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The Role of Diagnostic Tests in Constipation in Children

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1. Introduction

The diagnosis of constipation is usually suspected based on the presence of certain symptoms. These may include infrequent passage of stool, stools that are hard or difficult to pass or the presence of faecal incontinence. A careful clinical history and focussed physical examination are often all that is needed to confirm the diagnosis. There are a number of symptom-based classification tools that have been designed for diagnostic use in clinical practice. These classifications have evolved over time but there remains no universally accepted gold standard. The most recently published classification tool is the Rome III classification system.

In certain situations, further diagnostic investigations may be required to confirm the diagnosis or elicit further details to assist in optimal management of the patient. These investigations may range from simple blood tests or abdominal radiography to more complex measures of colonic transit and function that are only available in specialised centres.

This chapter will examine, evaluate and define the role of each of these tests using an evidence based approach. It will determine when it is appropriate to do physiological or other testing in constipated children and assist clinicians in selecting the most appropriate modality of investigation for their patients.

2. Physiology of defaecation

The process of defaecation relies upon a complex interplay between pelvic muscles, muscles of the internal and external anal sphincters and the autonomic and somatic nervous systems.

Faecal matter is moved from the colon into the rectum by peristaltic propagation. The presence of faecal matter in the rectum stretches the rectal wall and the puborectalis and levator ani muscles relax. Distension of the rectum induces a parasympathetic response involving contraction of the rectal walls and relaxation of the internal anal sphincter (recto-anal inhibitory reflex).

When faeces enter the anal canal, anal receptors are activated and the voluntary component of the process is initiated. In an appropriate environment and social situation, the external anal sphincter and puborectalis muscle relax and there is simultaneous contraction of the levator ani, abdominal and diaphragm muscles. At this time, defaecation occurs and faecal matter is evacuated from the body.

In instances where the environment or social situation is unsuitable for defaecation to occur, the external anal sphincter voluntarily remains contracted with the help of the pelvic floor muscles. This occurs for a few seconds until the rectal wall is able to adapt and distend to allow for storage of the additional rectal volume.

Some children may achieve voluntary bowel control around the age of 18 months but there is variability in the age of attainment of complete bowel control. Most children will achieve bladder and bowel control and be toilet trained by the age of 3 years.

3. Physiology of constipation

Some children will have an underlying organic cause for their constipation. These children may be identified by the presence of 'red flag' signs on history taking and examination or characteristic findings on diagnostic investigations. The underlying physiological process will differ based on the individual aetiology e.g. Hirschprung disease is caused by absence of enteric nerves and functional obstruction compared with mechanical obstruction in cases of anal stenosis or atresia.

However, in at least 90% of children, there is no underlying organic cause found for constipation and it is termed 'functional constipation'. Withholding behaviour plays an important role in the development of functional constipation in infants, toddlers and young children. These behaviours can originate from an experience of painful defaecation (e.g. related to passage of hard stools or anal fissures), a lack of regular routine with toileting or environmental factors including unfamiliar bathroom environment associated with time of commencement of school.

Withholding behaviours may manifest as grunting or back arching in infants or clenching of the buttocks and repetitive rocking / fidgeting actions in older children. When stool is withheld, the rectal wall adapts and distends to allow for the storage of faecal material. Over time, stool accumulates in the rectum and larger, harder faecal matter is formed, which is then associated with further pain on attempted defaecation. This cycle of persistent painful defaecation can lead to further retentive posturing and toilet avoidance. With time, increasing rectal distension can result in rectal insensitivity and faecal incontinence with a significant impact on the child's quality of life.

4. The use of diagnostic tests in constipation in adults

A recent review summarised the different diagnostic tests available for use in adult constipation (Rao & Meduri, 2011). Using the available evidence, graded recommendations were given for each diagnostic test. These recommendations were based on the presence and quality of evidence in favour of the test in addition to information on specificity, sensitivity, accuracy and predictive values.

Table 1 summarises these findings.

