Systematic review

Systematic review of atraumatic splenic rupture

P. Renzulli[1], A. Hostettler[1], A. M. Schoepfer[1,2], B. Gloor[1] and D. Candinas[1]

[1]Department of Visceral Surgery and Medicine, Inselspital, Berne University Hospital, and University of Berne, Berne, Switzerland, and [2]Farncombe Family Institute of Digestive Health Research, McMaster University, Hamilton, Ontario, Canada

Correspondence to: Dr P. Renzulli, Department of Visceral Surgery and Medicine, Inselspital, Berne University Hospital, and University of Berne, CH-3010 Berne, Switzerland (e-mail: pietro.renzulli@stgag.ch)

Background: **Atraumatic splenic rupture (ASR) is an ill defined clinicopathological entity.**

Methods: **The aim was to characterize aetiological and risk factors for ASR-related mortality in order to aid disease classification and treatment. A systematic literature review (1980–2008) was undertaken and logistic regression analysis employed.**

Results: **Some 632 publications reporting 845 patients were identified. The spleen was normal in 7·0 per cent (atraumatic–idiopathic rupture). One, two or three aetiological factors were found in 84·1, 8·2 and 0·7 per cent respectively (atraumatic–pathological rupture). Six major aetiological groups were defined: neoplastic (30·3 per cent), infectious (27·3 per cent), inflammatory, non-infectious (20·0 per cent), drug- and treatment-related (9·2 per cent) and mechanical (6·8 per cent) disorders, and normal spleen (6·4 per cent). Treatment comprised total splenectomy (84·1 per cent), organ-preserving surgery (1·2 per cent) or conservative measures (14·7 per cent). The ASR-related mortality rate was 12·2 per cent. Splenomegaly ($P = 0·040$), age above 40 years ($P = 0·007$) and neoplastic disorders ($P = 0·008$) were associated with increased ASR-related mortality on multivariable analysis.**

Conclusion: **The condition can be classified simply into atraumatic–idiopathic (7·0 per cent) and atraumatic-pathological (93·0 per cent) splenic rupture. Splenomegaly, advanced age and neoplastic disorders are associated with increased ASR-related mortality.**

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Introduction

Atraumatic splenic rupture (ASR) is rare but often life threatening. The first author and co-workers have recently published two case reports of ASR[1,2] and in the process of preparing the manuscripts recognized the ill defined nature of the condition. Little is known about patient characteristics, incidence and aetiology. There are no guidelines on management. Data on outcome and risk factors influencing ASR-related mortality are limited. Finally, the nomenclature of ASR is often ambiguous and contradictory, including descriptive terms such as 'true spontaneous', 'spontaneous', 'idiopathic', 'pathological', 'atraumatic' and 'occult'.

The present study aims to provide a description of patient characteristics, a specification of aetiological factors and an identification of risk factors for ASR-related mortality. A simplified disease classification is suggested and the principles of management are discussed.

Methods

A comprehensive literature review on ASR was performed. All publications from 1 January 1980 to 30 June 2008 were scrutinized. The literature search (Medline, EMBASE, Cochrane Library) was limited to patients over 18 years old, and restricted to papers written in English, German, French, Italian, Spanish or Japanese.

Splenic ruptures of predominantly iatrogenic or traumatic origin were excluded. These occurred in patients undergoing transoesophageal echocardiography[3], endoscopic retrograde cholangiopancreatography[4], colonoscopy[5], laparoscopic procedures[6], extracorporeal shockwave lithotripsy[7] and electroconvulsive therapy[8].

Patients with splenic infarcts without rupture and those with an isolated vascular pathology without splenic parenchymal laceration (for instance splenic vein rupture or splenic artery aneurysm/rupture) were also excluded[9].

All publications were reviewed and the following information gathered: language of publication, number of patients, sex, age, diagnostic methods, conservative or surgical treatment, presence of splenomegaly, splenic weight, splenic rupture-related mortality, aetiology and nomenclature/descriptive classification.

