

Meckel's diverticulum: misdiagnosis and late presentation

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Abstract: Meckel's diverticulum (MD) is a congenital anomaly of the gastrointestinal (GI) tract, occurring due to the incomplete obliteration of the omphalomesenteric duct during embryogenesis. Depending on the patient's age and symptomatology at presentation, scintigraphy is often the initial test of choice to detect MD. However, an increased incidence of false positive and negative tests has made MD difficult to diagnose in older children as well as in adults. MD can present in the older population with symptoms such as GI bleeding and anemia, which may mimic other GI pathologies, such as appendicitis, Crohn's disease, and Littre's hernia. Early diagnosis and appropriate management of symptomatic diverticula are vital to the prevention of complications, such as malignancy. The management of symptomatic MD is accomplished through diverticulectomy. Robijn et al proposed a scoring system based on the evaluation of the risk of complications of a nonresected MD against the complications that arise from resection. They suggested that patients with an asymptomatic MD and a risk score < 6 should be left alone, while a risk score > 6 indicates the need for resection. The aim of this study was to provide a review of the literature on MD, with an emphasis on the late presentation and misdiagnosis of this congenital anomaly.

Keywords: congenital anomaly, omphalomesenteric duct, vitelline duct, GI bleeding, appendicitis, Crohn's disease

Introduction to the characterization and prevalence of Meckel's diverticulum

First described by Fabricus Hildanus in 1598, Meckel's diverticulum (MD) is the most common congenital anomaly of the gastrointestinal (GI) tract.¹ It is formed due to the incomplete obliteration of the omphalomesenteric duct during early gestation and is often described by the "rule of twos."²

Based on the patient's age and symptomatology, the primary method of detecting MD is through the use of technetium-99m-pertechnetate (99mTc-pertechnetate) scintigraphy, followed by exploratory laparoscopy for confirmation. However, false test results continue to be a problem, and other imaging modalities such as computed tomography (CT) have been employed in situations where scintigraphy has failed to be diagnostic.^{1,3,4}

MD often remains asymptomatic and can present later in life, masquerading as other GI pathologies such as appendicitis, Crohn's disease, or Littre's hernia;⁵⁻¹⁰ herein lies the potential for misdiagnosis. Other symptoms and complications may also arise due to MD, such as GI bleeding, inverted MD, iron deficiency anemia, and malignancy.¹¹⁻¹⁴

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The management of MD remains controversial. While the resection of symptomatic diverticula is generally agreed upon, the management of MD that is not symptomatic remains debatable. Some studies suggest that asymptomatic diverticula should be left alone, as the chance they will become symptomatic in the future is relatively low.¹⁵ However, other studies have recommended preventive diverticulectomies.^{1,15}

Diverticulectomy is commonly performed via laparoscopy alone, or laparoscopic-assisted procedures.^{16,17} The greatest advantage of a laparoscopic-assisted procedure is the ability to perform an extracorporeal resection, which allows for palpation of the MD for ectopic tissue. This increases the likelihood of resecting all of the ectopic tissue and reduces the need for a subsequent procedure.¹⁸

The aim of this study is to provide a review of the literature on MD, with an emphasis on late presentation and misdiagnosis of this congenital anomaly.

History

The earliest description of this congenital anomaly was made in 1598 by Fabricius Hildanus.^{1,16,19} Nearly a century later, one of the first cases of the diverticulum in the terminal ileum was recorded by the German surgeon, Lavater, in 1671.²⁰ This outpouching of the small intestine was later illustrated in detail by Ruysch of Leyden, in 1707.^{1,20} Several decades later, Morgagni described the diverticulum in the duodenum, ileum, and the rectum. His descriptions included the developmental anatomy of the diverticula in the ileum, and he made connections between this developmental process and disease occurrence.^{1,20}

It wasn't until the early 1800s that Johann Friedrich Meckel made his contributions to our current understanding of the development of the diverticulum. Meckel described the clinical picture and the embryological features of this congenital anomaly in 1809, giving rise to its present name.^{1,16} He examined 22 pediatric cadavers and described the various stages of malformations such as the omphalomesenteric cyst, omphalomesenteric fistula, umbilical sinus, and mesodiverticular band. More importantly, Meckel connected these malformations to the incomplete obliteration of the omphalomesenteric duct.¹

Meckel's contributions were followed by a treatise he wrote on normal and pathological anatomy and his translation of Casper Fredrich Wolff's monograph, *Ueber die Bildung des Darmkanals in Bebrüteten (About the Formation of the Intestinal Canal in Embryos)*, which described the development of the intestine in a chick.¹⁶ He also wrote treatises on

the diverticulum in 1809, in which he reviewed the existing literature and described the development and anatomy of diverticula persisting in the terminal ileum, and connected pathologies to this developmental process.^{1,20} Ultimately, Meckel illustrated this intestinal diverticulum in his "Tabulae anatomico-pathologicae."²¹