Key:

Grade A1: Excellent evidence in favour of the test based on high specificity, sensitivity, accuracy and positive predictive values.

Grade B2: Good evidence in favour of the test with some evidence on specificity, sensitivity, accuracy and predictive values.

Grade B3: Fair evidence in favour of the test with some evidence on specificity, sensitivity, accuracy and predictive values.
Grade C: Poor evidence in favour of the test with some evidence on specificity, sensitivity, accuracy and predictive values.

Test		Evidence	Recommended Grade
Blood tests		No evidence	C
Imaging	Abdominal Xray	Poor	C
	Barium enema	Poor	C
	Defecography	Fair	B3
Anorectal Ultrasound		Poor	C
Magnetic Resonance Imaging		Fair	B3
Flexible sigmoidoscopy and colonoscopy		Poor	C
Gastrointestinal transit studies	Colonic transit with radiopaque markers	Good	B2
	Colonic transit with scintigraphy	Good	B2
	Wireless motility capsule	Excellent	A1
Manometric studies	Anorectal manometry	Good	B2
	Colonic manometry	Good	B2

Table 1. Summary of diagnostic tests and their recommended grade in adults (modified from Rao & Meduri, 2011).

Table 1 illustrates the variability in the quality of evidence supporting the use of investigations in the diagnosis of adult constipation. In practice, the diagnosis of constipation still relies heavily on careful history-taking and detailed clinical examination technique.

To date, there are limited studies assessing the role of diagnostic tests in constipation in children. A review by Baker and colleagues in 1999 graded the quality of evidence for limited investigations in children using methods of the Canadian Preventive Services Task Force. They found that the evidence for abdominal radiography, when interpreted carefully was II-2 (*evidence obtained from well-designed cohort or case-control analytic studies, preferably more than one centre or research group). The evidence for rectal biopsy and rectal manometry in reliable exclusion of Hirschprung disease was II-1 (evidence obtained from well-designed cohort or case-control trials without randomisation). Measurement of transit time using radiopaque markers was graded as II-2* (Baker et al., 1999).

There is limited availability of a supportive consensus guideline for choice of diagnostic tests in children with constipation. This makes the selection of investigations and their interpretation a challenge for many clinicians. This chapter aims to assess the data available with particular focus on the paediatric population group. It outlines both the benefits and limitations of each individual technique to allow clinicians to make an informed decision about when to employ their use.

5. The role of radiography

5.1 Abdominal radiography

The diagnosis of constipation is usually a clinical decision based on good history-taking and physical examination of the child. In clinical practice, abdominal radiography is still performed by some clinicians as part of their initial diagnostic assessment or as a tool to assess and monitor treatment response. There may be a role for abdominal radiography in certain circumstances and these will be discussed below.

Abdominal radiography may be indicated when the diagnosis of chronic constipation is strongly suspected but in doubt because of a lack of supportive evidence on history or examination. In particular, supportive physical signs (in the form of palpable abdominal faecal masses) may be difficult to elicit in obese children or children who have been frequently using stool softeners. Faecal impaction can be suspected clinically and is usually able to be confirmed by rectal examination. However, there may be exceptions where despite a strong suspicion of chronic constipation, there is no palpable faecal retention on rectal examination. The clinical practice guideline of the North American Society for Pediatric Gastroenterology and Nutrition recommends the use of a plain abdominal radiograph for diagnosing constipation in cases where there is uncertainty about the presence of constipation.

Rectal examination is an invasive procedure. Consequently, in some children it may be contraindicated (e.g. in the presence of a history of previous sexual abuse or by the degree of associated psychological distress and angst it causes the child). In these children there may be a role for abdominal radiography in place of a rectal examination to confirm faecal impaction and exclude bowel obstruction prior to the commencement of bowel washout.

