Primary aorto enteric fistula : report of 18 Belgian cases and literature review

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Abstract

Background and study aims : **We searched for Belgian cases of primary aorto enteric fistula (PAEF). After reviewing the literature we compared our data concerning incidence, types, pathogenesis, aetiology, clinical presentation, diagnostic modalities, treatment and prognosis of PAEF. We especially focus on the clinical picture and diagnostic options.**

Patients and methods : **We present our atypical case report. A questionnaire was send to 196 Belgian vascular surgeons in order to evaluate retrospectively the Belgian experience with PAEF. A Medline search of relevant literature from January 1980 to February 2006 was conducted.**

Results : **In total 18 Belgian cases of PAEF were detected usually originating from infrarenal abdominal aorta (83%), ending in the third or fourth part of the duodenum (67%) and affecting men (94%) with a mean age of 70 years old. Main cause is aneurysm (89%). Gastrointestinal bleeding is the main symptom (83%). Untreated, no one survives and overall mortality is 29%. Most patients are treated with in situ grafts (83%). With our experience we propose a diagnostic flow chart to obtain early diagnosis of PAEF.**

Conclusions : **PAEF is suspected when a patient presents with (considerable) (upper) gastrointestinal blood loss and has a known aneurysm, initial herald bleed or pulsating abdominal mass. In case of hemodynamic instability, prompt surgical exploration is mandatory. Hemodynamically stable patients must undergo contrast enhanced multislice computerized tomography rather than gastroduodenoscopy or arteriography to make early diagnosis. Surgery is the only definitive life saving treatment. Overall mortality is at least 30%. Late diagnosis, positive peroperative cultures and shock are indicators of poor prognosis.** (Acta gastroenterol. belg., **2008**, 71, **250-258**).

Key words : primary aorto enteric fistula ; aneurysm ; gastro intestinal hemorrhage ; herald bleeding ; abdominal pulsating mass.

Introduction

Primary aorto enteric fistula is a direct communication between the intravascular lumen of the native aorta or the iliacal branches and that of the digestive tract. An aorto enteric fistula caused by previous vascular operation is called secondary. PAEF was first described by Sir Astley Cooper in 1829. It is a rare condition but 100% lethal due to exsanguination if untreated (1,2).

Most literature about PAEF is to be found in surgical papers. Still, the vast majority of these patients presents initially to a general practitioner or an internal medicine or emergency doctor. Therefore we focus especially on clinical presentation and diagnostic options, addressing the question : When to think of PAEF and how to prove it ? Early diagnosis is life saving.

We present our own very atypical case report of PAEF. We obtained informed consent signature of this patient. We also add 17 Belgian cases of PAEF to the very limited list of previously published cases in literature. Thirteen of these Belgian cases have never been published before. This unique overview of the Belgian situation was made by extraction of data collected by means of a questionnaire we send to 196 Belgian vascular surgeons. The anonymous questionnaire contained questions about the duration of experience of each physician as a vascular surgeon, the number of cases of PAEF the surgeons treated in this period and the publication of these cases, if applicable. When the surgeon had treated such a patient personally, further questioning detailed the type, cause and clinical presentation of the fistula together with the age and gender of each patient. We also asked which diagnostic tool proved the fistula and what treatment was given. Finally we collected data about complications and survival rate of each patient.

In order to compare our data to literature, we reviewed thoroughly the key aspects of PAEF, using a Medline literature search for (mostly review) articles between January 1980 and February 2006. In 1984 Sweeney and Gadacz published a review of 189 patients, a compilation of all published cases of PAEF so far (3). Voorhoeve *et al.* described 62 patients in the period 1984-1993, extending the list of PAEF population to 251 patients (4). Saers *et al.* added 81 patients with PAEF published in English literature between 1994 and 2003 (5). Voorhoeve *et al.* identified 27 additional patients, operated on by Dutch surgeons, which Saers *et al.* did not include in their review because data were incomplete (6). Finally, a total of 359 patients is reported in the English literature until 2003. An important French review of 253 PAEF patients was made by Roux *et al.* in 1993 (7).

In conclusion we propose a rational diagnostic flow chart.

Case report

A 75 year old man with a history of bilateral preperitoneal inguinal hernia correction and lumbal foramen stenosis treated with corticosteroids, was admitted to our

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hospital because of tenesms and 3 subsequent episodes of rectal bleeding with clots within 24 hours. He experienced continuous but not severe pain in the hypogastric region with flatulence and anorexia for already 3 days. There was no fever. Further systemic anamnesis was normal.

During clinical examination we found a temperature of 38,4 degrees Celsius, a regular pulse of 84 beats per minute and a blood pressure of 136/80 millimetres of mercury. Deep palpation revealed a slight tenderness of the left inguinal fossa accompanied by hyperperistalsis. No percussion pain, pulsating abdominal mass or vascular murmur was found. Pulsations were felt on both feet. Palpation per anum revealed a little strain of fresh red blood.

Electrocardiography showed a low voltage sinus rhythm of 105 beats per minute with signs of old infarction in the inferior leads.

