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A N Assar, C K Zarins

ABSTRACT
Ruptured abdominal aortic aneurysm (AAA) is one of the most fatal surgical emergencies, with an overall mortality rate of 90%. Most AAAs rupture into the retroperitoneal cavity, which results in the classical triad of pain, hypotension, and a pulsatile mass. However, this triad is seen in only 25–50% of patients, and many patients with ruptured AAA are misdiagnosed. It is likely that different sites of rupture of AAA determine a variety of common and uncommon clinical presentations, the recognition of which can save many lives. This article reviews the different sites of rupture of infrarenal AAA and explores the evidence behind the various clinical presentations seen in patients with ruptured AAA.

“...an aneurysm of the abdominal aorta is very often diagnosed when not present, and when present the symptoms may be so obscure that the nature of the trouble is overlooked.”

These words of Sir William Osler still resonate today despite remarkable advances in medicine since Osler’s time.

Abdominal aortic aneurysm (AAA) is a common and life threatening disease that affects 5–9% of the population over the age of 65 years. AAA is more common in male smokers with a positive family history of aortic aneurysms. Most patients with AAA are asymptomatic unless they develop a complication. Rupture, the most common complication of AAA, is one of the most fatal surgical emergencies; it has an overall mortality rate of approximately 90%. Most clinicians know that patients with ruptured AAA present with a classical triad of back pain with or without abdominal pain, hypotension, and a pulsatile abdominal mass. However, this triad is present in only 25–50% of patients, and many patients present with symptoms and signs that suggest a different diagnosis. This is why misdiagnosis of ruptured AAA occurs in 50% of patients. It is likely that different sites of rupture of AAA determine a variety of common and uncommon clinical presentations seen in patients with ruptured AAA.

The purpose of this article is to review the different sites of rupture of infrarenal AAA and to explore the evidence behind the various clinical presentations seen in patients with ruptured AAA.

SITES AND CLINICAL PRESENTATIONS OF RUPTURED AAA
Infrarenal AAA ruptures posteriorly into the retroperitoneal cavity in approximately 80% of patients and anteriorly into the peritoneal cavity in approximately 20% of patients. Rarely, AAA may rupture into the abdominal veins or the bowel. This may or may not be associated with retroperitoneal rupture (box 1).

Anterior intraperitoneal rupture
Here, a tear in the anterior wall of the aneurysm results in sudden severe abdominal or back pain and collapse. The resultant bleeding into the peritoneal cavity is so rapid that exsanguination and death usually occur before the patient reaches the hospital. Infraperitoneal rupture is probably an underestimated cause of sudden death.

Posterior retroperitoneal rupture

Classical clinical picture
Rupture into the retroperitoneal cavity is the most common site of ruptured AAA (fig 1). A tear in the posterolateral aneurysm wall leads to retroperitoneal bleeding which manifests clinically as back pain with or without abdominal pain and hypotension. This tear is often sealed for a few hours, which allows time for the transfer of the patient to the hospital, diagnosis, and treatment. On examination, a pulsatile epigastric mass is often palpable, particularly in a thin patient. However, this mass may not be palpable in obese or distended patients or in those with severe hypovolaemia.

Unusual clinical picture
The temporary sealing of the tear in the aneurysm wall may rarely extend beyond a few hours. This results in a variety of misleading symptoms and signs because of the extension of the retroperitoneal haematoma and its compressive effects. There are reports in the literature of patients with ruptured AAA who presented hours and in some cases days after an initial attack of abdominal or back pain that was ignored (box 2). Note that in ruptured AAA the duration of symptoms may be unusually long, and that an abdominal pulsatile mass may be absent. Despite that, one should not rule out a ruptured aneurysm.

The three most frequent emergency presentations of ruptured AAA and their immediate management are:

- a patient known to have an AAA presents with a sudden onset of abdominal or back pain and hypotension
- a patient presents with the classical triad of pain, hypotension, and a pulsatile mass.

