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Psychological test usage in duchenne muscular dystrophy: An EU multi-centre study

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ABSTRACT

Aim: During the last two decades brain related comorbidities of Duchenne have received growing scientific and clinical interest and therefore systematic assessment of cognition, behaviour and learning is important. This study aims to describe the instruments currently being used in five neuromuscular clinics in Europe as well as the diagnoses being made in these clinics.
Method: A Delphi based procedure was developed by which a questionnaire was sent to the psychologist in five of the seven participating clinics of the Brain Involvement In Dystrophinopathy (BIND) study. Instruments and diagnoses being used were inventoried for three domains of functioning (cognition, behaviour and academics) and three age groups (3–5 years, 6–18 years and adulthood 18+ years).
Results: Data show wide diversity of tests being used in the five centres at different age groups and different domains. For the intelligence testing there is consensus in using the Wechsler scales, but all other domains such as memory, attention, behavioural problems and reading are tested in very different ways by different instruments in the participating centres.
Conclusion: The heterogeneity of tests and diagnoses being used in current clinical practice underlines the importance for developing a Standard Operating Procedure (SOP) to improve both clinical practice and scientific research over different countries and improve comparative work.

1. Introduction

Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD), are X-linked progressive muscular disorders occurring in a frequency of respectively about one case in 3500–9300, and one case in 16700–18500 new-born boys (Orpha: 98896; Orpha: 98895). Both diseases are caused by mutations in the dystrophin gene, resulting in partial expression of the dystrophin protein in BMD, and an absolute absence of dystrophin and in DMD(1–3). Lack of dystrophin causes progressive muscle weakness, and consequently fatal cardiac and pulmonary complications. Therefore, preservation of muscle plays a central role in daily functioning and has been the main focus of medical interest. In the last two decades there has been a growing interest in neurocognitive and behavioural comorbidities in dystrophinopathies. It has been shown that intellectual disability, autism spectrum disorders, attention deficit disorder and learning difficulties such as dyslexia, are more common in dystrophinopathies than in the typically developing population. Intellectual disability occurs approximately in 19–26% of the DMD patients,

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autistic features in up to 19%, obsessive compulsive features up to 25%, attention deficit disorder 11–32%, and language and speech delays almost to 25%. Moreover, males with DMD score one deviation below average on IQ (84.76) and significantly lower than males with BMD (92.11) [4]. A brain based explanation for this higher prevalence has been reported. Several isoforms of dystrophin (including Dp427, Dp140, and Dp71) are present in the central nervous system (CNS) and are thought to contribute to several higher order functions such as memory and learning [5–17].

Consistent screening of frequently comorbid neuropsychiatric and cognitive conditions in DMD patients is therefore important, [18]. This is to make sure that patients can be appropriately referred for psychiatric and psychological treatment and care, in accordance with the revised Standards of Care (SoC) in 2018 [19]. These Standards of Care describe the importance of early and adequate assessment for neuropsychological functioning in dystrophinopathies, but lack a description of what assessment procedures should be used to make a formal diagnosis.

Looking at the current literature there is a large number of different psychological instruments being used in DMD. A review of 32 studies on intellectual functioning in DMD boys [16] reported over 8 different instruments being used, resulting in a mean IQ of 80.2. In another review of Hellebrekers [20], they found a total of 61 instruments being used in 51 DMD studies. They concluded that different diagnostic instruments may lead to different prevalence rates of comorbidity depending on which tests were used.

In 2020 seven neuromuscular centres in six European countries (UK, Denmark, France Italy, Netherlands, Spain) started a multicentre study on Brain INvolvement in Dystrophinopathies (BIND, Horizon 2020 (847826), https://bindproject.eu/). The aim of this project is to improve understanding and measurement of dystrophin in the brain and working towards better treatment, care, and outcomes for all those living with Duchenne and Becker muscular dystrophies. As part of this study, we describe which tests are used in clinical practice for boys and men with DMD for three domains of functioning [1]: intelligence/neurocognitive functioning [2], behavioural/psychiatric functioning, and [3] learning disabilities over three age groups [1] preschool i.e. 3-5 years [2], school age: 6–18 years and [3] adulthood i.e. 18 years and older. Our aim is to describe a baseline of current practice with respect to psychological assessment and diagnoses being made in current clinical practice in 7 centres located in 6 European countries.

