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Idiopathic constipation in children clinical practice guidelines

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INFORMATION ABOUT CURRENT GUIDELINES

Constipation in children is a common reason for consulting medical professionals. It has a reported prevalence of 5%–30% in children depending on the criteria used.[1 2]

In 2010, the National Institute for Health and Care Excellence (NICE) published clinical practice guidelines on the diagnosis and management of constipation in children.[1] The aim of this guideline was to offer 'best practice advice on the care of children and young people with idiopathic constipation'.[1] The subsequent NICE quality standard (2014) consisted of a concise set of prioritised statements to achieve quality improvements in managing children with constipation.[2] Also in 2014, the European Society for Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) and the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) jointly published a clinical guideline on the diagnostic evaluation and treatment of functional constipation for children.[3] This review encompasses recommendations from both the NICE guidelines and the joint ESPGHAN/NASPGHAN guidelines, with guidance specifically from ESPGHAN/NASPGHAN shown in italics in the text below. Table 1 highlights the main differences between the NICE and joint ESPGHAN/NASPGHAN guidelines.

PREVIOUS GUIDELINE

In 1999, the NASPGHAN published a medical position paper providing recommendations on paediatric constipation.[4] This document served as the basic structure for the recently published joint ESPGHAN/NASPGHAN guidelines[3] (box 1).

KEY ISSUES THAT THE GUIDELINES ADDRESS

Definition of constipation

NICE guidelines have defined constipation as 'idiopathic' if it cannot be explained by any anatomical, physiological, radiological or histological abnormalities. The ESPGHAN/NASPGHAN guidelines recommended that the Rome III criteria for the definition of functional constipation be used for all ages (box 2).[5] Constipation is referred to as 'intractable' when it does not respond to sustained, optimal conventional medical treatment for at least 3 months. Faecal impaction is defined as a hard mass palpable in the lower abdomen identified on physical examination, a dilated rectum filled with a large amount of stool on rectal examination or radiological examination of the abdomen shows excessive stool in the distal colon. Box 3 highlights children who are at higher risk for developing idiopathic constipation.

Diagnosis

In children and young people, symptoms of constipation are often non-specific and there may be a delay in seeking advice because of feelings of embarrassment. In patients with typical symptoms, detailed focused history (including previous history of constipation) and physical examination (including inspection of spine, perineum and gait) should be sufficient to diagnose functional constipation. Digital anorectal examination is indicated in children with suspected faecal impaction, *when the diagnosis of functional constipation remains uncertain or in children with intractable constipation, to exclude underlying medical conditions*. Common presenting symptoms are summarised in table 2.

Investigations

- Functional constipation does not warrant any specialist diagnostic tests.

Table 1 Comparison of the guidelines

	NICE CG99, NICE CKS62 guidelines	Joint ESPGHAN/NASPGHAN guidelines
Diagnosis	'Idiopathic' constipation Diagnosis based on history and physical examination	'Functional' constipation, using Rome III criteria Diagnosis based on history and physical examination
Investigations	Plain abdominal radiography, abdominal ultrasound and colonic transit studies only by specialist services in intractable idiopathic constipation Testing for coeliac disease and hypothyroidism only suggested in intractable constipation or with evidence of faltering growth	Plain abdominal radiography when faecal impaction is suspected but physical examination unreliable/not possible Colonic transit study may help discriminate between functional constipation and functional non-retentive faecal incontinence and in situations when the diagnosis is not clear Testing for hypothyroidism, coeliac disease and hypercalcaemia only in presence of red flag symptoms
Role of diet and physical activity	A balanced diet and daily physical activity is recommended, but not sufficient as first-line treatment	Normal fibre and fluid intake is recommended along with normal physical activity
Laxatives	PEG orally using an escalating dose regimen as first-line for faecal disimpaction Add a stimulant laxative (or in combination with an osmotic laxative) if PEG alone does not work	PEG orally for 3–6 days as first-line treatment for faecal impaction. Lactulose may be used if PEG is not available Milk of magnesia, mineral oil and stimulant laxatives may be considered as additional or second-line treatment
Enema	Sodium citrate enemas to be used only if all oral medications for disimpaction have failed. Phosphate enemas for disimpaction only to be used under specialist supervision	An enema once per day for 3–6 days is recommended for children with faecal impaction, if PEG is not available. PEG and enema are equally effective for treatment of faecal disimpaction

ESPGHAN, European Society for Paediatric Gastroenterology, Hepatology and Nutrition; NASPGHAN, North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition; NICE, National Institute for Health and Care Excellence; PEG, polyethylene glycol.

