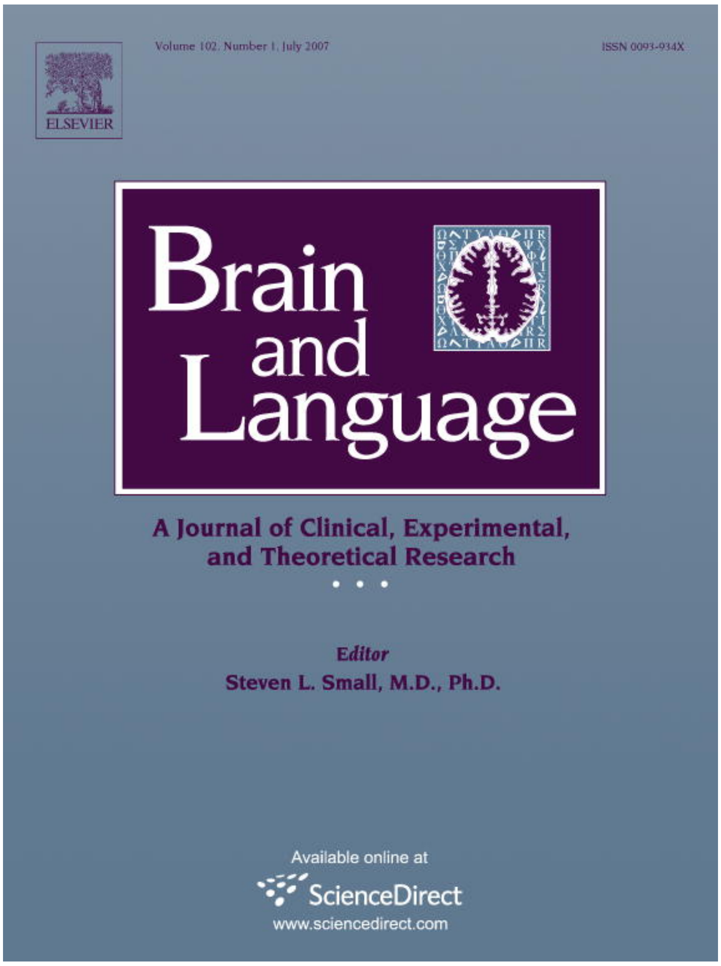
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Evaluation of narrative abilities in patients suffering from Duchenne Muscular Dystrophy

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Abstract

The present work investigated cognitive, linguistic and narrative abilities in a group of children suffering from Duchenne Muscular Dystrophy, an allelic X-linked recessive disorder caused by mutations in the gene encoding dystrophin. The patients showed mildly reduced IQ with lower Verbal than Performance Intelligence Quotient and were mildly affected in visual attention and short-term memory processing. At the linguistic assessment, neither receptive (word comprehension) nor expressive (naming tasks and fluency) lexical abilities were impaired. However, their narratives were qualitatively inferior with respect to those produced by a group of typically developing children. Their speech samples were characterized by the presence of fewer verbs and complete sentences. It is suggested that the reduced production of complete sentences is due to a selective problem in verb argument structure generation. Since the lack of dystrophin is assumed to produce effects on the maturation of the cerebellum, whose involvement has been recently suggested in verb and syntactic processing, these findings may lend indirect support to the hypothesis of a cerebellar-cortical circuit specialized in verb and sentence production.

Keywords: Duchenne Muscular Dystrophy; Discourse analysis; Dystrophin; Cerebellum

1. Introduction

Duchenne-type Muscular Dystrophy (DMD) is characterized by progressive degenerative myopathy. It is an allelic X-linked recessive disorder caused by mutations in the gene encoding dystrophin (Koening et al., 1987), a protein present in skeletal muscle and brain tissues (Ahn & Kunkel, 1993) whose absence may affect synaptic functions (Anderson, Head, Rae, & Morley, 2002) and cause region-specific hypometabolism (Lee et al., 2002). Dystrophin is relatively abundant in the cerebellum, in the hippocampus and in the cerebral neocortex, areas directly involved in higher cognitive functions (Bresolin, Castelli, Comi, & Felisari, 1994). Approximately 30–50% of DMD patients present with moderate to severe non-progressive mental retardation (Emery, 1993) and reduced Intelligence Quotient (IQ) of approximately one Standard Deviation (*SD*) below normal range (Cotton, Voudouris, & Greenwood, 2001).

Cognitive and intellectual deficits in patients suffering from DMD have been documented in a number of studies since the original observation of Duchenne (1868) who presented the case of a 7-year-old child suffering from a progressive muscle weakness associated with "poor

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language" and "dull intellect". Such mild mental retardation (Cotton et al., 2001) reflects a specific cognitive profile (Hinton, DEVivo, Nereo, Goldstein, & Stern, 2000). Marsh and Munsat (1974) documented an early impairment of Verbal (VIQ) rather than Performance (PIQ) intelligence in 34 male DMD patients assessed with the Wechsler Intelligence Scale for Children—Revised (WISC—R: Wechsler, 1993). Moreover, Karagan, Richman, and Sorensen (1980) found a basic language deficit characterized by poor expressive verbal skills and a deficiency in short-term memory "for patterns, numbers, and verbal labels" (p. 422). Such short-term memory deficits may play a critical role in the cognitive and linguistic development of DMD children (Wicksell, Kuhlgren, Melin, & Eeg-Olofsson, 2004). Indeed, Elliger, Dacheneder, and Schotensach (1990) showed the presence of a language delay in these children, as they are typically late speaking compared to accepted developmental milestones.

Linguistic deficits are reported in DMD sufferers also in reading abilities (Billard, Gillet, Barthez, Hommert, & Bertrand, 1998; Dorman, DesNoyers Hurley, & D'Avignon, 1988). Dorman et al. (1988) confirmed the dissociation between VIQ and PIQ with greater impairment of verbal components in a task assessing naming, verbal fluency and reading abilities. Billard et al. (1998) explained the impaired reading of non-words in a group of DMD children in terms of a language-based learning deficit suggesting that such a deficit may be caused either by a selective inability to analyze the phonetic pattern of a word, or in perceiving its visual shape. Moizard et al. (1998) reported a deficit in verbal intelligence rather than global mental deficiency in 49 patients with DMD with the absence of Dp71 and Dp140 brain dystrophin isoforms. A comparison of cognitive abilities and gene mutations revealed that patients with a VIQ below 70 and reading difficulties presented a gene deletion or duplication which was preferentially located in the distal part of the gene.

