well offer a key insight into the basis of resilience. The current study examines resilience among a group of children with a chronic, progressive and eventually fatal neuromuscular disorder, Duchenne muscular dystrophy (DMD). A diagnosis of DMD carries with it prolonged loss and burden; a seemingly healthy infant will grow weaker over years, lose the ability to walk as an adolescent and die as a young man. There is no known cure. Nonetheless, many children who live with DMD are well-adjusted and content youngsters. Finding what may contribute to their resiliency may help all affected children live better lives.

Resilience can be defined as “a dynamic process encompassing positive adaptation within the context of significant adversity”⁴. Resilience is “the ability to maintain a stable equilibrium” and is a concept differentiated from recovery^{5,6}. It is not a particular personality trait but a process by which positive adaptation occurs despite adversity⁴. In a classic 30 year longitudinal study, Emily Werner (1989) found that a number of “high risk” children were resistant to environmental disadvantages and deprived childhoods and developed into healthy adults⁴. The developmental research showed that multiple adverse conditions can influence the development of an individual and that individual protective factors differentiate adaptive versus maladaptive individual functioning⁷⁻⁹. Werner’s study challenged the common presumption of a direct connection between early childhood and later development and invoked a foundation for resilience research⁴. This has been applied to chronic illness, where it has been shown that individuals living with such physical adversity and who nonetheless adapt and adjust and do not have poor outcomes are considered “resilient”^{1,5,6}.

Individual, familial, and social factors have each been hypothesized to contribute to resilience. Individual protective factors include good physical health, strong intellect, a sociable outgoing style of interaction, and an appropriate level of self-confidence and self-esteem^{3,9}. At the family level, close parental relationships, adequate family income to meet needs, supportive connections, and parents who are warm, provide structure and have reasonable expectations of their children are all associated with a protective outcome^{3,10}. Lastly on a community level, protective factors include positive caring relationships outside the family like teachers and peers, involvement in organizations, and being enrolled in a good school¹¹⁻¹⁵. This area has been referred to as general social support.

Among children with chronic illness, as the severity and frequency of health problems increase, a child’s physical and psychological well-being has been shown to decrease⁷. Epidemiological and pediatric clinic samples have showed that 15-30% of chronically ill children have been shown to be “at risk” for developing emotional and behavioral problems⁸⁻¹⁰. Children with physical disorders show an increased risk for adjustment problems, both internalizing and externalizing symptoms, as well as a significantly lower self-concept¹⁰. Although the various chronic illnesses have distinct characteristics, trajectories and treatment, many share common psychological consequences. Research has shown an even higher risk of maladjustment in neurological disorders and those involving motor functioning^{8,11}. DMD meets criteria for a severe chronic disorder and is both neurological and affects motor functioning. As such, the concern for poor adjustment among

children affected with DMD is great. Additionally, understanding what may contribute to resilience among children with DMD is valuable and necessary information to ensure each child has the best possible quality of life.

DMD is a genetic neuromuscular disorder that occurs in about 1 in 3200 live male births. The disease is relatively uncommon, and its consequences devastating. Because of the lack of the protein dystrophin, muscle cells break down resulting in muscle weakness and an eventual loss of movement. Children grow weaker as they age, most losing the ability to walk in their early teens. Instead of increased independence with age, most boys become more dependent of others to perform daily tasks such as toileting and eating. Additionally, their lifespan is shortened. Most boys do not live longer than their mid-20's and survival depends mainly upon ventilatory support in the final stages of the disorder due to respiratory and cardiac complications¹². In addition to the physical presentation of the disorder, there has also been shown to be a cognitive component. Affected boys have a wide range of intellectual functioning, and as many as one third function in the intellectually disabled range¹³. Across all affected boys, specific cognitive deficits impacting language development and academic achievement have been found^{14,15}. Current standard of care for boys includes treatment with corticosteroids, physical therapy and good ventilatory support, as well as assessment and treatment of cognitive deficits and adjustment difficulties.^{16,17}.

