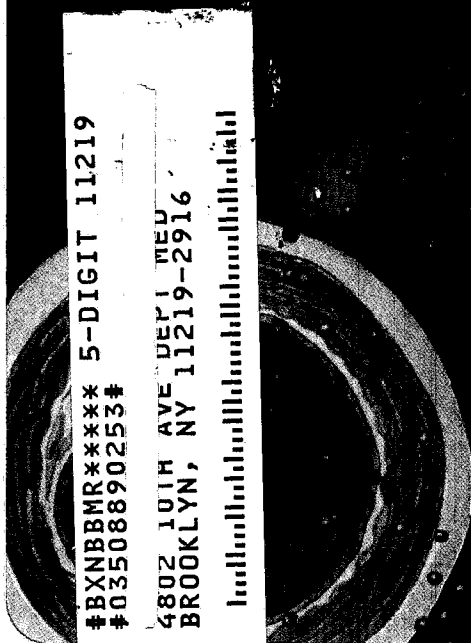


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**Case Report and Commentary**  
**from the University of Pennsylvania**



**Guest Editor: H. Ralph Schumacher, Jr., M.D.**  
Professor of Medicine, University of Pennsylvania School of Medicine

# Primary Aortoenteric Fistula

**This rare condition is one of the most difficult diagnostic challenges for the clinician confronted with gastrointestinal bleeding. If endoscopy shows only minor abnormal findings that do not explain the magnitude of the bleed, this condition must be considered.**

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hospital. She was resuscitated with packed red blood cells and crystalloid solution. Esophagogastroduodenoscopy was deferred due to an acute myocardial infarction. She was placed on H<sub>2</sub> blockers for a suspected bleeding peptic ulcer and discharged after observation and treatment of her myocardial infarction.

Three weeks later, the patient was admitted to another hospital for recurrent gastrointestinal bleeding. Diffuse gastritis was disclosed on esophagogastroduodenoscopy. A third minor bleeding episode occurred five days later during the same admission. Repeat endoscopy demonstrated diffuse gastritis and a lesser curvature ulcer. The electrocardiogram showed changes consistent with acute subendocardial myocardial infarction. The patient was maintained on antiulcer medication and released following treatment of her cardiac condition.

She remained stable for two weeks at which time rebleeding necessitated the present hospitalization. On admission to the family practice service, the patient was hemodynamically stable with a hematocrit of 26. She was given 3 U of packed red blood cells over the first two days of hospitalization. The patient underwent esophagogastroduodenoscopy on her third hospital day, which demonstrated

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► Primary aortoenteric fistula is a highly lethal condition. A 1984 review noted that 189 cases of primary aortoenteric fistulization were reported in the world's literature,<sup>1</sup> and several additional cases have since been reported.<sup>2-6</sup> These reports describe an entity that usually escapes timely detection despite a number of common presenting signs and symptoms. The case report that follows exemplifies many aspects of aortoenteric fistula and will hopefully aid in the future diagnosis and treatment of this grave condition.

## Case Report

A 75-year-old woman presented to the emergency department with a chief complaint of vomiting blood and having several bloody bowel movements. Her past medical history was significant for alcohol abuse, hypertension, two cerebrovascular accidents, peptic ulcer disease, and recent myocardial infarctions. Two months prior to this admission, the patient had experienced an upper gastrointestinal bleeding episode associated with hypotension (80/40 mm Hg) and was admitted to the

multiple, small, nonbleeding, mucosal lesions suspicious for angiodysplasia.

On the fourth hospital day, the patient was observed to have bright red blood draining from her nasogastric tube, as well as copious melanotic stool. An emergency surgical consult was requested. The patient was hemodynamically unstable, with a loss of consciousness and a drop in blood pressure to 70/0 mm Hg. Physical exam revealed an unconscious patient with palpable but thready pulses and no other remarkable findings. She was resuscitated with rapid infusion of 2 L of Ringer's lactate solution and 2 U packed red blood cells. Her blood pressure rose to 100/50 mm Hg, and she regained consciousness. Angiography was considered, however, the patient was deemed too unstable to tolerate angiogram and was prepared for emergency laparotomy. Despite maximal resuscitative efforts, the patient's blood pressure continued to drop, and she went into electromechanical dissociation and died prior to laparotomy.

Autopsy examination disclosed a fistula between the distal third portion of the duodenum and the abdominal aorta 5 cm inferior to the renal arterial openings (Figure 1). The duodenal mucosa was marked by a 2-mm round hole with leaking blood evident (Figure 2). A probe could easily be passed from the aorta to the duodenum. The aorta was severely atherosclerotic with fusiform aneurysmal formation, recent and old thrombi, and focal dissection of the aneurysmal wall. Microscopic examination of the aorta revealed ex-

tensive atherosclerosis with recent and old thrombi, focal dissection, and hemorrhage in the adjacent connective tissue toward the duodenal rent. The internal and external duodenal muscle layers were markedly thin or absent. The mucosa and submucosa showed chronic inflammatory changes, histiocytic reaction, degeneration, fibroblastic reaction, and focal perforation.

