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PSYCHOLOGY OF DUCHENNE MUSCULAR DYSTROPHY

AMSTERDAM SEPT 7TH

Dr. Jos Hendriksen PhD

Clinical psychologist and neuropsychologist

NETHERLANDS.

hendriksenj@kempenhaeghe.nl

Duchenne Parent Project

DUCHENNE CENTRUM NEDERLAND

Maastricht UMC+

NO CONFLICT OF INTEREST

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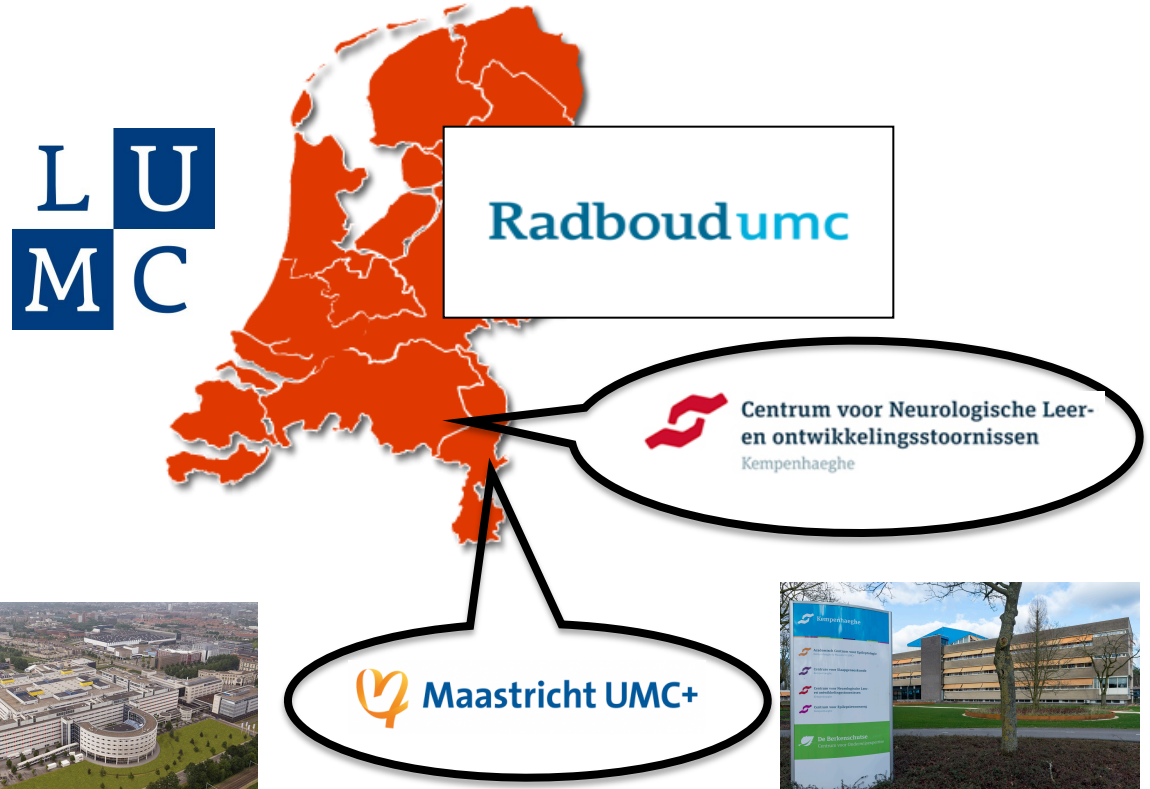
KEMPENHAEGHE CENTER FOR NEUROLOGICAL LEARNING DISABILITIES



Duchenne Centre Netherlands:
Leiden UMC
Radboud UMC
Heeze Kempenhaeghe/
Maastricht UMC

2 research lines with PhD trajects
- Non motor problems
- Diagnosis and treatment of problems in learning and behavior.

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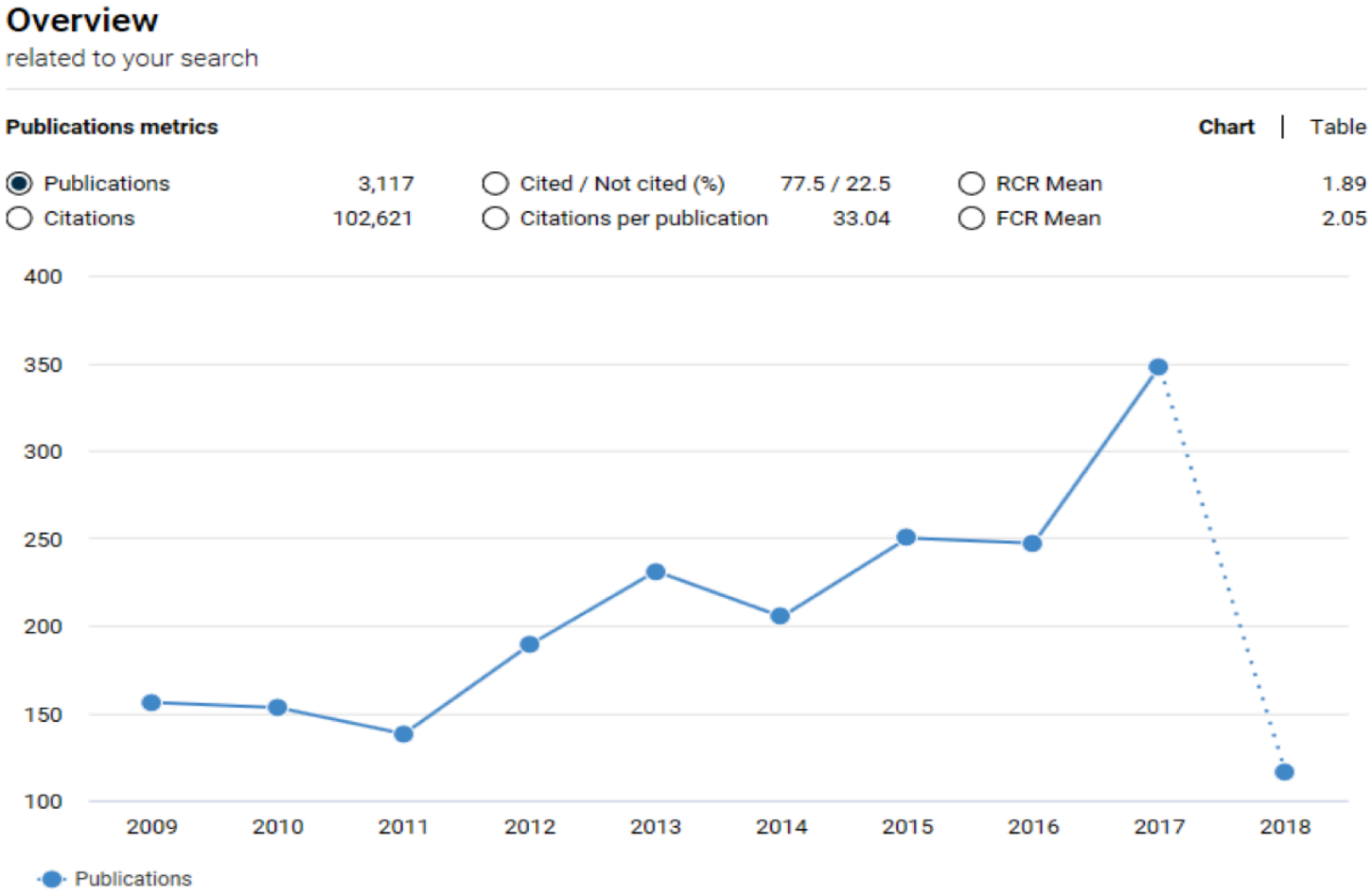
PART 1: INTRODUCTION: LEARNING AND BEHAVIOR IN DMD

LITERATURE SEARCH



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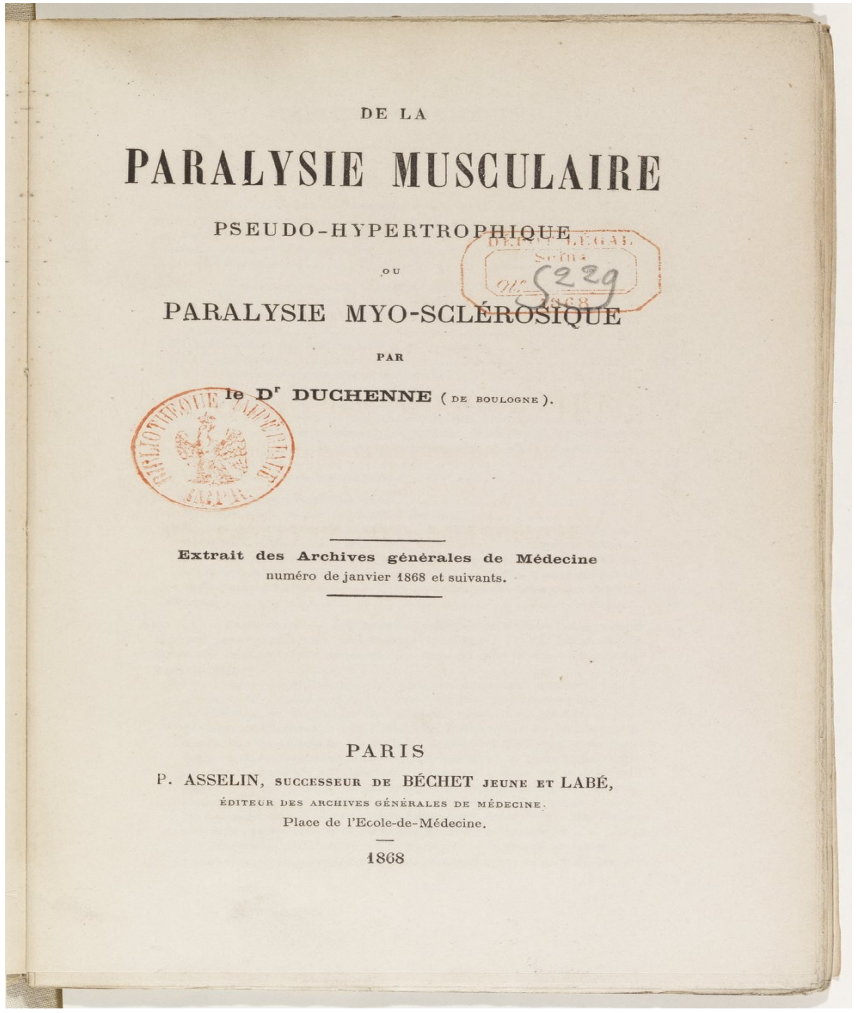
DUCHENNE DE BOULOGNE (1868)

LONG HISTORY



DE LA

PARALYSIE MUSCULAIRE

PSEUDO-HYPERTROPHIQUE

OU

PARALYSIE MYO-SCLÉROSIQUE

PAR

le Dr DUCHENNE (DE BOULOGNE).

Extrait des Archives générales de Médecine
numéro de janvier 1868 et suivants.

PARIS

P. ASSELIN, SUCCESSEUR DE BÉCHET JEUNE ET LABÉ,
ÉDITEUR DES ARCHIVES GÉNÉRALES DE MÉDECINE.
Place de l'Ecole-de-Médecine.

1868

Source gallica.bnf.fr / Bibliothèque nationale de France

13 patients:

6 low IQ
2 language deficit
2 epilepsy

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DYSTROPHIN ASSOCIATED DEVELOPMENTAL DISORDERS (2016)

150 YEARS LATER

DEVELOPMENTAL MEDICINE & CHILD NEUROLOGY ORIGINAL ARTICLE

Neurodevelopmental, emotional, and behavioural problems in Duchenne muscular dystrophy in relation to underlying dystrophin gene mutations

VALERIA RICOTTI[1] | WILLIAM P L MANDY[2] | MARIACRISTINA SCOTO[1] | MARIKA PANE[3] | NICOLAS DECONINCK[4,5] | SONIA MESSINA[6] | EUGENIO MERCURI[1,3] | DAVID H SKUSE[2] | FRANCESCO MUNTONI[1,*]

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150 YEARS LATER

DMCN Developmental Medicine & Child Neurology

Muscle and brain: a dyad with important diagnostic and therapeutic implications

JOS G M HENDRIKSEN | JOHAN S H VLES
Department of Neurological Learning Disabilities, Epilepsy Centre Kempenhaeghe, Heeze, the Netherlands.