Prevalence, anatomy, and embryology

Prevalence and anatomy

MD is often described in text books for medical students by using the "rule of twos." This rule simplifies the prevalence and anatomy of MD, as occurring in 2% of the population; twice as common in males; in individuals less than 2 years old; and usually protruding from the ileum approximately 2 feet from the ileocecal valve.² While this rule is convenient, Moore et al reported²² that, in fact, MD occurs in 2%–4% of infants; is three to five times more prevalent in males than in females; is usually 3–6 cm long; and is observed as a finger-like pouch arising from the antimesenteric border of the ileum, about 40 to 50 centimeters proximal to the ileocecal junction. The average size of the diverticulum is 2.9 cm long and 1.9 cm wide;¹ however, Limas et al²³ have reported "giant" MD, with dimensions as large as 16 cm long and 4 cm wide.

Embryology

The extraembryonic membrane, known as the yolk sac, is seen by the fourth week of gestation. At this time, the yolk sac becomes host to the formation of the neural tube, as somites rapidly develop over its caudal rim.¹ The yolk sac narrows as the body of the embryo folds and the ectodermal, mesodermal, and endodermal layers form. The GI tract arises from the endoderm and forms the foregut, hindgut, and midgut. Initially, the midgut remains open to the yolk sac, but, as development progresses, the yolk sac narrows further and reduces communication with the midgut. The yolk sac eventually reduces to a slim stalk, which is known as the omphalomesenteric duct, the vitelline duct, or simply as the yolk stalk.²⁴ Nutrients are transported between the yolk sac and the developing embryo through the yolk stalk.¹ By the sixth week of gestation, the yolk stalk and sac disappear along with the vitelline arteries.¹

Failure of the vitelline duct to regress may result in a variety of congenital anomalies (Figure 1). The classic MD is a finger-like outpouching of the ileal wall. An omphalomesenteric fistula can occur when the vitelline duct in the abdominal wall remains patent. An isolated omphalomesenteric cyst,

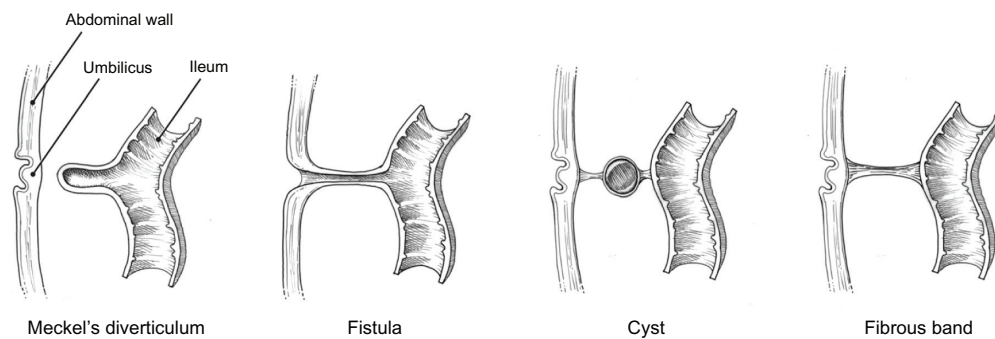


Figure 1 Illustration of the congenital anomalies that may arise from incomplete obliteration of the vitelline duct.

Note: From left to right: Meckel's diverticulum, omphalomesenteric fistula, omphalomesenteric cyst, and omphalomesenteric ligament.

suspended by ligaments, may also form. Finally, a fibrous band, known as an omphalomesenteric ligament, may form, connecting the ileum to the abdominal wall at the level of the umbilicus.²⁴

MD contains all three layers of the intestinal wall. It also has its own blood supply – a branch of the superior mesenteric artery, called the vitelline artery. This blood supply makes it vulnerable to infection and a candidate for obstruction, similar to the appendix.^{1,16}

Nearly half of all MD contain ectopic tissue. While the exact cause for this ectopic tissue is unknown, the occurrence of heterotopic gastric and pancreatic mucosa with an MD is common.¹ It has been suggested that prior to the union of the anterior and posterior pancreas during development, small buds are left in the foregut and move with subsequent gut development. Other ectopic tissues such as colonic, duodenal, jejunal, hepatic, and endometrial, have also been reported in the literature; however, these cases are rare and have not been known to result in complications.^{1,20}

Detection methods and recognition of late presentation

While MD remains mostly asymptomatic in the population, the probability of a symptomatic presentation is between 4%–6%. This incidence has been reported to decrease with age. MD can present with a variety of symptoms ranging from intestinal obstruction, bleeding, and perforation.^{1,23} In a 10-year study of the adult population with MD, Stone et al²⁵ reported that the most common presentation was diverticulitis, followed by obstruction. Acute bleeding was the least common presentation in this group, and the most common ectopic tissue found was gastric mucosa, as one would expect.