Other autopsy findings included severe coronary artery atherosclerosis, old posterior and posterolateral myocardial infarctions, and multiple superficial gastric antrum erosions.

### Discussion

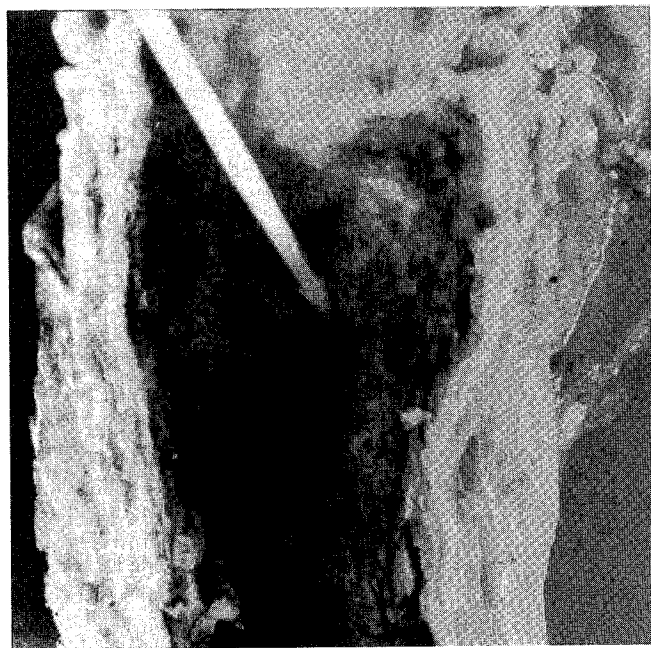
Successful management of primary aortoenteric fistula requires timely diagnosis and rapid treatment. As opposed to secondary aortoenteric fistulas, which by definition form following a prior abdominal operation (usually an aortic aneurysm repair), primary aortoenteric fistula arises de novo.

• *Presentation*—The presentation of primary aortoenteric fistula has many typical aspects, although the "classic" triad of abdominal pain, gastrointestinal hemorrhage, and a pulsatile abdominal mass is quite rare. In Sweeney's<sup>1</sup> 1983 review of 118 patients with primary aortoduodenal fistula, 32% of patients complained of abdominal pain and less than 25% had a palpable abdominal mass. Bleeding (hematemesis +/- melena) was noted in 64% as an initial symptom. Sweeney found shock to be a rare (< 5%) initial symptom and noted that in cases in which bleeding was the presenting symptom, over 70% of patients survived

at least 6 hours from the time of hemorrhage until death or operative intervention. Likewise, Steffes<sup>7</sup> reported that 50% of patients lived more than 24 hours between herald bleed and death, while 29% survived over one week. Other reported signs of primary aortoduodenal fistula worth noting are gastrointestinal bleeding with sudden hypotension and syncope, as well as concurrent massive upper and lower gastrointestinal bleeding.

• *Etiology*—The etiology of primary aortoduodenal fistula is unknown. One hypothesis postulates that aortic aneurysmal dilatation and pulsation causes inflammation and necrosis of adjacent bowel wall leading to fistulization.<sup>8</sup> Aortic dilatation, however, is not a requisite. At least seven cases of primary aortoduodenal fistula have been reported with no associated aneurysm.<sup>5-7</sup> The common pathologic feature in these cases was severe atherosclerotic disease. The microscopic findings in this case report correlate well with the above hypothesis since they reveal a long-standing pressure effect on the duodenal wall with atrophy of the muscular layers and perforation at the site of maximal pressure.

The pathophysiology of the herald bleed is most likely due to the aforementioned fistulization and bleeding, which produces profound hypotension. The subsequent drop in aortic pressure decreases aortic wall tension and allows the formation of a platelet plug and rudimentary fibrin deposition. With time, however, normalization of blood pressure and aortic pulsation invariably result in rebleeding.



