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ORIGINAL ARTICLE

Gastric volvulus in children: the twists and turns of an unusual entity

Sarah K. Oh · Bokyung K. Han · Terry L. Levin · Robyn Murphy · Netta M. Blitman · Carmen Ramos

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Abstract
Background Gastric volvulus in children is uncommon, and characteristic radiographic findings might not be recognized.
Objective To present the spectrum of clinical and imaging findings, correlate the type of gastric volvulus with clinical outcome, and identify imaging findings to aid in early diagnosis.
Materials and methods Medical records and imaging findings of ten children with gastric volvulus were reviewed. Imaging included abdominal radiographs, upper gastrointestinal (UGI) series, and CT. The diagnosis (organoaxial, mesenteroaxial or mixed type) was made on the UGI series (n=9) and CT (n=1), and confirmed surgically in seven children.
Results Patients were classified based on presentation: four acute, four chronic, and two neonatal. All of the acute group (three mesenteroaxial and one mixed type) had abnormal radiographic findings: three spherical gastric distension, four paucity of distal gas, three elevated left hemidiaphragm, one overlapping pylorus and gastric fundus, one unusual nasogastric tube course, and one situs inversus. All underwent emergent surgery. Three had diaphragmatic abnormalities. One had heterotaxy. Patients in the chronic group (three organoaxial, one mesenteroaxial) had long-standing symptoms. Most had associated neurologic abnormalities. In the neonatal group, organoaxial volvulus was found incidentally on the UGI series.
Conclusion A spectrum of findings in gastric volvulus exists. Mesenteroaxial volvulus has greater morbidity and mortality. Radiographic findings of spherical gastric dilatation, paucity of distal gas and diaphragmatic elevation are suggestive of acute volvulus, particularly in patients with predisposing factors.

Keywords Gastric volvulus · Organoaxial · Mesenteroaxial · Children

S. K. Oh (✉) · B. K. Han · T. L. Levin · N. M. Blitman
Department of Radiology,
Children's Hospital at Montefiore Medical Center,
111 East 210th St,
Bronx, NY 10467, USA
e-mail: soh@montefiore.org

R. Murphy
Department of Radiology, Morristown Memorial Hospital,
Morristown, NJ, USA

C. Ramos
Department of Pediatric Surgery,
Children's Hospital at Montefiore Medical Center,
Bronx, NY, USA

Introduction

Although gastric volvulus is considered rare, a growing body of case reports suggests that it is more common in the pediatric population than previously thought [1]. Gastric volvulus has a broad spectrum of clinical presentations. As such, diagnosis might be delayed or missed. In patients with an acute presentation, gastric volvulus is life-threatening and requires rapid recognition and emergent surgical treatment [2, 3]. Subacute and chronic presentations, on the other hand, might go unrecognized [4]. We categorized gastric volvulus into discrete subtypes with identifiable clinical and radiographic features, and correlated the type of volvulus with the clinical outcome. Abdominal radiographic findings suggestive of the acute fulminant form of this condition were identified.

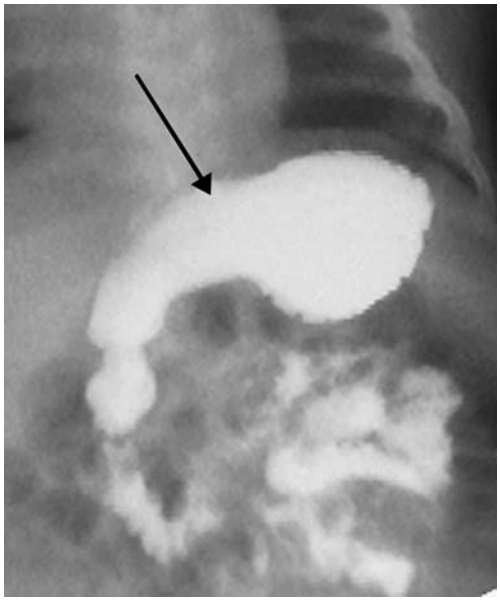

Fig. 1 Organoaxial volvulus (patient 9). Frontal view from a UGI series. The greater curvature (*long arrow*) lies to the right of and superior to the lesser curvature. The pylorus points inferiorly. There is no obstruction to the passage of contrast agent into the small bowel

