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Article

A CROSS-SECTIONAL STUDY OF SCHOOL EXPERIENCES OF BOYS WITH DUCHENNE AND BECKER MUSCULAR DYSTROPHY

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Abstract: The objectives of this study were to investigate types of supportive school services received and factors related to provision of these services. We conducted a cross-sectional study to describe the school experience of males with Duchenne and Becker muscular dystrophies. Study subjects were identified through the Muscular Dystrophy Surveillance, Tracking, and Research Network. Non-ambulatory males with Duchenne muscular dystrophy (DMD) were significantly more likely to use an instructional assistant and resource room

support when compared to ambulant males with DMD at the time of the caregiver interview. Males with DMD who received occupational therapy were more likely to use an instructional assistant, while those who received speech therapy were more likely to repeat a grade, use an instructional assistant, and receive resource room support. Males with DMD whose primary caregivers had less than 12 years of education were more likely to use an instructional assistant and resource room support. Non-ambulatory males with DMD should continue receiving appropriate school accommodations so they can keep pace with their physical demands. Males with DMD with speech deficits should be considered for various educational interventions.

Keywords: *physical impairments; cognitive impairments; speech-language pathology; physical therapy; occupational therapy*

Introduction

Duchenne and Becker muscular dystrophies (DBMD) are genetic conditions that occur almost exclusively in males and are characterized by progressive muscle weakness and atrophy. In 2010, prevalence of DBMD was estimated at 1.38 per 10,000 males, ages 5 to 24 years (Romitti et al., 2015). The two conditions have similar signs and symptoms; however, they differ in age of onset, rate of progression, and severity.

In boys with Duchenne muscular dystrophy (DMD), signs and symptoms are observed between ages 2 and 5 and worsen rapidly (Ciafaloni, Chinnery, & Griggs, 2014). Males with DMD experience a progressive loss of muscle function resulting in a number of physical challenges such as a decline in mobility with eventual need for a wheelchair by age 12, sleep apnea, limitations in performing daily activities, and respiratory and/or cardiac complications, all of which can hinder their ability to learn (Cyrulnik, Fee, De Vivo, Goldstein, & Hinton, 2007; Kornberg & Yiu, 2008; Yiu & Kornberg, 2015; Zebracki & Drotar, 2008). It has been shown that the distribution of the average intelligence quotient (IQ) among males with DMD is shifted approximately one standard deviation lower than the distribution of the normative population (Worden & Vignos, 1962). In addition, previous literature has described a specific cognitive profile associated with DMD, with higher rates of poor performance on tests of digit span, verbal comprehension, and story memory and comprehension (Hinton, De Vivo, Nereo, Goldstein, & Stern, 2000; Hinton, De Vivo, Fee, Goldstein, & Stern, 2004; Wicksell, Kihlgren, Melin, & Eeg-Olofsson, 2007).

In males with Becker muscular dystrophy (BMD), the skeletal muscle involvement is more mild than with Duchenne, the onset of skeletal muscle weakness occurs and is recognized after age five, and some individuals maintain the ability to walk until adulthood (Ciafaloni et al., 2014). A high incidence of learning difficulties was observed among males with BMD; however, individuals with BMD demonstrated a less homogeneous cognitive phenotype than that seen in individuals with DMD (Young et al., 2007). Currently there is no cure for DBMD, but treatment with corticosteroids, use of pulmonary support, and cardiac medication have changed the natural history of DBMD, and individuals are surviving into their thirties and forties. This has created an

increased focus on preparing individuals with DBMD for employment, independent living, and participation in society.

To our knowledge, there has been virtually no research done on the educational and school experiences of males with DMD or BMD and it is unknown what types of special education services these individuals receive. In the literature, we only identified two studies that reported results on school attendance among males with DBMD. A study on psychological and practical difficulties among parents of children with DBMD by Magliano et al. (2014) reported a school attendance rate of 94.7% among boys with DMD and 98.9% among boys with BMD. Similar estimates on school attendance were reported by Lopez-Hernandez et al. (2014). Another two studies evaluated therapies received by males with DBMD (Pandya, Campbell, Andrews, Meaney, & Ciafaloni, 2015; Zebracki & Drotar, 2008). The two studies reported similar prevalence of occupational therapy (OT) and physical therapy (PT) received; however, findings of the study by Zebracki and Drotar (2008) were obtained using a relatively small sample size (43 participants with DMD and 10 participants with BMD), while the estimates in the study by Pandya et al. (2015) were not calculated separately for DMD and BMD. In addition, neither of the two studies differentiated between therapies received at school or at facilities outside of school.

There is a paucity of data on types of supportive school services received by males with DBMD and factors related to provision of these services. Therefore, the objectives of this study were to describe accommodations and services provided and examine education-based factors and caregiver characteristics associated with service provision. More specifically, we evaluated the distribution of study subject and caregiver characteristics separately among males with DMD and BMD by four dependent variables: participation in general education classes, grade repetition, usage of instructional assistant, and usage of resource room support. In addition, using multivariable models, we evaluated the association between ambulatory status, therapy received, household income, and education level of the caregiver and the aforementioned dependent variables among males with DMD.

Method

Design and Population

We conducted a cross-sectional study to evaluate the school experiences of males with DBMD using caregiver reports. Individuals with DBMD were identified, both prospectively and retrospectively, beginning in 2004 through the Muscular Dystrophy Surveillance Tracking and Research Network (MD STARnet), a multisite surveillance system for identifying and following individuals with DMD and BMD. The surveillance methodology has been described elsewhere (Miller et al., 2006). Briefly, the MD STARnet cohort included individuals with DBMD born on or after January 1, 1982, in the states of Arizona (AZ), Colorado (CO), Iowa (IA), Georgia (GA), Hawaii (HI), and a 12-county area in western New York state (wNY), who were identified as having a childhood-onset dystrophinopathy and were diagnosed before their 21st birthday through neuromuscular clinics, hospitals and hospital discharge databases, private physicians, service sites for children with special care needs, and birth defects surveillance programs.

