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Duchenne and Becker Muscular Dystrophies Prevalence in MD STARnet surveillance sites: An Examination of Racial and Ethnic Differences

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Abstract

Introduction: Previous studies indicated variability in the prevalence of Duchenne and Becker muscular dystrophies (DBMD) by racial/ethnic groups. The Centers for Disease Control and Prevention's (CDC) Muscular Dystrophy Surveillance, Tracking, and Research network (MD STAR*net*) conducts muscular dystrophy surveillance in multiple geographic areas of the United States and continues to enroll new cases. This provides an opportunity to continue investigating differences in DBMD prevalence by race and ethnicity, and to compare the impact of using varying approaches for estimating prevalence.

Objective: To estimate overall and race/ethnicity-specific prevalence of DBMD among males aged 5 to 9 years and compare the performance of three prevalence estimation methods.

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Author Contributions

J. Mann, S. McDermott and B. Cai conceived of the study goals and design. Y. Zhang, K. James, and B. Cai conducted data analyses. Y. Zhang, J. Mann, and S. McDermott authored the original manuscript. K. Conway, P. Paramsothy, and T. Smith provided input and advice about study methods and read and edited drafts of the paper.

Disclosure Statement

The authors have no conflicts of interest to declare.

Statement of Ethics

This research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Informed consent was not obtained due to the nature of the research (clinical record review), and data collection was conducted under public health authority, institutional review board approval or exemption, or both for all sites.

Methods: The overall and race/ethnicity-specific 5-year period prevalence rates were estimated with MD STAR*net* data using three methods. Method 1 used the median of 5 yearly prevalence and Methods 2 and 3 calculated prevalence directly with different birth cohorts. To compare prevalence between racial/ethnic groups, Poisson modelling was used to estimate prevalence ratios (PRs) with non-Hispanic (NH) whites as the referent group. Comparison between methods was also conducted.

Results: In the final population-based sample of 1,164 DBMD males, the overall 5-year prevalence for DBMD among 5 to 9 years of age ranged from 1.92-2.48 per 10,000 males; 0.74-1.26 for NH blacks, 1.78-2.26 for NH whites, 2.24-4.02 for Hispanics and 0.61-1.83 for NH American Indian or Alaska Native and Native Hawaiian or Pacific Islander (AIAN/API). The PRs for NH blacks/NH whites, Hispanics/NH whites and NH AIAN/API/NH whites were 0.46 (95%CI: 0.36,0.59), 1.37 (1.17,1.61), and 0.61 (0.40,0.93), respectively.

Conclusions: In males aged 5 to 9 years, compared to the prevalence of DBMD in NH whites, prevalence in NH blacks and NH AIAN/API was lower and higher in Hispanics. All methods produced similar prevalence estimates; however, Method 1 produced narrower confidence intervals and Method 2 produced fewer zero prevalence estimates than the other two methods.

Keywords

Duchenne-Becker Muscular Dystrophy; Epidemiology; Race/Ethnicity

Introduction

Duchenne and Becker muscular dystrophies (DBMD) are genetic x-linked conditions caused by a mutation in the *DMD* gene that alters dystrophin production (1, 2). Variability in dystrophin production is associated with a spectrum of severity in DBMD (3). Duchenne muscular dystrophy (DMD) typically presents with onset of symptoms before age 5 years, rapid progression of severe muscle weakness that leads to impaired ambulation by age 12 years, and death due to respiratory failure or cardiac conditions in young adulthood (2, 4). Becker muscular dystrophy (BMD) has a later onset, slower progression and less severe muscle weakness, and longer life expectancy (5).

DBMD are rare conditions, which in the United States refers to conditions that affects fewer than 200,000 people nationally (6). Estimating prevalence of rare conditions can be challenging due to scarce epidemiological reports and measuring small populations (7). The first US population-based prevalence study of DBMD used data from the Muscular Dystrophy Surveillance, Tracking, and Research network (MD STAR*net*) (8). In this study, point prevalence among males born from 1983-2002 who resided in one of four MD STAR*net* sites was estimated by site and age group. The estimated point prevalence, per 10,000, in 2007 was 1.3-1.8 among those aged 5–24 years and 0.9–1.9 among those aged 5–9 years (8). More recently, period prevalence was estimated for 6 MD STAR*net* sites among males born from 1982–2011 who resided in at least one site (9). The period prevalence estimates, in 5-year periods from 1991 to 2010, for males aged 5-9 years ranged from 1.51-2.05 per 10,000.

Decisions about whether to calculate point prevalence or period prevalence may be impactful when conditions are rare and there are variable rates of detection or survival across age. For example, the MD STAR*net* prevalence estimates were shown to vary by geographical region (8), race/ethnicity (8), and age group (8, 9). Other studies have also reported variability in prevalence across racial/ethnic groups in different geographic areas (10–27). Therefore, we examined three approaches to calculating prevalence. Specifically, we estimated DBMD prevalence among males aged 5 to 9 years by combining two DBMD birth cohorts from MD STAR*net*. We also stratified by race/ethnicity and compared the performance of the three different prevalence estimation methods across these strata.