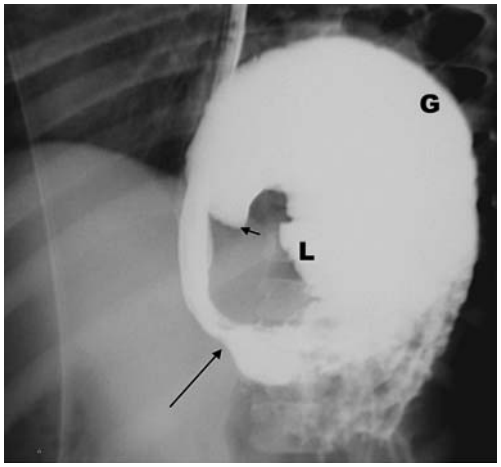

Fig. 3 Mesenteroaxial volvulus (patient 1). Single view from a UGI series. There is herniation of the stomach into the left chest through a Bochdalek hernia. There is close approximation and reversal of the normal relationship of the pylorus (*short arrow*) and the gastroesophageal junction (*long arrow*). The lesser (*L*) curvature maintains its rightward position relative to the greater (*G*) curvature of the stomach

Materials and methods

The medical records and imaging studies in ten children (eight boys, two girls) presenting with gastric volvulus between 2000 and 2005 were retrospectively reviewed. The study was approved by the Institutional Review Board (IRB). Ages ranged from 4 weeks to 17 years (mean 7 years). Images included abdominal radiographs, upper gastrointestinal (UGI) series, and CT. These were reviewed by three radiologists holding a Certificate of Added Qualification in pediatric radiology. Diagnosis was made by UGI series in nine children. In one child, findings of gastric volvulus were missed on abdominal radiographs, and CT was diagnostic.

Organoaxial (OA) volvulus was diagnosed when the greater curvature of the stomach was superior to the lesser curvature and the pylorus pointed inferiorly (Fig. 1). Partial OA volvulus was diagnosed when only a portion of the stomach demonstrated features of OA volvulus (Fig. 2). Mesenteroaxial (MA) volvulus was diagnosed when the pylorus was displaced superiorly and toward the left (shortening the distance between the pylorus and gastroesophageal junction), and the pylorus overlapped the gastroesophageal junction or gastric fundus (Fig. 3). Mixed-type volvulus was diagnosed when

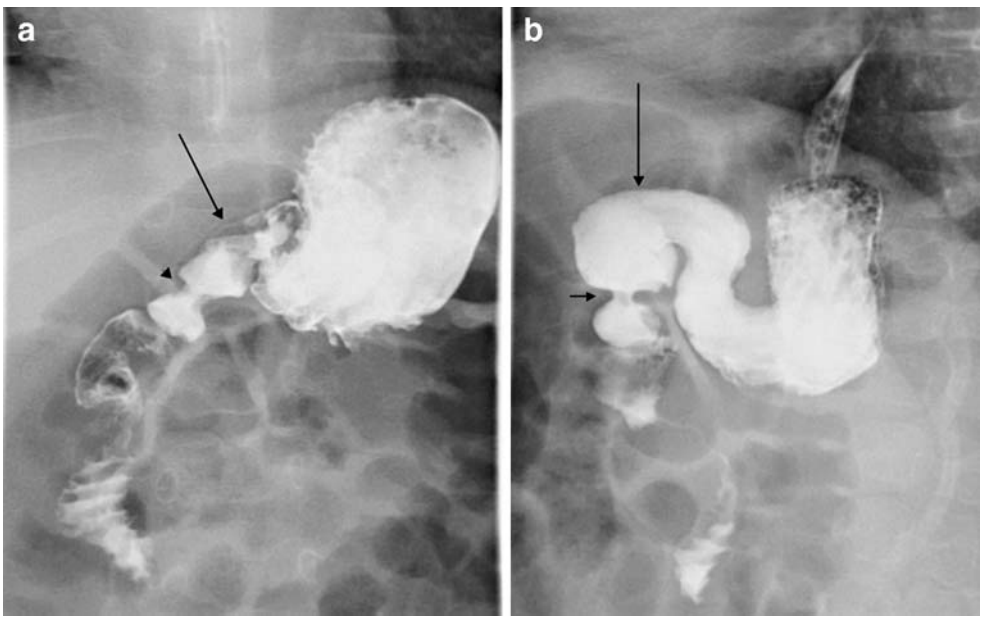

Fig. 2 Partial organoaxial volvulus (patient 5). AP (**a**) and lateral (**b**) views from a UGI series. The greater curvature (*long arrow*) lies to the right of and superior to the lesser curvature in the distal stomach only. The gastric fundus is normally positioned. Note the inferiorly pointing pylorus (*arrowhead* [**a**], *short arrow* [**b**]), a component of organoaxial volvulus