The primary method of detecting MD is through the use of technetium 99m-pertechnetate (99mTc-pertechnetate) scintigraphy, followed by exploratory laparoscopy for confirmation.

In the following section, we will discuss several modalities with which MD can be identified, either directly or through the exclusion of other pathologies.

99mTc-pertechnetate scintigraphy

Harden was the first to propose the use of 99mTc-pertechnetate to detect ectopic gastric mucosa in 1962. In 1970, Jewett et al recommended its usage clinically to determine the location of MD.¹ While this diagnostic tool is highly effective in diagnosing children with MD, its sensitivity, specificity, and accuracy falls greatly when applied to adults, with values of 62%, 9%, and 46%, respectively.^{26,27} This difference may be attributed to the area of involvement of the ectopic gastric mucosa; scintigraphy detects areas with larger ectopic gastric mucosa, which are most likely to hemorrhage earlier.¹

Due to the lower values of sensitivity and specificity, scintigraphy is prone to producing false positive and negative results. False positives arise when other sites containing ectopic gastric mucosa pick up the 99mTc-pertechnetate, as well as due to vascular anomalies, bowel ulcerations, inflammation, and obstruction.³ False negatives arise when the ectopic gastric tissue in the diverticulum is very minimal or when the scintigraphic activity is diluted due to bowel hypersecretion or sudden hemorrhage.^{1,3} The accuracy of scintigraphy may be increased by several methods that will be discussed later.

Scintigraphy is often the initial diagnostic test performed to detect MD, due to its noninvasive and relatively expedient nature, as opposed to laparoscopic exploration, which is invasive and requires significantly more time and resources. Still, the use of 99mTc-pertechnetate in cases of profuse intestinal bleeding has been questioned due to its lack of accuracy and difficult implementation in emergency situations.⁵ In addition, a negative scan warrants the need for intestinal exploration. Another deficit of 99mTc-pertechnetate is that

it cannot detect specific complications of an MD, such as Meckel's diverticulitis or Meckel's diverticular bands.⁵

Computed tomography and ultrasound

If an initial 99mTc-pertechnetate scan is nondiagnostic and acute bleeding has been ruled out, computed tomography and ultrasonography can be used to detect an inflamed MD in a noninvasive manner (Figure 2).^{1,4} CT scans have been shown to be useful for diagnosing intestinal obstruction and are an excellent tool in situations where diagnostic laparoscopy is contraindicated. Ultrasound has been used to visualize Meckel's diverticulitis, which appears similar to appendicitis and produces wall hyperemia on a color Doppler.⁴

Laparoscopy

Many practitioners favor laparoscopy for diagnosis of MD because of its ability to delineate common manifestations of MD as well as those less common, such as torsion.⁵ Chu et al²⁸ studied the use of laparoscopy for diagnosis and treatment of patients with MD. They supported laparoscopy as a valuable method for the diagnosis of MD, providing accuracy of results while remaining a minimally invasive technique. As noted earlier, accuracy is important as MD can manifest in a variety of ways: short or long, with or without ectopic tissue, and possibly with an ulcerated ileum. Taking this into consideration, Chu et al suggested that, at least for symptoms of abdominal pain and GI bleeding, laparoscopic management is superior to other methods of diagnosis. They did not, however, recommend it for patients presenting with

symptoms of obstruction where it would be difficult to inflate the abdominal cavity, which is a necessary step for proper visualization of abdominal contents.²⁸

On the other hand, Fagenholz and de Moya reported a successful case of laparoscopic identification and treatment of obstruction of the small bowel due to an MD.²⁹ The authors did not mention why their procedure worked despite opposing contentions held in the literature, but perhaps it was due to the fact that the obstruction required merely the resection of the MD and the phytobezoar within it. This procedure was similar to laparoscopy-assisted diverticulectomy. Thus, peritoneal inflation may have been easier to establish in this procedure than in other procedures for GI obstruction.²⁹

Angiography and colonoscopy

These techniques have been shown to be useful in specific situations. Practitioners have been able to identify active bleeding by angiography or CT angiography.³⁰ The small intestine accounts for 5% of GI bleeding.³¹

Misdiagnosis and unusual findings in MD diagnosis: a review of case reports

MD can present in patients with a wide variety of symptoms that may mimic those of other acute and chronic GI pathologies. These presentations often steer the diagnostic process off course and interfere with appropriate treatment protocols.

