BEHAVIOURAL ISSUES IN DMD

INTRODUCTION

It is well documented that some boys with Duchenne Muscular Dystrophy (DMD) can experience behavioural and learning difficulties. Various psychological, physical and social factors (such as needing to use a wheelchair) will affect a boy and his family at different periods in his life. This can sometimes lead to problems with behaviour, about which parents frequently seek guidance and support. The use of steroids in the management of Duchenne has also been associated with behavioural problems.

Parents can sometimes experience difficulties in managing their child’s behaviour. This can be linked to their own continuing grief following the diagnosis and to difficulties in establishing consistent routines at a time when parents find their world has been turned upside down.

This factsheet has been designed to give parents and carers some simple strategies for managing their child’s behaviour more effectively but is not meant to be a substitute for more specialist help.

WHAT ARE BEHAVIOURAL PROBLEMS?

These may present as;

- Moodiness, low mood, refusal to cooperate
- Bad temper, temper tantrums, screaming, swearing
- Frustration, throwing objects, hitting, biting
- Demanding behaviour, repeated requests until they get their own way

All of these behaviours in themselves are not unusual, it is only when these behaviours become more extreme or are affecting family life that they would be considered to be a problem.

AUTISTIC SPECTRUM DISORDERS & ADHD

A commonly held belief is that children with behavioural problems have some form of autism or hyperactivity. This is not usually the case.

Most children with DMD are not autistic; however autism does appear to be more common in children with DMD compared to their peers, which we do not yet fully understand. Autism is characterised by a reduced ability to communicate with others and repetitive behaviours. There is no cure for autism, but specialist help can provide strategies and interventions that will benefit autistic children.

Attention Deficit Hyperactivity Disorder (ADHD) can affect boys with DMD. This disorder causes the child to have difficulty paying attention, poor concentration, impulsive and hyperactive behaviour. Children with ADHD may require help from an educational psychologist, medication and/or behaviour therapy.

DIAGNOSIS AND EARLY CHILDHOOD

The early years of childhood for parents of a child with DMD can bring joy and pleasure as they see their child develop and achieve their milestones. This can be tinged with sadness because of the recent diagnosis. Some parents may doubt the diagnosis as they see their child develop or they may deny that anything is wrong. This is not unusual and for some parents this is their way of coping. Parents may be particularly vulnerable at the time of diagnosis and it is important that they are given
accurate information and support. It is possible that parents may feel overwhelmed as a procession of doctors, nurses, physiotherapists, social workers etc offer their help in the first few months following diagnosis. Others may find this helpful or feel they need even more support.

Parents can experience an overwhelming need to protect their child and may feel unable to discuss the condition with them or want to hide the diagnosis from them. Relationships within the family can become strained. There can be feelings of guilt and blame as DMD is a hereditary condition, though of course there is nothing that parents can do to change genetics.

It can also be a difficult time for siblings as there is increased attention given to the affected child. They may experience feelings of guilt that they have escaped the condition or envy of the attention he receives. There may also be a fear of “catching” the condition. It is important that brothers and sisters understand the condition, and feel involved.

At diagnosis, affected boys often have a very limited understanding of their diagnosis due mainly to their young age. However they will see their families showing signs of grief and upset and they can become anxious when they see this.

Parents often find it difficult to set boundaries as they normally would for a child, as a result of changes in the family routine, or feeling the need to ‘overprotect’ their child and so giving in to the child’s every request. This can lead to confusion and compound the child’s anxiety. This can be displayed as anger or challenging behaviour, such as tantrums.

**MANAGING BEHAVIOUR**

Behavioural problems in a child do not mean that you are a “bad parent”.

Be consistent. Give your child clear boundaries and expectations which you have agreed with your partner. If you don’t agree and you are both giving different messages to your child they will end up feeling confused. They will also learn to play one parent off against another.

Give your child lots of praise when things are done well. We often forget to praise.

Make clear (with a firm “NO”) which behaviours are unacceptable.

Give your child clear instructions. Rather than saying “I want you to be good”, explain exactly what you mean, eg “I would like you to help me by…”

Children can become frustrated when they are unable to do something. This can lead to anger, tears and temper tantrums. Recognise the situations when they become frustrated and avoid them if possible. If you are unable to, try to find ways to alleviate their frustration, for example helping them with a task (but not taking over), using aids to enable them to achieve their task or finding a compromise.

Tantrums inevitably happen when you are rushing and feeling stressed. A classic example is when supermarket shopping. Try to stay calm (easier said than done) and try not to worry about what other people think. However you handle the situation there will be somebody who “tut tuts”. But there will also be people who are sympathising.

There are several strategies for dealing with tantrums:

You could try distracting your child or simply removing them from the situation.

There are some schools of thought that you should allow the child to continue with their tantrum and to ignore them or walk away (not easy in a supermarket!)

Others believe that holding your child tightly in an embrace during the tantrum will calm them. (Although you may also find that it makes them fight harder).

