Challenging Behaviour

A guide to understanding **behaviour management** in Mucopolysaccharide and Related Diseases

What are Mucopolysaccharide Diseases?

Mucopolysaccharide (MPS) and Related Diseases are rare, genetic diseases which cause progressive physical disability and, in many cases, severe degenerative mental deterioration resulting in death in childhood. Sadly, at present, there is no cure for these diseases, only treatment for the symptoms as they arise.

There are 24 MPS and Related Diseases. Each of these conditions present in a variety of ways and many exhibit a wide spectrum of severity.

What is Challenging Behaviour?

The term 'challenging behaviour' has been used to refer to difficult or problem behaviours. Challenging behaviour typically risks the safety of the individual or others and has a significant impact on the person's or other people's quality of life.

Challenging Behaviour in MPS Diseases

Challenging behaviour is typically manifested in MPS III, Sanfilippo Disease, although it may also be seen in MPS II, Hunter Disease where there is involvement of the Central Nervous System (CNS).

MPS III is manifested in three stages. The first stage, usually in the pre-school years, is characterised by developmental delay and difficult, overactive behaviour. The second stage is characterised by extremely active, restless, difficult and challenging behaviour. The third stage is the quieter stage. The behavioural problems are still present but these are not as challenging, aggressive and hyperactive and often nursing needs take priority.

Typically, individuals with MPS III who display challenging behaviour are unaware of danger and unresponsive to discipline as they cannot understand what is required of them. This can be very difficult to manage.

Examples of behaviour in MPS III

- Extreme hyperactivity
- Aggression - biting, kicking, hitting
- Throwing objects
- No awareness of danger
- Limited communication and understanding
- Short concentration span
- Destructive
- Unresponsive to discipline
- Sleep disturbance
- Episodes of laughing/crying
- Need for routine/familiarity
- Obsessive behaviours, such as chewing
- Inability to play with toys
- Panic attacks
- Fear of people dressed up, such as clowns
- Dislike of loud noises
- Hallucinations

Challenging Behaviour and Learning Difficulties

In general, challenging behaviour is more common in people with learning disabilities. Most people without learning difficulties display lots of challenging behaviour very early in their lives but will soon develop a range of communication and social skills which enable them to obtain what they want and need more easily. Many children with learning disabilities do not develop such skills to anything like the same extent and are left with much the same needs as their peers but much less competent ways of getting them met.

Many cases of challenging behaviour appear to be effective ways for a person with a learning disability to control what is going on around them.

If the behaviour has just arisen or worsened, the possibility that it reflects some kind of biological or emotional disturbance which cannot be articulated in any other way should be considered. Children may bang their heads because their ear aches or hit out because they slept poorly the previous night. Understanding the variation in a person's challenging behaviour is often a key to promoting positive change.

Challenging Behaviour and Communication

Many people with learning disabilities have some difficulties with communication; this may be in understanding what other people are saying or in being able to express themselves. Communication is one of the most important ways in which we control our environment and influence other people. If a child's communication skills limit this control, frustration is likely and challenging behaviour may follow.

Abstract concepts and an awareness of time are particularly difficult to understand. Individuals may also have difficulties because they are given too much language to process and are only capable of understanding key words. Many people need to have information given to them in simple structure, using short, straightforward words or sentences.

Many factors may contribute to the difficulties people with learning disabilities have in getting other people to understand them, for example, because of difficulties in articulating speech or forming clear signs.

It is essential to have a good understanding of the ways in which a particular child or adult communicates. Other people need to make sure that they are communicating in a way that the person understands – by using simple, short sentences and trying to avoid saying something which could be misunderstood. Using single words associated with a particular object, picture or symbol is a particularly useful way of communicating.

Due to the degenerative nature of MPS diseases, individuals with MPS will gradually lose their speech and language skills as the disease progresses. This can make effective communication increasingly difficult. As the disease progresses communication may be through body language and facial expressions.

Prevention of Challenging Behaviour

The goal of prevention is worthwhile. On an everyday basis carers, parents and teachers can try to ensure that the person has what they need when they need it; help, attention, food, drink, preferred activities etc. It is very important that people are also given the opportunities and skills to get things for themselves or to ask for them.

Here are some other points which might be worth considering:

Do you think the behaviour could be evidence of a previously undetected problem

Is the child in pain? Or bored? Are they being asked to do things they find difficult?

Try to check things out for yourself. If you change something does that stop the behaviour?

Keep some sort of record of when the behaviour happens

If it is not safe to ignore the behaviour respond as calmly and blandly as possible to prevent the child hurting themselves or others

Use distraction and diversion to change the behaviour. Telling the child off for bad behaviour can be confrontational and can often make the situation worse

If you do have to respond, better to respond quickly than slowly – otherwise you are teaching the child to be more persistent

Try to move the surroundings that cause danger rather than removing the child

Dealing with Challenging Behaviour

Challenging behaviour can be an emotional experience for parents – you may feel very angry with the child or depressed about their behaviour. Don't be ashamed of this, don't bottle it up. Instead, talk about it with anyone who will listen and understand.

A characteristic of challenging behaviour is that it tends to be long term. In the absence of well thought out plans to manage out of control behaviours, the risks of injury for both parties are increased.

Reactive strategies provide carers with clear plans for how to respond to challenging behaviours. They are brought into play once challenging behaviours become apparent. Reactive strategies will not result in a long term change in behaviour pattern; their goal is simply to help carers achieve rapid, safe and effective control of out of control behaviours.