Abdominal radiography can be employed in the different circumstances described above. It may also be useful to give a measure of megarectum. The benefits of abdominal radiography include that it is an easily accessible, non-invasive and relatively inexpensive investigation. However, this modality does have some limitations that need to be factored in when it is being considered. Every radiograph performed provides the child with a small dose of ionizing radiation. In isolation, this is unlikely to have direct impact on their wellbeing, but cumulative doses of radiation may be potentially harmful for an individual's health.

The interpretation of abdominal radiography is variable. It has traditionally been largely subjective and open to individual interpretation. In an attempt to objectify the classification of this tool, a number of different scoring systems have been created including the Leech and Barr scoring systems. The Leech score assesses the large intestine in 3 segments: right colon, left colon and rectosigmoid colon and provides a score from 0 to 5 for each segment based on the amount of faeces present (0= no faeces visible to 5= severe faecal loading with bowel dilatation). An overall score out of 15 is obtained, with a score of 9 or greater being positive for constipation. The Barr score divides the large intestine into 4 segments: ascending colon, transverse colon, descending colon and rectum. It quantifies both the amount and consistency of the faeces (e.g. granular, rock-like) and gives a score out of 22 with a score of 10 or greater being positive for constipation (Pensabene et al., 2010).

Despite these formalised classification systems, studies comparing different scoring methods show that there is still a degree of inter-observer variability. This applies to interpretation by both paediatric clinicians and radiologists. Overall, individual scoring

systems used for interpretation of faecal loading on abdominal radiograph have a low sensitivity (Pensabene et al., 2010).

A recent systematic review assessed the relationship between clinical symptoms and signs of constipation and the presence of faecal loading on abdominal radiography. The availability of good quality literature was limited, however the study concluded that conflicting evidence exists for an association between the clinical and radiographic diagnosis of constipation. The high quality studies that were assessed in the review found that a radiographic diagnosis of constipation occurs almost as often in clinically constipated children as in clinically non-constipated children. Furthermore, they found that the results of rectal examination were not consistently associated with the presence of fecal retention on abdominal radiography. The review concluded that there is inadequate evidence to support the North American Society for Pediatric Gastroenterology and Nutrition clinical practice guideline and those clinicians who recommend a plain abdominal radiograph in cases of uncertainty of the presence of constipation in a child (Reuchlin-Vroklage LM et al., 2005).

There is a clear need for further clinical research to assess the precise role of abdominal radiography in the diagnosis of constipation in children. Current data provides conflicting opinion and challenging interpretation for clinicians. A better availability of future quality literature shall help to determine the indications for this investigation and validate its use in clinical practice.

5.2 Contrast radiography

5.2.1 Barium enema

A barium enema (lower gastrointestinal series) is a diagnostic procedure where opaque contrast medium (barium sulfate) is infused into the colon via a rectal enema tube. The flow of barium sulfate is captured by using fluoroscope xray pictures. The patient may be asked to move into different positions to obtain optimal detailed anatomical images of the gastrointestinal tract.

Barium enema can be useful in some instances for the identification of anatomical abnormalities including megacolon, megarectum or rectal masses. It may also be used as an initial screening for Hirschsprung's Disease (a condition characterised by the absence of ganglion cells in the myenteric plexus of the distal colon). The visualisation of a transition radiographic zone and delayed barium emptying is suggestive of Hirschsprung's Disease however not diagnostic. A rectal biopsy is still required to confirm this diagnosis. In neonates, the absence of a transition zone may be a normal variant making this test less useful in this particular age group.

Limitations associated with this modality include associated radiation exposure and the invasive nature of the procedure (requiring placement of an enema tube into the rectum).

Barium enema has little or no role as a routine investigation in the workup of children with chronic constipation but may be used to assess gastrointestinal anatomy in some patients.

5.2.2 Defaecating proctography

Defaecating proctography assesses the mechanics of defaecation in real time using fluoroscopy. A barium paste is manually infused into the rectum using specialised

equipment until there is adequate distension. The patient then moves to a portable plastic commode and their process of defaecation is recorded by an x-ray camera.