If these strategies do not work and the child’s behaviour becomes uncontrollable it may be best to leave well alone. Going too close may cause an accidental injury. It may be better to weather the storm until the situation calms down.
When the situation calms and the child has completely returned to normal then discuss the consequences of their behaviour. If the child has not calmed completely and you prematurely impose consequences you may inadvertently cause the child to become disruptive again.

Try not to give too many instructions at one time. Rather than saying ‘go upstairs, get washed and bring your shoes down with you’, give the instructions in smaller chunks.

Make your instructions clear.

Encourage your child to be prepared and organised.

Some parents find that the use of star charts recording good and bad behaviour can help to reinforce good behaviour.

Try not to take rude or aggressive behaviour personally; and recognise that you may not be the target for the child’s anger and it reflects their anxiety about something.

Set the child the same boundaries you would set for their brothers and sisters.

IT IS IMPORTANT TO REMEMBER THAT YOUR CHILD IS A NORMAL BOY WHO HAPPENS TO HAVE WEAK MUSCLES.

TALKING TO CHILDREN

We would recommend that families are honest with their child. Children can be confused at this time, not understanding why their family is upset. Answering their questions honestly will help with their understanding and in turn can improve their behaviour.

It is likely that your child will ask these questions at time when you least expect it.

Don’t panic and be honest

You can answer the question with a question. (This allows you to establish exactly what the child is asking and gives you a chance to catch your breath and think what you are going to say).

Usually a child is looking for a simple answer. Many parents fear that when their child starts asking questions that they will have to explain the diagnosis and prognosis in detail. This is not usually the case. Answer exactly what they asked you in a way that they can understand for their age.

Questions young boys may ask

Why do I have to go to hospital?
What is wrong with me?
Why can’t I run as fast/as far as my friends?
Why do I get cramps in my legs?
Why are mummy and daddy so upset?

Example Why can’t I run like my other friends?
Answer “You have poorly legs” or “you have weak muscles”.

This helps to give the child some understanding of what is happening to them.

Children tend to accept information in a matter-of-fact way and usually without being traumatised. Parents often ask “If I tell them about the condition how will they cope?” Remember that at this stage your son is far more interested in his latest toy, computer game etc than in his diagnosis. It can be helpful if you give an example of another child at school/nursery that wears glasses for poorly eyes, or needs to use an inhaler etc.

Things that we would discourage
• Giving false information to your child to try to make them feel better, such as “You will get better soon” or “these exercises will make you better”.

• Not giving an answer at all. No answer is in itself an answer and may lead a child to worry more inwardly, never having the courage to ask again. If parents do not talk because they are too hurt themselves or feel they are protecting the child, then the child may not talk because they know it upsets their parents.

INCREASING AWARENESS

As the boys become older they progressively lose mobility and this can lead to behavioural problems. They will become more aware of the physical changes in themselves and may feel that they have little control over their health. As a result they may attempt to gain control in other areas of their life by showing defiant behaviour or aggression.

Some children may learn that if they put up enough of a fight that they will get what they want. For others, feeling down and anxious can result in them being irritable and resistant. They may experience loss of self confidence and esteem which may lead to feelings of worthlessness.

Strategies

Be honest.

Encourage open communication within the family.

Trust is very important. A child needs to have someone to turn to, to answer their concerns over the years. Often “white lies” are told in an attempt to protect the child but this can backfire if the child later discovers the truth.

Try to find out how the child is feeling.

Try to involve the child in aspects of decision making with regard to health and social needs.

Continue the process of informing the child about his condition and making time for questions.

Try to have a “normal” family life.

Continue with consistency and the established house rules so the child has some boundaries.

All family members need to be consistent with the information given.

Try to prepare the child for physical changes before they happen. For example if a child says “I am going to join the football team when I am bigger“, this may be the time to say his legs might not be strong enough, but you will find you a team where the boys have weaker legs.

STEROIDS

In recent years there has been increasing evidence that the use of steroids with DMD can help with muscle strength, prolonging the time before the boys need to use a wheelchair and improving their breathing. It is now a commonplace treatment, but as with all medicines there are some side effects. One side effect reported by some parents is an increase in their child’s behavioural problems.

It is likely that some of these problems existed before the child started the steroids but were less severe, so that they were not noticeable or not considered to be a problem.

There are several options for parents to try if they feel that their child’s behaviour has become unmanageable.

1. Try giving the steroid at night
2. Decrease the dose
3. Intermittent steroid use
4. Stop steroid altogether
Although this has to be an individual decision that parents make, we would strongly recommend that parents try all options before stopping the steroids altogether, because of the proven benefits of steroids in DMD. We would also strongly recommend that you make this decision in consultation with the doctors caring for your son as steroids need to gradually decreased, rather than stopped suddenly.

**SCHOOL**

School is not only a place where children learn but also where they make friends and is very much a normal part of childhood.

Some boys with DMD are known to have what is called a ‘cognitive function deficit’, which means they have some difficulties with learning. Some of the boys have difficulty with reading and verbal skills, and a reduced attention span. However, they are often skilled in rote learning (learning by repetition) and long term memory.