In order to prove our tentative diagnosis of diverticulitis, a blood sample was taken and a contrast enhanced multislice computerized tomography (CT) was performed on a Philips Brilliance 16 Power CT after oral and retrograde contrast administration using an angio-CT protocol. Laboratory results showed leucocytosis ($15{,}9 \times 10^9$/litre) and an elevated C reactive protein of 19,4 milligram/decilitre). A normal haemoglobin level and haematocrite were noted. The most important finding on abdominal CT scan was an atherosclerotic saccular aneurysm of the right common iliac artery locally measuring more than 10 centimetres with a thrombus in its wall of 6,5 centimetres. The thrombus impressed the sigmoid colon with disruption of the aortic fat cover and contained multiple air bubbles in continuity with the bowel. This image was pathognomonic for a primary aorto enteric fistula from the right common iliac artery to the sigmoid colon (Fig. 1).

The patient was operated on immediately. Preoperative antibiotics were given and an axillo-bifemoral bypass graft was installed. This procedure was completely finished before performing xyphopubic laparotomy and transperitoneal approach to the infrarenal aorta, which was transsected. Afterwords the aortic stump was covered with peritoneal tissue. The entire aortoiliac strain was then removed up to the external iliac arteries. When dissecting the right internal iliac artery aneurysm, stool was found in the thrombus and a perforation of 2 centimetres diameter into the sigmoid colon was seen (Fig. 1). A Hartmann procedure was performed by the intestinal surgeons. The pathology report showed sigmoid diverticular abscedation with probably secondary perforation towards the iliac aneurysm.

In the immediate postoperative period the patient developed acute ischemia of the left leg due to thrombosis of the left femoral part of the axillobifemoral graft, for which thrombectomia and fasciotomia was performed. This event was followed by rhabdomyolysis, transient renal insufficiency acquiring haemodialysis, temporal paresis of the left leg, pneumonia, paroxysmal atrial fibrillation and Pseudomonas wound infection. Currently, 24 months after surgery, the patient is suffering from ischemic neuropathy of the left leg but generally doing very well .

Other belgian cases – results

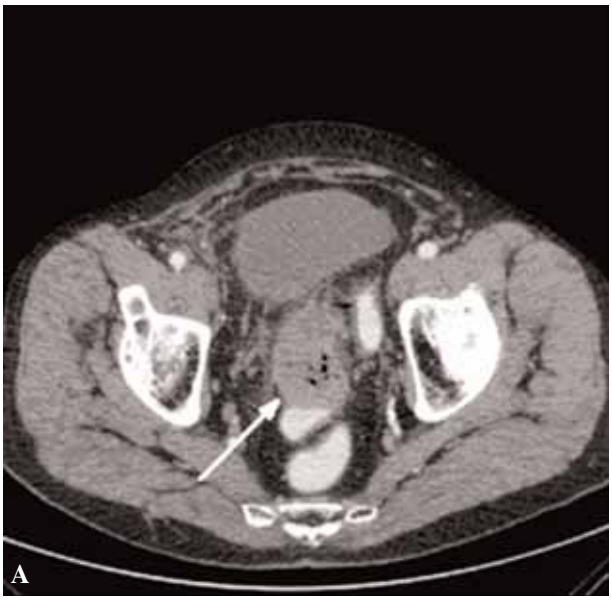
To evaluate the Belgian situation according to PAEF, we have send a questionnaire to a total of 196 Belgian vascular surgeons. 37% (n = 73/196) of the surgeons returned the anonymous form. In total these answers cover more than 1025 years of experience of individual vascular surgeons in Belgium between 1970 and 2006. Including our case, we collected 18 cases of PAEF of which 13 have never been published before (8,9,10,11) (Table 1 & 2). This means the minimal incidence of PAEF in Belgium is one case per 57 years of experience as a vascular surgeon, indicating the rarity of this pathology. 94% (n = 15/16) of the cases are men and the mean age is 70 years (range 50 to 90 years).

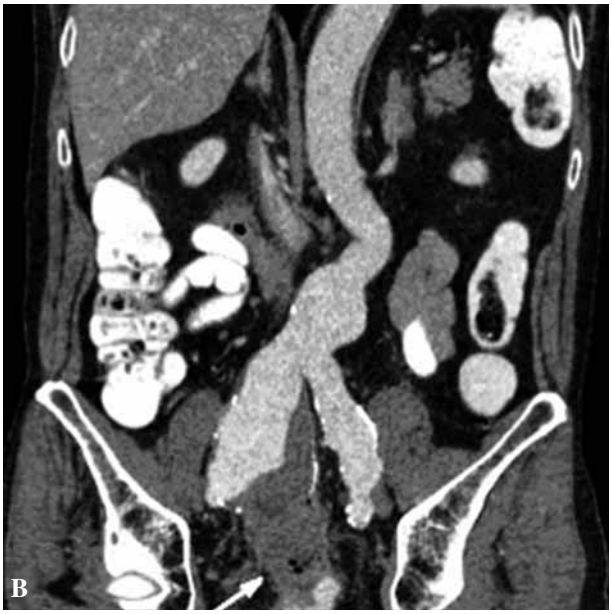
The vast majority, i.e. 83% (n = 15/18), of the fistulae originates from the abdominal aorta. The fistula ends in the third or fourth part of the duodenum in 67% (n = 12/18). Other origins mentioned are thoracic aorta (5,5%, n = 1/18), common iliac artery (5,5%, n = 1/18) and internal iliac artery (5,5%, n = 1/18). Sigmoid colon (11%, n = 2/18), stomach (5,5%, n = 1/18), oesophagus (5,5%, n = 1/18), duodenum first and second part (5,5%, n = 1/18) and ileum (5,5%, n = 1/18) are other fistula end points.