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LEIDEN UNIVERSITY MEDICAL CENTER

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BRAIN INVOLVEMENT

ALTERED BRAIN MORPHOLOGY IN DMD BOYS

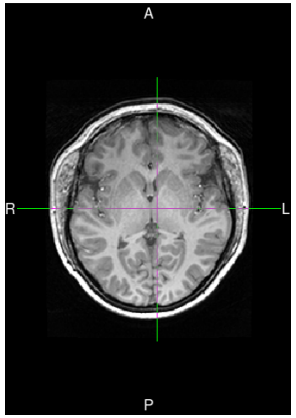
Reduced Cerebral Gray Matter and Altered White Matter in Boys with Duchenne Muscular Dystrophy

Nathalie Doorenweerd, MSc,[1,2,3] Chiara S. Straathof, MD,[3] Eve M. Dumas, PhD,[3] Pietro Spitali, PhD,[4] Ieke B. Ginjaar, PhD,[5] Beatrijs H. Wokke, MD,[3] Debby G. Schrans, MSc,[6] Janneke C. van den Bergen, MD,[3] Erik W. van Zwet, PhD,[7] Andrew Webb, PhD,[1] Mark A. van Buchem, MD, PhD,[1] Jan J. Verschuuren, MD, PhD,[3] Jos G. Hendriksen, PhD,[6,8] Erik H. Niks, MD, PhD,[3] and Hermien E. Kan, PhD[1,2]

Smaller grey matter volume

→ fewer neuronal cell bodies

Similar white matter volume with reduced integrity of the white matter



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PART 2: STANDARDS OF CARE

SOC: LANCET 2018

Psychosocial care:

- "should include surveillance and management of psychosocial aspects of the disease across the lifespan".

- "Should address social and cognitive development as wel as QOL and factors that affect patient and families across all environments (home, school, work)

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SCREENING ON MENTAL HEALTH

Routine mental health screening

- At each neuromuscular clinic visit, mental health and quality of life should be screened
- Screening can be informal and does not require comprehensive assessment
- An appropriate tool for paediatric patients is the Strengths and Difficulties Questionnaire;[26] for adult patients, the Patient Health Questionnaire 9-item depression scale (PHQ-9)[27] and the Generalized Anxiety Disorder 7-item scale (GAD-7)[28] are appropriate; for parents of patients aged 5–17 years, the Personal Adjustment and Role Skills Scale (PARSIII) is suitable[29,30] (scale and scoring programme is available on the Parent Project Muscular Dystrophy website)
- Screening can be conducted by a social worker or mental health professional or by other clinic staff with sufficient training or experience in this area (eg, a nurse or attending physician)
- If screening is positive, a referral should be made to a psychologist and psychiatrist for further assessment or treatment
- Every clinic should have a plan to assess and address suicidal ideation or other acute safety concerns
- Caregiver emotional adjustment should be monitored and intervention or support offered as needed
- Siblings of a person with DMD should be provided with opportunities to connect with other siblings of patients with DMD and with access to mental health services as needed

- SDQ: behavior
- PARS-III: adjustment

ADULT PATIENTS:
- GAD-7: anxiety
- PHQ9: depression

If screening is positive refferal to a psychologist/ psychiatrist should be made

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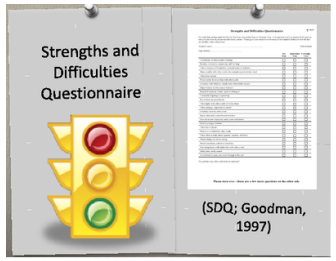
MENTAL HEALTH SCORE

STRENGHTS AND DIFFICULTIES QUESTIONNAIRE (SDQ)



To identify boys with mental health disorders
28 items about boys functioning:
-Emotional symptoms: “many worries”
-Conduct problems: “often has temper tantrums”
-Hyperactive behavior: “restless and overactive”
-Peer problems: “bullied by other children”
-Pro social behavior: “helpful if someone is hurt”

Scales	Normal	Borderline	Abnormal
Total difficulty score	0-15	16-19	20-40
Emotional symptom score	0-5	6	7-10
Conduct problem score	0-3	4	5-10
Hyperactivity score	0-5	6	7-10
Peer problem score	0-3	4-5	6-10
Prosocial behavior score	6-10	5	0-4

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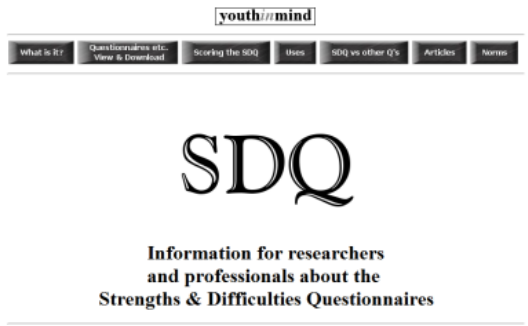
MENTAL HEALTH SCORE

STRENGHTS AND DIFFICULTIES QUESTIONNAIRE (SDQ)

Three versions:
- Parent report 2-17 years.
- Self report version 11-17 years
- Teacher report.



Adequate reliabilty and good predictive validity

85 languages (!) ; normative data in 10 countries

www.sdqinfo.com

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PSYCHOSOCIAL ADJUSTMENT

PARS-III

Original authors: Stein & Jessop (1990)
Adjustment and Coping strategies:
28 items:

1. Peer relations: making friends of his own
2. Dependency: asking for help
3. Hostility: responding to discipline
4. Productivity: keeping up with tasks
5. Anxiety/depression: acting afraid
6. Withdrawal: unaware of things going on

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PSYCHOSOCIAL ADJUSTMENT

PARS-III

N=322 boys and young men with Duchenne

Journal of Pediatric Psychology Advance Access published July 22, 2008

Psychosocial Adjustment in Males with Duchenne Muscular Dystrophy: Psychometric Properties and Clinical Utility of a Parent-report Questionnaire

Jos G. M. Hendriksen,[1,2,3] PhD, James T. Poysky,[4] PhD, Debby G. M. Schrans,[1,2] MSc, Eric G. W. Schouten,[5] PhD, Albert P. Aldenkamp,[1,3] PhD, and Johan S. H. Vles,[3] PhD, MD

[1]Kempenhaeghe Epilepsy Centre, Heeze, [2]Franciscusoord, Childhood Rehabilitation Centre, SRL, Valkenburg, [3]Department of Neurology, University Hospital Maastricht, [4]Baylor College of Medicine, Texas Children's Hospital and [5]Department of Psychology, Maastricht University

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PSYCHOSOCIAL ADJUSTMENT

PARS-III

Age 4-18 years

Good Reliability (alpha .91)
A score below 72 is indicative of higher risk for adjustment problems

Versions in English, Spanish, Turkish, German, Dutch, French, Tsjech, Greek. Hungarian.

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GENERALISED ANXIETY SCALE

GAD-7

GAD-7

Over the last 2 weeks, how often have you been bothered by the following problems?	Not at all	Several days	More than half the days	Nearly every day
1. Feeling nervous, anxious or on edge	0	1	2	3
2. Not being able to stop or control worrying	0	1	2	3
3. Worrying too much about different things	0	1	2	3
4. Trouble relaxing	0	1	2	3
5. Being so restless that it is hard to sit still	0	1	2	3
6. Becoming easily annoyed or irritable	0	1	2	3
7. Feeling afraid as if something awful might happen	0	1	2	3

Total Score ___ = Add Columns ___ + ___ + ___

If you checked off any problems, how difficult have these problems made it for you to do your work, take care of things at home, or get along with other people?

Not difficult at all	Somewhat difficult	Very difficult	Extremely difficult
☐	☐	☐	☐

Self report

Adult population

Reliable and valid

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DEPRESSIVE COMPLAINTS

PHQ-9

PATIENT HEALTH QUESTIONNAIRE-9

Over the last 2 weeks, how often have you been bothered by any of the following problems?	Not at all	Several days	More than half the days	Nearly every day
1. Little interest or pleasure in doing things	0	1	2	3
2. Feeling down, depressed, or hopeless	0	1	2	3
3. Trouble falling or staying asleep, or sleeping too much	0	1	2	3
4. Feeling tired or having little energy	0	1	2	3
5. Poor appetite or overeating	0	1	2	3
6. Feeling bad about yourself — or that you are a failure or have let yourself or your family down	0	1	2	3
7. Trouble concentrating on things, such as reading the newspaper or watching television	0	1	2	3
8. Moving or speaking so slowly that other people could have noticed? Or the opposite — being so fidgety or restless that you have been moving around a lot more than usual	0	1	2	3
9. Thoughts that you would be better off dead or of hurting yourself in some way	0	1	2	3

FOR OFFICE CODING

0 + _____ + _____ + _____

=Total Score: _____

If you checked off any problems, how difficult have these problems made it for you to do your work, take care of things at home, or get along with other people?

Not difficult at all	Somewhat difficult	Very difficult	Extremely difficult
☐	☐	☐	☐

Self report

Adult population

Reliable and valid

PM: item 9 = suicidal ideation

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LANCET 2018

- The neuromuscular care team should include a mental health professional (ie, psychologist or psychiatrist) with training and experience in assessing and treating psychiatric conditions in the context of chronic medical or neurodevelopmental conditions
- When mental health concerns are identified, the mental health professional should provide further evaluation of individuals with DMD and their family members, and provide cognitive or behavioural interventions to treat psychiatric conditions
- Standard, evidence-based practices should be used for those who need more formal mental health treatment
- Neuropsychological evaluations should be done when cognitive delays, difficulties with emotional and behavioural regulation, or concerns about social skills exist; re-evaluations should be done every 2–3 years to monitor developmental progress and response to interventions
- Neuropsychological evaluations should be considered within the first year of diagnosis to establish a baseline, or when transitioning to adulthood if government-based assistance might be necessary to promote functional independence

PSYCHOLOGICAL CARE:

- Evaluation of patients at risk;
- Neuropsychological evaluation if cognitive delay or behavior problems. Reevaluation within 2-3 years
- Baseline neuropsychological testing within first year of diagnosis and in transition to adulthood

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CARE CONSIDERATIONS BY STAGE OF DISEASE

LANCET 2018

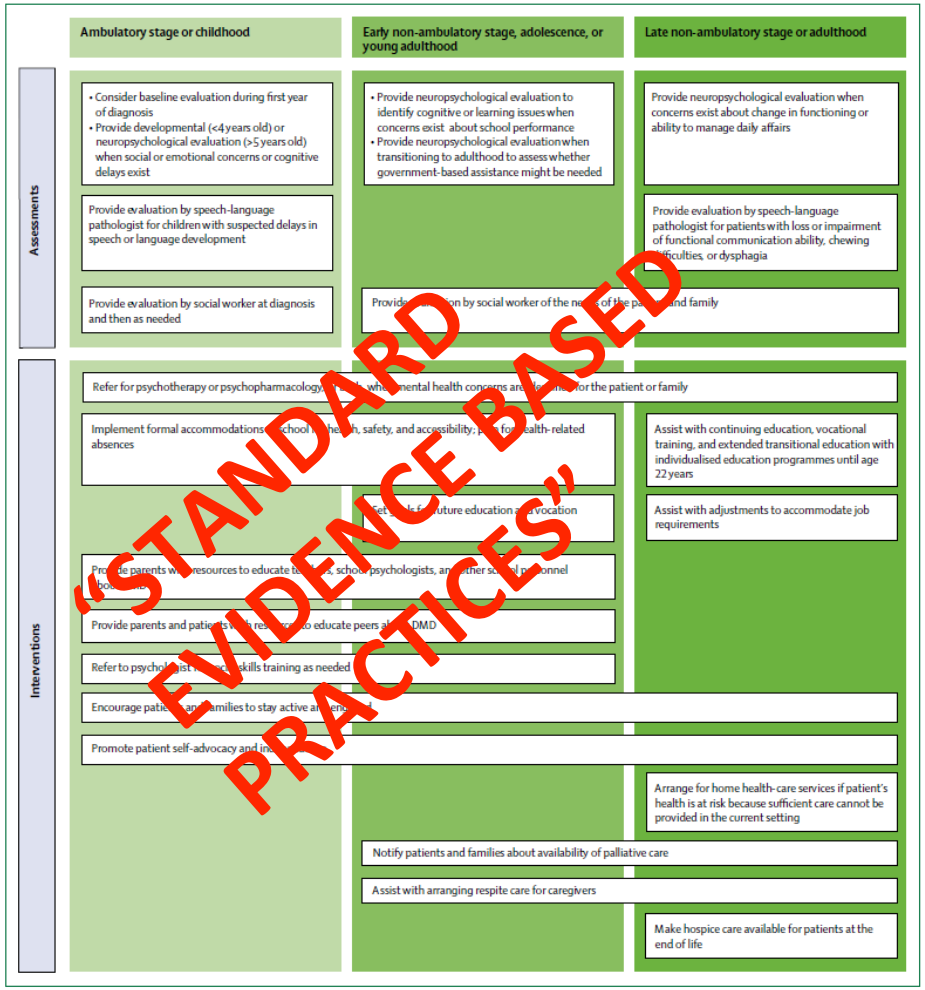

Figure 1: **Considerations for psychosocial care of individuals with Duchenne muscular dystrophy**
DMD=Duchenne muscular dystrophy.

