

Small bowel MRI in adult patients: not just Crohn's disease—a tutorial

Giedre Kavaliauskiene · Manon L. W. Ziech ·
C. Yung Nio · Jaap Stoker

Received: 31 January 2011 / Revised: 25 April 2011 / Accepted: 28 June 2011 / Published online: 13 July 2011
© The Author(s) 2011. This article is published with open access at Springerlink.com

Abstract

Objectives To provide an overview of less well-known small bowel and mesenteric diseases found at small bowel magnetic resonance (MR) enterography/enteroclysis and to review the imaging findings. MR enterography and enteroclysis are important techniques for evaluation of small bowel diseases. In most centres these techniques are primarily used in Crohn's disease, and most radiologists are familiar with these MRI findings. However, the knowledge of findings in other diseases is often sparse, including diseases that may cause similar clinical symptoms to those of Crohn's disease.

Methods We present a spectrum of less common and less well-known bowel and mesenteric diseases (e.g. internal hernia, intussusception, neuroendocrine tumour) from our small bowel MR database of over 2,000 cases.

Results These diseases can be found in patients referred for bowel obstruction, abdominal pain or rectal blood loss. Further, in patients with (or suspected to have) Crohn's disease, some of these diseases (e.g. neuroendocrine tumour, familial Mediterranean fever) may mislead radiologists to erroneously diagnose active Crohn's disease.

Conclusion Radiologists should be familiar with diseases affecting the small bowel other than Crohn's disease, including diseases that may mimic Crohn's disease.

Keywords MRI · Small bowel · Tumours · Congenital · Inflammation

Introduction

Magnetic resonance (MR) enterography and enteroclysis are important techniques for small bowel evaluation, combining good soft tissue contrast, detection of extra-enteric findings, lack of radiation exposure and repeated data acquisition for functional bowel evaluation [1]. Comparative studies between MRI and other small bowel imaging techniques to diagnose inflammatory diseases are often sparse, include a relatively small number of patients and often lack a good reference standard [2]. A meta-analysis of imaging techniques in inflammatory bowel diseases showed that there were no significant differences in diagnostic accuracy among ultrasound, computed tomography, scintigraphy and MR imaging in diagnosing inflammatory bowel disease [1]. Therefore it is preferable to use a non-invasive technique without radiation exposure (i.e. ultrasound or MR imaging) to detect small bowel lesions in patients with Crohn's disease. Advantages of ultrasound are availability and cost, but important advantages of MR imaging are the unrestricted overview, easy comparison between examinations and improved communication of results to the referring physician. Therefore MR imaging is the technique of choice in many centres.

Diagnosis of Crohn's disease and detection of disease activity is a major indication for small bowel MRI in most centres, and most radiologists are familiar with these MRI findings. However, there are many diseases that may cause similar clinical symptoms or radiological signs to those in Crohn's disease. Furthermore, MR enterography and enteroclysis play an increasing role in the detection of other small bowel diseases, including tumours [3, 4].

The purpose of this article is to present a spectrum of less well-known bowel and mesenteric diseases from our small bowel MR database of over 2,000 cases.

G. Kavaliauskiene (✉) · M. L. W. Ziech · C. Y. Nio · J. Stoker
Department of Radiology, Academic Medical Center,
Meibergdreef 9,
1105 AZ Amsterdam, The Netherlands
e-mail: giedre.kavaliauskiene@gmail.com

MR technique

Adequate luminal distension is mandatory for accurately assessing the small bowel. Three groups of contrast agents can be used to achieve distension, namely positive, negative or biphasic contrast agents. Positive oral contrast agents appear hyperintense on all sequences. They are based on gadolinium-chelate, ferrous or manganese ions. The high signal intensity of the lumen however can interfere with the high signal intensity of the bowel wall after intravenous contrast medium administration. Negative oral contrast agents are based on iron oxide particles. They appear as hypointense on all sequences. Biphasic oral contrast agents are now the most frequently used contrast agents. Usually they have low signal intensity on T1-weighted images (optimising the contrast between the enhancing bowel wall and hypointense lumen) and high signal intensity on T2-weighted images. Mannitol with tap water is frequently used as a biphasic contrast agent. Intravenous contrast agents (gadolinium 0.1 ml/kg) are recommended for the assessment of inflammation of the small bowel and can be helpful for diagnosing other small bowel diseases as well.

The main difference between MR enteroclysis and MR enterography is the use of naso-duodenal intubation under fluoroscopic guidance in enteroclysis for optimal distension of the small bowel, whereas with enterography the contrast agent is given orally. With MR enteroclysis better distension is achieved, although this does not automatically lead to a higher accuracy in the terminal ileum [5]. In practice, the proximal small bowel is often not well distended with MR enterography; therefore to depict the whole small bowel MR enteroclysis is the technique of choice.

Another method of assessing small bowel diseases is video capsule endoscopy. MR enterography or MR enteroclysis has to be performed before video capsule endoscopy to rule out small bowel obstruction. In patients without suspected small bowel obstruction, video capsule endoscopy can be performed. Some data exist on the comparison of MRI and video capsule endoscopy. A study by Gupta et al. [6] compared video capsule endoscopy with MR enteroclysis in 19 patients with Peutz-Jeghers syndrome. Although there was no significant difference between the two techniques with regard to accuracy, large polyps (>15 mm) were missed at video capsule endoscopy in three patients [6]. MRI (extraintestinal and mural lesions) and video capsule endoscopy (small mucosal lesions) can be used in a complementary manner [7].

Imaging of the small bowel is most often performed on 1.5-T MRI, although the examinations can be performed on 3-T MRI with slight adaptation of the sequences. All sequences should be performed in breath-holds. The patient's position in the MRI can be supine or prone. Prone positioning can give better loop separation because of

compression, but is less comfortable for the patient. We prefer using the supine position only.

Sequences

Half-Fourier single-shot RARE (HASTE) generates sequences with a strong T2-weighting. The acquisition times are short so breathing artefacts are minimal. Normal bowel wall has low signal intensity on HASTE images. Due to oedema (in inflammation), the signal intensity can increase. Flow artefacts due to peristaltic motion can occur with the HASTE sequence, but an anti-spasmodic agent can be given to prevent this. To distinguish fat from oedema (both have high T2 signal intensity), fat-suppression can be used. Balanced steady-state free precession (true-FISP) is a sequence that combines tissue contrast from both T1- and T2-weighting. The bowel wall has an intermediate to low signal intensity, and fluids have a high signal intensity. For this sequence no anti-peristaltic drugs are needed. T1-weighted sequences (VIBE) with intravenous contrast medium administration are performed to assess whether enhancing areas (with more perfusion) are present in the small bowel. This sequence is very sensitive to peristalsis so spasmolytics are mandatory. The authors use butylscopolamine bromide (Buscopan; Boehringer-Ingelheim, Ingelheim, Germany). Butylscopolamine bromide is not FDA-approved for this application, glucagon can be used as an alternative when Buscopan is contraindicated or not available.

Other sequences that can be used for small bowel assessment are diffusion-weighted imaging and cine imaging.

Imaging findings

GI tract rotation and fixation anomalies

Gastrointestinal (GI) tract congenital rotation and fixation anomalies are usually detected in neonates and children when they cause obstruction requiring surgical treatment. These congenital GI tract anomalies are rare findings in the adult population and can be an incidental radiological diagnosis. It is thought that the abnormal position of the bowel itself does not cause clinical symptoms. Symptoms occur with complications due to an abnormal mesentery position and fixation that may cause volvulus. Symptoms may vary from mild, non-specific complaints—leading to the incorrect diagnosis of irritable bowel syndrome or gastric ulceration—to acute midgut volvulus (rare in adults) [8, 9].

There are three main types of GI tract rotation and fixation anomalies: non-rotation, malrotation and reversed rotation. In non-rotation the midgut rotates only 180°

instead of 270° ; therefore the small bowel is on the right side of the abdomen and the colon on the left (Fig. 1a) [8]. The ileum crosses the midline from right to left before entering the caecum [10]. In malrotation the midgut rotation is incomplete and its severity depends on the localisation of the caecum, which varies from normal (on the right-hand side) to localised on the left-hand side. Reversed rotation is a rare rotation anomaly, where the transverse colon is situated behind the duodenum with the superior mesenteric artery between them [8].

