

# **Scottish Muscle Network**

Patient information leaflet

## **Bladder function and continence care in Duchenne Muscular Dystrophy (DMD)**

## Introduction

Duchenne Muscular Dystrophy (DMD) is normally associated with loss of muscle strength. Although we tend to think of this in terms of muscle in the body, arms and legs becoming weaker, bladder function can also be affected by weak muscles.

All young people with DMD are different and some have no toileting issues at all. However, some parents' report their sons have problems becoming toilet trained in the early years and that it can take longer than their friends. Some boys have regular 'accidents'.

DMD can affect the bladder muscle and the muscles of the pelvic floor, (which help keep the entrance to the bladder closed).

## What is bladder incontinence?

Bladder incontinence is when you pass urine without meaning too. Some people are unaware they have done this, whilst others know they are doing it but are unable to stop. Bladder problems can be embarrassing, and some boys will avoid going out if they are unsure if there will be suitable toilets available. If they have physical challenges and need help with their personal hygiene or even help to transfer onto the toilet, they will often avoid going to unfamiliar places and events.

There are a number of reasons why toileting may be difficult for your son and this leaflet explores some of the common problems and offers some solutions too. There is no doubt that finding the best way to manage these symptoms can improve family life.

## How do the bladder and kidneys work?

The kidneys are always working to make urine and a trickle of urine continuously passes from the kidneys into the bladder. If you drink more or eat foods with high water content such as fruit and vegetables, you will make more urine. If you are exercising or sweating, you will make less.

As the urine passes into the bladder, the bladder stores it. As the bladder is a muscle, it can expand to allow more urine in or contract to empty the urine out. The urethra (or sphincter) is where the urine comes out and this is normally kept closed until you are ready to go to the toilet. The pelvic floor muscles surround the urethra and help to keep this outlet closed.

As the bladder starts to fill, stretch receptors in the bladder muscle alert you to the fact you will need to empty your bladder and so when the time is right, your bladder muscle contracts, and the pelvic floor muscles and urethra relax allowing you to empty your bladder.

## What are the common bladder problems?

**Stress incontinence:** This is the most common type of incontinence and is usually caused by muscle weakness which is a key feature in neuromuscular disorders. When the bladder starts to fill, the pressure in the bladder may become too great for the weak pelvic floor muscles and these muscles cannot stay closed tightly enough to withstand the pressure from the filling bladder. Urine may then leak out when you cough, sneeze, or laugh. The bladder can also become overfull as the bladder muscle itself is weak and is more easily stretched.

**Urge incontinence:** This type of incontinence tends to take people by surprise. It happens when you have an urgent need to go to the toilet and sometimes urine leaks out before you get there. A common example is when you arrive home and suddenly need to go to the toilet and dribble a bit whilst you are approaching the toilet, or your bladder decides it needs to empty before you are ready to let go. This may be caused by the bladder being overfull (as the muscle is weak) or the fact that the bladder muscle is sending wrong signals to the brain (neurogenic bladder). This may have occurred over time as you have frequently held on too long i.e. some children don't like going in school so will hold on as long as possible until they get home, or some people don't like using public toilets so will wait. Whilst this is fine in the short term, repeatedly ignoring the bladder signals can lead to nerve dysfunction in the long term.

**Mixed incontinence:** It is possible to have a combination of stress and urge incontinence.

## Toileting difficulties

The following are examples of toileting difficulties and some solutions.

**Standing Balance:** For some young people with DMD standing still for more than a second or two can be challenging and this affects their balance. This can cause boys to “have an accident” when using a urinal or toilet in the standing position. In these circumstances, sitting down may be a better option and it is important to make sure that the toilet at school and home has handrails to assist the young person back into the standing position. An Occupational Therapist (OT) can assess the toilet height and position of handrails.

**Clothing:** If the young person has been used to toileting independently, loss of hand strength can make going to the toilet more difficult. Undoing buttons and zips take time and for many young people, going to the toilet is put off until the last minute. Suddenly there is an urgency, and this can make undoing clothes even more difficult. In these circumstances, it is important that school staff can recognise when this is happening and perhaps gently remind the young person to go to the toilet before the

interval or lunchtime. If the young person is wetting repeatedly, he may feel embarrassed and this needs to be handled sensitively.

In some cases, school staff may not be fully aware of the reasons behind the wetting instances and occasionally it is suggested that the young person wears a pad to school. **We do not recommend wearing pads as this encourages the young person to “let go” whenever they wish**, and they become used to wearing a pad. Trying to then re-establish a toileting pattern can be more difficult, therefore preventing this situation is a better option. However, pads may be an option if bladder training isn't possible.

If the young person begins to wet more frequently, you can start a toileting programme. It is normal to go to the toilet every two to three hours and it is important to give the young person time to fully empty their bladder. A toileting programme is when the young person is invited to go to the toilet at regular intervals throughout the school day. This can help to re-establish bladder control and the parent or carer can gradually increase the length of time between asking the young person if they need the toilet until the young person is able to ask by themselves in a more natural way. If the young person needs help with clothing, there are various options available to make things a bit easier and to help them remain independent when undoing clothing. An OT can give you more advice.