- Plain abdominal radiograph may be requested in intractable idiopathic constipation or *in a child in whom faecal impaction is suspected but physical examination is unreliable/not possible*.
- Ultrasound scan may be requested in intractable idiopathic constipation on specialist advice.
- Colonic transit study may be requested in intractable idiopathic constipation. *It may also be useful to discriminate between functional constipation, functional non-retentive faecal incontinence and in situations where the diagnosis is not clear.*
- Rectal biopsy is indicated where Hirschsprung's disease is suspected.
- *Anorectal manometry may be useful for evaluation of intractable constipation to evaluate the presence of a rectoanal inhibitory reflex as this excludes Hirschsprung's disease in older children and supports the diagnosis of idiopathic constipation.*
- Endoscopy or barium enema is not useful for the diagnosis of idiopathic constipation.
- *Routine testing for cow's milk protein allergy is not recommended.*
- Investigation for coeliac disease and hypothyroidism is suggested in intractable constipation and in cases where there is evidence of faltering growth with constipation.

Management

- Laxatives should be the first-line treatment for children with idiopathic constipation.
- For faecal disimpaction, polyethylene glycol (PEG) 3350 and electrolytes should be used as the first-line treatment with an escalating dose regimen to achieve response.
- Stimulant laxative (eg, sodium picosulfate, senna) should be added if PEG alone does not lead to disimpaction after 2 weeks of treatment.
- For children undergoing faecal disimpaction who do not tolerate PEG, a stimulant laxative singly or in combination with an osmotic laxative (lactulose or docusate) (for hard stools) can be used as a substitute.
- Rectal enema is indicated if all oral medications for disimpaction have failed.
- Start maintenance therapy with laxatives as soon as faecal disimpaction has been achieved. Maintenance dose of laxatives may be started at half the disimpaction

Box 1 Resources

- NICE (National Institute for Health and Care Excellence) (2010) clinical guidelines 99: https://www.nice.org.uk/guidance/cg99/resources/guidance-constipation-in-children-and-young-people-pdf
- ESPGHAN/NASPGHAN (European Society for Paediatric Gastroenterology, Hepatology and Nutrition/North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition) (2014) guidelines: http://www.naspghan.org/files/documents/pdfs/cme/jpgn/Evaluation_and_Treatment_of_Functional.24.pdf
- NICE (2014) quality standard 62: https://www.nice.org.uk/guidance/qs62
- ERIC (Education and Resources for Improving Childhood Continence): http://www.eric.org.uk/

Box 2 Rome III diagnostic criteria for functional constipation in children[5]

Must include two or more of the following in a child and should be fulfilled at least once per week for at least 2 months before diagnosis:

1. Two or fewer defecations in the toilet per week
2. At least one episode of faecal incontinence per week
3. History of retentive posturing or excessive volitional stool retention
4. History of painful or hard bowel movements
5. Presence of a large faecal mass in the rectum
6. History of large-diameter stools that may obstruct the toilet

dose and then gradually scaled down as per response. It can take several weeks to months before regular bowel habit is established or until toilet training is achieved.

- In children who are not faecally impacted, start maintenance therapy with PEG once the diagnosis of constipation is established. *A starting dose of 0.4 g/kg/day is recommended and the dose needs to be adjusted according to the clinical response.*
- Parental education, guidance and support are always required.
- Families should be informed that disimpaction treatment can initially increase symptoms of soiling and abdominal pain.
- It is necessary to ensure that a healthy balanced diet is maintained and sufficient fluids are consumed.
- Cow's milk exclusion from the diet should only be done on specialist advice on an individual basis. *A 2–4-week trial of avoidance of cow's milk protein may be indicated in children with intractable constipation.*
- Paediatricians need to be aware of the 'red flag' symptoms that indicate when further investigations or specialist advice may be required (see box 4).

WHAT DO I NEED TO KNOW?

What should I stop doing?

- Avoid using dietary interventions alone as first-line treatment for idiopathic constipation.

Box 3 Group of children who are at higher risk for developing severe constipation

Children and young people with:

- Physical disabilities, such as cerebral palsy (due to impaired mobility)
- Certain genetic disorders, for example, Down's syndrome
- Autism
- Previously treated for severe idiopathic constipation
- Under care of local authority

- Restrict use of enemas to cases where treatment with oral laxatives in optimal doses has failed in spite of being given regularly as directed.
- Do not routinely perform manual evacuation of the bowel under anaesthesia unless optimum treatment with oral and rectal medications has failed.
- Radiological investigations should not be routinely performed to diagnose idiopathic constipation.

What should I start doing?

- Start using a stool form chart (eg, Bristol stool form scale) to objectively describe stool consistency.
- Manage idiopathic constipation in children with oral PEG as first-line therapy.
- Perform full assessment (history and examination) of children and young people before making a diagnosis of idiopathic constipation.
- Patients undergoing disimpaction therapy should be reviewed within 1 week of initiating treatment to monitor response.
- Review patients (by phone or direct clinical review) within 6 weeks of starting maintenance therapy.
- Provide written information about laxatives to parents (and young people) at the initiation of therapy.
- Emphasise to parents (and community health professionals) that treatment failure with laxatives often occurs because of non-adherence or inappropriate dosing of laxative medications (eg, using low doses, abruptly stopping medications).

What can I continue to do as before?