"STANDARD EVIDENCE BASED PRACTICES"

Developmental and neuropsychological evaluation

Speech and language

Social work

Psychofarmaca and psychotherapy

Accomodations at school

Parent resources

Social skills

Self advocacy and independence

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LANCET 2018

Pharmacological interventions

- The neuromuscular team should include a psychiatrist or other physician with training and experience in providing medication to treat behavioural or emotional disorders in the context of chronic medical or neurodevelopmental conditions
- Standard prescribing practices should be followed
- Selective serotonin-reuptake inhibitors should be prescribed for depression, anxiety, and obsessive–compulsive disorder
- α-Adrenoceptor agonists (first choice) or atypical antipsychotics (second choice) should be prescribed for aggression and anger or emotional dysregulation
- Stimulants or α-adrenoceptor agonists should be prescribed for attention-deficit hyperactivity disorder

Medication to treat behavior or emotional disorders:
-Standard procedures
-SSRI's for depression, anxiety, OCD
-Alfa adenoreceptor agonists for ODD
-Stimulantia for ADHD

PART 3: EMPIRICAL EVIDENCE AND CLINICAL PRACTICES

CURRENT KNOWLEGDE

Evidence based knowledge is scarce and clinical practice is often based on expert opinion

LOW AVERAGE GENERAL IQ USING WECHSLER SCALE

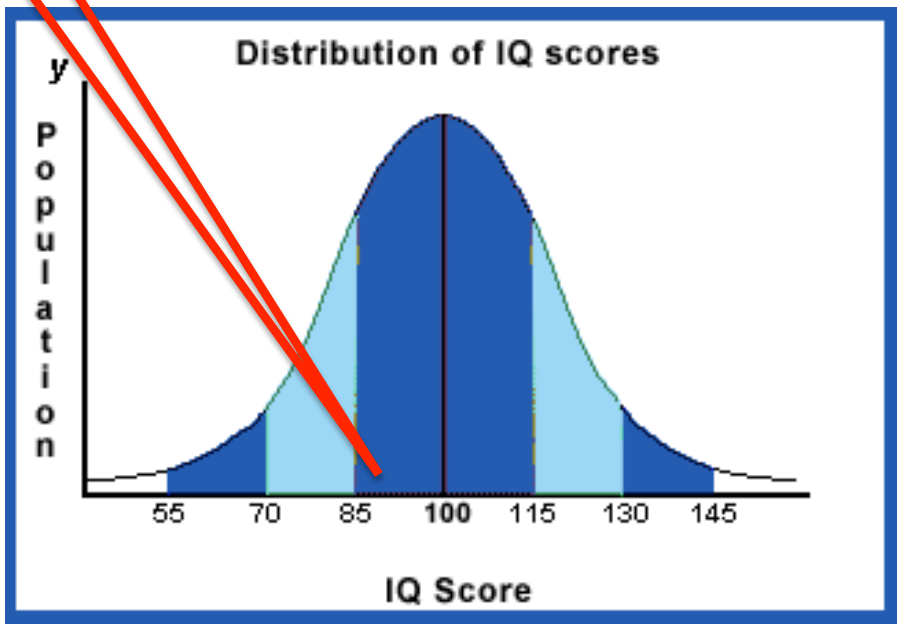
INTELLIGENCE

Performance IQ. **93**

Verbal IQ **88**

SCHRANS & HENDRIKSEN REVIEW (2012)

77 STUDIES (1954 – 2012);
N= 377 BOYS



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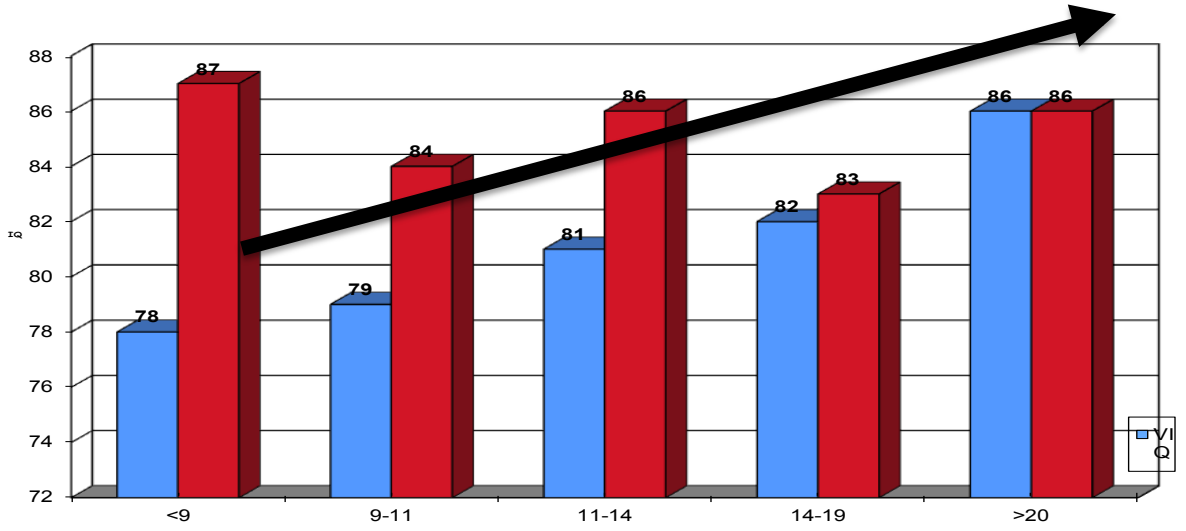
VIQ VERSUS PIQ



-No cognitive decline;

-Growing out of a language deficit

Association between intellectual functioning and age in children and young adults with Duchenne muscular dystrophy: further results from a meta-analysis

Sue M Cotton* BBSc GradDipAppSc MAppSc, Orygen Youth Health, Department of Psychiatry, University of Melbourne;
Nicholas J Voudouris PhD;
Kenneth M Greenwood PhD, School of Psychological Science, La Trobe University, Victoria, Australia.

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NEUROCOGNITION

GENERAL INTELLIGENCE RECONSIDERED:

No prospective follow up data available

General intelligence may be an underestimation of their potentials

There is more than a global deficit

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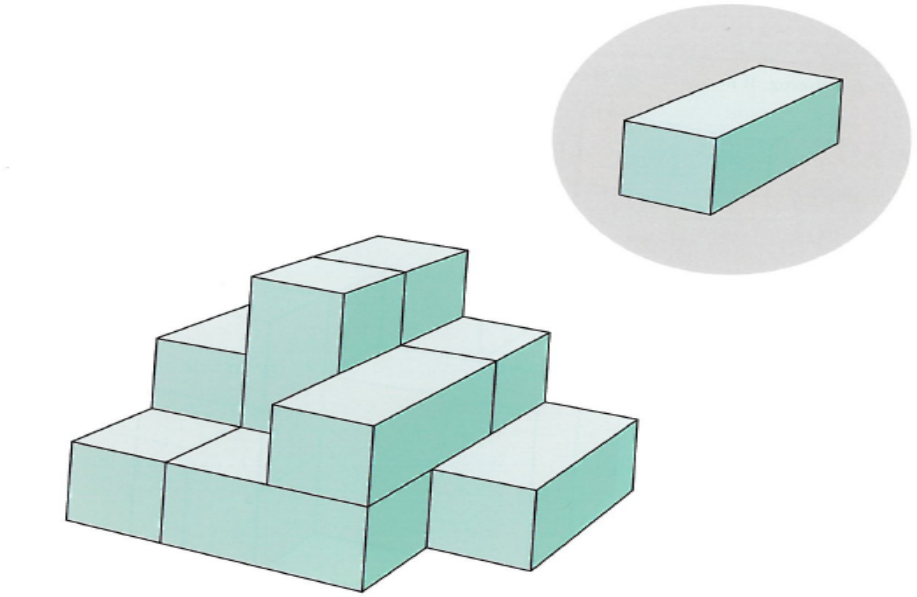
PERFORMANCE IQ: SPATIAL THINKING

STRENGHT



HOW MANY BLOCKS FIT IN THE BLOCK DESIGN?

	DMD
SPATIAL THINKING (Block counting)	107

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GROWING INTEREST

The relationship between deficit in digit span and genotype in nonsense mutation Duchenne muscular dystrophy

Mathula Thangarajh, MD, PhD, Gary L. Elfring, PhD, Panayiota Trifillis, PhD, Joseph McIntosh, MD, and Stuart W. Peltz, MD, on behalf of the Ataluren Phase 2b Study Group

Neurology® 2018;00:1-5. doi:10.1212/WNL.0000000000006245

Correspondence
Dr. Thangarajh
mthangar@
childrensnational.org

WEAKNESS

Cognitive profile in Duchenne muscular dystrophy boys without intellectual disability: The role of executive functions

R. Battini [a,b,1], D. Chieffo [a,1], S. Bulgheroni [c], G. Piccini [d], C. Pecini [b], S. Lucibello [a], S. Lenzi [b], F. Moriconi [a], M. Pane [a], G. Astrea [b], G. Baranello [c], P. Alfieri [d], S. Vicari [d], D. Riva [c], G. Cioni [b,e], E. Mercuri [a,*]

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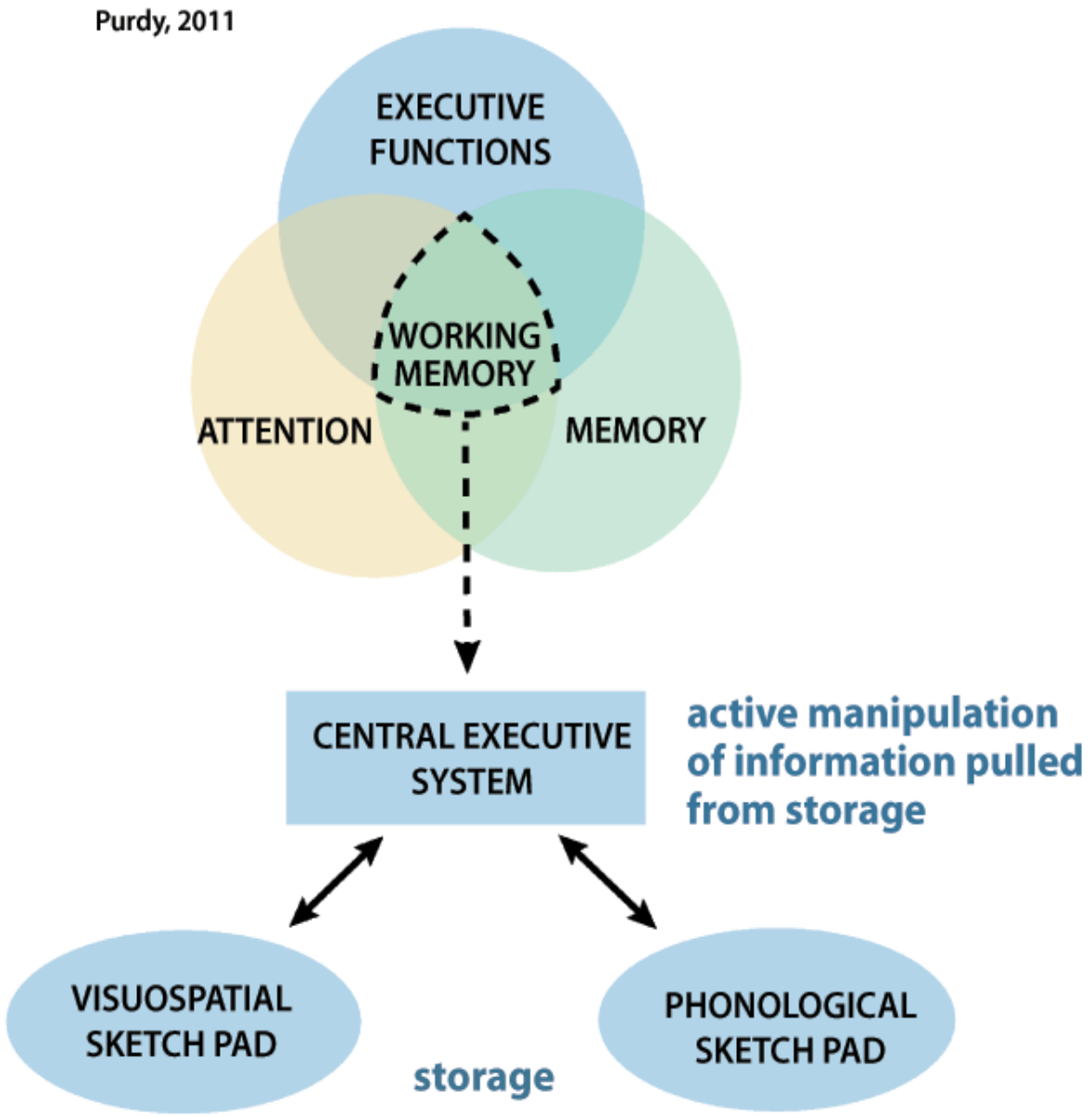
CONCEPTUAL FRAMEWORK