Defaecating proctography is not commonly performed in current practice. It may have a limited role in assessment of pelvic floor dysfunction (including anismus) in obstructed defaecation. Its main limitations include associated radiation exposure and invasive nature of the procedure.

6. The role of blood tests

Any child undergoing assessment for chronic functional or intractable constipation should have certain blood tests done to exclude an underlying organic cause. This is particularly the case where there are clinical signs or symptoms present that are suggestive of an underlying metabolic or pathological process.

Patients should undergo a complete blood count and biochemical profile, in particular looking at serum calcium levels to exclude hypercalcemia and blood glucose levels to look for evidence of diabetes. Thyroid function tests should be done to exclude hypothyroidism and a coeliac screen and total IgA to assess for evidence of coeliac disease. A diagnosis of coeliac disease can only be confirmed by endoscopy and small intestinal biopsy.

Less commonly, measurement of blood lead levels may be indicated to exclude lead toxicity as an aetiological factor.

7. The role of ultrasound

Ultrasound scan is a safe, non invasive and easily accessible mode of imaging. It is not currently widely utilised in children with constipation, but has a potential role in quantifying the degree of faecal loading / megarectum and monitoring of treatment response.

7.1 Pelvic ultrasound

Pelvic ultrasound scan can be used to visualise faeces of both hard and soft consistency. One group in the United Kingdom have successfully used pelvic ultrasound together with a scoring system in their outpatient management of constipated children since 2007 (Lakshminarayanan B et al., 2008). Their findings showed that the presence of faecal loading on ultrasound correlated highly with clinical symptoms of constipation on history taking. In addition, of the 269 patients (54%) with no palpable faeces on clinical abdominal examination, 31% of them showed significant faecal loading on ultrasound. This finding supports the notion that despite a thorough history and physical examination, some patients with constipation may still be missed. In patients in whom there remains an ongoing clinical suspicion of constipation, there may be a role for investigations such as ultrasound scan. Lakshminarayanan and colleagues successfully illustrated the use of this modality to diagnose and monitor treatment response of their patients in outpatient clinical practice.

Other studies have attempted to quantify the degree of constipation by using rectal diameter and other measurements on ultrasound scan (Karaman et al., 2010; Joensson et al., 2008). These groups found that a thicker mean rectal diameter correlated with a clinical diagnosis

of constipation by Rome III criteria. Furthermore, they found that the amount of faecal loading on ultrasound decreased by a significant amount after one month of treatment supporting a role for this modality in monitoring of treatment response.

Karaman and colleagues could identify no inter-observer difference between 2 different radiologists performing the ultrasound scans. This supports a degree of reliability between different users of ultrasound scan as an imaging modality. Further detailed studies to assess for inter-observer differences between paediatric clinicians and radiologists interpreting ultrasound in this context would be useful.

Pelvic ultrasound is appropriate for use in the outpatient setting. It has no associated radiation dose. It is a non-invasive procedure and in the hands of experienced staff, tends to be well tolerated by children. There are only limited studies available on the use of pelvic ultrasound in constipated children and further research would be beneficial to help ascertain the precise role for this modality in the assessment and long term monitoring of this patient group.

7.2 Endoanal ultrasound

Endoanal ultrasound involves insertion of an ultrasound probe into the anus allowing visualisation of the internal sphincter, external sphincter and puborectalis muscles. It can be used to provide information about the anatomical course of anal fistulae and some anal abscesses.

Endoanal ultrasound is relatively quick and simple to perform. There is no associated radiation dose. In some children it may not be well tolerated due to its invasive nature and may require the use of sedation or general anaesthetic in order to perform effectively. In addition, patients must undergo an enema a few hours beforehand to ensure adequate clearance of the rectal area prior to scanning.

8. The role of gastrointestinal transit studies

Colonic transit studies have traditionally provided information about total and segmental colonic transit time and overall colorectal motor function. There are 2 standard techniques performed: radio-opaque marker studies and radio-nuclide scintigraphy. Both techniques give similar information for ascending and transverse colon motility but radio-opaque marker studies generally produce faster total transit time (Southwell et al., 2009).

