Duchenne muscular dystrophy - learning and behaviour

Duchenne muscular dystrophy is a condition that causes
- muscle wasting - in all those affected
- changes to learning ability – in all, to some degree
- changes to behaviour – for some
- Mean IQ one standard deviation below average but most children are still within normal limits. For most children, the affects on learning will mean only a slight change in overall ability but for some, the effects will be much more significant.

Difficulties often reported in children with Duchenne muscular dystrophy
- Early speech and language problems
- Not seeming to understand complex instructions
- Poor or fleeting eye contact
- Falling behind peers with reading and spelling
- Difficulty coping with a change in routine
- Higher than normal level of tantrums or aggressive outbursts
- Very passive behaviour, difficulty communicating, socialising and making friends.

When tested against able-bodied siblings
No difference in skills
- Vocabulary
- Visuospatial skills
- Rote memory – verbal & visual
- Abstraction/ conceptualization

Regardless of IQ, performed more poorly in
- Digit span
- Comprehension
- Story memory
- Token test

This indicates
- Good single word comprehension

but
- Particular difficulties in comprehending complex verbal information
- Problems following ‘strings’ of verbal information
- Use of visual clues, pictures and routines can be helpful in overcoming these difficulties.
Non-progressive
Understanding complex language seems to be a delay rather than a deficit and with correct support the children are usually able to ‘catch up’.

Contributing factors
On testing, lower academic achievement scores than their siblings. Many factors could contribute

- fatigue
- mood
- physical ability
However, research showed decreased verbal span to be more of an influence than any of the other factors.

Slow language development – common difficulty
Once communicative speech develops

- know factual information
- have vocabulary
- problem solving skills
- on a level with peers

but

- comprehension likely to be comparable to a child two years younger, therefore more difficulty following verbal instructions
- Many children have poor reciprocal social language.

Phonological processing skills
Tests looking at estimates of intellectual function showed no differences between children with Duchenne muscular dystrophy and their siblings, however, the children with Duchenne muscular dystrophy did more poorly on

- phonological awareness
- phonological memory
- rapid naming
Indicates difficulty with phonological processing skills. Processing speed and complex attention may also be problematic.

Poor phonological processing/comprehension
In up to 40% - this combination of difficulties likely to cause problems with

- reading
- spelling
- maths
Specific advice will be needed.
Similar difficulties to those found in children with

- dyslexia
- reading disabilities
- strategies used for these children may be helpful
- better at sight reading than ‘sounding out’ words.
Self-esteem
Physical weakness means
► inability to keep up with their peers in the playground
► inability to be good at sports
If also experiencing difficulties
► processing what is being said by teachers
► learning to read and decode speech sounds
► in the classroom
this is doubly damaging to their self-esteem and may lead to frustration

How can we help?
Remember that problems are likely to be due to the condition. There are areas where these children do not usually experience difficulties
► build on these strengths
► help alleviate learning related frustrations
► rote learning is generally good
► repetition
► routines

Behaviour
Particular behaviours reported (by parents) are
► immaturity
► attention problems
► more social problems than unaffected siblings
Could be arising because
► not understanding as much as other children their age
► inability to follow complicated instructions causing decreased attention.

Autistic spectrum disorders
Research has shown that a higher than expected number of children with Duchenne muscular dystrophy score at a level suggesting autistic spectrum disorders (ASD) than in the general population.

The type of ASD qualities
► Failure to use nonverbal behaviours for social interactions (poor eye contact, lack of social smiling)
► Failure to develop peer relationships (lack of interest in children)
► Lack of shared enjoyment (not directing other’s attention to things of interest)
► Delay in spoken language
► Lack of varied, spontaneous make-believe play
► Lack of reciprocal conversation
► Circumscribed pattern of interest
► Compulsive adherence to rituals
► Restricted, repetitive and stereotyped patterns of behaviour were seen in some children, but this is not the predominant characteristic
It is thought likely that these issues will improve with age.
Other issues
Intellectual function within normal limits.
Specific difficulties noted in:
- the ability to identify affect and interpret facial expressions. This may lead to social interaction difficulties and behavioural issues so support and intervention should be provided to assist with this.
- switching from one task to another, particularly if they are absorbed.

This may appear as not seeming to listen or being rude and disinterested.
- Attention Deficit Hyperactivity Disorder (ADHD) reported by 12-24% of parents.
- These children tend to have poorer psychosocial adjustment.
- Remember the emotional responses of other family members are likely to have an effect on child's behaviour.

Recommendations
- Full assessment required
- Problems may not be obvious from a ‘quick look’.

Practical tips
- be aware of the problems
- be consistent
- when giving instructions, ask the child to look at you
- break down complex instructions – be clear and concise
- break down tasks into steps
- encourage children to be well-organised and prepared for tasks
- encourage routines and provide structure
- thoroughly prepare children for new situations
- give clear boundaries and explain what is expected of them
- give praise when things are going well
- explain the reason for a decision
- try to provide a compromise
- provide advanced warning of changes e.g. five minutes until...
- encourage early phonics learning
- ensure the teaching of subskills to assist reading and spelling
- use visual clues.

Learning and behaviour
- Evidence is based on children with Duchenne muscular dystrophy as a group
- Indication of the types of difficulties that these children may be at risk of
- Not all children with Duchenne muscular dystrophy will have significant problems

Each child is unique, individual assessment is vital and specific interventions should be tailored accordingly.

What is Duchenne muscular dystrophy? - Summary
Complex genetic condition
- affecting learning ability
- possibly affecting behaviour
these areas are non-progressive and can be improved with support
causes severe physical disability
life-limiting
effects can be slowed by medical/therapeutic interventions.

Useful literature
▶ An Introductory guide for families with a child newly diagnosed with Duchenne muscular dystrophy
▶ Duchenne muscular dystrophy – a guide for families with a child aged 5-12 years
▶ Duchenne muscular dystrophy – the Teenage years
▶ The Diagnosis and management of Duchenne muscular dystrophy – a guide for families
▶ Everybody’s Different, nobody’s perfect!
▶ Same but Different
▶ Hey, I’m here too! (for siblings/classmates)
▶ Behavioural issues in DMD
▶ Inclusive Education
▶ Guide to Transition

All available free of charge from the Muscular Dystrophy Campaign www.musculardystrophy.org
PPUK Learning and Behaviour toolkit - www.actionduchenne.org

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Here for you
The friendly staff in the care and support team at the Muscular Dystrophy UK’s London office are available on 0800 652 6352 or info@musculardystrophyuk.org from 8.30am to 6pm Monday to Friday to offer free information and emotional support.

If they can’t help you, they are more than happy to signpost you to specialist services close to you, or to other people who can help.

www.musculardystrophyuk.org