Neuromuscular Disorders 59 (2026) 106312

Contents lists available at ScienceDirect

Neuromuscular Disorders

journal homepage: www.elsevier.com/locate/nmd

Age-related differences in hot and cold executive functions in boys with Duchenne muscular dystrophy: longitudinal individual changes and age-group comparisons across childhood and adolescence

Jonas Gillenstrand [a,*], Malin Broberg [b], Anna-Karin Kroksmark [c], Jennifer Strand [b], Mar Tulinius [d], Anne-Berit Ekström [e]

[a] Department of Psychology, University of Gothenburg, Gothenburg, Sweden, Habilitation & Health, Region Västra Götaland, Gamlestaden, Sweden
[b] Department of Psychology, University of Gothenburg, Gothenburg, Sweden
[c] Department of Health and Rehabilitation, Institute of Neuroscience and Physiology, Sahlgrenska Academy, University of Gothenburg, Gothenburg, Sweden
[d] Department of Pediatrics Institute of Clinical Sciences, Sahlgrenska Academy, University of Gothenburg, Gothenburg, Sweden
[e] Department of Pediatrics Institute of Clinical Sciences, Sahlgrenska Academy, University of Gothenburg Gothenburg Sweden, Habilitation & Health, Region Västra Götaland, Sweden

ARTICLE INFO

Keywords:
Duchenne
Dystrophin
Developmental trajectories
Longitudinal study
Hot & cold executive function
Working memory
Age-related differences

ABSTRACT

This study investigated age-related differences in hot (affective-motivational) and cold (cognitive) executive functions (EF) in boys with Duchenne Muscular Dystrophy (DMD) across childhood and adolescence. In a cross-sectional design, 70 boys with DMD aged 5, 8, 11, and 14 years completed performance-based EF assessments, accompanied by parent-reported EF ratings. Longitudinal data were also collected from a subsample of 13 boys over a three-year period, with repeated assessments at the age intervals of 5–8, 8–11, and 11–14 years. At age 5, no significant EF impairments were observed. By age 8, however, significant deficits in hot EF tasks emerged, followed by impairments in cold EF at age 11. Cold EF performance indicated developmental delay rather than decline, as reflected in logits-based data. Longitudinal analyses using the Reliable Change Index revealed heterogeneous developmental patterns. Findings suggest that boys with DMD exhibit disrupted EF development, with increasing impairment through middle childhood and a potential positive trend from 11 to 14 years. These results underscore the importance of monitoring EF across a wider age range in this population.

1. Introduction

Duchenne muscular dystrophy (DMD) is an X-linked recessive neuromuscular disorder that affects approximately 1 in 3500–5000 liveborn males and is caused by mutations in the dystrophin gene on the X chromosome [1]. Dystrophin (DYS) protein is found in many human tissues, especially in muscle tissue, and insufficient functional DYS leads to progressive muscular weakness. Research also indicates that dystrophin plays an important role in the central nervous system, particularly in synaptic terminal integrity, synaptic plasticity, and cellular signal integration [2]. Furthermore, dystrophin isoforms have been identified in several brain regions, including the cerebral cortex, hippocampus, and cerebellum, suggesting that these areas are affected by dystrophin deficiency [3–5]. Although the precise role of dystrophin in the CNS remains unclear, evidence suggests that abnormal dystrophin likely influences brain circuitry and affects the development of neural structures and the brains developmental trajectories [6,7].

Several studies of boys with DMD have found that Full-Scale Intelligence Quotient (FSIQ) is generally normally distributed but about one standard deviation lower than the population norm [8–10]. Hinton [4, 11–14] and colleagues have argued that performance on digit span /working memory (WM) capacity regardless of IQ levels may reflect an underlying "core cognitive deficit" in children with dystrophinopathy, while others have suggested that the "core deficit" in DMD is better explained as a generalized executive skill deficit [15–19].

Executive functions (EF) are commonly described as top-down neurocognitive processes involved in the regulation of thoughts, actions, and emotions. These processes encompass cognitive flexibility, working memory, and inhibitory control [20–24]. Some researchers even argue that WM capacity only should be defined as a specific EF construct [22].

* Corresponding author.
E-mail address: jonas.gillenstrand@psy.gu.se (J. Gillenstrand).

https://doi.org/10.1016/j.nmd.2025.106312
Received 25 August 2025; Received in revised form 20 November 2025; Accepted 15 December 2025
Available online 16 December 2025

EF skills are crucial for modulating behaviour, solving problems, and devising strategies in response to changing external or social demands [25,26]. One influential developmental model from Anderson [20], proposes four interrelated executive domains, attentional control, cognitive flexibility, goal setting, and information processing, that together enable "executive control." According to this model, attentional control emerges early (in infancy) and develops rapidly during the preschool years, whereas cognitive flexibility, goal setting, and information processing undergo a major developmental spurt between ages 7 and 9, and reach relative maturity by around age 12. The model emphasizes a developmental trajectory of EF throughout childhood and adolescence.

Within the EF literature, some researchers also distinguish between "hot" and "cold" EF [24]. "Cold" EF refers to cognitive processes with little emotional or motivational involvement such as tasks assessing working memory, inhibitory control, or cognitive flexibility under neutral conditions - and is primarily associated with the prefrontal cortex [27,28]. "Hot" EF, by contrast, involves emotionally loaded or motivationally significant contexts, such as risky decision-making, and is more linked to orbitofrontal and medial brain regions [29]. While these categories are conceptually distinct, they frequently overlap in real-life tasks that require both cognitive and affective regulation. Notably, hot EF typically mature later than cold EF, particularly during adolescence, when decision-making in emotionally charged situations may develop at a slower pace compared to more cognitively oriented abilities [24].

Historically, it has been assumed that, unlike the progressive muscle deterioration, cognitive abilities in boys with DMD are stable over time and several studies have supported this assumption [17–19]. Other meta-studies have also proposed an increased capacity in verbal IQ [8] but not for performance IQ, between 9 and 20 years of age. However, some studies also suggest evidence for alternative developmental trajectories. For example, a longitudinal follow-up study by Hellebreker [30] indicated developmental stagnation in verbal span capacity over time, and investigations by Chieffo [31] and Gillenstrand [32] identified varying patterns of cognitive development throughout childhood and adolescence.

Although some studies have documented examples of both hot and cold EF difficulties in boys with DMD [16–19,33] no previous research has systematically examined age-related differences or longitudinal studies of EF in this population.

The aim of the present study was to examine age-related differences in hot and cold EF (including working memory capacity), in boys with DMD throughout childhood and adolescence. The investigation utilized both performance-based assessments and parental ratings of EF at 5, 8, 11, and 14 years of age, and further explored individual change across the intervals of 5–8, 8–11, and 11–14 years.

We hypothesized that significant differences would be observed across the four age groups (5 ± 0.5, 8 ± 0.5, 11 ± 0.5, and 14 ± 0.5 years) in both hot and cold executive functions. Furthermore, we hypothesize that longitudinal data will reveal significant individual differences in both hot and cold EF across the developmental intervals of 5–8, 8–11, and 11–14 years.

2. Patients and methods

2.1. Study design

This cross-sectional study compares parental self-reported and performance-based measures EF in boys with DMD, aged 5–14 years. All boys were tested by the same psychologist either at their local rehabilitation center or at the Regional Pediatric Rehabilitation Center in Gothenburg. During the child's performance-based assessment, parents completed survey forms and rated their child's EF skills. If both parents were present, they filled out a single questionnaire collaboratively; otherwise, one parent completed the questionnaire. Data on EF measures were part of a larger neuropsychological test battery, administered between 2017 and 2020. Two recent publications [32,33] reported partial findings from this cohort. The present study provides entirely new analyses, with the exception of background characteristics, which are also summarised here (Appendix B). The EF performance-based test was administered at the beginning of the assessment battery to minimize the effects of fatigue. All parents or guardians gave informed consent and all participants gave assent before enrolment. The study was approved by the regional ethical review board in Gothenburg (Ref. 737–16).