Signs highly suggestive of midgut rotation and fixation anomalies are the duodenojejunal junction (ligament of Treitz) and proximal loops of the jejunum on the right-hand side of the abdomen and the lack of the normal midline position of the horizontal part of the duodenum (Fig. 2a) [8]. An abnormal (reversed) superior mesenteric artery (SMA) and superior mesenteric vein (SMV) relationship (SMV located to the left of the SMA) may accompany GI tract rotation and fixation anomalies (Fig. 1b). However this sign should be used with caution as a normal mesenteric vascular anatomy does not exclude the possibility of malrotation (Fig. 2b), while a reversed position of mesenteric vessels can be seen in patients without malrotation [9, 10]. Rotation of the SMV and adjacent mesenteric fat around the SMA ('whirl sign') can be related, but is not specific to midgut volvulus and may occur after abdominal surgery or in internal hernias (Fig. 3) [10]. A 'whirl' sign indicates an urgent surgical pathological condition in the setting of closed loop obstruction and mesentery torsion with vascular strangulation. However, this sign should be used with care in asymptomatic patients when it is found as an incidental finding. As these patients do not complain of any symptoms, it is thought to be a mesenteric twist without complications (no closed loop obstruction or mesentery torsion).

Therefore a changed duodenal position should be considered the most reliable sign of rotation and fixation anomalies.

Fig. 1 A 31-year-old male patient presented with persistent abdominal pain. MR enteroclysis (a) coronal thick-slab single-shot turbo spin echo image demonstrates that all small bowel loops are localised on the right-hand side of the abdomen. (b) Axial true-FISP image demonstrates reversed superior mesenteric artery (SMA) and superior mesenteric vein (SMV) relationship: SMV (open arrow) to the left of the SMA (white arrow). MRI findings indicate midgut non-rotation



Meckel's diverticulum

Meckel's diverticulum is the most common congenital anomaly of the GI tract without association with other congenital anomalies. A higher prevalence of Meckel's diverticulum in Crohn's disease patients is considered [11].

Meckel's diverticulum is the result of incomplete atrophy of the omphalomesenteric duct. This is a true diverticulum on the anti-mesenteric side of the distal ileum containing three layers of the bowel wall. Ectopic gastric or pancreatic mucosa is present in 50% of all Meckel's diverticula [11, 12]. There is no gender predilection for the development of Meckel's diverticulum; however symptoms and complications are considered to be more common in men [11]. The clinical diagnosis is challenging, depending on the patient's age and the clinical findings, which may overlap more common diseases. The diagnosis is often suspected at cross-sectional imaging, scintigraphy or video capsule endoscopy.

Meckel's diverticulum itself is asymptomatic; clinical symptoms are caused by complications. Peptic ulceration with bleeding from heterotopic gastric mucosa located within or close to the diverticulum is the most frequent complication in the paediatric population, and it may appear in adults as well (Fig. 4) [11]. Meckel's diverticulum is diagnosed when a saccular, blind-ended structure continuous with the ileum is identified on imaging. An inflamed diverticulum has a distended lumen and surrounding infiltration.

Internal hernias

Internal hernia is a protrusion of viscera within the peritoneal cavity through a normal or abnormal peritoneal or mesenteric opening (congenital or acquired). Clinical manifestation may be non-specific, varying from mild abdominal complaints when the hernia is reducible to acute small bowel obstruction and strangulation when it is incarcerated [13–15]. The low

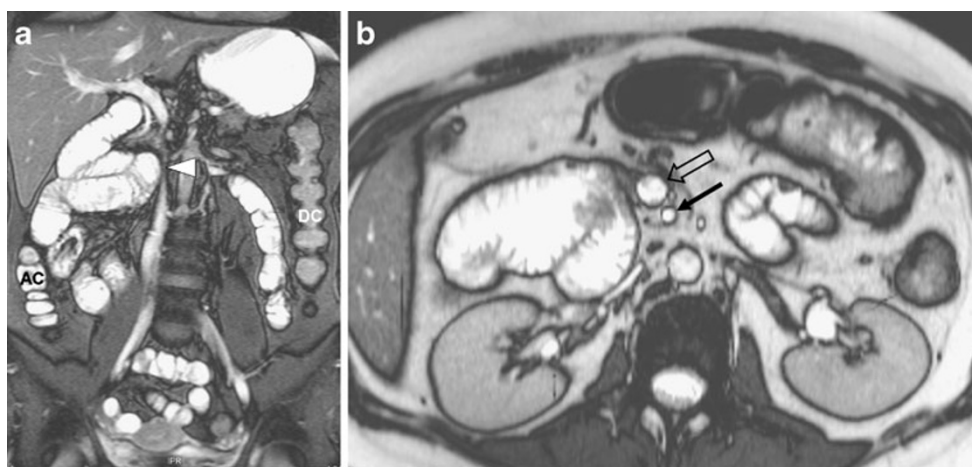


Fig. 2 A 37-year-old female patient presented with chronic abdominal pain and was suspected of having irritable bowel syndrome. MR enteroclysis (a) coronal fat-saturated true-FISP image demonstrates the duodenojejunal junction and proximal jejunum loops located on the right-hand side of the abdomen (arrowhead) and the normal

position of the ascending colon (AC) and descending colon (DC). (b) Axial true-FISP image demonstrates a normal relationship of the mesenteric vessels: SMV (open arrow) is to the right of SMA (black arrow). MR findings of GI tract malrotation

incidence of internal hernias and the non-specific clinical findings often make the diagnosis difficult but important because of the risk of acute bowel strangulation, which requires prompt surgical treatment.

There are different types of internal hernias depending on the orifice localisation. There are six main groups of internal hernias, which are listed in decreasing order of frequency: paraduodenal hernias (50–55% of internal herniations), pericaecal hernias (10–15%), transmesenteric hernias (8–10%), foramen of Winslow hernias (6–10%), intersigmoid hernias (4–8%) and paravesical hernias (<4%) [13, 16]. The most common are paraduodenal hernias, which may be left-

sided (more common) and right-sided. In left-sided paraduodenal hernias small-bowel loops herniate through a congenital defect in the descending mesocolon into a paraduodenal fossa. In right-sided paraduodenal hernia bowel loops herniate into Waldeyer's fossa, which is a rare peritoneal recess (observed in 1% of autopsies) located behind the superior mesenteric artery and inferior to the horizontal segment of the duodenum. Right paraduodenal hernia may be associated with midgut malrotation, although it may occur without any GI tract rotation anomaly [13, 16].

Not as common as paraduodenal hernias but important because of the higher risk of obstruction are transmesenteric hernias. These hernias occur as a result of small bowel herniation through a defect in the small bowel mesentery. It is the most common internal hernia in children, but not so common in adults. Transmesenteric hernia in adults is considered to be predisposed by previous abdominal surgery, trauma or abdominal inflammation. Generally, small bowel loops may herniate through an acquired or congenital small bowel mesentery defect. In transmesenteric hernias the herniated small bowel loops are not enveloped by any peritoneal lining, which is different from other internal hernias [15]. Lack of a hernial sack makes the diagnosis of transmesenteric hernias challenging and less obvious on imaging, especially when there are no signs of obstruction (Fig. 5). Therefore a cluster of herniated small bowel loops without dilation lying adjacent to the abdominal wall with adjacent mesentery vessel crowding and not displacing the nearby colon may be suggestive of a transmesenteric hernia. When this is associated with dilated small bowel loops and colon dislocation or a radiological 'closed loop' appearance, it is considered to be a sign of obstructed transmesenteric hernia [14–16].