Definitions

With respect to the nomenclature/descriptive classification, the terms used by the original authors within the title, keywords or abstract of the article were adopted. Splenic ruptures were therefore classified as 'spontaneous', 'pathological', 'atraumatic', 'occult' or 'without description/no classification'.

The assessment of splenomegaly was based on the authors' description (for example, 'enlarged spleen', 'splenomegaly') as well as on the reported spleen weight or size. 'Splenomegaly' was defined as a spleen weight above 200 g (resection specimen) or as a spleen size exceeding 110 × 70 × 50 mm (*in vivo* measurements by ultrasonography or computed tomography (CT)). This definition is based on the findings of two post-mortem series[10,11] and one ultrasonographic study[12]. The weight of the normal spleen in healthy adults was found to depend on body height and bodyweight (body mass index) and sex. The overall mean weight of the spleen was 156 g[10]. The mean(s.d.) weight of the normal spleen in men and women was found to be 156(87) and 140(78) g respectively[11]. Ultrasonographic determination of spleen size in 793 healthy adults showed that the dimensions were less than 110 × 70 × 50 mm and the calculated spleen weight (rotation ellipsoid formula) was less than 190 g in 95 per cent of all subjects[12].

A non-surgical conservative approach or an attempted conservative approach was classified as such only if it lasted for at least 24 h. Patients operated on within 24 h of the definitive diagnosis of splenic rupture were classified as undergoing primary surgical treatment.

Finally, 'mortality' was defined in relation to the event of splenic rupture as ASR-related mortality. Patients who survived the rupture were classified as 'survivors', even if they died from underlying disease within a 30-day follow-up period or during the hospital stay.

Statistical analysis

For statistical analysis a database was built in Excel™ 2000 (Microsoft Switzerland, Wallisellen, Switzerland). All statistical analyses were performed with the statistical package program Stata™ version 9 (Stata Corporation, College Station, Texas, USA). Data distribution was assessed with QQ plots. χ^2 tests were used for analysis of unpaired categorical data. A Bonferroni adjustment was performed in situations of multiple testing. Multivariable logistic regression modelling was performed to determine the primary associations between the outcome variables ASR-related mortality and choice of management, and dependent variables such as number of predisposing factors, major aetiological group, age and splenomegaly/splenic weight. Odds ratios (ORs), standard errors (s.e.) and 95 per cent confidence intervals (c.i.) were calculated. $P < 0{\cdot}050$ was considered statistically significant. In order to allow a meaningful statistical analysis, the aetiological factors for ASR were clustered into six major groups.

Results

A total of 632 publications reporting 845 patients with ASR were identified (*Appendix 1* (supporting information)). Some 466 publications (73·7 per cent) were in English, 58 (9·2 per cent) in Spanish, 55 (8·7 per cent) in French, 25 (4·0 per cent) in German, 15 (2·4 per cent) in Italian and 13 (2·1 per cent) in Japanese. Each publication reported a mean of 1·3 (range 1–41) patients. There was a mean of 30 newly described patients each year over the 28·5-year study period. There was a 2 : 1 male predominance (sex was specified for 801 patients, of whom 534 (66·7 per cent) were men). Age was reported in 799 instances. The mean(s.d.) age was 45(17) (median 45, range 18–86) years. The splenic rupture was classified by the authors as 'spontaneous' in 580 patients (68·6 per cent), 'pathological' in 78 (9·2 per cent), 'atraumatic' in 21 (2·5 per cent) or 'occult' in four (0·5 per cent). No description was given for 162 patients (19·2 per cent), and no classification was made. All patients denied any recent trauma.