In the two previous scenarios, a haemodynamically unstable patient should be immediately transferred to the operating theatre for emergency open repair. Recently, endovascular aneurysm repair (EVAR) has been successfully used to treat
ruptured AAA. In hospitals with capabilities for EVAR, a haemodynamically stable patient can undergo a preoperative computed tomography (CT) scan, and if the anatomy of the aneurysm is suitable, the patient can undergo endovascular repair for his/her ruptured aneurysm.

A patient is suspected of having a ruptured AAA, regardless of the symptoms and signs, and is haemodynamically stable. This patient should undergo a CT scan to confirm the diagnosis and assess his/her suitability for EVAR.

**Chronic contained rupture of AAA**

Although most patients with ruptured AAA have an acute presentation, some patients may escape detection for weeks or months after the aneurysm ruptures. This usually occurs when a retroperitoneal rupture leads to slow progressive bleeding which forms a large haematoma that is contained by the resistance of the periaortic tissues. Approximately 4% of ruptured AAA are contained ruptures (fig 2). They are also known as “sealed” or “spontaneously healed” aneurysms.

Since its first description by Szilagyi et al, a contained ruptured AAA has been frequently reported. Most patients with a contained rupture are haemodynamically stable with no manifestations of acute blood loss. However, a fundamental point needs to be remembered: a contained rupture is a **ruptured aneurysm in a stable patient** that may progress to free rupture at any time. Thus, urgent surgical treatment within 24 h, preferably after admission to an intensive care unit, is necessary. However, patients with more complex aneurysms may benefit from a full work-up, objective assessment, and planning. This includes assessing the patient’s suitability for EVAR which may delay surgery by more than 24 h, particularly when a specialised vascular team—for example, vascular anaesthetist, operating theatre staff—is needed.

Most patients with a contained rupture present with chronic back pain that may radiate to the groin. A long list of other reported presentations includes lumbar vertebral erosion, lumbar spondylitis-like symptoms, left lower limb weakness or neuropathy, left psas muscle haematoma, and obstructive jaundice. These presentations are puzzling and do not initially raise any suspicion for a ruptured aneurysm. To address this issue, Jones et al suggested some clinical and radiological criteria that may help with the diagnosis of contained ruptured AAA. These criteria include: (1) known AAA; (2) previous symptoms of pain that may have resolved; (3) stable condition and normal haematocrit; and (4) CT scan showing a retroperitoneal haematoma. Because most of these patients have chronic back pain, a magnetic resonance imaging scan may be the initial imaging test that they undergo. Thus, in a patient with a suspected contained rupture undergoing this type of scan, proper scrutiny of the aorta and the paraaortic tissues is essential to avoid missing the diagnosis.

**Rupture into the abdominal veins**

Rarely, AAA ruptures into the inferior vena cava or the left renal vein. This results in an aortocaval fistula or an aorta–left renal vein fistula, respectively.

**Aortocaval fistula**

A spontaneous aortocaval fistula most commonly occurs when an AAA erodes (ruptures) into the inferior vena cava (fig 3). Approximately 3–4% of patients with ruptured AAA have an aortocaval fistula. In these patients, the manifestations of rupture usually dominate the clinical picture and significantly diminish the chance of preoperative diagnosis. In fact, aortocaval fistulae are probably missed in 50% of patients and are discovered accidentally during elective repair of AAA. Trauma and surgery of the lumbar spine are other known causes of aortocaval fistulae.

The manifestations of an aortocaval fistula are variable because they depend on the size of the communication between the aorta and the inferior vena cava. Thus, temporary or permanent closure of this communication by an aortic mural thrombus or by a compressing aneurysm will change the clinical picture. Clinically, a patient with an aortocaval fistula presents with a classical triad of abdominal or back pain, a pulsatile...
abdominal mass, and a continuous bruit on abdominal auscultation.45 46 This triad is reported in 50–90% of patients in one series.46 However, it was seen in only 17% of patients in another series.53 Patients with an aortocaval fistula may also present with manifestations of high output heart failure such as dyspnoea, tachycardia, wide pulse pressure, cyanosis, and lower limb oedema.53 54 47 Additional symptoms and signs include angina, palpitations, hypotension, fever, oliguria, haematuria, pulsatile peripheral veins, and diminished lower limb pulses.35 32 44 45