Rates of behavioral problems are also increased in DMD. Children with DMD have an elevated risk for behavioral and emotional problems, including social skill deficits, depression and attention deficits¹⁸⁻²¹. Studies have shown that between 30% and 50% of boys with DMD have psychosocial problems²²⁻²⁴. These rates are similar to those found among individuals with other chronic illnesses, and may speak to the difficulties adjusting to living with the disorder.

Yet, even though rates of emotional problems among children with DMD are higher than the general population (as expected with any chronic illness), the majority of children affected do not have behavioral problems. We have seen this repeatedly in our samples. When parent behavior ratings from 86 children with DMD and their unaffected siblings were compared across behavior scales, no differences were observed in 80% of the cases¹⁹. Likewise, in a separate sample of 85 children with DMD who were examined for social skill deficits, 75% showed none²⁵. In a sample of 36 boys with DMD who completed the Child Depression Inventory there was no evidence of depression²⁶. Further when 74 boys with DMD were given a simple projective measure where each was asked to state three wishes that were objectively coded and analyzed, results indicated that the majority of responses were comparable to their healthy peers', reflecting good adjustment²⁶. Taken together, these data indicate that although the children with DMD all live with the physical consequences of the disorder, the majority do not have signs of detrimental psychological consequences. They may be considered resilient.

Additionally, it is noteworthy that self perception among many older individuals with DMD is in fact more positive than what their caregivers imagine. Bach, Campagnolo, and Hoeman (1991) found that even among the most severely disabled DMD individuals, those who could no longer survive without ventilator support, the majority reported positive affect. Despite having what their caregivers deemed a poor quality of life, most affected individuals reported that they are satisfied with their lives²⁷. Similarly, a cross-sectional survey of adult DMD patients revealed that although many individuals were concerned with a lack of competencies in social life, overall the majority reported their quality of life to be excellent with little worry about the disease or the future²⁸. Those individuals with DMD who are not overly preoccupied with their physical limitations and who have positive life satisfaction

may be thought of as those who are resilient. They neither define nor limit themselves by their illness.

Thus, although the disease has devastating effects on physical strength and life span, individual responses to it are variable. Given that individual, familial, and social factors have been shown to contribute to resilience in other adverse situations, it is hypothesized that these three factors likely contribute to outcome among individuals with DMD as well. Two studies have shown that family functioning plays a key role in moderating a child's experience in DMD. Reid (2001) found that levels of familial stress predicted psychosocial adjustment in 32 adolescent boys with DMD²⁹. The boys in the families who were reportedly less stressed were better adjusted and had a healthier sense of self. Similarly, Chen and Clark (2007) found that when examined among 80 families, family hardiness, caregiver health and family support all were factors associated with a family's ability to cope and function with DMD. Higher levels of family functioning greatly increased resilience and the ability to cope with the effects of the disorder in families of children with DMD³⁰.

The current paper examines this phenomenon further among a large group of boys with DMD. The goal is to determine what may contribute to the children's psychological state as reported by their parents. Working within the resilience model, it is hypothesized that family and social support will moderate the effects of individual symptoms of illness severity and influence positive adjustment in boys with DMD. . The current study, working with an already acquired dataset, will test whether measures of individual characteristics, parental distress and social competence contribute to children's behavior outcome. It is hypothesized that minimal parental distress and increased social competence will offer resilience and protect against the risk of behavioral and emotional problems associated with DMD.

METHODS

Participants

Subjects were participants in the ongoing study Cognitive Skills in Boys with Muscular Dystrophy (CPMC, IRB#AAAA5627), sponsored by the National Institute of Neurological Disorders and Stroke and the Muscular Dystrophy Association, Principal Investigator – V.J. Hinton. Diagnosis of DMD was based on clinical onset of progressive weakness before 5 years of age, and either molecular assessment of mutation in the DMD gene or muscle biopsy that was deficient in dystrophin and compatible with DMD.