2.2. Participants

All boys in Sweden aged 5–14 years with a confirmed DMD diagnosis between 2017 and 2020 were invited to participate. Potential participants ($n = 100$) were identified and approached via local physicians in the southern, eastern, and western healthcare regions (up to Härjedalen and Jämtland). Of these, 73 (73 %) agreed to participate and provided written informed consent. Three families were excluded due to language barriers or personal reasons, leaving data from 70 % of the total eligible population (Appendix A). Participants were divided into four age groups (5 ± 0.5 years, 8 ± 0.5 years, 11 ± 0.5 years, and 14 ± 0.5 years), corresponding to developmental stages used in our clinical follow-up schedule and were assessed in the year they turned 5, 8, 11, or 14, between 2017 and 2020. A subgroup of thirteen boys that were tested the first year 2017 were also re-tested 2020. Six of the boys contributed with longitudinal data between 5 - 8 years of age, five boys at 8 and 11 years of age and two boys at 11 and 14 years of age.

2.3. Measures

Standardized neuropsychological measures with robust normative data were selected for the assessment. The measures were carefully chosen to minimize the influence of motor demands; both the WISC-V and MEFS performance-based measurements were administered on an iPad, while the remaining tests required only verbal responses. In parallel with the performance-based assessments of boys with DMD, their parents completed a survey that included questions on their boy's motor functions, their family socioeconomic status (SES) and parental ratings of their boys EF.

2.3.1. Motor function

Information about upper and lower extremity function was collected by parental ratings of Brooke upper extremity functional classification and the Vignos scale [34,35]. Higher scores on both scales reflect greater motor impairments. The Brooke scale [36,37] (levels 1–6) assesses upper extremity function from full overhead arm movement (level 1) to an inability to bring the hands to the mouth or use them functionally (level 6). The Vignos scale (levels 1–10) evaluates lower extremity function from normal ambulation (level 1) to being confined to bed (level 10). As older patients with DMD lose muscle strength, dependency in activities of daily living increases correspondingly. A moderate positive correlation between Brooke and Vignos scores suggests an interrelation in upper and lower extremity function [38].

2.3.2. Socioeconomic status

Family SES was measured using a Swedish-adapted version of the Hollingshead Four-Factor Index of Social Status [39,40]. It includes factors such as marital status, employment status (retired/employed), educational attainment, and occupational prestige.

2.3.3. Cognitive abilities

FSIQ, General Ability Index (GAI) and Working Memory Index (WMI) were measured using the Wechsler Preschool and Primary Scale of Intelligence – Fourth Edition (WPPSI-IV) [41] for 5-year-olds, and the Wechsler Intelligence Scale for Children – Fifth Edition (WISC-V, iPad version) [42] for children aged 8, 11, and 14. Both are standardized with a mean (M) of 100 and a standard deviation (SD) of 15. The test-retest reliability for the WISC-V scales is: FSIQ = 0.92, GAI = 0.91 and WMI = 0.82. For the WPPSI-IV, the test-retest reliability for FSIQ is 0.93, with other composite scales ranging from 0.84 to 0.89. These values indicate highly robust and reliable retest measurements.

2.3.4. Executive functions

Measures of EF were collected using performance-based tests, parental ratings.

2.3.4.1. Performance-Based cold executive function test. The Minnesota Executive Function Scale (MEFS) is a standardized measure of EF and learning readiness [43]. It assesses attention, behavioural regulation, cognitive flexibility, and the ability to retain and process information. The MEFS is validated in >32,800 typically developing children aged 24–215 months in the United States (internal consistency = 0.94). A certified Swedish translator produced a Swedish version, which was back-translated to ensure accuracy. Test-retest reliability for MEFS is (0.829) indicating highly robust and reliable retest measurements.

MEFS is administered on an iPad, with the examiner reading instructions to minimize confounds related to reading difficulties. Children complete a card-sorting game in which they match cards to target boxes according to varying rules (e.g., colour, size, shape). Starting levels differ by age, and there are seven levels increasing in difficulty. Participants progress or regress through levels based on performance until stop criteria are met. The outcome variables include Standardized Score (SS) with $M = 100$, SD = 15, and a Total Score (TS) that measure accuracy and time efficiency and is related to an optimal score of 100 regardless of age, using Rasch-model analyse to generate personal ability logits [44].

2.3.4.2. Parental and self-reported executive functions test. The Behaviour Rating Inventory of Executive Function (BRIEF) includes multiple rating forms [45]. Parents completed the BRIEF-P, an 86-item questionnaire for children aged 5–18, reflecting performance in typical home or familiar environments. The BRIEF-P produces three composite scores:

1. Behavioural Regulation Index (BRI): Subscales of Inhibit, Shift, and Emotional Control. More likely to measure hot EF.
2. Metacognitive Index (MI): Subscales of Initiate/Task Completion, Working Memory, Plan/Organize, Organization of Materials, and Monitor. More likely to measure both hot and cold EF.
3. Global Executive Composite (GEC): A composite of all subscales above.

The normative data for the BRIEF-P are based on 1419 parents from diverse backgrounds in the United States, with reported internal consistency ranging from 0.80 to 0.98. BRIEF Parent T-scores ($M = 50$, SD = 10), derived from the US population norms. In our study T-scores were transformed into inverted Index Scores ($M = 100$, SD = 15) to facilitate easier comparison with other measures, such as IQ Index Scores and MEFS Standard Scores (both with $M = 100$, SD = 15). Meaning that T-scores exceeding 1 SD (>60), which are rated as clinically significant, are now reported as scores below 1 SD (≤85) on the inverted Index Score scale. The psychometric properties of the BRIEF are robust, with strong internal consistency across scales [45]. Test-retest reliability coefficients for the BRIEF parental scales are excellent and robust: GEC (0.98), MI (0.96), and BRI (0.96).

2.4. Statistical analysis

Descriptive statistics for age, family SES, FSIQ, GAI, WMI and parental ratings of upper and lower motor function (assessed using the Brooke and Vignos scales) and MEFS-TS are presented in Appendix B. A one-sample *t*-test was performed to evaluate whether the means of FSIQ, GAI, WMI, MEFS-SS, BRIEF scales of GEC, MI and BRI differed significantly from the Swedish age-related normative mean of 100 are reported in Appendix B. To compare Hollingshead SES factors with Swedish population means, the Mann–Whitney *U test* was utilized and results are reported in Appendix B.

Pearson bi-variate correlations were conducted to examine the relationships between performance-based measures EF (MEFS-SS and WMI) and parental ratings of EF (BRIEF-P: GEC, MI, and BRI), and covariates including age, lower motor function, upper motor function, FSIQ, GAI and family SES. The results of these analyses are reported in Table 1.

Shapiro-Wilk analyses were run to ensure the data were normally distributed for parametric analyses and Levene's test was conducted to assess the assumption of homogeneity of variances for FSIQ, GAI, WMI, MEFS Standard Score (SS), and BRIEF scales of GEC, MI and BRI across 4 age groups, 5, 8, 11 and 14-years old boys with DMD. The test was not significant for any of the dependent variables (FSIQ $p = 0.177$, GAI $p = 0.134$, WMI $p = 0.318$, MEFS-SS $p = 0.317$, BRIEF-GEC $p = 0.248$, BRIEF-MI $p = 0.245$ and BRIEF-BRI $p = 0.147$) indicating that the variances were approximately equal across groups. Therefore, we proceeded with a one-way analysis of variance (ANOVA) to examine group differences in FSIQ, GAI, WMI, MEFS-SS and for all three scales of BRIEF (GEC, MI and BRI) with post hoc Bonferroni test to examine differences between the age-groups. Cohen's d was also calculated to measure effect sizes between the age-groups (5-, 8-, 11- and 14-years old boys with DMD).

