



Paediatric liver ultrasound: a pictorial essay

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Abstract

Ultrasound scan is a painless and radiation-free imaging modality and, therefore, it is widely considered the first-choice diagnostic tool in the setting of hepatopathies in paediatric patients. This article focuses on the normal ultrasound anatomy of the liver in neonatal and paediatric age and reviews the ultrasound appearance of the most common diffuse and focal liver affections.

Keywords Ultrasound · Liver · Paediatric · Diffuse hepatopathies · Focal liver lesions · Cholestasis

Introduction

Ultrasound (US) scan is the first-choice diagnostic tool in the setting of hepatopathies in paediatric patients. According to their duration, less or more than 6 months, hepatopathies are, respectively, defined acute or chronic. In addition, there are focal and diffuse liver diseases. Diagnostic imaging plays a crucial role in the evaluation of liver affections as they may both present with non-specific clinical and laboratory picture or represent an incidental finding, especially when signs and symptoms arise late. Nevertheless, also imaging presentation

of liver injuries extensively overlaps, therefore, correlation with clinical data is mandatory [1, 2].

Sonography is a quick, painless and radiation-free tool; thus, it is particularly helpful in the diagnostic approach in children. In some cases, further imaging modalities are needed, while in other conditions, it is considered enough for the assessment of liver diseases or for their follow-up.

Anatomy, technique and ultrasonographic aspects

Liver is located in the right upper quadrant of the abdomen and is divided into a right lobe on the right-hand side of the falciform ligament and a left lobe on the opposite side, while the quadrate and the caudate lobes are near to the hilum. According to Couinaud Codification, each lobe is composed of many segments on the basis of portal and venous branches (Fig. 1), so that each one has its own vascular peduncle. The right portal vein separates anterior and posterior segments of the right lobe, the left portal branch, instead, identifies Segment II and Segment III on the left hepatic lobe. Segment I is an autonomously vascularised lobe situated behind the venous ligament [3]. A complete US study of the liver requires systematic study through longitudinal, axial and oblique scans; it also includes the evaluation of intrahepatic vascular structures (intrahepatic and portal veins) and the study of the gallbladder. When necessary, basic B-mode study should be completed by colour-Doppler technique [1, 4].

US examination of the liver is performed while the patient lies in the supine position with a convex probe.

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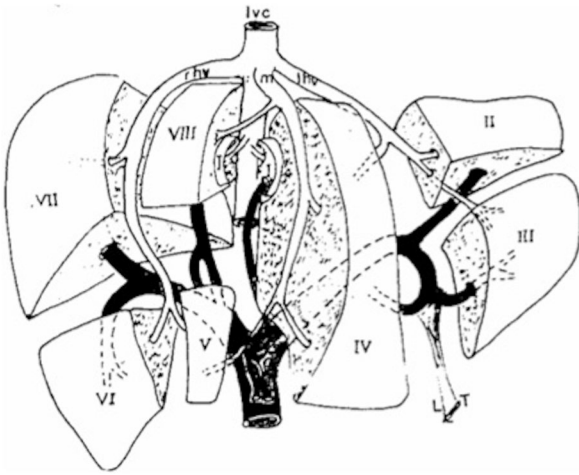


Fig. 1 Schematic overview of segmental anatomy of the liver. Commonly, segments are identified following portal and venous branches ramifications

However, as their abdominal wall is thin, in young children a high-frequency linear probe (5–20 MHz) may be used for obtaining clearer images with improved spatial resolution.

In addition, when possible, it is useful to acquire scans during deep breaths to allow better visualisation of abdominal organs.

A standardised approach is achievable: first, longitudinal and axial scans along the midline are acquired to evaluate the left lobe; secondly, longitudinal and oblique subcostal scans are performed to image the right lobe (Fig. 2).

Normally, the liver presents homogeneous echotexture with evenly distributed medium- to low-intensity echoes. Such homogeneous texture is disrupted by tubular anechoic structures representing vessels (portal vein and hepatic veins branches) crossing the hepatic parenchyma (Fig. 3). Hyper-echoic rim, due to peri-portal connective tissue, always surrounds the portal vein branches in the hepatic parenchyma [1, 2, 4].

Routinely, a longitudinal scan along the hemiclavear line is used to measure the longitudinal diameter of the right lobe, which is reported to evaluate the hepatic size (Fig. 4) [5–7]. Generally, the right lobe is bigger and extends to the lower pole of the ipsilateral kidney while the left lobe reaches the left of the aorta. By contrast, in neonatal period, the liver is relatively broad and may extend much over the midline with the left hepatic lobe being physiologically hypertrophic. Later, the volume of the right lobe increases more than the left one [1].

Furthermore, neonatal liver is characterised by bright echotexture and sharp margins, which progressively become smooth.

Table 1 reports normal range of hepatic diameter related to the patient age [7].

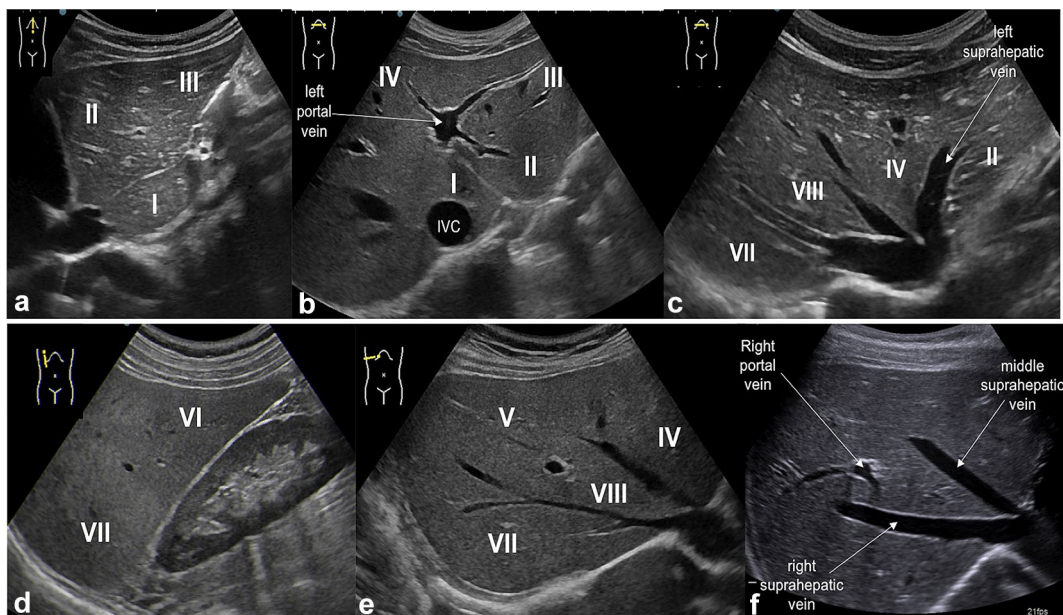


Fig. 2 US study of the liver: longitudinal and transverse scan along the midline (a, b) show the left hepatic lobe (segments II and III) separated from segment I (close to the inferior vena cava, IVC) and the left portal vein with its feeding branches for segments II, III and IV; in the oblique subcostal scan (c) there are right, middle and left

hepatic veins which separate segments VII, VIII, IV and II; longitudinal scan along hemiclavear line (d) shows segments VII and VI; oblique subcostal scans (e, f) show right portal vein separating segments VIII and V

