

PPUK Learning and Behaviour toolkit for Duchenne Muscular Dystrophy

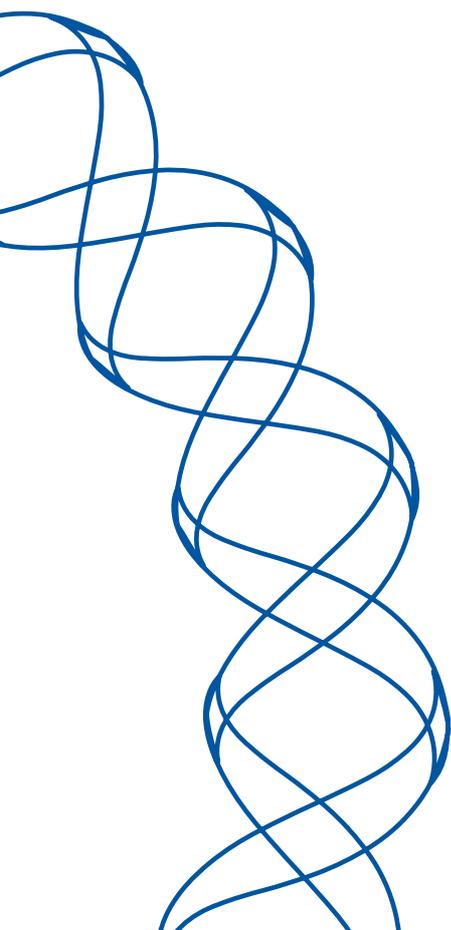


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duchenne
muscular
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DEDICATION AND ACKNOWLEDGEMENTS

Dedication

To all the families and boys with Duchenne Muscular Dystrophy who have inspired us all. We want this toolkit to help young men with DMD realise their potential and to lead longer and more fulfilled lives.

Acknowledgements

Thank you to all members of the working party who have given so much of their time and professional expertise. We would like to thank Veronica Hinton in particular for all her inspiration and research and help with editing this toolkit. We have made the final edit to the toolkit to ensure continuity but hope that we have kept the essence of the wonderful spirit of cooperation and sharing of ideas and best educational practice of the working group. It does not represent any one person or group's views and we hope that users will take the numerous contributions as starting points to improve the educational opportunities of boys with Duchenne.

We would like to thank all the PPUK families and supporters who have fundraised to make this possible.

There is no charge for this toolkit but PPUK would welcome any donations to help us with our work and further projects.

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**Nick Catlin and Janet Hoskin
London 2006**



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SECTION 1: OVERVIEW

1.1 DUCHENNE MUSCULAR DYSTROPHY AND PPUK

Duchenne Muscular Dystrophy (DMD) is a severe and progressive muscle wasting disease that occurs in about 1 in 3500 male births. It is caused by variations in the dystrophin gene on the X chromosome that results in a loss of the protein dystrophin in muscle cells. The severe physical symptoms of the condition almost exclusively affect boys and the gene variation is carried by mothers although some 30% of cases are new gene variations. In some cases women carriers can manifest milder symptoms of the condition. Becker Muscular Dystrophy (BMD) is a milder form of DMD.

The diagnosis is devastating for families as boys will see their muscles waste so that by around 10 years they will be in a wheelchair and without treatment will die on average at 19 years. Improved medical care is seeing young men living longer but services have to be fought for and are poorly coordinated across the UK. Yet DMD affects far more than just muscles. Cognitive and behavioural characteristics are also associated with the disease. Some boys will have severe learning difficulties, yet the majority will have a normal intellectual level. Many boys experience specific difficulties with learning that are at present going undiagnosed.

Further, DMD is associated with some behavioural characteristics, including limited social skills, attentional deficits and depression. Some behaviours are likely due to the underlying etiology, while others may be reactive responses to the condition. DMD impacts on more than just the affected individual; the diagnosis causes changes in families and family members roles. DMD brings with it physical, emotional and financial burdens. A child diagnosed with DMD will require multiple interventions on multiple levels, and the needs of the child will change with time. The goal of this document is to educate families, professionals and schools about the academic and behavioural concerns children with DMD may face.

Parent Project UK (PPUK) was set up by Duchenne families in the UK as a National Charity to campaign and to raise funds for a cure and more effective treatments for DMD. Since its launch in 2001 a number of PPUK families have reported that they have concerns with their sons' behaviour and their performance at school. At the PPUK 3rd International conference in London in October 2005 attended by 200 family members and international researchers and clinicians there was considerable interest in the research examining cognitive and academic difficulties among boys with Duchenne. Professor Veronica J. Hinton, a neuropsychologist who has been studying the cognitive, behavioural and psychosocial effects of the disease in the USA, spoke with families about emerging patterns to learning and behaviour problems within the Duchenne community.

As a result, PPUK commissioned a working group in the UK to publish a toolkit for parents, Educational Psychologists, Speech Therapists, SENCOs, Special Needs Teachers and SEN Support staff in schools.

The working group met with Veronica J. Hinton, in London on June 23rd 2006. This document is a compilation of information about the cognitive and behavioural characteristics associated with DMD, as well as interventions that may be helpful in improving the quality of life for all those affected.



1.2 WORKING GROUP

Annie Aloysius AAloysius@hhnt.nhs.uk is the Speech and Language Therapist for the Dubowitz Neuromuscular Centre at Hammersmith Hospital London, one of the leading muscle centres in the UK. She is a specialist in the care of children with Neuromuscular Disease has published clinical papers and written fact sheets for the Muscular Dystrophy Campaign.

Gill Backhouse g.backhouse@ucl.ac.uk is a Chartered Psychologist in the Dept. of Human Communication Science at University College London. Her main role has been in training Teachers & SLTs to identify & support children with specific learning difficulties. Gill has published the JCQ/PATOSS Guide to Access Arrangements During Examinations & co-edited Dyslexia: Assessing and reporting (Patoss/Hodder Murray).

Nick Catlin nick@ppuk.org

Nick is a founding member and the current CEO for Parent Project UK Muscular Dystrophy. Nick has a BSc (Hons) degree in Chemistry and a Diploma from UCL in Specific Learning Difficulties (Dyslexia). He was a SEN teacher for 20 years and Special Educational Needs Coordinator (SENCo) in Newham London before working full time for PPUK. Nick has a son with DMD.

Dr Michelle Eagle michelle.eagle@btinternet.com is a specialist physiotherapist at the Institute of Human Genetics, University of Newcastle upon Tyne. Michelle has published papers on clinical support for patients with Duchenne Muscular Dystrophy and is a member of the clinical team in Newcastle that is one of the leading muscle centres in the UK.

Dr Angela Fawcett a.fawcett@sheffield.ac.uk is a Reader in the department of Psychology at the University of Sheffield. Following experience of dyslexia in her family, she returned to study as a mature student. Her theoretical research on dyslexia with Rod Nicolson fed into the construction of best-selling screening tests for dyslexia from cradle to grave. Angela was chair of the 2004 British Dyslexia Association International Conference, and editor of Dyslexia: an International Journal of Research and Practice. Angela is Vice President of the British Dyslexia Association.

Professor Veronica Hinton hintonv@sergievsky.cpmc.columbia.edu Veronica is an Assistant Professor of Neuropsychology at Columbia University New York USA. She has published research on the selectively delayed development of verbal immediate memory and phonological processing skills of boys with Duchenne Muscular Dystrophy. Veronica's ongoing work includes examining the nature of the cognitive deficits of boys with DMD and relating this to the type of gene variation.

Janet Hoskin janet@ppuk.org is a practising specialist teacher for young people with dyslexia and specific learning difficulties in Waltham Forest, London. Janet has an MA from Cardiff University and a Diploma in Specific Learning Difficulties from the Hornsby International Dyslexia Centre. She is an Associate Member of the British Dyslexia Association and a member of PATOSS. She has a son with DMD and is a founding member and Director of Parent Project UK.

Cathy Szeplaki catherine.szeplaki@leedsth.nhs.uk is the Senior Paediatric Occupational Therapist working with the Leeds Neuromuscular Team. She works closely with children/young people and their parents/carers in their homes, schools and clinics in the Yorkshire region. She has a background working in a Child Development Centre and is currently working towards her MSc in Professional Health Studies.

Dr Angela Mijovic Angela.Mijovic@swlstg-tr.nhs.uk is a specialist registrar in child and adolescent psychiatry. Angela is based at St George's Hospital in London, where she has both clinical and teaching commitments. Her main research interest is related to neurodevelopmental disorders notably autism and attention deficit hyperactivity disorder. Angela is an honorary specialist registrar in research (in child and adolescent psychiatry) at Great Ormond Street Hospital for children in London, and honorary lecturer in child and adolescent psychiatry at St. George's, University of London.

Professor David Skuse dskuse@ich.ucl.ac.uk directs the Behavioural and Brain Sciences Unit at the Institute of Child Health, University College London. He also has a clinical appointment as a Consultant in Developmental Neuropsychiatry at Great Ormond Street Hospital for Children, where he runs a clinic for children with social and communication disorders. He directs the specialist-training program for residents in child psychiatry in Central and East London.

Since the workshop we have also co-opted:

Shana Cyrulnik shanasteiner@yahoo.com Shana is a postgraduate fellow at Columbia University in New York and has worked with Veronica Hinton to publish a recent paper on DMD and the role of the cerebellum in cognition and behaviour.

Dr James Poysky jxpoysky@TexasChildrensHospital.org is an Assistant Professor of Child Psychology at Baylor College of Medicine and a Clinical Neuropsychologist at Texas Children's Hospital, Houston, Texas, USA. His research and clinical interests are in cognitive functioning in neurodevelopmental disorders, and neurodevelopmental disorders in DMD. He has a son with DMD and is actively involved in the Parent Project Muscular Dystrophy organisation in the USA.