Appendicitis

Perforation is a rare complication of MD. However, when it does occur, it presents in a manner similar to acute appendicitis. Due to the emergent nature of acute appendicitis, MD pathology is often not diagnosed until surgery. It is recommended that proper imaging studies for MD, such as ultrasound and CT, be performed prior to a more invasive approach if the patient is not in acute distress and does not require immediate surgery for acute appendicitis.¹

MD is misdiagnosed as appendicitis on ultrasound imaging in approximately 11% of all MD cases.^{5,6} The presentation of patients with a perforated MD mimics that of patients presenting with acute appendicitis. The perforation is usually caused by ingested material such as wood splinters and fish bones.^{32,33} However, mucosal trauma and tumors, such as leiomyosarcoma, within the MD have also been observed to cause perforation.^{11,34-37}

Canelas et al reported a case in which a 52-year-old man with a perforated MD presented with right lower quadrant

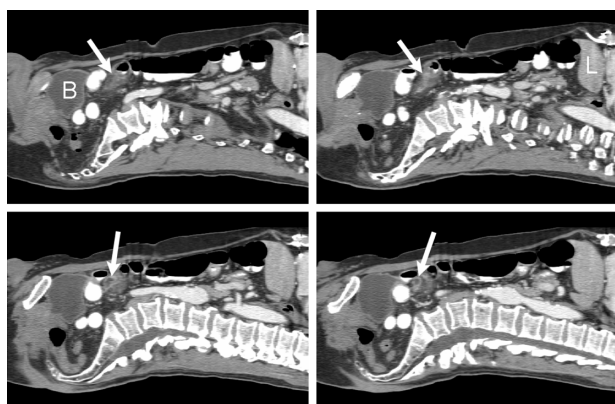


Figure 2 A sagittally reconstructed abdominal CT scan of a 75-year-old male with vague abdominal pain.

Notes: There is diffuse, mild small bowel dilatation and a fluid-filled projection from a pelvic small bowel loop, with an enhancing wall and evidence of inflammation (arrows). This is similar in appearance to an inflamed appendix; however, the location is more consistent with Meckel's diverticulum, which was found at laparotomy.

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Abbreviations: B, bladder; CT, computed tomography; L, liver.

abdominal pain, nausea, vomiting, and intermittent fever.³³ Similarly, A 12-year-old boy arrived at the emergency department with the sole symptom of severe periumbilical pain and was suspected to have a ruptured appendix.⁶

In 2011, Karabulut et al³⁸ found that out of 47 patients admitted for acute appendicitis that was negative upon exploratory laparoscopy, 13 were identified as cases of MD. This was the second most common diagnosis in this group, next to mesenteric lymphadenitis (Table 1).

Crohn's disease

MD perforation can mimic a variety of pathological conditions. Bondeson and Starck-Bondeson⁸ suggested that MD is a common finding among patients with terminal ileal Crohn's disease; however, the etiology of this is unknown.⁷ Wong et al¹¹ described a tall, thin, 21-year-old male with worsening constipation and increased flatus. The patient had a positive family history of Crohn's disease and reported a 10 kg weight loss, anorexia, decreased stool caliber, and dysuria. Diagnostic studies showed ileal thickening, ileocecal fistula, and a pelvic abscess, all thought to be due to Crohn's. However, exploratory surgery revealed a perforated MD adherent to an inflamed and fibrotic ileocecal fistula.¹¹ While MD masquerading as Crohn's disease is relatively rare, it is not uncommon to find the presence of MD in patients with Crohn's.

Similarly, Enc et al³⁹ described a 19-year-old female who presented with severe postprandial periumbilical pain for 1 year, anorexia, and a 10 kg weight loss. Enteroclysis revealed a 3 cm × 15 cm diverticular mass in the pericecal region, and the patient was treated for Crohn's disease. Conservative treatment failed, and the patient underwent an ileal resection, upon which a perforated MD was noted in addition to an ulcerated ileum. It is important to note that MD does not commonly present with anorexia, anemia, and ileocecal ulceration; however, MD should be considered in

the differential for Crohn's disease when these symptoms are present.³⁹

In a study examining patients undergoing hemicolectomy for Crohn's disease, Andreyev et al⁷ found that approximately 6% of patients between the ages of 18 and 56 years had MD with terminal ileal inflammation close to the mouth of the diverticulum. Only 41% of these diverticula were examined histologically, and none of the specimens showed evidence of heterotopic tissue. One patient had tested positive for MD preoperatively; however, this was considered to be a false positive due to the presence of Crohn's.