Aetiology

In 59 patients (7·0 per cent) the spleen was described as being completely normal and no aetiological factor for ASR was identified. In 711 patients (84·1 per cent) a single aetiological factor was found. In 69 patients (8·2 per cent) the splenic rupture was related to two underlying pathologies (for example, splenic tuberculosis and cavernous haemangioma of the spleen[13]), and in six (0·7 per cent) the splenic rupture was caused by a

Table 1 The six major aetiological groups

	No. of factors ($n = 926$)*
Neoplastic disorders	
Malignant haematological disorders	152 (16·4)
Acute myelogenous leukaemia[1–18]	21
Acute lymphoblastic leukaemia[19–30]	12
Various leukaemias[5,31–51]	24
Hodgkin's lymphoma[52–55]	4
Non-Hodgkin's lymphoma[5,32,34,41,56–97]	55
Myeloproliferative disorders[5,33,98–111]	24
Myelodysplastic syndromes[34,82,110,112–119]	12
Non-malignant haematological disorders	19 (2·1)
Histiocytosis[93,120–127]	9
Idiopathic thrombocytopenic purpura[33,128–130]	4
Various[5,131–135]	6
Primary neoplastic disorders	75 (8·1)
Angiosarcoma[136–155]	31
Peliosis[111,114,116,128,129,132,156–167]	18
Cystic lesions[33,163,168–170]	7
Haemangioma[33,171–176]	7
Various[33,177–185]	12
Secondary metastatic neoplastic disorders	35 (3·8)
Choriocarcinoma[186–192]	7
Lung cancer[33,189,193–196]	6
Melanoma[5,197–199]	4
Various[5,79,200–213]	18
Infectious disorders	
Viral infectious disorders	137 (14·8)
Infectious mononucleosis[4,5,163,214–282]	102
Cytomegalovirus infection[283–294]	13
Human immunodeficiency virus infection[75,104,185,295–298]	8
Dengue fever[246,299–304]	7
Various[5,305–310]	7
Bacterial infectious disorders	61 (6·6)
Endocarditis[5,208,311–318]	16
Q fever[319–325]	7
Tuberculosis[33,135,172,297,326–328]	7
Typhoid fever[329–331]	5
Various[5,243,332–355]	26
Protozoal infectious disorders	54 (5·8)
Malaria tertiana (*Plasmodium vivax*)[246,356–372]	23
Malaria tropica (*Plasmodium falciparum*)[298,356,373–389]	20
Various[190,390–400]	11
Fungal infectious disorders	1 (0·1)
Aspergillosis[1]	1
Inflammatory, non-infectious disorders	
Local inflammatory and neoplastic disorders	101 (10·9)
Chronic pancreatitis[5,88,104,401–435]	65
Acute pancreatitis[5,33,104,131,400,401,436–450]	20
Pancreatic cancer[104,432,451–454]	7
Various[5,33,455–457]	9
Amyloidotic disorders	35 (3·8)
Primary amyloidosis[458–475]	22
Secondary amyloidosis[327,476,477]	4
Various[123,478–484]	9
Vascular disorders	20 (2·2)
Wegener's granulomatosis[485–489]	5
Polyarteritis nodosa[490–492]	3
Various[33,493–499]	12

Table 1 (Continued)

	No. of factors ($n = 926$)*
Genetic disorders	16 (1·7)
Haematological disorders[4,104,106,208,243,500–506]	12
Storage diseases[124,127,228,243]	4
Autoimmune disorders	13 (1·4)
Rheumatoid arthritis[345,507,508]	5
Systemic lupus erythematodes[5,450,509,510]	4
Various[33,135,511]	4
Drug- and treatment-related disorders	
Drug-related splenic rupture	67 (7·2)
Granulocyte colony-stimulating factor[8,14,49–51,112,115,223,460–462,506,512–520]	22
Anticoagulation[5,33,202,315,316,521–536]	22
Thrombolytic therapy[173,249,537–542]	9
Various[7,16,17,110,165,177,461]	14
Dialysis-related splenic rupture	18 (1·9)
Haemodialysis[157,182,389,442,478,479,482,496,523,543,544]	13
Continuous ambulatory peritoneal dialysis[103,481,545–547]	5
Mechanical disorders	
Pregnancy-related splenic rupture	40 (4·3)
During pregnancy[174,381,526,529,548–558]	15
During labour and postpartum[18,30,169,170,559–563]	9
Postcaesarean section[134,188,562,564–567]	7
Intrasplenic pregnancy[568–576]	9
Congestive splenomegaly	23 (2·5)
Liver cirrhosis (portal hypertension)[5,33,133,181,546,577–582]	16
Hepatic inflow occlusion (Pringle manoeuvre)[583–587]	7
Normal spleen	
Normal spleen	59 (6·4)
With no triggering factor[104,106,163,208,478,588–617]	41
With triggering factor[228,523,578,602,618–632]	18