The cognitive deficits do not seem to be progressive. Sollee, Latham, Kindlon, and Bresnan (1985) compared younger (mean age = 7 years) and older (mean age = 13.3 years) patients diagnosed with DMD on a set of neuropsychological tests and found that younger patients performed worse than the older ones in selective areas of language ("naming") and attentional skills. More specifically, while younger patients were impaired in language and attentional skills, older DMDs did not show any linguistic or attentive impairment, but defective motor skills. The authors concluded that in patients with DMD both language and attentional skills may improve with age, but that such improvement in VIQ is mirrored by a deterioration in their PIQ, "due to loss of points for motor speed, not associated with cognitive deterioration". Therefore, there may be a dissociation between the muscular symptoms which get worse over time, and cognitive deficits which get better with increasing age. In a recent paper, Cotton, Voudouris, and Greenwood (2005) confirmed this hypothesis showing that VIQ increases significantly with age and that younger DMD patients (age $<$ 14 years old) show deficits in verbal reasoning and verbal processing, while older patients (age $>$ 14 years old) are less likely to present with such deficits.

It has been suggested that the verbal deficits reported in DMD patients may be due to selective impairment in short-term memory processing while coping with a neuropsychological test, i.e. in retaining preceding pieces of information over a few seconds as new information is presented, or alternatively an inability to maintain sustained attention to the task (Anderson, Routh, & Ionanescu, 1988). Such an interpretation was confirmed by a more recent study reporting DMD patients performing poorly on tests that required attention to complex verbal information such as comprehension and story recall (Hinton et al., 2000).

The reading deficits observed in DMD boys (Billard et al., 1998; Dorman et al., 1988) are similar to those described in developmental dyslexia (Nicolson, Fawcett, & Dean, 2001) which have been shown to be related to structural and/or functional abnormalities in the right cerebellum. Similarly, deficits in verbal working memory (Hinton et al., 2000) observed in DMD children have been shown to have a cerebellar focus (Desmond, Gabrieli, Wagner, Ginier, & Glover, 1997). The high concentration of dystrophin in the cerebellum and the hippocampus in normal subjects prompted the attention to these regions of the brain. A decreased uptake of fluorodeoxyglucose, evaluated by Positron Emission Tomography (PET), was observed in DMD children but not in a patient with Wernig–Hoffman disease with normal Full-Scale Intelligence Quotient (FIQ), suggesting that cerebellar hypometabolism is unrelated to motor deficits (Bresolin et al., 1994) but indicative of a lowered synaptic activity (Jueptner & Weiller, 1995) and altered bioenergetics in DMD patients (Misuri et al., 2000; Tracey et al., 1995). Glucose hypometabolism is more evident in the right post-central and middle temporal gyri, uncus, and the cerebellar lobule, as well as in the left hippocampal gyrus and cerebellar lobule (Lee et al., 2002). Biochemical studies on the mouse model of DMD (the mdx mouse) have shown an involvement of the "neuronal dystrophins" in NMDA receptor and/or $GABA_A$ receptor function, that might modulate neuronal excitability and network activity and consequently might play a specific role in memory functions involved in cognitive disorders described in DMD children (Knuesel et al., 1999; Magarinos, Verdugo, & McEwen, 1997; Rae et al., 1998; Tracey et al., 1995).

The purpose of the present study is to describe in detail the cognitive abilities of a group of DMD patients as well as their micro- and macrolinguistic abilities (Marini, 2001; Marini & Nocentini, 2003) while processing complex textual structures such as descriptions of visually presented cartoon-stories. The narratives produced by the group of DMD patients were compared to those provided by a group of normally developing children matched for chronological age and level of formal education. The elicited narratives were analyzed using a large array of measures that allowed to evaluate microlinguistic, macrolinguistic and informative aspects of discourse processing (Marini, Boewe, Caltagirone, & Carlomagno, 2005; Marini, Carlomagno, Caltagirone, & Nocentini, 2005). Here, it was hypothesized that such a detailed analysis would highlight aspects of impaired processing in the DMD subjects and plausible connections among attentional, visuo-spatial, memory and linguistic/communicative processing.

2. Materials and methods

2.1. Subjects

Sixty-one Italian-speaking participants matched for chronological age, education and gender were included in the study (Table 1). They formed two groups. The experimental group consisted of 21 male children suffering from DMD. The control group, formed by 40 healthy male children, was included in order to compare the narratives produced by the group of DMD participants with those uttered by typically developing children (see Section 2.2.4). The control participants, aged between 6 and 10 years old (mean 8 years and 5 months; *SD* 1.1), were randomly selected from mainstream schools. All participants performed within normal range on the block-design subtest of the WISC—R (which is known to have a high correlation with Performance IQ: $r = .7$, Wechsler, 1981) and on the tasks included in the "Test of Morpho-Syntactic Development" (TSM: Fabbro & Galli, 2001) which assess their linguistic abilities in sentence comprehension, morphological transformations and word and pseudo-word repetition. Their level of formal education ranged from 2 to 5 years (mean 3 years and 5 months of education) and all showed average school performance in language arts and reading. In a preliminary interview, their teachers confirmed that they showed normal cognitive and learning development. According to school records and parents' reports, moreover, none of them had a known history of psychiatric or neurological illness, learning disabilities, hearing or visual loss. Therefore, overlapping between chronological and mental age was assumed. In the neuropsychological and neurolinguistic tasks no control group was included as the scores from the DMD participants were considered with respect to general population norms. The two groups differed in mental age [$F(1, 60) = 12.029$; $p = .001$] (see Table 1).