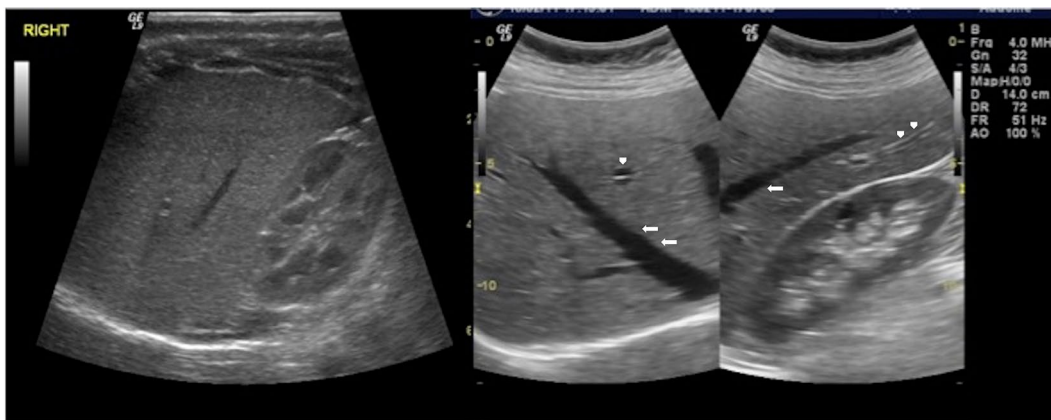


Fig. 3 US study of the liver, longitudinal and oblique scans: normal hepatic echotexture is characterised by homogenous low-to-intermediate echoes similar to the adjacent renal cortex; there are also

tubular anechoic structures representing intrahepatic veins (arrows) and portal vein branches (arrowheads)

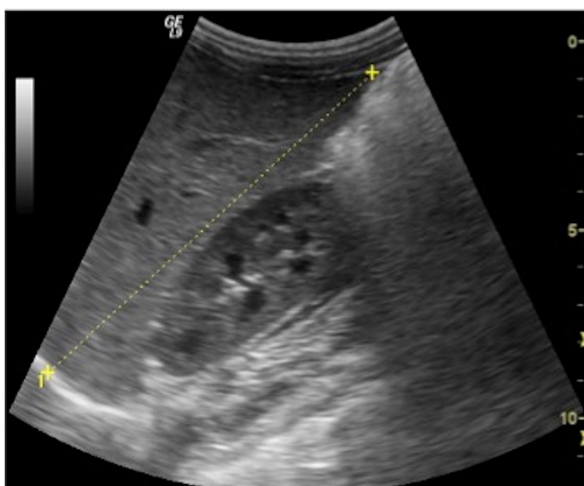


Fig. 4 US study of the liver: longitudinal scan along the right hemiclavear line is commonly used to measure the longitudinal diameter of the right hepatic lobe

Table 1 Normal range of hepatic diameter related to the patients age Modified from Konuş OL et al. [7]

Age versus longitudinal dimensions of right lobe of liver	
Age range (month)	Longitudinal dimensions (mm)
1–3	64
4–6	73
7–9	79
12–30	85
36–59	86
60–83	100
84–107	105
108–131	105
132–155	115
156–179	118
180–200	121

As well as the liver volume, with the child growth also increases the portal vein calibre, which is measured at the hilum, if possible during a deep breath (Fig. 5). The normal range is considered 3–5 mm in newborns and up to 7–11 mm in older children [8].

US study in newborns should also evaluate the presence of still patent ductus venosus, which may be a useful peripheral venous access through the umbilical vein (Fig. 6) [1].

Commonly, intra-hepatic bile ducts are not clearly identified at US study. Conversely, common hepatic duct and common bile duct are detected near to the hilum. There is not complete agreement about the normal calibre of common bile duct, however, it is generally considered pathological a diameter greater than 7 mm. Gallbladder lies on the inferior surface of the liver and is characterised by a normal length

of 1.5–3 cm in infants < 1-year-old and 3–7 cm in older children [9–11].

Diffuse hepatopathies

When considering differential diagnosis among numerous diffuse liver injuries, it is important to correlate clinical picture with laboratory findings and also with the age of the patient. This latter may be crucial as often there is different distribution of hepatopathies among various age groups. For example, newborns mostly suffer from metabolic disorders, cardiac failure, and hepatitis. On the contrary, in older children steatosis, cirrhosis, venous-occlusive disease or cystic fibrosis related hepatopathy are more common [1, 12].

Fig. 5 US of the liver, colour-Doppler study of the portal vein at the hilum. Portal flow is usually directed towards the liver, with normal mean velocity of 15–18 cm/s

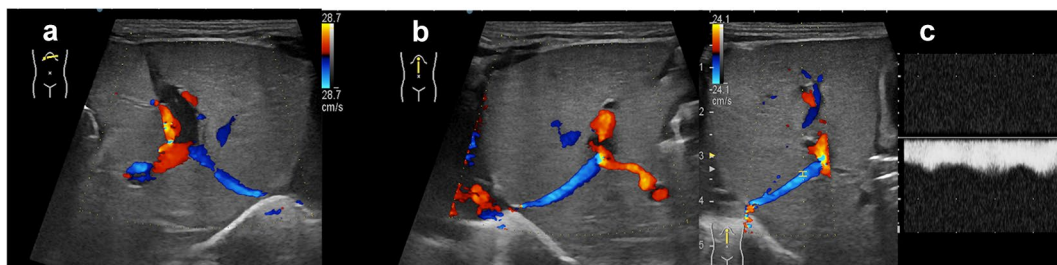
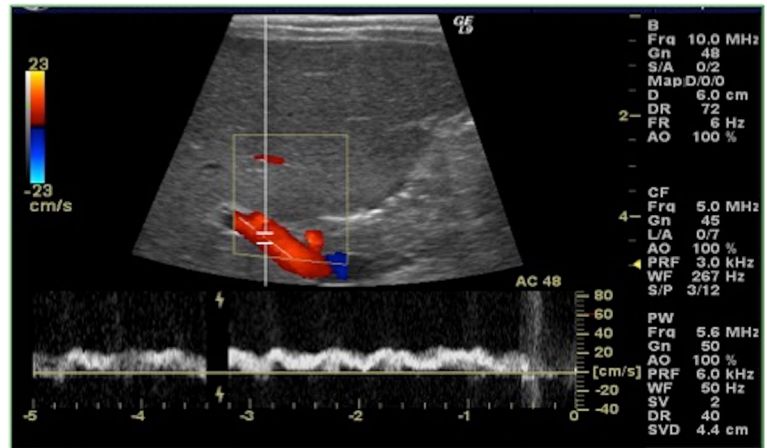


Fig. 6 US of the liver with colour-Doppler (a, b) and colour-pulse-Doppler (c) study in a newborn depicts patent ductus venosus (with hepatofugal flow in blue)

Acute hepatitis

Imaging is only marginally helpful in the evaluation of acute hepatitis. Actually, the diagnosis of this condition is mostly based on clinical and laboratory parameters. Aside from hepatomegaly, there are no specific US signs of hepatitis. The “starred sky” effect, given by a diffuse hypo-echogenicity of the liver with hyperechoic spots along the small portal branches, has been considered specific of acute hepatitis (Fig. 7), but it was demonstrated that it could be found also in healthy subjects. Gallbladder can show thickened walls, due to parietal hypertonus and/or associated oedema, sometimes with a triple-layer effect [1]. Sometimes, there is also ascites.