Values in parentheses are percentages. *Of 845 patients, 59 had a normal spleen (no aetiological factor), and 711, 69 and six patients respectively presented with one, two or three aetiological factors, giving a total of 926 aetiological factors for atraumatic splenic rupture. The references are listed in *Appendix 1* (supporting information).

combination of three aetiological factors (for example, haemophagocytic syndrome, splenic tuberculosis and sarcoidosis of the spleen[14]). The coincidence of several aetiological factors in a single patient resulted in a total of 926 aetiological factors for ASR in the 845 patients. These factors, divided into six major and subdivided into 18 minor aetiological groups, are listed in *Table 1*. The three commonest causes of ASR were malignant haematological disorders (16·4 per cent; for instance, acute leukaemia, non-Hodgkin's lymphoma), viral infectious disorders (14·8 per cent; for instance, infectious mononucleosis, cytomegalovirus infection) and local inflammatory and neoplastic disorders (10·9 per cent; for instance, acute and chronic pancreatitis).

Diagnostic procedures

At hospital admission, free intraperitoneal fluid was diagnosed by ultrasonography in 208 patients (24·6 per cent) or by CT in 194 (23·0 per cent). The diagnosis of haemoperitoneum was made by peritoneal lavage in 89 patients (10·5 per cent). The diagnostic method demonstrating the actual splenic rupture was reported in 731 patients. In these patients, the final diagnosis of splenic rupture was made by laparotomy in 309 patients (42·3 per cent), CT in 237 (32·4 per cent), ultrasonography in 136 (18·6 per cent), scintigraphy in five (0·7 per cent), laparoscopy in four (0·5 per cent), angiography in two (0·3 per cent) or post-mortem examination in 38 (5·2 per cent).

Knowledge of the underlying aetiology of ASR is crucial when deciding on treatment. Aetiology was known before hospital admission in 183 patients (21·7 per cent). In a further 169 patients (20·0 per cent) the correct diagnosis was made after admission, either before surgery (113 patients, 13·4 per cent) or before the initiation of non-surgical treatment (56 patients, 6·6 per cent). In a further 242 patients, the aetiology was identified either during (seven patients, 0·8 per cent) or after (235 patients, 27·8 per cent) surgery. Finally, in 22 patients (2·6 per cent)

the diagnosis was made at autopsy, and in 59 patients (7·0 per cent) no underlying aetiological factor for ASR was found. In 170 patients (20·1 per cent) no information was given about the exact time of diagnosis.

Splenomegaly

Splenomegaly was a common feature, affecting 465 patients (55·0 per cent). In 126 (14·9 per cent) the spleen was described as normal in size, and in 254 (30·1 per cent) no information on size was given. The weight of the resected spleen was reported in 320 instances (37·9 per cent). In 251 (78·4 per cent) of 320 patients, the measured splenic weight exceeded 200 g. The mean(s.d.) splenic weight was 704(677) (median 462, range 70–4000) g.