Professor Jeremy Turk MD BSc(HONS), FRCPsych, FRCPCH, DCH jturk@sgul.ac.uk is Professor of Developmental Psychiatry & Honorary Consultant Child & Adolescent Psychiatrist in the Division of Clinical Developmental Sciences at St. George's, University of London.

1.3 HOW TO USE THIS TOOLKIT

Nick Catlin and Janet Hoskin

Poor assessment of the learning or behaviour difficulties of boys with Duchenne Muscular Dystrophy (DMD) can lead to school failure or even unnecessarily being sent to a Special School. This toolkit has a strong focus on getting assessment right as early as possible and ensuring that parents and professionals work as a team to identify needs. It is our view that the mainstream school curriculum with trained specialist support staff offer the best place for most boys with DMD to learn. The rich curriculum and social network of peers without a disability offers boys with the optimum environment to learn and make friends for life.



You are probably reading this because you are concerned about your sons' behaviour or learning. Or for the first time you have met a young person in school or in the community who has Duchenne Muscular Dystrophy. This toolkit has been edited by Nick Catlin and Janet Hoskin from Parent Project UK, a National Duchenne Charity that has commissioned this resource, as well as Veronica J. Hinton, a researcher whose work has focused on understanding the cognitive, behavioural and psychosocial aspects of Duchenne. PPUK has brought together professionals who have expertise in learning and behaviour to develop this toolkit and educate families and teachers about ways to improve the lives of those affected with Duchenne. By sharing what we know of the disorder and the means to assess and treat potential areas of concern, we hope to improve the quality of life for all boys and families living with Duchenne.



As a whole this toolkit represents various research outcomes, opinions, assessment methods and resources for teaching. We recognise that it is not exhaustive and does not represent the views of any one person or group. It is important that all parents and professionals bring their own methods and experience when helping to assess and support young people with DMD. We hope that the toolkit can be a starting point for a more informed approach to assessing the needs of young people with DMD.

The toolkit is not a bible but a launch pad for assessing the needs of every young person with DMD. Every young person will be different and so it is very important to have a full assessment as early as possible to ensure that the person's needs are clearly identified.

1.4 QUESTIONS AND ANSWERS

Veronica Hinton and Shana Cyrulnik

1. Do boys with DMD/BMD have cognitive problems?

Yes. As a group, their IQs are about one standard deviation lower than expected for the general population. Most children with DMD/BMD still perform within the normal range, but more children with DMD have low intellectual skills than children in the general population. Further, children with DMD/BMD perform particularly poorly on tests of verbal skills.

2. Why would a muscle disease have associated learning problems?

It's called muscular dystrophy, but it's not just a muscle disease. The disease is caused by a genetic defect that influences the development of the muscles AND the brain. Children with DMD/BMD are missing a protein, called dystrophin, from both their muscles AND their brain. Without dystrophin in the brain, some areas (like the cerebral cortex and the cerebellum) may not function as efficiently, and this likely results in the observed learning problems that children with DMD/BMD have.

In the brain, dystrophin may be responsible for synaptic function. If a brain develops without dystrophin it may not transmit information as efficiently as a normally developing brain. This lack of efficiency may impact upon the coordination of cognitive information; that is to say, these children may have difficulty processing cognitive information in a quick and coordinated manner, something that might be rapid and effortless in other children. As such, a child with DMD may have difficulty when trying to process information which requires coordination of multiple pieces of information. For example, trying to process a set of instructions with many pieces, or trying to sound out a word with many phonemes.

3. Is there a specific neuropsychological profile associated with DMD/BMD?

Yes. All boys with DMD/BMD have a similar cognitive profile, regardless of overall level of intellectual function or degree of physical impairment.

This does NOT mean that all boys with DMD/BMD are impaired. Rather, across the subjects there are characteristic patterns of relative strengths and weaknesses.

4. What is the profile?

Boys with DMD/BMD are generally weak in attending to and comprehending complex verbal information. That is, when presented with complicated "strings" of verbal information, many children just don't "get it." A possible real life example: You say, "Go upstairs, brush your teeth, put your pajamas on and get into bed." You then go upstairs and find he's put the pajamas on



the bed. You may feel frustrated that he didn't do as he was told, but he feels he has, because he could only "get" part of what was said and knew it had something to do with pajamas and bed. He can understand the parts of this statement, it's when it's all put together that it is just too much.

Moreover, boys with DMD/BMD have difficulty discriminating speech sounds, or phonological processing. Youngsters may make sound substitutions in their speech, have difficulty repeating verbal sounds accurately, and then find learning to "sound out" words in beginning reading particularly problematic. In general, the children with DMD/BMD show reading difficulties similar to those shown in children with dyslexia or reading disabilities, and often cannot tell some sounds apart from others. It's a subtle, but real, finding that may have big implications for early language development and later reading acquisition.

5. Are there areas that they are strong in?

Yes! Boys with DMD/BMD have numerous strengths. They are strong in learning and rote memory. They also have good visual perceptual skills (that is, they can discriminate visual patterns, recognize incomplete pictures and construct puzzles well). They have excellent vocabularies. They are good at problem solving and abstract thinking.

It's very important to keep these strengths in mind when thinking about ways to help in school. Building on these may help alleviate lots of learning-related frustrations that these children may have.

6. Will the learning problems get worse over time?

No. Unlike the physical aspects of DMD, the cognitive issues are not progressive. Further, if anything, the cognitive issues improve over time. Language problems that the young children struggle with may be very limiting at age three or four. At ages five and six, children may have difficulty attending to the level of verbal information that their peers do, and that can result in "acting out" or problem behaviours. They also may struggle with learning the basic early reading skills. Once they are seven and eight, they seem to have a much better command of their verbal comprehension. And by nine or ten, the difficulties are much less problematic.

The difficulty comprehending complex language appears to be a **delay** rather than a **deficit**, so boys may "catch up" over time. Overall, boys with DMD/BMD appear to be about two years behind where they should be on this skill, yet they are age-appropriate on most other verbal skills.

Be aware, however, that children who do not grasp the basic letter-sound associations when they are first taught to read may do progressively worse as school demands increase. Reviewing phonic skills when the child with DMD/BMD is a little older and ensuring the child masters decoding will prevent this.

7. Are there specific behaviours associated with DMD/BMD?

Parents rate DMD/BMD boys as being more immature and having more social problems than their unaffected siblings. Overall, these areas are weakest for DMD/BMD young people. For some children with DMD/BMD (but not most) these problems are severe enough to fall on the autism spectrum of behavioural disorders, as they impact both on language and reciprocal social skills. More children with DMD/BMD have autism spectrum disorders than expected in the general population, yet most have only mild behaviour issues that do not warrant diagnosis. Some researchers have



suggested the children have more attention deficits than their peers. Some have also suggested the boys tend to be more depressed than other children, yet more recent findings have indicated that boys with DMD/BMD do NOT rate themselves as being more depressed than their siblings.

8. If there is an UNDERLYING difficulty in coordinating cognitive information how does it contribute to behaviour and school performance?

Language skills in general may be slow to develop. Parents are more likely to report delayed speech in boys with DMD/BMD than their siblings.

Overall on verbal tests, DMD/BMD subjects tend to do more poorly. Previous studies have reported lower verbal IQ than nonverbal IQ. Yet, their single word comprehension is strong. Tests that DMD/BMD children do poorly on require attending to long strings of serially presented information.

Parents rate their children as “immature” or having “attention problems.” Maybe these children are immature in that they don’t understand as much as other children their age. Maybe they’re inattentive in that they can’t follow complicated instructions.

School instruction generally relies on the ability to listen to and understand complex instructions, so poor comprehension of directions could impact on all areas of academic achievement.

Boys with DMD/BMD are better at sight reading than “sounding out.” The mechanics or “sounding out” requires many steps - associating the sound with the letters and then stringing the sounds together to derive the word meaning. Maybe the skills necessary for this are similar to those necessary for understanding complex instructions.

9. What should be done to help?

Remember that these findings are based on what we know about DMD/BMD children as a group. Individual assessment is crucial to determine each child’s specific cognitive strengths and weaknesses and plan the best possible interventions.

An educational or neuropsychological evaluation is a one-on-one, paper-and-pencil and question-and-answer evaluation. A qualified professional administers, scores and interprets the child’s tests. A child with BMD/DMD may do relatively poorly on tests of Digit Span, Sentence Repetition, Nonword Repetition, Story Memory and Sound Blending, compared to his performance on tests of Receptive Vocabulary, List Learning, Information, Matrices and tests of abstract thinking.

10. Is there a model for how everything fits together?

Yes, we think this:

- In DMD/BMD a gene on the X chromosome is mutated. Normally, the gene codes for a protein called dystrophin.
- The mutated dystrophin gene prohibits dystrophin from being made.
- The affected children develop without dystrophin – it is missing from their muscles and their brains.
- The consequence for the muscles is known and results in progressive weakness. For the brain, it’s less clear, but it results in lowered metabolism in particular brain areas, including the

cerebellum and the cerebral cortex. This is associated with a diminished ability to listen to and process verbal information. Children affected with DMD/BMD remember fewer digits and shorter sentences than their unaffected siblings.

- Young boys with DMD/BMD are slower to acquire early language skills than their peers and may have poor reciprocal social language.
- Once they develop communicative speech, the effects may not be too obvious. Yet, a boy with DMD/BMD's ability to comprehend complex language may be comparable to a child two years younger. Thus, even though the affected boy may know factual information, have a vocabulary and solve puzzles as well as his classmates, he may have considerable more difficulty following verbal instructions.
- If the children with DMD/BMD aren't processing school lectures quite as well as their peers, they may not be learning the basic academics as well. Further, they may have particular difficulty learning to read and decode speech sounds. This may be very frustrating.
- Overall, children with DMD/BMD do not have as strong academic skills as their peers. Although many things may contribute to this, like fatigue, mood, and physical ability, our study demonstrated that decreased verbal span accounted for more of the variance associated with children with DMD doing poorly in academics than any of the other influences.