Hepatic steatosis

In most of cases, in paediatric patients hyper-echogenicity of the liver is related to steatosis. This quite common changing is usually a reversible injury, characterised by an estimated prevalence of 10% [10]. It may present with a diffuse or focal pattern and is related to hepatocyte fat deposition depending on both abnormal alimentation, such as obesity, malnutrition, parenteral nutrition, nervous anorexia, or steroid

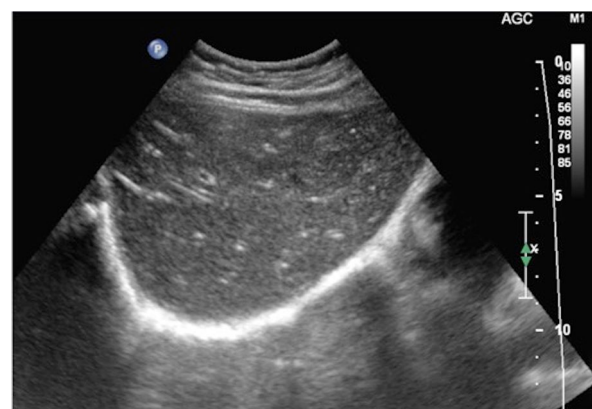


Fig. 7 US of the liver, oblique scan in patient affected by acute hepatitis: there is an increase of hepatic volume and slight decrease of echostructure, while portal branches wall appears brighter

therapy, or several metabolic/enzymatic disorders such as uncompensated diabetes mellitus, celiac disease, cystic fibrosis, Wilson disease, glycogenosis, fructose intolerance (Table 2) [13–16].

At US examination, diffuse fatty liver appears bright, markedly hyperechoic compared to the adjacent renal cortex

Table 2 Main causes of steatosis in paediatric age

Obesity	
Congenital Metabolic Disorder	<ul style="list-style-type: none"> • Wilson Disease • Glycogenosis • Fructose Intolerance
Steroidal Therapy	
Celiac Disease	
Cystic Fibrosis	
Uncompensated Diabetes Mellitus	
Parenteral Nutrition	
Nervous Anorexia	
Malnutrition	

and with blurred appearance of intra-hepatic vessel walls. The degree of these changes reflects the severity of the fat deposits. In particular, Grade I refers to diffusely bright echo-pattern with still well-defined diaphragmatic profile, Grade II to bright liver with shadowing of periportal echogenicity, Grade III to very bright liver with loss of diaphragmatic profile (Fig. 8) [16].

Sometimes, there are areas not involved in fat deposition, known as “fat sparing”, which could mimic focal lesions and need magnetic resonance imaging (MRI) to be clarified. Conversely, focal pattern of steatosis may result as geometrical or finger-shaped bright areas without any “mass effect” (Fig. 9). These areas are usually found close to the falciform ligament, the “porta hepatis” or the gallbladder fossa [17].

Cirrhosis

Liver cirrhosis is the result of chronic inflammation which leads to cellular regeneration and reparative process finally resulting in fibrosis. It is associated with many different conditions and it is considered rare in newborns. Most frequent forms of cirrhosis in paediatric population are biliary and post-necrotic [18–21]. US scan generally shows volume change of cirrhotic liver, with a small right lobe and hypertrophic caudate and left lobes. Moreover, margins become irregular, typically finely indented in micronodular pattern and roughly lobulated in macronodular one. Fibrosis

makes the overall hepatic echostructure appear diffusely and roughly heterogeneous (coarse pattern) (Fig. 10).

In addition, the portal vein calibre is usually increased due to portal hypertension, which is often found with ascites, splenomegaly, collateral vascular circles and slow or inverted spleno-portal flow [18]. The progressively decreased flow velocity in portal vein may lead to thrombosis. This situation may be followed by lysis of the thrombus and sometimes the cavernous transformation of portal vein (cavernoma), which is constituted by venous channels with hepatopetal flow (Fig. 11) [22]. Furthermore, the hepatic veins are reduced in calibre and may sometimes show parietal irregularities due to nodular regenerative pattern.

Finally, the complete loss of normal hepatic structure leads to extremely low protein synthesis, which contributes to the development of ascites. This latter is usually found in peri-hepatic, peri-splenic and pelvic spaces (Fig. 12).

The final step of hepatic changes in cirrhosis is diffused fibrosis. However, before this process becomes irreversible, there are several degrees of injuries that progressively accumulate.

Thus, recently, new diagnostic tools have been introduced to evaluate the mounting fibrotic damages in the liver. Elastography is one of these, which allows evaluation of hepatic fibrosis by measurement of hepatic stiffness. Even though biopsy has been largely considered the gold standard for the diagnosis of liver fibrosis, recently, many authors underlined several disadvantages connected to this procedure. In fact,

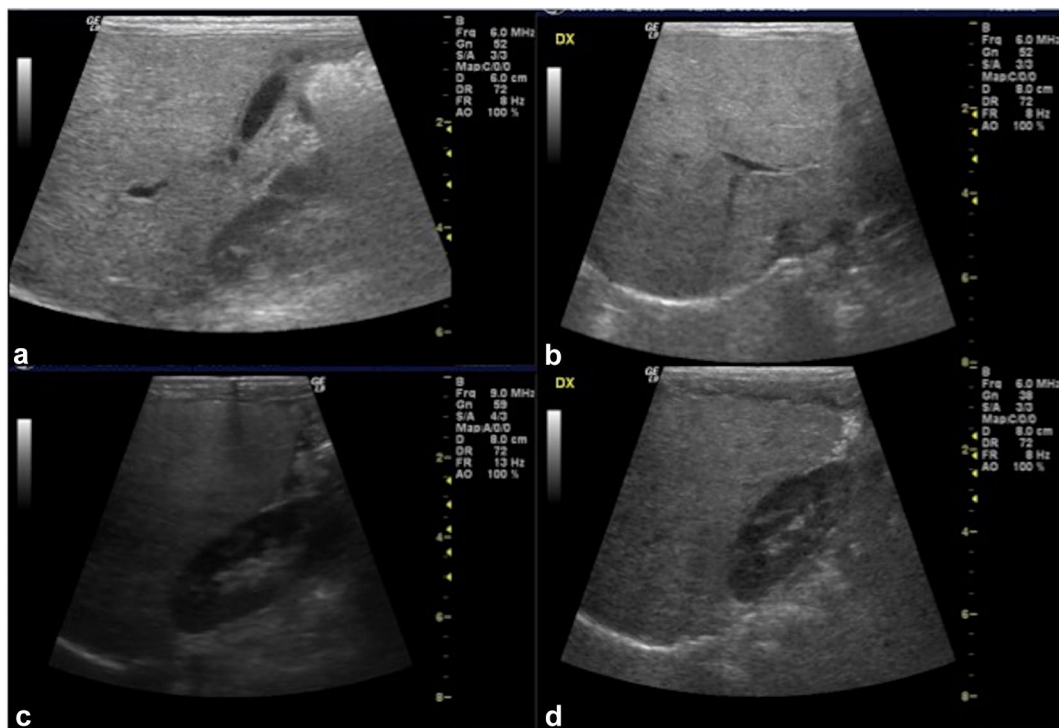


Fig. 8 US of the liver, longitudinal and oblique scans (a–d): the echogenicity of the liver is particularly bright, especially when compared to the adjacent renal cortex (c, d); moreover, there is blurred appearance of intrahepatic vessels walls and loss of diaphragmatic profile



Fig. 9 US of the liver, longitudinal, axial and oblique scan: there is hyperechoic liver parenchyma (particularly when compared to the cortex of right kidney) with quite well-defined area of relatively

hypoechoogenicity (a, b) due to fat sparing-area; in the right figure (c) there is a geometrical shaped bright area which does not cause any mass effect and is therefore in keeping with focal hepatic steatosis

it is invasive, with risk of haemorrhage, and, because of heterogeneous distribution of fibrosis, it is also associated to high risk of sampling errors. Therefore, elastography is progressively becoming the modality of choice for the assessment of hepatic fibrosis [18, 19]. Transient elastography is a non-invasive technique consisting of US transducer that follows the propagation and the velocity of a shear wave previously transmitted to the tissue. Actually, it has been demonstrated that stiffness is related to the degree of fibrosis

and it affects the wave velocity. One more elastographic technique is the point shear-wave elastography, based on the evaluation of the speed of the shear waves propagating through a defined ROI and which result from compression of the tissue by push pulse of standard transducer. A total of 10–12 ROIs are selected placing the probe on the intercostal spaces and mean, median and average velocities are then automatically calculated (Fig. 13). The usefulness of an accurate evaluation of the liver fibrosis is to understand if