Our study population included males with definite and probable DBMD diagnosis (Mathews et al., 2010). A male with documented clinical symptoms referable to a childhood-onset dystrophinopathy was categorized as definite if he met one or more of the following criteria: a positive genetic test for dystrophin mutation; a muscle biopsy demonstrating abnormal dystrophin; or an elevated creatine kinase (CK), family history of an X-linked dystrophinopathy, and an affected family member with a positive muscle biopsy or a dystrophin mutation. A male with documented clinical symptoms referable to a dystrophinopathy, elevated CK, and a family history of X-linked dystrophinopathy but no confirmatory genetic testing was categorized as probable.

Figure 1 shows the exclusion criteria used for this study. Excluded were (a) males younger than five years of age at the time of the interview ($n = 21$), (b) residents of Hawaii ($n = 2$) due to restricted ascertainment of individuals with DBMD, (c) males who were not enrolled in school from kindergarten through 12th grade ($n = 31$ deceased at the time of interview, $n = 13$ alive at the time of interview and younger than 18 years, and $n = 80$ alive at the time of the interview and 18 years old or older), (d) males who were homeschooled ($n = 2$), and (e) males who took postsecondary classes ($n = 4$).

Data Sources and Data Collection

Data collection consisted of abstraction of medical records, as well as annual follow-up abstraction until December 2011, the time of index male's death, or the time the index male moved outside of a site or catchment area. Abstracted data included demographic and diagnostic information, medical history, and family history of muscular dystrophy for the index males with DBMD. Surveillance data were supplemented with data collected through structured computer-assisted telephone interviews of eligible primary caregivers (primary caregivers of MD STARnet males with definite or probable DBMD diagnosis).

Survey sample. The survey was administered to 364 primary caregivers of 402 males with definite or probable diagnosis of DBMD (living or deceased). In the case of families with multiple affected males, we selected *a priori* to analyze the oldest sibling, in order to meet the statistical assumption of the independence of the observations. The interviews were conducted in two cycles from April 2007 to May 2008 and from April 2009 to March 2012. The cycles corresponded to annual pooling and continued identification of new cases. The dissemination methods were the same across cycles.

The 686 eligible primary caregivers were initially contacted by mail (GA, IA, and wNY), telephone (CO), through the primary health care provider (AZ), or in person (HI). Any differences in methods used to contact the caregivers were due to differences in site institutional review board requirements. An introductory packet was provided to the primary caregiver and contained a letter that described the study in detail, a project brochure, information on the rights of research subjects, and frequently asked questions, along with \$20 compensation. The telephone interviews took between 45 and 60 minutes, were conducted in English or Spanish, and collected information about socio-demographic characteristics of the primary caregivers such as education level and household income, residence history, social support, medical and

assistive technology services received, and information about schooling of the males with DBMD. Follow-up was done using a systematic protocol to obtain consent to conduct the interviews.

The response, cooperation, and refusal rates, calculated using the American Association for Public Opinion Research (AAPOR) calculator, were 53%, 66%, and 28%, respectively (Response Rates - An Overview - AAPOR, n.d.). Response rate was the number of primary caregivers who answered the survey divided by the number of eligible primary caregivers in the sample. The cooperation rate was the proportion of all primary caregivers interviewed to all eligible primary caregivers ever contacted, where contact was verified. It included partial interviews as respondents. The refusal rate was the proportion of all primary caregivers who refused to be interviewed, or who broke off an interview, to all potential primary caregivers. The procedures for data collection were approved by the institutional review board at each site.

Because DMD and BMD differ in their severity and rate of progression, we conducted separate analyses for each of these conditions. To include males with intermediate DMD and take into account the effect of corticosteroid treatment on prolonged ambulation, we categorized males who lost ambulation before 16 years of age as DMD and males who lost ambulation at or after 16th birthday as BMD. If ambulation status was not available at the time of the interview, or the child was too young to determine the type of dystrophinopathy based on ambulation status, age at first signs and symptoms was used to separate Duchenne from Becker (first signs and symptoms before age six years for DMD and after age of six years for BMD). We could not classify one male due to missing information on age at onset of first signs and symptoms. The final analytic sample consisted of 210 males with DBMD who were categorized as DMD ($n = 179$) and BMD ($n = 31$) (see Figure 1).

Variables

We evaluated the following independent variables: enrolled in general education classes (yes/no), grade repetition (yes/no), used instructional assistant (yes/no), and used resource room support (yes/no). A student who is enrolled in school but does not attend general education classes may attend school in the same building as other students, but is in a separate classroom all day. Grade repetition occurs when a child does not make academic progress or is not socially or emotionally moving ahead with his same-age peers. An instructional assistant's role in a general education classroom is to support a student to access the curriculum. A resource room is a classroom where an adapted educational curriculum is delivered to a student with a disability.

We selected these outcomes because, given enough resources, they can be improved (e.g. males with DMD may receive more instructional assistance and resource room support to meet the education requirements thereby reducing the need for grade repetition). Instructional assistant usage and resource room support are both examples of accommodations that improve access to the curriculum for students with special needs. The abstractors did not have access to the school records, so the survey was used to obtain information on school experiences and school needs for males with DBMD. The responses to these questions were used to derive our dependent variables and for exclusion criteria.