features of both OA and MA were present. Medical records were reviewed for clinical presentation, associated abnormalities and management. The patients were classified into three groups based on clinical presentation: acute fulminant, chronic intermittent, and neonatal groups. The type of volvulus was correlated with clinical outcome.

Results

Clinical types

The clinical findings are summarized in Table 1.

Acute fulminant type

Acute fulminant presentation was seen in four children, three with MA and one with mixed type (MT) volvulus. Diaphragmatic abnormality (including Bochdalek hernia or eventration) was present in three children. Heterotaxy/asplenia was seen in one patient with MA volvulus. All patients presented acutely with vomiting, abdominal distention and pain. Three underwent surgery within hours after the onset of symptoms. In one patient, diagnosis and surgical intervention were delayed, resulting in gastric necrosis and perforation.

Chronic intermittent type

Four children had a chronic intermittent presentation, with symptoms occurring over 3 months to 6 years. Three had associated neurologic abnormalities, including mental retardation, cerebral palsy, autism, and developmental delay. Three presented with OA volvulus, including one with partial OA volvulus (Fig. 2), and underwent gastropexy. The other patient, with a history of congenital diaphragmatic repair at birth and developmental delay, presented with MA volvulus without obstruction and did not undergo surgery because of other medical problems.

Table 1 Clinical findings

Type	Patient	Age	Sex	Subtype	Clinical presentation	Associated abnormalities	Management
Acute fulminant	1	17 months	M	Mesenteroaxial	Acute vomiting	Bochdalek hernia	Surgery: gastropexy
	2	2 years	M	Mesenteroaxial	Acute vomiting and abdominal distention	Complex congenital heart disease, heterotaxy, asplenia	Surgery: gastropexy
	3	8 years	M	Mesenteroaxial	Acute onset hematemesis	Trisomy 21	Surgery: partial gastrectomy and congenital diaphragmatic hernia repair
	4	17 years	F	Mixed type	Acute onset abdominal pain and distention, progressing to vomiting of foamy saliva[a]	Diaphragmatic eventration	Surgery: gastrectomy; jejunostomy; cervical esophagostomy
Chronic intermittent	5	6 months	M	Partial organoaxial	Intermittent nonbilious vomiting since birth		Surgery: gastropexy
	6	10 years	M	Organoaxial	Abdominal distention	Cerebral palsy, seizure disorder	Surgery: gastropexy
	7	10 years	M	Mesenteroaxial	Epigastric pain, and posttussive emesis	Congenital diaphragmatic hernia repair at birth. Developmental delay	No surgery
	8	13 years	F	Organoaxial	Postprandial abdominal pain, distention, and whole-food regurgitation	Pervasive developmental disorder, autism	Surgery: gastropexy
Neonatal	9	1 month	M	Organoaxial	Nasal flaring, rapid breathing, choking on saliva/formula since birth	Laryngotracheomalacia	No surgery
	10	1 month	M	Organoaxial	Feeding difficulty since birth	Larsen syndrome	No surgery

[a] Delay in diagnosis led to gastric necrosis and thrombosis of the left gastric artery.

Neonatal type

Two children had a benign clinical course with an incidental diagnosis of gastric volvulus on a UGI series performed for other indications. Both presented in the neonatal period and demonstrated intermittent OA volvulus.

Imaging

The imaging findings are summarized in Table 2.