Fig. 3 A 73-year-old female patient was complaining of nausea and vomiting. MR enteroclysis axial fat-saturated VIBE image after intravenous contrast medium demonstrates the 'whirl' sign (arrowhead), which indicates a mesenteric twist in the case of internal hernia. The patient had undergone Nissen fundoplication surgery previously. There were no signs of malrotation or small bowel obstruction

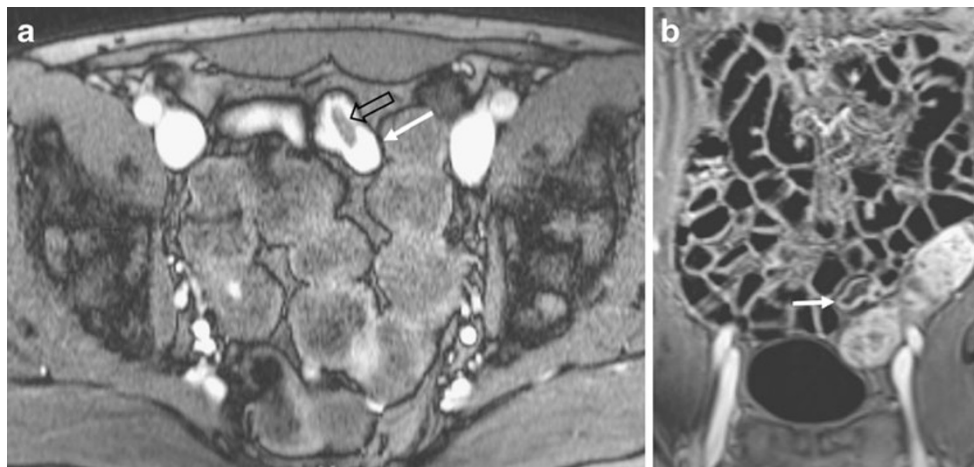


Fig. 4 A 30-year-old male patient complaining of rectal blood loss. MR enteroclysis (a) axial fat-saturated true-FISP image demonstrates a saccular, blind-ended structure of the small bowel in the lower abdomen (white arrow) with high-intensity fluid content and hypointense filling defect (open arrow)—a possible blood clot. (b) Coronal

fat-saturated VIBE image after intravenous contrast demonstrates the diverticulum with slightly thickened wall (white arrow), just above the urinary bladder wall. The diagnosis was Meckel's diverticulum as the source of bleeding

Intussusception

Intussusception is the invagination of a bowel loop with its mesenteric fat into the lumen of the adjacent bowel [17]. It is primarily a paediatric condition, but rarely (up to 5%) may occur in adult patients with predominance in the small bowel [18].

It is thought that up to 90% of adult intussusceptions have a pathological lead point and only 10% are idiopathic [17, 18]. However with the present widespread use of cross-sectional imaging, asymptomatic intussusceptions are

more frequently recognised, and these are often without a lead point. The latter are usually discovered incidentally as they do not cause obstruction and are transitional (Fig. 6). Intussusceptions with a lead point tend to be persistent with partial bowel obstruction symptoms. Aetiology of adult intussusceptions with a lead point ranges from benign (lipoma, polyp, especially in Peutz-Jeghers syndrome) to malignant lesions (metastasis, lymphoma, adenocarcinoma). Idiopathic intussusceptions and invaginations with benign lead points are more common in the small bowel, while intussusceptions with malignant lead points (e.g. adenocarcinoma, lymphoma) are more common in the colon [17–19].

Target-like (bowel-into-bowel) appearance of a bowel is pathognomonic for intussusceptions as it telescopes with its trapped mesenteric fat and vessels into the adjacent bowel (Figs. 6, 7 and 8). An intussusception with a lead point contains a target-like mass with a cross-sectional diameter usually greater than that of normal bowel loop (Fig. 7) and may be associated with bowel obstruction [17]. Cross-sectional imaging techniques help to distinguish intussusceptions with and without a lead point; however, exact determination of the underlying disease may remain challenging in some cases, including for MRI. The usual treatment of symptomatic intussusceptions with lead points in adults is surgery, with the underlying cause established at pathological examination [18].

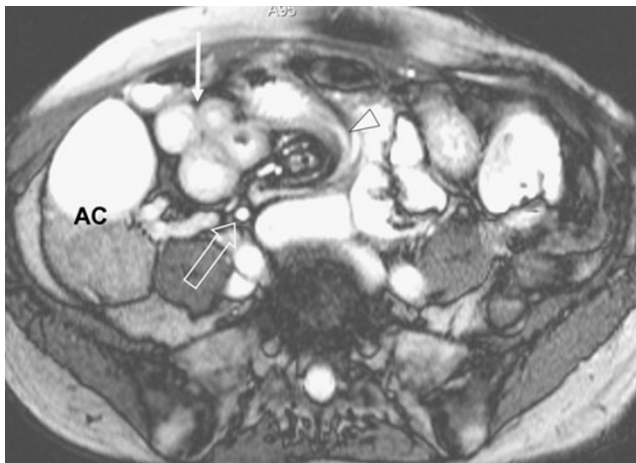


Fig. 5 The same 73-year-old female patient as in Fig 3. MR enteroclysis axial fat-saturated true-FISP image demonstrates a cluster of non-dilated small bowel loops (white arrow) lying adjacent to the abdominal wall, crowding of the mesenteric vessels (open arrow) and a collapsed bowel lumen at the site of the hernia (arrowhead), indicating transmesenteric internal hernia without mechanical obstruction. The ascending colon (AC) is not displaced

Peutz-Jeghers syndrome

Peutz-Jeghers syndrome is an inherited autosomal dominant disease characterised by two classic findings: mucocutaneous pigmentations and hamartomatous polyps of the GI tract. This

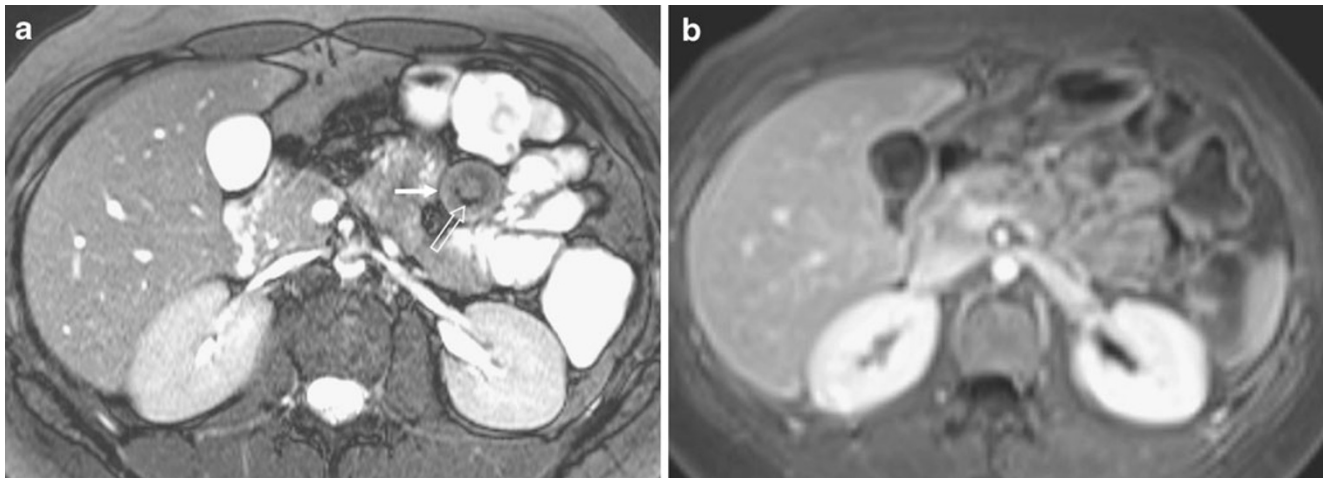


Fig. 6 An 18-year-old female patient complaining of abdominal pain; Crohn's disease was suspected. MR enterography **(a)** axial fat-saturated true-FISP image demonstrates a target-like small bowel appearance of a small bowel intussusception in the left upper quadrant (white arrow). A small amount of trapped mesenteric fat with vessels can be identified

(open arrow). **(b)** Axial fat-saturated VIBE image after intravenous contrast medium acquired 6 min later at the same level shows no evidence of intussusception. A diagnosis of a transient small bowel intussusception was made. No evidence of Crohn's disease

syndrome is usually diagnosed early in life because mucocutaneous pigmentation in the perioral region commonly appears in infancy or childhood [20].

Gastrointestinal tract Peutz-Jeghers polyps have malignant potential, contrary to mucocutaneous lesions. Hamartomatous polyps may occur anywhere in the GI tract with predominance in the small bowel (Fig. 8a and b), although most malignant degenerated polyps are detected in the stomach, duodenum and colon. There is an associated risk of development of extraintestinal malignancies like breast, pancreas, lung, ovarian or testicular cancer in Peutz-Jeghers syndrome [20, 21].

A common complication of Peutz-Jeghers syndrome is intussusception by a Peutz-Jeghers polyp. Although this concerns intussusceptions with a lead point, some of these intussusceptions resolve spontaneously (Fig. 8c) [20]. Other common complications of Peutz-Jeghers syndrome are obstruction and GI tract bleeding caused by polyp ulceration [20, 21].