Based on the findings from their study, Andreyev et al⁷ concluded that the prevalence of MD is three times higher in patients with Crohn's disease than in the general population. While heterotopic gastric mucosa may cause terminal ileal inflammation, this is rarely seen as the cause of inflammation in these patients, either due to low symptom prevalence or milder presentations, which do not require surgical intervention. Thus, the authors rejected the notion that gastric mucosa in MD causes terminal ileitis.⁷

In addition to these conclusions, Andreyev et al⁷ proposed three possible explanations for the association between MD and Crohn's disease. Firstly, MD and Crohn's disease may be linked by a common underlying factor, explaining why they occur together in many patients. Secondly, Crohn's disease may be present for many years before it becomes clinically significant; it is unlikely, however, that Crohn's can act on patients prior to the obliteration of the vitelline duct, which promotes retention of the diverticulum. Finally, it is possible that MD predisposes patients to Crohn's disease by one of two mechanisms. One mechanism is by way of trapped infectious agents within the diverticulum that increase GI permeability; this correlates with data that suggest increased permeability is one of the early signs of Crohn's disease and a marker of increased familial risk and relapse. The second mechanism by which MD may predispose patients to Crohn's disease is through the diverticulum's role in causing disruptions in gut motility, promoting ischemia, or even preventing proper lymphatic drainage. Any of these possibilities may predispose a patient to Crohn's disease.⁷

Littré's hernia

Littré's hernia refers to an incarcerated hernia that contains a small bowel diverticulum. This was first identified in 1700 by Alexis Littré.^{9,10} The two most common sites for this occurrence are in the inguinal and femoral regions.¹ Littré's hernia presents as a painful groin mass, without evidence of obstruction or peritonitis. Wollgast and Hilz reported

Table 1 Positive intraoperative findings of negative appendectomies in 47 patients

Diagnosis	Number of patients (%)
Mesenteric lymphadenitis	23 (15)
Meckel's diverticulum complications	13 (8.7)
Ovarian cysts	4 (2.6)
Ovarian torsion	2 (1.3)
Omental infarction	2 (1.3)
Tubo-ovarian abscess	1 (0.6)
Mesocyst	1 (0.6)
Acute pancreatitis	1 (0.6)

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35 cases of Littre's hernia before 1954, most of which were right-sided.¹⁰ Bowel necrosis, as well as Richter's hernia (herniation of the antimesenteric border of the small bowel), were ruled out in these patients, as none of them presented with fever or tachycardia. Page et al⁹ reported a case of a 77-year-old woman with a history of breast carcinoma who presented with nausea and vomiting. The symptoms were attributed to recent chemotherapy; however, the patient discovered a right groin bulge following a bout of emesis, which was confirmed by CT to be an inguinal hernia containing a loop of small bowel. The hernia was surgically reduced, and the hernia sac contained a cystic mass consistent with MD.⁹ Similarly, Baum and Olch⁴⁰ described a man with a left-sided groin mass, which was found to contain a gangrenous MD approximately 6 cm in length. Finally, a 61-year-old male presented with a right-sided lump in the groin and no other symptoms, and this was later revealed to be an MD within a femoral hernia.⁴⁰

GI bleeding

MD can often present with blood in the colon, particularly if the MD abnormality is located in an unusual location or if the ectopic tissue has caused ulceration. Ectopic gastric mucosa found in MD can often cause bleeding ulcers of the terminal ileum, from the ileocolic artery.^{12,13} GI bleeding associated with MD can present in fast, large volume episodes of painless, bright red blood loss, or as slow and occult blood loss with a guaiac-positive test result and, occasionally, anemia.¹³ GI bleeding can be explained in 62%–100% of cases by the ulceration of heterotopic gastric mucosa.⁴¹ However, Reddin and Rosenbaum stated that MD is an uncommon cause of GI bleeding.⁴²

In 2011, Yang et al¹² reported seven cases of MD with ectopic pancreatic tissue that occurred within the previous 30 years. These patients' ages ranged from 11–66 years (with a median age of 19.8 years). Yang et al¹² stated that lower GI bleeding was more common in older patients with MD with ectopic pancreatic tissue than in those patients who suffered from ulceration of the gastric mucosa. However, the pathophysiology of the bleeding is similar to that of gastric mucosa ulceration, as the mucosal secretions are responsible for causing GI ulceration as well as bleeding. Ectopic pancreatic tissue has also been linked to adenocarcinoma in some patients.⁴⁴

Oglevie et al¹³ reported a case of a 29-year-old who presented with sudden onset of painless rectal bleeding. Multiple diagnostic tests were negative and failed to identify the source of the bleeding. Finally, repeat contrast studies revealed an

MD in the right upper quadrant. Histopathological studies identified ulcerated ectopic gastric mucosa within the MD as the source of bleeding. Similarly, a 69-year-old female also presented with painless rectal bleeding.¹² A CT angiography was performed and detected active bleeding of the ileocolic artery. The source of bleeding was identified as an MD containing ectopic pancreatic tissue. Based on these findings, Yang et al¹² suggest that MD be considered in adult patients presenting with massive painless GI bleeding.