One hundred and sixty-five children ranging from 6 to 14 years old (mean age = 9.00; SD = 2.31) were eligible for inclusion in the study. 31% of the children were using a wheelchair. 36% of the children were receiving glucocorticoid therapy at the time of participation. The primary language of all the participants was English. Because DMD is an X-linked disorder, all participants were male. The ethnic composition of this group was: 89% Caucasian, 4% Hispanic, 4% African American, and 3% Asian.

Primary caregivers were requested to complete parental questionnaires (159 mothers, 5 fathers, and 1 grandmother). The parent's average education level was at least some college and the average household income fell in the \$60,000 and \$74,999 range.

Procedure

Participants were recruited through the Muscular Dystrophy Association (MDA) clinics of Columbia Presbyterian Hospital in New York City, Children's Healthcare of Atlanta, The Children's Hospital of Philadelphia, and through Washington University School of Medicine in St. Louis. Newsletters with the study description were also distributed to regional MDA

clinics and parent support groups. Participation interest forms were returned to the study personnel at Columbia Medical Center. Interested participants were then contacted by phone to set up a neuropsychological evaluation either at the hospital or in the family home. In cases where a home evaluation was necessary, the researcher traveled to the participant's location. Columbia University and New York Presbyterian Hospital Institutional Review Board annually approved the study for ongoing data collection.

Parents provided written informed consent before participation began. All children provided verbal assent and capable children also signed a written assent form. Children completed a battery of tests with a psychologist or research assistant who had been trained on the test battery, while the parents completed a developmental and family history forms, as well as a series of questionnaires about their child's emotional and behavioral development. Only the primary caregiver was asked to fill out all the questionnaires for consistency. All tests and questionnaires were scored twice to ensure data reliability.

Measures

For the purposes of the current study, select measures were chosen from the larger dataset for analysis.

Outcome

As a measure of general psychosocial adjustment, the total T score from the Child Behavior Checklist (CBCL) ³¹ was determined for each child. The scale includes one hundred and eighteen behaviors that children engage in, and parents rated their child on each item on a frequency scale from 0 (never) to 2 (very often). Individual scores were compared to a standardization sample of comparably aged peers, and a Total behavior T score was computed as a measure of the child's overall behavioral function. Scores that fall above the 96th percentile have been shown to reflect behavior more consistent with that of children who have been referred for clinical psychological treatment than the general population. The scale has been well validated to identify children who are clinically "at risk" for significant behavioral problems.

Individual

As measures of individual variability, participant's age, wheelchair use and estimated verbal intelligence level were used. Because of the progressive nature of the disease, age can be used as a proxy measure for physical ability; older boys are typically more physically limited than younger boys. Additionally, use of a wheelchair is a marker of physical limitation. As a measure of verbal intelligence, all boys were administered the Peabody Picture Vocabulary Test –III (PPVT-III) ³², an un-timed test of receptive vocabulary. The PPVT-III is appropriate for use across a wide age-range, as well as range of intellectual function, and does not involve any significant motor response that might confound performance among the physically disabled children. The raw scores were converted to age-referenced standard scores with a mean of 100 and standard deviations of 15. PPVT- III has a correlation of .69 with the Verbal IQ score of the Wechsler Intelligence Scales for Children-Revised and has a correlation of .70 with the Full Scale IQ score ³².

Family Support

As a measure of parent well-being and variability, the parent distress scale from The Parenting Stress Index-Short Form (PSI-SF) ³³ was used. The PSI-SF is a thirty-six item questionnaire that measures stress in parental functioning and is comprised of three subscales. To validate parental responses, a defensive responding scale is included with commonly endorsed items that serve as a means to find individuals whose answers may

reflect an overly unrealistic positive bias. The PSI-SF is considered “questionable” if parent’s score lower than 11 on the defensive responding scale, and, as such, only data from responders who scored above 11 on the defensive responding scale were used. Test-retest reliability for the total score and subscales of PSI-SF ranges from .68 to .85³³. For the current analyses, data from only one subscale, that measuring parental distress (PSI-PD), were included as a measure of parents’ level of adjustment, independent from their relationship with their child.