Colonic transit studies classify children with constipation into 3 subgroups:

1. Children with normal colonic transit time
2. Children with outlet obstruction
3. Children with slow transit constipation.

Colonic transit studies can be used to differentiate slow-transit constipation from pelvic dyssynergia. In clinical practice, this information can be useful to identify patients with motility disorders including Hirschsprung's disease and chronic intestinal pseudo-obstruction. It can also be used to help differentiate children with functional constipation and overflow incontinence from those with nonretentive faecal incontinence (Benninga et al. 1994). This is important as management differs between the two conditions.

Colonic transit studies may also have a role in predicting patient prognosis. One study showed a colon transit time of > 100 hours was associated with a poor treatment outcome at one year (de Lorijn et al., 2004). The range of normal colonic transit is 20-56 hours and there is little variation between children and adults (Southwell et al., 2009).

A more recent innovation is the use of wireless capsule technology. The additional benefits of radio-nuclide scintigraphy and wireless capsule monitoring are their capacity to give information about gastrointestinal transit in the stomach and small intestine as well as the colon. In cases of severe refractory constipation, this data may be useful in pre-operative assessment to aid decision-making about the portion of bowel for resection and the best position(s) for stoma creation. Information on gastric and small intestinal motility may also be useful in children with abnormal gastric emptying to help decide on appropriate methods of feeding (e.g. gastrostomy versus jejunostomy feeds).

8.1 Radio-opaque marker studies

This technique was first pioneered in adults in the late 1960s. There are a number of variations in its application including a few more commonly used methods. Firstly, there is the 'simple' radio-opaque marker test where a single capsule is swallowed (containing 20-50 markers) and a single abdominal radiograph is taken 4-5 days later showing the location of the markers. Alternatively, a single capsule (containing multiple markers) is ingested and radiographs are performed every 24 hours until the markers are no longer visible. A third technique is the 'multiple markers' test where a single capsule is ingested daily for 3 days (each containing a different shaped marker) and abdominal radiographs are taken at days 4 and 7 after ingestion. The different marker shapes help to identify their individual locations.

Delayed transit is defined as retention of more than 20% of markers at the time of abdominal radiograph (96 hours for the 'simple' test and 120 hours for the 'multiple markers' test) (Dinning and Di Lorenzo, 2011). Children should refrain from taking laxatives in the weeks before the study as these may affect bowel function and subsequent results.

Radio-opaque marker studies are inexpensive, relatively widely available and are useful in the identification of slow transit constipation. Their downside is the associated variability with the use of different methodologies and the lack of information gained regarding transit in the rest of the gastrointestinal tract.

8.2 Radio-nuclide scintigraphy

Radio-nuclide scintigraphy has been utilised since the mid 1980s. It involves oral ingestion of a labelled isotope followed by gamma camera scans at various intervals up to 5 days (depending on the specific method used). The progression of the isotope throughout colonic regions is plotted using graphs. It is possible to calculate the amount of isotope residue at each region for each time interval. Various measures of isotope retention can be used to diagnose and quantify delayed colonic transit. These include transit time in hours, % radioactivity retained, proximal colonic emptying and centre of mass.

Radio-nuclide scintigraphy is expensive and requires an appropriately equipped and trained specialist centre which may not be readily accessible to all clinicians. In addition, results of different studies may not be directly comparable due to differences in the method

of isotope administration (e.g. liquid versus solid markers) and varying measurements of interpretation used. However, radio-nuclide scintigraphy can be useful for classification of pathophysiological subtypes of constipation as outlined in the introduction to section 6.

8.3 Wireless capsule technology

This is a relatively recent innovation. It involves an ingestible gastrointestinal capsule, a receiver worn by the patient during the study period, a receiver docking station and display software. The capsule is able to identify and transmit information about intraluminal pH, pressure and temperature and has a battery life of 5 days.