Methods

MD STAR*net*, a population-based surveillance system funded by the Centers for Disease Control and Prevention, began data collection in 2004 in four sites (Arizona, Colorado, Iowa, and 12 counties in western New York). Georgia and Hawaii began data collection in 2006 and 2010 respectively. Eligible cases were diagnosed with DBMD before age 21 years, were born from January 1, 1982 through December 31, 2011 and resided in a catchment area at some point after diagnosis through December 31, 2011 or December 31, 2012 if identified after September 1, 2011 (first dataset). In 2015, data collection began in 6 sites (Colorado, Iowa, 21 counties in western New York, the Piedmont region of North Carolina, South Carolina, Utah/Nevada) (second dataset). Eligible cases were diagnosed with DBMD before age 21, were born from January 1, 2000 through December 31, 2015 and resided in a catchment area at some point during the study.

MD STAR*net* surveillance methods have been described in depth previously (9, 28, 29). For all sites, case sources included healthcare facilities, vital records, and state hospital discharge data. Potential cases were identified in medical records using International Classification of Diseases (ICD), 9th Revision, Clinical Modification (CM) code 359.1 and death certificates using ICD, 10th Revision, CM code G71.0. De-identified clinical data were reviewed by a neuromuscular physician at each site and the multi-site clinical review committee to assign a case definition (definite, probable, possible, asymptomatic, and manifesting female) (29). Definite DBMD was defined as documented clinical symptoms of DBMD and confirmatory diagnostic testing by DNA analysis or muscle biopsy in the index case or family member. Probable DBMD was defined as clinical symptoms and X-linked family history of DBMD but no diagnostic confirmation. Possible DBMD was defined as clinical symptoms without confirmatory diagnostic testing or family history. Asymptomatic DBMD was defined as confirmatory diagnostic testing without clinical symptoms and manifesting female was defined as confirmatory diagnostic testing with clinical symptoms in a female.

All sites, except Utah, obtained public health authority to collect surveillance data. All sites had Institutional review board (IRB) approval or exemption.

Study sample

Due to incomplete case ascertainment or truncated data collection, 45 cases from Hawaii or Nevada were not included. Details about the remaining sites are provided in Table 1. There

were overlapping cases between the two datasets among sites (CO, IA, 12 western counties of NY) that contributed to both datasets. We used data from the most recent dataset when inconsistencies were found in overlapping cases. We only included males who were classified as definite or probable DBMD and born from January 1, 1982 to December 31, 2010. Each case's self-defined race/ethnicity was identified from the medical record. If missing, parent's race/ethnicity from the birth certificate was used. Race/ethnicity was categorized into 5 groups: Hispanic (any race), non-Hispanic (NH) white, NH black, NH American Indian, Alaska Native, Native Hawaiian or Pacific Islander (AIAN/API), and other (including other, multiple, and unknown). The site that submitted data for pooling was used to assign surveillance site. Year of death was identified from medical or vital records. For patients not identified as deceased, year of loss-to-follow-up was assumed to be the year at last clinical visit (9).

We defined 5-year study periods as: P1 (1991–1995), P2 (1996–2000), P3 (2001–2005), P4 (2006–2010) and P5 (2011–2015).

The National Center for Health Statistics (NCHS) releases bridged-race population estimates for the July 1st population of the United States. We used the 1990–2017 NCHS online database as the denominator source for overall population and four race/ethnicity groups (Hispanic, NH white, NH black, and NH AIAN/API). Details can be obtained from https://wonder.cdc.gov/Bridged-Race-v2017.HTML (30).

Statistical analysis

Three methods were used to estimate prevalence (Supplement Table 1). Method 1 calculated period prevalence by taking the median of annual point estimates. Due to the different times of case enrollment by sites, denominators were composed from different populations. For example, in New York, patients born in 2000 would be collected from 12 original plus 9 additional counties. They would reach 5 years old in 2005. The denominator to calculate the prevalence in 2005 included males 5 to 9 years of age from the original 12 counties and 5-year-old males from the 9 new counties. In the following year, 2006, they would become 6 years old and a new group of 5-year-old males born in 2001 were also included. Therefore, the denominator in 2006 was now a combination of 5 to 9 years of age from 12 counties and 5 to 6 years of age from 9 counties.

In Method 2, the numerator was the total number of cases who were 5 to 9 years of age in at least one year during the study period. The denominator was the population at the last year of a study period. For example, males who were 9 years old in the first year would be 13 years old in the last year. Thus, the denominator was the population who were 5 to 13 years of age in the last calendar year. An important contrast between this method and Method 1 was that Method 2 calculated period prevalence directly.