Relationship between attention working memory and executive functions

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WORKING MEMORY

BOTTLE NECK THEORY

Transfer information from short term memory to long term memory

Digit span task:
2-3
4-8
5-6-1
7-9-2
……

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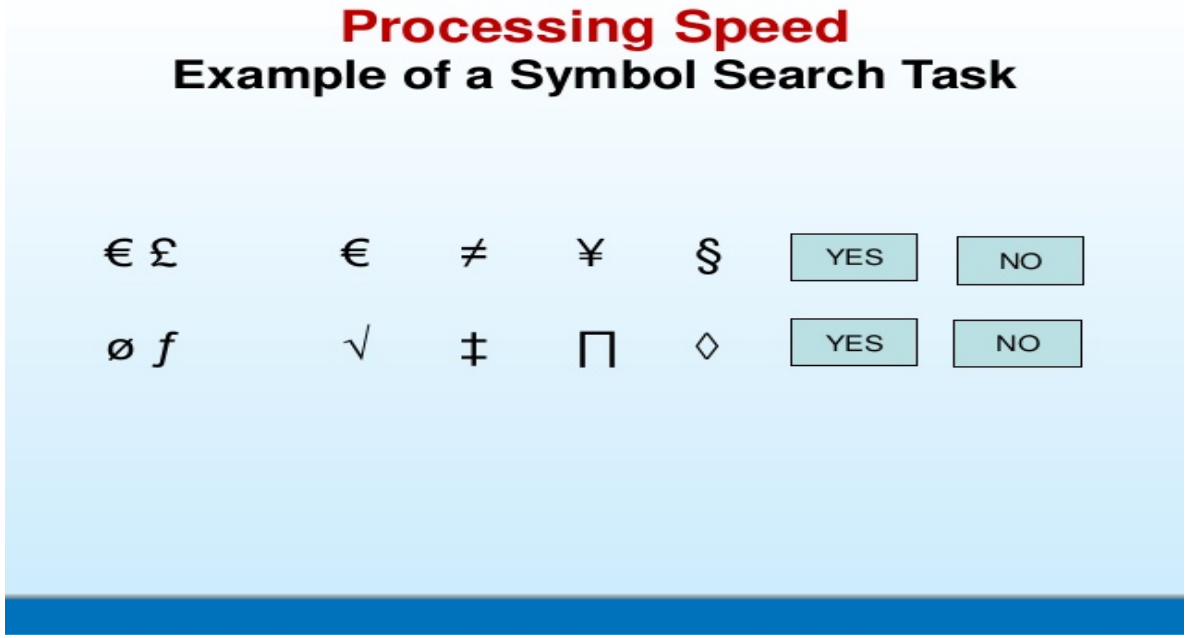
SPEED OF INFORMATION PROCESSING

ATTENTTION



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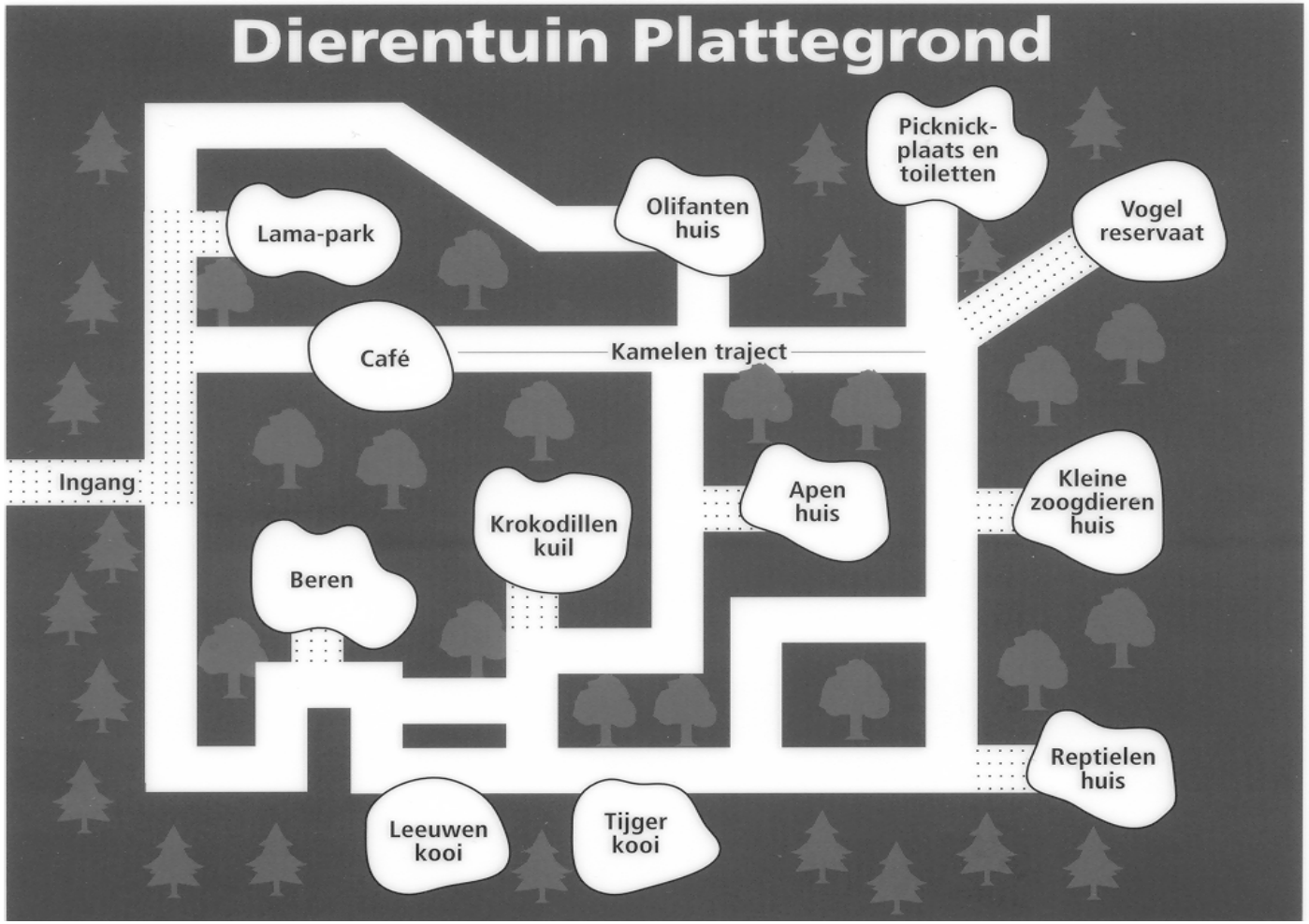
EXECUTIVE FUNCTIONS

FIND YOUR WAY IN THE ZOO:

GO FROM THE ENTRANCE TO THE PICKNICK AND IN THE MEANWHILE VISIT THE APES AND THE LAMAS



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WORKING MEMORY

WEAKNESS

Several studies in DMD:
Verbal working memory problems (i.e. limited verbal span in recalling digits, poor story recall and sentence repetition), are described as core neurocognitive deficits.

	DMD
SPATIAL THINKING (Block counting)	107
AUDITORY WORKING MEMORY (Wechsler number recall)	85

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AUTOMATISATION

Learning to read and learning artihmetics has to do with automatisation

LEARNING

جمعية النجاة الخيرية

صندوق إعانة المرضى

الرابطة الكويتية للدسلكسيا

(مركز محمد الخرافي لعسر القراءة)

QUICK AND AUTOMATIC DECODING OF SIGNS INTO SOUNDS

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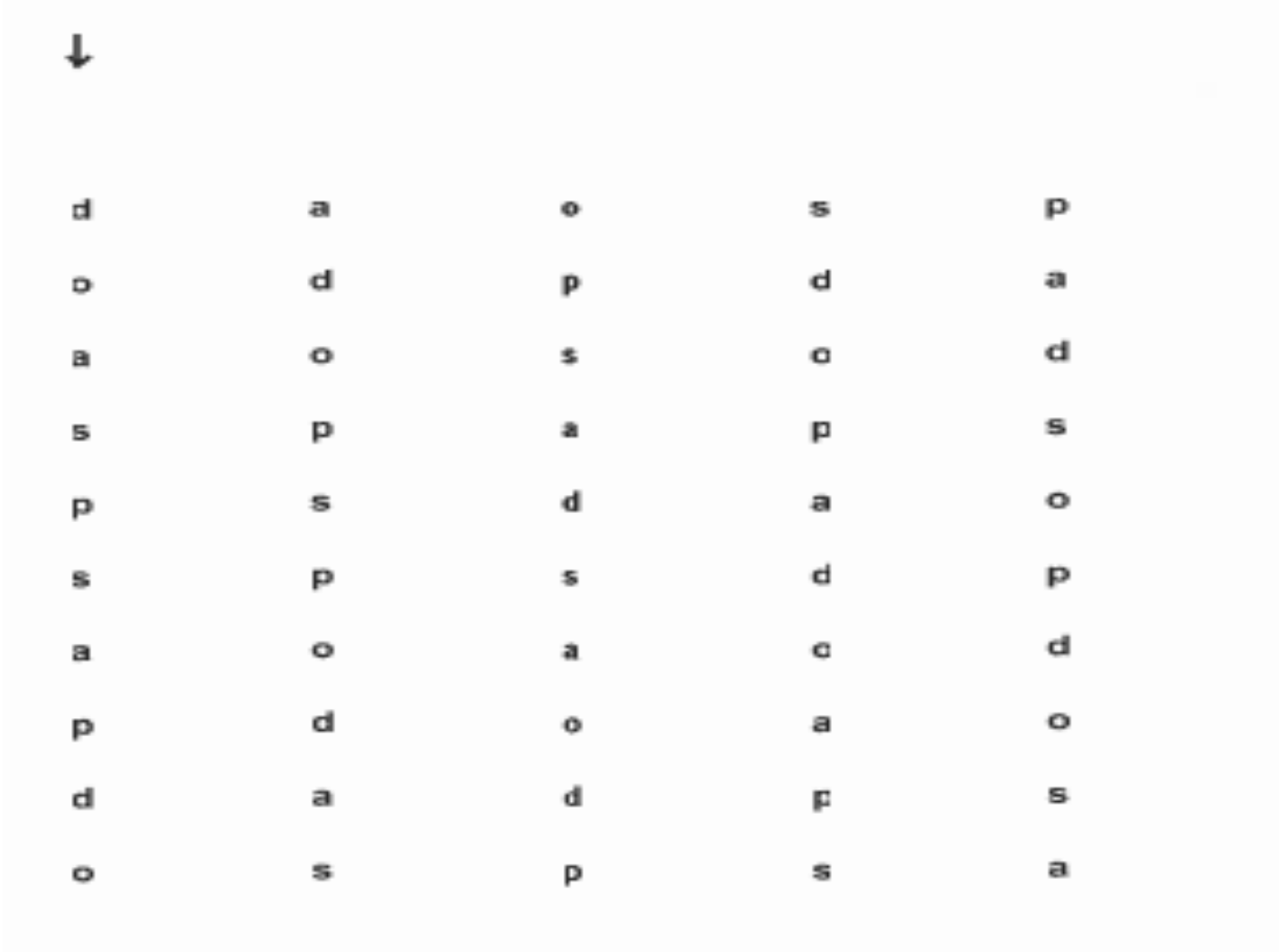
QUICKY NAME THESE LETTERS:

AUTOMATISATION



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DUCHENNE & DYSLEXIA: 27 -42%

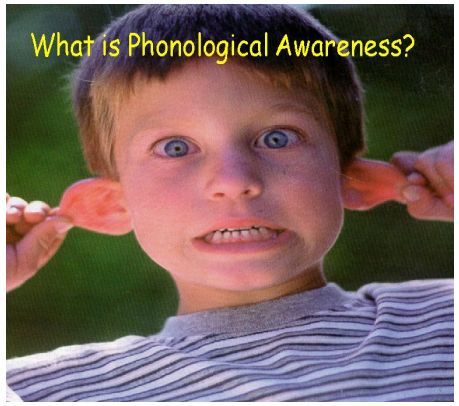
Population mean : 3%

AUTOMATISATION

ELSEVIER

Are Males With Duchenne Muscular Dystrophy at Risk for Reading Disabilities?