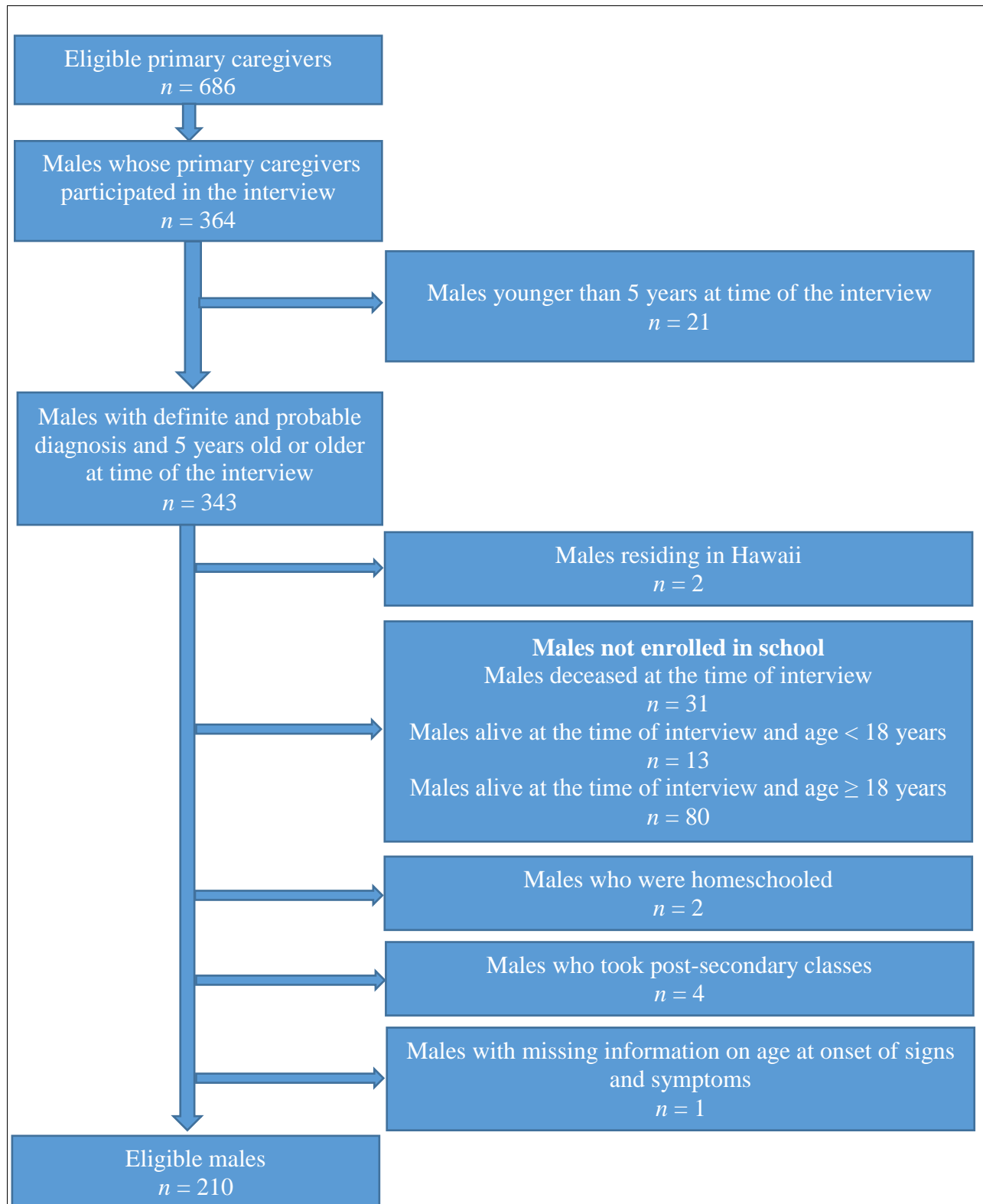


Figure 1. Sample exclusion criteria for males from MD STARnet cohort

The following factors were examined as independent variables: (a) age of male at the time of interview (5-10 years, 11-14 years, and ≥ 15 years), (b) race/ethnicity (White non-Hispanic, Black non-Hispanic, Hispanic, and Other race/ethnicity), (c) state of residence, (d) ambulation status (ambulatory, non-ambulatory, and not known), (e) types (occupational, physical, and speech) and location (only at school, school and medical practice or home, only at medical practice or home, and not received) of therapy received, (f) primary caregiver education level (≤ 12 years, >12 years), and (g) annual household income ($< \$30,000$, $\$30,000$ - $\$50,000$, and $> \$50,000$). The main goals of PT are to improve mobility, to strengthen the large muscle groups, to assist in increasing joint flexibility, and to prevent contractures and scoliosis. OT is focused on the development of fine motor skills that help with accomplishing activities of daily living such as using a computer or dressing. Speech therapy (ST) is recommended for a child with delayed or disordered language and/or articulation skills. We evaluated the ambulatory status and the types of therapies received because they are indicative of the severity of the condition. In addition, we were interested in whether the socio-economic status (SES) of the caregiver was associated with the school experience of boys with DMD and used caregiver education and household income as surrogates for SES.