To analyse longitudinal data from the 13 boys who completed both an initial test and a follow-up test between the ages of 5–8 or 8–11 or 11–14 years, the Reliable Change Index (RCI) was calculated. The RCI, based on the method outlined by Jacobson and Truax [46], was computed by dividing the difference between scores on Test 1 (T1) and test 2 (T2) by the standard error of measurement, incorporating the test-retest reliability coefficient of the test. An RCI value greater than 1.96 was interpreted as indicating a reliable change, while an RCI value of 1.96 or less suggested that the observed change could be attributed to measurement error rather than true change.

Within-subject longitudinal group differences between the age intervals (5–8, 8–11, 11–14 years and from 5–14 years) were analysed using paired *t*-tests. Cohen's d was calculated to assess effect sizes between each significant comparison [46].

Alfa level of significance was set to <0.05 for all analysis and all statistical analyses were performed using SPSS for Mac (version 29).

3. Results

The 70 boys with DMD included in the cohort participated in at least one performance-based executive function assessment at 5, 8, 11, or 14 years of age.

Parents completed questionnaires to assess family SES. For the 66 families who provided Hollingshead parental SES ratings (four families did not rate their family SES form), the mean score was 38.89 (SD = 15.49), which did not significantly differ from prior findings in the Swedish population (mean = 37.0, SD = 11.7) t (65 = 0.994, $p = 0.324$).

Upper and lower extremity motor function on the Brooke and Vignos scales, showed an expected decline with age, with increasing scores observed from 5 to 14 years. The mean age of participants and different age groups are shown in Appendix B.

Independent *t*-test was conducted to compare the cohort's performance to the population mean of 100 across our key variables, including FSIQ, GAI, WMI, MEFS-SS and the BRIEF scales of GEC, MI and BRI. The results indicated that all measures were significant lower. FSIQ ($M = 88.98$, SD = 18.42), (64) = −4.82, $p < 0.01$, with a medium effect size, Cohen's $d = -0.61$. GAI ($M = 91.77$, SD = 17,90), t (64) = −3.71, $p < 0.01$, with a medium effect size, Cohen's $d = -0.45$. WMI ($M = 84.58$, SD = 17.81), t (64) = −6.98, $p < 0.001$, with a large effect size, Cohen's

Table 1
Bivariate correlation between Age (months), Upper motor extremity function (Brooke), Lower motor extremity function (Vignos), Socioeconomic Status (Hollingshead SES), Full-Scale Intelligence Quotient (FSIQ), General Ability Index (GAI), Working Memory Index (WMI), Minnesota Executive Functions Scale (MEFS) Standard Score and the Behavioural Ratings of Executive Functions (BRIEF) Scales of General Executive Composite (GEC), Metacognition Index (MI) and Behavioural Regulation Index (BRI), (n 13), * = $p < 0.05$, ** = $p < 0.01$.

	Age	Brooke	Vignos	SES	FSIQ	GAI	WMI	MEFS	GEC	MI	BRI
Age	-										
Brooke	0.363**	-									
Vignos	0.641**	0.627**	-								
SES	−0.257*	−0.316*	−0.255	-							
FSIQ	−0.073	0.009	−0.070	0.313*	-						
GAI	0.004	0.068	−0.008	0.302*	0.981**	-					
WMI	−0.395**	−0.284*	−0.350**	0.243	0.827**	0.698**	-				
MEFS	−3.66**	−0.171	−0.269*	0.234	0.492**	0.439**	0.585**	-			
GEC	−0.154	0.178	−0.082	0.147	0.239	0.232	0.249*	0.020	-		
MI	−0.120	0.189	−0.66	−0.009	0.266*	0.263*	0.279*	0.027	0.923**	-	
BRI	−0.206	0.139	−0.114	0.254*	0.250*	0.217	0.347**	0.143	0.817**	0.653**	-
n	70	68	68	65	70	70	65	68	68	68	68
M	**125.42**	**1.96**	**4.44**	**39.08**	**83.63**	**86.83**	**84.63**	**85.95**	**86.41**	**91.95**	**81.85**
SD	39.18	1.11	3.16	15.54	27.50	27.46	17.75	13.58	19.54	17.53	22.32

$d = -1.27$. MEFS-SS ($M = 85.95$, SD = 13.58), t (67) = −8.53, $p < 0.001$, with a large effect size, Cohen's $d = -1.03$. BRIEF-GEC ($M = 86.14$, SD = 19.54), t (67) = −5.75, $p < 0.001$, with a medium effect size, Cohen's $d = -0.70$. BRIEF-MI ($M = 91.95$, SD = 17.53), t (67) = −3.79, $p <$ 0.001, with a small effect size, Cohen's $d = -0.46$. BRIEF-BRI ($M =$ 81.85, SD = 22.32), t (67) = −6.71, $p < 0.001$, with a large effect size, Cohen's $d = -0.81$.

To examine the relationships between variables across age-groups and assess patterns of co-variation, a bivariate Pearson correlation analysis was conducted (Table 1). WMI significantly correlated with all other covariates except family SES. MEFS-SS did not correlate with any of the parental ratings of EF (BRIEF, - GEC, -MI and -BRI). Age correlated with Brooke, Vignos, family SES, WMI and MEFS-SS and furthermore, family SES correlated with parental ratings of BRI but not MI or GEC. This analysis provides an overview of the strength and direction of associations between the measured variables offering insight into their developmental trajectories and potential interdependencies.

3.1. *Age-group differences*

When analyzing the independent variables across different age groups with independent *t*-test, no EF variables were found to be significantly lower than the population mean (100) at 5 years of age (Appendix B). However, at 8 years of age, all EF variables, with the exception of WMI, were significantly lower than the population mean and by 11 years of age, all EF variables showed significant reductions compared to the population mean. At 14 years of age, all EF variables, except for BRIEF-MI, remained significantly lower than the population mean but there was a positive trend for all EF variables from 11 to 14 years of age except for WMI.

A one-Way ANOVA revealed non-significant age differences between the age groups, in FSIQ, F (3,66) =. 19, $p = 0.905$., GAI, F (3,66) = 0.19, $p = 0.905$., GEC, F (3,64) = 1.91, $p = 0.134$ and MI, F (3,64) = 1.54, $p =$ 0.212.