Aneurysm is the cause in 89% (n = 16/18) of the cases of which 33% (n = 6/18) is infectious, proven by positive peroperative cultures. Escherichia Coli, beta lactamase negative Enterococcus, Streptococcus viridans and Staphylococcus species (sp) were mentioned as pathogenic organisms. Other causes mentioned are duodenal and sigmoid ulceration (13%, n = 2/16) and inflammatory disease (6%, n = 1/16) not further specified.

The leading symptom at presentation is gastrointestinal bleeding, found in 83% (n = 15/18) ; haematemesis is seen in 61% (n = 11/18), melaena in 33% (n = 6/18) and haematochezia in 5,5% (n = 1/18) The classical triad of abdominal or back pain, pulsating mass and gastrointestinal blood loss is found in only 5,5% (n = 1/18). Shock is present in 72% (n = 13/18) and 17% (n = 3/18) is diagnosed with fever or sepsis. Other symptoms are syncope (39%, n = 7/18), pulsating mass (33%, n = 6/18) and abdominal or back pain (22%, n = 4/18).

CT scan is used in 33% (n = 6/18) of which 83% (n = 5/6) is in shock, gastroduodenoscopy in 33% (n = 6/18) of which 67% (n = 4/6) is in shock and arteriography in 11% (n = 2/18) of which 50% (n = 1/2) is in shock. Immediate surgical exploration is performed in 50% of the cases (n = 9/18) of which 67% (n = 6/9) is in shock. Of all patients presenting with signs of shock, 46% (n = 6/13) are surgically explored immediately, 15% undergo only contrast enhanced CT (n = 2/13), 8% (n = 1/13)





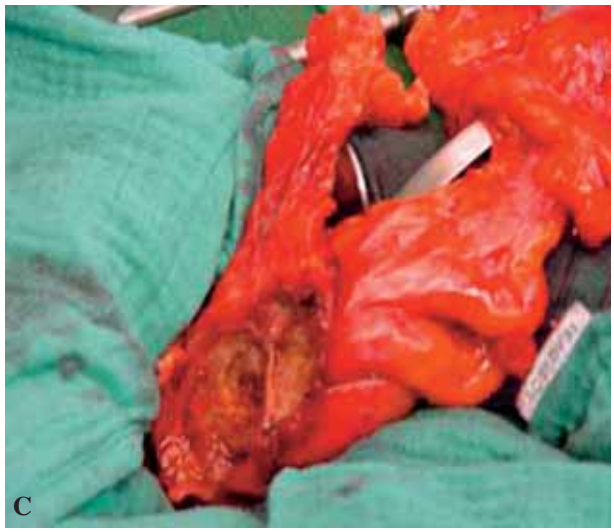


Fig. 1. — Pathognomonic image of our case of primary aorto enteric fistulisation from common iliac artery to sigmoid colon. Fig. 1. Panel A (arrow) shows CT axial reconstruction of thrombus in close proximity with the sigmoid with disruption of the fat covers and air-bubbles in the thrombus wall. Panel B (arrow) shows CT coronal reconstruction of the saccular aneurysm of the right common iliac artery with rupture of the atherosclerotic wall and a large thrombus with air-bubbles compressing the sigmoid. Panel C shows the resected sigmoid specimen with the ulcerated crater presenting the fistula outflow into the digestive tract.

have only gastroduodenoscopy, 23% have combined contrast enhanced CT and gastroduodenoscopy (n = 3/13) and 8% (n = 1/13) have gastroduodenoscopy combined with arteriography. Which diagnostic tool confirmed the diagnose of PAEF when a combination of diagnostic modalities was used, could not be extracted out of our data with certainty. Therefore we can not calculate detection ratios of the different diagnostic tools used in the Belgian series.

The mean treatment option is in situ grafting in 83% (n = 15/18). Extra anatomical bypass grafting is performed in 11% (n = 2/18) of which 100% proved to have positive peroperative cultures afterwords.

One patient, a 90 year old female, died preoperative due to palliative approach. Postoperative mortality within 30 days is 29% (n = 5/17). If peroperative cultures proved to be positive, 33% (n = 2/6) of the patients died within the first 30 days. When the peroperative cultures were negative, postoperative mortality within 30 days is 20% (n = 2/10). In presence of shock mortality within 30 days postoperative was 33% (n = 4/12). Mortality within 30 days after in situ grafting is 27% (n = 4/15) and 50% (n = 1/2) after extra anatomical bypass grafting.