To date, there are no large studies looking at the use of wireless capsule technology in children and little clinical experience from clinical application in this population group. Limited research available in adult patients has compared wireless capsule assessment of gastrointestinal motility with radio-nuclide scintigraphy. A recent study performed simultaneous whole gut scintigraphy and wireless capsule monitoring of whole gut transit in 10 adults. They found a very strong correlation between measurements of gastric emptying by the two different methods (Maqbool et al, 2009).

Potential benefits of this technique include no associated radiation exposure and its capacity to provide information about stomach and small intestine transit as well as colonic transit (by its ability to monitor pH and thus detect progression from the stomach into the small intestine).

In children, this technique may have some limitations. It relies upon the ingestion of a wireless capsule which some younger children may find difficult or unappealing. An alternative option is placement of the capsule into the stomach at the time of an endoscopy.

This technique is relatively new to clinical practice and further studies are needed to elucidate its precise role in the investigation of children with constipation.

9. Manometric studies

Manometry involves the measurement of pressures in various segments of the gastrointestinal tract to provide information on gastrointestinal function and motility. Both water perfused and solid state catheters have been used.

9.1 Anorectal manometry

Anorectal manometry assesses the motor function of the anal sphincters, sensory thresholds to rectal distension, recto-anal inhibitory reflexes and coordination of evacuation. Some degree of bowel preparation is required beforehand. A transducer is inserted through the anus with the child in the left lateral position and a sphincter pressure profile measured. Transient inflation of a balloon at the tip of the catheter assembly allows measurement of sensory thresholds, and also confirms normal reflex relaxation of the internal anal sphincter (rectoanal inhibitory reflex). Attempted evacuation of the rectal balloon allows examination of the normal coordination of evacuation as well as the response to withholding (squeezing).

Anorectal manometry is useful to exclude sphincteric damage, and rule out Hirschsprung disease. Blunted sensation to rectal distension and megarectum can also be defined.

Anorectal manometry also has a role in the identification of dyssynergic defaecation and altered rectal sensation in children with constipation (Rao et al., 2011). The normal process of defaecation involves relaxation of the puborectalis muscle and external anal sphincter combined with an adequate propulsive force. Pelvic floor dyssynergia occurs when there is abnormal pelvic floor muscle relaxation or even paradoxical contraction during the process of attempted defaecation. These children are unable to produce the coordination required for normal defaecation and frequently become constipated.

Identification of dyssynergic defaecation is important as it changes management intervention. These children do not require laxative management but will benefit from behavioural interventions and possibly from the use of biofeedback therapy (Chiarioni et al., 2006). Biofeedback therapy is a method of neuromuscular re-education. A computer and video monitor are used to display bodily processes to a patient that they are normally unaware of. Increased awareness of these behaviours provides an opportunity to consciously modify one's responses to a more acceptable pattern.

The most common problem associated with the use of anorectal manometry is that it is limited to certain specialised centres. Other complications may include a lack of standardisation in technique between different transducers and technical errors related to inadequate balloon sufflation or positioning (Benninga et al., 2004).

Anorectal manometry provides information on pathophysiological abnormalities underlying childhood constipation.

9.1.1 Rectal barostat test

The rectal barostat test consists of a highly compliant balloon placed in the rectum and connected to a barostat (computerised pressure-distending device). This device is able to record detailed information on tone, compliance and assess rectal sensation.

The information obtained by this technique can be useful in identification of megarectum, hyper- or hypo- compliant rectum. Rectal compliance, sensation and function are all closely inter-related.

9.2 Colonic manometry

Colonic manometry provides a detailed picture of overall motor activity of the colon. The American Neurogastroenterology and Motility Society recently recommended its use in assessment of severely constipated children unresponsive to medical therapy, with evidence of slow colonic transit and the absence of an evacuatory disorder (Camilleri et al., 2008). It can also be used to differentiate functional constipation from constipation secondary to an underlying neuromuscular cause.

