## Neurogenic bowel dysfunction: Clinical management recommendations of the Neurologic Incontinence Committee of the Fifth International Consultation on Incontinence 2013

### Abstract

**Background**: Evidence-based guidelines for the management of neurological disease and lower bowel dysfunction have been produced by the International Consultations on Incontinence (ICI). These are comprehensive guidelines, and were developed to have world-wide relevance.

**Aims**: To update clinical management of neurogenic bowel dysfunction from the recommendations of the fourth ICI, 2009.

**Materials and methods:** A series of evidence reviews and updates were performed by members of the working group. The resulting guidelines were presented at the 2012 meeting of the European Association of Urology for consultation, and modifications applied to deliver evidence based conclusions and recommendations for the scientific report of the fifth edition of the ICI in 2013.

**Results**: The current review is a synthesis of the conclusions and recommendations, including the algorithms for initial and specialized management of neurogenic bowel dysfunction. The pathophysiology is described in terms of spinal cord injury, multiple sclerosis and Parkinson's disease. Assessment requires detailed history and clinical assessment, general investigations, and specialized testing, if required. Treatment primarily focuses on optimizing stool consistency and regulating bowel evacuation to improve quality of life. Symptom management covers conservative and interventional measures to promote good habits and assist stool evacuation, along with prevention of incontinence. Education is essential to achieving optimal bowel management.

**Discussion:** The review offers a pragmatic approach to management in the context of complex pathophysiology and varied evidence base.

#### Introduction

Disorders of the central nervous system (CNS) are common, with worldwide estimates of the prevalence of spinal cord injury (SCI) at over 2.5 million, multiple sclerosis (MS) considerably greater than 1.5 million and Parkinson's disease (PD) approximately 3 million<sup>1,2</sup>. A large proportion of patients with CNS dysfunction experience frequent bowel symptoms, and these symptoms are amongst the most physically, socially and emotionally disabling<sup>3,4</sup>. Most of the literature deals primarily with the bowel dysfunction associated with three common CNS diseases – SCI, MS and PD.

Bowel dysfunction affects almost all patients with a chronic SCI – up to 95% report constipation, faecal incontinence is experienced at least once per year by 75% and daily by 5%, with 33% experiencing regular abdominal pain related to the level of injury<sup>5,6</sup>. Bowel dysfunction is less studied, but almost as prevalent in other neurological conditions. About one-third of MS patients suffer from constipation and one-quarter are incontinent at least once per week<sup>7</sup>. In patients with PD, constipation, in particular difficulty with defaecation, occurs in 37%<sup>8</sup>. One quarter of stroke survivors experience constipation, and 15% suffer with faecal incontinence<sup>9</sup>.

#### Methods

Using the previous review of the Neurologic Incontinence committee from the fourth International Consultation on Incontinence as a baseline<sup>10,11</sup>, an updated literature search for all published research was conducted from January 2008 to August 2012 using the key words: faecal incontinence,

neurogenic, neuropathic, neurologic, neurogenic bowel, bowel management, bowel care, conservative treatment, practice guideline, incontinence, constipation, cisapride, colonic, dietary fibre, laxative, suppositories, neurogenic constipation, faecal incontinence, surgery, sacral nerve stimulation, antegrade continent enema procedure, dynamic graciloplasty, artificial anal sphincter and colostomy. Databases searched included Medline, Embase, Cochrane Library, Biosis and Science Citation Index)Findings from such searches were presented at the annual scientific meeting of the European Association of Urology in 2012 for consultation and modifications applied for the scientific report of the fifth edition of the ICI in 2013. Levels of evidence (LOE) and grades of recommendation (GOR) were derived according to the modified Oxford system developed by the International Consultation on Urological Diseases<sup>12</sup>.

# Pathophysiology

The level and extent of injury or lesion are the most important factors in determining bowel symptoms in both SCI and MS. Gastrointestinal transit is under the complex regulatory interplay of the enteric nervous system and extrinsic autonomic innervation. Anal continence depends on an interplay between rectal sensory-motor function and the internal and external anal sphincters. According to standard clinical classification of SCI, lesions are classified as

- supraconal (above the conus medullaris, where inhibitory input is lost) slowed whole gut transit and hypertonia and hyperreflexia of the hindgut (i.e. distal to the splenic flexure)<sup>13</sup>.
  Rectal hypertonia results in reduced rectal compliance and predisposes to reflex defaecation and incontinence.
- within the conus or being located in the cauda equina (where excitatory sacral parasympathetic supply is lost) - efferent limb of the reflex arc to the hindgut is interrupted resulting in hypotonia and hyporeflexia<sup>14</sup>.