Multiple polypoid lesions in the GI tract may be detected in Peutz-Jeghers syndrome on imaging. Polyps may vary in location, appearance (sessile or pedunculated) and size (from small to large). Polyps tend to enhance after administration of contrast media, which helps to differen-

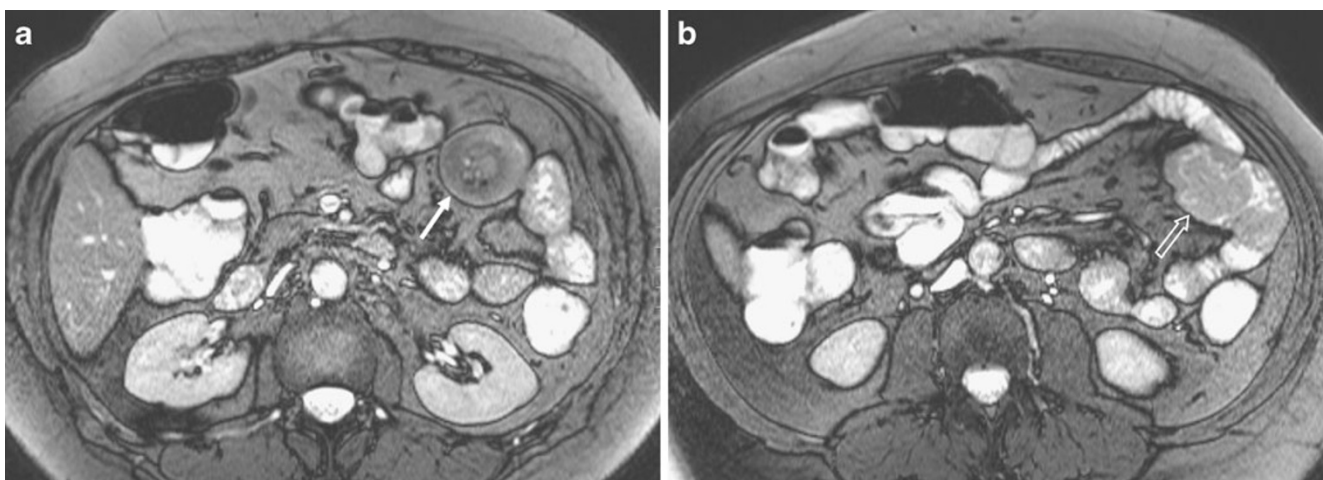


Fig. 7 A 45-year-old female patient with Peutz-Jeghers syndrome, anaemia and rectal blood loss. MR enterography **(a)** and **(b)** axial fat-saturated true-FISP images demonstrate intussusception with bowel-in-bowel appearance (white arrow) and a polyp as a lead point (open

arrow) at a lower level of the same sequence. The cross-sectional diameter of the intussusception at the level of the lead point (polyp) is greater than a normal bowel loop, and this is different from an idiopathic intussusception (see Fig. 6a)

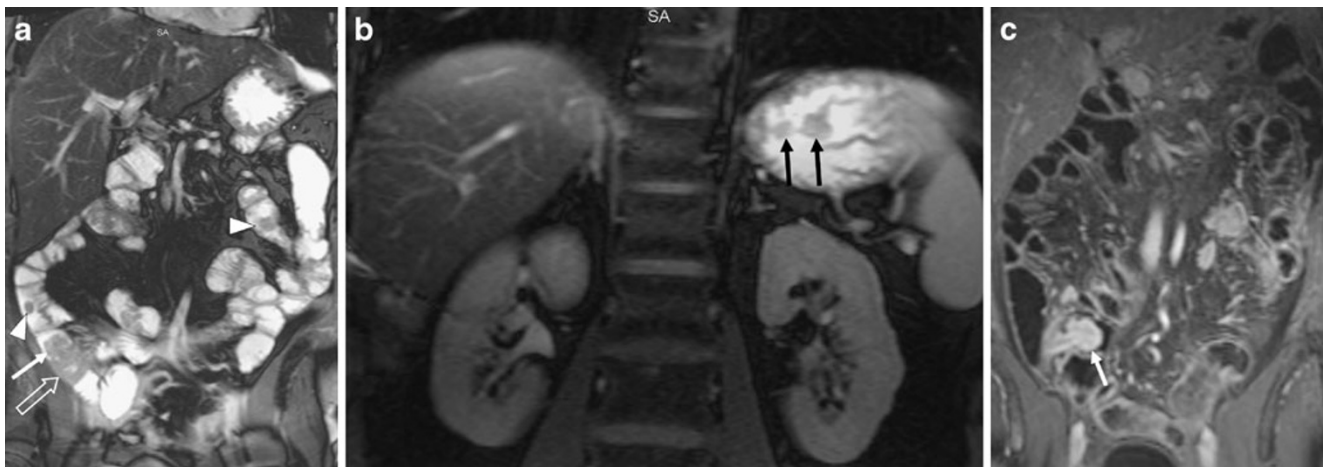


Fig. 8 A 48-year-old male patient with Peutz-Jeghers syndrome complained of abdominal pain. He had undergone Whipple surgery for malignant duodenal tumour previously. MR enterography (a) coronal fat-saturated true-FISP image demonstrates a polyp in the small bowel (white arrow), which caused intussusception (open arrow) of a short segment with bowel-in-bowel appearance. Other polyps (arrowheads) in different parts of the small bowel are seen. The same sequence (b)

image demonstrates two pedunculated polyps in the stomach (black arrows). Two months later MR enterography was performed again, (c) coronal fat-saturated VIBE image after intravenous contrast medium demonstrates the same contrast-enhancing pedunculated polyp in the small bowel in the right lower abdomen (white arrow). There are no signs of intussusception; it resolved spontaneously

tiolate polyps from bowel content. MR enterography or enteroclysis is performed to detect and monitor GI tract polyps (polyps larger than 1.5–2 cm are considered potentially malignant and are removed) and in patients with suspected intussusception in Peutz-Jeghers syndrome.

Multiple GI tract polyps are typical of Peutz-Jeghers syndrome, although they are not specific and may be found in other syndromes such as juvenile polyposis, familial adenomatous polyposis and Cronkhite-Canada syndrome [20]. Therefore the diagnosis is based not only on radiological findings, but also on clinical and histopathology results.

Gastrointestinal stromal tumour

Gastrointestinal stromal tumours (GIST) are the most common mesenchymal neoplasms of the GI tract. These tumours can be differentiated from smooth muscle and neural tumours by expression of tyrosine kinase growth factor receptors on histopathology. GIST can appear anywhere in the GI tract with the highest prevalence in the stomach (70%). In very rare cases they may be found in the omentum, mesentery or retroperitoneum. The clinical presentation of GIST may be variable—from asymptomatic to GI tract bleeding and not very commonly to highly aggressive behaviour with involvement of adjacent organs, metastases or peritoneal seeding [22, 23].

Gastrointestinal stromal tumours usually involve the muscular layer of the GI tract wall; therefore they have an exophytic growth pattern, but mucosa may be affected and ulcerations coexist in up to 50% [23]. Polypoid growth pattern may be evident in some cases and may cause intussusceptions

(Fig. 9). Large lesions often undergo cystic degeneration, bleeding, necrosis and sometimes a cavity communicating with the bowel lumen. Therefore there is a great difference in the imaging appearance of GIST depending on tumour size and localisation. However, usually they appear as well-circumscribed exophytic growing and heterogeneous masses with peripheral contrast enhancement (Fig. 10). The degree of necrosis and haemorrhage highly affects tumour intensity and homogeneity on MRI. Solid parts of tumour are hyperintense on T2 images and enhance after administration of intravenous contrast material (Figs. 9 and 10). There is disagreement in the literature concerning the correlation between the grade of heterogeneity and the malignant potential of GISTs. It is thought that tumours <5 cm are usually at low risk of malignancy and tumours >5 cm are malignant. However small tumours may be malignant and may metastasise [24]. The diagnosis of malignant GIST can be safely made on imaging when ingrowth into adjacent structures or metastases is found. Lymph node metastases are not common in GISTs.

The differential diagnosis of GIST includes other mesenchymal tumours (leiomyomas, leiomyosarcomas, schwannomas), neuroendocrine tumours, GI tract primary carcinomas, lymphoma or metastatic tumours, especially in the small bowel.