**Wheelchair:** Many boys can maintain independent toileting when in a wheelchair with the use of a bottle and if assisted, they can shuffle forwards and use the bottle without difficulty. When full, the bottle may be too heavy for the young person to hold and empty therefore, a carer should be there to take the bottle.

If the young person is embarrassed that he needs help to position the bottle, then this can also be problematic for maintaining good bladder care. Often boys with DMD have a small penis due to the fact it retracts in the forward sitting position and locating the penis in the bottle can be more difficult. Some boys have found a bottle with an extended neck or a female bottle with the wider opening is often easier to use. An Occupational Therapist or Continence Care Adviser can usually advise but often it comes down to trial and error.

Some boys have found that by reclining back in the chair (by sliding forward) the hip angle opens and makes it easier to use the bottle. There are a variety of bottles on the market and some males prefer to use the uribag (available on prescription from Fittleworth) which is a very small discreet bottle that expands for use and can be kept in the pocket.

Some families have found helpful advice via their local Continence Care (Enuresis) Clinic and if the young person is over the age of five years, a referral can be made via a healthcare professional.

**Bed wetting:** If your son is becoming a bit unsteady on his feet, especially if he wakes at night, using a bottle may be more sensible. Keeping a bottle at the side of

the bed to save him getting up in the middle of the night and in the dark could be a simple solution to bed wetting. Some young people go through phases of wetting the bed and it is not always related to their muscle problem. Staff from the Continence Care Clinic can investigate this and offer some practical solutions and advice.

**Neurogenic Bladder:** Some literature describes young men with DMD as having a neurogenic bladder. This means that rather than the muscle being affected, it is the nerves supplying the bladder that are not working properly. Neurogenic bladder problems and symptoms vary depending upon the cause. It is thought in DMD that young people become used to holding onto urine as they prefer to wait until they get home to go to the toilet. Holding onto urine too long can cause the nerves to mix up the signals of when the bladder is full.

Often this is because their carers at school or college may not be confident in helping them to the toilet and it can take a while to get used to doing things in a certain way. Some boys don't like going to the toilet as they feel toileting takes too much time out of their busy day when they could be socialising with their friends. Other young people report that if they are moved within the chair, it takes a long time to position them back comfortably.

If you regularly avoid going to the toilet when you need to, you overstretch your bladder. If the bladder is regularly overstretched:

- the stretch receptors in the bladder wall take longer to be stimulated
- the bladder becomes large and can accommodate much higher volumes of urine

This means that when the stretch receptors are finally stimulated, the bladder will be overly full, and they may not be able to wait long enough for the bottle to be positioned.

In cases like this, working with the young person and their family to establish a good bladder routine is essential. There are various options on the market such as a sheath, (a condom like glove that fits over the penis snugly) allowing the young person to urinate more frequently. The urine drains into a catheter bag which you attach to the leg which can be hidden under trousers.

Carers may need more in-depth training in moving and handling. They should discuss this with the family who understand the moving and handling needs of the young person.

Some young people will avoid drinking fluids through the day so that they don't have to go to the toilet during the day. Dehydration can lead to tiredness and fatigue and can cause issues with kidney stones. Establishing a good toileting regime is better than risking kidney stones, which are often painful when passed.

On the other hand, if the young person drinks large volumes of fluid, then they will need to go to the toilet more often and this can be a factor in bladder problems. However, it should be noted that restricting fluid intake can also irritate the bladder so it really is about finding a balance that is right for the young person as everyone different.

Certain drinks that contain caffeine such as coffee, tea, hot chocolate, and fizzy drinks can make the young person want to go to the toilet more often. This is because they are diuretics, and it might be worthwhile changing to caffeine-free drinks. Try not to drink late at night or just before bedtime to prevent waking up in the middle of the night to go to the toilet and disturbing sleep.

**Myoglobinuria:** Some children have reported episodes of myoglobinuria after periods of prolonged exercise. This is when the urine looks red and, in some cases, it can be mistaken for blood.

Myoglobin is a protein found in muscle. When a muscle is exercised, it requires oxygen. With continued activity, more oxygen is required, and myoglobin provides extra oxygen for the muscle to maintain this level of activity for a longer period. When muscle is damaged, the myoglobin is released into the bloodstream and passed via the kidneys into the urine. In large amounts, myoglobin can cause damage to the kidneys and episodes of myoglobinuria should be investigated so it is important to tell your doctor about this. The child will often tell parents that his urine is red or looks like Coca-Cola.

Myoglobinuria is a recognised side effect of steroids. It is felt that boys who are participating in steroid therapy may be more active than those who don't take steroids. It is thought that the increased activity places the dystrophin-deficient muscles under greater mechanical stress, which can lead to further muscle fibre damage and myoglobinuria.

## Conclusion

Bladder symptoms are common in boys with DMD, and it is important you discuss these symptoms with your neuromuscular team. There are a variety of ways to help manage these symptoms and it is best to let us know as soon as possible to avoid any undue stress.