The discovery of an aortocaval fistula during surgery is associated with major blood loss and the possibility of pulmonary embolisation with thrombus material from the aneurysm sac.50 Thus, unless the patient presents in extremis due to the rupture, every effort should be made to detect the fistula preoperatively.45 44 Abdominal CT scan is the definitive imaging test for the evaluation of AAA, and all patients suspected of having an aortocaval fistula should undergo a contrast CT whenever possible. Characteristic CT findings are loss of the fat plane between the aorta and inferior vena cava, vena caval effacement, and direct inflow of contrast from the aneurysm sac.53 54 Preoperative diagnosis can avoid unnecessary blood loss when the aneurysm is opened during surgery. Intraoperatively, a palpable thrill over the aneurysm, or an absent left renal vein which is usually felt in the left flank and radiates to the groin, mimicking ureteric colic.55 A pulsatile abdominal mass and a left sided continuous bruit are detected in approximately 60% and 70% of patients, respectively.53 Renal dysfunction is usually seen in 85% of patients.54 Barrier et al55 recently reported a male patient with an aorta–left renal vein fistula who presented with a large left sided varicocele, presumably due to left renal vein hypertension and impaired venous return from the left testicle. High output heart failure, similar to that seen in an aortocaval fistula, can also be seen in patients with an aorta–left renal vein fistula. The degree of heart failure depends mostly on the size of the fistula.51 Although extremely rare, an aorta–left renal vein fistula should be ruled out if a patient with an AAA develops haematuria, left loin pain, or manifestations of renal dysfunction.53 A contrast CT scan of the aorta can visualise the fistula, which should be looked for if a retroaortic left renal vein is seen.57 Preoperative diagnosis can avoid unnecessary blood loss when the aneurysm is opened during surgery. Intraoperatively, a palpable thrill over the aneurysm, or an absent left renal vein anterior to the aorta, should raise suspicion of an aorta–left renal vein fistula.56 An extremely rare cause of haematuria is an aortoureteric fistula. Here, the communication is usually between the ureter and a previously inserted aortic graft, but it can also occur with an aortoiliac aneurysm.58 The triad of unilateral hydronephrosis, intermittent haematuria, and previous aortic surgery should alert the clinician to this life threatening but potentially curable disease.55

Aorta–left renal vein fistula

Although Lord et al59 are credited with reporting the first case of aorta–left renal vein fistula complicating AAA, the first successful repair of this type of fistula was reported by DeBakey et al60 who repaired a traumatic fistula in a female patient. Aorta–left renal vein fistula is an extremely rare condition with fewer than 26 cases reported in the English literature up until 2007.53 It most commonly occurs when the wall of an infrarenal AAA erodes into the left renal vein. The left renal vein normally crosses in front of the abdominal aorta on its way to the inferior vena cava. In 1–2.4% of people, however, the vein crosses behind the aorta.53 A retroaortic left renal vein is involved in more than 90% of cases of aorta–left renal vein fistula.54 55