Supraconal injury tends not to alter anal tone, whilst the reduced tone of cauda equina lesions may relate to faecal bolus impaction as much as loss of sympathetic input. The striated external anal sphincter is under voluntary control from Onuf's nucleus in the ventral horn of the sacral spinal cord via the pudendal nerves. Thus, in complete SCI the voluntary control of the external anal sphincter is lost. Suprasacral complete SCI has been shown to result in the most severe degree of bowel dysfunction<sup>15</sup>.

The pathophysiology of bowel dysfunction in patients with PD is quite different from that of SCI or MS. Dystonia of the striated muscles of the pelvic floor and external anal sphincter explains the defecation dysfunction<sup>16</sup>; this aetiological factor is supported by the observation that pelvic floor dysfunction is alleviated with L-Dopa<sup>17</sup>. In addition to the pelvic dysfunction, colonic transit time is usually prolonged in patients with idiopathic PD<sup>16</sup>. The current theory of aetiology of PD is the Braak hypothesis which states that the earliest signs of Parkinson's are found in the gut's nervous system and the lower brain stem. The theory states that Parkinson's progresses to the substantia nigra and the white matter over time. The basis of this theory is supported by the "non-motor symptoms", such as constipation, loss of sense of smell and sleep disruption which often precede movement features"<sup>18</sup>.

### Assessment

Current bowel symptomatology is assessed, regarding bowel frequency, stool consistency, faecal incontinence and manoeuvres needed to achieve bowel management. This information is usually gathered from standard patient and carer history, but scoring systems exist which may supplement this. There are standard instruments (Cleveland Constipation score, St Mark's incontinence score, and recently a condition specific score has been developed for neurologic patients<sup>19</sup>. This Neurogenic Bowel Dysfunction (NBD) Score is validated in SCI, but not PD; it is currently being assessed in patients with MS.

Digital rectal examination is an essential component, allowing assessment of rectal filling, resting anal tone, ability to generate a voluntary contraction and it also gives a crude assessment of anal sensitivity. The place of more interventional physiological or radiological transit investigations is not established, but may be appropriate if there is any co-morbidity (prior anal surgery, obstetric history, pelvic organ prolapse). Plainly, patients with alarm symptoms should have necessary colonic imaging performed. Alarm symptoms in this patient group are more difficult to recognise, but any worsening of established bowel dysfunction, weight or blood loss warrants investigation.

Initial management of symptoms follows a conservative pathway as described below and summarised in figure 1. An overview of specialised management is provided in figure 2.

#### **Conservative treatment**

The primary aims of bowel care are twofold: to achieve bowel evacuation in a timely manner and to avoid faecal incontinence. The first step of this bowel care consists of optimizing stool consistency with adequate fluid and fibre intake, and stimulating evacuation of stool on a regularly scheduled basis with digital rectal stimulation. A range of other non-invasive interventions may supplement this: Valsalva or manually-generated external pressure, oral medications – stool softeners, stimulant laxatives and prokinetic agents; diet modification; biofeedback – a re-education strategy to inform change in bowel function; electrical stimulation and functional magnetic stimulations. The key to successful bowel management is intensive patient education and training. If conservative bowel management fails, surgical management may be necessary.

#### Bowel program/bowel care

Initial management for all subjects is medication review (especially drugs which may affect bladder function such as antimuscarinics, baclofen, codeine analgesia, non-steroidal anti-inflammatory drugs (NSAIDs) and antibiotics) and addressing any unusual dietary habits. In general, scheduled defecation should be attempted once a day or on alternate days. However, knowledge of bowel frequency prior to injury is important in deciding on the bowel program.

#### Specific techniques

Establishing a regular diet to optimize bowel motility is important and in general reducing fibre intake is helpful in improving the bloating and flatulence caused by slow whole-gut transit. Conversely, a higher fibre diet helps improve stool consistency and therefore prevent faecal soiling. Excessive quantities of caffeine, alcohol and foodstuffs containing the sweetener sorbitol can cause the stools to become looser and hence more difficult to manage<sup>20</sup>.

Promoting a sense of privacy and comfort, while exploiting gravity, to achieve a successful bowel regime is advised. Digital rectal stimulation (DRS) can be used to invoke a reflex contraction of the

colon and rectum, and hence a bowel action, although caution is advised as it may cause local trauma and induce autonomic dysreflexia (AD) in SCI individuals<sup>11</sup>. Finally, manual extraction of stool can be used and combined with a Valsalva manoeuvre to improve effectiveness<sup>20</sup>.

