

# An Extremely Rare Case of Primary Aorto-enteric Fistula in a Filipino Patient

John Daniel A. Ramos, M.D.\*; Ma. Teresa Abola, M.D.\* and Patrick Louie Maglaya, M.D.\*\*

## Abstract

**Introduction:** An aorto-enteric fistula is a fistulous communication between the duodenum and the aorta. The non-traumatic form, or primary aorto-enteric fistula (PAEF), is rare and fatal if untreated. This is a case of PAEF in a Filipino patient who presented with upper gastrointestinal bleeding (UGIB).

**Clinical Presentation:** A 62-year-old Filipino sought consult for hematemesis and melena. He had just been discharged the previous day and sent home on empiric H. pylori eradication therapy after a week of workup, which included an unremarkable esophagogastroduodenoscopy (EGD). He claimed to be hypertensive but was not taking any maintenance anti-hypertensive medication.

**Physical Findings:** Blood pressure was 80/50 mmHg, and cardiac rate of 94 bpm. He had pale palpebral conjunctivae, and pale nailbeds. Abdominal exam was unremarkable. Rest of physical exam was normal. Stat hemoglobin was 63 g/dL.

**Results:** Exploratory laparotomy revealed the primary aortoduodenal fistula at the anterolateral aspect of the

fourth segment of the duodenum (PADF). Patient was started on metoprolol and atorvastatin. Axillary femoro-femoral bypass, ligation of aorta, wedge resection of aortoduodenal fistula, duodenorrhaphy, tube jejunostomy completed was done. Post-operative course was complicated by peritonitis and sepsis, and eventually went into arrest on his third week.

**Significance:** This is the first case of PAEF in our institution, and possibly in the country. It is an extremely rare condition that has an annual incidence of 0.007 per million. Since its description in 1843, only 250 cases have been reported in literature.

**Recommendations:** A high index of suspicion is key to its diagnosis and management. Massive UGIB, a negative endoscopy, and known aortic aneurysm should raise the suspicion for PAEF, as prompt surgical intervention is the only chance for survival among these patients.

**Key words:** primary aorto-enteric fistula, PAEF, UGIB

## Introduction

An aorto-enteric fistula (PAEF) is a fistulous communication between the duodenum and the aorta.<sup>1,2</sup> The primary or non-traumatic form is extremely rare and invariably fatal when left untreated.<sup>3</sup> We report a case of PAEF in a Filipino patient who presented with upper gastrointestinal bleeding (UGIB).

## Clinical Presentation

A 62-year-old Filipino male consulted at the emergency room for hematemesis, amounting to about 750 mL (three cups), and melena of about 500 mL (two cups). This was associated with dizziness and generalized weakness; there was no abdominal pain. He denied any other associated

symptoms. He had just been discharged the previous day and sent home on empiric H. pylori eradication therapy after a week of workup, which included an unremarkable esophagogastroduodenoscopy (EGD), for a prior episode of hematemesis. He claimed to be hypertensive but was not into any maintenance anti-hypertensive medication. He had no other comorbidities. He had no previous surgeries. He denied any chronic use of non-steroidal anti-inflammatory drugs. He was a 46 pack-year smoker. He denied alcohol intake or abuse. Family history was unremarkable.

On presentation, he was pale and weak-looking. He had a blood pressure of 80/50 mmHg, and a cardiac rate of 94 bpm. He had pale palpebral conjunctivae, as well as pale nailbeds. Abdominal exam was unremarkable. Rest of physical exam was normal. Stat hemoglobin was 63 g/dL. Intravenous isotonic fluid resuscitation was done, packed red blood cell units were transfused, and patient was started on intravenous drip of proton pump inhibitor. Blood pressure improved to 100/60 mmHg, and hemoglobin level to 92 g/dL.

\*Section of Cardiology, Department of Medicine, Philippine General Hospital

\*\* Department of Surgery, Philippine General Hospital

Corresponding Author: John Daniel A. Ramos, M.D., Philippine General Hospital, Manila, Philippines  
Email: johndanielramos.md@gmail.com