2. Method

2.1. Participants

This study is part of an ongoing European project, Brain INvolvement in Dystrophinopathies (BIND). Psychologists from seven neuromuscular centres participated: University College London (UCL), University of Newcastle Upon Tyne (UNEW), Kempenhaeghe Centre for Neurological Learning Disabilities (KEM), Universidad Complutense de Madrid (UCM), Department of Neurology, Rigshospitalet, Copenhagen (RegionH), Imagine Institute des maladies genetiques Necker Enfant maladies foundation Paris (NEM), Universita Cattolica del Sacro Cuore, Rome (UCSC). In two of the seven centres, patients are not (routinely) assessed by a psychologist or psychiatrist. When there are any concerns regarding the cognitive, behavioural or emotional functioning of a child, he or she is referred to another health service. These centres (UCL and RegionH) were excluded from the analysis. On a yearly basis, approximately 100 males with dystrophinopathies are seen at UCSC, 20 at UCM, 20 at KEM, 10 at UNEW, and 10 at NEM for neurocognitive and behavioural assessments. Patients are referred for clinical care by health care specialists (e.g. neurologist, paediatrician, psychiatrist)), research activities (e.g. Duchenne Parent Project – Spain), and longitudinal monitoring (e.g. at UCSC).

2.2. Measure

An open-ended questionnaire was sent via e-mail to psychologists of the participating centres asking for [1] how often tests are being used (per age group and per domain (i.e. intelligence/cognition, behaviour and academics) rated on a four point scale: always, often, sometimes, never [2], which tests are being used per domain and per age group (an open ended question), and [3] which diagnoses are being made on the basis of these instruments mentioned (per age group and per domain).

2.3. Statistical analysis

Data analysis was performed using Microsoft Excel (2019). Initially, frequencies were calculated (see Tables 1–4). The open-ended questions were analysed by manually coding and transcribing, and describing test-name and psychiatric/neurodevelopmental diagnosis.

3. Results

With respect to how often testing was done on a regular basis, data on the combined five centres are described in Table 1. Boys in the age group 6 to 18 are most frequently assessed. For the age group over 18 years one of the five centres never assesses adults.

The number of different tests used in five centres is presented in Table 2. The names of the different tests are listed in addendum 1. As tests can be used in different age-groups and different domains, the number of 164 tests in Table 2 does not correspond with the number of 89 tests in the addendum. It can be seen that there is a vast majority of tests being used for the three domains of functioning and the different age groups. Especially for the domain learning and academics language specific instruments are being used.

For the age group 3–5 years of age on the cognitive domain the WPPSI (Wechsler Preschool and Primary Scale of Intelligence) is used most often: in the five centres). For the behavioural domain the CBCL (see addendum 1 for list of abbreviations) is used in three of the five centres. All other 32 test are used in only one of the centres and represent very different instrument e.g. Merrill Palmer Revised, Bells test, Griffith, and Leiter (cognition), ADOS, Conners-3. PARS-III, Vineland-II (behaviour) and Illinois test of psycholinguistic abilities, Prolexia (learning).

For the school aged boys [6–18] there is slightly more heterogeneity in the same tests being used at different centres. Of the 79 instruments being used in this age group (Table 2). Table 3 describes the tests being used in two or more centres. Also aim of the test, age ranges, and the normative samples available and/or used are described.

We also explored the diagnostic categories that are being made in the five centres on the basis of the instruments used via an open-ended question. In one of the five centres adults (older than 18) are referred to regular educational or health care services for diagnoses. The diagnosis of intellectual disabilities is made most often (in all of the centres). In Table 4 an overview of the most frequently made diagnoses is given for all three age groups.