Method 3 introduced birth cohorts for each study period, P1–P5: 1986–1990, 1991–1995, 1996–2000, 2001–2005, and 2006–2010. For instance, the numerator of prevalence for P1 (1991-1995) was the number of cases born from 1986-1990. The denominator was the population 5 to 9 years of age at the last year of a study period. The key difference that

distinguishes Method 3 from Method 2 is that in Method 3, case identification was restricted within a birth cohort. This method was used in a recent study by Romitti et al. (9).

For all three methods, cases who died or were lost-to-follow-up before each study period were subtracted from the numerator. The 95% confidence intervals (CIs) of prevalence were estimated with the Bootstrap percentile for Method 1 and were estimated with Wilson score intervals for Methods 2 and 3 using the 'boot' and 'DescTools' packages in R version 3.6.1 (31, 32).

To compare prevalence by race/ethnicity, a prevalence ratio (PR) was estimated by fitting Poisson regression, adjusting for study periods and using NH white males as the reference group. To evaluate differences in prevalence estimates across methods, Poisson models were fitted with the method indicator as the main variable of interest. We included race/ethnicity, study period, and a multiplicative interaction between method and race/ethnicity to control for potential confounding among these factors. Poisson models were estimated using SAS 9.4 (Cary, NC).

Patient characteristics were described in frequency and percentage, and by case definition, race/ethnicity, site, and eligibility. All tests were two-tailed, and type I error probability was set at 0.05.

Results:

Comparison of methods:

We did not find statistically significant differences in prevalence estimates across the three methods. However, the performance of methods varied on width of the 95% CIs and frequency of zero prevalence. Method 1 had the narrowest CI, followed by Method 2 (Fig. 1). For race/ethnicity-site-study period specific prevalence (supplemental table), zero estimates appeared 28 times for Method 1, 21 times for Method 2, and 30 times for Method 3. Given that Method 1 provided the most precise prevalence estimates, the following results described were estimated using Method 1.

Main results on DBMD prevalence:

Among the 1,164 DBMD males, most were classified as definite (93.4%) and NH white (61.2%) (Table 2).

The overall prevalence, per 10,000 males, of DBMD ranged from 1.92-2.48 for study periods P1–P5 (Table 3). In general, prevalence estimates were lowest for NH blacks and NH AIAN/API and highest for Hispanics (Table 3 and Fig. 1). Prevalence ranged from 0.74-1.26 for NH blacks and 1.78-2.26 for NH whites. Among Hispanics, prevalence estimates were 4.02 and 3.98 during P1 and P2, respectively, but became roughly comparable to NH whites from P3 through P5 (P3–P4: 2.24; P5: 2.57). In the NH AIAN/API racial group, prevalence ranged from 0.61 in P2 to 1.83 in P4. A detailed table of prevalence for DBMD and 95% CIs by race/ethnicity, site and study period are shown in the supplementary Table 2 and Figure 1.

Adjusted prevalence ratios by race/ethnicity groups showed NH blacks had 0.46 times the prevalence of NH whites; NH AIAN/APIs had 0.61 times the prevalence of NH whites; and Hispanics had 1.37 times the prevalence of NH whites. (Table 4)

Discussion:

There are two important findings in this study. First, by Method 1, the overall 5-year prevalence of DBMD ranged from 1.92-2.48 per 10,000 males aged 5–9 years. Prevalence estimates from Method 3, which aligned with that used by Romitti et al., agreed with estimates from 6 sites (9). The only difference was the decreased prevalence for the last study period (2006-2010), which could be explained by delayed diagnosis in younger ages.

Second, we also confirmed findings from other studies that demonstrate racial/ethnic differences in DBMD prevalence (10–27, 33). For example, a South African study with 143 DMD patients diagnosed during 1987–1992 found substantial variation in DMD minimum prevalence (per 10,000) in blacks of 0.04 compared to 0.12 for whites, 0.71 for Indians, and 0.22 for mixed ancestry (10). There are also studies that did not compare racial/ethnic groups, but their population were compromised by one major race/ethnic group. For example, DMD prevalence from studies in Japan and Hong Kong, where the majority race is Asian, have ranged from 0.50-0.98 (12, 16, 21, 27). Studies from European countries, where whites are the majority, have reported DMD prevalence ranges from 0.20-2.50 (11, 13–15, 17, 18, 22–26). A study in Puerto Rico, where Hispanics are the majority, found that the minimum prevalence for all age groups was 0.8 per 10,000 males in 2012 (33). Our new findings are in line with the previous analysis of MD STAR*net* data, but with 32.1% new cases (9). Prevalence of DBMD in NH blacks was less than half the prevalence of that as in NH whites, a difference that was robust across the three methods. Prevalence in NH AIAN/APIs was lower than that in NH whites and Hispanics. However, the small number of cases led to wider confidence intervals. Prevalence in Hispanics was higher than that in NH whites. We also found that the prevalence decreased in Hispanics from P1–P2 to P3–P5. As shown in Table 3, this decrease in prevalence was due to an increase in the population (denominator) of Hispanics that was not accompanied by a commensurate increase in DBMD cases.