Treatment

Treatment was specified in 774 patients (91·6 per cent). In 31 patients (3·7 per cent) no information was given about the exact treatment. In 38 patients (4·5 per cent) no specific treatment was given and the diagnosis was made at autopsy; two patients (0·2 per cent) died after establishment of a diagnosis of ASR but before effective treatment. Treatment modalities are outlined in *Table 2*.

Most patients (660 (85·3 per cent) of 774) had surgery within 24 h of the diagnosis of splenic rupture. One patient underwent secondary splenectomy after failure of organ-preserving surgery (splenorrhaphy). The ASR-related mortality rate in the operated group was 7·4 per cent (49 of 660 patients). A total of 114 patients (14·7 per cent) received primary non-surgical conservative treatment for at least 24 h after the diagnosis of splenic rupture. At least 16 (17 per cent) of 92 patients underwent secondary splenectomy in the event of rebleeding and haemodynamic instability. The ASR-related mortality rate in the conservatively treated patient group was 4·4 per cent (five of 114). The two diseases that were treated conservatively most frequently were infectious mononucleosis (32 of 101, 31·7 per cent) and malaria (15 of 50, 30 per cent). The respective success rates for non-operative management were 25 (78 per cent) of 32 and 12 (80 per cent) of 15. The overall splenic salvage rate for all treatment modalities combined was 106 (13·7 per cent) of 774.

Table 2 Treatment of splenic rupture in 774 patients

	No. of patients	Reoperation*	ASR-related mortality
Primary surgical treatment†	660 (85·3)		49 (7·4)
Total splenectomy	651	–	48
Organ-preserving surgery	9	1	1
Primary non-surgical treatment‡	114 (14·7)		5 (4·4)
Conservative treatment§	107	16	5
Conservative treatment with splenic arterial embolization	7	0	0

Information was not available for 31 of the 845 patients, and in 40 patients the rupture was diagnosed only at autopsy ($n = 38$) or before effective treatment was started ($n = 2$). Values in parentheses are percentages. *Secondary splenectomy after organ-preserving surgery (for example, splenorrhaphy, partial splenectomy) or conservative treatment; †surgery within 24 h of diagnosis; ‡conservative treatment for at least 24 h; §secondary splenectomy rate was not reported in 22 patients. ASR, atraumatic splenic rupture.

Atraumatic splenic rupture-related mortality

Information concerning ASR-related mortality was available for 786 patients; no information was given for 59. The overall ASR-related mortality rate, irrespective of treatment modality, was 96 (12·2 per cent) of 786.

Further statistical analyses were based on six major aetiological groups (*Table 1*). The main patient characteristics are detailed in *Table 3*. Initially a univariable analysis was undertaken. To account for the differences in patient characteristics, a multivariable logistic regression analysis was performed subsequently.

The χ^2 test showed a significantly increased ASR-related mortality rate in patients with more than one aetiological factor ($P = 0·016$), in patients aged over 40 years ($P = 0·011$) and in patients with splenomegaly ($P = 0·035$). ASR-related death was not related to sex ($P = 0·238$)

Table 3 Patient characteristics for the six major aetiological groups ($n = 926$)

	No	Men (%)	Mean(s.d.) age (years)	ASR-related mortality (%)
Neoplastic disorders	281	67·4	52(16)	21·4
Infectious disorders	253	75·8	35(15)	8·7
Inflammatory, non-infectious disorders	185	69·1	48(13)	9·5
Drug- and treatment-related disorders	85	57	52(14)	13
Mechanical disorders	63	25	36(13)	17
Normal spleen	59	63	47(15)	2

ASR, atraumatic splenic rupture.

Table 4 Multivariable logistic regression analysis of risk factors for atraumatic splenic rupture-related mortality in 845 patients

	Odds ratio	s.e.	*P*
Age > 40 years	1·94 (1·20, 3·13)	0·47	0·007
Male sex	0·77 (0·49, 1·19)	0·17	0·239
Primary surgical treatment	1·85 (0·72, 4·76)	0·89	0·199
Splenomegaly	2·34 (1·04, 5·26)	0·97	0·040
More than one aetiological factor*	1·12 (0·42, 3·01)	0·57	0·820

Values in parentheses are 95 per cent confidence intervals. *Sixty-nine patients had two aetiological factors, and six had three factors.