4. Discussion

We found a tremendous heterogeneity in the assessments used: e.g. 79 tests are used for psychological and cognitive assessment of school aged DMD boys in five centres. The most frequently used instruments were Wechsler intelligence Scales, Tower of London, Conners behaviour rating and Child behaviour checklist (CBCL). This wide variety of instruments is probably related to a combination of the increased attention that has been devoted to cognitive abilities in DMD, and to school concerns, as also demonstrated by the fact that cognitive abilities are most often assessed during school age. The assessment of cognitive abilities was often limited to general scales such as the Wechsler scale. More specific aspects of cognition, such as testing of language, which is

Table 1
Assessment frequency (1 = not at all; 2 = sometimes; 3 = often; 4 = always) on regular base in three age groups and domains of functioning.

	3–5 years (n = 5)			6–18 years (n = 5)			>18 (n = 5)		
	Mean	Median	Range	Mean	Median	Range	Mean	Median	Range
IQ and cognition	3,6	4	2–4	3,8	4	3–4	2,6	3	1–4
Behaviour/psychatric	2,8	3	1–4	3,6	4	3–4	2,8	4	1–4
Learning/academics	1,8	1	1–3	3,4	3	3–4	2,2	1	1–4

Abbreviations used: n = number.

Table 2
Number of tests being used over the three age groups and domains of functioning.

	Age 3-5	Age 6-18	Age >18
IQ and cognition	12	38	25
Behaviour/psychiatric	13	26	16
Learning/academics	9	15	10
Total	34	79	51

an important function in DMD, were only routinely performed in two centres.

Other aspects of neurobehavioral and neuropsychological functioning were often assessed with common tests, such as the CBCL, which provides information on a broad range of behavioural and emotional problems in children. However, the CBCL has recently been found to be not entirely suitable for DMD boys as some of the items are less applicable to them due to progressive muscle wasting [20].

When trying to identify the frequency of diagnoses made per age group, we observe that neurodevelopmental diagnoses (e.g. ADHD and ASD) are made more frequent in younger children with DMD, school aged boys are more often diagnosed with cognitive deficits or learning problems, and (young) adults more often with anxiety and depression. Different rates in diagnoses per age group may reflect a cumulative list of clinical symptoms when growing older and the possibility of already being diagnosed with a neurodevelopmental disorder, or reflect a change in psychological functioning (e.g. more anxiety and depression) during transition in adulthood. A longitudinal study design is needed to further investigate the relation between age and brain related comorbidities.

Different review studies [9,16,20] confirm our findings in seven EU neuromuscular centres showing an abundance of different tests being used for neuropsychological problems, resulting in different prevalence rates of brain related comorbidities in boys and young men with DMD.

The current analysis was extremely useful to understand the complexity of the topic. Despite the limitation of a relatively small number of centres participating to the survey, that may not reflect what is used in other centres or countries, the data collected are representative of expert centres for neuromuscular disorders who were selected because of proven experience in testing DMD boys and men.

The heterogeneity of tools used, highlights on one hand the complexity of brain related comorbidities in dystrophinopathies [18] and on the other hand the need to identify a kit of tools that would allow a comparison of the results. Clinical features of CNS involvement in DMD are complex and there is no “one size fits all” assessment procedure for the psychological functioning. The impact of brain related comorbidities on quality of life for both individuals with DMD and their carers, is becoming increasingly evident. Combined also with the fact that life expectancy is prolonged due to better anticipatory care of the musculoskeletal, cardiac and respiratory complications, makes the development of a Standard Operating Procedure (SOP) for assessment of psychological functioning in DMD imperative.

Work is in progress to finalize a tool kit to be used in the BIND study to uniform the procedures and to identify a core set of tools to be used in a clinical setting for clinical purposes, and other tests that may be more suitable for research settings to better understand the complexity of CNS involvement and the correlation among different cognitive and behavioural variables. The BIND test battery includes among others the Wechsler Intelligence Scales and tools that assess: (working) memory,

Table 3
Tests used in two or more centres.

