The final article in this issue focuses on less common diseases that surgeons are called on for management options. These five topics—volvulus, carcinoid, lymphoma, gastric varices, and gastric outlet obstruction (GOO) from peptic ulcer disease—are frequently used to evaluate surgical knowledge. Knowledge of these topics is useful for residents preparing for an in-training examination or board certification. Patients with these diseases require multidisciplinary management with oncologists and/or gastroenterologists, and mastery of these topics allows surgeons to effectively participate in the multidisciplinary care of these patients and advocate for surgical management when appropriate.

GASTRIC VOLVULUS

Gastric volvulus is a rare condition defined as an abnormal rotation of all or part of the stomach. Volvulus may occur along the vertical axis, mesoaxial torsion, or more commonly along an organoaxial direction. The less common mesoaxial torsion is associated with a less than 180° twist and the stomach situated below the diaphragm;
it is not discussed further. Organoaxial torsion patients have a diaphragmatic defect and a greater than 180° torsion and demand immediate surgical attention. The diagnosis of volvulus should be suspected based on the findings of Borchardt triad, initially reported more than a century ago, that include severe upper abdominal pain, retching with little vomitus, and inability to pass a nasogastric tube for decompression.¹ Not all patients with gastric volvulus demonstrate acute obstructive symptoms requiring emergency surgery; instead, a chronic condition that is minimally or asymptomatic might be discovered incidentally on a radiographic study performed for an unrelated purpose. Often the first tests performed in patients with chest or abdominal complaints are plain films that may demonstrate an intrathoracic stomach. Upper gastrointestinal (GI) contrast study has remained the gold standard and if performed with the stomach in the twisted state depicts an upside-down stomach.² CT scanning showing a double bubble with a transition line is gaining popularity as the imaging modality of choice.³ In the acute setting with severe symptoms, upper GI and CT scanning sensitivity can be limited by a patient’s inability to tolerate oral contrast material. Even on a CT without oral contrast, the clearly intrathoracic twisted stomach is evident.

The treatment of gastric volvulus is surgical and the goal of surgical therapy is reduction, decompression, débridement, and prevention of recurrence. Through a midline laparotomy, the stomach is reduced and assessed for viability. The stomach’s rich vascularity makes nonviability uncommon. Nonviable portions of the stomach are resected and a tube gastrostomy tube or suture gastropexy can be performed to prevent recurrence. Gastrostomy tubes placed in these situations are ideally situated in the left midabdomen, lower than where a typical open gastrostomy is placed to help fix the typically dilated stomach within the abdominal cavity. Care should be taken to place the tube low on the abdominal wall; however, undue tension created by pexing the stomach too low on the abdomen can result in dislodgement of the tube with intraperitoneal leak.⁴,⁵ Surgery for acute gastric volvulus is a surgical emergency and mortality is closely associated to delay in diagnosis. Surgery for chronic volvulus is elective and is performed to alleviate symptoms and prevent future complications. Minimally invasive techniques have been described and may reduce the morbidity associated with the open procedure.⁶,⁷ Because associated diaphragmatic defects should be expected, surgeons who use a laparoscopic approach must be facile with advanced techniques, including mesh placement and suture fixation. Endoscopic reduction has been described in selected groups of patients.⁸–¹⁰ Because of the increased risk of gastric perforation during endoscopic reduction, this technique should be limited to patients medically unfit for surgery. The outcome for gastric volvulus is dependent on timely diagnosis and proper management. The reported mortality rate from acute gastric volvulus is 15% to 20% and for chronic gastric volvulus is 0% to 13%.⁴,¹¹

