Doppler Sonography of the Spleen

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Abstract

Although the spleen is highly vascularised, the rate of focal lesions is far less frequent than in other abdominal organs, such as the liver or kidneys, possibly due to protective factors from the phagocytic and immunological competence of the spleen. Common pathological findings like splenic cysts, infarction or trauma can be diagnosed by ultrasonography with a high sensitivity and specificity. Other pathologies of the spleen such as focal splenic lesions are often unspecific and need further diagnostic evaluation.

In this chapter on Doppler sonography of the spleen in infants and children, the main indications, normal findings and the most common splenic pathologies in children are described. Special subsections deal with the ultrasound assessment of splenic involvement in infectious diseases, benign and malignant focal lesions, vascular pathologies (splenic aneurysms, infarction, peliosis), congenital alterations (wandering spleen, splenunculi, polysplenia) and trauma.

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Abbreviations

CDS	Colour-coded Doppler sonography
CEUS	Contrast-enhanced US
DS	Doppler sonography
IVC	Inferior vena cava
PH	Portal hypertension
PRF	Pulse repetition frequency (synonym: pulse repetition rate, velocity range, scale)
PW-DS	Pulsed-wave Doppler sonography
RI	Resistive index (synonym: resistance index, Pourcelot index)
TAV	Time average velocity (cm/s)
US	Ultrasonography
Ved	End-diastolic velocity
Vs	Peak systolic velocity

7.1 Introduction

The spleen may be affected in a wide variety of diseases in children. Nevertheless, it is an often neglected organ in diagnostic imaging as primary splenic lesions are rare.

Focal and diffuse alterations of the splenic parenchyma rarely originate from splenic diseases, but represent mostly secondary involvement of the spleen in systemic diseases. The spleen is the largest single lymphatic organ of the body. It has a diaphragmatic and a visceral surface.

Within the spleen two different functional and morphological systems are combined: the red pulp and the white pulp. The red pulp is building extensive vascular sinusoids with phagocytic, haematopoietic and, in humans less importantly, blood pooling functions. The white pulp is consisting of lymph follicles with immunological activities. In newborns and early infancy, the red pulp predominates. Later, the white pulp increases with the child's growth and immunological stimuli. However, with ultrasonography (US), no differentiation of red and white pulp is possible.

Although the spleen is highly vascularised, the rate of focal lesions is far less frequent than in other abdominal organs such as the liver or kidneys, possibly due to protective factors from the phagocytic and immunological competence of the spleen. Common pathological findings such as splenic cysts, infarction or trauma can be diagnosed by ultrasonography with a high sensitivity and specificity. Other pathologies of the spleen such as focal splenic lesions are often unspecific and need further diagnostic evaluation (Bachmann and Görg 2004; Sivit and Siegel 2002).

7.2 Normal Findings

7.2.1 Embryology and Vascular Anatomy of the Spleen

The spleen originates from the dorsal mesogastrium in the fifth embryonic week and is lobulated in embryonic life. It is protected by the 9th through the 11th rib. The visceral peritoneum forms the splenic capsule and adheres to the gastrosplenic and splenorenal ligaments as well as to the greater omentum. The splenic artery is the biggest branch (diameter up to 0.5 cm in adolescents and adults) (Fig. 7.1) (Görg 2011) of the coeliac trunk. The splenic artery can be displayed by colour-coded Doppler sonography along its entire, in older patients often serpentinous, course (Fig. 7.2a, b). Branches of the splenic artery supply the corpus and tail of the pancreas (Aa. pancreaticae magnae and dorsales), the fundus and great curvature of the stomach and the major omentum until the splenic artery reaches the spleen. Here, the splenic artery divides into several branches at the splenic hilus and divides within the spleen into segmental, subsegmental and subsubsegmental arteries, which extend into the splenic capsule (Fig. 7.3).

The splenic vein drains the blood from the spleen in 2–6 branches at the hilus and forms, together with the superior mesenteric vein, the portal vein at the splenoportal confluence. The inferior mesenteric vein is the biggest vein which drains into the splenic vein (variance of 30 % into the superior mesenteric vein). Furthermore,

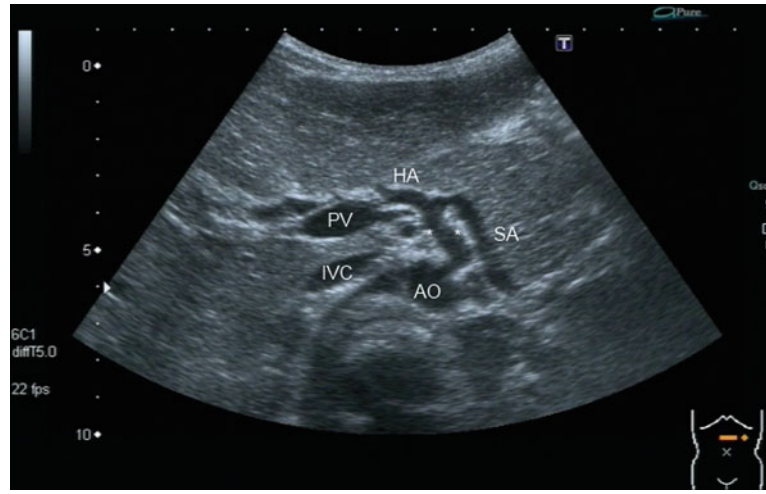


Fig. 7.1 Normal anatomy of the coeliac trunk and its branches. The transverse section through the upper abdomen shows the aorta, coeliac trunk and its main branches, the hepatic and the splenic artery as indicated. *AO* aorta, *IVC* inferior vena cava, *HA* hepatic artery, *SA* splenic artery, *PV* portal vein. The *markers* (*) are set at the coeliac trunk

the left gastric vein, the small gastric veins (Vv. gastricae breves) and the gastroepiploic veins drain into the splenic vein. The inflow of the inferior mesenteric vein into the proximal pancreatic part of the splenic vein can only rarely be visualised by ultrasonography.

7.2.2 Ultrasonographic Approach

The easiest approach for visualisation of the splenic vessels is a transverse section through the upper abdomen. The artery runs at the superior border of the pancreas, usually localised more cranially than the vein. The middle portion of the artery can also be shown in longitudinal sections through the upper abdomen with the transducer slightly angled to the left side (Fig. 7.4a–f). The artery is always neighboured by the splenic vein, which can be imaged behind the pancreatic corpus and tail in transverse and longitudinal sections (Fig. 7.5a–d). Using B-mode ultrasonography (US) alone, both vessels cannot be exactly distinguished. With the help of the different Doppler techniques (colour or spectral Doppler), however, artery and vein can easily be differentiated. From the left lateral intercostal approach, the splenic hilus and the intrasplenic vessels can be demonstrated (Fig. 7.6a, b).

The investigation of the spleen can be performed non-fasting and without special preparation in the supine position or lying on the right side.

Transducers: The appropriate transducer frequency has to be chosen depending on the age/size of the patient and the indication. In neonates, infants and for the detection of smaller focal lesions, high-resolution linear transducers up to 15 MHz are optimal (Fig. 7.7a, b). Older children, especially obese adolescents, are best investigated with a curved array transducer <6 MHz, followed by an additional approach with a high-frequency transducer in order to detect smaller lesions, discontinuation of the splenic surface in cases of trauma and superficial minor vascular pathologies, which may not be detected with low-frequency curved array scanning (Fig. 7.9).

7.2.2.1 Colour and Pulsed Doppler Sonography (CDS and PW-DS)

Using colour-coded Doppler sonography (CDS) in a transverse mid-abdominal section, the flow in the splenic artery behind the pancreas can be displayed blue as the flow is directed away from the transducer (Fig. 7.4b, d). This translates into a negative flow in the splenic artery in *pulsed waved* (*synonym*: *spectral*) *Doppler sonography* (*PW-DS*) behind the pancreatic body (Fig. 7.4c,

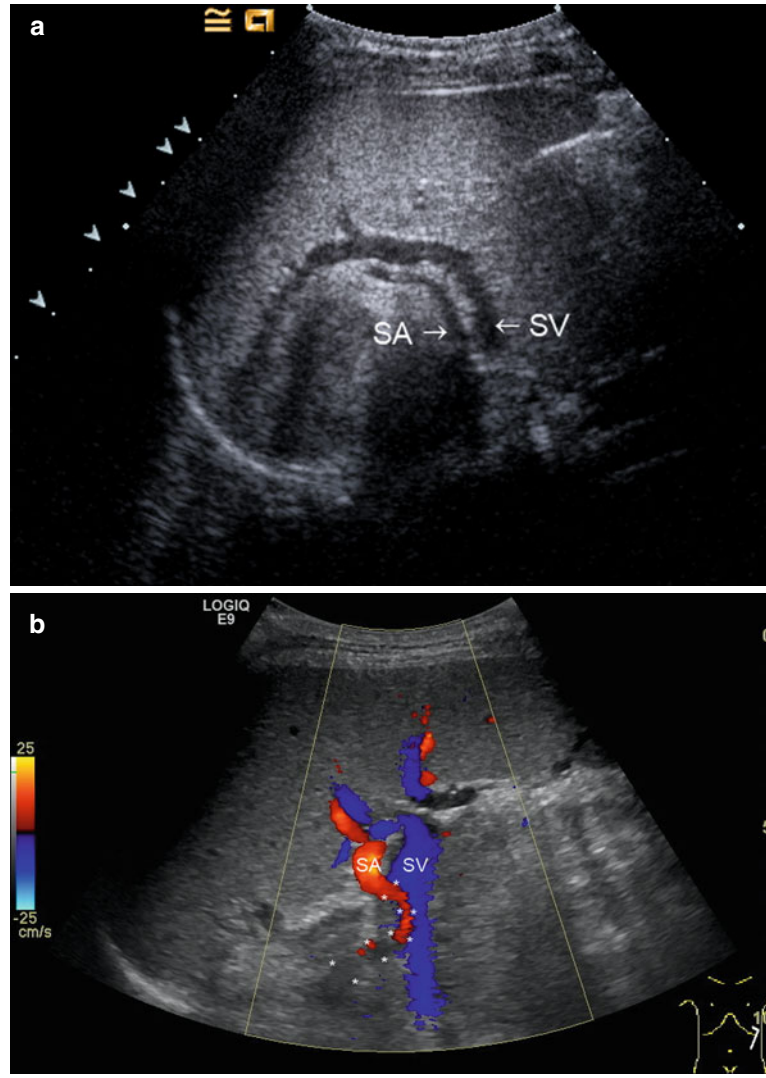


Fig. 7.2 (**a**) The splenic artery (*SA*) is located more cranially than the splenic vein (*SV*). Along the course of the splenic vein, the tail of the pancreas can be visualised. Oblique intercostal section through the left lateral upper abdomen. (**b**) Colour-coded Doppler sonography of the vascular supply of the spleen. Slightly serpentinous course of the splenic artery (*red*) in a 17-year-old adolescent with primary biliary sclerosis (the course of the splenic artery (*SA*) is marked with *). The splenic vein (*SV*) is displayed blue. Oblique intercostal section through the left lateral upper abdomen

f) and a positive flow after the artery has passed the pancreatic tail (Fig. 7.4e).

In the splenic vein, however, the flow behind the body of the pancreas is directed towards the transducer and is therefore displayed red (Fig. 7.5b). Pulsed Doppler shows a forward flow displayed above the baseline (Fig. 7.5c). Similar to the flow in the portal vein, a nearly continuous positive flow with small undulations can be seen (Figs. 7.5c and 7.8).

The *main indications* for Doppler sonography (DS) of the spleen are listed in Table 7.1.

7.3 Splenic Trauma

The spleen is often injured in accidents, although it is shielded by the ribs of the left hemithorax, which usually protects the spleen from laceration. Enlarged spleens which tower below the

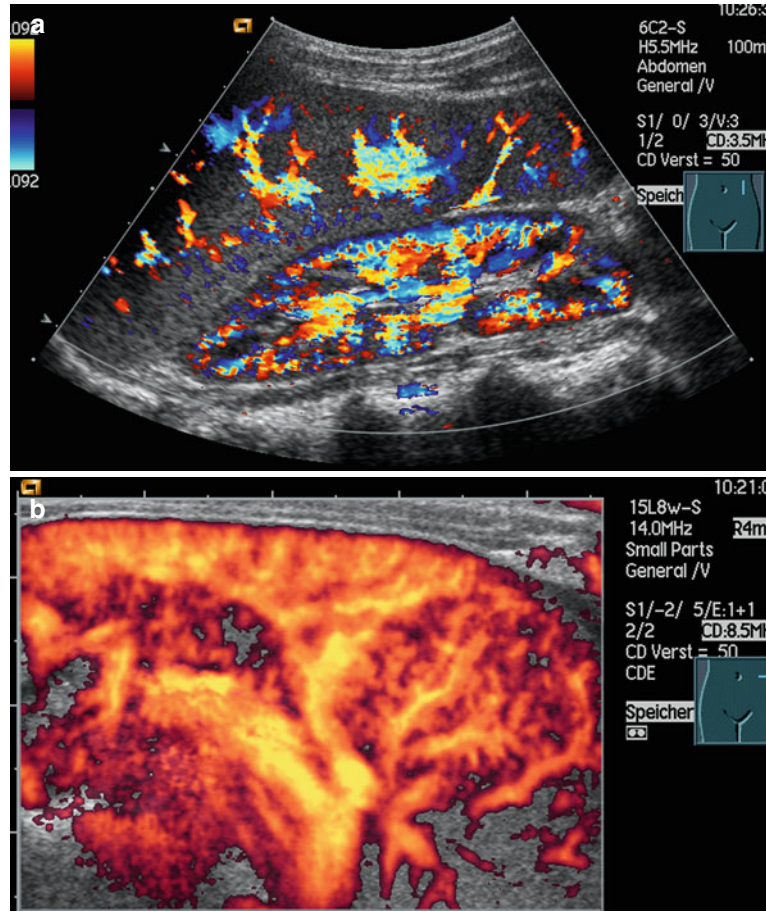


Fig. 7.3 Colour (**a**) and power (**b**) Doppler of the spleen. The splenic artery divides within the spleen into segmental, subsegmental and subsubsegmental arteries, which extend to the splenic capsule. Longitudinal (**a**) and transverse (**b**) section through the spleen

costal arch are extremely vulnerable. In blunt abdominal trauma, injury of the spleen must always be ruled out as the spleen is the most common injured abdominal organ accounting for up to 45 % of all visceral injuries (Tataria et al. 2007). An enlarged spleen is far more at risk of being injured after blunt abdominal trauma. Sonographic signs of possible spleen injury are free abdominal fluid with internal echoes, splenomegaly, reduced respiratory movement, inhomogeneous texture of the spleen, subcapsular haematoma, anechoic lacerations within the parenchyma or disruption of the splenic capsule (Hofmann 2005) (Figs. 7.9a–d and 7.10a, b). In many emergency rooms, a FAST (*f*ocal *a*bdominal *s*onography for *t*rauma) protocol with scanning of the four abdominal quadrants for free fluid, as a sign for haemoperitoneum, is done and later followed by a complete and detailed US investigation after deciding on surgical or conservative therapy for a trauma patient. However, in a study by Richards in 2002, 12 % of children with abdominal injury had no free fluid. Contusions, lacerations, ruptures, fractures, subcapsular and perisplenic haematoma (Figs. 7.9b and 7.10a) of the spleen may be distinguished. Haemoperitoneum and active bleeding are the most likely indications for surgical intervention. Due to the thicker splenic capsule in children, operations in splenic trauma are less frequently compared to adults. In paediatric patients, haemodynamic stability is the