1. Photo of opened abdominal aorta 5 cm below renal arteries. Arrow points to defect in aorta leading to duodenum



2. Opened duodenum (third portion), with fistulous tract evident.

• *Diagnosis*—The diagnosis of primary aortoduodenal fistula relies primarily on a high index of suspicion since the time span from presentation to death is relatively short and time spent on various tests is often wasted. Based on current practice, most stable patients with upper gastrointestinal bleeding will undergo upper endoscopy. While this test is only marginally helpful in the diagnosis of primary aortoduodenal fistula, it is indispensable in the identification of other far more common sources of gastrointestinal hemorrhage. The major caveat of esophagogastroduodenoscopy is to correlate the findings with the patient's signs, symptoms, and overall condition. Endoscopic reports often

note gastritis, duodenitis, and angiodysplasia. It is the responsibility of the attending physician to correlate these findings with the clinical situation. In this case report, multiple endoscopic procedures disclosed several abnormal findings, none of which, however, were likely to cause the massive bleeding that was seen. This type of error is widely apparent. Grigsby noted an esophagogastroduodenoscopy finding of a small nonbleeding varix of the gastroesophageal junction, which was treated with H<sub>2</sub> blockers.<sup>4</sup> The patient died of primary aortoduodenal fistula within one week. Similarly, Sweeney noted "antral gastritis,"<sup>1</sup> Pritchett noted "duodenal angiodysplasia,"<sup>2</sup> and both Per-

rott and Jaroch documented gastroesophageal tears.<sup>5,7</sup> Finally, it is essential that the third and fourth portions of the duodenum be well visualized during esophagogastroduodenoscopy because the majority of cases of primary aortoduodenal fistula occur at these sites.

The abdominal plain x-ray and upper gastrointestinal series are of little help in diagnosing primary aortoduodenal fistula, though a calcified aortic aneurysmal wall may hint at the diagnosis. Sonography will similarly alert the clinician to the presence of an aortic aneurysm, but it will not aid in the detection of fistulization.

Although arteriography would seem to be the most useful modality in diagnosing primary aortoduode-

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nal fistula, it is only positive during active bleeding, a situation in which the patient is usually too unstable to undergo such an invasive procedure. The bleeding scan suffers a similar drawback because it requires both active bleeding and prolonged time under a detector.

Computerized tomography (CT) has been used in the evaluation of secondary aortoenteric fistulas with some success.<sup>9-11</sup> Reports have documented intraluminal aortic gas, extravasation of contrast material, and generalized inflammation of the affected area, all leading to the diagnosis of aortoenteric fistulization. Overall, the CT scan with intravenous contrast appears to be the most useful current diagnostic modality. It can detect and assess an aortic aneurysm, as well as provide clues to enteric fistulization as mentioned above.

• **Treatment**—Primary aortoenteric

fistulas are uniformly fatal if not treated. The treatment of choice is exploratory laparotomy, replacement of the diseased aortic segment with a prosthetic graft, and primary repair of the enteric defect. Cultures should be obtained to assess the need for antibiotic coverage, although such coverage is usually not needed. This is in contrast to secondary aortoenteric fistulas, in which case antibiotic coverage and extra-anatomic bypass are routinely necessary due to a contaminated field.

### Summary

Primary aortoenteric fistulas are the cause of severe, usually fatal, gastrointestinal bleeding. They are commonly associated with aortic aneurysms, which may otherwise be asymptomatic, and they are extremely difficult to diagnose unless a high index of suspicion is maintained. Esophagogastrodu-

denoscopy with visualization of the third and fourth portions of the duodenum can be helpful, however, common findings such as angiodysplasia or gastritis should not be easily accepted as the cause of this type of massive bleed. The endoscopy must be correlated with the clinical picture. No patient should be discharged home on H<sub>2</sub> blockers with a diagnosis of upper gastrointestinal bleeding of unknown etiology until primary aortoenteric fistula is ruled out. Currently, the best diagnostic modality is CT scan with intravenous contrast. Primary aortoenteric fistula is a highly fatal condition, but death can be prevented if the condition is suspected early and treated aggressively.

### References

1. Sweeney MS, Gadacz TR: Primary aortoduodenal fistula: Manifestations, diagnosis, and treatment. *Surgery* 96:492-497, 1984.
2. Yano H, Jimi A, et al: Primary aortoduodenal fistula: Report of an autopsy case. *Gastroenterol Jpn* 22:218-221, 1987.
3. Grigsby WS, Eitzen EM, Boyle DJ: Aortoenteric

fistula: A catastrophe waiting to happen. *Ann Emerg Med* 15:731-734, 1986.

4. Jaroch MT, Diehl JT, Zippert AM: Primary aortoduodenal fistula without abdominal aortic aneurysm. *Cleve Clin Q* 552:579-581, 1985.

5. Gad A: Aortoduodenal fistula revisited. *Scand J Gastroenterol* 24:97-100, 1989.

6. Perrott C: Aortoenteric fistula without aortic dilatation: A case report. *J Emerg Med* 7:349-351, 1989.

7. Steffes BC, O'Leary JP: Primary aortoduodenal fistula: A case report and review of the literature. *Am Surg* 46:121-129, 1980.

8. Elliot JP, Smith RF, Szilagyi E: Aortoenteric and paraprostatic fistulas. *Arch Surg* 108:479-488, 1984.

9. Bower TC, Cherry KJ, Pairlero PC: Unusual manifestations of abdominal aortic aneurysms. *Surg Clin North Am* (Aug)69(4):750-751, 1989.