Statistical Analyses

Descriptive statistics (frequency counts and percentages) were calculated for all independent variables. For males with DMD, crude prevalence ratio (PR) estimates were calculated for evaluating the associations between study subject and caregiver characteristics, and the outcomes. Because the outcomes did not meet the rare event assumption of a prevalence, ($< 10\%$), Poisson regression models with a robust error variance were conducted to estimate adjusted prevalence ratios (aPR) and 95% confidence intervals (Petersen & Deddens, 2008). All multivariable regression models were adjusted for male's age at interview, race/ethnicity, and state of residence. When evaluating the association between household income and the dependent variables, we adjusted for the number of family members supported by the household income along with other confounders. For each type of therapy received, due to small counts in some categories, we created two groups: therapy received and not received. Lastly, we compared the study subject and caregiver characteristics between males with DBMD who were reported as not attending general education classes to males with DBMD reported as attending general education classes. This sub-analysis was conducted to evaluate how comparable this population was to males who attended general education classes, and to evaluate potential characteristics that may explain the differences between these two samples, and, as a result, the potential selection bias. Data management and analyses were conducted using SAS 9.3 software.

Results

Descriptive Statistics

Among the 179 males with DMD, 73.2% were between five and 15 years of age, 69.8% were White non-Hispanic, and 58.1% were non-ambulatory (Table 1). Fifty-four percent were reported as receiving OT in school, 49.2% received PT, and 49.8% received ST. Ninety percent of males were reported as attending general education classes while grade repetition was

reported for 26.3% of subjects with DMD. Use of an instructional assistant was reported for 59.2% and use of a resource room was reported for 58.7% (Table 1). With respect to subjects' primary caregivers, 69.3% reported having more than 12 years of education and 49.7% reported an annual income of >\$50,000. Among the 31 males with BMD, over 71% were age 11 years and older, 61.3% were White non-Hispanic, and 67.7% were ambulatory at the time of the interview (Table 1). About 32.2% received OT in school, 48.4% received PT, and 29.0% received ST. Among primary caregivers of males with BMD, 54.8% reported more than 12 years of education and 67.7% reported an annual income >\$50,000 (Table 1).

Table 1

Distribution of study subject and primary caregiver characteristics in the MD STARnet cohort

Study Subject Characteristics	DMD (<i>n</i> = 179)		BMD (<i>n</i> = 31)	
	<i>n</i>	%	<i>n</i>	%
Age groups				
5-10 years	71	39.7	9	29.0
11-14 years	60	33.5	11	35.5
≥15 years	48	26.8	11	35.5
Race/ethnicity				
White non-Hispanic	125	69.8	19	61.3
Black non-Hispanic	9	5.0	3	9.7
Hispanic	28	15.6	6	19.4
Other	7	3.9	0	
Unknown	10	5.6	3	9.7
State				
AZ	43	24.0	5	16.1
CO	34	19.0	5	16.1
GA	40	22.4	7	22.6
IA	32	17.9	8	25.8
western NY	30	16.8	6	19.4
Ambulation status				
Ambulatory	58	32.4	21	67.7
Non-ambulatory	104	58.1	6	19.4
Don't know	17	9.5	4	12.9
Occupational therapy				
Only at school	74	41.3	9	29.0
School and MP/Home	23	12.9	1	3.2
Only at MP/Home	29	16.2	2	6.5
Not received	50	27.9	19	61.3
Missing	3	1.7	0	
Physical therapy				
Only at school	56	31.3	10	32.3
School and MP/Home	32	17.9	5	16.1
Only at MP/Home	69	38.6	9	29.0

Not received	21	11.7	7	22.6
Missing	1	0.6	0	
Speech therapy				
Only at school	78	43.6	9	29.0
School and MP/Home	11	6.2	0	
Only at MP/Home	13	7.3	2	6.5
Not received	75	41.9	20	64.5
Missing	2	1.1	0	
General education classes				
Participates	162	90.5	30	96.8
Does not participate	17	9.5	1	3.2
Grade repetition				
Yes	47	26.3	6	19.4
No	130	72.6	25	80.6
Missing	2	1.1	0	
Instructional assistant				
Yes	106	59.2	9	29.0
No	72	40.2	22	71.0
Missing	1	0.6	0	
Resource room				
Yes	105	58.7	11	35.5
No	73	40.8	20	64.5
Missing	1	0.6	0	
Caregiver and Household Characteristics				
Education level of caregiver				
>12 years	124	69.3	17	54.8
≤ 12 years	44	24.6	11	35.5
Missing	11	6.2	3	9.7
Household income				
<\$30,000	59	32.9	7	22.6
\$30,000-\$50,000	24	13.4	2	6.5
>\$50,0000	89	49.7	21	67.7
Missing	7	3.9	1	3.2

MD STAR_{net} = Muscular Dystrophy Surveillance Tracking and Research Network; DMD = Duchene muscular dystrophy; BMD = Becker muscular dystrophy; MP = medical practice; AZ = Arizona; CO = Colorado; GA = Georgia; IA = Iowa; NY = western New York State

Crude Analysis

Participation in general education classes. Among males with DMD, residing in CO was significantly associated with participating in general education classes (Table 2). No other statistically significant estimates were observed.

Grade repetition. Of the 47 males with DMD who were reported as repeating a grade, 91.5% repeated a grade in elementary school, 4.2% repeated a grade in junior high school, and 2.1% repeated a grade in senior high school. The likelihood of repeating a grade was significantly higher among males with DMD who received ST at school, or at school and medical practice/home compared to males who did not receive any ST. No other significant associations with grade repetition were observed (Table 2).

Instructional assistant usage. Males with DMD 11 years of age and older were more likely to use an instructional assistant (Table 3). These individuals were significantly more likely to use an instructional assistant if they received OT or ST at school, or at school and medical practice/home. Males with DMD who were ambulatory at the time of the interview were significantly less likely to use an instructional assistant compared to those who were non-ambulatory (Table 3).