Jos G.M. Hendriksen, PhD*† and Johan S.H. Vles, PhD, MD‡



Contents lists available at ScienceDirect

Research in Developmental Disabilities

ELSEVIER

Reading impairment in Duchenne muscular dystrophy: A pilot study to investigate similarities and differences with developmental dyslexia

Guja Astrea [a,*], Chiara Pecini [a], Filippo Gasperini [a], Giacomo Brisca [b], Marianna Scutifero [c], Claudio Bruno [b], Filippo Maria Santorelli [a], Giovanni Cioni [a,d], Luisa Politano [c], Anna Maria Chilosi [a], Roberta Battini [a]

[a] Department of Developmental Neuroscience, IRCCS Fondazione Stella Maris, Viale del Tirreno 331, 56128 Calambrone, Pisa, Italy
[b] Center of Myology and Neurodegenerative Disorders, Department of Neuroscience, IRCCS G. Gaslini Institute, Via Gerolamo Gaslini, 5, 16148 Genoa, Italy
[c] Department of Experimental Medicine, Cardiomyology and Medical Genetics, Second University of Naples, Piazza Miraglia, 80131 Naples, Italy
[d] Department of Clinical and Experimental Medicine, University of Pisa, Italy

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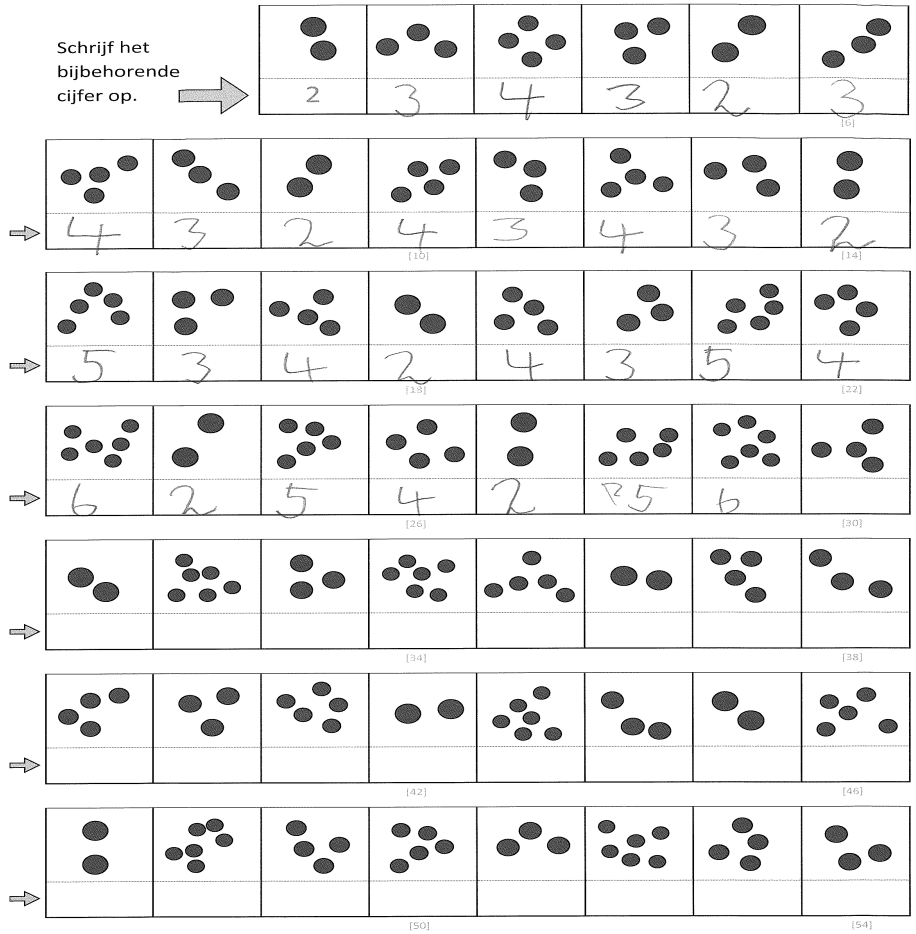
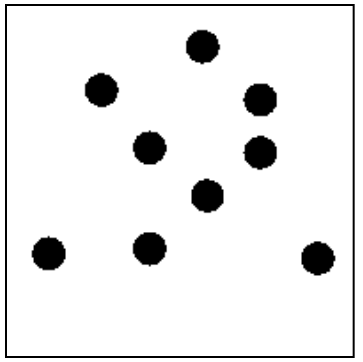
QUICKY NAME THE NUMBERS:

AUTOMATISATION





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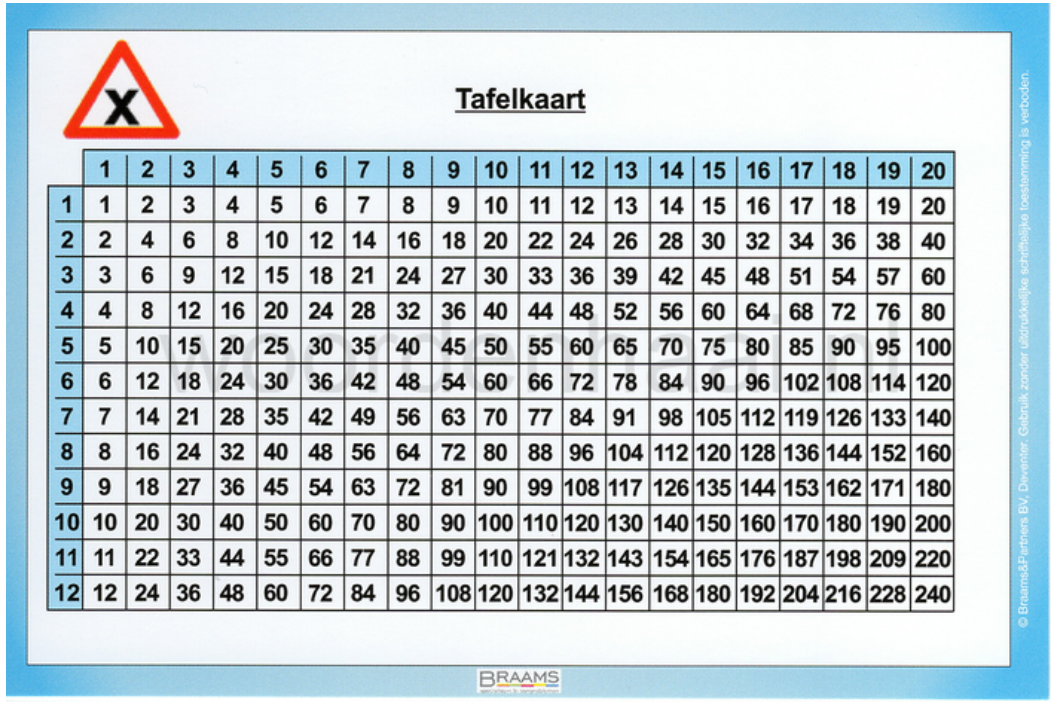
DIFFICULTIES WITH MATHEMATICS

AUTOMATISATION

No data in DMD population



	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20
1	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20
2	2	4	6	8	10	12	14	16	18	20	22	24	26	28	30	32	34	36	38	40
3	3	6	9	12	15	18	21	24	27	30	33	36	39	42	45	48	51	54	57	60
4	4	8	12	16	20	24	28	32	36	40	44	48	52	56	60	64	68	72	76	80
5	5	10	15	20	25	30	35	40	45	50	55	60	65	70	75	80	85	90	95	100
6	6	12	18	24	30	36	42	48	54	60	66	72	78	84	90	96	102	108	114	120
7	7	14	21	28	35	42	49	56	63	70	77	84	91	98	105	112	119	126	133	140
8	8	16	24	32	40	48	56	64	72	80	88	96	104	112	120	128	136	144	152	160
9	9	18	27	36	45	54	63	72	81	90	99	108	117	126	135	144	153	162	171	180
10	10	20	30	40	50	60	70	80	90	100	110	120	130	140	150	160	170	180	190	200
11	11	22	33	44	55	66	77	88	99	110	121	132	143	154	165	176	187	198	209	220
12	12	24	36	48	60	72	84	96	108	120	132	144	156	168	180	192	204	216	228	240

?

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NEUROPSYCHIATRIC DISORDERS IN DMD

BEHAVIOR

- **ADHD:** Attention Deficit Hyperactivity Disorder
- **ASD:** Autism Spectrum Disorder
- **OCD:** Obsessive Compulsive Disorder

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ATTENTION DEFICIT ADHD

INHIBITION IS CORE CONCEPT

WHEN I WANT HIM TO STOP HE GOES ON AND ON, AS IF THERE IS NO BRAKE ON HIS BEHAVIOR

Three core symptoms:

- Inattention
- Impulsivity
- hyperactivity

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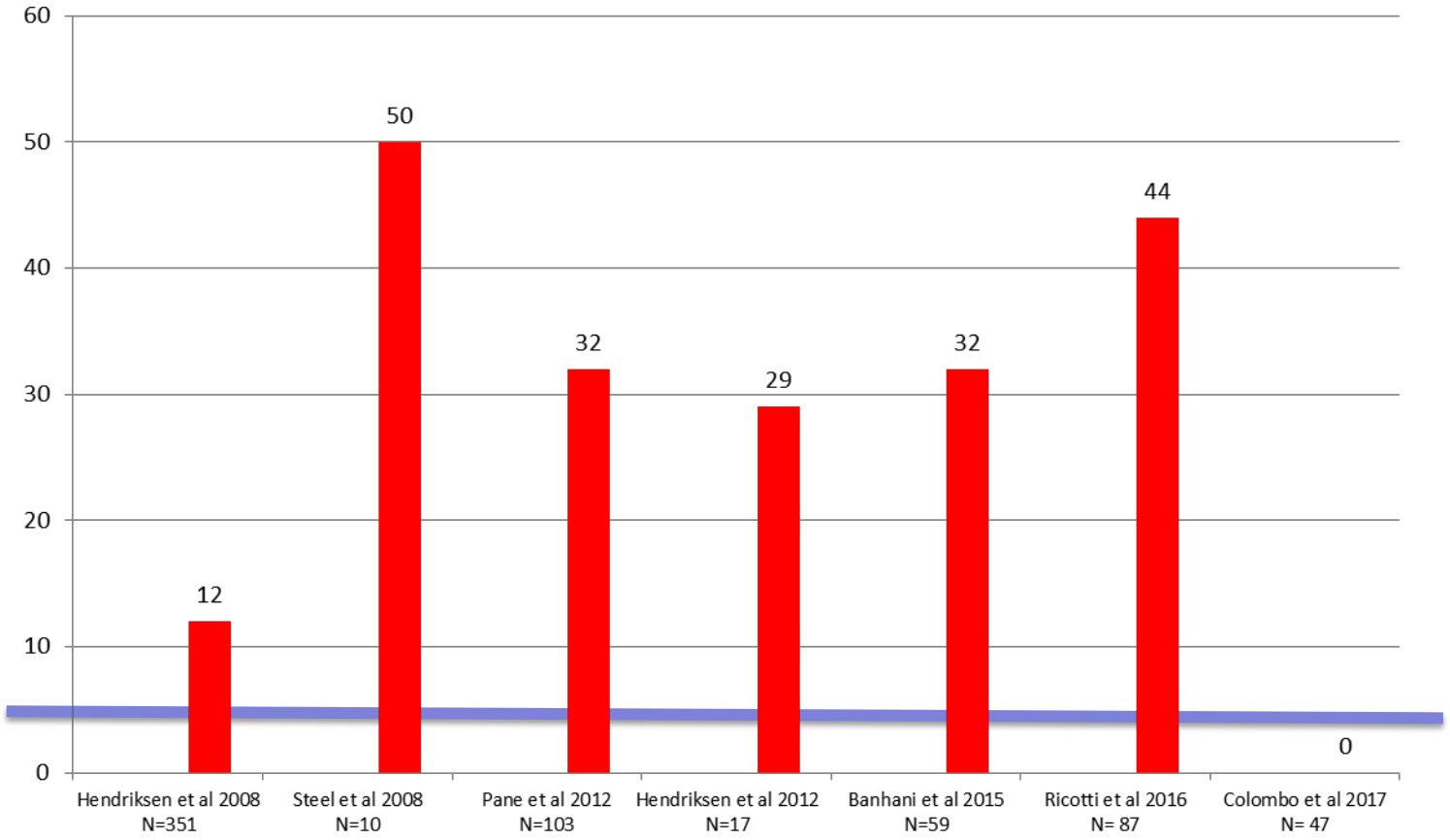
ATTENTION DEFICIT