These learning difficulties are not progressive and the boys can make steady progress through school.

For many parents the process of their child starting school can be difficult emotionally. It may be the first time their child has been away from home and exposed to other people without their parent’s protection. For a parent of a boy with DMD these fears can be amplified.

Try to be aware of potential problems such as difficulty in access, bullying, toileting problems, playground exclusion, falling behind in school work and increasing loss of mobility. These can all lead to low self esteem, frustration and anger, which can manifest as behavioural problems.

**What to do when your child begins school**

Inform the school that your child has DMD.

Liaise with the SENCO (Special Educational Needs Coordinator) and Head teacher to organise any extra support for your child that may be needed.

Ideally a Statement of Special Educational Needs (in Scotland, a Co-ordinated Support Plan) should be set up and reviewed on an annual basis.

The school needs to have an understanding of the condition. Written information such as the publication “Inclusive Education Guidelines” (from the Muscular Dystrophy Campaign) can help, as can speaking with a member of the team caring for your boy or an MDC Regional Care Advisor.

The school needs to be aware of the child’s physical limitations and that they may experience fatigue in the afternoon and as the week progresses.

The school environment needs to be considered and access may need to be created.

If a child attends a mainstream school it is important that he attends social activities with other less able children, to help boost his self esteem. If he is constantly comparing himself to able bodied boys he will never measure up and this could lead to frustration, anger and low mood.

Activities could include; drama club for disabled children, IT, chess club, wheelchair football, art, music. When children are involved in these activities they are all equal in physical ability. This can help your child to see past their disability and concentrate on his inner abilities and strengths. This in turn could help boost his self esteem and give him the opportunity to make friends and develop a wider social network.

Be prepared for other children to ask about your boy’s condition. It can be helpful to discuss with the school how these questions will be answered and what information is to be given. Remember that information given to other children will eventually make its way back to your son.
USING A WHEELCHAIR

Moving to using a wheelchair can be a difficult time for both parents and boys. It can be seen as a negative milestone, as it is the point when the child is no longer able to walk and has to rely on the wheelchair for mobility.

Although the wheelchair can provide relief for a child who has been struggling with mobility it can be a time when their behaviour becomes problematic. Before getting the wheelchair they have been dependent on others for their needs. The wheelchair offers them some independence and control. It is not unusual for them to begin ramming the wheelchair into furniture, people, objects as a way of expressing their anger and frustration. It can also be used as a way of expressing their personality such as mischievous tendencies.

This can be dangerous behaviour, so parents need to very firm about this.

Moving to a wheelchair is a very visible sign for a child that they are struggling physically, and families should try to find out how their child is feeling and give them time to express their anxieties.

ADOLESCENCE ONWARDS

Anxiety about the future becomes more of an issue for the boys as they move into adolescence and become increasingly aware of the progressive nature of their condition. The usual issues of adolescence will often be compounded. The boys go through a normal puberty and can experience frustrations due to the assumption that they don’t experience puberty as other boys do or because of a lack of opportunity.

It is a time of change and the boys need increasing control over diet/sleep/friendships/clothes/privacy/explorations like any other teenager. This coincides with a time when they have increasing dependence for self-care needs. There is a conflict between what they want and what they are able to.

It is also a time for planning the future, exams, employment, and independent living.

Possible feelings of teenagers with DMD

Loss of confidence.

Witnessing brothers and sisters attaining independence and leaving him behind.

Feeling protected from the future and from the outside world. Parents may find it difficult to allow him independence.

Limited social opportunities for disabled teenagers.

Embarrassment due to disability, now very visible with use of wheelchair and needing assistance with care.

He may blame his parents and/or feel resentment towards his siblings.

Strategies

Recognise the teenager’s need for control; they are now becoming a young adult. Remember they are a normal young man with weak muscles.

Continue to talk about the condition. Boys will access information elsewhere even if you don’t talk to them, and this may lead to feelings of isolation. Our experience is that when questions are answered honestly less harm is done. Withholding information can lead to anxiety and behavioural problems. If you as a parent do not talk to your child about the condition, others might - this could be their peers, through the internet or by overhearing a conversation - and this information will not necessarily be correct. It is our experience that once it is out in the open, it is much easier for the family.

Life expectancy. There is no easy way to talk about this or a right way. We would encourage families to tell their child that their condition is life shortening. They may also want to see healthcare
professionals independently of their parents to enable them to talk about issues that they feel they can’t with their parents, for fear of hurting them.

SUMMARY

Behavioural problems can occur at any stage, but tend to occur more in periods of transition. For example, losing the ability to run or walk, and having to use a wheelchair, can be linked to how the boys are feeling about themselves. Remember not all boys will have behavioural problems and even if they do experience them this does not mean that the problems will persist throughout their childhood. Behavioural problems don’t have to be a sign of bad parenting. Talk to the people involved in your child’s care about their behaviour and if simple strategies are not helping seek more specialist help. There will be difficult times but there will also be many good times.