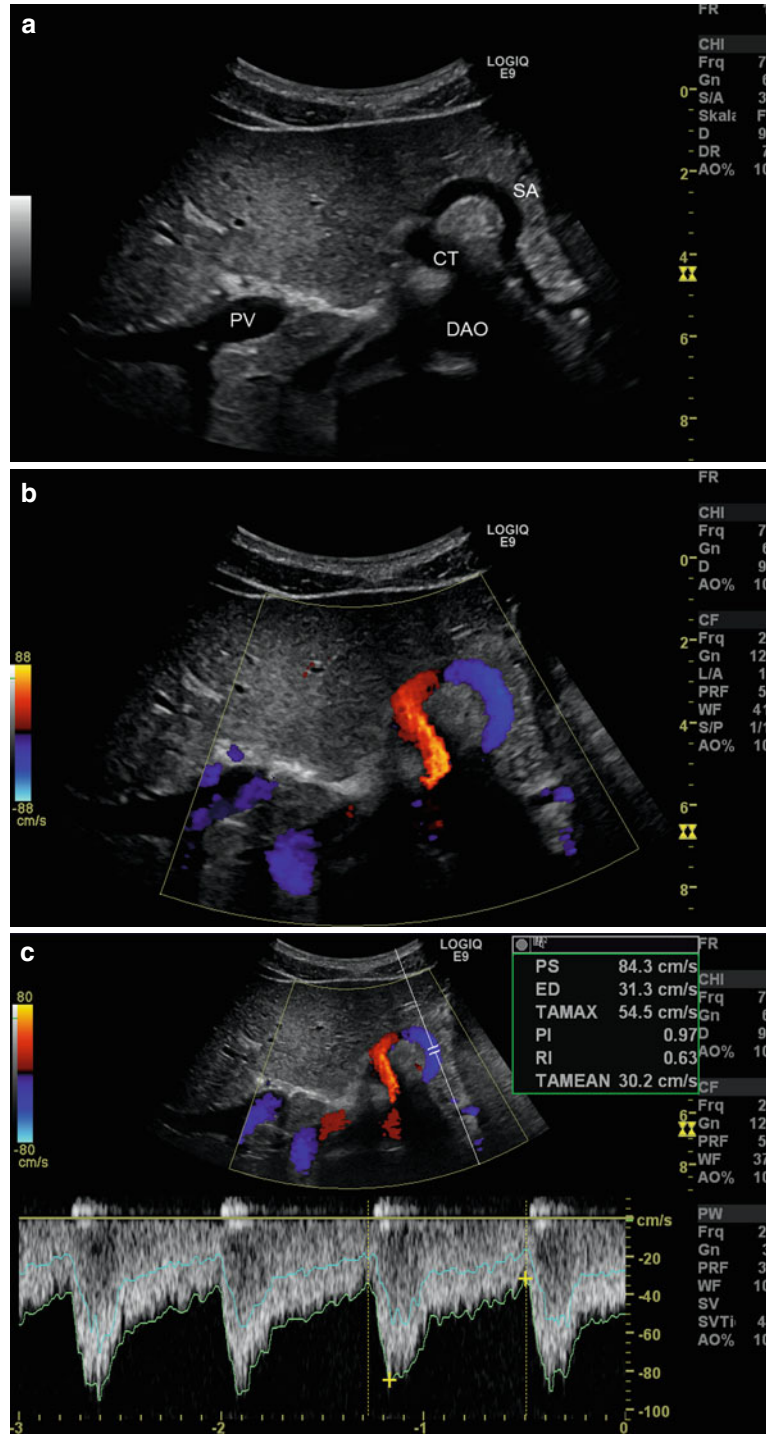
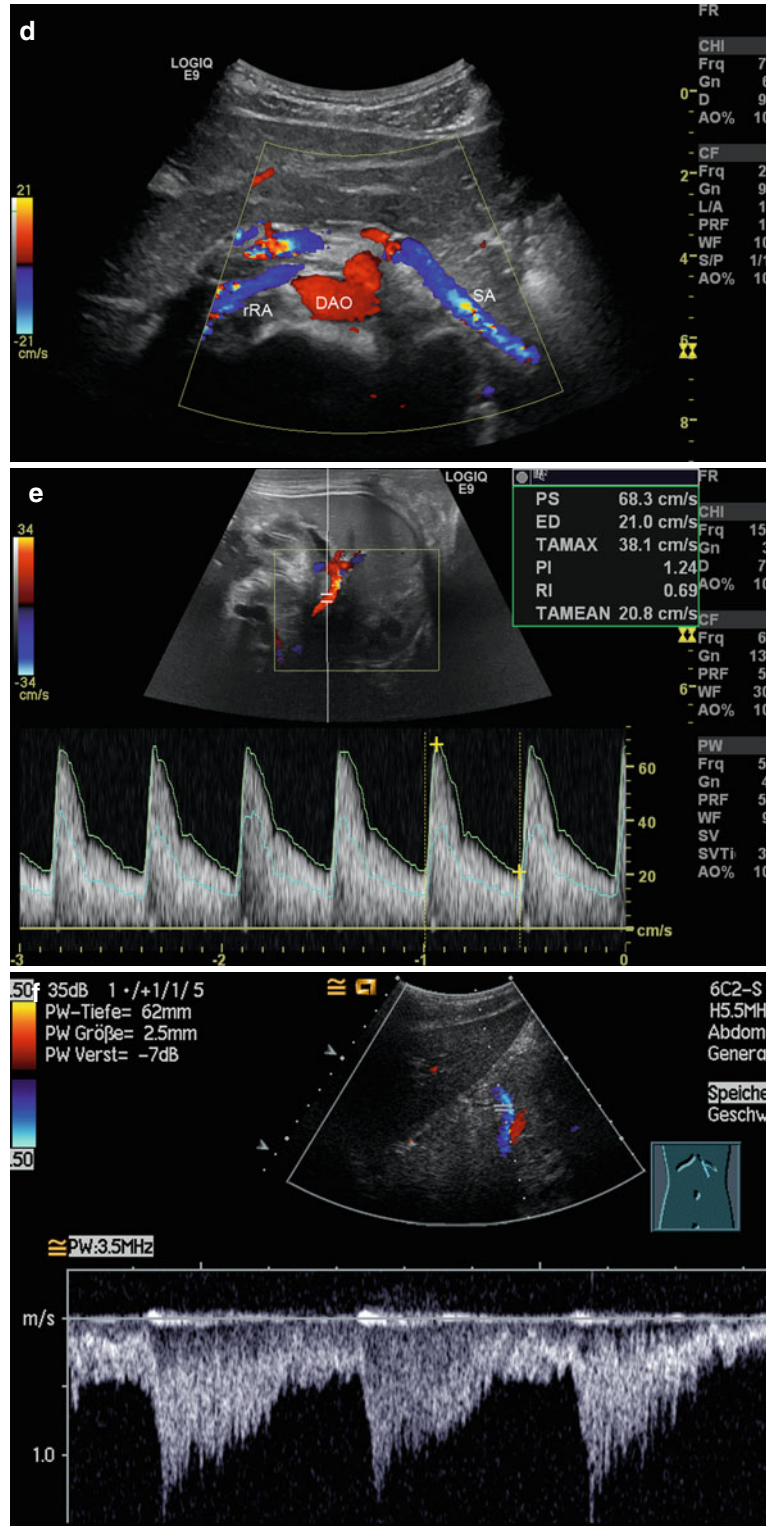


Fig. 7.4 Course of the splenic artery from its origin from the coeliac trunk (*CT*) to the splenic hilus. *SA* splenic artery, *PV* portal vein, *DAO* descending aorta. Cross section. (**a**) 2D image of the proximal splenic artery in a 16-year-old boy. (**b**) Colour Doppler: the coeliac trunk is displayed *red*, the splenic artery blue. (**c**) Spectral Doppler of the flow in the proximal splenic artery shows a systolic-diastolic forward flow which is displayed below the baseline, as the flow is directed away from the transducer. (**d**) Colour Doppler of the flow in the splenic artery (*SA*) in a transverse section through the middle upper abdomen. *rRA* right renal artery, *DAO* descending aorta. (**e**) Spectral Doppler of the flow in the distal splenic artery at the splenic hilus. The flow curve is displayed above the baseline, as the flow is directed towards the transducer and the spleen. (**f**) The middle portion of the splenic artery can also be shown in longitudinal sections with the transducer slightly angled to the left side. The flow in the splenic artery in this section is directed away from the transducer and therefore displayed blue and depicted below the baseline in the PW-Doppler. The PW-Doppler shows a systolic-diastolic forward flow due to low peripheral resistance

Fig. 7.4 (continued)



most important determinant for conservative treatment. Following trauma, splenic healing can rapidly be observed. This was experienced in the last decades of predominately successful conservative management of splenic injuries. Because of devastating post-splenectomy infections with encapsulated bacteria such as *Streptococcus pneumoniae*, *Neisseria meningitidis* and *Haemophilus influenzae*, splenectomy should be avoided whenever possible.

Long-term sequelae of splenic injury may be scarring of parts of the spleen, development of posttraumatic pseudocysts, polysplenia and calcification (Fig. 7.11). In 15–30 % of patients, a

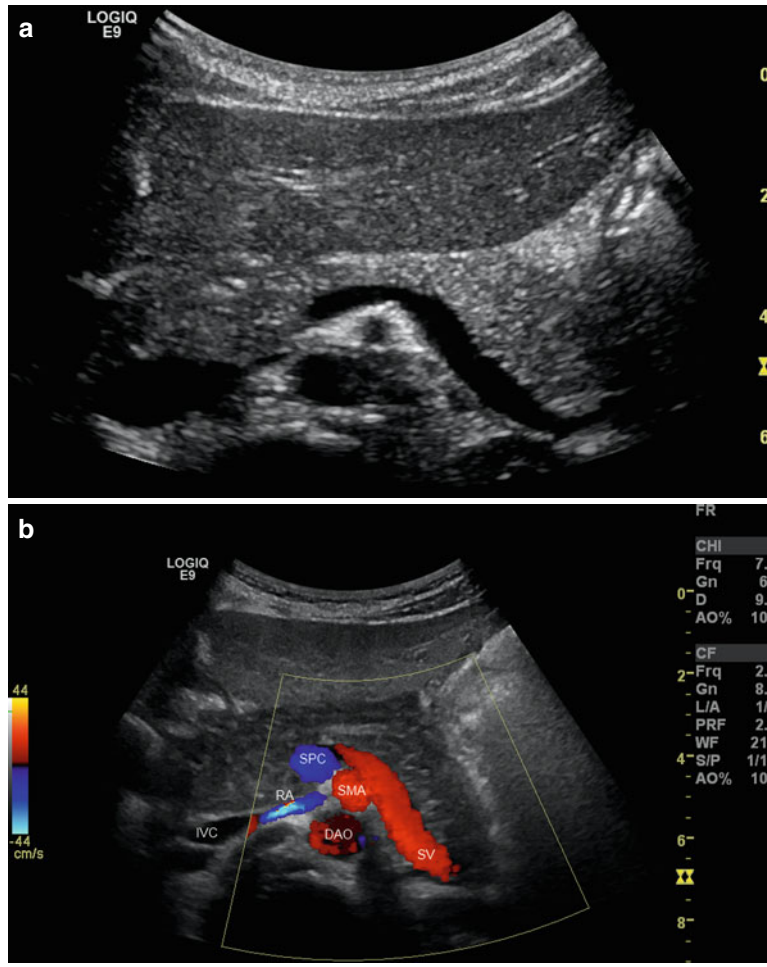
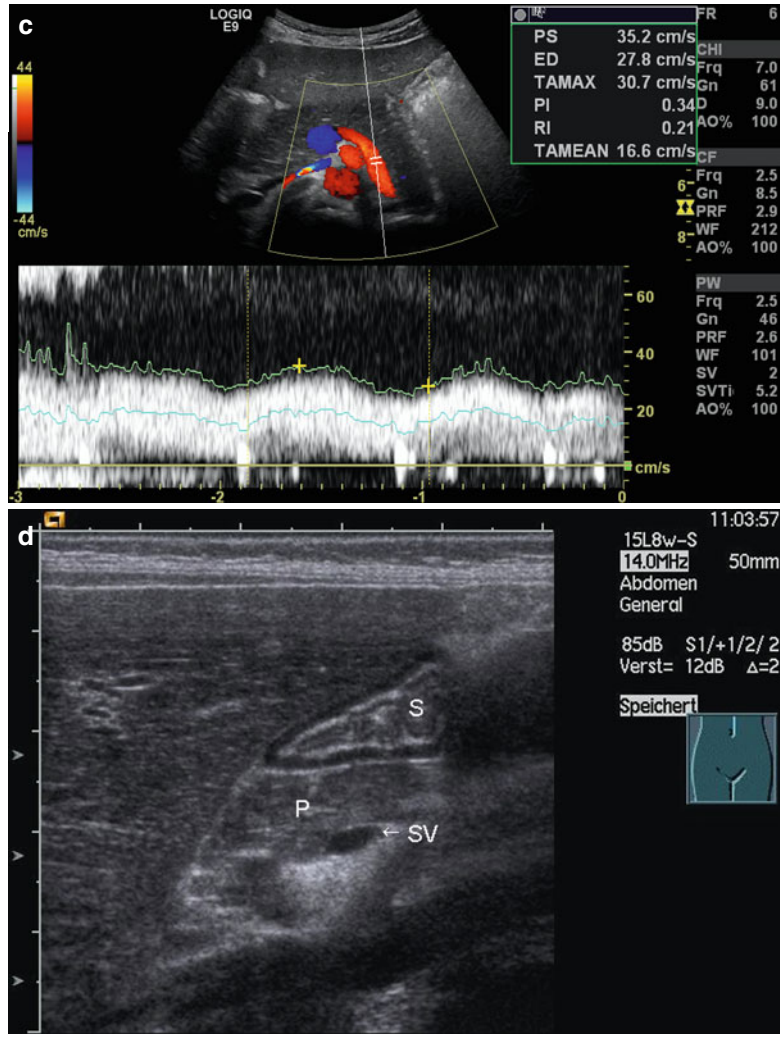


Fig. 7.5 (**a**) The splenic vein can be imaged behind the body and tail of the pancreas in B-mode. Transverse section through the upper abdomen. (**b**) Colour Doppler of the flow in the splenic vein in a transverse section through the upper abdomen. The flow in the splenic vein (*SV*) along the pancreatic tail is directed towards the transducer and therefore displayed *red*. Close to the pancreatic head, the flow in the splenoportal confluence (*SPC*) is directed away from the transducer and therefore displayed blue. The aorta (*DAO*), the inferior vena cava (*IVC*), right renal artery (*RA*) and the superior mesenteric artery (*SMA*) are shown. (**c**) Spectral Doppler of the flow in the splenic vein shows an antegrade flow with a time average maximal velocity (TAMAX) of 30.7 cm/s and a mean time average velocity (TAMEAN) of 16.6 cm/s. Transverse section through the upper abdomen. (**d**) Longitudinal section through the middle upper abdomen shows the pancreatic body (*P*) and the splenic vein (*SV*). The stomach is marked with "S"

Fig. 7.5 (continued)



two-phase delayed splenic rupture may be expected within 2 weeks. Delayed complications, such as splenic abscesses and pseudoaneurysms of the splenic artery and its branches, have been observed (Lynn et al. 2009). To detect complications, a short follow-up by B-mode sonography and CDS should be scheduled (Goletti et al. 1996). In cases of suspected splenic injury, CDS with low flow settings and a low wall filter should be used (Fig. 7.12a). If a high-frequency curved array probe is used, power Doppler is especially helpful for the diagnosis of focal hypoperfusion of the organ (Fig. 7.12b). Areas without perfusion can be distinguished from the surrounding normal vascularised parenchyma. Lacerations are displayed as focal hypoechoic lesions. Power Doppler reveals missing perfusion, often in a triangular shape. Acute posttraumatic injury and lacerations of the spleen may not be visualised with B-mode due to diffuse swelling of the organ. In the detection of traumatic splenic injury, CEUS (contrast-enhanced ultrasonography) is much more sensitive than mere B-mode and Doppler US alone (see Sect. 7.8, Fig. 7.13; Manetta et al. 2009; Weskott 2013).

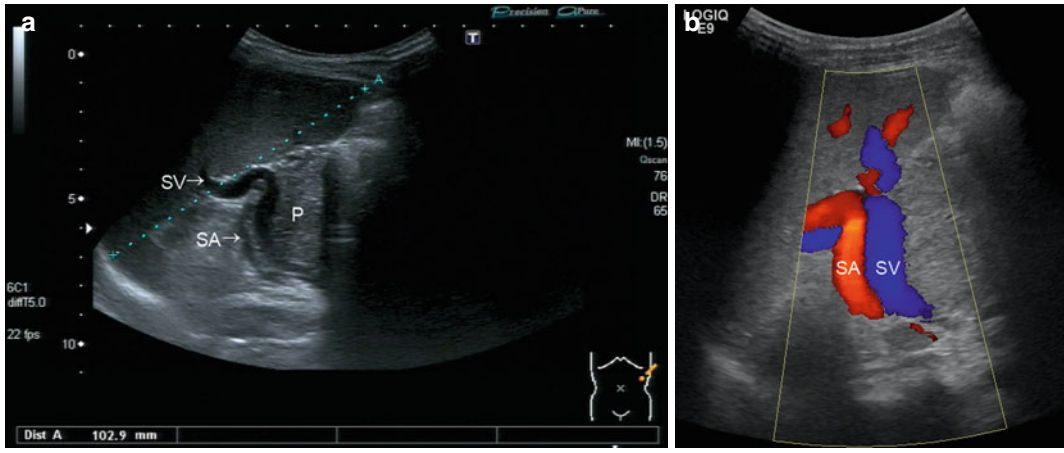


Fig. 7.6 (**a**) B-mode section through the splenic hilus. The length of the spleen is measured. The tail of the pancreas can be visualised caudally to the splenic vein (*P*). The splenic artery (*SA*) is smaller in size and located more cranially than the splenic vein (*SV*). Left lateral intercostal approach. (**b**) Colour Doppler of the flow in the splenic hilus. The flow in the splenic vein (*SV*) is directed away from the transducer and coded in blue; the flow direction in the splenic artery (*SA*) is towards the transducer and therefore coded in *red* colour. Left lateral intercostal approach