PREVALENCE OF ADHD IN DMD:



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ATTENTION DEFICIT ADHD

ELSEVIER Official Journal of the European Paediatric Neurology Society

Original article

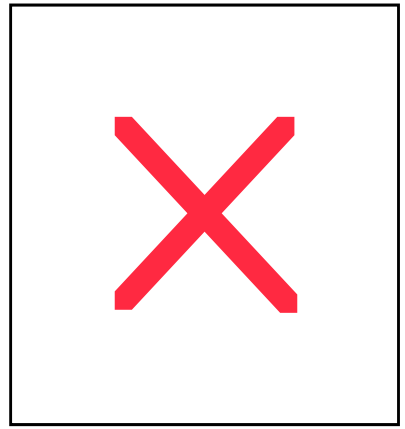
Assessing mental health in boys with Duchenne muscular dystrophy: Emotional, behavioural and neurodevelopmental profile in an Italian clinical sample

Paola Colombo [a], Maria Nobile [a,*], Alessandra Tesei [a], Federica Civati [b], Sandra Gandossini [b], Elisa Ma[illegible] [a], Massim[illegible] Molteni [a], Nereo Bresolin [c], Grazia D'Angelo [b]

RISK OF FALSE NEGATIVES

.. no ADHD comorbidity was found.

.. CBCL, SDQ are not as useful, as children with DMD cannot display the same type of hyperactivity symptoms, such as fidgetiness, due to their motor problems.



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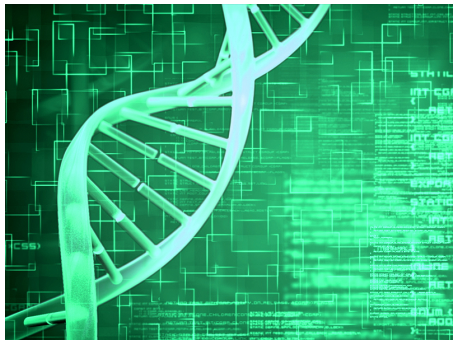
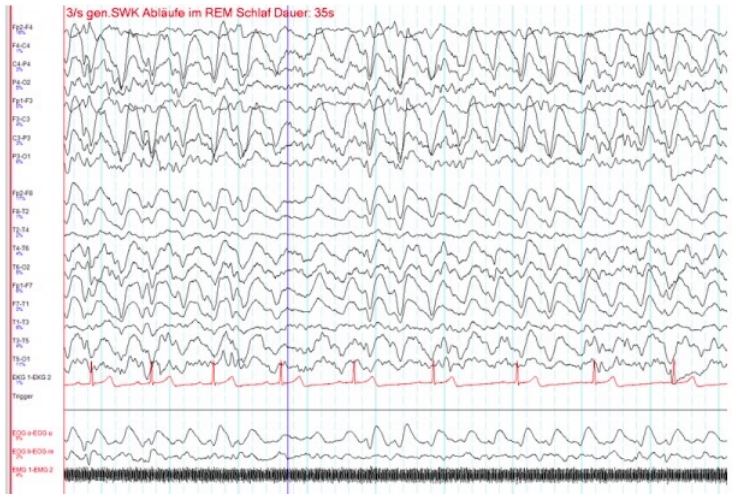
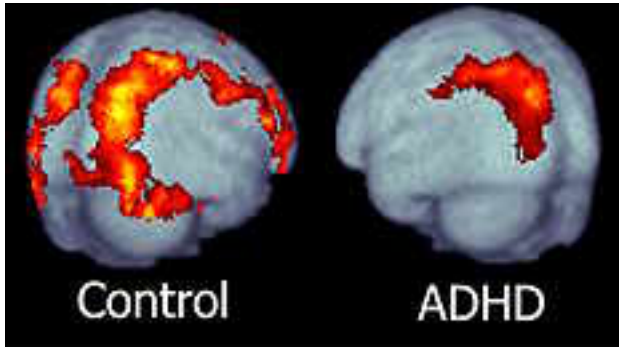
ATTENTION DEFICIT ADHD

ADHD = CLINICAL DIAGNOSIS
NO BIOMARKERS AND NO TESTS







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ATTENTION DEFICIT ADHD

IOWA CONNERS: SCREENING

Please circle the number that best describes your boys behavior **TODAY**

0= not at all; 1= Just a little; 2= pretty much; 3=Very much

1.	Fidgeting	0	1	2	3
2.	Hums and makes other noises	0	1	2	3
3.	Excitable, impulsive	0	1	2	3
4.	Inattentive, easily distracted	0	1	2	3
5.	Fails to finish things he starts-short attention span	0	1	2	3
6.	Quarrelsome	0	1	2	3
7.	Acts 'smart'	0	1	2	3
8.	Temper outbursts - behavior explosive and unpredictable	0	1	2	3
9.	Defiant	0	1	2	3
10.	Uncooperative	0	1	2	3

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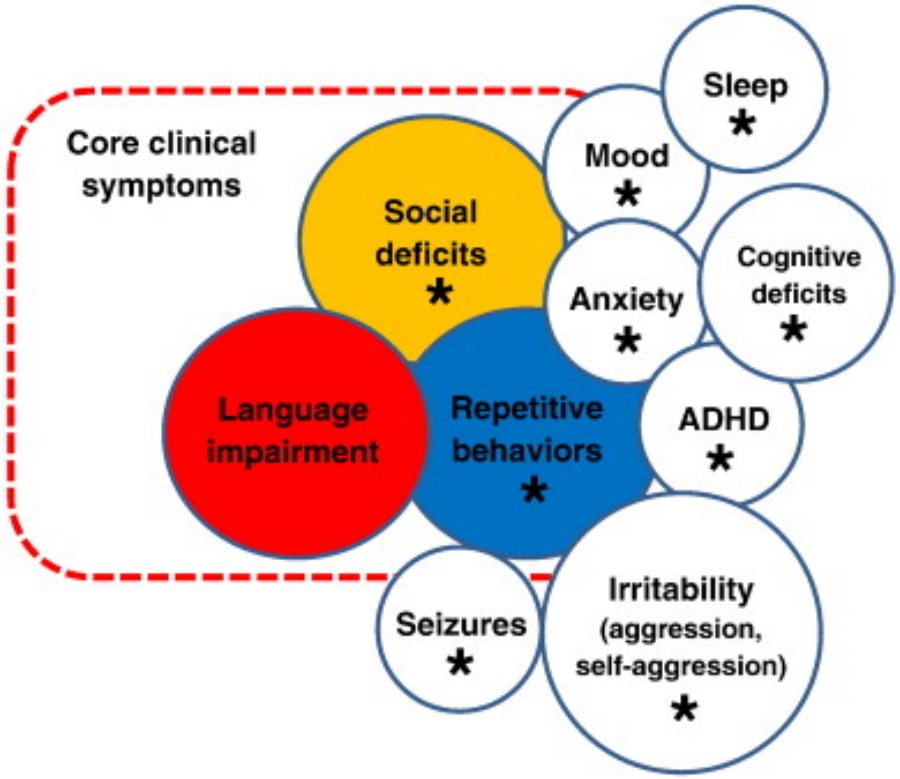
POPULATION MEAN = 0.5%

AUTISM SPECTRUM



REPETITIVE BEHAVIORS= OBSESSIONS

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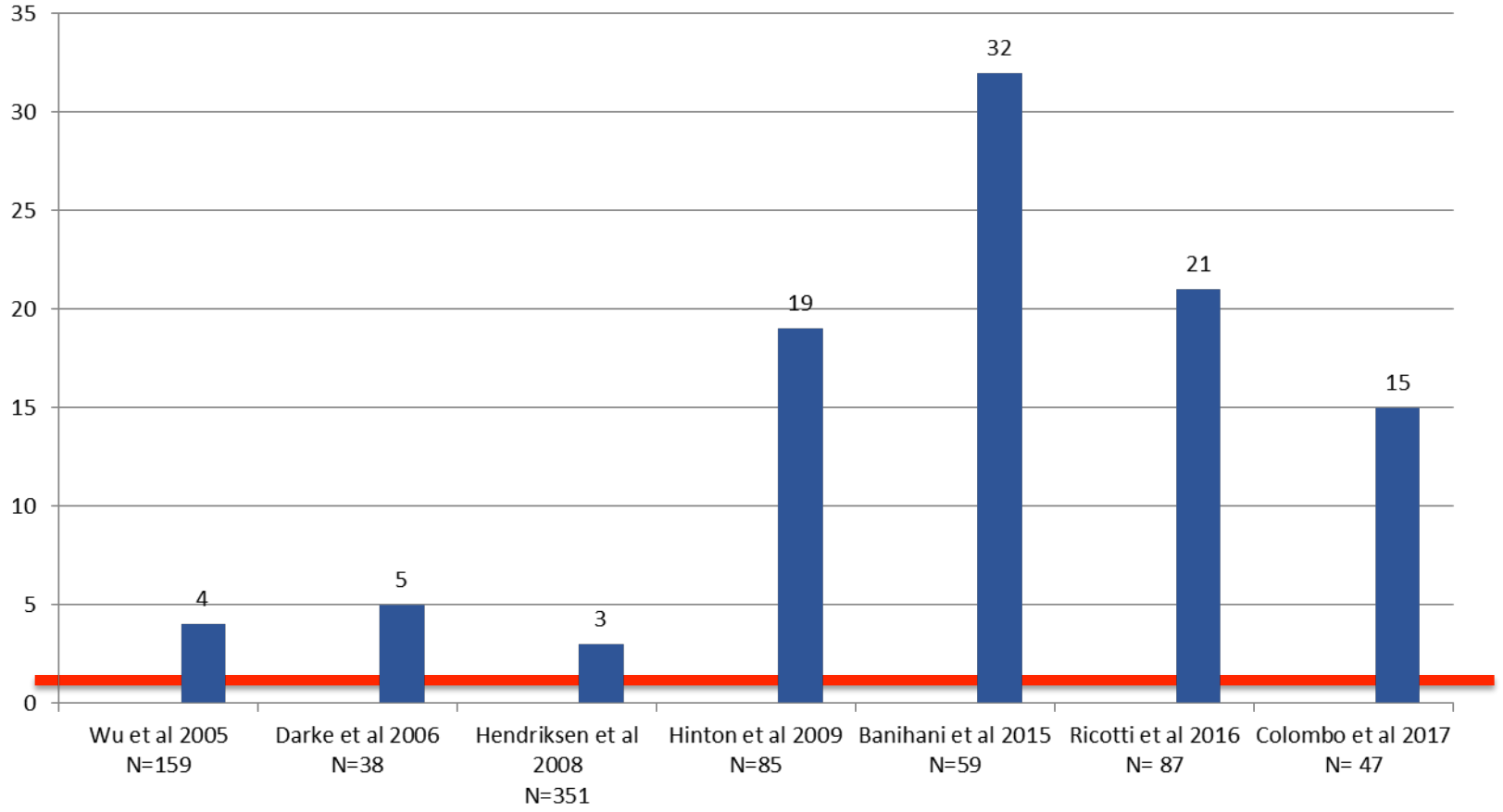
PREVALENCE OF ASD IN DMD 4-32%:

AUTISM SPECTRUM



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HOW DIAGNOSIS ASS WAS MADE:

AUTISM SPECTRUM

Wu (2004)	Clinical history on basis of DSM-IV classification
Darke et al (2006)	Social Communication Questionnaire and Children's Communication Checklist
Hendriksen et al (2008)	Clinical history on basis of parent report
Hinton et al (2009)	Social Communication Questionnaire
Bahnihani et al (2015)	DSM-IV and Autism Diagnostic Observation Schedule (ADOS)
Ricotti et al (2016)	Social Communication Disorder Checklist
Colombo et al (2017)	Autism Diagnostic Observation Schedule (ADOS)

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OBSESSIVE COMPULSIVE DISORDER

Original Article

Journal of Child Neurology
2018, Vol. 33(9) 572-579

DOI: 10.1177/0883073818774439
journals.sagepub.com/home/jcn
SAGE

Descriptive Phenotype of Obsessive Compulsive Symptoms in Males With Duchenne Muscular Dystrophy

Angela J. Lee, BA[1], Edward T. Buckingham, MD[2], Aaron J. Kauer, MD[2], and Katherine D. Mathews, MD[3]

OCD

N = 107; prevalance = 15%
Hendriksen et al (2008)= 5%

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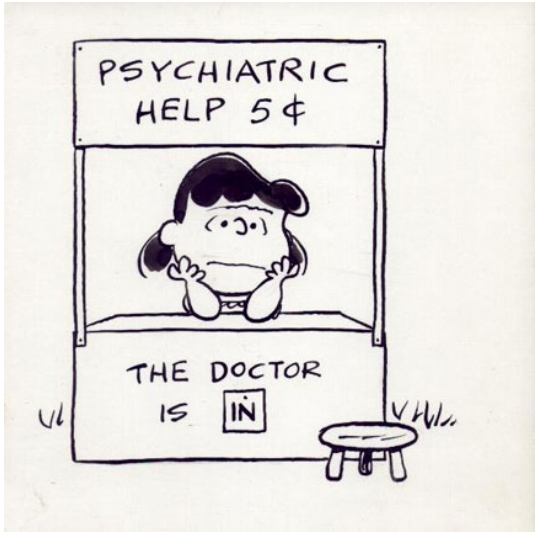
TREATMENT OPTIONS



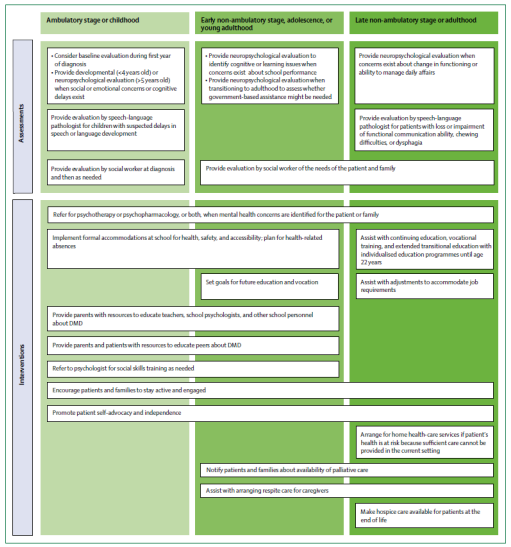


Figure 1: Considerations for psychosocial care of individuals with Duchenne muscular dystrophy
DMD=Duchenne muscular dystrophy.

STANDARDS OF CARE:

- Psychotherapy
- Psychopharmacology
- Formal schoolplanning
- Vocational training
- Psychoeducation
- Social skills training

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PSYCHOPHARMACA

Available online at www.sciencedirect.com

ScienceDirect

Neuromuscular Disorders 26 (2016) 659–661

www.elsevier.com/locate/nmd

Case report

Diagnosis and treatment of obsessive compulsive behavior in a boy with Duchenne muscular dystrophy and autism spectrum disorder: A case report

Jos G.M. Hendriksen [a,b,*], Sylvia Klinkenberg [a,b], Phillipe Collin [a,c], Brenda Wong [d], Erik H. Niks [e], Johan S. Vles [a,b]

Case report on a clinical significant and positive effect of Fluoxetine (20mg/day) on OCD behavior.

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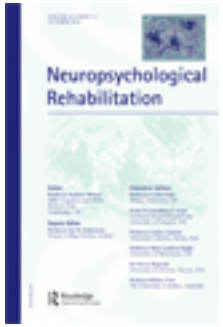
PSYCHOPHARMACA

IN PROGRESS:
STUDY ON EFFECT METHYLPHENIDATE IN DMD AND ADHD (N=10)

Lionarons, Hellebrekers et al 2018
European Journal of pediatric Neurology



CAN WORKING MEMORY BE TRAINED?

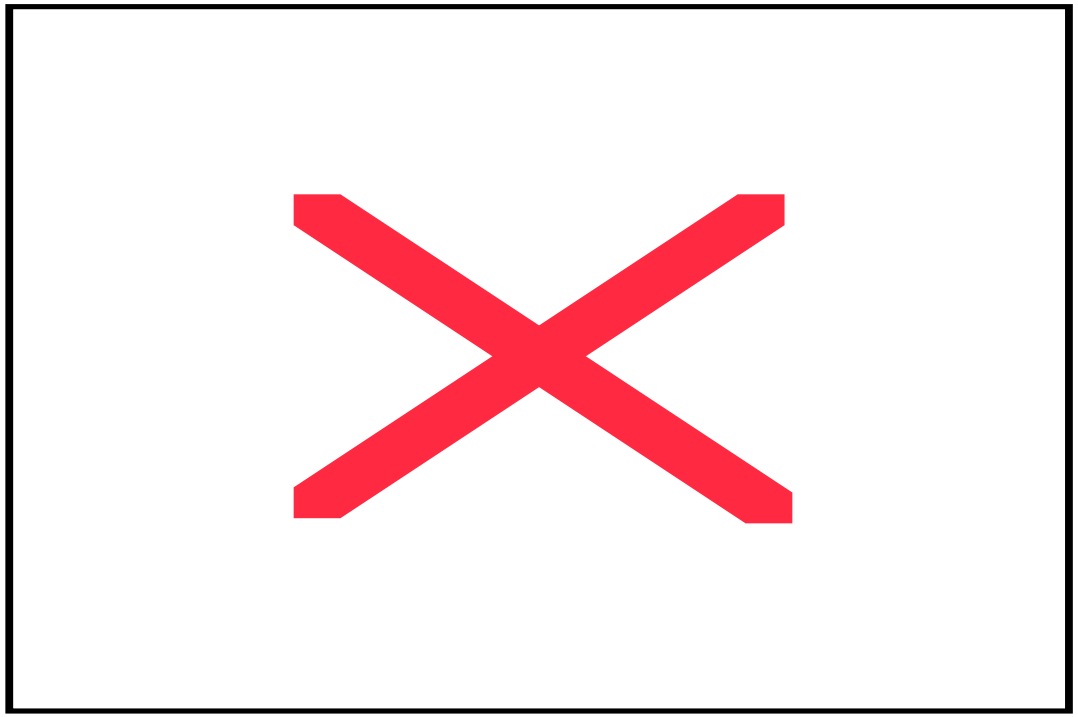
Efficacy of working memory training in children and adolescents with learning disabilities: A review study and meta-analysis

Janneke C. A. W. Peijnenborgh[abf], Petra M. Hurks[c], Albert P. Aldenkamp[bdef], Johan S. H. Vles[abd] & Jos G. M. Hendriksen[adf]

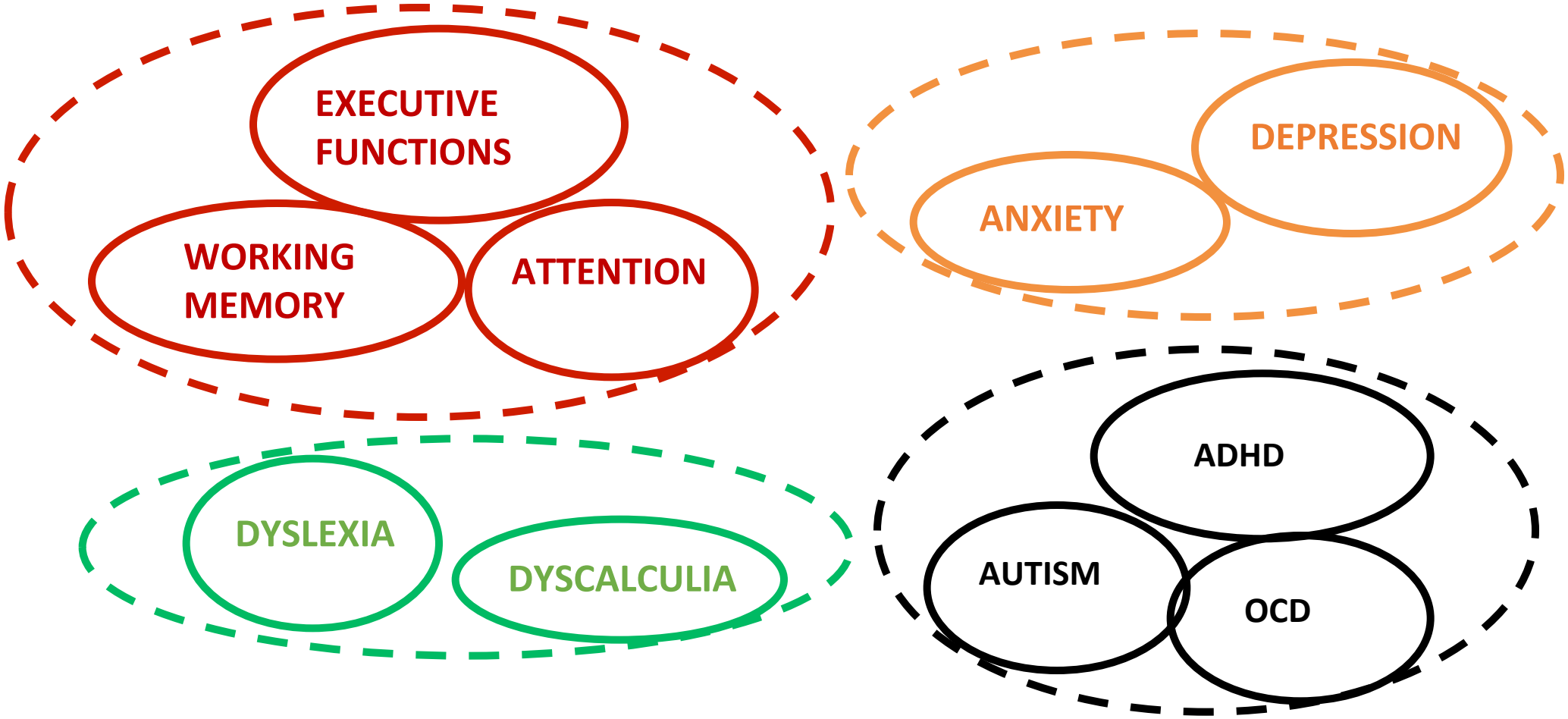
[a] Center of Neurological Learning Disabilities, Kempenhaeghe Epilepsy Center, Heeze, The Netherlands

Limited but promising results that merit further attention

SUMMARY AND CONCLUSIONS



SYSTEMATIC SCREENING FOR THE BIG TEN LEARNING BEHAVIOR DUCHENNE



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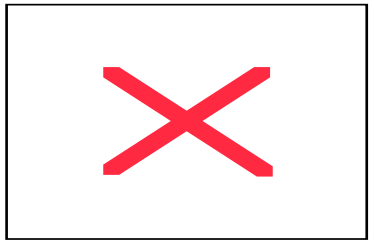
GOLD STANDARD

STEP 1 = ADEQUATE SCREENING
STEP 2 = MAKING A DIAGNOSIS



We need to develop a gold standard of screening and detecting mental health problems in boys and young men with DMD.

Physical limitations may result is false negatives (Colombo et al 2017)

and also false positives (!)