Inverted MD and iron-deficiency anemia

Iron-deficiency anemia is a common finding among patients with GI bleeding and MD ulceration.¹¹ However, there have been cases reporting inverted Meckel's diverticula in which patients present with microcytic anemia in the absence of bleeding. James et al⁴⁵ described a 52-year-old patient with an inverted MD presenting with remarkable iron deficiency, without any evidence of gross or occult bleeding. Resection of the diverticulum corrected the anemia, and the patient was subsequently symptom free. Histological studies did not reveal a source of the bleeding. Shelton et al⁴⁶ presented a similar case of a 48-year-old female with iron deficiency anemia, fatigue, and abdominal bloating. CT scan of the abdomen revealed an endoluminal mass in the distal ileum. The mass was discovered to be an inverted MD containing ectopic pancreatic tissue, upon surgical resection. A source of bleeding was not found; however, the patient's anemia resolved postoperatively.⁴⁶

Inverted MD does not always present in the aforementioned way. Steinwald et al⁴⁷ suggested that GI bleeding is more commonly associated with right-sided MD and that an inverted MD is more likely to cause bowel obstruction. This inversion may serve as a lead point for ileo-ileal intussusception and can also simulate a pedunculated small bowel polyp.¹ The anemia resulting from MD is usually secondary to a GI bleed.^{41,45,49,50} The etiology of anemia presenting with a nonbleeding MD remains unknown.

Malignancies within MD

The prevalence of ectopic tissue within an MD is between 10%–60% and usually remains asymptomatic.¹⁴ However, ectopic tissue has been associated with an increased risk of malignancy, especially pancreatic adenocarcinoma.^{7,44,51,52} Malignancy accounts for approximately 0.5%–3.2% of the complications associated with MD. The most common malignancy associated with MD is carcinoid tumor.^{53–56} Others include adenocarcinoma, pancreatic carcinoma, intraductal papillary mucinous neoplasm, GI stromal tumors (GIST)

and leiomyosarcomas, lymphoma, lipoma, adenomyoma, and villous adenoma.^{44,57–69} Table 2 provides a comprehensive list of demographics concerning MD-related malignancy.

Anderson⁵³ reported a case involving a 62-year-old male who presented with right upper quadrant abdominal pain, nausea, indigestion, and fatty food intolerance. Imaging studies were unremarkable, with the exception of a gallbladder ejection obstruction. Upon surgical exploration, the patient was diagnosed with MD with carcinoid tumor.⁵³ Carcinoid tumor associated with MD is usually diagnosed after bowel resection.^{14,36,70,71} Moyana⁵⁴ found that jejunoileal carcinoid tumors most similarly resemble the type associated with MD, on immunohistochemical analysis. The survival

rate for patients with locally-staged, small bowel carcinoid tumors in the absence of MD is approximately 75%.⁷² Olson et al⁷³ reported a 5-year survival rate of 83% in patients with a combination of MD and carcinoid tumor. The 5-year survival rate declined to approximately 50% in patients with metastases.

In addition to carcinoid tumors, 12% of MD-associated tumors are GIST.⁷⁴ Mitchell et al⁷⁵ documented a 39-year-old male with severe abdominal pain in the right iliac fossa, with localized peritonism, fever, and tachycardia. The patient had a previous appendectomy, and after a CT scan located a small bowel obstruction, a diagnosis of tumor or MD was suggested. Khoury and Aulicino⁶³ reported a 28-year-old male with severe abdominal pain, nausea, and vomiting. CT scan showed small bowel obstruction resembling a cystic mass. MD was the preoperative diagnosis. Chou et al⁷⁶ reported a 76-year-old female with lower abdominal cramping for two days, nonbloody diarrhea, and vomiting. CT scan revealed small bowel perforation and a distended diverticulum with fluid accumulation. Laparoscopy confirmed a perforated MD. Each of these patients was diagnosed with GIST upon resection of the MD.