We do not have sufficient information available to test or make conclusions on the reasons why prevalence declined among Hispanics. We can speculate that possible contributing factors may include changes in demographics among the Hispanic population in the United States and across the MD STAR*net* participating states (34, 35) or delayed diagnosis among Hispanics (36, 37). Another possibility is purely random variation; while the trend for decline in prevalence is statistically significant ($p = 0.0056$), purely random variation is within the realm of reasonable possibility. Additional research with a sufficiently large Hispanic population will be needed to identify cases to evaluate whether the decline in DBMD prevalence among Hispanics is sustained and, if so, which of the above factors may contribute to it.

A strength of this study is that it is currently the largest population-based study of DBMD prevalence in the United States. Patients were identified from 8 sites, which added a number

of people from race/ethnicity minority groups, although the underlying population demographics do not necessarily reflect those of the entire United States. Another strength of the study was the use of a novel method (Method 1) and two other approaches (Method 2 & 3) to calculate prevalence. Methods 2 and 3 are the most widely used approaches with the denominator defined as the average population or population at the mid-intervals of the period (8). Both require a constant population assumption, which is valid when there is not a substantial change in the underlying population. Method 2 performs well in reducing chances of zero estimates, since more cases are included in the numerator. However, Method 1 does not require an assumption of constant population and can work with data from any length of time and over various population dynamics. Another advantage for Method 1 is the straightforward calculation and the narrowest confidence intervals based on point estimates. In this study, although results using the three methods were comparable, Method 1 is preferred due to its increased precision. However, an understanding of the pros and cons for each method is needed and the methodology should be chosen according to the characteristics of interest in future studies. Statistical simulations should be conducted with other study populations to compare methods under numerous conditions.

Despite study strengths, there are also limitations to this study. First, surveillance was limited to 8 US sites. It is possible that prevalence in these sites differs from that in other states. Second, surveillance was conducted primarily at neuromuscular clinics in the selected geographic areas. Although it is considered standard of care for individuals with DBMD to receive care in specialized clinics; we do not know whether some cases may have received care in settings not included in surveillance activities. Racial/ethnic differences in accessing to healthcare services might introduce selection bias in the estimated prevalence as well as for the prevalence ratios. Finally, for racial/ethnic specific prevalence, we did not include cases who were reported as other, multiple or missing race/ethnicity which may result in an underestimation of prevalence within these groups and biased prevalence ratios.

This study confirmed racial/ethnic differences in DBMD prevalence among males aged 5 to 9 years. We also demonstrated agreement in prevalence estimation across three methods with the smallest confidence interval when the midpoint was used. The prevalence calculation approaches described can be used for other rare conditions.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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References:

1. Flanigan KM. Duchenne and Becker muscular dystrophies. Neurologic clinics. 2014;32(3):671–88, viii. [PubMed: 25037084]
2. Ryder S, Leadley RM, Armstrong N, Westwood M, de Kock S, Butt T, et al. The burden, epidemiology, costs and treatment for Duchenne muscular dystrophy: an evidence review. Orphanet journal of rare diseases. 2017;12(1):79. [PubMed: 28446219]
3. Monaco AP, Bertelson CJ, Liechti-Gallati S, Moser H, Kunkel LM. An explanation for the phenotypic differences between patients bearing partial deletions of the DMD locus. Genomics. 1988;2(1):90–5. [PubMed: 3384440]
4. Yiu EM, Kornberg AJ. Duchenne muscular dystrophy. Journal of paediatrics and child health. 2015;51(8):759–64. [PubMed: 25752877]
5. U.S. Department of Health & Human Services NIoH, NCATS. Becker muscular dystrophy 2016 [2019/7/16]. Available from: https://rarediseases.info.nih.gov/diseases/5900/becker-muscular-dystrophy.
6. U.S. Department of Health & Human Services NIoH, NCATS. FAQs About Rare Diseases 2017 [2019/7/16]. Available from: https://rarediseases.info.nih.gov/diseases/pages/31/faqs-about-rare-diseases.
7. Auvin S, Irwin J, Abi-Aad P, Battersby A. The problem of rarity: estimation of prevalence in rare disease. Value Health. 2018;21(5):501–7. [PubMed: 29753345]
8. Romitti P, Puzhankara S, Mathews K, Zamba G, Cunniff C, Andrews J, et al. Prevalence of Duchenne/Becker muscular dystrophy among males aged 5-24 years-four states, 2007. Morbidity and Mortality Weekly Report. 2009;58(40):1119–22. [PubMed: 19834452]
9. Romitti PA, Zhu Y, Puzhankara S, James KA, Nabukera SK, Zamba GKD, et al. Prevalence of Duchenne and Becker muscular dystrophies in the United States. Pediatrics. 2015;135(3):513–21. [PubMed: 25687144]
10. Ballo R, Viljoen D, Beighton P. Duchenne and Becker muscular dystrophy prevalence in South Africa and molecular findings in 128 persons affected. South African Medical Journal. 1994;84(8):494–7. [PubMed: 7825085]
11. Bundey S A genetic study of Duchenne muscular dystrophy in West Midlands. Journal of medical genetics. 1981;18(1):1–7. [PubMed: 7252996]
12. Chung B, Wong V, Ip P. Prevalence of neuromuscular diseases in Chinese children: a study in southern China. J Child Neurol. 2003;18(3):217–9. [PubMed: 12731646]
13. Darin N, Tulinius M. Neuromuscular disorders in childhood: a descriptive epidemiological study from western Sweden. Neuromuscular Disorders. 2000;10(1):1–9. [PubMed: 10677857]
14. Hughes M, Hicks E, Nevin N, Patterson V. The prevalence of inherited neuromuscular disease in Northern Ireland. Neuromuscular disorders. 1996;6(1):69–73. [PubMed: 8845721]
15. Jeppesen J, Green A, Steffensen BF, Rahbek J. The Duchenne muscular dystrophy population in Denmark, 1977–2001: prevalence, incidence and survival in relation to the introduction of ventilator use. Neuromuscular Disorders. 2003;13(10):804–12. [PubMed: 14678803]
16. Kanamori M, Morton NE, Fujiki K, Kondo K, Rao D, Borecki I. Genetic epidemiology of Duchenne muscular dystrophy in Japan: classical segregation analysis. Genetic epidemiology. 1987;4(6):425–32. [PubMed: 3428571]
17. Leth A, Wulff K, Corfitsen M, Elmgreen J. Progressive muscular dystrophy in Denmark: natural history, prevalence and incidence. Acta Pædiatrica. 1985;74(6):881–5.
18. Merlini L, Stagni SB, Marri E, Granata C. Epidemiology of neuromuscular disorders in the under-20 population in Bologna Province, Italy. Neuromuscular disorders. 1992;2(3):197–200. [PubMed: 1483045]
19. Monckton G, Hoskin V, Warren S. Prevalence and incidence of muscular dystrophy in Alberta, Canada. Clinical genetics. 1982;21(1):19–24. [PubMed: 6978204]
20. Morton N, Chung C. Formal genetics of muscular dystrophy. American journal of human genetics. 1959;11(4):360. [PubMed: 14424475]

21. Nakagawa M, Nakahara K, Yoshidome H, Suehara M, Higuchi I, Fujiyama J, et al. Epidemiology of progressive muscular dystrophy in Okinawa, Japan. Classification with molecular biological techniques. Neuroepidemiology. 1991;10(4):185–91. [PubMed: 1745328]
22. Norwood FL, Harling C, Chinnery PF, Eagle M, Bushby K, Straub V. Prevalence of genetic muscle disease in Northern England: in-depth analysis of a muscle clinic population. Brain. 2009;132(11):3175–86. [PubMed: 19767415]
23. Peterlin B, Zidar J, Meznarič-Petruša M, Zupančič N. Genetic epidemiology of Duchenne and Becker muscular dystrophy in Slovenia. Clinical genetics. 1997;51(2):94–7. [PubMed: 9111995]
24. Prot J Genetic-epidemiological studies in progressive muscular dystrophy. Journal of medical genetics. 1971;8(1):90. [PubMed: 5098074]
25. Talkop Ü-A, Kahre T, Napa A, Talvik I, Sööt A, Piirsoo A, et al. A descriptive epidemiological study of Duchenne muscular dystrophy in childhood in Estonia. European Journal of Paediatric Neurology. 2003;7(5):221–6. [PubMed: 14511626]
26. van Essen AJ, Busch H, te Meerman GJ, Leo P. Birth and population prevalence of Duchenne muscular dystrophy in The Netherlands. Human genetics. 1992;88(3):258–66. [PubMed: 1733827]
27. Yasuda N, Kondô K. No sex difference in mutations rates of Duchenne muscular dystrophy. Journal of medical genetics. 1980;17(2):106–11. [PubMed: 7381863]
28. Miller LA, Romitti PA, Cunniff C, Druschel C, Mathews KD, Meaney FJ, et al. The muscular dystrophy surveillance tracking and research network (MD STARnet): Surveillance methodology. Birth Defects Research Part A: Clinical and Molecular Teratology. 2006;76(11):793–7. [PubMed: 17036307]
29. Mathews KD, Cunniff C, Kantamneni JR, Ciafaloni E, Miller T, Matthews D, et al. Muscular Dystrophy Surveillance Tracking and Research Network (MD STARnet): case definition in surveillance for childhood-onset Duchenne/Becker muscular dystrophy. Journal of child neurology. 2010;25(9):1098–102. [PubMed: 20817884]
30. Centers for Disease Control and Prevention. Bridged-Race Population Estimates 1990-2017 Request 2018 [2019/9/18]. Available from: https://wonder.cdc.gov/Bridged-Race-v2017.HTML.
31. Canty A, Ripley B. Package 'boot'. Version; 2019.
32. Signorell A, Aho K, Alfons A, Anderegg N, Aragon T, Arppe A. DescTools: Tools for descriptive statistics. R package version 099. 2016;18.
33. Ramos E, Conde JG, Berrios RA, Pardo S, Gómez O, Mas Rodríguez MF. Prevalence and genetic profile of Duchene and Becker muscular dystrophy in Puerto Rico. Journal of neuromuscular diseases. 2016;3(2):261–6. [PubMed: 27854217]
34. Ríos M, Romero F, Ramírez R. Race reporting among Hispanics: 2010. Population Division Working Paper. 2014;102.
35. Guzman B The Hispanic population. Census 2000 brief. 2001.
36. Counterman KJ, Furlong P, Wang RT, Martin AS. Delays in diagnosis of Duchenne muscular dystrophy: An evaluation of genotypic and sociodemographic factors. Muscle & Nerve. 2020;61(1):36–43. [PubMed: 31573675]
37. Holtzer C, Meaney FJ, Andrews J, Ciafaloni E, Fox DJ, James KA, et al. Disparities in the diagnostic process of Duchenne and Becker muscular dystrophy. Genetics in Medicine. 2011;13(11):942–7. [PubMed: 21836521]