The “abdominal pain, haematuria, silent left kidney” syndrome described by Mansour et al44 summarises the clinical features of aorta–left renal vein fistula. Haematuria is the most important clinical feature in this condition,52 53 followed by pain which is usually felt in the left flank and radiates to the groin, mimicking ureteric colic.55 A pulsatile abdominal mass and a left sided continuous bruit are detected in approximately 60% and 70% of patients, respectively.53 Renal dysfunction is usually seen in 85% of patients.54 Barrier et al55 recently reported a male patient with an aorta–left renal vein fistula who presented with a large left sided varicocele, presumably due to left renal vein hypertension and impaired venous return from the left testicle. High output heart failure, similar to that seen in an aortocaval fistula, can also be seen in patients with an aorta–left renal vein fistula. The degree of heart failure depends mostly on the size of the fistula.51 Although extremely rare, an aorta–left renal vein fistula should be ruled out if a patient with an AAA develops haematuria, left loin pain, or manifestations of renal dysfunction.53 A contrast CT scan of the aorta can visualise the fistula, which should be looked for if a retroaortic left renal vein is seen.57 Preoperative diagnosis can avoid unnecessary blood loss when the aneurysm is opened during surgery. Intraoperatively, a palpable thrill over the aneurysm, or an absent left renal vein anterior to the aorta, should raise suspicion of an aorta–left renal vein fistula.56 An extremely rare cause of haematuria is an aortoureteric fistula. Here, the communication is usually between the ureter and a previously inserted aortic graft, but it can also occur with an aortoiliac aneurysm.58 The triad of unilateral hydronephrosis, intermittent haematuria, and previous aortic surgery should alert the clinician to this life threatening but potentially curable disease.55

Rupture into the bowel

An aortoenteric fistula is an abnormal communication between the abdominal aorta and the bowel; it may be primary or secondary. A primary aortoenteric fistula connects an infrarenal AAA to the bowel, most commonly the duodenum (aortoduodenal fistula). This condition is often fatal but fortunately rare, with an estimated incidence at autopsy of 0.04–0.07%.59 A secondary aortoenteric fistula is a late postoperative complication due to erosion of a prosthetic aortic graft into the duodenum. This condition is more common, occurring in 0.5–2.3% of patients after aortic surgery.55 The third and fourth parts of the duodenum are most commonly involved in an aortoenteric fistula because this duodenal segment is closely applied to the anterior wall of the aorta, being fixed posteriorly by the ligament...
of Treitz. However, communications with other parts of the gastrointestinal tract have also been reported.\(^{60} {61}\)

Patients with a primary aortoduodenal fistula commonly present with upper gastrointestinal haemorrhage (haematemesis, melaena, haematochezia).\(^{54}\) Abdominal pain and a pulsatile abdominal mass may also be present; however, patients rarely have all three findings.\(^{60} {61}\) In their review of reports published in English over a 15 year period, Dossa et al.\(^{60}\) found that gastrointestinal haemorrhage was the most common presentation in patients with primary aortoduodenal fistula, occurring in 96% of patients. Massive haemorrhage is uncommon initially; patients usually experience an episode of small brisk bleeding which stops spontaneously. This “herald bleed” is characteristic of an aortoduodenal fistula.\(^{60} {61}\)

Oesophago-gastroduodenoscopy (OGD) is likely to be the first diagnostic test done in patients with upper gastrointestinal haemorrhage, even if the cause of the bleeding is an aortoduodenal fistula.\(^{60}\) OGD is useful to rule out the much more common causes of bleeding such as gastroduodenal ulcers and oesophageal varices. If bleeding is due to an aortoduodenal fistula, OGD may not be precise in visualising the fistula because of failure to pass the endoscope into the third or fourth parts of the duodenum.\(^{60}\) Consequently, in patients with known or previously treated AAA presenting with unexplained upper gastrointestinal haemorrhage, CT has emerged as the most important initial diagnostic test to rule out an aortoduodenal fistula.\(^{54} {55}\) Highly suggestive findings on CT include loss of the fat plane between the aorta and duodenum and the presence of air in the retroperitoneal cavity.\(^{59}\) Unless a primary gastrointestinal source of bleeding has been unequivocally identified in a bleeding patient with AAA, an aortoduodenal fistula should always be ruled out. Currently, the preoperative diagnosis of aortoduodenal fistulae is reached in only 50% of patients.\(^{59}\)

RUPTURED AAA IN CHILDREN

Aortic aneurysms are extremely rare in children, and their aetiology is different from those in adults. In children, aortic wall infection, vasculitis, and connective tissue disorders are important causative factors for AAA.\(^{63} {64}\) Umbilical vein catheterisation is also a well recognised cause of childhood AAA, possibly through infection.\(^{65} {66}\) Most AAAs in children present as painless pulsatile masses; however, a few alarming cases of rupture have been reported.\(^{60} {61}\) Ruptured AAA is not often suspected in children; however, given its fatal nature, clinicians should immediately rule out ruptured AAA in children if it is suspected.

