Management of chronic constipation in children☆

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Abstract

Children are commonly affected by constipation. Optimal management of chronic constipation requires a good understanding of the underlying pathophysiology. The presentation and management of constipation varies by age. This review aims to give the reader a clear guide to diagnosis, investigation, pharmacological and non-pharmacological management of chronic constipation in children. We describe the features typically evident in the clinical history and how the pathology can interrupt normal physiology. We outline the age dependent presentation and management of chronic, functional constipation based on the best available evidence and examine the NICE guideline for laxative use in children.

Keywords age dependent presentation; children; chronic constipation; clinical diagnosis; investigations; laxative treatment; non pharmacological treatment; red flag symptoms and signs

Introduction

Constipation, derived from the Latin 'constipare', meaning 'to cram together', is the commonest gastrointestinal disorder comprising up to 25% of referrals to tertiary paediatric gastroenterology clinics. In primary, secondary and tertiary care, there are more consultations for constipation management than for other periodic, chronic conditions such as asthma or migraine. Chronic constipation is a heterogeneous group of disorders, and is often late-presenting. It is defined by infrequent and or difficult passage of stools, and is a clinical diagnosis that should be based on symptoms that fulfil the ROME III criteria (see Table 1).

Prevalence

The reported prevalence of constipation varies from 0.8 to 28% and the condition has a wide geographic variability, with the highest reported prevalence in the USA and the lowest in Finland. Pathogenesis is multifactorial with research focussing on environmental factors, behavioural problems and genetic predisposition. Environmental factors such as activity level and diet but also low maternal education level or social circumstance play a part. The association with behaviours is complex because constipation can be both caused by and cause changes in behaviour. Significantly higher rates of constipation have been reported amongst mothers of constipated children as opposed to fathers or siblings of a constipated child. A genetic component is likely to be part of the pathogenesis of functional constipation but no mutations in specific genes have been linked.

Like many other functional gastrointestinal disorders the pathophysiology and prognosis are variably understood by medical practitioners. This results in a large variety of strongly held beliefs and management strategies. To understand constipation in childhood it is necessary to have a good knowledge of normal physiology, the wide range of normality and the role of diet and behaviour.

Diagnosis of functional constipation is as per the ROME III criteria

Infants up to 4 years should have at least two symptoms for 1 month prior to diagnosis and those over 4 years at least two of the following symptoms present for the previous 2 months:*

1. Two or fewer defaecations per week
2. At least one episode of faecal incontinence per week
3. Retentive posturing or stool retention
4. Painful or hard bowel movements
5. Presence of a large faecal mass in the rectum
6. Large diameter stools that may obstruct the toilet

* Without objective evidence of a pathological condition and without fulfilling irritable bowel syndrome criteria.

Table 1

Physiology of defaecation

Enteric content enters the colon via the ileocaecal valve. Stools are formed by the progressive absorption of water, and are propelled along the colon to the rectum. Stool is stored until a socially acceptable time to defecate. The rectum stores and eliminates stool through a complex mechanism involving pelvic floor muscles, the autonomic and somatic nervous systems. The anorectal angle, formed by the anal sphincter complex and puborectalis muscle is crucial to successful storage and defaecation. This angle is 85–105° at rest. The rectum is usually empty but distension of the rectal wall with stool descending from the sigmoid colon causes a temporary reflex relaxation of the internal anal sphincter allowing stool to come into contact with sensitive receptors in the anal canal. The rectoanal inhibitory reflex results in a contraction of the internal sphincter, inhibiting defecation. The process, however, alerts the individual to the presence of stool, liquid or flatus in the rectum. An indication of the exquisite sensory innervation of the anorectum is the ability to distinguish between distension caused by solid, liquid or gas.

Once a child has an opinion about the appropriate time to respond to this signal, a voluntary process of defaecation is either begun or overruled. If the sensation on passing stool is pain (for example when the stool is both large and hard) then even very young children can resist the urge to push. Such stool withholding – often misinterpreted as straining to evacuate stool – is frequent in toddlers. If the individual decides the time is right

☆ This article is based on a previously published article of the same name, published in this journal issue 22(10): 401–8.