Table 1
Means (and standard deviations) of chronological age, level of education (years) and mental age in the two groups of participants

	Controls	DMD
Chronological age	8.5 (1.1)	8.3 (1.1)
Education	3.5 (1.1)	2.9 (1.2)
Mental age	8.5 (1.1)	7.4 (1.4)

The DMD participants were diagnosed according to international standard criteria (Jennekens, Ten Kate, de Visser, & Wintzen, 1994): progressive muscular weakness; increased muscle plasma enzymes; muscular biopsy identifying degeneration and absence of the dystrophin protein; alterations on the dystrophin gene (deletion in the DMD gene or other genetic alteration as duplications and point mutations); family history of X-linked muscle disease. The DMD children were aged between 5.7 and 10.2 years (mean chronological age 8.3 years). At the time of the evaluation, twelve children were still ambulant while the remaining nine patients could not walk anymore. In these nine patients the loss of walking capability manifested between 4.5 and 8.4 years. None of the children suffered from additional diseases such as deafness, severe visual impairment, family history of mental retardation, epilepsy, cerebral palsy, behavioural and/or psychiatric disturbances (e.g. emotional problems or depression). All parents gave informed consent.

2.2. Procedures

2.2.1. Intelligence test

General (verbal and performance) intelligence of DMD participants was assessed administering age-appropriate Wechsler Intelligence Scales: the WISC—R (Wechsler, 1993) for children older than six, and the WPPSI (Wechsler, 1996) for younger children. Results are expressed as IQ scores (mean, 100; *SD*, 15) for the Verbal and the Performance Scale, and as scaled scores (mean, 10; *SD*, 3) for the single subtests.

2.2.2. Neuropsychological assessment

The neuropsychological assessment focused on those cognitive functions which are needed to fulfil the specific linguistic tasks employed in the experiment: i.e. visual attention, verbal memory, visual memory and cross-modal memory.

Visual attention, visual/verbal cross-modal memory and verbal learning were assessed administering specific subtests of the NEPSY Developmental Neuropsychological Assessment (Korkman, Kirk, & Kemp, 1998). Visual attention was assessed with a visual search task in which the subject is required to mark target pictures on structured or non-structured arrays of pictures on large sheets. Both accuracy and speed were recorded. The total scores computed from speed and accuracy scores are expressed as *z*-scores. Visual/verbal cross-modal memory was assessed by means of the "Memory for names" subtest. The subject must learn and recall the names of eight children whose faces are shown in line drawings in three immediate recall trials. Total scores are computed from the total number of correct responses on the three trials and expressed as *z*-scores. Verbal learning, supra-span memory and resistance to interference were assessed administering the "List Learning" subtest. The test assesses the immediate recall of word lists over five repeated trials without interference and one with verbal interference, as well as the delayed recall 30 min after the presentation of the stimuli. Total scores are computed from the first five trials and the delayed trial and expressed as *z*-scores.

Visual memory was assessed with the "Abstract Visual Memory" subtest taken from the TEMA battery (Italian version of TOMAL, Test of Memory and Learning: Reynolds & Bigler, 1994, 1995). This subtest assesses immediate recognition of abstract, non-verbalizable pictures. The subject is first shown the picture and then requested to recognize it among a number of distracters. The total number of correct responses is recorded. Results are expressed as *z*-scores.

2.2.3. Linguistic assessment

The participants' general linguistic abilities were analyzed administering the "Batteria 4–12" ("Battery for linguistic assessment of children from 4 to 12 years", Fabbro, 1999), the Italian adaptation of the "Batterie d'évaluation du langage oral de l'enfant aphasique" (ELOLA) (De Agostini et al., 1998). It is a testing battery for linguistic assessment in children aged from 4 to 12 years. The battery includes tests examining phonological, lexical and syntactic abilities in comprehension, production and repetition. All linguistic tests provide age-related Italian norms (expressed as means and standard deviations for each age-group, usually for each year of age between 4 and 12 years).

As to comprehension, subjects' verbal auditory discrimination was assessed administering the "Same–Different Judgement" test, a phonemic identity judgement task. The subject is requested to say if a couple of words, either identical or constituting a "minimal pair" includes "same" or "different" words. Semantic comprehension was assessed administering the Picture Identification and the word comprehension tests. The former requires the child to match each of 30 words read by the examiner with one out of four pictures (the target, two phonological distracters and a semantic distracter). The latter was adapted from the British Picture Vocabulary Scale (De Agostini et al., 1998; Dunn, Dunn, & Whetton, 1982), and requires matching each of 25 words read by the examiner with one out of four pictures (the target and three semantic distracters). Morpho-syntactic comprehension was assessed with the "Test of Grammatical Comprehension for Children" (TCGB: Chilosi & Cipriani, 1995) where the subjects are required to match each of 76 sentences of increasing complexity with one out of four pictures. Syntactic comprehension was assessed with the Italian version of the Token Test for children (De Agostini et al., 1998; Di Simoni, 1978).

The participants' production was assessed in a Noun and Verb Naming task requiring subjects to name 36 object pictures and five pictures representing actions (De Agostini et al., 1998; Riva, Nichelli, & Devoti, 2000) and a test of semantic fluency (Fabbro, 1999; Riva et al., 2000) where subjects are prompted to name, during 90 s, as many words as possible belonging to two semantic categories: animals and house objects. The largest number of correct words produced in 60 consecutive seconds is recorded as test score.

Repetition abilities were assessed by administering a Sentence Repetition task requiring the subjects to repeat a list of sentences of increasing length and complexity (Ferrari, De Renzi, Faglioni, & Barbieri, 1981; Vender, Borgia, Cumer Bruno, Freo, & Zardini, 1981). An additional test of word and non-word repetition was taken from the "Test of Morpho-Syntactic Development" (TSM: Fabbro & Galli, 2001).

2.2.4. Assessment of narrative abilities

The narrative assessment was performed on the storytellings elicited with a picture–story description task. Each subject was asked to produce a narrative elicited with the help of a cartoon story (the "Nest Story", Paradis, 1987) consisting of a series of six drawings presented on the same page. Since for this picture description task no normative data are currently available, the narratives produced by the group of DMD participants were compared to those uttered by the group of 40 healthy controls. In order to avoid poor performance due to short-term memory limitations (Anderson et al., 1988; Wicksell et al., 2004), the cartoon story remained visible until the subject had finished his description. Each story telling was tape-recorded and subsequently transcribed verbatim by one of the authors (AM) and by two experienced speech therapists including phonological fillers, pauses, false starts and extraneous utterances. These transcriptions were compared in order to obtain highly reliable transcripts for the analysis. Discrepancies were discussed and resolved before the narratives were analyzed further.