Table 5 Multivariable logistic regression analysis of risk factors for atraumatic splenic rupture-related mortality in relation to 926 aetiological factors

	Odds ratio	s.e.	*P*
Neoplastic disorders	2·63 (1·76, 3·94)	0·54	0·008
Infectious disorders	1·74 (0·85, 3·16)	0·41	0·092
Inflammatory, non-infectious disorders	0·58 (0·32, 1·04)	0·17	0·065
Drug- and treatment-related disorders	0·96 (0·48, 1·92)	0·34	0·904
Mechanical disorders	1·42 (0·70, 2·89)	0·52	0·235
Normal spleen	0·11 (0·02, 0·81)	0·11	0·030

Values in parentheses are 95 per cent confidence intervals.

or choice of primary treatment, surgical or conservative ($P = 0·193$). A subgroup analysis examined the influence of a change in management on outcome. Patients undergoing a primarily conservative treatment approach had a significantly increased ASR-related mortality rate if they had to undergo secondary surgery, in comparison to patients who were successfully treated conservatively ($P < 0·001$).

The multivariable logistic regression analysis found splenomegaly (OR 2·34 (95 per cent c.i. 1·04 to 5·26); $P = 0·040$) and age above 40 years (OR 1·94 (95 per cent c.i. 1·20 to 3·13); $P = 0·007$) to be significantly associated with an increased ASR-related mortality rate. Male sex, primary surgical treatment and multiple aetiological factors were not related to mortality (*Table 4*). Multivariable analysis of the six major aetiological groups showed that neoplastic disorders were associated with an increased ASR-related mortality rate (OR 2·63 (95 per cent c.i. 1·76 to 3·94); $P = 0·008$) compared with other aetiologies. A normal spleen and no aetiological factor, however, was associated with a significantly decreased ASR-related mortality (OR 0·11 (95 per cent c.i. 0·02 to 0·81); $P = 0·030$) (*Table 5*).

Discussion

Recent review articles have identified a relatively small number of patients with ASR and have provided only summative and approximate information about aetiology[15–17]. The present study is a more comprehensive analysis of the recent literature on ASR in the adult population. Being a review, however, is something of a limitation, but because of the rarity of ASR there is no other realistic approach. One major problem was missing data, causing subsequent analyses to be limited to those characteristics with sufficient information. In this respect, it proved impossible to include the American Association for the Surgery of Trauma (AAST) grades of splenic injury[18]. For the same reason, the amount of haemoperitoneum as a potential risk factor for failure of non-operative management could not be assessed[19].

ASR is often life threatening. The present study calculated the overall ASR-related mortality rate as 12·2 per cent. Risk factors for death have hitherto not been identified[15–17], but the present analysis suggests that splenomegaly and age above 40 years are major elements. Examination of the six major aetiological groups found that neoplastic disorders were also significantly associated with a fatal outcome. Atraumatic rupture of a normal spleen without aetiological factors (atraumatic–idiopathic splenic rupture) was, on the other hand, associated with a significantly decreased ASR-related mortality rate.

In the present study, initial treatment was total splenectomy (84·1 per cent), organ-preserving surgery (1·2 per cent) or conservative (14·7 per cent); 17 per cent of this last group eventually had a delayed splenectomy because of rebleeding. In comparison to patients with blunt abdominal trauma, the proportions undergoing organ-preserving surgery or non-surgical management are low[19]. Furthermore, there is a rather low success rate for non-operative treatment, and transcatheter arterial embolization of splenic lesions was uncommon. The present study appears to indicate that treatment guidelines for isolated splenic rupture secondary to blunt abdominal trauma cannot be simply applied to ASR; 93·0 per cent of the latter was associated with a pathologically altered spleen. Furthermore, patients with ASR are generally older than those with traumatic splenic rupture.