Social Support

As a measure of the child’s social support, the Social scale from the CBCL was used. In addition to the behaviors scales described above, the CBCL also provides information regarding the child’s participation and competence in activities, academic achievement, and social networks. The Social scale includes the number and frequency of peer relations, and the behavior of the child with others.

Data Analysis

For the Boys with DMD, mean age, PPVT standard scores, PSI-SF Parental distress score, CBCL Social score, and CBCL Total score were calculated. All defensive responders from the PSI-SF were removed from the group analysis (n= 19). The remaining group consisted of 146 children, ranging in age from 6 to 14 (mean age = 9.01, SD = 2.33). This group’s demographic characteristics remained the same as the original sample.

Descriptive analyses were conducted on each variable of choice to ensure a wide range of responses that represent both high functioning and low functioning scores. To ensure the data were normally distributed for parametric analyses, Kolmogorov-Smirnov analyses were calculated. Only those variables with sufficient variance were included in the analyses. A linear regression analysis was then performed using the Total T behavior score as the outcome measure. PPVT, age, PSI-PD, and Social T score were entered as the independent variables. The contributions of individual variables (age, PPVT), family variables (PSI-PD), and social environment variables (Social T competence scale from the CBCL) were determined.

Additionally, in order to ensure behavioral outcome data are “clinically relevant,” the outcome data were split at a T value of 67. This value was based on findings that scores in this range are at the 96th percentile. T values greater than 67 have been found to discriminate between normal and clinically referred populations; scores greater than T of 67 reflect behavioral problems more similar to those observed among children referred for psychiatric treatment than the normal population. A logistic regression was then performed on this dichotomous total T behavior score as the outcome. The contributions of individual variables (age, PPVT), family variables (PSI-PD), and social environment variables (Social T competence scale from the CBCL) were determined.

RESULTS

Descriptive data on each of the variables are presented in Table 2. As anticipated, each variable shows a wide range of values, and these numerical values can be translated to clinically significant values using standard “cut points.” Most importantly, the outcome behavior measure indicated a range from “normal” to “clinically at risk” scores, with 16% of the group scoring in the “at risk” range (or having a T value >67). For the purposes of the current study, scores below 67, considered a good behavioral outcome, were described as “resilient,” as they imply adequate adjustment. Other scales also show a clinically relevant range of values. PPVT-III scores range from 40 to 140, with 9% of the group falling in the

“intellectually disabled” range (Standard score < 70), and the parent distress scale ranging from 16 to 58 with 23% falling in the “at risk” range (Raw score > 33). Results from the Kolmogorov-Smirnov analyses indicated that all variables met the assumptions for parametric analyses. The one exception to this was age, where $K-S = 1.86, p = .002$. Data were skewed such that the majority of the children were in the 6 to 9 years of age range.

Results from the linear regression analysis examining the contribution of individual, family and social variables on the range of behaviors displayed by the children with DMD were significant (omnibus $F = 5.47, p = .000$) (Table 3). Analysis of individual contributions showed that both the family and social variables contributed significantly, while the individual variables did not. Thus, both parent distress and social environment are associated with a continuum of children’s behavior.

When outcome data are cut into a dichotomous variable indicating children’s behavior that is “clinically at risk” vs. “normal” and the analyses are rerun using a logistic regression, the overall results are also significant (Chi-square = 11.21, $p = .02$) (Table 4). Analysis of individual contributions indicated that only the social variables were significantly associated with the outcome. Individual and family contributions were not. Thus, social factors appear to be strongly related to good behavioral outcome, or resilience.

DISCUSSION

Although all the children who participated in this study live with a chronic, progressive, and eventually fatal neuromuscular disorder, a remarkable 84% were not found to be psychosocially “at risk” by a standard child behavior measure. These children are resilient. They live with adversity yet their behaviors reflect normal function. Further, among the children in this sample, two things were strongly associated with good outcome: social support and parent adjustment. These two factors contributed more to behavioral outcome than even the disease characteristics.