Similarly, Chandramohan et al⁷⁴ presented a case of a 65-year-old male with constipation of 4-months duration and rectal bleeding for one month. A lobulated mass compressing the anterior wall of the sigmoid colon was found on CT examination. Exploratory laparotomy found a mass within an MD, which was later confirmed as GIST.⁷⁴ Finally, Sozen and Tuna⁷⁷ presented a 62-year-old female with periumbilical pain in the right iliac fossa and localized peritonism. A palpable mass was present in the right lower quadrant, and air pockets on X-ray indicated an obstruction. Upon surgical intervention for suspected appendicitis, a perforated MD with nodular mass was found.⁷⁷

Due to the nonspecific presentation of MD and associated complications, it is important to know that tumors associated with MD can present in a plethora of ways. The average age of presentation with symptomatic GIST is usually within the sixth decade. These are most commonly found in the stomach (60%–70%), and fewer (20%–30%) are found in the small intestine.⁶³

False test results and misdiagnosis

Just as with atypical symptoms and presentation, MD can also be misdiagnosed due to diagnostic errors. As mentioned previously, the 99mTc-perchnetate scan is the initial test of choice for detecting MD but may produce misleading results, especially in the adult population.^{42,78}

Table 2 Patient characteristics of all cases of Meckel's tumors

Patient characteristics	Number of patients (n = 158)	Percentage (%)
Gender		
Male	101	64%
Female	57	36%
Race		
African-American	148	94%
European	6	4%
Other	4	2%
Major pathological subtypes		
Carcinoid	121	76.5%
Adenocarcinoma	18	11.4%
GIST/leiomyosarcoma/sarcoma	17	10.8%
Lymphoma	2	1.3%
Tumor behavior		
Benign	0	0%
Malignant	158	100%
Tumor size		
<1 cm	14	48.3%
1–5 cm	6	20.7%
>5 cm	6	20.7%
Unknown	3	10.3%
Stage		
Localized	106	67.1%
Regional (positive nodes)	24	15.2%
Metastatic	16	10.1%
Unstaged	12	7.6%
Surgical treatment		
Performed	151	95.6%
Recommended but not performed	6	3.8%
Not recommended	1	0.6%
Radiation		
No radiation	156	98.8%
Beam radiation	1	0.6%
Unknown	1	0.6%

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Abbreviation: GIST, gastrointestinal stromal tumors.

Summers et al⁷⁸ reported a case in which a 47-year-old male presented with bloody diarrhea, fatigue, nausea, and vomiting. A ^{99m}Tc-pertechnetate scan produced noticeable radiotracer uptake in the mid-abdomen. Although this was an unusual location for MD, it became the most likely diagnosis until laparoscopy confirmed that this was in fact, an enlarged appendix containing a carcinoid tumor.⁷⁸ Similarly, Reddin and Rosenbaum⁴² discovered a false positive scan for MD in a 32-year-old patient presenting with rectal bleeding – a carcinoid tumor was found without MD. In a similar case, Tulchinsky⁴³ reported a patient with low-volume, intermittent small bowel bleeding who tested positive on a ^{99m}Tc-pertechnetate scan for MD but who was later diagnosed with a bleeding ileal ulcer without MD, by capsule endoscopy.

False negatives arise when the ectopic gastric tissue in the diverticulum is very minimal or when the scintigraphic activity is diluted due to bowel hypersecretion or sudden hemorrhage.^{1,3} Hod et al⁷⁹ suggested that a ^{99m}Tc-pertechnetate scan should be postponed if a barium procedure was recently performed. The accuracy of the scan can be improved if the test is repeated when the barium contrast is known to be fully cleared from the system.⁴⁴ Several other methods of increasing the accuracy of scintigraphy have also been proposed: the use of a more water-soluble isotope of technetium may increase uptake; histamine blockers may be used to inhibit pertechnetate secretion after uptake; and glucagon has been shown to prevent wash-out of the pertechnetate by peristalsis.^{1,20}

Treatment and prognosis

The management of MD is controversial, especially pertaining to diverticula that remain asymptomatic. According to some studies, the rate of complications arising from surgical removal of the diverticulum is approximately 1%, with a 5%–6% lifetime risk of complications, such as peritonitis.¹ Other studies suggest that asymptomatic MD should be left alone, while symptomatic diverticula should be removed at any time up to the age of 80 years.¹⁵

Artigas et al⁸⁰ conducted a 15-year retrospective study and concluded that asymptomatic MD should not be resected due to the likelihood that the diverticulum would not become symptomatic in the future. They also noted that the chance of the diverticulum becoming symptomatic decreased with age.^{1,80} However, other studies have recommended preventive diverticulectomy. An epidemiological study performed at the Mayo Clinic looked at 50 years' worth of data and concluded that all incidentally discovered diverticula should be resected at any age.^{1,15}

Robijn et al⁸¹ proposed a scoring system based on the risk of complications of a nonresected MD against the complications that arise from resection. They identified the most common risk factors for asymptomatic diverticula, which increase the risk of future complications. A Risk Score was created, based on the following four risk factors: male gender, young age (<45 years), diverticula length > 2 cm, and presence of a fibrous band (Table 3).⁸¹ The presence of ectopic tissue in the diverticulum has been associated with an increased risk of bleeding and ulceration but was not included in the Risk Score proposed by Robijn et al. They suggested that patients with an asymptomatic MD and a risk score < 6 should be left alone, while a risk score > 6 indicates the need for resection.