The choice of treatment is, just as for traumatic splenic injuries, determined by the haemodynamic stability, the amount of blood product used, the degree of haemoperitoneum and the extent of the splenic injury as described by the AAST classification[18,19]. For ASR, further consideration must be given to the underlying pathology, which may be limited to the spleen or have a systemic element. Patients with ASR of malignant aetiology should generally undergo immediate total splenectomy, although in some circumstances transcatheter arterial embolization may be used as a temporary stabilizing measure. Patients with ASR of non-malignant aetiology might be treatable

by organ-preserving surgery or a non-surgical approach, with or without transcatheter arterial embolization. It is self-evident that further risk factors, such as advanced age or anticoagulant treatment (for example, for a mechanical heart valve), need to be taken into consideration. Even if all preconditions for non-operative management are met, however, the relatively high failure rate must not be forgotten. Finally, there are patients with ASR of unknown aetiology for whom the standard treatment should probably be total splenectomy.

The principle of performing a total splenectomy, even in haemodynamically stable patients, can be justified for three reasons. First, histological examination of the spleen will establish the aetiology of the ASR as well as any underlying systemic disease[17]. Second, a significant number of malignant diseases may cause ASR, thereby prohibiting any organ-preserving approach. Third, the splenic function might already be compromised by a pathological alteration or infiltration of the splenic parenchyma, resulting in functional hyposplenism[20,21]. In such circumstances the removal of a non-functional spleen is justified and will not increase the risk of an overwhelming postsplenectomy infection.

The nomenclature of ASR has tended to be vague[1]. The most frequently used term in the literature is 'spontaneous rupture'. Unfortunately, some authors use it for all splenic ruptures that occur 'spontaneously', that is without adequate trauma, irrespective of the histopathological findings[15,22], whereas others use it as a synonym for 'idiopathic rupture', that is splenic rupture occurring without trauma in a histopathologically normal spleen[16,17,23]. This latter group may also be described as 'true spontaneous'. The second commonest term is 'pathological rupture', analogous to 'pathological fracture' in diseased bones, making a reasonable reference to the fact that ASR almost always occurs in a diseased spleen[16,17]. The term 'atraumatic rupture' is a logical correspondence to the much more frequently encountered condition of traumatic splenic rupture. Finally, there is the rare use of the term of 'occult rupture'. For some, this is synonymous with 'pathological rupture'[16,24], whereas for others it refers to a rupture with a preserved splenic capsule, resulting in a closed subcapsular haematoma without intra-abdominal bleeding[25,26].

To clarify the nomenclature, the authors propose a simplified classification of splenic rupture. A first distinction should be made between 'traumatic rupture' in the presence of an adequate trauma, and 'atraumatic rupture' without an adequate trauma. Atraumatic rupture in a pathologically altered spleen with increased fragility may occur spontaneously, without a triggering factor, or may be triggered by a minor physical event, such as sneezing, coughing, vomiting, straining during defaecation or muscular exertion[27,28]. A further classification can be made, however, as 93·0 per cent of patients with ASR have histopathological changes within the spleen and can therefore be described as having 'atraumatic–pathological splenic rupture'. The remaining 7·0 per cent without an aetiological factor and with no histopathological changes in the spleen can be classified as 'atraumatic–idiopathic splenic rupture'; this diagnosis can be made only by exclusion. One might wonder, however, whether the latter group of patients really constitutes a distinct clinicopathological entity. It may be that they are misclassified as a result of an unrecognized trauma or an unrecognized pathological process within the spleen, the traces of which might have been destroyed during surgical removal of the organ or even by the rupture itself[16,17,23,29].

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SUPPORTING INFORMATION

Additional Supporting Information may be found in the online version of this article:

Appendix S1: References of Table 1.