



Myotonic Dystrophy  
SUPPORT GROUP

# Bowel problems in Myotonic Dystrophy

by  
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# Myotonic Dystrophy

Congenital Myotonic Dystrophy is not only associated with problems of the limb muscles but with the smooth muscles that line the gut and form the apparatus for controlling bowel movements (the anal sphincter). Gastro-oesophageal reflux, indigestion and swallowing problems, also abdominal pain, constipation, diarrhoea and faecal incontinence are symptoms resulting from gut smooth muscle being affected in various areas. In some instances these complaints may come to light before problems of limb weakness or muscle relaxation are apparent.

## **Extent of the problem**

Reports have shown that up to a quarter of adults with Myotonic Dystrophy consider their bowel problems to be the most disabling of the symptoms associated with their condition. However, information is lacking on the frequency of these problems in children and young people with Congenital Myotonic Dystrophy, and

importantly the impact on themselves, their families and carers.

A number of families attending the Paediatric neuromuscular clinic at Guy's Hospital told us about the difficulties they experienced when toilet training their children with Congenital Myotonic Dystrophy, in dealing with episodes of severe constipation, and coping with the embarrassment experienced by school age children with faecal incontinence. We sought to investigate the frequency of bowel problems in the children and young people attending clinic to assist us in designing effective management plans. 15 young people with Congenital Myotonic Dystrophy aged between 3 and 22 years and their families helped us with a questionnaire.

The number of times individuals went to the toilet ranged from once a week to 42 times a week. 8 of the 15 young people were constipated (infrequent, painful passage of hard

stool) and required laxatives to maintain their bowel action, 1 child required regular enemas. 2 teenage boys at secondary school reported daily episodes of incontinence of stool. Only 2 felt they could 'hold on' to pass stool for longer than 15 minutes, while 11 of the 15 weren't able to hold on at all or for more than 5 minutes. 7 young people experienced frequent abdominal pain.

In 4 of the 15 individuals, bowel problems were apparent before other symptoms of Myotonic Dystrophy and in 3 of 15, bowel problems were considered the worst of the problems experienced because of the condition. On a scale of 1 (least of problems) to 5 (worst of problems), 11 graded their bowel problems 3 or more.

### **Dealing with the problems**

The results of underlying chronic constipation must be dealt with by the use of stool softeners (e.g. Movicol paediatric solution) and stimulating agents (e.g. Senna), to clear out the bowel. It is most important to introduce a regular toileting

routine, that is, time spent in the loo before and after school, to try and reduce the amount of the day when there are stools in the bowel, which could potentially lead to accidents. Constipation and incontinence can co-exist and 'holding on' can be a big problem. Stool bulking agents, for example, methylcellulose, or Movicol paediatric solution, may be of benefit to assist appreciation of the presence of a stool in the lower bowel and using stimulant laxatives such as senna or picosulphate may help in establishing a predictable time of day when passing stool is most likely to occur.

Where medicines are unsuccessful in controlling severe constipation and soiling, and in spite of compliance with treatment and toileting routines, regular rectal enemas or an ACE procedure (antegrade continence enema) may significantly improve quality of life. The ACE is a surgical intervention that permits catheter access via a stoma<sup>1</sup> button on the abdominal wall to the bowel and enema administration to

enable regular controlled and predictably timed evacuation of large bowel contents.

In some instances colostomy may be considered preferable to lifelong faecal incontinence if other management strategies are unsuccessful.

Not everyone involved in the care and supervision of children and young people with Myotonic Dystrophy may be aware that bowel problems are a major feature of the condition, warranting discussion, reassurance and understanding, support and active medical management to improve quality of life.

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- 1. For people with Myotonic Dystrophy and having weak fingers cleaning of a stoma area may be difficult.**



## Other publications available from the Myotonic Dystrophy Support Group:

- Anaesthesia and Sedation for patients with Myotonic Dystrophy
- Basic Information for Midwives
- Congenital Myotonic Dystrophy
- Excessive Daytime Sleepiness and Myotonic Dystrophy
- Facts for patients, family members and professionals
- Myotonic Dystrophy and the Eye
- Myotonic Dystrophy Support Group
- Relatives Information
- The Heart and Myotonic Dystrophy
- Why do we get new families with Myotonic Dystrophy?

Reference "Lower gastrointestinal tract disturbance in Congenital Myotonic Dystrophy." Kerr TP, Robb SA, Clayden G. European Journal of Paediatrics; August 2002, 161 (8), 468-469



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