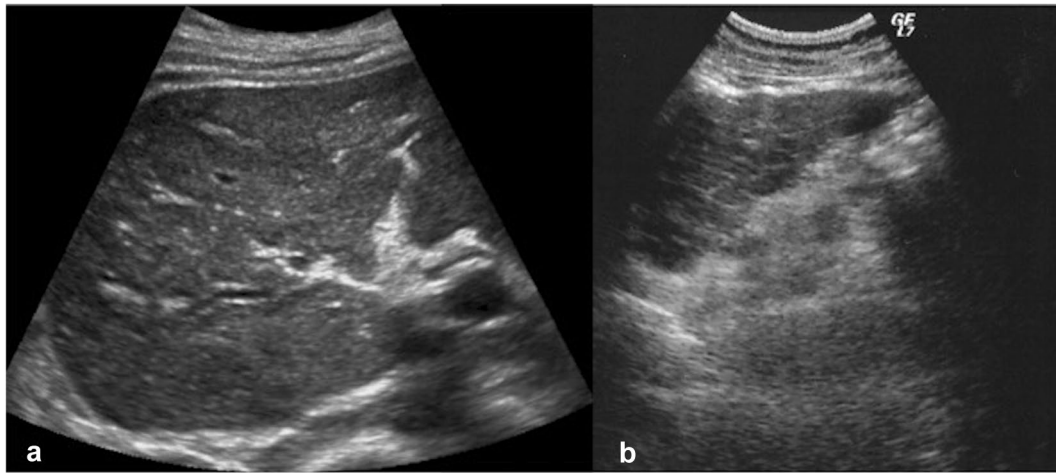


Fig. 10 Abdominal US study shows changes in hepatic cirrhosis, with evidence of volume decrease of the right lobe, irregular margins and roughly heterogeneous echogenicity (coarse pattern) in the right (**a**) and left lobe (**b**)

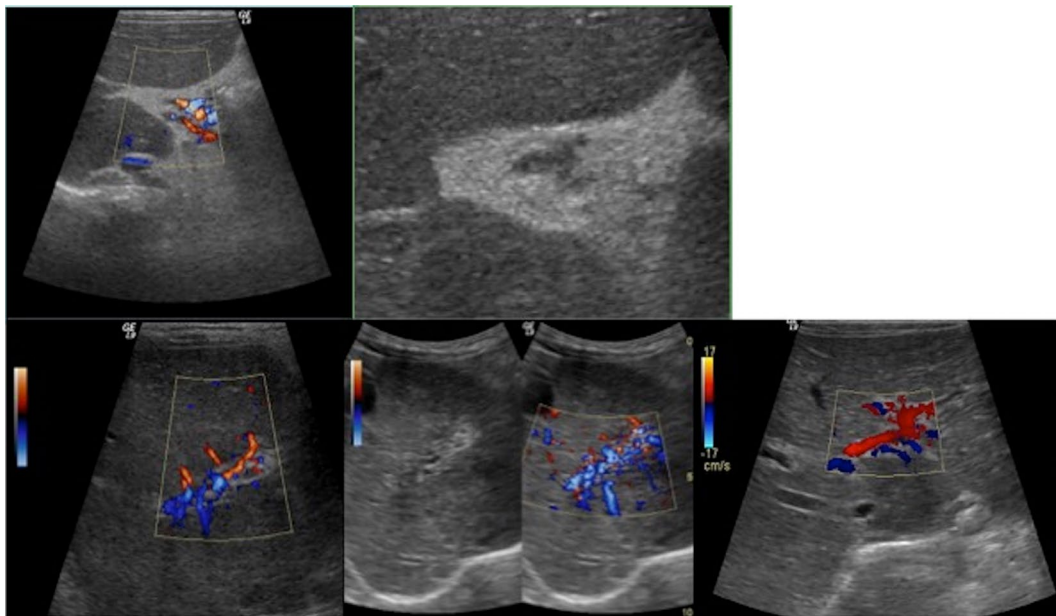


Fig. 11 US evaluation in cirrhotic liver with colour-Doppler study: at the hilum, the portal vein is replaced by portal cavernoma, composed by many small tortuous vessels as consequence of portal hypertension and vein thrombosis

there is the possibility for the patient to benefit from medical treatment [23–25].

Cystic fibrosis

Hepatic manifestations of cystic fibrosis are encountered in older patients surviving to respiratory complications. At US study, liver may be affected by a wide spectrum of alterations, varying from increased volume with hyperechoic structure to a coarse pattern with irregular margins and concurrent hyperechoic areas. In addition, there are injuries to

the biliary tree that becomes irregular with tapering of intra-hepatic branches and also strictures and beading (Fig. 14) [1, 26].

Venous-occlusive disease

Venous-occlusive disease is characterised by damage to the central veins of hepatic lobule resulting in hepatic congestion. It usually follows a primary injury to sinusoidal endothelial cells due to bone marrow transplantation or, very rarely, chemo- or radio-therapy. Ultrasonographic findings

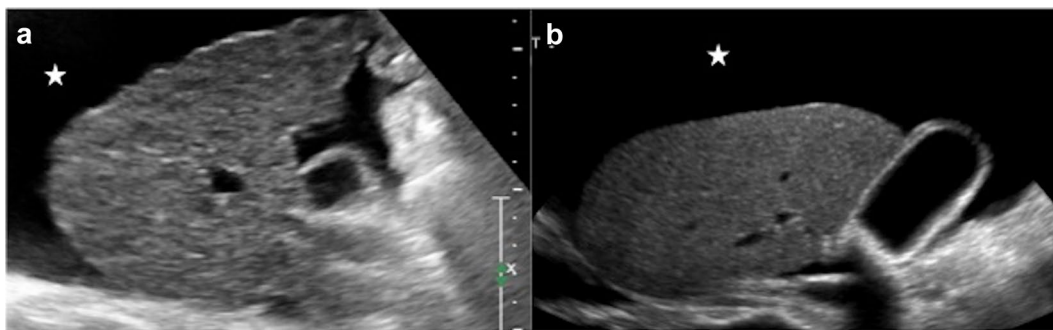


Fig. 12 Abdominal US in advanced cirrhotic condition shows hepatic changes do to regeneration process and fibrosis (a, b), with also ascites, that is identified as anechoic fluid around the perihepatic space (star)

Fig. 13 Elastography of cirrhotic liver allows to evaluate stiffness of the parenchyma, which is related to the degree of fibrosis, as it affects the wave velocity measured by the probe



Fig. 14 US study of the liver in patient with cystic fibrosis shows diffuse echostructure changes with alternation of hypoechoic and hyperechoic areas

are non-specific and the diagnosis may be suspected when there is an increase of liver volume associated to splenomegaly, ascites, gallbladder wall thickening, decreased hepatic vein calibre (<3 cm), portal vein dilatation. Progressively,

liver volume reduces, except for caudate lobe because of its autonomous vascularisation. At colour-Doppler reversed portal flow may be detected [27, 28].