The stories were subjected to a quantitative, in-depth textual analysis involving both micro- and macrolinguistic aspects of narrative production. The analysis focused on four main aspects of linguistic processing: verbal productivity, lexical and syntactic organization, informativeness and textual organization (Marini & Carlomagno, 2004). Both simple values and ratios were considered to account for the difference across narratives. The scoring procedure was performed independently by two raters and then compared. Differences were resolved through discussion.

Verbal productivity was measured as the number of units produced by each subject during the story telling. The unit count included all verbalizations, irrespective of their linguistic or contextual correctness or appropriateness (Haravon, Obler, & Sarno, 1994; Marini, Boewe et al., 2005). The total number of well-formed words excluding phonological fillers, phonemic paraphasias and phonetic errors was then computed. The number of words was used to obtain a measure of speech rate in terms of words per minute (words/m').

The lexical and syntactic organization was measured in terms of lexical and morpho-syntactic processing. The subject's lexical diversity was measured by means of the Type-

Token Ratio (TTR). This index was computed by dividing the number of different open-class words (types) by the total number of open-class words produced (tokens). Lexical processing was assessed in three ways. At a first step, the analysis concerned the lexeme level (Levelt, Roelofs, & Meyer, 1999) of word processing and ratios of phonological selection and of production of phonological paraphasias were computed. The ratio of phonological selection (Marini, Boewe et al., 2005; Marini, Carlomagno et al., 2005; Marini, Caltagirone, Pasqualetti, & Carlomagno, 2007) was obtained by dividing the number of words by the number of units. Thus, the ratio of phonological selection allowed assessing the ability of the subjects to retrieve phonologically well-formed words. The ratio of phonological paraphasias was then computed dividing the number of phonemic paraphasias by the number of units produced in each description. Finally, the lemma level was assessed in terms of production of semantic paraphasias and paragrammatic errors. The ratio of semantic paraphasias was calculated by dividing the number of semantic paraphasias by the number of words (Marini, Boewe et al., 2005). Higher values represent more semantic errors per word. These errors were scored when a target word was substituted by a semantically related word (Haravon et al., 1994). An example for a semantic paraphasia is the word "mother" in the sentence "*here he's talking to his mother*", where the speaker intended to say "wife". The ratio of paragrammatic errors included grammatical errors with bound morphemes (for example, *questo è una coppia* "this *[masc]* is a couple [fem]"—should be *questa*) and function words (for example, *batte da una porta* "he is knocking *from* a door"—*da* instead of *a*). Higher values in the ratio of paragrammatisms in the descriptions represent more paragrammatisms per word.

Morpho-syntactic organization was evaluated in terms of Mean Length of Utterance (MLU), verb production and proportion of complete sentences. For each story description, the total number of utterances was assessed following the criteria established in the Shewan Spontaneous Language Analysis System (Shewan, 1988). Under the assumption that longer utterances reflect the attempt to produce articulated sentences, a ratio measuring the mean length of utterance (MLU) was calculated dividing the total number of words by the number of utterances. Verb count was introduced in the analysis because verb processing can be considered a more valid measure of morpho-syntactic development. Indeed, since verbs generate the argument structure needed to produce or understand grammatically corrected sentences, a good production of verbs may reflect the achievement of more mature morpho-syntactic skills (Marini, Boewe et al., 2005). Moreover, a count of complete sentences was performed. A sentence was considered grammatically complete if all the arguments required by the verb were inserted correctly in the body of the sentence and if there were no omissions or substitutions of free or bound morphemes. Production of complete sentences was measured by dividing the number of grammatical sentences by the number of utterances (Saffran, Berndt, & Schwartz, 1989).

The informative content of each narrative was measured in order to obtain numerical evaluation of lexical-semantic appropriateness in describing the gist of the story. The lexical-semantic appropriateness was determined counting the Lexical Information Units (LIUs), i.e. words that were not only phonologically well-formed but also appropriate from a grammatical and pragmatic point of view (Marini, Boewe et al., 2005; Marini, Carlomagno et al., 2005). Therefore, all those words that were classified as semantic or verbal paraphasias, fillers, paragrammatisms or present in tangential utterances (i.e. utterances that were somehow deviating from the gist of the story) were excluded from the LIUs count. The ratio of lexical informativeness was obtained dividing the amount of LIUs by the amount of words.

As for discursive organization, the macrolinguistic measures included degrees of local coherence and local coherence errors. As discussed in Marini and Boewe et al. (2005) both positive and negative measures of coherence abilities were included under the assumption that positive and negative measures are not mirror images of each other. Indeed, a high coherence error rate does not automatically imply a low degree of coherence. The degree of local coherence measured the extent to which each utterance of the story was conceptually related to the preceding one. Local coherence is realized through cohesive ties like anaphoric pronouns, number and gender agreement between pronouns or NPs across utterances, cohesive function-words and lexical items which are semantically related (Halliday & Hasan, 1976). The degree of local coherence was determined by dividing the number of cohesive ties in each story by the number of utterances. The ratio of local coherence errors was measured by dividing the total number of local coherence errors by the number of utterances produced. Missing or ambiguous referents and semantic shifts were counted as local coherence errors. A semantic shift was scored when there was an abrupt stop within an utterance, after which the flow of thoughts was not continued in the following utterance but, instead, a new concept began. For instance, in "*he's trying to... and here he is on a landing*" the first utterance remained unfinished, the second utterance introduced a new scene. Missing referents were instances in which the referent of a pronoun or the implicit subject of a verb (Italian is a pro-drop language) were not unambiguously clear or were incorrect. For example, consider the following sequence of utterances: "*Qui stanno litigando furiosamente. Poi dice: (...)*" ("here they are fighting furiously. Then [*implicit pronoun*] says: (...)"). In the second utterance there was a missing referent because it was not clear whom the verb "dice" referred to.