The strongest contribution to child behavior was social support. Social support, as defined here by number and quality of peer relations as well as involvement in peer groups and activities, was related on a continuum of behaviors. Additionally, limited social support positively identified those children deemed “at risk” for clinically significant behavior problems.

The results of the current study demonstrate that strong social networks go hand-in-hand with resilient behavior in boys with DMD. Increased participation in organizations, number of friendships, and positive peer relations were associated with a decreased risk of a boy with DMD developing behavioral problems. The findings are supported by work of others that has shown that positive peer social perceptions and relationships effectively aid a child in the ability to cope with major stressors such as chronic medical illness^{11,34}. The sample of boys with DMD examined showed an inverse relationship between the problem behavior scale and the social scale; as social opportunities and involvement increases, problem behaviors decrease. In a setting of a child affected by an incurable illness such as DMD, this relationship may be particularly salient, as it offers a protective factor that can be modified. Enhancing a child’s social opportunities is something that can be done in most all situations, even when biological cures are lacking.

It is proposed that social support leads to resilience by positively intervening in the process by which stressful situations lead to maladjustment. Prior research has attempted to conceptualize social support by examining the many aspects and sources of support³⁵. Social embeddedness, enacted support, and perceived quality of support have all been shown to contribute to the construct of social support. Social embeddedness refers to the

relationships children have with others including family, friends and the community. Enacted support is positive feedback, guidance and the emotional support received. Among medically ill children, the degree of social support has repeatedly been shown to be a discriminating factor between adjustment and maladjustment. Across multiple chronic illnesses including diabetes, cerebral palsy, juvenile arthritis, cystic fibrosis and limb deficiencies, social support significantly influenced positive adjustment and decreased internalizing and behavior problems^{11,36-38}. In pediatric cancer patients, higher perceived social support in classmates was consistently associated with lower depressive symptoms, state anxiety, social anxiety, externalizing and internalizing behavior problems and an overall higher sense of self-esteem^{11,39}. In studies of children with limb deficiencies, lack of social support significantly predicted depressive symptoms and maladjustment^{11,38}. A child's understanding of social support is consistent with coping theory where perceptions and interpretations of social networks may protect against emotional distress while handling adversity^{11,34}.

The current sample also showed a positive association of family variables with outcome. Parents' rating of distress was associated with children's behaviors such that those who rated themselves as less distressed were more likely to have children with healthy behaviors. The finding was observed on a continuum, but did not selectively identify children at behavioral risk.

This too is consistent with prior findings. Family variables have been shown to protect against maladjustment in cases of chronic illness and in adverse environments. Parents who are supportive, involved, and have positive attitudes increased a stress-resilient outcome in their children³. Family cohesion, stability, and financial status also predicted psychological outcomes for a child⁴⁰. And among families living with DMD, family functioning has been shown to be positively associated with child's outcome^{29,30}.

Interestingly, in the current study, individual factors did not show an association with outcome. Unlike other work that has shown intellect to be associated with psychosocial outcome³, there was no sign that a child's intellectual level was related to his behavior profile in this sample of children with DMD. Further, and perhaps even more striking, proxy measures of physical ability (age and wheelchair use) showed no association with the child's behavior. This finding highlights the notion that a child's perception of disability may well be more relevant than any objective measure when it comes to understanding resilience. When examined within the context of the Lazarus and Folkman model, these results support the theory that a child's response to his illness is modified by environmental factors (such as his parents' coping skills) that are separate from his actual disease state^{11,34}. Put differently, if resilience is "a positive adaptation to the adversity"⁴¹ then the quantifiable nature of the adversity may be less important to the child than the positive adaptation to it.