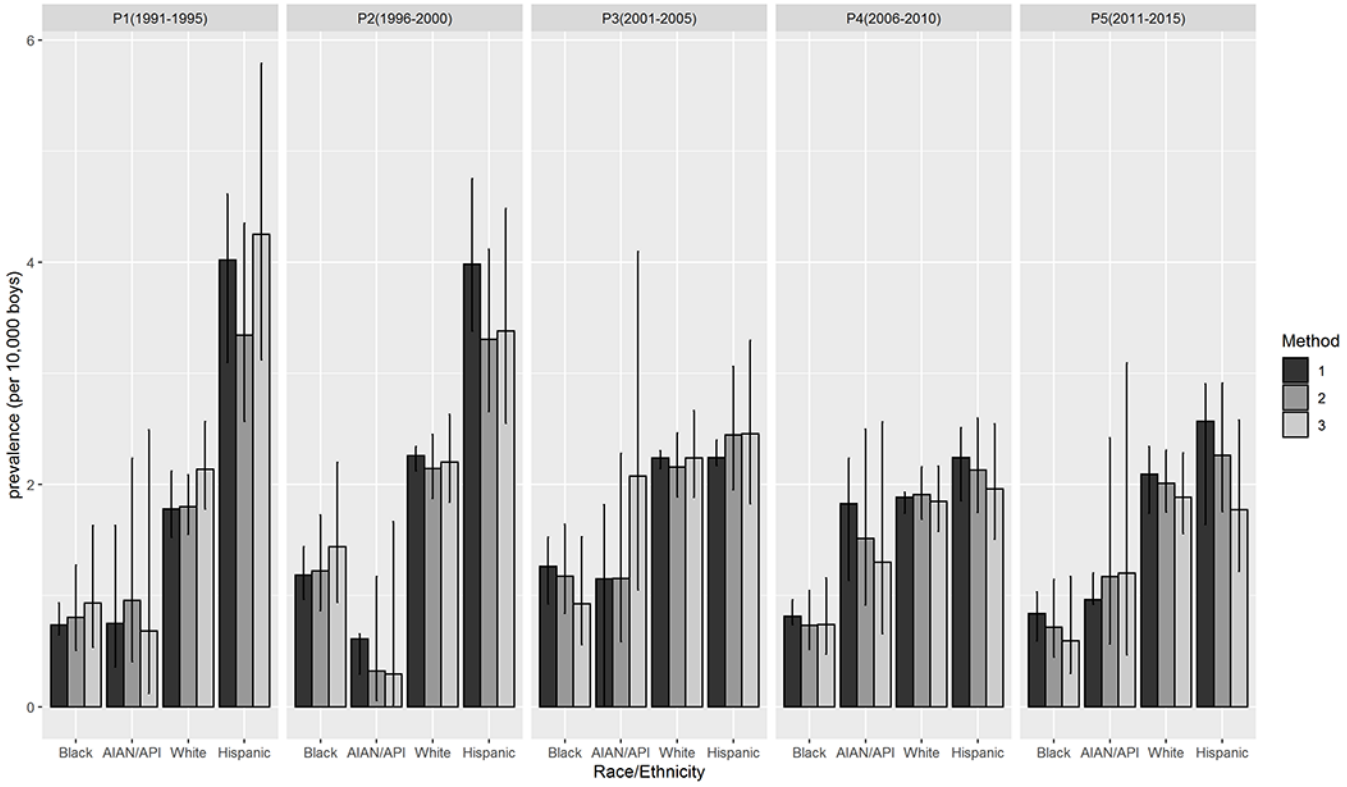

Fig. 1.