Radiographs

Abdominal radiographs, acquired in five children (four with acute fulminant volvulus and one with chronic intermittent volvulus) showed abnormalities. Four children showed a single large spherical gas bubble (Fig. 4); in three, this was in the left upper abdomen or mid-abdomen, and in one (a patient with situs inversus) this was in the center to the right side of the abdomen. In the child in whom a spherical gas bubble was not present on the abdominal radiograph, the stomach had been decompressed by placement of a nasogastric tube. The tube followed an unusual course and outlined a MA volvulus (Fig. 5).

A paucity of distal bowel gas was seen in all children with acute fulminant volvulus, indicating gastric outlet obstruction. Overlap of the pylorus and gastric fundus and elevation of the left hemidiaphragm were also present. In one child with acute fulminant MA volvulus, situs inversus was demonstrated on the abdominal radiograph. MT volvulus was suggested in one patient in whom close approximation of the gastroesophageal junction and pylorus (typical of MA volvulus) as well as inversion of the greater curvature (typical of OA volvulus) were present (Fig. 6).

Upper gastrointestinal series

A UGI series performed in nine patients was diagnostic and determined the type of volvulus in each (Fig. 7). A UGI series was not performed in the one patient with acute fulminant volvulus in whom the diagnosis was missed on initial radiographs and made on CT imaging.

Table 2 Imaging findings

Type	Patient	Age	Sex	Radiograph	UGI series	CT
Acute fulminant	1	17 months	M	Elevation left diaphragm (Fig 4a), spherical gastric distention (Fig 4a)	MA volvulus herniated into chest (Fig. 3). (+) gastric outlet obstruction	
	2	2 years	M	Spherical gastric distention, heterotaxy, gastroesophageal junction/pylorus reversal	MA volvulus (Fig. 7b). (+) partial high-grade gastric outlet obstruction	MA volvulus, gastric distention, heterotaxy, asplenia
	3	8 years	M	Abnormal nasogastric tube course, elevation left diaphragm (Fig. 5a)	MA volvulus (Fig. 5b). (+) gastric outlet obstruction with nasogastric tube decompression	
	4	17 years	F	Spherical gastric distention, double air-fluid levels, gastroesophageal junction/ pylorus reversal (Fig. 6)	MT volvulus. (+) high-grade gastric outlet obstruction	MT volvulus, hypoperfusion liver and spleen, gastric pneumatosis, eventration left diaphragm (Fig. 8)
Chronic intermittent	5	6 months	M		Partial OA volvulus (Fig. 2). (−) gastric outlet obstruction	
	6	10 years	M	Spherical gastric distention, double air-fluid levels	OA volvulus. (−) gastric outlet obstruction	
	7	10 years	M		MA volvulus. (−) gastric outlet obstruction.	
	8	13 years	F		Intermittent OA volvulus (Fig. 7a). (−) gastric outlet obstruction	
Neonatal	9	1 month	M		Intermittent OA volvulus (Fig. 1). (−) gastric outlet obstruction	
	10	1 month	M		OA volvulus. (−) gastric outlet obstruction	

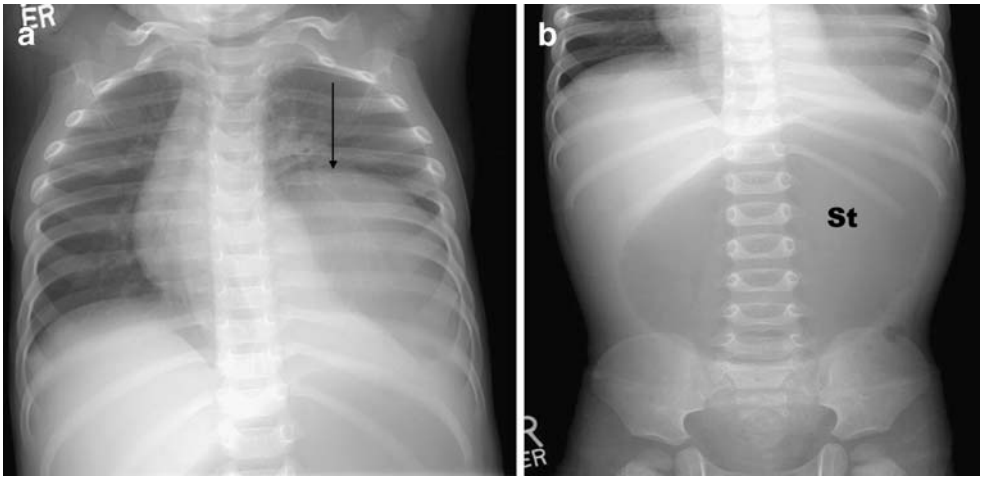

Fig. 4 Plain radiographs of mesenteroaxial volvulus (patient 1). **a** Chest: the left hemidiaphragm (*arrow*) is elevated. **b** Abdomen: note marked spherical distention of the stomach (*St*). There is no distal gas, indicating gastric outlet obstruction