11. Research and other references are given at the end of the toolkit in Section 6



SECTION 2: PARENTS AND CARERS - GETTING THE BALL ROLLING

2.1 CHECKLIST

Nick Catlin and Janet Hoskin

Remember that DMD/BMD is not just a muscle wasting disease. It will also affect your son's learning and possibly behaviour. But this will not be progressive and with proper professional assessment and support your son will be able to make progress.

With better medical care young men with DMD are living longer and there are an increasing number going on to further and higher education including taking degrees at University. Educational opportunity is very important for a person who will be making their way in life with a physical disability. It will also provide routes to employment and skills vital to keep young men with a positive outlook on life.

Lack of dystrophin in the brain is likely to affect all boys to some degree. For most boys it will mean just a slight change in their overall abilities but for some it could mean profound learning and behaviour problems.

Is your child experiencing:

- Problems with early speech and language
- Not seeming to understand complex instructions (difficulty "minding" you?)
- Poor or fleeting eye contact
- Falling behind his peers with reading and spelling
- Difficulty coping with change in routine
- More than a normal level of tantrums or outbursts of aggressive behaviour
- Very passive behaviour and finds it difficult to communicate and to socialise and make friends

2.2 WHAT TO DO:

- Ask your muscle consultant at the clinic, your GP, your community Paediatrician or your school SENCo to refer your son for a full Educational Psychologist and Speech and Language Therapy assessment for a Special Educational Needs (SEN) Statement. This can be organised at a pre school stage. If you have an SEN Statement request a review.
- Give everyone involved with assessments and education a copy of this toolkit and ensure that they understand that at present there is no cure for DMD.
- Involve the Special Educational Needs Coordinator SENCo or Head Teacher at your school in helping you to get these assessments undertaken. Tell your school SENCo your concerns about reading, behaviour or other issues.
- There should be a "Parent Partnership Officer" employed by the Local Education Authority who is there to assist families in the process of obtaining the right education for their child. Ask them to attend meetings with you to take notes and help you write the Parental Advice for the SEN Statement. Make sure that you clearly state your son's needs as your advice is taken very seriously and you can influence the level of support that he will receive.
- If you are having difficulties getting assessments or need expert advice make contact with the ACE Advisory Centre for Education www.ace-ed.org.uk or the Independent Panel for Special Educational Advice www.ipsea.org.uk .

- A Local Education SEN Officer will write the SEN Statement following the assessments and meetings. Make sure that you read carefully the draft SEN Statement and don't be afraid to suggest changes. If you are unsure about the content or are unhappy about the SEN Statement and provision seek further advice and help.
- Make sure that your child's SEN Statement identifies educational and behavioural needs as well as those for physical disabilities. The SEN Statement can still be used to trigger extra help for your child. Remember you have a legal right to see all assessments that have been written regarding your child. You are able to ask which tests the Educational Psychologist has made on your child and why these have been chosen. (Refer to 3.3).
- From the SEN statement your child will be entitled to specialist support e.g. Speech and Language Therapy sessions, Specialist teacher sessions (dyslexia or Specific Learning Difficulty) Behaviour support teacher, Advisor for Autism, Occupational Therapist, Physiotherapist, classroom support or teaching assistant time. We would suggest that in class support covers at least 50% of the timetable depending on identified needs. Your son might also need short sessions of 1:1 teaching outside of the normal classroom to help with reading and spelling. (Refer to Section 4.2 Reading and Spelling).
- Every year the School has a legal obligation to hold an annual review of the Statement to which you will be invited. You should provide written parental advice for each review that includes your own observations on progress and remaining difficulties. If you are concerned at any time about your son's progress you can request a Review meeting. Always take notes.
- Make sure that the classroom teachers and SEN support staff are aware of your son's needs and have been trained ideally in working with students with specific learning difficulties.
- Ask to see your child's Individual Educational Plan and make sure that the targets are realistic and are really being met. Make sure the IEP is reviewed every term with the SENCo.



SECTION 3: ASSESSMENT

3.1 INTRODUCTION

Nick Catlin and Janet Hoskin



We strongly recommend that any assessments and programmes for learning, speech and language or behaviour are delivered with the full cooperation of parents and professionals. In the case of Duchenne Muscular Dystrophy, families who are trying to cope with a very difficult prognosis of a severe muscle wasting disease for which there is no cure may find it very stressful to now discover that these problems can be compounded by problems at school with learning and behaviour. By optimising the partnership between professionals and parents, huge steps will be taken to offer boys with DMD the training and support that they need.

Educational Psychologists, Speech Therapists, Clinical Psychologists, Teachers with a Specific Learning or Dyslexia diploma, Specialist behaviour support teachers and Autism advisors will play a key role in the cognitive and learning assessment of boys with DMD. We acknowledge that professionals will have their own preference for test batteries and assessment tools. We hope that the following suggestions might add to or compliment tools for investigating the possible cognitive and behaviour problems presented by boys with DMD. We strongly recommend: **Dyslexia? Assessing and reporting The PATOSS Guide** Edited by Gill Backhouse and Kath Morris Hodder Murray 2005 as an excellent guide to testing and assessment.

3.2 GENERAL OVERVIEW AND ASSESSMENT AT DIFFERENT STAGES

James Poysky

Behaviour and cognitive functioning can vary significantly in children with Duchenne muscular dystrophy (DMD). In general, among boys with DMD, IQ and academic achievement scores are about one standard deviation lower than their unaffected peers. When their performance is compared to normative data, unaffected siblings or children with a different neuromuscular disorder, as a group children with DMD consistently score more poorly. Yet the range of scores is great, such that most score in the “normal” range, but as many as 30-35% may score in the “mentally deficient” range. Verbal reasoning appears more affected than nonverbal reasoning. Progressive cognitive decline does not occur, and there is evidence that VIQ scores may improve with age. Weaknesses in language use/processing, including auditory comprehension, expressive vocabulary, and story recall have been reported. Rate of learning for information presented over multiple trials appears to be within normal limits, as does long-delay recall. In contrast, deficits have been identified in auditory/verbal short-term memory, also referred to as span memory, working memory, or the amount of information one is able to take in at any given time. Processing speed/complex attention may also be problematic. Research examining visual short-term memory has resulted in equivocal findings. Visual-perceptual skills appear to generally be intact. Basic reading, mathematics, and spelling skills may be deficient, and have been correlated with weaknesses in verbal working memory. Up to 40% of boys with DMD may have dyslexia, with associated problems in phonological processing.



Emotional and behaviour problems are associated with DMD, and prevalence rates are comparable to those in other neurodevelopmental disorders. Some behavioural difficulties may be associated with the underlying etiology, while others may be reactive responses to the progressive course of the illness. Most data rely on parent reports.

34-40% report significant social behavioural problems and estimates of the occurrence of autism range from 4-19%. There is emerging evidence that some of the deficits of autism improve with age in DMD, including interest in relationships, sharing interests with others, and emotional/social reciprocity. 12-24% of parents report their child with DMD may have attention-deficit/hyperactivity disorder (AD/HD). DMD boys with a diagnosis of AD/HD typically have poorer psychosocial adjustment than boys without a diagnosis of AD/HD. 8-50% of children with may have symptoms of emotional distress, such as depression and/or anxiety, and these may be associated with parental coping strategies. That is, the better the child's parents are coping with the illness, the less likely the child will present as depressed. It is extremely important to recognise that in a condition like Duchenne, all family members are affected, and similarly, all family members' emotional states may impact on one another.

The following categories are provided as general guidelines to keep in mind when assessing boys and young men with DMD. They are not mutually exclusive and considerable overlap should be expected. The list of measures is not exhaustive and may not be always be appropriate, depending on the child, clinical setting, or presenting concerns.

Parent Interviews with the Educational Psychologist and SENCo

Parent interviews are invaluable for understanding any child and can offer tremendous insight into his individual strengths and weaknesses. Parents of boys with DMD are often aware from a very early stage of problems that their child is having with physical developmental delay and with speech and language. They are also having to deal day to day with any difficult behaviour if it exists. Parents might be wary of flagging up these issues so as not to cause a fuss or simply because they don't want their child to be labelled with a learning or behaviour problem in school. But parental insights for DMD assessments are crucial in unravelling any potential problem.

Ages 0-5 years

Boys with DMD (up to 35%) may initially be referred to health care professionals due to concerns about language and cognitive delays. As such, comprehensive neurodevelopmental evaluation for younger children with DMD is important. This should include expressive and receptive language, visual-spatial problem solving, and fine motor skills, in addition to gross motor skills. Tests that provide instructions in simple language or that allow for repetition of directions are particularly valuable for ensuring that the skill being tested is measured, rather than the child's ability to follow the instructions. Screening for an autism spectrum disorder should be considered for all toddler-age boys with DMD, with more comprehensive autism assessment conducted as needed. Significant symptoms of impulsivity associated with AD/HD may start to become problematic during this period, as well as other externalising problems such as oppositional/argumentative behaviour and aggression. Of note, due to the physical limitations of DMD, impulsivity may be more predominant than hyperactivity in AD/HD hyperactive-impulsive and combined types. Rigid thinking patterns and problems with adaptability may be significantly disruptive and interfering at this age. Given the potential for dyslexia, evaluation of phonological awareness and rapid naming at 4 or 5 years of age is recommended.