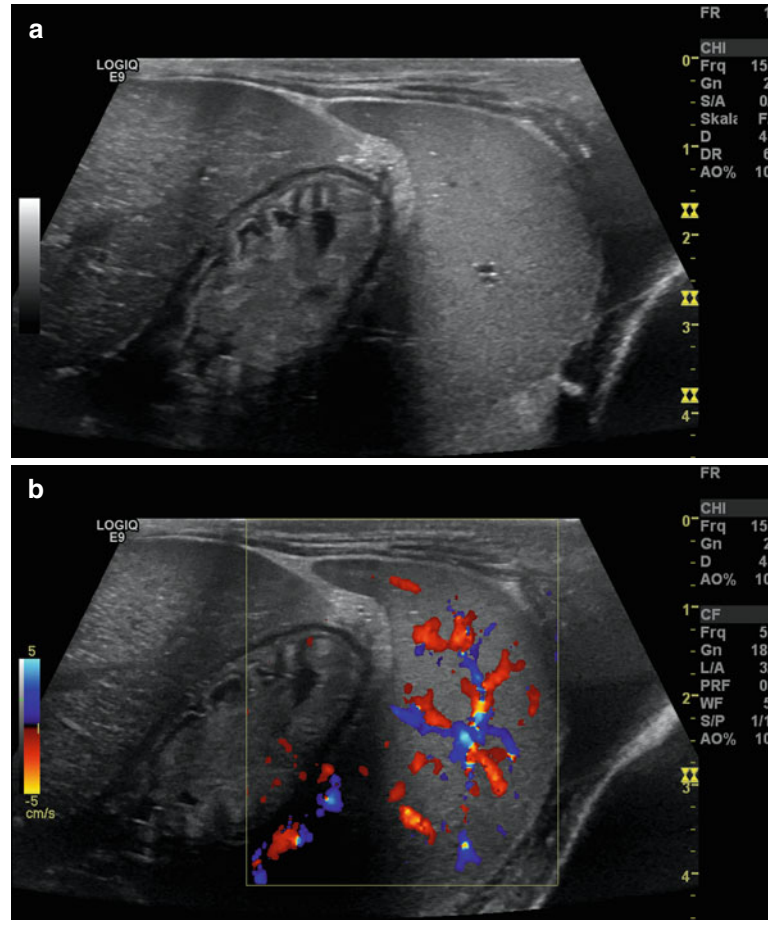


Fig. 7.7 Newborn with hepatosplenomegaly due to myelodysplastic syndrome. In newborns and infants and for the detection of smaller focal lesions, high-resolution linear transducers up to 17 MHz are optimal for visualisation of the parenchyma. (**a**) "Kissing phenomenon" of the liver and spleen. The stomach is in between. B-mode image. Transverse section through the spleen. (**b**) Colour Doppler image of the splenic perfusion. Transverse section through the spleen

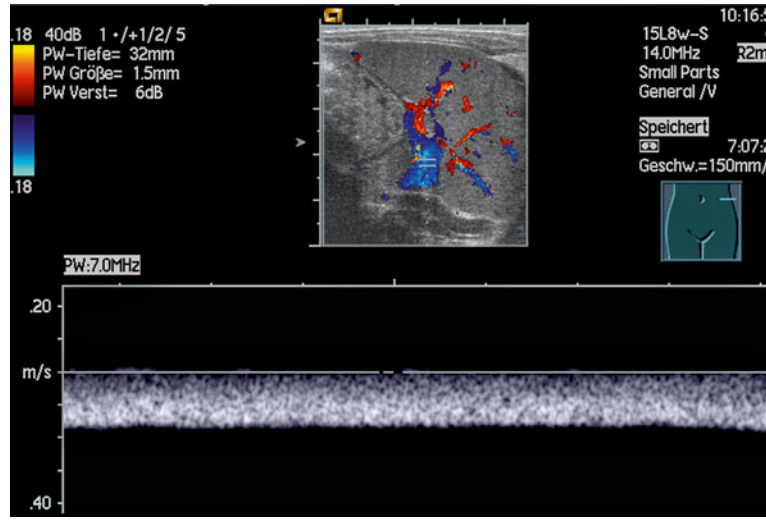


Fig. 7.8 Pulsed-wave Doppler sonography in the splenic vein with a laminar flow profile. Transverse section through the spleen

Table 7.1 Main indications for Doppler sonography (DS) of the spleen

Differential diagnosis of splenomegaly (acute and chronic infections, haematological and immunological diseases, portal hypertension, storage diseases)
Differential diagnosis of reduced splenic size (hyposplenia/asplenia)
Diffuse alterations of the spleen (diffuse benign or malign infiltration, systemic inflammatory or infectious diseases)
Vascular alterations (thrombosis, infarction, aneurysm)
Trauma
Focal lesions of the spleen

7.4 Focal Lesions of the Spleen

7.4.1 Introduction

Focal lesions of the spleen are rare (0.2 %, Bachmann and Görg 2005; Chen 2005; Görg 2011). Bachmann et al. described 98 adult patients with focal lesions and classified them as avascular, hypovascular, isovascular, hypervascular and arteriovenous "high flow", using the surrounding splenic tissue as in vivo reference. Most of the focal splenic lesions (68.4 %) were avascular, 15.3 % appeared hypovascular, 8.2 % isovascular, 5.1 % hypervascular and in 3.1 % an arteriovenous "high-flow" pattern was found. Alongside the diagnosis of intrasplenic pseudoaneurysm, the practical utility of CDS in differentially diagnosing focal spleen lesions is low (Bachmann and Görg 2005). However, US has a role in the detection and follow-up of focal lesions. Cystic and solid lesions are distinguishable in B-mode scanning.

7.4.2 Benign Splenic Lesions

7.4.2.1 Cystic Lesions

Cystic lesions are most commonly *dysontogenetic* (*simple cysts*) followed by non-epithelialised *pseudocysts* (*synonym*: *secondary cyst*) and *parasitic cysts* (hydatid cyst, usually caused by *Echinococcus granulosus*).

Congenital, dysontogenetic cysts (synonyms: simple cyst, real cyst, epidermoidal cyst, primary cyst) are usually anechoic, round and solitary. They usually show a hyperechogenic epithelial lining at their borders to the splenic parenchyma (Fig. 7.14a, b), a dorsal acoustic enhancement and only occasionally thin septa.

Secondary cysts are diagnosed following trauma, infection or infarction. They are mostly not completely round; multiple and intraluminal septa are present more frequently. Doppler sonography shows no signals within cysts (Fig. 7.15a, b). The difference between a primary and a secondary cyst depends mainly on the presence of an epithelial wall surrounding the cyst. The differentiation is not always reliable with US.

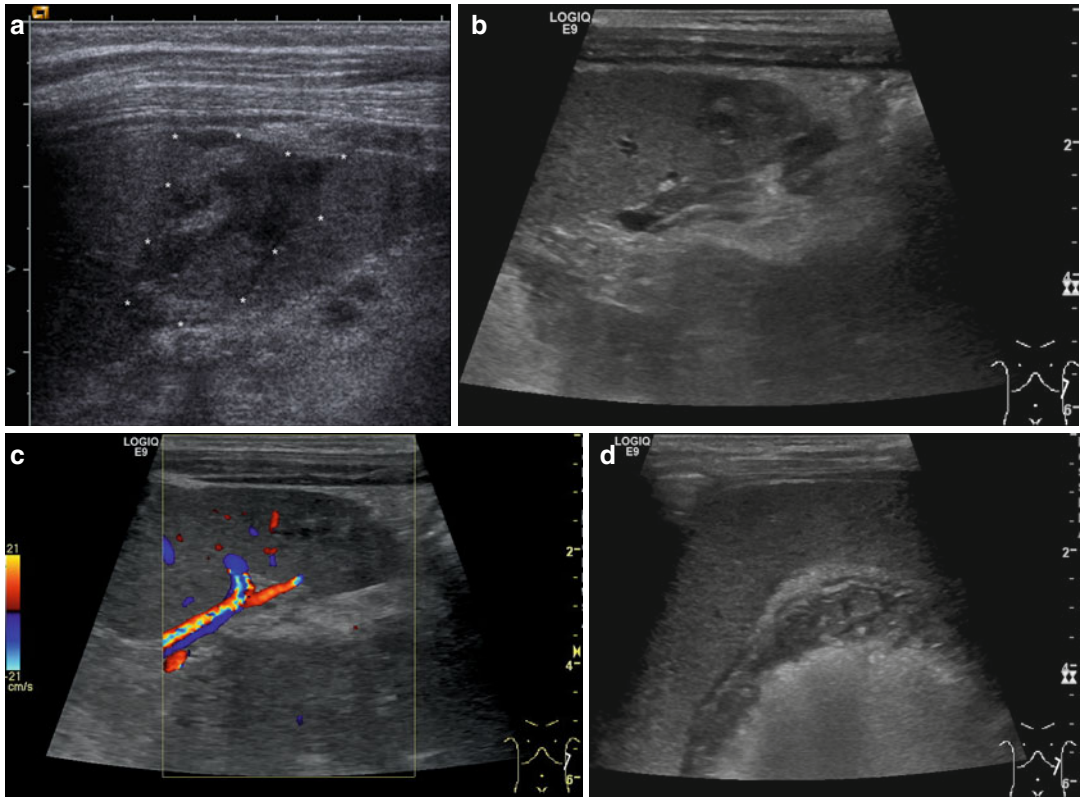


Fig. 7.9 (**a**) Splenic rupture in a child with a traffic accident. The B-mode image shows a hypoechoic rectangular area marked with *. Transverse section through the spleen. (**b**) Contusion of the apical splenic area in an 8-year-old child after bicycle accident with blunt abdominal trauma. Around the apex of the spleen, a hyperechoic delineation of a perisplenal haematoma can be seen. (**c**) Same patient. CDS shows only the perfusion of major vessel branches in the central part of the spleen, but no perfusion within the region of contusion. (**d**) Same patient. An echoic rim of blood is located between the spleen and the stomach wall in a B-mode scan, oblique section

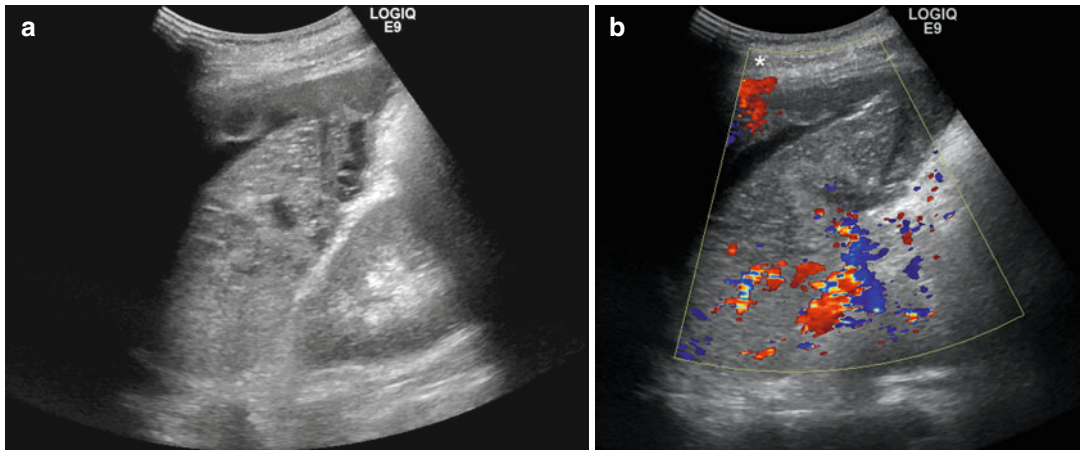


Fig. 7.10 Splenic trauma with a fracture through the lower pole and gross perisplenic haematoma. (**a**) B-mode. An echogenic band of blood is seen between spleen and kidney and echofree/echorich haematoma all around the splenic borders. Longitudinal section through the spleen. (**b**) CDS shows perfusion only in the dorsal parts of the spleen. CDS motion artefacts on the ventral side are marked with *. Longitudinal section through the spleen

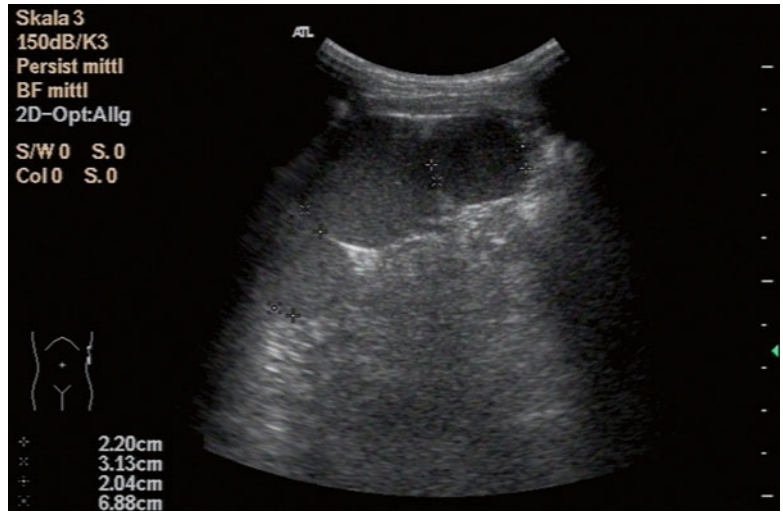


Fig. 7.11 Traumatic polysplenia. Four splenic residuals are left in this child after a traffic accident. Longitudinal section through the spleen

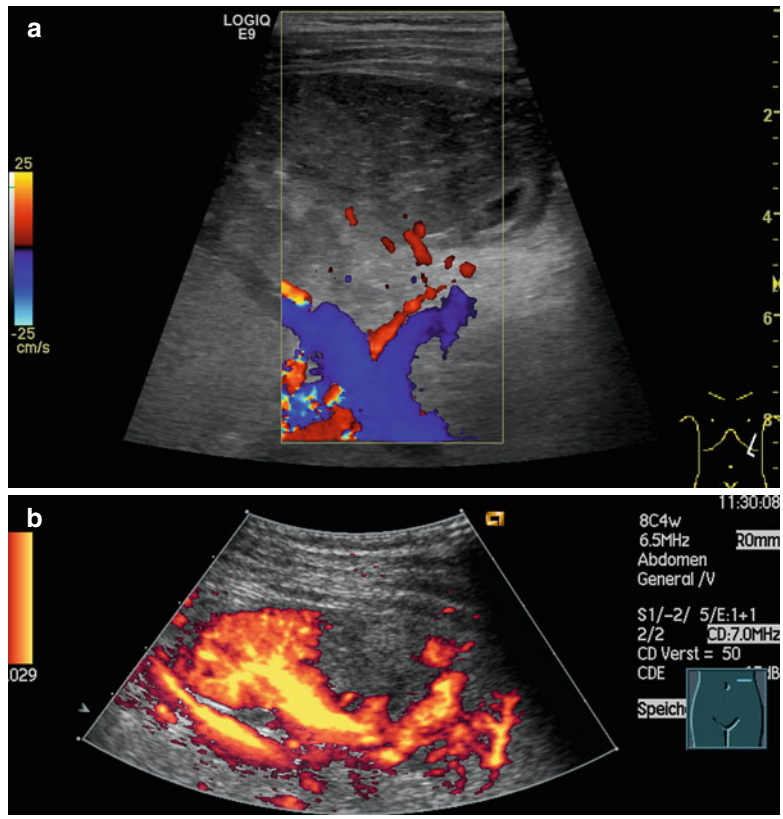


Fig. 7.12 Trauma of the spleen. (**a**) Due to diffuse haematoma and splenic contusion, the colour-coded Doppler sonogram shows no vessels in the lower pole of the spleen, intercostal oblique section. (**b**) Power Doppler reveals no perfusion in the traumatised triangular subcapsular splenic area. Transverse section through the spleen

The intracystic fluid of secondary cysts may have an increased echogenicity due to cholesterol crystals, inflammatory debris or haemorrhage. Parietal calcifications are more common in pseudocysts. Surgical enucleation of large cysts may be necessary, but procedures should always be organ preserving (Czauderna et al. 2006). Complications of nonparasitic splenic cysts include intracystic haemorrhage, rupture, peritonitis and hypersplenism (Czauderna et al. 2006).

Echinococcal parasitic infections have to be considered in the differential diagnosis of cystic

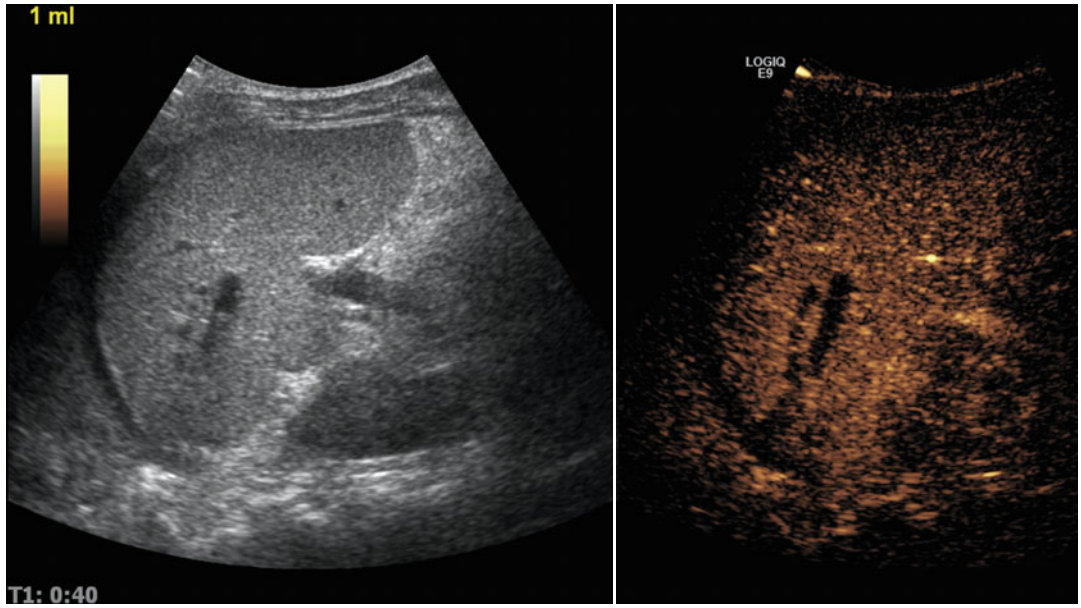


Fig. 7.13 Contrast-enhanced ultrasonography (CEUS) in an adolescent with two parallel lacerations of the spleen after a traffic accident. Subphrenic haematoma around the spleen. Intercostal oblique section

lesions of the spleen, although splenic involvement is rarer than hepatic or pulmonary manifestations (Fig. 7.16). Splenic echinococcal involvement is reported only in 0.9–8 % of patients, usually in infants with multi-organ disease.