Chemical stimulants such as suppositories and enemas may supplement the above by causing a reflex contraction of the rectum. Implicitly, there is little point in using these agents when the rectum is empty on digital checking<sup>20</sup>. Stimulants range in potency, from glycerine suppositories, through micro-enemas, to larger volume stimulant enemas.

# Assistive techniques for defecation

Abdominal massage with the heel of the palm, in a circular motion from right to left may help increase bowel transit and movement of content towards the rectum<sup>20</sup>. In small controlled trials, positive effects were seen in patients with MS<sup>21</sup> and with SCI<sup>22</sup>. Anal stimulation with pulsed water irrigation to break up stool impactions and to stimulate peristalsis is a safe and effective method for individuals with SCI who develop impactions, or do not have an effective bowel routine<sup>23</sup>.

Several studies have been published on transanal/transrectal irrigation (TAI). Christensen et al. (2009)<sup>24</sup> described how 163 of 348 patients (47 percent) had a successful outcome from treatment with TAI after a mean follow-up of 21 months, although success rates varied between patients with different underlying pathology. Amongst factors correlating with positive outcome were neurogenic bowel, low rectal volume at urge to defecate, low maximal rectal capacity, and low anal squeeze pressure increment. Two non-fatal bowel perforations were found in approximately 110,000 irrigation procedures.

# Appliance/assistive techniques for faecal incontinence

Anal plugs are one option although previous studies have yielded conflicting results(23). Whilst anal plugs may provide a benefit to the majority of patients, it does not suit all eligible patients, with *in situ* plug retention being a problem for some<sup>25</sup>.

Neuromodulation, electrostimulation or magnetic stimulation are techniques that offer an alternative intervention option for neurogenic bowel dysfunction in children and adults. Studies to date suggest successful incorporation of magnetic stimulation as an adjuvant treatment among individuals with SCI<sup>26</sup>, improved sphincter pressures and rectoanal inhibitory reflex and increased frequency of defecation with electrostimulation in children<sup>27</sup>, and in small numbers, neuromodulation demonstrated preliminary potential for some neurogenic constipation<sup>28</sup>.

# Conclusions

- In reflex bowel, digital rectal stimulation relaxes the external anal sphincter and increases peristaltic contractions by facilitating an excitatory anorectal (ano-colonic) reflex, and enhances bowel movement and evacuation (LOE 3)
- Abdominal massage has beneficial effects on neurogenic bowel dysfunction, including defaecation function and faecal incontinence (LOE 3)
- Transanal irrigation is a safe method to improve constipation and faecal incontinence in individuals with neurogenic bowel dysfunction (LOE 2)
- An anal plug can help control fecal incontinence in some neurological patients (LOE 3)
- Different forms of electrical stimulation/neuromodulation seem promising for faecal incontinence and defecation management in neurological patients (LOE 3)

- To increase adherence rate with bowel care program/clinical practice guideline, implementation strategies should be addressed to care providers (LOE 3)
- Recommendations
- Multi-faceted programs are the first approach to neurogenic bowel management and are supported by lower levels of evidence (B). They may consist of toileting, rectal stimulation (digital or with water stream), manual faeces extraction, transanal irrigation and other assistive techniques (B)
- Diet can help but multi fibre is not necessarily indicated in patients with upper motor neuron lesion (B)
- It should be taken into consideration that autonomic dysreflexia when using mechanical stimulation and assistive techniques can occur in neurological patients with a high spinal cord lesion (B/C)

# Surgical treatment

The mainstay of current treatment in neurogenic faecal incontinence is adopting a conservative approach towards reversing the systemic effects and optimizing the mechanics of defecation through the use of laxatives and irrigation approaches. Surgery should be normally reserved for patients who have failed conservative therapy. This section focuses on specific aspects of faecal incontinence surgery in neurogenic patients. Options for surgical treatment of neurogenic bowel dysfunction are limited consisting of; 1) sacral neuromodulation, 2) antegrade continent enema procedure, 3) dynamic graciloplasty, 4) artificial anal sphincter, 5) elective colostomy, 6) postanal repair.

Sacral neuromodulation has been reported to restore continence in patients with intact muscle structure. The overview of the studies shows that electrical nerve stimulation is effective in *partial* SCI, however, there are no reports for complete spinal cord lesions. Kutzenberger et al (2005)<sup>29</sup> reported a 17-year experience with sacral deafferentation (SDAF) and implantation of sacral anterior root stimulator (SARS). Of 464 paraplegics receiving a SDAF-SARS with a mean follow-up of 6.6 years, 401 paraplegics used it for defecation (frequency 4.9 per week).