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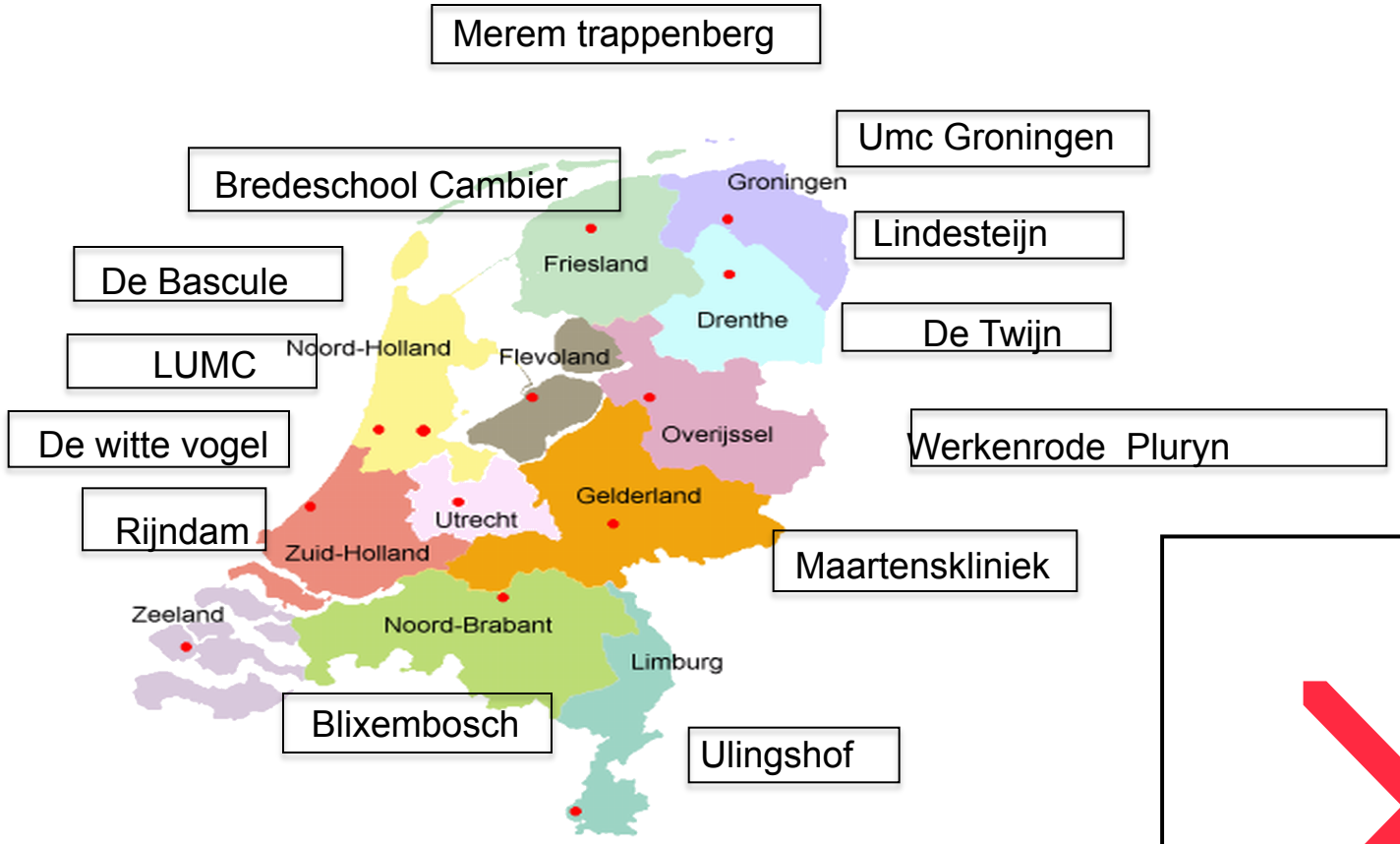
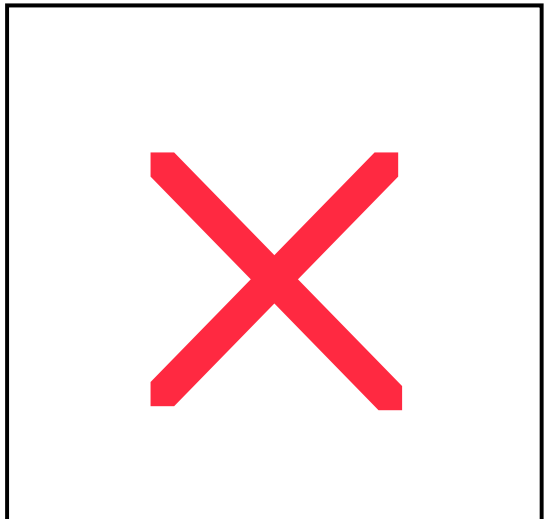
DUTCH FORUM FOR PSYCHOLOGISTS AND EDUCATIONALISTS

Nationwide and uniform protocol





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Duchenne Parent Project

TO CONCLUDE

- **MIND THE GAPS IN OUR KNOWLEDGE**
  (DIAGNOSTICS AND TREATMENT OPTIONS). PROTOCOLS NEED TO BE DEVELOPED
- **BEWARE OF FALSE POSITIVE AND FALSE NEGATIVE SCREENING & DIAGNOSIS**
- **DEVELOP A GOLD STANDARD TAKING IN ACCOUNT PHYSICAL AND NEUROCOGNITIVE LIMITATIONS**
- **LETS WORK TOGETHER TO DEAL WITH NON MOTOR COMORBIDITIES**

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TEAM

Prof. dr. J.S.H. Vles, child neurologist
Prof. dr. C.G. Faber, neurologist
Dr. J.G.M. Hendriksen, psychologist
Dr. S. Klinkenberg, child neurologist
Dr. G. Hoogland, researcher
D. Hellebrekers, psychologist
J. Lionarons, MD physician
Dr. R. Hendriksen, physician

Duchenne Parent Project

SPIEREN VOOR SPIEREN
Helpt kinderen met een spierziekte

Maastricht UMC+

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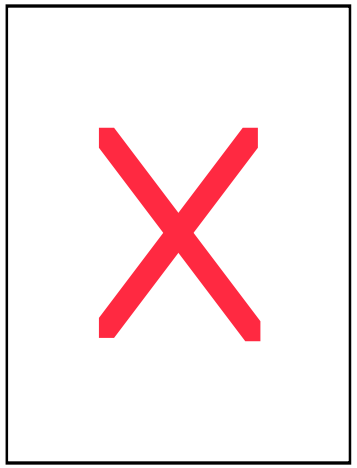
THANK YOU FOR YOUR ATTENTION



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DAILY HASSELS:
DAILY STRESSES OF LIFE WITH DMD

GROWING UP

Fatigue

Not able to endure activity
Bodily pain

Asking for help
Waiting for help

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LUMC
Leiden University Medical Center

Centrum voor Neurologische Leer- en ontwikkelingsstoornissen
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32 DUCHENNE;

WEAKNESS

	DMD
SYMBOL SEARCH (Wechsler scale)	102
SPATIAL THINKING (Block counting)	107
AUDITORY WORKING MEMORY (Wechsler number recall)	85

Reduced Cerebral Gray Matter and Altered White Matter in Boys with Duchenne Muscular Dystrophy

Nathalie Doorenweerd, MSc,[1,2,3] Chiara S. Straathof, MD,[3] Eve M. Dumas, PhD,[3] Pietro Spitali, PhD,[4] Ieke B. Ginjaar, PhD,[5] Beatrijs H. Wokke, MD,[3] Debby G. Schrans, MSc,[6] Janneke C. van den Bergen, MD,[3] Erik W. van Zwet, PhD,[7] Andrew Webb, PhD,[1] Mark A. van Buchem, MD, PhD,[1] Jan J. Verschuuren, MD, PhD,[3] Jos G. Hendriksen, PhD,[6,8] Erik H. Niks, MD, PhD,[3] and Hermien E. Kan, PhD[1,2]

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ATTENTION DEFICIT

AQUIRED ADHD IN DMD IS OVERSHADOWED BY MUSCLE PROBLEMS

EUROPEAN JOURNAL OF PAEDIATRIC NEUROLOGY 19 (2015) 521–524

ELSEVIER

Official Journal of the European Paediatric Neurology Society

Original article

Diagnostic overshadowing in a population of children with neurological disabilities: A cross sectional descriptive study on acquired ADHD

J.G.M. Hendriksen [a,b,c,*], J.C.A.W. Peijnenborgh [a,b,d], A.P. Aldenkamp [b,c,d,e], J.S.H. Vles [a,c,d]

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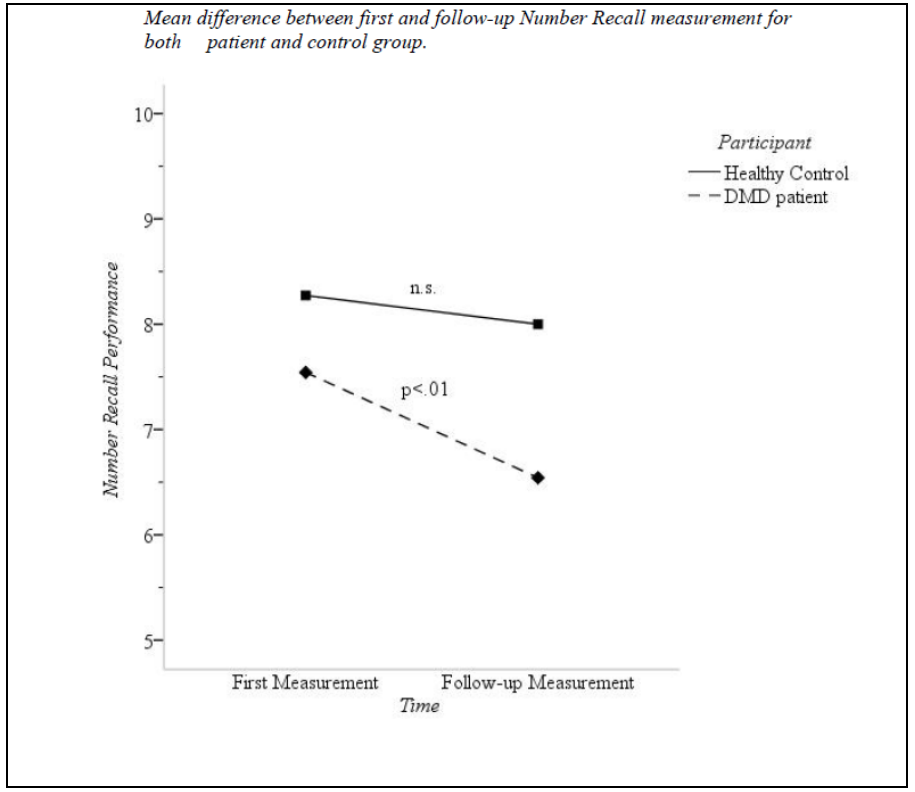
DIAGNOSTIC OVERSCHADOWING

POPINT OF THE STORY

THE PRESENCE OF ONE MEDICAL CONDITION INTERFERES WITH THE AWARENESS OF ANOTHER CONDITION

LONGITUDINAL FOLLOW UP OF WM

WEAKNESS



24 DMD boys
12 healthy controls

significant decline in NR performance over time ($t(23)=2,937$; $p<.01$)

Hellebrekers, Hendriksen et al submitted for publication

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DUCHENNE AND AUTISM

AUTISM SPECTRUM

Original Article

Journal of Child Neurology
Volume XX Number X
Month XXXX xx-xx
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10.1177/0883073807309775
http://jcn.sagepub.com
hosted at
http://online.sagepub.com

Neuropsychiatric Disorders in Males With Duchenne Muscular Dystrophy: Frequency Rate of Attention-Deficit Hyperactivity Disorder (ADHD), Autism Spectrum Disorder, and Obsessive–Compulsive Disorder

Joseph G. M. Hendriksen, PhD, and Johan S. H. Vles, MD, PhD

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ATTENTION AND EXECUTIVE FUNCTION

WEAKNESS

Even in boys without intellectual impairments attention and executive functions are believed to to be impaired

Available online at www.sciencedirect.com

ScienceDirect

Neuromuscular Disorders 28 (2018) 122–128

www.elsevier.com/locate/nmd

Cognitive profile in Duchenne muscular dystrophy boys without intellectual disability: The role of executive functions

R. Battini [a,b,1], D. Chieffo [a,1], S. Bulgheroni [c], G. Piccini [d], C. Pecini [b], S. Lucibello [a], S. Lenzi [b], F. Moriconi [a], M. Pane [a], G. Astrea [b], G. Baranello [c], P. Alfieri [d], S. Vicari [d], D. Riva [c], G. Cioni [b,e], E. Mercuri [a,*]

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SERIOUS OBSESSIONS AND COMPULSIONS

e.g. Taking prednisolon was always preceded by saying 10 times “peanut butter” and taking exactly 10 sips of water

OCD