Resource room support. Males with DMD 15 years of age and older were less likely to receive support in a resource room. Subjects who received OT at school and medical practice/home, were 1.69 times as likely to use a resource room as males with DMD who did not receive OT. A 68% and 92% increase in the likelihood of using resource room was observed among males with DMD who received ST only at school, or at school and medical practice/home, compared to those who did not receive OT, respectively (Table 3). With respect to education level of the caregiver, the prevalence of resource room use was higher among males whose primary caregivers had less than 12 years of education compared to males whose primary caregiver had at least 12 years of education.

Multivariable Analysis

Table 4 shows the aPR estimates of multivariable Poisson regression models with robust error variance used to evaluate the associations between ambulation status, therapies, primary caregiver education, household income, and the four dependent variables of interest. Compared to males with DMD who were ambulatory at the time of the interview, those who were not ambulating were 75% more likely to use an instructional assistant and 54% more likely to use a resource room. Males with DMD who received OT in school were 1.5 times as likely to use an instructional assistant and 37% more likely to use resource room support as those who did not receive OT. Significantly elevated aPRs were observed for the association between ST received in school and grade repetition, instructional assistant use, and resource room support. Lastly, males whose primary caregivers had less than 12 years of education were significantly more likely to use an instructional assistant or resource room support compared to males with DMD whose primary caregivers had at least 12 years of education.

Separately, we evaluated males with DBMD who did not have reported school enrollment at the time of the interview and were excluded from the results above. Out of 124 males who were reported as not being enrolled in school, 31 (25%) were deceased at the time of the caregiver interview. Out of 93 for whom no participation in general education classes was reported, but were alive at the time of the interview, six (6.45%) were homeschooled. Ninety percent were 15 years old or older ($p < 0.001$), their primary caregivers had a lower educational level than those of males who were enrolled in school ($p = 0.03$), and their annual household income was lower than that of males enrolled in school ($p = 0.17$) (Table 5).

Discussion

We conducted a cross-sectional study to investigate types of supportive school services received by males with DBMD and factors related to provision of these services.

Males with DMD

The current study observed that, of males with DMD enrolled in school, 90.5% were reported as participating in general education classes, 26.3% were reported as repeating a grade, 59.2% were reported as having an instructional assistant, and 58.7% were reported as using the resource room. The current study showed that there was no significant difference in participation in general education classes, grade repetition, use of an instructional assistant or resource room among racial/ethnic groups, among residents of different states, and between various levels of household income. Our results suggest that socioeconomic factors do not play a strong role in the educational experience of boys with DMD.

A study by Magliano et al. (2014), conducted in Italy in eight treatment centers for individuals with muscular dystrophies, reported a similar prevalence of males with DMD who attended school (94.7%) as we observed (90.5%). Our findings of high levels of participation in general education classes suggest that children with relatively severe physical needs are being integrated into the regular classroom with their age-appropriate peers.

The adjusted analysis showed that non-ambulatory males with DMD had a significantly greater prevalence of using an instructional assistant and resource room support compared to those who were ambulating, suggesting that schools need to plan to allocate greater resources for boys with DMD as they grow older and more physically disabled. Instructional assistant use and resource room support are both examples of accommodations made to improve access to the curriculum for students with special needs. In addition, the Muscular Dystrophy Association (MDA) determined that a variety of classroom accommodations and adaptive equipment, such as adaptive seating to support upright posture, raised desktop or wheelchair tray as desk, adapted keyboards, software and calculators, special pencil grip, and use of tape recorder for note-taking can help males with DMD maximize their physical abilities and excel academically (Heller, Mezei, & Avant, 2009; Muscular Dystrophy Association, n.d.). To improve classroom access, it is recommended that males with DMD receive preferential seating and are allowed extra time to

Table 2

Crude prevalence ratio (PR) estimates of the association between study subject and caregiver characteristics and general education classes and grade repetition among DMD cases in the MD STARnet cohort

Study Subject Characteristics	General Education Classes				PR (**P value)	Repeated Grade				*PR (**P value)
	Participates (N = 162)		Does not participate (N = 17)			Repeated (N = 47)		Never repeated (N = 130)		
	n	%	n	%		n	%	n	%	
Age groups										
5-10 years	65	40.1	6	35.3	Referent	15	31.9	54	41.5	Referent
11-14 years	55	34.0	5	29.4	1.00 (0.980)	19	40.4	41	31.5	1.46 (0.210)
≥15 years	42	25.9	6	35.3	0.96 (0.490)	13	27.7	35	26.9	1.25 (0.500)
Race/ethnicity										
White non-Hispanic	115	71.0	10	58.8	Referent	29	61.7	94	72.3	Referent
Black non-Hispanic	9	5.6	0		---	2	4.3	7	5.4	0.94 (0.930)
Hispanic	22	13.6	6	35.3	0.86 (0.120)	8	17.0	20	15.4	1.21 (0.570)
Other	6	3.7	1	5.9	0.93 (0.650)	3	6.4	4	3.1	1.82 (0.200)
Unknown	10	6.2	0		----	5	10.6	5	3.9	2.12 (0.030)
State										
AZ	36	22.2	7	41.2	Referent	13	27.7	29	22.3	Referent
CO	33	20.4	1	5.9	1.16 (0.040)	9	19.2	25	19.2	0.86 (0.670)
GA	36	22.2	4	23.5	1.08 (0.400)	12	25.5	27	20.8	0.99 (0.990)
IA	30	18.5	2	11.8	1.12 (0.160)	6	12.8	26	20.0	0.61 (0.250)
Western NY	27	16.7	3	17.7	1.08 (0.430)	7	14.9	23	17.7	0.75 (0.480)
Ambulation status										
Non-ambulatory	92	56.8	12	70.6	Referent	31	66.0	73	56.2	Referent
Ambulatory	54	33.3	4	23.5	1.05 (0.310)	12	25.5	45	34.6	0.71 (0.240)
Don't know	16	9.9	1	5.9	1.06 (0.380)	4	8.51	12	9.2	---
Occupational therapy										
Not received	47	29.0	3	17.7	Referent	10	21.3	40	30.8	Referent
Only at School	67	41.4	7	41.2	0.96 (0.470)	20	42.6	54	41.5	1.35 (0.380)