In contrast, significant age differences were observed for WMI F

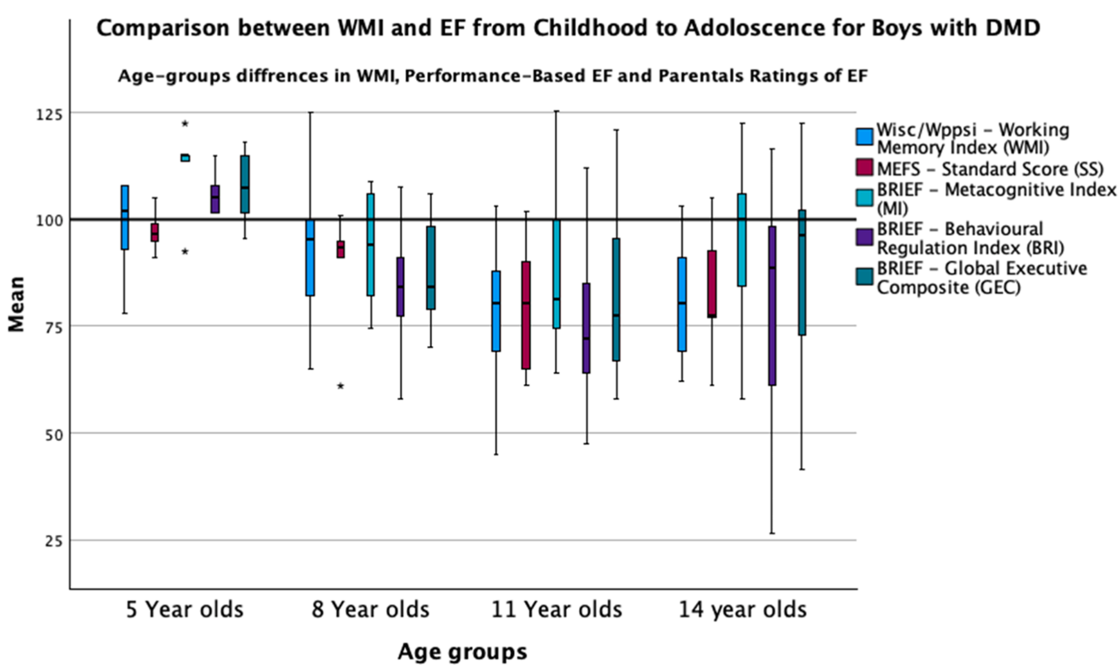

Fig. 1. Distribution of index and standard scores ($M = 100$, SD = 15) with 95 % CIs for cold and hot EF measures across age groups in boys with DMD. Cold EF includes the WISC/WPPSI Working Memory Index (WMI), MEFS standard score (SS), and BRIEF Metacognitive Index (MI). Hot EF is represented by the BRIEF Behavioural Regulation Index (BRI). The BRIEF Global Executive Composite (GEC) reflects parental ratings integrating both EF domains. Boxplots illustrate age-group differences from childhood to adolescence. Asterisks (*) indicate extreme values.

(3,61) = 3.51, $p = 0.019$., BRI F(3,63) = 3.053, $p = 0.033$ and in MEFS-SS F (3,63) = 6.058, $p = 0.001$. Post hoc comparison using the Bonferroni correction revealed significant differences in WMI between 5- and 14-year-olds, and in MEFS-SS between 5- and 11-year-olds, as well as between 5- and 14-year-olds. Age group differences for the BRIEF scales, including the GEC, MI and BRI, along with the Wisc/Wppsi - WMI, and MEFS-SS, are presented in Fig. 1.

Age-related differences in hot and cold EF were observed (Fig. 1). At 5 years of age, all age-normed measures within this sample were within the expected developmental range but subsequently, a more pronounced decrease was observed in hot EF skills measured by the parental ratings on the BRIEF (partly measured in GEC and especially measured in BRI) between 5 and 8 years of age, compared to cold EF skills (measured by MEFS-SS, WMI and BRIEF - MI) which showed a steeper decline between 8 and 11 years of age. Both hot and cold EF skills reached their lowest levels at 11 years of age. Thereafter, an indication of positive developmental progress was observed, with performance gradually approaching age-related norms. Despite this improvement, it is important to emphasize that the positive trends seen in the WMI, MEFS-SS, and BRI originate from baseline scores that were more than one standard deviation below age-related norms.

Furthermore, MEFS-TS, which assesses both accuracy and time efficiency, using Rasch analysis ability logits, indicates a continuous age-related difference in performance-based cold EF skills from 5 (M = 53.83, SD = 14.97) to 8 (M = 67.71, SD 20.47) to 11(M = 70.83, SD = 11.29) and 14 years of age (M 75.50 = 10.22), with no evidence of EF skill decline for boys with DMD.

3.2. Longitudinal individual change

Thirteen boys with DMD completed the assessments on two occasions: six boys were assessed at 5 and 8 years of age, five at 8 and 11 years of age, and two at 11 and 14 years of age. RCI scores were calculated for each individual and are summarized in Table 2. Delays in individual EF skills were most frequently observed in the WMI and the BRIEF scales of GEC and MI between 5 and 8 years of age (50 %). The BRIEF-BRI showed a range of changes, with 1/3 of participants demonstrating increased scores, 1/3 decreased scores, and 1/3 showing no change. Between 8 and 11 years of age, there was substantial variability in changes across all domains, as reflected in the RCI calculations (Table 2). From 11 to 14 years of age, FSIQ, GAI, WMI and MEFS-SS indicated only positive age-related differences. Furthermore, the RCI analysis of MEFS-TS indicated an overall improvement in executive function (EF) skills between the two measurement points across all participants and age groups, supporting the evidence of no individual EF skill decline in boys with DMD (Fig. 2).

Notably, when comparing RCI scores to age-normed means for the whole group at the first T1, and second assessments T2, all domains at “5 to 8 years” of age were within the age-appropriate average range (90 – 109, percentile 25 to 75) [40], except for WMI at T2 that were just under the average range (88 Index points), (Table 3).

A paired-sample *t*-test was also conducted to compare mean scores between T1 and T2 on the dependent variables of FSIQ, GAI, WMI, MEFS-SS and BRIEF scales of GEC, MI and BRI. Only WMI scores between T1 and T2, between 5 and 8 years of age were significant. However, the effect size, as measured by Cohen’s d, was large ($d = 1.29$) indicating a substantial difference between WMI scores between 5 and 8 years of age. Additional indications suggested medium effect size differences between T1 and T2 measurements on MEFS-SS; however, this difference did not achieve statistical significance.

3.3. Discussion

The aim of the present study is to investigate age-related differences of hot and cold EF (including WM capacity) in boys with DMD throughout childhood and adolescence. Examining both age-group differences and longitudinal individual changes.

3.4. Age-group differences

We hypothesized that significant age-group differences would emerge in both hot and cold EF, measured through performance-based EF tests and parental ratings of EF. Consistent with this hypothesis, significant age-group differences were observed in WMI, MEFS-SS and BRI, indicating substantial age-related variations across both hot and cold dimensions of EF. We also saw significant lower results compared to population mean for all the EF measurement (WMI, MEFS-SS, BRIEF scales of GEC, MI and BRI) at 11 years of age.

Table 2
Reliable Change Index (RCI) values across measurements, where values exceeding ±1.96 are considered significant. A positive RCI (+) indicates reliable improvement, a negative RCI (−) indicates reliable delay, and values between −1.96 and +1.96 indicate no reliable change. The table also presents the mean (M) and standard deviation (SD) for each age group, as well as the overall total, (n-13).