The key to effective use of reactive strategies is a detailed knowledge of the pattern of behaviour. Most people show us signs that they are becoming agitated or distressed before they lose control. Learning to recognise these early signs is the basis for early intervention, and the earlier carers intervene, the more probable it is that serious behavioural outbursts can be avoided.

Distracting the person or defusing the situation is one of the key ways of dealing with challenging behaviour to prevent it worsening and becoming too difficult to manage safely.

Individuals with MPS diseases who exhibit challenging behaviour will tend to have very short concentration spans. A high degree of flexibility will be needed to best manage this to prevent the behaviour becoming disruptive.

Physical interventions are any means in which force is applied to restrict movement and mobility. This is an emotive topic which generates numerous ethical and practical concerns. In 1996, the British Institute for Learning Disabilities produced a set of policy guidelines. Some key principles include:

- Physical interventions should only be used in the best interests of the person with learning disabilities
- They should only be used in conjunction with other strategies to help people learn to behave in non-challenging ways
- They should be individualised and subject to regular review
- They should employ minimal force and not cause pain

Medication

In MPS Diseases it is important to point out that any medication used is often unsuccessful as the individual's brain is already severely damaged by the time the diagnosis is made. However, medication should not be totally ruled out as it may be partially effective. However, this may be limited as each individual responds differently and doses will vary. It is important to consult with your specialist consultant.

Melatonin is commonly used for sleep disturbance, however, for some this can have little or no effect. Some families have found herbal medicines to be effective, for example, Omega 3. Others have found changes to their child's diet have helped with behaviour.

Environmental & practical approaches to behaviour management

Due to the underlying brain disease associated with MPS III, treatment of behaviour by modification techniques or medication is extremely limited. Experts in the medical management of MPS diseases recommend alterations to the environment rather than attempts to change the behaviour of the children. 1:1 care and supervision should be maintained at all times.

Adaptations to your home - ***Ensure a safe environment both inside and out***

Explore adapting your home, for example, with a ground floor en-suite bedroom and bathroom

Use soft furnishings and padding for walls

Put mattresses or padded flooring on hard floors to cushion any falls

Consider using barn style doors and/or child safety gates for extra security

Use window locks for the tops of doors

Use safety laminated glass for windows

Insert spyholes in doors so you can check up on your child without disturbing them

Install cameras and monitors

Remove any unnecessary hazards such as ornaments, pictures etc

Ensure sharp corners are protected

Protect wooden surfaces such as window sills with plastic edging strip as some children like to chew and gnaw at the edges

Place televisions high up on wall mounted brackets

Use specialist seating and feeding chairs

Make use of specialist toys and sensory equipment

Consider using vinyl flooring which is easiest to clean, or durable washable carpet

Situate electrical sockets and light switches out of reach and use protectors where possible

Radiators can be boxed to eliminate the danger of burns

When you are outdoors

Avoid hard surfaces in your garden. Use grass, soft play or some other protective ground covering

Remove all sharp objects

Use high, safe fencing and ensure gates have locks

Use an appropriate wheelchair or buggy

Use handling belts/reins if appropriate

Consider car seats with five point harnesses to keep your child safe when travelling in the car

Use car door safety locks or central locking for doors and windows

In an educational setting

Implement a varied flexible curriculum that is individual to the child

Put in place 1:1 support that offers flexibility in order to encourage and keep the child focused

Use more than one carer to give 1:1 support on a rotational basis

Change the environment where practical

Recognise a change of activity may be required every 5-10 minutes

Use familiar people/routines

Introduce a soft play/safe area and plenty of space to run around if appropriate

Focus on objects and activities the child likes and enjoys to help maintain any skills learnt for as long as possible

Provide regular physiotherapy and hydrotherapy to help maintain fine and gross motor skills whilst accommodating limitations

Give regular praise, encouragement and reward

Ensure good home – school liaison to ensure sharing of information and approaches

About the MPS Society

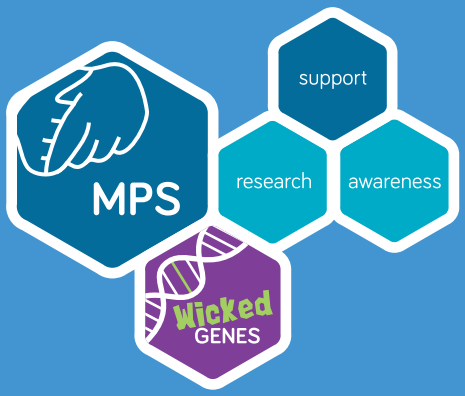
The Society for Mucopolysaccharide Diseases is a registered charity, founded in 1982, which represents from throughout the UK children and adults suffering from MPS and related diseases, their families, carers and professionals.

The Society produces a range of publications including a quarterly MPS Magazine. Regional clinics, information days and conferences are organised throughout the UK. The Society's advocacy team provides a unique, needs-led, individual advocacy service to individuals suffering from MPS and related diseases, their families and carers.

The Society can support you in providing information to families, schools, hospices, occupational therapists and housing officers, for example, by writing education reports and training in schools and the provision of housing reports and copies of adaptations already undertaken.

For further information about the work of the Society and the service we provide please contact us.
This booklet is not intended to replace medical advice or care.

Acknowledgement: The Challenging Behaviour Foundation, www.thecbf.org.uk



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