School and MP/Home	19	11.7	4	23.5	0.89 (0.210)	6	12.8	16	12.3	1.36 (0.500)
Only at MP/Home	27	16.7	2	11.8	0.99 (0.880)	10	21.3	18	13.9	1.79 (0.130)
Missing	2	1.2	1	5.9	--	1	2.1	2	1.5	--
Physical therapy										
Not received	20	12.4	1	5.9	Referent	6	12.8	15	11.5	Referent
Only at School	47	29.0	9	52.9	0.88 (0.100)	20	42.6	36	27.7	1.25 (0.570)
School and MP/Home	28	17.3	4	23.5	0.92 (0.310)	7	14.9	25	19.2	0.77 (0.580)
Only at MP/Home	67	41.4	2	11.8	1.02 (0.710)	14	29.8	53	40.8	0.73 (0.460)
Missing	0		1	5.9	---	0		1	0.8	---
Speech therapy										
Not received	71	43.8	4	23.5	Referent	10	21.3	65	50.0	Referent
Only at School	70	43.3	8	47.1	0.95 (0.260)	29	61.7	48	36.9	2.82 (0.002)
School and MP/Home	9	5.6	2	11.8	0.86 (0.310)	5	10.6	6	4.6	3.41 (0.006)
Only at MP/Home	11	6.8	2	11.8	0.89 (0.360)	2	4.3	10	7.7	1.25 (0.750)
Missing	1	0.6	1	5.9	---	1	2.1	1	0.8	---

Caregiver and Household Characteristics

Education level of caregiver										
>12 years	117	72.2	7	41.2	Referent	30	63.8	92	70.8	Referent
≤ 12 years	37	22.8	7	41.2	0.89 (0.100)	13	27.7	31	23.9	1.20 (0.510)
Missing	8	4.9	3	17.7	---	4	8.5	7	5.4	---
Household income										
<\$30,000	51	31.5	8	47.1	Referent	17	36.2	42	32.3	Referent
\$30,000-\$50,000	21	13.5	3	17.7	1.01 (0.900)	8	17.0	16	12.3	1.16 (0.680)
>\$50,0000	83	51.2	6	35.3	1.08 (0.200)	20	42.6	67	51.5	0.80 (0.430)
Missing	7	4.3	0		---	2	4.3	5	3.9	---

*Two cases had Repeated Grade = "Do not know" and were excluded; ** Z statistic; DMD = Duchenne muscular dystrophy; MD STARnet = Muscular Dystrophy Surveillance Tracking and Research Network; MP = medical practice; AZ = Arizona; CO = Colorado; GA = Georgia; IA = Iowa; NY = western New York State; PR = Prevalence Ratio

Table 3

Crude prevalence ratio (PR) estimates of the association between various study subject and primary caregiver characteristics and usage of school aid and resource room among DMD cases in the MD STARnet cohort

Study Subject Characteristics	Instructional Assistant				*PR (**P value)	Resource Room Support				*PR (**P value)
	Has an assistant (N = 106)		Does not have an assistant (N = 72)			Receives support (N = 105)		Does not receive support (N = 73)		
	n	%	n	%		n	%	n	%	
Age groups										
5-10 years	30	28.3	41	56.9	Referent	46	43.8	24	32.9	Referent
11-14 years	42	39.6	17	23.6	1.68 (0.001)	37	35.2	23	31.5	0.94 (0.630)
≥15 years	34	32.1	14	19.4	1.67 (0.002)	22	21.0	26	35.6	0.70 (0.040)
Race/ethnicity										
White non-Hispanic	75	70.8	50	69.4	Referent	72	68.6	53	72.6	Referent
Black non-Hispanic	6	5.7	2	2.8	1.25 (0.300)	5	4.8	4	5.5	0.96 (0.910)
Hispanic	15	14.2	13	18.1	0.89 (0.550)	17	16.2	10	13.7	1.09 (0.590)
Other	4	3.8	3	4.2	0.95 (0.880)	6	5.7	1	1.4	1.48 (0.020)
Unknown	6	5.7	4	5.6	1.00 (1.000)	5	4.8	5	6.9	0.88 (0.660)
State										
AZ	27	25.5	16	22.2	Referent	27	25.7	16	21.9	Referent
CO	22	20.8	12	16.7	1.03 (0.860)	20	19.1	14	19.2	0.94 (0.720)
GA	17	16.0	22	30.6	0.69 (0.100)	23	21.9	16	21.9	0.94 (0.720)
IA	21	19.8	11	15.3	1.05 (0.800)	17	16.2	15	20.6	0.85 (0.410)
Western NY	19	17.9	11	15.3	1.01 (0.960)	18	17.1	12	16.4	0.96 (0.810)
Ambulation status										
Non-ambulatory	75	70.8	28	38.9	Referent	64	61.0	39	53.4	Referent
Ambulatory	25	23.6	33	45.8	0.59 (0.001)	33	31.4	25	34.3	0.92 (0.520)
Don't know	6	5.7	11	15.3	---	8	7.6	9	12.3	---
Occupational therapy										
Not received	21	19.8	29	40.3	Referent	24	22.9	25	34.3	Referent
Only at School	50	47.2	24	33.3	1.61 (0.010)	46	43.8	28	38.4	1.27 (0.170)