Recommended Measures:

- Mullen Scales of Early Learning (developmental assessment)
- Child Development Inventory (development rating scale)
- Parent's Evaluation of Developmental Status (development rating scale)
- Wechsler Preschool and Primary Scale of Intelligence (IQ)
- Preschool Comprehensive Evaluation of Language
- Preschool Language Scale (expressive/receptive language)



- Comprehensive Test of Phonological Processing (phonological awareness, rapid naming)
- Modified Checklist for Autism in Toddlers (autism screen)
- Childhood Autism Rating Scale (autism rating scale)
- Social Communication Questionnaire (autism rating scale)
- Behaviour Assessment System for Children (broad behaviour rating scale)

Ages 6-11 years

Difficulty mastering academic material is a common complaint during the primary years. Academic difficulties may be due to specific and general learning disabilities. Using IQ tests that have verbal reasoning, nonverbal reasoning, processing speed, and working memory indexes may provide valuable information, but combining them into a Full Scale IQ may lead to misinterpretation of a child's strengths and weaknesses. The use of a nonverbal IQ test that has limited time and motor demands should be considered. This is particularly the case when using a discrepancy model to determine if a child meets criteria for a specific learning disability. When assessing reading, the use of tests that measure word recognition and decoding speed is recommended, as both have been shown to be problematic in DMD. Problems with verbal working memory and attention may be problematic for boys with DMD of this age, but may be overlooked by parents and teachers because they are not disruptive. Evaluation using parent/teacher rating scales and objective measures is recommended. Similarly, evaluation of executive functions should be considered, including planning, organisation, mental flexibility, and initiation.

As the boy's awareness increases, acute emotional reactions and externalising behaviours can occur in response to their disorder. These problems can occur at any age, but the years just prior to the loss of ambulation appear to be particularly problematic. As such, DMD boys ages 8 to 10 are at increased risk for having poorer psychosocial adjustment. Evaluations should also monitor for perseverative behaviours secondary to mental inflexibility/rigidity. Multiple physical, psychosocial, and cognitive factors may be contributing to social problems, and comprehensive evaluation is recommended. Assessment of parent stress level and coping style may also be helpful in determining appropriate interventions.

Recommended Measures:

- Wechsler Intelligence Scale for Children (broad IQ)
- PHAB (Phonological Assessment Battery)
- WRAT 4 (Wide Range Achievement Test)
- WRIT (Wide Range Intelligence Test)
- BPVS (British Picture Vocabulary Test)
- Leiter International Performance Scale (nonverbal IQ)
- Raven's Progressive Matrices (nonverbal IQ)
- Woodcock-Johnson Tests of Academic Achievement (broad academics)
- Test of Word Reading Efficiency (word and nonword reading fluency)
- Gray Oral Reading Test (reading fluency and comprehension)
- Children's Memory Scale (verbal and visual learning and memory)
- Symbol Digit Modalities Test – Oral version (processing speed/complex attention)
- Digit Span (verbal working memory)
- Token Test for Children
- Behaviour Assessment System for Children (broad behaviour)
- Child Behaviour Checklist (broad behaviour)
- Children's Yale-Brown Obsessive-Compulsive Scale (obsessive-compulsive symptoms)
- Behaviour Rating Inventory of Executive Functions (executive functions)
- Children's Depression Inventory (depression)
- Multidimensional Anxiety Scale for Children (anxiety)



Ages 12 and higher:

As more and more responsibility is expected of boys with DMD (as they grow older), weaknesses in verbal working memory and executive functions may interfere with academic, occupational, and social functioning. This may be true for young men who have had no previous history of problems in these areas. Previous recommendations for evaluation of these problems remain applicable.



Psychosocially, adolescence can be a difficult time for young men with DMD. This may be due to their physical limitations, as well as disruptions in the developmental cycle secondary to medical treatments (such as short stature or delay of puberty as a result of steroid treatment). Symptoms of depression and anxiety should be routinely monitored for. Distress related to issues of dependence/independence should be assessed. Families may also need assistance in preparing for how to address end of life issues.

Recommended Measures:

Many of the measures from the previous section may be applicable.
Personality Inventory for Youth (psychosocial and emotional adjustment)
Beck Depression Inventory (depression)
Wechsler Memory Scale (verbal and visual learning and memory)
Delis Kaplan Executive Function System (executive functions)

3.3 ADVICE FOR EDUCATIONAL PSYCHOLOGISTS

Gill Backhouse

Parental reports together with emerging research findings (Section 6) point to a very real need for the recognition that many – possibly the majority - of boys with Duchenne Muscular Dystrophy (DMD) have learning difficulties which should be considered independently from their other developmental problems. All too often their medical condition and prognosis has tended to overshadow concerns about educational progress. Denied the ability to keep up with their peers in the playground, sporting activities and so-on, it is doubly damaging to their self-esteem and well-being if boys with DMD also struggle to maintain adequate progress in the classroom.

Educational Psychologists (EP's) have a valuable role to play in promoting understanding and awareness of the likely educational needs of such boys and the importance of **early** assessment and **effective** support.

Whilst research is still at an early stage, a robust finding so far has been that despite a wide range of ability in the DMD population, typical and highly specific cognitive 'deficits' exist in the following areas:

- Verbal memory span / working memory
- Phonological processing

Consequent on these difficulties problems with **phonics** and the acquisition and development of age-appropriate **reading** and **spelling** skills typically ensue. Many will therefore present as dyslexic.

A broad range of difficulties with **comprehension** - whether listening or reading – is also reported, but it is unclear at this stage whether this is consequent on reduced verbal memory span or a specific language problem per se. SLT assessment will help to tease out the relative impact of these two factors.

Assessment of IQ

Please see comments by James Poysky (Section 3.2 Ages 6-11 years).



Reading

Decoding is likely to be a weakness (due to poor phonological processing); also **comprehension** which will impair the use of 'top-down' processing. Therefore, a careful analysis of reading skills & strategies is valuable.

- **Speed & accuracy of single word reading** – sight words cf. decoding skills. The Test of Word Reading Efficiency (TOWRE) is particularly useful for this. Any disparity between decoding skills & word recognition will instantly reveal to the SENCo where the thrust of remedial support should be focused. The Nonword Reading Test, Graded Nonword Reading Test, or Nonword subtest in the PhAB may be used to explore **decoding** skills alone.

If decoding skills are weak, it will be extremely important to explain to school staff that encoding i.e. spelling instruction, is by far the most effective way of forging an understanding of how words are written in an alphabetic code.

- **Reading comprehension.** Designed for weaker readers, the Diagnostic Reading Analysis is useful since it includes a listening comprehension passage and then on the basis of just three passages for the child to read, provides scores for accuracy and fluency as well as comprehension

A miscue analysis will reveal the boy's strategies when reading continuous prose. Teachers & parents are likely to know whether he confuses similar-looking words, or 'reads for meaning' but often are not sure how to target support that selectively targets a strength in order to compensate for a weaker aspect.

Paired reading methodology (Section 4.3) can generally be used to improve sight vocabulary, fluency and comprehension. Its advantage is that parents & classroom assistants can be trained to deliver and decoding skills are bypassed. However, it does not target phonics, spelling or word attack skills – all of which are likely to remain a challenge.

Spelling

Careful assessment of **phonological skills** that will feed into practical advice regarding spelling tuition is essential. (If the boy is being seen by a SLT, development of these skills may well be under review.) Depending on the information available in school, it may also be necessary to check sound - symbol correspondence for both reading and writing, including the digraphs, **th, sh, ch, ng**.

Tests of phonological processing readily available in the UK include:

Phonological Abilities Test (5-7 years only), PhAB (6 – 14 Years), Sound Linkage (7+ years and includes a comprehensive scheme of support); CTOPP. Although this has a wide age range (5 – 25 years), excellent Rapid Naming and phoneme deletion subtests, some parts are unsuitable for UK children due to American vocabulary.

The SENCo will find guidance regarding the level of segmentation that boys can manage, extremely useful, since this has obvious implications for the level at which spelling support needs to be targeted.

E.g.

- The boy may need practice in clapping/counting **syllables** and learn the rule that when spelling each one must include a vowel, vowel digraph or **y**.
- Or he may need lots of auditory/oral work on **alliteration and rhyming** in tandem with spelling instruction at **onset & rime** level. (For regular words, this approach demands far less working memory capacity.)



- He may be ready for a more advanced phonemic approach and be able to differentiate clusters at the beginning of words, yet still need practice in hearing the two sounds in a cluster at the **end** of a word.

Inspection of his English work book will soon reveal whether he spells alphabetically or not when writing at sentence/text level, as well as secure/insecure grasp of spelling patterns and rules.

Classroom assistants frequently do not understand the importance or nature of **multi-sensory teaching methods** and so advice which includes specific guidance regarding the use of plastic letters, felt-pens & white boards etc. is strongly recommended.

Furthermore, the necessity of setting finely graded targets and incorporating a good deal of **overlearning and reinforcement** (using games, IT and so-on) should also be emphasised since support – even when appropriately structured - is often delivered at too fast a pace for the dyslexic child.

Listening Skills

Reduced verbal memory span will clearly lead to problems across the curriculum and school staff will benefit from standard advice such as:

- Adults should speak in short sentences & encourage child to repeat instructions /put into his own words / visualise self carrying them out.
- Adult to be careful about 'order of mention' E.g. Not "After you've put your maths books and pencil cases away in your trays and collected your English work books, you can choose a reading book" But "First, put your maths books and pencil cases away in your trays; Second, collect your English work books. Then, choose a reading book"

As ever, the busy SENCo will appreciate practical advice which is clear and generic – what to do and how to do it – which she can utilise in the context of the school's own resources (support materials & staff) & extend to suggestions for the home setting. Clear objectives, broken down into prioritised SMART targets are particularly welcomed!

Boys with DMD are already identified by virtue of their medical condition. The knowledge base regarding their cognitive profiles and associated learning difficulties is restricted at present, particularly with regard to numeracy development. PPUK will welcome any observations from EPs who have a boy with DMD in their case-load, that contribute to our understanding of their particular needs.