		Measures						
		Intelligence Quotient		**Working Memory**	**Parental Ratings of EF**			**Performance-Based EF**
Individuals	Age-Span	FSIQ	GAI	WMI	GEC	MI	BRI	MEFS
1	5 to 8 years	4.00*	1.26	−0.56	−8.00*	−6.84*	−2.36*	0.82
2		−0.33	−0.79	−2.22*	3.00*	1.65	3.06*	1.81
3		−0.67	−1.10	−2.89*	1.33	0.71	1.41	−0.18
4		−1.67	0.31	0.44	−4.67*	−4.48*	0.71	−0.36
5		−4.33*	−2.51*	−2.22*	−3.33*	−2.83*	−2.12*	−0.64
6		3.33*	3.30*	−0.22	6.67*	5.89*	3.54*	2.09*
Mean		**−0.28**	**0.08**	**−1.28**	**−0.83**	**−0.98**	**0.47**	**0.59**
SD		2.71	2.03	1.34	5.44	4.62	2.57	1.17
7	8 to 11 years	−0.33	−0.47	0.00	−8.00*	−5.66*	−3.54*	0.00
8		0.00	1.10	−1.67	−0.33	−0.71	0.71	−1.09
9		0.33	0.31	0.00	6.67*	6.60*	−0.24	0.00
10		−2.67*	−3.14*	−0.33	4.67*	3.30*	0.94	
11		−0.50	0.00	−0.56	1.00	−0.24	1.65	−0.82
Mean		**−0.63**	**−0.44**	**−0.51**	**0.80**	**0.66**	**−0.09**	**−0.48**
SD		1.18	1.62	0.69	5.66	4.60	2.04	0.56
12	11 to 14 years	3.17*	1.89	2.33*	1.67	−0.71	3.77*	0.91
13			1.10		1.33	−0.47	−1.65	0.91
Mean		**0.25**	**1.49**	**1.00**	**1.50**	**−0.59**	**1.06**	**0.91**
SD		4.12	0.56	1.89	0.24	0.17	3.83	0.00
TOTAL Mean		**-0.33**	**0.10**	**−0.63**	**0.15**	**−0.29**	**0.34**	**0.29**
SD		2.24	1.75	1.36	4.90	4.08	2.35	1.02

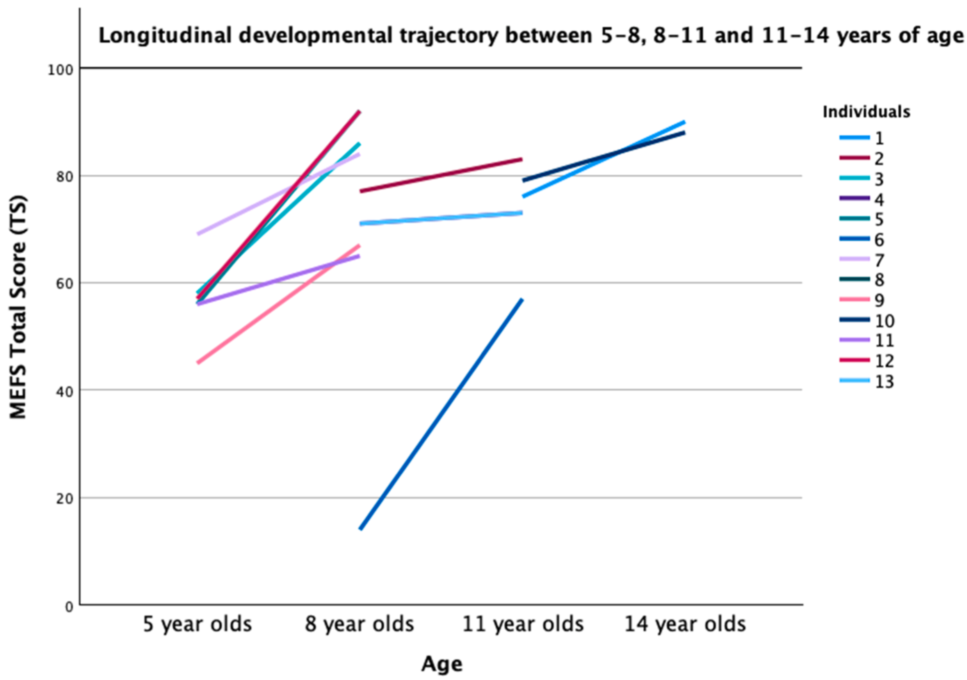

Fig. 2. MEFS Total Score (TS) in logits, reflecting combined accuracy and reaction time, where 100 represents the highest possible performance. (No age-related normative data available.) Higher scores indicate better executive function performance.

Table 3
Measurements of FSIQ, GAI, WMI, GEC, MI, BRI, and MEFS obtained from longitudinal data across two testing time points (Test 1 and Test 2) for boys assessed within three age ranges: 5–8 years, 8–11 years, and 11–14 years. The table also includes the overall mean (M) and standard deviation (SD) for each measure. *Values that deviate by ± 1 SD (15 points) from the standardized mean of 100 are considered significant, ($n = 13$).

		FSIQ		GAI		WMI		GEC		MI		BRI		MEFS	
ID	Age-Span	Test 1	Test 2	Test 1	Test 2	Test 1	Test 2	Test 1	Test 2	Test 1	Test 2	Test 1	Test 2	Test 1	Test 2
1	5–8	87	99	93	101	99	94	102	78*	114	85*	82*	72*	97	106
2		103	97	104	99	105	85*	118*	129*	123*	130*	108	121*	99	119*
3		121*	117*	125*	118*	108	82*	105	109	115*	118	88	94	105	103
4		81*	86	89	91	78*	82*	111	97	115*	96	103	100	91	87
5		113	100	119*	103	108	88	115*	105	115*	103	115*	106	96	89
6		80*	89	70*	91	93	91	96	118*	93	118*	102	117*	95	118*
Mean	**5–8**	**97.50**	**98.00**	**101.40**	**100.50**	**98.50**	**87.00**	**107.50**	**106**	**112.50**	**108.33**	**99.67**	**101.66**	**97.17**	**103.67**
SD		17.39	10.88	22.43	9.96	11.61	4.90	8.54	17.60	10.11	16.62	12.40	17.72	4.67	13.71
7	8–11	93	91	94	91	97	97	99	75*	106	82*	82*	67	95	95
8		108	108	100	107	125	110	103	102	109	106	90	93	94	82*
9		53*	55*	61*	63*	45*	45*	82*	102	85*	112	82*	81*	61*	61*
10		80*	64*	90	70*	65*	62*	76*	90	85*	99	69*	73*		65*
11		118*	115*	111	111	112	107	96	99	103	102	87	94	91	82*
Mean	**8–11**	**90.40**	**86.60**	**91.20**	**88.40**	**88.80**	**84.20**	**91.20**	**93.60**	**97.60**	**100.20**	**82.00**	**81.60**	**85.25**	**77.00**
SD		25.42	26.43	18.65	21.49	33.17	29.06	11.61	11.50	11.70	11.28	8.03	11.95	16.26	13.91
12	11–14	86	105	89	101	79*	100	88	93	100	97	69*	85*	90	100
13		79*		86		76*		78*	69*	82*	75*	73*	66*	80*	90
Mean	**11–14**	**82.50**	**105.00**	**87.50**	**101**	**77.50**	**100**	**83.00**	**81.00**	**91.00**	**86.00**	**71.00**	**75.50**	**85.00**	**95.00**
SD		4.95		2.12		2.12		7.07	16.97	12.73	15.56	2.83	13.44	7.07	7.07
Total Mean		**92.46**	**93.83**	**94.69**	**95.50**	**91.54**	**86.92**	**97.61**	**98.38**	**103.46**	**101.77**	**88.46**	**89.92**	**91.17**	**92.08**
SD		19.35	18.77	17.80	15.88	21.99	18.36	13,52	16.88	13.52	15.55	14.74	18.05	11.22	17.61

Fig. 1 illustrates separate age-related differences for hot and cold EF and a combined measurement (of both hot and cold EF) in BRIEF – GEC. The results indicate that behavior regulation difficulties emerge earlier with impairments becoming more apparent compared to age-related norms by 8 years of age. In contrast, cold EF skills (measured by WMI, MEFS – SS and BRIEF – MI) demonstrated a steeper developmental decline later between 8 and 11 years of age. These findings suggest that behavior regulation problems may precede delays in cold EF development. This also aligns with studies that suggests that hot EF lags behind cold EF [24,47]. But the differences are much more evident for boys with DMD with lower scores compared to age-related norms.