Name of the test	n	Aim of the test	Age range	Norms	
Wechsler Intelligence Scale for Children	5	Intelligence	6–17	Yes	DK, FR, IT, NL, UK
Wechsler Preschool and Primary Scale of Intelligence	5	Intelligence	2,5-7	Yes	DK, FR, IT, NL, UK
Wechsler Adult Intelligence Scale	4	Intelligence	16–85	Yes	DK, FR, IT, NL, UK
D-KEFS Tower test	4	Executive functioning	8–89	Yes	US
Conners-3	4	ADHD	6–18	Yes	US
Child behaviour Checklist	4	Behaviour and emotion	1,5-5, 6-18	Yes	US
Modified Wisconsin Card Sorting Test	3	Executive functioning	18–90	Yes	US
Rey complex figure test	3	Visuo-construction, visual memory	18–90	Yes	US
D-KEFS Trail Making Test	3	Executive functioning	8–89	Yes	US
Behaviour Rating Inventory of Executive function	3	Executive functioning	5–18	Yes	DK, FR, NL, UK
D2 Test of Attention	2	Visual attention	9–80	Yes	EU, UK
Language fluency task	2	Language fluency	*	*	
D-KEFS Colour Word Interference test	2	Executive functioning	8–89	Yes	US
NEPSY-II	2	Cognitive functioning	3–16	Yes	NL, UK, US
Children Memory Scale	2	Memory functioning	5-8, 9-16	Yes	UK
Raven’s Progressive Matrices	2	Nonverbal intelligence	4–70	Yes	FR, NL, SC, SP, UK
State-Trait Anxiety Inventory	2	Anxiety	12–99	Yes	NL, UK
Social and Communication Disorders Checklist	2	Autism	3–19	Yes	US
Strengths and Difficulties Questionnaire	2	Mental health	2–18	Yes	DK, IT, UK, US
SCL-90 Symptom Checklist	2	Mental health	12+	Yes	US
Paediatric Quality of Life Inventory	2	Quality of Life	2–18	Yes	US
Rapid arithmetic	2	Arithmetic	*	*	
Rapid reading	2	Word reading	*	*	

Note. * = country specific.
Abbreviations used: ADHD = Attention Deficit Hyperactivity Disorder; DK = Denmark; EU=European Union; FR= France; IT= Italy; n = number of centres; NL = the Netherlands; SC; Scandinavia SP= Spain; UK= United Kingdom, US= United States.

Table 4
Diagnoses being made in different centres at three age groups.

Diagnoses	Age 3-5	Age 6-18	Age >18
Intellectual Disability	5	5	4
Autism Spectrum Disorders	4	2	–
ADHD/risk of ADHD	4	3	–
Learning disorder/risk of learning disorder	4	5	2
Language disorder	3	–	–
Behavioural disorder	2	–	–
Emotional disorder	3	–	–
Anxiety	–	3	4
Depression	–	3	4
Executive function deficits	–	3	–
Memory deficits	–	2	–
Obsessive compulsive disorder	–	2	2

attention, executive functioning, language, academics, ASD, ADHD, OCD, anxiety and depression, as these domains seem predominantly affected in dystrophinopathies [9,18,21]. Collecting population-specific information on utility, validity, and normative data in dystrophinopathies is ongoing.

Methodological challenges that come with cross-cultural research in psychology need to be considered. When comparing measurement outcomes across multiple cultures it is hard to obtain equivalence, which refers to the comparability level of measurement outcomes [22,23]. To address the issue of languages, and cultural differences, we aim to include instruments that show good psychometric properties of the different translated versions and normcores in multiple countries. Our proposed method in attempting to statistically control for cultural bias includes analysing confounding factors such as years of education or socioeconomic status of study populations [23]. Nonetheless, gathering sufficient normative and generalizable data across multiple populations, languages, and especially minorities remains a challenge.

With this EU multi-centre study we hope to set a first step in standardizing assessment procedures to identify comorbidities, in a way that advances research and collaborations, and standards of care in dystrophinopathies.