One study looked at the indications for colonic manometry in a group of 146 children referred to a tertiary centre in the USA (Pensabene et al., 2003). The 4 main indications identified for use of colonic manometry were:

1. Clarification of pathophysiology of lower gastrointestinal symptoms (68%)
2. Clarification of pathophysiology of persisting lower gastrointestinal symptoms after surgery for Hirschsprung's Disease (14%)

3. Confirmation of diagnosis of intestinal pseudo-obstruction (11%)
4. Assistance with decision about re-anastomosis of a diverted colon (7%).

Colonic manometry is performed by using colonic catheters that incorporate multiple recording ports or sensors. These provide a feedback of information on intraluminal pressures to a recording system. The catheters can be water-perfused or solid state, with most paediatric centres favouring the former (Dinning PG & Di Lorenzo C, 2011). Catheters are usually placed by colonoscopy or via radiological guidance such as fluoroscopy after adequate colonic clearance using bowel preparation. They can be placed into the mouth, nose, anus (most common) or through an existing stoma. The study period generally lasts around 5 hours but may be up to 24 hours in duration.

Water-perfused catheters consist of flexible tubing with multiple recording ports connected to a pneumohydraulic infusion pump that provides a constant flow of water. Colonic wall contractions occlude the ports and impede water flow and this resistance to flow is transmitted as pressure change to external transducers. These catheters are relatively cost effective and some are autoclaveable and re-usable. On the downside, patients are confined to bed for the duration of the study and uncertainty exists regarding whether large amounts of infused water may have deleterious effects in young children (Dinning et al., 2010).

Solid-state catheters comprise strain gauges embedded into a flexible tube. Each gauge connects to a recording system by fine wiring. Used with portable recorders, these catheters allow the patient to be ambulant because they don't rely on constant water flow. However they do tend to be more expensive than water-perfused catheters (Dinning et al., 2010).

There are 2 types of normal colonic motor activity:

1. Low- amplitude tonic and phasic contractions (mixing of luminal contents)
2. High-amplitude propagated contractions (propulsion of stool from colon to rectum).

In addition, colonic motility should increase after a meal (gastro-colonic response) and respond to other physiological stimuli such as morning waking and exercise (Hussain SZ & Di Lorenzo C, 2002). The presence of these features of normal colonic motility in the context of constipation suggests a behavioural cause or functional constipation.

In the case of abnormalities, colonic manometry can distinguish between an underlying neuropathy and myopathy. Weak or absent colonic contractions in the absence of generalised colonic dilatation suggests a colonic myopathy. Absent, disordered or abnormal colonic contractions combined with an absent gastro-colonic meal response suggests the presence of an underlying neuropathy.

Colonic manometry may also have a role in monitoring of treatment response in constipation management. A study by Pensabene et al in 2003 showed that the results of colonic manometry resulted in recommendations to adjust management plans (mostly surgical intervention) in 93% of 146 children. Of the 98 patients that were able to be contacted for follow up data, 88% had parents who believed that these interventions had been a positive impact on their child's health.

Colonic manometry is a useful diagnostic tool in childhood chronic constipation and a number of international centres are trained in this technique. Compared with adults, there are less underlying systemic diseases or drugs affecting colonic motility in children and the results tend to be easier to interpret in this younger population.

Limitations related to this diagnostic procedure include variation related to use of different catheter types, placement techniques and protocols in different centres. It is also associated with a degree of invasiveness and so may not be tolerated well by some children.

10. Electromyography

Electromyography records the electrical activity of skeletal muscles and can be used to assess for evidence of abnormal skeletal muscle function. Combined with manometry, it can be useful to confirm the presence of paradoxical sphincteric contraction during attempted defaecation.

11. The role of biopsy

The most common biopsy performed in relation to childhood constipation is a rectal biopsy. This is done in cases of suspected Hirschsprung's disease to confirm a diagnosis. The child may or may not have had other previous investigations including barium enema or anal manometry.