GASTRIC CARCINOID

Gastric carcinoid tumors are rare and comprise approximately 8.7% to 30% of GI carcinoids and approximately 1% of all gastric neoplasms.¹² Also known as neuroendocrine tumors, they are derived from the enterochromaffin cells of the gastric corpus that mediate the secretion of histamine, which stimulates their unique receptors on parietal cells to stimulate acid production. The tumors are submucosal and most commonly occur in the fundus or antrum of the stomach. Three subtypes are described. Type I lesions are the most common (approximately 80% of the total) and are associated with the hypergastrinemia found in chronic atrophic gastritis,
pernicious anemia, autoimmune atrophic gastritis, and chronic antacid treatment with histamine blockers and proton pump inhibitors. Hypergastrinemia results in enterochromaffin cell hyperplasia, which may give rise to type I gastric carcinoids. Tumors are more commonly seen in women over age 50 and usually small in size and asymptomatic at time of diagnosis. Type II lesions (approximately 5% of the total) are associated with gastrinomas, are equally distributed in men and women, and can be found in patients with Zollinger-Ellison syndrome or in patients with multiple endocrine neoplasia, type I. Type III (approximately 15% of the total) gastric carcinoids occur sporadically, presenting as solitary large tumors, and are not associated with hypergastrinemia. They occur more commonly in men and have high malignant potential; approximately 50% of patients have metastatic disease at the time of diagnosis. Biochemical analysis includes measuring 24-hour urine levels of 5-hydroxyindoleacetic acid, which is a serotonin metabolite. A pentagastrin provocative test may be performed if the 24-hour urine levels of 5-hydroxyindoleacetic acid is inconclusive. Because of their small size and asymptomatic nature, most carcinoid tumors are found incidentally during esophagogastroduodenoscopy performed for other reasons. Endoscopic ultrasonography can be used to determine size and depth of the lesion. CT and MRI can be used to identify the primary tumor, local lymph node involvement, and metastatic spread. Somatostatin receptor scintigraphy (95% sensitive) is used to localize carcinoid tumors not seen on other imaging modalities. Type I lesions less than 1 cm and confined to the mucosa may be removed endoscopically. Surveillance endoscopy is recommended to monitor for recurrence. Antrectomy is recommended for type I gastric carcinoids when there are more than 5 lesions or 1 lesion greater than 1 cm. For older patients discovered to have an indolent disease, however, such as a type I gastric carcinoid, a valid argument can be made for consideration of observation and stopping antacid hypergastrinemia causing medications if a patient’s symptoms allow. Type II tumors less than 1 cm may be treated endoscopically; however, multiple lesions and tumors greater than 1 cm should be removed by partial gastrectomy. Because type III lesions often present with local lymph node involvement, treatment includes partial or total gastrectomy with extended lymph node resection. Type I gastric carcinoids are generally benign and slow growing and have an excellent 5-year survival rate of greater than 95%. Type II lesions are of intermediate malignant potential and overall have a good prognosis, 70% to 90% five year disease free survival. Type III lesions have the most malignant potential and a 5-year survival rate of less than 35%. The differentiation of type I from type III carcinoids is critically important because an aggressive approach to type III lesions is indicated whereas nonoperative observational management may be appropriate for type I lesions. A larger lesion that cannot be explained by a condition or medication associated with gastrin excess should be approached like a gastric adenocarcinoma, with radiographic staging and consideration of aggressive local and regional resections.

GASTRIC LYMPHOMA

The stomach is the most common site of primary extranodal non-Hodgkin lymphoma (NHL) of the GI tract. The two main histologic subtypes are mucosa-associated lymphoid tissue (MALT) NHL and diffuse large B-cell NHL. Chronic gastritis associated with Helicobacter pylori infection has been shown to be the cause of MALT and atrophic gastritis has been proposed as a risk factor for diffuse large B-cell NHL. As with most patients with NHL, there is a male predominance. Symptoms are nonspecific and include abdominal pain, dyspepsia, nausea, vomiting, and anorexia. Constitutional symptoms are uncommon. Upper GI bleeding may be the presentation in up
to 20% to 30% of patients; gastric obstruction and perforation occur less frequently.
Diagnosis is obtained by endoscopic biopsy. Gastric MALT is characterized by the
presence of lymphoepithelial lesions that are formed by invasion of single glands by
aggregates of neoplastic cells with centrocyte morphology. Lymphoma diffuse large
B-cell is characterized by a centroblastic morphology. Staging procedures for
primary gastric lymphomas include laboratory analysis for lactate dehydrogenase;
β2-macroglobulin; CT of the abdomen, pelvis, and chest; and bone marrow aspiration
and biopsy. Positron emission tomography has been shown beneficial for diffuse large
B-cell to examine the volume of disease at time of diagnosis. Three staging systems
are described and used to guide therapy: the Ann Arbor classification system, the
Lugano staging system, and more recently the Paris staging system (Table 1). The
Paris staging system is a modified TNM classification system that describes the depth
of tumor infiltration the extent of nodal involvement and the extent of local tissue infil-
tration by lymphoma.