CT

A CT scan was obtained in two children with the acute fulminant gastric volvulus. In both, the diagnosis was not made on initial plain radiographs. Although CT was not necessary in either child and it delayed surgery, adjunctive findings in one patient included heterotaxy (asplenia) and gastric distention, and in the other gastric pneumatosis, hepatic and splenic hypoperfusion, and a high-lying left kidney secondary to elevation of the left diaphragm (Fig. 8).

Discussion

The stomach is a dynamic structure and is anchored by the gastrohepatic, gastrophrenic, gastrocolic, and gastrosplenic ligaments. These ligaments allow the stomach to expand and contract yet maintain enough tension to prevent torsion along its long or short axis [5]. Congenital absence of one or more ligaments, ligamentous laxity and abnormal gastric distention all predispose to gastric volvulus [4, 6, 7]. Other abnormalities associated with gastric volvulus include a wandering spleen [8–11], intrathoracic kidney [12], malrotation with asplenia [5, 8] and diaphragmatic eventration [12–14]. Postsurgical interruption of gastric ligaments following liver transplantation [15] and laparoscopic Nissen fundoplication [16] can also predispose to gastric volvulus.

OA volvulus occurs when the stomach twists along its long axis, such that the greater curvature is positioned superior to and to the right of the lesser curvature. MA volvulus occurs when the stomach folds along its short axis, such that the greater and lesser curvatures are in their usual

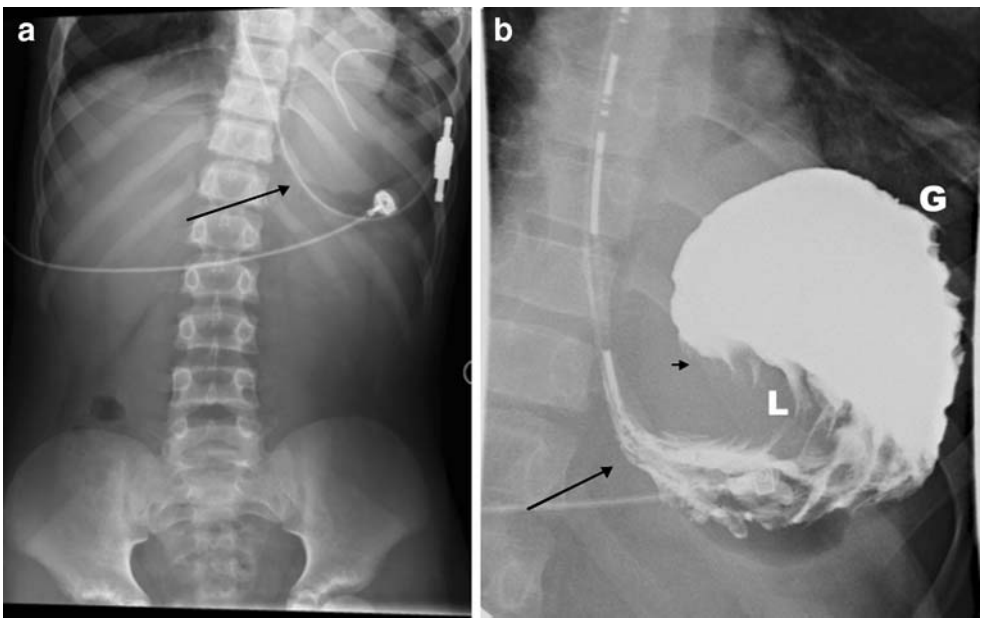

Fig. 5 Mesenteroaxial volvulus (patient 3). **a** Abdominal radiograph. The stomach has been decompressed by a nasogastric tube, which has an unusual course (*arrow*) suggesting a mesenteroaxial volvulus. The left hemidiaphragm is elevated and there is a paucity of distal gas. **b** Corresponding image from a UGI series. Note the close approximation of the pylorus (*short arrow*) and the gastroesophageal junction (*long arrow*), the relative positions of which are the inverse of normal. The lesser curvature (*L*) and greater curvature (*G*) maintain their normal relationship. These findings confirm a mesenteroaxial volvulus