- Provide education, guidance and reassurance to parents of otherwise healthy children without 'red flag' symptoms (see box 4) and explain the need for continuing laxatives.
- Be aware of 'red flag' symptoms in children with constipation (see box 4) and appropriately investigate these or refer on to appropriate specialists.

What should I do differently?

- Children and young people with idiopathic constipation who do not respond to initial optimal treatment initiated by primary/secondary health professionals within 3 months should be referred to a specialist with an interest in the diagnosis and treatment of constipation.
- *Conducting an MRI scan of the spine is only necessary for patients with intractable constipation with coexistent neurological abnormalities or spinal malformations.*

UNRESOLVED CONTROVERSIES

- The use of probiotic treatment in children with idiopathic constipation.
- *The role of a multidisciplinary team in the treatment of children with idiopathic constipation needs further evidence* and the benefits of including other health professionals such as psychologists remains uncertain.

Table 2 Symptoms and signs that may be associated with constipation

Symptoms	Signs
▶ Infrequent bowel activity	▶ Abdominal distension
▶ Foul-smelling wind and stools	▶ Hard faeces palpated in abdomen
▶ Excessive flatulence	▶ Faecal mass palpated in suprapubic region
▶ Irregular stool texture	▶ Hard faeces palpated on digital anorectal examination
▶ Passing occasional enormous stools or frequent small pellets (rabbit droppings)	▶ Anal fissures
▶ Withholding or straining to stop passage of stools	▶ Visible hard faeces at anal opening
▶ Soiling or overflow	▶ Reluctance to allow abdominal examination as apprehensive of pain
▶ Bleeding associated with hard stool	
▶ Abdominal pain, bloating or discomfort	
▶ Poor appetite that improves with passage of large stool	
▶ Lack of energy	
▶ Unhappy, angry or irritable mood and general malaise	
▶ Painful defaecation	

- *The use of behavioural therapy or biofeedback in the treatment of childhood constipation.*
- *The use of cow's milk protein-free formula to empirically treat infants with constipation and the need for allergy testing in such cases.*
- *The efficacy and safety of novel therapies need to be established in children. Drugs such as lubiprostone, linaclotide and prucalopride have been shown to be effective in adults with constipation.* Early studies have shown encouraging results with prucalopride and lubiprostone in treating children with functional constipation.[6 7] *The role of transcutaneous electrical stimulation in intractable constipation needs further research.*

Several other issues not mentioned in the guideline are summarised in box 5.

Box 4 Red flag symptoms that needs specialist opinion

- Reported from birth or first few weeks of life
- Failure or delay in passage of meconium (>48 h after birth (in term baby)) suggestive of cystic fibrosis or Hirschsprung's disease
- Ribbon or toothpaste like stool suggestive of anal atresia
- New-onset weakness in legs, locomotor delay
- Abdominal distension with (bilious) vomiting suggestive of malrotation or volvulus
- Faltering growth and/or coarse facies suggestive of (congenital) hypothyroidism

Box 5 Critical review

- Doses of laxative higher than recommended in the British National Formulary for Children (BNFC) will be necessary for managing children with faecal impaction and subsequent maintenance therapy as recommended by the National Institute for Health and Care Excellence (NICE) guidelines.
- Parents need to be empowered in adjusting doses of laxatives up and down to achieve regular bowel habit without soiling.
- Radiological investigations may be necessary in unclear cases or where the family may be reluctant to accept a clinical diagnosis but should only be requested by specialist services.
- Involvement of paediatric gastroenterologists will be necessary only in a small number of cases (failure of response in faecal disimpaction in spite of optimum oral or rectal medication, intractable constipation) to confirm the diagnosis of constipation and initiate appropriate therapy. Ongoing management can then be led by the primary or secondary care professionals.
- *Do not treat 'infant dyschezia'* (characterised by at least 10 min of straining and crying before successful passage of soft stools in healthy infants aged <6 months) with laxatives.[5]
- Well-written parent information is available from ERIC (Education and Resources for Improving Childhood Continence) and clinicians may find these as useful while counselling parents.

Clinical bottom line

- Idiopathic constipation is common in paediatric practice. It can present with infrequent defaecation, hard faeces, abdominal pain, poor appetite or overflow spurious diarrhoea.
- First-line management of constipation (either with or without faecal impaction) is to use oral laxatives and PEG should be the treatment of choice.
- Dietary and lifestyle modifications may be used as adjunctive measures in addition to laxative therapy.
- Radiological investigations should be reserved for children with intractable constipation and those with faecal impaction where the diagnosis is difficult to make clinically.
- Idiopathic constipation which remains unresponsive to optimal treatment for 3 months should be referred to a specialist with a special interest in childhood constipation.
- Children and young people with 'red flag' symptoms should prompt further evaluation and not be empirically commenced on laxatives.

Correction notice The paper has been amended since it was published Online First. In the section titled UNRESOLVED CONTROVERSIES in the second bullet point the word "community" was inadvertently retained, this has now been deleted.

Contributors All the authors contributed equally towards writing the manuscript. CHS provided expert opinion in addition to editing the manuscript.

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