Treatment of low-grade MALT lymphomas confined to the stomach can be accom-
plished by eradication of *H pylori* with antibiotics. Complete remission can be seen in
up to 70% of patients. Relapse is associated with *H pylori* reinfection and, therefore,
endoscopic follow-up is recommended. For patients unresponsive to antibiotics or for
the subset of *H pylori*–negative cases (approximately 10%), radiation therapy alone in
patients with early stages (I and II) can achieve greater than 95% complete response
rates. If patients have contraindications to radiation therapy, a single-agent chemo-
therapy (rituximab) can also be used.

### Table 1
Comparison of Lugano, Ann Arbor, and Paris staging systems in primary GI lymphomas

<table>
<thead>
<tr>
<th>Lugano Staging System for Gastrointestinal Lymphomas</th>
<th>Ann Arbor Stage</th>
<th>TNM Staging System Adapted for Gastric Lymphoma</th>
<th>Tumor Extension</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage IIE  Confined to G1 tract</td>
<td>I_E</td>
<td>T1 N0 M0</td>
<td>Mucosa, submucosa</td>
</tr>
<tr>
<td>I_E1 = mucosa, submucosa</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I_E2 = muscularis propria, serosa</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stage IIE  Extending into abdomen</td>
<td>II_E</td>
<td>T1-3 N1 M0</td>
<td>Perigastric lymph nodes</td>
</tr>
<tr>
<td>II_E1 = local nodal involvement</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>II_E2 = distant nodal involvement</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stage IIE  Penetration of serosa to involve adjacent organs or tissues</td>
<td>II_E</td>
<td>T4 N0 M0</td>
<td>Invasion of adjacent structures</td>
</tr>
<tr>
<td>Stage III-IV Disseminated extranodal involvement or concomitant supradiaphragmatic nodal involvement</td>
<td>III_E/IV</td>
<td>T1-4 N3 M0/T1-4 N0-3 mL</td>
<td>Lymph nodes on both sides of the diaphragm/distant metastases (eg, bone marrow or additional extra nodal sites)</td>
</tr>
</tbody>
</table>

Advanced-stage MALT lymphomas (stage III and IV) are rare and, because of its indolent nature, chemotherapy is not curative; therefore, asymptomatic patients can be observed without treatment. Indications for systemic therapy include candidate for clinical trial, symptomatic disease, GI bleeding, threatened end-organ function, bulky disease, steady progression, and patient preference.

Treatment of diffuse gastric large B-cell lymphoma is rituximab plus chemotherapy with anthracycline-based regimens (cyclophosphamide, vincristine [Oncovin], hydroxydaunorubicin, and prednisone [CHOP]; cyclophosphamide, epirubicin, vincristine [Oncovin], and prednisone [CEOP]; and cyclophosphamide, mitoxantrone [Novantrone], vincristine [Oncovin], and prednisone [CNOP]).

Surgery for primary gastric lymphoma is indicated for the following: primary radical treatment, severe bleeding or perforation, and as palliative treatment. Because surgery is associated with higher rates of morbidity when compared with radiation alone and chemotherapy alone, the treatment strategy has shifted toward organ preservation. The first-line chemotherapy regimens are combination chemotherapy that includes rituximab and CHOP (R-CHOP) or bendamustine and rituximab. Despite its aggressive nature, the 5-year survival rate of diffuse gastric large B-cell lymphoma is good, ranging from 73% to 90% after primary systemic therapy. The 5-year survival rate for patients with low-grade gastric MALT is greater than 91%, with 5-year survival rates ranging from 50% to 70% in patients with high-grade gastric MALT tumors.