However, there were also similarities between the hot and cold EF skills. For 5-year-olds both cold and hot EF were within average range, and at 11 years of age both cold and hot EF was rated most problematic/ and performance reached lowest age-normed results, and furthermore at 14 years of age, all EF showed a positive trend of catching up to age-related norms. This age-related difference is also similar in relation to our [32] previous finding regarding behavioural problems. With parental ratings of behaviour problems within expected range at 5 years of age, followed by a decline in the portion of average ratings at 8 years of age and fewest ratings within the average range recorded at 11 years of age, followed by an upward trend of average ratings between 11 and

14 years of age.

Notably, in this study, the MEFS-TS (person ability logits estimate derived from Rasch analysis) did not indicate any developmental decline in cold EF skills, as all boys with DMD showed progressive improvements of accuracy and time efficiency at all ages. This pattern suggests a potential developmental delay between 8 and 11 years of age, followed by evidence of catching-up towards normative EF levels between 11 and 14 years.

This raises a compelling question: could a possible developmental delay in both hot and cold EF during this period increase the risk of behavioural problems? And can EF training for boys or compensation for EF skills minimize the outcome of behavioural problems?

Our overall findings are also consistent with previous research indicating that FSIQ and WMI scores are, on average, approximately one standard deviation below the mean [8–10]. However, when examining age-related differences (as illustrated in Fig. 1), it is critical to note that these group averages provide limited insight into developmental change. This highlights the importance of adopting a developmental perspective. For example, while FSIQ and WMI scores among 5-year-olds may fall within age-appropriate norms, the scores for 11-year-olds are approximately one standard deviation below the mean. Notably, these patterns are not apparent when examining only the overall mean scores for the full sample.

Furthermore, our results also highlight the possibility for age-related differences in hot and cold EF in boys with DMD. These age-related differences could guide proactive, tailored interventions that support strategies helping boys and their parents address delays in hot EF skills around 8 years of age and, subsequently, delays in cold EF skills around 11 years of age.

Notably, WMI in our study were very closely connected to age-related differences in MEFS-SS supporting the idea that they both could be part of the same construct measuring cold EF skills. A confounding factor is that WMI measurements between 5 and 8 years of age were conducted using two different tests: the WPPSI-IV for 5-year-olds and the WISC-V for participants aged 8–14 years. The WPPSI-IV includes only visuo-spatial WM tasks, which are often less impaired in boys with DMD compared to verbal WM capacity. In contrast, the WISC-V-WMI also incorporates verbal WM, which may underlie the observed differences in performance between these age-groups. And we did not use only verbal-span as a separate measurement in our study because of the limitation in age-span data (only for 8 to 14 years of age).

Furthermore, the bivariate correlations revealed no significant relationship between MEFS-SS and the parental BRIEF ratings, suggesting that these measures assess distinct dimensions of EF. However, WMI was significantly correlated with both MEFS-SS and all BRIEF parental rating scales, including GEC, MI, and BRI. WMI also correlated with age, as well as upper and lower motor extremity functions, though age is likely to be a confounding underlaying factor for the significant correlations with the Brooke and Vignos measures. Our study also confirmed that the physical decline in boys with DMD, measured by the Brooke and Vignos scales, was not associated with any of the parental ratings of hot EF or the performance-based measure of cold EF - MEFS-SS (except WMI), these results also align with meta-analysis, confirming no significant correlation between the physical and cognitive impact in boys with DMD [9].

Another interesting finding was that family SES was significantly correlated with BRI but not with MI or GEC, suggesting that lower SES may also be associated with an increased prevalence of behaviour regulation difficulties. Longitudinal research suggests that individual differences in both hot and cold EF are considerable stability across time [48,49]. One likely possibility is that key aspects of children's environment tend to remain stable and that EF is associated with the contexts in which children develop including their family SES [24].

3.5. Longitudinal individual change

We hypothesized that longitudinal data would reveal significant individual differences in WM capacity and in both hot and cold EF across the age intervals of 5–8, 8–11, and 11–14 years. Delays in individual EF skills were most commonly observed in WMI and BRIEF scales of GEC and MI between 5 and 8 years of age (Table 3). However, only WMI showed significant differences between 5 and 8 years when comparing results from Test 1 and Test 2 using a paired *t*-test. Despite the small sample size, Cohen's d indicated a large effect size, underscoring the magnitude of the difference. However, results must be interpreted with caution due to the limited sample size and the possible confounding variable of using more visual versus verbal WMI test in WPPSI then WISC.

Some individual changes within the sample further supported age-related differences observed across the age-groups. Notably, the MEFS-SS measure did not show a decline between 5 and 8 years of age and demonstrated a positive developmental trend between 11 and 14 years of age. Age-related differences for MEFS-TS ability logits followed the expected pattern of improvement, with scores progressively increasing at 5, 8, 11, and 14 years of age. These findings suggest a developmental delay rather than a decline in cold EF functions in boys with DMD. Additionally, the BRI showed no impairments at 5 years of age, but as expected, impairments became more pronounced between 8 and 14 years.

Limited evidence was found to support a clear decline in behavior regulation between 5 and 8 years of age. Specifically, 50 % of the participants exhibited an increase in behavior regulation difficulties during this period, while the other 50 % showed a decrease, indicating substantial variability among individuals. Similarly, large individual differences were observed in working memory performance between 8 and 11 years of age. These findings highlight the heterogeneity of age-related differences within the sample. Given the small sample size of 13 participants, broad generalizations should be approached with caution. But at the same time significant individual changes observed are unlikely to be solely attributable to test-retest variability or random chance.

One of the most consistent findings, both in terms of age-related differences and individual changes, was the positive developmental trend observed between 11 and 14 years of age. However, it remains unclear whether this trend will continue beyond 14 years or if it will plateau. When compared to developmental models, such as Anderson's model [20], which suggests a steep increase in EF skills between 6 and 14 years of age, our findings raise the possibility that developmental delays between 8 and 11 years could contribute to greater discrepancies from age-related norms during this period. If a developmental spurt occurs later in boys with DMD, it could indicate a catch-up phase beyond 11 years of age.

These observations underscore the need for further longitudinal research and cross-sectional studies with a larger and more diverse sample, extending the age range up to 20 years. Such research could provide a more comprehensive understanding of individual growth trajectories in both hot and cold EF, while clarifying the long-term developmental outcomes for boys with DMD.

3.6. Conclusions

This study contributes to the understanding of age-related differences of EF in boys with DMD. In addition, it provides information about separate age-releted differences for hot and cold EF throughout childhood and adolescence in boys with DMD. Our result confirms that WMI and MEFS are closely related at different ages supporting the assumption that they both could be representative for cold EF measurement. However, WMI correlated with a wider range of variables showing support for a more central involvement in many cores intellectual and executive processes.

Clinically, these findings highlight the importance of adopting a proactive, individualized approach that considers complex age-related development in boys with DMD, addressing difference in both of hot and cold EF skills.

3.7. Further research

Further research is needed to determine whether the positive developmental trends observed in both the age-group differences and individual changes reflect a delayed developmental spurt in EF or merely a short-term positive trend. Extending the age range of studies to include individuals up to 20 years of age would provide a more nuanced understanding of the executive developmental period and its potential variability.

We suggest that studying both hot and cold EF functions may provide a more comprehensive understanding of the broader vulnerabilities associated with DMD [49]. These vulnerabilities include genetic isoforms, neuropsychiatric conditions such as attention-deficit/hyperactivity disorder (ADHD), autism spectrum disorder (ASD) and obsessive-compulsive disorder (OCD). Furthermore, children with DMD may also exhibit behavioral difficulties such as oppositional/argumentative tendencies rising from developmental crises, environmental challenges and/or lower EF skills [50]. Additionally, using a range of different measurement tools is essential to fully capture the complexity of EF, especially when comparing atypical with typical development. Or as Zelazo and Carlson [51] so elegantly describes it; Ecologically adapted measures of EF skills can complement standardized measures that capture important age-related and individual differences and should remain a cornerstone of research on the topic (of EF). We believe this type of model will facilitate a deeper understanding of how multiple, simultaneous, and interacting causal influences, operating at many levels of analysis (cultural, social, cognitive, neural, and molecular), work together to produce conscious control.