Focal liver lesions

Common paediatric benign liver tumours are infantile haemangioma, mesenchymal hamartoma, focal nodular hyperplasia and adenoma; hepatoblastoma and hepatocarcinoma are most frequent malignant focal liver lesions in children [1, 29–31]. US study generally allows the detection of the tumour both in symptomatic patients and in cases of incidental findings. Further diagnostic modalities may, however, be necessary to better characterise the lesion.

Haemangioma

Hepatic haemangiomas are the most frequent hepatic lesions in neonates, detected in patients younger than 6 months. They are also divided into three different sub-types, characterised by different behaviours: focal, multifocal or diffuse. Focal hepatic haemangiomas are congenital variant similar

to rapidly involuting congenital haemangioma (RICHs) as they fully disappear usually by 14 months. Multifocal hepatic haemangiomas develops after birth and after a proliferative phase tend to decrease in a longer period (6–10 years). Diffuse lesions replace the entire parenchyma. They all can be asymptomatic or be associated to mass effect or severe complications mostly related to significant arterio-venous shunts and consequent cardiac failure. At US study, focal haemangiomas are detected as heterogeneous mass mostly hypoechoic with central area of necrosis or fibrosis and some anechoic region related to dilated vessels (Fig. 15). All the variants of infantile age haemangioma at colour-Doppler may show high-flow prominent vascular structures, sometimes with well depicted arterio-venous shunts. The most important differential diagnosis is between focal hepatic haemangioma and hepatoblastoma, which is similar in location, main features and epidemiology. However, the lack of persistent elevation of blood alpha-fetoprotein, and, for the diffused form, the presence of very large mass associated with heart failure, will suggest the diagnosis of focal hepatic haemangioma [30, 32–35].

Mesenchymal hamartoma

This is the second most common benign liver tumour in paediatric population after infantile haemangioma. Most of cases are diagnosed by 5 years and are associated to asymptomatic abdominal swelling. It is constituted by mixed components including biliary ducts, hepatocytes, vessels and mesenchymal tissue, with a variable amount of fluids. At US, quite often it is described as a multicystic mass, with anechoic components separated by thin or thick echogenic septa. Rarely, solid component may predominate. Differential diagnosis includes hepatoblastoma, which is however typically associated to high alpha-fetoprotein blood levels [30].

Focal nodular hyperplasia

Focal nodular hyperplasia (FNH) is more frequently found in adults rather than in children. Nevertheless, it represents up to 2% of all primitive liver tumours in paediatric population and it is usually recognised incidentally in asymptomatic

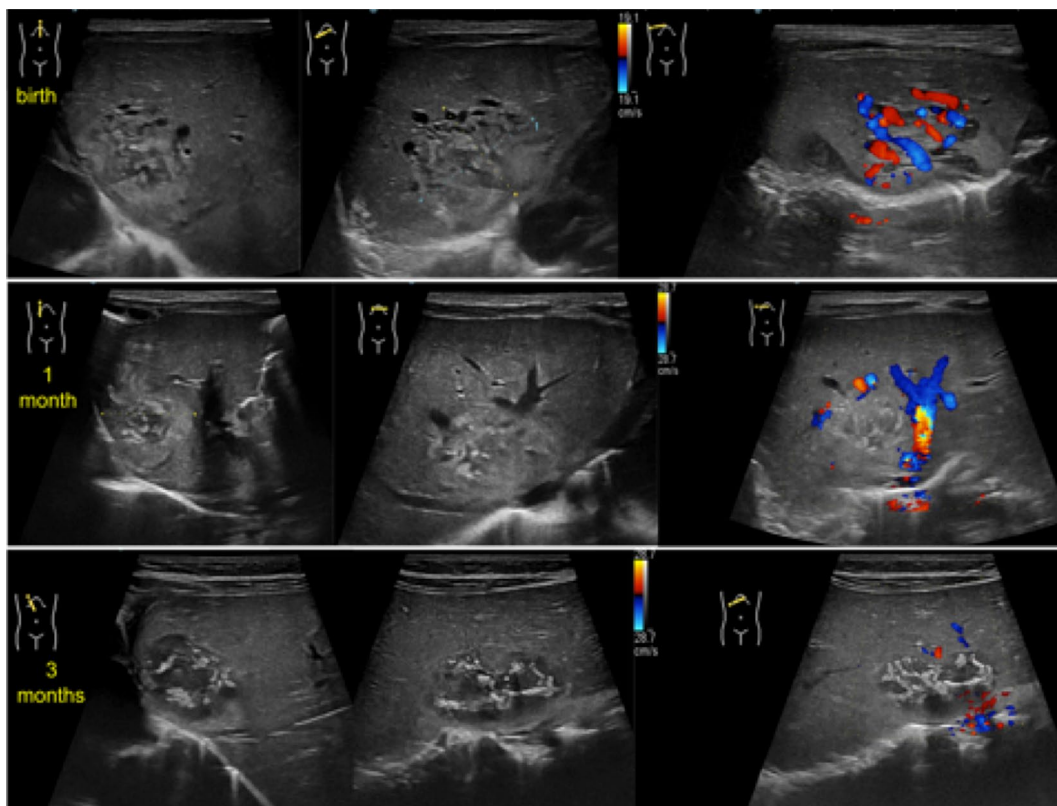


Fig. 15 US of the liver, oblique B-Mode imaging and colour-Doppler study at birth (upper line), at 1 month (central line) and at 3 months (lower line): there is a large heterogeneous mass, measuring about 20 mm, mostly iso-hypo-echoic, with internal anechoic tubular structures. Colour-Doppler images shows internal vessels characterised by both arterial and venous flow. At follow-up, B-mode evaluation shows

slight reduction of diameter with increase of the internal anechoic necrotic component. Moreover, colour-Doppler tool demonstrates significant reduction and finally disappearing of internal vascular flow. These features were in keeping with the diagnosis of focal hepatic haemangioma

2 to 5 years old children [30]. This lesion is characterised by hyperplastic hepatocytes separated by Kupffer cells and sinusoids contained in fibrous septa. These latter radiate from a central vascular axis, which is responsible for the peculiar central stellate scar. At US exam FNH is usually homogenous, with sharp margins, variably iso- hypo- or hyper-echoic, occasionally with hyperechoic central scar (Fig. 16). Colour-Doppler study may detect vascular flow within the scar and its peripheral branches which therefore assume a spoke-wheel like appearance [30, 36, 37].

Adenoma

Liver adenoma is a rare finding in children. Most frequently, they are related to systemic conditions such as glycogenosis or chronic steroid therapies. It is usually not associated with specific symptoms, however, in few cases it has been described as abdominal mass. At US it is typically a well-defined lesion relatively hyperechoic, particularly in lipid-rich variants or when there is an amount of blood within (Fig. 17). Less commonly, in steatotic bright liver, the lesion may result relatively hypoechoic. Doppler evaluation may

demonstrate peripheral and internal vessels with no central arterial flow (in contrast to FNH) [30, 38].