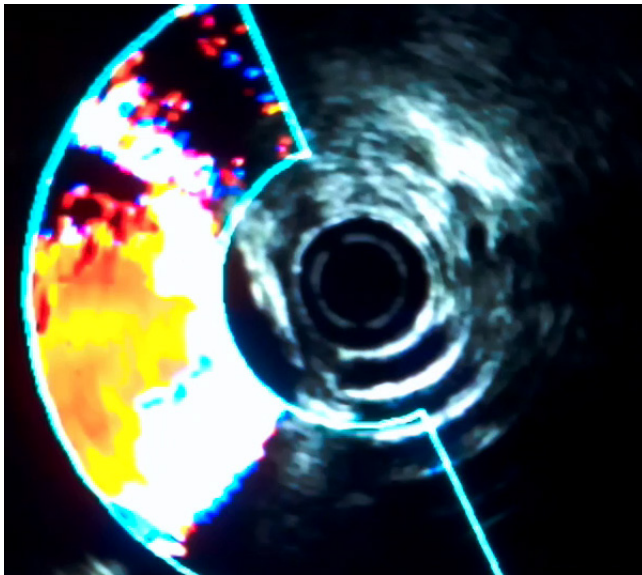


**Figure 1.** shows a 6x6 cm. hypoechoic lesion seen on endoscopy with color flow on Doppler Ultrasound, that was extrinsically compressing the area between the second and the third parts of the duodenum.

An abdominal computed tomography (CT) scan with contrast revealed an infra-renal abdominal aortic aneurysm, measuring 7.6 cm x 5.9 cm x 7.0 cm, with a 1.8 cm mural thrombus on its left aspect (Figure 2). The aneurysm was intimately related to the third segment of the duodenum with an outpouching that bored into the duodenal wall, but no frank contrast extravasation to the duodenum was seen.

On 3-Dimensional reconstruction of the aorta, this outpouching of the aorta was clearly demonstrated. Patient was started on metoprolol and atorvastatin.

Prompt surgical referral was done, and patient underwent exploratory laparotomy which revealed the primary aortoduodenal fistula at the anterolateral aspect of the fourth segment of the duodenum (Figure 3). Subsequent axillary femoro-femoral bypass, ligation of aorta, wedge resection of aortoduodenal fistula, duodenorrhaphy, tube



**Figure 2.** A. Abdominal CT Scan within intravenous contrast showing the axial view of the infra – renal abdominal aortic aneurysm, measuring 7.6 cm x 5.9 cm x 7.0 cm. Note the presence of a 1.8 cm mural thrombus (arrow) and an outpouching intimately related with the duodenum (circle)  
B. Three-dimensional reconstruction of the aneurysm showing the same outpouching (circle) compressing the area between the second and the third parts of the duodenum.

When patient was more stable, a repeat EGD with endoscopic ultrasound was done; this time, there was a note of a 6 x 6 hypoechoic lesion, with color flow on Doppler ultrasound (Figure 1), that was extrinsically compressing the area between the second and third parts of the duodenum. This lesion had a central umbilication, with note of blood actively oozing from it. An aortoduodenal fistula was considered even though the patient had no history of prior aortic disease or intraabdominal manipulation.

jejunostomy completed the surgery. The fistulous tract tissue was sent for histology and revealed an atheromatous aortic wall tissue that went deep into the duodenal wall. Patient initially had an unremarkable postoperative course during the first week; then had a stormy hospital course when he suffered a dehiscence of the duodenal repair site, contracted peritonitis, and eventually succumbed to severe sepsis from an intraabdominal infection and pneumonia on his 19<sup>th</sup> postoperative day.