The current examination of behavioral and emotional problems has several limitations, most notably the limited measures used to quantify each variable. The CBCL was used to determine behavioral outcome, our measure of resilience. Although a valid reflection of a child's behavior, the CBCL nonetheless offers a narrow view by being based solely on parental report without the additional resources of the child's, teacher's or clinician's perspective. Moreover, the CBCL has been shown to be less sensitive to adjustment problems in children with chronic illness due to physical symptoms^{5,42,43}. This was addressed in the current study by establishing cutoffs at the 96th percentile, relative to the normative sample, increasing sensitivity to identify problem behaviors. The social competence scale of the CBCL, used in this study as a measure of social support, has the same limitations of being based solely on parent report. The PSI Parent Distress scale used as a family variable measure, offered insight into the parent's affective state, yet reflected

only one parent's perspective with no details about family structure or financial resources. Likewise the child variables of IQ, age and wheelchair-use, are narrow indices of child functioning. The measures chosen are all adequate indices, and the findings are conclusive, yet the study cannot reflect the broad complexities of each child's world

Nonetheless, within these constraints, results from the current study show that general social competencies and parental psychosocial functioning contribute to children's ability to "maintain stable equilibrium," or resilience, whereas individual factors that may be considered measures of degree of adversity (including physical and intellectual characteristics of DMD) did not. These findings offer hope in the face of an incurable illness. They suggest that providing opportunities for friendships and social support and aiding parents' adjustment will contribute to children's resilience, ensuring they can live well, even while living with the significant burdens associated with DMD.

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References

1. Sinnema G. Resilience among children with special health-care needs and among their families. *Pediatr Ann.* 1991; 20:483–486. [PubMed: 1945545]
2. Patterson JM. Family resilience to the challenge of a child's disability. *Pediatr Ann.* 1991; 20:491–499. [PubMed: 1834978]
3. Condly SJ. Resilience in Children: a review of literature with implications for education. *Urban Education.* 2006; 41:211–236.
4. Werner EE. Children of the Garden Island. *Sci Am.* 1989; 260:106–111. [PubMed: 2928762]
5. Harris ES, Canning RD, Kelleher KJ. A comparison of measures of adjustment, symptoms, and impairment among children with chronic medical conditions. *J Am Acad Child Adolesc Psychiatry.* 1996; 35:1025–1032. [PubMed: 8755799]
6. Hysing M, Elgen I, Gillberg C, et al. Chronic physical illness and mental health in children. Results from a large-scale population study. *J Child Psychol Psychiatry.* 2007; 48:785–792. [PubMed: 17683450]
7. Waters E, Davis E, Nicolas C, et al. The impact of childhood conditions and concurrent morbidities on child health and well-being. *Child: Care, Health and Development.* 2008; 34:418–429.
8. Hysing M, Elgen I, Gillberg C, et al. Emotional and behavioural problems in subgroups of children with chronic illness: results from a large-scale population study. *Child Care Health Dev.* 2009
9. Glazebrook C, Hollis C, Heussler H, et al. Detecting emotional and behavioural problems in paediatric clinics. *Child Care Health Dev.* 2003; 29:141–149. [PubMed: 12603359]
10. Lavigne JV, Faier-Routman J. Psychological adjustment to pediatric physical disorders: a meta-analytic review. *J Pediatr Psychol.* 1992; 17:133–157. [PubMed: 1534367]
11. Wallander JL, Varni JW. Effects of pediatric chronic physical disorders on child and family adjustment. *J Child Psychol Psychiatry.* 1998; 39:29–46. [PubMed: 9534085]
12. Eagle M, Baudouin SV, Chandler C, et al. Survival in Duchenne muscular dystrophy: improvements in life expectancy since 1967 and the impact of home nocturnal ventilation. *Neuromuscul Disord.* 2002; 12:926–929. [PubMed: 12467747]
13. Cotton SM, Voudouris NJ, Greenwood KM. Association between intellectual functioning and age in children and young adults with Duchenne muscular dystrophy: further results from a meta-analysis. *Dev Med Child Neurol.* 2005; 47:257–265. [PubMed: 15832549]
14. Hinton VJ, De Vivo DC, Nereo NE, et al. Selective deficits in verbal working memory associated with a known genetic etiology: the neuropsychological profile of duchenne muscular dystrophy. *J Int Neuropsychol Soc.* 2001; 7:45–54. [PubMed: 11253841]