Hepatoblastoma

Hepatoblastoma is the most frequent primary malignant hepatic tumour in children, being up to 48% of all malignant lesion of the liver. In most of cases, the diagnosis is made by 5 years and the prognosis is unfortunately poor. It typically presents with a large isolated mass in the right lobe and is associated, in up to 90% of cases, with elevated alpha-fetoprotein blood levels [1]. US evaluation can depict a well-defined lesion, mostly hyperechoic but heterogeneous, with internal cystic areas because of necrotic degeneration and with hyperechoic calcific spots (Fig. 18). Doppler study shows high vascularisation. Most important differential diagnosis is with infantile haemangioma as both of them are solid lesions with significant vascularisation, sometimes with calcifications, and they may be found by 1 year. However, hepatoblastoma is characterised by a more aggressive behaviour. Actually, there



Fig. 16 US study of the liver, longitudinal and transverse B-mode images (a, b) and longitudinal colour-Doppler scans (c): there is a well-defined large mass, mostly iso-echoic, showing internal and

peripheral vascular flow at colour-Doppler study. This latter takes a spoke-wheel-like appearance which is typical for focal nodular hyperplasia

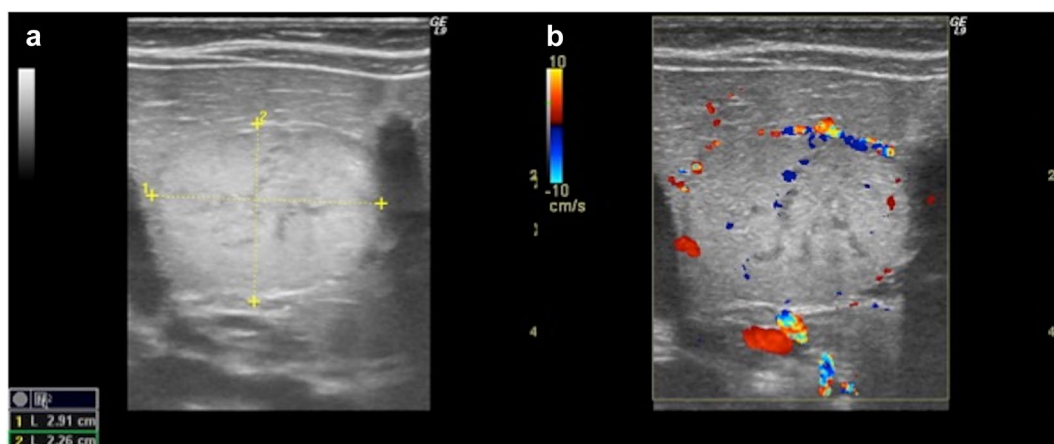


Fig. 17 US study of hepatic adenoma with B-mode and colour-Doppler images: there is a well-defined large hyperechoic mass, with some internal anechoic areas, which shows peripheral and internal vessels (a, b)

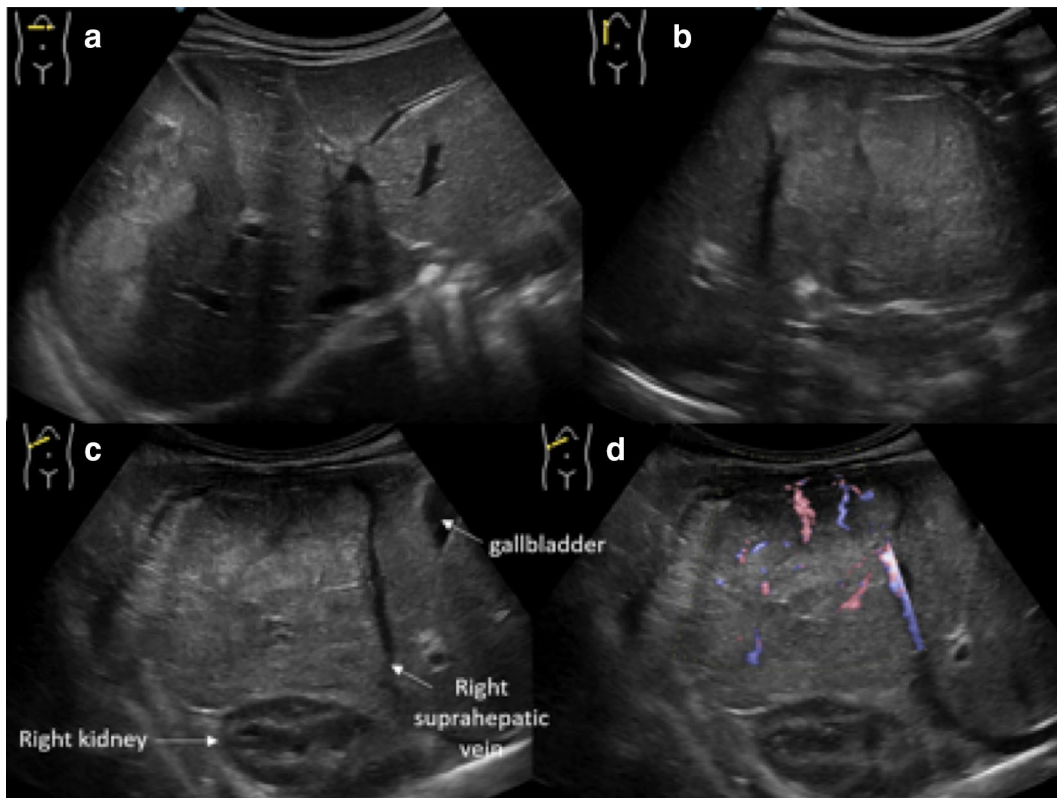


Fig. 18 Ultrasonographic images of hepatic hepatoblastoma: there is an ill-defined heterogeneous mass, involving almost completely the right lobe, which shows hyper- and isoechoic areas (a–c); many internal vessels are identified at colour-Doppler image (d)

could be encasement or even invasion of portal branches. Moreover, elevated alpha-fetoprotein blood levels are suggestive for hepatoblastoma [1, 31, 39].

Hepatocarcinoma

Hepatocarcinoma is the second most common primary malignant hepatic tumour in children and therefore it represents up to 20% of lesions in this group [1]. It has been described in association with several predisposing conditions such as primary biliary atresia, glycogenosis type I, hepatitis by virus B or C. As well as for adults, even for children hepatocarcinoma may present as isolated mass, multifocal or diffuse. At US study, small lesions appear hypoechoic to the liver, bigger ones are more heterogeneous, with hyper- or hypo-echoic internal areas, respectively, due to fat infiltration and/or haemorrhages or necrosis (Fig. 19). Doppler usually shows arterial internal flows. Main differential diagnosis is with other vascularised solid lesions, particularly with adenoma and FNH [31, 40].

Liver abscess

Liver abscess is a quite rare diagnosis in developed countries, nonetheless, it is to consider among differential diagnoses of focal liver lesions. In particular, umbilical region infections or complicated appendicitis should be considered in children with depressed immunity, who are more vulnerable to sepsis. Other known predisposing conditions are congenital abnormalities of biliary tree, which can be associated to obstruction and therefore determine bacterial proliferation. In most of cases pyogenic bacteria (80%) are involved, generally associated to single abscess that frequently is localised in the right hepatic lobe. Histologically, it is constituted by central necrotic and purulent portion with well-defined fibrotic wall. Clinical picture is usually characterised by abdominal pain and fever, less commonly there is vague pain, fatigue and weight loss. At US evaluation, pyogenic abscess appears as roundish lesion with hypo to anechoic centre, sometimes with mixed echoes, delimited by thick wall. Less frequently are found fungal abscesses that usually are multiple small lesions scattered throughout the liver parenchyma. Their