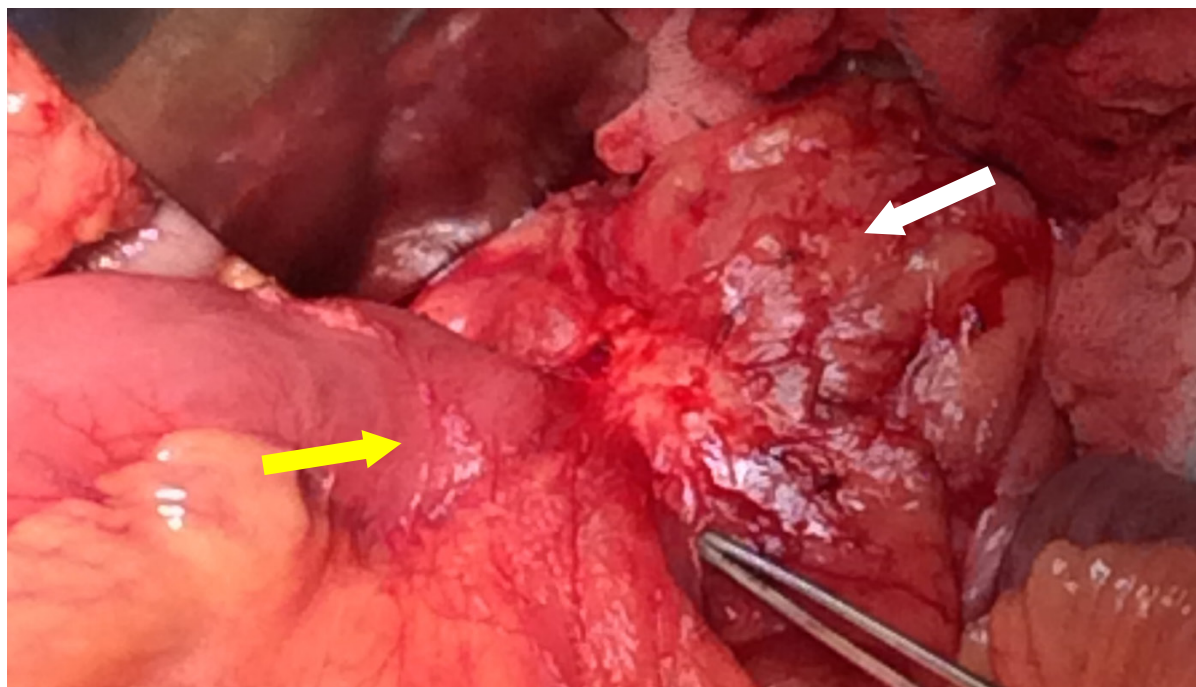


**Figure 3.** is an intraoperative image showing the primary aortoduodenal fistula (tip of forceps) connecting the aorta (white arrow) with the fourth part of the duodenum (yellow arrow)

## Discussion

In this paper, we report the first case of PAEF in our institution, and possibly in the country. Aorto-enteric fistulae are direct communications between the aorta and the gastrointestinal tract, and are of two types, primary and secondary.<sup>1</sup> Secondary aortoenteric fistulae are the more common type, with incidence of up to 2.3% to 4.0%, and are usually due to complications of abdominal aortic aneurysm repair or surgery.<sup>1,2</sup> An aorto-enteric fistula that occurs in the absence of trauma or abdominal aortic aneurysm surgery.<sup>1</sup> It is an extremely rare condition that has an annual incidence of 0.007 per million.<sup>3</sup> Since its description in 1843, only 250 cases have been reported in literature.<sup>1,2</sup> In autopsy studies, incidence is between 0.04% to 0.07%.<sup>1,2</sup>

The cause of PAEF is unclear but mechanical erosion and infection are thought to play a role in its pathogenesis.<sup>1</sup> PAEF's commonly arise from an abdominal aortic aneurysm of which 85% are atherosclerotic.<sup>4</sup> Repeated mechanical trauma between the pulsating aorta and a gastrointestinal segment causes progressive erosion of the former into the latter by ischemic and necrotic mechanisms.<sup>2</sup> The segments most commonly involved are the third and fourth parts of the duodenum (80%), because of their proximity to the aorta; followed by the jejunum and ileum (10%), large bowel (6.0%), and stomach (4%).<sup>5</sup> Another theory is that low-grade infection by bacteria and tuberculous organisms with abscess formation subsequently erodes through the

bowel wall.<sup>4</sup> Other causes implicated in fistula formation are peptic ulcer, gallstones, radiation, inflammatory bowel disease, and carcinoma; but these are much rarer.<sup>1,2,6</sup>

The most common clinical features of PAEFs are upper gastrointestinal bleeding (UGIB) (64%), abdominal pain (32%), and a pulsatile abdominal mass (25%), constituting a classic triad.<sup>1</sup> But diagnosis has usually been proven difficult and PAEFs are usually missed because this triad is found only in 23% of cases;<sup>6</sup> in another series, the triad was found in even fewer patients, at 11%.<sup>7</sup> For our patient, he presented only with UGIB, manifesting as hematemesis and melena. He did not complain of abdominal pain nor had a pulsatile abdominal mass on physical examination, and this may have contributed to a missed diagnosis in his prior work-ups. This difficulty in diagnosis has very important implications in terms of management and prognosis. PAEF is associated with a very high mortality when left untreated, approaching almost 100%.<sup>1-3</sup>

In 94% of cases, the clinical picture is that of a "herald bleed", defined as an episode of minor and brisk bleeding, usually overt UGIB, that stops spontaneously; then followed hours or days later by a massive catastrophic bleeding event.<sup>2,7</sup> This was consistent with our patient's presentation. This herald bleeding is the result of an initial small fistula transiently occluded by thrombus formation. If the fistula continues to expand or the occluding thrombus is removed, massive hemorrhage then follows.<sup>2</sup> The first hematemesis

could have been the patient's herald bleed. He was discharged after a week during his initial admission after a negative EGD and with apparent resolution of the bleeding. This was however followed by a bigger bleeding episode the next day, prompting re-admission. It has been observed that 70% of patients survive at least six hours after the initial bleeding episode and up to 50% survive 24 hours.<sup>2</sup> Thus, a herald bleed has been suggested as an opportunity for prompt intervention.<sup>2, 8</sup>

The diagnostic work-up is generally the same as that for UGIB. Endoscopy is used as first step in the work-up, and can help identify other causes of bleeding.<sup>2</sup> But a negative endoscopy does not rule PAEF out,<sup>1</sup> as what was demonstrated in this case. The acute angle between the third and fourth parts of the duodenum can be very difficult for visualization of a fistula distal to the third part.