15. Hinton VJ, Fee RJ, Goldstein EM, et al. Verbal and memory skills in males with Duchenne muscular dystrophy. *Dev Med Child Neurol.* 2007; 49:123–128. [PubMed: 17254000]
16. Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. *Lancet Neurol.* 9:77–93. [PubMed: 19945913]
17. Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and management of Duchenne muscular dystrophy, part 2: implementation of multidisciplinary care. *Lancet Neurol.* 9:177–189. [PubMed: 19945914]
18. Hendriksen JG, Vles JS. Neuropsychiatric disorders in males with duchenne muscular dystrophy: frequency rate of attention-deficit hyperactivity disorder (ADHD), autism spectrum disorder, and obsessive-compulsive disorder. *J Child Neurol.* 2008; 23:477–481. [PubMed: 18354150]
19. Hinton VJ, Nereo NE, Fee RJ, et al. Social behavior problems in boys with Duchenne muscular dystrophy. *J Dev Behav Pediatr.* 2006; 27:470–476. [PubMed: 17164619]
20. Darke J, Bushby K, Le Couteur A, et al. Survey of behaviour problems in children with neuromuscular diseases. *European Journal of Paediatric Neurology.* 2006; 10:129–134. [PubMed: 16766215]
21. Poysky J. Behavior patterns in Duchenne muscular dystrophy: report on the Parent Project Muscular Dystrophy behavior workshop 8-9 of December 2006, Philadelphia, USA. *Neuromuscul Disord.* 2007; 17:986–994. [PubMed: 17720499]
22. Hendriksen JG, Poysky JT, Schrans DG, et al. Psychosocial adjustment in males with Duchenne muscular dystrophy: psychometric properties and clinical utility of a parent-report questionnaire. *J Pediatr Psychol.* 2009; 34:69–78. [PubMed: 18650207]
23. Hendriksen JGM, Vles JSH. Neuropsychiatric disorders in males with Duchenne muscular dystrophy: frequency rate of attention-deficit hyperactivity disorder (ADHD), autism spectrum disorder, and obsessive-compulsive disorder. *Journal of Child Neurology.* 2008; 23:477–481. [PubMed: 18354150]
24. Darke J, Bushby K, Le Couteur A, et al. Survey of behaviour problems in children with neuromuscular diseases. *Eur J Paediatr Neurol.* 2006; 10:129–134. [PubMed: 16766215]
25. Hinton VJ, Cyrulnik SE, Fee RJ, et al. Association of autistic spectrum disorders with dystrophinopathies. *Pediatr Neurol.* 2009; 41:339–346. [PubMed: 19818935]
26. Nereo NE, Hinton VJ. Three wishes and psychological functioning in boys with Duchenne muscular dystrophy. *J Dev Behav Pediatr.* 2003; 24:96–103. [PubMed: 12692454]
27. Bach JR, Campagnolo DI, Hoeman S. Life satisfaction of individuals with Duchenne muscular dystrophy using long-term mechanical ventilatory support. *Am J Phys Med Rehabil.* 1991; 70:129–135. [PubMed: 2039614]
28. Rahbek J, Werge B, Madsen A, et al. Adult life with Duchenne muscular dystrophy: observations among an emerging and unforeseen patient population. *Pediatr Rehabil.* 2005; 8:17–28. [PubMed: 15799132]
29. Reid DT, Renwick RM. Relating familial stress to the psychosocial adjustment of adolescents with Duchenne muscular dystrophy. *Int J Rehabil Res.* 2001; 24:83–93. [PubMed: 11421396]
30. Chen J-Y, Clark M-J. Family function in families of children with Duchenne muscular dystrophy. *Family & Community Health.* 2007; 30:296–304. [PubMed: 17873636]
31. Achenbach, TM. Manual for the Child Behavior Checklist/4-18 and 1991 Profile. University of Vermont, Department of Psychiatry; Burlington, VT: 1991.
32. Dunn, LM.; Dunn, DM. Peabody Picture Vocabulary Test (PPVT-IV). ed Fourth. Pearson Assessments; Minneapolis, MN: 2007.
33. Abidin, RA. Parenting Stress Index-Short Form (PSI-SF): Professional Manual. Psychological Assessment Resources, Inc.; Odessa, FL: 1990.
34. Lazarus RS. Coping theory and research: past, present, and future. *Psychosom Med.* 1993; 55:234–247. [PubMed: 8346332]
35. Belle, D. Children's social networks and social supports. ed 2nd. John Wiley & Sons, Inc.; Ontario: 1989.
36. Wallander JL, Varni JW. Social support and adjustment in chronically ill and handicapped children. *Am J Community Psychol.* 1989; 17:185–201. [PubMed: 2528286]