One case of fistulisation from the pulmonary artery to the oesophagus due to ulceration of a lung tumour and one case of fistulisation from a lienal artery aneurysm to the stomach were also mentioned. Both cases presented with haematemesis and shock. As these cases did not match to our definition of PAEF, they were not included in the analysis. The 18 Belgian cases will be further referred to as the 'Belgian cohort'.

Discussion

Incidence

PAEF is known to be a very rare pathology as we indicated in the Belgian cohort. A Medline search up to February 2006 reveals less than 500 case reports worldwide, published so far. In the Belgian cohort 1 case per 57 years of experience as a vascular surgeon was found. Large autopsy studies point out an incidence of 0,04% to 0,07% (2,4,5,7,12) (Table 2). Tareen *et al.* found an incidence of 0.1 to 0.8% (13). As Voorhoeve *et al.* already indicated, this means that the published case reports only

Table 1. — **Eighteen Belgian cases of primary aorto enteric fistulisation**

Case	Age (years)	Sex (M/F)	symptoms	Diagnostic tools	Cause	Origin fistula	End fistula	Treatment	Cultures	Outcome Within 30 days
1	75	M	H,P,Fe	CT	A,U	CIA	Sigm	EA	E.Coli	Alive
2	72	M	Me,S	Ex	I	IIA	Ileum	IS	-	Alive
3	?	?	He,P,S	Ex	A	AA	III-IV	IS	-	Alive
4	?	M	He,S,Sy	Ex	A	AA	III-IV	IS	-	Alive
5	?	M	Pu,S,Fe,Sy	Ex	U	AA	Sigm	EA	+	Dead
6[A]	71	M	Pu	Ex	A	AA	III-IV	IS	-	Alive
7[A]	54	M	He,Fe,Sy	Ex	A	AA	III-IV	IS	BLNE	Alive
8	?	M	S,Me	Ex	A	AA	I-II	IS	/	Dead
9	?	?	S	Ex	A	AA	III-IV	IS	-	Alive
10	70	M	He,S,Sy	CT	A	AA	III-IV	IS	-	Dead
11	68	M	He,Me,Pu,S	Ga,Ar	A	AA	III-IV	IS	-	Alive
12	72	M	He,Me,Pu	Ga,Ar	A	AA	III-IV	IS	-	Alive
13[B]	74	M	He,S,Sy	Ga,CT	A	TA	Oesop	IS	-	Alive
14[C]	50	M	He,P,S,Sy	CT	A	AA	Stom	IS	+	Alive
15	79	M	Me,Pu	Ex	A	AA	III-IV	IS	-	Dead
16[D]	66	M	He,S	Ga	A	AA	III-IV	IS	Str,Stap	Alive
17	90	F	He,Pu,P,S,Sy	Ga,CT	A	AA	III-IV	/	/	Dead
18	72	M	He,Me,S	Ga,CT	A	AA	III-IV	IS	+	Dead

[A] reference 8, [B] reference 9, [C] reference 10, [D] reference 11

A : aneurysm, AA : abdominal aorta, Ar : arteriography, BLNE : beta lactamase negative enterococcus, CIA : common iliac artery, CT : computerized tomography, EA : extra anatomical bypass graft, Ex : surgical exploration, F : female, Fe : fever or sepsis, Ga : gastroduodenoscopy, H : hematochezia, He : hematemesis, I : inflammatory disease, IIA : internal iliac artery, , IS : in situ graft, M : male, Me : melaena, Oesop : oesophagus, P : back pain or abdominal pain, Pu : pulsating abdominal mass, S : shock, Sigm : sigmoid, Stap : Staphylococcus species, Stom : stomach, Str : Streptococcus viridans, Sy : syncope, T : tumour, TA : thoracic aorta, U : ulceration, I-II : duodenum part one or two, III-IV : duodenum part three or four, ? : not mentioned, / : no or none, + : positive not further specified, - : negative not further specified.

present the tip of the iceberg. This is probably due to a publication bias in favour of successfully treated patients and to patients dying of an unrecognised PAEF. However, PAEF case report publication should be stimulated as our knowledge of this life threatening clinical picture depends largely on this literature (6).

Pathogenesis

Three factors can play a role in the pathogenesis of PAEF : an inflammatory response, mechanical stress and anatomic orientation.

An inflammatory response in the vascular and more seldom in the intestinal wall is often found to cause a considerable weakness of this wall (2). Mostly this inflammatory response is caused by aneurysms. Resident cells in the aneurysm wall are shown to produce (metallo)proteinases leading to leucocyte infiltration causing production of more proteinases, chemokines and cytokines in the vascular wall. This leads to elastic fibre degradation in the media and collagen degeneration in the adventitia. Leucocyte recruitment might also be a consequence of adherent thrombus formation leading to hypoxia in the media with neovascularisation (12,14). Other factors causing inflammatory responses and weakening of the wall are pathogenic micro organisms, tumours, radiation, foreign bodies and inflammatory diseases (14).

Mechanical stress due to repetitive pulsatile pressure caused by an aneurysm might lead to erosion or even necrosis with considerable weakening of the digestive tract wall (2,5,13).

Finally, the third factor in the pathogenesis is anatomic orientation. The proximity of the aorta to some parts of the digestive tract is favourable for PAEF formation. Anatomical proximity can also be caused by fibrotic adherence to the bowel wall by an inflammatory response (2,5,13,14).