3. Results

3.1. Cognitive and neuropsychological assessment

The results from the cognitive and neuropsychological assessment showed a general reduction in almost all cognitive functions in DMD patients (see Tables 2 and 3). Their

Table 2
Performance of the DMD participants at the verbal, performance and full IQ tests

	Mean (*SD*)	Range	*p*-value
Verbal IQ	84.81 (17.03)	51–118	.001
Performance IQ	90.19 (16.61)	49–113	.014
Full-Scale IQ	85.81 (17.00)	51–112	.001

FIQ scores (mean 85.8) were about one *SD* below average, with a relative discrepancy between the Verbal Scale (mean 84.8) and the Performance Scale (mean 90.2). More precisely, 12 out of 21 patients (57.1%) had a VIQ lower than PIQ, while for the remaining 9 patients (42.9%) VIQ was higher than PIQ. The two groups characterized by opposite profiles did not differ in chronological age or FIQ. A *t*-test against average values was performed on the three measures of intelligence. The analysis revealed that the DMD patients were significantly below normal level in FIQ ($t(20) = 3.82$, $p = .001$), PIQ ($t(20) = 2.71$, $p = .014$) and VIQ ($t(20) = 4.09$, $p = .001$).

The results of the neuropsychological and linguistic evaluation are presented as mean *z*-scores for each cognitive and linguistic parameter. In order to determine whether the reduction in general intellectual abilities (reduced FIQ) may have affected the patients' performance on those tests assessing linguistic and narrative abilities, the *z*-scores were corrected by subtracting the effect of FIQ estimated by means of a linear regression analysis. A *t*-test was then performed to determine which of these corrected variables were significantly below average. The results of the neuropsychological assessment, including non-corrected and corrected *z*-scores are reported in Table 3. Scores from the Digit Span, Picture Arrangement and Picture Completion subtests of the WISC—R (measuring verbal short-term memory, logical/sequential analysis and visual analysis abilities, respectively) are also reported, as they are assumed to influence performance on the narrative task. For all the variables corrected for IQ that were found in a *t*-test to be still significantly below average, *p*-values are reported as well. IQ scores and WISC—R subtests did not undergo any transformation.

As to the neuropsychological assessment, when the *z*-scores were not corrected for IQ the DMD patients performed about one *SD* below average on cognitive tasks measuring short-term memory span ($z = -1.04$), Picture Identification ($z = -1.09$) and visual attention ($z = -1.06$). However, when the *z*-scores were corrected for FIQ the subjects performed within one *SD* below normal range on all measures. Nevertheless, their performance was significantly below average in the tasks assessing short-term memory span (digit span) ($t(14) = 3.73$, $p = .002$) and visual attention abilities ($t(15) = 4.39$, $p < .001$).

At the linguistic assessment, results change critically whether the *z*-scores are corrected for FIQ or not. When considering *z*-scores, the DMD patients performed well below average on verbal auditory comprehension ($z = -3.25$), syntactic comprehension ($z = -2.56$) and morpho-syntactic comprehension ($z = -1.55$). On the other measures their scores fell within one *SD* or even above average. Also in this case, however, when the *z*-scores were corrected for FIQ, the linguistic profile of the patients changed drastically. Indeed, they fell over one *SD* below average only on the Token Test measuring syntactic comprehension ($z = -1.22$), while on the other tasks the DMD children performed within one *SD* or even above average. At a *t*-test their performance was significantly below average only in two tasks, assessing morphosyntactic ($t(20) = 2.87$, $p = .01$) and syntactic comprehension ($t(20) = 2.08$, $p = .05$), respectively. Verbal auditory discrimination, word comprehension, lexical and phrasal repetition and lexical production were unimpaired.

3.2. Assessment of narrative abilities

A multivariate analysis of variance with *group* (1, Controls; 2, DMD participants) as fixed factor and *mental age* and *years of formal education*[1] as covariate was performed on the following 13 dependent variables: words; speech rate; ratio of phonemic paraphasias; ratio of phonological selection; Type-Token Ratio; verbs; mean length of utterance; ratio of complete sentences; ratio of paragrammatic errors; lexical informativeness; ratio of semantic paraphasias; degree of cohesion; ratio of local coherence errors. The criterion for significance was $\alpha = .05$. The results will be presented in two separate sections: a microlinguistic analysis, and a macrolinguistic analysis.

3.3. Microlinguistic analysis

The mean values for each group at each microlinguistic measure are reported in Table 4. Although the two groups did not differ in Speech Rate, the control group produced more Words [$F(1,57) = 5.677$; $p = .021$] and fewer phonological paraphasias [$F(1,57) = 6.458$; $p = .014$] than the DMD group. This corresponded to higher values in phonological selection for the controls than the DMDs [$F(1;57) = 13.255$; $p = .001$]. The two groups did not differ in Type-Token Ratio, MLU and ratio of paragrammatic errors but the control group produced less semantic paraphasias [$F(1,57) = 4.508$; $p = .038$], more verbs [$F(1,57) = 9.091$; $p = .004$], and correspondingly more complete sentences [$F(1,57) = 16.693$; $p = .0$].

3.4. Macrolinguistic analysis

The mean values for each group at each macrolinguistic measure are reported in Table 5. The controls scored better on lexical informativeness than patients suffering from DMD [$F(1,57) = 13.022$; $p = .001$]. The textual organization of the story-tellings produced by the control group

[1] Education level was included as covariate because the mean difference between the two groups (2.9 vs. 3.5 years of education), even if non-significant, may still be an important difference at this age.