Conflict of interest

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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Appendix 1. List of instruments

Domain	Name of the test	Aim of the test	Age range
Cognition and intelligence			
	1. Wechsler Intelligence Scales	Intelligence	3-6, 6–17, 17-89
	2. Leiter International Performance Scale (Leiter-3)	Intelligence	2–18
	3. Griffiths Scales of Child Development (Griffiths-III)	Intelligence	Birth-6
	4. Kaufman Assessment Battery for children (KABC-II)	Intelligence	3–22
	5. Nouvelle Echelle Metrique de l'Intelligence (NEMI-2)	Intelligence	4.5–12.5
	6. Raven's Progressive Matrices (Raven's 2)	Nonverbal intelligence	4–70
	7. Wechsler Non Verbal Scale of Ability	Nonverbal intelligence	4–21
	8. Bayley Scales of Infant Development (Bayley-III)	Intelligence, development delays	16 days-42 months
	9. Merrill Palmer Revised (M-P-R)	Developmental delays and learning	Birth-6.5
	10. A developmental NEuroPSYchological Assessment (NEPSY-II)	Cognitive functioning	3–16
	11. Cuestionario de Madurez Neuropsicológica Infantil (CUMANIN)	Developmental delays	3–7
	12. Developmental Profile 3 (DP-3)	Developmental delays	Birth-13
	13. Psychoeducational Profile (PEP-III)	Development in children with autism	2–7.5
	14. Peabody Picture Vocabulary Test (PPVT-III)	Language	2.5–90+
	15. Clinical Evaluation of Language Fundamentals (CELF-5)	Language	5–22
	16. PEPLA psycholinguistics assessment	Language	N.A.
	17. Language fluency tasks	Verbal fluency	N.A.
	18. Rey auditory learning task (RAVLT)	Verbal learning and memory	7–89
	19. California Verbal Learning Test (CVLT-II)	Verbal learning and memory	16–89
	20. Test de Aprendizaje Verbal España-Complutense (TAVEC)	Verbal learning and memory	16+
	21. Children Memory scale (CMS)	Memory and learning	5-8, 9-16
	22. Wechsler Memory scale (WMS-IV)	Memory functioning	16–90
	23. Rey Complex Figure Test (RCFT)	Visuo-construction, visual memory	18–90
	24. Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery VMI)	Visual motor integration	2–100
	25. Bells test	Visual neglect	N.A.
	26. D2 Test of Attention	Visual attention	9–80
	27. Escalas Magallanes de Atención Visual (EMAV)	Visual attention	5-8, 9+
	28. Test of Everydag Attention for Children (TEA-Ch)	Attention	6–16
	29. Continuous performance test (CPT)	Attention	N.A.
	30. Sustained attention task	Attention	N.A.
	31. Atención Global Local (AGL)	Attention	12–18
	32. Test for Attentional Performance (TAP-II)	Attention	18–89
	33. FePsy	Attention	7–70
	34. Paced Auditory Serial Attention Test (PASAT)	Attention	N.A.
	35. Delis-Kaplan Executive Function System (D-KEFS)	Executive functioning	8–89
	36. Five digit task (FDT)	Executive functioning	7+
	37. Behaviour Rating Inventory of Executive Function (BRIEF)	Executive functioning	5–18
	38. Behavioral Assessment of Dysexecutive Syndrome (BADS-C)	Executive functioning	8-16, 16-87
	39. Stroop Color Word Test (SCWT)	Executive functioning	3-75+
	40. Modified Wisconsin Card Sorting Test	Executive functioning	18–90
	41. Tests des Labyrinthes (LABY 5–12)	Executive functioning	5–12

(*continued on next page*)

(*continued*)