ISOLATED GASTRIC VARICES

Isolated gastric varices occur in patients with segmental or left-sided portal hypertension due to splenic vein thrombosis. This condition must be differentiated from gastric varices seen in association with esophageal varices in the setting of portal hypertension. The management of patients with combined gastric and esophageal varices should be focused on the treatment of the underlying portal hypertension and is not discussed further in this article. Isolated gastric varices result from benign and malignant pancreatic pathology that cause splenic vein thrombosis. Direct tumor extension or the fibrosis associated with chronic inflammatory conditions can obstruct the splenic vein, causing thrombosis. Continued arterial flow to the spleen engorges the gland, resulting in splenomegaly that can only decompress through the normally small-caliber short gastric veins. These small veins dilate and can ulcerate through the gastric mucosa and cause upper GI bleeding. Patients with gastric varices develop large dilated submucosal veins in the stomach and may present with hematemeses, hematochezia, or melena.

The risk of bleeding from isolated gastric varices is low when compared with that of gastroesophageal varices. When isolated gastric varices are incidentally detected at endoscopy and proved the result of splenic vein thrombosis, treatment is unnecessary due to a low risk of bleeding. When upper GI endoscopy is used to evaluate a bleeding patient, however, the finding of gastric varices without esophageal varices is virtually diagnostic of splenic vein thrombosis. Abdominal imaging with contrast-enhanced CT or MRI is essential to evaluate the pancreas to differentiate benign from malignant etiologies. Laboratory analysis should include complete blood count, coagulation factors, and liver function test, and cancer antigen 19-9 levels may be helpful if benign versus malignant causes are not apparent on imaging studies. Initial treatment should focus on airway protection followed by large bore venous access and resuscitation with blood products. The treatment of choice for bleeding isolated gastric varices is splenectomy, which effectively eliminates the collateral outflow.
When bleeding gastric varices results in splenic vein thrombosis from pancreatic carcinoma of the body or tail of the gland, curative resection is highly unlikely and splenectomy in this situation should be judiciously used.\textsuperscript{33}

**BENIGN GASTRIC OUTLET OBSTRUCTION**

Patients with GOO may have either benign or malignant causes of blockage. In general, patients with malignant causes of GOO have advanced disease, a limited life expectancy, and imaging studies that demonstrate widespread metastases.\textsuperscript{34} When patients have signs and symptoms of GOO without evidence of metastatic cancer, endoscopy is typically performed to determine whether the cause of obstruction is benign or malignant. The distally obstructed stomach that does not permit easy passage of the endoscope may not allow for access to the tumor for biopsy. Therefore, surgeons should not be fooled by benign biopsies that may reflect the difficulty with sampling the tumor and should rely on abdominal imaging in addition to endoscopic findings to distinguish cancer from benign causes of obstruction. The topic of surgery in the setting of stage IV cancer is discussed by Patel and Kooby in the article on gastric adenocarcinoma elsewhere in this issue and not discussed further in this article. When imaging demonstrates isolated GOO and endoscopy fails to document cancer, patients should be evaluated for either surgical or endoscopic management of what is most likely a benign cause of obstruction, most frequently a result of chronic neglected peptic ulcer disease. Although balloon dilation and medical management of GOO, including *H. pylori* eradication, have been reported, the ultimate need for surgery increases with longer follow-up because some patients who initially respond to nonoperative management relapse and ultimately require surgery.\textsuperscript{35,36} Gibson and colleagues\textsuperscript{37} reported a surgical series of 24 patients treated by surgery for GOO from ulcer disease. This group from Memphis found that only one-third of patients were *H. pylori* positive, nonsteroidal anti-inflammatory drugs were a common cause of GOO from ulcer disease, and that all but one patient was dramatically improved by surgery. The choice of operation is either resection of the obstructed antrum and pylorus or bypass. Because a gastrojejunal bypass is ulcerogenic with acid-rich secretions bathing an unprotected jejunum, resection is preferred as long as the proximal duodenum can be safely divided and closed. Surgeons’ confidence in their ability to achieve a satisfactory duodenal closure drives the intraoperative decision as to whether resection or bypass is most appropriate. Ulcerogenic bypass procedures are preferred to the morbidity associated with the complications of a difficult duodenal stump and the potential for a leak and development of a duodenocutaneous fistula. Patients treated by gastrojejunostomy as a bypass of a benign obstructed stomach require acid reduction in the form of vagotomy or lifelong medication. Because the development of GOO by ulcer occurs over a protracted time course with symptoms that likely are ignored by patients, many of these patients are noncompliant and may do better with vagotomy instead of relying on lifelong medication.

**REFERENCES**