5-year prevalence of Duchenne and Becker muscular dystrophy among males aged 5 to 9 years by race/ethnicity and three estimating methods, the Muscular Dystrophy Surveillance, Tracking, and Research network (MD STAR*net*). The lines at the top of each bar are 95% confidence intervals. AIAN = American Indian or Alaska Native & Native Hawaiian. API = American Pacific Islander.

Table 1.

Sites participating in Muscular Dystrophy Surveillance, Tracking, and Research network (MD STAR*net*)

State	Surveillance Region	Eligible Dates of Birth*
Arizona, Georgia	Statewide	January 1, 1982 – December 31, 2010
Colorado, Iowa	Statewide	January 1, 1982 – December 31, 2010
Western New York	12 counties (Allegany, Cattaraugus, Chautauqua, Erie, Genesee, Livingston, Monroe, Niagara, Ontario, Orleans, Wayne, and Wyoming)	January 1, 1982 – December 31, 2010
	AND	
	9 additional counties (Cayuga, Chemung, Onondaga, Oswego, Schuyler, Seneca, Steuben, Tompkins, and Yates) added in the 2014 – 2019 funding period	January 1, 2000 – December 31, 2010
South Carolina, Utah	Statewide	January 1, 2000 – December 31, 2010
North Carolina Piedmont Region	33 counties (Alamance, Anson, Cabarrus, Caswell, Chatham, Davidson, Davie, Durham, Forsyth, Franklin, Gaston, Granville, Guilford, Iredell, Lee, Lincoln, Mecklenburg, Montgomery, Moore, Orange, Person, Randolph, Richmond, Rockingham, Rowan, Stanly, Stokes, Surry, Union, Vance, Wake, Warren, and Yadkin)	January 1, 2000 – December 31, 2010

*Cases within the range of dates of birth comprised the analytic sample.

Table 2.

Characteristics of males with Duchenne and Becker muscular dystrophies in the Muscular Dystrophy Surveillance, Tracking, and Research network (MD STAR*net*), N=1164

Characteristics		**N**	**%**
Case status	Definite	1087	93.4
	Probable	77	6.6
Race/Ethnicity *	Non-Hispanic white	712	61.2
	Non-Hispanic black	83	7.1
	Non-Hispanic AIAN/API	27	2.3
	Hispanic	243	20.9
	Other	< 10	-
	Multiple	16	1.4
	Unknown	77	6.6
State **	AZ	215	18.5
	CO	236	20.3
	GA	242	20.8
	IA	145	12.5
	NC	64	5.5
	wNY	143	12.3
	SC	58	5.0
	UT	61	5.2
Eligibility	January 1, 1982 – December 31, 2010	790	67.9
	January 1, 2000 – December 31, 2010	374	32.1

* AIAN = American Indian or Alaska Native & Native Hawaiian. API = American Pacific Islander.

** AZ = Arizona, CO=Colorado, GA = Georgia, IA = Iowa, NC = North Carolina, wNY = western New York State, SC = South Carolina, UT = Utah.

Table 3.

5-year period prevalence of Duchenne and Becker Muscular Dystrophies for males aged 5 to 9 years by race/ethnicity and three estimating methods in the Muscular Dystrophy Surveillance, Tracking, and Research network (MD STAR*net*)