Usually a suction-method biopsy is performed. This is a simple and quick procedure, though does carry a risk for haemorrhage. In neonates, it can be performed at the bedside without the need for a general anaesthetic. Occasionally there may be difficulty obtaining an adequate tissue specimen by this technique (as it is a blind procedure). In these circumstances, a full thickness surgical biopsy may need to be performed.

There may be a role for other gastrointestinal tract biopsies to demonstrate abnormal histology in the presence of chronic constipation (e.g. neuronal intestinal dysplasia: qualitative and quantitative abnormalities of the myenteric plexus).

12. Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) of the spine may be performed in children with constipation if the patient history or physical examination is suggestive of spinal pathology. A recent study looked at the result of MRI-spine scans performed on 88 children with intractable constipation. They found that 9% (8 children) had spinal abnormalities on MRI that required surgery (75% of these were a tethered cord). Of these 8 children, 5 had no abnormal signs on physical examination. (Rosen et al., 2004). This study highlights the importance of a thorough history and physical examination in any child presenting with chronic constipation. In particular, one should assess the lower limb neurological status and sacral area for any abnormalities (skin discolouration, naevi, sinuses, hairy patch, central pit or bony abnormalities).

MRI and MR defecography (dynamic pelvic MRI) have also been used in the assessment of anorectal disorders. They have the advantage of being able to simultaneously image anatomy and motility function and a low dosage of radiation exposure. However, there is a high associated cost and very limited data exists on the role of this investigation in children.

13. Miscellaneous Investigations

There are some other investigations that may be indicated in assessment for underlying causes in chronic constipation. These include the following:

1. Sweat test to exclude cystic fibrosis. In particular, this condition may be suspected in cases of constipation associated with failure to thrive, recurrent chest infections or a history of delayed passage of meconium or meconium plug.
2. Allergy testing (IgE antibodies to cow's milk protein antigens) for cow's milk protein allergy. This may be suspected in the context of constipation if the patient has a strong history of allergies, anal fissures / excoriation or abdominal discomfort.

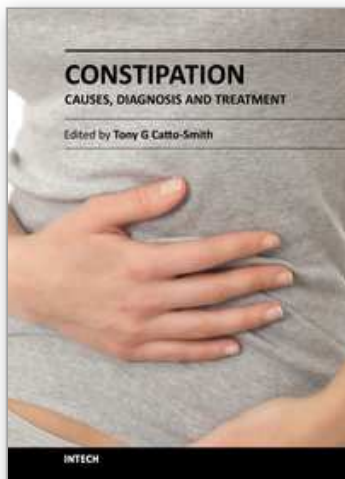
14. Conclusion

Most children with chronic constipation do not require any investigations other than a thorough history and physical examination. In patients with intractable constipation or red flags suggestive of an underlying organic aetiology, specialised testing may be indicated and of use. The choice of investigations should be made with specific consideration to the diagnosis being considered.

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Constipation - Causes, Diagnosis and Treatment

Edited by Dr. Anthony Catto-Smith

ISBN 978-953-51-0237-3

Hard cover, 172 pages

Publisher InTech

Published online 07, March, 2012

Published in print edition March, 2012

Constipation is common in both adults and children. Estimates would suggest a median prevalence of around 12-16% in the general population. While regarded as a minor nuisance in some cases, its consequences can be severe, with a substantial impact on quality of life. Secondary faecal soiling has a profound psychological effect at all ages. This book provides contributions from authors with a range of backgrounds which clarify the pathogenesis, diagnosis, and therapy of constipation for the general population and also for certain high risk groups.

How to reference

In order to correctly reference this scholarly work, feel free to copy and paste the following:

Anthony G. Catto-Smith and Kathleen H. McGrath (2012). The Role of Diagnostic Tests in Constipation in Children, Constipation - Causes, Diagnosis and Treatment, Dr. Anthony Catto-Smith (Ed.), ISBN: 978-953-51-0237-3, InTech, Available from: <http://www.intechopen.com/books/constipation-causes-diagnosis-and-treatment/the-role-of-diagnostic-tests-in-constipation>

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