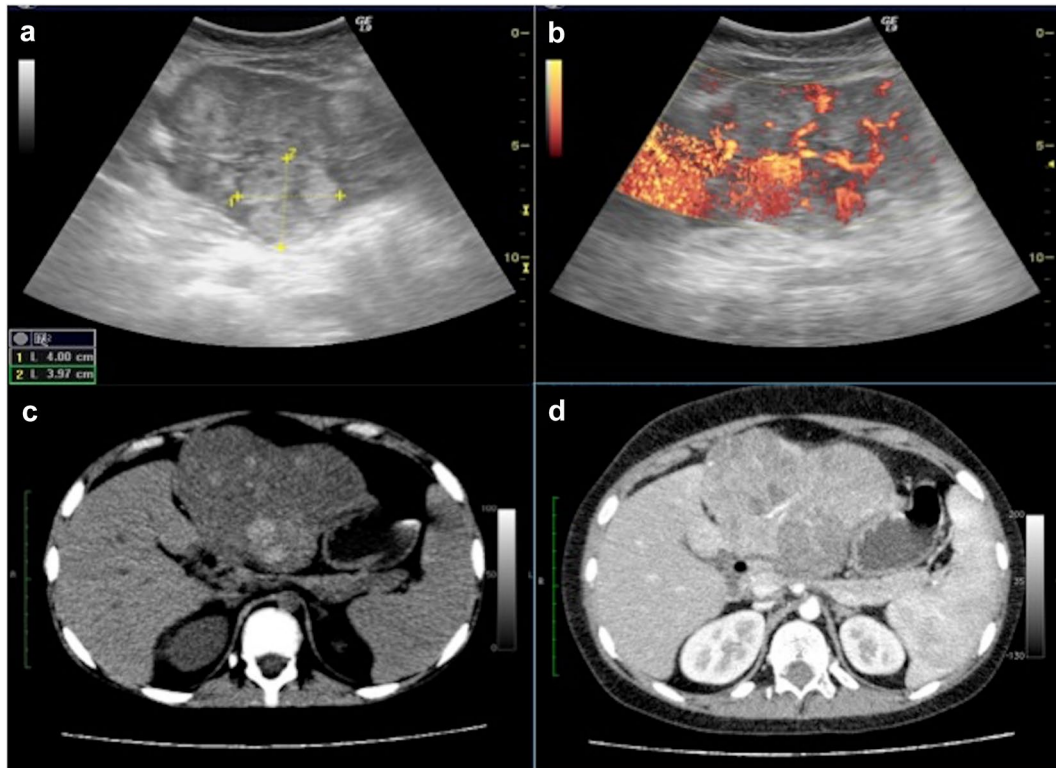


Fig. 19 Hepatocarcinoma at US B-Mode and Doppler images (a, b) and axial computed tomography and contrast-enhanced computed tomography images (c, d). At US the tumour appears as large heterogeneous lesion that bulges the liver surface, with significant internal

vascular flow (a, b). Computed tomography images show large lesion with hyperdense components at un-enhanced scan (c), which demonstrates heterogeneous enhancement (d)

US appearance may vary from target lesions to small anechoic ones. Moreover, there is the amoebic abscess, the result of a rare parasitic infection. At US evaluation it is difficult to differentiate from other liver abscesses as it is characterised by central anechoic component, which can show some low-level internal echoes, and variably thick wall (Fig. 20) [41].

Hepatic complications associated with umbilical venous catheter

As previously mentioned, US study in newborns should also evaluate the presence of still patent ductus venosus, which may be a useful peripheral venous access through the umbilical vein.



Fig. 20 US study (a, b) and computed tomography axial scans venous phase (c) of amoebic abscess: there is a large well-defined lesion involving VII/VIII segments, which shows internal hypo and

anechoic areas at US. Computed tomography scans demonstrates internal fluid component with only enhancement of the irregularly thick peripheral wall

Usually, the right progression of umbilical venous catheter (UVC) from single umbilical vein to the inferior vena cava, passing through the ductus venosus, is found by abdominal radiography [42]. Some authors proposed US as radiation-free imaging modality to assess the correct position of the catheter tip at the level of ductus venosus or inferior vena cava distal to the right atrium [40]. In addition, US may become necessary when the catheter erroneously perforates the wall of intrahepatic vessels, secondary causing intrahepatic hematoma. This is an uncommon complication of malpositioned catheter which could be suspected when there is poor blood return from the catheter or there is difficult administration of fluids through it. US study may detect the presence of well-defined hepatic mass, initially anechoic and later heterogeneous, which normally resolves spontaneously and may progressively become calcific (Fig. 21). Anyway, US monitoring is advisable [42, 43].

Cholestasis

There are several conditions associated to cholestasis. Here we will focus on biliary atresia and choledochal cyst. Biliary atresia is a condition mostly found in newborns by 3 months, presenting with persistent jaundice. Conversely, choledochal cysts may not be associated with jaundice and present only in more advanced age. Diagnostic imaging plays an important role in the diagnosis and management of these conditions. Actually, if not recognised, they may lead to hepatic transplant by 1 year of life.

Choledochal cysts

Choledochal cysts represent congenital dilatations of the biliary tree. There are many variants of choledochal cysts, all related to congenital conditions. The clinical picture is variable, related to the age of the patient: obstructive jaundice is the main sign in children, while abdominal pain is more common in adults. Only the 8% of children present symptoms by 10 years. In older children and teenagers, it can be recognised the typical triad including fever, jaundice and abdominal pain. Rarely, choledochal cysts may present as palpable abdominal mass [44–46]. The current classification (Todani's) includes (Fig. 22): type I, the most common variant that is characterised by cystic or fusiform dilatation of extrahepatic bile duct (Fig. 23); type II, that is quite rare, represents a diverticulum of the extrahepatic bile duct; type III is a cystic dilatation of extrahepatic bile duct within the duodenal wall; type IV is a cystic dilatation of both main biliary duct and intrahepatic biliary tree; in type V there is a diffused cystic dilatation of intrahepatic biliary ducts (also known as Caroli's disease) [47].

Caroli's disease is characterised by biliary ducts dilatation, which appears at US as intrahepatic single or multiple cysts. Portal branches may be partially or totally rounded by dilated ducts; frequently, there is also dilatation of the main bile duct.

US is the imaging modality of choice for the first evaluation of intra and extra-hepatic biliary ducts. Main differential diagnosis of choledochal cysts is the ileal duplication cyst, which is depicted as a round anechoic formation close to the

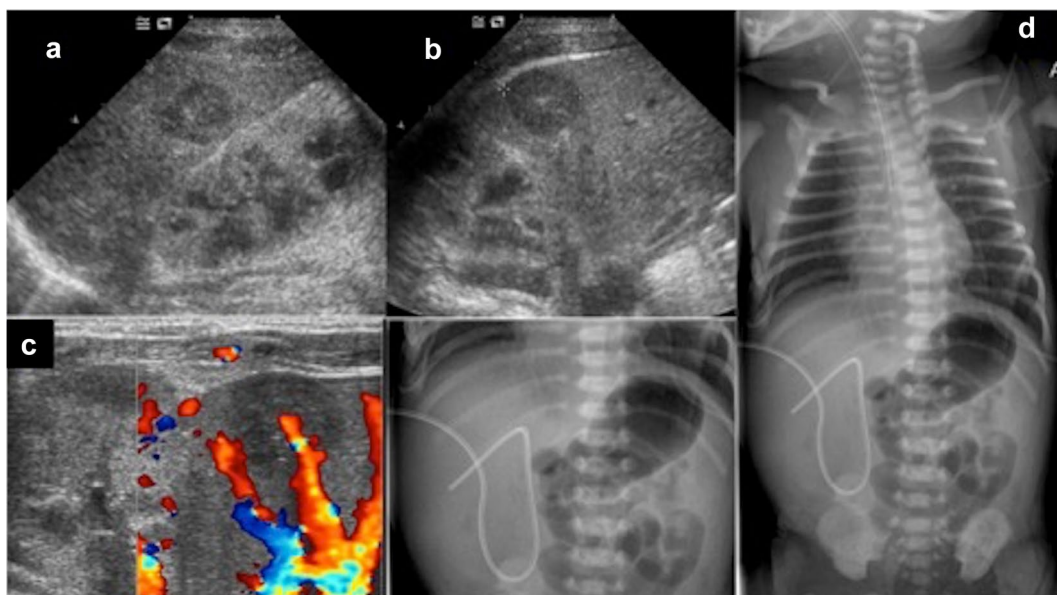


Fig. 21 US study of the liver demonstrates the presence of roundish hypo- anechoic area, probably due to parenchymal hematoma caused by displaced UVC (a–c). Frontal abdominal radiography of a newborn showing the UVC ending at the level of right portal vein (d)