Neuroendocrine tumours of the GI tract

Neuroendocrine tumours arise from the diffuse GI tract endocrine system (gastrin, secretin, enterochromaffin-like, somatostatin-producing cells, etc.) outside the pancreas and thyroid. These tumours may occur anywhere in the GI tract,

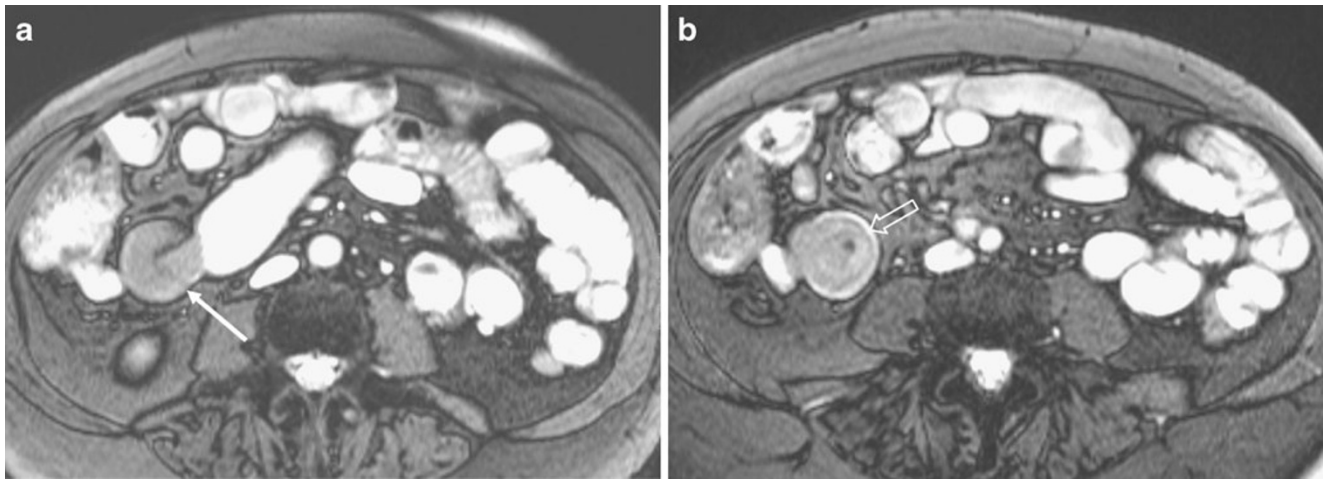


Fig. 9 A 68-year-old female patient with abdominal pain and anaemia. MR enterography axial fat-saturated true-FISP images (a) and (b) demonstrate a small bowel intussusception (white arrow). There is a hyperintense, heterogeneous mass (open arrow) as a lead

point visible at a lower level of the same sequence. Small bowel resection was performed, and histopathology demonstrated GIST as the cause of the intussusception

although they most frequently involve the appendix and small bowel, particularly the distal ileum [25–28]. Neuroendocrine tumours of the small bowel may be multiple in about 40% of cases and may be associated with a second primary malignancy [25, 27].

There are multiple classifications of neuroendocrine tumours as they present a heterogeneous group. In 2000, the World Health Organisation presented a new classification of neuroendocrine tumours. According to this classification GI tract neuroendocrine tumours are divided into main three groups: (1) well-differentiated tumours (or carcinoids) with benign or uncertain malignant potential, (2) well-differentiated endocrine carcinomas (or malignant

carcinoids) with low-grade malignancy and (3) poorly differentiated endocrine carcinomas with high-grade malignancy. As these groups cannot be distinguished on imaging, neuroendocrine tumours should be considered to have malignant potential [25, 26].

Neuroendocrine tumours may have different clinical and biological behaviours. Clinical manifestation depends on the bioactive substances—hormones produced by the tumour (due to GI endocrine cell type). Tumours may cause mechanical bowel obstruction and abdominal pain. The carcinoid syndrome with cutaneous flushing, bronchospasm, abdominal pain and diarrhoea develops in approximately 10% of patients with neuroendocrine tumours. It most frequently appears in patients

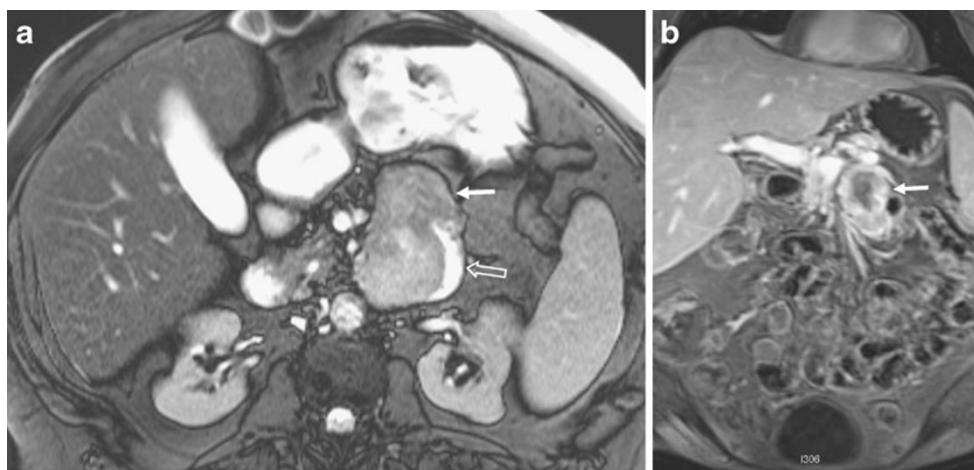


Fig. 10 A 66-year-old female patient complaining of abdominal pain. MR enterography (a) axial fat-saturated true-FISP image demonstrates a heterogeneous, exophytic growing mass (white arrow) at the duodenojejunal junction with bowel lumen compression (open arrow).

(b) Coronal fat-saturated VIBE image after intravenous contrast medium administration better demonstrates the heterogeneity of the tumour (white arrow) with a central necrotic part. GIST was suspected on MRI, and this diagnosis was confirmed on histopathology

with metastasised ileal neuroendocrine tumours when bioactive substances pass into the systemic circulation [25, 26].

Commonly neuroendocrine tumours are of uncertain or low-grade malignant potential, although some of them are highly invasive. Approximately 58–64% of patients with ileal neuroendocrine tumours have disease spread to the regional lymph nodes or to the liver at the time of diagnosis [26]. Less common sites to metastasise are bones, skin and thyroid [25].

Radiological appearance of neuroendocrine tumours is variable depending on their localisation, biological behaviour pattern and size. Small bowel neuroendocrine tumours may vary from a small submucosal lesion to a large intraluminal mass. Usually they appear as discrete, moderately contrast-enhancing masses on post-contrast MRI. Local production and release of bioactive substances rather than infiltration growth of tumour are considered to cause submucosal and adjacent mesenteric retraction or so-called desmoplastic reaction, which demonstrates intense contrast enhancement [26–28]. Small neuroendocrine tumours may cause small bowel wall kinking—the ‘hair pin’ sign—because of their submucosal localisation and desmoplastic reaction (Fig. 11). Less commonly these tumours may manifest as uniform or concentric mural thickening, enhancing after gadolinium administration without any evident lesion with mass effect. At histopathology this area of thickening may correspond to several small submucosal lesions [26, 27].

Differential diagnosis of small bowel neuroendocrine tumours should include other malignancies such as small bowel metastatic disease, lymphoma, small bowel adenocarcinoma, GIST and non-neoplastic diseases such as

Meckel’s diverticulum or Crohn’s disease predominantly in neuroendocrine tumours presenting without an evident mass [26].

Lymphoma

Primary or extranodal lymphoma is a rare disease, although GI tract primary involvement (particularly non-Hodgkin’s lymphoma) is the most common extranodal lymphoma. The stomach is most commonly involved, but other parts of the GI tract may also be affected. Lymphoma is considered to account for 20% of all primary small bowel malignancies. The terminal ileum is considered to be the most frequently affected site in the small bowel, as there is a relative abundance of lymphoid tissue [29–31].

Infiltrative and polypoid forms are two main growth patterns of primary GI tract lymphoma; lymphoma rarely presents as a large exophytic mass extending into adjacent tissues [32]. Infiltration is usually circumferential, homogeneous, may be nodular to a variable degree (up to bulky mass) and over a different length. Multifocal involvement of small bowel is possible and is thought to be more common in primary T-cell lymphoma [29].