Tests

- Comprehensive Test of Phonological Processing (CTOPP); 1999; Wagner R., Torgesen J. & Rashotte C.; Harcourt Assessment
- Diagnostic Reading Analysis; 2004; Crumpler, M. & Mc Carty, C.; Hodder & Stoughton
- Graded Nonword Reading Test; 1996; Snowling, M, Stothard, S. & McLean, J.; Harcourt Assessment
- Nonword Reading Test; 2004; Crumpler, M. & Mc Carty, C.; Hodder & Stoughton
- Phonological Abilities Test (PAT); 1997; Muter, V., Hulme, C. & Snowling, M. J.; Harcourt Assessment
- Phonological Assessment Battery (PhAB); 1997; Fredrickson, N., Frith, U. & Reason, R.; NFER Nelson
- Sound Linkage; 2001; Hatcher, P.; Wiley
- Test of Word Reading Efficiency (TOWRE); 1999; Torgesen, J. K., Wagner, R. K. & Rashotte, C. A.; Harcourt Assessment

3.4 SPEECH AND LANGUAGE ASSESSMENTS

Annie Aloysius

Speech and Language Therapists may come across children affected by Duchenne Muscular Dystrophy in the schools or clinics they work in or they may be referred directly to them for a communication assessment. When assessing these children it is essential that SLT's have an awareness of the specific cognitive and language difficulties they may have. For example verbal memory span and story recall may be specifically impaired impacting on their abilities to score on concept conceptualisation tasks etc.

Awareness of potential areas of weakness as highlighted by recent research will enable targeted screening and more focused assessment.

Screening

The Token Test for Children (TTFC) by Frank DiSimoni is a rapid screening test that measures subtle receptive language dysfunction in children. It is not usually used by SLT's but is useful as a screening tool. It gradually increases the complexity of information recall and sequencing highlighting the difficulties these children may have with holding and recalling increasing amounts of verbal information.

Specific assessments

The Clinical Evaluation of Language Fundamentals (CELF3) useful subtests:

- Concepts and Directions
- Recalling sentences
- Listening to paragraphs

These subtests may highlight the increasing difficulties with language comprehension and recall as the volume and complexity of information to be processed increases. By contrasting performance on these tests to that on receptive vocabulary tests, the selective difficulties observed in children with DMD will be brought out. This demonstrates the difficulty with the amount of information being processed rather than specific concept comprehension issues. Results may be low average for their chronological age or up to 2years delayed in these areas. This reflects a delay in these skills rather than a specific disorder.

The Renfrew Bus Story (Renfrew 1977) that tests consecutive speech will demonstrate the child's difficulties for story memory and recall. Recall is often not cohesive or well sequenced but the main themes of the story are present.

SLT's should be able to highlight/screen these areas through targeted assessment. Further detailed assessment would then be required to further assess specific cognitive impairments in relation to their impact on reading and phonological awareness. Referral on to educational psychology or equivalent professional for a more detailed cognitive assessment to demonstrate strengths and weaknesses in a range of modalities is appropriate.

Information from this multidisciplinary assessment would then be collated and used to form the basis of an appropriate intervention programme

Early screening and assessment by SLT may ensure appropriate targeted interventions to prevent early literacy and numeracy problems and possible early educational failure. Awareness of education staff of



specific difficulties may enable adaptations to the learning environment to enable children to succeed in particular activities and tasks.

There is a higher incidence of autistic spectrum impairments with specific difficulties noted in ability to identify affect and interpret facial expressions. This can result in social interaction difficulties and behavioural issues it is therefore also essential that these areas are screened appropriately and appropriate support and intervention provided.

3.5 SCREENING CHILDREN WITH DUCHENNE FOR READING ASSOCIATED DEFICITS

Angela Fawcett

Duchenne muscular dystrophy is a disorder associated with gradual loss of muscle power, in addition to cognitive deficits. Recent advances in our treatment of this disorder has meant a longer survival time for children suffering from Duchenne. In tandem with the changes in the prognosis for these children, a greater recognition of the cognitive deficits from which they suffer is crucial to their overall quality of life.

In terms of developing reading, research has shown that the timing of intervention is crucial, and that the earlier support can be provided the most effective and cost-effective it can be. The problem has been that parents and teachers are naturally somewhat overwhelmed by the diagnosis of Duchenne, and may emphasise the physical rather than the cognitive aspects of the disorder.



In recent research a potential link has been found between the cognitive symptoms of Duchenne and those found in developmental dyslexia. If we consider the symptoms, both can be characterised by problems in phonological skills, as well as problems with verbal memory based skills of all types. It is hypothesised that for both disorders the cerebellum may play a crucial role. Both groups of children (those with Duchenne and those with developmental dyslexia) show problems in literacy, particularly reading, spelling, and phonology, and in memory tests such as Digit Span. Children with Duchenne present with a lowered intelligence typically around one standard deviation below the mean, and so in some ways their profile is similar to children with generalised learning difficulties.



In order to ensure that children with Duchenne receive the best possible support, screening tests that have been devised to investigate the profiles of children with dyslexia might be particularly useful. These tests are designed to provide a profile of skills in children aged from 3.5 upwards, with the **Dyslexia Early Screening Test** (Nicolson and Fawcett, 2003) for ages 4.5-6.5, and the **Dyslexia Screening test, Junior and Secondary** (Fawcett and Nicolson, 2005) for 6.5-16.5 the best tuned to the age range within which Duchenne falls. These tests have been designed to provide a quick and easy method of testing performance across a basket of skills known to be associated with problems in literacy. The complete test takes no more than 30 minutes, and has been designed to be fun for children to complete and easy for teachers to interpret. Results can be compared with norms for children of the same age to establish a pattern of strengths and weaknesses. Results can then also be compared with the characteristic profile of dyslexia, to identify areas of commonality and difference.

In addition to cognitive skills, these tests include norms for motor skills tests such as bead threading, copying and postural stability. Professional/occupational therapist advice might be needed on whether or not it is appropriate to administer the Postural Stability test to this group.



These tests have been designed to investigate performance across a raft of skills, and make minimal demands in terms of perseverance, which makes them particularly suitable for use with Duchenne children. It should be noted that there are also a range of other tests that assess phonological ability and literacy, for example the computer based COPS tests. Several of these have been recommended for use by Speech Therapists, and can be used in conjunction with the screening tests to give further information on areas which need further investigation.

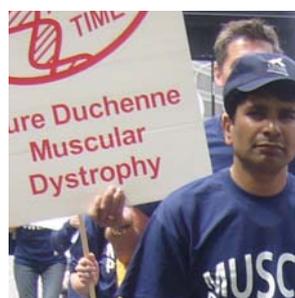
Armed with this profile of need, it should be possible for an Individual Education plan (IEP) to be devised tailored to the needs of each child with Duchenne. The program should be structured and cumulative and avoid reliance on verbal memory. A technique such as Toe by Toe might be appropriate for some children with Duchenne. This aspect of the remediation should be discussed with the special needs advisor and form part of the Statement of Educational Needs to which these children should be entitled.

References

Nicolson, R.I. and Fawcett, A.J (2003). *The Dyslexia Early Screening Test*. 2nd edition. The Psychological Corporation, London.

Fawcett, A.J. and Nicolson, R.I. (2005). *The Dyslexia Screening Test.- Junior*. The Psychological Corporation, London.

Fawcett, A.J. and Nicolson, R.I. (2005). *The Dyslexia Screening Test.- Secondary*. The Psychological Corporation, London



SECTION 4: INTERVENTION

4.1 INTRODUCTION

Nick Catlin and Janet Hoskin

Research has shown that the boys' IQ scores will be about one standard deviation lower. For most boys this is not very significant. They will progress well in school and many will be able to undertake the full school curriculum and sit GCSE Examinations. The main issue then being physical support for which you should contact the Occupational Therapists, Physiotherapists and IT services as the boys progressively lose muscle strength and the use of skeletal muscle.

For some boys the impact of DMD on their learning and behaviour is significant but can be quite subtle and thus difficult to identify. Hence the need for careful assessment.

Many boys will perform poorly on tasks relating to verbal skills. Check Educational Psychologists and Speech and Language Therapists reports for low verbal skill scores in tests. They might only show only a slight delay but this can be significant.

This can impact directly on speech. Boys might be finding it hard to pronounce words or find the right word that they want to say. This can result in communication difficulties and frustration.

Young people with DMD might also find it difficult to understand complex instructions or they simply do not seem to listen. This often happens when boys are very absorbed in one specific task. They don't seem to be able to switch from one task to another very easily. Make sure that everyone who works with him understands this and takes it into account when giving out instructions to the whole class for example. Often this lack of minding what you say can seem to be rude or disinterested. Ask boys to look at you and listen carefully and be prepared to give help with a set of complex instructions.

These problems with verbal skills may impact on developing phonological awareness and therefore cause a delay with learning to read. Unlike the muscle wasting this is not a progressive problem and can be helped with good teaching. It is always better to intervene as early as possible.



4.2 GETTING STARTED – STRATEGIES FOR HOME AND SCHOOL

Nick Catlin and Janet Hoskin

Reading and Spelling

From Veronica Hinton's research (1.4 Questions and Answers, Question 4) we know that many boys with DMD will have difficulty discriminating speech sounds often called phonological processing. This is a similar profile to people who experience dyslexia (sometimes called 'specific learning difficulties').

Sub skills you MUST learn

Writing is a code that is made up of letters that represent sounds. In order to read effectively you have to be able to crack the code. This does not come automatically – certain subskills have to be developed. **Carmen and Geoffrey McGuinness ('The Reading Reflex' Penguin 1998)** list these as the ability to:

- read a group of letters from left to right
- match visual symbols to auditory sounds
- blend discrete sound units into words
- segment sounds into words
- understand that two or more letters can represent one sound
- understand that most sounds can be represented in more than one way
- understand that some components of the code can represent more than one sound

For most of us we perform these subskills unconsciously and automatically when we're reading something. Let's give you the McGuinness's example from Dr Seuss:

This mess is so big so **deep** and so tall,
There's no way to **clean** it up, no **way** at all

Just in this children's extract you have used all the subskills mentioned above. For example you've seen that some sounds like 'th' are represented by two letters, some letters like 'o' can represent two different sounds such as in 'no' and 'to'. The sound 'ee' can be represented by the letters 'ee' and 'ea'. It is possible to store some words in your memory, but most people can only memorise two to three thousand words. In order to read at a higher level using a wide vocabulary it is essential to develop these sub-skills. If you don't believe us, try the McGuinness's 'wider vocabulary' version of Dr Seuss:

This gallimaufry is multitudinously gargantuan, puissantly capacious sand ineffably Junoesque and in consequence of such Protean tribulations and in such psychotic contravention of stereotypical consuetudinary hygiene, there exists the infinitesimal exiguity of a satisfactory resolution to this cataclysmic dilemma.