The presence of an ulcer or erosion adjacent to a thrombus in the duodenum, with or without an extrinsic pulsatile mass, is highly suggestive of PAEF.<sup>1</sup> The second EGD, with endoscopic ultrasound, proved to be very crucial in the subsequent work-up, as it demonstrated the pulsatile aortic aneurysm extrinsically compressing the duodenum, and the presence of blood oozing from the former into the latter. Abdominal CT scan with intravenous contrast is the most superior diagnostic modality for PAEF, as it is less invasive, convenient, expedient, and poses no risk in dislodging the aortic thrombus.<sup>1,7,8</sup> CT scan can demonstrate air in the retroperitoneum and in the thrombus, loss of fat plane between aorta and duodenum, and, in very rare cases, extravasation of contrast from the aorta into the involved segment of bowel; all are pathognomic for fistula formation.<sup>9</sup> None of these were demonstrated in our patient's scan. CT can still miss PAEF, owing to low and variable sensitivity.<sup>1</sup> The sensitivity of CT has been estimated to be 50%.<sup>9</sup> Hence, a high index of clinical suspicion is still paramount. Gastrointestinal bleeding combined with a negative endoscopy in the presence of an aneurysmal aorta suggests PAEF and requires urgent evaluation by CT.<sup>7</sup>

Surgery is the only chance for survival of patients with PAEF.<sup>1,2,3</sup> Surgery has been found to improve survival,<sup>8</sup> and this was promptly done for the patient. Despite prompt management, complication rates are still high. Perioperative complications occur in 50%<sup>1</sup> of patients, with post-operative mortality rate greater than 30%.<sup>8</sup> Endovascular techniques have been explored and are generally viewed as temporary measures and as bridge therapy prior to definitive surgical management.<sup>1</sup> Our patient was able to tolerate surgery. Unfortunately, he developed complications thereafter that caused his demise.

## Conclusion

In summary, we reported a case of PAEF in a Filipino patient who presented with upper gastrointestinal bleeding (UGIB). To our knowledge, this is the first case of PAEF in our institution. Because of its rarity, a high index of suspicion is key to its diagnosis and management. Work-up is straightforward, but the diagnosis is still often initially missed. Massive UGIB, a negative endoscopy, and known aortic aneurysm should raise the suspicion for PAEF, as prompt surgical intervention is the only chance for survival among these patients. Postoperative complications, however, remain high.

## References

1. **Xiomeritis, Konstantinos, Ilias Dalainas, Michalis Stamakos, Konstantinos Filis.** Aortoenteric Fistulae: Present-Day Management. *Int Surg.* 96: 266-273, 2011.
2. **Cumpa, Edgard MD, Ross Stevens, MD, Kim Hodgson, MD, Fernando Castro, MD.** Primary Aortoenteric Fistula. *South Med J.* 95(9), 2002.
3. **Weerakkody, Yuranga and Avni Skandhan.** Aorto-enteric fistula. Downloaded: <http://radiopaedia.org/articles/aorto-entericfistula>
4. **MacDougall, Louise, John Painter, Terry Featherstone, Claus Overbeck, Shyju Paremal, and Suvadip Chatterjee.** Aorto-enteric fistulas: a cause of gastrointestinal bleeding not to be missed. *BJMP June.* 3(2): 317, 2010.
5. **Abernathy, W., and J.H. Sekijima.** Aortoenteric fistula. *New England Journal of Medicine.* 336: 27-28, 1997.
6. **R Khalid, I Shapey, D Andrew, G Miller.** A Unique Clinical Presentation of Primary Aortoenteric Fistula. *The Internet Journal of Surgery.* 9(1), 2006.
7. **Saers SJ and MR Scheltinga.** Primary aortoenteric fistula. *Br J Surg.* 92(2):143-52, 2005.
8. **Shehzad Khalid, Amjid Riaz, and John Meyrick-Thomas.** Primary aortoduodenal fistula – a rare clinical entity. *J R Soc Med Sh Rep.* 1:7, 2010.
9. **Raman SP, A Kamaya, M Federle, and EK Fishman.** Aortoenteric fistulas: spectrum of CT findings. *Abdom Imaging.* 38(2):367-75, 2013.