Preservation of the surrounding fat tissue and a long involved segment help to differentiating lymphoma from adenocarcinoma (Fig. 12). However, a high-grade lymphoma may infiltrate the mesenteric fat. Lymphoma may ulcerate, perforate or create fistulas into the adjacent mesenteric or adjacent bowel loops, which makes it hard to distinguish from Crohn’s disease on MRI. Polypoid growing lymphoma may act as a lead point and cause intussusception. Obstruction of the small bowel is not

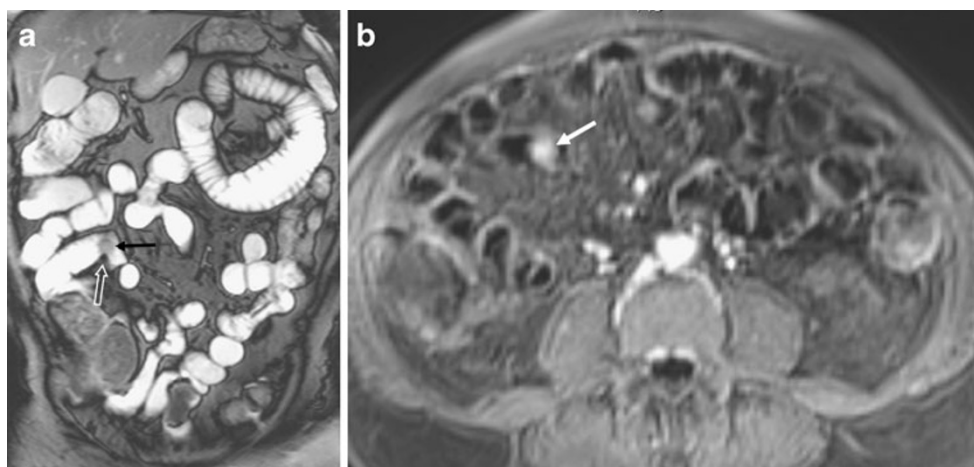


Fig. 11 A 76-year-old male patient with anaemia and rectal blood loss. MR enteroclysis (a) coronal fat-saturated true-FISP image demonstrates a small bowel lesion (black arrow), causing small bowel wall kinking—the ‘hair pin’ sign (open arrow). The sign was also seen in other sequences, differentiating it from bowel motility. (b) Axial fat-

saturated VIBE image after intravenous contrast medium administration demonstrates that the lesion (white arrow) moderately enhances. A neuroendocrine tumour may be suspected on MRI; the diagnosis was confirmed on histopathology

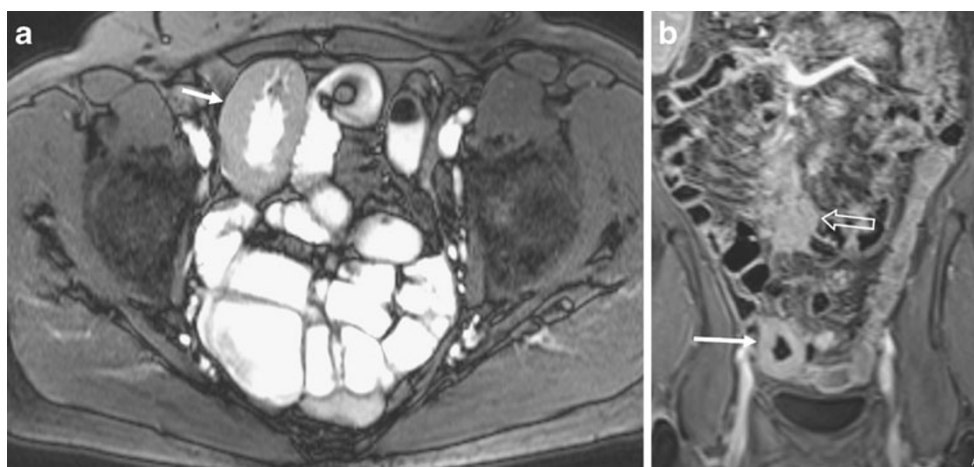


Fig. 12 A 52-year-old female patient with known non-Hodgkin's lymphoma complained of nausea and intermittent vomiting. MR enterography (a) axial fat-saturated true-FISP image and (b) coronal fat-saturated VIBE image after intravenous contrast medium administration demonstrate substantial circumferential small bowel wall

thickening (white arrow), (b) with evident contrast enhancement but with no luminal obstruction. Mesenteric lymphadenopathy was present (open arrow), but no fat tissue stranding adjacent to the tumour. MRI findings indicate small bowel lymphoma

common in lymphoma as it does not develop a desmoplastic reaction. Lymphoma may cause aneurysmal dilatation of the lumen because of lymphomatous involvement of the muscular layer and nerve plexus [28–33]. Lymphoma lesions often show contrast enhancement on MRI.

The GI lymphoma is a 'great mimicker' and has a variety of radiological appearances. Therefore the diagnosis is made at histopathology, although some radiological findings such as adjacent fat tissue preservation, long or multiple segment involvement, aneurysmal dilatation and a bulky mass without obstruction should raise the suspicion of primary GI tract lymphoma.

Primary adenocarcinoma of the small bowel

Small bowel adenocarcinoma is a rare disease. About half of the cases are found in the duodenum, while a location in the jejunum is more common than in the ileum [28, 32]. Crohn's disease is thought to be one of the risk factors for developing small bowel adenocarcinoma, especially in the ileum. There is no clear correlation between tumour size and its invasiveness [28].

Adenocarcinoma of the small bowel appears on MRI as a heterogeneous, moderately enhancing eccentric mass or circumferential lesion that may narrow the bowel lumen

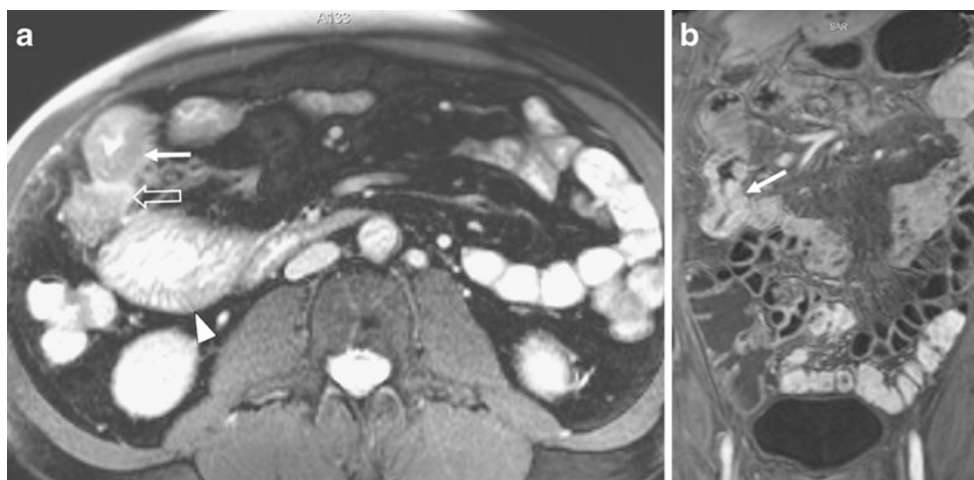


Fig. 13 A 56-year-old patient with Crohn's disease and a worsened clinical condition for 3 months. Active Crohn's disease was suspected clinically. MR enterography (a) axial fat-saturated true-FISP image and (b) coronal fat-saturated VIBE image after intravenous contrast medium administration demonstrate circumferential irregular small bowel wall thickening—tumour with adjacent fat infiltration (white

arrow) and regional lymphadenopathy (open arrow). Prestenotic bowel dilatation is demonstrated in (a) (arrowhead). MRI findings demonstrated a small bowel primary tumour as a cause of the symptoms; histopathology revealed adenocarcinoma. No evidence of Crohn's disease