School and MP/Home	15	14.2	7	9.7	1.62 (0.030)	19	18.1	4	5.5	1.69 (0.003)
Only at MP/Home	17	16.0	12	16.7	1.40 (0.140)	15	14.3	14	19.2	1.06 (0.810)
Missing	3	2.8	0		---	1	0.9	2	2.7	---
Physical therapy										
Not received	12	11.3	9	12.5	Referent	12	11.4	9	12.3	Referent
Only at School	37	34.9	19	26.4	1.16 (0.490)	33	31.4	22	30.1	1.05 (0.820)
School and MP/Home	20	18.9	12	16.7	1.09 (0.700)	23	21.9	9	12.3	1.26 (0.290)
Only at MP/Home	36	34.0	32	44.4	0.93 (0.730)	36	34.3	33	45.2	0.91 (0.680)
Missing	1	0.9	0		---	1	0.9	0		---
Speech therapy										
Not received	35	33.0	40	55.6	Referent	32	30.5	43	58.9	Referent
Only at School	54	50.9	24	33.3	1.48 (0.006)	56	53.3	22	30.1	1.68 (0.001)
School and MP/Home	8	7.6	2	2.8	1.71 (0.007)	9	8.6	2	2.7	1.92 (0.001)
Only at MP/Home	9	8.5	4	5.6	1.48(0.080)	7	6.7	6	8.2	1.26 (0.420)
Missing	0		2	2.8	---	1	0.9	0		---
Caregiver and Household Characteristics										
Education level of caregiver										
>12 years	68	64.2	55	76.4	Referent	66	62.9	58	79.5	Referent
≤ 12 years	30	28.3	14	19.4	1.23 (0.110)	30	28.6	13	17.8	1.31 (0.040)
Missing	8	7.6	3	4.2	---	9	8.6	2	2.7	---
Household income										
<\$30,000	35	33.0	23	31.9	Referent	38	36.2	20	27.4	Referent
\$30,000-\$50,000	17	16.0	7	9.7	1.17 (0.340)	13	12.4	11	15.1	0.83 (0.370)
>\$50,000	50	47.2	39	54.2	0.93 (0.610)	50	47.6	39	53.4	0.86 (0.250)
Missing	4	3.8	3	4.2	---	4	3.8	3	4.1	---

*One case had Instructional Assistant= "Do not know" and was excluded; *One case had Resource Help = "Do not know" and was excluded; ** Z statistic; DMD = Duchenne muscular dystrophy; MD STARnet = Muscular Dystrophy Surveillance Tracking and Research Network; MP = medical practice; AZ = Arizona; CO = Colorado; GA = Georgia; IA = Iowa; NY = western New York State; PR = Prevalence Ratio

Table 4

Adjusted prevalence ratio (aPR) of the association between various study subject and primary caregiver characteristics and public school enrollment, grade repetition, usage of school aid, and resource room help among DMD cases in the MD STARnet cohort

	General Education Classes		Repeated Grade		Instructional Assistant		Resource Room Support	
	aPR	*P value	aPR	*P value	aPR	*P value	aPR	*P value
Ambulatory status (ref= ambulatory)	0.97	0.390	1.29	0.610	1.75	0.009	1.54	0.020
Occupational therapy (ref = not received)	0.95	0.410	1.40	0.290	1.50	0.010	1.37	0.050
Physical therapy (ref = not received)	0.97	0.580	0.85	0.680	0.98	0.910	1.03	0.870
Speech therapy (ref = not received)	0.94	0.220	2.57	0.005	1.44	0.003	1.66	0.001
Caregiver Education (ref >12years)	0.93	0.130	1.46	0.580	1.49	0.030	1.36	0.040
Household income (ref < 50,000)	1.06	0.100	0.66	0.340	0.94	0.620	0.89	0.450

aPR = adjusted Prevalence Ratio; *Z statistic; DMD = Duchenne muscular dystrophy; MD STARnet = Muscular Dystrophy Surveillance Tracking and Research Network; Adjustment was done for: age at interview, race/ethnicity, and state of residence. In addition, when looking at the household income, we adjusted for the number of family members supported by that income.

Table 5

Distribution of DBMD case and primary caregiver characteristics by participation in general education classes, MD STARnet cohort

Study Subject and Caregiver Characteristics		Alive and Reported No General Education Classes (<i>n</i> = 93)		Attended General Education Classes (<i>n</i> = 210)		*P value
		<i>n</i>	%	<i>n</i>	%	
Age groups						<0.001
	5-10 years	8	8.6	80	38.1	
	11-14 years	1	1.1	71	33.8	
	≥15 years	84	90.3	59	28.1	
Race/ethnicity						0.290
	White non-Hispanic	64	68.8	144	68.6	
	Black non-Hispanic	4	4.3	12	5.7	
	Hispanic	15	16.1	34	16.2	
	Other	0	0.0	7	3.3	
	Unknown	10	10.8	13	6.2	
State						0.790
	AZ	23	24.7	48	22.9	
	CO	16	17.2	39	18.6	
	GA	24	25.8	47	22.4	
	IA	19	20.4	40	19.0	
	Western NY	11	11.8	36	17.1	
Education level of caregiver						0.030
	>12 years	47	50.5	141	67.1	
	≤ 12 years	34	36.6	55	26.2	
	Missing	12	12.9	14	6.6	
Household income						0.170
	<\$30,000	27	32.3	66	31.4	
	\$30,000-\$50,000	16	17.2	26	12.4	
	>\$50,000	34	36.6	110	52.4	
	Missing	16	14.0	8	3.8	

*Chi-square test; DBMD = Duchenne and Becker muscular dystrophy; MD STARnet = Muscular Dystrophy Surveillance, Tracking, and Research Network; SD = Standard Deviation; IQR = interquartile range; AZ = Arizona; CO = Colorado; GA = Georgia; IA = Iowa; NY =western New York state

transition between classes. Physical assistance for transfers and physical tasks is also recommended when necessary (EndDuchenne.org - Leading the fight to end Duchenne muscular dystrophy - Parent Project Muscular Dystrophy, n.d.).