Isolated splenic parasitic cysts are even rarer. In early stages of echinococcal infection, a hyperperfused rim may be seen with DS. Hydatid sand, daughter cysts and infolded membranes may be visible within the cyst, but they can also appear purely cystic or solid. In later stages, calcifications may occur (Dilli et al. 2011). Another cause of parasitic cysts is porcine tapeworm (Taenia solium) infections (Benter et al. 2011).

Lymphangiomas consist of multiple vascular canals filled with lymphatic fluid. Therefore, they have a hypo- or anechoic, pluriseptate appearance with possible debris or calcification inside (Fig. 7.17a, b). Usually, no vessels can be detected inside lymphangiomas. In rare cases however, vessels can be seen within the septae of a lymphangioma. Lymphangiomas grow slowly. They may appear single or multiple (Paterson et al. 1999), mostly with a multicystic, "honeycomb" appearance, rarely like a solid tumour or a solitary cyst. Unlike haemangiomas, the capsule and trabeculae of the spleen, where lymphatic tissue is concentrated, are often involved. The clinical manifestation may range from a small incidental lesion to a large multicystic abdominal mass requiring surgical intervention (Abbott et al. 2004). In *lymphangiomatosis*, multiple organs are involved, most often the liver, mediastinum, axilla, neck and retroperitoneum.

7.4.2.2 Solid Lesions

Solid lesions show echoes within the lesions and may show perfusion in CDS and pulsed Doppler sonography. In CDS two-thirds of focal splenic lesions appear to be avascular (Bachmann and Görg 2005). The importance of imaging solid splenic lesions is to differentiate them from malignant lesions. For all imaging modalities (US, CT, MRI, CEUS), a definite differential diagnosis of solid splenic focal lesions is difficult.

Although rare, the most frequent benign solid *splenic lesions* are *haemangiomas*.

Haemangiomas usually appear hyperechogenic and in CDS often without signals (Taibbi et al. 2012). Their appearance in the spleen is

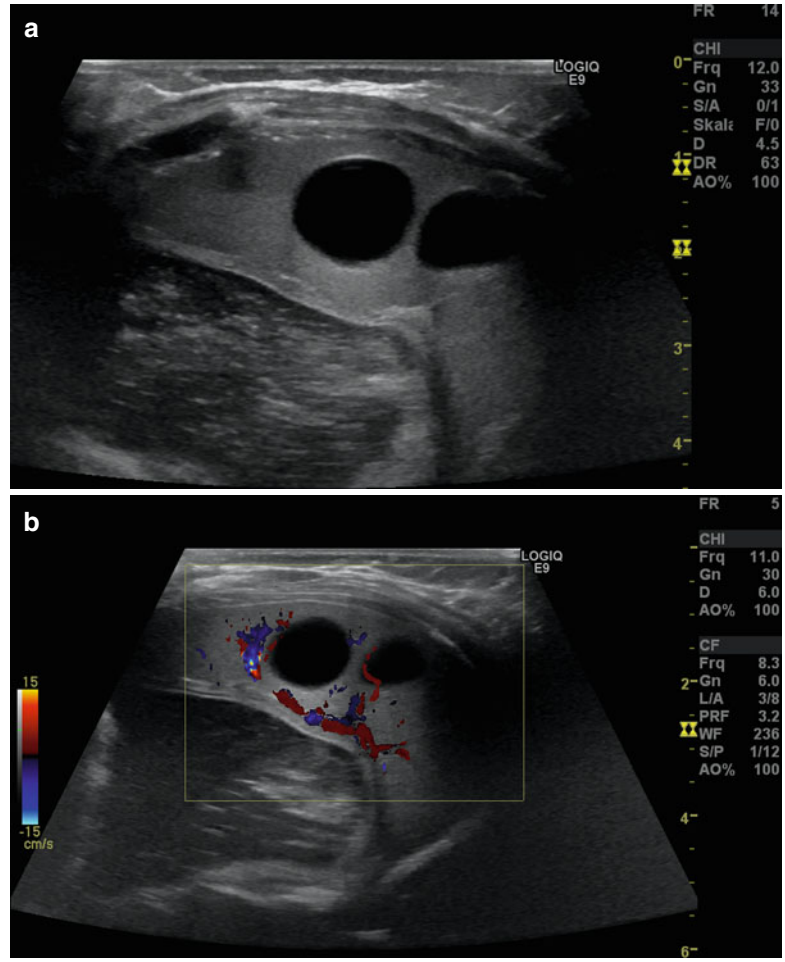


Fig. 7.14 Two simple cysts in a 5-week-old infant as an accidental finding (synonyms: dysontogenetic, simple, real, epidermoidal, primary cyst). (**a**) B-mode with a high-frequency 14 MHz probe. Transverse section. (**b**) Colour-coded Doppler sonogram (CDS) of the same patient. The two dysontogenetic cysts show no internal Doppler signal. Vessels can only be displayed in the normal neighbouring parenchyma. Transverse section

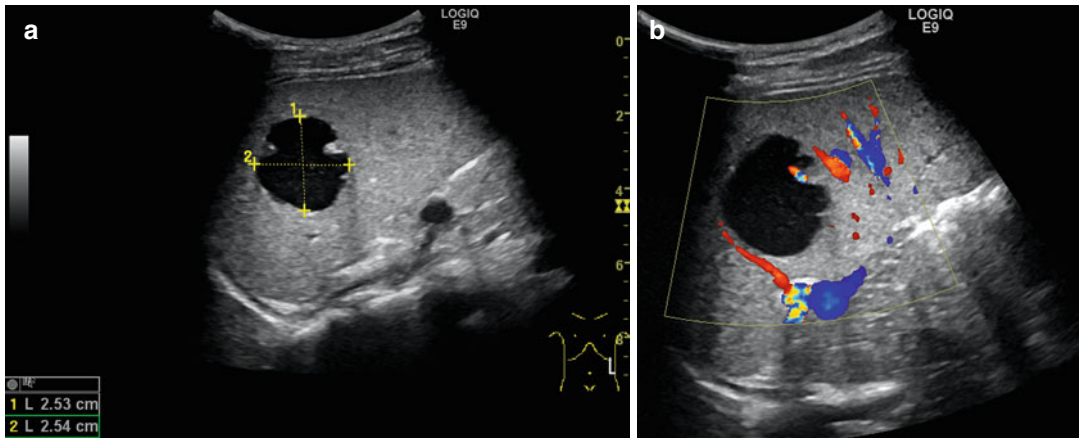


Fig. 7.15 An 8-year-old girl with upper abdominal pain and incidental finding of a splenic cyst. (**a**) B-mode US shows an irregularly delineated cyst measuring 2.5 cm in diameter. Fine echoes can be seen within the cyst and a small parenchymal tongue at the border. Longitudinal section. (**b**) Colour-coded Doppler sonogram (CDS) shows no signals within the cyst, but vessels can be displayed in the neighbouring parenchyma. Longitudinal section

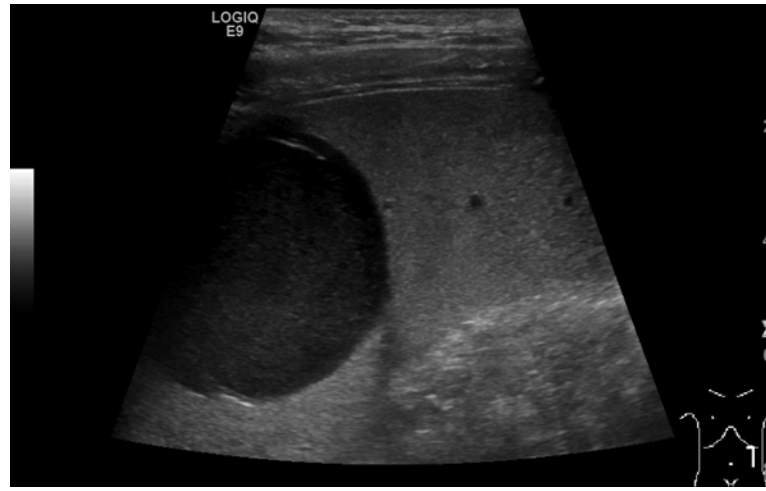


Fig. 7.16 Echinococcal cyst of the spleen (*E. granulosus*). The image shows multiple internal echoes, pseudomembranes and debris within the cyst. Longitudinal section

non-specific on B-mode and CDS. Depending on the vessel size inside the haemangioma, capillary and cavernous types can be distinguished. The cavernous type has a combination of solid and cystic components and appears more sponge-like. Haemangiomas may occur solitary or multiple and are found more frequently in association with Klippel-Trénaunay-Weber, Turner or Beckwith-Wiedemann syndromes. *Splenic haemangiomas* are usually asymptomatic, but complications include bleeding, rupture and Kasabach-Merritt syndrome, a life-threatening consumptive coagulopathy due to disseminated intravascular coagulation starting in the abnormal haemangioma vessels with thrombocytopenia and anaemia (Peddhu et al. 2004). Most lesions are small and found incidentally in asymptomatic patients (Abbott et al. 2004); calcifications may occur.

CDS may reveal flow within the haemangioma; however, splenic haemangiomas often do not show the typical rim enhancement with centripetal inflow into the centre of the lesion as seen in hepatic haemangiomas especially in contrast-enhanced ultrasonography (CEUS) (Taibbi et al. 2012). *Capillary haemangiomas* and *hamartomas* seem to be the most likely diagnosis in hyperechogenic lesions, but in all cases a careful ultrasonographic follow-up is warranted (Görg et al. 2006). Even pathological differentiation of hamartomas from haemangiomas may be difficult in the individual case (Abbott et al. 2004).

Hamartomas (*splenoma*, *splenadenoma or* "*FNH of the spleen*") are non-neoplastic lesions, which are composed of different splenic components apart from the white pulp. The *hamartoma/splenoma* was first described by the pathologist von Rokitansky in 1861. It consists of irregular red pulp with sinusoidal-like vessel structures and fibrous tissue (Günter 2005; Hartmann et al. 2008; Abramowsky et al. 2004), therefore also called "focal nodular hyperplasia (FNH) of the spleen". They are typically solitary and asymptomatic; calcifications can be found. Their size varies up to 20 cm (Hartmann et al. 2008) and splenomegaly is usually present. CDS may depict hypervascularity with multiple intralesional colour flow signals distributed in a radial fashion (Görg and Schwerk 1994 and Peddhu et al. 2004). Hamartomas are usually well-described nodular lesions, which tend to compress the adjacent parenchyma. Calcification secondary to ischaemia or haemorrhage is rare.

Mostly the discovery of a hamartoma/splenoma is incidental; in one case out of 170 described, a spontaneous rupture occurred (Günter 2005). Hamartomas are thought to be congenital in origin, and splenic hamartomas have been associated with further hamartomas in other organs such as in patients with tuberous sclerosis or Wiskott-Aldrich-like syndromes (Abbott et al. 2004). However, the aetiopathology remains unclear.

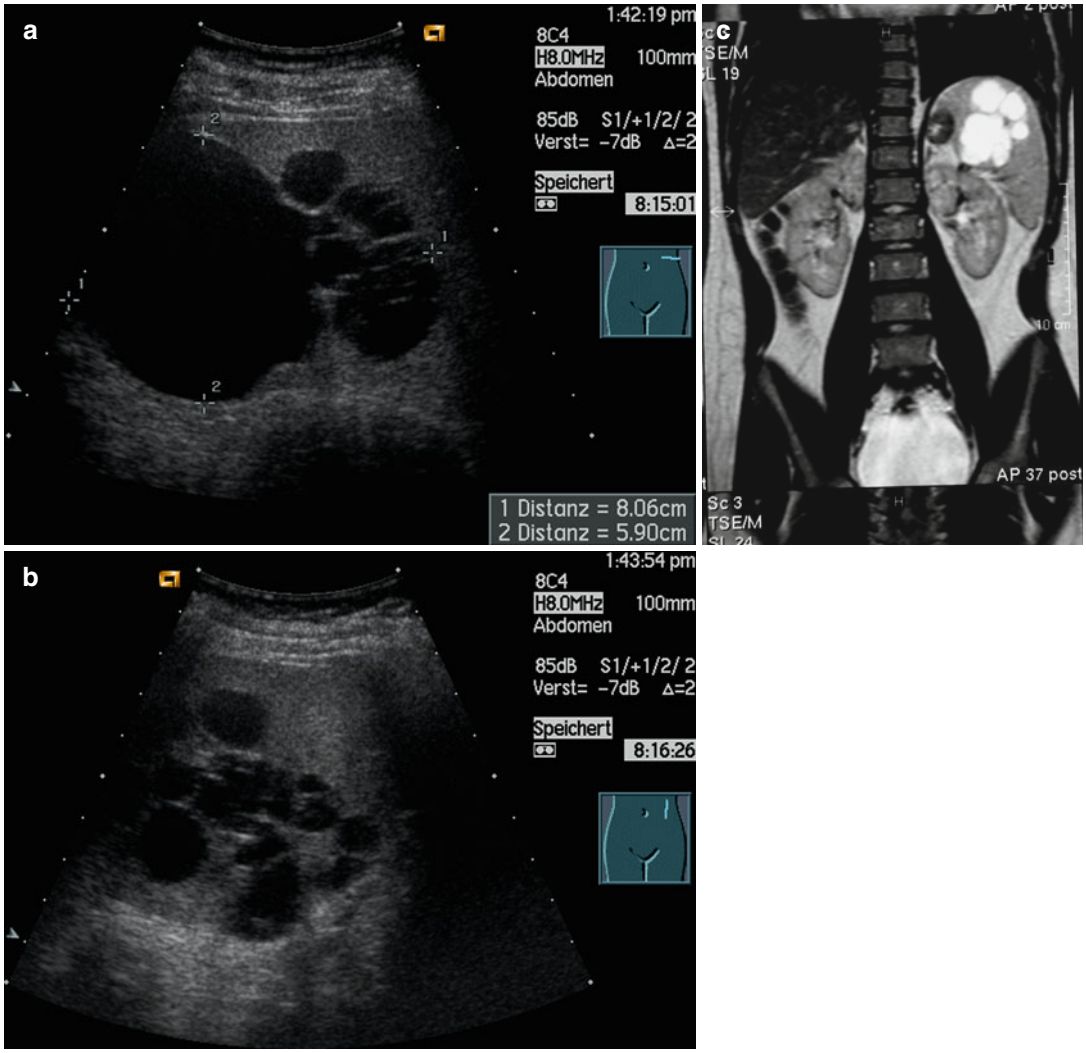


Fig. 7.17 Lymphangioma of the spleen. Lymphangiomas consist of multiple vascular canals filled with lymphatic fluid. (**a**) B-mode: multiple anechoic round lesions with thin septation. Usually no vessels can be detected inside the lymphangiomas. Transverse section. (**b**) Multiple cystic lesions which are separated by thin septations. Longitudinal section through the spleen. (**c**) Abdominal MRI of the same patient with lymphangioma of the spleen

Haemangioendotheliomas are benign vascular tumours with sometimes significant morbidity (organ enlargement, Kasabach-Merritt syndrome and congestive heart failure). The US appearance in the spleen is non-specific; they are mostly described as hypoechoic. Anechoic areas may represent intralesional necrosis compared to the normal splenic parenchyma. Haemangioendotheliomas are hypovascularised in CDS. Calcifications are not a specific feature.

More rare benign manifestations of focal splenic lesions are lipomas and angiomyolipomas (Fig. 7.18).

Littoral cell angioma of the spleen is a rare vascular tumour seen at any age with no gender predisposition. Originally, littoral cell angiomas were thought to be benign, but also malignant features have been described (Abbott et al. 2004). Splenomegaly is almost always present. Littoral cell angioma originates from the red pulp sinuses.

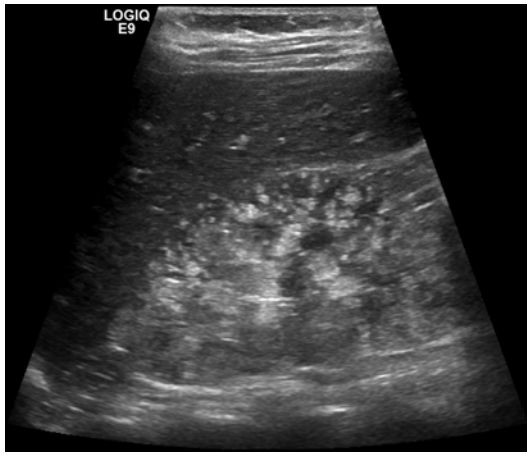


Fig. 7.18 An 8-year-old girl with tuberous sclerosis. Multiple angiomyolipomas are seen as hyperechoic roundish lesions within the kidneys and a few solitary echogenic lesions within the spleen. Longitudinal section

In US, lesions are usually multiple. They may have the same echogenicity as the spleen or may be hyper- or hypoechoic. They may be innumerable and confluent, then appearing diffusely with heterogeneous echotexture of the spleen.