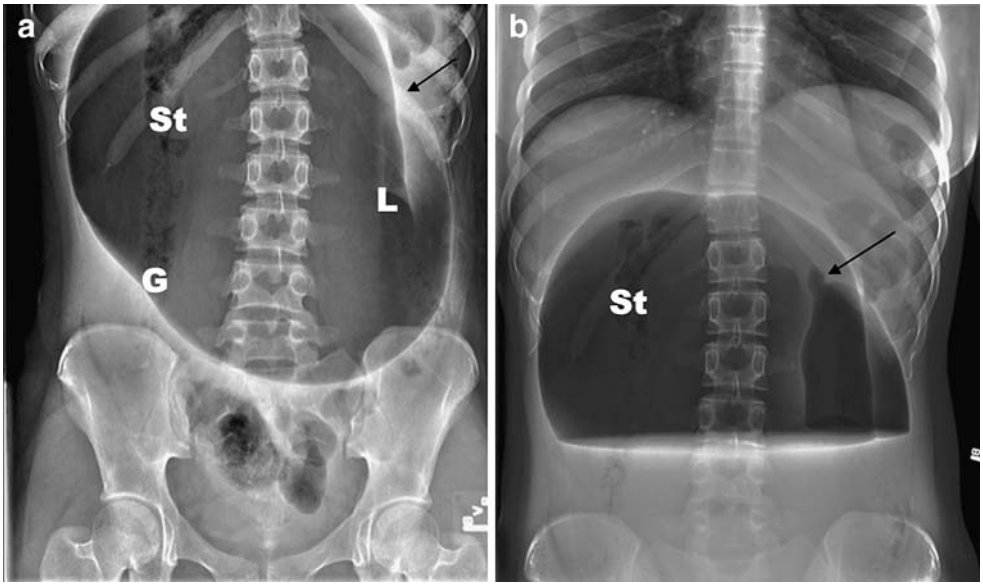

Fig. 6 Plain radiograph. Mixed-type volvulus (patient 4). Supine (**a**) and upright (**b**) views of the abdomen. There is marked spherical distension of the stomach (*St*) with two air-fluid levels. Features of both organoaxial and mesenteroaxial volvulus are present: the pylorus (*arrow*) projects over the fundus, typical of mesenteroaxial volvulus, and the greater curvature (*G*) of the stomach lies to the right of the lesser curvature (*L*), typical of organoaxial volvulus

positions relative to each other [12]. Reversal of the relationship of the gastroesophageal junction and pylorus is present. MT volvulus includes features of both OA and MA volvulus. MA and MT volvulus often present acutely, while OA volvulus might be more indolent. In severe cases, closed-loop obstruction, strangulation and gastric ischemia can occur. This is more frequent in MA volvulus [17] and was not seen in any patient with OA volvulus in this series. MA volvulus is more often associated with diaphragmatic abnormalities.

In the acute fulminant type of gastric volvulus, which includes MA and MT volvulus, there is close approximation of the gastroesophageal junction and pylorus (Fig. 3). This creates a narrow pedicle about which the stomach can twist, leading to gastric obstruction and ischemia. Diaphragmatic abnormalities (Fig. 5) as well as congenital and surgical factors can contribute to abnormal gastric fixation and predispose to twisting of the stomach. In contrast, in OA volvulus, the gastroesophageal junction and pylorus, which form the long axis of the stomach, maintain their normal anatomic positions and are not in close proximity (Fig. 1). The stomach might flip upward along its long axis, but because it does not twist on itself there is no risk of ischemia. Obstruction, however, can occur. Gastric distention might serve as a predisposing factor.