In our study, about one-half of males with DMD were reported as receiving OT or PT at school. Zebracki and Drotar (2008) and Pandya et al. (2015) also reported high prevalence of recipients of occupational and physical therapies, however their study did not differentiate between therapies received at school or other facilities. Interestingly, in a study on variation in educational services receipt among U.S. children with developmental conditions (autism spectrum disorder, developmental delay, and intellectual disability), Lindly, Sinche, and Zuckerman (2015) observed that children with intellectual disability were more likely to have received one or more school-based therapies (OT, PT, social skills training, ST, or language therapy) than those with autism and developmental delay, while children with developmental delay were less likely than the rest of the sample to have received one or more school-based therapies. While we did not measure the level of intellectual delay/disability in our study sample, we do know that 20-50% of boys with DMD are diagnosed with cognitive deficits (Wicksell et al., 2007). Evaluating cognitive deficits and the receipt of therapies for cognitive disorders was beyond the scope of our study. Therefore, future studies should evaluate whether all students with cognitive disorders due to DMD and who need therapy, are receiving it at school, and examine the characteristics associated with having or not having their therapy needs identified in a timely manner.

In the current study, 57% of males with DMD were reported as receiving ST. In a study by Cyrulnik et al. (2007) on delayed developmental language milestones in children with DMD, speech delay was observed in 38% of boys with DMD. One explanation for the high prevalence of ST use in our study is that, although receiving ST could be an indicator of speech delay, the interview used in the current study was not designed to evaluate any language milestones. In our study, males with DMD who received ST had a statistically increased prevalence for grade repetition, instructional assistant use, and resource room use when compared to non-recipients.

Higher rates of speech and language deficits among children with DMD are well documented. Deficiency in phonological processing combined with problems in short-term memory could result in learning disability (Bushby et al., 2010). Hinton et al. (2004) recommended educational interventions such as breaking detailed verbal instructions into smaller segments, repeating verbal information, and phonological awareness training. The survey used in our study did not distinguish among the conditions for which ST was provided.

Lastly, the multivariable model showed that males with DMD whose caregivers had less than 12 years of education were significantly more likely to use an instructional assistant and resource room support than those whose caregivers had at least 12 years of education. No literature to date has explored the relationship between the education level of caregivers of children with DMD and the use of school-based services.

Males with BMD

Out of 31 males with BMD in our sample, 97% reported participation in general education classes, a prevalence similar to that observed by Magliano et al. (2014). In the current study, 19% reported grade repetition, and 29% and 35.5% respectively reported instructional assistant and resource room use. In addition, between one-third and one-half of males with BMD reported receiving PT, OT, or ST in school.

Strengths

This is the first study to date that describes school experiences and the accommodations and services that males with DMD or BMD receive in school. MD STAR_{net} is a comprehensive population-based cohort identified through neuromuscular clinics, hospitals and hospital discharge databases, private physicians, service sites for children with special care needs, and birth defects surveillance programs in six states. Definition/classification of DBMD was reviewed by experienced clinicians and diagnosis was confirmed by laboratory data and/or family pedigree. Health data were collected using medical records and therefore were not subject to recall bias.

Limitations

The males identified through the MD STAR_{net} cohort may not be representative of the entire DBMD population in the U.S., since the participant states were not randomly selected. Additionally, a small number of males with DBMD may not have interacted with the specialty clinics and, therefore, those cases were not captured. The survey response rate of 53% may limit the generalizability of our findings to the entire MD STAR_{net} population. However, we compared selected study subject and parental characteristics between eligible MD STAR_{net} families and interviewed families, and we observed similar distribution of case status, child year of birth, vital status, maternal and paternal age at interview, race/ethnicity, and maternal and paternal education level. Therefore, the potential selection bias caused by the relatively low response rate is minimal.

The survey data did not differentiate between types of schools (e.g., public, private, or parochial) or the location of schools (rural versus urban), characteristics that may impact the resources available to meet the needs of children with disabilities. The survey did not collect information related to speech, language, learning, cognitive, and behavioral characteristics that may impact the school experience and learning/school success. Also, although many of the males with DMD and BMD were enrolled in general education classes, the survey did not collect information on the amount of time they spent in those classes. Regarding the various types of therapy received at school, the interview did not determine whether these males qualified for any of these therapies. Children may qualify for, but choose not to receive, therapies at school.

Implications for Practice and Future Research

Schools can use these data to plan and allocate their resources to ensure that classroom accommodations and adaptive equipment that can help males with DBMD are being provided. Teachers and school-based therapists can use these data to ensure they understand the progress of DBMD and its impact on classroom activities. Parents can use these results to advocate for resources that boys with DBMD may need in order to thrive in the school setting. Policymakers can use these results to ensure that laws pertaining to special education appropriately accommodate the needs of this medically and cognitively complex population. Since our study did not examine compliance, future studies should also survey schools to identify compliance with current laws regarding special needs accommodations, barriers to identification of educational needs in children with progressive physical disabilities as well as potential cognitive deficits, and provision of services to boys with DBMD. In addition, our study did not examine the quality of the services received by boys with DBMD. Future studies should more extensively survey families of boys with DBMD to evaluate the quality of educational assistance at home and at school. Combined, these data can serve to provide adequate assistance in school and improve the academic achievement of boys with DBMD.

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