Eventually these three factors can cause considerable weakening of the vascular and intestinal wall resulting in aortoenteric fistulisation.

Aetiology

Six general types of causes of PAEF have been described so far. The vast majority, i.e. about 80 to 85%, is caused by aneurysms (5,7,13,14,15) (Table 2). In the Belgian cohort 89% of PAEF was due to aneurysm of which 33% were infected. In general only up to one quarter to half of the PAEF aneurysms are infected, proven by positive bacterial cultures, possibly because the blood flow through the fistula tract is directed towards the digestive tract, thereby flushing away the micro organisms out of the fistula (5,13,16). Infiltrating tumours also cause PAEF. Probably due to their anatomical proximity to the aorta in the retroperitoneal space, pancreatic tumours have been described frequently (2). Other causes are ulceration, e.g. by foreign bodies (e.g. tooth pick), inflammatory diseases (e.g. Takayasu Disease), radiotherapy, and infection (2,5,13,14,15,17).

Although infection is often a situation associated with aneurysms, we found no infection as solitary cause of PAEF in the Belgian cohort. Only rare cases of solitary infection are described, especially in younger patients.

Table 2. — **Characteristics of primary aorto enteric fistula : review data compared to Belgian cohort**

		Aorto enteric fistula	Review data (%)	18 Belgian cases (%)
EPIDEMIOLOGY			Incidence 0,04-0,8%	1 case per 57 surgical years*
CLINICAL FEATURES		Men	75-80	94
		Mean age (years)	62-64	70
		Gastrointestinal bleeding	64-94	83
		Haematemesis	61-78	61
		Melena	46-49	33
		Haematochezia	15	5,5
		Shock	33-62	72
		Pulsating mass	17-54	33
		Abdominal or back pain	32-64	22
		Classical triad°	11-28	5,5
		Syncope	10	39
		Fever or sepsis	7-17	17
TYPE	ORIGIN FISTULA	Infrarenal AA	Vast majority	83
		Other	Vast minority	17
	END FISTULA	Duodenum III-IV	54-86	67
		Other	14-46	33
AETIOLOGY		Aneurysm	80-85	89
		Infectious aneurysm	25-50	33
		Not infectious aneurysm	50-75	67
		Other	15-20	11
DIAGNOSTIC MODALITIES		Surgical exploration	> 99	50
		Abdominal CT °°	61-94	33
		Gastroduodenoscopy	18-29	33
		Arteriography	0-26	11
MORTALITY		Untreated	100	100
		Overall	>30	29
		Positive peroperative culture	>30	33
		Negative peroperative culture	<30	22
		Presenting with shock	>30	33
		After EA bypass grafting	>30	50
		After in situ grafting	20-40	27

Data collected from literature review are pointed out and compared to data from the Belgian cohort. Diagnostic mean detection rate data from literature review are pointed out in the left column. The right column gives the percentages of use of the different diagnostic modalities in the analysis of 18 Belgian cases. Mortality rates within 30 days postoperatively are indicated. AA : abdominal aorta, EA : extra anatomical, * as a vascular surgeon, ° abdominal or back pain, gastrointestinal blood loss and abdominal pulsating mass, °° contrast enhanced computerized tomography, > : more than, < : less than.

Previous intravascular procedures and a depressed immunocompetence are described as risk factors for infective PAEF (13,16). Causative micro organisms reported so far are : Klebsiella sp., Salmonella sp., Staphylococcus sp. and E. Coli most commonly, but also Streptococcus sp., Clostridium sp., Lactobacillus sp., Bacteroides, Mycosis, E. Faecalis, Mycobacteria sp., T. Pallidum (syphilis) and Arizona Hinshawii (2,5,7,13, 15). The prevalence of infective causes, especially tuberculosis and syphilis, has strongly diminished over the last decades due to effective antibiotical treatment (5,7). In our series only six positive cultures were found ; E.Coli, Enterococcus, Streptococcus sp. and Staphylococcus sp.

Types

The vast majority of PAEF, 83% in the Belgian cohort, originates from the infrarenal part of the abdominal aorta (Table 2). Sometimes the thoracic aorta is the origin and only rare cases of iliac artery aneurysm originating fistula have been described as in our case report : up to 1996 only 16 cases of PAEF originating from a solitary iliac aneurysm have been described (18). Due to the anatomical orientation of the third part of the duodenum, retroperitoneally fixed just anterior to the abdominal aorta, this is the most common bowel segment involved in PAEF, present in 54% to 86%, compared to 67% in our series (2,5,7,13,15,17,19). Because of the

proximity over a relatively long distance to the thoracic aorta the oesophagus is the second most frequent type of PAEF, present in up to one third of the patients (5). The stomach, first and second duodenal part, small and large bowel and sigmoid are less frequent places of presentation of PAEF (2,17). Therefore a primary fistula between the iliac artery and the sigmoid, as described in our case, is extremely rare. No other such cases were reported in the Belgian cohort.

Clinical presentation

The big pitfall in clinical presentation of PAEF is the not specific, not constant and insidious character of the symptoms. This is the main reason why 46% of PAEF patients were diagnosed post mortem in a review of 253 patients by Roux *et al.* (7).