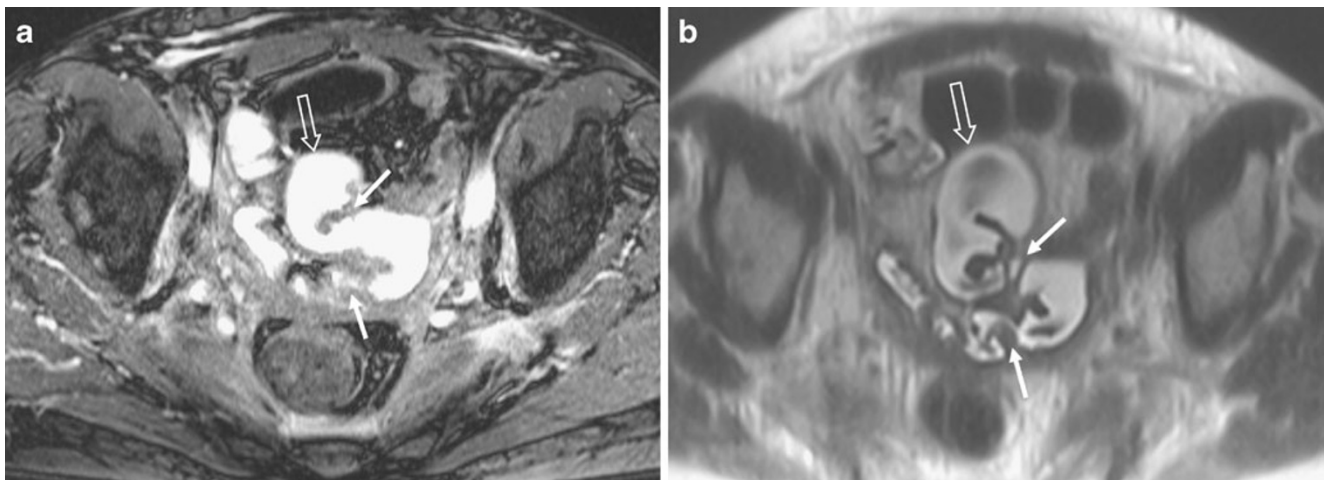


Fig. 14 A 65-year-old female patient, 6 months after pelvic radiotherapy for endometrial cancer with clinical signs of bowel obstruction; tumour disease recurrence was suspected. MR enteroclysis **(a)** axial fat-saturated true-FISP image demonstrates multiple irregular strictures (white arrows) of the distal small bowel with slight bowel wall thickening, prestenotic small bowel dilatation (open arrow)

and no mass. **(b)** Axial HASTE image acquired at the same level after 20 min demonstrates an identical configuration of the fixed small bowel loops as there is no motility in these loops. The diagnosis of radiation enteritis was made based on the medical history and findings at MR enteroclysis.

(Fig. 13). Therefore small bowel obstruction in adenocarcinoma is more common than in lymphoma. A circumferential adenocarcinoma is more commonly found in the distal small bowel, while duodenal and proximal jejunal adenocarcinomas are more often polypoid, heterogeneous contrast-enhancing lesions. The latter may predispose for intussusception [28, 32]. Large ulcerated masses can mimic lymphoma; adjacent fat infiltration may help in differentiation as this is more common in adenocarcinoma.

Radiation enteritis

Radiotherapy plays an important role in oncology management, although various complications may occur depending on the area of radiation, delivered dose and number of fractions.

Abdominopelvic radiation most commonly affects the rectum, distal small bowel and distal colon as these areas are part of the radiation field of most pelvic cancers. The small bowel is the bowel part most sensitive to radiation.

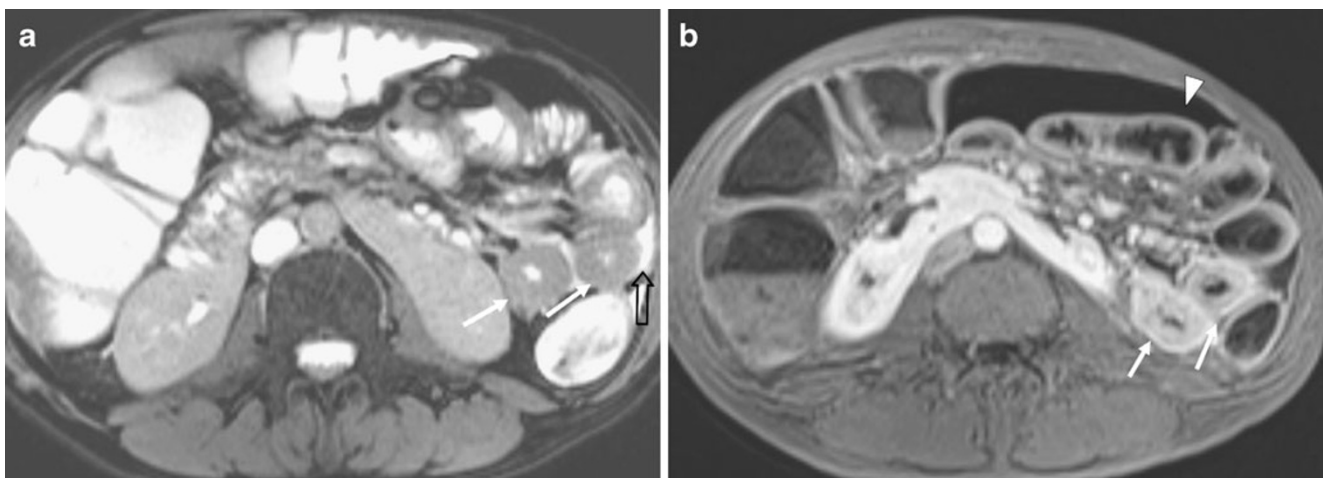


Fig. 15 A 44-year-old male patient with a known history of familial Mediterranean fever and Crohn’s disease. He had had surgery for small bowel obstruction a few days previously and then complained of abdominal pain and fever. MR enterography **(a)** axial fat-saturated true-FISP image demonstrates a thickened wall of adjacent small bowel loops (white arrows) with a small amount of free fluid in the

peritoneal cavity (open arrow) and some mesenteric fat infiltration. **(b)** Axial fat-saturated VIBE image after intravenous contrast medium administration demonstrates thickened enhancing wall and particularly serosa (white arrows) of the same small bowel loops. Free air (arrowhead) after the recent operation and a horseshoe kidney are seen

Because of its mobility it is less damaged than theoretically expected. However, previous abdominal surgery, adhesions and bowel fixation increase the risk of possible small bowel complications from radiotherapy [34, 35].

Bowel mucosa and submucosa are the most vulnerable parts of the bowel wall. Radiation changes depend on the radiation dose and the time elapsed after radiotherapy. Immediately after radiotherapy mucosal hyperaemia, oedema and inflammation may develop with mucosal ulcerations. Later submucosal obliterative endarteritis may lead to ischaemia and progressive fibrous changes resulting in contraction and thickening of the bowel wall and mesentery. Therefore chronic post-radiation changes in the small bowel may appear as thickened wall and narrowed adherent, fixed bowel loops in a previous radiation field (Fig. 14) [34–36]. It may be difficult to differentiate radiation-induced changes from inflammatory and malignant conditions or adhesions, but fixed bowel loops and the lack of a mass in an area of previous radiotherapy are highly suggestive of radiation enteritis [36]. Malignancy may be suspected when mass-like thickening and lymph node enlargement are detected.

Familial Mediterranean fever

Peritonitis (serositis) of small bowel loops may occur in common diseases causing acute abdominal pain (e.g. appendicitis), in post-surgical patients and in rarer diseases such as familial Mediterranean fever. This is a hereditary autosomal recessive disease with the highest prevalence in Turkey, and lower prevalence in Israel, Armenia and other countries of the Middle East. Familial Mediterranean fever is not strictly limited to that area as it is also found in Greece, North African countries, Italy, Germany, France, the USA and Japan. It is considered that the prevalence of the gene associated with familial Mediterranean fever mutation in different ethnographic groups predisposes such distribution. Distribution is related to world migration as most ancient mutations appeared in the Middle East—in the former area of Mesopotamia—and are thought to be spread all over the world. Lack of familial Mediterranean fever cases in other countries may be explained by mild disease forms according to different types of mutation or they are misdiagnosed as other more common diseases [37].

Clinically recurrent attacks of fever with serositis (commonly focal peritonitis with abdominal pain) and synovitis are common for familial Mediterranean fever. Usually the first attacks appear at an early age with no clinical manifestations between disease relapses [37, 38].

The diagnosis of familial Mediterranean fever is usually made on clinical findings. There are no specific diagnostic tests except for genetic tests; therefore the condition is easily missed in non-prevalent areas. Acute attacks of familial Mediterranean fever are thought to be self limiting; thus it is

most important to differentiate them from other acute conditions requiring prompt surgical treatment. Adhesion-induced small bowel obstruction is considered a life-threatening complication of familial Mediterranean fever [38].

At MRI the thickened wall of adjacent bowel loops with contrast enhancement, particularly serosal, is the result of focal peritonitis (Fig. 15) and should raise the suspicion of possible familial Mediterranean fever, especially when there is no other cause of peritonitis in a patient of prevalent descent. Different amounts of free fluid, mesenteric vessel engorgement and lymphadenopathy may also be present.

Conclusion

Radiologists should be familiar with less well-known diseases affecting the small bowel other than Crohn's disease as these diseases may be detected in patients referred for small bowel MR enterography or enteroclysis. This also includes malignant and inflammatory diseases that may mimic Crohn's disease.