37. Meijer SA, Sinnema G, Bijstra JO, et al. Social functioning in children with a chronic illness. *J Child Psychol Psychiatry*. 2000; 41:309–317. [PubMed: 10784078]
38. Tyc VL. Psychosocial adaptation of children and adolescents with limb deficiencies: a review. *Clinical Psychology Review*. 1992; 12:275–291.
39. Varni JW, Katz ER, Colegrove R Jr. et al. Perceived social support and adjustment of children with newly diagnosed cancer. *J Dev Behav Pediatr*. 1994; 15:20–26. [PubMed: 8195433]
40. Northam EA. Psychosocial impact of chronic illness in children. *J Paediatr Child Health*. 1997; 33:369–372. [PubMed: 9401876]
41. Luthar SS, Cicchetti D, Becker B. The construct of resilience: a critical evaluation and guidelines for future work. *Child Dev*. 2000; 71:543–562. [PubMed: 10953923]
42. Perrin EC, Stein RE, Drotar D. Cautions in using the Child Behavior Checklist: observations based on research about children with a chronic illness. *J Pediatr Psychol*. 1991; 16:411–421. [PubMed: 1941423]
43. Canning EH, Kelleher K. Performance of screening tools for mental health problems in chronically ill children. *Arch Pediatr Adolesc Med*. 1994; 148:272–278. [PubMed: 8130859]

Table 1

Participant characteristics

n = 165	Boys with DMD	n = 146	Boys with DMD
% Male	100.0	% Male	100.0
% White	89.4	% White	89.1
Child Age, years (M+/-SD)	9.00+/-2.31	Child Age, years (M+/-SD)	9.01+/-2.33
% in wheelchair	31.4	% in wheelchair	31.3
Average Mother Education	Some college	Average Mother Education	Some college
% Steroid Use	36.4	% Steroid Use	36.5

Table 2

Performance profile of the 146 boys with DMD

	Range	Mean	SD	K-S
IQ estimate	40-140	100.43	20.89	1.21
PSI-PD	16-58	28.72	6.84	0.91
Social competence	20-55	38.97	8.17	0.93
CBCL Total T	26-80	57.31	10.32	0.93

K-S, Komogorov-Smirnov test

Table 3

Linear regression analysis on Total behavior scores of the boys with DMD

	Beta	t	p
IQ estimate	0.10	1.28	.20
Age	−0.00	−0.02	.98
PSI-PD	0.20	2.50	.01*
Social scale	− 0.27	− 3.29	.00*

Dependent variable: CBCL Total T score

n = 146

omnibus F = 5.47, p = .000

*
p < .05

Table 4

Logistic regression analysis on the “clinically at risk” vs. “normal” boys with DMD

	B	S.E.	df	p
Age	0.09	0.09	1	.31
Constant	-1.23	1.90	1	.52
IQ estimate	0.01	0.01	1	.22
PSI-PD	0.02	0.03	1	.43
Social scale	- 0.07	0.03	1	.00*

Outcome T $\geq$ 67

n = 146

Chi-square = 11.21, p=.02

*
p < .05