Langerhans cell histiocytosis with proliferation of bone marrow-derived histiocytes causes diffuse or focal hypoechoic nodules in the spleen, which vanish after therapy (Paterson et al. 1999) (Fig. 7.19a–c).

7.4.3 Malignant Manifestations in the Spleen

In children and adults, the most frequent malignant manifestations in the spleen are *lymphomas* (Fig. 7.20a), which may be diffuse, nodular (with small size <3 cm and large size >3 cm) or bulky with extremely large and inhomogeneous lesions. Lymphomas of low malignancy present in B-mode US diffuse or with small nodules, whereas highly malignant lymphomas tend to have larger nodules or bulky disease (Weskott 2012; Benter et al. 2011). The echogenicity of the lesions may be hypoechoic or more rarely (<10 %, Görg 2011) hyperechoic. Doppler US may depict increased vascularity within the lesions (Fig. 7.21); however, there are no reliable differential diagnostic signs. In children with known lymphoma, hypoechoic focal splenic lesions are highly suspicious of being of malignant origin because of the low incidence of focal lesions in the spleen. Thus, the incidental finding of a focal hypoechoic lesion within the spleen should also be suspicious of being a lymphoma.

Contrast-enhanced US (CEUS) is more helpful than Doppler sonography in distinguishing benign from malignant splenic lesions (Fig. 7.21b) (Sutherland et al. 2011; von Herbay et al. 2009; Stang et al. 2011; Chiavaroli et al. 2011). Splenic lymphomas may show iso-, hyper- or hypoenhancement in the early phase of CEUS. In the late phase, all lesions demonstrated rapid wash-out after 60 s (von Herbay et al. 2009). Splenomegaly is usually associated. Normal texture and size of the spleen do not rule out lymphoma manifestation within the spleen. Calcification may occur after chemotherapy.

In the differential diagnosis of malignant focal splenic lesions, angiosarcomas, leiomyosarcomas (Fig. 7.22) and metastases from other paediatric tumours have to be considered. All of these tumours of the spleen are extremely rare.

Angiosarcoma is the most common non-lymphatic, aggressively malignant splenic tumour, usually diagnosed incidentally in older adults (Hartmann et al. 2008; Neuhauser et al. 2000). Unlike hepatic angiosarcoma, primary angiosarcoma of the spleen has no association with exposure to carcinogens.

The spleen is an uncommon organ for metastasis. Most *metastases* are hypoechoic, but occasionally also hyperechoic lesions with a "target sign" may be found (necrotic anechoic centre and hyperechoic rim). Splenic metastases are usually accompanied by a splenomegaly.

7.5 Vascular Pathologies

7.5.1 Splenic Aneurysms

Splenic vein aneurysms have rarely been reported in children and adults. They may occur during the course of systemic infections.

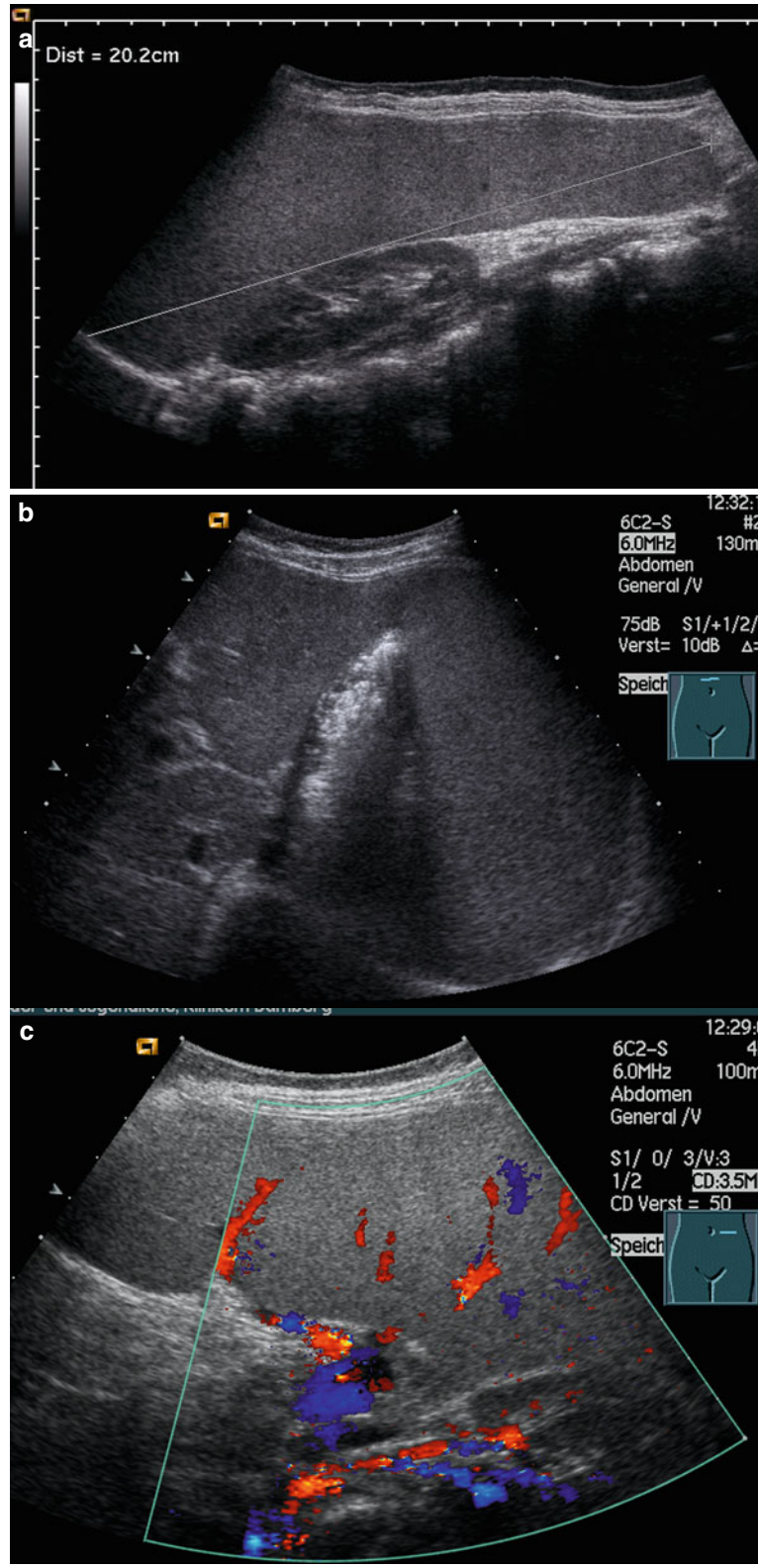


Fig. 7.19 Patient with splenomegaly and Langerhans cell histiocytosis. Diffuse hypoechoic nodules within the spleen. (**a**) Longitudinal "panorama" view of the enlarged spleen. (**b**) Kissing phenomenon due to the splenomegaly. Transverse section. (**c**) Colour-coded Doppler sonogram (CDS) reveals a normal perfusion of the spleen. Transverse section

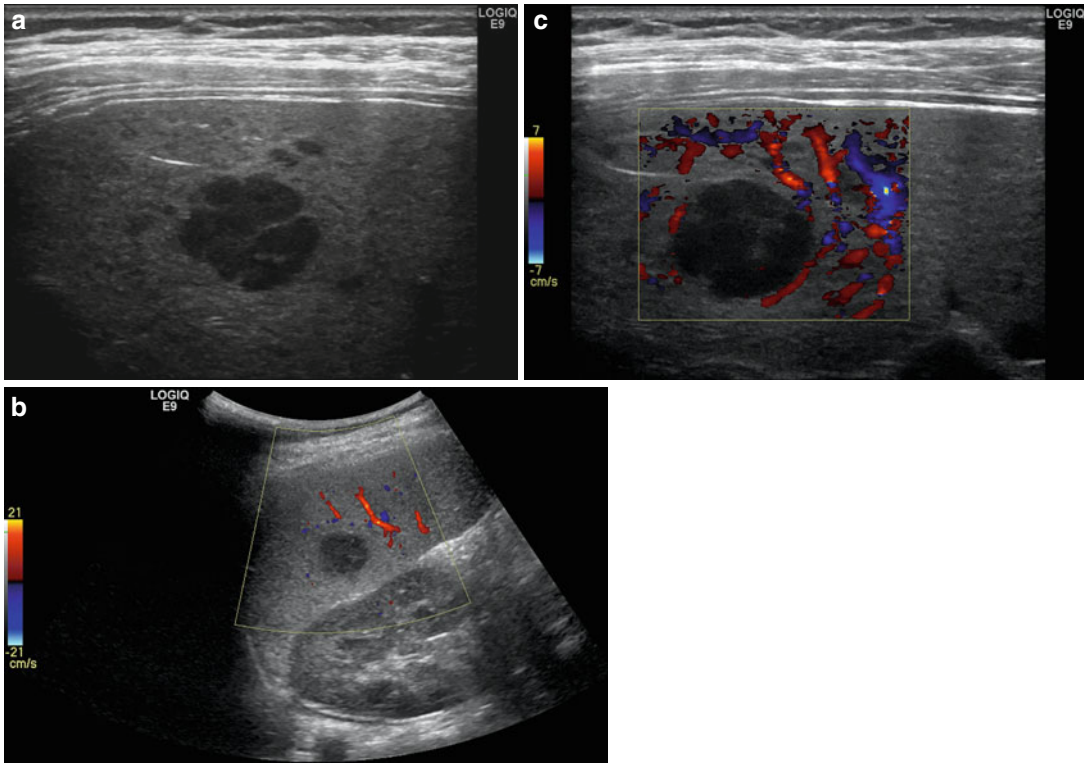


Fig. 7.20 Hodgkin's disease in a 5 ½-year-old boy. (**a**) The 2D image shows diffuse infiltration of the spleen. Additionally, a larger focal hypoechoic lesion (measuring 1.6 cm in diameter) and multiple tiny hypoechoic lesions can be shown. Longitudinal section. (**b**) CDS shows no internal vascularity of the focal lesion. Longitudinal section. (**c**) Colour Doppler of the vascularisation of the spleen. Diffuse splenic infiltration with a nodular lymphoma (>3 cm) in the spleen which shows no detectable intralesional vascularisation. Longitudinal section

Tolgonay reported spontaneous resolution of a splenic vein aneurysm in a patient with leukaemia (Tolgonay et al. 1998). After appropriate chemotherapy, the spleen diminished in size, and this decrease was accompanied by regression of the aneurysm. Colour Doppler sonography enables the noninvasive detection, diagnosis and follow-up of splenic vein or artery aneurysms.

7.5.2 Splenic Pseudoaneurysms

Pseudoaneurysms may occur after traumatic injury (Safavi et al. 2011). Although rare, traumatic splenic artery pseudoaneurysms can be life threatening. Yardeni presented a 10-year-old boy with a large splenic artery pseudoaneurysm. The pseudoaneurysm was successfully angiographically embolised, and subsequent abdominal CT demonstrated successful resolution of the pseudoaneurysm with a small residual splenic cyst (Yardeni et al. 2004). Unlike splenic artery pseudoaneurysms in adult patients, the severity of the splenic injury does not have predictive value for development of splenic artery pseudoaneurysm in children. Abdominal pain is the most frequent symptom of splenic artery pseudoaneurysm, but some children are asymptomatic at the time of diagnosis. Therefore, the possibility of splenic artery pseudoaneurysm should be ruled out even in the asymptomatic child with mild splenic injury.

As pseudoaneurysms may expand in a splenic haematoma and cause delayed splenic rupture, early diagnosis and treatment are crucial. Abdominal sonography may show free intraperitoneal fluid and an enlarged spleen with a heterogeneous area occupying parts of the organ (Fitoz et al. 2001).

Colour Doppler sonography may show flow within the lesions suggesting pseudoaneurysms.

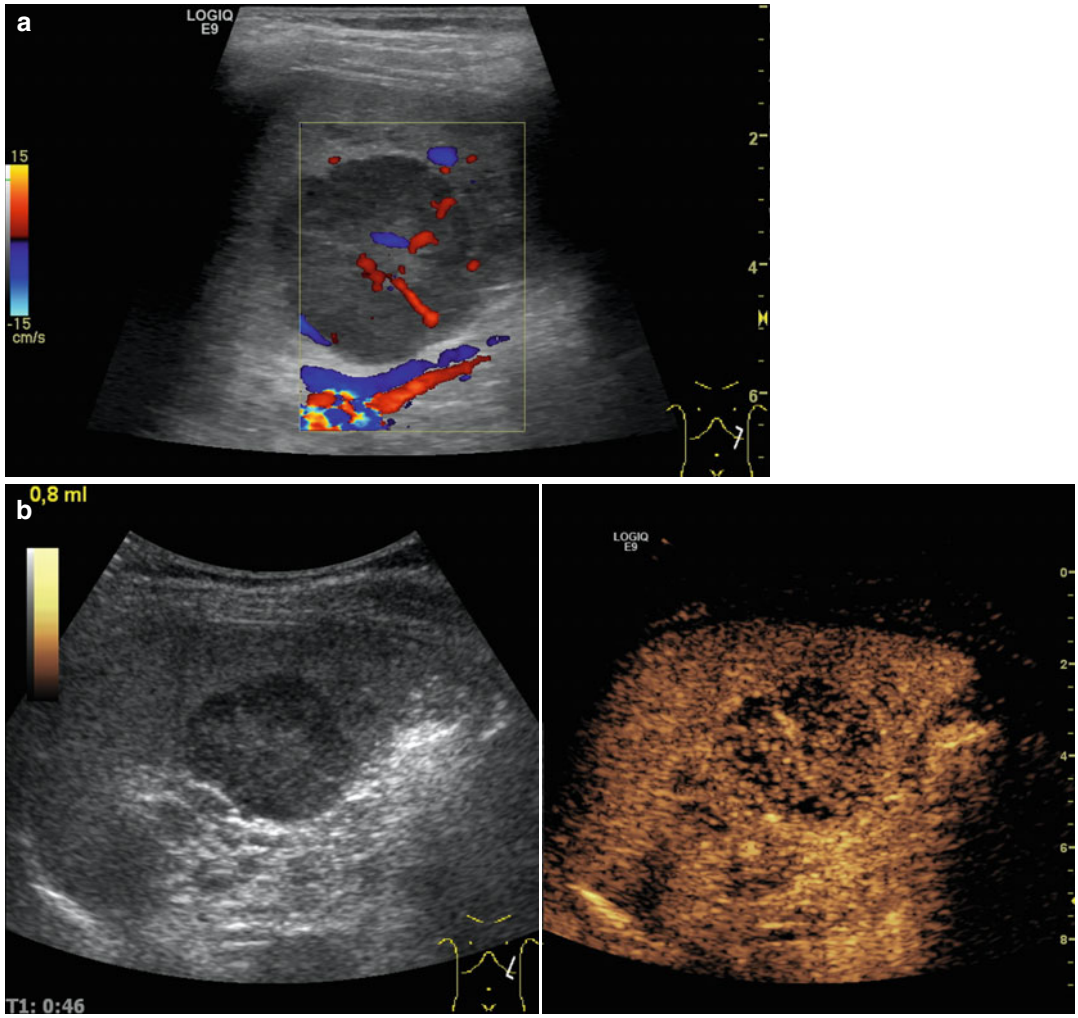


Fig. 7.21 Hodgkin's lymphoma of the spleen in a 9-year-old boy with back pain. (**a**) A single large sized nodular lesion >3 cm and multiple small lesions with vessels inside the nodule seen in CDS. Longitudinal section. (**b**) Contrast-enhanced ultrasonography (CEUS) shows a wash-out phenomenon 46 s after IV injection of the contrast agent (SonoVue®). The contrast agent leaves the areas of the Hodgkin's lymphoma within the spleen faster than in the unaffected parenchyma. Longitudinal section

Pulsed Doppler demonstrates turbulent arterial flow with high-flow amplitude.

In children with posttraumatic splenic pseudoaneurysm, spontaneous thrombosis is reported (Raghavan et al. 2004).