NEW THERAPEUTIC OPTIONS FOR RUPTURED AAA

The introduction of EVAR and the rapid advances in endovascular technology have changed the way vascular surgeons approach aortic aneurysm repair. EVAR has gained acceptance as an alternative to elective open repair of AAA in patients with suitable aneurysm anatomy. Two multicentre randomised controlled trials that compared open repair with EVAR for infrarenal AAA demonstrated a threefold reduction in perioperative mortality after EVAR.\(^{70} {71}\) Emergency endovascular repair for ruptured AAA is now technically feasible, and several case series have been reported in the literature over the past few years.\(^{72}\) Because the suitability for EVAR is assessed by a preoperative CT scan, there is concern that the haemodynamic state of patients with ruptured AAA would preclude a CT scan. However, two relatively recent studies suggest that most patients with ruptured AAA would be able to undergo a preoperative CT en route to the operating theatre.\(^{73} {74}\) The current lack of level 1 evidence to support endovascular repair for ruptured AAA is addressed by two ongoing multicentre randomised controlled trials comparing emergency open repair with endovascular repair for ruptured AAA. These are the French Ruptured Aorta-iliac Aneurysms: Endo vs Surgery (ECAR) Trial\(^{75}\) and the UK Immediate Management of the Patient with Aneurysm Rupture: Open Versus Endovascular Repair (IMPROVE) Trial.\(^{76}\) Although the results of these trials will not be available for a few years, they will answer an important question that is eagerly awaited by the vascular community.

A relatively new and promising endovascular technique used in patients with ruptured AAA is the aortic occlusion balloon. Using local anaesthesia, this compliant balloon is inserted through the femoral artery, and under fluoroscopy, is positioned in the supracoeliac aorta.\(^{77}\) In haemodynamically unstable patients, the balloon is immediately inflated inside the aorta.
to provide control of the circulation and prevent blood loss through the ruptured aneurysm. If the patient is stable, the balloon is not inflated, but is ready to be used should the patient become unstable during induction of general anaesthesia, or at any stage before clamping the aorta during open repair or deploying the stent graft during EVAR. The balloon is also incorporated into one of the commercially available stent grafts. The development of new endovascular techniques and the application of existing effective ones to patients with ruptured AAA may help reduce the mortality rate from this fatal emergency. It is likely that current and future research will be able to address a number of unanswered questions related to ruptured AAA (box 3).

SUMMARY

Rupture is the most common and most fatal complication of AAA. AAA ruptures posteriorly into the retroperitoneal cavity, anteriorly into the peritoneal cavity, and, rarely, into the abdominal veins and the bowel. These different rupture sites determine a variety of common and uncommon symptoms and signs seen in patients with ruptured AAA. The recognition of these various clinical presentations is likely to lead to an earlier diagnosis and a timely intervention, both of which can have a positive impact on the high mortality rate associated with ruptured AAA. In addition, endovascular techniques may improve the survival of patients with ruptured AAA in the years to come.

MULTIPLE CHOICE QUESTIONS (TRUE (T)/FALSE (F); ANSWERS AFTER THE REFERENCES)

1. Abdominal aortic aneurysm:
   A. Affects 1% of the population over 65 years
   B. Occurs equally in men and women
   C. Can be treated by endovascular techniques
   D. Is rarely symptomatic
   E. Is more common in smokers

2. Regarding ruptured abdominal aortic aneurysm:
   A. It is the most common complication of an abdominal aortic aneurysm
   B. It has an overall mortality of approximately 30%
   C. It is most commonly retroperitoneal
   D. It is characterised by a triad of pain, hypotension, and a pulsatile mass
   E. It can be easily diagnosed clinically in almost all cases