Acknowledgement Giedre Kavaliauskiene was ESOR/ESGAR fellow at the Academic Medical Centre, University of Amsterdam, The Netherlands, when writing this manuscript. The authors acknowledge ESOR/ESGAR for providing the opportunity to write this paper.

Open Access This article is distributed under the terms of the Creative Commons Attribution Noncommercial License which permits any noncommercial use, distribution, and reproduction in any medium, provided the original author(s) and source are credited.

References

1. Horsthuis K, Bipat S, Bennink RJ, Stoker J (2008) Inflammatory bowel disease diagnosed with US, MR, scintigraphy, and CT: meta-analysis of prospective studies. *Radiology* 247:64–79
2. Tolan DJM, Taylor SA, Halligan S (2010) MRI of the small bowel: clinical role. In: Stoker J (ed) *MRI of the gastrointestinal tract*. Springer Heidelberg Dordrecht London, New York, pp 149–171
3. Masselli G, Poletini E, Casciani E, Bertini L, Vecchioli A, Gualdi G (2009) Small-bowel neoplasms: prospective evaluation of MR enteroclysis. *Radiology* 251:743–750
4. Tolan DJ, Greenhalgh R, Zealley IA, Halligan S, Taylor SA (2010) MR enterographic manifestations of small bowel Crohn disease. *Radiographics* 30:367–384
5. Negaard A, Paulsen V, Sandvik L, Berstad AE, Borthne A, Try K et al (2007) A prospective randomized comparison between two MRI studies of the small bowel in Crohn's disease, the oral contrast method and MR enteroclysis. *Eur Radiol* 17:2294–2301. doi:10.1007/s00330-007-0648-4
6. Gupta A, Postgate AJ, Burling D, Ilangovan R, Marshall M, Phillips RK et al (2010) A prospective study of MR enterography versus capsule endoscopy for the surveillance of adult patients with Peutz-Jeghers syndrome. *AJR Am J Roentgenol* 195:108–116. doi:10.2214/AJR.09.3174
7. Crook DW, Knuesel PR, Froehlich JM, Eigenmann F, Unterweger M, Beer HJ et al (2009) Comparison of magnetic resonance enterography and video capsule endoscopy in evaluating small

- bowel disease. *Eur J Gastroenterol Hepatol* 21:54–65. doi:10.1097/MEG.0b013e32830ce7a7
8. Berrocal T, Lamas M, Gutiérrez J, Torres I, Prieto C, del Hoyo ML (1999) Congenital anomalies of the small intestine, colon, and rectum. *Radiographics* 19:1219–1236
 9. Kanazawa T, Kasugai K, Miyata M, Miyashita M, Mizuno M, Nagase F et al (2000) Midgut malrotation in adulthood. *Intern Med* 39:626–631
 10. Lee NK, Kim S, Jeon TY, Kim HS, Kim DH, Seo HI et al (2010) Complications of congenital and developmental abnormalities of the gastrointestinal tract in adolescents and adults: evaluation with multimodality imaging. *Radiographics* 30:1489–1507
 11. Levy AD, Hobbs CM (2004) From the archives of the AFIP Meckel diverticulum: radiologic features with pathologic Correlation. *Radiographics* 24:565–587
 12. Malik AA, Shams-ul-Bari WKA, Khaja AR (2010) Meckel's diverticulum—Revisited. *Saudi J Gastroenterol* 16:3–7
 13. Takeyama N, Gokan T, Ohgiya Y, Satoh S, Hashizume T, Hataya K (2005) CT of internal hernias. *Radiographics* 25:997–1015
 14. Zissin R, Hertz M, Gayer G, Paran H, Osadchy A (2005) Congenital internal hernia as a cause of small bowel obstruction: CT findings in 11 adult patients. *Br J Radiol* 78:796–802
 15. Selçuk D, Kantarci F, Oğüt G, Korman U (2005) Radiological evaluation of internal abdominal hernias. *Turk J Gastroenterol* 16:57–64
 16. Mathieu D, Luciani A; GERMAD Group (2004) Internal abdominal herniations. *AJR Am J Roentgenol* 183:397–404
 17. Kim YH, Blake MA, Harisinghani MG, Archer-Arroyo K, Hahn PF, Pitman MB et al (2006) Adult intestinal intussusception: CT appearances and identification of a causative lead point. *Radiographics* 26:733–744
 18. Guillén Paredes MP, Campillo Soto A, Martín Lorenzo JG, Torralba Martínez JA, Mengual Ballester M, Cases Baldó MJ (2010) Adult intussusception—14 case reports and their outcomes. *Rev Esp Enferm Dig* 102:32–40
 19. Renzulli P, Candinas D (2010) Idiopathic small-bowel intussusception in an adult. *CMAJ* 23;182(3):E148
 20. Rufener SL, Koujok K, McKenna BJ, Walsh M (2008) Small bowel intussusception secondary to Peutz-Jeghers polyp. *Radiographics* 28:284–288
 21. Kopacova M, Tacheci I, Rejchrt S, Bures J (2009) Peutz-Jeghers syndrome: diagnostic and therapeutic approach. *World J Gastroenterol* 15:5397–5408
 22. Choumouzi D, Sinakos E, Papalavrentios L, Akriviadis E, Drevelegas A (2009) Gastrointestinal stromal tumors: a pictorial review. *J Gastrointestin Liver Dis* 18:379–383
 23. Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M (2003) Gastrointestinal stromal tumors: radiologic features with pathologic correlation. *Radiographics* 23:283–304
 24. Stamatakos M, Douzinas E, Stefanaki C, Safioleas P, Polyzou E, Levidou G et al (2009) Gastrointestinal stromal tumor. *World J Surg Oncol* 7:61
 25. Chang S, Choi D, Lee SJ, Lee WJ, Park MH, Kim SW et al (2007) Neuroendocrine neoplasms of the gastrointestinal tract: classification, pathologic basis, and imaging features. *Radiographics* 27:1667–1679
 26. Levy AD, Sobin LH (2007) From the archives of the AFIP: Gastrointestinal carcinoids: imaging features with clinicopathologic comparison. *Radiographics* 27:237–257
 27. Horton KM, Kamel I, Hofmann L, Fishman EK (2004) Carcinoid tumors of the small bowel: a multitechnique imaging approach. *AJR Am J Roentgenol* 182:559–567
 28. Buckley JA, Fishman EK (1998) CT evaluation of small bowel neoplasms: spectrum of disease. *Radiographics* 18:379–392
 29. Ghai S, Pattison J, Ghai S, O'Malley ME, Khalili K, Stephens M (2007) Primary gastrointestinal lymphoma: spectrum of imaging findings with pathologic correlation. *Radiographics* 27:1371–1388
 30. Rubesin SE, Gilchrist AM, Bronner M, Saul SH, Herlinger H, Grumbach K et al (1990) Non-Hodgkin lymphoma of the small intestine. *Radiographics* 10:985–998
 31. Ziech MLW, Stoker J (2010) MRI of the small bowel: enterography. In: Stoker J (ed) *MRI of the gastrointestinal tract*. Springer Heidelberg Dordrecht London, New York, pp 117–134
 32. Gore RM, Mehta UK, Berlin JW, Rao V, Newmark GM (2006) Diagnosis and staging of small bowel tumours. *Cancer Imaging* 29;6:209–212
 33. Lohan DG, Alhajeri AN, Cronin CG, Roche CJ, Murphy JM (2008) MR enterography of small-bowel lymphoma: potential for suggestion of histologic subtype and the presence of underlying celiac disease. *AJR Am J Roentgenol* 190:287–293
 34. Bluemke DA, Fishman EK, Kuhlman JE, Zinreich ES (1991) Complications of radiation therapy: CT evaluation. *Radiographics* 11:581–600
 35. Capps GW, Fulcher AS, Szucs RA, Turner MA (1997) Imaging features of radiation-induced changes in the abdomen. *Radiographics* 17:1455–1473
 36. Cronin CG, Lohan DG, Browne AM, Alhajeri AN, Roche C, Murphy JM (2009) MR enterography in the evaluation of small bowel dilation. *Clin Radiol* 64:1026–1034
 37. Ben-Chetrit E, Touitou I (2009) Familial Mediterranean Fever in the world. *Arthritis Rheum* 61:1447–1453
 38. Zissin R, Rathaus V, Gayer G, Shapiro-Feinberg M, Hertz M (2003) CT findings in patients with familial Mediterranean fever during an acute abdominal attack. *Br J Radiol* 76:22–25