Obviously gastrointestinal bleeding is the most common presentation of PAEF : up to 94% in a review by Saers *et al. (*5) and 83% in our series (Table 2). Three quarters of the patients present with haematemesis due to upper gastrointestinal bleeding. About one half of the patients have melaena and a minority, as in our case, presents with haematochezia (5,15). Often this gastrointestinal bleeding has a typical biphasic evolution. The initial transient and mostly self limited herald bleed, also called sentinel haemorrhage as observed in our case patient , is followed by a massive bleed that can cause abrupt exsanguination. The herald bleed is thought to be caused by low local blood flow and hypotension causing a thrombus formation that temporary plugs the fistula tract at one hand and secondary spasm of the bowel musculature on the other hand (2,5,12,13,14,17,18,19,20). The time interval between the herald and massive bleed can range from hours to months, but in 70% of patients this interval is more than 6 hours and in 50% more than 24 hours (3,5,7,15,17). The importance of gastrointestinal bleeding as a sign has increased over time as Sweeney *et al.* found gastrointestinal bleeding in only 64% (3,14). This might indicate that physicians are more aware that a sentinel haemorrhage must raise strong suspicion for the diagnosis of PAEF (5). Due to massive bleeding, one third up to two thirds of patients present with shock (5,7). The Belgian cohort patients presented more with shock, up to 72%.

The classical triad of gastrointestinal blood loss, abdominal or back pain and pulsating abdominal mass is present in only 11% to 28% and has declined over the years, compared to 5,5% in the Belgian cohort (2,3,5,7,15). Saers *et al.* point out this might indicate that the role of clinical examination has become less important in practice (5). Still, in 17 to 54% a pulsating abdominal mass is detectable and half of patients complain about abdominal or back pain (3,4,5,7).

Other less frequent symptoms at presentation are syncope in 10% and fever or sepsis in 7 to 17%. PAEF is three to four times as frequent in males and the mean age is between 62 and 64 years old (5,7). Laboratory results show leucocytosis in one quarter of the patients and up to two thirds have lower than normal haemoglobin levels or a reduced haematocrit (5,13).

Our case was atypical as it presented with less specific symptoms such as left iliac fossa pain, fever and haematochezia, mimicking a diagnosis of diverticulitis.

Considering this clinical presentation, PAEF must always be in the differential diagnosis of gastrointestinal bleeding. Still, a high index of suspicion of patients having PAEF until proven otherwise must be present when the patient presents with (considerable) (upper) gastrointestinal blood loss and has one of the following three problems : known aneurysm, initial herald bleed or pulsating abdominal mass (2,4,5,13,15,17,19,20). The incidence of PAEF in patients with known abdominal aneurysm and gastrointestinal haemorrhage can be up to 18% (6,14). Therefore, if a patient is known with aneurysm, this should always be well indicated in his or her medical file.

Diagnosis

As PAEF is a life threatening condition, the right choice of diagnostic investigation is critical : a quick, non-invasive method with a high detection rate of PAEF is ideal. However there are many other more frequent causes of gastrointestinal blood loss to exclude. This makes the diagnosis of a PAEF a difficult challenge which is reflected in the controversy about the best diagnostic approach found in the literature. Most authors agree that plane abdominal x-ray, abdominal ultrasonography, colonoscopy and leucocyte scans are of little or no value in the search for PAEF. Four diagnostic options can be considered : gastroduodenoscopy, CT scan, arteriography and surgical exploration (2,5,6,13,19) (Table 2).

Surgical exploration by laparotomy and complete mobilisation of the duodenum with aortic control is the only and highly sensitive diagnostic way for hemodynamically unstable patients with a high index of suspicion (4,15,20,21). Though, in our Belgian cohort only 46% of PAEF patients had immediate surgical exploration without preoperative diagnostic investigations when presenting with signs of shock. This might indicate that half of the patients had no high index of suspicion and thereby confirming the not specific, not constant and insidious character of the symptoms. Another explanation is PAEF was not in the differential diagnose of the examinating physician or diagnostic certainty was seeked before initiating invasive treatment. Only hemodynamically stable patients are eligible for the other diagnostic investigations, taking into account that a negative investigation never rules out the presence of PAEF (5,18).

Gastroduodenoscopy is often the first diagnostic option. The main rationale is the exclusion of other causes of (upper) gastrointestinal bleeding, In 19 to 23% of PAEF patients gastroduodenoscopy reveals co-existing digestive tract pathology that leads to diagnostic mistakes (7,13,14). The mean detection rate for

PAEF during gastroduodenoscopy ranges only between 18 and 29% (5,7,14,15). In the Belgian cohort gastroduodenoscopy was used as one of the diagnostic tools in 33%. The rather low detection rate of gastroduodenoscopy is due to the fact that a PAEF is mostly present in the third part of the duodenum which is technically more difficult to visualise during endoscopy and not routinely done. Therefore the use of a side-viewing endoscope has been advised (2,5,7). An active bleeding site, adherent blood clot, ulceration combined with a blood filled stomach, erosion or necrosis or granuloma accompanied by an excentric protruding pulsating mass through the duodenum or oesophagus or accompanied by a known aneurysm and known aneurysm with negative findings in stomach and first two parts of duodenum have all been described as suspicious for the presence of PAEF. A possible dislodgement of the (temporary) fistula thrombus is a possible danger during endoscopy (2,5,13,17).