3. An abdominal aortic aneurysm can rupture:
   A. Into the inferior vena cava
   B. Into the left renal vein causing left loin pain and haematuria
   C. Into the bowel, partially the first part of the duodenum
   D. Intraperitoneally, resulting in a haematoma that can be contained for days or weeks
   E. Into the duodenum causing gastrointestinal bleeding

4. An aortoovascular fistula
   A. Most commonly occurs between an aortic prosthetic graft and the inferior vena cava
   B. Typically presents with pain, a pulsatile mass and a continuous bruit on auscultation
   C. Can result in high output heart failure
   D. Is best treated by antihypertensive medications
   E. Can cause major bleeding during aortic surgery

5. Regarding ruptured abdominal aortic aneurysm:
   A. All patients require an abdominal CT scan before surgery
   B. An abdominal CT scan should be requested for a stable patient with a suspected aneurysm
   C. An abdominal CT scan is mandatory for a patient with a known aneurysm, presenting with hypotension and a pulsatile mass and having open surgery
   D. An aortoovascular fistula is typically diagnosed on a non-contract abdominal CT scan
   E. Can never occur in children due to the absence of atherosclerosis

Competing interests: None.

REFERENCES


Answers

1. (A) F; (B) F; (C) T; (D) T; (E) F

2. (A) T; (B) F; (C) T; (D) T; (E) F

3. (A) T; (B) T; (C) F; (D) F; (E) T

4. (A) F; (B) T; (C) T; (D) F; (E) T

5. (A) F; (B) T; (C) F; (D) F; (E) F

6. (A) F; (B) T; (C) T; (D) T; (E) T

7. (A) T; (B) T; (C) T; (D) T; (E) T

8. (A) T; (B) F; (C) T; (D) F; (E) T

9. (A) T; (B) T; (C) T; (D) F; (E) T

10. (A) F; (B) T; (C) T; (D) F; (E) F

11. (A) T; (B) T; (C) T; (D) T; (E) T

12. (A) F; (B) T; (C) T; (D) T; (E) T

13. (A) T; (B) T; (C) T; (D) T; (E) T

14. (A) T; (B) T; (C) T; (D) T; (E) T

15. (A) T; (B) T; (C) T; (D) T; (E) T

16. (A) T; (B) T; (C) T; (D) T; (E) T

17. (A) T; (B) T; (C) T; (D) T; (E) T

18. (A) T; (B) T; (C) T; (D) T; (E) T

19. (A) T; (B) T; (C) T; (D) T; (E) T

20. (A) T; (B) T; (C) T; (D) T; (E) T

21. (A) T; (B) T; (C) T; (D) T; (E) T

22. (A) T; (B) T; (C) T; (D) T; (E) T

23. (A) T; (B) T; (C) T; (D) T; (E) T

24. (A) T; (B) T; (C) T; (D) T; (E) T

25. (A) T; (B) T; (C) T; (D) T; (E) T

26. (A) T; (B) T; (C) T; (D) T; (E) T

27. (A) T; (B) T; (C) T; (D) T; (E) T

28. (A) T; (B) T; (C) T; (D) T; (E) T

29. (A) T; (B) T; (C) T; (D) T; (E) T

30. (A) T; (B) T; (C) T; (D) T; (E) T

31. (A) T; (B) T; (C) T; (D) T; (E) T

32. (A) T; (B) T; (C) T; (D) T; (E) T

33. (A) T; (B) T; (C) T; (D) T; (E) T

34. (A) T; (B) T; (C) T; (D) T; (E) T

35. (A) T; (B) T; (C) T; (D) T; (E) T

36. (A) T; (B) T; (C) T; (D) T; (E) T

37. (A) T; (B) T; (C) T; (D) T; (E) T

38. (A) T; (B) T; (C) T; (D) T; (E) T

39. (A) T; (B) T; (C) T; (D) T; (E) T

40. (A) T; (B) T; (C) T; (D) T; (E) T

41. (A) T; (B) T; (C) T; (D) T; (E) T

42. (A) T; (B) T; (C) T; (D) T; (E) T