Fig. 22 Schematic overview of choledochal cyst (Todani's classification) Modified from Soares et al. [44]

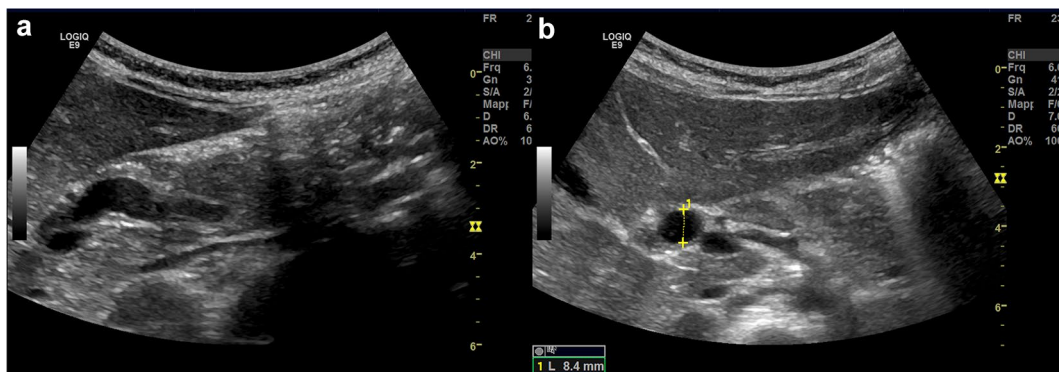
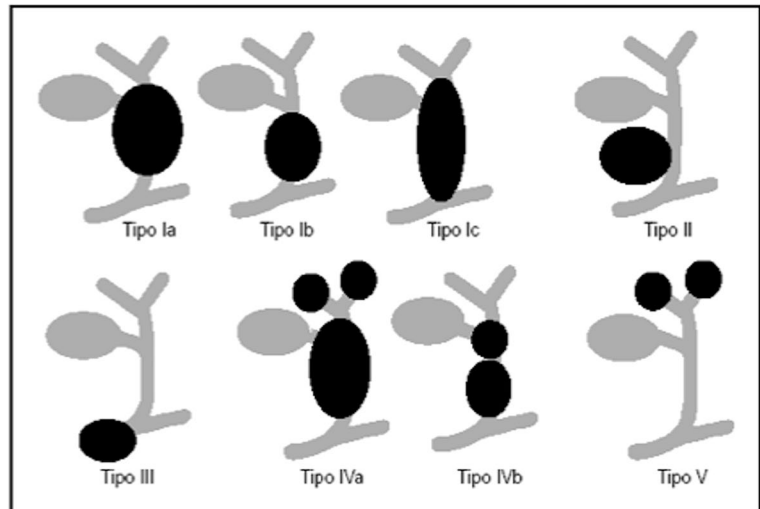


Fig. 23 US study of choledochal cyst type I shows fusiform dilatation (a) of extrahepatic bile duct measuring 8.4 mm (b)

hepatic hilum and the gastric antrum. However, ileal duplication cyst shows the peculiar stratified wall sign that cannot be recognised in the choledochal cyst [48]. Other cystic lesions to include in the differential diagnosis are simple cysts of the liver, cystic teratoma and pancreatic pseudocysts, which may require further diagnostic techniques to be excluded. In addition, magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) are also useful to obtain a more precise biliary tree anatomy, which is crucial for surgical plan preparation. [49].

Biliary atresia

Biliary atresia is characterised by a progressive obstruction of extrahepatic biliary ducts with subsequent possible involvement of intrahepatic ducts too. It has been postulated a multifactorial aetiology including genetic, infective and toxic factors leading to an obstructive progressive process. If not surgically treated by 45–60 days of life, it is associated

to a poor prognosis because of the development of hepatic cirrhosis. Clinical picture is characterised by jaundice, hepatomegaly, acholic stools and brown urine [50–52].

Differential diagnosis between biliary atresia and other causes of obstructive jaundice is crucial to undertake the best therapeutic management and avoid severe complications.

Abdominal US plays a pivotal role in the screening of infantile cholestasis: biliary atresia can be suspected when gallbladder is not visualised (ghost gallbladder) and if there is a hyperechoic tissue anterior to the main portal vein, probably representing fibrotic residuum of atretic biliary ducts (triangular cord sign) (Fig. 24). This latter sign is associated to a sensibility of 78.2% and specificity of 100% in the diagnosis of biliary atresia [53–58].

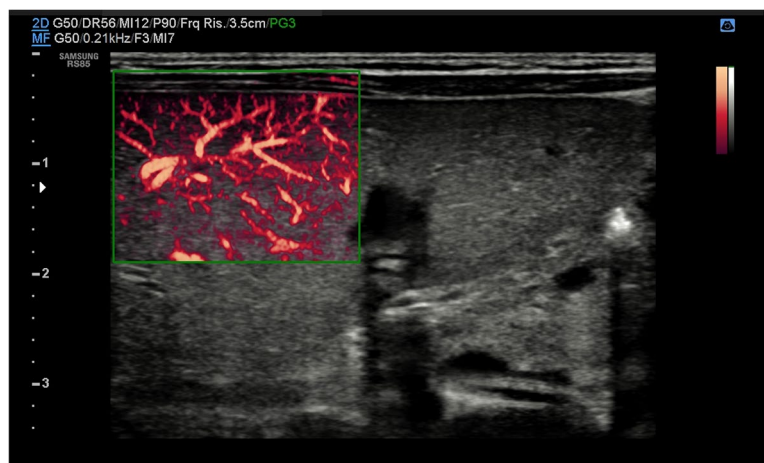
In addition, there could be US signs of increased hepatic vascular resistances, such as increased portal vein calibre and increased flow in the subcapsular arteries (Fig. 25) [58, 59].

However, US study has some limitations: extrahepatic bile duct may sometimes not be visualised even in normal

Fig. 24 US study of the liver with B-mode (a) and colour-Doppler (b) in a newborn with biliary atresia shows hyper-echoic tissue (arrows) just near to the hepatic hilum



Fig. 25 US study of the liver with B-Mode and power Doppler modalities in a newborn with biliary atresia: there is hepatic arterial flow extending to the hepatic surface (subcapsular flow)



newborns; a not clearly evaluable gallbladder may be also related to other causes of cholestasis; there are, on the contrary, cases of biliary atresia with normal gallbladder (about 20%); depending on operator's expertise and US scanner quality, triangular cord sign could be sometimes missed [60].

Despite the fact that MRCP and ERCP are potentially useful in the diagnosis of biliary atresia, their routine use in infants is still not accepted. Therefore, definitive diagnosis is often made with cholangiography which may also provide accurate evaluation of biliary tree [61, 62].

Conclusions

Ultrasonography is an essential imaging modality for the first detection and the management of hepatic injuries in children. Therefore, it is important to be aware of basic

normal ultrasonographic anatomy and of most common liver focal or diffuse pathologies in order to recognise them and eventually ask for laboratory tests and/or further diagnostic modalities.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed consent All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, and its late amendments. Additional informed consent was obtained from all patients for which identifying information is not included in this article.

Human and animal rights This article does not contain any studies with human or animal subjects performed by any of the authors.

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