The 100 high frequency words that the National Literacy Strategy promotes might get your son through Year 1 with flying colours but he will soon begin to flag with increasingly complex reading material or more difficult spellings.

How can we teach these subskills to our children? (The McGuinness Way)

1. Choose some simple 3 letter (CVC) words eg. hat. Break them up into their different sounds and write the letters that represent those sounds onto individual cards /pieces of paper – so you will have one card with 'h', one with 'a' and one with 't'. Have a picture from a magazine or book of a hat and say you are going to make the word 'hat'. Get your child to make the word by putting the sounds together in the correct order. (Pic 1).
2. Now get your child to read aloud the individual sounds and THEN the whole word. (Pic 2).
3. Get your child to write each sound on a small whiteboard or a piece of paper, saying each sound as he writes it. (Pic 3).

The above method is multi-sensory. It is teaching your child to blend and segment sounds by using sight, sound and movement. Our experience of teaching young people with dyslexia/specific learning difficulties is that it is BY FAR the most successful way of doing it.

Pic 1.



Pic 2.



Pic 3.



4. Your child needs to move on to more complex code, understanding that some sounds can be represented by more than one letter eg. 'oa' in 'boat' ('th', 'sh' etc.) – so you would have cards with b-oa-t and a picture of a boat. (Pic 4).



Pic 4.

5. He needs to understand that sometimes the same letters make different sounds eg. the 'ea' in 'bread' is different from the 'ea' in 'gleam'.

6. Sometimes the same letters can make different sounds eg. 'oe' in 'toe' makes the same sound as 'ow' in 'snow'.

In both 5 and 6 the only way you can teach which one to choose is by letting him decide which would make a real word eg. you have to choose 'gleam' because there is no such word as 'glem'.

7. Boys with reading difficulties will need lots of overlearning of these sub-skills. It is often necessary to go back to very easy three letter words to instil the practice of breaking words up into individual sounds so that they can go on to tackle more complex language.

8. Words need to be broken up into syllables. Help the boys to see that longer words are made up of small chunks that contain a vowel. This will help them successfully try much harder words.

Steps 1-8 are crucial to develop the ability to read and spell words. However, at the same time it is really important that boys are kept interested in reading all sorts of material. Vocabulary is a strength for boys with DMD and it is important to play to this and develop their use of words and knowledge through reading to them, museum visits, encyclopaedias, the internet, discussion of TV programmes and of course through the richness of the mainstream curriculum. Don't be scared to read to them when the language is too complex and don't dumb language down when you are talking – it's all important in developing their spoken language which will affect their ability to read it. Listening to story tapes or CDs is a great way to encourage love of stories. Also reading them books of popular films is a way to develop vocabulary eg. Robin Hood, Harry Potter, Lord of the Rings, The Narnia series, Peter Pan etc.

Paired reading (Section 4.3) can help develop fluency and confidence using texts that match interests and vocabulary. However, paired reading methods alone will NOT be enough to learn to read and spell effectively. Steps 1 -8 must be taken.

Young people with reading difficulties do not often come to the table eager to improve their reading and spelling. Children as young as six who have phonological problems already often feel that they are failing. It is our experience that you need to be well organised, clear about what you are teaching and firmly in control when you are working together. Short regular sessions of 20 mins are ideal and be sure to finish on a positive note. It's simply not good enough to say your child 'can't do it' or that they 'get upset' when you try to work with them. It is no good dodging the issue of reading and writing because you feel sorry for them or you don't want to burden them with more problems. You will make them much happier when they feel confident about their ability to read and write. These are key skills for success in school and for the future. Many young people who fail to learn to read and write at 5 or 6 years become frustrated and can display poor behaviour as a consequence. Developing their literacy skills massively improves their self esteem.

If you would like help or advice on working with these multi sensory methods we recommend that you take time to read The Reading Reflex. But please contact us for specialist advice or training. Janet Hoskin janet@ppuk.org or Nick Catlin nick@ppuk.org



Maths

Many people who experience dyslexia-type difficulties also have problems with mathematics. In particular, one to one correlation, sequencing, directional and spatial awareness, use of short term or working memory eg. Holding numbers in your head and doing mental calculations, understanding mathematical language. It is important to make mathematical problems as concrete as possible eg. Using money/ bricks for number work. There has been very little research specifically on DMD boys in this area. A mathematical assessment may help to indicate strengths and weaknesses. Unicorn Maths (www.unicornmaths.com) provides excellent training and resources for children with dyscalculia.

Secondary School

Boys who are still experiencing difficulties in secondary school should be offered further assessment and diagnosis of learning needs. They might need to revisit the reading and spelling programmes outlined above and be offered specialist support. Study skills will be important as boys need to be able to tackle new material independently without panicking. Techniques such as highlighting key words, looking for sub-headings, understanding exactly what questions are asking you to do are all important skills here.

Boys will be given time concessions for examinations called 'access arrangements'. Boys who have problems with reading or comprehension may qualify for a reader. Slow writing may mean a word processor or scribe could be used. Many boys will also qualify for rest breaks and extra time. Make sure you discuss the child's specific needs with the SENCo. We recommend '**Dyslexia: Assessing the need for Access Arrangements during Examinations. A Practical Guide Second Edition**' by Gill Backhouse, with Elizabeth Dolman and Caroline Read published by PATOSS

In further and higher education young men should qualify for extra help both with physical but also learning needs eg. Helping with planning of assignments and dissertations etc.

4.3 PAIRED READING

Gill Backhouse

Reading is a complex skill and the only way to improve it is by regular practice. When a child's reading does not improve steadily, the daily reading routine becomes an unwelcome chore, often leading to high levels of frustration and anxiety for all concerned. Paired Reading is a well-proven method of promoting reading skill in a stress-free way.

The theory

Skilled readers recognise a very large proportion of the words they encounter in print. These make up their *sight vocabulary*. How does the SIZE of the sight vocabulary increase? For each word encountered, but not recognised, the reader may:

- use phonics to try and sound it out; and / or
- guess / predict what it is likely to be. This is called *reading for meaning*.
- be told the word by a helper. This method is called 'Look and say'.

The first two strategies may or may not be successful. They interrupt the flow of the reading and the



sense of the text may be lost. The third method, however, is ALWAYS successful - assuming the helper is an accurate reader!

Paired Reading builds up the sight vocabulary by the Look and say method. Considerable gains in confidence and lessening of anxiety about reading are frequently associated benefits.

The Method For Infant / Primary pupils

Choose a well-illustrated book - fact or fiction. Make sure it is written in language that the child understands. Ideally, he should be able to read 90 - 95% of the text unaided, but where reading ability does not match age/interest level, this can be difficult. It is vital that the child **enjoys** the book, so it is usually better to choose one that is interesting or funny, but a little too hard, than one which is easy but boring.

Look at the book together. Discuss the title and the 'blurb' on the cover; then thumb through the pages, pointing out the names of the main characters, looking at the pictures, and outlining the story. This aids *reading for meaning*. Now start to read the book properly. **Read a short section to the child**, whilst running your finger under the print - so that he is actually looking at each word as he hears it. A 'short section' means somewhere between a sentence and a paragraph or two. This will depend on the age and ability of the child.

You and the child should now read the SAME passage ALOUD together - perhaps he can point to the words by this stage.

Now, the child should read the SAME passage ALOUD by himself. If he hesitates (allow 3-4 seconds), or makes a mistake, **tell** him the word or phrase straight away, encourage him to **repeat** it and then carry on.

Continue reading in this way, section by section, discussing the content at appropriate intervals, to ensure comprehension.

The child has now read all the words he didn't know, correctly, three times in quick succession. His sight vocabulary will gradually become larger. You will find that long/unusual words (e.g. aeroplane, dinosaur) are learned more quickly than small 'easy' ones that look a bit like several others (e.g. sharp/ sheep/ sleep).

As the child becomes more confident and skilled, he may prefer to leave out stage 4 or to do Shared Reading with you. Here, helper and child read alternate passages, with the helper supporting, as in stage 5.

The Rules

Establish a routine - a special time and place for reading. A little each day (say 10 minutes) is much better than a lot once a week.

Be positive and patient. An encouraging atmosphere is vital, particularly if the child is anxious or frustrated about his reading.

SHOW that you are pleased with the child for trying to read. PRAISE whatever aspects of his reading behaviour you can - for example, reading with expression; getting lots right; reading long or difficult words; intelligent guesses; being interested in the story; not wriggling; or whatever!

Keep a record of time spent reading and reward (merit points / stickers etc.) for each 10 minutes or whatever is appropriate for the child. Your child's class teacher will probably



have provided a reading record book already, so the efforts made at home can be acknowledged at school too.

NEVER imply that a child could do better, either by word or facial expression (watch your eyebrows!) Do **NOT** tell him to “Sound it out”, “Break it up”; “Look at it again”, or “Think carefully”, or point out that he knows a word because he read it on the page before. **Just quietly tell him what it is, time and time again, for as long as necessary. Total reassurance and support is the order of the day.**

Keep up the momentum of **daily** practice for at least the first 6 weeks.