Clinical presentation was found to correlate strongly with volvulus type. In our series, children with the acute fulminant type of volvulus presented with acute onset of vomiting and

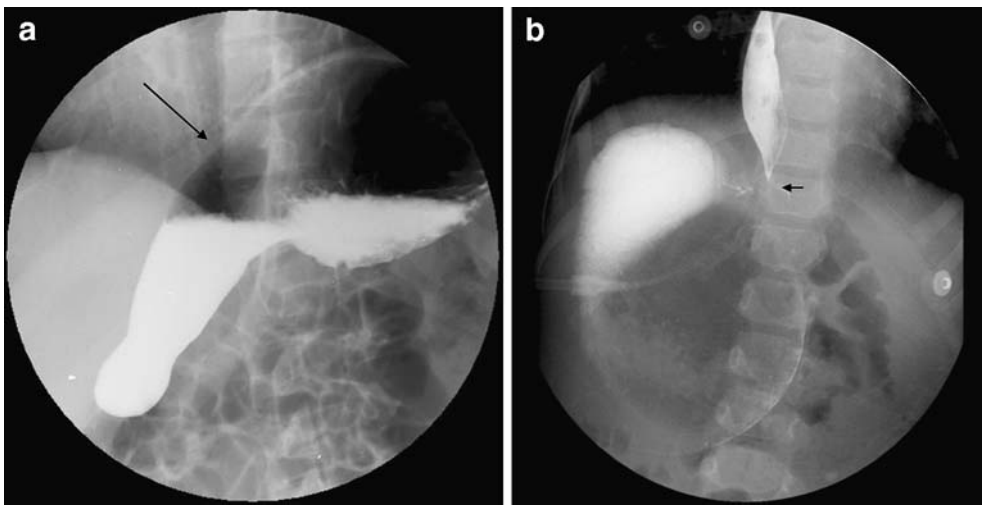

Fig. 7 UGI series. **a** Organoaxial volvulus (chronic intermittent type, patient 8). Upright view shows two air-fluid levels characteristic of organoaxial volvulus. The greater curvature (*long arrow*) lies above and to the right of the lesser curvature. There is a normal distal bowel gas pattern. **b** Mesenteroaxial volvulus (acute fulminant type, patient 2). The spherically distended stomach is seen in the right upper quadrant in this patient with heterotaxy. In contrast to organoaxial volvulus, the gastroesophageal junction and pylorus are in close approximation (*small arrow*). There is a paucity of distal bowel gas

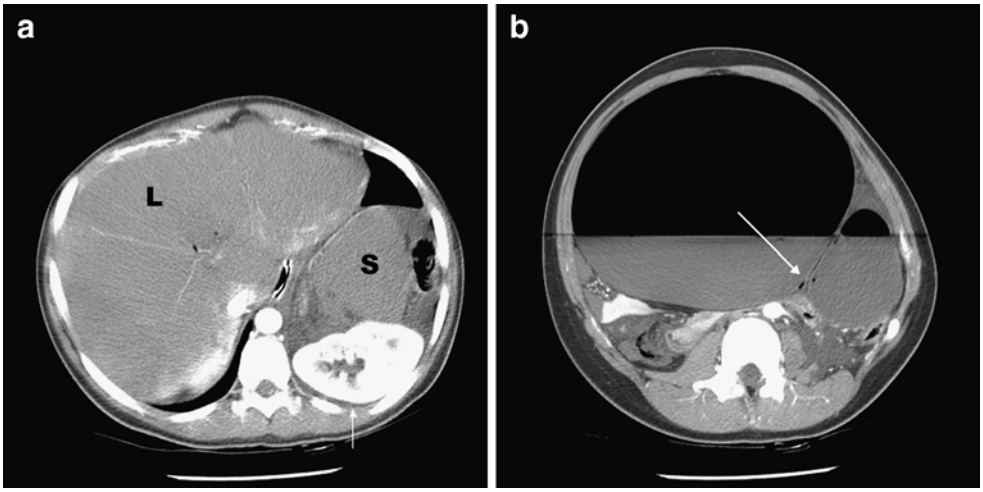

Fig. 8 Axial contrast-enhanced CT images. Mixed-type volvulus (patient 4). **a** Upper abdomen. There is hypoperfusion of the liver (*L*) and spleen (*S*) as well as a high-lying left kidney (*arrow*). **b** Mid-abdomen. Extravasation of intravenous contrast agent is suspected by the presence of high-density material around the liver. A markedly distended stomach is present with two air-fluid levels, as well as gastric pneumatosis (*arrow*)

abdominal distention lasting hours to days prior to surgical intervention. All were either MA or MT in configuration. All of these children had predisposing factors, including diaphragmatic abnormality and asplenia. Most underwent emergent surgery within hours of presentation.