Table 3
Means (and standard deviations) of the scores obtained by the DMD group on the cognitive and neuropsychological tasks

Neuropsychological and linguistic assessment	Mean (*SD*) *z*-scores	Mean (*SD*) corrected *z*-scores	*p*-value
Digit span (WISC—R subtest)	−1.04 (1.08)	—	.002
Picture arrangement (WISC—R subtest)	−0.33 (0.97)	—	NS
Picture completion (WISC—R subtest)	−0.30 (1.05)	—	NS
Picture identification	−1.09 (1.85)	−0.22 (1.56)	NS
Visual attention	−1.06 (0.80)	−0.86 (0.79)	.001
Abstract visual memory	−0.48 (0.63)	−0.14 (0.60)	NS
Memory for names (cross-modal)	−0.44 (0.95)	−0.09 (0.87)	NS
List learning	0.07 (0.80)	0.20 (0.80)	NS
Verbal auditory discrimination	−3.25 (6.64)	−0.76 (5.50)	NS
Word comprehension (BPVS)	0.11 (1.38)	0.69 (1.28)	NS
Syntactic comprehension (Token Test)	−2.56 (3.28)	−1.22 (2.69)	.05
Morpho-syntactic comprehension (TCGB)	−1.55 (1.66)	−0.76 (1.22)	.01
Sentence repetition	−0.47 (1.19)	−0.07 (1.11)	NS
Word and non-word repetition	0.13 (1.06)	0.41 (1.04)	NS
Noun naming	−0.16 (1.41)	0.39 (1.28)	NS
Semantic fluency	−0.05 (0.62)	0.15 (0.57)	NS

For each variable, non-corrected *z*-scores are reported along with *z*-scores corrected for IQ. *p*-values are reported for corrected *z*-scores (normal *z*-scores for WISC—R subtests) that were found to be significantly below average (*t*-test).

Table 4
Results of the microlinguistic analysis

Parameter	DMD	Controls
Words*	56.10 (21.7)	76.3 (25.9)
Speech rate	100.84 (34.3)	112.3 (28.5)
Phonological paraphasias*	0.96 (1.61)	0.09 (0.41)
Phonological selection*	95.6 (4.10)	98.9 (1.35)
TTR	79.8 (9.79)	77.6 (9.97)
Verbs*	9 (2.66)	12.9 (3.87)
Semantic paraphasias*	0.99 (1.37)	0.25 (0.52)
MLU	6.33 (1.76)	7.56 (1.74)
Paragrammatic errors	1.72 (1.60)	1.12 (1.15)
Complete sentences*	0.68 (0.22)	0.88 (0.11)

*Indicates when the group-related difference is significant.

presented with higher levels of cohesion [$F(1,57) = 10.463$; $p = .002$] and fewer violations of local coherence [$F(1,57) = 17.782$; $p = .0$.

3.5. Evaluation of chronological age effects within the group of DMD participants

In order to determine whether chronological age affects performance on the narrative task, the DMD participants were divided into two age-groups: the first group included 12 DMD patients aged 5.7–8.08 years (mean, 7.6; *SD*, .9); the second group was formed by 9 DMD patients aged 9–10.02 years (mean, 9.2; *SD*, .4). A one-way ANOVA with *group* as fixed factor and the *nine measures* which had been found to be impaired in the DMD group as dependent variables (words; ratio of phonemic paraphasias; verbs; ratio of phonological selection; lexical informativeness; ratio of semantic paraphasias; ratio of complete sentences; degree of cohesion; ratio of local coherence errors) was performed. The analysis showed the absence of age-related differences between the two groups of DMD participants (all *p*-values >.05). An additional one-way ANOVA showed that the two subgroups did not differ in FIQ, VIQ or PIQ.

Table 5
Results of the macrolinguistic analysis

Parameter	DMD	Controls
Lexical informativeness*	77.72 (12.20)	90.14 (7.35)
Degree of cohesion*	0.54 (0.23)	0.76 (0.16)
Local coherence errors*	0.31 (0.30)	0.06 (0.09)

*Indicates when the group-related difference is significant.

3.6. Correlations between measures of narrative abilities and cognitive functions

In order to better understand the origin of the specific difficulties found for DMD children in the narrative production, correlations were performed among those neuropsychological functions that were found to be weakened (i.e. digit span and visual attention) and those aspects of the narrative production that were impaired (see Tables 4 and 5). Since differences in mental age may have caused spurious correlations to emerge (indeed, practically all variables were highly correlated with mental age), partial correlations controlling for mental age were computed, using raw scores (since no *z*-scores were available for narrative variables).

Significant positive correlations emerged between visual attention and production of verbs ($r = .713$; $p = .009$) and between visual attention and degree of local coherence ($r = .640$; $p = .025$). It should be noted that the correlation between visual attention and the ratio of lexical informativeness approached significance ($r = .553$; $p = .062$). These results suggest the possibility that the quantity and quality of the narrative descriptions of picture–stories may be influenced by the accuracy of visual inspection. More difficult to explain are the negative correlations found between the digit span subtest of the WISC—R and the degree of phonological selection ($r = -.778$; $p = .01$), as well as phonological paraphasias ($r = .658$, $p = .011$). A tentative interpretation could be that those participants who have a better verbal memory also produce more complex descriptions and are therefore more prone to phonological errors. This interpretation is supported by the absence of correlation between the aforementioned variables when MLU instead of mental age was controlled for.

4. Discussion

The present work investigated cognitive, linguistic and narrative abilities in a group of DMD participants compared to a group of normal controls. All aspects of linguistic processing were measured, namely verbal production, comprehension and repetition. The narrative assessment included micro- and macrolinguistic analysis of the cartoon-story descriptions provided by the subjects.

Several studies on intellective functions in DMDs report a mild mental retardation in the Full-Scale Intelligence Quotient (Anderson et al., 1988; Emery, 1993), often associated with lower Verbal than Performance Intelligence Quotient (Dorman et al., 1988; Marsh & Munsat, 1974). Such a dissociation is assumed to reflect a selective deficit in specific aspects of linguistic processing (Elliger et al., 1990; Karagan et al., 1980) in DMD patients younger than 14 years of age (Cotton et al., 2005). The present study found that such general reduction in intellectual abilities is associated with a considerable heterogeneity among boys with DMD. The mean scores were approximately one *SD* below normal range with FIQ averaging 85.8 (range 51–112). These data indicate that the DMD patients constitute a rather differentiated group with individuals affected by mild mental retardation (defined by an FIQ range comprised between 50 and 70; see Hallahan & Kauffman, 1988) along with subjects with normal or even above-average intelligence quotients. As to Verbal vs. Performance Intelligence Quotients, the DMD boys' Verbal IQ (mean 84.8; range 51–118) was on average more affected than their Performance IQ (mean 90.2; range 49–113), thus confirming previous reports (see also Cotton et al., 2001). However, a certain degree of individual variability could be observed in the sample. More precisely, no more than 57.9% of the patients in the present sample showed lower VIQ than PIQ. No differences emerged with respect to chronological age and FIQ, suggesting that these two kinds of profiles can be found in all age groups and at different levels of global intellectual functioning. Previous findings suggesting that the discrepancy between VIQ and PIQ tends to be reduced in older subjects (Cotton et al., 2005; Sollee et al., 1985), therefore, were not confirmed by the present data, although it should be considered that the subjects included in this study were generally younger than those described in previous studies.