7.5.3 Splenic Infarction

Splenic infarction frequently occurs in patients with myeloproliferative diseases, endocarditis with cardiac emboli, storage disorders such as Gaucher disease and sickle cell anaemia. Various sonographic patterns of splenic infarction exist, but little is known about tumour-associated splenic infarction in cancer patients. In cancer patients with splenic infarction, an acute complete infarction is the most common pattern. It is caused predominantly by a hypercoagulable state and is associated with an extremely short survival rate (Görg et al. 2004). Özcan et al. (2006) described an 11-year-old girl with splenic infarction. The coeliac trunk and common hepatic artery were patent, whereas the splenic artery could not be visualised. Multiple collaterals were found at the splenic hilus region. The diagnosis

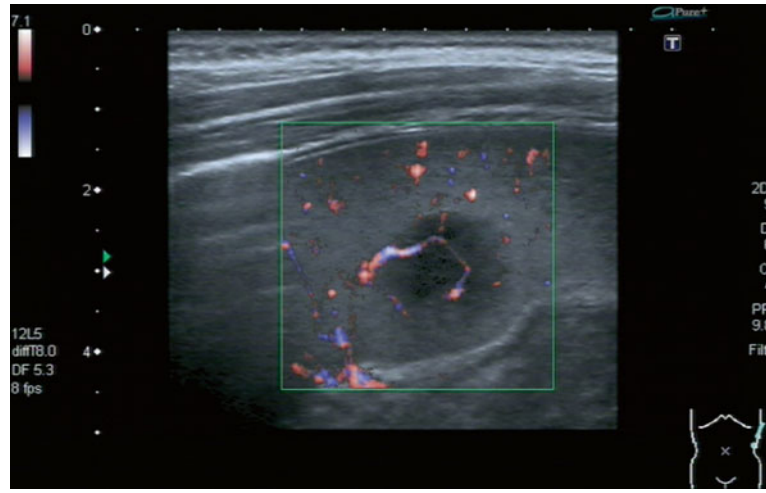


Fig. 7.22 Intracranial leiomyosarcoma with metastasis in the spleen in an 11-year-old girl with Fanconi anaemia and bone marrow transplantation. The diagnosis was proven histologically after splenectomy. Bidirectional power Doppler shows vascularisation of the focal lesion. Longitudinal section

of splenic artery occlusion was made. Massive gastric bleeding from submucosal gastric collateral vessels, secondary to the splenic artery occlusion, has been reported.

On CDS, identification of prominent collateral vasculature in the peripancreatic region and splenic hilus may be a clue to the diagnosis.

Splenic infarction may be associated with left upper quadrant abdominal pain, fever, chills, nausea, vomiting, pleuritic chest pain and left shoulder pain.

Splenic infarctions impose as triangular subcapsular hypoechoic lesions without perfusion (Fig. 7.23). The branches of the splenic artery are noncommunicating end arteries; therefore, sudden occlusion always leads to infarction. CDS or power Doppler shows absence of colour signals in the affected area, suggesting a lack of perfusion.

Splenic infarction has a high tendency to complete healing or the development of chronic infarction.

Chronic infarction develops in 17.5 % of patients with infarctions. It occurs predominantly in patients with sickle cell anaemia and myeloproliferative disease (Görg and Zugmaier 2003).

Two types of chronic infarction can be discerned (Görg and Zugmaier 2003):

Type I morphology can predominantly be found in homozygous sickle cell anaemia. It is sonographically characterised by a small or normal-sized spleen with diffuse, enhanced echogenicity and foci with diminished echogenicity.

Type II morphology is predominantly found in myeloproliferative diseases. It is characterised by an enlarged spleen with a homogeneous echotexture and solitary, triangular or hyperechoic splenic foci near the surface of the spleen. With colour-coded Doppler sonography, chronic infarcts are characterised by reduced flow signals or the absence of flow (Görg and Zugmaier 2003). Spontaneous splenic ruptures can complicate chronic infarcts in 21 %.

A functional hypo- or asplenia ("autosplenectomy") may result after multiple splenic infarctions; after bone marrow transplantation, amyloidosis, radiation or associated with autoimmune diseases such as coeliac disease with a reduced size, isoechoic or hyperechoic parenchyma and presence of Howell-Jolly-bodies in the erythrocytes.

Colour Doppler sonography (CDS) revealed an absent flow in 17 %, a hilar flow in 71 % and hilar and parenchymal vascularisation in 12 % of patients with hypo-/asplenia in a study by Görg et al. (2003).

7.5.4 Peliosis

Peliosis is a rare condition of the reticuloendothelial system with multiple blood and thrombi-filled spaces, most often within the liver or in conjunction with hepatic and splenic manifestation. The appearance in US may be

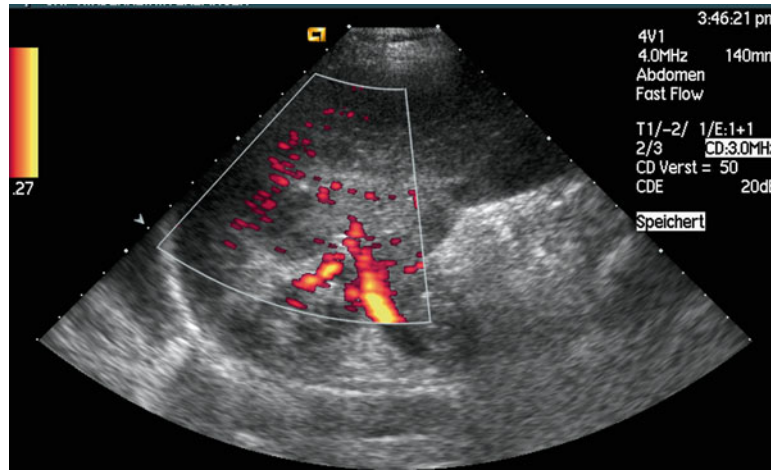


Fig. 7.23 Infarction of the spleen in a child with spherocytosis. Inhomogeneous echogenicity of the spleen. The splenic hilus is displayed echogenic, whereas the splenic periphery is speckled and inhomogeneous. Power Doppler sonogram shows only very low perfusion in the region of the splenic hilus, whereas the periphery of the spleen is not perfused. Longitudinal section (Courtesy Prof. Th. Rupprecht, Bayreuth)

hyper- or hypoechogenic with various enhancement patterns after applying contrast media. Calcifications are not described. Peliosis of the spleen is nearly always secondary to an extrasplenic disease (Hartmann et al. 2008) and may be associated with disseminated tuberculosis, HIV, haematological malignancies, transplantation, anabolic hormones and steroid therapy (Paterson et al. 1999; Abbott et al. 2004). Aetiology and pathogenesis of peliosis are poorly understood. If peliotic lesions are located close to the capsule, rupture and life-threatening bleeding with fatal outcomes have been reported (Shimono et al. 1998; Benjamin and Shunk 1978). Fine-needle biopsy should therefore be avoided; the definite diagnosis and therapy is usually made with splenectomy. In CDS, peliosis appears as hypervascularised, blood-filled nodules or cysts with a size up to 1 cm.

7.5.5 Flow in the Splenic Vessels in Congestive Conditions

Splenomegaly is a common sign in congestive conditions, mostly due to chronic liver cirrhosis or portal hypertension after portal vein thrombosis. Bolognesi reported in 2012 that an estimation of the congestion in patients with right heart or congestive failure is possible by evaluation of the splenic pulsatility index and its relation to the hepatic vein diameter.

7.5.6 Flow in the Splenic Vessels in Portal Hypertension

Due to congestion in portal hypertension, splenomegaly is present in most of the children. The splenic vein and artery may increase in diameter and the course of the vessels may be more serpentine (Figs. 7.24 and 7.27c). Normally, the flow in the splenic vein is directed from the spleen to the liver. In portal hypertension however, flow in the splenic vein may either be reversed (Fig. 7.25) or biphasic with flow into portosystemic shunts (Barakat et al. 1998) (Fig. 7.26).

The development of variceal collateral flow at the splenic hilus is characterised by multiple echofree areas at the splenic hilus (Fig. 7.27a). Colour Doppler shows flow within the echofree areas characterising them as dilated veins (Fig. 7.27b, d, e). Pulsed-wave Doppler reveals the flow pattern and velocity.

Further details concerning the flow in the splenic, mesenteric and portal vein are described in the section on liver circulation and portal hypertension.

7.5.7 Thrombosis of the Portal Venous System after Splenectomy

Post-splenectomy thrombosis of the portal vein, mesenteric vein and splenic vein occurs in about 5 % of all patients after splenectomy. Possible

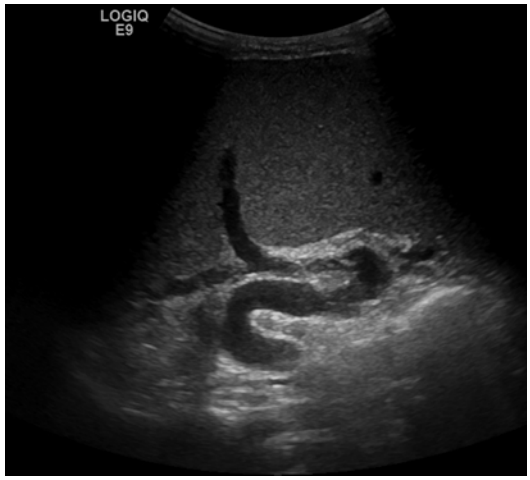


Fig. 7.24 B-mode US of portal hypertension in a child with Jeune syndrome. Elongated and tortuous splenic vein with dilated diameter. Oblique section through the spleen

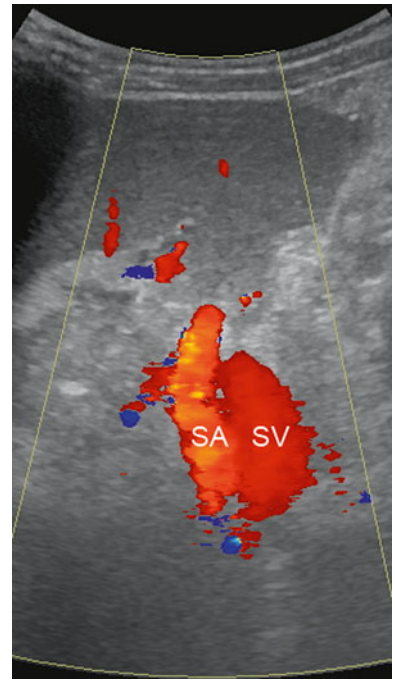


Fig. 7.25 Portal hypertension in a child with biliary atresia. Reverse flow in the splenic vein (*SV*) displayed in a *red* colour like the splenic artery (*SA*) (towards the transducer). Oblique intercostal section

risk factors are thrombocytosis and thrombophilic disorders (Stamou et al. 2006).

Age, gender, type or length of the operation and use of preoperative and postoperative thrombosis prophylaxis with low molecular weight heparin did not prove to be significant factors in the occurrence of post-splenectomy thrombosis (Stamou et al. 2006).

After laparoscopic splenectomy, thrombosis of the portal vein and its tributaries occurs more often (18.9 %) (Romano et al. 2006). In 8 % the thrombus extended from the splenic vein to occlude the portal axis. This complication was symptomatic in 11 %, whereas in the rest of the cases, the thrombosis was diagnosed incidentally in asymptomatic patients. Thrombosis occurred even as late as 2 months after splenectomy. Splenomegaly was the only significant factor predictive of thrombosis. Only those patients who were detected early with portal or splenic vein thrombosis had recanalisation of the veins with anticoagulant therapy (Brink et al. 2003). Patients with splenomegaly, who underwent laparoscopic splenectomy, are at special risk of thrombosis of the portal system and should undergo strict imaging surveillance and aggressive anticoagulation therapy (Romano et al. 2006).

Colour Doppler and pulsed-wave Doppler are able to show missing flow in the superior mesenteric vein, the splenic vein and the portal vein (Brink et al. 2003).

7.6 Splenic Involvement in Infectious Diseases

Various infectious diseases cause splenomegaly and usually no distinct morphological or vascular patterns help in the differential diagnosis. However, in *Epstein-Barr virus infection*, the increase in the splenic volume is especially rapid and marked (Fig. 7.28a, b). In rare cases spontaneous rupture of the organ may appear (Fig. 7.28a). 2D image shows a linear- or triangular-shaped zone of decreased echogenicity (Fig. 7.28a). Colour Doppler or power Doppler reveals the perfusion of the organ. At the site of rupture, no flow can be shown (Fig. 7.28b). Splenomegaly may be sudden and reversible in acute infections and sustained in chronic infections. Other causes of splenomegaly are infections with *malaria* with a homogenous enlarged spleen (Fig. 7.29a, b).

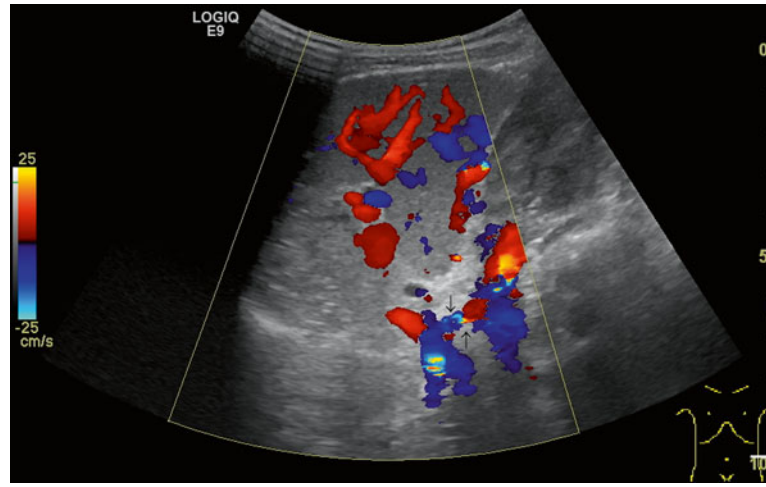


Fig. 7.26 Portal hypertension in a child with biliary atresia. Small-sized splenorenal shunts between the splenic and renal parenchyma and a large shunt between the splenic and renal vein (marked with *arrows*). Cross section through the spleen

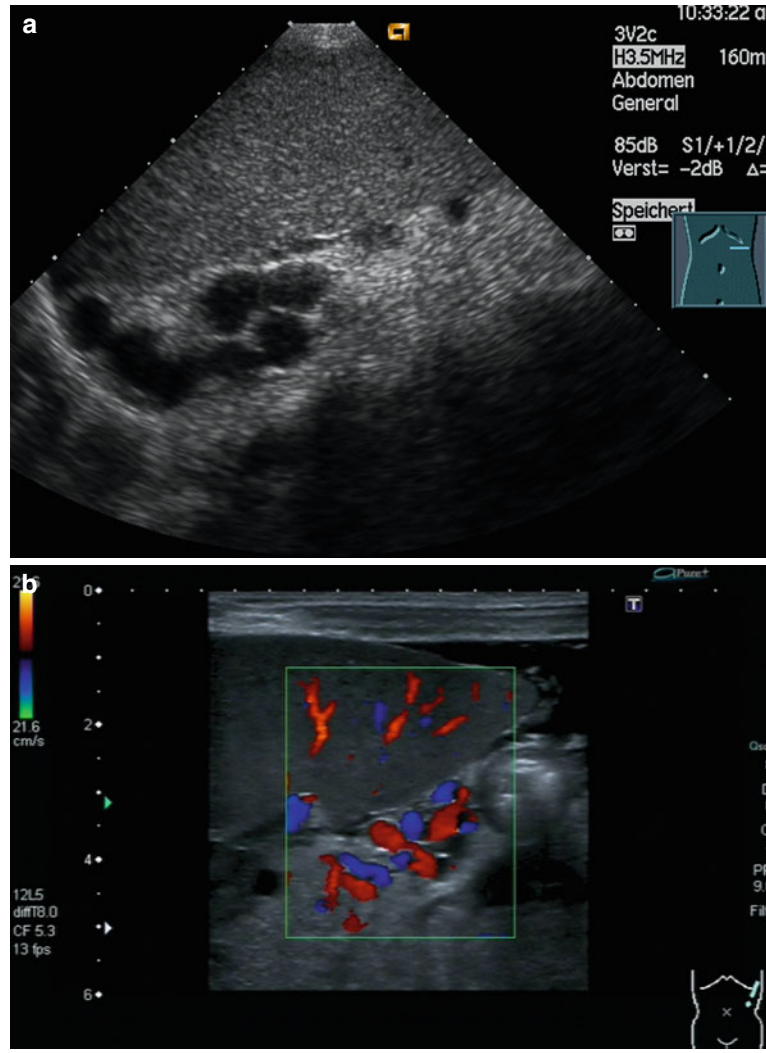
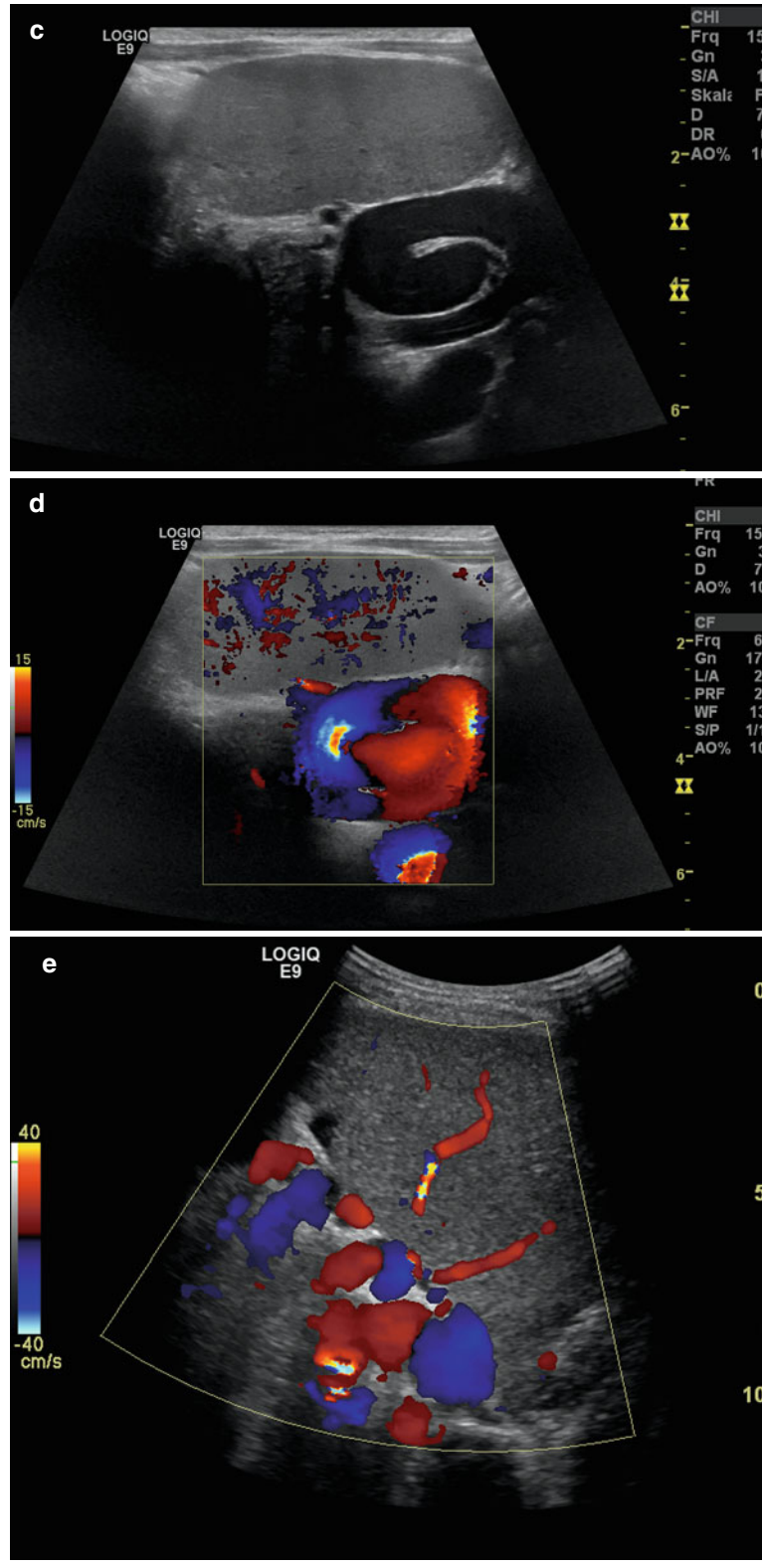