Race/ethnicity*	Study period	Method 1			Method 2			Method 3		
		Cases	Population	Prevalence** (95%CI)	Cases	Population	Prevalence (95%CI)	Cases	Population	Prevalence (95%CI)
Overall ***	1991–1995	162	752433	2.15 (1.85–2.38)	288	1417806	2.03 (1.81–2.28)	189	789934	2.39 (2.07–2.76)
	1996–2000	210	847258	2.48 (2.34–2.56)	361	1573587	2.29 (2.07–2.54)	208	867378	2.40 (2.09–2.75)
	2001–2005	192	850041	2.26 (2.23–2.33)	376	1684479	2.23 (2.02–2.47)	220	951211	2.31 (2.03–2.64)
	2006–2010	229	1192141	1.92 (1.64–2.05)	431	2298969	1.87 (1.71–2.06)	252	1442192	1.75 (1.54–1.98)
	2011–2015	177	874693	2.02 (1.59–2.27)	298	1577459	1.89 (1.69–2.12)	149	876783	1.70 (1.45–2.00)
Non-Hispanic white	1991–1995	94	527729	1.78 (1.53–2.12)	176	977459	1.80 (1.55–2.09)	115	538052	2.14 (1.78–2.57)
	1996–2000	123	544425	2.26 (2.12–2.34)	216	1007593	2.14 (1.88–2.45)	120	545463	2.20 (1.84–2.63)
	2001–2005	128	571482	2.24 (2.15–2.30)	219	1015040	2.16 (1.89–2.46)	128	571482	2.24 (1.88–2.66)
	2006–2010	120	636694	1.88 (1.74–1.94)	254	1330985	1.91 (1.69–2.16)	155	838217	1.85 (1.58–2.16)
	2011–2015	118	563715	2.09 (1.74–2.34)	204	1014802	2.01 (1.75–2.31)	105	556539	1.89 (1.56–2.28)
Non-Hispanic black	1991–1995	< 10	122246	0.74 (0.65–0.93)	18	223499	0.81 (0.51–1.27)	12	128528	0.93 (0.53–1.63)
	1996–2000	17	143251	1.19 (0.97–1.44)	32	261903	1.22 (0.87–1.72)	21	145987	1.44 (0.94–2.20)
	2001–2005	18	142526	1.26 (0.93–1.53)	34	289530	1.17 (0.84–1.64)	15	161989	0.93 (0.56–1.53)
	2006–2010	19	232988	0.82 (0.74–0.96)	30	408970	0.73 (0.51–1.05)	19	256501	0.74 (0.47–1.16)
	2011–2015	11	131095	0.84 (0.59–1.03)	17	237685	0.72 (0.45–1.15)	< 10	134648	0.59 (0.30–1.17)
non-Hispanic AIAN/API	1991–1995	< 10	26715	0.75 (0.36–1.63)	< 10	52321	0.96 (0.41–2.24)	< 10	29268	0.68 (0.12–2.49)
	1996–2000	< 10	32732	0.61 (0.29–0.66)	< 10	62184	0.32 (0.06–1.17)	< 10	33998	0.29 (0.02–1.67)
	2001–2005	< 10	34773	1.15 (0.00–1.82)	< 10	69198	1.16 (0.59–2.28)	< 10	38548	2.08 (1.05–4.10)
	2006–2010	< 10	43765	1.83 (1.14–2.24)	15	99056	1.51 (0.92–2.50)	< 10	61578	1.30 (0.66–2.56)
	2011–2015	< 10	31107	0.96 (0.92–1.20)	< 10	59721	1.17 (0.57–2.42)	< 10	33254	1.20 (0.47–3.09)
Hispanic	1991–1995	33	82088	4.02 (3.10–4.61)	55	164527	3.34 (2.57–4.35)	40	94086	4.25 (3.12–5.79)
	1996–2000	48	120526	3.98 (3.38–4.75)	80	241907	3.31 (2.66–4.12)	48	141930	3.38 (2.55–4.48)
	2001–2005	33	147201	2.24 (2.17–2.40)	76	310711	2.45 (1.95–3.06)	44	179192	2.46 (1.83–3.30)
	2006–2010	51	227597	2.24 (1.85–2.51)	98	459958	2.13 (1.75–2.60)	56	285896	1.96 (1.51–2.54)

Race/ethnicity*	Study period	Method 1			Method 2			Method 3		
		Cases	Population	Prevalence** (95%CI)	Cases	Population	Prevalence (95%CI)	Cases	Population	Prevalence (95%CI)
	2011–2015	38	147998	2.57 (1.64–2.91)	60	265251	2.26 (1.76–2.91)	27	152342	1.77 (1.22–2.58)

*AIAN = American Indian or Alaska Native & Native Hawaiian. API = American Pacific Islander.

**prevalence per 10,000 males.

***cases who were reported as other, multiple or missing race/ethnicity were included in the overall prevalence.

Table 4.

Duchenne and Becker muscular dystrophy prevalence ratios and 95% confidence intervals for males aged 5 to 9 years by race/ethnicity and estimation method, the Muscular Dystrophy Surveillance, Tracking, and Research network (MD STAR*net*)

Method	Race/Ethnicity*	Prevalence Ratio (95% CI)	Chi-Square	Degree of freedom	P value
1	non-Hispanic white (reference)	1.00			
	Non-Hispanic black	0.46 (0.36–0.59)	37.72	1	<.001
	Hispanic	1.37 (1.17–1.61)	14.57	1	<.001
	non-Hispanic AIAN/API	0.61 (0.40–0.93)	5.15	1	0.023
2	non-Hispanic white (reference)	1.00			
	Non-Hispanic black	0.46 (0.37–0.57)	47.63	1	<.001
	Hispanic	1.28 (1.11–1.48)	11.62	1	<.001
	non-Hispanic AIAN/API	0.54 (0.36–0.80)	9.20	1	0.002
3	non-Hispanic white (reference)	1.00			
	Non-Hispanic black	0.45 (0.34–0.59)	32.38	1	<.001
	Hispanic	1.25 (1.05–1.50)	6.02	1	0.014
	non-Hispanic AIAN/API	0.58 (0.36–0.94)	4.98	1	0.026

*AIAN = American Indian or Alaska Native & Native Hawaiian. API = American Pacific Islander.