A contrast enhanced multislice CT scan currently has the best detection rate of all preoperative investigations (2,5,7,15). Saers *et al.* found a detection rate up to 61% comparing to gastroduodenoscopy (25%) and arteriography (26%) (5). It must be considered that the literature review included case reports as early as 1987 where CT was first used as a diagnostic modality. Since the first case report where CT was used successfully, CT technology has undergone an important evolution and the sensitivity in detecting PAEF has improved up to 94% (21). Today contrast enhanced multislice CT is considered to be gold standard in preoperative diagnosis of PAEF. Contrast enhanced multislice CT is a quick, non-invasive tool and provides diagnostic information superior to those of other diagnostic modalities such as gastroduodenoscopy and angiography (7). Contrast enhanced multislice CT also serves as an aid for surgical planning and avoids danger of thrombus dislodgement (21). Air bubbles in the aortic wall as in our case, bowel wall thickening overlying an aneurysm, disruption of aortic fat cover as in our case, contrast in close contact or penetrating the bowel and presence of air in the retroperitoneal space have been described in patients with PAEF (2,5,6,13, 17,18). These signs render contrast enhanced multislice CT the most sensitive diagnostic modality for PAEF. Reflecting the superior sensitivity, the use of CT scans has tripled over the last two decades in patients suspected of having PAEF (5). In the Belgian cohort 33% underwent contrast enhanced CT.

Finally, we believe arteriography actually has a minor role in the diagnosis of PAEF. The value of arteriography in this context is known to be limited. The decline in use of arteriography is also seen in our Belgian cohort ; only 11% of the cases underwent arteriography. Detection rates described are low ; 0 to 26% (5,7,14). Arteriography is helpful only during active bleeding, and a rate of 1 millilitre per minute is required for visualization (22). Extravasation of contrast agent into the digestive tract is seen as proof of PAEF presence (2,5). But the investigation is invasive, time consuming and has no diagnostic advantages over contrast enhanced multislice CT. There even is important danger of fistula thrombus dislodgement.

Based upon the above mentioned literature data and the results in the Belgian series we propose a diagnostic flow chart (Fig. 2). If there is no raised index of suspicion for PAEF in an acute (upper) gastrointestinal bleed, it seems rational to perform endoscopy before CT scanning. A positive index of suspicion must lead to hemodynamic status evaluation. Unstable patients are eligible for immediate explorative surgery. Stable patients are good candidates for initial contrast enhanced multislice CT scan as it provides an accurate measure of aortic size and identifies aortic rupture or communication with the bowel. Gastroduodenoscopy can follow multislice CT scanning if necessary. We consider arteriography to be an exceptional option with limited significance.

Treatment

Surgery and antibiotics are the most important modalities in PAEF therapy (2,4,5,6,7,13,15,19). In a patient suspected of having PAEF, hypotension must not be treated too aggressively as hypotension induces thrombus formation in the fistula tract thereby preventing exsanguination. Systolic blood pressures should be kept between 60 to 100 millimetres of mercury. This idea of 'controlled hypotension' has already been proven life saving in abdominal aneurysm rupture (5,18).

Over the last decades the percentage of patients having surgery has increased (5). Urgent surgery is the only definitive life saving option to treat PAEF and exists of a vascular part and a digestive part (2,5,7,13). Based on clinical presentation or diagnostic imaging a choice needs to be made between in situ grafting, extra anatomical bypass grafting or endovascular treatment for the vascular part. Primary aortic repair by means of patch or aortic suture is no longer considered a beneficial therapy (7). Laparatomy with disconnection and closure of the fistula, followed by in situ vascular graft insertment eventually after aneurysmectomy is actually considered to be the safest option (2,5,6,13,14,17). An antibiotic impregnated graft gives better results, as does the interposition of tissue between graft and bowel, e.g. wrapping of an omental flap around the vascular anastomosis to prevent secondary fistulisation (2,4,5,13). Only when extensive bowel sepsis or severe peritonitis is suspected, as in our case, extra anatomical bypass grafting with disconnection and fistula closure with aneurysmectomy is indicated (2,4,5,7,13,14,15,17). To install the bypass before laparotomy seems reasonable as it diminishes the ischemia time of the lower limbs resulting in less lower limb amputations, but this issue is still discussed (7,14).