# Conclusion

• SNM is a minimally invasive procedure, and seems to be an option for faecal incontinence and constipation due to functional deficit of the anal sphincter without structural defect in *incomplete* neurological lesions (LoE3/4)

Recommendation

• Studies on large series with long-term follow-up are needed to determine the role of SNM in the treatment of faecal incontinence associated with neurological lesions and identify those patients most likely to benefit

The original antegrade continent enema (ACE) procedure was developed by Malone et al<sup>30</sup> who reported successful results in five children with intractable faecal incontinence. This procedure has been applied mainly to the paediatric population with neurogenic bowel dysfunction and anorectal anomaly, and successful outcome was achieved in 70-100 %<sup>31–33</sup>. Modifications have been reported including application among adult neurological patients with faecal incontinence, and similar success rates (83-100 %) were reported. Overall, stoma stenosis is the most common complication, affecting 10-41 %.

### **Conclusion and recommendation**

 Antegrade continence enema stomas are effective for controlling faecal incontinence and constipation associated with neurogenic bowel dysfunction especially in children (LoE 3; Grade B). Patients should be properly selected to determine appropriate motivation.

Satisfactory continence has been reported with dynamic graciloplasty in 56% to 81% of patients<sup>34–36</sup>. However, all studies presently available include small numbers of neurological patients, or there is no subgroup information on the outcome in neurological patients.

### **Conclusions and Recommendations**

• In practice, dynamic graciloplasty has been essentially abandoned in all but specialist centres, as it seems to be associated with high complication rates, and outcome appears to correlate to surgeon's experience (C).

Implantation of an artificial anal sphincter (AAS), also referred to as artificial bowel sphincter (ABS) was first reported in 1987<sup>37</sup> as a modification of the device used for the treatment of urinary incontinence. The reported success rates obtaining acceptable continence range from 41 to 90%<sup>38–40</sup>. Most studies have a small number of neurogenic patients or do not indicate the number of neurogenic patients included. O'Brien et al. (2000)<sup>41</sup> observed improvements in symptom and quality of life scores when comparing standard care with the AAS in fourteen patients, leading the authors to conclude the AAS is safe and effective when compared with supportive care alone, although 15-30% may require permanent explantation.

## **Conclusions and Recommendations**

- Implantation of the artificial anal sphincter may be undertaken in neurogenic faecal incontinence, except in patients with previous perianal infections or with a thin and scarred perineum, in whom muscle transposition is preferable.
- Due to the relatively high risk of treatment failure and of complications requiring re-operation, patient selection and counselling should be very strict (C)

Several retrospective studies on the effect of colostomy formation in SCI patients showed a significant decrease in the average time spent on bowel care per week and improvement of QoL<sup>42</sup>. The early and long-term complication rates reported are 6 to 15%, and 15 to 37.5%, respectively. The commonest long-term complication is mucus discharge per rectum. One of the more frequent, persistent, problematic complications is diversion colitis<sup>43</sup>.

# Conclusion

• Elective colostomy may be an option for some SCI patients with severe uncontrolled faecal incontinence (C)

Postanal repair is inexpensive in comparison with graciloplasty and sacral nerve stimulation, and has low morbidity. Additionally, this procedure is useful in the elderly or those with significant comorbidities. Although long-term continence has been found to deteriorate, the procedure can result in a satisfactory outcome in the long-term in a proportion of patients<sup>44</sup>.

### **Conclusion and Recommendation**

• Postanal repair results in satisfactory outcome in the long-term in patients with neurogenic sphincter weakness. However, this is a single centre experience, which needs further confirmation (C).

# Conclusions

In both SCI and MS, the level and extent of injury or lesion is the most important factor in determining bowel symptoms. In PD however, dystonia of the striated muscles of the pelvic floor and external anal sphincter explains defecatory dysfunction. Assessment requires detailed history and clinical assessment, supported by general investigations and specialised testing if required. Treatment primarily focuses on optimizing stool consistency and achieving bowel evacuation on a regularly scheduled basis to avoid faecal incontinence. Symptom management includes conservative measures to promote good habits and assist evacuation of stool, through to surgical interventions reserved for those where conservative management has failed. Education is key to engaging patients to accomplish optimal management of their bowel symptoms in order to establish acceptable quality of life.

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### Figure 1 Legend

Algorithm to describe recommended initial assessment and therapy for neurogenic lower bowel dysfunction.

#### Figure 2 Legend

Algorithm to describe recommended specialised assessment and therapy for neurogenic lower bowel dysfunction.