The results from the neuropsychological and linguistic assessment were treated as *z*-scores matched to normal values. When *z*-scores were taken into account, the DMD group showed reduced processing abilities in several aspects of the cognitive and linguistic domain. However, when the *z*-scores had been corrected for IQ, the DMD participants were mildly affected only in visual attention and in two linguistic measures, i.e. morpho-syntactic and syntactic comprehension. Since these functions were confirmed to be significantly below average after the correction for FIQ, such aspects of cognitive/linguistic functioning are probably to be considered *specifically* impaired in DMD children. Phonological abilities, as measured by auditory discrimination, picture identification and repetition tasks showed no impairment after IQ had been taken into account. Similarly, visual memory abilities, verbal learning and cross-modal memory were less than one *SD* below average before correction, and perfectly on average after correcting for IQ. However, the DMD boys performed significantly below normal range (over one *SD* below average) on the digit span subtest. This finding confirmed previous reports about a deficiency in short-term memory for patterns, numbers, and verbal labels (Karagan et al., 1980; Wicksell et al., 2004).

Interestingly, neither receptive (word comprehension) nor expressive (naming tasks and fluency) lexical abilities were impaired. These results contrast with previous studies where verbal impairments were found in tasks assessing naming and verbal fluency (Dorman et al., 1988) and poor expressive verbal skills were reported (Karagan et al., 1980). Such discrepancy may be due to the fact that in those studies the patients' responses at tasks assessing basic linguistic functions had not been corrected for general intellectual abilities.

The third main finding of the present work concerns the assessment of the patients' narrative abilities. To the authors' knowledge, no attempt has been made to characterize both micro- and macrolinguistic abilities of DMD individuals. The statistical analysis showed that, even controlling for mental age, the narrative speech of the DMD patients was qualitatively inferior with respect to that produced by the group of typically developing children and that such linguistic deficits were not dependent on chronological age. Their speech samples were shorter, characterized by the presence of fewer words. They produced less verbs but more phonological and semantic paraphasias than the control group. As to their morpho-syntactical and syntactical skills, the reduced proportion of complete sentences was not due to a generic inability to produce long utterances, as the mean length of utterance ratio in their speech was comparable to that of the normally developing control children. Rather, an inspection of the speech samples produced by the group of DMD participants revealed that the majority of the incomplete sentences uttered by the patients were missing function and content words, especially verbs. Such association between reduced verb and function words production and reduced proportion of grammatically complete sentences suggests the possibility that both verb and sentence production may be the expression of an underlying common deficit. More specifically, a selective problem in verb argument structure generation and thematic role assignment may undermine the ability to produce complete sentences (Garrett, 1988; Levelt, 1989; Levelt et al., 1999; Marshall, Pring, & Chiat, 1998; Saffran, Schwartz, & Marin, 1980). Interestingly, such behaviour resembles that of agrammatic aphasic patients, whose speech is often characterized by verb retrieval deficits associated with impaired sentence production (Berndt, Mitchum, Haendiges, & Sandson, 1997a, 1997b; Miceli, Silveri, Villa, & Caramazza, 1988). It is noteworthy that the weakened ability to generate verbs may have determined the reduced level of lexical informativeness in DMDs' speech and their problems in generating adequate cohesive and coherent extra-sentential links among the utterances.

The apparent discrepancy between the lack of linguistic deficits in production at the general linguistic assessment and the presence of selective linguistic impairments at the assessment of the patients' narrative abilities suggests that standardized linguistic tests may not be sensitive enough to capture an individual's communicative performance. Indeed, in story description tasks speakers tend to be more fluid communicators and to make use of several linguistic skills in a communicatively oriented interaction. As a consequence, discourse analysis allows to control for the interaction among the various processing levels (such as the interaction between verb processing, argument structure generation, sentence production and inter-utterance integration) and incorporates measures that are not taken in consideration in traditional linguistic batteries, such as indexes of informativeness and of the individuals' ability to generate cohesive and coherent ties among utterances. It is noteworthy that measures of lexical informativeness, based on the Correct Information Units (CIUs) analysis by Nicholas and Brookshire (1993), have been shown to have ecological validity. For example, Jakobs (2001) reported that changes in informativeness of connected speech samples by non-fluent aphasic patients after therapy were perceived by naïve listeners. Moreover, measures of discourse informativeness and efficiency are functionally based, in that they capture pragmatic and organizational aspects of discourse structure (Armstrong, 2000; Jakobs, 2001; Marini et al., 2007). Indeed, cartoon-story description tasks may increase demands of message level processing thus leading to a massive drain on cognitive resources that complicate lexical access and grammatical encoding in natural communicative situations (Linebarger, McCall, & Berndt, 2004; Marshall et al., 1998). Therefore, the linguistic samples obtained in story description tasks can be long and complex enough to allow researchers or clinicians to evaluate a patient's talkativeness in a more spontaneous context than standard linguistic tasks.