Fig. 7.27 Portal hypertension in a 14-year-old patient with neonatal portal vein thrombosis. Multiple collaterals at the splenic hilus. (**a**) Multiple anechoic structures at the splenic hilus. Cross section through the spleen. (**b**) Colour Doppler reveals flow within the anechoic tubular structures and identifies them as portosystemic shunts (collaterals). Longitudinal section through the spleen. (**c**–**e**) Large spiral twisted splenic vein in an adolescent girl with neonatal portal vein thrombosis and excessive collateral network at the splenic hilus. (**c**) 2D image shows a spiral twisted dilated splenic vein. (**d**, **e**) colour Doppler of the flow in the dilated vein (**d**) and in varices at the splenic hilus (**e**)

Fig. 7.27 (continued)



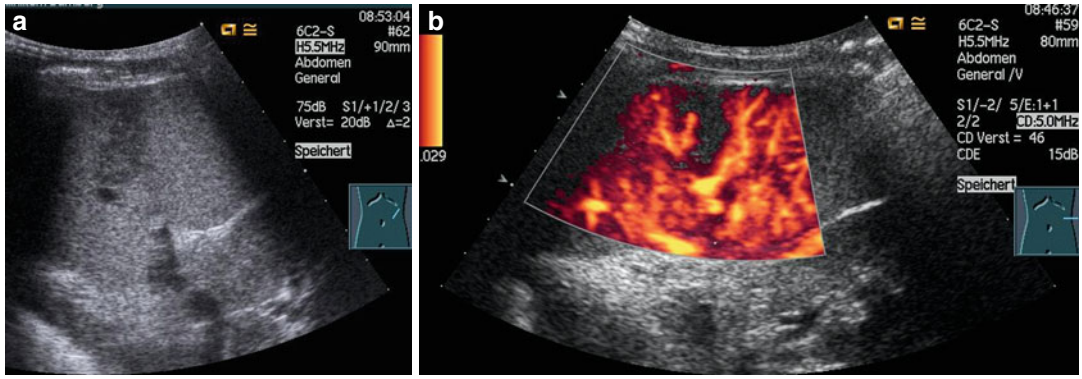


Fig. 7.28 Spontaneous splenic rupture in a child with Epstein-Barr virus infection and splenomegaly due to the rapid increase of the splenic volume. (**a**) Triangular hypoechoic area below the capsule in the region of splenic laceration in the B-mode. Longitudinal section through the spleen. (**b**) Power Doppler of the perfusion of the spleen. The normal splenic vascularisation is displayed orange. The laceration shows no flow. Transverse section

Colour Doppler reveals increased vascularity. Power Doppler demonstrates good perfusion of the organ with no lack of focal perfusion. Pulsed Doppler reveals normal arterial and venous flow.

Cat-scratch disease is caused by infections with *Bartonella henselae* after bites or scratches from cats and dogs and is common in children (Weinspach et al. 2010). About 50 % of cats are seropositive; cat flees are thought to transmit *Bartonella henselae*. Immunologically compromised children especially may develop systemic manifestations with multiple small abscesses in the spleen or liver; otherwise the clinical course is usually benign and self- limiting within a few months. Typically, regional lymphadenopathy is present (Fig. 7.30); other non-specific signs are abdominal lymphadenopathy, hepatosplenomegaly and fever. The enlarged lymph nodes are oval shaped and perfused by hilar vessels (Fig. 7.30).

Abscesses within the spleen resemble those in the liver and may occur solitary or multiple and are usually ill defined. Possibly due to the immunological competence of the spleen, abscesses in the spleen are far less frequent than in the liver, especially in nontropical countries (Fotiadis et al. 2008). Abscesses may be secondary to trauma, ischaemic infarcts or following infections. Gram-negative bacteria by haematogenic seeding are more common than gram-positive. Amoebic abscesses can be seen in certain epidemiological conditions. Especially, children with haemoglobinopathies and immunologically compromised children, such as patients with congenital or acquired immunodeficiency, intake of immunosuppressive medication, diabetes mellitus or chronic granulomatous disease (Fig. 7.31a, b), are at risk. In phases of neutropenia, the lesions and the hypervascularised rim zone around the abscess, which can be demonstrated by CDS, may transiently disappear. Usually the abscesses are hypoechogenic and may contain echogenic internal debris or gas bubbles (Fig. 7.31a) (Sutherland et al. 2010); later on calcification can develop. Colour Doppler usually shows no internal vessels in the abscess (Fig. 7.31b).

Postinfectious granuloma within the splenic parenchyma may be seen as multiple hyperechoic lesions after tuberculosis, histoplasmosis, mycobacterium avium and Pneumocystis carinii infection, especially in patients with acquired immune deficiency syndrome (Benter et al. 2011). These miliary lesions may calcify. Isolated splenic tuberculosis is extremely rare (Zhan et al. 2010).

Splenic lesions in *visceral leishmaniasis* (*kala-azar*) appear as multiple, small, hypoechogenic spots within the splenic parenchyma and are associated with hepatosplenomegaly (Saxena et al. 2011). In endemic countries, visceral leishmaniasis is an important differential diagnosis in diffuse splenic lesions.

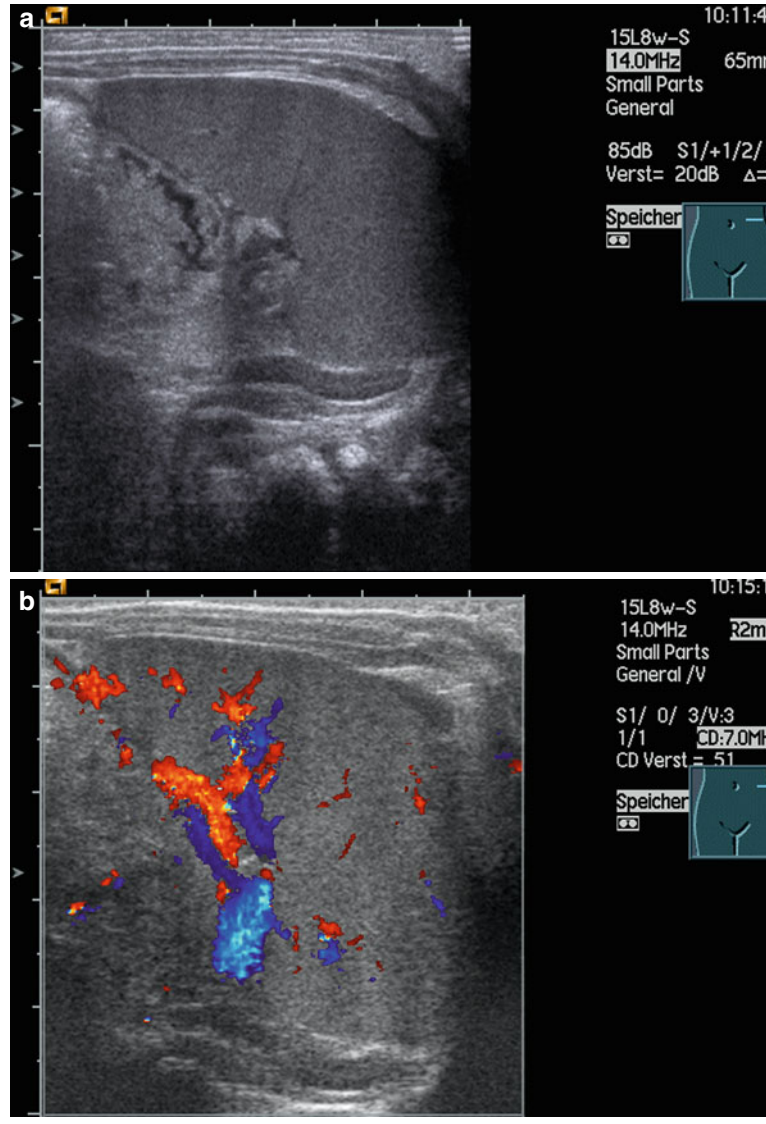


Fig. 7.29 Congenital malaria infection in a newborn at the age of 4 weeks with marked splenomegaly. (**a**) Marked increase of the size of the organ with homogenous internal reflexes. Transverse scan. (**b**) Colour Doppler demonstrates normal internal vessels of the organ. Transverse scan

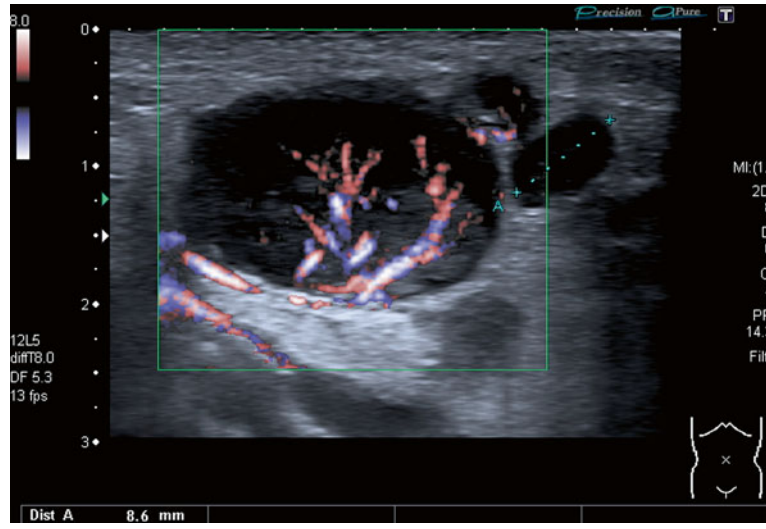


Fig. 7.30 Cat-scratch disease in a 10-year-old girl with lymphadenopathy and fever. No splenic involvement was found. Markedly increased cervical and axillary lymph nodes with decreased echogenicity and hyperperfusion. Power Doppler of a huge axillary lymph node and several smaller surrounding lymph nodes. Longitudinal section in the axilla

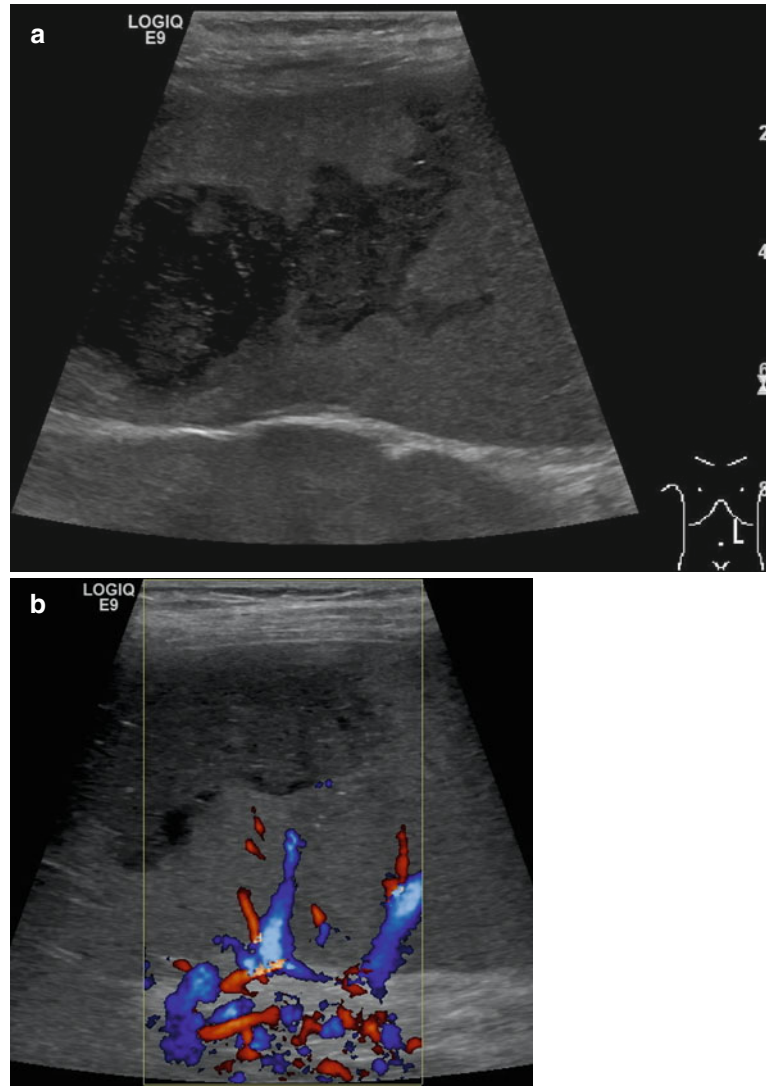


Fig. 7.31 Splenic, hepatic and renal abscesses in a 17-year-old adolescent with chronic granulomatous disease (CGD). (**a**) Hypoechoic lesions (two of several abscesses) in the spleen. B-mode. Longitudinal section. (**b**) Bidirectional power Doppler sonography shows no perfusion within the abscesses. Same section

In disseminated *mycotic infections*, especially found in immunologically compromised patients with lesions of the mucosal barriers, broad spectrum antibiotic treatment, chemotherapy and indwelling catheters, the spleen may be affected as well as the liver and, less frequently, the kidneys (Fig. 7.32). Mostly multiple small echogenic lesions with variable appearance are found; *Candida albicans*, *Cryptococcus neoformans* and *Aspergillus* spp. are the infectious agents. Ultrasonography is valuable in detection and follow-up of lesions. 2D images can show either hyperechoic or hypoechoic lesions which show no Doppler flow within the lesions.

7.7 Congenital Splenic Alterations

7.7.1 Wandering Spleen

A wandering spleen is a rare clinical entity resulting from congenital maldevelopment or acquired laxity of the spleen's suspensory splenogastric and the splenorenal ligaments (Ayaz et al. 2012; Danaci et al. 2000; Paterson et al. 1999).