3.8. Limitations

This study has several limitations. First, the sample size is relatively small, particularly in terms of the longitudinal data. However, this is the first study to investigate age-related differences of EF in boys with DMD, both through cross-sectional age-group comparisons and longitudinal assessments. Although the small sample size limits the generalizability of the findings, the study provides valuable insights into how EF, particularly the distinction between hot and cold EF, can inform the development of tailored interventions for boys with DMD and their families. Additionally, our findings highlight the importance of recognizing age-related periods of heightened vulnerability, which could be critical for early intervention strategies.

A second limitation is the reliance on age-related norms rather than a control group. However, the study utilized robust assessment tools with high test-retest reliability, and the tests were carefully selected to minimize the impact of motor impairments on performance. Combining measurement from age normed scores with ability logits scores also strengthens the validity of the results in the study.

Finally, the measurement of WMI included both visual and verbal components due to limitations in available tests. The results may have differed if we had been able to assess verbal working memory capacity in isolation. However, we did not have access to reliable and validated tests designed specifically to assess verbal working memory in 5-year-old children. This limitation highlights the need for future studies to consider more targeted assessments when possible.

Declaration of generative AI and AI-assisted technologies in the writing process

During the preparation of this work the author(s) used [ChatGPT/40] in order to use the following prompt in ChatGPT:

Please review the following text for grammar, punctation, sentence structure, and academic tone. Ensure that the language is formal and suitable for a scientific article. Provide specific suggestions for improvement and highlight any unclear or awkward sections.

Text: (Insert text here) After using this tool/service, the author(s) reviewed and edited the content as needed and take(s) full responsibility for the content of the publications.

Funding source

The study was financially supported by grants from the Swedish state under the agreement between the Swedish government and the county councils; the ALF-Agreement (SU 2019- 02662). Financial support was also received from contributors during the following period: Insamlingsstiftelsen för muskeldystrofiforskning (SMDF) 2013, 2015, 2018, 2019, 2024; Riksförbundet för Rörelsehindrade Barn och Ungdomars (RBU) Forskningsstiftelse 2014; Petter Silferskiölds Minnesfond 2014–2015; Sunnerdahls Handikappfond 2014, 2015, 2017 2018; Neuroförbundet 2015; Muskelkampen 2015, 2017; Norrbacka-Eugeniastiftelsen 2017–2019; Drottning Silvias Jubileumsfond 2018; Stiftelsen Bo-Göran Bremerts minne 2017–2022; Linnéa och Josef Carlsson stiftelse 2019, 2024.

CRediT authorship contribution statement

Jonas Gillenstrand: Writing – review & editing, Writing – original draft, Visualization, Validation, Software, Resources, Project administration, Methodology, Investigation, Funding acquisition, Formal analysis, Data curation, Conceptualization. **Malin Broberg:** Writing – review & editing. **Anna-Karin Kroksmark:** Writing – review & editing. **Jennifer Strand:** Writing – review & editing. **Mar Tulinius:** Writing – review & editing. **Anne-Berit Ekström:** Writing – review & editing, Supervision.

Declaration of competing interest

None.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.nmd.2025.106312.

References

[1] Bushby KMD, Finkel RMD, Birnkrant DJMD, Case LEDPT, Clemens PRMD, Cripe LMD, Constantin CP. Diagnosis and management of Duchenne muscular dystrophy, Part 1: diagnosis, and pharmacological and psychosocial management. Lancet Neurol 2010;9(1):77–93. https://doi.org/10.1016/S1474-4422(09)70271-6.

[2] De Stefano ME, Ferretti V, Mozzetta C. Synaptic alterations as a neurodevelopmental trait of Duchenne muscular dystrophy. Neurobiol Dis 2022;168:105718. https://doi.org/10.1016/j.nbd.2022.105718.

[3] Anderson JL, Head SI, Rae C, Morley JW. Brain function in Duchenne muscular dystrophy. Brain 2002;125(Pt 1):4–13. https://doi.org/10.1093/brain/awf012.

[4] Cyrulnik SE, Hinton VJ. Duchenne muscular dystrophy: a cerebellar disorder? Neurosci Biobehav Rev 2008;32(3):486–96. https://doi.org/10.1016/j.neubiorev.2007.09.001.

[5] Kreis R, Wingeier K, Vermathen P, Giger E, Joncourt F, Zwygart K, Steinlin M. Brain metabolite composition in relation to cognitive function and dystrophin mutations in boys with Duchenne muscular dystrophy. NMR Biomed 2011;24(3):253–62. https://doi.org/10.1002/nbm.1582.

[6] Gorecki DC, Lukasiuk K, Szklarczyk A, Kaczmarek L. Kainate-evoked changes in dystrophin messenger RNA levels in the rat hippocampus. Neuroscience 1998;84(2):467–77.

[7] Kim TW, Wu K, Black IB. Deficiency of brain synaptic dystrophin in human Duchenne muscular dystrophy. Ann Neurol 1995;38(3):446–9.

[8] 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(4):257–65. https://doi.org/10.1017/s0012162205000496.