10. Ibrahim IM, Raccuia JS, Zafar A: Primary aortoenteric fistula-diagnosis by computerized tomography. *Arch Surg* 124:870-871, 1989.

11. Kokora JS, Rushton FW, Cranston PE: New computerized tomographic signs of aortoenteric fistula. *Arch Surg* 119:1073-1075, 1984.

## Drs. Carpenter and Sachs Comment

The rarity of aortoenteric fistula and the relative frequency of other conditions producing the same clinical manifestations conspire to make this one of the most difficult diagnostic and therapeutic challenges in clinical medicine and surgery: A successful outcome depends on recognition or strong suspicion of the condition and prompt surgical intervention, usually under circumstances of critical illness and hemodynamic instability.

Aortoenteric fistulas are of two types, primary and secondary. Primary fistulas, first described by Sir Astley Cooper in 1829,<sup>1</sup> are direct communications between the native aorta and the enteric tract. They may be caused by aortic disease (aneurysm, atherosclerosis, infection) or diseases of the bowel (ulcer, pseudocyst, infection, neoplasm). In an autopsy series of 10,000 consecutive cases, only 7 primary aortoenteric fistulas were detected.<sup>2</sup> Secondary aortoenteric fistulas between

the aorta or an aortic prosthesis and the bowel following aortic reconstructive surgery are more common. The first report of a secondary aortoenteric fistula appeared only one year after the first successful repair of an abdominal aortic aneurysm.<sup>3</sup>

While aortoenteric fistulas have been described in virtually every region of the gastrointestinal tract, the most common location is the duodenum. The intimate association of the duodenum and the abdominal aorta suggests an erosive mechanism for primary fistulization. Infection, too, may play a role, as evidenced by the prevalence of bacteria cultured from the abdomen during operations to repair these fistulas.<sup>4</sup> It is unclear, however, whether infection is the cause or the result.

Unfortunately, even the excellent diagnostic approach outlined by Moskovitz and colleagues in this report, which included gastrointestinal endoscopy, particularly esophagogastroduodenoscopy, CT scanning, and arteriography, produces a very low diagnostic yield. In patients with a known or suspected abdominal aortic aneurysm and gastrointestinal hemorrhage, prompt laparotomy indicated by a high index of suspicion is essential as both a diagnostic and therapeutic step.

The first successful repair of a primary aortoenteric fistula was described by Heberer in 1957.<sup>5</sup> It is necessary for the surgeon to obtain vascular control, eliminate the communication between the bowel and the aorta, and repair the enteric and vascular defects. The operation will

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need to be individualized, but several guiding surgical principles apply. In the most common situation, a fistula to the duodenum, the relative sterility of duodenal contents allows for standard aortic reconstruction in situ. In a review of 14 such primary fistulas, only one patient died from a disruption of a proximal aortic anastomosis, and this was due to infection with gram-negative bacteria.<sup>6</sup> The contamination associated with primary aortoduodenal fistula is usually limited and can be successfully eliminated by debridement of the region at the time of in situ graft placement. If,

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**Guest Editor**



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however, extensive contamination or sepsis should be encountered when managing a primary aortoenteric fistula, in situ replacement of the aorta would be contraindicated, and extra-anatomical reconstruction, such as axillobifemoral bypass, should be employed with vascularized coverage of the aortic closure. Surgical management of secondary aortoenteric fistulas nearly always requires extra-anatomical reconstruction.

**Summary**

It is hoped that, through heightened awareness of aortoenteric fistula, early operative interven-

tion may lead to a reduction in the high mortality associated with this condition. The key to suspecting the diagnosis is recognizing that the bleeding patient might have significant aortic disease. ◀

**References**

1. Cooper A: The lectures of Sir Astley Cooper on the principles and practice of surgery, p. 156. London: Westley, 1829.
2. Ferguson MJ, Ardon MS: Gastrointestinal hemorrhage, secondary to rupture of aorta: A review of four duodenal and three esophageal cases. *Arch Intern Med* 117:133, 1966.
3. Brock RC: Aortic Homografting: A report of six successful cases. *Guys Hospital Report* 102:204, 1953.
4. Kleinman LH, Towne JB, Bernhard VM: A diagnostic and therapeutic approach to aortoenteric fistula: Clinical experience with twenty patients. *Surgery* 86:868, 1979.
5. Heberer E: Diagnosis and treatment of an aneurysm of the abdominal aorta. *Germ Med Monthly* 2:203, 1957.
6. Daugherty M, Shearer GR, Ernst CB: Primary aortoduodenal fistula: Extra-anatomic vascular reconstruction not required for successful management. *Surgery* 86:399, 1979.

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