Torsion of a wandering spleen is a rare cause of abdominal pain in children and has been described in increased frequency in children with prune belly syndrome with deficient abdominal

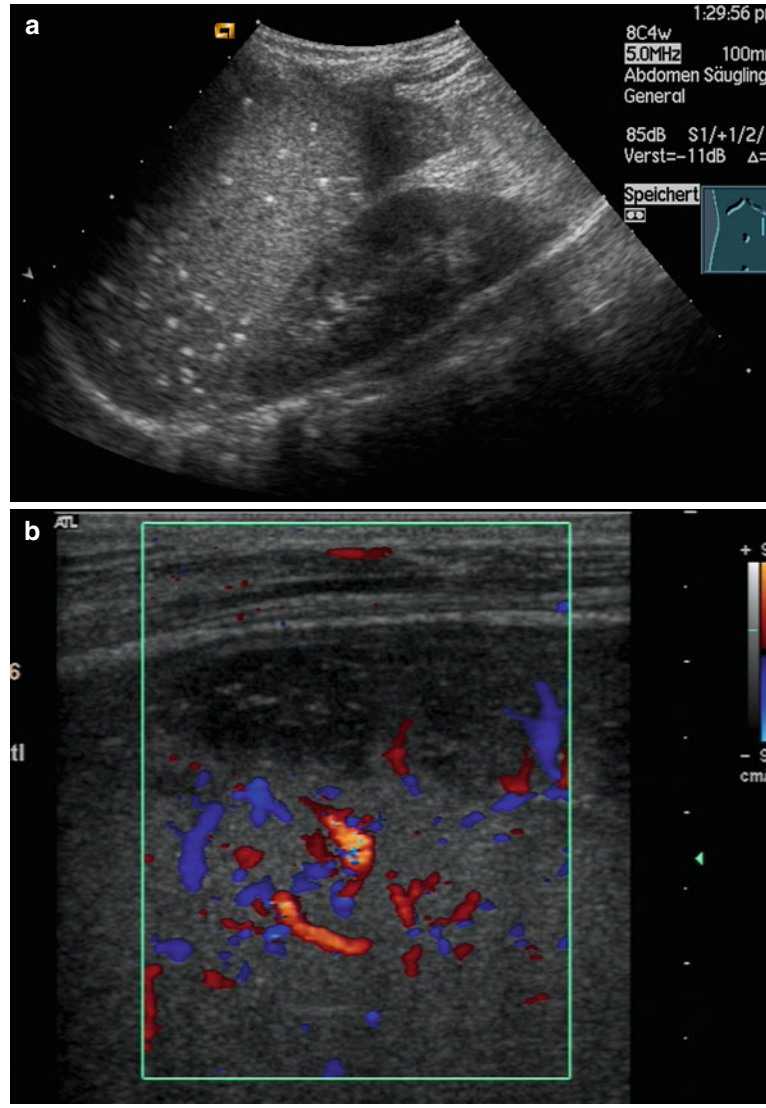


Fig. 7.32 Systemic fungal infection in a child with bone marrow transplantation. (**a**) In homogenous splenic texture. Left side longitudinal section through the spleen and kidney. (**b**) In the subcapsular splenic regions, small abscesses with no splenic perfusion can be visualised. Intercostal section with a high-frequency probe (9 MHz) and CDS

muscles (Teramoto et al. 1981). The most common presentation is acute abdominal pain, although signs and symptoms vary widely. Due to the risk of splenic infarction, rapid and accurate diagnosis is essential (Fig. 7.33e) (Di Crosta et al. 2009). The right decubitus position can help to identify the migration of the spleen (Chen et al. 2012). A rare complication is haemoperitoneum caused by acute splenic torsion of a wandering spleen (Lopez-Tomassetti Fernandez et al. 2006).

Wandering spleen and splenic torsion can be diagnosed by Doppler ultrasound (Fig. 7.33c, e) (Romero and Barksdale 2003). Confirmatory findings would include absence of the spleen in its normal location and demonstration of a splenic mass elsewhere in the abdomen or pelvis (Fig. 7.33).

Beside the location, the size and echogenicity of the spleen are investigated. Torsion of the spleen may be characterised by an increase in size of the organ and a change in echogenicity. The grey-scale US shows the displaced spleen as a homogeneous, hypoechoic mass suggestive of an enlarged, ectopic spleen in the central abdomen (Fig. 7.33a, b) (Danaci et al. 2000).

Spectral analysis and CDS demonstrate a normal vascular branching pattern and high diastolic

flow due to low resistance in the vascular bed. The parenchymal resistance index of the mass is similar to that of the native spleen (Vural et al. 1999).

The aim of colour Doppler is the demonstration of normal or abnormal internal arteries and vessels. Complete torsion is characterised by a lack of demonstrable flow within the splenic parenchyma either on colour flow images or power Doppler (Fig. 7.33e) (Danaci et al. 2000; Nemcek and Miller 1991).

Pulsed Doppler may show pathological flow profiles in the arteries and veins. If torsion occurs, firstly, the peak flow falls in the splenic vein, then the diastolic forward flow in the splenic artery is decreased, due to an increase in the peripheral resistance (Nemcek and Miller 1991).

If no flow can be shown with colour or power Doppler, diagnosis of suspected torsion can be confirmed by contrast-enhanced ultrasonography (CEUS) (Fig. 7.33f). Treatment options include splenopexy or splenectomy.

7.7.2 Accessory Spleens (Splenunculi)

Accessory spleens are common (10–30 %). They may be solitary or multiple and usually don't

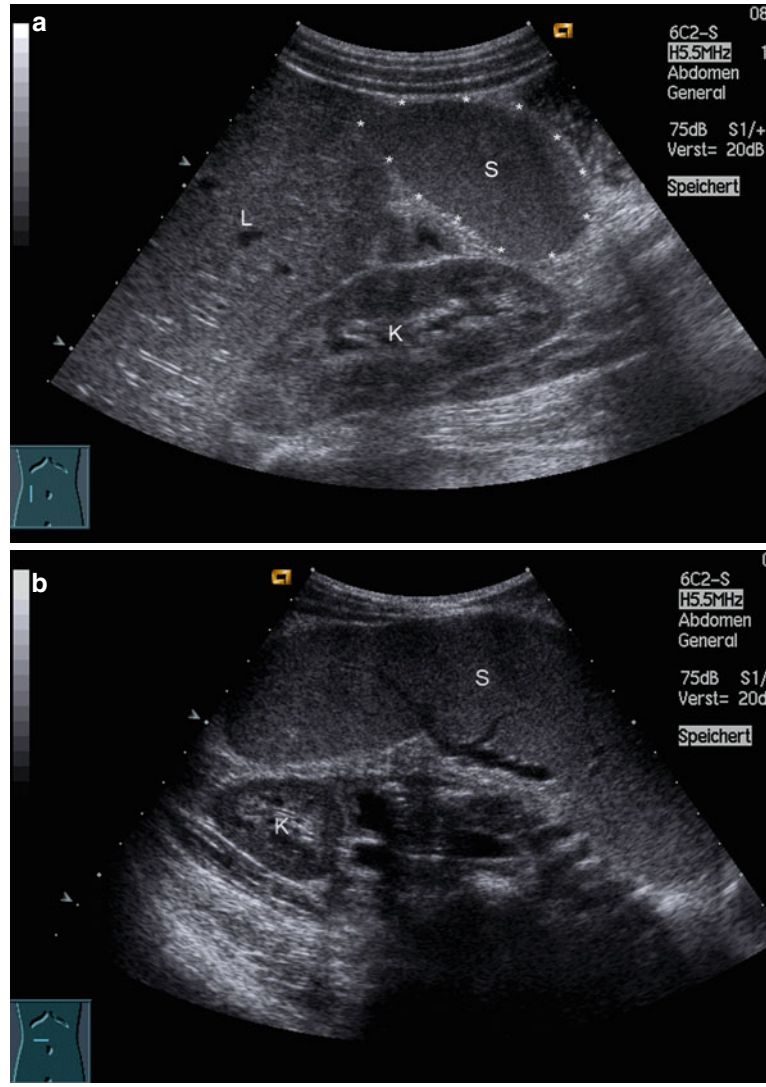


Fig. 7.33 Wandering spleen. (**a**) 2D image of a patient with intermittent upper abdominal pain and a wandering spleen. The image shows splenic parenchyma (*S*) between the liver (*L*) and the right kidney (*K*). Longitudinal section through the upper right abdomen. (**b**) Enlarged spleen (*S*) in the right upper abdomen anterior to the kidney (*K*). Transverse section through the upper abdomen. (**c**) CDS shows a normal vascularisation of the wandering spleen (*S*). Longitudinal section. (**d**) MRI of the same patient with a wandering spleen displays the spleen (*S*) in the right middle abdomen (*L*) liver. Splenopexy was performed later on. (**e**) Torsion of a wandering spleen in a 13-year-old girl with recurrent abdominal pain. Acute severe abdominal pain. The spleen was located in the lower abdomen and showed no perfusion with colour Doppler. (**f**) CEUS was performed to confirm the suspected diagnosis of torsion of the spleen: No perfusion of the organ could be shown (Courtesy Dr. Schulz, Erlangen)

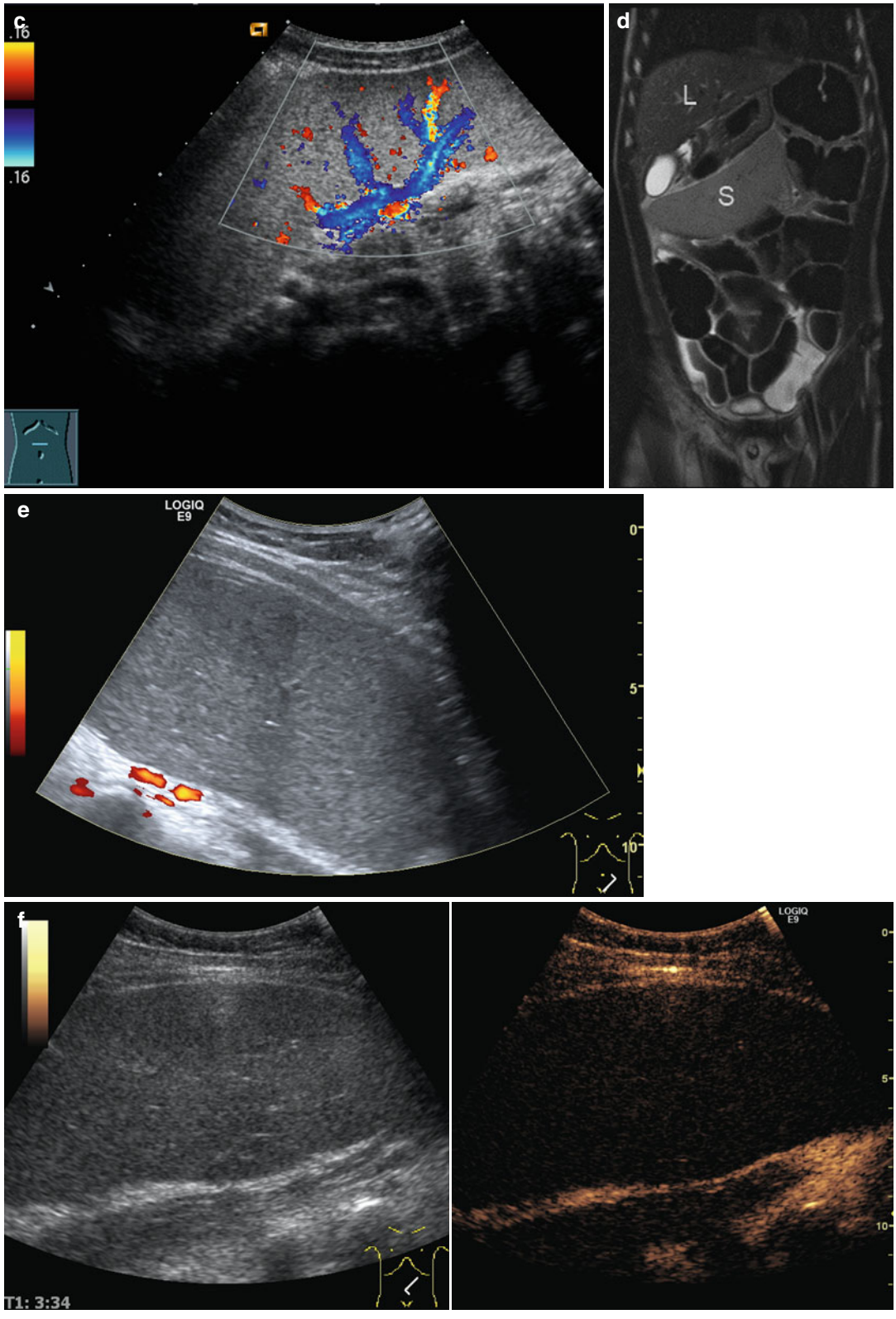


Fig. 7.33 (continued)

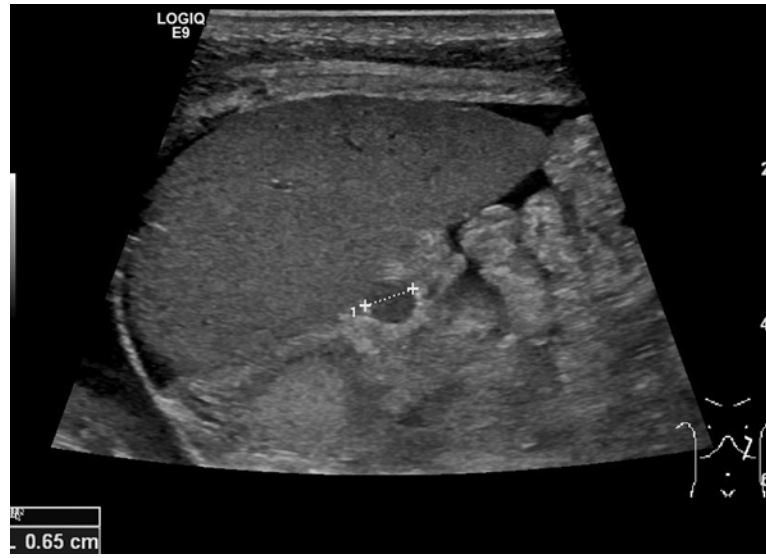


Fig. 7.34 Splenunculus. A round, small accessory spleen is localised in the splenic hilum. The parenchyma of the splenunculus has the same texture and echogenicity as the main body of the spleen. Additionally, splenic collaterals can be seen in this child with portal hypertension. Longitudinal section

measure more than 4 cm (Fig. 7.34). They are most often located at the hilus region (Paterson et al. 1999; Peddhu et al. 2004; Elsayes et al. 2005). They have the same echogenicity and echotexture as the main spleen and are capable of hypertrophy. They may vary from a few millimetres to several centimetres in size and range from 1 to 6 in number. Intrapancreatic splenunculi are rare, but accessory spleens may be found anywhere in the abdomen.

7.7.3 Splenosis

After traumatic injury of the spleen, a diffuse spreading of splenic parenchyma in the abdominal cavity, called splenosis, may occur. Splenosis may protect children from bacterial infections after splenectomy. The nodules may mimic lymphomas or metastases (Ksiadzyna 2011).

7.7.4 Polysplenia

In patients with polysplenia, there is more than one hilus (Fig. 7.35) and multiple splenic nodules. Polysplenia may accompany congenital heart disease. In children with situs inversus and/or heterotaxy syndrome, the spleen is located on the right side usually together with the stomach as both originate from the dorsal mesogastrium. The anatomy of the splenic vessels vary according to specialities of the liver anatomy (e.g. horizontal or butterfly liver with central hilus) and anatomical variations of the great abdominal vessels, including vascular signs of malrotation in half of the patients with malposition or rotation of the superior mesenteric vein around the superior mesenteric artery.

7.8 The Role of Contrast-Enhanced US (CEUS) in Splenic Lesions

Contrast-enhanced ultrasonography (CEUS) has been suggested as a useful tool in characterising incidentally detected splenic lesions (Chiavaroli et al. 2011; Sutherland et al. 2011). Up to now, it is off-label use even for use in extrahepatic indications in adults.

CEUS with second-generation contrast agents has been shown to improve the differentiation between benign and malignant splenic tumours (Stang et al. 2011) and to improve visualisation of splenic metastasis (Neesse et al. 2010).

Most splenic haemangiomas showed isoenhancement after intravenous contrast agents, one-third had a hypoenhancement and 11 % a wash-out; only a minority of the haemangiomas

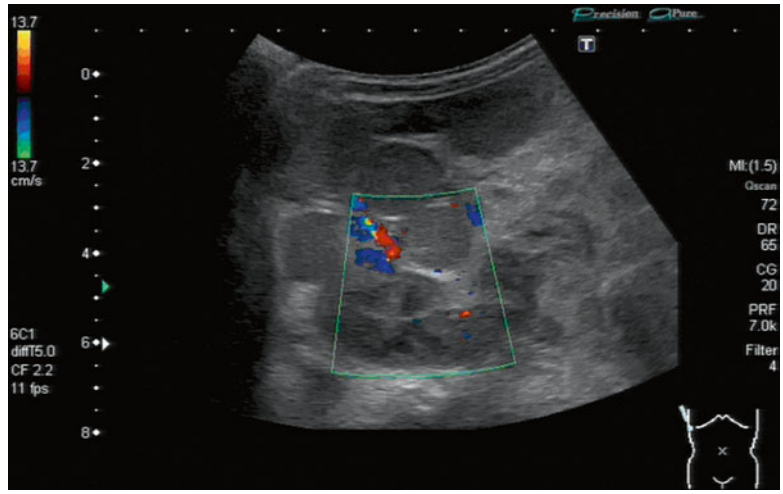


Fig. 7.35 Polysplenia and situs inversus. Syndromatic biliary atresia with polysplenia, situs inversus, aplasia of the inferior vena cava and persistence of the azygos vein. Using CDS the main hilar splenic vessels and small branches to the individual small spleens can be detected. Right lateral longitudinal section

showed a peripheral globular rim enhancement and centripetal fill-in as typically seen in liver haemangiomas (Taibbi et al. 2012).

Rim enhancement may be seen in pyogenic splenic abscesses (Weskott 2013; Paterson et al. 1999). In patients with splenic trauma, CEUS is more sensitive in detecting splenic lesions than conventional US and CDS (Weskott 2013) and may be used as a follow-up imaging technique after interventions of splenic trauma (Dormagen et al. 2011).

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