The digestive part obeys a simple principle : suture in healthy tissue after removal of the fistula end. Large defects or necrotic parts of the digestive tract might lead

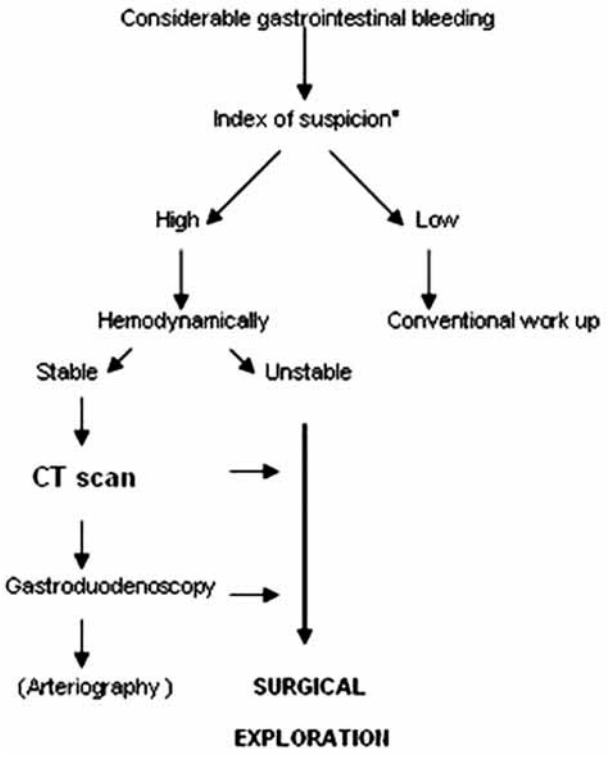


Fig. 2. — Diagnostic flow chart for early diagnosis of primary aorto enteric fistula.
Fig. 2. °index of suspicion : known aortic aneurysm, previous herald bleed or pulsating abdominal mass.

to partial resection with end-to-end anastomosis or temporarily deviation stoma, while smaller defects can be treated by means of simple suture (6,15). Closure of the fistula tract alone is never considered an option as it induces sepsis and multiple organ failure (5).

Only when conventional surgery is not suitable or the patient is hemodynamically too unstable, endovascular or percutaneous treatment might be feasible (5). Endovascular stenting for vital arteries and embolisation for non vital arteries by means of fibrin glue, balloon occlusion, coils, particles or cryano-acrylate injection have been successfully described (1,23). A potential drawback from this treatment is the risk of overwhelming sepsis as the infection source remains in situ. As this treatment might also be not curative, patients need to undergo later definitive repair after recovery on an elective basis. Still, the last word about endovascular treatment has not been said because this treatment option knows important evolutions currently (5,7). Finally, as no prospective studies are available, determining the right surgical option remains difficult.

Systemic and preferably intravenously given antibiotics against gram negative and positive strains should be administered whenever a PAEF is suspected, even before surgery (5,13). When cultures of the aorta or periaortic site, taken during surgery, prove to be negative, antibiotics are discontinued after 7 to 10 days. Positive cultures must lead to an antibiotic treatment regimen of at least 4 to 6 weeks, possibly longer depending on clinical or biochemical evolution and tailored to the cultured organisms (2,5,6,7,13,15). After endovascular treatment a prolonged period of antibiotic treatment is feasible, sometimes life long as the infection source remains in situ (5).

Prognosis

PAEF has a poor prognosis. No patient survives this situation if left untreated (1,2,5,13) (Table 2). Over the years overall mortality after surgical treatment has diminished, but remains higher than 30%, comparable to 29% in the Belgian cohort (2,5,13,14,15). This rather low overall mortality rate in our series might be due to a raportation bias in favour of successfully treated patients. Mortality rates of in situ grafting remain stable around 20 to 40%, comparable to 27% within 30 days postoperative in our series (5,14,15). Extra anatomical bypass grafting is associated with higher mortality, up to 50% in the Belgian cohort, possibly because this treatment is often limited to patients presenting with important signs of ongoing infection (5,7,14). Positive bacterial cultures, shock and time loss due to non conclusive diagnostic procedures have been proven to raise mortality (2,13,14,15,19). In our series this trend was also confirmed : peroperative positive cultures raised mortality within 30 days to 33%, compared to 20% when peroperative cultures were negative. In presence of shock 33% of the patients died within 30 days postoperative.

Death is usually caused by sepsis or multiple organ failure (5,13). Post operative complications occur in at least 40% of the cases, including : acute renal insufficiency, graft infection in 5-25%, proximal anastomotic rupture in 10%, aortic stump blow out in extra anatomical bypass grafting in 10 to 30% and early lower limb ischemia leading to limb amputation in 6% after in situ grafting and up to 20% after traditional extra anatomical bypass grafting (2,14,15). Secondary aortoenteric fistulisation is rather rare according to Goshtasby *et al.*, but Tareen *et al.* report an incidence of 14% (13,15). Close follow up by means of clinical examination, laboratory testing and CT scanning initially every 3 to 6 months to monitor for pseudo aneurysm formation or other signs of graft disruption or infection seems feasible (5).

Conclusion

PAEF is suspected when a patient presents with (considerable) (upper) gastrointestinal blood loss and has a known aneurysm, an initial herald bleed or a pulsating abdominal mass. If hemodynamically unstable, prompt surgical exploration is gold standard. Hemodynamically stable patients must undergo contrast enhanced multislice CT rather than gastroduodenoscopy or arteriography to make early diagnosis. Surgery is the only definitive life saving treatment. Overall mortality is at least 30%. Late diagnosis, positive peroperative cultures and shock are indicators of poor prognosis.

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