Domain	Name of the test	Aim of the test	Age range
	42. Tower of London Test	Executive functioning	6+
	43. Trail Making Test (TMT)	Executive functioning	8–90
	44. Go/no-go task	Response inhibition	N.A.
	45. Perception of faces tests (faces)	Social cognition	N.A.
Emotion and behaviour			
	46. Strengths and Difficulties Questionnaire (SDQ)	Mental health	2–18
	47. SCL-90 Symptom Checklist (SCL-90)	Mental health	12+
	48. Development and Well-Being Assessment (DAWBA)	Psychiatric diagnoses	5–17
	49. Kiddie Schedule for Affective Disorders and Schizophrenia (K-SADS)	Affective disorders	6–18
	50. Minnesota Multiphasic Personality Inventory (MMPI)	Personality and psychopathology	14-18, 18-80
	51. Neo Five factor inventory (NEO-FFI-3)	Personality	12-20, 21+
	52. Big Five Questionnaire for children (BFQ-C)	Personality	9–14
	53. Child behaviour Checklist (CBCL)	Behaviour and emotion	1.5–5, 6-18
	54. Behavior Assessment System for Children (BASC-3)	Behaviour and emotion	2–22
	55. Adaptive Behavior Assessment System (ABAS)	Adaptive skills	Birth-90
	56. Vineland Adaptive Behavior Scales, Second Edition (Vineland-II)	Adaptive behavior	Birth-90
	57. Personal Adjustment and Role Skills Scale (PARS-III)	Psychosocial adjustment	5–17
	58. Paediatric Quality of Life Inventory (PedsQL)	Quality of Life	2–18
	59. State-Trait Anxiety Inventory (STAI)	Anxiety	12–99
	60. Revised Children's Manifest Anxiety Scales (R-CMAS)	Anxiety	14+
	61. Generalized Anxiety Disorder (GAD-7)	Anxiety	13+
	62. Social and Communication Disorders Checklist (SCDC)	Autism	3–19
	63. Social Responsiveness Scale (SRS-2)	Autism	2.5+
	64. Autism Diagnostic Observation Scale (ADOS)	Autism	1+
	65. Autism Diagnostic Interview- Revised (ADI-R)	Autism	2+
	66. Autism Spectrum Quotient (AQ)	Autism	16+
	67. Childhood Asperger Syndrome Test (CAST)	Autism	5–11
	68. Conners-3	ADHD	6–18
	69. Evaluación del trastorno por déficit de atención con hiperactividad	ADHD	6–12
	70. Sistema de Evaluación de Niños y Adolescentes (SENA)	Emotional and behavioural problems	3–18
	71. Multiscore Depression Inventory for Children (MDI-C)	Depression	8–17
	72. Children's Depression Inventory (CDI)	Depression	8–21
	73. Beck Depression inventory (BDI)	Depression	13+
	74. Beck Hopelessness Scale (BHS)	Hopelessness	17–80
	75. Child Revised Impact of Events Scale (CRIES-8)	Trauma	8–18
Academics			
	76. Wechsler Individual Achievement Test (WAIT-III)	Academic abilities	4–51
	77. Batterie Modulable Tests Informatisée (BMT-i)	Learning abilities	6–12
	78. Prueba de Lenguaje Oral Navarra (PLON)	Language development	3–6
	79. Illinois Test Psycholinguistic Abilities (ITPA-3)	Oral and written language	5–12
	80. PROLEXIA (Diagnóstico y Detección Temprana de la Dislexia)	Dyslexia	4+
	81. PREDISCAL (Screening de Dificultades Lectoras y Matemáticas)	Reading and mathematics	7–12
	82. PROLEC (Batería de Evaluación de los Procesos Lectores)	Reading	6–12
	83. Lecture de Mots et Compréhension (LMC-R)	Reading and comprehension	Children
	84. Rapid reading	Word reading	N.A.
	85. Batteria per la Valutazione della Scrittura e della Competenza Ortografica (BVSCO)	Writing and spelling	6–14
	86. Rapid arithmetic	Arithmetic	N.A.
	87. AC-MT (Test per la Valutazione dei Disturbi di Calcolo)	Mathematics	11–14
	88. UDN-II (évaluation dynamique de la pensée logique)	Mathematics and logical thinking	4–16
	89. ZAREKI	Dyscalculia	8–14

Note. Age range is given in years if not specified otherwise. Abbreviation used: N.A. = not applicable.

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