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then increased intrarectal pressure comes from straining of intra-abdominal muscles and pelvic floor muscles to push faeces towards the anal canal. The puborectalis muscle relaxes to allow the descent of the pelvic floor, straightening the anorectal angle and inhibiting the internal and external anal sphincters, allowing faeces to be expelled. In newborn babies and very young infants the voluntary element of control is not yet developed so defaecation occurs following initial distension of the rectal wall.

Pathophysiology of constipation

Whilst the majority (more than 90%) of children with chronic constipation will be considered to have functional, idiopathic constipation, exclusion of organic causes is important (summarised in Table 2). Constipation is also an important side effect of several classes of medicines and is commonly encountered in children receiving opiates, antacids or iron.

Coeliac disease is commonly thought of as causing diarrhoea, but constipation is seen, possibly due to anorexia or changes in ileal function or gut motility. Constipation is very common in children and adults with cystic fibrosis, where there is an association with low total fat absorption and a history of meconium ileus.

Neuromuscular conditions can affect the gut. Smooth muscle cells and intestinal cells of Cajal play a major role in normal gut motility. These cells ensure regular contractions of the colonic wall and propulsion of content. Constipation is often seen in patients with cerebral palsy and Duchenne muscular dystrophy. In patients with spinal muscular atrophy (SMA) proximal muscle weakness is a cardinal feature. In most of these patients constipation is a problem because of reduced abdominal muscle tone as well as disturbed innovation of the myenteric plexus.

Children with significant developmental delay are more prone to constipation for a wide variety of reasons depending on their underlying disorder. For example, children with cerebral palsy often have dysmotility problems. Disorders that affect the enteric nervous system such as hereditary sensory and autonomic neuropathy (previously known as Riley—Day syndrome) are associated with constipation. Normal gastrointestinal motility is disturbed by abnormal autonomic function.

Constipation can be a feature of disorders that affect water/electrolyte balance such as diabetes insipidus. This can lead to reduced water content i.e. harder stools or as a result of muscle weakness caused by electrolyte imbalance. Other endocrine diseases such as MEN3 and hypothyroidism can present with constipation.

It is important that there is a general awareness of the wide range of rare pathologies that may present with constipation as effective management depends on an understanding of the underlying pathophysiology. Constipation should be regarded as a symptom and not a disease. Anorectal malformations and Hirschsprung's disease are amongst the commoner pathologies underlying very early onset childhood constipation.

History

Pitfalls

Although the ROME III criteria (Table 1) appear self-explanatory, history-taking can be difficult. The key features in history are described in Box 1. Care must be taken. Often history depends upon reports by parents or other carers, and may be subject to over- or under-reporting bias. Functional constipation may often present late, or with abdominal pain or spurious diarrhoea. A large faecal mass in the rectum gives the sensation of incomplete evacuation and children may try to open their bowels several times a day. If only small amounts of soft/liquid stools are passed around the sides of the obstructive faecal mass, this is termed overflow, or overflow diarrhoea. Anal canal trauma from passage of hard or large stool can present with bright red rectal bleeding or severe anal pain. Pain may exacerbate the problem, as it will inhibit defecation.

Key features in history taking

- Delay in passage of meconium
- Age at onset
- Relation to toilet training
- Toileting history-stool frequency, consistency, pain, soiling, presence of blood
- Stool withholding behaviour
- Urinary symptoms (13% of those with constipation have urinary symptoms)
- Abdominal pain
- Diet-history of exacerbation e.g. with cow's milk or a poor diet low in fruit and fibre
- General health and developmental milestones
- Family and social history

Box 1

Organic causes of childhood constipation

Structural colorectal	Spinal cord	Systemic	Neuropathic lesions of intestine	Drugs	Other important causes
Anal stenosis	Spina bifida	Diabetes	Hirschsprung disease	Opioid analgesia	Coeliac disease
Post NEC/IBD stricture	Sacral agenesis	Hypothyroidism	Intestinal neuronal dysplasia	Antacids	Cystic fibrosis
Chronic intestinal pseudo- obstruction	Spinal cord tumours	Hypo/hypercalcaemia		Iron	Cows milk protein allergy
		Neurofibromatosis		Cholestyramine	
		Cerebral palsy			

Table 2