The goal of a Meckel's diverticulectomy is to resect the entire diverticulum along with ectopic gastric tissue, pancreatic epithelium, or peptic ulcers of the nearby ileum.⁵ This procedure is usually performed using laparoscopic surgery or laparoscopy-assisted surgery.

Laparoscopic surgical intervention

The goal of laparoscopic surgical intervention for a Meckel diverticulectomy is to introduce a laparoscope and conduct an intracorporeal resection of the MD, all ectopic tissue, and any ulceration in the ileum.¹⁷ If the base is narrow and has no mass in the ileum, then procedure calls for a simple wedge resection of the diverticulum and a transverse closure of the ileum, using a linear stapler. If the base is too wide or the patient has palpable ectopic tissue or inflammation, the involved bowel should be resected as well. Assuming this

Table 3 Risk Score proposed by Robijn et al, to determine the need for resection in patients with asymptomatic Meckel's diverticula

Risk factor	Points
Gender	
Male	3
Female	1
Age	
<45 years	2
≥45 years	1
Length of diverticulum	
<2 cm	2
≥2 cm	1
Presence of fibrous band	
Yes	3
No	0
Risk score	Total points

Reproduced with permission from Robijn J, Sebrechts E, Miserez M. Management of incidentally found Meckel's diverticulum a new approach: resection based on a Risk Score. *Acta Chir Belg.* 2006;106(4):467–470.⁸¹

wide MD is in the ileum, an ileoileostomy would then be performed to reconnect the separated parts of the ileum.¹⁷

The use of laparoscopy for MD has been debated due to the risk of recurrence if any ectopic tissue is left behind. Upon examination of the external appearance of the diverticulum, some have concluded that a simple transverse resection can be done for long diverticula, as these will have their ectopic gastric tissue at the distal end.⁸² However, in short diverticula, the ectopic tissue can exist in nearly any area and should be removed through an ileal resection with end-to-end anastomosis or through a wedge resection done after exteriorization, in order to ensure the removal of all ectopic tissue and avoid future complications necessitating a second surgery.⁸²

Mukai et al,⁸² among others, have set measurable guidelines defining MD as long or short. These guidelines have been based on the knowledge that heterotopic tissue will grow from the pluripotent yolk sac, at the distal end of a MD. Varcoe et al⁸³ believe that a height to diameter ratio > 2 indicates a long MD and should be resected using a simple transverse approach, whereas a value < 2 indicates a short MD. They suggested that an MD should be resected via an ileal resection with end-to-end anastomosis.⁸³ Mukai et al⁸² believe a height to diameter ratio > 1.6 indicates the presence of heterotopic gastric epithelium in the distal end of the diverticulum, whereas a height to diameter ratio value < 1.6 indicates a clinically significant risk of having epithelium in nearly all areas of the diverticulum.

Laparoscopic-assisted transumbilical Meckel's diverticulectomy (LATUM)

LATUM involves fewer incisions than traditional laparoscopic surgery. An incision is made at the umbilicus, into which the laparoscope is introduced with a trocar.¹⁶ The laparoscope is then used to bring the MD out through the incision. In this way, the base of the diverticulum may be palpated for ectopic tissue, greatly diminishing the risk of residual tissue being left in the ileum and decreasing the need for a subsequent surgical procedure.¹⁶ Thus, the greatest advantage of LATUM is the ability to perform an extracorporeal resection, which allows for palpation of the MD and greater success at detecting ectopic tissue than does the laparoscopic technique on its own, as well as remaining less invasive than an open procedure.¹⁸

Conclusion and future directions

MD is the most common congenital anomaly of the GI system. Despite this, it is often asymptomatic and therefore, left undetected until later in life. This diverticulum can

present in a myriad of ways, often mimicking appendicitis, and is easily misdiagnosed. Left undiagnosed, significant GI distress can result, including an increased potential for GI bleeding, nonhemorrhagic iron deficiency anemia, and many types of GI cancer.

The key to prevention of complications is early diagnosis and treatment. While treatment of asymptomatic diverticula remains debatable, untreated symptomatic MD can have dire consequences. Thus, it is important for clinicians to include MD as a possible differential diagnosis, when presented with older patients displaying nonspecific GI symptoms.

Disclosure

The authors declare that they had no conflicts of interest in this work.

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