[9] Cotton S, Voudouris NJ, Greenwood KM. Intelligence and Duchenne muscular dystrophy: full-scale, verbal, and performance intelligence quotients. Dev Med Child Neurol 2001;43(7):497–501. https://doi.org/10.1017/s0012162201000913.
[10] Snow WM, Anderson JE, Fry M. Regional and genotypic differences in intrinsic electrophysiological properties of cerebellar Purkinje neurons from wild-type and dystrophin-deficient mdx mice. Neurobiol Learn Mem 2014;107:19–31. https://doi.org/10.1016/j.nlm.2013.10.017.
[11] Hinton VJ, De Vivo DC, Nereo NE, Goldstein E, Stern Y. Poor verbal working memory across intellectual level in boys with Duchenne dystrophy. Neurology 2000;54(11):2127–32. https://doi.org/10.1212/wnl.54.11.2127.
[12] Hinton VJ, De Vivo DC, Nereo NE, Goldstein E, Stern Y. 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(1):45–54. https://doi.org/10.1017/s1355617701711058.
[13] Hinton VJ, Fee RJ, Goldstein EM, De Vivo DC. Verbal and memory skills in males with Duchenne muscular dystrophy. Dev Med Child Neurol 2007;49(2):123–8. https://doi.org/10.1111/j.1469-8749.2007.00123.x.
[14] Leaffer EB, Fee RJ, Hinton VJ. Digit span performance in children with dystrophinopathy: a verbal span or working memory contribution? J Int Neuropsychol Soc 2016;22(7):777–84. https://doi.org/10.1017/S1355617716000461.
[15] Battini R, Chieffo D, Bulgheroni S, Piccini G, Pecini C, Lucibello S, Mercuri E. Cognitive profile in duchenne muscular dystrophy boys without intellectual disability: the role of executive functions. Neuromuscul Disord 2018;28(2):122–8. https://doi.org/10.1016/j.nmd.2017.11.018.
[16] Fee RJ, Montes J, Stewart JL, Hinton VJ. Executive skills and academic achievement in the dystrophinopathies. J Int Neuropsychol Soc 2018;24(9):928–38. https://doi.org/10.1017/S1355617717001333.
[17] Donders J, Taneja C. Neurobehavioral characteristics of children with duchenne muscular dystrophy. Child Neuropsychol 2009;15(3):295–304. https://doi.org/10.1080/09297040802665777.
[18] Mento G, Tarantino V, Bisiacchi PS. The neuropsychological profile of infantile Duchenne muscular dystrophy. Clin Neuropsychol 2011;25(8):1359–77. https://doi.org/10.1080/13854046.2011.617782.
[19] Wicksell RK, Kihlgren M, Melin L, Eeg-Olofsson O. Specific cognitive deficits are common in children with Duchenne muscular dystrophy. Dev Med Child Neurol 2004;46(3):154–9. https://doi.org/10.1017/s0012162204000283.
[20] Anderson P. Assessment and development of executive function (EF) during childhood. Child Neuropsychol 2002;8(2):71–82. https://doi.org/10.1076/chin.8.2.71.8724.
[21] Diamond A. Executive functions. Annu Rev Psychol 2013;64:135–68. https://doi.org/10.1146/annurev-psych-113011-143750.
[22] McCabe DP, Roediger HL, McDaniel MA, Balota DA, Hambrick DZ. The relationship between working memory capacity and executive functioning: evidence for a common executive attention construct. Neuropsychology 2010;24(2):222–43. https://doi.org/10.1037/a0017619.
[23] Miyake A, Friedman NP. The nature and organization of individual differences in executive functions: four general conclusions. Curr Dir Psychol Sci 2012;21(1):8–14. https://doi.org/10.1177/0963721411429458.
[24] Zelazo PD, Carlson SM. Hot and cool executive function in childhood and adolescence: development and plasticity. Child Develop Perspect 2012;6(4):354–60. https://doi.org/10.1111/j.1750-8606.2012.00246.x.
[25] Fogel Y, Rosenblum S, Josman N. Environmental factors and daily functioning levels among adolescents with executive function deficits. Br J Occup Ther 2020;83(2):88–97. https://doi.org/10.1177/0308022619876557.
[26] Yang Y, Shields GS, Zhang Y, Wu H, Chen H, Romer AL. Child executive function and future externalizing and internalizing problems: a meta-analysis of prospective longitudinal studies. Clin Psychol Rev 2022;97:102194. https://doi.org/10.1016/j.cpr.2022.102194.
[27] Eslinger PJ, Flaherty-Craig CV, Benton AL. Developmental outcomes after early prefrontal cortex damage. Brain Cogn 2004;55(1):84–103. https://doi.org/10.1016/S0278-2626(03)00281-1.
[28] Bunge SA, Wright SB. Neurodevelopmental changes in working memory and cognitive control. Curr Opin Neurobiol 2007;17(2):243–50. https://doi.org/10.1016/j.conb.2007.02.005.
[29] Bechara A. The role of emotion in decision-making: evidence from neurological patients with orbitofrontal damage. Brain Cogn 2004;55(1):30–40. https://doi.org/10.1016/j.bandc.2003.04.001.
[30] Hellebrekers DM, Doorenweerd N, Sweere DJ, van Kuijk SM, Aartsma-Rus AM, Klinkenberg S, Vles JS, Hendriksen JG. Longitudinal follow-up of verbal span and processing speed in duchenne muscular dystrophy. Eur J Paediatr Neurol 2020;25:120–6.
[31] Chieffo DPR, Moriconi F, Pane M, Lucibello S, Ferraroli E, Norcia G, Mercuri E. A longitudinal follow-up study of intellectual function in Duchenne muscular dystrophy over age: is it really stable? J Clin Med 2023;12(2):403. https://doi.org/10.3390/jcm12020403.
[32] Gillenstrand J, Ekström A-B, Kroksmark A-K, Tulinius M, Broberg M. Behavioural strengths and difficulties in relation to intellectual functions and age in Swedish boys with Duchenne muscular dystrophy. Child Neuropsychol 2023;29(6):959–72.
[33] Gillenstrand J, Broberg M, Kroksmark A-K, Tulinius M, Ekström A-B. Concordance between parental and self-rated and performance-based measures of executive functioning in boys with Duchenne muscular dystrophy: the role of parental life satisfaction and carrier versus non-carrier status. Child Neuropsychol 2024:1–20. https://doi.org/10.1080/09297049.2024.2433810.
[34] Vignos PJ, Spencer GE, Archibald KC. Management of progressive muscular dystrophy of childhood. J Am Med Assoc 1963;184(2):89–96.
[35] Lord JP, Portwood MM, Lieberman JS, Fowler JWM, Berck P. Upper extremity functional rating for patients with Duchenne muscular dystrophy. Arch Phys Med Rehabil 1987;68(3):151.
[36] Brooke MH, Griggs RC, Mendell JR, Fenichel GM, Shumate JB, Pellegrino RJ. Clinical trial in Duchenne dystrophy. I. The design of the protocol. Muscle Nerve 1981;4(3):186–97.
[37] Lue Y-J, Lin R-F, Chen S-S, Lu Y-M. Measurement of the functional status of patients with different types of muscular dystrophy. Kaohsiung J Med Sci 2009;25(6):325–33. https://doi.org/10.1016/S1607-551X(09)70523-6.
[38] Savaş D, Şimşek TT. Functional level and its relationship to upper extremity function, pain, and muscle stiffness in children with Duchenne muscular dystrophy. Ir J Med Sci 2023;192(4):1867–73. https://doi.org/10.1007/s11845-022-03162-z.
[39] Hollingshead AB. Four factor index of socioeconomic status. Yale J Sociol 1975;8:21–5238.
[40] Olsson MB, Hwang PC. Influence of macrostructure of society on the life situation of families with a child with intellectual disability: sweden as an example. J Intellect Disabil Res 2003;47(4–5):328–41. https://doi.org/10.1046/j.1365-2788.2003.00494.x.
[41] Wechsler D. WPPSI-IV: technical and interpretive manual. Pearson; 2012.
[42] Wechsler D. Wechsler intelligence scale for children-iv (WISC-IV) manual. San Antonio: The Psychological Corporation; 2004.
[43] Carlson S, Zelazo P. Minnesota executive function scale: test manual. Reflect Sci 2014.
[44] Kline TJ. Psychological testing: a practical approach to design and evaluation. SAGE Publications, Inc.; 2005. https://doi.org/10.4135/9781483385693.
[45] Gioia GA, Isquith PK, Guy SC, Kenworthy L. Behaviour rating inventory of executive function: BRIEF. Psychol Assess Resour 2000.
[46] Speer DC. Clinically significant change: jacobson and Truax (1991) revisited. J Consult Clin Psychol 1992;60(3):402–8. https://doi.org/10.1037/0022-006X.60.3.402.
[47] Ferguson CJ. An effect size primer: a guide for clinicians and researchers. Prof Psychol Res Pr 2009;40(5):532–8.
[48] Polderman TJC, Posthuma D, De Sonneville LMJ, Stins JF, Verhulst FC, Boomsma DI. Genetic analyses of the stability of executive functioning during childhood. Biol Psychiatry 2007;76:11–20.
[49] Darmahkasih AJ, Rybalsky I, Tian C, Shellenbarger KC, Horn PS, Lambert JT, Wong BL. Neurodevelopmental, behavioral, and emotional symptoms common in Duchenne muscular dystrophy. Muscle Nerve 2020;61(4):466–74. https://doi.org/10.1002/mus.26803.
[50] Birnkrant DJ, Bushby K, Bann CM, Apkon SD, Blackwell A, Colvin MK, Ward LM. Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan. Lancet Neurol 2018;17(5):445–55. https://doi.org/10.1016/S1474-4422(18)30026-7.
[51] Zelazo PD, Carlson SM. Reconciling the context-dependency and domain-generality of executive function skills from a developmental systems perspective. J Cognit Develop 2023;24(2):205–22. https://doi.org/10.1080/15248372.2022.2156515.