Personnel

The person ‘pair’ reading with a child must be trained to adhere to the method and have the ability to remain calm, positive and encouraging at all times. He/she must also have the time to read with the child every day. Clearly, the person best placed to do this is a parent or other family member. In school, parent volunteers, welfare assistants and older pupils can also train in paired reading techniques. The child can also pair-read with a taped or CD-ROM version of a book, or act as helper to a younger child (very good for self-esteem)



4.4 ADVICE FOR SENCOS AND SPECIALIST TEACHERS

Nick Catlin and Janet Hoskin

It is possibly the first young person with Duchenne Muscular Dystrophy that you have met. Your role in assessing the needs and supporting the young person and their family is crucial to their success in school. We recognise that much of what we are advocating here will be in place in many SEN Departments in schools. But often SEN staff might miss learning and behaviour problems associated with DMD as it is largely seen as a problem relating to a physical disability. It is vital that you play the key role in ensuring that all assessments have been completed as recommended in the toolkit (Section 3) and that you are able to bring together the key professionals (in particular the Educational Psychologist, Specialist Dyslexia or SpLD teachers and Speech and Language Therapists) to provide advice and support to school staff.

Make sure that professionals provide guidance for clear teaching and learning programmes that should be designed following their assessments and observation sessions. Good classroom teaching in the early years with extra LSA support can provide an excellent foundation for boys with DMD to begin to overcome problems with reading and spelling. If boys are withdrawn from lessons (and many will need some intensive early input) we strongly recommend that this must be for short periods of 1:1 teaching of specific skills (See Sections 4.2 and 4.3) or multi-sensory programmes designed by you or other support teaching staff. Make sure that you have some measures of progress and at the very least ask for regular reassessments of reading and spelling ages for the Annual SEN Review. Please try to involve parents and family members in these literacy programmes and offer training and support.

We have assumed that the school is catering for the young persons physical difficulties here and they will have full access to the school campus. Schools must now comply with the SEN and Disability Act (2001) which amended the Disability and Discrimination Act (1995) <http://www.dfes.gov.uk> and have already adapted buildings for those students with a physical disability or have plans in place to make these changes. We strongly recommend that the school undertakes a Risk Assessment audit that ensures measures are in place to minimise risks young people with a physical disability might encounter

around the school. Seek advice and support from Occupational Therapists and Physiotherapists in determining risk and helping to enrich the school curriculum for these young disabled people.

We also hope that schools can be encouraged to keep young people with DMD in mainstream education as we firmly believe that this is where they will benefit from the full curriculum of mainstream schools and being included and educated alongside their peers.

Checklist

- Interview parents and note any concerns with speech, language, behaviour or difficulties with reading and spelling. It is very important not to be judgemental about these parental concerns. Do not dismiss them lightly or overemphasise problems that are flagged up. These are very important observations that can help with diagnosis and future learning and behaviour programmes.
- Observe young person's learning and behaviour for yourself in class
- Check to see if a full Educational Psychologist's assessment has been completed (Section 3.3)
- Check to see if a Speech Therapist's report has been completed (Section 3.4)
- If not ask your Educational Psychologist and SLT for a full assessment. This should include the full battery of tests and in- school observations.
- Has the young person ever been referred to a behaviour support teacher or autism service? (Section 5)
- Boys with DMD should have a SEN Statement to meet their physical needs. In most cases the SEN Statement must also identify learning, speech and language and behaviour issues
- Boys will need in class support. In most cases this will cover a **minimum** of 50% of the school week
- Most boys will need SHORT periods of 1:1 withdrawn teaching sessions using a well designed multi-sensory programme for reading and spelling (Section 4.2)
- It is vital that Learning Support Assistants are properly trained in working with students with specific learning difficulties (dyslexia) (Section 4.5)
- All DMD boys should have an IEP that identifies specific and measurable targets. These should be reviewed termly with staff and parents.



4.5 ADVICE FOR LEARNING SUPPORT ASSISTANTS

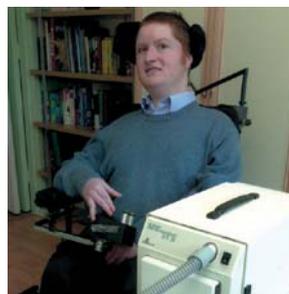
Nick Catlin and Janet Hoskin

Your role in supporting young boys with DMD in the classroom is vital to both their educational achievement and success in maintaining good social relationships. It is important that you are able to know when **not** to help and intervene. Let boys make mistakes both in terms of their academic work and in relationships with peers. They have to do everything that normal classmates do in a lesson or in the playground. It is very important to encourage independence.

But you will be able to build a close relationship and be able to offer help with well designed programmes to support reading and spelling.

Generally make sure that you

- Have been trained to help with 1:1 reading and spelling games and especially those to help with blending and segmenting phonemes for younger boys
- Have been advised on any support needed with behaviour including those within the autism spectrum
- Speak to all school staff to make them aware of planned strategies to deal with behaviour difficulties
- Have been trained to offer paired reading support (Section 4.3)
- Give instructions clearly to boys and that they can understand what the teacher is expecting them to do
- Give boys plenty of space to make mistakes
- Support in class and make sure that boys understand the meaning of and spell key words in lessons eg photosynthesis in Science
- Help the boys to develop knowledge and vocabulary from the lessons
- Help boys with comprehension type questions and how to formulate answers
- Get the boys to organise themselves with essential school equipment, timetables, diaries, laptop, homework, lunch, getting to the toilet etc. They must learn to cope themselves with you in the background as a pair of hands when needed
- Are involved in monitoring targets for a boys Individual Education Plan and advise the SENCo on new learning and behaviour targets at assessment meetings
- Develop a good relationship with the boys. Many of the boys have a very good vocabulary and varied interests that they will be keen to talk about.



SECTION 5: BEHAVIOURAL CONCERNS

Angela Mijovic, Jeremy Turk, Veronica Hinton and James Poysky

Not all boys with DMD will have behaviour difficulties. In fact, research has demonstrated most boys surveyed had a very healthy attitude towards life and do not necessarily get depressed by their condition; younger boys wish for the same things their peers wish for (toys and trips to fun places) and older individuals on ventilator support report better quality of life than their caregivers think they have. Parents have reported to PPUK that their boys have a remarkable ability to want to get on with life and have the usual needs and interests of any group of young people their age. However there is evidence that young people with DMD are more likely to experience behaviours associated with the autism spectrum than other young people within the general population. Children with DMD may also have attentional deficits and, like all children with chronic illness, are at increased risk for developing mood disorders. It is important to discuss carefully with families just how boys are behaving and try to suggest positive interventions.

5.1 PERVASIVE DEVELOPMENTAL DISORDERS

Children with DMD are at increased risk for having “*pervasive developmental disorders*,” or “*autistic spectrum disorders*.” There is a greater than expected association of autism with DMD, yet most children with DMD are NOT autistic. There is no laboratory test for autism - the diagnosis is established by clinical findings during interview with child and family, and through direct observation of and interaction with the child. The core features of these pervasive developmental disorders consist of

- qualitative impairments in socialisation
- impairments in verbal and nonverbal communication
- restricted and repetitive patterns of behaviours.

Various diagnostic interviews are used by professionals to help arrive at diagnosis. Two commonly used ones are the Autism Diagnostic Interview (ADI) and the Diagnostic Interview for Social & Communication Difficulties (DISCO). These are detailed questionnaires which require trained professionals to complete. Autism and Asperger Syndrome are both autistic spectrum disorders, although diagnoses of Asperger Syndrome are usually given to individuals with higher levels of intellectual functioning and good expressive language skills.



Children with DMD often have some mild behaviours that are consistent with the diagnosis, but are not sufficient for it. Those who do meet criteria for autism spectrum disorders may not be recognized until after school commences when their multiple differences from other children become obvious. They may be perceived as “odd”, “quirky”, “unusual”, “socially aloof”, “eccentric”, “like a little professor”, or “lacking common sense.”

Qualitative impairments in socialization

These depend upon the child’s age, developmental level and how severely they are affected. Children with autism often tend to ignore other people or appear insensitive to others’ needs, thoughts or feelings. They seem to be unable to impute independent mental states to others and have great difficulty in expressing and understanding emotions. They tend not to make the usual eye contact or use appropriate facial expressions or other normal “non-verbal communications” such as changes in facial expression, gesture and posture in social situations. They are less likely to use gestures such as pointing to communicate. They tend to find it difficult to cooperate, share or take turns with others. They usually prefer to play alone and show little interest in imaginative play. They get on best with

understanding adults. Socializing with other children and forming friendships is hard for them.

Impairments in verbal and nonverbal communication



Not being able to communicate properly is a particularly disabling aspect of autism, and often the one that first causes concern. Nearly all affected children have language problems - both in understanding ("comprehension") and in speaking ("expressive language"). More severely affected children may never learn to speak or to communicate in other ways. If they do begin to talk, children may simply echo what they have just heard or may speak in other odd ways.

Restricted and repetitive patterns of behaviours

Children with autism or Asperger syndrome prefer familiar routine and tend to resist change, which they find difficult and unpleasant. They may also have unusual interests, such as maps or electronic gadgets. They may be very sensitive to sounds, tastes, smells, certain sights and textures such as tight or irritating clothing. They may also have odd body movements such as hand-flapping, finger-twiddling or body rocking. Any attempt to stop these activities and interests can cause furious protest and distress (see Section 5.4 Challenging Behaviour). When upset, these children may have tantrums or injure themselves. Conversely, some children have special talents or gifts for drawing, music or calculation.

Many more children with DMD might have a number of **autistic tendencies** without reaching the diagnostic thresholds for autism or Asperger disorder. They might be better described as having social and communicatory difficulties of a neurodevelopmental nature similar to those witnessed in autism. Increasingly they are described clinically as having an "autistic spectrum disorder".

The evidence base for treatment in autism is limited. No treatment will lead to a cure. However, psychoeducation i.e. information about autism, its causes, prognosis and possible intervention strategies is probably the essential part of any treatment for autism, and special educational input focusing on the triad of impairments described above is known to be beneficial.

Children with autism and an IQ of 70 or under will require specialist programmes to meet their needs. Students with Asperger syndrome will benefit from attending a mainstream classroom, but may often need extra support. The most important aspects pertain to the knowledge of autism on the part of the teachers. All children with autism, Asperger syndrome or autistic tendencies need a considerably greater amount of structure, predictability and routine, and systematic, well planned skills training than is usually required educationally.