Although both neonatal and chronic intermittent gastric volvulus are usually of the OA type, they fall into separate distinct categories. The neonatal type of gastric volvulus is not clinically significant. It is seen in the neonatal period and is likely related to circulating maternal hormones causing lax ligamentous attachments [1, 2, 7, 18]. It has been described as a "floppy stomach" rather than a true gastric volvulus [19]. It is asymptomatic and is often discovered incidentally. Although there were only two children in our series with this form of gastric volvulus, it is probably more common than our series suggests. Surgical intervention is not indicated, and neither infant in this series underwent surgery.

In contrast, chronic intermittent gastric volvulus is seen largely in patients with associated neurologic disorders and mental retardation. The occurrence of OA volvulus in this patient population might be related to the chronic gastric distention often seen in these patients [20] and attributed to aerophagia [5]. Chronic gastric distention has been proposed as a predisposing factor to the development of OA volvulus [17]. These patients might be symptomatic and surgery indicated depending on symptom severity. Surgical gastropexy has been reported to be of benefit in patients with persistent and severe symptoms [21]. Nonemergent surgical gastropexy was performed in three of the four patients with chronic intermittent volvulus in this series for relief of recurrent symptoms. Of these patients, none had recurrent gastric volvulus, although two experienced recurrent episodes of constipation and ileus, one requiring subsequent gastrostomy tube placement for presumed superior mesenteric artery syndrome. Management of this group of patients remains controversial [19, 21].

Review of abdominal radiographs in the patients with acute fulminant volvulus revealed common findings highly suggestive of the diagnosis. Marked spherical gastric distention with paucity of distal bowel gas indicating gastric outlet obstruction were present in all but one patient, in which the stomach had been decompressed by placement of a nasogastric tube. Other radiographic findings frequently present on abdominal radiographs included elevation of the diaphragm and two air-fluid levels in the stomach. In some patients, the configuration of the volvulus was suggested on the abdominal radiograph; projection of the pylorus over the gastric fundus and an unusual course of the nasogastric tube suggested MA configuration, and inversion of the greater curvature relative to the lesser curvature suggested OA configuration. In one child, findings on abdominal radiographs (Fig. 6) consistent with MT volvulus including marked spherical gastric distention, overlap of the pylorus on the gastric fundus, paucity of distal bowel gas, and inversion of the greater curvature relative to the lesser curvature were not appreciated. The diagnosis of gastric volvulus was made by CT. Delay in diagnosis led to left gastric artery thrombosis, gastric necrosis and perforation. At surgery, evidence of MT configuration was seen. The child underwent total gastrectomy.

Although findings on abdominal radiographs can be highly suggestive of the diagnosis of gastric volvulus, UGI series remains the diagnostic study of choice. In this series, UGI demonstrated the type of volvulus in all patients in whom it was performed and confirmed the radiographic findings of gastric outlet obstruction when present. CT, which was performed in two children with acute fulminant volvulus, is not indicated in the initial diagnosis of gastric volvulus and can delay emergent surgery.

The limitations of this study include its retrospective nature and small sample size. The natural rate of occurrence of this entity in childhood makes a randomized prospective study from a single center difficult. Although the sample

size was small, it compares favorably with prior studies that have focused primarily on selected case reports. A further limitation is that three of the children while diagnosed on UGI series did not undergo surgery for definitive diagnosis.

Conclusion

The spectrum of gastric volvulus ranges from the acute fulminant form to the chronic intermittent form, to the neonatal form. The first has potentially devastating consequences. In patients with known predisposing conditions, abdominal radiographs demonstrating a large spherical bubble in the upper abdomen with paucity of distal bowel gas in conjunction with left hemidiaphragmatic elevation should alert the physician to the possibility of the acute fulminant form and expedite emergent surgical intervention. Other helpful findings are an unusual nasogastric tube course, two air-fluid levels, overlap of the pylorus with the gastric fundus, and inversion of the greater curvature of the stomach.

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