The second point of interest concerns the plausible reasons for the peculiar linguistic/narrative behaviour observed in the DMD participants. A possibility is that such results may reflect the influence of other cognitive factors on the specific task of story description. However, the fact that no specific impairment was found in the Picture Arrangement and Picture Completion subtests of the WISC—R suggests that the DMD patients had the cognitive skills required to process the information presented in a picture–story format. Even though the positive correlations between visual attention and both verb production and degree of local coherence suggest the possibility that the low production of verbs may be influenced by the accuracy of visual inspection, it is noteworthy that the reduced visual attention processing was within one *SD* below average level and thus can not be regarded as pathological. An alternative explanation may lie in the specific bioenergetics and morphological abnormalities inducing altered modulation of neuronal excitability and network activity described in the brain of dystrophin-deficient mice and humans (Bresolin et al., 1994; Lee et al., 2002; Knuesel et al., 1999; Rae et al., 1998; Tracey et al., 1995). At the morphological level, the absence of full-length dystrophin is accompanied by impaired synaptic clustering of $GABA_A$ receptors, suggesting a role of dystrophin in the regulation of GABAergic transmission in a subset of inhibitory synapses (Anderson et al., 1988; Knuesel et al., 1999) and also in the increased sensitivity to hypoxia of the hippocampal pyramidal neurons (Mehler, Haas, Kessler, & Stanton, 1992; Vaillend, Billard, & Laroche, 2004). The observation of macroscopic brain abnormalities in DMD autopsies such as reduced brain weight and ventricules enlargement (Bresolin et al., 1994; Rae et al., 1998), with a preferential loss of neuronal populations (i.e. Purkinje cell loss) that normally express dystrophin, and small cortical ischemic infarct support the hypothesis that the lack of dystrophin increases neuronal susceptibility against hypoxia-induced injury (Jagadha & Becker, 1988; Rosman & Kakulas, 1966). Such lack of dystrophin may also produce effects on the correct maturation and consequent functioning of certain structures of the brain, such as the cerebellum which is particularly rich in dystrophin and α-dystroglycan (Blake & Kroeger, 2000; Lee et al., 2002; Lidov, Byers, Watkins, & Kunkel, 1990; Muntoni, Mateddu, & Serra, 1991; Rae et al., 1998). Nevertheless, a region-specific hypometabolism, unrelated to a motor deficit, is suggested by the observation of a decreased uptake of fluorodeoxyglucose in the cerebellum and in the left hippocampal gyrus, evaluated by PET, in DMD children and not in age-matched controls or in a patient with Wernig–Hoffman disease with normal IQ (Bresolin et al., 1994; Lee et al., 2002; Misuri et al., 2000; Tracey et al., 1995). Moreover, the reading deficits observed in DMD boys are similar to those described in phonological dyslexia which is associated with abnormalities in the right cerebellum (Billard et al., 1998; Dorman et al., 1988; Rae et al., 1998). Similarly, deficits in verbal working memory in DMD children have been found to have a cerebellar focus (Desmond et al., 1997; Hinton et al., 2000; Ravizza et al., 2006). Recent evidence from functional neuroimaging and human lesion studies suggests the possibility that the cerebellum is involved not only in motor control, but also in more basic aspects of word production such as verbal working memory (Desmond et al., 1997; Fiez, Petersen, Cheny, & Raichle, 1992; Paulesu, Frith, & Frackowiak, 1993), verbal fluency (Schlösser et al., 1998), and verb production (Grabowski et al., 1996; Martin, Haxby, Lalonde, Wiggs, & Ungerleider, 1995; Petersen, Fox, Posner, Mintun, & Raichle, 1989; Raichle et al., 1994) (For a review see Fabbro, 2000; Fabbro, Moretti, & Bava, 2000). In particular, it has been suggested that the cerebellum may play a role in word search, whereas the prefrontal cortex may underlie the selection of words from among competing alternatives (Marvel, Schwartz, & Isaacs, 2004) and that the right postero-lateral cerebellar hemisphere may be implicated in a front-cerebellar circuitry dedicated to verb production (Sach, Seitz, & Indefrey, 2004; Papathanassiou et al., 2000; Desmond & Fiez, 1998; Martin, Haxby, Lalonde, Wiggs, & Ungerleider, 1995; Petersen et al., 1989). For example, in a PET study by Petersen et al. (1989), a group of healthy participants were asked to pronounce aloud appropriate verbs in response to written concrete nouns. In a control condition, they had to simply read the nouns aloud. Activation of the right lateral cerebellum was found in the verb generation condition but not in the control condition. Martin et al. (1995) report selective activations associated with action naming in the left fronto-parietal cortex, the middle temporal gyrus and the cerebellum. More recently, in a PET study by Sach et al. (2004), verb-production was related to an activation in the left inferior frontal gyrus, nucleus lentiformis, thalamus and superior medial cerebellum. Results from human lesion studies further support these findings. For example, single case reports have correlated agrammatic-like symptoms involving difficulties in verb and sentence processing to right cerebellar cortex lesions (Fiez et al., 1992; Justus, 2004; Silveri, Di Betta, Filippini, Leggio, & Molinari, 1998; Silveri, Leggio, & Molinari, 1994). Fiez et al. (1992), for example, described a patient with an extensive right cerebellar lesion who presented with language disturbances in verb generation tasks. Silveri et al. (1998) report poor verbal short-term memory in a patient who had undergone right cerebellar hemispherectomy and hypothesize that the cerebellum is involved in a neuroanatomical loop subserving the rehearsal system (cortico-ponto-cerebellar and cerebello-thalamo-cortical loop), a component of the working memory system which is responsible for inner repetition of verbal information (see also Fabbro, 2000; Schmahmann & Pandya, 1997). In a study from Justus (2004), a group of cerebellar patients was impaired in discriminating between grammatical and ungrammatical sentences, particularly when the error was of subject–verb agreement as opposed to word order. Thus, a damage to the cerebellum might disrupt the cortico-ponto-cerebellar-thalamo-cortical loop resulting in weakened ability to use grammatical morphology. However, not all available data support this claim. Indeed, in a recent study from Richter et al. (2004) 10 patients suffering from mild to severe cerebellar ataxia showed preserved learning on a verb generation task (for similar results see also Helmuth, Ivry, & Shimizu, 1997 and Gebhart, Petersen, & Thach, 2000). Nevertheless, narrative production is a much more exacting task than verb generation. In particular, the speech production task of story telling, may increase demands of message-level processing and such a drain on cognitive resources might complicate lexical access and grammatical encoding (Linebarger et al., 2004; Marini et al., 2007; Marshall et al., 1998) thus inducing the DMD patients to produce incomplete sentences in order to realise their communicative intentions.

In conclusion, even if no neuroimaging evaluation was performed on the DMD participants and therefore no firm conclusion can be drawn from present data, our findings may lend indirect support to the hypothesis of a cerebellar-cortical circuit specialized in verb and sentence production (Dogil et al., 2002; Fabbro et al., 2000). Future investigations directed at assessing the role played by the cerebellum in various structural and functional aspects of discourse processing are needed to help clarifying such aspects of verbal production.

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