Many pharmacological (drug) treatments are available for symptoms associated with autism like attention deficit hyperactivity disorder (ADHD), epilepsy, behavioural outbursts, mood difficulties, obsessive-compulsive problems and sleep issues. Advice from highly specialised child and adolescent psychiatrists is required in these circumstances.



5.2 ATTENTION DEFICIT DISORDERS

Children with DMD may be at increased risk for presenting with attention deficit disorders. Attention deficits are usually first diagnosed during primary school years. Sometimes, symptoms continue into adolescence and even adulthood. A child with Attention Deficit Hyperactivity Disorder (ADHD) has

problems paying attention and concentrating and/or with hyperactivity and impulsive behaviour. Despite good intentions, a child may be unable to listen well, organize work and follow directions. Cooperating in sports and games may be problematic. Acting before thinking can cause problems with parents, teachers and friends. These children may be restless, fidgety and unable to sit still. Problems related to ADHD appear in multiple areas of a youngster's life and can be very upsetting to the youngster, the family, and people at school. For children with DMD, it may be difficult to disentangle language deficits and limited social skills from attentional deficits, as all may present as a child who is unaware of the world around him.

Treating ADHD includes psychological managements comprising psychoeducation, improving parenting skills, behaviour therapy, anger management and medication. The most frequently used and best-researched medication is methylphenidate (Ritalin, Equasym, Concerta), which is an extremely safe intervention with a response rate of 70-80%. Side effects such as insomnia, poor appetite, headaches, tummy aches, dizziness, nervous tics and emotionality may limit its use in a few individuals but in these instances other beneficial medical and psychological treatments are available.

5.3 MOOD DISORDERS

It would be expected that as boys with DMD get older and begin to suffer the consequences of severe muscle wasting they risk gradually becoming more socially withdrawn, with low mood and social isolation. Children with progressive and chronic illness have been shown to be at increased risk for developing depression. Research of children with supportive families suggests that most boys with DMD do not have significant signs of depression. It is important, however, to consider during an assessment that boys may be experiencing mood disorders. Feelings of depression can be more powerful and unpleasant than the short episodes of unhappiness that we all experience from time to time. Here are some symptoms that are suggestive of a young person feeling depressed:

- loss of enjoyment and interest in life
- fatigue or loss of energy
- irritability, restlessness, and/or agitation
- difficulty in making decisions
- feeling guilty
- loss of appetite and weight (some find they do the reverse, eat more and put on weight)
- difficulties getting to sleep, and then waking up earlier than usual
- loss of self-confidence and self-esteem, feeling worthless
- negative view of the future or inability to think about the future
- poor concentration and memory
- tearfulness
- suicidal thoughts

Sometimes, particularly in children, physical symptoms or irritability can be the first signs of depressed mood. These symptoms can cause considerable distress and interfere with the child's everyday life. About 1 in 10 people who suffer from depression will also have periods when they are elated and overactive.

The general demeanour of boys with DMD (limitation of facial expression, gesture and general animation secondary to widespread muscle weakness) may make diagnosing depression challenging. Therefore it is important to establish the diagnosis based on the presence of symptoms rather than the appearance or interaction of the boys at interview. A specialist child psychiatric opinion may be useful.



Treatment

Most people with depression are treated successfully with some form of “talking” therapy, antidepressants or both. 4 out of 5 people with depression will get completely better without any help and this usually takes 4-6 months. However, treatment is important as it shortens the duration of depression and prevents:

- the recurrence of illness
- the risk of suicidal thoughts or even actions

Research with children with chronic illness has shown that children’s reactive responses to their condition are closely associated with their parents’ coping skills. As such, family therapies may be a useful means in helping all family members cope with the myriad of difficulties associated with DMD.

Although behaviour problems can occur at any age, they are typically more prevalent at a younger age. In many cases, there are multiple factors that are contributing to the behaviour problems.

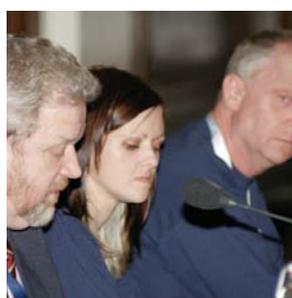
5.4 OPPOSITIONAL-DEFIANT BEHAVIOUR

Many, but not all, children who demonstrate significant oppositional behaviour have problems with behavioural regulation. More specifically, they may be impulsive and have significant difficulty “putting the breaks” on their behaviour and thinking before they act. In addition, in the same manner in which they have problems controlling their impulses, they may also have difficulty controlling their emotional reactions. Impulsive children typically become frustrated, impatient, and angry more easily than other children their age, and have a difficult time defusing their anger. Similarly, these children have a difficult time learning from their mistakes, and typically do not respond as readily to punishment as other children. Cognitive factors may also contribute to oppositional behaviour. Children who are more disorganised and easily overwhelmed may react in an oppositional manner. In addition, oppositional and argumentative behaviour may be the result of problems with mental flexibility or the ability to be adaptive in their thinking. They are often described as “stubborn” or “hard headed”, and may miss the bigger picture or have difficulty taking another’s perspective. Impulsivity and cognitive factors are typically considered to be biological in origin, but other factors that are more psychosocial in nature can contribute to these behaviour problems. Some children may learn that they can get what they want if they put up enough of a fight. In addition, emotional distress such as depression or anxiety can cause a child to become irritable and oppositional. There is also the possibility that a child with a progressive neuromuscular disease may feel as if they have little control over their health and self-care. As a result, they may attempt to gain control in other areas of their life by employing commanding or oppositional strategies. However, it should not be assumed that these strategies are purposeful or consciously employed.

5.5 AGGRESSION

Problems with behavioural regulation (as mentioned above 5.4) are often the antecedents to aggressive behaviour. In addition, cognitive factors likely play a role. In this manner, aggression is more likely to occur when the problem solving demands of a certain situation exceed a child’s ability level. More

specifically, a social conflict develops and the child is not able to control their emotions/impulses



and/or generate solutions to the conflict. In addition, aggression may also be “learned”. A child may learn that they get what they want if they are aggressive (as in the case of schoolyard bullies). They may also learn about aggression from things that they observe, such as from the media or from older brothers. In some cases, boys may learn about aggressive behaviour from parents or other adults in the community.

Comprehensive assessment is crucial in the treatment of behavioural problems. While analysis of parenting strategies is helpful, it is important to understand that behaviour problems are not always the result of “bad parenting”. Many children with behaviour problems do not respond to “typical” parenting strategies. Behavioural therapy/interventions may assist with certain aspects of behaviour. However, when biological factors such as poor behavioural regulation and cognitive deficits are prevalent, pharmacological and/or other interventions may be necessary before the child is able to benefit from the behavioural therapy.

5.6 PARENTS’ PRACTICAL STRATEGIES FOR MANAGING BEHAVIOUR

Nick Catlin and Janet Hoskin

- Make sure boys are looking at you when you give instructions
- Don’t give too many instructions at any one time
- Encourage boys to be well organised and prepared for tasks
- Thoroughly prepare boys for new situations by explaining what is likely to happen and what is expected of them i.e. going into assembly, going on a school trip, starting a new class, meeting family members, going shopping. Explain what will happen at these events and what you want them to do. Ask yourself does he really understand what I mean when I say “I want you to be good”. Better to say before visiting the supermarket “I want you to help me push the trolley and choose the fruit that we need”. Tantrums and outbursts will happen (usually when you are in a rush or feeling stressed) but try to remain positive and not worry too much about what you think other people might be thinking.
- Give boys clear boundaries and expectations for behaviour and give them lots of praise when things are going well
- If your son is diagnosed with an autism spectrum disorder make sure that you are given specialist advice and support
- Boys may have problems with mental flexibility. They may be rigid in their thinking and have a “one track mind”. As such, they may have trouble with transitions, changes in expectations, or become easily frustrated if things don’t go their way. In these situations, several things may help: 1) explaining why you have said, “No”, rather than merely expecting they will follow your instructions. 2) Helping them problem-solve a compromise (“We can’t do that now, but if you follow my instructions we can do it later today”, etc.) 3) Provide advance warning of changes or transitions, E.g. “five minutes until...”



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Resources

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Toe by Toe Multi Sensory Reading Manual for Teachers and Parents Keda and Harry Cowling www.toe-by-toe.co.uk

Ruth Miskin Literacy Read Write Inc www.ruthmiskinliteracy.com

Jolly Phonics by Sue Lloyd Sarah Wernham and Christopher Jolly www.jollylearning.co.uk

Usborne First Readers, Usborne Little Encyclopedias www.usborneonline.org



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Contact Us:

PPUK
Epicentre
41 West Street
London
E11 4LJ
02085569955

Nick Catlin (Parent and CEO) 07920723490 nick@ppuk.org

Janet Hoskin (Parent and Director) janet@ppuk.org

JOIN THE DMD REGISTRY TODAY!

The DMD Registry provides a National Duchenne database of boys' gene variation information and other essential medical information for clinicians, health professionals and researchers to help accelerate the development and delivery of new treatments for Duchenne and Becker Muscular Dystrophy.

"We are moving towards very exciting times when clinical trials are becoming a reality. DMD is a rare disease and any trials of drugs are going to require recruitment of patients from a wide area which otherwise could be a bottleneck hindering the progress of research. The DMD Registry will benefit researchers and clinicians identifying new targets for drug trials and will accelerate the progress of this research."

Professor Kate Bushby Action Research Professor in Neuromuscular Genetics University of Newcastle.

By registering you will gain access to key genetic and medical information currently stored by the NHS about you or your child. This information will help you to make choices about becoming involved in future research or clinical trials. Once you have registered we will inform you of the progress of the latest research and let you know of any new treatments for DMD.

To register go to the website www.dmdregistry.org or phone Atia on 0208 5569955 for a Registration form.



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020 8556 9955
charity no: 1101971

